GENERAL PRACTICE

Part 2

Dear Student,

Welcome to "part general practice - Pediatrics- which is divided into 12 chapters including; Perinatology, Respiratory Problems, Cardiology, Hematology, Oncology, Infection, Endocrinal Diseases, Gastroenterology, Metabolism, Urology, Genetics & followed by Multiple choice Questions, in a simple, illustrative way to make it easy to understand also it contains illustrative diagrams, photos and tables. Most of the topics represent the major categories of diseases e.g. what investigations to do on facing new couple going to marry, how to do fetal assessment, estimation of gestational age, the normal and abnormal findings of baby at birth, the congenital anomalies including the AD, AR, X- Linked and the multifactorial disorders. Birth injuries. How to do resuscitation of the baby. The technique of umbilical catheterization. The at risk pregnancy and intrauterine infection, prematurity, neonatal jaundice and how to dealwith, the neonatal convulsions and how to manage, the respiratory problems of neonate and how to do mechanical ventilation, the congenital heart diseases and treatment of heart failure, the common childhood malignancies of blood, bones, brain, eye & kidney. The vaccination, infant development, child abuse, and poisoning in the last chapter. Therefore, after reading this book, you are encouraged to read books on different medical fields, because the book conceived when the previous curriculum was being implemented. In writing this book. I have tried to make it as clear & as brief as possible. Since too much brevity may compromise understanding, i have been a bit "liberal" in some areas including some details ẃ are necessary for the general practitioner's understanding. This is done to encourage understanding rather than memorization. It is intended to be a text-book for medical students junior doctors & even those beyond can use this book for review.

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ALPHABETICAL INDEX

ABBREVIATIONS & ACRONYMS

<	Less Than
>	More Than
-ve	Negative
é	With
éout	Without
6 MP	6 Mercapto Purine
AA	Amino Acids
A1AT	α-1 Antitrypsin deficiency
ABGs	Arterial Blood Gases
A/C	Assist / Control Ventilation
ACEIs	Angiotensin Converting Enzyme Inhibitors Drugs
AD	Autosomal Dominant
ADHD	Attention Deficit Hyperactivity Disorder
AFE	Amniotic Fluid Embolism
AFV	Amniotic Fluid Volume
AIDS	Acquired Immune Deficiency Syndrome
ALL	Acute Lymphocytic Leukemia
AML	Acute Myeloid Leukemia
Amp	Ampule
antiHBc	antibodies against Hepatitis B core antigen
antiHBc IgG	antibodies against Hepatitis B core antigen type IgG
antiHBc IgM	antibodies against Hepatitis B core antigen type IgM
antiHBe	antibodies against Hepatitis B e antigen
antiHBs	antibodies against Hepatitis B surface antigen
APHge	Ante Partum Hemorrhage
APTT	Activated Partial Thromboplastine Time
AR	Autosomal Recessive
AS	Aortic Stenosis
ASD	Atrial Septal Defect
ATN	Acute Tubular Necrosis
AUB	Abnormal Uterine Bleeding
AZT	Azidothymdine
BBB	blood brain barrier
BET	Blood Exchange Transfusion

ß-HCG	Beta Human Chorionic Gonadotrophine
BL	Blood
BP	Blood Pressure
BPD	Broncho Pulmonary Dysplasia
BS	Blood sugar
BT	Bleeding Time
BV	Blood Volume
BW	Birth weight/Body Weight
CDL	Chronic Lung Disease
CHD	Congenital Heart Defect
CHO	Carbohydrates
Cl	Clostridium
CMV	Cyto Megalo Virus
CNS	Central Nervous System
CoA	Coarctation of Aorta
Conc	Concentration
Cong	Congenital
COP	Cardiac Out Put
CP	Cerebral Palsy
CPAP	Continuous Positive Airway Pressure
CPD	Cephalo Pelvic Disproportion
CRL	Crown Rump Length
CRP	C Reactive Protein
CRS	Congenital Rubella Syndrome
CS	Caesarean Section
CSF	Cerebro Spinal Fluid
CT	Clotting Time
DBP	Diastolic Blood Pressure
DDAVP	Trademark for preparations of Desmopressin Acetate
DIC	Disseminated Intravascular Coagulopathy
DM	Diabetes Mellitus
DNA	Deoxyribonucleic Acid
DSB	Direct SERUM bilirubin
ECF	Extra Cellular Fluid

ECMO	Extra Corporal Membrane Oxygenation
eGFR	Estimated Glomerular Filtration Rate
ELISA	Enzyme Linked Immune Sorbent Assay
EPT	Extremely Pre Term
ETT	Endo Tracheal Tube
F4	Fallot Tetralogy
FBS	Fasting Blood Sugar
FTA-ABS	Flourescent Treponema Antibodies Absrption Test
FiO2	Fraction or % of oxygen in the air that is inspired
FPD	Feto Placental Dysfunction
FH	Foetal Heart
FHb	Foetal Haemoglobin
Freq	Frequency
FT	Full Term
FTA-ABS	Fluorescent Treponimal Absorbed Test
G6PDD	Glucose 6 Phosphate Dehydrogenase Deficiency
GDM	Gestational Diabetes Mellitus
GIT	Gastro Intestinal Tract
GnRH	Gonadotrophin Releasing Hormone
HAART	Highly Active Anti Retroviral Therapy
HB	Hepatits B
Hb	Hemoglobin
HBcAg	Hepatitis B core Antigen
HBeAg	Hepatitis B e Antigen
HBIG	Hepatits B Immunoglobulin
HBsAg	Hepatitis B surface Antigen
HBV	Hepatitis B Virus
HCV	Hepato Cellular Carcinoma
HCG	Human Chorionic Gonadotrophine
Hct	Hematocrite
HF	Heart Filure
HFO	High Frequency Oscillator
HIE	Hypoxic Ischemic Encephalopathy
HIV	Human Immune deficiency Virus
HR	Heart Rate

Hr	Hour
HSCR	Hirschsprung Disease
HSV	Herpes Simplex Virus
Hx	History
ICC	Intra Cranial Calcifications
ICF	Intra Cellular Fluid
ICHge	Intra Cranial Hemorrhage
ID	Intra Dermal
IEM	Inborn Errors Metabolism
I/E ratio	Inspiration/Expiration ratios
IgM	Immunoglobilin M
IM	Intra Muscular
INo	Inhaled Nitiric Oxide
Infec	Infection
IP	Incubation Period
ITP	Immune Thrombocytopenic Purpura
IUCD	Intra Uterine Contraceptive Devices
IUCP	Intra Uterine Contraceptive Pills
IUFD	Intra Uterine Fetal Death
IUGR	Intra Uterine Growth Retardation
IUI	Intra Uterine Infection
IV	Intra Venous
IVF	Intra Venous Fluids
L	Liter
LBW	Low Birth Weight
LFTs	Liver function tests
LGA	Large for Gestational Age
LNs	Lymph Nodes
LPM	Litter Per Minute HBV
MAS	Meconium Aspiration Syndrome
Mo	Month
MM	Mucous Membrane
MR	Mental Retardation
NEC	Necrotising Entero Colitis.

NGT	Naso Gastric Tube
NHS	Neonatal hepatitis syndrome
NICU	Neonatal Intensive Care Unit
NN	Neonate
NND	New Natal Death
NNHS	Neonatal Hepatitis Syndrome
NSAIDs	Non Steroidal Anti Inflammatory Drugs
NTD	Neural Tube Defect
NTE	Neutral Thermal Environment
O_2	Oxygen
OGTT	Oral Glucose Tolerance Test
PaO2	Partial pressure (arterial) of Oxygen in Haemoglobin.
PCR	Polymerase Chain Reaction
PDA	Patent Ductus Arteriosus
PEEP	Positive End Expiratory Pressure
PID	Pelvic Inflammatory Disease
PIP	Peak Inspiratory Pressure
PPBS	Post Prandial Blood Sugar
PPHge	Post-Partum Hemorrhage
PPT	Precipitating
PPV	Positive Pressure Ventilation
Preg	Pregnancy
PRM	Premature Rupture Membrane
PS	Pulmonary Stenosis
PSV	Pressure Support Ventilation
Pt	Patient
PT	Pre Term / Prothrombin Time
RD	Respiratory Distress
RDS	Respiratory Distress Syndrome
RDW	Red Blood Cells Distribution Width
RES	Reticulo Endothelial System
RFTs	Renal Function Tests
R/O	Rule Out
RPR	Rapid Plasma Reagin

RR	Respiratory Rate
Rx	Treatment
RR	Respiratory Rate
$	Syphilis
SB	Serum Bilirubin
SBP	Systolic Blood Pressure
SC	Sub Cutaneous
Sec	Second
SEM	Skin Eye Mouth
SGA	Small Gestational Age
SIMV	Synchronized Intermittent Mandatory Ventilation
Sol	Solution
Spo2	Saturation (peripheral) of oxygen in blood
STDs	Sexually Transmitted Diseases
SVC	Superior Vena Cava
Sy	Syndrome
TA	Truncus Arteriosus
Ta	Tricuspid Atresia
TAS	Trans Abdominal Sonography
T & D	Total & Direct
TFTs	Thyroid Function Tests
Ti	Time inspiration)
TM	Trimester
TPN	Total Parenteral Nutrition
TSB	Total Serum bilirubin
TT	Tetanus Toxoid
Tv	Tidal volume
TVS	Trans Vaginal Sonography
UDPGT	Uridine 5'-Di-Phospho-Glucuronosyl Transferase
U/S	Ultra Sonography
URTI	Upper Respiratory Tract Infection
UTI	Urinary Tract Infection
UVC	Umbilical Vein Catheter
VC	Vaso Constriction.

VDRL	Venereal Disease Research Laboratory Test
VLBW	Very Low Birth Weight
VSD	Ventricular Septal Defect
VWD	Von Willebrand`s Disease
VWF	Von Willebrand`s Factor
W	Week
Ŵ	Which
Wk	Week
Wt	Weight
yr	Year
ZDV	Zidovudine

CHAPTER 1

PERINATOLOGY 1

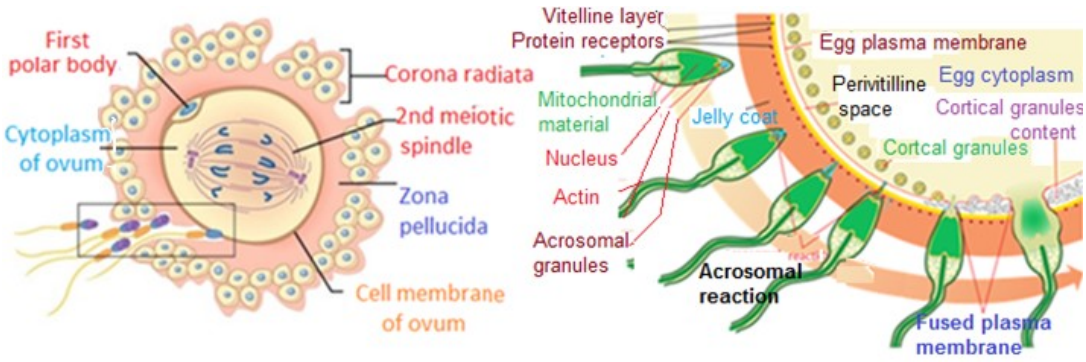
- Premarital counselling
- Conception
- Body & Physiologic changes in pregnancy
- Stages of Labour
- Foetal Assessment
- Estimation of Gestational age
- Primitive Reflexes
- Baby after Birth
- Birth Injuries
- Birth Asphyxia
- Resuscitation

PREMARITAL COUNSELLING

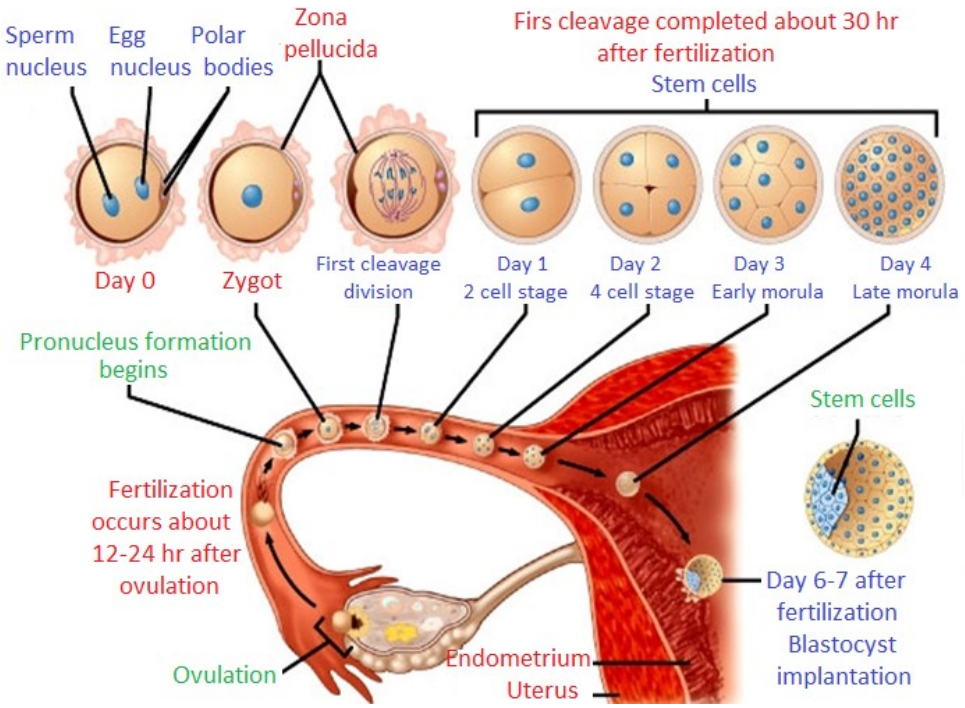
Include the following investigations:-

- Complete blood picture for anemia (may affect fertility).
- TORSCH screening for IUI.
- HIV antibodies (IgM) for AIDS.
- FBS & 2 HPPBS for diabetes mellitus may affect fertility.
- Semen analysis (man) for fertility.
- Rh grouping for Rh incompatibility.
- Chromosomal studies for hereditary diseases & AR disorders.
- Hemoglobin electrophoresis for thalassemia, sickle cell anemia.
- Thyroid function tests "TSH & T_4" as ⇧ or ⇩ can affect fertility.
- Hormones affecting fertility as; FSH, secreted by the anterior pituitary, promotes the formation of ova or sperm.
- Coagulation profile: BT, CT, PT, APTT "may cause dysmenorrhea/prolonged menses.
- Urine for vaginitis.

CONCEPTION



The Sperm Acrosomal Enzyme digest the egg`s coat, only one sperm penetrate the ovum.



Stages from day 0 up to day 7 of fertilization (up to implantation in uterus)

Human fertilization is the process during ẃ a male gamete (sperm) unites ē a female gamete (oocyte) to form a single cell (ZYGOTE). The egg can fertilized for about 24 hrs after ovulation. Sperm remain viable for up to 48 hrs within the female reproductive tract. This gives a 3 day "window" for fertilization: 2 days before & 1 day after ovulation. Sperm swim up the female reproductive tract, aided by muscular contractions of the uterus stimulated by prostaglandins in semen. The oocyte may also secrete a chemical that attarcts sperm. Freshly ejaculated sperms are unable to fertilize the 2ry oocyte & they must undergo a series of changes known as capacitation, sperms that have undergone capacit-

ation become hyperactive & highly motile. Capacitation occurs in the female reproductive tract, It takes about 7-8 hrs, during ẃ the membrane around the acrosome becomes fragile & its enzymes are released. It requires the combined action of many sperm to allow one sperm to penetrate the oocyte. When the 1st sperm enters the egg, the cell depolarizes causing the release of Ca+ ions inside the cell. This stimulates the release of granules that cause changes in the zona pellucida to prevent entry of other sperm.

Fertilization occurs through the following stages:-

Stage 1: Passage of sperm through Corona Radiata.

Stage 2: Penetration of Zona Pellucida.

Stage 3: Fusion of plasma membranes of the oocyte & the sperm, entry of sperm contents into the oocyte.

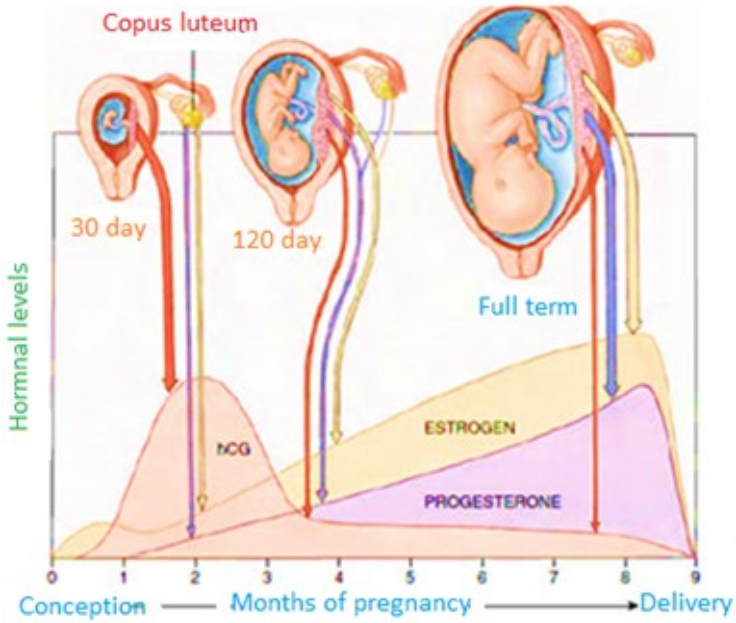
Stage 4: Second meiotic division of the 2ry oocyte & formation of female pronucleus.

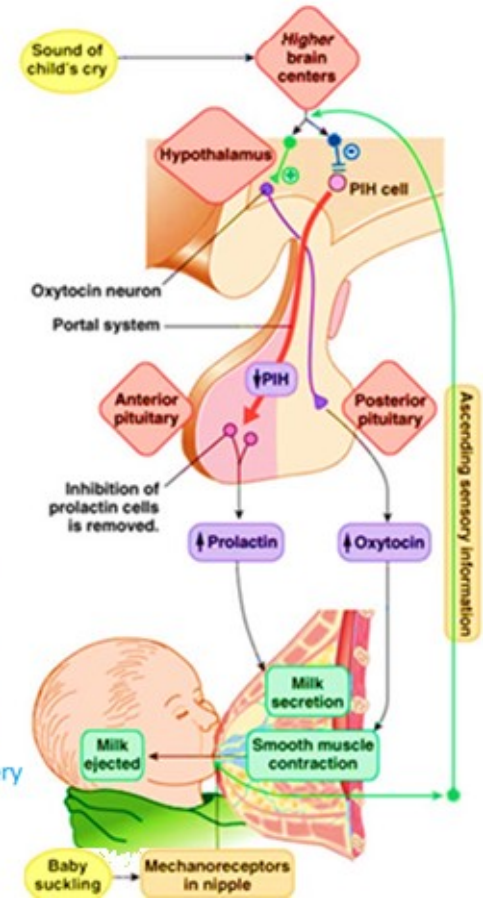
Stage 5. Formation of male pronucleus.

Stage 6. Fusion of pronuclei & ormation of the zygote & preparation of first mitotic division, repeated mitotic division of the zygote, begins about 30 hrs after fertilization. There is rapid ⇧in the number of cells, ẃ are called (blastomeres) become smaller ē each division.

Implantation: the blastocyst remains free in the uterus for a short time during ẃ the zona pellucida disintegrates. Blastocyst nourished by glycogen from glands of the endometrium. At about the 6th day after ovulation blastocyst implants (orients cell mass toward endometrium & secretes enzymes ẃ allow it to penetrate (digest) the endometrial wall. This nourishes the blastocyst for about a week after implantation. As early as 8-12 days after fertilization, the blastocyst begins to secrete HCG hormone. The HCG keeps the corpus luteum active until the placenta can produce estrogens & progesterone. The presence of HCG is the basis for pregnancy tests.

PREGNANCY HORMONES





Pregnancy hormones include

Human Chorionic Gonadotrophine: produced by placenta that maintains the corpus luteum during pregnancy

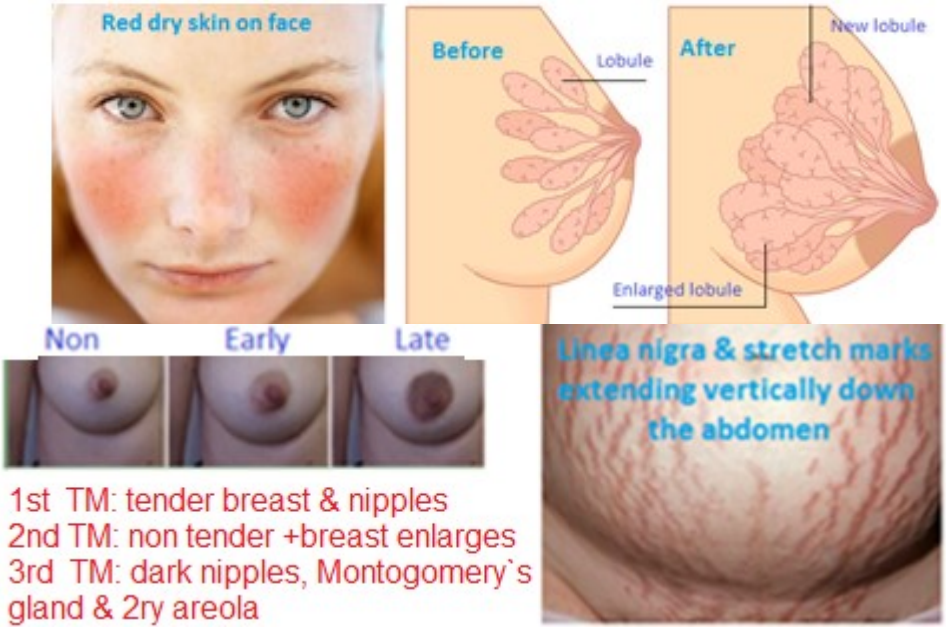
Estrogen: produced by the ovaries & later by placenta, ⇧ blood flow to uterus, maintain the uterine lining & stimulate growth of the ductus of breast.

Progesterone: produced the ovaries, the placenta & the adrenal gland, ready the uterus for implantation, relaxes uterine smooth muscle to prevent spontaneous abortion, prevent maternal immunological response to the fetus, stimulate growth of alveoli & ductal system of the breast.

Prolactin: released after birth from anterior pituitary, stimulate milk production.

Oxytocin: released from posterior pituitary, causes ⇧ contraction of the uterus during labor & stimulates ejection of milk into the ducts (let down reflex).

BODY & PHYSIOLOGIC CHANGES IN PREGNANCY



Cardiovascular changes

- Sodium & water retention. Total body water ⇧ (40%).
- ⇧ COP (30-50%).
- ⇩ BP (mean 105/60 mmHg in 2nd TM).
- ⇧ Maternal heart rate (up 15-20 bpm).
- ⇩ Systemic vascular resistance (vasodilatation + high flow, low-resistance circuit of the uteroplacental circulation).

Respiratory changes

- Mechanical: diaphragm rise 4cm, less negative intrathoracic pressure. No impairment in diaphragmatic or thoracic muscle motion. Lung compliance unaffected.
- Physiologic: O_2 consumption ⇧ 15-20% & 50% of this ⇧ is required by the uterus. Progesterone directly stimulates breathing. 70% of pregnant women experience ⇧ desire to breathe.

Gastrointestinal changes

- Mechanical: pressure from growing uterus on stomach lead to reflux & heart burn.

Pressure from growing uterus on lower portion of colon & rectum ⇨ constipation.

•Physiologic: relaxation of sphincter muscle between esophagus & stomach. Progesterone (smooth muscle relaxant) causes ⇩ in GI motility & delayed gastric emptying.

Metabolic changes

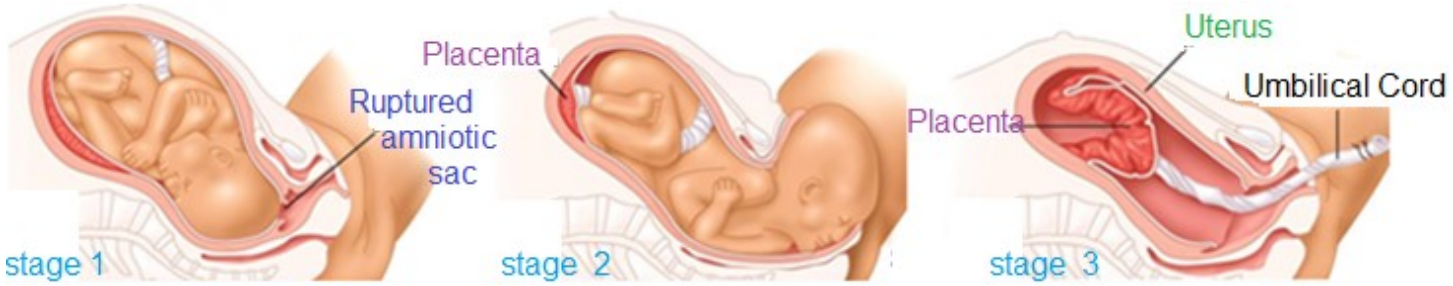
Caloric requirement: for a pregnant woman is about 300 kcal higher than that for non-pregnant woman's basal needs. Placental hormones affect glucose & lipid metabolism to ensure that fetus has ample supply of nutrients.

Lipid metabolism: ⇧ lipolysis (preferential use of fat for fuel, in order to preserve glucose & protein), ⇧ serum triglyceride (300%) & cholesterol (50%) spares glucose for fetus, since lipids do not cross the placenta.

Glucose metabolism: ⇩ insulin sensitivity & ⇧ insulin resistance due to hormones secreted by the placenta that are "diabetogenic": [GH, Human placental lactogen, Progesterone, Corticotrophin releasing hormone]. Transient maternal hyperglycemia occurs after meals because of ⇧ insulin resistance.

STAGES OF LABOR

Labor is a journey ẃ can take a long time, every women`s labor is different,it include;

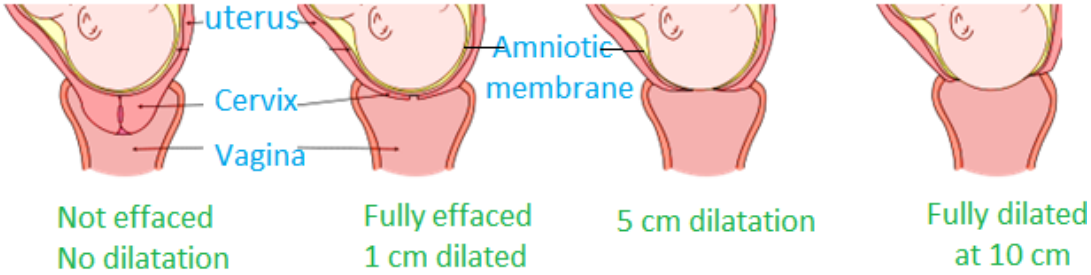


*The first stage

The latent phase: also called prodromal labour or prelabour, during ẃ the cervical effacement "thining" takes place (the uterus contracte & make the cervix become flat & soft),

it can last several days or wks before active labour starts, especially for primiparae, ended by cervical dilatation 3-4 cm & during ẃ the women may or may not have active contractions.

The active phase: strong, painful contractions tend to occur/3-4 min é the cervical dilatation from 3 cm ⇨ 10 cm, rupture of membranes or a bloody show may or may not occur at or around this stage. The duration of active phase averages some 8 hrs for primi & shorter for multiparae (at a rate of 1-3 cm dilatation hourly).



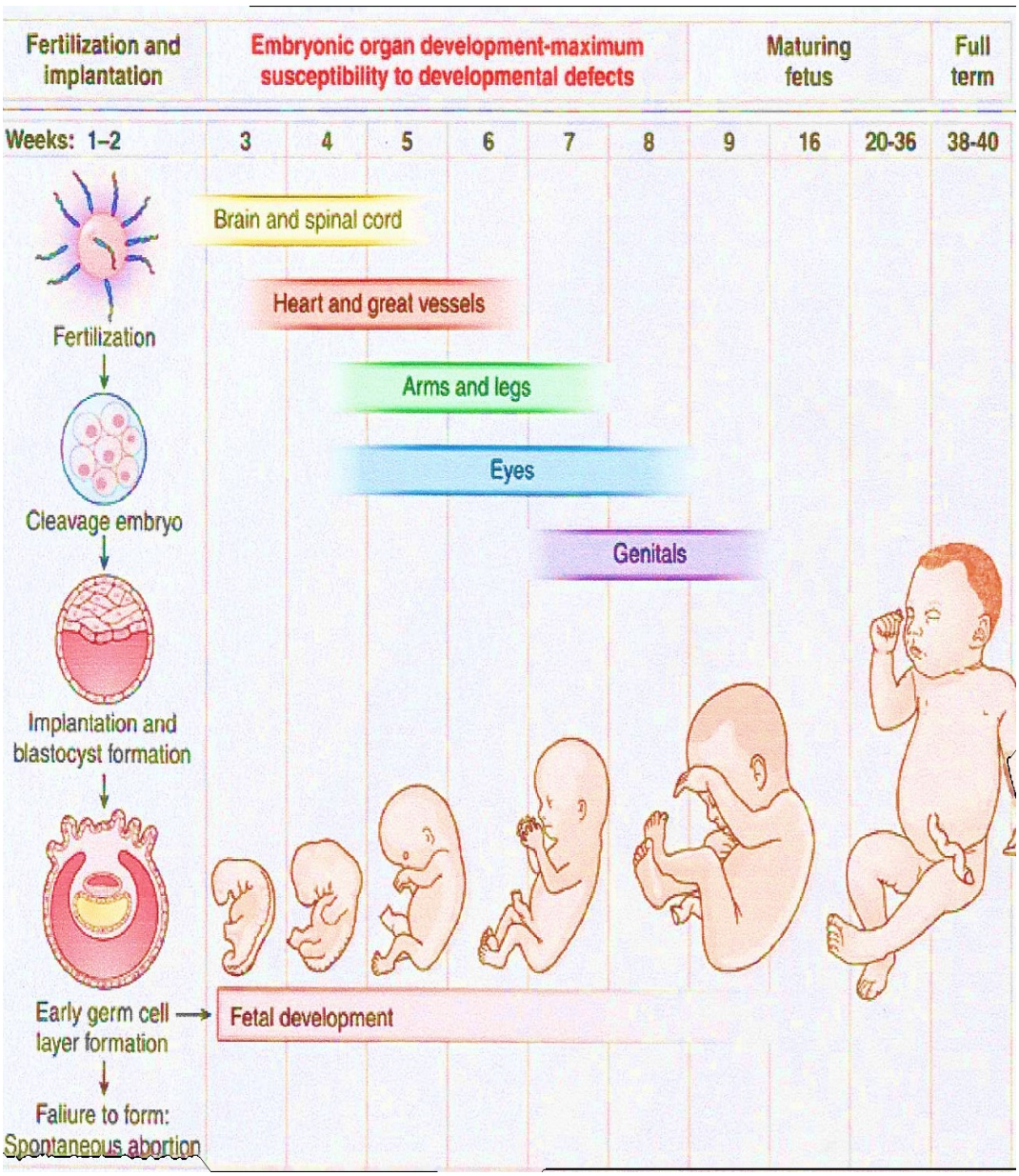
*The second stage

Start from full cervical dilatation, as the pressure on the cervix ⇧, the Ferguson reflex increases uterine contractions, the fetal head is fully engaged in the pelvis, below the level of pelvic inlet then below level of pubic arch, the appearance of fetal head at the vaginal orifice is termed “crowning”. Normal duration of this phase is 45-120 min in primi & 15-45 min in multiparae, the 2nd stage ended by the delivery of the baby.

*The third stage

Start from delivery of baby up to placental expulsion, the average estimated time is about 10-12 minute. The umbilical cord is clamped as early as 1 minute after the birth of the baby, while holding the baby 20 cm below the level of the mother, some prefare to do milking of the cord towards the baby before clamping as this help to ⇧ Hct & thus reduce the need for transfusion espicially in PT babies. The umbilical cord falls after 1-3 weeks from delivery.

FOETAL ASSESSMENT



The aim is assessment of growth & detection of birth defects as ē; NTD, Down sy, fragile X sy, cleft palate, Tay Sachs disease, sickle cell anemia, thalassemia, cystic fibrosis, or muscular dystrophy….

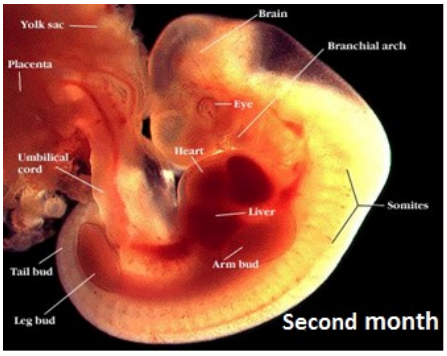
Foetal assessment includes

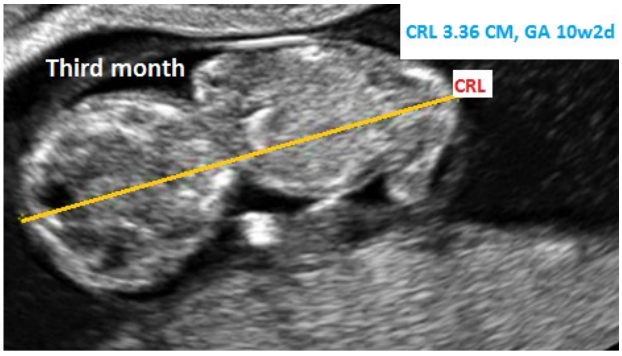
❁Ultrasonography ❁Amniocentesis ❁Chorionic villus sampling ❁Foetal blood cells in maternal blood ❁Maternal serum α fetoprotein ❁Maternal serum ß-HCG ❁Maternal serum Estriol ❁Electronic foetal heart monitoring ❁Biophysical profile.

Ultrasonography

The introduction of obstetric U/S in early 1970`s led to marked improvement in evaluation of fetal, placental anatomy, fetal growth, (the most accurate technique for estimating gestational age). Most pregnant women have 1st TM scan followed by another in the 2nd TM, for estimation of gestational age, growth, multiple pregnancies, fetal anomalies, placenta praevia. It is obligatory by law in many countries to be done twice for every pregnant women, it`s noninvasive, harmless to both fetus & mother, the developing embryo can first be visualized at about 6 weeks gestation. Through U/S we can determine the following:- gestational age, size & position of fetus, size & position of placenta, amount of amniotic fluid & fetal anatomy.

Ultrasonography 1st trimester



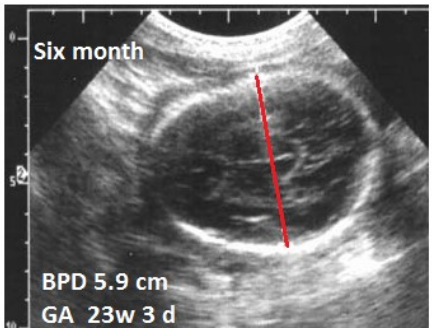


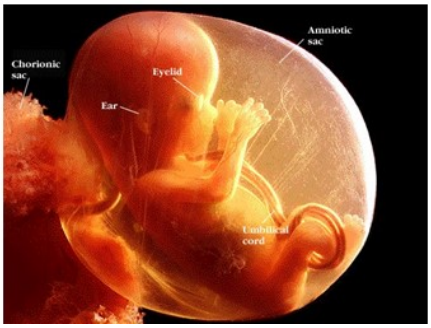
<u>Detection of mean sac diameter,</u> by calculating means of 3 sacs diameters, gestational age determined by consulting a table. An alternative method is adding 30 to the mean of 3 sacs size in millimeters to give gestational age in days. If cardiac activity detected but the embryo not measurable, the gestational age is about 6 weeks.

<u>Measuring crown rump length (CRL)</u>: longest demonstrable length of embryo excluding limbs & yolk sac. Correlation between it & gestational age is excellent up to 12 weeks amenorrhea. By the end of the 1st month, the embryo is about 0.1 inch long. The heart, ẃ is no larger than a poppy seed, has begun beating. By the end of 2nd month, the embr-

yo is about ¼ inch long & has distinct, slightly webbed fingers. Veins are clearly visible. The heart has divided into right & left chambers.

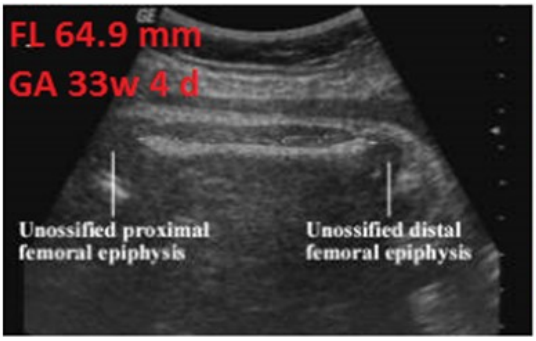
Ultrasonography 2nd trimester





Measuring the BPD, HC & Femur length, virtually linearly relate to gestational age. By gestational age 17-20 wk, the fetus is 3 inch long, covered é layer of thick, downy hair "lanugo". His Heart beat can be heard clearly. Mother may feel baby's first kick.

Ultrasonography 3rd trimester



Measuring BPD & femoral length, ossific center of distal femoral epiphysis at 32-33 wk gestation, visualization of **proximal tibial epiphysis** means gestational age 35 wk, if diameter of ossific center >7mm, it means gestational age 37 wk. By gestational age 25-28 wk, the **eyebrows & eyelids are visible.** The baby`s lungs are filled é amniotic fluid & he has started breathing motions. If mother talk or sing, he can hear (hearing surroundings). By gestational age 33-36 wk, baby gaining about ½ pound/ wk & layers of fat are piling on. He has probably turned head-down in preparation for birth. **The weighs by gestational age 33-36 wk is around 3-4 pounds.**

Chorionic Villus Sampling: From 10-12 wks gestation, catheter passed through the vagina to uterus, done transvaginal or transabdominal, fetal cell sample taken from placental chorionic villi under TV screen, it carry higher risk factor for fetal morbidity 1% above what would normally be expected. Help identify chromosomal anomalies as down sy, cystic fibrosis, sickle cell anemia. Considered to be 98% accurate in the diagnosis of chromosomal defects.

Amniocentesis: From 14-20 wks gestation, a needle passed through mother`s lower abdomen into amniotic cavity inside uterus under TV screening. It carries risk factor of fetal mortality 0.5% above what would normally be expected. The fetal cells from amniotic fluid mostly derived from fetal skin, grown in culture for chromosome analysis, biochemical & molecular biologic analysis.

Fetal Blood Cells in maternal blood: detecting fetal DNA present in maternal blood. Become available for select trisomes as Down & Edward syndromes (T 21, T 18).

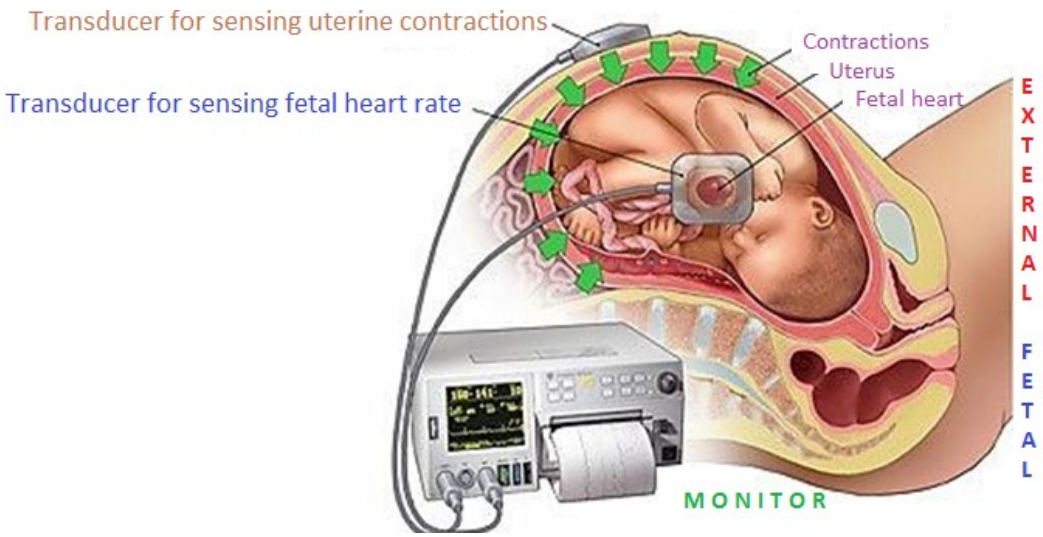
Maternal Serum α fetoprotein: ⇧ In case of multiple gestations, placental abruption, neural tube defect, abdominal wall defect, principal tumors that secrets α FP (as yolk sac carcinoma, neuroblastoma, hepatocellular carcinoma). ⇩ In case of Down, or Edward syndrome, or diabetic mother.

Maternal ß- Chorionic Gonadotrophine: represents the trophoblastic cell mass & is an indirect measurement of embryo development at early implantation stage. Very high level suggests molar pregnancy. Elevation + absence of fetus on sonar = Hydatiform mole. Elevation + ⇩ in α fetoprotein = Down syndrome.

Maternal Serum Estriol: ⇩ in Down syndrome, Adrenal hypoplasia é Anencephaly.

Electronic Fetal Heart Monitoring

Include: non stress test, stress test & Bio physical profile.



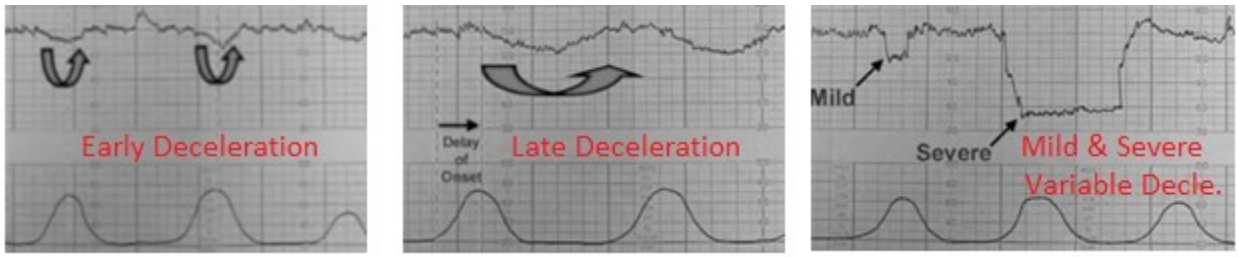
1-Non stress test

A cardiotocograph used to monitor FH using transducer for sensing FH & another for sensing uterine contraction on mother`s abdomen, to detect the following:-

Reactive (normal): ⩾2 FH accelerations within 20 min é or éout fetal movement discernible by the mother. Accelerations defined as 10 beats above baseline for at least 10 sec.

Non-reactive: no reactive criteria found within 30 min.

2-Stress test



Performed near the end of pregnancy to determine how well the fetus well cope ē labor contractions. Induce contractions through nipple stimulation or IV Pitocin, monitor FHR abnormalities using cardiotocograph, it include the following:-

Early Deceleration: the normal FHR is 120-160 bpm, normally ⇧during uterine contracti-on tion &return to normal at end of contraction this is called acceleration pattern. Early deceleration defined as ⇩ of FHR é uterine contraction in a mirror image pattern caused

by vagal stimulation as result of pressure over the anterior fontanel during contraction. It is not associated é fetal distress & is reassuring.

Late Deceleration: FHR ⇩ at the end of uterine contraction to <120 bpm, it means fetal hypoxia, the mother to be given O_2 by mask, lateral position, no oxytocin.

Variable Deceleration: series of decelerations varies in intensity, duration & relation to uterine contraction, FHR ⇩ to 90 bpm, means cord compression or cord around baby`s neck, it is very serious & need urgent C.S.

3-Biophysical Profile

Evaluation of fetal wellbeing, through scoring system using electronic FHR monitor & fetal U/S record for 20-30 min, done in case of non-reactive stress test, include:-

- 1- Fetal breath movements.
- 2- Gross body move.
- 3- Fetal tone.
- 4- Reactive FHR.
- 5- Amniotic fluid volume.

Biophysical profile	Normal {score = 2} during 20-30 min.	Abnormal (score=0)
Foetal breathing move.	At least 1 episode of at least 30 sec duration	Absent
Gross body movement	At least 3 body/Limb movements.	≤ 2
Foetal tone	1 episode of extension or flexion (Limb, trunk)	No
Reactive FHR	At least 2 episodes of 15 beat FHR acceleration	No
Amniotic fluid volume	≥ 1 fluid pocket 1 x 1 cm in 2 directions	No

Manning`s score: *Normal Score=8-10, Score < 8 is a bad sign & need early intervention.*

Conditions that can be detected prenatally

- Chromosomal abnormalities: Down, Edward, or Patau syndromes.
- Anatomical anomalies: skeleton, limbs, heart, bladder, kidneys, brain, duodenal atresia, omphalocele, gastroschisis.
- Tumors: neck, thorax, or abdomen.
- Diaphragmatic hernia.
- Inborn errors metabolism: lipid storage, tay sachs, gaucher, nieman pick, fabry`s disease, generalised gangliosidosis, mucopolysaccharidoses, hurler, hunter, san fillippo, morquio's sy, amino acid disorders, homocystinuria, tyrosinaemia, maple syrup urine, non-ketotic hyperglycinaemia, methyl malonic acidosis.
- Others: thalassemia, sickle cell, hemophilia A, VWD, galactosaemia, glycogen storage type II & IV, osteogenesis imperfect, lesch nyhan & menkes sy, α 1-antitrypsin deficiency, congenital nephrosis, CAH, xerodera pigmentosum, severe combined immune deficiency, acid phosphatase or adenosine deaminase deficiency.

ESTIMATION OF GESTATIONAL AGE

Gestational age is the time measured from 1st day of women`s last menstrual period to the current date, measured in wks, duration of normal pregnancy range from 38-42 wks. Infants born < 37 wks are considered PT. Infants born > 42 wks are considered post term, estimation of gestational age includes the followings:

1-Expected date of delivery: the simple method after knowing date of 1st day of last menstrual cycle is to add 7 to the day mentioned by the lady & deduct 3 from the month, for example if 1st day of last menstrual cycle is 10/10/2016, so the expected date of delivery will be 17/7/2017.

2-Ultrasonography: discussed before.

3-New Ballard Score: include 12 criteria (6 physical & 6 neuromuscular) as follow:-

The 6 physical criteria

① Skin: ranges from sticky, red to smooth, cracking or peeling

② Lanugo: soft dawny hair on baby`s body especially across shoulder & upper back, most common in preterm.

③ Plantar surface: creases on soles range from absent to covering entire soles.

④ Breast: thickness, size of breast tissue & areola (darkened ring around nipple)

⑤ Eyes/Ear: eyes fused or open, ear cartilage & stiffness.

⑥ Genitalia: Male: presence of testes, appearance of scrotum -from smooth to wrinkled. Female: appearance, size of clitoris & labia.

	-1	0	1	2	3	4	5
Skin	Sticky, friable, transparent	Gelatinous, red, translucent	Smooth, pink; visible veins	Superficial peeling and/or rash; few veins	Cracking, pale areas; rare veins	Parchment, deep cracking; no vessels	Leathery, cracked, wrinkled
Lanugo	None	Sparse	Abundant	Thinning	Bald areas	Mostly bald	
Plantar surface	Heel-toe 40–50 mm: –1 < 40 mm: –2	> 50 mm, no crease	Faint red marks	Anterior transverse crease only	Creases anterior 2/3	Creases over entire sole	
Breast	Imperceptible	Barely perceptible	Flat areola, no bud	Stippled areola, 1–2 mm bud	Raised areola, 3–4 mm bud	Full areola, 5–10 mm bud	
Eye/Ear	Lids fused loosely: –1 tightly: –2	Lids open; pinna flat; stays folded	Slightly curved pinna; soft; slow recoil	Well curved pinna; soft but ready recoil	Formed and firm, instant recoil	Thick cartilage, ear stiff	
Genitals (male)	Scrotum flat, smooth	Scrotum empty, faint rugae	Testes in upper canal, rare rugae	Testes descending, few rugae	Testes down, good rugae	Testes pendulous, deep rugae	
Genitals (female)	Clitoris prominent, labia flat	Clitoris prominent, small labia minora	Clitoris prominent, enlarging minora	Majora and minora equally prominent	Majora large, minora small	Majora cover clitoris and minora	

The 6 Neuromuscular criteria

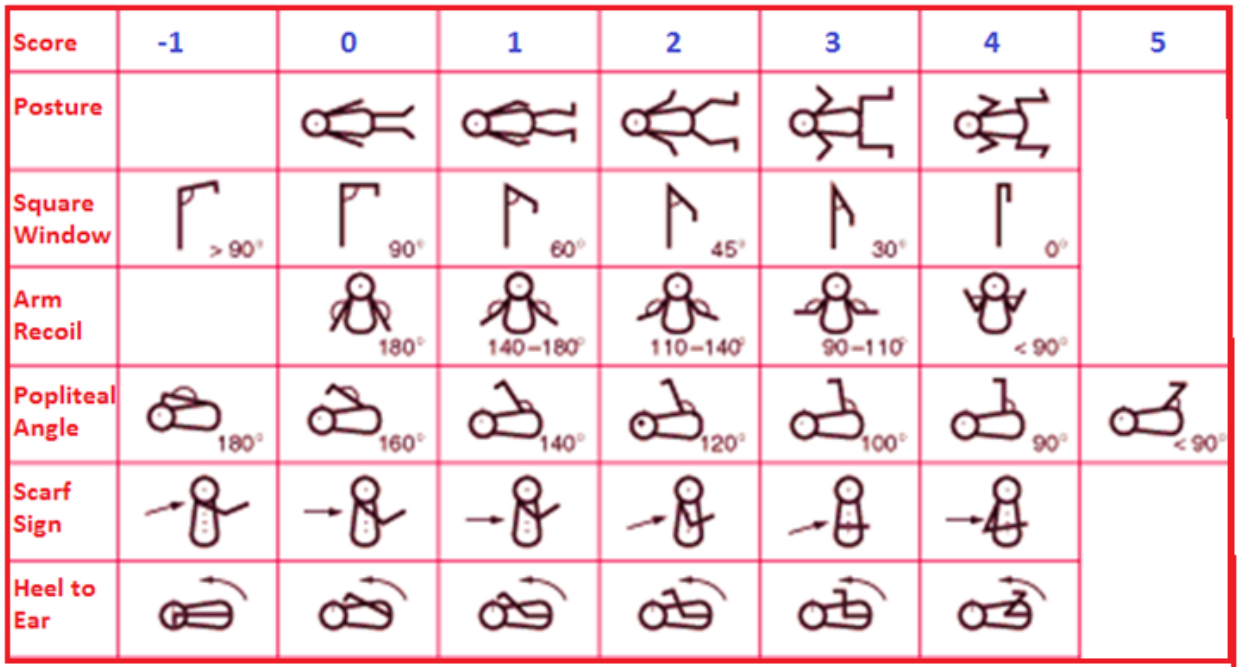
1-Posture: how does the baby hold his arms & legs? 2-Square window: how much baby`s hand be flexed toward wrist?

3-Arm Recoil: how much baby`s arm spring back to flexed position?

4-Popliteal Angle: how far baby`s knee extend ?

5-Scarf Sign: how far baby`s elbow can moved across baby`s chest?

6-Heel to Ear: how close can baby`s feet moved to the ear?



Maturity Rating

Score	Weeks
−10	20
−5	22
0	24
5	26
10	28
15	30
20	32
25	34
30	36
35	38
40	40
45	42
50	44

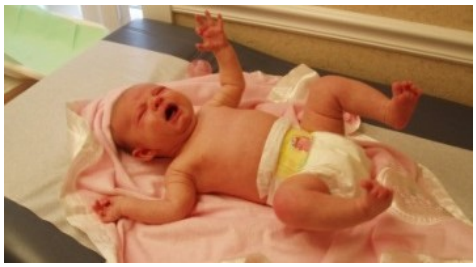
Calculating the sum of score in the above 2 table gives the Billard score ẃ determine the gestational age of the baby.

- The maximum score is equal to 50 ẃ denotes 44 wks gestational age.
- Score equal to 40 is equal to 40 wks gestation.
- The minimum score is equal to 10 ẃ denotes gestational age of 20 wks.

PRIMITIVE REFLEXES

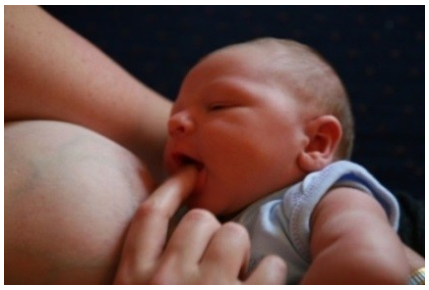
Primitive reflexes are seen in newborns. Develop in utero, as automatic movements & changes, directed from brain stem, require no cortical involvement (thought). The primitive reflexes needed for survival & growth. Inhibited by age one year. May appear later in life due to illness, particularly those affecting frontal lobes of brain.

MORO REFLEX



The reflex is elicited by excessive information in any of baby`s senses, as head suddenly shifts position, loud noise, bright light, sudden rough touch. Legs & head extend while the arms jerks up & out. Peaks in 1st month of life, begins to disappear around 2 months of age. It is the baby alarm reflex, as the newborns higher centers are not sufficiently developed to make rational decision whether a circumstance is threatened or not. Bilateral absence of reflex may linked to damage to the infant`s CNS. Unilateral absence means injury due to birth trauma as fracture clavicle, or injury to brachial plexus. Persistence of reflex indicates immaturity of CNS.

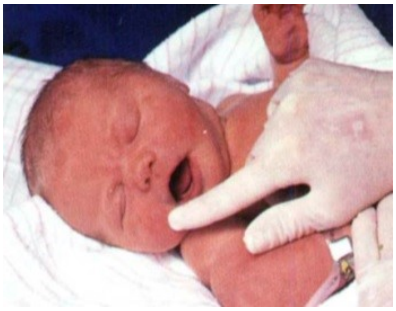
SUCKING REFLEX



Touching roof of baby mouth ē finger, or mother touch it ē her nipple, baby start sucking.

Disappear by age 2-3 months & sucking will be a result of conscious effort. This reflex does not begin until about 32 wk gestation & is not fully developed until about 36 wk gestation, so PT babies may have a weak or immature sucking ability.

ROOTING REFLEX



Turn head toward anything that strokes his cheek or mouth, searching for the object by moving his head in steadily decreasing arcs until the object was found.

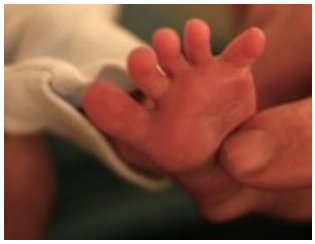
PALMAR GRASP

When an object is placed in the infant`s hand & strokes their palm, the fingers will close & they will grasp it ē palmar grasp, the grip is strong but unpredictable, though it may be able to support child weight or may release his grip suddenly ēout warning.

PLANTER GRASP

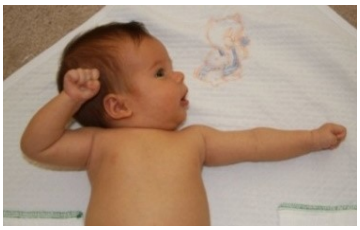
Pressing a finger against the sole of a foot just behind the toes, the response consists of flexion & adduction of all toes.

PLANTER REFLEX/BABINSKI SIGN



When the sole of the foot is stimulated ē blunt instrument, or finger, baby smaller toes fan out & his big toe will dorseflex slowly. This happens because the corticospinal pathways that run from brain down the spinal cord are not fully myelinated at this age. The reflex disappeared by the end of 1st yr. It is non-pathological. While Babinski's sign refers to its pathological form due to brain or spinal cord disease when elected after age 1 year or any age after as a result of neurological damage.

TONIC NECK REFLEX



Fencing reflex because the characteristic position of the infant arms & head resembling that of classically trained fencer. If the face turned to one side, the arm & leg on the side of turn will extend, while the arm & leg on the opposite side will flex.

STEEPING REFLEX

Walking or dance reflex. Carefully support the baby underneath his arms, lean him slightly forward & lower his feet onto a hard flat surface, he will make a walking step.

GALANT REFLEX

Using finger nail, gently stroking one side of neonate spinal column from head to buttocks, neonate trunk curve toward the stimulated side. The reflex inhibited between 1st -3rd month of life.

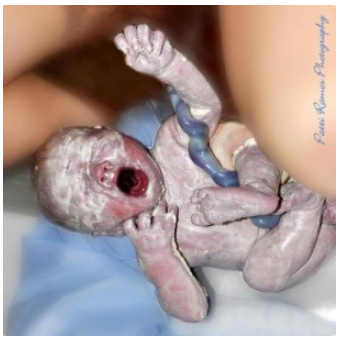
SWIMMING REFLEX

Putting baby under 6 months of age in water, he will move his arms & legs & hold his breath, looking like natural swimmer. The reflex slowly diminishes from around 9th month of age. Placing the infant in water can be very risky procedure, infant can swallow large amount of water while performing this task.

BABY AFTER BIRTH

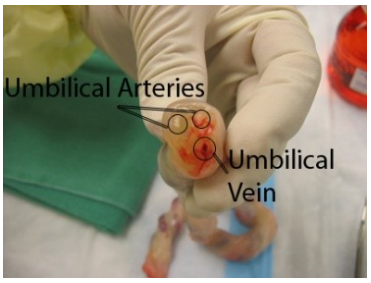
The baby is covered in utero ē a sebaceous secretion called 'Vernix Caseosa'. His respire-tions is 30-40/min, mostly abdominal & shallow at first. The Skin temperature is 36.4 - 37.0 °C. His abdomen is convex. The liver is usually palpable 1-2 cm below rib margin. Kidneys are often just palpable. Limbs are warm & well-rounded. The whole skin should rapidly become pink. Length is 50 cm & weight is 3.5 kg, the eyes are dry. The newborn baby seldom weeps. Sclera is often blue at birth. Pulse (apex) 120-140/min. Stump of umbilical cord tied or clamped, it should obliterate in 3-4 days & separate in 6-9 days. In male baby the testes should have descended into the scrotum by term. Pressure from tight birth canal might cause baby skull bones to shift, overlap, elongation, cone shaped, particularly if mother had long labor or vacum extraction. Cleaning é sterile water, cotton once his body temperature stabilized, mild baby shampoo may be used, cleaning eyes é sterile cotton & warm water, antibiotic eye drops as prophylactic for neisseria gonococci, Vit K 1 mg PO or 0.5 mg IM. The healthy baby is alert, active for 1-2 hr, then goes into sleep for another 1-2 hr to recover from labor stress, the 2nd period of activity lasts for 1-2 hr during ẃ he dema- nd feeding. Bonding process, presence of baby beside mother during the first few hrs is important.

VERNIX CASEOSA



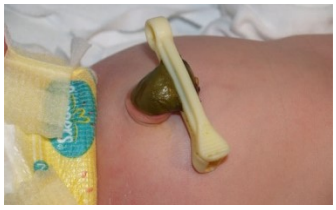
All newborn babies covered é vernix caseosa as white, sticky & waxy. It protects baby`s skin from the amniotic fluid. After birth vernix is easy enough to wipe away or disolves into the skin soon after birth. Top layer of newborn skin flakes off shortly after birth, dry, peeling of skin in the 1st few wks. Skin may covered by fine, **downy hair** at birth (Lanugo hair) especially on back, shoulders, forehead & is common in PT, typically disappear with-in several wks.

UMBILICAL CORD



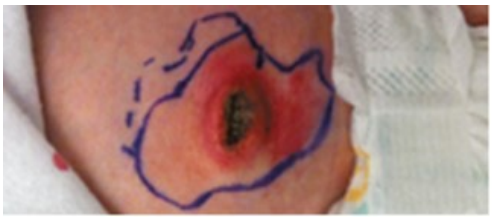
Cutting umbilical cord usually done after 30 sec from delivery, while baby position lower than mother abdomen. Some prepare to do milking once or twice towards the baby, as early clamping can deprive baby from about 50 ml of blood. The blood volume of NN is about 80 ml/Kg, equal to 1/11 of his BW (in adult it is approximately 5 L -70 ml/Kg), so 50 ml of blood in placenta represent 30% of total blood volume in PT weigh 2 Kg. The stump of umbilical cord usually yellowish green in color at birth, dries out & falls after 1-3 wk, the color change from yellow green to brown to black.

MECONIUM STAINED UMBILICAL CORD



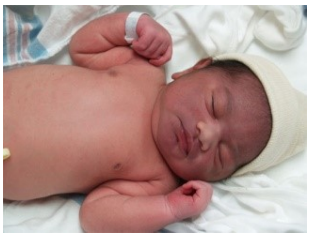
The above photo shows a cord of about 7 hrs old, but the normal light yellow color is not visible, even though the cord is still plump. The presence of meconium in amniotic fluid gives the cord its dark green color. Although meconium staining has no direct effect on the infant, the presence of meconium is often associated ē in utero stress.

UMBILICAL INFECTION



Contamination at birth, from infected amniotic fluid, home deliveries, poor hygienic condition Alternatives of treatment include; Flucloxacillin: PO + IV ŵ is better than IM, syrup 250 mg, amp 500 mg, dose 150 mg/Kg ÷3 for 10 days, or Cefotaxime: amp 250 mg, IV or IM, dose 50 mg/Kg ÷2 for 10 days, or Aminoglycosides: amp 20 mg, IV or IM, dose 3-5 mg/kg ÷ 2 for 3 days.

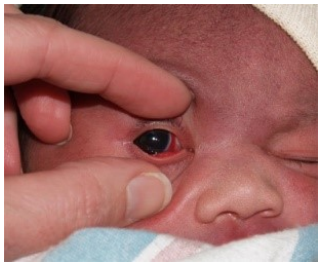
FACIAL BRUISING



Marked bruising of the face can occur during delivery. It is more common when there is a tight nuchal cord, when the delivery is precipitous or difficult, or when the infant is large. When the infant is bundled, this facial appearance could be mistaken for cyanosis, but ē

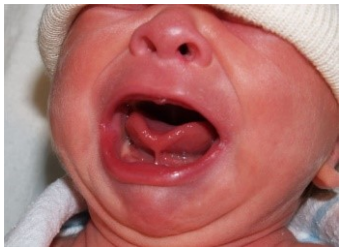
quick comparison to the color of the rest of the body, the diagnosis is obvious. This type of bruising resolves over the course of several days.

SUBCONJUNCTIVAL HEMORRHAGE



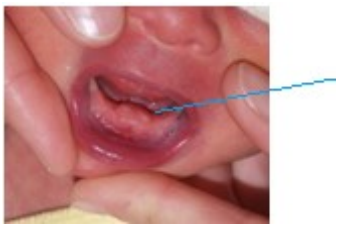
Is a frequent finding in normal newborns, results from the breakage of small vessels during the pressure of delivery. The red area may be large or small but is always confined to the limits of the sclera. It is asymptomatic, does not affect vision & spontaneously resolves in several days. Note also the numerous petechiae on the forehead. This infant had significant facial bruising as well as the eye finding.

ANKYLOGLOSSIA



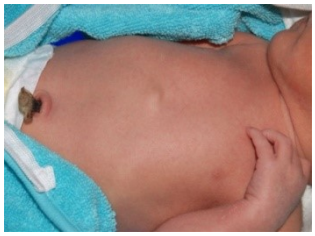
Tongue-tie seen in 4% of newborns. Many babies ē this condition can breastfeed ēout difficulty, but in some cases, a tight frenulum makes latching on difficult. In such cases, frenotomy may be indicated. The Hazelbaker assessment tool for lingual frenulum function is one tool that may be used to grade the severity of the tongue tie objectively. There are no prospective trials on the outcome of speech in those infants, so this is not currently an evidence based reason to clip the frenulum in the nursery. The major impact on breast feeding, however, is well documented. Mostly not require surgery. The tongue grow forward in the first year.

NATAL TEETH



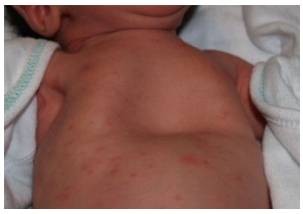
Natal teeth usually occur in this location in the mandibular gum (Photo). In this case, eruption cysts are still completely covering the teeth, but ē palpation, 2 firm teeth can be appreciated. They are also partially visible as faint white streaks within the cysts. Natal teeth occur in 1:2000 - 1:3500 newborns. They are usually part of the primary dentition of the child, so they should not be removed unless they are mobile, presenting an aspiration risk, or causing secondary tongue ulceration.

PROMINENT XIPHOID



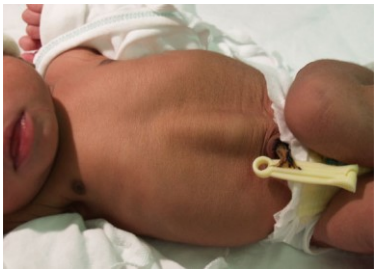
This visible (photo), firm lump in the midline of the chest is a frequently observed finding in newborns. It is simply a prominence of the xiphoid process & does not represent an abnormality. With time, this becomes less noticeable

PECTUS EXCAVATUM



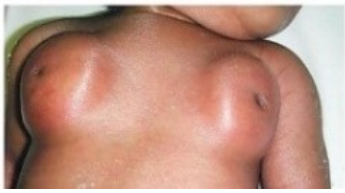
With deep inspiration, the sternum appears to almost collapse into the chest cavity. While connective tissue disorders, such as Marfan's sy, may be associated ē this finding, pectus excavatum is more commonly a benign, isolated entity.

DIASTASIS RECTI



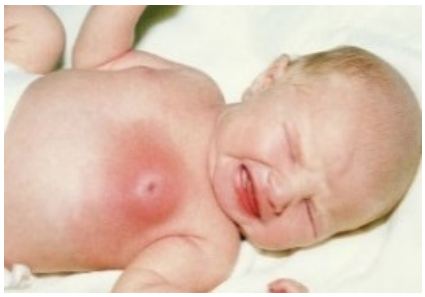
A vertical bulge down the midline of the abdomen can be seen in many newborns when intra-abdominal pressure⇧. In this photo, a shadow lateral to the bulge can be seen going up the left side of abdomen, starting near the umbilical clamp. Diastasis recti is caused by a relative weakness of the fascia between the two rectus abdominus muscles. It is not a herniation & is not pathologic. With time, Spontaneous resolution is still expected.

BREAST & GENITALIA



Before birth, mother hormones pass to the baby, this lead to swollen breast at birth in both boys or girls. Typically disappears in 2-4 week. In addition, girls may also have a light vaginal discharge & swollen vulva ẃ may last for several days.

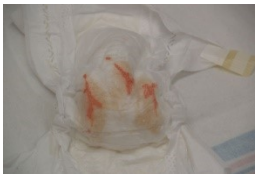
BREAST ABSCESS



Commonly staphylococcal. Once there is pus it must evacuated. Technique of evacuation is as follow; antiseptic, local or spray anaesthesia, go é closed artery forceps, open it inside, squeeze, clean, change dressing every 12 hrs. Alternatives of antibiotics include:-

Flucloxacillin PO or IV is better, sy 250mg, amp 500mg dose 150 mg/Kg÷3 for 10 days or Cefotaxime (3rd geneneration Cephalosporine) amp 250 mg, 50 mg/kg÷2 IV or IM. or Aminoglycosides amp 20 mg IM or IV, 3-5 mg/Kg ÷ 2 for 3-5 days.

URATE CRYSTALS

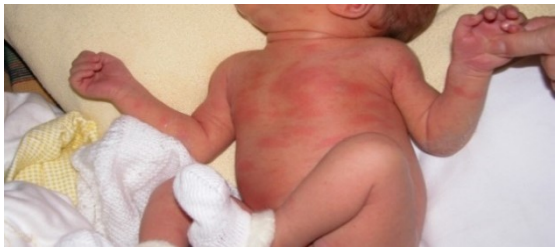


Often mistaken for blood in the urine, urate crystals are a frequent intermittent finding in the first week. The characteristic appearance of pink-orange material is sufficient to make the diagnosis. Are typically found in the setting of concentrated urine & may indicate dehydration.

NORMAL VAGINAL BLOOD

This is normal vaginal withdrawal bleeding that occurs in some female infants. Similar to withdrawal bleeding in adolescents, this typically occurs on the 3rd day after birth, continues for a few days, then stops.

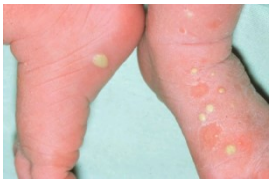
ERYTHEMA TOXICUM NEONATORUM



The most common rash in newborns. Seen on face, trunk, or extremities. characterized by macular erythema, papules, vesicles & pustules & it resolves ēout permanent sequelae. It is a benign self-limited, asymptomatic disorder of unknown etiology, occurring pri-

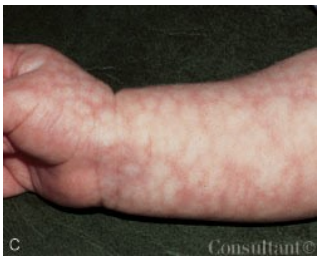
marily in healthy newborns in the early neonatal period. Seen in 50% of infants, fades within 5-7 days, but recurrences may occur for several wks. Smear of pustule reveals eosinophils. No treatment.

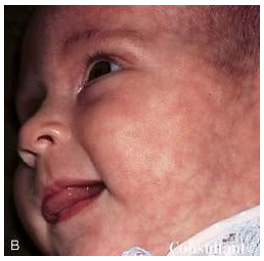
TRANSIENT NEONATAL POSTURAL MELANOSIS



Self-limiting dermatosis of unknown etiology. Usually presents at birth. Pustule on non-erythematous base, crusts over several days ẃ desquamates & leaves a hyperpigmented macule ē collarets of fine scale. Hyperpigmentation fades in 3 wks to 3 months. Smear of pustule reveals neutrophils. No specific treatment.

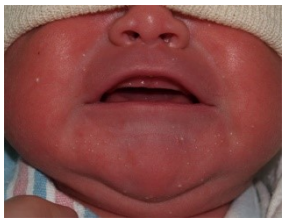
CUTIS MARMORATA





Is normal finding seen in NN (Geographical skin), due to vasomotor instability (variable vascular constriction & dilation). The skin is red, white marbled appearance, most obvious when skin in cool & resolves ē warming.

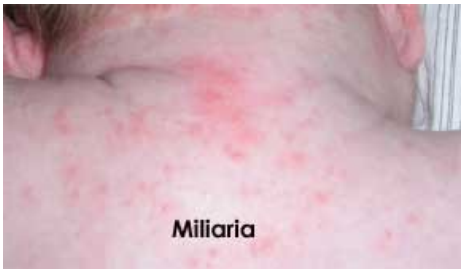
MILIA

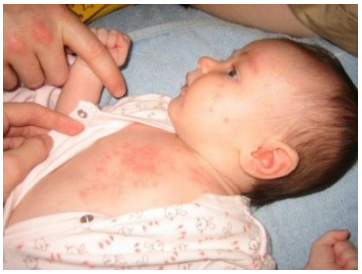


White papules present at birth & have no inflammatory component, appears on this baby's chin & cheeks, seen in up to 40% of newborns. Milia are keratin filled epithelial

cysts, spontaneous exfoliation & resolution is expected within a few wks. Parents will occasionally mistake those lesions for neonatal acne. Acne, even though caused by maternal hormones, does not generally appear until after 2 wks of age. Milia usually resolve during the 1st month of life.

MILIARIA





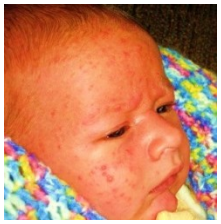
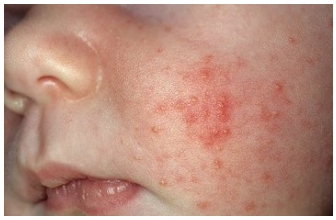
Miliaria crystallina: superficial 1-2 mm vesicles on non-inflamed skin.

Miliaria rubra (prickly heat): occur in response to thermal stress. Common during hot humid weather & é over dressing. Appear as small red papules & pustules, usually erupt in crops in the scalp, face & trunk result from sweat gland dysfunction. Results from obstruction to the flow of sweat & sweat gland rupture.

SEBACEOUS HYPERPLASIA

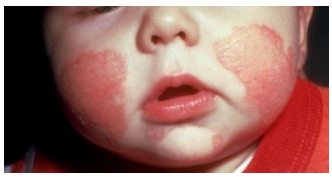
In contrast to milia, the *raised lesions* on the nose in this newborn are sebaceous hyperplasia. The lesions are more yellow than milia & are the result of maternal androgen exposure in utero. Sometimes referred to as "miniature puberty of the newborn", maternal hormone exposure may also cause vaginal withdrawl bleeding in infant girls & neonatal acne in boys. Sebaceous hyperplasia is a benign finding & spontaneously resolves ē time.

BABY ACNE

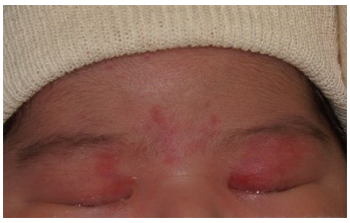
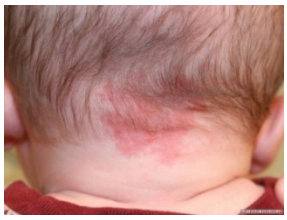
It does not generally appear until *after 2 wks of age.* Red or white papules on forehead, cheeks. Result from exposure to maternal hormones during pregnancy. No treatment required as the condition disappear on its own within few wks.

BABY ECZEMA



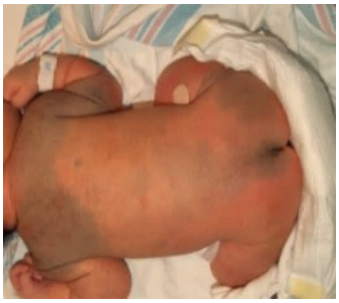
Patches of red, scaly & itchy skin. Result from rough fabrics, food allergy, bubble baths. Management is prophylactic.

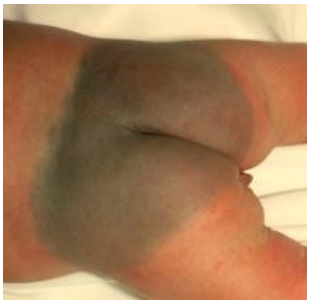
SALMON PATCH

Is vascular malformation. Seen in 60% of infants, may also be found on the nape of the neck in newborns, these lesions become less intense ē time, but are frequently visible into adulthood. When lesions are present only on the eyelids, they are sometimes mistaken for bruising, although the lids may be quite edematous, bruising in this location is very rare.

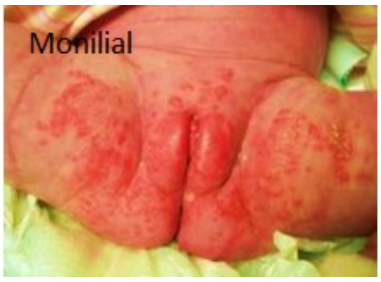
MONGOLIAN SPOTS

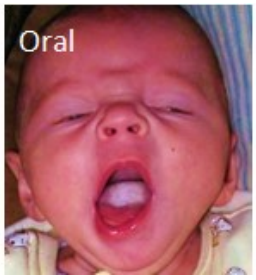




Large, flat, bluish or grey mark on back, buttocks (may cover the entire buttocks). Commonly seen in Asian, African descent. It fades during early child hood.

DIAPER DERMATITIS & ORAL MONALIASIS



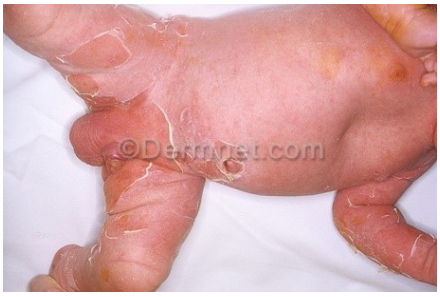


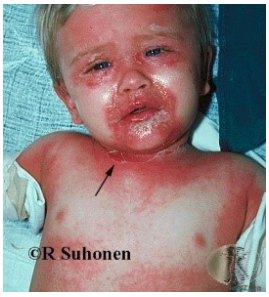
Candidal dipar dermatitis appears as confluent bright red & plaques, scattered pustules, overlying scales & satellite lesions on the periphery. Involving the skin folds. Flourishes in warm moist environment. Babies who have recently taken antibiotics are more likely to develop a yeast infection. Oral monaliasis appear as creamy white sores over the tongue & in mucosal buccal cavity. Treated by local antifungal skin ointment or oral drops, or gel for oral monaliasis.

Prolonged contact é urine or feces result in diaper dermatitis appear as bright red color not affecting the skin folds & is prevented by changing diapers when they are wet or soiled, allowing diaper area to dry between changes & using topical barrier ointment as zinc oxide.

Irritant/Contact diaper dermatitis: characterized by presence of red eroded papules.

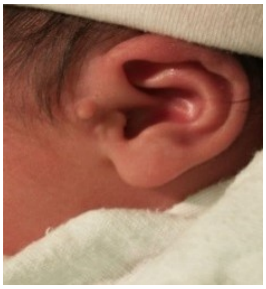
STAPHYLOCOCCAL SCALDED SKIN SYNDROME

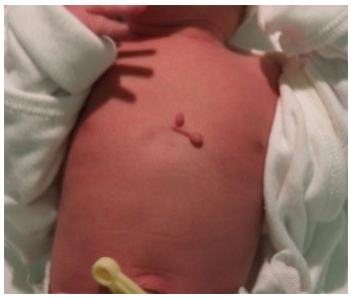


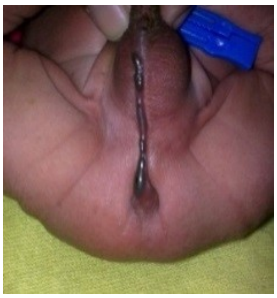


Severe staph infection of skin, associated é prodrome of fever, malaise, sore throat & fluid loss. Mortality rate is 3% in kids. If the baby in hospital, you should isolate him & identify possible staph carrier. Management include; parenteral antibiotic (Flucloxacillin, 50-100 mg/Kg ÷ 3 for 5-7 days).

SKIN TAGS

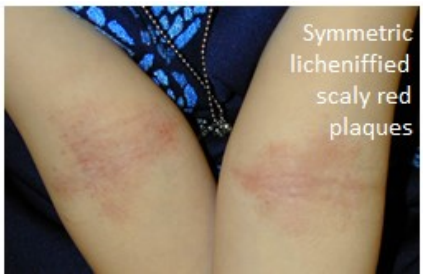






Common on ears. Usually tied off or clipped. May be associated é anal atresia.

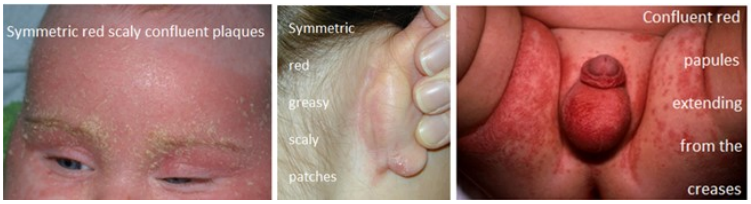
ATOPIC DERMATITIS



Is a chronically relapsing skin disorder ē an immunologic basis. The clinical presentation varies from mild to severe. In the worst cases it may interfere ē normal growth.

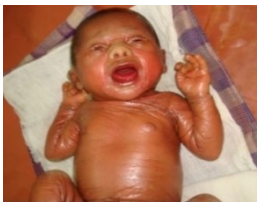
Treatment: adequate skin hydration, avoidance of allergenic precipitants, topical anti-inflammatory oint, systemic antihistamines & antibiotic coverage of secondary infections.

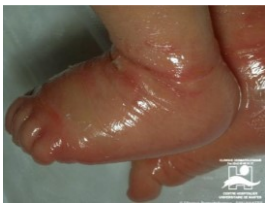
SEBORRHEIC DERMATITIS



Seen in 50% of babies, appear as thick, yellow, crusty greasy patches on scalp, may extend to forehead & eye brows. Researchers are still studying what causes this common skin disease. It appears that the cause is complex. Many factors seem to work together to cause it. These factors may include the yeast that normally lives on the skin, genes, living in a cold & dry climate, stress, & a person's overall health. Treated by baby or olive oil & in severe cases medicated shampoo "Nizapex" gives good results for scalp affection.

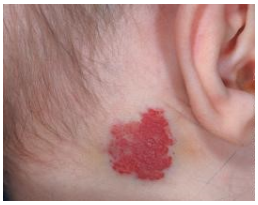
COLLODIAN BABY





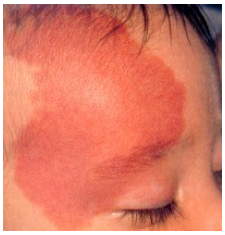
Baby born encased in thick cellophane like membrane. Most go on to develop ichthyosis. The membrane is then shed, leaving either normal skin or, more often, one of the forms of nonvullous congenital ichthyosiform erythroderma or lamellar ichthyosis. (a group of scaling disorders). Barrier function of skin is affected by cracking & fissuring, in addition to ⇧ of insensible water loss, heat loss & ⇧ risk of infection. Complications are minimized by placing baby in high humidity, neutrally thermal environment. Desquamation usually complete by 2-3 weeks of life.

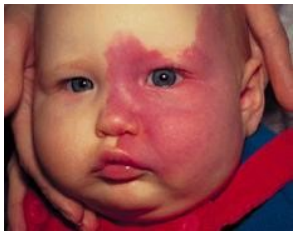
HAEMANGIOMA



Congenital vascular malformation. Occur in 10% of all newborns. Presents in the first few months of life. Marked vascular overgrowth resulting in bright red discoloration & definite elevation. Rapid growth for the first 6-12 months, then a plateau period, then slow involution. 50% involute by age 5, 90% by age of 9 years. Refer to dermatology if lesion involves a vital structure or if there are multiple lesions.

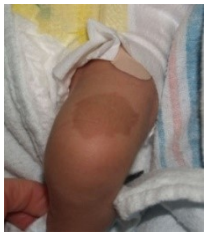
PORT WINE STAIN

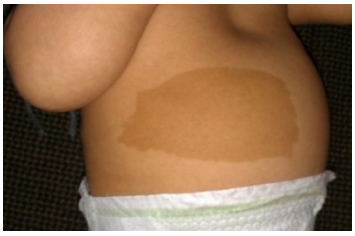




Purplish - red vascular malformation present at birth. Lesions do not enlarge but remain flat & persist. When involves ophthalmic branch of the 5th cranial (trigeminal) nerve, it can be associated be a constellation termed Sturge-Weber syndrome associated ē intracranial vascular abnormalities, manifested by seizures, MR, hemiplegia, glaucoma & in such condition it usually require CT scan & MRI for diagnosis.

CAFÉ AU LAIT SPOTS





Seen in 10% of babies. Result from ⇧ amount of melanin. Appear as flat, brown, round or oval lesions é smooth edges. in some areas of skin. May ⇧ in number é age. It is usually of little or no significance but may indicate neurofibromatosis if > 4-6 cm or > 6 lesions are present. Neurofibromatosis is an AD, occur in 1/3000 live birth, need further investigations as MRI of brain & spine, CXR, & XR of spine & abdomen.

BIRTH INJURIES

Injuries to the newborn that result from mechanical forces (i.e compression, traction) during the birth process. Even though most women give birth in modern hospitals surrounded by medical professionals, 7/1000 births result in birth injuries. In general birth injuries account for fewer than 2% of neonatal deaths.

Predisposing factors

- Prolonged or rapid delivery.
- CPD, small maternal stature.
- Deep transverse arrest of presenting part of the fetus.
- Oligohydramnios.
- Abnormal presentation (breech, shoulder).
- Use of midcavity forceps or vacuum extraction.
- VLBW or EPT infant.
- Large babies > 4,000 gms.
- Fetus anomalies.

Classification

Soft tissue: •Abrasions •Petechial •Ecchymosis •Lacerations •SC fat necrosis.

Skull: •Caput succedaneum •Cephalohaematoma •Subgaleal or ICHge •Linear fracture.

Face: •Subconjunctival Hge •Retinal Hge.

Peripheral nerve: •Brachial plexus palsy •Unilateral vocal cord paralysis •Radial nerve palsy •Lumbosacral plexus injury.

Cranial nerve & spinal cord injuries: Facial palsy.

Musculoskeletal injuries: •Clavicular fractures •Fractures of long bones •Sternocleiodomastoid injury.

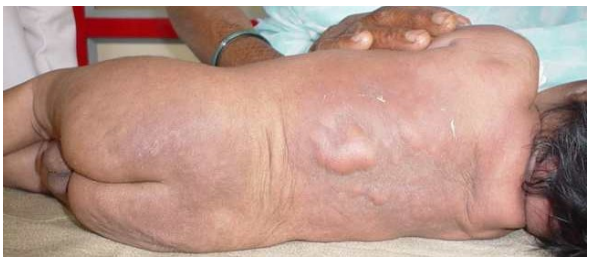
Intra-abdominal injuries: to Liver/Spleen/Adrenal/or Renal Hge.

Recognition of trauma necessitates a careful physical & neurologic evaluation of the infant to establish whether additional injuries exist. Occasionally, injury may result from resuscitation. Look for symmetry of structure & function should be assessed. Specifics such as cranial nerve, individual joint range of motion & scalp/skull integrity.

ABRASIONS & LACERATIONS

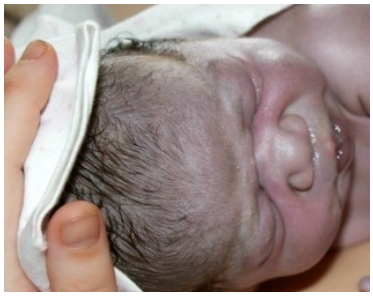
Sometimes may occur as scalpel cuts during CS or during instrumental delivery. Infection remains a risk, but most uneventfully heal. Treatment consists of careful cleaning, application of antibiotic ointment & observation. Lacerations occasionally require suturing.

FAT NECROSIS



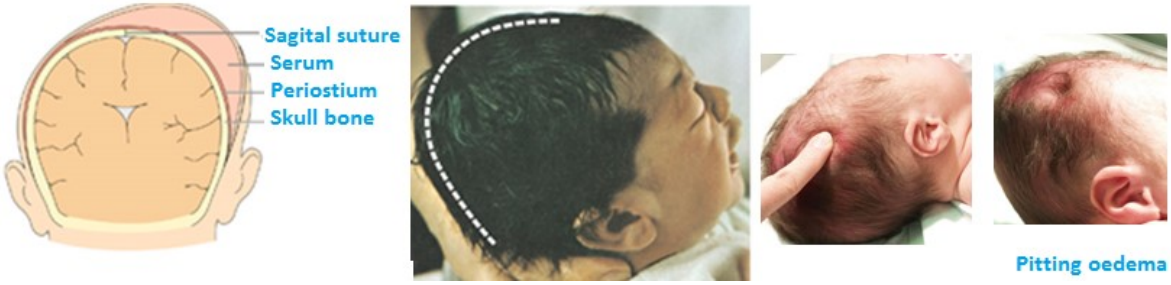
Well-circumscribed firm nodule é purplish discoloration. Usually occurs after forceps use, but can occur at other sites of trauma. Resolves spontaneously within weeks.

TRAUMATIC CYANOSIS OF FACE



Result from cord around neck, causing cyanosis of face, petechiae, ecchymosis on face, while rest of body is normal. Condition resolve spontaneously.

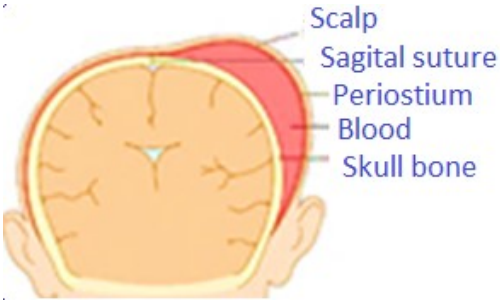
CAPUT SUCCEDANEUM

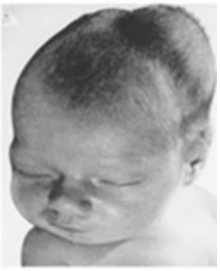


Collection of fluid (serum) under the scalp. Present at birth, as diffuse tissue edema over-

lying more than one bone (crossing suture line), has illdefined edge, soft in consistency, pits on pressure, discoloration of scalp, distortion of face, sometimes is ecchymotic, edematous. Involving the portion presenting during vertex delivery é instrument use or in case of CPD. No specific treatment & caput usually disappear within 1-2 days.

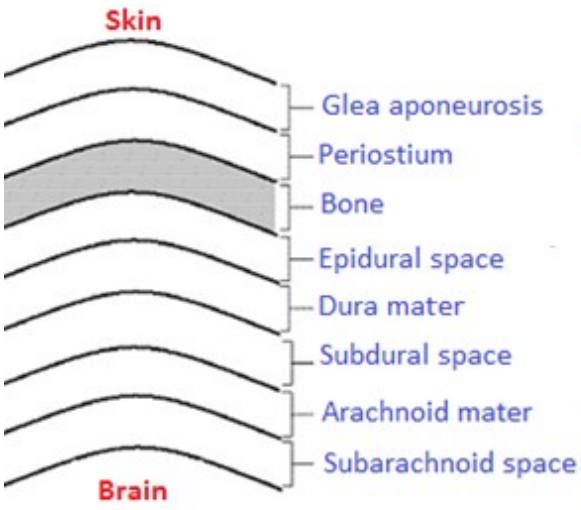
CEPHALOHAEMATOMA

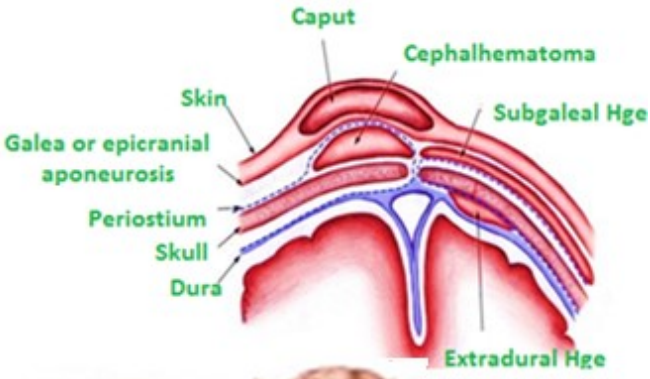


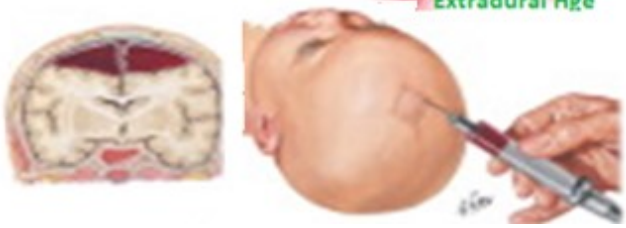


Collection of blood between the surface of a skull bone & the periostium. Not crossing suture line. Sub periosteal hematoma, commonly over parietal bone, limited by sutures, has well defined edge, elastic in consistency, not pitting on pressure. Result from difficult vacuum or forceps extraction. No discoloration of overlying scalp. Usually not visible until several hrs after birth, as subperiosteal bleeding is slow. Underlying skull fracture -linear- occasionally associated, the sensation of central depression may be suggestive but not indicative of fracture. Resolved within 2-12 wks, depending on their size, it begins to calcify by end of 2nd wk. May be associated é ICHge. No Rx usually needed, but phototherapy may be needed if it is massive, or for hyperbilirubinemia & rarely require blood transfusion.

SUBGALEAL HEMORRHAGE

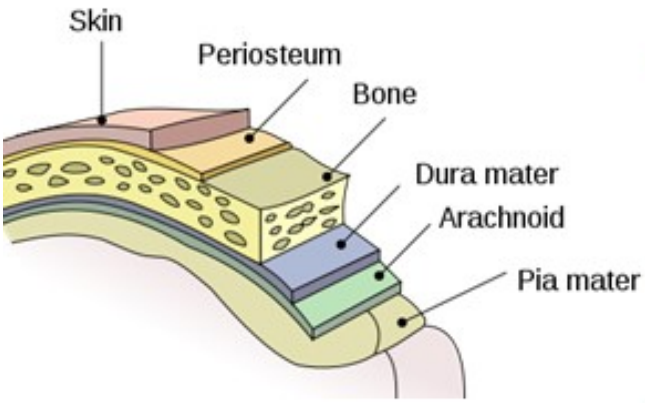


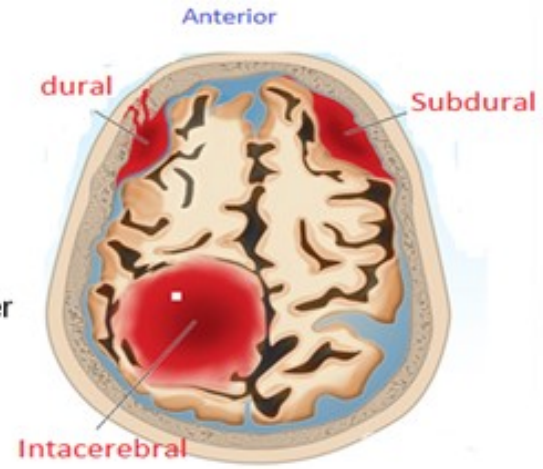




Subgaleal hemorrhage is a rare but potentially lethal condition found in newborns. It is caused by rupture of the emissary veins, ẃ are connections between the dural sinuses & the scalp veins. Blood accumulates between the glea aponeurosis & the periosteum of the scalp. This potential space extends forward to the orbital margins, backward to the nuchal ridge & laterally to the temporal fascia. In term babies, this subaponeurotic space may hold as much as 260 mL of blood. Subgaleal hemorrhage can therefore lead to severe hypovolemia & up to one-quarter of babies who require neonatal intensive care for this condition die, as blood tracks between the fibres of the occipital & frontal muscles causing bruising behind the ears, along the posterior hair line & around the eyes. May presented é shock, pallor, tachycardia & hypotension. Within 30 min of hemorrhage, the Hb & PCV start to drop rapidly. Monitoring of the bleeding times & coagulation is important. Treatment include; assessment of the level of consciousness. Assessment of the level of Hb & Hct. The ⇧ of SB is expected due to blood lyses. Aspiration of blood may required by expertise.

NEONATE INTRACRANIAL HEMORRHAGE





Types

Intracranial Hge: may result from birth trauma, asphyxia, primary hemorrhagic disturbance, or congenital vascular anomaly. Its incidence ⇧ é LBW & occur in 90% of babies 500-750 gm BW, 20% of those 1000-1500 gm BW. Rarely present at birth, 90% of cases occur between 1st - 3rd day of life & 10% of cases occur after the 1st wk of life.

Extra cerebral Hge: result from difficult labor, affect tentorium cerebella or flax cerebra venous sinus.

Intra ventricular Hge: common é preterm asphyxia, especially those <34 wk as the vasculature of the brain is very thin, or result from respiratory acidosis as for each 10 mmHg ⇧ in PCo_2, there is 50% rise in cerebral blood flow, or secondary to hypertonic solution as Na^+Hco_3 8.4%, or Ca^+ gluconate 10%, or Glucose 25%, ŵ suddenly change blood osmolality of fragile blood vessels of the immature brain.

Causes: sudden compression or decompression of the head as in breech, precipitous labor, cephalopelvic disproportion, forceps delivery, or fracture skull. In NN period 40% of convulsions are due to IC Hge, 40% are due to HIE, 20% are due to other causes as congenital anomalies of the brain, hypoglycemia, hypocalcaemia, hypomagnesaemia, meningitis, or febrile convulsions.

Predisposing Factors

•Prematurity due to physiological hypoprothrombinaemia, fragile blood vessels &liability to trauma. •Asphyxia causes anoxia of vascular wall. •Coagulopathy disorders.

Sites

Subdural; from damage of superficial veins where vein of Galen & inferior sagital sinus combine to form straight sinus, usually due to birth trauma.

Subarachnoid; damage to the vein of Galen as a result of tear in the dura at the junction of the falex cerebra & tentorium cerebella, usually due to birth trauma.

Intraventricular/Intracerebral; fetus is usually PT exposed to hypoxia.

Grades

① Germinal matrix.

② Intra ventricular.

③ Dilated ventricles.

④ Parenchymal bleeding.

Clinical Picture

- Lethargy, reluctant to feed, poor sucking. Flaccidity or spasticity.
- Altered consciousness, somnolence.
- RD (extrapulmonary), apneic attacks, irregular or periodic breathing, gasping.
- Decrease or absence of Moro reflex.
- No eye movements & pupil may be fixed & dilate.
- Convulsions, opisthotonos, rigidity or twitches.
- Vomiting.
- High pitched cry.
- Bulging & tense anterior fontanel.

Investigations

- Transfontanelle cranial U/S (as a routine for every infant < 37 wks gestation).
- CT scan & MRI.
- L P may reveal bloody CSF.
- Coagulopathy studies (BT, CT, PT, APTT) to R/O bleeding disorders.

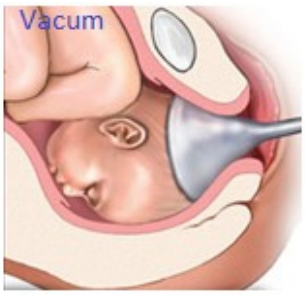
Management

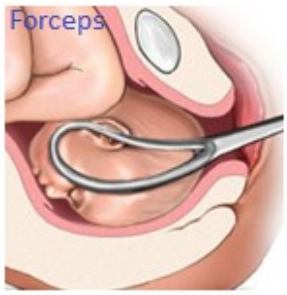
✲Minimal handling. ✲Temp control. ✲Fluid restriction to 60-80 ml/kg/day. ✲Furosomide/Lasix 40 mg amp, 1 mg/Kg/day. ✲Decadrone/Fortecortine 8 mg/2ml, 0.5mg/Kg/ dose /6 hrs, for 2 days or Mannitol 20%, 1 gm/kg/day (or 20 ml/Kg), slowly IV over 20 min, for 2 successive days é monitoring serum osmolality to be <300 mosm/L for mannitol to be effective. ✲Antibiotics: Penicillin G 1000.000u/vial, dose 100.000u/Kg, IV÷4, or Ampicillin 1000 mg/vial, 100 mg/Kg/day÷4 IV or Claforan amp 500mg/12 hrs, dose 50 mg/Kg ÷ 2 IV. ✲Vit. E 25 mg/day for 1wk. ✲Vit K: if bleeding associated é coagulation defect. ✲Packed RBCs: in case of severe anemia. ✲Shock: fluid resuscitation. ✲Metabolic acidosis: slow administration of $NaHco_3$. ✲Seizures aggressively treated é anticonvulsant: Phenobarbitone amp 200 mg/5 ml, loading dose 15mg/Kg IV over 10-15 min (high dose may cause apnea/resp depression), maintenance dose 5 mg/Kg/day÷4. Phenytoin: adding 2nd drug may needed, amp 250 mg, loading dose 15-20 mg/Kg IV, é maximum infusion of ½ mg/Kg /min, high dose cause ⇩ of BP & arrhythmia, maintenance dose 4-8mg/Kg/day. Valium: 5 mg amp, 0.25mg/Kg/dose, IV/IM stat, maintenance 0.25 mg/Kg/day ÷ 4.

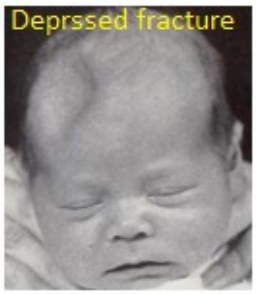
Prophylactic measures

- ⊙ Vit K 10 mg IM to mother in late pregnancy or early labor
- ⊙ Episiotomy, especially in PT & breech delivery.
- ⊙ Experienced obstetrician for forceps delivery.-

SKULL FRACTURE





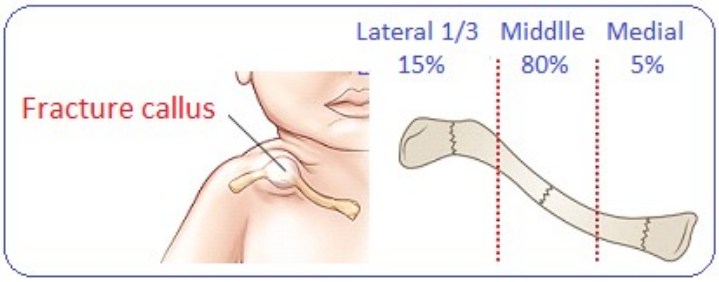


Incidence 3.7/100.000 births. 75% of cases occur é instrumental deliveries. May result from difficult labor, fetal skull pressed against symphysis or sacral promontory or ischial spine. May result from spontaneous labor or precipitous labor. Also may be seen é CS from lifting hand pressure by obstetrician, upward hand or thumb pressure over opposite side. With skull fracture the risk of development of leptomeningeal cyst due to associated dural tear ẃ leads to herniation of pia & arachnoid layers (leptomeninges) through the dural tear, also the pulsating CSF lead to progressive erosion of skull around fracture site. Any skull fracture can cause underlying ICHge, but 50% of internal Hge have no fracture. Fractures are usually linear, involving parietal bone, often associated é cephalohematoma. Skull fractures may cause no symptoms & require no Rx unless it is associated é intracranial injury, or depressed fracture forming the so called Ping-Pong fracture. Depressed fractures cause palpable (sometimes visible) step-off deformity, ẃ must be differentiated from the palpable elevated periosteal rim occurring é cephalohematoma. The linear fractures, or bigger depressed fractures >2cm require neurosurgeon consultation. Smaller one <2cm may corrected by breast pump suction or vacuum extractor suction using pressure ½ Kg/cm^2.

Investigations

•Skull X ray. • Cranial U/S. •C-T scan skull or MRI is diagnostic. •Coagulation profile: for bleeding disorders & ē suspicion of ICHge (BT, CT, PT, APTT).

FRACTURE CLAVICLE

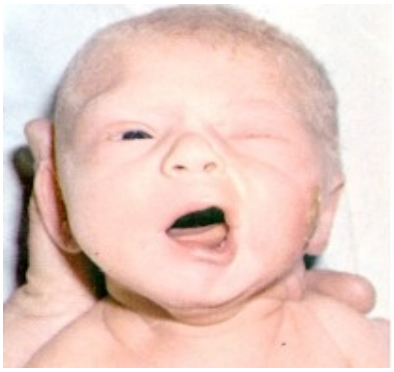


Result from difficulty in delivery of the shoulder in vertex presentations, or the presence of extended arm in breech delivery. Causing the following; Loss of Moro reflex, not moving the arm freely on the affected side. Crepitus feeling & bony irregularities may be palpated. Discolouration is occasionally seen over the fracture site. Sternocleidomastoid muscle spasm may notice. Lifting the baby under the arms is painful.

Investigations: X-Ray clavicles confirm the diagnosis.

Treatment: treated by immobilization of the affected side, figure 8-bandage for 3 weeks, hard lump develop where the bone is healing & this lump may be the only sign that the newborn had a broken collar bone.

FACIAL NERVE PALSY



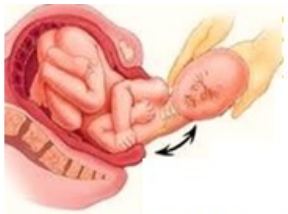
The 7th cranial nerve is mixed nerve, have a motor & a sensory root, originate from the pons, pass é auditory nerve (8th cranial nerve) inside the internal auditory canal, comes out through the styelomastoid foramen, it is motor to the muscles of expression & sensory to anterior 2/3 of the tongue. 80-90% are associated é birth trauma,while 10-20% are

associated é developmental lesion. It result from compression of the nerve between the facial bones & the mother`s pelvic bones, or as a result of compression of the nerve by blade of forceps in forceps delivery, resulting in edema & hematoma around the nerve. It is unilateral & temporarily.

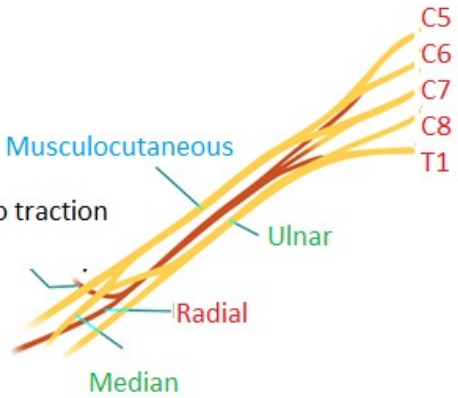
Clinical picture: absence of nasolabial fold, permanently open eye, absent blinking on affected side. The angle of mouth is deviated towards the healthy side.

Treatment: is conservative & recovery usually within the 1st month. Protect cornea é moisturizing eye drops.

BRACHIAL PLEXUS INJURIES

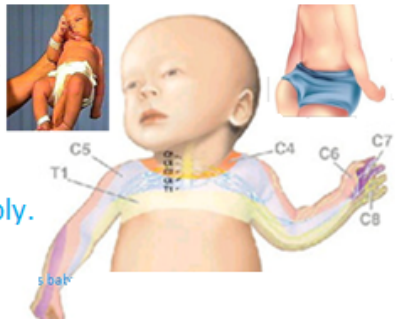


Brachial plexus is stretched due to traction



Overtraction on neck & excessive ⇧ in the angle between neck & shoulder, as in case of shoulder dystocia. It include the following types:

ERB-DUCHENNE PALSY (C5-C6)



Baby demonstrates the findings of a left sided ERB`S paralysis
Asymmetric position of the arms. The left arm is not flexed & hangs limbly.

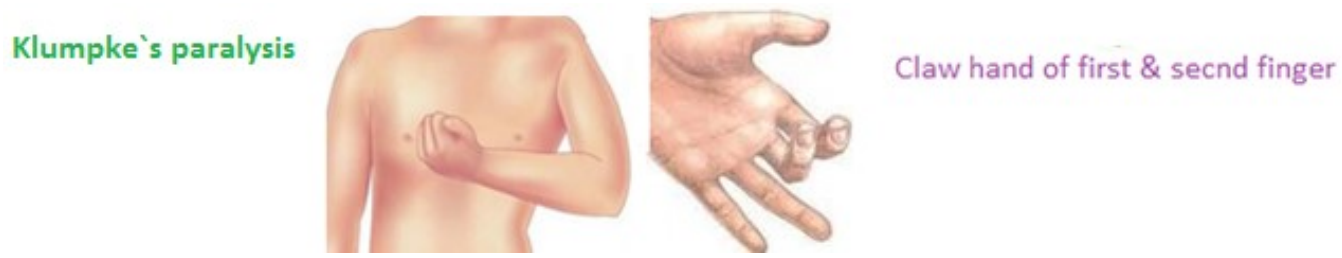
The commonest form of brachial plexus injuries, due to injury to C_5, C_6, & occasionally C_7 roots. Injury lead to paralysis of deltoid, infraspinatus & flexors muscles of the forearm.

Result in absent of Moro reflex on the affected side while the grasp reflex is intact. The upper limb drops beside the trunk, adducted shoulder, extended elbow, internally rotated limb, é flexed wrist (policeman`s or waiter`s tip hand). Abnormal positioning of the scapula (winging) & may be associated é sensory loss on lateral aspect of arm.

Management

- Physiotherapy
- Massage exercise &
- Faradic stimulation.

KLUMPIK'S PARALYSIS (C 7-8, T1)

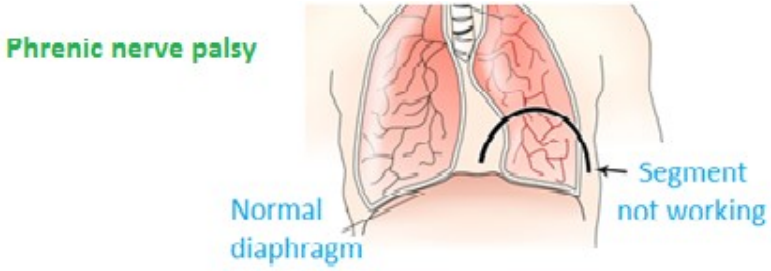


It is less common, due to injury to the 7th, 8th cervical roots. It leads to paralysis of the intrinsic muscles of the hand & weakness of the wrist & fingers flexors (claw hand). Elbow flexed. Forearm supinated. ⇩ Sensation in the palm of the hand. Absent grasp reflex. If the sympathetic fibers of the 1st thoracic root are injured it lead to Horner syndrome (ipsilateral ptosis, meiosis & anhydrosis).

Management

- Physiotherapy
- Massage exercise &
- Faradic stimulation.

PHRENIC NERVE PALSY (C 3, 4, 5)



Called kofferate syndrome. The phrenic nerve is the nerve that controls the diaphragm ẃ helps the baby to breathe. Normally as the baby inhale, the phrenic nerve tells the diaphragm to contract, ẃ enlarges his chest cavity & creates suction that draws air into his lungs. Because his nerve is damaged, it actually does the opposite effectively only gives him use of 1½ of his lungs instead of 2. This is the reason he's been breathing so hard. Result from hyperextension of neck during labor, causing overstretching or avulsion of 3rd, 4th, 5th cervical root ŵ supply phrenic nerve, leading to diaphragmatic paralysis. Cause recurrent episodes of cyanosis, irregular & labored breathing. Pneumonia can be suggested mistakenly. Breathing is completely thoracic, no bulging of abdomen.

U/S or fluoroscopic examination will show elevation of diaphragm on paralyzed side -sea saw movement of diaphragm. Recovery within 1-3 months in most cases.

Rarely surgical plication of diaphragm indicated.

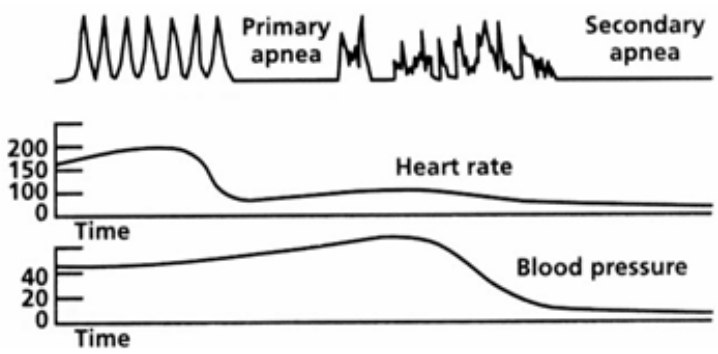
KERER'S PARALYSES

- Total brachial plexus injury.
- Entire arm paralysis.
- The most disturbing of all brachial plexus injury.
- Adynamy.
- Muscle hypotonia.
- +ve "scarf" sign..

BIRTH ASPHYXIA

For neonates undergoing transition from intra to extra uterine life approximately 10% of newborns require some assistance to begin breath at birth & only 1% require extensive resuscitative measures. Birth asphyxia is failure to initiate, sustain breathing at birth. If baby asphyxiated he took few gasps during the first min of life then develop *primary apnoea* after that he may take very few gasps in next 10 min before passing into *secondary apnoea* "terminal apnea", understanding this cycle is very important to interfere before 2ry apnoea takes place, the cardiac activity continue for about 10 min after the last gasp, the baby react to hypoxia by bradycardia, but cyanosis is late sign of hypoxia in NN due to presence of FHb ẃ possess marked affinity to O_2 causing left shift of O_2 Hb dissociation curve. Cyanosis only become visible when PaO_2 is <40 mmHg or é formation of >5 gm/dl of reduced Hb, if cyanosis is of cardiac origin it will not respond to O_2 therapy. The most liable area of brain to be affected from prolonged asphyxia in NN is parasagittal area & the asphyxiated baby is very susceptible to infection, so we put him on antibiotics.

Heart rate and Blood Pressure changes during 1ry & 2ry Apnea



Conditions ⇧ the risk of Asphyxia

Maternal Condition: DM, anaemia, AP Hge, preeclampsia, hypertension, maternal cardiac condition, chronic renal disease, blood group incompatibility, PRM & evidence of amnionitis, maternal drug or alcohol ingestion, previous NND.

Labour & Delivery Conditions: forceps or vacuum extraction, CS, cord prolapse or compr-

ession, abnormal presentation, CPD, maternal hypotension or Hge.

Fetal Condition: pre or postmaturity/IUGR, multiple births, oligo/polyhydramnios, low biophysical profile, meconium in amniotic fluid, hydrops fetalis or fetal malformation.

APGAR SCORE

The first test given to NN after birth, designed for quick evaluation of NN physical condition & the need for emergency care. Developed in 1952 by American Anaesthesiol-gist, Virginia Apgar. The test usually given to a baby twice; once at 1 min after birth, second at 5 min after birth & sometimes, if there is concern about baby`s condition or low score, test scored for 3rd time at 10 min, it include the following;

*A activity *P pulse *G grimace *A appearance *R respiration

Apgar score	0 Points	1 Point	2 Points	Points totaled
Activity (muscle tone)	Absent	Arms and legs flexed	Active movement	
Pulse	Absent	Below 100 bpm	Over 100 bpm	
Grimace (reflex irritability)	Flaccid	Some flexion of Extremities	Active motion (sneeze, cough, pull away)	
Appearance (skin color)	Blue, pale	Body pink, Extremities blue	Completely pink	
Respiration	Absent	Slow, irregular	Vigorous cry	

Severely depressed	0-3
Moderately depressed	4-6
Excellent condition	7-10

RESUSCITATION

At least 2 trained staff required for adequate resuscitation to perform PPV & chest compression if needed.

- In term or preterm babies cord should not clamped earlier than 1 min after delivery as placenta contain 100-200 ml of blood & early clamping deprive the baby from about 50 ml blood (the baby total blood volume is 70 ml/kg).
- Neonate born through clear amniotic fluid who start breathing on their own, suctionning of mouth, nose should not performed.
- In presence of meconium stained amniotic fluid, intrapartum suctioning of mouth, nose at delivery of head not recommended.
- Neonate born through meconium stained fluid, who start breathing on their own, tracheal suctioning not recommended.
- Neonate who do not start breathing, despite thorough drying & additional stimulation, PPV should started within 1 min after birth, using air rather than 100% O_2 & face mask.

With no response think in:-

- Hypovolemic or cardiogenic shock.
- Severe anemia.
- Diaphragmatic hernia.
- Pneumothorax.

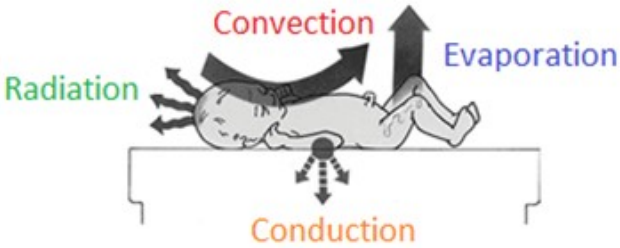
Discontinue resuscitation: if there is no detectable heart beats for 10 minutes.

Therapeutic hypothermia: 33.5-34.5^{o}C implemented within 6 hrs of birth in term infants at highest risk for brain injury from moderate to severe hypoxic encephalopathy & é further treatment in NICU, is associated é significantly fewer deaths & less neurodevelopmental disability, both cooling methods (systemic vs selective head cooling) were shown to

be effective, continue for 72 hrs after birth, rewarm over at least 4 hrs, carefully monitor adverse effects of cooling, as thrombocytopenia & hypotension.

SUGGESTED SEQUENCE OF RESUSCITATION

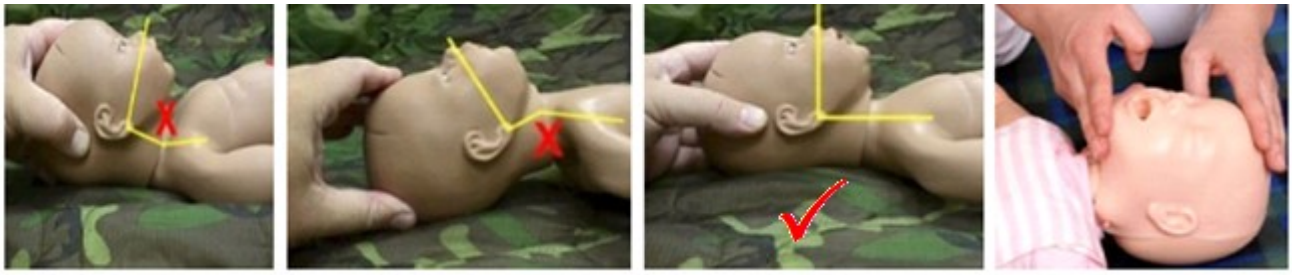
1-Keep baby warm



Prevention of cooling reduces mortality, morbidity in NN, for every 1^0C ⇩ in temp in PT infants the odds of dying ⇧ by 28%. The NN lose heat by; evaporation, radiation, convection & conduction. Babies are born small & wet, get cold very easily especially if they remain wet & in draught, preventing heat loss during resuscitation is essential, several factors ⇧ heat losses in NN; his high ratio of skin surface area to body weight, also the ⇧ heat loss & evaporative fluid loss results in massive heat losses, the thin fetal skin é blood vessels that are near to surface provides poor insulation ẃ lead to further heat loss, additionally, NN especially PT or critically ill or depressed infants have limited capacity to change body position for heat conservation, unable to accomplish flexed positioning. His limited energy stores, limited capacity for metabolic heat production, largely because of ⇩ SC brown fat stores, ẃ is more prominent in PT & SGA, in addition to incapability of effective shivering, ẃ is major source of heat production in adult, contraibutes to get cold very easily. Cold stress can lead to metabolic acidosis, hypoglycemia, cerebral injury. The goal during resuscitation tion is to keep the axillary temp ≥ 36.5 ^{o}C, baby should be dried é prewarmed blankets or towels, remove the wet towels, placed on prewarmed heat source, open bed warmers, ẃ use radiant heat. The PT <500 gm is best pla-

ced éout drying, into food-grade plastic wrapping under radiant heater, woolen head cap should be used, this process provide significant stimulation, allow time to assess tone ,RR, HR. The infant temp should be documented as soon as possible after birth & every 10-15 min thereafter until continuous temp stabilized, another source of heat loss is the use of unheated non-humidified O_2 or gas during resuscitation.

2-Positioning



Ensure patency of baby airway, place the baby on his back é the head in neutral position (neck neither flexed nor extended), in some babies the occiput is prominent, so place some support under shoulders (2 cm thickness blanket or towel) é care not to over extend the neck, if the baby very floppy, it may be necessary to apply chin lift or jaw thrust as in the photo.

3-Suctioning

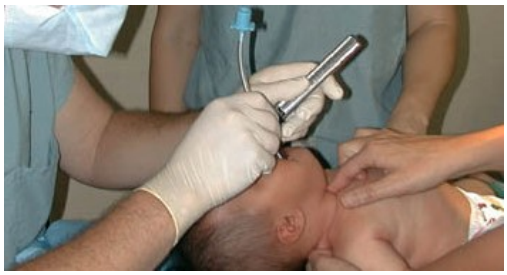
In the last 5 yrs, value of suctioning baby`s nose, mouth at birth as a routine procedure, has been questioned, it is now believed that most healthy babies do not require any suctioning & are quite capable of clearing their airways on their own, the fact that babies, on average are born é up to 75-100ml of amniotic fluid in their lungs already (being absorbed within 24 hrs after birth), means that another 1-2 ml from their nose &/or mouth will not make much difference, suction using suction bulb needed only if there is airway obstruction. Air way obstruction may be caused by particulate meconium but can also caused by blood clot, or thick tenacious mucous or vernix, even in deliveries where

meconium staining is not present, suction better done under direct vision using suction catheter 6, 8 FG, connected to suction source pressure 50-100 mmHg, when thick meconium or mucous cannot removed by suction bulb, vigorous suction of nose é catheter can lead to edema of nasal tissues & resultant RD after infant leave the delivery room, suction duration each time < 4 sec to avoid parasympathetic stimulation & reflex bradycardia. Suction is done through ETT using straight blade laryngoscope size 0 or 1 & ETT size 2.5-3 FG (simply the diameter is equal to top of finger or nostril of baby).

Size of ETT & depth of insertion according to BW

Weight	ETT tube size	Depth of insertion
1 Kg	2.5	7 cm
2 Kg	3	8 cm
3 Kg	3.5	9 cm
4 Kg	4	10 cm

(NB: Adult female 7 - 8 size & 20 -22 cm depth & Adult males 8 - 8.5 size & 20 -22 cm depth.



9-10 cm at the lip for this term infant

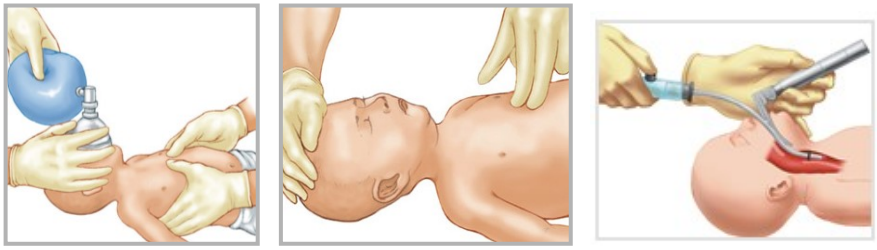
Technique of intubation: tip of laryngoscope passed over tongue sweeping it out of the way, the epiglottis is visualised, gently lifted by the laryngoscope blade, the vocal cords seen behind the epiglottis, ETT is held by right hand, introduced through the visualised vocal cords, the centimeter mark at the lip margin is noted, site confirmed by CXR, the end of ETT to be at the level of 1st thoracic vertebrae (above the carina ẃ is site of bifurcation of trachea), if suction done through nose, the distance of suction catheter to pass is equal to double length of ETT. Lip reference mark (cm) = 6 + BW.

4-Assess

Tone, color, respiration, apex beat, heart rate. The healthy baby cry within few sec of delivery, have good HR within a few min of birth (120-150/min), pulse also can assessed by feeling it at the base of the umbilical cord. The less healthy baby appear blue at birth, less good tone, may have HR < 100/min & may not establish adequate breathing by 90-120 sec. Very ill baby will looks pale, floppy, not breathing, slow, very slow heart sound.

The 1st sign of improvement during resuscitation is the ⇧ in HR ẃ remain the most sensitive indicator of resuscitation efficacy. Pulse oximeter should be considered during resuscitation if babies are persistently cyanosed or have labored breathing, to achieve O_2 saturation 92-96 % in term infant & 88-92% in PT through 100% O_2. If Baby gasping or not breathing, open the airway & give 5 inflation breaths (pressure 20-30 mmHg), can be repeated in case of no response, using infantile self-inflating bag, assure sealing of mask & the characteristic noise as the valve opens, consider using pulse oximeter & listen to the heart, or feel umbilical cord. If HR is not detectable or slow (< 60/min), start active resuscitation.

Active resuscitation



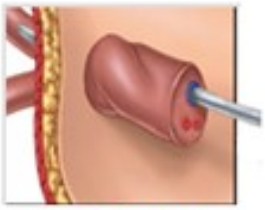
Active resuscitation

Chest compression, 3 compressions to each 1 breath é cooperation of one of the attending staff, cardiac massage done through, either 2 finger or 2 thumb techniques, to depress lower half of sternum (1 finger breadth below the inter nipple line or approximately 1/3 of the antroposterior diameter of chest), 100% O_2 to be used & if resuscitation is suc-

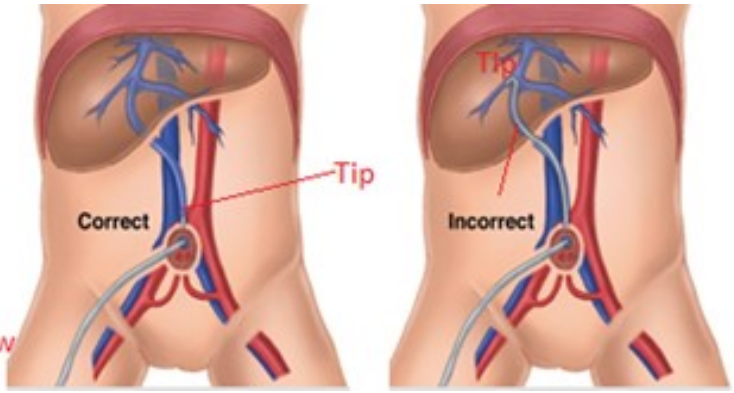
cessful O_2 can be blended é air to achieve target O_2 saturation 80% at 5 min & 90% at 10 min. If you notice no improvement, put umbilical catheter.

Normal Oxygen saturation at birth

1 minute	60-65%
2 minutes	65-70%
3 minutes	70-75%
4 minutes	75-80%
5 minutes	80-85%
10 minutes	85-95%



Into the vein
Notice the 2 arteries below



Umbilical Catheterization

Technique of umbilical catheterization

Prepare abdomen, umbilicus é antiseptic sol. (surgical preparation, alcohol then pethidine), cover é sterile towels, using scalpel cut the cord horizontally, 1.5-2 cm from abdominal wall, 2 thick walled small arteries & 1 thin wall large vein identified, dilating the umbilical vein using forceps, clearing thrombus, 3.5 FG catheter is used for PT & 5 FG catheter for FT, flush the catheter é heparinized saline, attach it to 3 way stopcock, measure & mark 5 cm from tip of catheter, advance catheter gently towards liver, only 1-2 cm beyond the point at ẃ good blood return is obtained, this approximately 4-5 cm in FT, secure catheter é suture through the cord & tape bridge plaster, remove catheter when resuscitation is complete & peripheral vascular access obtained.

Medications

Adrenaline: indicated if HR < 60 after 30 sec of coordinated ventilation & compressions. Amp 1 ml, 1/1000 conc, dilute é distilled water to conc of 1/10.000, 0.1 mg/ml, 0.1-0.3 ml/Kg (1 ml FT, 0.5 ml PT & 0.25 ml EPT), given through umbilical vein or ETT. Dose may repeated é no response after 5-10 min in case of asystole.

Atropine: amp 1 ml contain 1 mg, 0.1 mg/Kg (0.1 ml/Kg) to be given as IV bolus in case of persistent bradycardia.

Ca gluconate: 10%: ampule 10 ml, 0.3 ml/Kg slowly & strictly IV.

Dopamine/Dubutamine: if perfusion is permanently poor, shock, hypotension. Dose calculated according to BW X 3 = number of mg to collected from vial Dopamine (200mg in 5ml amp = 40 mg/ml) by insulin syringe, add 50 ml glucose 5%, give 2 ml/ hr, this is equal to 2ug/Kg/ min, the dose can doubled to 5 ug/Kg/min, this ⇧ blood supply to internal organs. Dose 6-10 ug/ Kg/min will have the same previous effect in addition to +ve inotropic & +ve chronotropic effect on heart. Much higher dose from 11-15 ug/Kg/ min causes the same previous effect in addition to peripheral VC.

Fluids (volume replacement): indicated é the no response to resuscitation or é evidence of blood loss. include the following;

- Dextrose 10%: 250 mg/kg (2.5 ml/Kg).
- Normal saline (0.9%): 10-20 ml/Kg as bolus given over 10-20 sec, if there is clear history of blood loss, using isotonic crystalloid rather than albumin is preferred for emergency volume replacement until blood is ready, this often produce rapid response & may repeated safely.
- Ringers or Blood: as alternatives. The route is IV (through the umbilical vein).

Sodium Bicarbonate: indicated é documented or assumed metabolic acidosis. Concentra-

tion 4.2% (0.5 meq/ml) at a dose of 2 meq/kg, given IV through the umbilical vein, amp 20 ml of 8.4%, the ideal is to use conc 4.2%, it should be given only after establishment of adequate ventilation & circulation, for correction of persistent metabolic acidosis, be directed by ABG levels, may induce acute iatrogenic gradient between plasma & brain cells or alveoli causing intraventricular or pulmonary Hge.

Naloxone (Narcan): indicated in case of severe respiratory depression after PPV has restored a normal HR & color. History of maternal narcotic administration within the past 4 hr, amp 1 ml contain 0.4mg, dose 0.1 ml/kg IM or IV bolus. It can repeated after 5 min.

Hypoglycemia: BG < 45-60 mg/dl, 5 ml/kg of Dextrose 10% IV.

Other drugs: rarely used include:-

- Glucagon 1 ml = 1 mg, 0.1 ml/Kg as IV bolus.
- Dexamethasone amp 8 mg/2 ml, 0.5 mg/Kg/dose.
- Mannitol, Lasix, Phenobarbitone, all are rarely used.

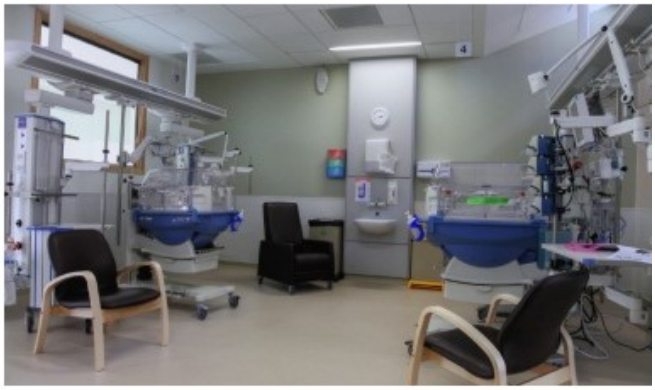
ESSENTIAL IN THE DELIVERY ROOM AND THEATRE

Checking mother file, all equipment, instruments, medications, all are functioning & are ready for use: ❁Firm padded resuscitation surface ❁Overhead warmer. ❁Light for the area. ❁Clock é timer in seconds. ❁Warmed towels or blankets. ❁Polyethylene bag (plastic bag) or sheet big enough for baby <500 gm or < 32 wks gestation & bonnet (head cover). ❁Stethoscope, NN size preferred. ❁Pulse oximeter + disposable neonatal probe. ❁Air & O_2 supply é blender attached. ❁Resuscitation bag & mask (Laerdal) é suitable mask fitted & flow driven T-piece (Neopuff). ❁Suction machine (pressure <100 mmHg). ❁Suction catheters, size 6, 8, 10, 12FG. ❁Laryngoscope é straight blade, size 00, 0, 1. ❁Oropharangyeal airways, size 00, 0. ❁Uncuffed ETT size 2.5, 3, 3.5 FG. ❁Supplies for fixing the ETT (eg. scissors, tape). ❁Feeding tubes for gastric decompression, size 6, 8 FG.

❁Umbilical venous catheter, size 3.5, 5 F. ❁Peripheral canulation insertion equipment (butter fly size 23F & IV cannula size 18, 20, 22F). ❁Scalpel, silk suture (3/0). ❁Small clamps, Needle holder, Scissors. ❁Syringes & needles different sizes, 3-way stopcock. ❁Sterile gauze, adhesive plaster, Pethidine & alcohol solution. ❁Blood gas syringes, tubes (Plain, EDTA, Florid). ❁Intraosseous needle, 50mm length available if umbilical venous catheter unsuccessful in term infant, or é emergency or failure of other methods (site 1-3 cm below tibial tuberosity or 2-3 cm above external condyle femur). ❁Adrenalin 1mg/1ml (1/1000) amp, (dilute to 1/10.000 conc to reach conc of 0.1 mg/ml. ❁$NaHco_3$ (8.4%, 20ml =20meq) & Ca gluconate amp (10% 1 gm/10ml, 1ml =100mg) ❁Naloxone (0.4mg/1ml) & atropine amp (1mg/1ml). ❁Dubutamine amp (200mg/5ml) ❁G 5 & 10%, normal saline, distilled water. ❁Decadrone amp (4mg/1ml)❁Group O-ve blood available in fridge of the theatre. ❁Spare batteries, bulbs. ❁Portable incubator ready to use.

SPECIFICATIONS OF NEONATAL SPECIAL CARE UNIT

- Temp: 20 -22 ^{0}C, humidity 40%
- The presence of ventilation filters.
- Basin every 8 steps from incubator.
- Between each other incubator distance 2.5 meters.
- Every incubator has area of 8 square meters.
- One nurse for every 1-2 incubators.



Cleaning of the incubator

Taking out all content, cleaning each separately ē soap & water, incubator cleaned ē alcohol after washing ē soap & water & leave to dry. Humidifier every 24 hrs to be washed ē soap & water, filled again ē distilled water, special instructions of manufacturing company to be followed.

Baby in incubator

- Be sure that the baby is under the NTE.
- Oxygen is 30- 40 %, to start ē.
- Humidity is 40-60%.

Examination of baby: immediately after birth, check for apex beat, absence of apparent congenital anomaly as spina bifida, cleft palate, imperforate anus, followed by thorough medical examination after 24 hrs.

Care of the baby after birth: cleaning é sterile water & cotton once his body temp stabilized (mild baby shampoo may be used), cleaning his eyes ē sterile cotton & warm water, antibiotic eye drops as prophylactic for neisseria gonococci, Vit K PO 1 mg or IM 0.5 mg. Healthy baby after birth is alert & active for 1-2 hrs then goes into sleep to recover from stress of labor for 1-2 hrs then again, the 2nd period of activity lasts for 1-2 hrs during ẃ he demand feeding. .

Facing the mother: madam your baby is very nice & in good health, he/she is crying, playing & laughing, his/her body weight is…, look at him, touch him & after sometime you are going to feed him.

Remember: in general nonverbal cues as tone of voice, body language, or comment to other staff often give parents a massage different from the spoken words, parents usually coming to hospital very concerned, worried, so reassuring them is needed.

Always be

Be professional.

Be consistent.

Be yourself.

Never appear to be rush & try to establish confidentiality.

Feeding: start feeding once mother condition allowing, for the value of colostrum during the first few days, also good nutrition of mother & mobilization of mother as early as possible is recommended.

Artificial feeding: under certain circumstances artificial milk is allowed, e.g. PT, illness of mother, maternal AIDS, breast milk jaundice, presence of hare lip &/or cleft palate, multiple pregnancy. The feeds to be prepared under aseptic technique, sterilization of utensils used by boiling for 5 min & disinfectant may be used, water to be boiled, preparing nearly the exact amount to be given & using it within 4 hrs from time of starting feeding, it may be kept in refrigerator after preparation for 24 hrs & used within 4 hrs from opening it, mother can prepare the whole 24 hrs feeds & to keep them in refrigerator to be used within 24 hrs & each bottle to be used within 4 hrs from taking it out of refrigerator, the amount of feed in ml can be calculated as (body weight in grams ÷ 36, for a total of 6 feeds/day), but in the first 2 months of life feeding is usually a demand feeding.

Expressed milk: allowed for mother é contracted nipple, or baby who is unable to suck, PT, cleft palate. The milk is collected under aseptic technique using breast pump ẃ must be cleaned thoroughly each time to be used, sterilization using disinfectant. The expressed milk may kept in refrigerator, used within 2 days from time of collection or during 6 months if frozen to **<** -20 ^{0}C.

Solace: you knowing, that the situation was so.., in fact the child tried hard to live & we tried various ways by doing…….. but as a result of....., the child cannot afford life, i know how much you are sad & that the next days or wks will be hard for you, but gradually é the passage of days you are going back to your normal life & forgotten those days.

CHAPTER II

PERINATOLOGY 2

- At High Risk Pregnancy
- Indications For Caesarean Section
- Intra Uterine Infection
- Prematurity
- Small For Gestational Age
- Large For Gestational Age
- Post Term Baby
- Intra Uterine Foetal Death
- Neonatal Jaundice
- Necrotizing Enterocolitis
- Neonatal Convulsion
- Neonate Intracranial Haemorrhage
- Cerebral Palsy

AT HIGH RISK PREGNANCY

Is one in ẃ some condition puts the mother, the developing fetus, or both at higher-than-normal risk for complications during or after the pregnancy & birth.

Causes

1- Personal Causes

- Age <18 yrs old: ⇧ incidence of abortion, PT labor, IUGR & preeclampsia.
- Age >35 yrs old: ⇧ incidence of Down syndrome, preeclampsia, IUGR, FPD.
- Lives far from hospital or health facility: ⇨ birth trauma, asphyxia & hypothermia.
- +ve consanguinity: history of congenital malformation, repeated abortion.
- Smoking: ⇧ incidence of abortion, PRM, PT, LBW & placental abruption.
- Long duration of marriage ē infertility & use of ovulatory drugs: ⇧ incidence of anxiety ē pregnancy/ labor, multiple pregnancies, PT labor & ectopic pregnancy.

2- Obstetrical Causes

- Parity ≥ 5: ⇨ prolonged or obstructed labor, uterine rupture & FPD.
- No Spacing: ⇨ nutritional deficiencies & weak general health.
- Previous IUFD or NND: ⇨ recurrence of risk factors, fetal malformation & IUGR.
- Previous SGA: recurrence of risk factors, IUGR & FPD.
- Previous LGA: ⇨ recurrence of risk factors, gestational DM, birth trauma.

- Previous Fetal Malformation: ⇨ hereditary disorders & cong. anomalies.
- Previous Abortion: ⇨ recurrence of risk factors.
- Previous PT Labor: ⇨ persistence of risk factors.
- Previous CS: ⇨ uterine rupture.
- Previous Retained placenta or PP Hge: ⇨recurrence of the problem.
- Previous Rh isoimmunisation: ⇨ still birth & feto-maternal incompatibility.
- Duration of Labor < 4 hr: delivery on way to hospital, NN asphyxia, hypothermia.
- Previous Instrumental Delivery: ⇨ prolonged or obstructed labor, uterine rupture.

3- Past history

- Hypertension: ⇨ exaggerated hypertension, FPD & renal affection.
- Heart disease/murmur: ⇨ heart failure & pulm edema.
- TB or intake of anti-TB drugs: ⇨ teratogenicity of anti-TB drugs, IUI.
- Epilepsy or intake of antiepileptic drugs: ⇨ teratogenicity, & traumatic seizures.
- Chronic illness: may affect the pregnancy or vice versa.
- Previous myomectomy: ⇨ uterine rupture, abnormal placentation, APHge & PPHge.
- Previous cerclage: ⇨ incompetent cervix, ⇧ incidence of abortion & PT labor.
- Uterine anomalies, fibroid, or pelvic masses: ⇨ 2nd TM miscarriage, premature labor, IUGR, APHge, PPHge, abnormal placentation, uterine rupture, abdominal pain (fibroid degeneration).

4- Family history

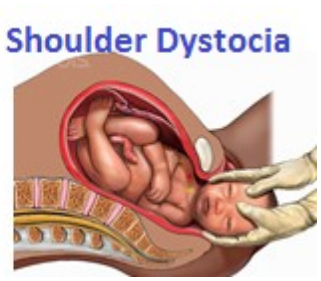
- Fetal abnormalities: ⇧ incidence of fetal anomalies.
- Twins/multiple pregnancies of mother &sister: ⇧ incidence of multiple pregnancy.
- Hypertension: ⇨ pregnancy aggravated or induced hypertension.
- DM: ⇨ gestational diabetes, spontaneous abortion, LGA & cong. anomalies.

5-On-going maternal or fetal problem

- Unknown last menstrual period: ⇨ postdate, failure diagnosis of IUGR.
- Gait "limping": ⇨ CPD & obstructed labor.
- Color "pallor": anemia é pregnancy, IUGR, & PT labor.
- Color "jaundice": biliary colic, obstructive jaundice & Cholecystitis.
- Maternal weight >90 kg: ⇨ gestational diabetes, hypertension, macrosomic infant, dystocia, CS, wound or episiotomy infection.
- Maternal weight <45 kg: ⇨ IUGR, PT labor & FPD.
- Hyperemesis gravidarum: ⇨ hypertension, tachycardia, dehydration, weight loss, electrolyte imbalance.
- Non immune against tetanus: ⇨ tetanus neonatorum.
- Absent fetal movement: IUFD & molar pregnancy.
- Marked changes in frequency/intensity of fetal movements: IUFD, IUGR.
- Uterine size <gestational age: IUFD, IUGR, oligohydramnios.
- Uterine size > gestational age: DM, multiple pregnancy, Polyhydramnios.
- Vaginal bleeding in early pregnancy:ectopic pregnancy,threatened/missed abortion.
- BP >140/90 mmHg: ⇧ incidence of eclampsia, IUGR, FPD, renal disease.
- Amniotic fluid >2 L (Polyhydramnios): PPHge, anencephaly, oesophageal/duodenal atresia , Down sy., neuromuscular disorders, fetal macrosomia, preterm labor.
- Amniotic fluid <500 ml at 32-36 wks gestation (oligohydramnios): renal agenesis, hypoplasia, or aplasia, ureteral or pulm. hypoplasia, IUGR & IUFD.
- 3rd TM bleeding: IUFD, IUGR, placenta praevia, APHge or PPHge.
- Sudden gush of vaginal water: ⇨ PROM, preterm baby, cord prolapse, Chorioamnionitis , NN infection.

- Hb < 11 gm: anemia in pregnancy, PT baby & IUGR.
- Proteinuria > +1: UTI, renal disease, & preeclampsia.
- Glycosuria: gestational DM.
- Rubella: ⇨ severe fetal damage if infection occurred during 1st months, heart damage, cataract, deafness, hepatosplenomegaly & MR.
- Herpes: ⇨ spontaneous abortion, reactivation of infection, PT baby, IUGR, IUFD & NN herpes infection.
- No head engagement at 40 wks gestation: ⇨ malpresntation & CPD.
- Bacteriuria > 100.000 bacteria in urine culture: ⇨ UTI & Pyelitis.

DIABETIC MOTHER



Gestational DM

First diagnosed during pregnancy seen in 10% of all pregnancies & it indicates predisposition to later development of type II DM. The chance of recurrence in future pregnancies is 50%.

Risk factors: •Maternal age >25 yrs •BMI >25 •Race/Ethnicity: Latina, Native American, South & East Asian} •Positive personal/Family history. •History of LGA baby.

Types: Type A1: normal FBG & 1-2 hrs after meals, but abnormal OGTT. Diet modification is sufficient to control BG levels.

Type A2: abnormal FBG &/or after meals + abnormal OGTT. Additional treatment é insulin or other drugs is required.

Pregestational DM

Type 1 DM is autoimmune process that destroys pancreatic B cells.

Type 2 DM is acquired insulin resistance related to obesity.

Clinical implications

- Obstetric complications: ⇧ incidence of miscarriage. ⇧ incidence of congenital malformations 4 X higher than in general population. Association é hypertensive disorders of pre-gnancy & preeclampsia. Association é premature delivery, IUFD, traumatic delivery (e.g. shoulder dystocia), vacuum or forceps-assisted delivery.
- Fetal Macrosomia: disproportionate amount of adipose tissue concentrated around the shoulders & chest.
- Neonatal metabolic abnormalities: hypoglycemia, hyperbilirubinemia/jaundice.
- RDS, Organomegaly, Polycythemia, ⇧ Perinatal mortality.
- Predisposition to childhood obesity.

Management

Insulin: significant benefit of insulin treatment. Prior to insulin use, the perinatal mortality was 65%, while after introduction of insulin, the perinatal mortality ⇩ to 5%. During management the Hb A1C level should be ≤ 6%, levels between 5-6% are associated é fetal malformation rates equal to those observed in normal pregnancies (2-3%). Goal of normal or near-normal Hb A1C level for at least 3months prior to conception. Hb A1C concentration near 10% is associated é fetal anomaly rate of 20-25%.

BG goals during pregnancy is FBS <100 mg/dl, 1-HPPBS <140, 2-HPPBS am <200 mg/dl. Nocturnal BG level should not go <60mg/dl. Abnormal PPBS measurement is more predictive of adverse outcomes than pre-prandial measurements. Optimize glycemic control through frequent insulin dose adjustments.

In type 1 DM, often have insulin pump. In type 2 DM give SC insulin. Fetal monitoring starting at 28-32 wks, depending on glycemic control. U/S to be done to assess the fetal growth at 36 wks. Delivery at 38-39 wks. For known case of DM Type II, to stop oral hypoglycemic agents & change to insulin. Reassure that the risk of congenital abnormality due to insulin is small. The SC insulin requirements ⇧ rapidly, especially from 28-32 wks of gestation. During the 1st TM the dose of insulin is 0.6-0.8 U/kg/day & during the 2nd TM is 0.8 - 1.0 U/kg/day & in the 3rd TM is 1.0 -1.2 U/kg/day.

Nutrition: caloric requirements: 30 kcal/kg/day, distributed as: 20% at breakfast, 20% at lunch, 30% at dinner & 30% for snacks (to avoid hypoglycemia).

Caloric composition: 50% from complex, high-fiber COH, 20% from protein & 30% from primarily unsaturated fats.

HYPERTENSIVE MOTHER

Complicates 10% of pregnancies. ⇧ of SBP ≥140 mmHg &/ or DBP ≥ 90 mmHg, on 2 occasions at least 6 hrs apart.

Chronic Hypertension

SBP ≥140 mmHg, DBP ≥90 mmHg, or both, presents before 20th wk of gestation or persists >12 wks postpartum.

Causes

- Primary “Essential”.
- Secondary from other medical condition e.g. renal disease.

Management

ECG should be obtained in women é long-standing hypertension. Base-line laboratory tests including; urinalysis & culture, serum creatinine, glucose & electrolytes, in order to R/O renal disease & identify comorbidities such as DM. Women é proteinuria on urine di-

pstick should have a quantitative test for urine protein. Avoid treatment in women é uncomplicated mild essential hypertension as BP may ⇩ as pregnancy progresses. May taper or D/C medications for women é BP <120/80 in 1st TM. Reinstitute or initiate Rx for persistent DBP >95 mmHg, SBP >150 mmHg, or é signs of hypertensive endorgan damage. Medications of choices are oral Methyldopa (centrally acting anti hypertensive) & Labetalol (cardioselective B blocker)

Preeclampsia: new onset of hypertension & proteinuria after 20 wks gestation. SBP ≥140 mmHg or DBP ≥ 90 mmHg & proteinuria of ≥ 0.3 gm in a 24 hrs urine. In case of preeclampsia before 20 wks, think in molar pregnancy !.

Eclampsia: generalized convulsion &/or coma in the setting of preeclampsia, é no other neurological condition. Severe preeclampsia must have one of the following;

- CNS dysfunction (blurred vision, scotoma, altered mental status, severe headache).
- Symptoms of liver capsule distention (right upper quadrant or epigastric pain).
- Nausea, vomiting.
- Hepatocellular injury (serum transaminase concentration at least twice normal).
- SBP ≥ 160 mmHg or DBP ≥ 110 mmHg on 2 occasions at least 6 hours apart.
- Thrombocytopenia (<100,000 platelets/ml).
- Proteinuria ≥ 5 gm in 24 hours. or Oliguria < 500 mL in 24 hours.
- Severe IUGR.
- Pulmonary edema or cyanosis.

Preeclampsia superimposed on chronic hypertension

Preexisting hypertension é the following additional signs/symptoms:-

- New onset proteinuria.
- Hypertension & proteinuria beginning prior to 20 wks of gestation or sudden ⇧ in BP.

- Thrombocytopenia.
- ⇧ Aminotransferases.

Management: definitive treatment is delivery. The major indication for antihypertensive drugs is prevention of stroke, in case of DBP ≥110 mmHg or SBP ≥160 mmHg.

Medications of choices are; IV Labetalol or Hydralazine, SR Nifedipine (calcium channel blocker) in the acute stat & Oral Methyldopa or Labetalol as long term Rx.

Gestational hypertension

Mild hypertension éout proteinuria or other signs of preeclampsia. Develops in late pregnancy, after 20 wks gestation. Resolves by 12 wks postpartum. Can progress into preeclampsia, often when hypertension develops < 30 wks gestation. The indications for & choice of antihypertensive treatment are the same as for women é preeclampsia.

Risk factors for hypertension é pregnancy

▲Null parity. ▲Preeclampsia in a previous pregnancy. ▲Age >40 yrs or <18 yrs. ▲Family history of pregnancy-induced hypertension. ▲Chronic hypertension. ▲Chronic renal disease. ▲Anti-phospholipid antibody sy or inherited thrombophilia. ▲Vascular or connective tissue disease. ▲DM (pregestational/gestational). ▲Multifetal gestation. ▲High BMI. ▲Male partner whose previous partner had preeclampsia. ▲Hydrops fetalis.

Evaluation of hypertension in pregnancy

History: complaint, Hx of preeclampsia, past medical Hx, past family Hx, past obstetrical Hx, past gynecological Hx, social Hx, medications, allergy Hx.

Physical examination: vital signs, vision, CVS, respiratory system, abdominal examination (epigastric or RUQ pain), neuromuscular & extremities (reflexes, clonus, oedema). Fetal assessment (U/S, biophysical profile, NST).

Laboratory investigations: CBC, RFTs, LFTs, Coagulation profile, Urine protein.

Management of hypertension in pregnancy

Depends on severity & gestational age.

Observation

- Restricted activity & Close maternal & fetal monitoring (NST/U/S).
- BP monitoring & observation for manifestations of preeclampsia.
- Routine weekly or biweekly blood work.

Medical Rx

Acute Rx: IV Labetalol, Hydralazine, SR Nifedipine.

Expectant Rx: oral Labetalol, Methyldopa, Nifedipine. Eclampsia prevention ($MgSO_4$). The contraindicated drugs during pregnancy include; ACEIs & ARBs.

Delivery: vaginal delivery vs CS depends on severity! May need to administer ante-natal corticosteroids depending on gestational stage!

Rh NEGATIVE MOTHER

known as Rhesus incompatibility, Rhesus disease, RhD hemolytic disease of NN. When Rh-ve mother gets pregnant to Rh +ve fetus, she may be sensitized to Rh antigen & develop antibodies, these will cross the placenta causing hemolysis of fetal RBCs. The rhesus system comprises number of antigens C, D, E, c, e. The Rh isoimmunisation is due to D antigen in > 90% of cases. The individual having the antigen on the human RBCs is called Rh +ve & in whom it is not present is called Rh -ve.

Incidence: 15 % of Caucasians, 5 % African Americans & 2 % of Asians are Rh -ve.

Mechanism of antibody formation in the mother

Antibody formation occurs by isoimmunization, ŵ is defined as the production of immune antibodies in an individual in response to an antigen derived from another individual

of the same species provided that the first one lacks the antigen. This occurs in two stages; sensitization & immunization. In ABO-Blood groups, a naturally occurring anti-A & anti-B are present in the serum. But in Rh group there is no such naturally occurring antibodies. So for the first time when Rh +ve fetal RBCs enter mother's blood, they remain in the circulation for their remaining life span. There after they are removed by the RES & are broken down é liberation of antigen ŵ triggers the isoimmunization. Since it takes as long as 6 months for detectable antibodies to develop, the isoimmunization in 1st pregnancy is unlikely. If the fetomaternal bleed is < 0.1 ml, the antibody production sufficient to produce isoimmunization is unlikely. The main effect of Rh antibodies is on the baby in the form of hemolytic disease of the newborn. If the baby is Rh +ve & the mother is Rh -ve, in the sensitized mother the antibody becomes attached to the antigen on the surface of fetal erythrocytes. The effected fetal cells are rapidly removed from the circulation by the RES. Depending upon the degree of agglutination & destruction of the fetal RBCs. Various types of fetal hemolytic diseases appear including the following:-

Congenital anemia of NN: is mildest form of the disease where hemolysis is going on slowly. The destruction of RBCs continues up to 6 wks after ŵ the antibodies are not available for hemolysis. So the neonate may require blood transfusion.

Icterus gravis neonatorum: the baby is born alive éout evidence of jaundice but soon develops it within 24 hrs of birth. If bilirubin level ⇧ to critical level of 20 mg/100ml then bilirubin crosses the blood brain barrier to damage the basal nuclei of the brain producing clinical manifestations of Kernicterus & may require exchange transfusion.

Hydrops fetalis: excessive destruction of the fetal RBCs leads to severe anemia, tissue anoxaemia & metabolic acidosis. These have got adverse effects on the fetal heart, brain &on the placenta. Hyperplasia of the placental tissue occurs in an effort to ⇧ the transfer

of oxygen. As a result of fetal anoxaemia there is damage to the liver leading to hypoproteinemia ŵ is responsible for generalized edema, ascites & hydrothorax. Fetal death occurs sooner or later due to cardiac failure. Baby is either still born or macerated & even if it is born alive dies soon after.

Affection in the mother: ⇧ incidence of preeclampsia. Polyhydramnios. Big size baby. Hypofibrinogenemia due to prolonged retention of dead fetus. Repeated abortions.

Diagnosis

Past history: previous transfusions, previous normal fetus & in subsequent pregnancies baby presenting ē hemolytic disease, or Hx of receiving Anti D after delivery.

Signs: Generalized edema. Jaundice may be present.

On abdominal examination: polyhydramnios may be present. Size of the uterus may be > the expected. In case of IUFD, the FHS are absent.

Investigations

CBC, ABO group, Rh group, retic count, positive direct Coombs test is a sign of Rh incompatibility, SB & U/S.

Management

Sensitization means that Rh -ve mother's blood is exposed to Rh antigen. It usually occurs at previous pregnancy. Also it may occur through miscarriages, abortion, ectopic pregnancies &blood transfusions. The mother's blood will then produce antibody against the Rh antigen. In further pregnancy, when this blood containing the Rh antibody enters into fetal body, it causes hemolytic diseases. Rhesus disease does not usually affect the first born child.

Why is that? Because the mother's blood is not sensitized yet to produce the antibody. If a pregnant woman is Rh -ve & has not yet been sensitized, she usually will be given an in-

jection of *Rh immunoglobulin at 28 wks gestation.* This will prevent the sensitization for the rest of the pregnancy. It is recommended by most practitioners.

What does it actually do? This anti D immunoglobulin will seek to destroy any antigen present in the mothers blood. Thus the antigen will not be exposed & no antibody will be created. After birth the cord blood samples from baby is collected & send for blood grouping test. If it comes +ve, then the mother needs another round of *Anti -D Immunoglobulin (300 mcg) within 72 hrs after delivery* to prevent sensitization.

POLYHYDRAMNIOS

Presence of >2 liters of amniotic fluid. 20 % of cases associated é fetal malformations. The fetal prognosis worsens é more severe polyhydramnios & presence of congenital anomalies. The normal Amniotic fluid volume at 8 weeks is equal to 15 ml & ⇧ 30 ml every week. At 17 weeks it is about 250 ml & ⇧ 50 ml every week. At 28-38 weeks it is about 750-1000 ml & ⇩ after the 34th week of gestations. At 42 weeks of gestation the amniotic fluid is < 500 ml.

Etiology

•Idiopathic. •Fetal anomalies: immune/nonimmune hydrops fetalis, problems é swallowing, oesophageal or duodenal atresia, anencephaly, NTD, neuromuscular diseases, Down syndrome. •DM. •Multifetal gestation. •IUI. •Placental haemangiomas.

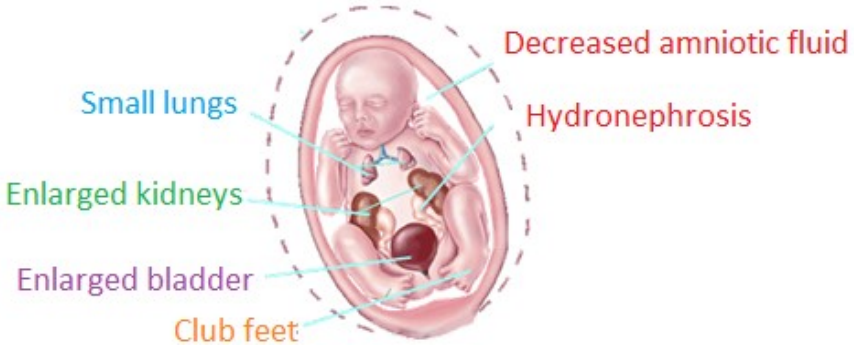
Diagnosis

*Fundal height > gestational age. *Difficulty palpating fetal parts & hearing FHS. *Tense uterine wall *U/S is conclusive.

Management

- Mild to Moderate case: rarely requires treatment.
- Hospitalization, bed rest.
- Amniocentesis for chromosomal studies.
- NSAID analgesia & blood sugar control.

OLIGOHYDRAMNIOS



Etiology

*Postdate. *IUGR. *Fetal Anomalies: obstruction of fetal UT/renal agenesis, pulmonary hypoplasia. *PRM. *Drugs as ACEIs & NSAIDs.

Diagnosis

*Fundal height < gestational age.

*⇩ Foetal movement.

*FHR abnormality.

*U/S is diagnostic.

*Presence of <500 ml at term.

Management: according to the cause.

Potter syndrome:

1/5000 live births, oligohydramnios of mother, renal agenesis, pulmonary hypoplasia, abnormal facial features. Baby usually die within 2 days.

AMNIOTIC FLUID EMBOLISM

Is sudden, unexpected, rare, life threatening complication of pregnancy. It has a complex pathogenesis & serious implications for both mother & infant. Associated é high rates of mortality & morbidity. Diagnosis is by exclusion, suspect AFE when confronted é any pregnant women who has sudden onset of respiratory distress, cardiac collapse, seizures, unexplained fetal distress & abnormal bleeding.

What's the meaning of AFE: it is a complex condition characterized by the abrupt onset of pulmonary embolism, shock & DIC, ŵ is due to the entering of amniotic fluid into the maternal circulation. Ricardo Meyer (1926) reported the presence of fetal cellular debris in the maternal circulation. Until 1950, only 17 cases had been reported. AFE was not listed as a distinct heading in causes of maternal mortality until 1957 when it was labeled as obstetric shock. Since then more cases have been documented, probably as a result of ⇧ awareness. Overall incidence of AFE is 1 in 8,000 pregnancies. It represents 16% of maternal deaths in UK & 10% in USA. 75% of survivors are expected to have long-term neurological deficits. If the fetus is alive at the time of the event, nearly 70% will survive the delivery but 50% of the survived neonates will incur neurological damage.

Time of event

•During labor. •During C/S. •After normal vaginal delivery & has been reported to occur as late as 48 hrs following delivery. •During the 2nd TM.

Etiology

- ⇧ Intrauterine pressure: uterine hypertonus, tetanic, or using oxytocin.
- Open uterine blood vessels: traumatic or é laceration.
- Membrane changing: IUFD, dystocia.
- Amniotic fluid itself: different constitutions as é allergic reaction.

Risk factors

•Advanced maternal age. •Multiparity. •Meconium. •Cervical laceration. •IUFD. •Very strong frequent or uterine tetanic contractions. •Placenta accrete. •Polyhydramnios. •Uterine rupture. •Maternal history of allergy. •Chorioamnionitis. •Macrosomia. •Male fetal sex. •Oxytocin (controversial).

Clinical presentation

The classic clinical presentation has been described by 5 signs that often occur in the following sequence:

(1) RD. (2) Cyanosis. (3) CVS collapse (cardiogenic shock). (4) Hge. (5) Coma.

A sudden drop in O_2 sat can be the initial sign of AFE during CS & > 1/2 of pts die within the 1st hour. Of the survivors 50% will develop DIC ŵ may manifest as persistent bleeding from incision or venipuncture sites. About 10% of pts will develop seizures.

Diagnosis

In 1941, Steiner & Luschbaugh described histopathologic findings in the pulmonary vasculature in 8 multiparous women dying of sudden shock during labor. The findings include; mucin, amorphous eosinophilic material & in some cases squamous cells. The presence of squamous cells in the pulmonary vasculature once considered pathognomonic for AFE is neither sensitive nor specific (only 73% of pts dying from AFE had this finding).

•Detection of sialyl Tn antigen in the serum of pt ē AFE is a direct way to detect the release of meconium - or amniotic fluid - derived mucin into the maternal circulation, is simple, noninvasive & sensitive test (NeuAc α 2-6GalNAc α 1-O-Ser/Thr) recognized by monoclonal antibody TKH-2.

•CXR may be normal or show effusions, enlarged heart, or pulmonary oedema.

•ECG may show a right strain pattern é ST-T changes & tachycardia.

- CBC: is the basis of laboratory diagnosis, platelet < 100 $\times 10^9$/l or gradually ⇩.
- PT: >15 second.
- Fibrinogen: <1.5 gm/L & ⇧ FDP.
- Plasma protamine para coagulation test: +ve.
- Obtrite RBC in blood smear.
- ABG.

Management

Goals of Management

*Restoration of CVS & pulmonary equilibrium.

*Maintain SBP > 90 mmHg & Arterial PO_2 > 60 mmHg..

*Maintain urine output >25 ml/ hour.

*Correct coagulation abnormalities.

*As intubation & CPR may be required, the following must be within reach: resuscitation tray é intubation equipment, DC shock & emergency medications.

Immediate Measures

▲Set up IV Infusion, Airway control ⇨ ETT & O_2 administration.

▲Maximal ventilation & oxygenation. ▲CBC, ABG, PT, PTT, fibrinogen, FDP.

▲Treat hypotension by ⇧ circulating volume & COP é crystalloids. After correction of hypotension, restrict fluid intake to maintenance levels since ARDS follows in up to 40-70% of cases.

▲Steroids may indicated (but no evidence as to their value).

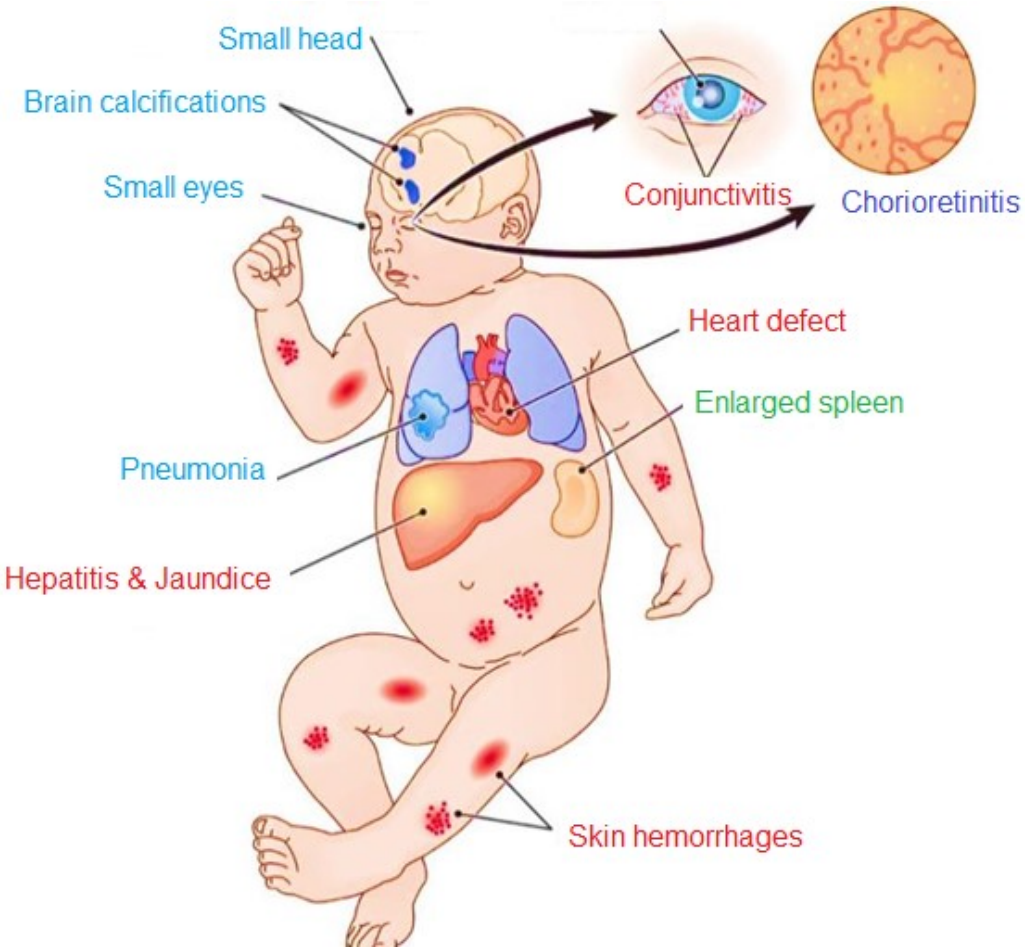
▲Dopamine infusion if women remain hypotensive (myocardial support). Other investigators have used vasopressors as Ephedrine or Levarterenol é success (as they ⇩ the systemic vascular resistance).

INDICATIONS FOR CAESAREAN SECTION

Recommended when a vaginal delivery might pose a risk to the mother or baby, not all of the listed conditions represent a mandatory indications & in many cases the obstetrician must use discretion to decide whether a CS is necessary, the indications include:-

- Failure to progress in labor 30% of cases.
- Arrest of descent, or dilatation.
- Repeated CS 30% of cases.
- Malpresntation 10% of cases. Transverse lie, breech, brow, or facementum lie.
- Non-reassuring FH pattern 10% of cases.
- Cord prolapse.
- VLBW.
- Large baby >4 kg.
- Multiple pregnancies.
- Conjoined twins
- Fetal congenital anomalies. Precious baby. Previous uterine rupture.
- Placenta Praevia, Placenta abruption, Placenta accrete.
- Uterine anomalies, contracted pelvis, obstructive tumor.
- Failed labor induction.
- Failed instrumental delivery.
- Chronic illness. HIV. ITP. Maternal genital herpes.
- Abdominal cerclage.
- Reconstruction vaginal surgery (e.g. fistula repair).
- Improper use of technology (electric fetal monitoring).
- Lack of obstetric skill (in performing, breech birth, or multiple births).

INTRA UTERINE INFECTION

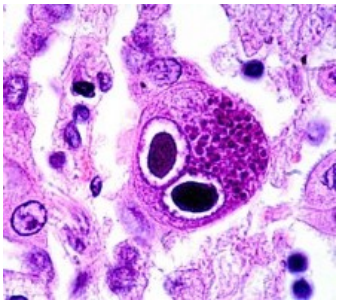


Is major cause of PT labor, represent an approximately 25% of all PT births. The earlier the gestational age at delivery, the higher the frequency of IUI. It is considered as one of the major maternal insults during pregnancy. The incidence of IUI during pregnancy is estimated to be about 14% when laboratory methods of detection are used. The commonest IUI are; toxoplasmosis, rubella, syphilis, CMV, herpes simplex, hepatitis B, HIV (TORSCH) & others include; Coxsackie, Varicilla zoster, Parvo viruses.

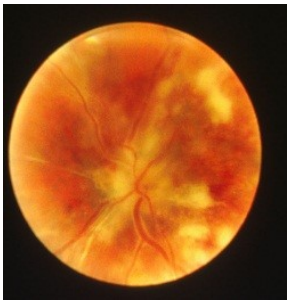
Preventive measures

Are necessary since lesions caused by some IUI are permanent & damaging. TORCH`S screening for every pregnant women is a medical acronym for a set of perinatal infection transmitted from the mother transplacentaly or during labor.

CONGENITAL CYTOMEGALO VIRUS INFECTION

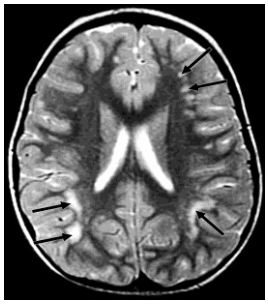


Cytomegalo cells



Chorioretinitis

Deafness



Cerebral calcification

In 1920, Good Pasture correctly postulated the viral etiology of the histopathological changes, probably in tissues from a congenitally infected infant & he used the term cytomegalia to refer to the enlarged, swollen nature of the infected cells, the virus was first isolated in 1956, it is 1 of 8 human herpes viruses. The incidence of congenital CMV infection varies widely throughout the world ranges from 0.2-2.2% of live births. It is considered one of the most serious infections during pregnancy. There is significant risk increase of adverse fetal effects if infection occurs during the first half of pregnancy. It was estimated that 50-80% of adults in USA have had a CMV infection by the age 40 yrs, it is one of the STDs & once CMV is in a person`s body, it stays there for life. Infection acquired to the baby either transplacentaly, or during labor through contact é infected cervical secretions, or during breast feeding & through contaminated blood transfusion.

Clinical picture

The Mother: usually have no symptoms, or mild manifestations of flu like as fever, muscle ache, headache, but infection can be very serious in people who have received organ transplants or immunocompromised people.

The Baby: is asymptomatic; in 90% of cases, infant appear healthy at birth, but 10-15% of those babies may develop late squeals, especially hearing defects after a period of months or even years.

Symptomatic baby; in 5% of cases, severe fetal damage & in rare cases death due to abortion, or é manifestations of; SGA, hepatosplenomegaly, petechiae (purple skin splotches or rash or both), thrombocytopenia, prolonged NN jaundice, pneumonitis, microcephaly, occasionally cerebral calcification, later complication include; CP, epilepsy, MR, visual impairment, chorioretinitis, optic atrophy, delayed psychomotor development, expressive language delay, learning disabilities & deafness.

Diagnosis

*Culture from body fluids, or tissue biopsy specimen (blood, saliva, throat swab, CSF, urine, stool, vaginal secretions, breast milk, semen of father), culture monitored for development of CMV-associated cytopathic effect.

*PCR.

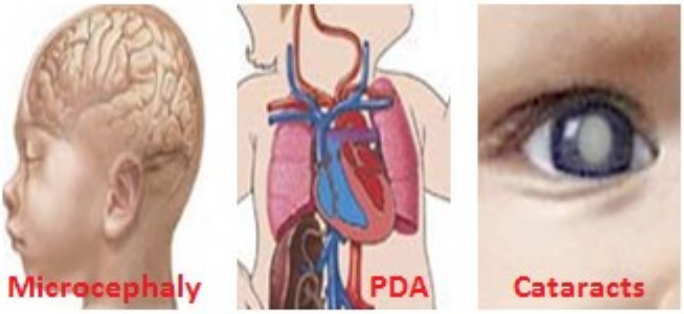
*CMV IgM.

*ELISA (Enzyme Linked Immune Sorbent Assay) test is diagnostic.

Treatment

No specific treatment, there is some evidence that Ganciclovir (500 mg amp) -antiviral- may prevent hearing loss & developmental outcome in infants born é symptomatic CMV infection, the dose from age 6 months -16 years is equal to 5 mg/kg/12 hours IV for 7-14 days, or PO (250 mg cap) 15 mg/Kg twice daily & one of its main side effects is bone marrow suppression.

CONGENITAL RUBELLA SYNDROME



The name Rubella is derived from a Latin term meaning"Little Red", was 1st described in 1941 by Australian Ophthalmologist Norman Gregg, who had noticed an unusual number of infants é cataracts following rubella epidemic in 1940. In 1964–1965 worldwide outbreak occur result in an estimated 12.5 million cases & 20.000 cases of congenital rubella syndrome (CRS) & 2100 NNDs of CRS in USA only. The virus was isolated in 1961 & vaccine developed in 1969, this is the shortest time period from virus identification to vaccine ever. The introduction of the MMR vaccine & screening program for pregnant mothers improve dramatically the incidence of CRS in countries applying this system. Worldwide it was estimated that >100.000 infants born é CRS annually, as it is still common in many developing countries. The infection caused by rubella virus ẃ is a member of the rubivirus group, it`s IP is 2-3 weeks & there is 90% chance of passing infection transplacentaly if mother get infected during the 1st TM, but fetal damage is rare after 12 weeks gestation.

Clinical Picture

Sensorineural deafness (in 80% of cases). MR (55% of cases), Cataract, Retinopathy, Microophthalmia (50% of cases). PDA- (50% of cases). Meningoencephalitis (25% of cases). DM type1 (20% of cases). LBW. SGA. Hepatosplenomegaly. Generalized lymphadenopathy. NN jaundice. Thrombocytopenia. Abnormal skull. Microcephaly. Microganthia. IC calcification. Schizophrenia & autism.

Diagnosis

Mother: detection of IgM Rubella specific in mother saliva sample, or maternal blood is both sensitive & specific, indicates primary infection & ⇧ in IgG titer over 2 weeks usually occurs

Baby: isolation of rubella virus from blood, nose, throat, or urine. Detection of IgM rubella specific & PCR +ve for rubella virus.

Management

Pregnancy termination if rubella specific IgM is positive in the 1st 16 weeks. Screening programme used in many western countries for all adolescent girls in their secondary school to detect rubella antibodies & if negative, the girl must be given MMR vaccine.

CONGENITAL HERPES SIMPLEX

NN infection é HSV was 1st reported in 1935 by Dr. Hiss M. who reported a case of hepatoadrenal necrosis é intranuclear inclusion bodies. It is one of the STDs, infection also can transmitted through skin to skin contact. The NN infection, mostly caused by type 1 HSV, while 25% caused by type 2 HSV, the risk is more higher if the 1st attack (painful & itchy) occur after the 28th week of pregnancy.

85% of transmission occur during birth when a baby come in contact é infected genital secretion in the birth canal. 5% occur through transplacentaly. 10% acquired the infection postnataly. The incidence of congenital Herpes 1/3000-20000 live births in USA.

Clinical picture

Mortality rate in NN is 100%, the diagnosis can be difficult, but should be suspected in NN when one of parent have positive history of herpes infection, or pregnant mother é itching, discharge, vesicles in vulva, lower abdominal pain, inguinal lymphadenopathy, or in case of baby é irritability, lethargy, fever, poor feeding at the 1st week of life. The baby infection is either:-

*Localized Skin, Eye, Mouth: the skin lesions appear as small, fluid filled vesicles, these vesicles rupture, crust over & finally heal, often leaving a mild scar.

*Encephalitis: presented é seizures, tremors, lethargy, irritability, poor feeding & bulging fontanels.

*Disseminated form: is another presentation involving the CNS, lung, liver, adrenals, SEM. The transplacental transmission may presented é micro or hydrocephalus, chorior-etinitis & IC calcification.

Diagnosis

*Isolation of the virus by culture from; blood, CSF, oropharynx, urine, stool, skin vesicles, eye or nose secretions.

*Positive PCR for HSV.

*ELISA test for HIV.

Management: it has been recommended that CS should be performed if acute lesions are present at the onset of labor.

Mother: é proven HSV infection in the late 3rd TM, or mother é active recurrent genital herpes, oral Acyclovir 400 mg tid from 36th week of gestation until delivery & breast fee-ding is allowed, as the HSV not transmitted through breast milk.

Baby: é proven HSV infection, IV Acyclovir 60 mg/kg/day ÷ 3 equal doses for 2-3 weeks.

CONGENITAL HEPATITIS B

The WHO has estimated >350 million people over the world are chronically infected é HBV. In adults, infection transmitted through; drug abuse, high risk sexual activities, multiple sexual partner, sexual partner é viral hepatitis, infected blood or blood product transfusion, infected needle & tattoos. The IP of the disease may vary from 6 wks up to 6 months. If pregnant woman is a HBV carrier é HBeAg +ve, her NN has a 90% likelihood to be infected & become a carrier. 25% of those babies will die later during adulthood from chronic liver disease or liver cancer. The availability & extensive use of HBV vaccine has dramatically ⇩ the number of incident infections in many countries. The HBV, is large virus & does not cross the placenta, hence it can`t infect the fetus unless there have been breaks in the maternal fetal barrier, most of cong. HBV infection (90-95%) occur during delivery from abrasions in the infant`s skin or mucosa or from small maternofetal bleeds across the placenta during labor, while transplacental transmission occur in 5%. The mode of delivery does not influence the vertical transmission. Hepatitis B is the only STD to have protective vaccine.

Clinical picture

Almost all infections in the neonates are asymptomatic but >90% become chronic carriers & at high risk for chronic liver disease, cirrhosis & hepatocellular carcinoma during adulthood compared é only 5-10% of individuals acquiring HBV infection as adolescents or adults.

Risk factors for HBV transmission

Maternal HBeAg +ve, high maternal HBV DNA, threatened preterm labor & threatened abortion.

Investigations

Mother: acute HB or persistent carrier state, will show; +ve HBsAg, +ve HBe Ag & +ve anti HBc (IgM or IgG).

HBs Ag: is the surface antigen of hepatitis B virus, it indicates current HB infection, commonly referred to as the Australian antigen, this is because it was first isolated by the American research physician & Nobel prize winner, Baruch Blumberg in the serum of Australian person, it was discovered to be part of the virus that cause hepa-titis by virologist Alfred Prince in 1968.

HBeAg: appear during 3-6 wk of infection & it indicate that pt is infectious & its persis-tence >10 wk indicates chronic infection.

HBc Ag: not detectable in blood but in liver cells (biopsy).

anti HBc IgM: denotes early acute infection.

anti HBc IgG indicate chronic infection.

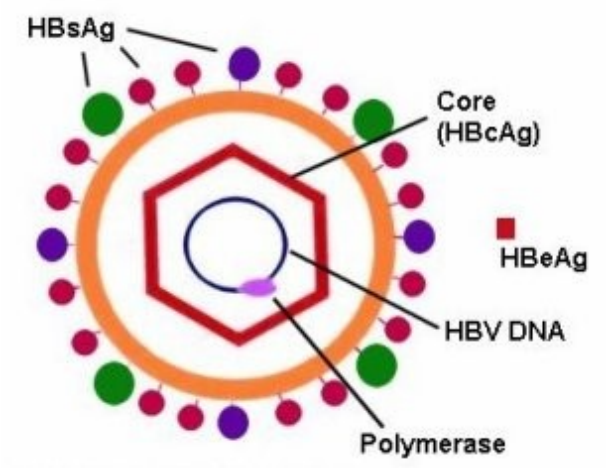
anti HBe: prognostic for resolution of infection.

anti HBs: indicate clinical recovery & subsequent immunity.

Disease stage	Serological markers
Acute disease	HBsAg , antiHBc IgM
Chronic disease	HBsAg
Infectivity	HBsAg , HBeAg , Viral DNA
Recovery	antiHBe, antiHBs
Carrier state	HBsAg* , antiHBc (total)
Immunity	antiHBs , antiHBc (total)
Past immunization	AntiHBs

*Persisting for more than 6 months

Infant: +ve HBsAg is indicative of acute infection & it`s persistence for >6 months is indicative of chronic infection.

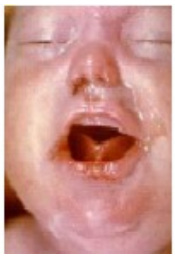


Management

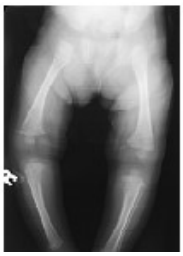
Mother: screening of all pregnancies for HBsAg, if pregnant women is +ve she should be given hepatitis B immunoglobulin. No antiviral agent has been approved for use during pregnancy, "the risk-benefit equation of using antiviral depends upon age of mother, the trimester of pregnancy & her stage of liver disease".

Baby: should be given HB hyper immunoglobulin at birth, followed by HBV vaccination, the first dose to be given within 12 hours of birth, second dose at 1 month age & third dose at 6 months age, this regime is 95% effective in prevention of HBV perinatal transmission. Heptavax, is the 1st generation of hepatitis B vaccine in 1980`s was made from HBsAg extracted from the plasma of HB pts. Current vaccine is made from recombinant HBsAg in grown yeast. Breast feeding by HBsAg +ve mother is not known to ⇧ the risk of transmission & therefore not contraindicated.

CONGENITAL SYPHILIS

Snufflcs 

Desquamation 

Periostitis 

Syphilis is a STD, has been recognized since antiquity, in infancy it was described as early as 1497 & the causative microbe Treponima Palladium, was discovered in 1905, congenital syphilis still serious, under diagnosed & is a threat for children in poor countries. WHO estimate that there were 2 million syphilis infections among pregnant women annually. In 2007, WHO, launched an initiative to eliminate syphilis, that set targets of at least 90% of pregnant women being tested for syphilis & at least 90% of sero positive pregnant women to receive adequate Rx by 2015. Congenital syphilis is caused by passage of bacteria from mother to the child during fetal development especially before 16th week of gestation or at birth. Untreated syphilis results in a high risk of a poor outcome pregnancy e.g. miscarriage, preterm labor, IUGR, stillbirth. Some infants é congenital syphilis have symptoms at birth, but most develop symptoms later.

Clinical picture

Early signs of cong syphilis: those appearing in the first 2 years of life are in the form of;

*Bone lesions: osteochondritis, periostitis, pseudo paralysis (secondary to pain or fracture) affecting long bones, cranium, ribs, spine…….. (78% of cases)

*Hepatosplenomegaly, jaundice, lymphadenopathy…….. (71%)

*Skin lesions: extremely variable; macular, vesicobullous, bullous (pemphigus syphiliticus), desquamation (washer woman skin palms, soles), rash in mouth or genital area, paronychia, mucous patch, condylemata lata……(68%)

*Fever …….. (42%).

*Failure to thrive……..(33%)

*CNS involvement: leptomeningitis, seizures, hydrocephaly……..(23%)

*Pneumonitis……..(17%)

*Snuffles……(14%) sero sanguineous discharge from nose, saddle shaped nose (collapse of the bony part of nose).

*Chorioretinitis, uveitis & glaucoma.

The late signs of cong. syphilis: appearing later over the first 2 decades of life include;

*Hutchinson`s teeth (centrally notched, widely spaced shaped upper central incisors).

*The triad of Hutchinson teeth, keratitis & deafness occur in 63% of cases).

*Rhagades: linear scar at the angles of mouth & nose é secondary infection.

*Skin scaring around the mouth, genitals & anus.

Investigations: •Detection of Treponima Palladium (in blood or secretions) •FTA-ABS Flourescent Treponema Antibodies Absrption Test. •PCR •VDRL •CSF •Bone X Ray.

Management

Aqueous Crystalline Penicillin G 100.000-150.000 u/kg/day IV divided into 2 doses for 10 days, or Procaine Penicillin G 50.000 u/kg/day IM as a single daily dose for 10 days. In case of allergy, children should be desensitized.

Desensitization technique

① 0.1 ml 1/20 conc + 0.05 ml adrenaline SC watching for local or systemic reactions.

② 0.1 ml 1/10 conc + 0.05 ml adrenaline SC after 30 min.

③ 0.01 ml full conc + 0.05 ml adrenaline SC after 30 min.

④ 0.1 ml full conc + 0.05 ml adrenaline SC after 30 min.

⑤ 0.5 ml full conc + 0.05 ml adrenaline S.C after 30 min.

⑥ Full dose IM after 30 min. é the presence of adrenaline & forticort ready to use.

CONGENITAL HUMAN IMMUNE DEFICIENCY VIRUS

STD, worldwide there were 2.5 million new cases in 2011 & about 34.2 million people are living é HIV around the world, once a person is infected, the virus remains in the body for life, there is no cure for AIDS but there are drugs that help control the virus, enabling people to live a full & healthy lives. HIV may be transmitted from mother to baby at any time during pregnancy, or labor in 65% of cases, or as a result of breast feeding. HIV infection progressing to AIDS over 8-10 years.

Factors ⇧ the risk of transmission

•Acute stage of mother illness. •High maternal viral load. •Low maternal CD4 count. •PRM or premature delivery. •Abruptio placenta. •Vaginal delivery. •Breast feeding.

Clinical picture

Recurrent bacterial, fungal, or viral infection, wasting, delayed milestones.

Investigations

During pregnancy, the fetus passively acquires maternal HIV antibodies across the placenta, this does not mean the fetus is infected, it can take up to 12-18 months for a baby to clear these maternal antibodies. PCR test is very accurate & the best test to diagnose HIV infection in babies, by detection of HIV-DNA-PCR (qualitative) or HIV-RNA-PCR (quantitative) ẃ is more valuable. A positive PCR test must be confirmed by repeat test to confirm infection. CD4 cells sometimes called T cells, are a type of lymphocyte, they are an important part of the immune system. When HIV infects humans, the cells it infects most often are CD4 & when someone is infected for a long time, the number of CD4 they have drop dramatically (normal value of CD4 cells is 500-1600/ml^3 blood = 20-40%) & any one é CD4 < 200 (< 14%) is considered to have serious immune damage. The T lymphocyte cells (CD4) stimulate the production of interleukin 2 ŵ in turn stimulate the production of

T-killer & T suppressor cells, both act to regulate the defensive mechanism against infection, also T lymphocyte stimulate the production of B lymphocytes & plasma cells ŵ in turn stimulate the production of complement (neutralization of bacteria), opsonin (opsonization of bacteria) & immunoglobulin, all responsible for cellular & humoral immunity in the body.

Body defensive mechanisms

Physical immunity: intact skin, mucous membrane, ciliary function & bacterial flora.

Cellular immunity: macrophage ŵ engulf bacteria & produce lactoferin (chelate iron from bacteria), lysozyme (kill bacteria), B & T lymphocytes.

Humoral immunity: including; immunoglobulins, complements & opsonin.

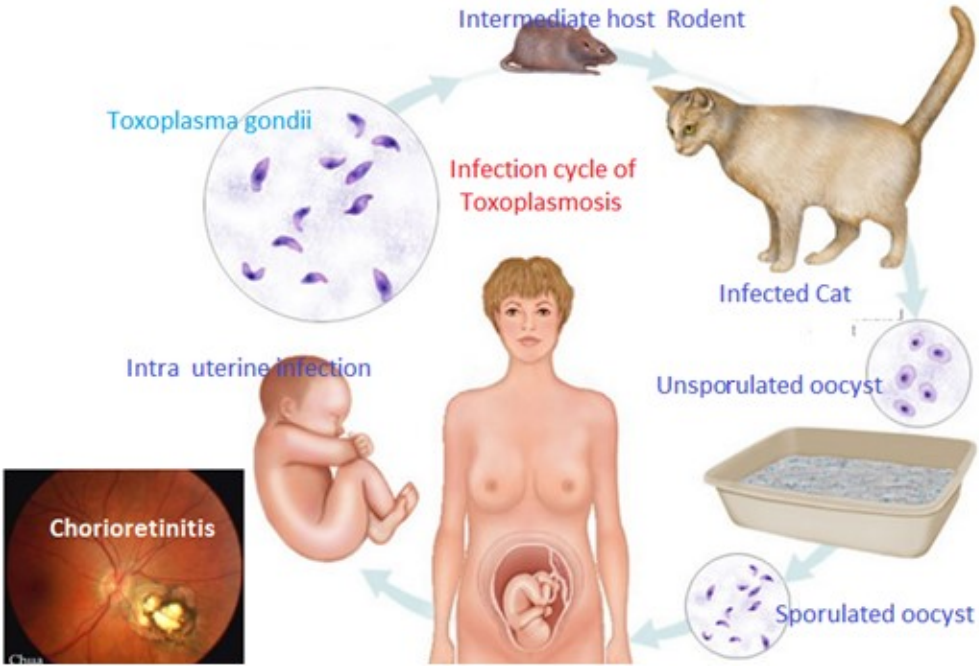
Management

Pregnant mother: HIV treatment is life long, if pregnant mother proved to have HIV infection or her CD4 count < 500, antiretroviral drug therapy, Zidovudine "ZDV" 250 mg PO twice daily or Azidothymdine "AZT" after the 14th week of gestation & to continue throughout pregnancy in addition to intrapartum intravenous ZDV (amp 20 ml = 200 mg), starting 4 hours prior to CS, at a loading dose of 2 mg/kg over one hour then 1 mg/kg hourly until the umbilical cord is clamped. HIV may become resistant to ZDV or AZT over time & for this reason it may be used in conjunction é other anti HIV drugs, called Highly Active Anti-Retroviral Therapy (HAART) include; Efavirenz +Tenofaver + Emtricitabine, given according to certain protocol & follow up schedule for CD4 & PCR.

Baby: if pregnant mother proved to have HIV infection, baby must be given Zidovudine syrup (5 ml contain 50 mg), starting 8 hours after birth in a dose of 2 mg/kg/6 hours for 6 weeks, if baby is PT or unable to tolerate feeds, IV infusion used & the baby is closely monitored by PCR monthly for the first 4 months, then PCR & CD4 every 3-4 months until

age of 18 months to completely R/O HIV infection. Prophylactic for pneumocystis carinii pneumonia, Cotrimoxazole syrup 0.5 ml/kg/day should be initiated when infants are 6 weeks old & continued for at least 4 months. No BCG or live vaccines should be given.

CONGENITAL TOXOPLASMOSIS



Caused by infection é the protozoan parasite, 1st identified & isolated from African rodent in 1908 by Drs Niclle & Manceaux. In 1909 the parasite was named Toxoplasma Gondii. In 1923 Janku reported parasite cysts in the retina of an infant who had hydrocephalus, seizures & bilateral micro ophthalmia, while the first adult infection was recorded in 1940. Human infection may be acquired by ingestion of oocyte excreted by cats & contaminating soil or water, or by eating tissue cysts that remain viable in undercooked meat of infected animals. There are considerable geographic differences in prevalence rates. The sero +ve (IgG) women in the childbearing period is about 50-80% in Latin America & 30-50% in Middle East. It's rare that a women who got toxoplasmosis before getting pregnant will pass the infection on to her baby, but if she catch it during pregnancy & remain untreated there`s a chance that she could pass infection on to her developing fetus &

Babies who become infected during their mother`s first trimester tend to have the most severe symptoms. Infection in adult are usually either asymptomatic or associated é self-limited symptoms as fever, malaise & lymphadenopathy.

Clinical picture

90% of babies born é cong toxoplasmosis have no symptoms early in infancy, but large % of them will show signs of infection months or years later. include;

- Prematurity.
- Persistent jaundice, hepatosplenomegaly, anemia.
- MR.
- Hydrocephaly or microcephaly.
- Intracranial calcification, chorioretinitis, blindness, epilepsy
- Skin rash (tiny red spots/petechiae).

Investigations

Pregnant women: +ve toxoplasma specific IgM & IgG indicate recent infection but +ve toxoplasma specific IgG only indicate old infection ŵ leads to a lifelong antibody.

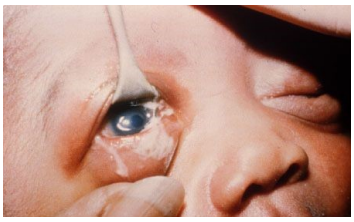
Infant: toxoplasma antibodies (IgG) are passed from the mother to the baby through the placenta & could be of maternal origin, while +ve toxoplasma specific IgM (can`t pass the placenta) indicates infected baby.

Management

Pregnant women: é confirmed infection, Pyrimethamine: loading dose 100mg/day divided into 2 doses for 2 days & the maintenance dose is 50mg/ daily.

Infant: Pyrimethamine: loading dose 2 mg/kg/day for 2 days, then 1 mg/kg/day for 2-6 month, maintenance every Monday, Wednesday & Friday for 1 year + Sulphadiazine 100 mg/kg/day ÷ 2 doses for one year + Folic acid 10 mg/ 3 times weekly for one year.

CONJUNCTIVITIS



Conjunctivitis in general include the following causes:-

*Chemical: occurs in the 1st day of life, result from conjunctival irritation by blood or meconium during labor.

*Gonococcal: manifest in the 2nd-3rd day of life, the conjunctival discharge is purulent (ophthalmia neonatorum). Diagnosed by culture of the baby conjunctival discharge, swab from mother urethra, cervix & vagina. Condition treated by; Penicillin 6 hourly, Mephenicol eye drops ½ hourly for 6 hours, then 4 hourly 5 days, Mephenicol ointment twice daily (in addition to treatment of the mother).

*Chlamydia infection: occur in the fifth day of life, associated é severe congestion (Erythromycin is effective treatment).

*Viral: characterized by severe congestion, subconjunctival He, it takes about 1-2 weeks to subside, symptomatic treatment (Visine, Tresolin, Dexapolyspectran eye drops).

Clinical Finding	Bacterial	Viral	Allergic
Bilateral eyes	50% to 74%	35%	Mostly
Discharge	Mucopurulent in younger children	Mild, watery, or "sleepers" only	Rare
Redness	Common in older children, uncommon in infants and toddlers	Usually	Usually
Acute otitis media	32% to 39%	10%	No
Pruritic	No (but many rub eyes)	No	Major

PREMATURITY

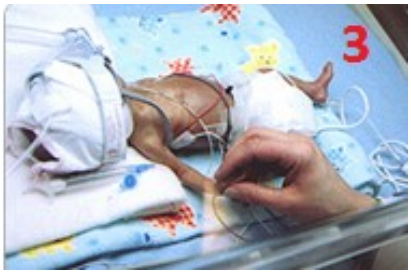
november 17: world prematurity day

Babies born before the 37th wk of gestation are born prematurely & are sometimes given the nickname "preemies". They are often of LBW i.e. BW < 2.5 Kg at birth. Late PT babies who are born between 35-37 wks gestation may not look premature & may not be admitted to ICU, but they are still at risk for severe problems than FT.

The normal development during pregnancy is as follow;

- 4th month: fetus is about 1/8 Kg (150 gm).
- 5th month: 1/4 Kg (250 gm).
- 6th month: 1/2 Kg (500 gm).
- 7th month: 1 Kg (1000 gm).
- 8th month: 2 Kg (2000 gm).
- 9th month: 3 Kg (3000 gm).

Prematurity classified into:-



❄1- <u>Mild:</u> baby who is born at 33-36 wks gestation &/or have BW 1500-2500 gm.

❄2-<u>Moderate:</u> baby who is born at 28-32 wks gestation &/or have BW 1000-1500 gm.

❄3-<u>Extreme:</u> baby who is born before 28 wks gestation &/or have BW < 1000 gm.

Incidence

Prematurity is the greatest risk factor for infant mortality & is the world`s single biggest cause of neonatal death & the second leading cause of child deaths after pneumonia, 65% of infants who died before the age of one year were born prematurely & the NND account for 40% of all deaths among children under 5 years of age. 50% of all premature birth have no known cause, black women are nearly twice as likely to have their babies prematurely compared to white women. Every year, about 15 million babies are born prematurely & > 1 in 10 of all babies born are premature.

Maternal Risk Factors for Prematurity

Age of the mother: < 16 yrs or > 35 yrs.

Chronic illness of the mother: as DM, hypertension, kidney disease, malnutrition, SLE, may become out of control during pregnancy & in some situations the only way to stop the worsening of the condition is to deliver the baby & sometimes the labor will begin too early on its own.

Stress: chronic, high level of psychological stress.

Short term between pregnancies: the risk of preterm birth is 2 times higher than normal if pregnancies are **<** 6 months apart.

Multiple pregnancies: cause the uterus to over distend, ẃ can cause labor to start early & the risk ⇧ é the number of babies.

Placenta abruption: placenta starts to separates from the uterus before the baby is born, can cause extreme blood loss in mom & baby & can be fatal & emergency delivery of the baby is necessary.

PRM: amniotic fluid sac breaks before baby reach full term, some studies link this é infection in the uterus, or from placenta praevia.

History of premature delivery.

Congenital anomalies of the uterus, or uterine fibroid.

Cervical incompetence: weakened cervix that begins to open early.

Preeclampsia: ⇧BP & presence of proteinuria develop after 20th wk of gestation.

IUI: Toxoplasmosis, Rubella, Syphilis, CMV, Herpes & HIV (TORSCH).

Use of Tobacco, Nicotine: causes blood vessels in the uterus to contract ŵ can prevent nutrients & O_2 from getting to the baby or contribute to early labor, also use of Cocaine or Amphetamine.

Lack of prenatal care & low socioeconomic state.

Preterm characteristics

- Abnormal breathing patterns, shallow, irregular, apneic attacks é period < 10 sec.
- Feeding problem due to trouble sucking or coordinating swallowing & breathing.
- Less activity than FT baby, lower muscle tone & less body fat.
- Soft flexible ears cartilage.
- Thin, shiny skin, transparent (often you can see veins under skin). Lanugo hair.
- Enlarged clitoris (female), small scrotum that has no ridges & undescended testicles (male), as usually the testes descend by the 38th wk of gestation.

Preterm complications:

- RDS: the alveoli of lungs began to form at 26-28 wks of gestation, while the surfactant ŵ is important in ⇩ surface tension of alveoli & prevention of alveolar wall collapse during expiration started to be produce at 34 wks gestation.
- Retrolental fibroplasia & blindness: if O_2 level is too high or ē prolonged period.
- Hypothermia: less able to shiver & to maintain his body temp.
- Hypoglycemia or hypocalcaemia: convulsions that may result in brain damage.

- NN jaundice: kernicterus in PT occurs at a lower level of bilirubin than in FT. Kernicterus was found at autopsy of PT babies at levels of 6-7 mg/dl of SB.
- Intra ventricular Hge: é its severe long term effects as CP or deafness ŵ may affect as many as 10-15% of significantly PT babies.
- Neonatal infection.
- NEC.

Management

Pregnant mother: whom at risk for premature labor between 26-34 wks gestation, to be given either betamethasone or dexamethasone to promote fetal lung maturity, betamethasone 12 mg/24 hrs IM for 2 doses or dexamethasone 6 mg/12 hrs IM for 4 doses, tocolytics may delay labor a few days & antibiotics if an infection is suspected or present.

Preterm baby: Kangaroo care is placing a preterm in an upright position on a mother`s bare chest allowing tummy to tummy contact & placing the PT in between the mother`s breasts. The baby`s head is turned so that the ear is above the parent`s heart. That is in mild case in ŵ the preterm does not need incubator care.

Incubator care: baby is admitted to high risk nursery & placed in an incubator, for controlling temp & keeping the baby warm, the babies grow fastest if they are kept warm.

Respiratory assistance; O_2 hood: this is a clean plastic box that is placed over the baby`s head & is attached to a tube that pumps O_2 to the baby.

CPAP: for babies who can breathe on own but need help getting air to their lungs.

Ventilator: for babies unable to breath spontaneously, connected to the ETT é monitoring ventilator for FiO_2 %, Tv, PIP, PEEP &RR.

Monitoring: all babies are attached to a heart & breathing monitors while they are in the NSCU, these monitors sound & alarm if there is a big change in the baby`s HR, RR, the

baby is also attached to a pulse oximeter ŵ records the O_2 level in the baby skin, there are also temp alarms for the warming beds & incubator.

Nutrition: providing adequate nutrition to PT infants is a challenging because of inability to suck & swallow & the immaturity of bowel function & the high risk of NEC. Disorders of fluid & electrolyte are common in NN & a proper understanding of the physiological changes in body water & solute after birth is essential to ensure a smooth transition from aquatic in utero environment, the newborn kidney has a limited capacity to excrete excess H_2O & Na^+. An overload of fluid or Na^+ in the first week of life may result in condition like NEC & PDA, also after birth there is a sudden efflux of fluid from the ICF to ECF compartment, this ⇧ in ECF compartment floods the NN kidneys eventually resulting in a salt & H_2O diuresis, hence the physiological weight loss in the 1st week of life, this is more evident in preterm than term baby. Term infants are expected to lose 10% as compared to 15% in preterm. The preterm baby < 34 wks gestation is unable to coordinate sucking & swallowing in addition that he/she has limited energy stores ŵ become rapidly depleted é starvation. Infants receiving only IV glucose may lose protein stores at a rate of up to 1.0 gm/Kg/day.

Method of feeding depends on the PT individual needs:

- Orogastric or nasogastric tube feeding for PT baby who is ready to digest breast milk or formula but unable yet to suck, swallow & breath in a coordinated manner.
- IV line: in the scalp, arm, or leg, for short term nutritional support of PT baby é resp problem or complications.
- Central line: for long term nutritional support, using larger veins in the neck, arm, leg for the delivery of nutrients & medicines that irritate smaller veins.
- Umbilical Catheter: for critical cases é breathing problems or complications (the safest

& most effective way for babies requiring long term nutritional support).

Intra Venous Fluid: for mild PT babies anticipated to tolerate enteral feeding & é no breathing problem we follow the following schedule;

Day 1: use G 10%, 60- 80 ml/ Kg /day (Na^+ free fluid).

Day 2-5: G 5% + Saline (ratio 4/1) é daily ⇧ 20 ml/Kg up to a maximum of 150-180 ml/ Kg /day to be reached by the end of the 1st wk +Ca gluconate 10% 1 ml/Kg/day (amp 10 ml) + Kcl 20% (amp 5 ml) 1 ml/Kg/day to be added after ensuring adequate urine output equal to 1 ml/Kg/hour & Kcl level is < 5.5 meq/l + the daily requirement of vitamin & trace elements using ½-1 amp/day of pediatric multivitamin formula, the 5 ml amp contain (vit A 2300 IU, vit D 400 IU, vit E 7 IU, vit K 200 mcg, vit C 80 mg, vit B_1 1.2 mg, vit B_2 1.4 mg, vit B_6 1 mg, vit B_{12} 1 mcg, biotin 20 mcg, folic acid 14 mcg, niacin 17 mg, pantothenic acid 5mg) + additional allowances of IVF equal to 20 ml/Kg/day for PT under radiant heat & phototherapy to compensate for fluid loss.

Total parenteral nutrition: in situations where adequate nutritional support can`t be achieved & fat & glycogen stores have been exhausted, infants begin to catabolize protein stores for energy, for example PT infant 1 Kg weight, the fat contributes only to 1% of BW, as compared to a term infant 3.5 Kg where about 16% weight is fat, in addition, such PT infants often do not tolerate enteral feeding due to their small stomach capacity & immature GIT & gastric empting & intestinal transit times are significantly delayed as compared to the term infant, the gut motility does not begin until 32-34 weeks gestation. The primary goal of TPN is to provide energy & nutrients in a sufficient quantities to allow normal growth. Before using TPN we may try syringe pump of milk & changing milk every 4 hours. TPN require central vein access ŵ allow for administration of a solution é higher osmolality, either through SVC or UVC ŵ generally placed after birth & removed

within 2 weeks due to ⇧ risk of infection. Most PT infants <1500 gm BW will need TPN, also TPN is needed for larger PT infants when it is anticipated that full enteral feedings will be delayed for > 3-5 days to meet energy & nutritional requirements. Using TPN necessitate the presence of fluid & nitrogen balance charts, ŵ include the fluid intake & output. The fluid output include; urine (70%), insensible water loss (20%) & stool (10%). The nitrogen chart record of daily intake of nitrogen (as recorded over the AA solution bottle used, how many grams of nitrogen in the amount of amino acids we are giving) & the nitrogen output calculated as follow (24 hours urine for nitrogen ÷ 2.14) + 2.

How we prepare the solution

- Calculate total requirements of fluids & calories according to BW, the PT will need 100-150 ml & 150-180 calories/day.
- In the first 5 days give Glucose 10 or 20% + Amino Acids sol. + Vit & Trace elements. The Amino Acids sol. calculated as; BW (Kg) X 0.6 to get amount of nitrogen (gms)/day, read the content of nitrogen recorded on the bottle of Amino Acids solution & accordingly determine the amount of Amino Acids sol. to be given. Taking into consideration that each gm of protein gives 4 calories. The PT needs 2.5 gm/Kg/day of protein to support growth at a rate comparable é the intrauterine rate.
- For each gm nitrogen give 160 calories glucose 10 or 20%. Each gm of COH gives 3.75 calories & the daily requirement is 10 gm/kg/day.
- Add the daily requirement of vit & trace elements (pediatric multivitamins formula vial).
- From day 6 start to add intralipid 20%, start é 0.5 gm/Kg/day & ⇧ gradually 0.5 gm every 2 days to a maximum of 5 gm/Kg/day & monitor triglycerides to be kept < 100 mg /dl (each gm lipid gives 10 calories & that the daily requirement of lipids is 5 gm/Kg).
- Do daily estimation of: Ca, Mg, Ph & triglycerides until all indices are stable.

- Do weekly estimation of: LFT, SB, [total & direct], albumin & triglycerides.

Caloric requirements

Infants have high metabolic rate & energy requirements per unit of BW than children & adult, baby need about 60 Kcal/Kg/day just to prevent catabolism & sum of 110-120 Kcal/Kg/day to prevent catabolism & to allow growth. A baby should gain 20-30 gm/day over the first month of life, while the EPT will gain about 13-16 gm/day. In TPN for PT we start é 80-100 Kcal/Kg/day because energy is not needed to cover fecal losses, nor is energy being utilized for thermogenic effect of food.

Fluids requirements

Start by total fluid of 80 ml/Kg/day, ⇧ gradually 10-20 ml/Kg/day to a maximum of 150 ml/Kg/day to be reached by end of 1st week. In EPT start by 105 ml/Kg/day &⇧ gradually 10-20 ml/Kg/day to a maximum of 150 ml/Kg/day to be reached by the end of 1st week.

Electrolytes requirements

- Na^+ 2-5 meq/Kg/day.
- K^+ 2-4 meq/Kg/day.
- Ca^+ 2-4 meq/Kg/ day (60 mg elemental Ca/Kg/day).
- Ph^+ 1-2 meq/Kg/day (48 mg/Kg/day).

Vitamins & Trace elements requirements: ½-1 amp/day, of ped multivitamin formula.

Heparin: 0.5 unit for each ml of TPN recommended as it ⇩ the formation of fibrin sheath around the catheter, may reduce phlebitis & ⇧ the duration of catheter patency, also stimulate release of lipoprotein lipase, ŵ may improve lipid clearance.

Spending time under bilirubin lights: phototherapy help baby system to break down excess bilirubin, because liver can`t process it all, baby wear protector eye mask, his fluid intake ⇧ by 20% to compensate for fluid loss.

Receiving blood transfusion: because the preemies may have an underdeveloped ability to make his/her own RBCs, blood transfusion may be needed to ⇧ blood volume, especially if the baby had several blood samples drawn for him.

Medications & Precautions

Surfactant instillation in trachea for babies < 30 wks gestation, Alveofact 50 mg/1.2 ml/ day given through the ETT for 4 consequent days during ŵ the baby on ventilator & ⇧ the PIP for 30 seconds after instillation. Antibiotics are given to cover both gram positive & negative bacteria & strict adherence to the nosocomial infection control protocol.

Nasogastric feeding & expressed breast milk

Do not start feeds until babies are hemodynamically stable & showing interest in feeding, once we decided to start enteral feeding, we select the method, amount & frequency. Initiate feeds é breast milk whenever possible, this ⇩ the risk of NEC 4 folds, the expressed breast milk is the ideal to initiate é, so that the infant gets the benefits of feeding colostrum, if not possible, feeding through milk pump é either breast milk or PT formula using amount equal to 20-40 ml/Kg/day & changing the milk /3 hours, as continuous drip through the NGT. If we decided to give 2 hourly feeding we start é 2 ml/feed & gradually ⇧ the amount 1 ml/12 hours to a maximum of 20 ml/feed to be reached by the end of 1st week, at the same time we have to do test feed before each feed (is very important), if there is residual milk > 25% of the previous feed we have to ⇩ the amount of subsequent feed or delay it. In case of good tolerance we shift to ½ strength then full strength PT formula & when baby is strong enough to suck, breast or bottle feeding is often possible

NB: NEC is a potentially life threatening infection & the risk factor ⇧ é prematurity, early feeding, high concentrated milk formula, IUGR, infection & umbilical catheterization.

SMALL FOR GESTATIONAL AGE

SGA refers to a, fetus that has failed to achieve a specific biometric or estimated weight threshold by a specific gestational age, it describe fetus or NN whose weight &/or crown heel length is at least -2 SD below the mean for an appropriate reference population. It is becoming increasingly recognized that being born SGA carries an elevated risk of developing metabolic disease in later life, particularly obesity, insulin resistance & dyslipidemia.

Incidence: depending on geographical region, it is between 8-28% of all infants born world-wide, including low, very low & ELBW.

Causes

Maternal factors: •Preeclampsia. •Hypertension. •APHge. •Chronic illness. •Chronic infection. •SLE. •Anaemia. •Malnutrition. •Malignancy. •Abnormal uterus. •Uterine fibroids. •Abnormal placenta. •Placental infarcts. •Partial abruption. •Placental hematoma. •Age < 16 or > 35 yrs. •Drug use. •Smoking. •Alcohol intake.

Fetal factors: ▲Multiple births. ▲Congenital malformation. ▲IUI. ▲IEM.

Environmental problems: ✖High attitude. ✖Pollution. ✖Toxic substance.

Clinical Picture

✳SGA. ✳Wasted look. ✳Dull hair. ✳Poor skin turgo. ✳Hypothermia: cold skin of trunk & extremities. ✳Poor feeding & sucking. ✳Shallow respiration & cyanosis. ✳Diminished activity & weak cry. ✳Hypoglycaemia.

Complications

- Perinatal asphyxia.
- Polycythemia.
- Aspiration pneumonia.
- Hypothermia.
- Hypoglycemia.
- Hypocalcaemia.

Diagnosis

*U/S: CRL & BPD.

*Date of last menstrual period.

*Inadequate maternal weight gain & fundal height <expected for gestational age.

*Non reassuring NST.

*Biophysical profile.

*Assessment of placental function.

Management

To guard against hypoglycemia & hypothermia. It is extremely important to treat hypoglycemia to prevent CNS damage, as the brain need glucose, early feeding is recommended, using glucose 10% either PO/IV, it is extremely important to ensure normal body temperature & the immediate precautions to be taken immediately after birth. In normal term baby delivered into warm environment, rectal temp may drop by 1-2^{0}C shortly after birth & may not achieve normal stable body temp until the age of 4-8 hours, while in LBW baby the ⇩ in body temperature may be much greater & more rapid unless special precautions are taken immediately after birth as the baby lose 0.25 ^{0}C/minute if not protected. So SGA Baby should be warmed quickly by wrapping in warm towel & use of

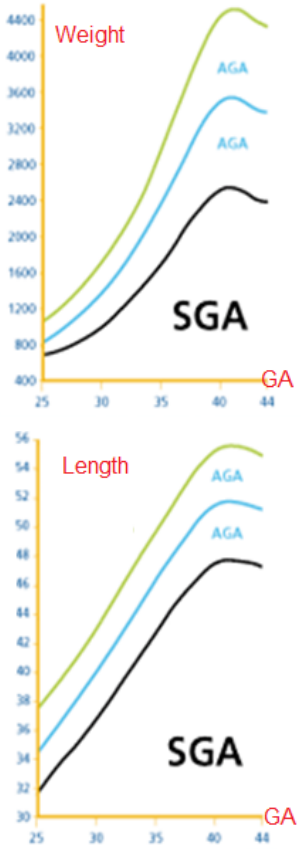
extra clothes or blankets to keep the body warm, if baby in incubator increasing the incubator`s temperature to achieve the NTE, use hot water bottle (50 $^0$C). Also the given food or even IVFs should be warmed in addition to avoidance of baby exposure to direct source of air draft & to check baby`s temperature frequently.

*Mild hypothermia: baby`s temp < 36 ^{0}C.

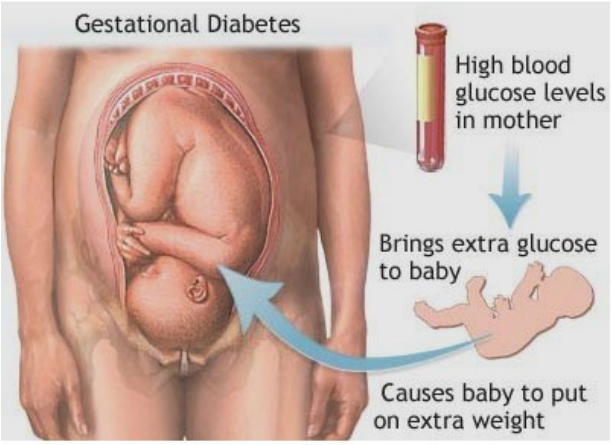
*Moderate hypothermia: baby`s temp <35.5 ^{0}C.

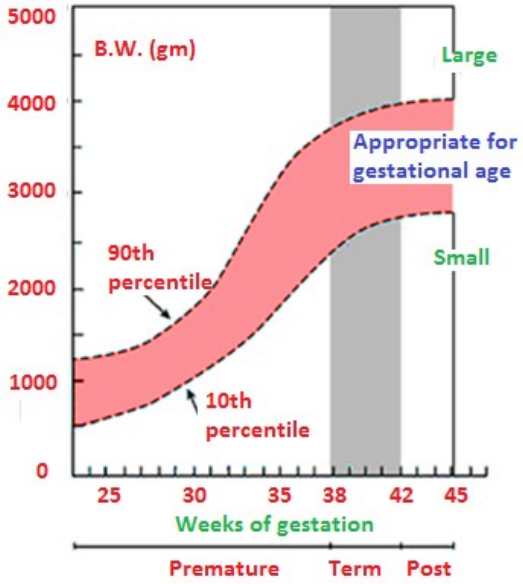
*Severe hypothermia: baby`s temp < 35 ^{0}C.

The effect of cold stress include; hypoxia & ⇧ of O_2 need, cold stress also cause ⇩ in surfactant production leading to respiratory distress, hypoglycemia, metabolic acidosis, jaundice, hyperviscosity & polycythemia (more RBCs than normal, as the baby responds to hypoxia by making more RBCs), partial exchange transfusion may be needed for polycythemia, using albumin 5% if Hb > 20 gm & Hct >65.



LARGE FOR GESTATIONAL AGE





Infant whose BW is ≥ the 90th percentile (> +2 SD) on the intra uterine growth curve, the baby may be PT, Post Term, or Term. LGA does not mean post maturity. Other than genetically determined size, the major cause of an infant`s being LGA is maternal DM. The macrosomia results from the anabolic effects of high fetal insulin levels produced in response to excessive blood glucose during gestation as insulin act during intrauterine life as the growth hormone. The less well controlled the mother diabetes during pregnancy, the more severe the fetus macrosomia. A rare non genetic cause of macrosomia is Beckwith-Wiedemann syndrome ŵ characterized by macrosomia, omphalocele, macroglossia & hypoglycemia.

Causes

∗Miscalculation of the date of conception. ∗Maternal DM. ∗Genetic predisposition. ∗Beckwith Wiedemann syndrome. ∗Infants é erythroblastosis fetalis.

Clinical picture

✵Large. ✵Obese. ✵Plethoric. ✵Poor motor skills. ✵Poor feeders. ✵Difficult to arouse. ✵RDS. & ✵Hyperbilirubinemia.

Complications

✻Perinatal asphyxia. ✻Difficult labor. ✻CS. ✻Birth injuries. ✻Hypoglycemia. ✻Polycythemia. ✻⇧ Susceptibility to infection.

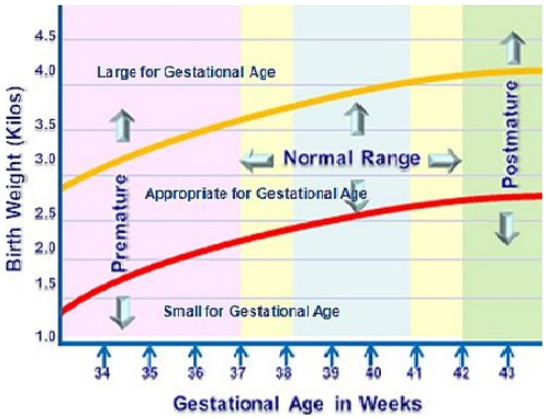
Management

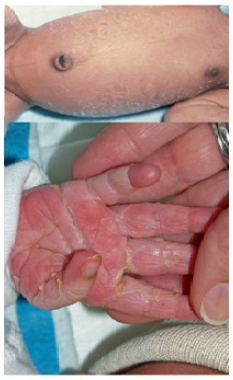
Induction of labor at 37 wks is recommended.

Monitor for hypoglycemia & Polycythemia (Hb & Hct).

Early feeding to keep blood sugar > 25 mg/dl.

POST TERM BABY





Post term pregnancy is a common situation seen in 5-10% of all pregnancies, it cause anxiety for both women & obstetricians, because it perceived as being a cause of ⇧ risk to the fetus.

Definition

Post term baby is ≥42 weeks (≥ 294 days), while post maturity syndrome is IUGR associated é meconium stained amniotic fluid, oligohydramnios, fetal distress, loss of SC fat, dry cracked skin reflecting placental insufficiency.

Causes

- The commonest cause is error in calculation of gestational age.
- History of post term delivery in previous pregnancies.

- Congenital anomalies like anencephaly ẃ disrupt fetal pituitary adrenal axis & rare maternal enzyme deficiency (placental sulphatase).
- In most cases the cause is not known.

Clinical picture

- Dry peeling skin & green/yellow coloring from meconium staining.
- Overgrow nails.
- Abundant scalp hair.
- Visible creases on palms & soles.
- Minimal fat deposit.

Complications

*The placental function declines sometime around term, this exposes the fetus to a state of relative hypoxia ẃ can affect the fetal growth & the biophysical parameters of fetal wellbeing.

*In pregnancies where placenta continues to function well beyond due date, fetus continue to grow almost at the same rate as in 3rd TM.

*⇧ Maternal morbidity é large for date or macrosomic babies occurs because of ⇧ incidence of prolonged labor, shoulder dystocia, this result in an ⇧ risk of; pelvic floor trauma, instrumental deliveries & CS in 25% of cases.

*Fetal hypoxia, ⇩ liquor are associated é ⇧ incidence of meconium stained liquor & abnormal FHR pattern during labor.

*⇧ Risk of PPHge & endometritis.

*Still birth rate ⇧ significantly, it is 0.35/1000 pregnancies at 37 week, while it ⇧ to 2.1/1000 pregnancies at 43 week gestation.

*Meconium aspiration, asphyxia before, during & after delivery.

*Cord compression (fetal hypoxia).

*Fractures & peripheral nerve injury as result of difficult labor.

*IC Hge, pneumonia & septicemia.

INTRA UTERINE FETAL DEATH

Seen in 6.9/1000 births. Foetal death prior to the complete expulsion or extraction from the mother`s womb after 20 weeks gestation or fetal weight >500 gm when the gestational age is unknown. Include; early fetal death 20-27 weeks & late fetal death ≥ 28 weeks.

Causes

Maternal factors: •Advanced maternal age (>35 yrs). •Chronic illness of mother: DM, hypertension, nephritis. •Preeclampsia. •Post term pregnancy. •Antepartum asphyxia. •Placenta abruption. •Multiple pregnancy. •Rh isoimmunisation. •Previous history of still birth. •Obesity (risk of DM, hypertension, placental dysfunction). •Race: the black women has higher still birth rate. •Low socioeconomic or educational status. •Smoking, tobacco. •Drug abuse.

Fetal factors: •Congenital malformations. •IUGR or Prematurity. •Male chromosomally has poor survival rate than female. •IUI usually occur in fetus weight <1000 gm.

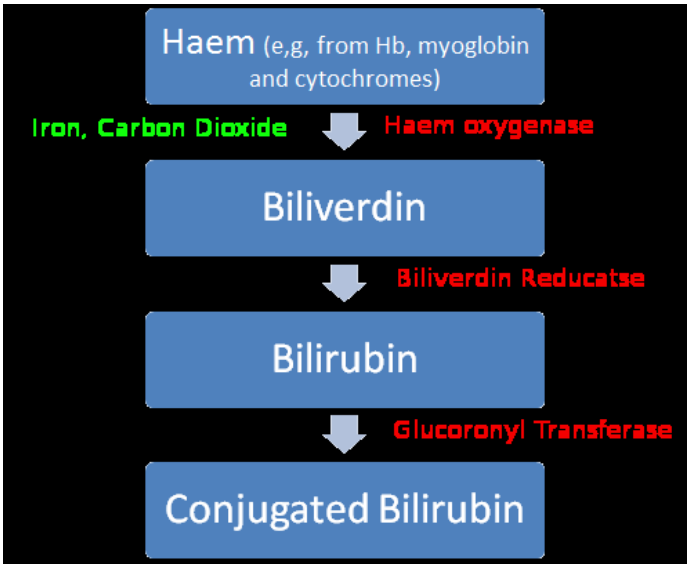
Diagnosis

- Mother reports loss of fetal movements.
- Fundal height on palpation is < the estimated gestational age, or fundal height regress es as compared to previous documentation.
- Absence of FHS.
- U/S confirmatory (absence of FH activity).

NEONATAL JAUNDICE

One of the most common conditions requires medical attention in NN. Yellowish discoloouration of skin & sclera because of ⇧ serum level of bilirubin ŵ may be indirect (unconjugated) or direct (conjugated). Occur in 60% of term infant & 80% of preterm on first week of life. In most cases there is no underlying disease, but some will suffer from pathological jaundice. Physiologic jaundice is a normal phenomenon during transition becomes concerning when levels continue to rise. The neurotoxic compartment of bilirubin is the unconjugated form.

Bilirubin Metabolism



Most bilirubin in NN (85%) come from catabolism of Hb, the rest (15%) come from myoglobin & cytochromes. One gram of Hb gives about 35 mg bilirubin. Hb breakdown into biliverdin + carbon monoxide in the RES, biliverdin acted upon by biliverdin reductase enzyme to be reduce to lipid soluble unconjugated bilirubin (indirect bilirubin), ŵ combine é Y, Z proteins (Ligadin) transporting it to liver where conjugation takes place é glucuronide by effect of glucuronyl transferase enzyme to form water soluble conjugated bilirubin (direct bilirubin), ŵ excreted in intestine, where part of it reunconjugated & rea-

bsorped into blood stream to pass to liver (enterohepatic circulation), another part acted upon by bacteria in gut changing it to urobilinogen ŵ again reabsorbed into circulation, excreted in urine, another part acted upon by bacteria in gut changing it to stercobilinogen, excreted in stool.

Unconjugated Hyperbilirubinemia

Physiological jaundice

Most common cause of jaundice in NN period. It usually appear in 2nd - 3rd day of life in FT infant & 3rd - 4th day in PT. Peak usually 6-8 mg/dl & not >12 mg/dl in 3rd day in FT. It is usually 10-12 mg/dl & not >15 mg/dl in 5th day in PT. Usually disappear by 4-5 days (rarely by 7-10 days) in FT & usually by 7-9 days (rarely by 10 days - 2 weeks) in PT. The rise of bilirubin should be not >5 mg/dl/24 hours or not > 0.5 mg/dl/hour.

Causes of physiological jaundice: ⇧ production of bilirubin due to ⇧ RBC volume/kg body weight & ⇩ RBC survival (70-90) days in infant versus 120 days in adult, the breakdown of HbF as it is replaced by Hb A, the ⇧ ineffective erythropoiesis & ⇧ turnover of non Hb hem proteins, also absence of intestinal flora & immaturity of liver (conjugation), ⇩ hepatic excretion of bilirubin. Relatively low activity of glucuronyl transferase. ⇧ of enterohepatic circulation by high level of intestinal B-glucuronidase, ⇩ intestinal bacteria, ⇩ gut motility é poor evacuation of bilirubin laden meconium. Defective uptake of bilirubin from plasma by ⇩ ligandin (for conjucation) due to ⇩ of UDPG-T.

Physiological jaundice may be exaggerated (⇧ Peak & Duration) by:

•Prematurity. •Breast feeding. •Male sex. •Cephalohaematoma. •Cutaneous bruising. •Polycythemia. •Weight loss. •Dehydration. •Caloric deprivation. •Delay bowel movement. •Maternal DM. •Drug (Vit K3, Novobiocin, Oxytocin). •Trisomies (21, 18, 13).

The jaundice should not be regarded as physiological jaundice & should regarded as path-

ological jaundice & must be investigated if:

- It appears in the 1st 24 hours of life.
- TSB increasing >5 mg/dl/24 hour or > 0.5 mg/dl/ hour.
- TSB > 12 mg/dl in FT or > 14 mg/dl in PT.
- Duration of jaundice is >10-14 days or present at or beyond age 2 weeks.
- Direct bilirubin >2 mg/dl at any time or pale stool, dark urine +ve bilirubin.

ABO incompatibility

It is unconjugated hyperbilirubinemia, mother usually type O blood group, baby type A or B, confirmed by Anti A or Anti B in maternal circulation (IgG type), in addition to positive coombs test & reticulocytosis.

Rh incompatibility

15% of mothers are Rh -ve, if the baby is Rh +ve, D antigen pass to maternal circulation forming anti D ŵ in subsequent pregnancies pass to fetal circulation causing hemolysis, first baby usually escape, diagnosis confirmed by Rh grouping, +ve coombs test & reticul-ocytosis. Management of Rh incompatibility include the Rx of mother during pregnancy & during the subsequent ones by giving Anti D human gamma globulin within 72 hr from delivery & in the next pregnancy to give 1st dose in last TM & the 2nd dose to be given within 72 hr from labor, in addition to testing Rh antibodies in subsequent pregnancy.

Polycythemia

Causes: delayed clamping or excessive milking of umbilical cord during labor, SGA, Beckwith & Down syndromes, thyrotoxicosis, congenital adrenal hyperplasia.

Clinical picture: plethoric face, peripheral cyanosis, respiratory distress & may cause NEC, renal vein thrombosis, acute tubular necrosis, or cerebral infarcts.

Diagnosis: Hb > 20 gm/dl., Hct > 65.

Management: partial exchange transfusion using albumin.

$$\text{Volume(mL)} = \frac{\text{Initial Hct} - \text{Desired Hct}}{\text{Initial Hct}} \times \text{Weight (kg)} \times \text{Blood Volume}$$

Blood volume = 70-90 ml/kg for term & 85-110 ml/kg for preterm infants

Breast milk jaundice

One of the causes of prolonged NN unconjugated hyperbilirubinemia when baby may or may not has physiological jaundice at beginning, develop significance ⇧ of bilirubin between 1st & 2nd wk (usually after 7th day) of life, reach maximum 10-30 mg/dl during 2nd - 3rd wk. The cause is unclear & number of theories have been suggested e.g. during the first few days baby on breast feeding is not getting enough nutrients & fluids necessary to help their body to breakdown & excrete bilirubin, the delayed passage of meconium, the ⇧ of enterohepatic circulation leading to hyperbilirubinemia, presence of 5 α 20 pregnandiol in mother milk, ŵ is abnormal metabolite of progesterone inhibit conjugation of bilirubin. If breast feeding continue the jaundice persist for 3-10 wks at lower level, but if we stop breast feeding there will be rapid ⇩ to reach normal level within few days éout return of hyperbilirubinemia when restart breast feeding. Before Rx of breast milk jaundice we have to exclude other causes of unconjugated hyperbilirubin-emia as hemolysis, hypothyroidism. For Rx we have to stop breast feeding for 1-2 days. Sometimes phototherapy may be needed, very rarely kernicterus has been reported.

Excessive swallowing of maternal blood

Baby may vomit blood as a result of excessive swallowing of maternal blood during labor. Diagnosed by APT test; 3 drops of vomited blood + 0.5 ml water +1 ml sodium chloride, if brown color developed it indicates that blood is of maternal origin (Hb A) resulting from swallowing maternal blood & is reassuring. If no color change occur this means that blood in the vomit is of fetal origin (Hb F) & will need further investigations for the cause.

Red blood cells enzymes deficiency

1- Glucose 6 phosphate dehydrogenase deficiency

X Linked Recessive, Chromosome, Gene G6PD, Location X q 28.

Incidence

Common in Negros & Middle East. Occur in 11-13% of African Americans. Estimated 400 million people worldwide carry the gene.

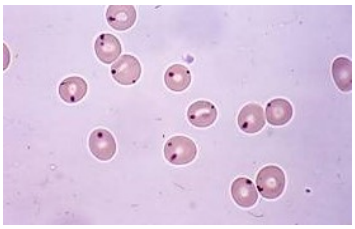
Result from break down of RBCs, ẃ can be triggered by infections, severe stress & certain foods. Episodic haemolysis from **fava beans**, oxidant drugs (especially Primaquine, Sulpha, Amiodarone, Antimalarial, Nitrofurantoin, Antihistaminics, Antituberculous, Aspirin). G6PDD is a significant cause of mild to severe jaundice in NN.

Clinical picture

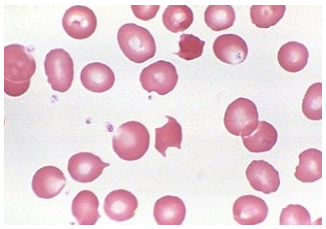
- Anaemia. • Fatigue. • Tachycardia. • Shortness of breath. • Dark urine. • Splenomegaly.

Diagnosis

CBC/Blood smear & G6PD levels. Pts are less susceptible to malaria.



B. Smear- G6PD, showing Heinz Bodies (denatured haemoglobin)



Blood Smear- G6PD, showing Bite cells. Prussian blue staining (detects hemosiderin)

Differential Diagnosis

Sickle cell disease (painful crisis). Pyruvate kinase deficiency (hemolysis not precipitated by drugs or infections).

Management

- Avoidance of precipitating factors.
- Gradually improve by age.

2- Pyruvate kinase deficiency

Is AR inheritance, common in people of Northern Europe. The enzyme required for production of ATP in RBC`s, deficiency leads to ⇩ RBC`s life span & hemolysis. Diagnosed by estimation of blood level of pyruvate kinase.

Abnormalities of shape of RBCs

Hereditary Spherocytosis, Elleptocytosis, or Stomatocytosis

Is AD inheritance. Incidence: 1/5000 life birth. Unusual in NN period. Characterized by; blood hemolysis & hyperbilirubinemia. Diagnosed by blood smear examination, marked ⇧ in reticulocytic count & osmotic fragility.

Sepsis: secondary to hemolysis. Impairment of bilirubin conjugation.

Sequestration: blood in body cavities as the body metabolizes Hb, as in case of cephaloh-aematoma, subdural, subgaleal hematoma & é excessive bruising.

Transient Familial NN Hyperbilirubinemia

Disorder of conjugation, NN develop severe non hemolytic hyperbilirubinemia, their serum contains high conc. of glucuronyl transferase inhibitors ŵ ⇩ by about 14 days of life & consequently hyperbilirubinemia resolves, so no specific Rx usually needed.

Intestinal obstruction

Causes ⇧ of enterohepatic circulation, as in pyloric stenosis, cong. atresia, malrotation, Hirshsprung, cystic fibrosis (each gm of meconium contain 1 mg of bilirubin).

Deficiency of bilirubin receptors or carriers

Deficiency of Y & Z “Lignadin“ or competitive inhibition by drugs as valium or Lasix.

Crigler Najjar Syndrome

Familial non hemolytic unconjugated hyperbilirubinemia, is inherited deficiency of conj-ugation of bilirubin. Different degrees of ⇩ in hepatic Uridine 5'-Di-Phospho-Glucuronos-

yl Transferase UDPG-T, it include 2 types;

Crigler Najjar type 1

AR, complete abscence of the enzyme, rare, incidence 1/1000.000 live births. Both parents show abnormal LFTs. Clinically presented é severe unconjugated hyperbilrubine mia é no hemolysis, develop in the first 3 days of life ŵ may reach 25-35 mg/dL during the 1st month & may continue after that. kernicterus is common.

Diagnosis

- Based on High level of unconjugated bilirubin é no hemolysis
- Bilirubin in bile < 10 mg/dl in contrast to normal 50-100 mg/dl.
- No response to Phenobarbitone (differentiated from type 2).
- Definitive diagnosis is established by measuring hepatic glucuronyl transferase activity in liver specimen. Done by close biopsy not open because surgery & anesthesia may precipitate kernicterus.

Treatment

May need continuous phototherapy & repeated exchange transfusion to prevent kernicterus. After the NN period the risk of kernicterus still present but é level >35 mg/dl so we have to put the baby on phototherapy (usually we put children during night). Cholestyramine or agar used to bind photobilirubin product & thus interfere é enterohepatic circulation. Rx of infection or any illness to prevent development of kernicterus. Orthotopic hepatic transplant cure disease. Plasmapheresis. Metacloporphyrin may be used ŵ prevent hemeoxygenase so inhibit formation of bilirubin. Genetic engineered enzyme replacement, Liver direct gene therapy & hepatocyte transplant remain option in future.

Crigler Najjar type 2

AD, partial defect in hepatic UDPG-T, one of the parents show abnormal LFTs. Jaundice

usually not exceeding 20 mg/dl. Persist into adulthood. May be present similar to Type 1 or may be less severe even occasionally éout NN manifestation. When present in NN period unconjugated hyperbilirubinemia usually occurred in the first 3 days. TSB level may compatible é physiological or pathological level. Jaundice persists in & after 3rd week between 1.5-22 mg/dl. Stool color is normal. Bilirubin in bile is nearly normal.

Treatment: responds dramatically to phenobarbital 5 mg/kg/day for 7-10 days.

Conjugated hyperbilirubinemia

Indicate liver disease, the direct SB >20% of total SB.

Neonatal hepatitis syndrome

Nonspecific hepatic inflammation, develop 2ry to IUI, IEM, endocrinal disorders as hypothyroidism, hypopituitarism. Characterized by dark urine, pale yellow stools, poor feeding, hepatosplenomegaly, bleeding from vit K deficiency & abnormal LFTs.

Biliary Atresia

Either atresia, hypoplasia, or choleducal cyst. Early diagnosis carry good prognosis as it prevent intrahepatic damage. Investigations include; U/S abdomen, LFTs, operative cholangiography is diagnostic (checking BT, CT & platelet before doing it).

Cystic fibrosis

AR, common in Europe, 1/2500 live births. Affect lungs, digestive tract & pancreas by viscous secretions. Characterized by; poor growth, steatorrhoea (pancreatic insufficiency), chest infection, cough, wheezing, dyspnea, meconium ileus, salty taste skin.

Diagnosis: sweat chloride test at age 2-6 wks, measure conc. of Cl^+ & Na^+ excreted in sweat (collect 40 mg of sweat, it is +ve if contain >50 meq/l).

Intrauterine Infection

- Toxoplasmosis (IgM toxoplasma specific antibodies).
- Rubella (IgM rubella specific antibodies).
- CMV (IgM CMV specific antibodies).
- Herpes (viral culture of vesicle scraping).
- Syphilis (FTA-ABS test, VDRL).
- Hepatitis B (HBsAg, anti HBc IgM, HBV DNA-PCR).

Endocrinal disorders

Congenital hypothyroidism

Many countries do screening program for congenital hypothyroidism on 7th-10th day of life, if TSH is high, confirmatory test by T_3 & T_4 to be done. Hypothyroidism should be exclude in every infant.

Incidence: 1/3000 live births.

Clinical picture: usually present é unconjugated hyperbilirubinemia, but may be conjugated & associated é NHS. Characterized by; prolonged jaundice, lethargy, reluctant to feed (physically &mentally constipated baby), big tongue, cold skin & delayed milestones.

Investigations: TSH, T_3, T_4, & also bone age for detection of the delayed bone age.

Management: Rx of jaundice & compensation for hypothyroidism by using Eltroxin 50, 100 ug tab, 4 ug/Kg/day as early as possible to protect the baby from MR & to allow normal growth & development.

Hypopituitarism

Pituitary adrenal dysfunction. 50% of cases associated é NHS. May be due to hypothalamic deficiency of anterior/posterior pituitary function.

Chromosomal anomalies

Trisomy 21, 18 & Alagille sy, diagnosed by its characteristic features & karyotype studies.

Causes of jaundice in the 1st 24 hr

- Hemolysis (ABO & Rh incompatibilities, G6PD, pyruvate kinase def, spherocytosis).
- Concealed Hge (cephalohaematoma, hepatic & splenic Hge).
- Sepsis.
- IUI.

Causes of prolonged jaundice

"jaundice is prolonged if the duration > 2 wk & may persist in & beyond 1st month".

- Breast milk jaundice.
- IUI.
- Idiopathic NN hepatitis.
- Biliary atresia.
- Hemolysis & inspisiated bile syndrome follows hemolytic anemia..
- Hypothyroidism.
- Hereditary glucuronyl transferase deficiency (Crigler Najjar syndrome).
- Intestinal obstruction (Pyloric stenosis).
- Hyper alimentation & cholestasis.
- IEM "Galactosaemia".

Clinical criteria to assess neonatal jaundice

Body area	Bilirubin range mg/dl
Face	4-8
Upper trunk	5-12
Lower trunk & Thigh	8-16
Arms & Lower legs	1-18
Palms & Soles	>15

Routine Investigations for NN jaundice

Hb.
Retic count.
Serum bilirubin total & direct.
Coombs test direct & indirect.
Rh grouping.

Specific Investigations for NN jaundice

Haemolysis: Rh grouping, CBC, retic count, RBCs appearance, direct coombs, G6PD.

Infection: TORSCH screening.

Endocrinal: thyroid function tests: TSH, T_3, T_4..

Metabolic disorders: reducing substance in urine, amino acids.

Biliary atresia, Choleducal cyst: U/S abdomen.

Others: Hb electrophoresis, G6PD, UDPGT enzyme.

Kernicterus

Is a neurological syndrome result from the deposition of unconjugated (indirect) bilirubin in the brain cell especially basal ganglia, so unconjugated bilirubin is toxic to CNS & when bilirubin exceed bilirubin binding capacity of albumin, the free bilirubin will cross BBB & diffuse to brain cell & cause cell damage & Kernicterus. The precise level of indirect bilirubin ŵ is toxic to brain & the duration of exposure was unknown but Kernicterus is unusual & rare in healthy FT at bilirubin <25 mg/dl é no hemolysis. There is some risk factor that ⇧ the possibility of Kernicterus at lower level of bilirubin ŵ may damage BBB or ⇩bilirubin binding capacity of albumin as hemolysis, hyperosmolality, IV Hge, acidosis, hypoalbuminemia, hypothermia, drug, hypoglycemia, hypoxia, sepsis, asphyxia, meningitis & prematurity. So for example LBW infant develop Kernicterus at lower SB level than 20-25 mg/dl or even 6-7 mg/dl in VLBW as seen in autopsy.

Clinical manifestations

Symptoms of Kernicterus usually appear at age 2-5 days in FT & 7th day in PT, but it can occur at any time in NN period. The early signs are usually indistinguishable from sepsis, asphyxia, hypoglycemia & ICHge, include poor feeding, lethargy, loss of Moro reflex, hypotonia, high pitch cry, irritability. Then at end of 1st & 2nd week the infant become gravely ill, prostrated, RD, pulmonary Hge, bulging fontanels, ⇩ tendon reflex, twitching of face or limbs, hypertonia of extensor muscles, opisthotonos, shrill high pitch cry, convulsion, rigidity is rare. Many infant who reach this stage die & most of survival usually develop later complete neurological syndromes but may appear to recover for 2-3 month (appear é little abnormality).

Later in the 1st yr opisthotonos, muscle rigidity, ⇧deep tendon reflexes, irregular movement, hypertonia, obligatory tonic neck reflex & convulsion.

In 2nd yr opisthotonos, seizure abate but irregular involuntary movement rigidity, hypertonia or in some hypotonia ⇧ steadily.

By 3rd yr complete neurological syndrome develop ŵ include; chorioathetosis, involuntary muscle spasm, extrapyramidal signs, fit, MR, dysarthritic speech, high frequency hearing loss, spastic quadriplegia. Pyramidal signs, hypotonia, ataxia may occur in few.

Prevention

Screening for hyperbilirubinemia &presence of risk factors in the first 24- 48 hr of life to detect infants at high risk for severe jaundice by physical exam & investigations. Early measurement & follow up of SB level in any jaundiced baby & Rx accordingly. Check bilirubin in jaundiced baby in the 1st day of life & to be evaluated for possible hemolytic disease. Avoid visual assessment in estimation of jaundice severity. Parental communication to concerns about infant's skin color & education about potential risks & neurotoxicity.

Mothers should be advised to nurse infant every 2-3 hrs in order to ensure adequate hydration & caloric intake & to avoid routine supplementation é water or glucose water. Rx of condition that ⇧ risk of Kernicterus, as sepsis, acidosis. Prevention of Rh isoimmunisation where any pregnant Rh -ve woman, giving her human anti D globulin when she delivered Rh +ve baby or develop abortion.

Management of neonatal jaundice

Phototherapy

Serum Bilirubin	Day one <2.5 kg	Day one >< 2.5 kg	Day two <2.5 kg	Day two >< 2.5 kg	Day three or more <2.5 kg	Day three or more > 2.5 kg
0- 4 mg/dl						
5 - 9	Phototherapy					
10 – 14	Exchange		Phototherapy			
15 – 19	Exchange				Phototherapy	
20 or more	Exchange					

Used for FT baby é SB > 14 mg/dl in 2nd or 3rd day of life. Blue light, wave length 450-460 nm from distance of 45 cm. Phototherapy result in photo isomerization (formation of photo bilirubin) ŵ is nontoxic isomer, water soluble excreted in urine & stool. Either through continuous method: exposure 18 hrs, taking baby out for each time of feeding,

or intermittent method: 15 min on phototherapy, 45 min off. Eyes must be protected by mask. Fluid intake to be ⇧ 20% to compensate for fluid loss from heat.

Blood Exchange Transfusion

This procedure, used most commonly to treat severe unconjugated hyperbilirubinemia (SB > 19.5 mg/dl).

Technique of blood exchange transfusion

- All BET are to be conducted in NICU level III.
- Before exchange: 10% albumin, 1 gm/Kg IV over 2 hrs, to ⇧ bilirubin conjugation.
- Exchange should be done under radiant warmer.
- Donor blood, warmed to temp < 37 ^{o}C.
- Monitoring baby BP, RR, HR, general condition.
- BET kit contains catheters, stopcock, waste bag, Ca gluconate sol. 10%.
- Fresh whole blood, same baby group & Rh. In emergency we use group O Rh-ve.
- Exchange volume generally twice infant BV this remove 88% of infant RBCs.
- BV in FT infant is 70-90 ml/Kg & PT infant it is 85-110 ml/Kg, for purpose of simplicity approximate 90 ml/Kg will be used for both.
- Complete aseptic conditions, sterilization of area thoroughly, put sterile towels to cover abdomen, insert catheter, push-pull technique through umbilical vein, tip of catheter should be in IVC just above diaphragm.
- Withdraw 10 ml in FT & 5 ml in PT each time over 2 minutes duration & push them to waste bag using the stopcock, take same amount from fresh whole blood infuse them slightly faster to infant using the stopcock. In case of crying, irritability or ⇧ HR, slow down the procedure.
- Total duration for exchange is 2-4 hrs.

- Ca gluconate 10%, 1 ml IV every 100 ml blood exchange to be given.
- Check BS of infant every 30 min during exchange.
- At end of exchange, blood sample se-nt for Na^+, glucose, TSB & DSB, Hb & Hct.
- Rebound ⇧ of SB occurs after 2 hrs then it ⇩. Check it 6 hourly/
- Feeding allowed 2-4 hrs after exchange.

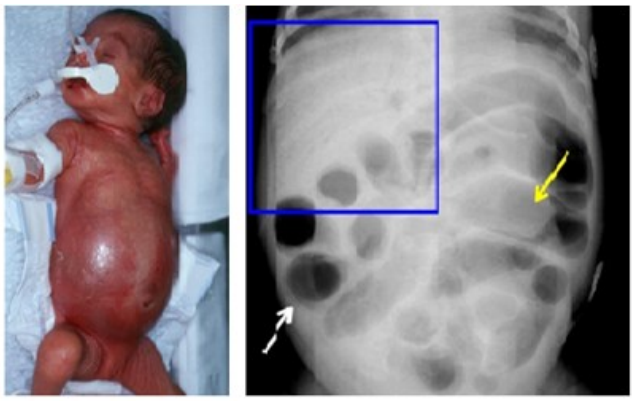
Complications of BET

- Hypocalcaemia (citrated BL), Hypokalaemia (old blood).
- Hypoglycaemia.
- Blood group incompatibility.
- Embolism.
- NEC.
- Cardiac arrhythmia.

Criteria for control

- Total SB < 13 mg.
- Daily rise of SB < 0.5 mg.
- Direct SB < 1.5 mg.

NECROTIZING ENTEROCOLITIS



Multiple dilated loops of bowel (yellow arrow), linear radiolucency is seen paralleling the bowel wall indicating air in the wall (white arrow),air in the portal venous system (blue box).

Seen in 3/1000 of live births, 90% of cases are seen in PT & 10% in term babies. It is the most common GIT emergency in PT, of unknown etiology but may occur as epidemic in the word raising the question of being bacteriological in origin, represent 5% of all admission to the NICU, result from inflammation & necrosis of intestinal walls, commonly affect terminal ileum & proximal colon, onset within the 1st-3rd wk of life, perforation is serious complication requiring immediate surgery & occurs in late stage, commonly occurs within 48-72 hrs after pneumatosis intestinalis or portal venous gas detection.

Precipitating factors

- Prematurity: inadequate perfusion of gut mucosa, perinatal hypoxia.
- SGA: more é BW < 1500 gm.
- PRM.
- Placenta abruption.
- Low Apgar score.
- Maternal preeclampsia.
- Antenatal cocaine abuse
- Gastroschissis.
- Cardiopulmonary diseases; cyanotic CHD,PDA, RDS, lead to ⇩COP & ⇩perfusion of GIT.
- Aggressive advancement of enteral feeding, early introduction of feed, over feeding,

hyperosmolar feeding, or hyper osmolality of the IVFs.

✲ Umbilical catheterization, blood exchange transfusion.

✲ Polycythemia & thrombocytopenia.

✲ Hypothermia & Septicemia ✲ Hypothyroidism.

Clinical picture

Bell`s staging criteria include; "systemic, intestinal & radiological assessment"

▲ Stage I suspected: temp instability, lethargy, poor feeding, mild abdominal distension, vomiting ŵ may be projectile, bile stained, normal abdominal X ray.

▲ Stage II definite: metabolic acidosis, thrombocytopenia, ⇩ intestinal sounds, abdominal wall oedema & tenderness, abdomen XR shows ileus & pneumatosis intestinalis.

▲ Stage III advanced: hypotension, toxic, apnea, DIC, neutropenia, occult or bloody stool, anuria, marked abdominal tenderness, distension, abdominal wall erythema, petechiae or bruises, abdominal masses & characteristic doughy sensation of abdomen, absent intestinal sounds, abdominal X ray shows portal vein gas, ascites, lastly perforation & pneumoperitoneum. DIC occur generally in severe illness as late stage of NEC, asphyxia or sepsis. DIC characterized by consumption of all clotting factors é ⇧ in FDPs & thrombocytopenia, all clotting tests are prolonged (PT, CT, APT), it carries poor prognosis".

Investigations

🕊 X ray Abdomen: erect, supine, lateral horizontal, daily for early detection of intestinal perforation, immediate surgical consultation, pneumatosis intestinalis (presencece of submucosal, subserosal air on intestinal wall), portal vein gas (extension of gas into portal vein), pneumoperitoneum (air in abdominal cavity) or under diaphragm.

🕊 CBC: thrombocytopenia & neutropenia.

🕊 Electrolytes: hyponatremia, hyperkalemia & hypoproteinemia.

Management

✻NPO ✻Nasogastric suctioning two hourly: for gastric decompression. ✻Removal of umbilical catheter & placement of peripheral line. ✻Fluid chart: for the intake & out-put ✻TPN: starting from the 3rd day to assure adequate nutritional growth. ✻Check CBC, platelet, electrolytes every 12-24 hrs. ✻IV antibiotics to cover a broad range of aerobic & anaerobic intestinal bacteria: Ampicillin 1000 mg vial, 100 mg /Kg/day ÷ 4 doses IV + Gentamicin 3 mg /Kg/day ÷ 2 IV or IM + Flagyl 15 mg/Kg/day as continues infusion IV, bottle 500 mg in 100 ml sol + Oral Neomycin, 500 mg tab. 25 mg/Kg/ day ÷ 4 for 3 days. ✻Dopamine infusion: improve intestinal, renal perfusion & ⇧ COP. Dopamine amp 10 ml contain 250 mg, dose 2-5 ug/Kg/min. How to calculate the dose of Dopamine? BW X 3 = amount to be given in milligrams, to be diluted é 50 ml glucose 5% & give through infusion set, at a rate of 2 ml/hr, ŵ is equal to 2 ug/ min, dose can ⇧ up to 5ug/min. Higher doses of Dopamine > 5ug/min possess inotropic & chronotropic effect on heart + peripheral vasoconstriction. ✻Blood transfusion or fresh frozen plasma or 5% albumin 10-20ml/Kg. ✻Platelet concentrate transfusion (1 unit/5 kg BW). ✻Severe cases: ABGs, acid base regulation, O_2 supply & mechanical ventilation may needed. ✻Surgical interv-ention é the occurrence of perforation.

Prophylactic measures

- Avoidance of premature birth.
- Antenatal steroids for at risk babies (PT < 34 wk).
- Early institution of minimal enteral feeding (human milk have a protective factors & significantly lower the risk) &avoidance of hypertonic formula.
- Limiting duration of empiric antibiotics to < 5 days & prompt Rx of polycythemia.
- Placement of umbilical artery catheters é tip below level of inferior mesenteric artery.

NEONATAL CONVULSIONS

Seizures in the NN period constitute a medical emergency.

Incidence: 1.5-3.5/1000 live term births & 10-130 /1000 live PT births.

Causes

1- HIE (40-60%): the commonest cause 2ry to perinatal asphyxia, usually present within the first 24 hrs of life.

2- IC Hge (30%): seen in the second to seventh day of life, include intraventricular Hge (mostly in PT), intracerebral, subdural, or subarachnoid He (mostly in term babies).

3- Infection (5%): bacterial or viral meningitis, encephalitis, meningoencephalitis secondary to IUI may presented as seizures in the NN period, septicemia, tetanus, severe RD.

4- Metabolic (3%): hypoglycemia, hypocalcaemia, hypo/hypernatremia, pyridoxine dependency, IEM as nonketotic hyperglycinemia, gamma aminobutyric acid transaminase deficiency, glucose transporter type 1, cerebral creatine deficiency.

5- Congenital: chromosomal anomalies, congenital brain anomalies, cerebral dysgenesis, hydrocephaly, microcephaly, neurodegenerative disorders, neuronal migration defects (lissencephaly, pachygyria, schizencephaly) are rare causes of seizures in NN.

6-Miscellaneous (4%): as é polycythemia, maternal narcotic withdrawal. In older age Rey`s syndrome ẃ is associated ē hepatic encephalopathy, severe hypoglycemia, hyperammonemia & abnormal LFTs. Tetany is associated ē low Ca^{+}, high Ph^{+} & normal serum alkaline phosphatase & X ray skull may show basal ganglia calcification.

Classification

Subtle seizures: 50% of cases of seizures, are the commonest type of seizures in the NN period, it may be difficult to differentiate it from extremes of NN behavior, many subtle seizures are thought to arise from the basal ganglia as a result of diminished cortical inhi-

bition, further depression of the cortex é anticonvulsant may not alter these seizures, more common in FT than PT infants, occur in babies é severe global insult e.g. HIE & IV Hge. Occur in the form of:-

*Ocular movements é eyelid blinking or fluttering, nystagmus, jerky movements, rolling up of eyes, fixation of gaze.

*Oral-buccal-lingual movements; sucking, smacking, chewing & tongue protrusions.

*Progressive movements; crowing, swimming, pedaling, bicycling, limb boxing.

*Autonomic phenomena; tachycardia, bradycardia. *Apnea

*Complex purposeless movements; sudden arousal é crying & limb hyperactivity.

Colonic seizures: 25% more common in term babies, consciousness usually preserved, occur é HIE & birth trauma, often signals focal cerebral injury (cerebral artery infarction), or metabolic disease, either colonic-focal, multifocal, or rhythmic jerks (1-3/sec.) localize in a small part of the body, face, limbs, or axial muscles, or twitching migrate haphazardly from one limb to another.

Myoclonic seizures: 20% carry the worst prognosis, more frequent in PT, associated é the most severe brain damage, seen in developmental defects & anencephaly, seen also é drug withdrawal (especially opiates), are rapid, single or arrhythmic repetitive jerks, may affect a finger, limb, or whole body, may mimic the Moro reflex (resemble salaam spasm) or startling responses, most likely associated é EEG changes.

Tonic: 5% more common in preterm & often signals severe ICHge, also seen in kernicterus. May resemble decerebrate (tonic extension of all limbs) or decorticate posturing (flexion of upper limbs & extension of lower limbs), deviation of the head, eye signs, heavy breathing, apnea. 30% have EEG correlation, often difficult to treat é anticovulsants.

Diagnosis

Careful study of maternal chart, history of perinatal asphyxia, or birth trauma, maternal drug abuse, family history of seizures, maternal diabetes, presence of bulging fontanels may suggestive of meningitis or IC Hge, the wide variety of presentation depending upon the etiology includes; seizures, irritability, lethargy, hypotonia, apneic attacks, tachycardia, hypothermia, focal neurological signs, cranial nerve palsy, horizontal deviation of eyes, +ve Kernig`s sign (inability to extend the knee when the leg flexed at the hip), +ve Brudziniski`s sign (flexion of head accompanied by flex- ion of lower limbs).

Diagnosis according to time of onset of seizures

Time of onset: during the 1st -3rd day of life

- IC Hge
- Hypoglycemia
- Hypocalcaemia
- Hypo/Hypernatremia
- Pyridoxine deficiency
- Congenital cerebral malformations
- Narcotic withdrawal.

Time of onset during the 4th -7th day of life

- Meningitis.
- Encephalitis.
- Hypomagnesaemia.
- IUI (TORSCH).
- Kernicterus.
- Tetanus.
- Developmental malformation.
- Pyridoxine dependency.

Differential Diagnosis

Normal behavior: stretching, nonspecific random movement, random sucking, coughing or gagging, benign neonatal myoclonus ŵ occurs during active sleep (rapid eye movements).

Jitteriness: occurs primarily in response to minor stimuli in the form of rapid, oscillatory, tremors & movements cease when limbs is held or flexed, associated é no ocular movements or deviation, may occur while the baby is awake or sleep.

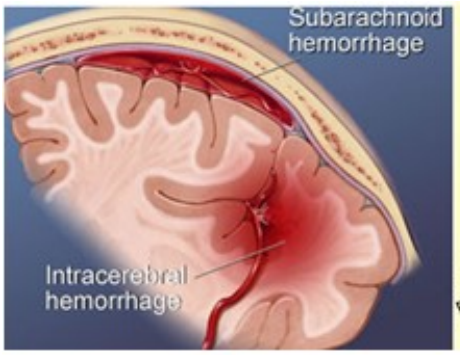
Benign familial NN seizures: typically occur in first 48-72 hrs of life, does not continue after NN period, +ve family history of seizures & normal development.

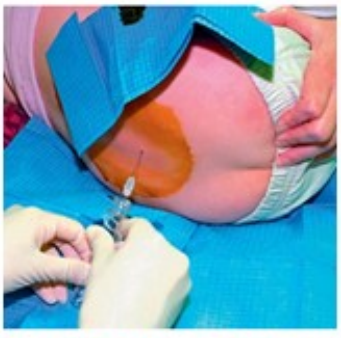
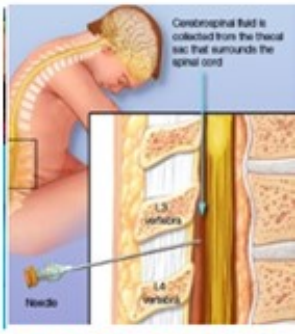
Benign idiopathic NN seizures: typically presents at day 5th also called 5th day fits, multifocal in type & no cause detected.

Neonatal Seizures Vs Jitteriness		
Characteristic	Seizures	Jitteriness
Can External Stimulus Initiate?	No	Yes
Movements	Irregular & Jerky	Symmetrical Fine Tremors
Associated ⇧ In Heart Rate	Yes	No
Associated Breath Holding	+/ -	No
Movements Be Easily Stopped?	No. Self-Limited Movement	Yes. Gently Holding Limb

Investigations

- CBC, Blood culture.
- Blood glucose, Serum Ca, Mg.
- TORSCH screening.
- Serum & Urine amino acids
- Cranial U/S: is an excellent tool for detection of IV Hge & parenchymal Hge.
- MRI: valuable in detection of subarachnoid/subdural Hge & malformations.
- EEG: may detect burst–suppression pattern, Low voltage, invariant pattern.
- CSF: should be done in all cases as seizures may be the 1st sign of meningitis, it should not be omitted even if another etiology such as hypoglycemia is present because meningitis can often coexist but may be withheld temporarily if severe cardiopulmonary compromise is present in infants é severe birth asphyxia.






CSF collected from the thecal sac that surrounds the spinal cord

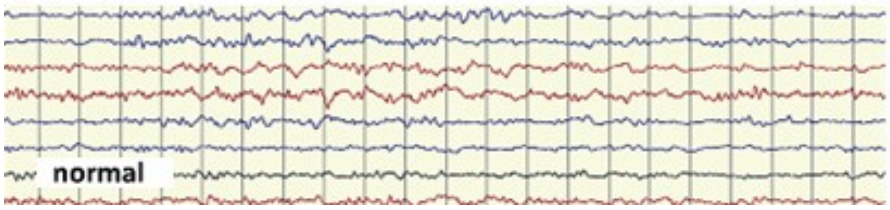
Lumbar puncture technique

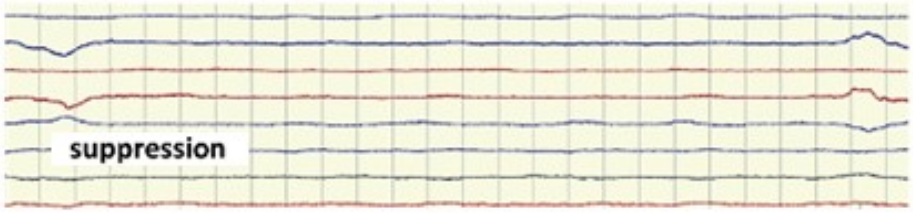
Under complete aseptic conditions direct the LP needle, caliber F.G. 21 or 22, between L_3 & L_4 vertebrae & towards the umbilicus, the upper border of iliac crest is at the level of 5th vertebrae, so go up to touch the spine of 4th vertebrae, then introduce the needle into the disc space between the 4th & 3rd vertebrae, (if CSF was found to come out under high pressure stop the procedure to avoid conning of cerebellum), collect CSF, in 3 test tubes, 10 drops each, one for cells, other for biochemistry & the last for culture/sensitivity. Take blood sample for BS.

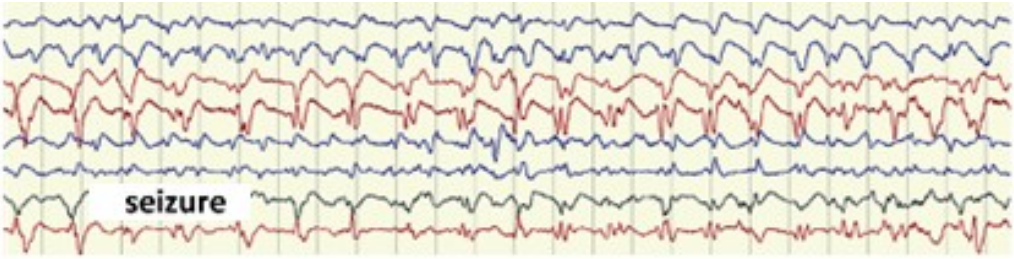
C.S.F. types of meningitis

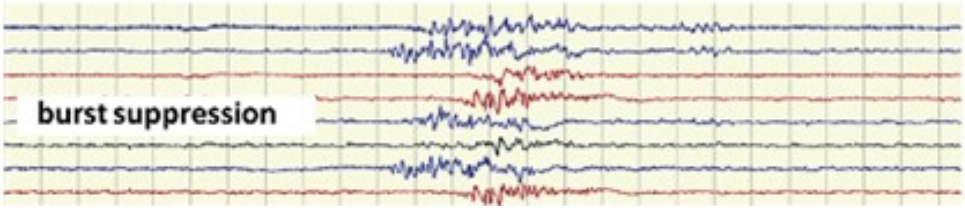
	Cells/mm	Protein	Sugar	Chloride	Bacteriology
Normal value	0-5 mononuclear	20-40mg%	40-80mg%	690-720mg%	
Purulent meningitis	≥500-3000 mainly polymorphs.	⇧⇧	<1/2 B. Sugar	Normal	Isolation by smear & culture
TB Meningitis	⇧50-500 mainly lymphocytes.	⇧	⇩	⇩	smear&culture & guina pig inoculat
Aseptic viral meningitis	⇧ 50-500 mainly lymphocyte	⇧ or Normal	Normal	Normal	Virological Studies

Examples of normal & abnormal NN ECG









Management

Hypothermia has recently become a standard Rx for HIE & has been shown to improve outcome, the Therapeutic hypothermia should be considered for infants born at term or near term é evolving moderate to severe HIE (é its protocol & follow up). Induced hypothermia (33.5-34.5^{o}C) implemented within 6 hrs of birth in term infants at highest risk for brain injury & é further Rx in NICU is associated é significantly fewer deaths & less neurodevelopmental disability, both cooling methods (systemic versus selective head cooling) were shown to be effective, started within 6 hrs from birth, continue for 72 hrs after birth & rewarm over at least 4 hrs, carefully monitor for known adverse effects of cooling as thrombocytopenia & hypotension.

Anticonvulsant: treat é anticonvulsant if the seizure is prolonged (> 3 min), frequent, or associated é cardiopulmonary disturbance.

Phenobarbitone: loading dose 20 mg/ Kg I.V. over 10-15 min.(high dose may cause apnea/respiratory depression), maintenance dose 2.5-5 mg/Kg once daily, 70% of seizures will abate é Phenobarbitone only.

Phenytoin: adding 2nd drug as Phenytoin may needed. Loading dose 15-20 mg/Kg I.V. é a maximum infusion of ½ mg/Kg/min. (high dose may cause hypotension & cardiac arrhyt-

hmia), maintenance 4-8 mg/Kg/day.

Diazepam: is generally avoided in NN period due to its short duration of action, narrow therapeutic index & because of the prese nce of sodium benzoate as a preservative.

Midazolam, Paraldehyde: are rarely used.

Correct metabolic disturbances

Hypoglycemia, glucose 10% 2 ml/Kg I.V. (0.2 gm/Kg) as a bolus followed by continu-ous infusion at up to 8 mg/Kg/ min.

Hypocalcaemia, if serum Ca^+ < 7 mg /dl, Ca^+ gluconate 10%, 100 mg/Kg slowly over 1-3 min & strictly I.V. to avoid extravasation & severe tissue necrosis, monitor HR as it cause bradycardia & cardiac arrest, followed by maintenance 500 mg/Kg/ day I.V. or P.O. can be added to feedings 1-2 ml/feed é a maximum 12 ml/day.

Hypomagnesaemia, if serum Mg < 1.5 mg/dl, $MgSO_2$ 10%, 25mg/Kg IV or IM & mainten-ance 25 mg/Kg/day ÷ 4 IV or IM an over doses cause bradycardia & heart block.

Hyponatraemia, Na^+ deficit is calculated as; 0.7 X BW X (desired Na^+- actual Na^+), replace half the deficit over 12 hrs.

Pyridoxine dependency, 10 mg IV & repeat every 10 min till control, maintenance 5 mg/ Kg/day orally.

Prognosis: -Normal outcome 56%. -Neurological sequel 30-40%. -Death 15-25%. - Chronic seizures disorders 15-20%.

Outcome depends on:

⦿Level of maturity. ⦿Etiology. ⦿Neurological examination. ⦿EEG/Imaging studies.

Good Prognosis

⊙ Uncomplicated hypoglycemia. ⊙Narcotic withdrawal. ⊙Subarachnoid Hge.

Poor prognosis

✲Low Apgar score ≤ 6 at 5 minutes. ✲Onset of seizures within 24 hrs of life. ✲Presence of myoclonic attacks. ✲Abnormal EEG. ✲≥ 3 days uncontrolled fits.

CEREBRAL PALSY

A disorder of movement & posture, caused by a non progressive injury to the immature brain, during fetal life, infancy, or childhood up to 5 yrs of age (the period of brain development). It is one of the most common disabling conditions affecting children, damage is to the cerebral cortex, cerebellum, or spinal cord, it is non-curable & life long condition, may be cong or acquired & 50% of cases are of unknown etiology.

Incidence: 1.5-2.5/1000 live births.

Etiology

Prenatal causes 70% •Developmental malformation, the brain fails to develop correctly •Genetic or chromosomal cause •Parent`s age as related to sperm & egg viability •Maternal history of MR, seizures, hyperthyroidism, two or more prior fetal death or sibling ē motor deficits •Premature placental separation •IUGR •Prematurity •Multi ple pregnancy •APHge •Rh incompatibility •Polyhydramnios •Drug, alcohol, or toxin exposure •IUI (TORCH) •Infection in the birth canal or uterus (chorionitis).

Natal etiology of CP 5-10% •Prolonged labor •Birth asphyxia (HIE) •Instrumental delivery • Infection in the birth canal.

Post natal causes: •Encephalitis •Meningitis •NN convulsions (hypoglycemia, hypocalcemia, hypomagnesemia) •Kernicterus •Trauma (accidental) •IEM.

Types

According to the number of limbs involved

Quadriplegia: equal involvement of all 4 limbs, trunk, neck & head often affected, pt often has problems controlling the mouth & tongue muscles, unable to walk, skeletal

deformities, bladder, bowel problems.

Diplegia 50%: UMNL of all 4 limbs, legs more severely affected than arms, may be symmetric or asymmetric. Difficulty in moving lower part of the body due to stiffness of the legs, difficulty straightening fully at the hips, difficulty ē balance when standing or walking, pt often has a tilted head & shoulders back in an attempt to achieve an upright position creating an exaggerated curve in the lower back & when walking they move the trunk excessively to compensate for stiffness of the legs.

Hemiplegia 30%: UMNL affect one side of the body, the arm usually more affected than leg, neglect of the affected side, problem reaching & grasping ē affected hand, lack of feeling on the affected side of the body, the pt usually has a bent arm (flexed) & all hand is fisted. The leg is stiffened & walking on tip toes & affected side can be smaller due to tight muscles & lack of growth.

Triplegia: three limbs involved, usually both arms & one leg.

Monoplegia: one limb affected (usually arm), rare & usually occur after meningitis.

According to movement disorder

Spastic CP 80%: UMNL, result from damage to the motor areas of cerebrum, most people ē spastic diplegia eventually walk, hip dislocation & crossed eyes are common, ⇧ muscle tone primarily of flexors & internal rotators, ẃ might lead to permanent contractures & bone deformity, the muscles are tight, movements are stiff especially the legs, arms & back.

Athetoid CP (dyskinetic) 10%: occur when there is damage to the basal ganglia (masses of gray matter composed of neurons located deep within the cerebral hemisphere of the brain), results in overflow of motor impulses to the muscles, slow, writhing movements that are uncoordinated & involuntarily, affect movements of the entire body, involves

slow uncontrolled body movements & low muscle tone & hard to sit straight & walk.

Ataxic CP 10% condition that occurs when there is damage to the cerebellum (ẃ normally regulates balance & muscle coordination), disturbed sense of balance & depth perception, poor muscle tone, staggering walk &unsteady hands, diagnosed when the child attempt to walk, muscles show abnormal degrees of hypo tonicity, lack of balance & coordination necessary for proper arm & leg movement causing wide based gait to be exhibited, child show difficulty ē performing basic motor skills & pattern that include locomotors movements (as running, jumping, skipping).

Mixed CP: include both the ataxic & athetoid CP.

According to the area of brain damage

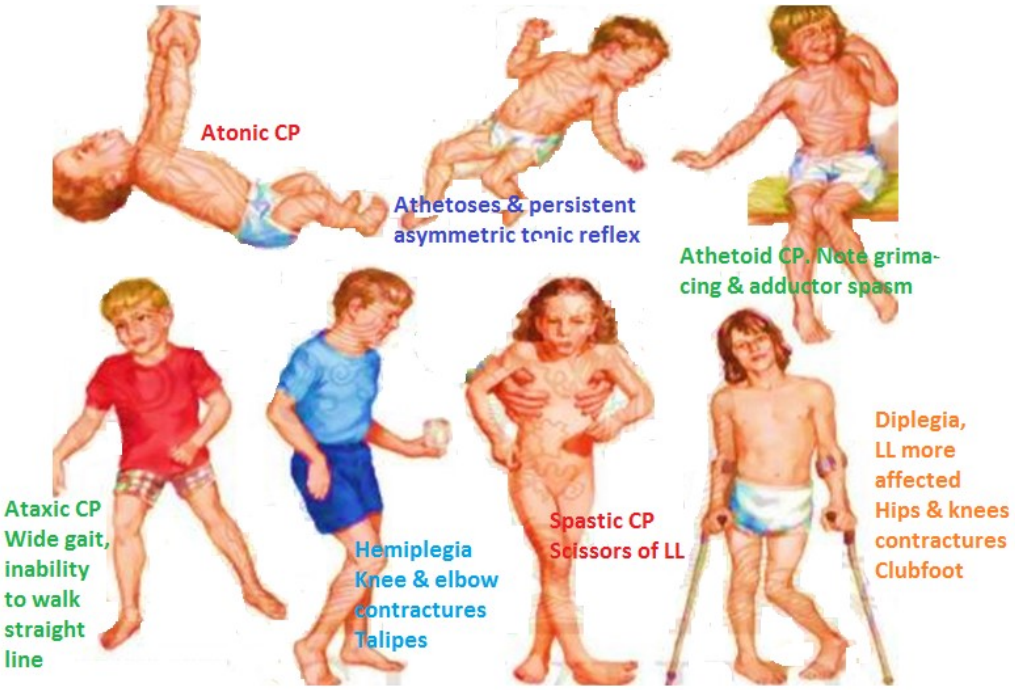
Pyramidal: motor area of cerebral cortex.

Extra pyramidal: basal ganglia and cerebellum.

Mixed: include pyramidal & extrapyramidal involvement.

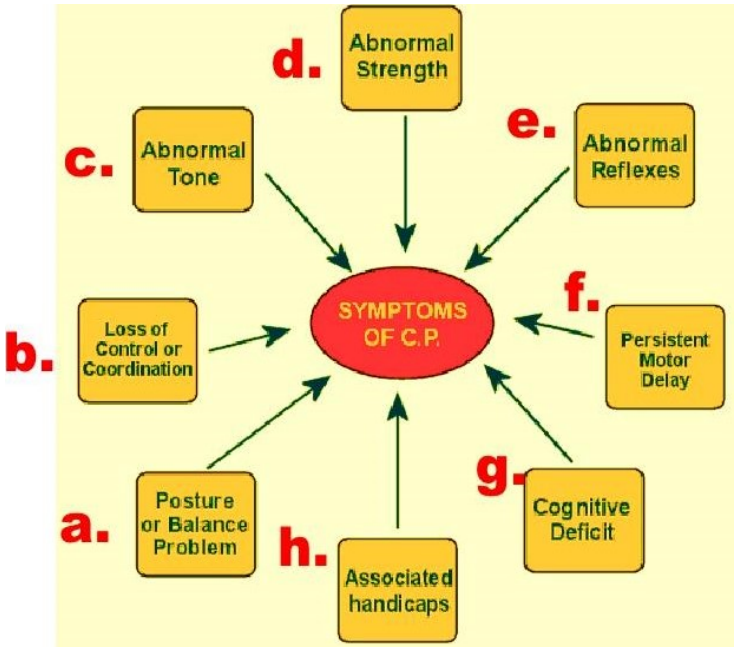
According to the degree of severity

Either mild , moderate, or severe.



Clinical picture: Early signs: stiff or floppy posture & poor head control, lethargy or irritability. Weak suck, feeding difficulty, abnormal or prolonged primitive reflexes. Delayed milestone & behavi-oural symptoms; poor ability to concentrate, unusual tenseness.

Associated problems: MR (60)%. Seizures / Epilepsy (33%). Hearing & visual problems (10%). Sensory integration problems, communication disorder. Skeletal deformities/ Dental problems. Bladder & bowel control problems.



Management

Baby ē CP have normal life span as it is not progressive, the effect of CP may change over time, some may improve, some may get worse, but no cure.

Aim at managing symp-toms. #Team approach.

#Physiotherapy, speech therapy.

#Recreational & occupational therapy focuses on helping the child learn skills for daily living, feeding, dressing.

#Adaptive equipment. Orthopaedic & dental care (for scoliosis, & dental problems)

#Medications, include: Glycopyrolate (to reduce drooling & salivation). Diazepam, Baclo-ofen, Dantrolene sodium, Tazanidine (for treatment of stiff, rigid or spastic muscles).

Chapter III

RESPIRATORY PROBLEMS

- Transient Tachypnea Neonate
- Respiratory Distress Syndrome
- Mechanical Ventilation
- Meconium Aspiration Syndrome
- Persistent Pulmonary Hypertension
- Neonatal Pneumonia
- Pneumothorax
- Congenital Diaphragmatic Hernia
- Stridor
- Tracheo Oesophageal Fistula
- Epiglottitis
- Asthmatic / Wheezy Bronchitis
- Whooping Cough

Diagnosis of respiratory problems

Presence of at least 2 of the following features in NN are essential:-

① Tachypnea (RR >60/Min).

② Retraction (intercostal &/or subcostal).

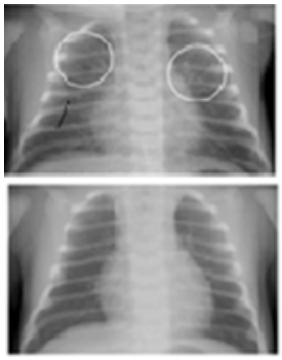
③ Expiratory grunting.

Etiology

In Term Baby: ▲Transient tachypnea ▲Aspiration pneumonia ▲Pneumothorax. ▲Pulmonary hypoplasia ▲Pleural effusion ▲Airway problem.

In PT Baby: ∗HMD ∗Pneumonia ∗Air leak syndrome ∗Pulmonary hypoplasia. ∗Bronchopulmonary dysplasia ∗Laryngomalacia.

TRANSIENT TACHYPNEA NEONATE



Streaky, peihilar linear densieies (white circle) Indistinctness of blood vessels & fluid in the minor fissure (black arrow)

3 day`s after - of the same baby show complete clearing of the fluid & normal CXR

About 1% newborns develop TTN, known as wet lung disease. Pulmonary edema from delayed resorption & clearance of fetal alveolar fluid (before birth, fetus lungs are filled é fluid, while inside the mother fetus does not use lungs to breath & all his O_2 requirements comes from blood vessels of placenta). It is mild RD immediately after birth ŵ usually eases after few days é Rx. Seen in NN whom mother sedated or following CS, precipitous labor due to lack of gradual compression & squeezing of chest wall ŵ takes place during normal delivery, seen also in baby of diabetic or asthmatic mother, SGA babies, excess maternal fluid administration, delayed cord clamping. More in sex male. This is diagnosis

of exclusion when no other cause found for infant tachypnea. May not diagnosed until symptoms subside, this may not until 3 days after birth.

Clinical Picture

- Tachypnea, RR > 60/min.
- Intercostal, subcostal recession & working alanasi.
- Cyanosis: the skin around the mouth & nose has a bluish tinge.

Investigations

- CXR.
- Blood C/S: looking for bacteria/other microorganism.
- ABGs.
- CBC.

Management

- NPO.
- O_2 by mask, tent, or CPAP.
- Maintenance of body temperature.
- Antibiotics because it is difficult to distinguish it from early pneumonia & until test results come back.
- Daily requirements of IVFs.

RESPIRATORY DISTRESS SYNDROME

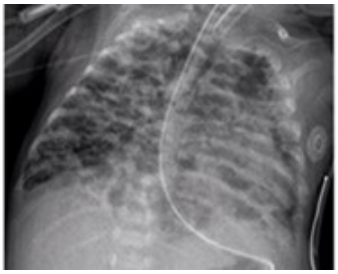
known as HMD, or SDS. In 1920_s Kurt Von Neergward, Swiss physiologist, postulated the existence of a substance in the lungs that reduces surface tension allowing the lungs to open. In 1950_s John Clements, USA pulmonary physiologist, showed that this substance was surfactant. Finally in 1959 Mary Avery & Jere Mead, both working at Harvard at the time, demonstrated that surfactant was lacking in the lungs of PT babies, ŵ was the base cause of resp failure in some of these infants. Further study found that the deficiency of surfactant was a consequence of either insufficient production by the immature lungs or a genetic mutation in one of the surfactant proteins (rare genetic form not associated é

PT birth & occurs in FT babies). It was found also that the RDS is more frequent in infants of diabetic mothers. Surfactant is complex lipoprotein composed of 6 phospholipids & 4 apoproteins usually produced by type II pneumocytes by the age of 34 wk gestation.

Epidemiology

Major cause of morbidity & mortality in preterm infants. Incidence & severity of RDS are related inversely to gestational age. At 26-28 wk gestation the incidence is 50%. & at 30-31 wks gestation it is < 30%. According to the weight the overall incidence is as follow:-

- 501-750 grams: 71%.
- 751-1000 grams: 54%.
- 1001-1250 grams: 36%.
- 1251-1500 grams: 22%



Diffuse ground glass, reticulogranular, develop 6 - 12 hours after birth, similar to congenital pneumonia

Clinical Picture

Features observed	0	1	2
Chest movement	Synchronized Resp.	Lag of Resp.	See - Saw Resp.
Intercostal retraction	Non	Just visible	Marked
Xiphoid retraction	Non	Gust visible	Marked
Nares flaring	Non	Minimal	Marked
Expiratory grunt	Non	Audible by stethoscope	Audible by unaided ear

✳Tachypnea, diminished air entry, expiratory grunting, intercostal/subcostal recession, nasal flaring, ⇩ breathing sounds, expiratory rhonchi, fine rales, cyanosis, apneic attacks ŵ is normal finding in PT if <10 sec, but in RDS it is prolonged, associated é fatigue & hypoxia. Apnea may be seen also é hypocalcaemia, hypoglycemia, IC Hge, HIE, meningitis, convulsions, pneumonia & cardiac diseases.

✳Cyanosis is either central in the buccal mucous membrane, or peripheral seen in the nail beds, is late sign of hypoxia in NN due to presence of Hb F ŵ possess marked affinity to O_2 causing left shift of O_2 Hb dissociation curve, cyanosis only become visible when PO_2 is < 40 mmHg, ē formation of 5 gm/dL of reduced Hb. Cyanosis of cardiac origin will not respond to O_2 therapy. Other causes of cyanosis include; polycythemia, sepsis & CNS depression.

✳Combined metabolic + respiratory acidosis ŵ add in prevention of surfactant production by lungs & leads to depression of myocardium activity & diaphragmatic contraction & ⇧ pulmonary vascular resistance.

✳Persistence of PDA will add to the problem as blood pass from aorta to pulmonary artery (left to right shunt) causing pulmonary hypertension, accentuation of pulmonary 2nd sound over the pulmonary area & ⇧ of pulmonary vascular resistance.

✳Oliguria & constipation, small amount urine < 1 ml/Kg/hr & may not pass meconium until 3rd or 4th day of life.

Differential Diagnosis

- Air leak: interstitial emphysema, pneumomediastinum, Pneumopericardium.
- Pneumonia (often group B beta hemolytic streptococci).
- Congenital anomalies: diaphragmatic hernia, lobar emphysema, bronchogenic cyst.
- TTN (usually after CS).

- Metabolic problem as; hypothermia, or hypoglycemia.
- Hematological problem as; anemia, or polycythemia.

Complications

- BPD, retinopathy from prolonged use of O_2 or high O_2 conc.
- Persistent PDA.
- Persistent pulmonary hypertension.
- Pulmonary Hge (⇧ in very preemies, especially following surfactant therapy).
- NEC.
- Hospital acquired infection.

Investigations

ABG: mixed acidosis (⇩ PaO_2, ⇧ PCo_2, ⇩ PH ,⇩ HCO_3). For every 10 mmHg ⇧ in PCo_2 there is associated 50% ⇧ in cerebral blood flow & high risk for IC Hge.

CXR: diffuse ground glass/reticulogranular appearance always develop 6-12 hrs after birth, similar to that of congenital pneumonia.

ECHO cardiography: for detection of PDA, the direction & degree of shunting, pulmonary hypertension & other structural heart defect.

CBC Blood C/S Glucose & Electrolytes Renal & LFTs.

Management

Incubator care: Place infant in lateral or prone position rather than supine, to provide clear airway, repeated suctioning of pharynx not required & may cause apnea & hypoxia

Control body temperature to be maintain in NTE range, core temp should be maintained at 37^0C to minimize O_2 consumption & acidosis. Peripheral temp at 36^0C or more.

Monitoring: O_2 saturation in blood (SpO_2) using pulse oximeter to be kept at 88-92%, also monitoring HR, RR & apnea.

Oxygen supply: warm & humidified O_2 to keep SpO_2 >88% may prevent use of ventilator in moderate PT babies. Excess or prolonged use of O_2 cause RLF, BPD, IC Hge. Excess of Co_2 in blood (hypercapnia) cause also IC Hge as cerebral blood flow ⇧ 50% for every 10 mmHg ⇧ in $PaCo_2$, in such case redness of skin due to vasodilatation may be seen, The O_2 supply is either through; Nasal catheter 1-6 liters/min (40-50%), or O_2 mask 6 liters/ min (40-50%), or O_2 Box 10 L/min (100%).

Surfactant administration: for babies <30 weeks gestation, a liquid surfactant instelated into the trachea to the lungs to help their maturation. Alveofact amp 50 mg/1.2 ml for 4 consequent days while the baby on ventilator & ⇧ the PIP for 30 sec after instillation, surfactant is important in lowering surface tension of the alveoli, prevention alveolar collapse during expiration, it is started to be produced at 34 weeks gestation.

Sedation: usually recommended to infant to avoid discomfort.

NPO & Nasogastric suction of gastric content.

IVFs: Day 1: G 10%, 60-80 ml/Kg/day. Day 2-5: G 5% + Saline (ratio 4/1), daily ⇧ of 20 ml/ Kg up to maximum of 150-180 ml/Kg/day by end of 1st week + Ca^+ gluconate 10% amp 10 ml, 1 ml/Kg/day + Kcl 20% amp 5 ml, 1 ml/Kg/day to be added after passing urine (urine output equal to 1 ml/Kg/hr) + Daily requirement of Vit & trace elements, 5 ml amp of pediatric formula, ½-1 amp/day. (Vit A 2300 U, Vit D 400 U, Vit E 7 U, Vit K 200 mcg, Vit C 80 mg, Vit B_1 1.2 mg, Vit B_2 1.4 mg, Vit B_6 1 mg, Vit B_{12} 1 mcg, Biotin 20 mcg, folic acid 14 mcg, niacin 17 mg, pantothenic acid 5 mg) + Additional allowances of IVFs, 20 ml/Kg/day if baby under phototherapy to compensate for heat loss.

Antibiotics: Crystalline Penicillin 1000.000 U/vial, 100.000 U/Kg/day ÷ 4, I.V. or Ampicillin 1000 mg/vial, 50-100 mg/Kg/day ÷ 4, I.V., for 5-7 days, or Cefotaxime (3rd generation Cephalosporin.) amp 250mg, 50 mg/Kg ÷2, IV. or IM, for 5-7 days (not recommended as it

enhance fungal infection) + Aminoglycosides amp 20 mg IM, or IV. 3-5 mg/Kg÷2 X 3D.

Feeding: after stabilization of condition & respiratory status, initial small volume of gastric feeds 1-2 ml/4 hrs (preferably breast milk) via gastric tube or milk pump to initially stimulate gut development & progress carefully according to the clinical situation.

NB: the RDS typically worsens over the first 48-72 hrs, then gradually improves é Rx.

Ventilator care

Indications

- Cyanosis that persist in spite of maximum O_2 therapy.
- Severe recurrent apnea.
- Respiratory failure: PCo_2 >70 & pH is <7.2

Normal blood gases

- PH: 7.3 -7.4
- PO_2 : 80 -110
- PCO_2 : 35-45
- HCO_3 : 24-26
- BE: - 4 : + 4.

Stabilization of ventilator settings

- O_2: 40-60%.
- RR: 60/min
- PIP: 15 cmH_2O, gradual⇧ 5 by 5, maximum to 25 cmH_2O
- PEEP: 2 cmH_2O, gradual ⇧ 2 by 2, maximum to 10 cmH_2O
- Ti: 0.3 sec.

Technique

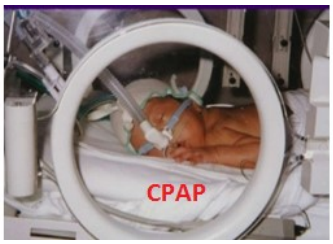
- Frequent nasogastric suction.
- Ensure synchronacy between baby respiration & ventilator cycles.
- Do ABGs/6 hrs. Pulse oximeter to keep O_2 conc > 88% (⇩ 15% from given reading to get actual reading of PaO_2 mmHg in blood).
- With severe hypotension, give Dopamine if perfusion is permanently poor, shock, or renal failure. Dose = BW X 3 = number in mg of dopamine (vial 250 mg) to be collect by insulin syringe, then add 50 ml G 5% & give 2 ml/hr ŵ is equal to 2 ug/Kg/minute, the dose can doubled up to 5ug/Kg/minute, this ⇧blood supply to internal organs, higher dose 6-

10 ug/kg/min have the same previous effect in addition to +ve inotropic & chronotropic effect on heart. Much higher dose from 11-15ug/Kg/minute, cause the same previous effect + peripheral vasoconstriction.

- In case of severe cases é PaO_2 < 40 mmHg on O_2 70%, we may use pulmonary vasodilators as; Tolazine 1 mg/Kg IV, can be repeated/hr, putting in mind its main side effect as hypotension, internal Hge from his histamine like action.
- Aminophylline amp 250mg, 5 mg/Kg/day ÷ 3 IV infusion, have the same side effects.
- $NaHco_3$: amp 8.4% 25 ml contain 20 meq, used in metabolic acidosis (when base deficit is > -5) 1-2 meq/Kg/day =1 ml/Kg/day, calculated according to the base deficit (Base deficit X BW X 0.4 =meq of $NaHco_3$ to be given).
- Keep Hb >13 gm/dL: packed RBCs (Normal Hb -Baby Hb X BW X 3.5) or whole blood transfusion (Normal Hct - Baby Hct) ÷ Donner Hct) X BV)- Blood volume = BW X 80 ml. Keep serum proteins >2 gm/dl: 10% albumin, or fresh plasma 10-20 ml/Kg/12 hrs.
- Vit E: for baby <1500 gm, 25-100 U/day for 7 days.
- Fundoscopy: before discharge for RLF.

Weaning from ventilator

Started when clinical condition & ABGs are stable for 12 hrs, gradual lowering of FiO_2, PIP & RR, separately in the same order of increasing it & putting the baby on FiO_2 0.4 (40%) using CPAP, follow up by ABG & CXR 6 hourly for 24 hrs. One shot Forticortin IV 0.5 mg/Kg may be given é the removal of ETT. Then put O_2 by mask or head-box, reassess clinically & ABG.



CPAP: prevent alveolar collapse at end expiration. Indications: 🕊 FiO_2 >0.4 🕊 PaO_2 < 50 mmHg. 🕊 Pressure: 4-6 cm H_2O

CMV: Indications: 🕊 CPAP >8 cm H_2O 🕊 FiO_2 > 0.6 🕊 PaO_2 < 50 mmHg 🕊 $PaCO_2$ >60 mmHg 🕊 PH< 7.25 🕊 Frequent apnea 🕊 PH < 7.25 🕊 Frequent apnea.

Modes of Ventilators

	TARGET	NAME	Indication
Control / Assist Control Mode	Volume	Volume Control	ARF
	Pressure	Pressure Control	ARDS
	Volume Target Pressure Reg Flow	PRVC / VV +	ARF with dyssynchrony
SIMV Mode	Volume / Spont	SIMV – Vol Cont	ARF
	Pressure / Spont	SIMV – Press Cont	infrequent
	Vol or Pres / Support	VC or PC with PS	ARF
Support Mode	Volume	Volume Support	Wean
	Pressure	Pressure Support	Wean

Complications of mechanical ventilation

- Barotrauma: from ETT, high Tv, or high PEEP: lip damage, mouth ulceration, laryngeal edema, hypotension, bradycardia, PVCs, aspiration pneumonia, rupture alveoli, pneumothorax, pneumomediastinum, pulmonary interstitial emphysema.
- O_2 toxicity: from high FiO_2, formation of singlet O_2 (free radicles) ŵ is toxic to all tissues, causing RDS, BPD, retinopathy.
- ⇩ COP: from ⇧ PEEP, RR, or Tv, causing ⇧ of the intrathoracic pressure & ⇩ of the venous return to heart, renal impairment from ⇩ of venous return, ⇧ fluid retention, impairment of metabolism of certain drugs as the acid base balance impaired.
- Diaphragm paralysis: become lazy from having all the work done for him.

•Ventilation associated pneumonia: from prolonged intubation, long stay on ventilator, mainly G-ve bacteria, pseudomonas aerogenosa (aerobic, rod-shaped) is the major cause of nosocomial infection.

•⇧ of ICP: due to ⇩ of venous return from head & may cause ICHge .

•Respiratory alkalosis •Gastric distension.

Prevention of RDS

✳intubation of infant born ≤ 30 wks gestation.

✳Prophylactic natural surfactant therapy is administrated through the ETT as soon as the infant is stable after intubation. Prophylactic surfactant therapy is not recommended for infant >30 wks gestation. Do not delay surfactant for CXR. No CXR is necessary to confirm proper tube placement.

✳Antenatal steroids should be given to any pregnant women at 24-34 weeks of gestation é intact membranes at high risk for PT delivery.

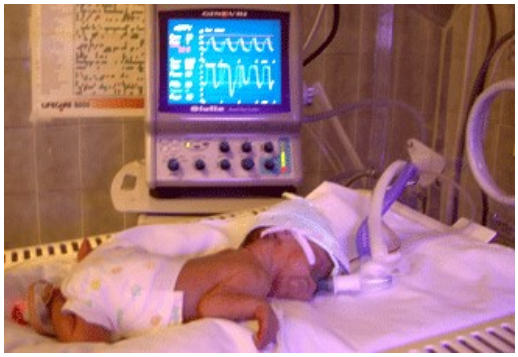
✳Delaying premature birth. Tocolytics may delay delivery by 48 hrs & therefore enable time for antenatal corticosteroids to be given. Same protocol for the baby, dose of 0.2 mg/Kg twice daily IM for 2 successive days it induce pharmacological closure of PDA, but in newborn may cause internal Hge & ⇧ in serum creatinine. After administration of surfactant & if the infant is active & exhibit spontaneous respiratory effort, extubation & stabilization on CPAP rather than continued intubation & mechanical ventilation.

✳Good control of maternal diabetes & avoidance of hypothermia in the neonate are also important prophylactic measures for prevention of RDS.

Prognosis

VLBW < 501 gms survival rate is 10% & 100% risk of BPD & very high risk of retinopathy, BW 1001-1500 grams survival rate is ~ 96% & few develop BPD & ROP.

MECHANICAL VENTILATION



Excessive airway pressure & tidal volume can lead to more harm & lung injury, "ventilator induced lung injury", & contribute to ⇧ mortality.

"Ventilators are the medical worst invention of the 20th century". Most PT infants born before 30 wks gestation receive some form of respiratory support. Although mechanical ventilation is frequently a lifesaving therapy, its use ⇧ the risk of lung injury, particularly in PT infants in whom the incidence of BPD remains high. If the lungs are not working at all well &/or the baby cannot manage to do all their breathing, they will need more help. This can be given by a mechanical ventilator ẃ can help the baby`s efforts to breath, or if necessary take over the breathing function completely. The ventilator is connected to a supply of air & $O_2$ & these are mixed in the ventilator to give the right levels for the baby`s need. To connect the baby to the ventilator, a tube is inserted through the mouth into the trachea (ETT), the tube is kept in place by attaching it to a bonnet or to tape ẃ is secured to the baby`s upper lip, occasionally the tube well need changing if it becomes blocked é mucous or dislodged. Whilst the baby is on a ventilator mucous collect in the airways & suction catheter is used to clear the airway to get rid of the mucous every few hours making the baby more comfortable. A +ve pressure ventilation can delivered by 2 kinds of machines using 2 different principles, including; the conventional mechanical ventilation & the advanced high frequency ventilation, the first is more familiar mode ẃ

is used in most of the NICU setups, but in certain diseases & in worsening respiratory distress diseases, HFV is now becoming an increasingly popular mode due to its lesser side effects & clinical advantage é efficient ventilator expertise & vigorous monitoring. Many PT or sick babies develop breathing difficulties, these difficulties occur because the lungs are not fully developed & the baby`s brain is not yet mature enough to control effective, regular breathing so they require some active help é breathing. CPAP is less hazardous, it is an external way to give NN air/$O_2$ éout placing an ETT, only using a mask fitted over the NN`s nose or set of prongs placed into the nasal passages, the tube is attached to a machine & humidifier.

Indications of mechanical ventilation

Lung immaturity: some EPT babies have not had sufficient time for their lungs to mature & this could mean that they struggle to breathe.

Apneic spells: this is when baby`s breathing pattern is sporadic (long pause between breathing >10 sec. unresponsive to medical Rx.

Lack of surfactant: ẃ is naturally produced substance in the body, it is important in reducing surface tension of alveoli & prevention of alveolar collapse during expiration, started to be produce at 34th wk of gestation & it`s absence result RDS.

Pneumonia: congenital pneumonia can be caused by numerous microorganisms, but the most commonly cultured infectious agents are group B-beta hemolytic streptococci & Escherichia coli among infants é early onset disease (first 3-5 days of life).

Meconium aspiration: not occur before 34th wk of gestation (time for development of su-

cking & swallowing reflex), may cause birth asphyxia, inhaled meconium can cause airway obstruction, air trapping & over distension of lungs, pneumothorax, chemical pneumonitis, 2ry bacterial infection, atelectasis & ⇧ in pulm vascular resistance.

Persistent pulmonary hypertension in NN: disorder characterized by ⇧ pulmonary vascular resistance resulting in shunting of blood away from the pulmonary vascular bed through the fetal channels (ductus arteriosus & foramen ovale), the resultant venous admixture cause profound hypoxemia, ⇩ tissue O_2 delivery, metabolic acidosis & further pulmonary vasoconstriction.

Broncho pulmonary dysplasia: the etiology & pathogenesis are clearly multifactorial, include; the effect of PPV (Baro trauma), over distension of lung by large Tv ventilation (volu-trauma), repetitive opening & closing of the lung units (Atelect trauma) & the effects of oxidant stress & inflammation (Bio trauma). Rx of BPD include: mechanical ventilation, bronchodilator, diuretics & steroids.

Clinical Criteria for mechanical ventilation

RD: severe retraction (intercostal, subcostal, suprasternal) & nasal flaring.

Central cyanosis: cyanosis of mucosa on $FiO_2 > 0.4$-0.7 (40 -70%).

Refractory apnea: unresponsive to medical Rx.

Laboratory Criteria for mechanical ventilation

Severe hypercapnia: $PaCO_2 > 55$-60 mmHg ē pH < 7.2-7.25. Severe hypoxemia: $PaO_2 <$ 40–50 mmHg or $SpO_2 < 85\%$ on $FiO_2 > 0.4$-0.7(40-70%)

Contraindications of mechanical ventilation

- < 32 wk gestational age or BW <400 gm.
- Cong. anomalies incompatible é survival.
- Severe prolonged HIE.

Definitions

FiO_2: amount of O_2 delivered to the infant. Natural air contain 20.9% O_2 ẃ is equivalent to FiO_2 0.21, the O_2 enriched air has a high $FiO_2 > 0.21$ up to 1.00 ẃ mean 100% O_2.

RR: number of breaths/min, ventilator is to deliver.

Tv: amount of air move into or out the lungs during single respiratory cycle, delivered é each ventilator breath.

PEEP: how much air pressure still in the alveoli at the end of expiration, it prevent the alveoli from collapsing during exhalation.

PIP: is the pressure exerted against the infant`s airway during the breath, PIP affect also MAP ẃ is the average pressure exerted in the airway & begins from the begining of inspiration until the beginning of next inspiration.

I/E: in normal spontaneously breathing NN the I/E is about 1/3 to 1/4 e.g. if RR is 30/min, so each respiratory cycle (insp + exp) will take 2 sec, by mean that "T_I", is 1/3 -1/4 of the respiratory cycle equal to 0.7-0.5 sec, the ⇧ of T_I will ⇧ the oxygenation of Hb (PaO_2).

Flow Rate: the minimum flow is at least 2 times an infant`s minute ventilation (Tv X RR), approximately 0.2–1 L/min, but the usual operating range for mechanical ventilation is usually 4-6 L/min, much of this flow not delivered to the infant but rather is used to drive ventilator.

CPAP: the commonest form of help given to support baby`s breathing, it is non-invasive type of ventilation, it is spontaneous breathing é +ve air way pressure, indicated if PaO_2 <50-60 mmHg in FiO_2 >0.6 (60%), it is a technique of assisting breathing by maintaining air pressure in the lung &air passages constant &above atmospheric pressure throughout the breathing, ẃ stops the lungs collapsing when the baby breaths out. The air/ O_2 is given through small soft tubes placed just inside the nose or by a mask over the nose, the

baby does all his own breathing but this is made easier by having the lungs kept partially expanded by the CPAP. It is of benefit to infants who have lungs prone to collapse, such as the stiff lungs é RDS, usually we start é pressure 4-6 cmH_2O & ⇧ 2 cmH_2O every 15 min to a maximum of 10 cmH_2O, & to maintain O_2 saturation (Spo_2) 92-96%. Most babies cope é the nasal tube very well although sometimes the nose can get a bit sore, also get slightly swollen tummy because the machine blows into stomach as well as the lungs.

CPAP settings		
Flow	:	4 - 6 L / min
FiO_2	:	0.4 - 0.6 (40-60%)
Pressure	:	4 -10 cmH_2O

Weaning: commence weaning when; SpO_2 >96%, RR stabilized, grunting ceased, recession reduced & improvement of ABG.

Suggested weaning method: CPAP may be removed after 24-48 hr of apnea free interval ,at first ⇩O_2 gradually in steps of 5% by 5% until FiO_2 21-23%, then ⇩ the pressure to a minimum of 4 cmH_2O in a steps of 1 by 1 every 2-4 hr, é checking of ABG.

Bi-level CPAP

If the baby cannot quite manage on CPAP alone, they can sometimes be helped by being given a small amount of extra pressure (a breath) through the prongs several times a minute. This works well so long as the baby can do most of the breathing & the lungs are working moderately well.

Hazards of CPAP; Generally associate é using high pressure:-

- ⇩ Of COP due to ⇧ of intrathoracic pressure & ⇩ of venous return to heart.
- ⇩ Of pulmonary blood flow 2ry to compression of pulmonary vessels.
- ⇩ Of glomerular filtration rate, Na^+ excretion & urine output.
- Air leak syndrome e.g. pneumothorax & pneumomediastinum.

- ⇧ of ICP.
- Nasal obstruction, necrosis or erosion of nasal septum.
- Gastric distension.

Contraindications of CPAP

*Severely apneic baby or NN é poor respiratory efforts.

*Trachioesophageal fistula or untreated air leaks or congenial diaphragmatic hernia.

*Cleft palate & chonal atresia, or baby é persistent pulmonary hypertension.

*Baby who can`t maintain an adequate spontaneous Tv. Baby é alveolar instability.

*⇧ Of ICP.

*CVS instability.

Desired arterial blood gas ranges

Premature		Term
* 7.25-7.35	pH	7.35 -7.45
* 45-59	$PaCo_2$	35-50
* 50-70	PaO_2	60-80
* 80-92	SpO_2	92-97

History of infant ventilators

1970`s Volume controlled ventilators. 1980`s Pressure controlled ventilators. 1990`s Reintroduction of Volume controlled ventilators

Types of Ventilators

*Pressure cycled: allows air to flow into lungs until the preset pressure has been reached, once this pressure is reached, a valve closes & expiration begins. The volume of air delivered varies é changes in lung compliance &/or resistance.

*Volume cycled: allows air to flow into lungs until the preset volume has been reached, the Tv is delivered despite changes in compliance or resistance.

*Time cycled: allow air to flow into lungs until the preset time then expiration begins.

Intermittent Mandatory Ventilation

Intermittent breaths delivered to a set pressure (IMV-PC) or Tv (IMV-VC), at a fixed rate & it is not synchronized to baby. The consequences of asynchrony include:-

•Fighting the ventilator. •Inconsistent Tv delivery. •⇧ work of breathing. •Inefficient gas exchange. •Barotrauma, thoracic air leaks. •Disturbances in cerebral perfusion.

Synchronized Intermittent Mandatory Ventilation (SIMV)

Is the most widely used forms of ventilation in NICU, sensor attached between the ventilator tubing & ETT, it`s either pressure or volume limited, using either pressure or flow sensor. It deliver the preset pressure or volume & rate while allowing the baby to breath spontaneously in between ventilator breaths, set the amount of O_2, pressure or Tv & RR/min, the ventilator gently pushes air/O_2 mix into the lungs, it allows time for the air & CO_2 to come out. Breaths from the ventilator can be set so that they well be triggered by the baby`s own breaths, for example, if the rate is set at 30 bpm, the ventilator will cycle every 2 sec, each time it is supposed to cycle it will look for spontaneous breath & well start or delay the breath if spontaneous effort is detected within the timing window, the baby spontaneous respiration are never interrupted & the baby can breathe spontaneously through the ventilator or circuit between mandatory breath. SIMV is either: pressure or Tv controled, each at a fixed rate. The more advanced types includes:-

SIMV- Pressure Control é Pressure Support (SIMV-PC/PS): in addition to SIMV-PC, any breath baby takes over the set ventilator rate is supported by amount of pressure (usually half of PIP or twice PEEP).

SIMV- Volume Control é Volume Support (SIMV-VC/VS): in addition to SIMV-VC, any breath baby takes over the set ventilator rate is supported by a set amount of volume (usually 50-75% of the Tv).

Assist/Control Ventilation

Baby receives a set rate & volume, at the same time baby may initiate spontaneous breaths as well, by mean that if ventilator set rate is 12 breaths/min, the baby is guaranteed 12 full breaths, but if his spontaneous breathing is 20 bpm he will receive 20 full breaths. The ventilator also assist every breath the baby taking according to the set & continue during apnea.

High Frequency Ventilator

The rationale of HFV is that the provision of tiny gas volumes at a rapid rates, results in much lower alveolar pressure. The high rate of breath delivery maintain open alveoli, there are several types of HFV devices, including:-

- High Frequency Positive Pressure Ventilation HFPPV" ẃ uses RR of 60-100 bpm.
- High Frequency Jet Ventilation "HFJV" ŵ uses RR 100-600 bpm.
- High Frequency Oscillatory Ventilation "HFOV" ŵ causes rates up to 4000/min.

Sedation & paralytics are usually required for the baby to tolerate this sitting & the chest will feel like it is vibrating.

Indications for HFV

- Rescue following failure of conventional ventilation.
- Air leak syndromes; pneumothorax, pulmonary interstitial emphysema.
- To ⇩ Barotrauma when conventional ventilator settings are high.

Experimental studies

- Only six manual inflations of 35-40 ml/Kg given to preterm lambs injuries lungs.
- Significant of lung oedema & transcapillary albumin flux in rats ventilated é high Tv contrast to rats ventilated é low Tv & high pressures.
- Adult human study shows that lower Tv ⇩ mortality.

Ventilator settings

*PIP: 15- 25 cmH_2O, start low at 15 to prevent barotraumas, ⇧ gradually 5 by 5 up to maximum of 25.

*PEEP: 2-6 cmH_2O, start low at 2 & ⇧ gradually 1 by 1 up to a maximum of 6, adjust to maintain acceptable PaO_2 & SpO_2.

*FIO_2: 0.4-1.0 start low at 0.4 (40%), adjust to maintain the target SpO_2, ⇧ 0.1 by 0.1 (10% by 10%). *Tv: 4-8 cc/Kg BW, start low at 4, adjust to maintain target PO_2, ⇧ 1 by 1. *RR: 30-50/min. *Flow: 6-8 l/min. *I/E: 1-1.5 - 1-2. *T_i : 0.3 - 0.4 sec.

Strategies for mechanical ventilation

- To ⇧ PaO_2 or SpO_2: ✓⇧ FiO_2 T_i or PEEP or PIP or change I/E towards 1-2.
- To ⇩ $PaCO_2$: ✓⇧ RR or Tv or PIP.

Always check for:-

- Chest movement, air entry, presence of retraction, hyperinflated chest, or wheeze.
- Level of ETT at lips, visible secretions in ETT, any kinking or disconnection or any warning alarms on ventilator.
- Assess baby`s own respiratory drive; depth & rate.
- Signs of baby fighting the ventilator; air hunger, asynchrony, gross difference bet-ween ventilator & baby`s breathing rate.
- Signs of pain, agitation, or abnormal posturing.
- Abnormal HR, BP, temp & signs of excessive sedation.

Sedation

Helps the patient tolerate the constant irritation of ETT in their mouth, pharynx & trach-ea, éout some form of sedation & analgesia, it is common for pt to “fight” the ventilator. This fighting ⇧ work of breathing & may cause further lung injury, daily interruption are

frequently described as ”sedation vacations” & have been shown to ⇩ time pt stay on mechanical ventilation (Midazolam, Morphine).

Weaning: started when clinical condition & ABGs are stable for 12 hrs, gradual lowering of FiO_2 ,PIP & RR, separately in the same order of increasing it & putting the baby on FiO_2 0.4 (40%) using CPAP, follow up by ABGs & CXR 6 hourly for 24 hrs, one shot of Forti-cortin IV 0.5 mg/Kg may be given é the removal of ETT, then O_2 by mask or head box, reassess clinica- lly & ABGs..

Risks of mechanical ventilation: the next complications are important to put into consideration during dealing é mechanical ventilation.

Barotrauma & complications of the ETT: complications of ETT include; damage to lips, ulceration of mouth/lips, laryngeal edema, hypotension, bradycardia, PVCs, aspiration into the lungs, others related to high Tv or high PEEP include; stretch of alveoli up to rupture é resultant of; pneumothorax, pneumomediastinum, pneumopericardium, pul-monary interstitial emphysema.

O_2 toxicity: from high FiO_2 & the formation of singlet O_2 (free radicles) ẃ is toxic to all tissues, causing; RDS, BPD & retinopathy.

⇩COP: result from ⇧ of PEEP, RR, or Tv, resulting in higher intrathoracic pressure ẃ cause ⇩ of COP & venous return to the heart, also renal impairment due to ⇩ veno- us return, ⇧ fluid retention, also the metabolism of certain drugs are altered as the acid base balance is impaired.

Diaphragm paralysis: become lazy from having all the work done for him.

Ventilation associated pneumonia: from prolonged intubation, mainly caused by gra-m – ve bacteria especially pseudomonas aerogenosa (aerobic, rod-shaped & is a major agent of nosocomial infection).

⇧ In ICP: due to ⇩ of venous return from head & may cause IC Hge.

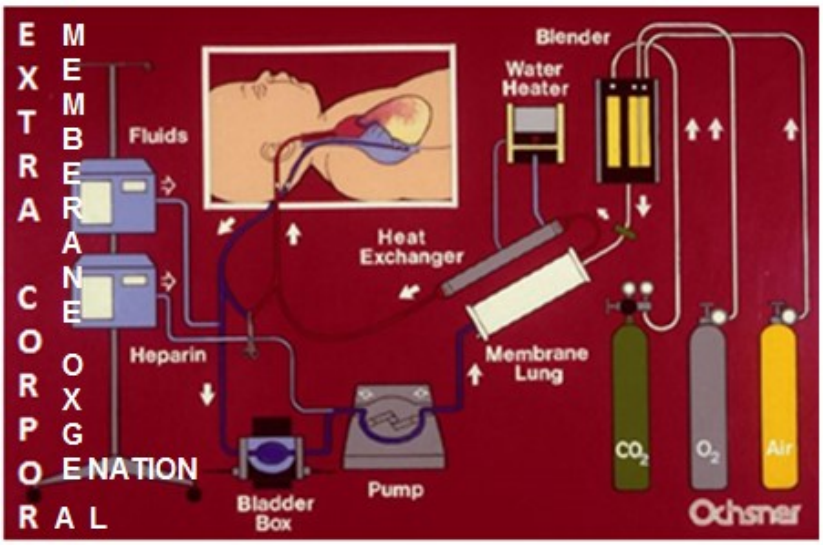
Respiratory alkalosis & Gastric distension.

Ways preventing added injury

- Minimize invasive therapy.
- Optimize lung volume
- Target CO_2 & O_2 .
- Manage fluid & nutrition to enhance normal development.

Extra Corporal Membrane Oxygenation

Needs highly specialized NSCU & team approach 24/7. It is a technique providing both cardiac & respiratory support to babies whose, heart & lungs are so severely diseased or damaged that they can no longer servers their function, is similar to a heart-lung machine, internal Canulation of baby receiving ECMO performed by a surgeon, there are several forms of ECMO, the most common of Ŵ are venoarterial & venovenous, in both modalities, blood drowned from the venous system are oxygenated outside the body, newborn can`t place on ECMO if they are under 4.5 pounds & ECMO can have dangerous side effects, in addition to that the large catheter inserted in the baby`s neck provide a fertile field for infection & resulting in fatal sepsis.



MECONIUM ASPIRATION SYNDROME

Is a serious condition in ŵ newborn breathes mixture of meconium & amniotic fluid into the lungs around the time of delivery, can be detected in 8-25 % of all birth after 34 weeks gestation, this is usually happens when the babies are under stress. Meconium is the early feces passed by a newborn soon after birth, before the baby started to digest breast milk or formula, it is viscous, dark, green substance composed of intestinal epithelial cells, lanugo, mucous & intestinal secretions as bile, it is sterile & does not contain bacteria. MAS did not occur before 34 weeks gestation (time for development of sucking & swallowing). The fetal distress during labor causes intestinal contraction & relaxation of anal sphincter ŵ allow meconium to pass into the amniotic fluid, the inhaled meconium can cause airway obstruction, air trapping, over distension of the lungs, pneumothorax, chemical pneumonitis, atelectasis, deactivation of surfactant, secondary bacterial infection & ⇧ pulmonary vascular resistance.

Risk factors for MAS

✻Post maturity. ✻Difficult or prolonged labor. ✻FPD. ✻Infant of diabetic mother. ✻Pre-eclampsia. ✻Maternal hypertension. ✻Oligohydramnios. ✻Maternal drug abuse especially tobacco, cocaine.

Clinical Picture

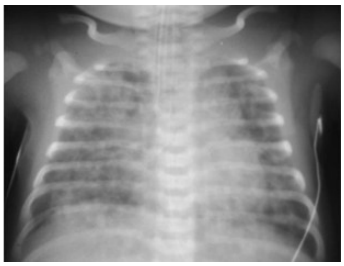
🕊Tachypnea, intercostal retraction. 🕊Yellow-green staining of finger nails, umbilical cord & skin. 🕊End expiratory grunting. 🕊Barrel shaped chest in the presence of air trapping. 🕊Auscultated rales & rhonchi (in some cases).

Investigations

- CXR: radiographic findings are different, over expansion of the lungs, barrel shaped chest, widespread coarse, patchy infiltrates (atelectasis, consolidation), other findings

include, areas of emphysema (air trapping), spontaneous pneumothorax, pneumomediastinum, small pleural effusion, or no air bronchogram from obstruction of trachea or main bronchus •ABG •Blood culture & sensitivity.

Frontal chest show large, ropey & strand like densities, barrel shaped chest in a post mature infant.



Differential Diagnosis

➤Birth asphyxia é pulmonary hypertension &/or hemorrhagic pulmonary edema.

➤Sepsis/Pneumonia ➤Pneumothorax ➤RDS ➤TTN ➤Cong diaphragmatic hernia.

Complications

✳Persistent pulmonary hypertension. ✳Air leak: pneumothorax, pneumomediastinum. ✳Complications of asphyxia: HIE, DIC, thrombocytopenia & renal failure.

Management

Normal term infants born through meconium stained amniotic fluid éout history of maternal group B streptococcal infection or other infection, whom are vigorous at birth & manifest no resp. distress, can be allowed to stay é the mother as a normal newborn. In presence of risk factor as chorioamnionitis, PRM, postmaturity, oligohydramnios, or FHR abnormalities, broad spectrum antibiotics to be started (Ampicillin + Gentamicin), discontinue if 48 hrs blood cultures are -ve.

Mild MAS: in presence of thick meconium staining & fetal distress, suctioning is better done under direct vision using suction catheter 12-14 FG, connected to suction source & é pressure 50-100 mmHg, the procedure is repeated until meconium is no long seen in the suction content & admit to SCU where incubator care & keep baby in NTE, give O_2 by

hood or nasal cannula, using FiO_2 < 40% to maintain PsO_2 88-97%, nutritional support, start IVFs & NPO & antibiotics (Ampicillin + Gentamicin), discontinued if 48 hrs blood C/S are -ve , usually infants in this category recover in 3-5 days.

Moderate MAS: CPAP if FiO_2 to be raised above 0.4 (40%) to maintain O_2 saturation within normal limits (CPAP should not be used in presence of air leaks & air trapping on CXR), antibiotics (Ampicillin + Gentamicin) IV for 2-3 days, then start TPN using amino acids then Intralipids later from the 7th day. If infants not responding to the above measures, he should be intubated & mechanically ventilated.

Criteria for ventilating the baby: •Cyanosed & need O_2 >60% (FiO_2 >0.6). •⇧ of RD. •Frequent apnea. •Deterioration of ABG: PaO_2 <50, $PaCo_2$ >70, HCO_3 ⇩ & pH < 7.25.

Surfactant: instillation in trachea as MAS cause deactivation of the natural surfactant

Inotropes:"Dopamine & Douputamine" may be used as many of such asphyxiated infants at birth may have myocardial depression, ⇩ COP, BP & tissue perfusion.

Severe MAS: infants not responding to the above procedures & who is refractory to the mechanical ventilation, is best managed at level IV NSCU, to give inhaled nitric oxide or ECMO. iNO can be started after ECHO confirmation of pulmonary hypertension & is useful in the management of pulmonary hypertension associated é MAS, it act by relaxing smooth muscles of pulm vessels causing vasodilatation, promoting bronchodilatation & is more effective when combined é HFV. Starting ē conc of 20 ppm for 4 hrs, then 5 ppm for 20 hrs, along é conventional or HFV. ABG & Methaemoglobin are measured at 4, 24, 96 hrs, treatment continued at 5 ppm until FiO_2 is <0.7, PaO_2 is >60 mmHg & pH <7.55. Multicenter clinical trials on iNO therapy had typical duration of <5 days & in case of no response, place the baby on ECMO.

PERSISTENT PULMONARY HYPERTENSION

Persistent pulmonary hypertension of newborn is syndrome of marked pulmonary hypertension that causes hypoxemia & right to left shunt, seen in 1/1000 births. The marked pulmonary hypertension affects pulmonary arteries & capillaries in the lungs ẃ become narrowed, blocked, or destroyed, this make it harder for blood to flow through the lungs & raises pressure within the arteries in the lungs, this put burden on the right ventricle ẃ must work harder to pump blood through the lungs. In neonates pulmonary hypertension is not defined by a specific pressure of the pulmonary circulation & the diagnosis is confirmed regardless of the pulmonary arterial pressure as long as it is accompanied by right to left shunt & absence of CHD.

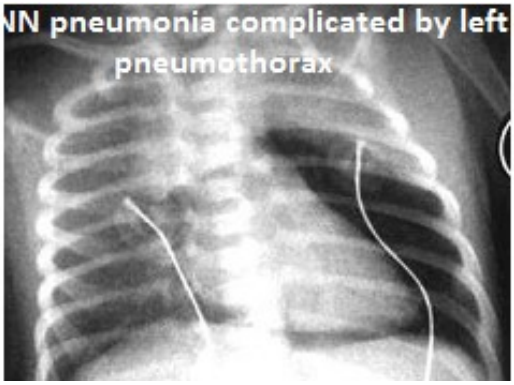
Causes

- HMD.
- Meconium aspiration.
- TTNN.
- Congenital pneumonia.
- Hypo plastic lungs.
- Polycythaemia.
- Heart failure.
- Idiopathic.

Clinical picture

Signs of RD (dyspnoea, tachypnea, intercostal, subcostal recession, working ala nasi), cyanosis & grunting.

NEONATAL PNEUMONIA



Incidence is 5-50% Live Births. The organisms found in fetal lung usually are those commonly found in the maternal vagina. Pneumonia in the early NN period usually occurs

in cases where membrane rupture for >6 hrs before delivery, also in case of prolonged or complicated labor & in PT infants. Pneumonia that became clinically evident within 24 hrs of birth may originate at 3 different lines:-

1- True congenital pneumonia; the mother has a blood stream infection, or ascending infection from birth canal & aspiration of infected amniotic fluid.

2- Intrapartum pneumonia: during passage through birth canal.

3- Postnatal pneumonia, in some of the cases, are nosocomial.

Etiology

- Group B strept: the most common cause of NN pneumonia.
- Escherichia Coli: the most common cause among VLBW.
- Other bacteria includes; non-typable haemophilus influenza, klebsiellas, pseudomonas, listeria monocytogenous, enterococci, staph aureus & chlamydia. In children the commonest cause is viral, then comes streptococcal pneumonia, staph aureus. Strept pneumonia tend to affect right lung & to cause pleural effusion.

Clinical picture

The features are very nonspecific & precise symptoms should not be sought, however generalised features may include:-Lethargy & RD.

- Tachypnea, Expiratory grunting & Apnea.
- Intercostal, subcostal, suprasternal retraction & working ale nasi.
- Diminished air entry over area of consolidation or effusion.
- Fine crepitations in some babies.
- Bradycardia, poor feeding & temperature instability.

Investigations

- CXR: may not apparent in the early stage, or resemble HMD & may show lobar pneum-

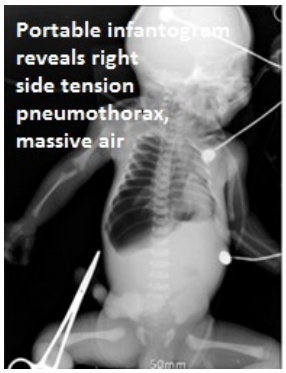
onia or interstitial infiltrates, or associated é complications e.g. pneumothorax.

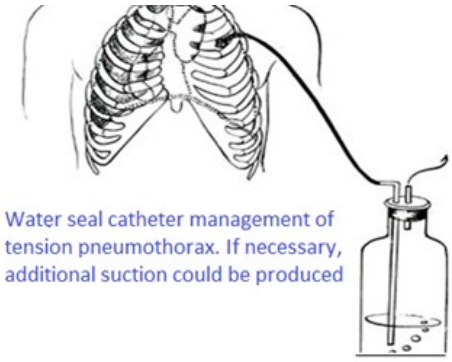
- CBC & Blood C/S.
- Tracheal aspirate C/S.
- Nasopharyngeal swab C/S.

Treatment

Ampicillin + Gentamicin é or Cefotaxime + Vancomycin. should be started as soon as possible. Duration depends upon the follow up investigations & clinical response.

PNEUMOTHORAX





Seen in 1-2% of all newborns. Presence of air or gas in pleural cavity separating visceral pleura from parietal pleura. It is either cong. or acquired. 25% of cases are 2ry to CPAP or mechanical ventilation. 15% are 2ry to MAS. The surfactant therapy can ⇩ the risk of pneumothorax in newborns during mechanical ventilation. Some otherwise healthy babies may develop spontaneous air leak as a result of rupture of congenital bleb or bullae that does not cause symptoms or distress.

Clinical Picture

- Sudden deterioration & desaturation.
- ⇧ RD & ⇩ chest movements.
- ABG show hypoxia &/or metabolic acidosis.
- Unequal or ⇩ air entry in the affected side.
- Hyperresonance on the affected side.
- Hyperinflation on the affected side.
- Mediastinal shifting.
- Displaced apex beats.
- Liver & spleen pushed down.

Diagnosis

- CXR: air in places outside normal lung airway, lung collapse, shifting of mediastinum to opposite side, jet black appearance of the affected side é absence of lung marking. May associated é subcutaneous emphysema & pneumomediastinum.
- Transillumination: fibroptic light probe placed on baby chest wall, the affected side transmits brighter light (hyperleucency) this procedure often used as an emergency.

Management

Drainage is often matter of urgency especially when air collection is under pressure (tension pneumothorax) or when associated é clinical deterioration, may require immediate drainage (needle aspiration or intercostal catheter insertion). Pneumothorax ŵ diagnosed, as incidental finding on CXR may not require active drainage.

Needle aspiration

Care must be taken to avoid laceration of lung or puncturing blood vessel, larges size butter fly needle, 21 gauge (green) or 23 (blue), 3 way stopcock, connected to 10 ml syringe, alcohol skins wipe & 1 pair sterile gloves

- Infant supine, prepare area é alcohol wipes, insert needle into pleural space directly over top of the rib in the 2nd or 3rd LICS at mid clavicular line, or 4th LICS at anterior axillary line, until air is aspirated into syringe then expel air through the 3 way stopcock.
- Following needle aspiration, insertion of ICC is -chest tube- under local anaesthesia é suction machine 10-20 cmH_2o usually required for ongoing management.

Catheter-torcher chest drainage

Done by surgeon, using sterile surgical instrument pack, under local anesthesia, transverse incision to be done parallel to 2nd or 3rd left rib, blunt dissection of intercostal muscles, putting thoracic tube number 10-20 attached to suction machine, drainage under

water seal é continuous suctioning pressure of 10-20 cmH_2o, evaluation by repeated CXR & clinical response, é the stoppage of air bubbling, clamp in site for 24 hrs, re-evaluate & then remove.

CONGENITAL DIAPHRAGMATIC HERNIA

Suspected in pregnant mother ē hydramnios. It is a developmental defect in the diaphragm (of unknown cause), result from failure of closure of the pleuroperitoneal canal in developing embryo ŵ usually completed by the 8th wk gestation ē resultant absence of portion of diaphragm (rarely complete absence), allowing abdominal contents to migrate into the chest, compressing the ipsilateral developing lung, leading to pulm hypoplasia & hypertension. The defect usually poster lateral (Bochdalek hernia), or anterior (Morgagni hernia). In 80% of cases of diaphragmatic hernia occur in the left side. Hiatus hernia in older children may be presented ē recurrent chest infection & failure to thrive. hiatus hernia is either sliding esophageal hernia or paraoesophageal hernia, diagnosed by CXR ē thin barium swallow film under TV screen by expertise. In highly specialized center they do PH monitor of esophagus over 24 hrs.

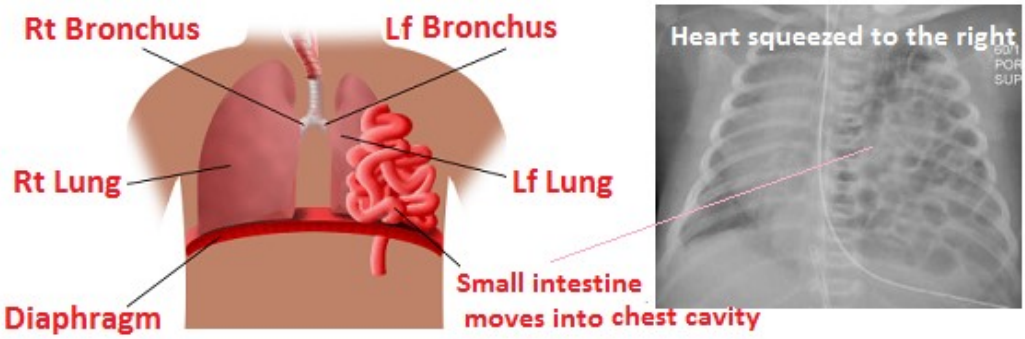
Incidence

1/3000 births. 28% of cases associated é other congenital anomalies, commonly NTD, cardiac defect, esophageal atresia, omphalocele & trisomy 13, 18 & 21. This defect is very commonly seen in stillborn infants.

Clinical Picture

- Severe RD at time of birth or respiratory deterioration hrs after birth, or sudden deterioration of Apgar score after using O_2 mask due to induced stomach inflation ẃ cause pressure on the lungs.

- ⇩ Or absent breathe sounds in one side of the chest, usually the left (80%) é the heart shifted to the right & audible bowel sounds over the chest.-
- Large chest (barrel shaped) & asymmetry of chest wall.
- Narrow abdomen (scaphoid) & feels less full on palpation.



Diagnosis

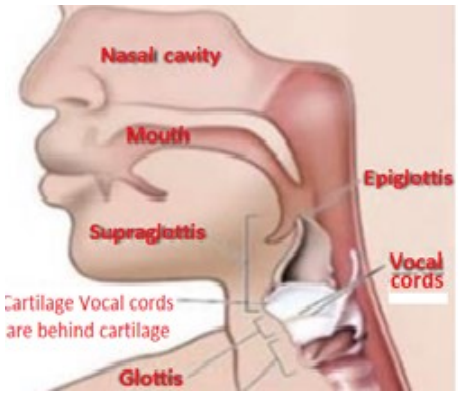
- Prenatal diagnosis: U/S in 2nd TM, polyhydramnios 80% cases.
- Chest X ray: bowel loops in chest, mediastinal shift.

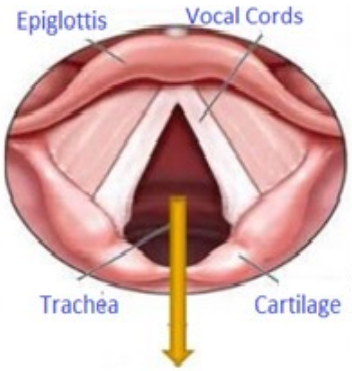
Management

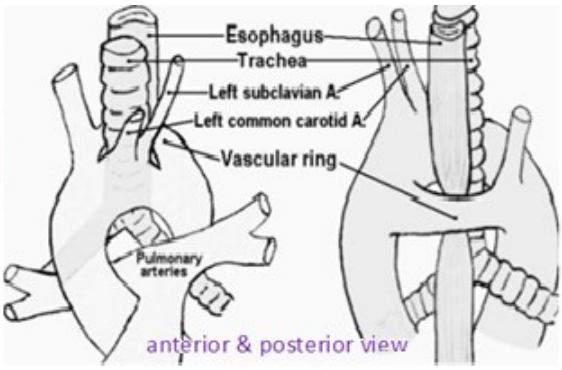
In delivery room: in case of prenatally diagnosed case, or case diagnosed during resuscitation, avoid bag mask ventilation, immediately intubate the baby & put nasogastric tube connected to continuous suction to prevent bowel distension & further compression & transfer immediately to pediatric surgery dep. in portable incubator.

In NICU: gentle lung ventilation, using HFV, either jet or oscillating ventilator, adapt ventilator to obtain O_2 saturation 85-90%, pH >7.2, avoid high pressure, continuous monitoring of ABG, BP, perfusion, the use of surfactant may be beneficial & transfer to pediatric surgery department as the baby will need urgent surgical correction, the operation is called Nissan fundoplication operation.

STRIDOR







VASCULAR RING

Stridor is symptom not diagnosis or disease. It is harsh vibrate sound of different pitch (high, low) produced by turbulent air flow through narrow or obstructed airway. The URT divided into:-

- Supraglottic (nose, nasopharynx, oropharynx & hypopharynx).
- Glottic (larynx).
- Subglottic (extra thoracic trachea, intrathoracic trachea & bronchus).

Types

- Inspiratory stridor: suggests supraglottic or glottic obstruction.
- Expiratory stridor: suggests subglottic.
- Biphasic stridor: suggests subglottic or glottis.

Causes

- Supraglottic: chonal atresia, microganthia (Pierre Robin), macroglossia in Beckwith-Weidemann, Down, glycogen storage disease, congenital hypothyroidism, lingual thyroid & thyroglossal duct cysts.
- Glottic: laryngomalacia (commonest cause), laryngeal cyst, web, papillomata, vocal cord paralysis (2nd most common cause), Arnold chiari sy, IC Hge, ⇧ ICP, HIE, trauma during delivery (traction), or post intubation.

⊙Subglottic: tracheomalacia, congenital tracheal stenosis, subglottic Haemangioma, cystic hygroma, laryngotracheoesophageal cleft, vascular ring, mediastinal mass (teratoma, lymphoma, lymphadenopathy).

Diagnosis

⊙History of birth: trauma, intubation, HIE, RD degree, cyanotic episodes, type of stridor, ability to feed, chonal patency, jaw & tongue size.

⊙CXR (P/L): detection of air way passage, soft tissue mass.

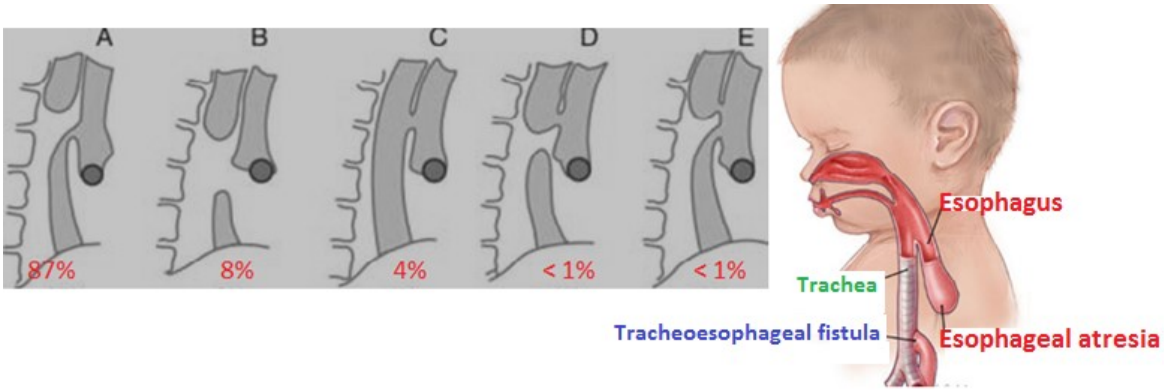
⊙Barium swallow: detection of any compressors over trachea, esophagus, detection of vascular ring as oblique running filling defect in mid thoracic region.

⊙Endoscopy: localize site of lesion & vocal cord movement.

Management

60% of cases are laryngomalacia, appear since birth, inspiratory stridor, resolve spontaneously within months, exaggerated é presence of URTI. Other cases treated according to the cause, surgery may require.

TRACHEO-ESOPHAGEAL FISTULA



Failure of embryonic esophagus & trachea to separate correctly during 3rd wk gestation, abnormal connection in one or more places between esophagus & trachea.

Incidence: 1/5000 Live Births.

May associated ē other anomalies (VACTERL), the presence of 3 of the following, seen in

1/10.000 Births: V Vertebral: NTD. A Anal: imperforate anus. C Congenital heart (VSD, ASD ,PDA). T Tracheoesophageal fistula. E Esophageal atresia. R Renal agenesis, ureteral anomalies, hypospadias. L Limb: polydactyly, wrist or knee anomalies.

The Role is (once there is one congenital anomaly, always look for another).

Diagnosis

Prenatal: hydramnios in 85% of cases of TOF.

Newborn: Cough, Chocking, Cyanosis (3C), Excessive salivation, drooling, Vomiiting, coincident é onset of feeding , Inability to pass down orogastric tube into stomach (type A). CXR ē advancing radiopaque feeding tube through nose, tube curled up in the upper pouch of esophagus (site of oesophageal atresia), absence of air/gas bubb-les in stomach, intestine (type B,D). Presence of air/gas, gastrointestinal distension (type A,E). Barium swallow using <1 ml water soluble contrast media via nasogastric tube, by expertise under TV screen for localisation of fistula (type E). The type H-fist-ula is the most difficult to diagnose & latest in presentation. Bronchoscopy/Endosco- py. U/S for diagnosis of any associated congenital nomalies.

Management

While awaiting surgery, infant condition stabilized, preoperative care concentrating on avoiding aspiration pneumonia.

- Elevating the head to avoid reflux aspiration of stomach contents.
- Using suction catheter to remove mucous, saliva that could be inhaled, suction every 5-10 min to keep proximal pouch clear.
- Withholding oral feed.
- IVFs, then TPN when necessary, or gastrostomy tube for feeding.

CROUP

Acute laryngotracheobronchitis is viral infection, affect age group 3 month - 3 year.

Clinical picture

Preceded by URTI for few days followed suddenly by, stridor ẃ is inspiratory & expiratory, hoarseness of voice stridor is harsh sound originate from vocal cords due to narrowing of larynx or trachea as result of inflammation or edema.

Investigations

- CBC.
- Blood culture.
- CXR (PA, L) in 1st attack to R/O foreign body inhalation.

Management

Never try to touch pharynx. ETT rarely needed (1% of cases). Warm moist steam/tincture benzoin compositum. Adrenaline 1/10.000, 0.1ml/Kg SC, may repeated after 15 min ē duplication of dose if needed, may be given through nebulizer. Fortecortine/ Decadrone amp 8 mg/2 ml, 0.5 mg/Kg/dose, IM/IV repeated 6 hourly if needed. Phenadon syrup 1 X 2 for two days. O_2 supply. IVFs & hydration.

EPIGLOTTITIS

Of sudden onset, caused by Haemophilus influenza bacteria, affect age group 3-5 yr.

Investigations

- CBC.
- Blood culture.
- CXR (PA, L) in 1st attack to R/O foreign body inhalation.

Management

Never try to touch the pharynx. Admit to hospital as 60% need ETT by anaesthiologist in the operating theatre. Chloramphenicol syrup 150 mg, amp 100 mg, 25mg/Kg/ day ÷ 4, for 10 days, or Augmentin syrup (Amxacillin + Clavulinic acid) 156 mg, 50mg/ Kg÷3 for 5 days. or Claforan (3rd gen. Cephalosporin) amp 500 mg/12 hrs IV, IM, 50 mg/Kg/day ÷ 2,

give the first 2 doses parentrally, then oral as Orelox syrup 40 mg, 8mg/Kg ÷ 2 X 5 days. Warm moist steam/Tincture Benzoin Compositum. Adrenaline 1/10.000 ,0.1ml/Kg SC, repeated after 15 min ē duplication of dose if needed. Fortecortine/Decadrone amp 8mg /2 ml, 0.1 mg/Kg/dose, IM/IV repeated 6 hourly if needed. Phenadon syrup 1 X 2 tsp for two days.

ASTHMATIC or WHEEZY BRONCHITIS

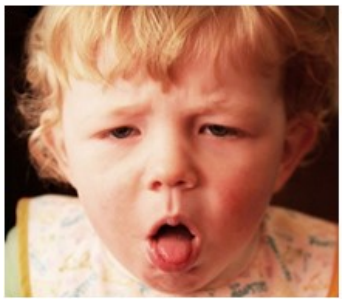
Investigations: CXR to R/O foreign body inhalation (if 1st attack).

Management

- O_2 supply: mask or catheter.
- Combivent/Atrovent vial using nebulizer, for children ½ vial, can be repeated after 30 min, then 8 hourly (selective beta agonist)
- Adrenaline: 1/10.000, 0.1 ml/Kg SC, may repeated after 15 minutes ē dose duplication.
- Fortecortine/Decadrone 8 mg amp/2 ml, 0.1 mg/Kg/dose IM/IV, may repeated 6 hourly.
- Phenadon syrup 1 tsp X 2 daily.
- Ventolin (salbutamol) syrup 125 mg, 1 tsp X 3.
- Minophylline ped syrup 125mg, sup 300mg 1 X2 daily or Ivyrospan syrup: 1 tsp X 3 daily
- Bronchicum syrup 1 tsp X 3 daily may be used as expectorant.
- Singulair sachet 4 mg, 1sachet over ¼ cup of water daily, as prophylactic from age of 6 months to 14 yrs, continue for 6 months, for adults 1 tab 10 mg daily for same period.

Bronchopulmonary aspergillosis is sort of fungal infection of lung, presented ē manifestations of bronchial asthma & ⇧ eosinophilic count in addition to lung infiltrates on CXR. Treated ē Diflucan amp 100mg in 50ml given through IV drip, or syrup 25mg,5mg/Kg/day ē monitoring of creatinine level in blood + Prednisolone 5 mg tab.

WHOOPING COUGH



Etiology: Gram -ve bacteria, Pordetella Pertussis, incubation period 1-2 wks, infective during whole period of illness & 5 days after, one attack lead to permanent immunity while immunization does not.

Clinical picture

Repeated dry irritant cough ended by whoop each time, vomiting after the attack, may associated ē sub conjunctival Hge, may complicated by bronchopneumonia, lobar collapse. Whooping cough include 3 stages; prodromal stage for 2 wks, catarrhal stage for 2 wks & convalescent stage.

Investigations: nasopharyngeal swab culture.

Management

- Erythromycin sy 200 mg/tsp, 25mg/Kg÷3, or Zithromax sy 250mg/ tsp, amp 500 mg, 10 mg/Kg once daily for 3 days (Azithromycin, Macroloides).
- Toplexil, or Codilar, or Tussilar.
- All members of family to be given prophylactic Erythromycin.

Chapter IV

CARDIOLOGY

- Congenital Heart Diseases
- Examination of Cardio Vascular System
- Ventricular Septal Defect
- Patent Ductus Arteriosus
- Atrial Septal Defect
- Pulmonary Stenosis
- Coarctation of Aorta
- Aortic Stenosis
- Tetralogy of Fallot
- Transposition of Great Arteries
- Truncus Arteriosus
- Tricuspid Atresia
- Total Anomalies Pulmonary Venous Return
- Heart Failure

CONGENITAL HEART DISEASES

CHD is a heart problem that`s present at birth caused by improper development of the heart during fetal development.

Incidence: 1% of babies are born ē CHD, 30% of them require intervention to prevent death in the 1st year of life. 90% of cases have no known cause while 5% of cases are related to chromosomal abnormality & 2% are related to environmental factors. It is cyanotic in 22 % & acyanotic in 78 % of cases.

Simple way to classify Congenital Heart Diseases.

- Acyanotic (Left to right shunt): VSD, PDA, ASD.
- Cyanotic (right to left shunt): the 5 " T_s ": T4, TGA, TA, Ta, TAPVR.
- Obstructive: AS, PS, COA.

Commonest group of life threatining Congenital Heart Diseases.

VSD (30:50%) -PDA (10%) -ASD (6%) -PS (6%) -CoA (6%) -AS (5%) -F_4 (5%) -TGA (5%).

EXAMINATION OF CARDIO VASCULAR SYSTEM

Inspection: nutritional status, RR, recessions, cyanosis (central or peripherall), pallor, clubbing of fingers, dysmorphism (top 3 syndromes: Down's Williams, Digeorge or Turner's , Noonan's), visible pulsations (hyperdynamic apex beat), chest wall deformity.

Palpation

- Apex precordium: thrills (like stroking a cat), turbulence, heaves heart.
- Femoral pulse: if we feel the femoral pulses does this R/O CoA.
- Liver: >2 cm BLCM.

Auscultation: heart sounds (1 & 2), murmur (systolic or diastolic), murmur intensity (loud or soft) & where is it loudest ?

Innocent or Pathological murmurs

Innocent murmur: the diastolic murmur is never innocent. Innocent murmur is present in at least 50% of normal children.

Still`s murmur: low pitched, vibratory, systolic ejection, ⇧ ē supine position.

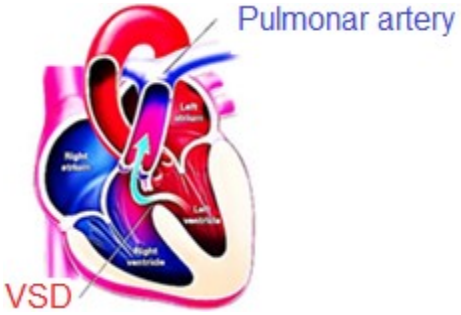
Venous Hum: continuous murmur in supraclavicular region ⇩ on lying down or ē pressure on neck.

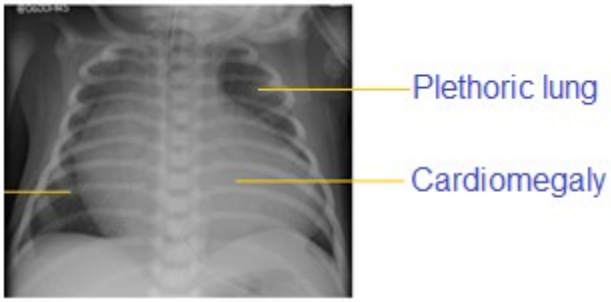
NB: # A baby ē PDA & high pulmonary pressure may have a completely normal examination in the first few wks of life even if there is significant problem ē the heart.

50% of babies ē CHD has no murmur on examination & absence of a murmur does exclude the presence of potentially serious heart disease.

ACYANOTIC CONGENITAL HEART DISEASES

VENTRICULAR SEPTAL DEFECT





VSD is communication between the 2 ventricles, if the defect is small it may passed unnoticed & closed spontaneously in the first few months of life, or even after decades of life, but 15% of cases become clinically symptomatic ē the presence of moderate to large VSD (>5mm in size) & presented ē CHF in the 3rd-4th wk of life. VSD is considered one of the most common causes of CHF after the NN period. In VSD, the O_2 content in Rt ventricle is greater than in Rt atrium.

Incidence: 20% of CHD. Isolated VSD seen in 2-6 /1000 Live births.

Clinical picture

●Dyspnea. ●Tachypnea. ●Difficult breathing. ●Failure to gain weight. ●Sweating while feeding. ●Frequent respiratory infection. ●Pansystolic (holsystolic) murmur along lower sternal border. ●Palpable systolic thrill along left sternal border ●Displaced apex beat as the heart enlarges.

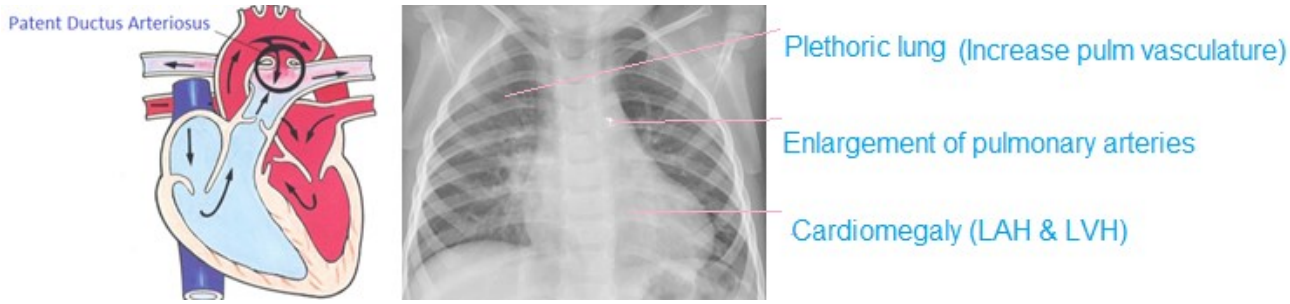
Investigations

●ECG: Left atrial hypertrophy; broad notched P wave in lead II & peak duration ≥ 0.04 seconds, terminal P negativity in lead V_1 ē duration ≥ 0.04 seconds & depth ≥ 1mm. Left ventricular hypertrophy; tall R in V_5 ,V_6 > 25mm & strain pattern V_5 , V_6 ē depressed ST segment inverted T wave in severe hypertrophy, deep S in V_1, V_2, ē or ēout left axis deviation, or biventricular hypertrophy.

●Chest X ray: cardiomegaly & plethoric lung.

●ECHO: localize the size of VSD, cardiac dilatation, evaluation of left ventricular func-tion, detection of presence or absence of other associated defect in the heart.

PATENT DUCTUS ARTERIOSUS



PDA is persistent communication between the descending thoracic aorta & the pulmonary artery that results from failure of normal physiological closure of the fetal ductus. PDA undergoes fairly rapid initial constriction during the first hrs after delivery.

The final functional closure over 1-8 days. In PT infants closure may be delayed up until the time of full gestational age & beyond, a widely PDA is an important & fairly frequent cause of serious illness in the neonate, symptoms of heart failure, growth retardation & may prone to lower respiratory tract infection. The frequency of PDA inversely proportional to advancing gestational age. PDA is present from birth but may not presented until adulthood, when symptoms of endocarditis, pulmonary hypertension, or heart failure may prevail.

Incidence: 1/2000 births. 10% of CHD. The female/male ratio is 2 : 1

Clinical picture

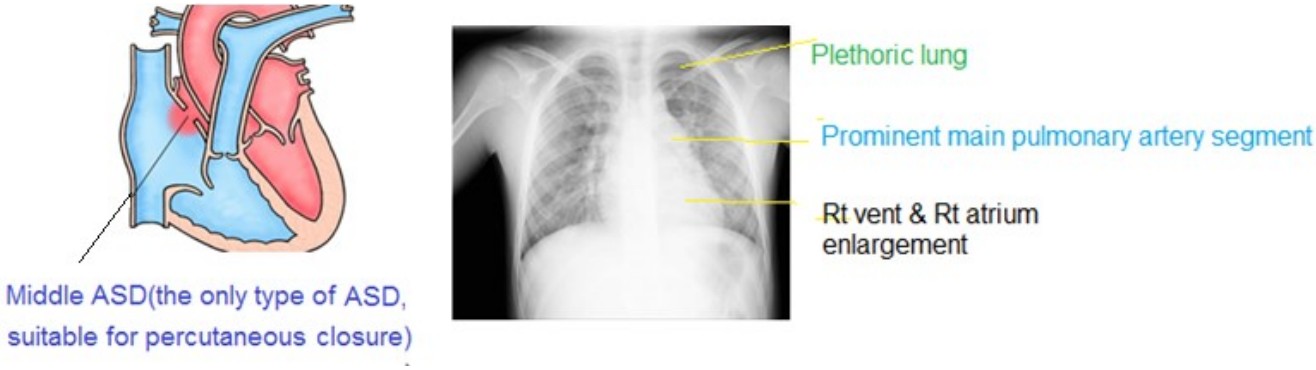
The symptoms depend upon the size of the ductus & how much blood flow it carries, it may cause no symptoms & detected by the heart murmur. The turbulent flow of blood through the PDA puts a person at a higher risk for a serious infection (endocarditis). The symptoms include:-

- Poor feeding & impaired growth.
- Sweating while feeding (diaphoresis).
- Dyspnea (shortness of breath).
- Tachypnea (rapid breathing).
- Bounding pulses (peripheral pulse ⇧ in amplitude).
- Widened pulse pressure >25 mmHg.
- Continuous “machinery” murmur, usually heard most clearly at the left upper sternal border & left infraclavicular area are characteristics.
- ⇧ susceptibility to chest infection & endocarditis.

Investigations

- Chest X ray: enlargement of pulmonary arteries & veins, cardiac enlargement (LAH & LVH) & ⇧ pulmonary vasculature (plethoric lung).
- ECG: LAH & LVH
- Echocardiography: confirmatory, demonstration of flow of blood through PDA, it`s size, presence of cardiomegaly & presence of other associated cardiac anomalies.

ATRIAL SEPTAL DEFECT



ASD is a defect in the wall between the 2 upper chambers of the heart. Include 4 typ-es; the commonest (80% of cases) is the ostium secundum, others include, ostium pr-imum, sinus cavernous & coronary sinus ASD. The left to right shunt (because the left atrial pressure is higher than that in right atrium), cause a large volume of blood than normal to be handled by the right side of the heart, this extra blood passes through the pulmonary artery into the lungs causing higher blood flow than normal in lungs.

Incidence: the 3rd most common CHD after VSD & PDA, account for 6-8 % of CHD. Girls have ASD twice as often as boys.

Clinical picture

Isolated ASD in infancy usually asymptomatic & are most often detected at the time of preschool physical examination, sometimes these defects are detected when ech-

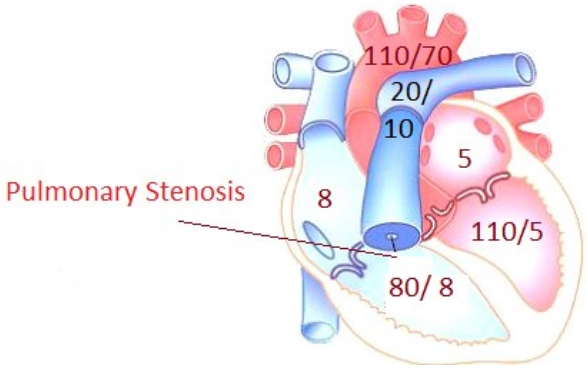
ocardiography are performed for some unrelated reason. A few babies do present ē symptoms of heart failure in infancy, this is uncommon. While ASD is generally well tolerated in infancy & childhood, development of exercise intolerance & arrhythmia in later childhood & adolescency & the risk of pulmonary vascular obstructive disease in adulthood make these defects important. The symptoms include;

- Soft ejection systolic murmur, grade I-II/IV, secondary to ⇧ blood flow across the pulmonary valve, heard best at upper left sternal border.
- Mid diastolic flow rumble (ē the bell of stethoscope), best at the lower left sternal border due to large volume flow across tricuspid valve.
- Fixed splitting of the 2ndHS (fixed, does not vary ē respiration) is the most character-istic sign of ASD).

Investigations

- Chest X ray: varying degrees of enlargement of right ventricle & atrium depending on the size of the shunt, prominent main pulmonary artery segment, plethoric lung (appearance of vessels in the distal lung), cardiac enlargement is often best appreciated on the lateral view because the Rt ventricle protrudes anteriorly as its volume ⇧.
- ECG: shows volume overload pattern on the right ventricle; the QRS axis may be normal or exhibit right axis deviation.
- Echocardiography: detect even small ASD, 100% accuracy, measurement of the size & description of the precise location of ASD.
- Cardiac Catheterization: for detection of O_2 saturation & pressure gradient in different chambers of the heart & in the main blood vessels.

PULMONARY STENOSIS



PS accounts for 7% of CHD, most cases are asymptomatic unless PS is severe. Commonly associated ē Noonan`s sy.(male Turner). Classified into 4 types; Valvular ẃ is the commonest & occur in 85% of cases, Supravalvular, Subvalvular (infundibular) & Branch peripheral PS (affecting either the left or right branch of pulmonary artery).

Mild PS: if the valve area is >1 cm/m^2 & the transvalvular gradient is 30-50 mmHg, or the peak right ventricular systolic pressure < 75 mmHg.

Moderate PS: if valve area is 0.5-1 cm/m^2 & the transvalvular gradient is 50-70mmHg , or right ventricular systolic pressure 75-100 mmHg.

Severe PS: if the valve area is <0.5 cm/m^2 & the right ventricular systolic pressure gradient is >75 mmHg.

In PS the pressure in the Rt ventricle ⇧ while in the pulmonary artery pressure ⇩.

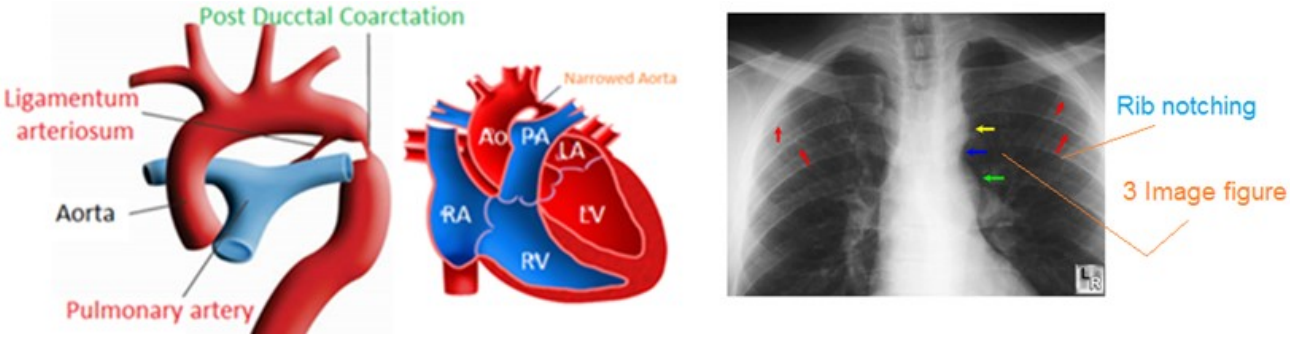
Clinical Picture

•Dyspnoea & tachypnea. •Lethargy & feeding difficulty. •Pale, cool, or clammy skin •Ejection systolic murmur, loudest in the left sternal border (2nd-4th ICS) & radiating toward the left shoulder. •Ejection click often precede the systolic murmur. •Wide splitting of the 2nd HS as a result of delay in right ventricular ejection. •Inaudible pulmonary closure sound (P_2) in severe PS. •There may be a thrill, best felt when the pt leans forward & breathes out. •Prominent α wave in the jugular pulse.

Investigations

- ECG: evidences of RVH : tall R wave in V_1 equal to or larger than S wave in that lead (reversed R/S ratio), deep S in V_6, strain pattern in V_1 , V_2 , V_3 ē depressed ST segment, inverted T wave in severe hypertrophy ē or ēout right axis deviation.
- CXR: usually shows a normal heart size. In severe PS there may be ⇧ in heart size & dilatation "post stenotic" of the main pulmonary artery.
- ECHO cardiography: define where the stenosis lies & how severe it is, evaluation of right ventricular function, systolic pressure & transvalvular gradient.

COARCTATION OF AORTA



CoA is a congenital narrowing of upper descending thoracic aorta, the heart must work harder to keep the blood flowing passed the narrowed area, ẃ may be predu- ctal, ductal, or postductal, most commonly at the site of insertion of the ductus arte- riosus, additional cardiac abnormalities are common including bicuspid aortic valve ẃ occur in 80% of cases, VSD in 40% of cases & ASD, or TGA. The risk ⇧ ē Turner, Sturge Weber, William & Di-George syndromes. The narrowed aorta produces ⇧ in left ventricular overload, wall stress, LVH & CHF.

Incidence

6% of infants ē CHD & the 5th most common cause of CHD in infants. Is 2-5 times more frequently in males than females.

Pathophysiology

Narrowed aorta produces ⇧ Lf ventricular afterload, wall stress, LVH & CHF. The associated pathology include:-

1. Collateral circulation

*Inflow primary from branches of both subclavian arteries (internal mammary A, vertebral A, costocervical, thyrocervical trunks).

*Outflow: into descending aorta, two pairs of intercostal arteries.

2. Aneurysm formation of intercostal arteries: 3rd&4th rib notching (rare before age 10 yr)

3. Coronary artery dilatation & tortuosity: due to LVH.

4. Aortic valve: bicuspid aortic valve (27-45%) & aortic stenosis (6-7%).

5. Intracranial aneurysm: Berry type intracranial aneurysm in some pts.

6. Associated cardiac anomaly: seen in 85 % of cases ē CoA.

Clinical Picture

Symptoms depend on how much blood can flow through the aorta, the presence of other heart defects may also play a role, around 50% of newborn ē this problem will have symptoms in the first few days of life, in milder cases, symptoms may not develop until the child have reached adolescence, symptoms include:-

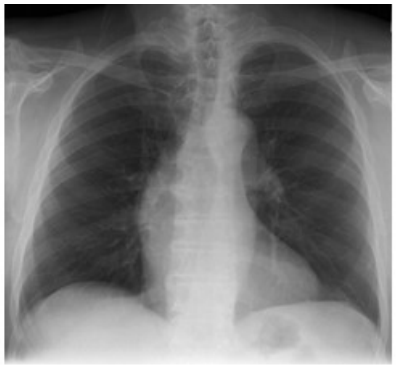
- Dyspnea. • Tachypnea. • Sweating. • Cold feet & legs. • Easy fatigability & poor feeding. • Failure to thrive, poor growth. • Nose bleeding. • Leg cramps. • Dizziness or fainting. • Headache. • High BP in the arms & occasionally the left arm pressure is lower than the right arm pressure if the origin of the left subclavian artery is involved in the coarctation. • BP difference between arms & legs (>20 mmHg). • Weak or delayed pulse in the legs. • Systolic murmur (harsh) or abnormal whooshing sound caused by turbulent blood flow, heard best in the left infraclavicular area & under the left scapula.

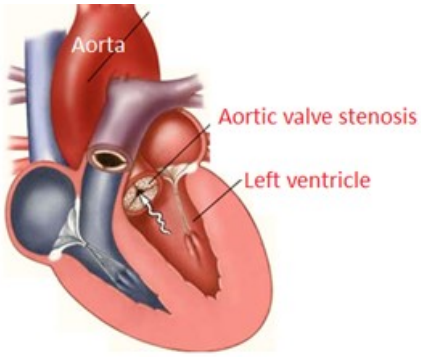
Unfortunately in infants <6 months of age ē CoA & cardiac failure, the diagnosis can be mistaken for sepsis or pulmonary disease in almost 50% of cases.

Investigations

- ECG: may be normal, or show LVH, Left atrium abnormality, 1st degree heart block, complete or incomplete RBBB or biventricular hypertrophy.
- Chest X ray: cardiomegaly, narrowing in the aorta at the site of the coarctation, pos-tstenotic dilatation results in the "3" image is often seen, notching (grooves) on the ribs, commonly seen after 5 yrs of age, results from erosions in the ribs 2ry to tortuous pulsating intercostal arteries. Pulmonary edema if associated ē HF.
- Echocardiography: localize the site & severity of the coarctation, flow. Color doppler measure the peak pressure gradient across the obstruction & left ventricular dimensions & function are assessed by M-mode & detection of other heart defect such as a bicuspid aortic valve ẃ occur in 80% of cases.

AORTIC STENOSIS





Chest x-ray shows prominent of the right mediastinal border occupied by the ascending aorta. The descending aorta is unfolded but of normal calibre. Heart size is normal. No lung or pleural abnormality

AS include supravalvular, subvalvular &valvular types, the aortic valve has 3 flaps, called "cusps" or "leaflets" that open & close during systole & diastole. Baby ē mild AS shows no symptoms & those ē moderate & severe AS can experience dyspnea, tachypnea.

Older children & adults experience dizziness, fainting attacks & easy fatigability, exertional dyspnea, angina & syncopal attacks. The left ventricle initially compensate for the ⇧ resistance caused by AS by thickening to help to eject blood through the stenotic aortic valve. Isolated AS rarely become symptomatic until the aortic valve area is <1 cm & the mean gradient is >40 mmHg or the aortic jet velocity is >4 m/second. Supra valvular AS commonly seen in William`s syndrome.

Incidence: 5 % of CHD & 4 times more likely to occur in boys than girls.

Clinical Picture

- Tachypnea. Dyspnea.
- Sweating while feeding.
- Liver enlargement.
- Palpable left ventricular heave or thrill.
- Pulmonary rales.
- Absent or ⇩ 2nd HS & narrow pulse pressure. .
- Systolic murmur is loudest over the 2nd right ICS & suprasternal notch ē or ēout thrill, transmitted to neck & apex.
- Aortic ejection click.
- Gallop rhythm.

Investigations

- ECG: is frequently normal but may show LVH.
- CXR: slight LVH, plethoric lung..
- Echocardiography: confirmatory, degree of valve obstruction, evaluation left ventricular function & filling pressure, transvalvular gradient, detection of coexisting abnormalities of other valves.

CYANOTIC CONGENITAL HEART DISEASES

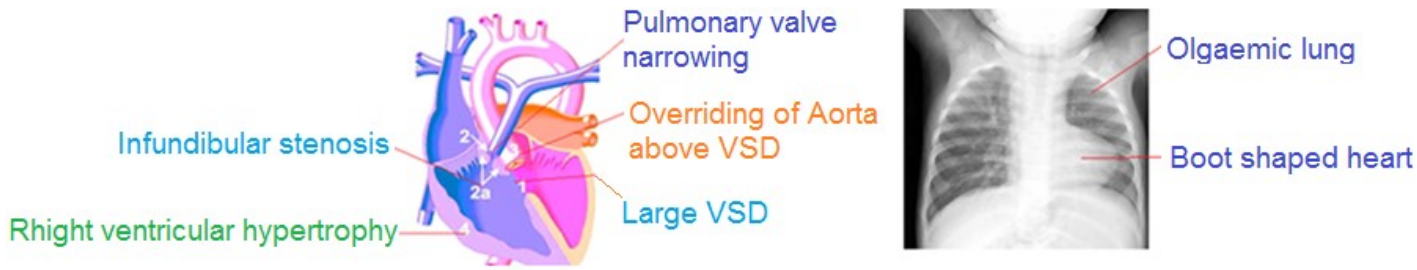
Cyanosis: is it of cardiac or pulmonary in origin?

Hyperoxia test: neonates ē CHD usually do not have significantly ⇧ of PaO_2 during administration of 100% oxygen.

- The O_2 saturation in cyanotic heart diseases is <90% (pulse oximeter) & PaO_2 <60.
- The degree of cyanosis; depend upon the amount of pulmonary blood flow.

The 5 T_s are the most common cyanotic CHD: T4, TGA, TA, Ta, & TAPVR.

TETRALOGY OF FALLOT



Incidence: 2-3/10.000 live births. The most common CHD beyond infancy.

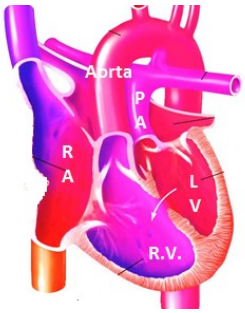
Clinical picture

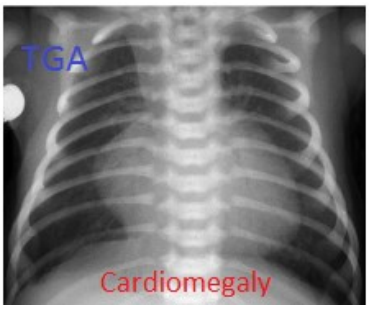
- Central cyanosis & Cyanotic spills.
- Clubbing seen from 3-6 months of age.
- Harsh ejection systolic murmur over the pulmonary area & LSB.
- Thrill along LSB.

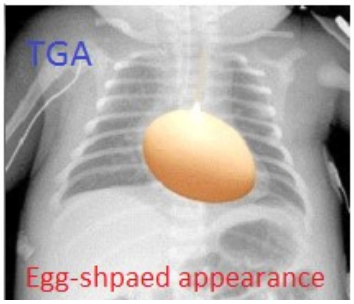
Diagnosis

- CXR: boot shaped heart & oligemic lung.
- ECHO: for anatomy of great vessels, overriding of aorta, PS, RVH & VSD.
- ECG: RVH ē/ēout right axis deviation.

TRANSPOSITION OF GREAT ARTERIES







TGA account for 5% of CHD, the aorta leaves the right ventricle (rather than the left as in normal heart) & takes unoxygenated blood to the body, while the pulmonary artery leaves the left ventricle & take oxygenated blood to the lung (the position of pulmonary artery & aorta are reversed), so that most of the blood returning from the lungs return to the lungs again "lung-heart-lung" & most of blood returning from the body return to the body again, "body-heart-body" ēout being routed to the lungs for oxygen. Infants born ē TGA survive only if they have one or more connections that let oxygen rich blood reach the body, either through ASD, VSD, or PDA. The TGA require surgery, usually in the first wk of life.

Clinical picture

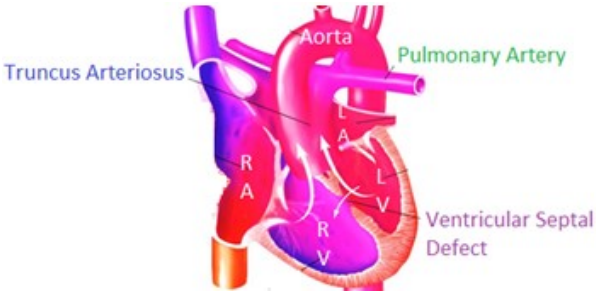
- Cyanosis: if the infant has an intact ventricular septum, cyanosis at birth (at least by 48 hrs because by then the ductus arteriosus has closed), all babies has a PDA at birth that may allow enough mixing to prevent severe cyanosis initially, but as the ductus arteriosus closes, as it typically will in the first hours or days of life, cyanosis become more severe. If infant has a large VSD, less severe cyanosis will be noticed & associated ē CHF.
- Tachypnea: in response to the low O_2 levels.
- Silent heart: no murmur, or are not typical nor always present unless other lesion present e.g. VSD.
- Palpable right ventricular impulse.

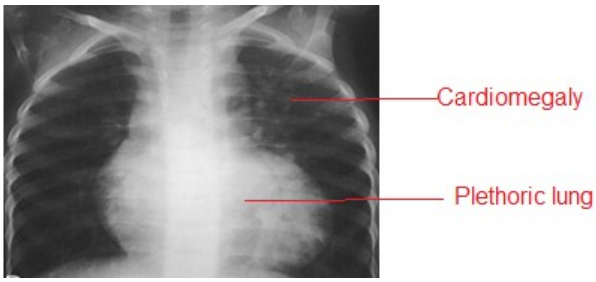
- Engorged neck veins.
- Enlarged liver become apparent in the neonatal period. If untreated, over 50% of infants ē TGA will die in the first month of life & 90% in the first yr. Babies will often develop signs & symptoms of CHF over the course of the first wks or months of life.

Diagnosis

- ECG: may show, RVH, Rt axis deviation & may show myocardial damage due to ischemia
- CXR: narrowed superior mediastinum gives to the cardiac silhouette characteristic egg-shaped appearance, cardiomegaly ē ⇧ pulmonary vascular markings may be found if VSD is present.
- ECHO: anatomy of vessels, presence of other associated anomaly e.g. VSD, ASD, or PDA, are easily seen.

TRUNCUS ARTERIOSUS





TA seen in 2-4% of severely sick neonates ē CHD. The pulmonary & aortic arteries are combined, only one artery arises from the heart & that this artery "TA" gives rise to the coronary arteries, pulmonary artery & aorta, result in ⇧ of blood flow into the lungs & it is always override a large VSD.

Clinical picture

•Cyanosis. •Dyspnea. •Tachypnea. •Excessive sweating. •Pansystolic murmur (VSD), at the left sternal border. •Cardiomegaly.

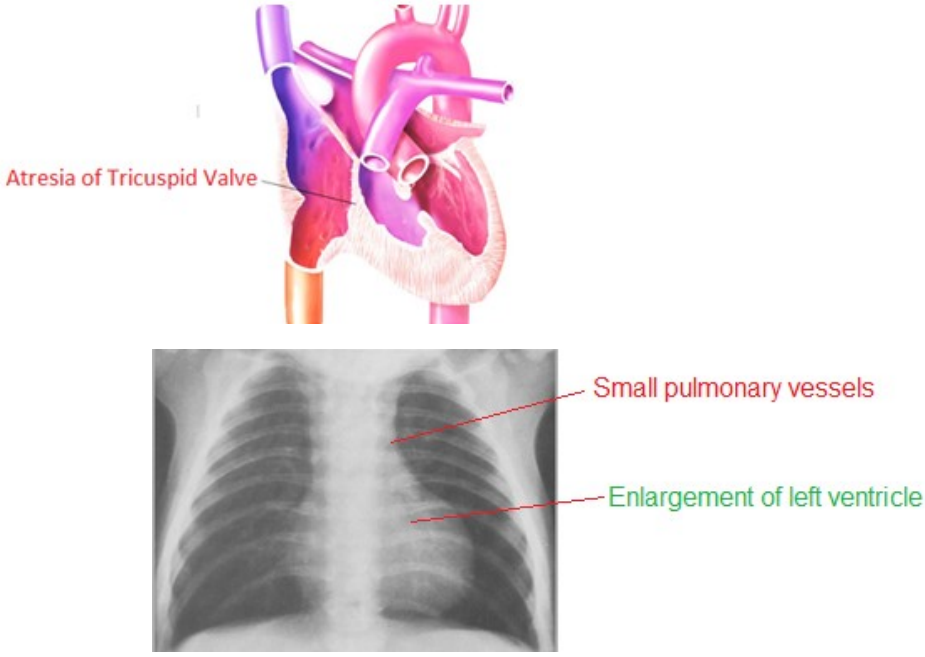
Investigations

•CXR: cardiomegaly, plethoric lung, the combination of right sided aortic arch, strongly suggests TA, however further confirmatory investigations are always needed.

•ECG: RVH & right axis deviation.

•ECHO: anatomy of great vessels, truncal valve, aortic arch & VSD are easily seen.

TRICUSPID ATRESIA



Ta is a congenital agenesis or absence of the tricuspid valve, during the first 8 wks of gestation, resulting in no direct communication between the right atrium & right ventricle, missing tricuspid valve result in an undersized or absent right ventricle & wellhave ⇩ blood flow into the lung. The blood that return from the body to the right atrium cannot directly enter the right ventricle & most pass through the hole of ASD into the left atrium & then to the left ventricle. An opening may be present in atrial or ventricular level, also

a PDA allow blood to pass through from the aorta to the pul-monary artery & receive O_2 from the lungs. 70% of cases have normal relationship of great vessels & 30% have transposition of great arteries.

Incidence: 5/100.000 Live births & the 3rd most common form of CHD.

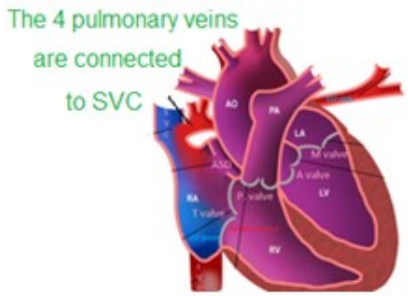
Clinical picture

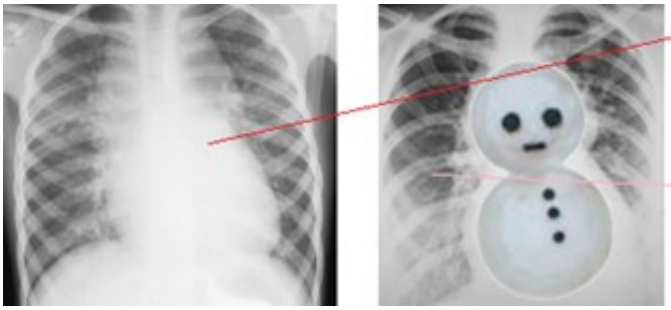
Nearly 50% of babies ē Ta present ē symptoms on the day of birth & 80% will have symptoms by the end of the first month of life, the magnitudes of pulmonary blood flow determine the timing of & type of clinical presentation. Infants ē pulmonary oligemic exhibit cyanosis in the first few days of life, tachypnea & acidosis, hypoxic spells are not common in the neonate although the spells can occur later in infancy, infants ē pulm-onary plethora usually present ē signs of heart failure within the first few wks of life; dyspnea, fatigue, difficult to feed & a holosystolic murmur on LSB is suggestive of VSD, or soft ejection systolic murmur & splitting of the 2nd HS ẃ is characteristic of ASD.

Investigations

- Chest X ray: enlargement of Left ventricle, a concave left border & small pulm. ves- sels, the aorta is continuous ē the cardiac shadow in left anterior oblique views.
- ECG: LVH & left axis deviation (the only cyanotic type of CHD ē this finding).
- ECHO: confirm the presence of Ta & VSD.
- Cardiac catheterization: is an invasive procedure that gives very detailed informati- on about the structures inside the heart, done under sedation, a small, thin, flexible tube (catheter) is inserted into a blood vessel in the groin & guided to the inside of the heart, BP & O_2 measurements are taken in the 4 chambers of the heart, as well as the pulm artery & aorta & contrast dye may be injected for more visualization.

TOTAL ANOMALIES PULMONARY VENOUS RETURN





TAPVR, the pulmonary veins have no connection ē the left atrium, they drain directly or indirectly into the right atrium. There is total mixing of the systemic venous blood & the pulmonary venous blood within the heart. The systemic circulation dependent on shunting through ASD or patent foramen ovale.

Clinical Picture

•Cyanosis of skin, lips, or nails. •Tachypnea & Feeding difficulties. •Not growing as fast as normal. •Frequent respiratory infection. •Some children does not start having symptoms until later in infancy.

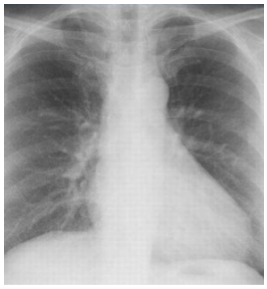
Investigations

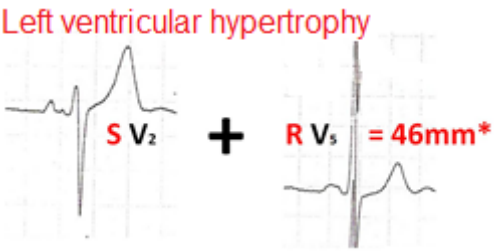
•Chest X ray: there is cardiomegaly with increased pulmonary arterial markings. There is dilation of both the Lf & Rt innominate veins & the right superior vena cava producing the classical "snowman" or "figure of 8" appearance. The superior mediastinum is enlarged secondary to dilation of the right vena cava, innominate vein & ascending vertical vein

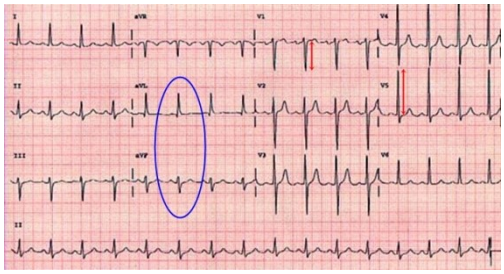
•ECG: shows RVH.

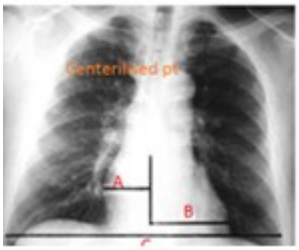
•ECHO: RVH, ASD, or patent foramen ovale ē left to right shunt.

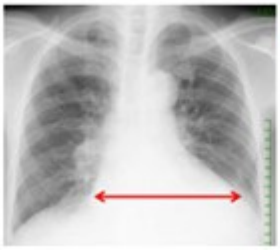
HEART FAILURE

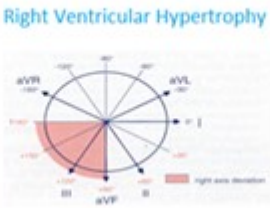


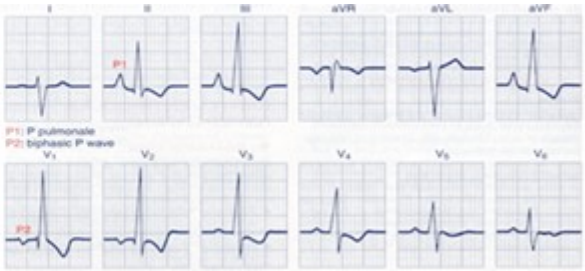












CHF refers to clinical state of systemic & pulmonary congestion resulting from inability of the heart to pump as much blood as required for the adequate metabolism of the body, the clinical picture of CHF result from combination of relatively ⇩ COP & compensatory responses to ⇧ it.

Clinical picture

- Feeding difficulties: important clue in detecting CHF in infants, interrupted feeding (suck-rest-suck cycles), inability to finish the feed, forehead sweating during feeds due to activation of sympathetic nervous system (a very useful sign).
- Tachypnea: > 60/min from age 0-2 months, & > 50/min in 2 months to 1 yr of age & > 40/min in 1-5 yrs of age, it is happy tachypnea ēout much retraction. Grunting (form of +ve end expiratory pressure). Fever ē pulmonary infection may produce tachypnea & in cyanotic CHD, tachypnea may be due to associated brain anoxia & not CHF, the Rx for the 2 conditions is entirely different.
- Tachycardia: > 160/min. in infants, > 100 /minute in old child. Tachycardia ē absence of fever or crying & accompanied by rapid RR & hepatosplenomegaly is indicative of heart

failure. Consider supra ventricular tachycardia if heart rate >220/min. in infants & >180/ minute in older child.

- Cardiomegaly: consistent sign of impaired cardiac function, secondary to ventricular dilatation &/or hypertrophy, may be absent in early stages especially ē myocarditis, arrhythmia, restrictive disorders & pulmonary venous obstruction.
- Hepatosplenomegaly: lower edge of the liver is palpable >2 cm below right costal margin, may be associated ē mild elevation in the bilirubin level & LFTs changes. The ⇩ in liver size after initiation of Rx is an excellent criterion of response to Rx. Usually in such circumstances the spleen is palpable.
- Jugular venous pulsation: seen only in older children & adolescents.
- Pulmonary rales: of not much use in detecting CHF in infants, rales may be heard at both lung bases, when present, are difficult to differentiate from those due to the pulmonary infection ẃ frequently accompanies heart failure.
- Peripheral edema: is a very late sign of failure in infants & children. Facial edema is most common in infants & children, while presacral & posterior chest wall edema in young infants, it indicates a very severe degree of failure. Daily weight monitoring is useful, rapid ⇧ in Wt >30 gm/day in neonate may be a clue to CHF & also useful in monitoring response to Rx. Cold extremity, ⇩ BP, skin mottling, all are signs of impending shock
- Pulse: either pulsus alternans (strong & weak contractions of failing myocardium), pulsus paradoxus (⇩ of pulse volume & BP ē inspiration) are frequently observed in infants ē severe CHF.
- Apical Pulse: visible, diffuse apical pulsation in RVH. Visible, localized apical in LVH.

Investigation

- CXR: look for heart size, contour, pulmonary vasculature, presence/absence of pleural

effusion. In RVH, an angle seen between apex & diaphragm, while in LVH no angle. The earliest sign of heart failure will be cardiomegaly (before pulm. edema). Cardiomegaly is the ⇧ of cardiac shadow > 50% of chest diameter as shown in the diagram below. The cardiac shadow calculated as; horizontal line from must concave point to mid vertical line of centralised pt (a) + horizontal line from must convex point to mid vertical line of centralised pt (b), it is equal to sum of (a) + (b).

- ECHO: the 1st sign of heart failure on ECHO will be enlargement of the filling chambers (left atrium for left sided heart failure, right atrium for right sided heart failure), &/or ⇩ of ventricular contractility.
- ECG: RVH; R wave in V_1, V_2, V_3 > 25 mm, right axis deviation. LVH; R wave in V_5,V_6 > 25 mm, deep S in V_1,V_2, inverted T & left axis deviation
- Electrolytes & CBC: including Ca^+, Mg^+, K^+. & CBC. This helps us to R/O the presence of anemia & electrolyte disturbances. Baby often have mild hyponatremia resulting from ⇧ renal water retention rather than a true -ve sodium balance, mild hyponatremia, ther-efor, does not need to be treated, & administering supplemental sodium may actually worsen the baby`s fluid retention & heart failure. Ca^+ should be administer when hypoca-lcaemia is documented (Ca^+ gluconate 10% 1 ml = 100mg/Kg IV), same as for Mg^+ (Mg^+ SO_4 50% 25 mg/Kg IV). K^+ is important especially when we start to use furosemide, hypo-kalemia may cause cardiac arrhythmia, cardiac arrest, polyuria, paralytic ileus & muscle weakness (dose of K^+cl^- 20% is 2 ml/Kg).

Management

- Diuretics: Furosemide (Lasix), 0.5-1 mg/Kg/day, dose can be ⇧ to 3 mg/Kg/day in severe CHF.
- Inotropic: Dopamine infusion 5-10 mcg/Kg/hr.

- Correction of acidosis: through administration of fluid &/or $NaHco_3$.
- Digoxin: Digitalis Glycoside. digitalizing dose; PO 8-10 mcg/Kg/day, or IV 80 % of the oral dose, maintenance dose is approximately ¼ of total daily dose divided bid, its half-life is 36 hrs, so given once or in two divided doses daily, it is well absorbed through GIT, initial effect can be seen within 30 min. after oral administration & within 15 min after IV administration, adjust the dose in pt ē renal failure. Give ½ the total digitalizing dose immediately & the succeeding 2 quarter doses at 12 hrs intervals, later ECG monitoring. The dose of digoxin is almost never increased but may be decreased in the presence of toxicity or renal failure. Signs of cardiac toxicity include; arrhythmia, bradycardia, AV block & PVCs as premature QRS complex of abnormal shape & duration. Hypokalemia & hypercalcaemia ⇧ toxicity of digoxin &discontinue digoxin if any new rhythm disturbance noted.

CHAPTER V

HAEMATOLOGY

- Anaemia
- Thalassemia
- Spherocytosis
- Sickle Cell Anaemia
- Bleeding Disorders
- Haemorrhagic Disease of New born
- Nasal Bleeding
- Hematemesis
- Henoch Schonlein Purpura
- Idiopathic Thrombocytopenia Purpura

ANAEMIA

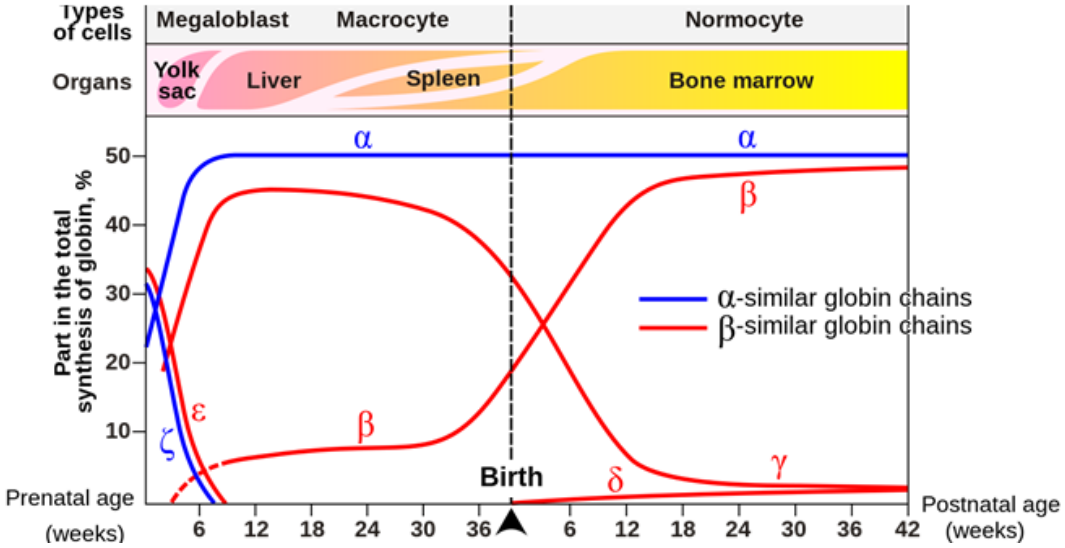
Anaemia definition

Deficiency of either erythrocytes or Hb or both in the circulating blood, this result in a reduced capacity to deliver O_2 to the tissues (⇩in the O_2 carrying capacity of the blood), leading to tissue hypoxia, producing symptoms such as weakn ess & shortness of breath. (An-without-emiablood).

Blood formation:

1- During the Intra uterine life; blood cells are formed in the liver & spleen up to the fifth month, after the fifth month the bone marrow share in the formation of these cells.

2- After birth; formation of these cells will be restricted to the bone marrow.



Normal life span of RBCs: 120 days. Normal size of RBCs: mature RBC is 7.8 micron across, 1.7 micron thick (8 X2 micron).

WHO criteria for anaemia: •Men ē Hb <13gm/dL & Hct <41 •Women ē Hb 〈12gm/dL & Hct 〈36

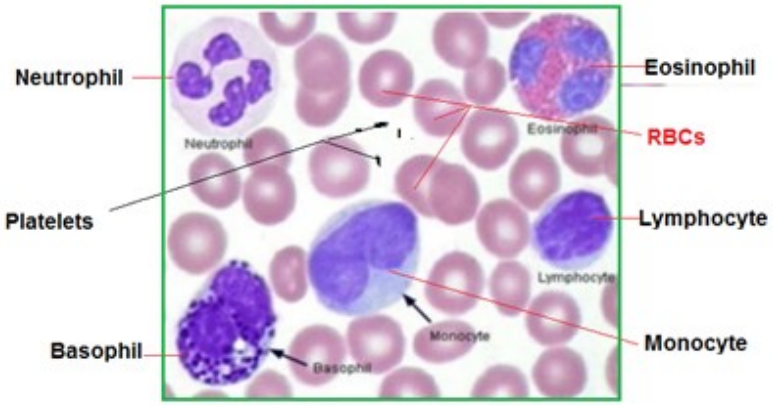
Normal Life Span of RBC: 120 days.

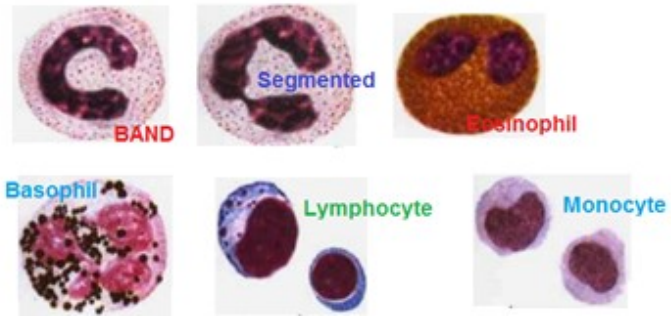
Normal Size of RBC: mature RBC is 7.8 micron across, 1.7 micron thick (8X2 micron).

Age & Normal level of Hb & MCV

Age	Hb (g/dl)	Lower MCV limit	Upper MCV limit
1-4	11.2	72	85
5-7	11.5	75	87
8-10	11.8	76	89
12-14	12	76	89
15-17	12 (F), 13 (M)	78	92
Over 18	12 (F), 14 (M)	80	95

Normal Peripheral Blood Smear





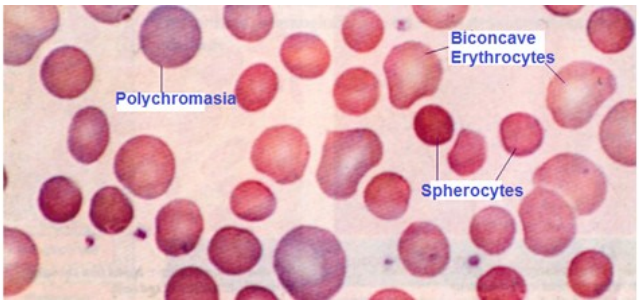
Abnormal Peripheral Blood Smear

RBC fragments (Schistocytes)	Microangiopathic haemolytic anaemia
Spherocytes	Hereditary spherocytosis
Hyper segmented polymorphs	Megaloblastic anaemia
Target cells	Hemoglobinopathies, Alcoholics
Tear drops cells	Myelofibrosis, Thalassemia
Howell jolly bodies	Splenectomy, Function asplenia
Sideroblasts	Myelodysplasia, Alcoholics
Burr cells	Uremic

Spherocytosis

Many small size RBCs & lack of the central zone of pallor

* Hereditary spherocytosis
* Autoimmune haemolytic anaemia



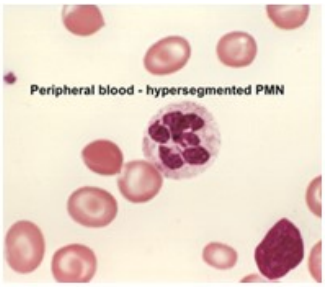
Hyper segmentation

Hyper-segmented polymorph nuclear cells

6 lobes

suggest Megaloblastic anaemia

(Vit. B 12, Folate deficiency)



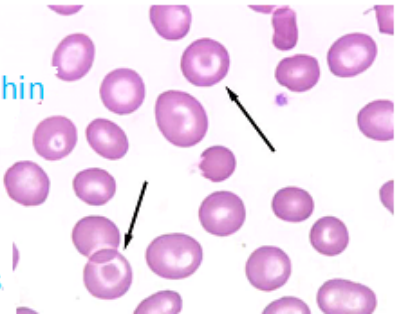
Target cells

Target cells

RBCs have an area of ⇧ staining appear within the area of central pallor,

seen in sickle cell anaemia, thalassemia,

post splenectomy, liver diseases, alcoholics



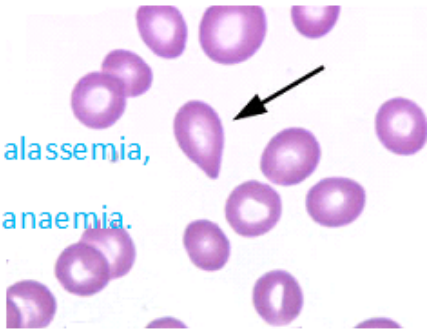
Tear drop cells

Tear drops cells

Seen in

Myelofibrosis, Myeloid metaplasia, Thalassemia,

Pernicious anaemia & some haemolytic anaemia.

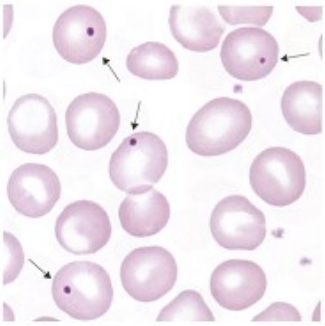


Howell jolly bodies

Howell jolly bodies

Seen in

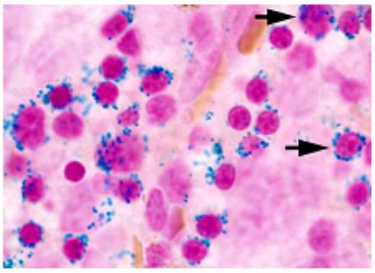
Splenectomy, Functional asplenia.



Sideroblasts

Sideroblasts

erythroblast that contains one or more aggregates of non hime iron appears asPrussian blue-stainable granules, seen in Myelodysplasia, Myeloid leukaemia, Malignancy, Rheumatoid arthritis, and Alcoholics

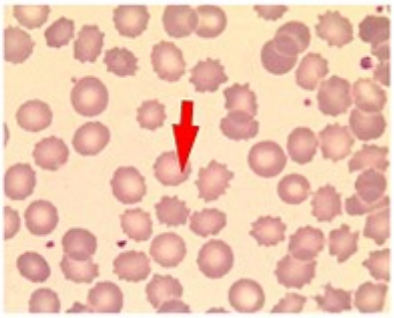


Burr cells

Burr cells

Peripheral blood film showing Burr cells,

seen in Uremic patient



Causes

(1) Physiological anemia

Common in the 1st & 2nd month of life. (no interference except if Hb level <7 gm/dl, Hct < 20 using Ferrous drops & in severe cases whole blood transfusion).

(2) Impaired production

a) Disturbance of proliferation & differentiation of stem cells, include; •Pure red cell aplasia •Aplastic anemia, affect all kinds of blood cells, Fanconi anemia is a hereditary disor-

der ē aplastic anemia & variable other abnormalities. Anemia of renal failure as a result of insufficient erythropoietin production. Anemia of endocrinal disorders; hypothyroidism. Chemotherapy, causing bone marrow suppression.

b) Disturbance of proliferation & maturation of erythroblasts, include; Pernicious anemia, is a form of megaloblastic anemia due to Vit B 12 deficiency. Folic acid deficiency. Iron deficiency anemia. Thalassemia. Anemia of renal failure (also causing stem cell dysfunction). Anemia of chronic inflammation. Myelophthisic anemia, is a severe type of anemia resulting from the replacement of bone marrow by other materials, such as malignant tumors or granulomas.

(3) ⇧ Destruction

A- Intrinsic causes; Defects of RBCs membrane: as in case of Hereditary Spherocytosis or Elleptocytosis. Defects of Hb: as in case of; Thalassemia, Sickle cell anemia. Hypersplenism, sequestration of blood (portal hypertension). Defects of RBCs metabolism as in case of G6PDD or Pyruvate kinase deficiencies. Paroxysmal nocturnal hemoglobinuria.

B-Extrinsic causes; Antibody mediated: Rh incompatibility. Transfusion reaction. Autoimmune hemolytic anemia; the immune system mistakes RBCs for foreign invaders & begins destroying them as in case of; SLE, CLL, Hodgkin's lymphoma, Rheumatoid arthritis. Drug induced. Mechanical trauma: Burns, Haemodialysis, Malaria, Microangiopathic hemolytic anemia as in case of; DIC, TTP. Hemolytic uremic syndrome. Heart surgery.

(4) Blood loss

A) Acute blood loss as result of trauma, surgery. Immediately after blood loss, the Hb is normal. After fluid replacement, the Hb ⇩ but the cells looks normal. Reticulocytosis start to appear after 2-3 days.

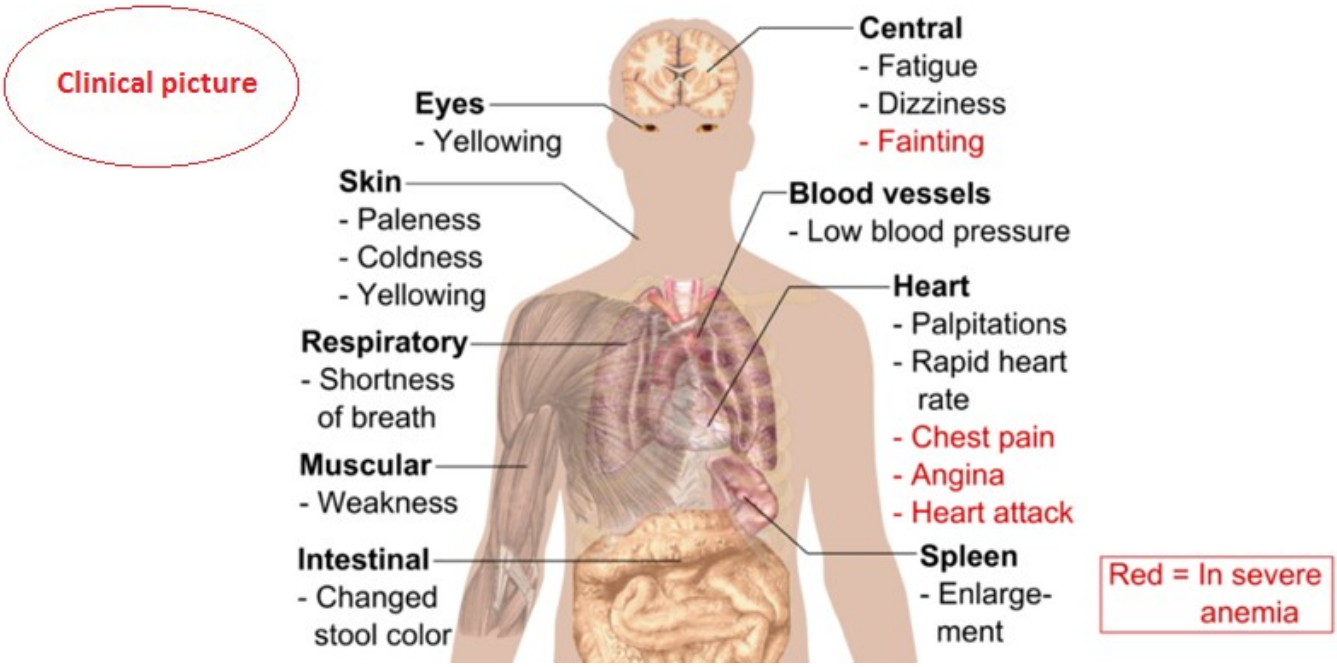
B) Chronic blood loss as a result of GIT Hge, piles, parasite infestation (nematodes, hookworms), gynecological disturbances, repeated blood sampling from premature, is different, as it cause iron deficiency anemia.

(5) Fluid overload

Hypervolemia cause ⇩ Hb conc & apparent anemia. Anemia of pregnancy is induced by blood volume expansion.

Clinical picture

•Pallor •Fatigue •Shortness of breath •⇩ BP •Tachycardia •Chest pain •Melena.



Diagnosis

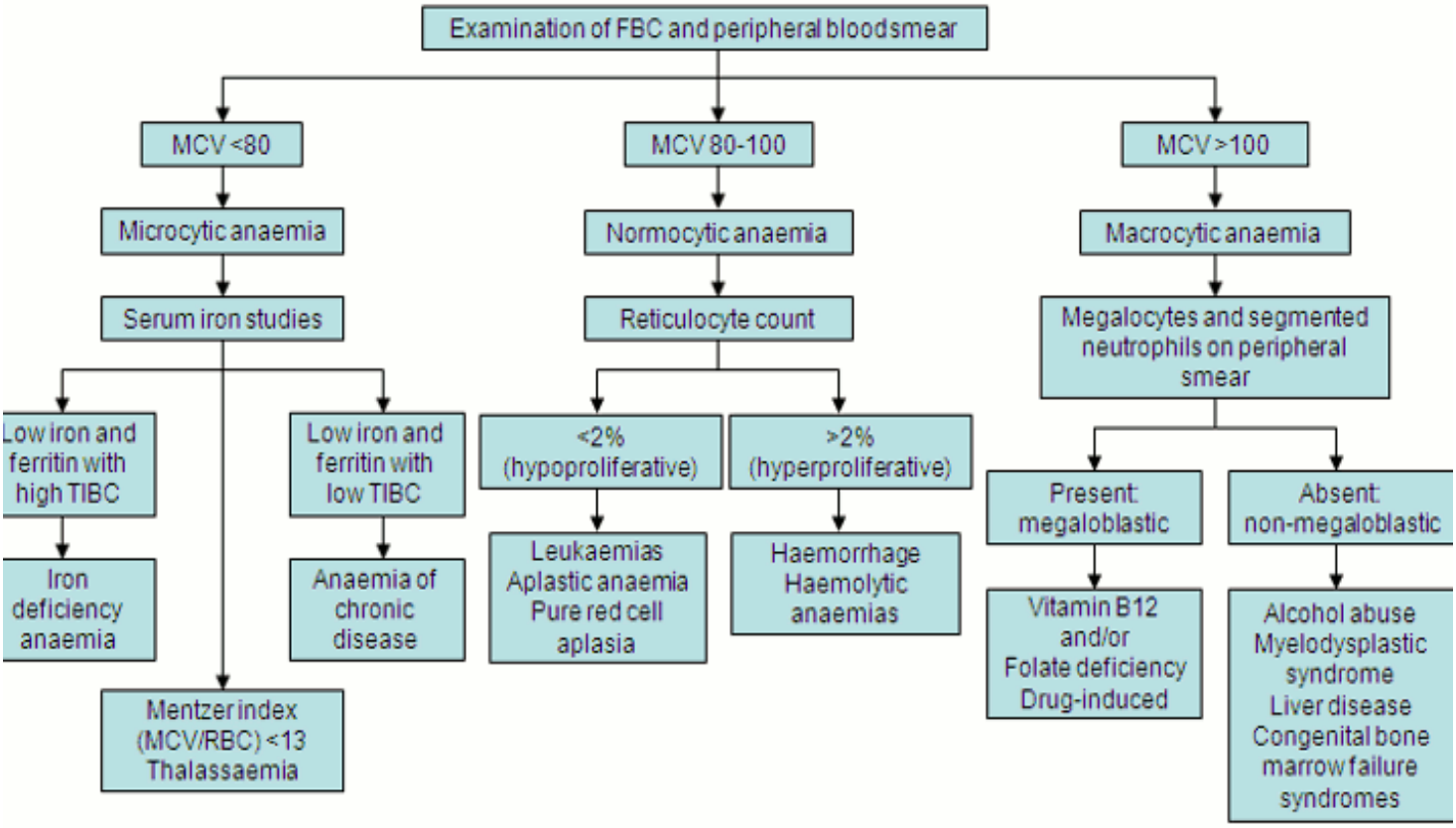
1-Determination of the morphological type: microcytic, hypochromic, normocytic normochromic, macrocytic, normochromic.

2- RBCs parameters: MCV, MCH, MCHC. RDW normally 12-17 is an index of RBC size variant, ⇧ ē iron deficiency, normal ē thalassemia & anemia of chronic disease.

3-Reticulocyte count: ⇧ >2-3% or 100,000/mm^3 total, seen in blood loss & hemolytic processes, although up to 25% of hemolytic anemia will present ē normal Retic count

due to immune destruction of RBC precursors. Retic counts are most helpful if extre-mely low < 0.1%, or > 3% (100,000/mm^3 total).

Anemia work up



IRON DEFICIENCY ANEMIA

Iron absorption: iron is absorbed in proximal small intestine.

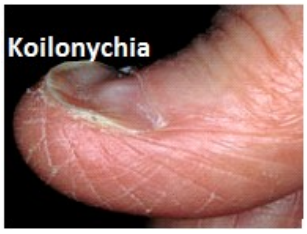
Causes of iron deficiency anemia

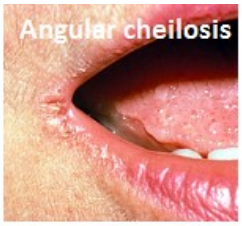
Malnutrition. Pregnancy/lactation. Normal growth. Blood loss. Intravascular hemoly sis. Chronic Hge (piles). Malabsorption. Pica (tendency to eat, clay, crunchy materials)

Clinical picture

Pallor: skin, conjunctiva. Weakness. Shortness of breath. Koilonychia (known as spoon shaped nails, seen also in B12, or folic acid deficiencies). Angular cheilosis (bilateral fissu-ring at corners of mouth). Atrophic glossitis (red large swollen tongue, seen also in B12

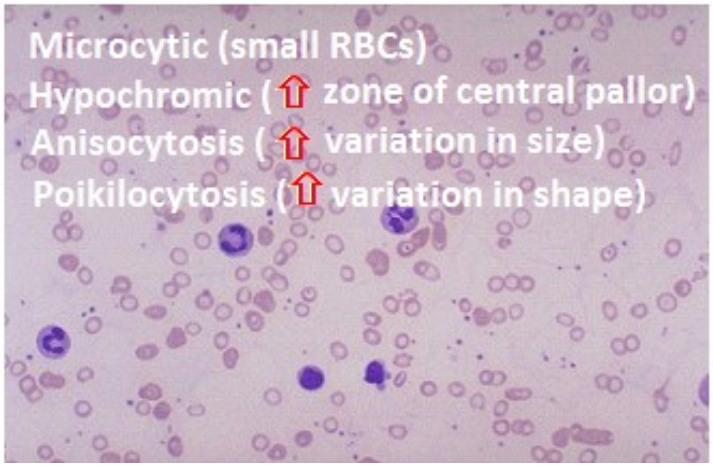
or folic acid deficiencies).





Investigations

- CBC: Hb, Hct.
- Blood smear: microcytic hypochromoc anaemia (MCV, MCH, MCHC, RDW, Retic).
- SI: ⇩ (< 60 microgram /dL).
- TSIBC: ⇧ (>360 microgram/dL).
- Serum ferritin: ⇩ (<20 nanograms/mL), can be falsely normal ē inflammatory state.



Management

Iron drops 15 mg/dropper, 6 mg/Kg/day÷2 (therapeutic dose), side effects include; constipation, black stool, +ve hem occult test # Iron syrup (50 mg/tsp) # Iron-Folic acid tab (iron 100 mg + folic acid 350 mcg). # Theragran hematinic syrup (66 mg iron/tsp) is multivitamin ē iron. # Vit C: 35 mg/day, drops 100 mg/1 ml, facilitate iron absorption.

After 10 days from starting Rx the SI start to ⇧ 0.1 mg/day, repeat investigation after one month & continue Rx for 6 months.

MEGALOBLASTIC ANAEMIA

Result from defective synthesis of deoxyribonucleic acid (DNA) in all proliferating cells, most commonly result from lack of Vit B12, or Folic acid.

PERNICIOUS ANAEMIA

Failure of secretion of intrinsic factor from parietal cells of gastric mucosa as a result of atrophic gastritis in old age (pernicious anemia is a disease of elderly 5th-8th decade), or in younger age secondary to gastrectomy (total or partial), also may be associated ē autoimmune diseases & the formation of antibodies to intrinsic factor in diseases like Hashimoto, Graves, Vitiligo, DM, Myasthenia gravis. Another causes are parasite infestation (fish tape warm- Diphyllobothrium latum), malabsorption, crohn`s disease of small intestine, ileal resection (as Vit B12 after combination ē intrinsic factor will be absorbed from the ileum) & deficient dietary intake of Vit B12. Pernicious anemia may associated ē genetic predisposition & pt have ⇧ risk of gastric cancer.

Clinical picture

Manifestations of anemia as mentioned above, in addition to neuropsychiatric symptoms; spastic ataxia, psychosis, loss of vibratory sense, dementia.

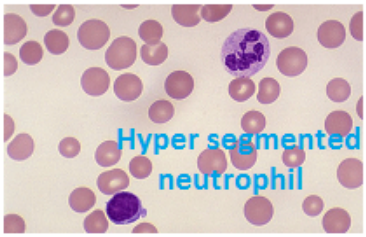
Diagnosis

- CBC: ⇩Hb, Hct & RBCs count. MCV >100 fl. MCH ⇧. MCHC normal. Reticulocytopenia (striking reticulocytosis after parenteral administration of Vit B12). Normal or low WBCs count. Normal or low platelet count.
- ⇩ Level of serum B12.
- ⇧ level of serum homocysteine.
- Serum antibodies to intrinsic factor & anti parietal cells.
- Abnormal schilling test, oral labeled B 12 & IM unlabeled B12 at the same time, 24hour

urine to assess absorption (diagnosed by checking antibody levels rather than schilling test).

Blood smear

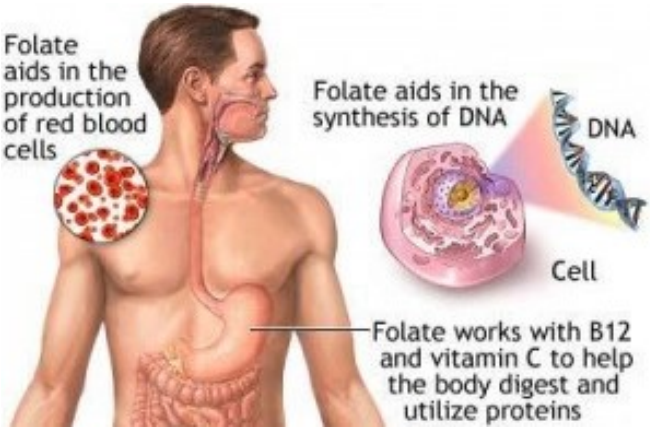
Macrocytosis
+
Hypersegmented neutrophil
(6 lobes instead of the usual 3-4 seen in B12 def. or Folic acid def.)



Management

Hydroxyl Cobalamin, initial dose 1000 mcg of IM/day for 1 wk, maintenance dosage 1000 mcg IM/3 months.

FOLIC ACID DEFICIENCY ANAEMIA



Folic acid is necessary for RBCs production & neural tube formation in embryo, repe-ated studies have shown that woman who get 400 mcg daily prior to conception & during early pregnancy reduce the risk that their baby will be born ē a sever NTD (spina bifida, anencephaly, encephalocele), these defects occur during the first 28 days of pregnancy-usually before a woman even know she`s is pregnant. Folic A nor-mally absorbed in duodenum & proximal jejunum. Deficiency often occur ē ⇩ oral in-take of Folic acid, or ⇧ demands, or ⇧ utilization or impaired absorption. Found in malabsorption, coeliac disease, regional enteritis, amyloidosis, alcoholics because enzyme required for degluta-

mation of folate is inhibited by alcohol. Often found in pregnant women & in case of desquamating (exfoliative) skin disorders & in pts ē sickle cell disease. Daily requirements of folic acid ⇧ in case of hemolytic diseases & exfoliative skin diseases. Normal daily requirements of folic acid is 400 mcg/day

Clinical picture

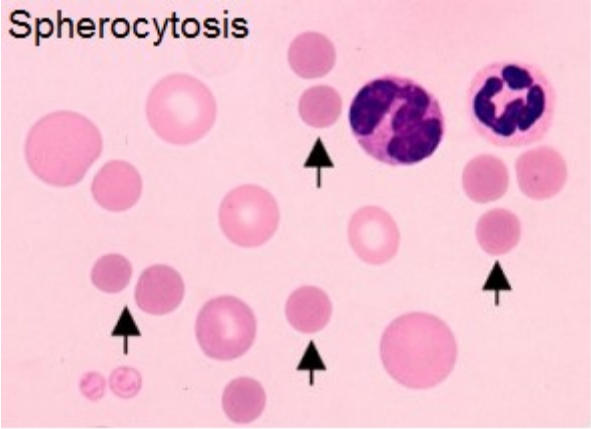
Symptoms similar to vit. B12 deficiency, but nervous system function remain normal.

Diagnosis

- CBC & Blood smear.
- ⇧ Serum Homocysteine.
- Low folate.
- Normal Methylmalonic acid.

Management: Folic acid 5 mg tablet /day.

SPHEROCYTOSIS

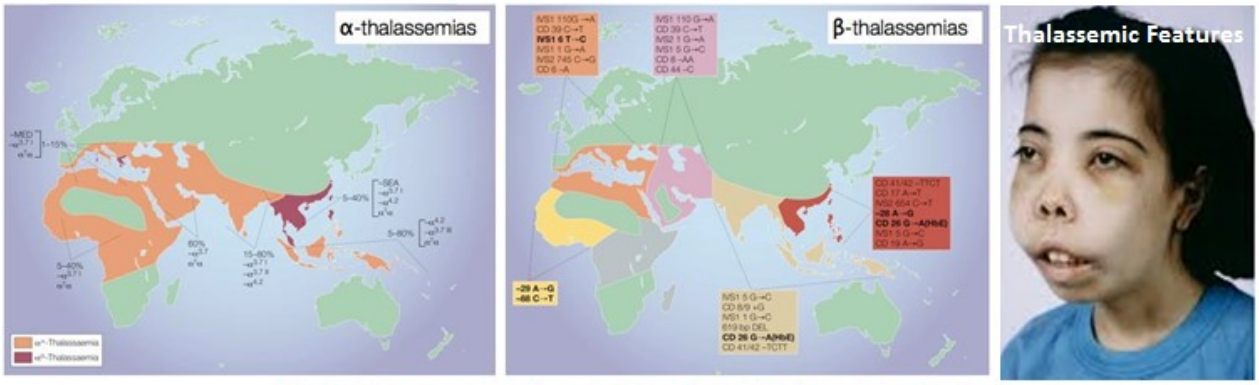


In hereditary spherocytosis, there is a lack of spectrin, a key RBC cytoskeletal membrane protein. This produces membrane instability that forces the cell to the smallest volume. The spherocytes do not survive as long as normal RBC's. This is shown by the ⇧ of osmotic fragility & blood smear.

BETA THALASSEMIA

Chromosome 11, Gene HBB,Location 11 P 15.5

In Greek Thalassic (Sea) & Emia (Blood), described in 1932.



Defective production in B chain of haemoglobin.

Incidence: prevalent in Mediterranean people ē highest incidence in Cyprus & Maldives where the carrier rate is 18% of population.

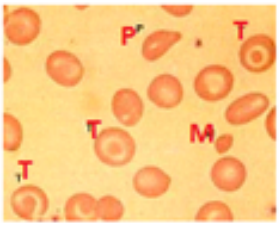
Genotype	Clinical presentation	Hb A (%)	Hb A2 (%)	Hb F (%)
B^+ / B	B-Thalassemia Minor or Normal	95	3.5-7	0-6
B^0 / B	B-Thalassemia Minor	95	3.5-7	0-6
B^+ / B^+	β-Thalassemia Intermedia	25-65	1-4	30-70
B^0 / B^0	β-Thalassemia Major	0	1-4	>95
B^0 / B^+	β-Thalassemia Major or Intermedia	< 20	1-4	>75

Clinical Picture

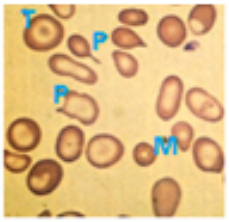
•Pallor. •Symptoms started by age 6 months. •Mongoloid features, prominent check bone (expansion of marrow cavity of bones of skull & face, protrusion of upper jaw from extra medullary erythropoiesis). •Growth retardation. •Deposition of iron anywhere in the body (sidrosis). •Hepatosplenomegaly. •Poor prognosis & children die before adolescency.

Diagnosis

▲Hb electrophoresis: ⇧ Hb F 80-90% by age 3 months + ⇩Hb A (normally Hb F is <10 % by age 3 months & Hb A represent 90%) ▲CBC: severe microcytic hypochromic an-emia, target cells, reticulocytosis, leucopenia & thrombocytopenia ▲⇧ Serum iron ▲Carrier state: Hb A_2 > 3.5 %, Hb F is zero & HbA is normal.



Peripheral blood smear in Beta-Zero Thalassemia Minor showing Microcytes (M), Target cells (T) & Poikilocytosis(P)



Peripheral blood smear in Beta-Zero Thalassemia Major showing more Anisop-oikilocytosis(P), Target cells (T), marked microcytosis(M), prominent hypochromia.

Management

✱keep Hb >10 gm/dl, packed red cells transfusion, nearly will need 2 units/month (Normal Hb-pt Hb) X BW X 3.5 (or 10-15 ml/Kg). One unit packed cells ⇧ Hb level 1 gm/dl.

✱Iron chelation: Desferoxamine ampule 500 mg, 20 mg/kg IV over 5 hrs, diluted ē glucose 5% 50 ml or IM daily for 5 days/wk.

✱Vit C tab or effervescent daily 1 X 1. ✱Folic acid 5 mg tab/day.

✱Splenectomy: massive splenomegaly interfering ē breathing , post splenectomy im-munization by Pneumovax & Meningovax (usually required by age of 8 yr).

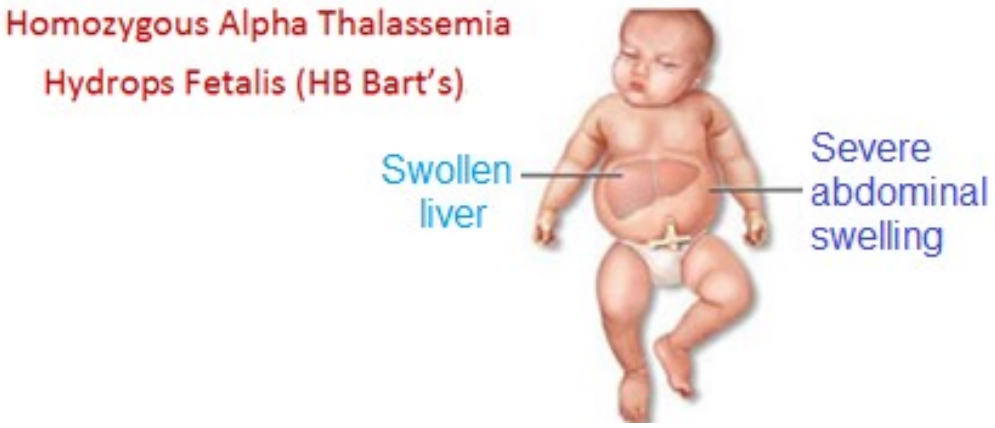
✱Genetic induction of Hb A by Erythropoietin amp 4000 U, twice weekly for one yr.

✱Bone marrow transplantation, curative, highly specialized centers.

✱Premarriage counseling & public awareness.

ALPHA THALASSEMIA

Chromosome 16, Genes HBA1and HB A 2, Location 16 p 13 - 3.



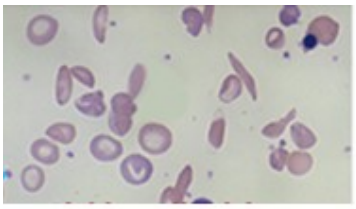
- Defective production of α chain of Hb
- Human cells contain 2 copies of Hb A_1 & 2 copies of Hb A_2, by means that the disease is under control of 4 genes.
- Absence of the 4 genes (Homozygous α Thalassemia) result in production of Hb Bart's (Hydrops Fetalis) ẃ is incompatible ē live.
- Absence of the 3 genes (Heterozygous α Thalassemia) results in production of Hb H.
- Absence of the 2 genes (α Thalassemia trait) result in microcytic hypochromic anemia.
- Absence of 1 gene (α Thalassemia Silent) is asymptomatic.

Genotype	Clinical presentation		Hb A %	Hb A2%	Hb F %	Hb H%	Hb Bart%	Hb Portland%
αα / αα	Normal		95	< 3	1.5			
-α / αα	Silent Trait (α^+ Thalassemia)	Silent Carrier (Thalss. 1)	95	2-3.5	1.5– 3	Traces	0 - 3	
-α / -α	α- Thalass Minor (α^0 Thalassemia)	Carrier (Thalss. 2)	Slightly ⇩	1.5-3.5	1.5 –3	<2	2 - 8	
-- / -α	Hb-H Disease (α^0 / α^+) Thalassemia	(Thalss. 3)	⇩	1-2	<2	2-40	< 5	
-- / --	Hb Bart`s (α^0 / α^0) Thalassemia	Die during Pregnancy or After labour	Absent or Trace	Absent or Trace	Absent or Trace	<5	70-80	10-15

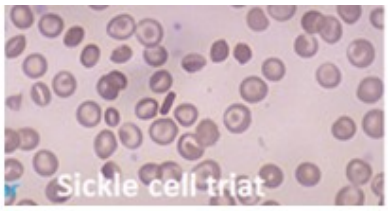
SICKLE CELL ANAEMIA

Chromosome 1, Gene HBB, Location 11 p 15.4

Discovered by, Cardiologist Dr .James B. Herik, USA, in 1904.



RBCs change shape to long, thin sickle forms that sludge in capillaries



Presence of " target cells"- RBCs with central reddish dot. The rectangular RBCs is indicative of Hb crystals

Autosomal recessive disorder, it is abnormality in the shape of RBCs. Usually started by age 6 months ē the replace- ment of Hb F by Hb SS or Hb SA. (Normally at birth, about 80 % is HbF & 20 % is HbA & by age 3 months, about 90 % is HbA & 10% is HbF). In sicklers Hb is either: Hb SS 80 - 90% (sickle cell anemia) or Hb SA 50% S &50% A (sickle cell trait). In sickle cell crisis, the abnormal Hb SS is prone to crystallization when O_2 tension is low, the RBC's change shape to long, thin sickle forms that sludge in capillaries, further decreasing blood flow & O_2 tension. Pt ē sickle cell trait (Hb AS) are much less likely to have this happen. Pt ē Hb SC, both Hb S & Hb C are present, the RBC's may sickle, but not as commonly as ē Hb SS disease. The Hb C leads to the formation of “target cells”-RBC's that have a central red-ish dot & Hb C crystals ẃ is also characteristic for Hb C disease.

Incidence: 75 % of cases occur in Africa, where carrier rate about 10-40%

Clinical picture

Commonest presentation is vasoocclusive crises affecting hands & foot (Dactylitis) present as painful, symmetric swelling of hands & feet or recurrent painful episodes of Abdominal Pain from affection of any internal organ, crises occur nearly on daily basis, may affect kidney, spleen, may cause Priapism, or acute chest pain, may affect brain

(cerebrovascular occlusion)-stroke- or affect the retina- Angioid Streaks. The pt is very susceptible to infection, especially malaria in endemic areas, so we give him an antimalarial drugs in daily basis.

Diagnosis

☞Hb electrophoresis: detect presence of Hb SS, or AS.

☞CBC: severe normocytic normochromic anemia (Hb 5-7gm/dl), reticulocytosis, neutrophilia & thrombocytosis.

☞Blood smear: target cells, Hb C crystals (rectangular RBCs), Howell jolly bodies ẃ may indicate hyposplinism, anisocytosis, poikelocytosis.

☞ESR: ⇩ sickle cells fail to form rouleaux.

☞ Sonar Abdomen: may show evidence of internal organ damage.

☞ CT scan Brain: may show multiple tinny infarcts.

☞Liver/Renal function tests: evidences of organ damage.

☞ Fundus examination: may show Angioid Streaks.

Management

☝ Avoidance precipitating factors as fever, dehydration, hypoxia, acidosis &infection.

☝ IVFs: over hydration using 150 % of the maintenance daily requirements.

☝ Analgesics: Morphia, Codeine, Aspirin for those > 5 years (to avoid Reye`s sy), Paracetamol drops 100 mg/dropper, syrup 250 mg, maximum total daily dose is 1200 mg.

☝Prophylactic antibiotics ☝Vit. C & Folic acid daily requirement.

☝ Vaccination: the routine vaccination + pneumococcal vaccine.

☝ Blood transfusion: if Hb < 5 gm, using whole blood = (Normal Hct - Pt. Hct) ÷ Donner Hct) X Blood volume (BL. Vol.= B.Wt. X 80 ml).

☝ Hematopoietic cell transplant: curative.

GLUCOSE 6 PHOSPHATE DEHYDROGENASE DEFICIENCY

X Chromosome, Gene G6PD, Location X q 28

Incidence: Common in Negros & Middle East. Occur in 11-13% of African Americans. Estimated 400 million people worldwide carry the gene. G6PDD is significant cause of mild to severe jaundice in newborns.

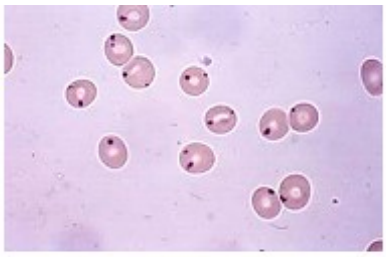
Result from break down of RBCs, ẃ can be triggered by infections, severe stress & certain foods. Episodic hemolysis from **fava beans**, oxidant drugs as; Primaquine, Sulpha, Amiodarone, Antimalarial, Nitrofurantoin, Antihistaminics, Antituberculous, Aspirin.

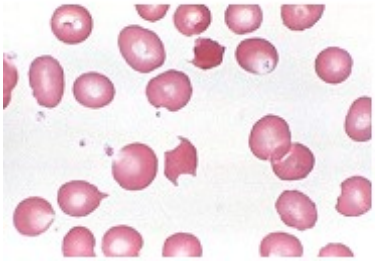
Presentation: Anemia. Fatigue. Tachycardia. Shortness of breath. Dark urine. Splenomegaly. Notes**:** pts are less susceptible to malaria.

Diagnosis

- ⊙ CBC (macrocytic anemia, reticulocytosis) & Blood Smear (Henz bodies, bite cells).
- ⊙ G6PD level is diagnostic, usually paradoxically normal during hemolytic episode.
- ⊙ ⇧Serum bilirubin.

Blood smear- G6PD, showing Heinz Bodies (denatured haemoglobin).





Blood smear- G6PD, showing Bite cells.
Prussian blue staining (detects hemosiderin)

Differential Diagnosis

-Sickle cell disease (painful crisis).

-Pyruvate kinase deficiency (hemolysis not precipitated by drugs or infections).

Management

- Avoidance of precipitating factors.
- Gradually improve by age.

BLEEDING DISORDERS

Normal clotting mechanism

Body response to injured blood vessel by the following mechanisms:

1- Immediate vasoconstriction occur (release of serotonin, histamine, PG etc..) ẃ causes vasoconstriction of the micro vascular bed).

2- Platelet plug (need intact, functioning platelet & VWF to binds damaged vessel).

3- Functioning clotting factors to activate clotting cascade ē generation of fibrin clot.

4- Fibrinolysis (clot breakdown).

Platelets

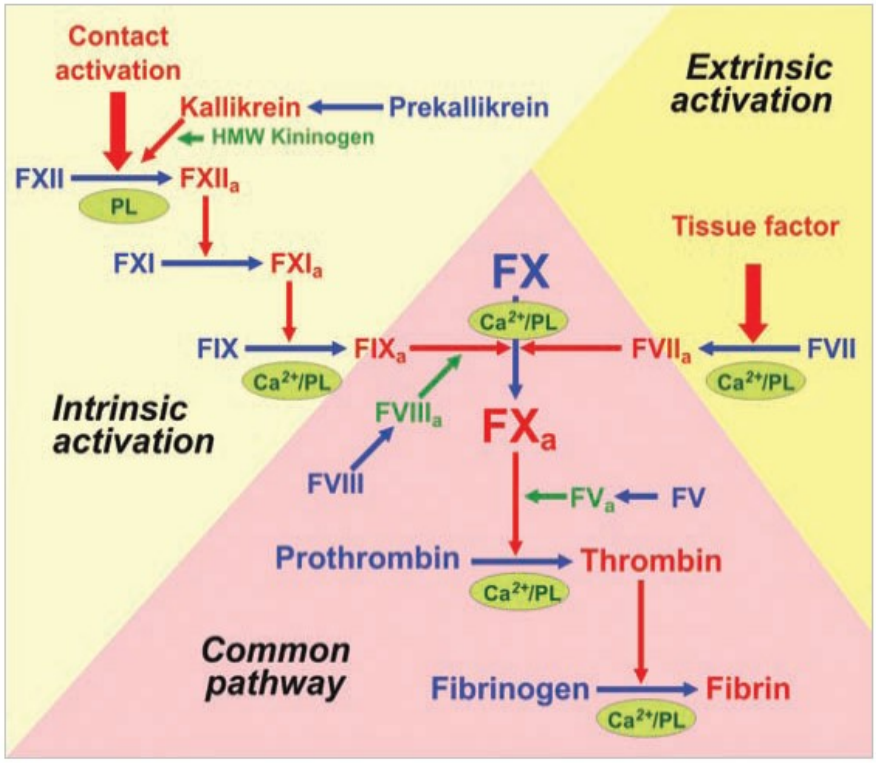
Is very small cell ēout a nucleus, size 2-3 micron, its life span 8-10 days & its normal reference value 150.000-400.000/cmm, platelet cell contain many receptors on its surface ẃ combine ē the clotting factors during the process of hemostasis.

Clotting

Is the function of platelet, when it aggregate & adhere to the site of blood vessel injury forming platelet plug (primary hemostasis).

Coagulation

Is the process by ẃ blood change from a liquid to a gel by deposition of clotting fact- ors to fill the micro spaces between the deposited platelets causing cessation of blo-od loss from a damaged vessel. Its chain reaction, if one of the factors is missing this chain reaction cannot proceed.



The extrinsic pathway

Once tissue damage occur, it liberate tissue factor ẃ combine to factor FVII & in the presence of calcium & phospholipid, factor FVII activated to form active $FVII_a$, the tissue $FVII_a$ complex start the common pathway by activation of FX. The efficacy of extrinsic pathway measured by PT.

The intrinsic pathway

Include activation of factors FXII, FXI, FIX) in addition to FVIII. The efficacy of intrinsic pathway measured by PTT.

The common pathway

Activation of factors I, II, V, X. It end by formation of fibrin clot.

Causes

Vascular abnormalities

Hereditary hemorrhagic telangiectasia. Ehlers Danlos sy. (Hereditary). Senile Purp-

ura. Henoch-Schonlein sy. Scurvy, when Vit C fall < 10 mg/dl, Vit C is essential for synthesis of collagen. Steroids. Cushing sy. Amyloidosis.

Thrombocytopenia

Pt usually presented ē petechiae (do not blench ē pressure), ecchymosis, purpura, endothelial bleeding (nasal bleeding, menorrhagia). Idiopathic Thrombocytopenic Purpura or Thrombotic Thrombocytopenic Purpura. Heparin Induced Thrombocy-topenic Purpura. Hemolytic Uremic Sy (⇧ destruction). Autoimmune: SLE, HIV infecti- on (⇧ destruction). Disseminated Intravascular Coagulopathy (⇧ destruction of plat-elets, prolongation of all clotting tests & ⇧ fibrin degradation products). Hyperspleni-sm. Severe infection. Leukemia. Lymphomas. Malignancies.

Thrombocytopathy

Normal platelet count but abnormality in platelet function, shape, adhesion, aggreg-ation as in Bernard syndrome (AR, abnormality in platelet membrane glycoproteins, abnormal platelet adhesion, prolonged bleeding time + normal PT, PTT). Glanzman sy (AR, abnormality in platelet membrane glycoproteins + abnormal platelet aggregation. Gray sy (AR, large cell, stain gray ē Wright`s stained smear).

Factor deficiencies

Hemophilia A (classical hemophilia represent 85% of all hemophiliacs, deficiency of FVIII, incidence 1/10.000 males). Hemophilia B (Christmas disease, deficiency of FV IX). Hemophilia C (deficiency of FV XI). Von Willebrand`s disease (deficiency of VWF & amount of FVIII. VWF released from endothelial lining of blood vessels & from platelets.

Vit K deficiency (deficient intake or defective formation by intestinal bacteria, or malabs-orption, it is essential for the formation of Vit. K dependent factors 10, 9, 7. Liver irrhosis (all clotting factors are produced by liver except FVIII & VWF). Severe infection. DIC.

Other causes

Oral anticoagulants (Warfarin, Heparin). Broad spectrum antibiotics.

Clinical features of bleeding disorders

	Platelet disorders	Factor deficiencies
Site	Skin, MM (epistaxis, gum, GIT, vagina)	Deep in soft tissues (joints ,muscle)
Petechiae	Yes	No
Ecchymosis	Small, superficial	Large, deep
Hemarthrosis	Extremely rare	Common
Bleeding after cuts	Yes	No
Bleeding after surgery	Immediate (usually mild)	Delayed 1-2 days (often severe)

Investigations

*CBC.

*Platelet count: moderate thrombocytopenia 50.000-100.000/cmm. Severe thrombocytopenia < 50.000/cmm).

*Platelet morphology .

*Bleeding time: reflect efficiency of platelets & integrity of endothelial cells of blood vessels (normal reference value is 1-8 minutes).

*Prothrombin Time: measure the effectiveness of the extrinsic pathway, FVII, FX (normally 10-15 sec.).

*Partial Thromboplastine Time: measure the effectiveness of the intrinsic pathway, FXII, FXI, FIX, FVIII (normally 25-40 sec.).

*Thrombin Time: measure the time for prothrombin “F II” to convert fibrinogen to fibrin, (normally 9-13 second).

HEMORRHAGIC DISEASE OF NEWBORN

Clinical picture

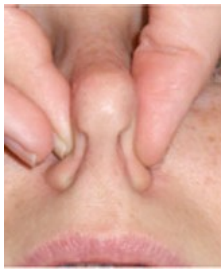
Early type occur during 1st wk of life, late type occur during 3rd- 4th wk of life, bleeding either from umbilical cord or hematuria, due to deficiency of Vit K dependent factors (II, III, IX, X) as a result of absence of gut bacterial flora ẃ produce vit. K in the sterile gut of neonate & the fact that vit. K not crossing the placenta or excreted through breast milk.

Investigations: prolonged PT & APTT.

Management

Konakion amp 0.2mg/ml or 10 mg/ml, 1mg IM, as prophylactic to every baby at birth.

NASAL BLEEDING



Investigations

- CBC.
- Coagulopathy profile.
- Serum creatinine.
- LFTs.
- Rarely may need MRI of head or C-T scan nose & sinuses.

Management

#Head chest position, compression of nose by fingers for 5-10 minute.

#Anterior nasal pack using strip of cotton or gauze 10-12 cm length, soaked in adrenalin

1/10.000 concentration.

#Posterior nasal pack by NET specialist.

#Cauterization or surgical ligation of bleeding vessel.

HEMATEMESIS

Causes

Bleeding disorders as hemophilia, VWD, Christmas disease, sickle cell disease, thrombocytopenia, gastritis, peptic or duodenal ulcer, portal hypertension, liver disease, Biliharziasis, Mallory Wiss Syndrome.

Investigations

- CBC.
- Coagulation profile (BT, CT, PT, APTT).
- Platelet count.
- LFTs.
- Kidney function; urea, creatinine.
- Urine & Stool analysis.

Management

Monitor BP. # Pt may need anti shock measures.

NPO & start IVFs (Ringer`s).

Stomach wash ē iced saline 50 ml, leave for 2 min, repeat/5 minutes for 30 minutes.

Frequent gastric suction /½ hr.

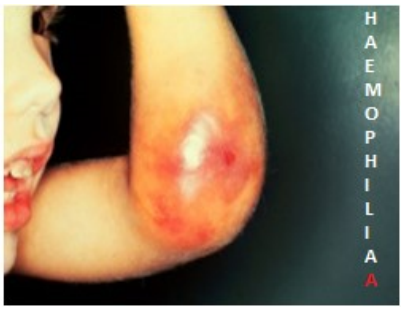
Zantac amp. 50 mg IM /12 hrs (antacid).

Dicynone amp. 250 mg, IM, 2 amp stat then 1 amp./6 hrs until bleeding stop (act on capillaries, ⇧ it's resistance).

Endoscopy is diagnostic.

HEMOPHILIA A

X Chromosome, Gene F 8, Location X q 28



Incidence: 1/5000 male Births. Impairment of body ability to control blood clotting, stopping bleed when blood vessel is broken, due to deficiency of Factor VIII.

Clinical picture

⊙70% of cases not bleed from circumcision during the 1st year of life.

⊙Child is easily traumatized, bruising especially in knees, elbows when baby start to crawl (6-9 months age).

⊙Repeated Hemarthrosis is common & bleeding may be internal if FVIII < 5%, may cause damage to any internal organ & tissue & may be life threatening.

⊙In NN period baby may presented ē IC Hge as FVIII does not cross the placenta during pregnancy.

Investigations

- Normal coagulation profile except prolonged APTT ẃ measure the intrinsic pathway of coagulation (common pathway + Factors I, II, V, X) + Factors VIII, IX, XI, XII), it rises to the double of the normal value.
- Low level of factor VIII is diagnostic. With Mild hemophilia the F VIII level is 5 -50% of normal. Moderate hemophilia when F VIII level is 1-5% of normal. Severe hemophilia when the VIII is < 1% of normal. Internal bleeding usually occur when F VIII level is < 5%. In case of VWD, the BT is prolonged (due to the associated thrombocytopenia).

- The mother of the hemophilic child usually have low factor VIII (30-70%).

Management

- Prophylactic measures for avoidance of trauma.
- In normal situations we aim to ⇧ FVIII level to 30- 40%, but ē major operation we have to ⇧ it to 100%.
- Fresh plasma, ⇧ FVIII up to 5%, 10-20 ml/Kg/12 hrs.
- Cryoprecipitate, ⇧ FVIII up to 25% (contain FVIII 100 U + fibrinogen) 1 bag/5 Kg BW.
- FVIII conc. infusion, ⇧ FVIII up to 100%. The dose of F VIII conc. calculated as: Number of units desired to ⇧ FVIII to the level needed ÷ 2 X BW (Kg). OR 20 units/Kg twice daily, (each bottle of FVIII labeled ē the number of units it contain).
- Post infusion level FVIII.
- No aspirin or antihistaminic.
- Measuring FVIII antibodies & 40 days to be passed between repeated FVIII transfusions or until all antibodies disappeared.

VON WILLEBRAND DISEASE

Chromosome 12, Gene VWF, Location p13.3

Named after Dr. Erik Adolf Von Willebrand, a Finnish paediatrician in, 1926

Classification

A- Quantitative deficiency of VWF.

<u>Type 1:</u> Partial quantitative deficiency of VWF.

Type 2: Virtually complete deficiency of VWF.

B- Qualitative deficiency of VWF

<u>Type 2A:</u> Qualitative variants ē ⇩ platelet dependent function associated ē the absence

of high & intermediate molecular weight VWF multimers.

Type 2B: Qualitative variants ē ⇧ affinity for platelet GPIb.

Type 2M: Qualitative variants ē ⇩ platelet dependent function not caused by absence of high -molecular weight VWF multimers.

Type 2N: Qualitative variants ē markedly ⇩ affinity for factor VIII.

Incidence: 1/100 population

Clinical picture

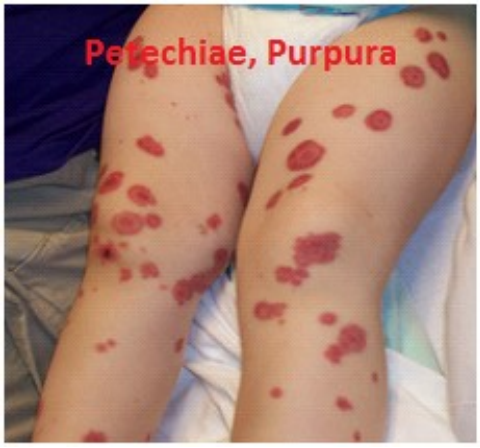
Presented ē epistaxis. Mucocutaneous bleeding. Skin bleeding. Women may experien ce heavy menses or excessive blood loss during labor. ⇩ VWF & F VIII. Prolonged BT, APTT. Thrombocytopenia. Abnormal platelet aggregation. .

Management

Desmopressin (1-deamino-8-d-arginine vasopressin) is a synthetic analog of vasopressin originally designed for the Rx of diabetes insipidus. DDAVP cause⇧ of VWF & FVIII plasma concentrations in pts ē mild hemophilia A &VWD by provoking release of stored VWF. DDAVP is cheap & carries no risk of transmitting blood-borne viruses. DDAVP (Emosint®, Minirin®) is usually administered SC, when a concentrated formulation is available, or IV at a dose of 0.3 µg/kg (for IV administration DDAVP is diluted in 50-100 mL saline & infused over 30 min). This treatment ⇧ plasma VWF-FVIII levels 2-4 times above the basal levels within 30-60 min. In general, hemostatically useful VWF-FVIII levels are measured in plasma for 6–8 hr. Infusions can be repeated every 12–24 hr depending on the type & severity of the bleeding episode. The drug is also available as an intranasal spray (Octostim®) ẃ can, however, result in variable adsorption ē a lower ⇧ in FVIII/VWF. DDAVP is usually effective in pts ē type 1 VWD & baseline VWF & FVIII levels > 10 U/dL, while in other VWD types there is significantly less response toDDAVP.

In type 2B, DDAVP is contraindicated because of the transient appearance or aggravation of thrombocytopenia leading to an ⇧ risk of bleeding, although a few pts have clinically benefited from its use. Pts ē type 3 VWD are unresponsive to DDAVP. Tachycardia, headache & flushing are frequent, mild adverse-effects of DDAVP & can often be attenuated by slowing the rate of infusion or by using the SC route. Tachyphylaxis (the progressive reduction in responsiveness after repeated treatments) is the main limitation to the use of DDAVP & should be consid- ered when repeated doses are anticipated. Hyponatremia & volume overload due to the antidiuretic effect of DDAVP occur rarely, but small children who have received closely repeated infusions are particularly at risk. To avoid this complication, fluid intake should be limited during DDAVP treatment. Finally, this drug should be used cautiously in pts ē uncontrolled hypertension, recent myocardial infarction or stroke, or suffering from angina, as thrombotic events have been reported to occur following its use. DDAVP can also be safely used at the time of parturition in responsive VWD women ē low FVIII C & VWF RCo levels, & it has been safely used in the first trimester of pregnancy to cover invasive procedures such as villocentesis & amniocentesis. The replacement therapy: those pts in whom a test infus- ion ē Desmopressin is not able to achieve clinically useful FVIII &/or VWF levels are candidates for replace- ment therapy. Purified, viral inactivated, plasma derived VWF/ FVIII products are most frequently used nowadays for VWF replacement therapy. The quantity of the ristocetin cofactor activity (VWF: RCo) in comparison to the F VIII C content varies by product.

HENOCH SCHONLEIN PURPURA



Unknown etiology. More in girls 5-15 years. Characterized by abdominal pain + purpura of lower limbs + Nephritis. Usually started as urticaria ẃ fades, replaced by purpuric rash, symmetrical in both lower limbs.

Investigations

#CBC.

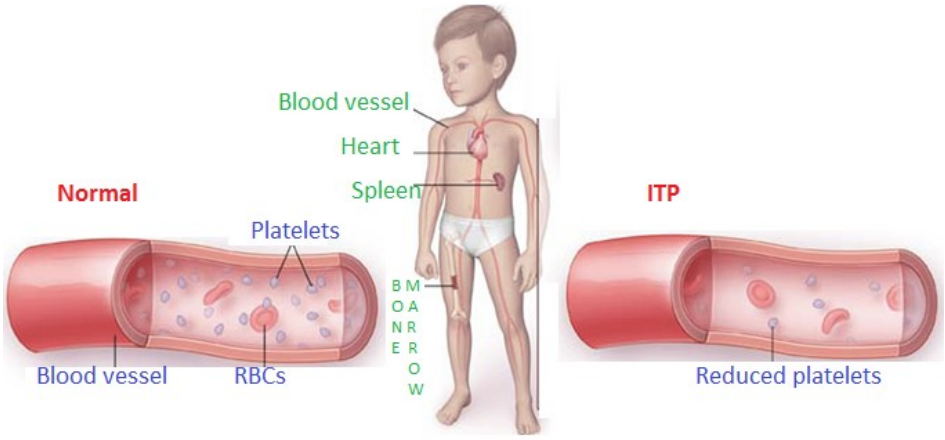
#Coagulation profile tests: all are normal (BT, PT, APTT).

#Urine: microscopic hematuria.

Management

- No specific treatment.
- Prednisolone tablet 5 mg 1X3 for 5 days, then 1X2 for 5 days, then 1X1 for 5 days.
- Admission to hospital for few days, as pt susceptible to GIT Hge, intussusception.

IDIOPATHIC THROMBOCYTOPENIA PURPURA



Thrombocytopenia may be idiopathic in first 1-4 years of life, is a self-limiting, no platelet transfusion unless platelet < 50.000/cmm.

First line agents for the management of primary ITP

Agent & Dose	Response
Corticosteroids: Adults: Prednisolone 1-2 mg/kg/day for 4 wk, Children: no standard regimen exists, but shorter courses are preferred.	Initial rates; Adults: 70-80%, Children 80-90%; Time: 1 wk; Durability; 10-30% of adults have a durable remission.
IVIG: 0.8-1 gm/kg/day given for 1-2 day	Initial rates: Adults: 80%, Children: 80-90%; Time: 24-48 hrs; Durability: typically 3-4 wk based on antibody half- life.
Anti-D: 50-70 ug/Kg for one dose	Initial rates; Adults: 80%, Children; 50-80%; Time: 24-48 hrs; Durability: 3-4 wk based on antibody half-life.

Investigations

- CBC: low platelet count.
- Bleeding time: impaired clot retraction, prolonged BT (normally 2-8 min) it measure the efficiency of platelets & integrity of blood vessels.
- One of parents show low platelet count.

CHAPTER VI

ONCOLOGY

- Leukaemia
- Brain tumours
- Bone Tumours
- William' s tumour
- Neuroblastoma

PEDIATRIC CANCER

Pediatric cancer is the second leading cause of death in children. 1/350 children diagnosed annually. Or the incidence per year would be 15-16 cases/100 000 children per year. About 11000 new cases in children under 20 yrs of age each year in the whole world. Pediatric cancer considered in the past as hopeless diseases now 70% of children ē cancer can be cured definitively

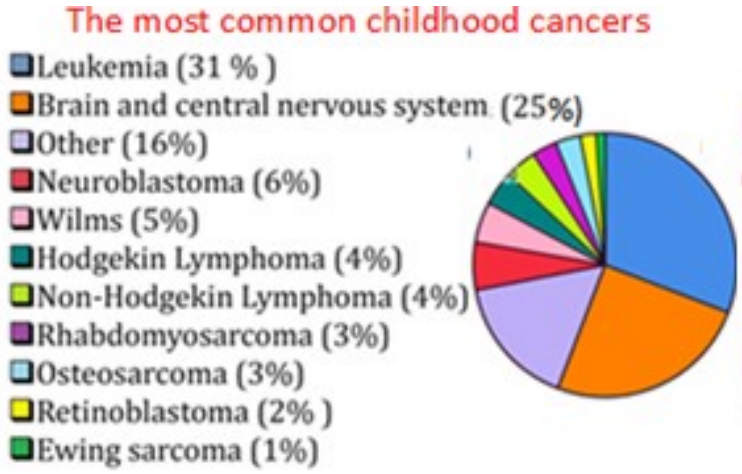
ACUTE LEUKEMIA

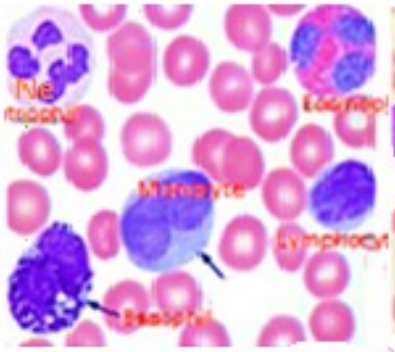
Characterized by presence of immature WBCs in the marrow & peripheral blood. There are 2 types (ALL & AML). Such classification & the further subtyping is done by using the following techniques:-

•Morphological. •Immunological. •Cytochemical characteristics.

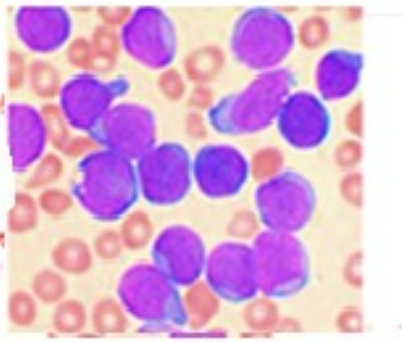
The differentiation between ALL & AML is critical because each have different; natural history, prognosis & response to Rx. ALL is common in children & 85% of cases of ALL occur in children. 90% of leukemia that occur in children are ALL. The lymphoblasts are found both in bone marrow & blood in case of ALL & are characterized by the following feature microscopically:

•Smaller size (10-15 mm) than myeloblasts. •Thin rim of dark blue cytoplasm é no granules. •Nucleus is round or convoluted, centrally located & has 1-2 nucleoli.





Normalblood film



ALL: many more immature lymphocytes than you would typically expect to see

Classification of ALL

Morphological classification (FAB):

① **L 1:** small monomorphic cells (childhood), é small nucleolus.

② **L 2:** large heterogeneous cells (adults) é ≥ 1 prominent nucleoli.

③ **L 3:** uncommon (constituting <5% of ALL), Burkett cell-type, large. vesicular nucleus é basophilic often vacuolated cytoplasm.

Immunological classification

① **Common ALL:** 75 % of ALL, derived from precursors of B-cell.

② **T-ALL:** 20% of ALL, common in adolescent males, associated é high WBC count, anterior mediastinal mass & CNS involvement.

③ **B-cell ALL:** 5%, extra medullary presentation & metabolic abnormal.

Classification of AML

Myeloblasts predominantly make up AML. These cells are larger than lymphoblasts & are characterized by:-

- Lower nuclear to cytoplasmic ratio.
- Prominent multiple nucleoli.
- Auer rods (stick like structures in cytoplasm) seen in 50% of AML.
- Granules in cytoplasm.

The FAB group divided AML into 8 subtypes based on the followings:-

- Degree of differentiation.
- Maturation of predominant cells towards granulocytes, monocytes, erythrocytes or megakaryocytes.

The 8 subgroups nominated as;

- M0 (undifferentiated): composed of primitive cells éout cytochemical stains (it represe-

nt 3% of AML).

•M1 (éout maturation): few if any azurophilic granules (20% of AML).

•M2 (é maturation): blasts é promyelocytes granules, Auer rods, present strong avidity to peroxide- ase & Sudan black (25% of AML).

•M3 (promyelocytes): hyper granular promyelocytes often é Auer rods per cell.

•M4 (myelomonocytic): monocytoid appearing cells in peripheral blood, strong avidity to nonspecific esterase (20% of AML).

•M5 (monocytic): it include 2 subtypes:-

M5a – undifferentiated.

$M5_b$- differentiated é 80% promyelocytes & monocytes.

Both have avidity to nonspecific esterase (20% of AML).

•M6 (erythroleukemia): erythroblasts >50% of all nucleated cells, avidity to periodic acid schiff stain (5% of AML).

•M7 (acute megakaryocytic): megakaryoblasts are >30% of all nucleated cells, activi- ty to PAS (5% of AML).

M_0	Minimally differentiated
M_1	éout maturation
M_2	é maturation
M_3	Hyper granular promyelocytic
M_4	Myelomonocytic
M_5	a) Monoblastic & b) Monocytic
M_6	Erythroleukaemia
M_7	Megakaryoblastic

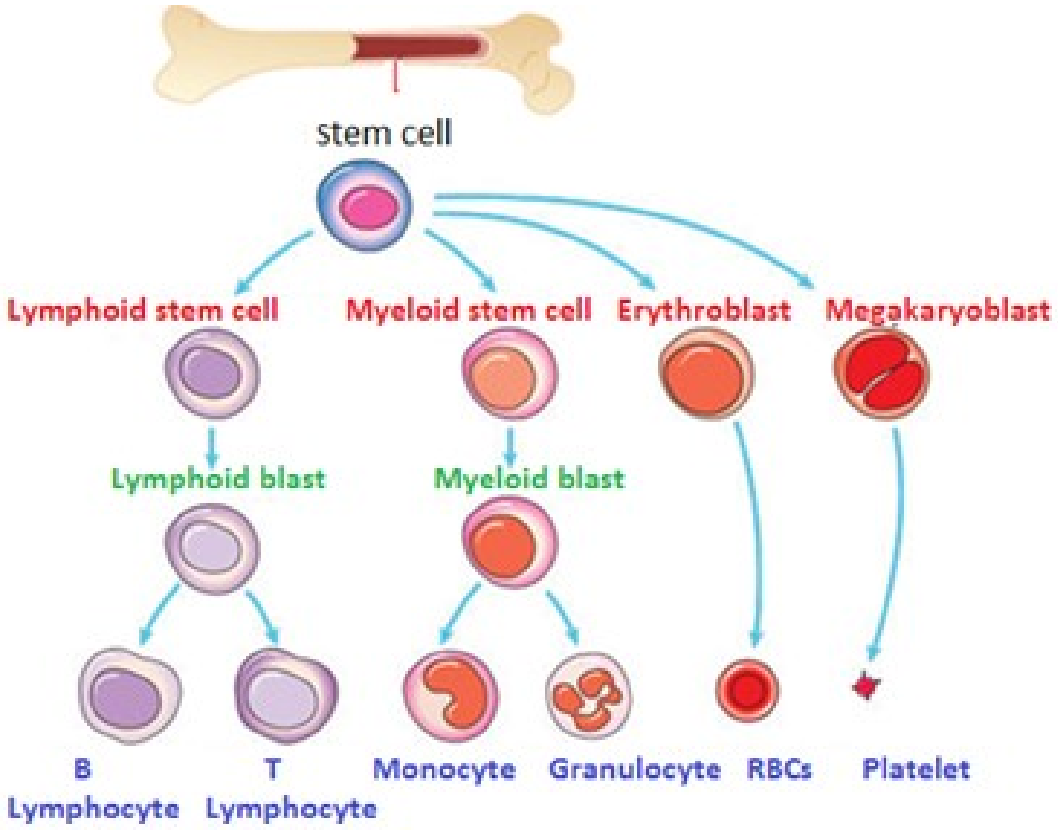
Pathophysiology

Acute leukemia characterized by clonal proliferation of immature hematopoietic cells The most important characteristic is the defect in maturation beyond lymphoblast in ALL or beyond myeloblast or promyelocyte in AML, the proliferation of these immature cells in the bone marrow leads to:-

•Appearance of blast cells in circulation where they aren't normally seen & occlusion of microcirculation by blast cells (leukostasis).

•Infiltration & enlargement of the tissues i.e. LNs, liver, spleen, skin, gum, viscera & CNS.

•Accumulation of blasts in bone marrow has 2 major effects on hematopoiesis causing bone marrow failure through suppression of the normal hematopoiesis & replacement of the normal elements in the bone marrow. The bone marrow changes lead to ⇩ of normal blood cells in circulation causing; infection due to ⇩ WBCs, anemia due to ⇩ RBCs & bleeding due to ⇩ of platelets.



Clinical features

The bone marrow failure, in both ALL & AML share many clinical features. In the majority of cases the initial symptoms are present for <3 months, including:-

- **Symptoms of anemia** such as tiredness, weakness, shortness of breath on exertion.
- **Recurrent infection** due to:⇩neutrophil count & functionally abnormal neutrophils.
- **Bruising &/or Bleeding** related to ⇩ platelet count.
- **Occasionally, LNs enlargement.**
- **Symptoms relating to enlargement of the liver & spleen.**
- **Symptoms related to hypoperfusion** of lungs & brain due to occlusion of microcirculation of these organs by blast cells.

Physical findings

- Pallor.
- Bruises, petechial Hge & purpura.
- Signs of infection like fever.
- Peripheral/generalized lymphadenopathy in ALL, uncommon in AML.
- Hepatosplenomegaly in ALL & small % in AML .
- Weight loss.
- Testicular involvement in ALL
- Bone pain & sternal tenderness due to expanding malignant cell mass, occur in >50% of pts é acute leukemia.
- Symptoms related to CNS (meningitis) & kidneys involvement.

Investigation

The definitive diagnosis of acute leukemia is made on the basis of peripheral blood fi lm & bone marrow aspirate. The following tests should be done:-

CBC: will show ⇩ Hb, while the WBC counts often very high, but occasionally ⇩ or norm-

al, in addition to ⇩ of platelet count.

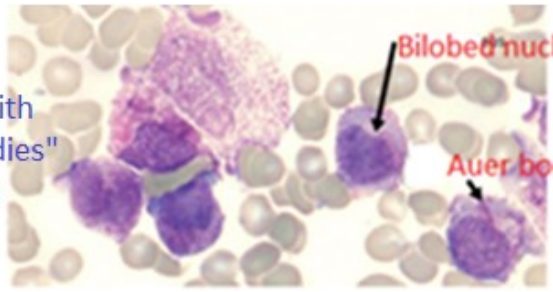
Blood film: will show characteristic leukemic cells, ẃ are distinct from each other morphologically through the following:-

- **Type of cells;** lymphoblast/myeloblast.
- **Cell size;** smaller or larger.
- **shape of nucleus;** is it round/central/irregular or eccentric.
- **Nucleoli;** number is it 1 or 2 & not prominent? or >2 & prominent?
- **Auer rods;** are they absent/or present ?
- **Cytoplasm;** does it have dark/or blue rim ? is it pale blue/or granular ?

Criteria for differentiation in blood film

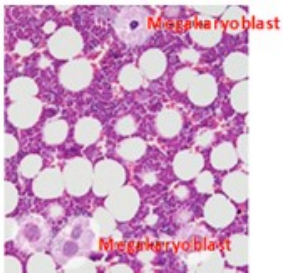
	Lymphoblast (ALL)	Myeloblast (AML)
Cell size	Smaller	Larger
Nucleus shape	Round, central	Irregular, eccentric
Nucleoli	1- 2 in number, not prominent	> 2, prominent
Auer rods	Absent	Present in 50%
Cytoplasm	Dark blue rim	Pale blue, granular

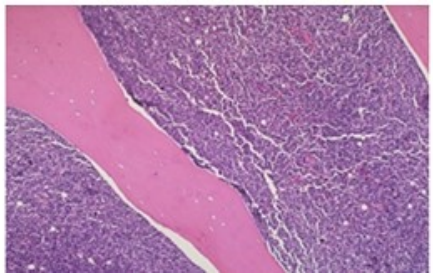
AML
Very large immature myeloblasts with many neucleoli. Linear red "Auer bodies" composed of crystallized granules



Bone marrow biopsy & aspirate

Normal Bone Marrow





ALL highly cellular marrow ~100% cellularity

The normal cellularity of bone marrow range from 30-70%, ẃ include many mature red cells & myeloid cells in variant stages of differentiation, also the presence of megakaryocytes, erythroid islands & granulocytic precursors.

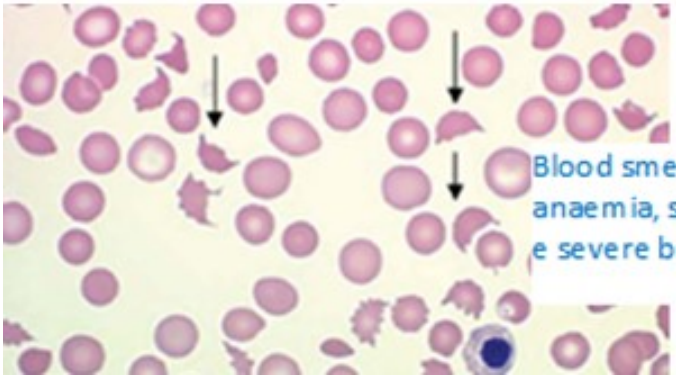
The above right microscopic picture shows high cellularity of bone marrow smear in ALL where 30% or more of all nucleated cells are blast cells. The left picture shows the normal bone marrow cellularity. Notice that the marrow between the pink bone trabeculae seen (in the right slide) is nearly 100% cellular & consists of leukemic cells of ALL that have virtually replaced or suppressed normal hematopoiesis. Thus, though the marrow is quite cellular, there can be peripheral cytopenia. Biopsy mostly done on the sternum using special needle, smear prepared & stained é wright stain, shows; ⇧ cellularity é abnormal lymphoid or myeloid blast cell population.

Hypermetabolism: ⇧ Uric acid in 50% of pts due to rapid cellular turnover & ⇧ LDH.

Radiology: CXR show mediastinal mass in T-cell ALL. X ray bones may show osteopenia or lytic lesion in 50% of pts.

Blood electrolytes: ⇩ or ⇧ K^+, ⇩ Mg^+,⇩ Ph^+

DIC: é consumption of all clotting factors & ⇧ of FDP is most common é promyelocytic leukemia & in small % of monocytic leukemia & ALL.



Disseminated intravascular coagulopathy

Blood smear of a patient e Microangiopathic haemolytic anaemia, schistocytes (arrows) are also noted in patient e severe burns, DIC, dysfunctional prosthetic valves.

Management

The aim of Rx for ALL is to destroy the leukemic cells & enable bone marrow to work normally again. Chemotherapy is the main Rx for ALL. Usually a combination of chemotherapy & steroids are given according to a Rx plan.

Chemotherapeutic agents: drugs that have the capacity to kill leukemic cells.

① ***Remission induction phase:*** this phase aimed at destroying as many leukemic cells as possible. This phase lasts 4-6 wks. Bone marrow test is taken at end of this phase to confirm whether or not the pt still has leukemia. The sample is looked for & when there is no evidence of leukemia, the pt condition referred to as being in remission. In this phase 2 or 3 drugs are used including:- Vincristine 1.4 mg/m^2 IV once weekly for 4 weeks. Prednisone 1 mg/kg BW PO daily. & - L. Asparaginase or Adriamycin.

② ***Intensification phase:*** after complete remission if there is no further Rx given, leukemic cells will expand & lead to relapse, so in this phase intensive chemotherapy is given to ⇩ the total number of residual malignant cells to 10G cells or less. The drugs used include:- Methotrexate 15 mg/m^2 I.M. daily for 3-5 days. followed by Cystarabine 100 mg/m^2 IV twice daily for 3-5 days.

③ ***Maintenance phase:*** next phase of Rx aim to maintain remission & prevent spread of leukemic cells into brain & spinal cord. Here lower dose chemotherapy is given over several years, it include:- MTX 15 mg/m^2 once or twice weekly I.M. -6 M.P 1-2.5 mg/kg daily PO & -Cyclophosphamide: 200 mg/m^2 PO weekly.

④ ***CNS & Testicular prophylaxis:*** involves injecting a drug, usually MTX, directly into spinal fluid (intrathecally) during lumbar puncture. Occasionally, radiotherapy to the brain is also necessary. In this phase local chemotherapy or radiation is given to sites of frequent relapse. This phase include; intrathecal MTX 6-12 mg/m^2 as 5 injections (twice

weekly). Cranial irradiation 1800-2400 R. If child is < 2 yrs age, they will not be given this treatment, but in some situations it may be necessary for boys.

Side effects of Rx for ALL

Many cancer treatments will cause side effects. This is because while Rx is killing cancer cells, they can also damage some normal cells. The side effects of chemotherapy are; hair loss, reduction of the number of blood cells produced by bone marrow result in ⇧ risk of bruising, bleeding & infection. Loss of appetite & weight, feeling nauseated & vomiting. The steroid can also cause side effects as ⇧ appetite, weight gain, mood changes, irritability & muscle weakness.

Summary of Rx of ALL & AML

Phase	ALL	AML
Remission	Vincristine + Prednisolone + L-Asparaginase for 4 wks	Daunorubucin+ Cytosine Arabinoside, for 4 wks
Intensification	Combination of 6-MP & MTX + BMT.	2-3 intensive cycles or high dose of Cytosine Arabinoside + BMT.
Maintenance	Oral 6MP & MTX	No benefit
Prophylaxis	Brain radiation combined + intrathecal MTX	No benefit because CNS relapse occurs only é systemic relapse

Supportive treatment

Applicable for both ALL & AML including:-

①***For pts é severe anemia & thrombocytopenia:*** especially when platelet <20.000 /ml é the risk of bleeding may be transfused é whole blood & platelet conc.

②***Infections are common in acute leukemia:*** so appropriate preventive measures should routinely be employed to prevent infections in such immunocompromised pt including:-

•Isolation of staff & visitors by the use of face masks.

•Careful hand washing before coming in contact é a pt.

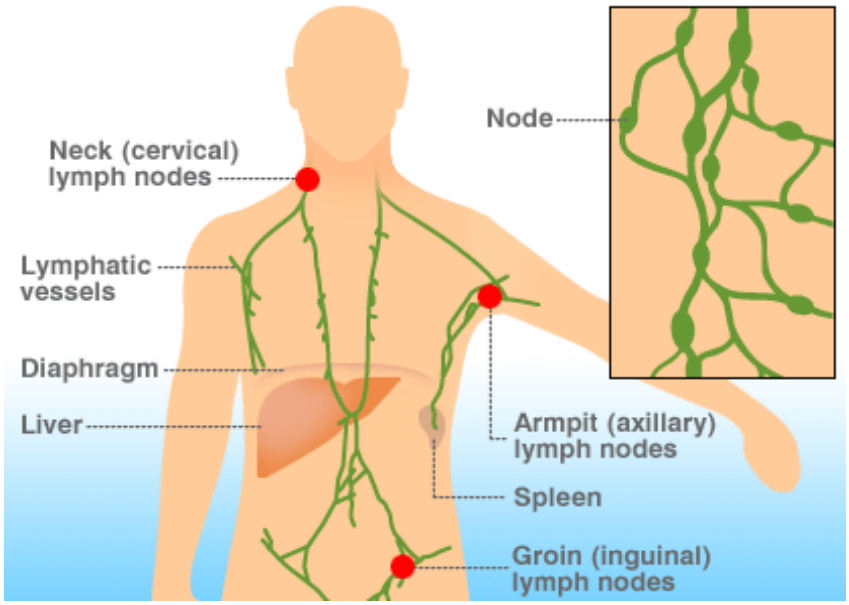
•Advise the pt to eat only cooked foods.

•When infections occur, gram -ve sepsis is the commonest presentation, ẃ requires prompt evaluation & use of empirical antibiotic until definitive diagnosis made by blood culture, after ẃ the antibiotic can be modified depending C/S results.

③***Bone marrow Rx:*** bone marrow or stem cell transplants for children's cancers, only used for children é ALL that's likely to come back following standard chemotherapy. Can also be used for children whose leukemia reoccurred following standard Rx.

Drug doses & duration	Dose	Route	Regimen
Induction (4weeks)			
Vincristine	1.5 mg/m^2	IV	Weekly for 4 weeks
Prednisolone	40 mg/m^2	Oral	Daily for 4 weeks
L-Asparaginose	6000 u/m^2	IM	3 xWeekly for 3 wks
Daunorubicin	45 mg/m^2	IV	Daily for 2 days
Intensification (1 week)			
Vincristine	1.5 mg/m^2	IV	1 dose
Daunorubicin	45 mg/m^2	IV	Daily for 2 days
Prednisolone	40 mg/m^2	Oral	Daily for 5 days
Etoposide	100 mg/m^2	IV	Daily for 5 days
Cytarabine	100 mg/m^2	IV	2 x daily for 5 days
Thioguanine	80 mg/m^2	Oral	Daily for 5 days
CNS prophylactic (3 wks)			
Cranial irradiation	24 Gy		
Methotrexate	I.T. weekly (3wks)		
Maintenance (2 years)			
Methotexate	20 mg/m^2	Oral	Weekly
6-Mercaptopurine	75mg/m^2	Oral	Daily
Prednisolone	40mg/m^2	Oral	days/month
Vincristine	1.5 mg/m^2	IV	Monthly

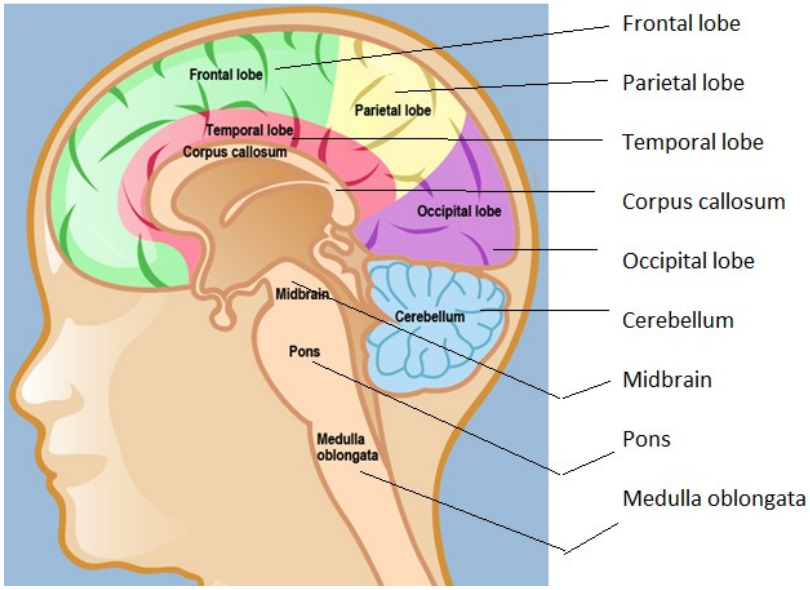
LYMPHOMAS



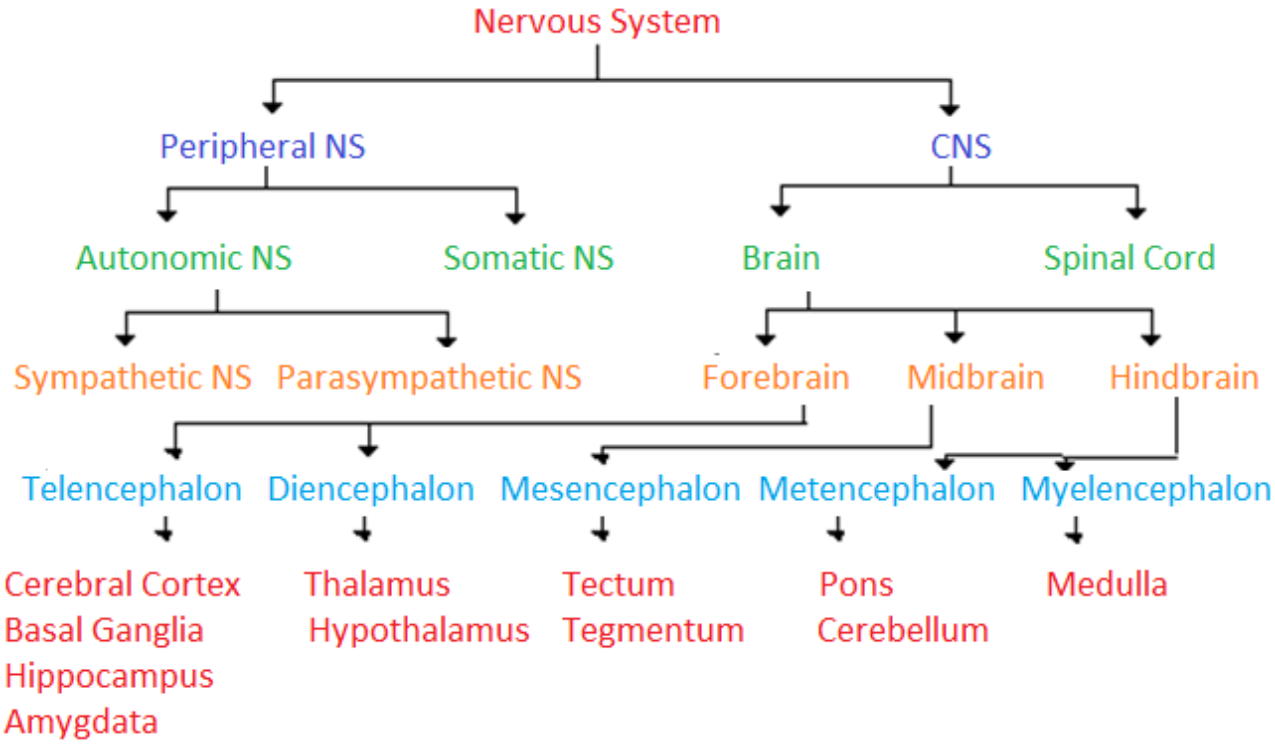
Lymphoma are cancers originating from the lymphatic cells of the immune system, typically seen as solid tumors. The lymphatic system is part of the body`s immune system & helps fight infections & other diseases. The lymph system is made up of thin tubes that branch into all parts of the body. Lymph vessels carry lymph, a colorless, watery fluid that contains lymphocytes. Along the network of vessels are groups of small, bean-shaped organs called LNs, found in clusters in the under arm, pelvis, neck & abdomen. Because lymphatic tissue is found in many parts of the body, lymphoma can start almost anywhere. In 1832, Thomas Hodgkin, a British pathologist published the first description of lymphoma, a specific form ẃ is named after him ”Hodgkin lymphoma”. Since then many other form of lymphoma have been described, all are grouped under a single label” non-Hodgkin lymphoma”. However, the latest lymphoma classification by the WHO in 2008 considers the ancient arrangement absolete because the different lymphomas grouped under NHL have very little in common é each other. Hence NHL label is slowly being abandoned considering its minimal relevance.

BRAIN TUMORS

Abnormal growths or masses that occur in the brain tissue. Most common solid tum- or in children under the age of 15 years. Represent about 20% childhood cancers.



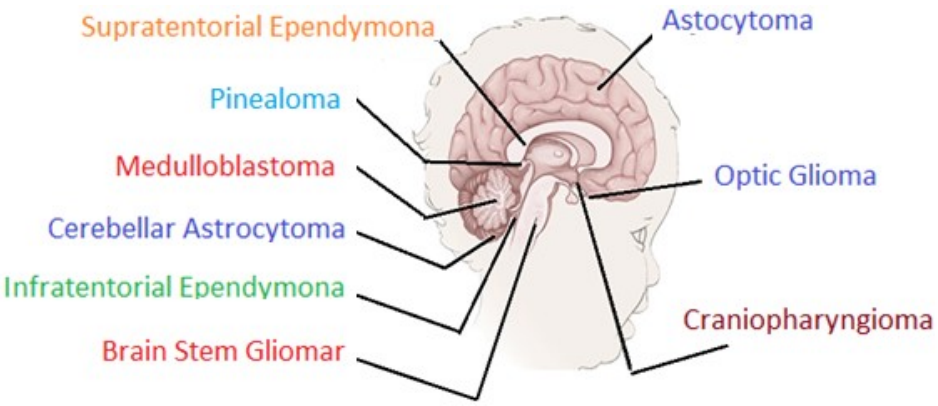
What is the nervous system



Brain Function: Control center. Movement, speech, memory, emotion, thoughts, Interpretation of sensory information (i.e., taste, vision, hearing, touch)

Spinal Cord: main pathway for information connecting the brain & peripheral NS.

Types of brain tumors



The commonest types of brain tumors in children are Medulloblastomas & Gliomas.

Medulloblastoma

Arise form in the cerebellum, accounts for 15% of brain tumors in children, referred to as a infratentorial primitive neuroectodermal tumor, can metastasize to the spinal cord & is highly aggressive.

Brain stem Glioma

located in the middle of the brainstem, majority of the tumors cannot be surgically removed, occur in school-aged children, symptoms include: endocrine problems, paralysis of nerves/muscles of the face & respiratory changes.

Clinical picture

- ⇧ Head size in infants.
- Seizures.
- Morning headache or headache that goes away after vomiting.
- Unusual sleepiness.
- Alterations in vision, hearing & speech.
- Frequent nausea.
- Personality changes.

Prognosis

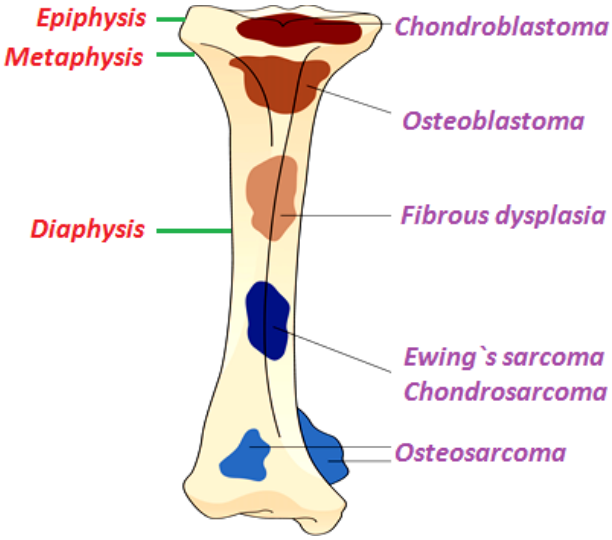
The 5 yrs survival about 80%, depending upon the age, tumor location, & whether it has infiltrated other brain tissue.

BONE TUMORS

Bone tumors are classified into:

- Primary bone tumors.
- Secondary bone tumors (Metastasis).

Most are classified according to the normal cell of origin & apparent pattern of differentiation.



Bone-Forming tumors: ▣ Osteoma. ▣ Osteoblastoma. ▣Osteosarcoma

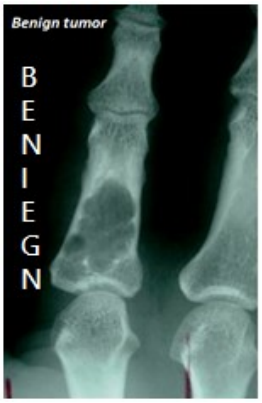
Cartilage-Forming tumors: ▣ Chondroma. ▣ Osteochondroma. ▣Chondrosarcoma.

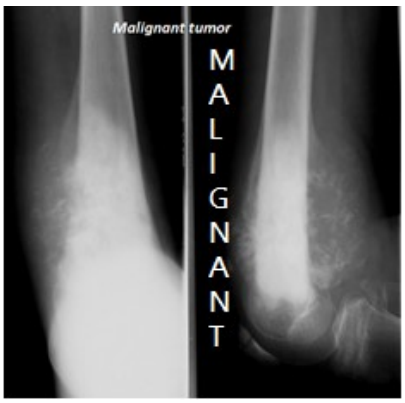
Miscellaneous tumors: ▣ Ewing's sarcoma. ▣ Giant cell tumor of bone.

Radiographic features

*Benign: well circumscribed, no reaction & sclerotic border.

*Malignant: ++++ reaction, large, permeative, destructive & moth eaten.





OSTEOMA

Is benign lesion of bone that in many cases represent developmental aberrations or reactive growths rather than true neoplasms.

OSTEOSARCOMA

Most common primary malignant tumor of bone. 1/500.000 of population.

Age: 10-20 yrs: 75% in pts < 20 yrs of age (primary type) & 25% old age (secondary to Paget disease). Site: metaphysis of long bones of limbs (60% occur around the knee).

M : F ratio is 1.6 : 1. Risk factors include; Paget disease of bone, Ionizing radiation, Fibrous dysplasia, Chronic osteomyelitis, Bone infarcts & Mutation of TP53 gene.

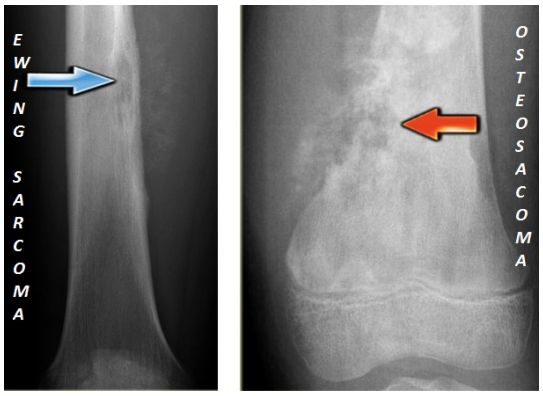
Clinical features

Pain, swelling, usually attributed to minor unnoticed trauma. Can affect any bone in the body. pathological fracture, marked ⇧ in the serum alkaline phosphatase, early hematogenous spread to the lungs, liver & brain.

Diagnosis

- X ray bone shows characteristic lytic lesions, or bony mass.
- Biopsy is diagnostic.
- Scanning for distance metastasis (Chest X Ray).

EWING SARCOMA



Primary malignant small round cell tumor.

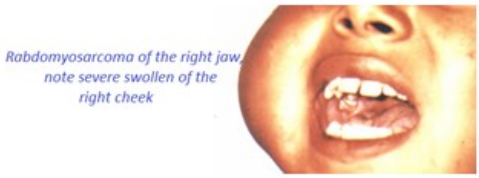
Ewing sarcoma has the youngest average age at presentations (10-15 years).

Boys slightly more often affected than girls.

Site usually in the diaphysis of long bones especially femur followed by tibia & humerus.

The pathways of spread include; direct extension, lymphatic or vascular dissemination, intraspinal seeding.

RHABDOMYOSARCOMA



Soft tissue sarcoma that arises from undifferentiated mesenchymal cells in muscle, tendons, bursa & fascia, or fibrous, connective, lymphatic or vascular tissue. 2/3 of cases are diagnosed by 10 yrs of age. It represent 5% of neoplastic diseases in children.

Histologically; Embrional & Botroid (75%) , Alveolar + Pleomorphic (20%), & Undifferentiated (5%).

Localization

- Head & Neck 40%.
- Pelvis + Urinary Tract 25%
- Limbs 20%.
- Other rare Localizations 15%; (diaphragm, abdominal walls, viscera & every region originated from mesenchyme arising in striated muscle.)

Stages

- Stage I: limited tumor excised completely.
- Stage II: grossly removed tumor ē microscopic residual disease

- Stage III: incomplete removal or only biopsy ē gross residual tumor.
- IV Stage VI: metastatic disease at diagnosis.

Investigations

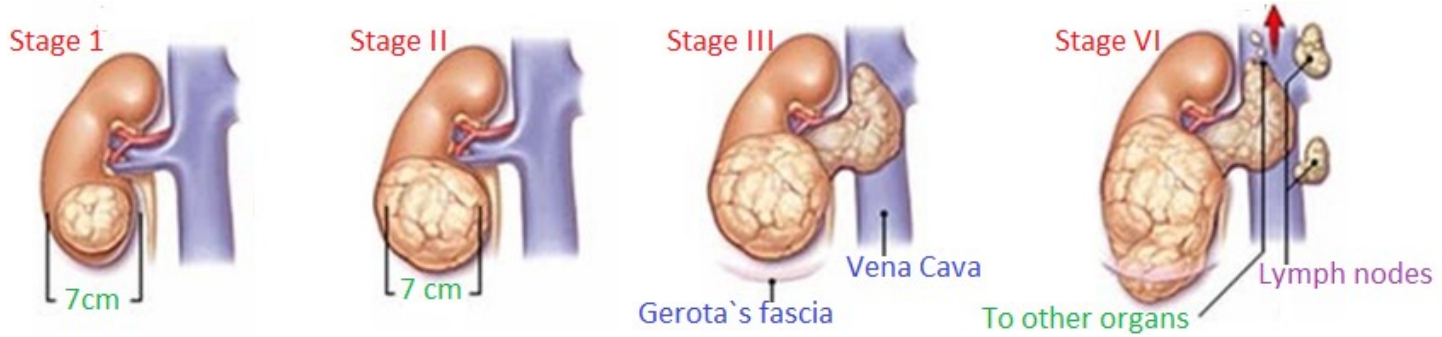
- Hb, Platelet, WBC, LDH.
- CT & MRI of primary tumor.
- Chest X-ray & CT.
- Bone Scan.
- Tumor Histology.

Treatment

- Surgery.
- Radiotherapy.
- Chemotherapy - Vcr, Holoxan.
- Farmarubicin, VP16, Endoxan.
- Carloplatin, Actinomycin D.

Prognosis: long term survival rate 70-80%

WILM' S TUMOR



Embrional tumor of the kidney. is the most common childhood abdominal malignancy, peak at age 0 - 3 yrs. May be unilateral or bilateral.

Pathogenesis

2 forms; Sporadic -No known genetic predisposition & inherited as AD. Both forms linked to the deletion or inactivation of genes on the short arm of Chromosome 11. 18% of children ē Wilms have other congenital anomalies.

Incidence: 7.8 /1000.000 children..

Clinical Manifestations

90% have enlarging asymptomatic upper abdominal mass that is firm, nontender, smooth & encapsulated. its direction of spread is vertical & not crossing the mid line of the body, most commonly mother notice abdominal swelling of her baby, may associated ē hematuria in 20 % of cases, & may be associated ē urinary anomalies, aniridia or hemihypertrophy.

Diagnosis

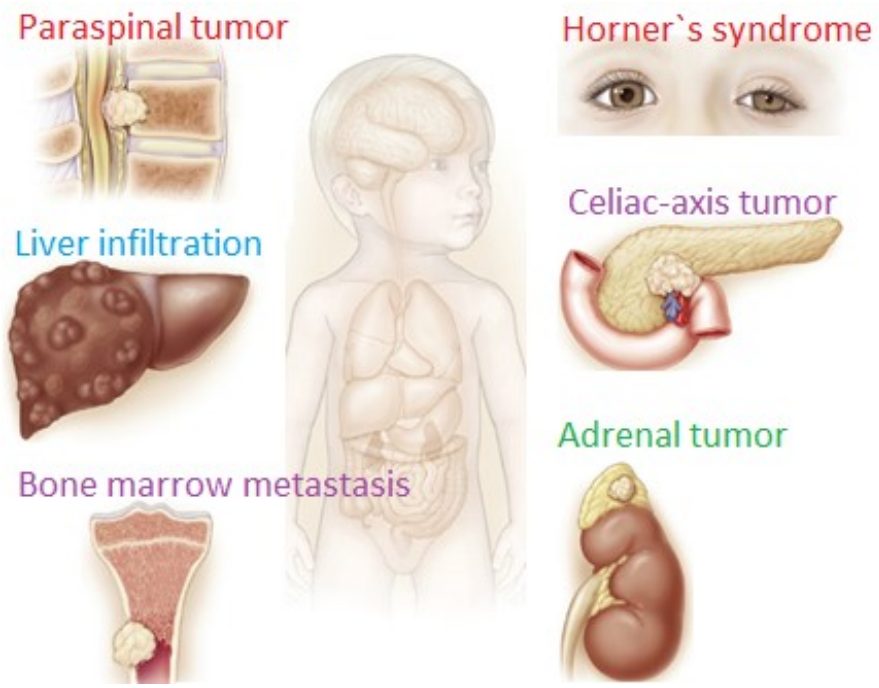
- U/S abdomen.
- CT scan abdomen.
- Skeletal survey for metastasis.

Treatment

- Nephrectomy.
- Radiation (except in Stage I & II).
- Chemotherapy.

Prognosis: 95% cure rate for Stage I to III.

NEUROBLASTOMA



The most common extra-cranial malignant solid tumor in infancy. It is neuroendocrinal tumor, arise from the sympathetic nervous system anywhere in the body. In western countries it is obligatory by law for every infant to have U/S abdomen during 1st year of live. The peak of neuroblastoma is from age 0–5 years.

Incidence

1/1000 live birth. 8% of neoplastic diseases in children. Unfavorable histology (Neuroblastoma) 90%. Favourable histology (Ganglio neuroblastoma) 10%.

Localization

- 70% Abdomen (1/2 of cases from suprarenal gland).
- 15% Mediastinum.
- 3% Neck.
- 8% Paravertebral Region.
- 4% Other rare regions (olfactory region, Multiple primary tumors CNS ets).

Stages (Evans et al.)

- Stage I: tumor limited to the organ or structure of origin. Excised completely.
- Stage II: tumor ē regional spread, not crossing the midline.
- Stage III: tumor crossing the midline, bilateral L Ns may be involved. Complete excision is impossible.
- Stage IV: tumor ē distant metastasis (bone"50% of cases", LNs, organs, soft tissues).

Clinical picture

Effect of the enlarged sympathetic chain over the adjacent organ, or as a result of metastasis in different parts of the body. Pancytopenia. Anaemia. Proptosis. Horners syndrome. PUO. Hypertension. Diarrhea. Convulsions (symptoms of primary localization or metastatic localization - Paraneoplastic symptoms).

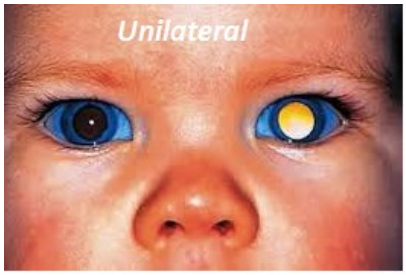
Investigations

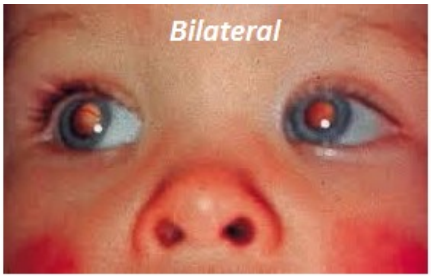
- CBC.
- Catecholamine in urine: HVA & VMA.
- U/S abdomen: direction of spread of is horizontal, may contain calcifications.
- Bone scanning: for metastasis.
- CT scan abdomen.

Management

- Surgery; survival is better when radical excision is done.
- Radiotherapy. No RT in I, II. stages.
- In III & IV stages RT in tumor & metastasis, Redions ē dose 15-35 GY.
- Chemotherapy: L. R. - Vcr, Endoxan, Farmarubicin. H. R. - Vcr, Endoxan, VP16, Cisplatin, Carboplatin, Holoxan, Farmarubicin.

RETINOBLASTOMA





Rare congenital tumor that originates in the retina of one or both eyes. 2 forms;

- Inherited - diagnosed during the first yr, often involves both eyes.
- Acquired - diagnosed at age 2-3 yrs, 60% unilateral.

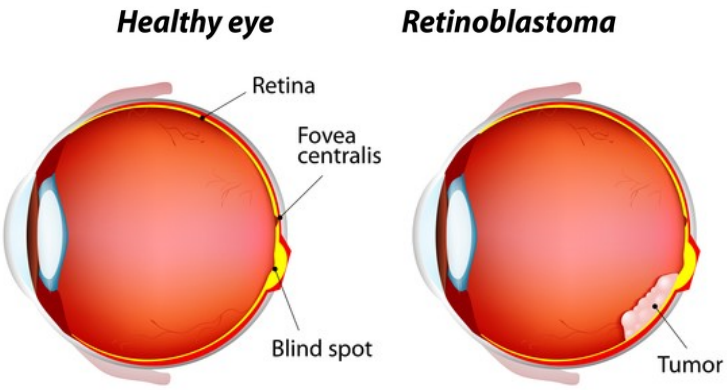
Clinical picture

Primary sign -leukokoria, a white pupillary reflex called “cat’s eye reflex”. Others - strabismus, red, painful eye & limited vision.

Treatment

Radiation for small tumors, large or multiple tumors require inoculation.

Prognosis: 90% long term survival rate.



CHAPTER VII

INFECTION

- Introduction
- Chicken Pox
- Measles
- German Measles
- Fifth Disease
- Roseola Infantum
- Kawasaki Disease
- Sepsis/Septicaemia/Meningitis
- Neonatal tetanus
- Osteomyelitis
- Polio
- Mumps
- Tonsillitis
- Scarlet Fever
- Erysipelas
- Infectious Mononucleosis
- Diphtheria
- Otitis Media

Introduction

The human body defensive mechanisms: include the following:-

Physical mechanism: intact skin, mucous membrane, ciliary function, bacterial flora.

Cellular mechanism:

Macrophage: engulf bacteria, produce lactoferin ẃ chelate iron from bacteria, produce lysozyme ẃ kill bacteria.

B-lymphocytes ẃ produce opsonin causing opsonization of bacteria & complement ẃ neutralize bacteria.

T-lymphocytes ẃ stimulate production of more B lymphocytes & more plasma cells. Also produce interleukin 2 ẃ stimulate production of T killer & T suppressor cells, both regulate the defensive mechanism.

Plasma cells produce complement, opsonin, immunoglobulins.

Humoral mechanism: immunoglobulins, opsonin, complement. Baby at birth have only IgG from his mother (the only immunoglobulin ẃ cross placenta due to its low molecular weight), then baby start to produce his own immunoglobulins.

Bacteria

Gram +ve: Staphylococci, Streptococci, Diphtheria, Clostridium, Anthrax.

Gram -ve: E Coli, Pseudomonas, Proteus, Salmonella, Shigella, Klebsella, Pordetela Pertussus, Chlamydia Trachomatis.

Incubation period

✲ DICES: 1-7 days; Diphtheria, Influenza, Cytomegalovirus, Erysipelas, Scarlet fever, Herpes simplex.

✲ TP MEWs: 1-2 wks; Tetanus, Polio, Measles, Enterica (typhoid), Whooping cough.

✲ CRUMPs: 2-3 wks; Chicken pox, Rubella (German measles), Roseola, Mumps.

Order of appearance of rash

- Very: day 1 Varicella (chicken pox, Herpes).
- Sick: day 2 Scarlet fever.
- People: day 3 Poxvirus.
- Must: day 4 Measles.
- Take: day 5 Enterica (Typhoid).
- Entire: day 6 Typhus.
- Rest: day 7 Relapsing fever.

Isolation period for common contagious infections

- Measles: 4 days before until 5 days after rash appears .
- German measles: 7 days before until 5 days after rash appear.
- Chickenpox: 5 days before rash until all sores have crusts (5-7 days).
- Roseola: from onset of fever until rash is gone (2 days).
- Fifth disease: (Erythema infectiosum): 7 days before rash until rash begins.
- Mumps: 5 days before swelling until swelling gone (7 days).
- Whooping cough: onset of runny nose until 5 days on antibiotic.
- Diphtheria: onset of sore throat until 4 days on antibiotic.
- Infectious mononucleosis: onset of fever until fever is gone (7 days).
- Scarlet fever: onset of fever or rash until 24 hrs on antibiotic.
- Meningitis: onset of symptoms & for 1 to 2 wks.
- Bronchiolitis: onset of cough until 7 days.
- Influenza: onset of runny nose until fever is gone.
- Croup (viral): onset of cough - stridor - until fever is gone.
- Tuberculosis: 2 wks on drugs (most childhood TB not contagious).

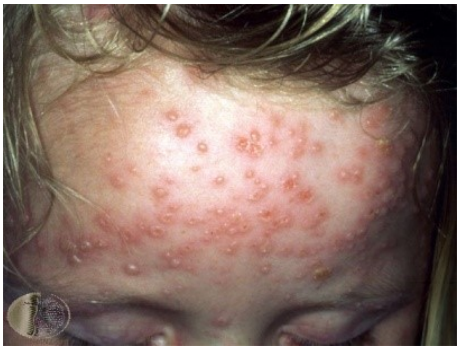
Immunoglobulins: amp 2 ml, 0.2 ml/Kg IV/IM for 3 successive days, given in case of;

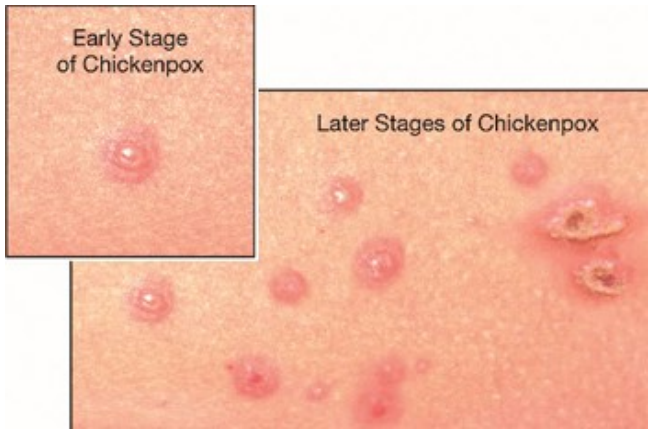
- Kawasaki disease
- Chicken pox within 3 days from exposure
- AIDS
- ITP.

Fever with rash

- Chicken pox: polymorphic, centripetal rash.
- Measles: monomorphic, centrifugal, kopliks spots.
- German measles: monomorphic, centrifugal.
- Fifth disease: slapped check rash.
- Roseola infantum: papular in trunk, rapidly spread to whole body.
- Kawasaki disease: papular & plotchy rash.
- Meningococcal meningitis: reddish to purple skin rash in whole body.
- Typhoid fever: rose spots on 5th day on the trunk.
- Endocarditis: petechial or echymotic generalized rash, Subconjunctival Hge, soft palate petechie, Hge within nail bed (splinter Hge), painful SC nodes on palm (osler`s)

CHICKEN POX





Caused by Varicella-Zoster Virus (also known as VARICELLA). Highly contagious. Most people infected by age 5 years. Immunity ⇧ ē age. Outbreaks usually winter/spring. Epidemics every 2-5 yrs. Spreads via respiratory tract. The IP 2-3 wks. Infective from 5 days before rash, until crusts fall off. Days 1 & 2 are the most infective.

Clinical Picture

The virus first infects mucous membrane of URT. Viral proliferation occurs in LNs for 2-4 days after the initial infection. 4-6 days after initial infection, virus enters blood stream, this is followed by a second round of replication in body's organs especially in spleen & liver. The prodromal stage is of short period, 1 day of fever, malaise & URTI. The Rash is itchy & scratchy, centripetal & polymorphic, start in abdomen, may be one spot in face, or thumb, then next day it cover the whole body.

Complications

•Secondary infection (scratching). •Secondary Bacterial infection especially Group A Streptococcal. •Encephalitis. •CNS-cerebellar ataxia, myelitis, vasculitis. •Osteomyelitis. •Sepsis. •Otitis media.

Management

#Isolation until rash completely crusted.

#Keep skin clean by frequent baths or, once the fever has subsided, showers. Cool, wet compresses or tepid water baths help to relieve itching.

#Antihistamines may used to help relieve the itching.

#Acyclovir is used for severe infection involving the lungs or the brain & in persons ē a depressed immune system. Oral Acyclovir 200 mg, dose 15 mg/Kg/day ÷ 3 X 5 days.

Prevention

✳Children between 12-18 months should receive a dose of chickenpox vaccine, Varicella-zoster immune globulin.

✳Many countries have passed legislation requiring the chickenpox vaccine for child care & school entry.

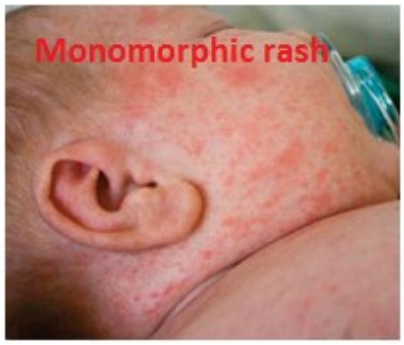
✳Healthy children older than 13 & adults who have no history of chicken pox & have

never been immunized against the disease, should be given the vaccine.

✳ Contacts can be given within 3 days from exposure dose of hyperimnoglobulin 0.2 ml/Kg (amp 2 ml).

✳ Avoid pregnant women, neonates, immunocompromised.

MEASLES



Highly contagious viral illness. First described in 7th century. The IP 1-2 weeks. Affect age group 2-5 years. Its peak is Marsh. Infective 4 days before & 5 days after rash appears. Infective 5 days before & after rash.

Clinical picture

Severe manifestations of URTI; fever, cough, running nose, conjunctivitis for 3-5 days. Appearance of Kopliks spots on the third day opposite the lower first & second molars (one day before appearance of rash). Rash is monomorphic, centrifugal start in face. The temperature decline ē appearance of rash.

Investigations: measles specific antibodies IgM.

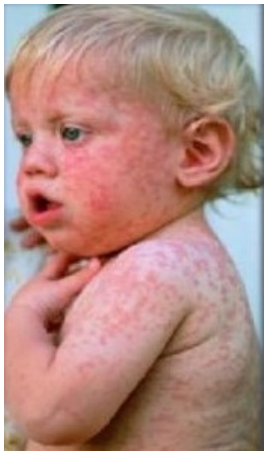
Complications

Diarrhea, otitis media, pneumonia, encephalitis

Management

- Symptomatic treatment.
- Isolation 5 days before until 5 days after rash appears.

GERMAN MEASLES



Rubella comes from the Latin words for “little red”. Creates dense groups of small skin rash. Produces swollen LNs, runny nose & fever. Spread through respiratory droplets from coughing & sneezing. Also spread through body secretions & excretions. The IP 2-3 wks. infection during pregnancy (1st TM) lead to; miscarriage, fetal death, premature delivery, or live birth ē congenital defects (CRS) ē its classic triad; Cataract, Cardiac abnormalities & Deafness..

Clinical picture

- Short prodromal stage 1-2 days.
- Mild fever, cough, running nose, enlarged tender posterior auricular Lymph nodes.
- Rash is monomorphic & centrifugal (start in neck & face).

Management

No specific therapy.

Isolation period is 7 days before until 5 days after rash appear.

Symptom-based treatment.

Immunoglobulin does not prevent rubella virus infection, only for pregnant woman who exposed to rubella, whom will not consider termination of pregnancy under any circumstances (IM immunoglobulin within 72 hrs of rubella exposure).

Prevention: rubella vaccine contains live attenuated rubella virus grown in human diploid cells, combined ē measles & rubella (MR) or measles, mumps & rubella (MMR) formulations, or tetravalent measles, mumps, rubella & varicella (MMRV) vaccine. One dose induces seroconversion in 95% of persons >1 year of age.

FIFTH DISEASE

Fifth disease is a common childhood infection, caused by parovirus, its IP is 1-3 weeks. Most commonly affects young children & often occurs in several members of the family or school class. Infective from 14 days before & ceasing at the onset of the rash. No recommendation to keep away from school once well.

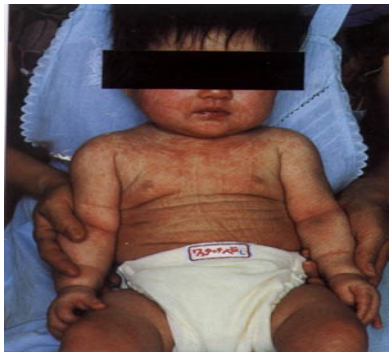
Clinical picture

Causing a slapped cheek appearance, rash appear first on face as if he is slapped in his face. The rash then spread to arms & chest as red spotty rash, continue for 1-3 weeks. 30% of cases have no symptoms. May associated ē fever, rash continue for 1-3 weeks. Once rash appear, the child will be not infective.

ROSEOLA INFANTUM

Caused by Herpes virus (HHV6 &7), also called 6th disease. Higher incidence during spring spring & fall months, the IP 5-15 days. Most adults excrete HHV-6 &7 in saliva & may serve as primary sources for virus transmission to children. Prodromal period is usually asymptomatic; mild URI signs, mild cervical or less frequently occipital lymph nodes may be

noted. Some chidren may have mild palpebral edema.Clinical illness is generaly heralded by high temparature ẃ persists for 3-7 days ; resolves abruptly. A rash appears within 12 - 24 hrs of fever resolution, rash appear as rose colored, distinctive on the trunk → to neck ,face, proximal extremities (centripetal, maculopapular). The rash fade in 1-3 days. Some childr- en may become irritable & anorexic. Seizures may occur in 15% of cases. Rhinorrh- ea, sore throat,abdominal pain, vomiting & diarrhea may be associated ē the disease. The characteristic enanthem consist of the soft palate &the base of the uvula. The enanthem may be present on the fourth day in 2/3 of pts ē roseola.



Diagnosis: Specific test for HHV 6-7. - Virus culture. - PCR. - Antigen detection

Management: no specific treatment. Antipyretics (never use aspirin). Cetal drops 100 mg, syrup 250 mg, supp 120mg, max dose 1200mg/D. Marcofen pediatic syrup or supp 100 mg, 1 X 2 for baby >6 months age. HHV-6 is inhibited by Ganciclovir (but not Acyclovir).

KAWASAKI DISEASE

Etiology: disease of unknown etiology. ? Viral. More in boys < 5 years old.

Clinical picture: high fever up to 2 wks, generalized vasculitis, congested throat, conjunct-

ivitis, cracked lips, cervical lymphadenitis, blotchy purpuric rash, oedema, inflammation/ redness & oedema of both hands & feet, peeling of skin over tip of fingers takes place in 2nd week, truncal rash, strawberry tongue, desquamating perineal rash, arthritis, iritis, carditis ẃ may result in myocardial infarction or heart failure.

Investigations

- CBC: leucocytosis, thrombocytosis • ESR: ⇧ • CRP: ⇧ (normally it is < 1 mg/dL).
- ECHO cardiography: repeat after 2 wks then after 2 months.

Management

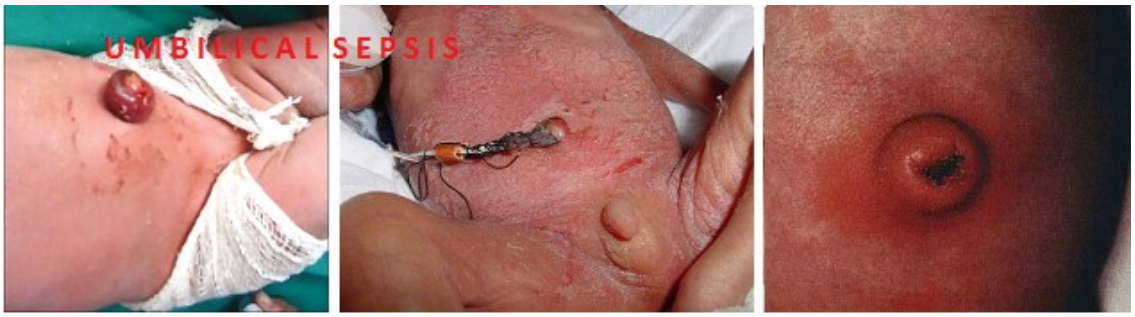
High dose of cortisone 30mg/Kg/day for 1-3 days, prednisone/prednisolone tab 5 mg, Soluprid tab 20 mg.

Immunoglobulins IV or IM, Gama globulin 5 %, 2.5 mg/100 ml, 1 ml/Kg/day for 3 successive days to ⇩ the risk of coronary artery aneurysm.

No aspirin.

Plavix 75mg daily for long duration.

SEPSIS/SEPTICEMIA/MENINGITIS



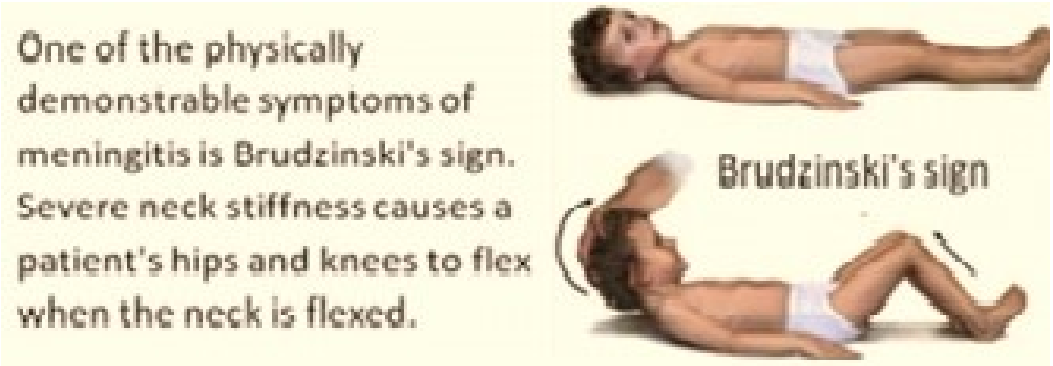
Severe skin & systemic infection of baby born after prolonged rupture membrane for several days, baby was very foul smelling, skin desquamating & erythematous ē evidence of fissuring, culture of skin swab shows growth of E coli, group B streptococci, staphylococcus aureus or candida albicans. The commonest organism in the NN period is

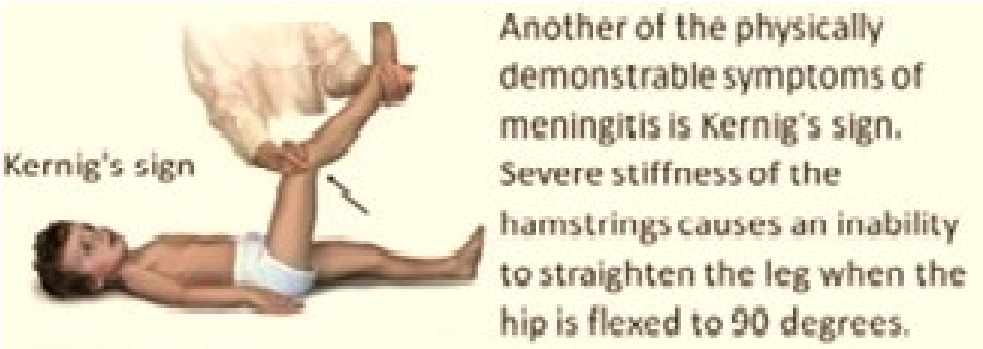
group B streptococci & E coli, then come gram -ve bacteria. 10% of pregnant mothers are found to be colonized in vagina ē group B streptococci & 25% of their babies found to acquire this bacteria, mortality rate of neonatal sepsis is 20-80%, surviving infants may have significant neurologic squeals. Bacteria gain access into the blood stream, may cause overwhelming infection ēout much localization (septicemia) or may get localized to lung (pneumonia) or meninges (meningitis). Early onset sepsis presenting in 1st wk of life, through organisms in mother genital tract or in delivery room (nosocomial infection), commonly associated ē LBW, PRM, chorioamnionitis, foul smelling liquor, difficult or prolonged labor, meconium aspiration syndrome, maternal infection during pregnancy & often manifest as pneumonia, less commonly septicemia or meningitis. Late onset sepsis is presenting after 1st wk of life, result from acquired infection from home or hospital, hands of care providers, present ē septicemia, pneumonia, or meningitis, commonly associated ē LBW, lack of breast feeding, superficial skin infection (pyoderma), home deliveries especially in rural areas, traditional practices as circumcision away from hospital, umbilical sepsis, aspiration of feed, disruption of skin integrity ē needle pricks & use of IVFs.

Clinical Picture

Mostly vague, in early onset sepsis the baby may presented ē RD, apneic spells or gasping, may be the only manifestation of septicemia. Hypothermia is common & fever is infrequent. Lethargy, inactive & reluctant to feed. In late onset sepsis the baby, ,who had been active & sucking well, gradually or suddenly, become lethargic, inactive, refuse sucking, may presented ē diarrhea, vomiting, abdominal distension, poor capillary perfusion, jaundice, sclerema. Critical neonate may develop shock, cyanosis, bleeding & renal failure. With meningitis baby develop irritability, high pitched cry, excessive crying, seizu-

res, blank look, neck retraction, bulging fontanel, focal neurological signs.





Mortality: 5-10% (90% if associated ē DIC).

Diagnosis

Apply sepsis screening; ⩾ 2 +ve tests of the following investigations:

① Cultures blood, CSF, urine, pleural effusion, pus: diagnostic.

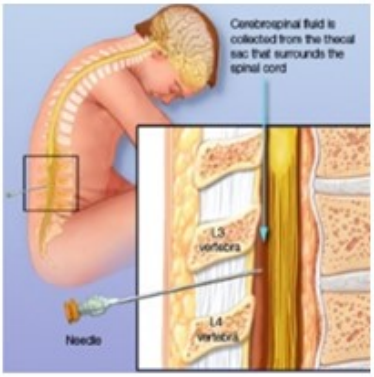
② CBC: leucopenia <5000/cmm, neutropenia <1800/cmm, or immature/total neutrophils (I/T) >0.2 (immature cells = band cells + myelocytes + metamyelocyte) ẃ means that bone marrow pushes premature cells into circulation for fighting infection.

③ CRP +ve: high degree of sensitivity for NN sepsis, can be affected by asphyxia, shock, meconium aspiration, PRM.

④ ESR: >15 mm 1st hr.

⑤ CSF: 10-15% of late onset sepsis may have associated meningitis also lumbar puncture is indicated ē 1st attack of convulsion.

Technique of lumbar puncture



	Bacterial	Viral	TB
Cells	10-100,000	<2,000	250-500
	polys	lymphs	lymphs
Glucose	low	normal	very low
Protein	N-INC	N-INC	N-INC
G-Stain	gen +ve	-ve	+ve Zn

Feel the upper border of iliac crest it's at the level of L5 vertebrae, go up to touch spine of L4 vertebrae & under complete aseptic conditions introduce needle (size FG 21, 22, for infants &children) into the disc space between L4-L3 vertebrae, directing the needle towards the umbilicus, if CSF was found to come out under high pressure stop the procedure to avoid conning of cerebellum. Collect CSF in 3 tubes, 10 drops in each for cells, biochemistry & culture. Take blood sample for BS after you finish to compare é CSF sugar (normally the CSF sugar is <2/3 of blood sugar), ⇩ CSF sugar seen in bacterial meningitis.

C.S.F. types of meningitis

in TB meningitis, CSF shows early polymorph, then lymphocytes become predominant, CT scan brain may show exudate in basal cisterna (see part 1, page 53).

Tuberculin test

Negative in the first 3 months of life even ē infection, in adults it takes few wks after infection to turn +ve, read after 72 hrs.

Mantoux test

Confirmatory, 5 U of PPD, read reaction after 72 hrs, 0-4 mm = -ve, 5-9 mm=? +ve & repeat, 10 mm or more is +ve test & in such case do PCR for tuberculosis.

Management

- Provide warmth if baby hypothermic, ensure consistently normal body temperature
- Nothing per oral if baby very sick, or has abdominal distension.
- IVFs: normal saline 10 ml/Kg over 5-10 min if perfusion is poor as detected by capillary refill time (> 3 sec), repeat same dose if perfusion still poor over the next 45 min.
- Dopamine if perfusion is permanently poor, dose calculated as; baby weight X 3 = mg to be collected from dopamine 250mg vial by insulin syringe, add to it 50 ml of glucose 5% & give 2 ml/hr ẃ is equal to 2ug/Kg/min, it can be doubled to 5 ug/Kg/min, this ⇧ blood supply to internal organs. Higher dose from 6-10 ug/Kg/min cause ⇧ of blood supply to internal organs in addition to +ve inotropic, chronotropic effect on heart. Much higher dose 11-15ug/Kg/min cause the same previous effects + peripheral vasoconstriction.
- Glucose 10% 2 ml/Kg stat (hypoglycemia always associated ē sepsis & septicemia).
- Antibiotic Rx.

Septicaemia/Pneumonia

Antibiotic	Dose	Frequency/Age <7 days	Frequency/Age >7 days	Route	Duration
Ampicillin or Penicillin or	100 mg/Kg /day 1000.000 U/Kg/D	12 hrly 6 hourly	8 hrly	IV, IM IV, IM	7-10 days
Cloxacillin +	100 mg/Kg /day	12 hrly	8 hrly	IV	7-10 days
Gentamicin or	3-5 mg/Kg /day	12 hrly	8 hrly	IV, IM	7-10 days
Amikacin	7.5 mg/Kg /day	12 hrly	8 hrly	IV, IM	7-10 days

Meningitis

Ampicillin +	100 mg/Kg/day	12 hrly	8 hrly	IV	3 weeks
Gentamicin or	3-5 mg/Kg/day	12 hrly	8 hrly	IV	3 weeks
Claforan +	50 mg/Kg/day	12 horly		IV, IM	10 days
Vancomycin	50 mg/Kg/day	12 horly		IV, IM	10 days

Meningococcal meningitis

Vaccinate all children age 1-5 yrs whom are in hospital & to all health workers in hospital, or give Rifampicin syrup 100 mg, dose of 20 mg/Kg/day ÷ 2 for 2 days.

Viral meningitis

Antiviral inhibit viral duplication in case of H. Simplex, H. Zoster, Varicella, EBV or CMV infection. Zovirax syrup 200 mg. For children 6 month -12 yrs give 200 mg X 3 X 5 day or amp 250 mg infusion over over 1 hr/8 hr for 5 days. Or Acyclover amp 250 mg, 15 mg/Kg /day ÷ 3, IV over 1 hr for 10 days.

Tuberculous meningitis

Rifampicin amp 250 mg IM, dose 20mg/Kg/day÷ 2 for 2 wks + Streptomycin 1000mg amp , 25 mg/Kg/day ÷2, IM for 2 wks. Or Rifam plus cap (Rifampicin + Isoniazide) 150 /100 for children, for 7 months if sputum is +ve for AFB.

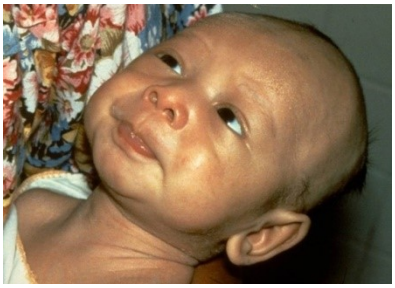
- Anticonvulsant: Valium amp 5, 10 mg, 0.25 mg/Kg/dose stat IV or IM, suppository 5, 10 mg, daily maintenance is 0.25 mg/Kg/day÷4.

Phenobarbital amp. 200 mg, syrup 20 mg, first dose is 15 mg/Kg IV or IM, maintenance 5 mg/Kg/day÷2-4.

Epanutine amp 250mg, syrup 30mg, starting dose 15mg/Kg, maintenance 5mg/Kg/day÷4.

- Vit. K 1mg IM (to guard against bleeding).
- Oxygen by hood or mask, if baby cyanosed, grunting, or distressed.
- Gentle physical stimulation, if baby apneic.
- Dexamethasone in case of septic shock, 0.5 mg/Kg/dose, IV, 6 hourly for 2 days.
- Exchange transfusion may be needed if there is sclerema.
- There is no role of immunoglobulin's therapy in neonatal sepsis.

NEONATAL TETANUS



Caused by exotoxin produced by gram +ve bacteria "clostridium tetani", ẃ produces 2 exotoxins; tetanolysin its action not known, tetanospasmin ẃ is neurotoxin bind to CNS interfering ē the neurotransmitter release to block inhibitor impulses causing the cli-nical manifestations of the disease. Cl. tetani is anaerobic, motile, rod, forms ovale colorless terminal spores (tennis racket or drumstick shape), the spores are very resistant to heat, chemicals, radiation, drying, can survive for long time in envir-onment (months up to years, decades), it is found worldwide in soil, in inanimate environment, in animal, human intestine/faeces, skin surfaces, contaminated substances including heroin. The disease occurs sporadically, affects unimmunized, partially immunized, fully immunized who fail to maintain adequate immunity ē booster doses of vaccine. Tetanus neonat-orum caused by unvaccinated mother, home deliveries, unhygienic cutting of the umb-ilical cord. The disease is common in 3rd world countries for the high % of home deli-veries, lack of vaccination, poor sanitary conditions, causing several hundred thousands deaths every year.

Clinical picture

Presents most often after the 7th day of life (IP 3-21 days) ē short history of failure to feed, spasms are typical but the diagnosis can be mistaken for meningitis or sepsis. Generalized rigidity, painful, paroxysmal convulsions, spasm of voluntary muscles invo-lving masseters (lock jaw), facial muscles (risus sardonicus), muscles of back, neck (opist-

hotonos position), difficult swallowing, fever in 40% of cases & retained consciousness.

Diagnosis

- Spatula test: touching the oropharynx ē spatula, baby develop reflex spasm of masset-ers, bite the spatula.
- Clinical symptoms & the medical history (as home delivery, unimmunized mother).

Management

- IV line.
- Separate room.
- Minimal handling.
- Penicillin G 200.000 U/Kg/day÷4 IV, for 10 days.
- Tetanus human immunoglobulin 5000 U IM.
- Metronidazole (Flagyl) active against various anaerobic bacteria & protozoa, 15 mg /Kg/day ,IV.
- Valium amp 5, 10 mg, 0.25 mg/Kg/dose IV/IM stat, suppository 5 mg, then 0.1-0.2 mg/ 3-6 hrs IV.
- Fortecortine/Decadrone amp. 8 mg/2 ml, 0.1 mg/Kg/dose, IV or IM, repeated 6 hourly, then gradual weaning.
- Mechanical ventilation may be needed in severe cases in addition to muscle relax- ant.

The spasms may continue for 3-4 wks & complete recovery may take months.

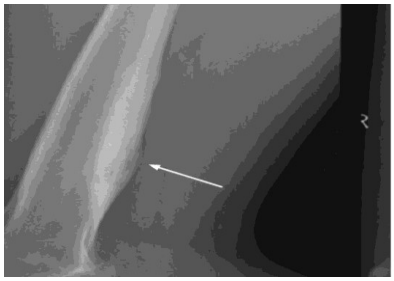
The NN mortality even ē Rx is 80%. Clinical tetanus does not produce state of immunity therefore infant who survive will require active immunization ē tetanus toxoid.

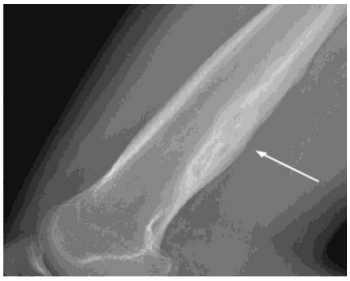
Prevention

Immunization of pregnant women ē tetanus toxoid, 2 doses of tetanus toxoid (TT) to all pregnant women between 16-36 wks pregnancy ē interval of 1-2 months between doses,

highly recommended in developing countries, if pregnant woman previously immunized, a booster dose is sufficient. If pregnant woman not immunized, the NN should be protected against tetanus by given tetanus human immunoglobulin 750 U within 6 hrs of birth.

OSTEOMYELITIS





Can affect healthy child or occur 2ry to trauma, blood stream infection. Affect any bone in the body, commonly the knee.

Clinical picture

- Local pain.
- Redness.
- Swelling.
- Tenderness.
- Fever.

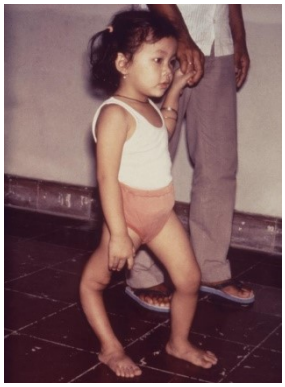
Investigations

◈CBC. ◈CRP: follow up. ◈ESR: follow up. ◈X ray bone: repeat after one wk, it takes 1-2 weeks for changes to appear as osteolytic lesions, periosteal reaction as a result of new bone formation. ◈Bone biopsy for identification of pathogen may needed.

Management

Flummox (Amoxicillin + Flucloxacillin) PO & IV, syrup 250 mg, amp 500 mg, dose 150 mg/Kg ÷ 3 for 10 days or Claforan amp. 250 mg, 50 mg/Kg ÷ 2 IV/ IM for the same period.

POLIO



Caused by Polio virus. Incubation period 1- 2 wks, characterized by selective affection of motor nerves, AHC of spinal cord. Any group of muscles can be affected as LMNL (loss of muscle tone, power, reflexes, absence of Babiniski sign, presence of fasciculations, intact sensation). May affect respiratory muscles. Bulbar palsy when affect bra-in medulla & cranial nerves 9, 10, 11, 12, resulting in dysphagia, dysphonia.

Clinical picture

Prodromal stage: URTI for few days, followed by paralytic stage, affected limb is painful at first but ē repeated passive movement pain subside. The sensation is intact. 2/3 of cases will have residual neurological seqelae.

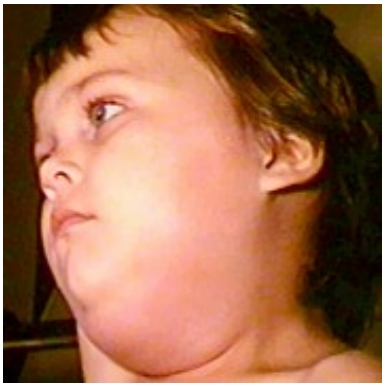
Investigations

- Isolation of virus from stool in prodromal stage, or blood in the paralytic stage.
- PCR.
- Polio antibodies (IgM, IgG).
- CSF: ⇧cells, ⇧ proteins & normal sugar.

Management

- Complete rest for 2 wks in the maximum functioning position of the affected limb
- Ventilator care in case of affection of respiratory muscles.
- Physiotherapy after the 2 wks.

MUMPS



Viral infection. The IP 2-3 wks, peak is in June, July, August, single attack cause permanent immunity. Infects salivary glands, especially the parotid gland. Spread through airborne transmission of respiratory droplets by sneezing & coughing. Also spread from direct contact ē saliva.

Clinical picture

Primary symptom is swollen checks. The swelling is elastic, painful, better seen than felt, at angle between mastoid process & mandibule, pushing ear lobule upward, stensen duct opposite the upper second molar teeth is red & edematous. Can also cause fever, headache & malaise may be presented or complicated by encephalitis or orchitis (common) or oophritis (rare).

Management

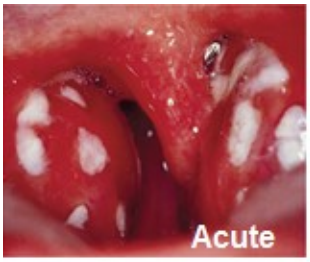
☆ Mouth hygiene care; Tantum Verde mouth wash 1 tsp as gargle 3-6 times daily, or Sulfa Buracyl 1 tsp over one cup of warm water gargle 3-6 times daily.

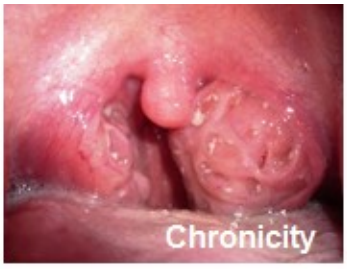
☆ Mumps is infective for 10 days after appearance of swelling.

☆ It is important to do audiogram after pt cure because it is commonly cause unilateral nerve deafness.

☆ It is Important to do urine analysis after cure as it may cause diabetes mellitus.

TONSILLITIS





Mainly a disease of childhood but is also seen in adults. May occur primarily as infection of the tonsils themselves or may secondarily occur as a result of URTI following viral inf.

Organisms

- Beta-haemolytic streptococcus.
- Staphylococcus.
- Haemophilus influenza.
- Pneumococcus.
- The part played by viruses in acute tonsillitis is unknown.

Clinical picture

Common in cool rainy months, of sharp onset, painful swallowing, tender cervical LNs, if viral will be associated ē running nose cough, hoarseness, important sign of chronicity is the marked flush of the medial border of anterior faucial pillars. Quinsey is rare complication (peritonsilar abscess), pushing tonsil medially towards uvula ass-ociated ē swollen soft palate.

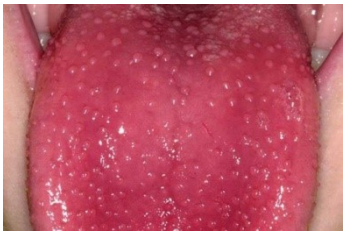
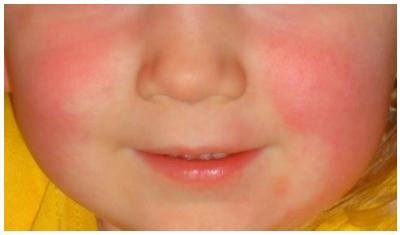
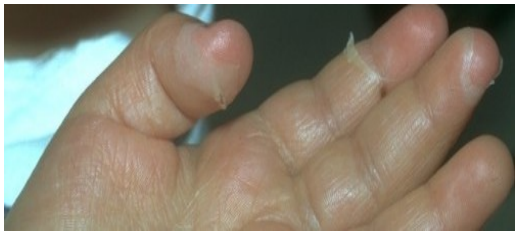
Management

•Cetal drops 100 mg, syrup 250 mg, suppository 120 mg, maximum total daily dose 1200 mg. Marcofen pediatric suppository 100 mg, 1 X 2 for baby >6 months.

•Augmentin syrup 156, 312, 457 mg, tab 612, amp 600, 50 mg /Kg ÷3 X 5 days (Beta-lactamase Inhibitor).

•Tonsillectomy in chronic cases, associated ē improve appetite, activities, happiness.

SCARLET FEVER

Group A B hemolytic streptococci, affect commonly school age children, 2-4 days post streptococcal pharyngitis.

Clinical picture

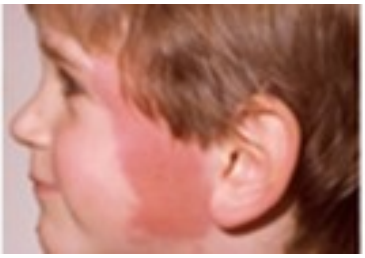
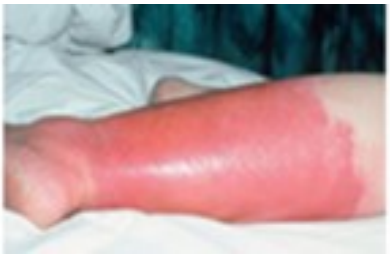
Presented ē fever, headache, sore throat, white strawberry tongue, flushed face ē circumoral pallor, unwell, rash may extend to whole body, rough 'sand paper' skin, desquamation after 5/7 days, particularly soles & palms.

Investigations

- Throat swab .
- ASO titer.

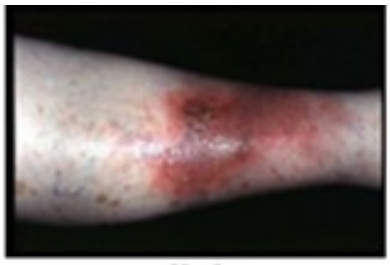
Management: Penicillin 10 days.

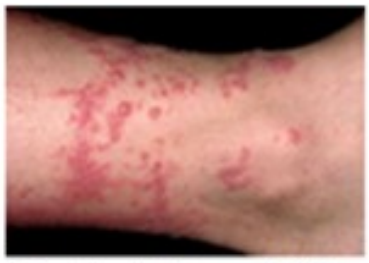
ERYSIPELAS

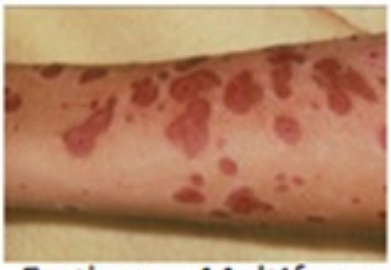



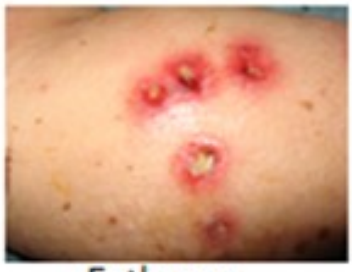
Cellulitis & Erysipelas are skin infections that develop as a result of bacterial entry via breaches in the skin barrier. Cellulitis & Erysipelas manifest as areas of skin erythema , edema, & warmth. They differ in that erysipelas involves the upper dermis & superficial lymphatics, whereas cellulitis involves the deeper dermis & SC fat. Erysipelas is caused by strept pyogenes, or staph aureus , may be complicated by sepsis.

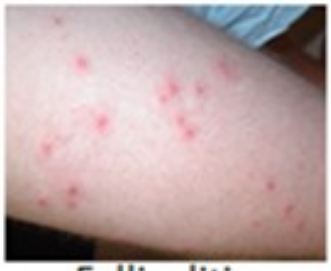
Erysipelas	Cellulitis
Abrupt onset ē fever	A low grade fever may be present ē a less abrupt onset
The skin is bright red	The skin is dull red
A spreading, hot, tender plaque ē well-defined border	The border is less well defined, fades into the surrounding skin
Vesicles & bullae may be present	No blisters

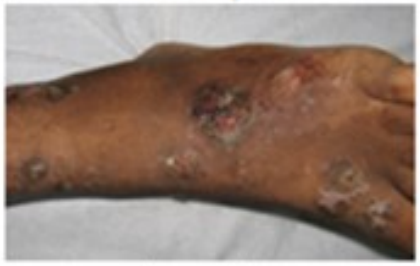

Cellulitis


Contact Dermatitis


Erythema Multiforme


Ecthyma

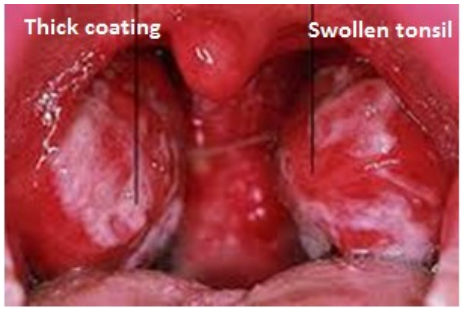

Folliculitis


Impetigo

Management

Penicillin G ampule 1000.000 U, 100.000U/Kg/day ÷ 4 IV, for 2 wks. Flummox syrup 250 mg, cap 250, 500 mg, amp 1000 mg, IV/IM, 50-100 mg/ Kg ÷ 3 X 10 days (Amoxicillin + Flucloxacillin). Avil retard tab. 1 X 1 daily. Reparil tab. 1 X 3 daily (antioxidant, anti-inflammatory). Dermovate cream 1 X 2 daily (steroid).

INFECTIOUS MONONUCLEOSIS



Epstein Barr Virus, The IP 1-2 months, common in age 2-4 yrs & in adolescent.

Clinical picture

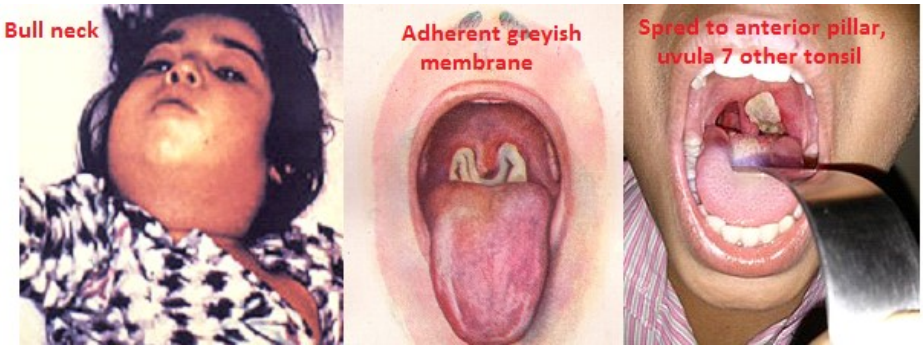
Marked inflammation & congestion of tonsils, fever continue for 2-4 wks, generalized lymphadenopathy in 90% of cases, hepatosplenomegaly in 50 % of cases, in addition to malaise & fatigue.

Investigations

- CBC: leucocytosis, >20.000/cm^3 mostly lymphocytes, atypical lymphocyte.
- Antibodies: IgM for EBV in acute illness, IgG for EBV in chronic illness.
- Paul Bunnel test: for a titer 1/200.
- Monospot test: positive.

Management: no specific treatment.

DIPHTHERIA



An acute toxic infection caused by Corynebacterium diphtheria & rarely toxigenic strains of Corynebacterium ulcerans. Aerobic, non- capsulated, non-spore forming, mostly non-

motile pleomorphic, gram +ve bacilli. Differentiation of C. Diphtheria from C. Ulcerans is based on urease activity, C. Ulcerans is urease +ve. There is 4 types of C. Diphtheria biotypes including; Mitis, Intermedius, Belfanti & Gravis. The differentiation is by colonial morphology, hemolysis & fermentation reactions.

Clinical picture

Pharyngeal (commonest), nasal or conjunctival, pt looks toxic, feverish, dirty white membrane over tonsil, gradually spread to anterior pillar, uvula & other tonsil, trial of removal result in underneath bleed.

Investigations

☆ Pharyngeal swab urgent for culture. ☆ CBC.

Management

Diphtheria antitoxin ½ IV, & ½ IM 20.000-80.000 U/day until complete recovery, after doing allergy test by diluting to conc. 1:100 - 1:1000, 0.1 ml, ID, observe for 1 hour for any local reaction, if sensitive use adsorbed purified toxoid, if not available do desensitization as follow;

0.1 mL 1/20 conc. + 0.05 ml adrenaline SC. After 30 min

0.1 mL 1/10 conc. + 0.05 ml adrenaline SC. After 30 min

0.01 ml full conc. + 0.05 ml adrenaline SC. After 30 min

0.1 mL full conc + 0.05 ml adrenaline SC. After 30 min

1/2 mL full conc + 0.05 ml adrenaline SC. After 30 min

Full dose IM ē the presence of adrenaline, decadron ready for use in case of need.

Penicillin G 200.000U/Kg/day ÷ 4 for 10 days, or

Erythromycin 500 mg X 4, 50 mg/Kg/day ÷ 4 for 10 days in case of penicillin allergy.

Isolation until 3 throat swabs/day are -ve.

OTITIS MEDIA

Peak is 6-12 months, 50% at age 3 year, 25 million visit per year to pediatricians for otitis media in USA, bacterial in origin in 60% of cases, viral 15% & nonspecific in 25% most bacterial infection are due to streptococcal pneumonia & Haemophilus influenza, common ē tonsillitis & URTI, postmortem examination of prematures shows 75% of them had otitis media.

Clinical picture

Fever not required for diagnosis, visualize the drum for loss of luster, presence of inflammation or dilated vessels. Check for swelling or tenderness behind ear pinna (over mastoid process) ẃ suggest mastoditis.

Secretory otitis media: lead to conductive deafness, common in age group 2-5 years, uni or bilateral, nerve conduction is better than air conduction. Treated ē nasal decongestant, antihistaminic & myringotomy ē no improvement, ventilation tube (grommet) ẃ extruded by itself.

Swimming pool otitis media: chemical irritation, lead to obstruction of Eustachian tube & ear pain, resolve within 1-2 weeks, .Treated ē Otal drops, Oto calm drops, anti-allergic syrup, in addition to antibiotic.

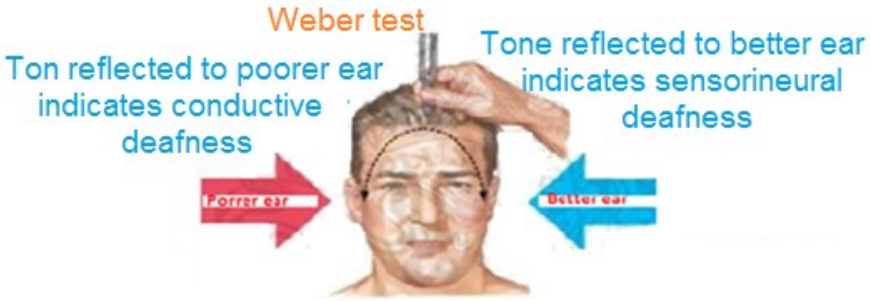
Acoustic neuroma: in adults presented ē loss of equilibrium & balance, associated ē nerve deafness & nystagmus, MRI is diagnostic. Treated surgically by excision of the benign tumor ẃ is attached to the 8th cranial nerve.

Conductive & Nerve deafness differentiation

Weber test: tuning fork over middle of top of head, hearing is better on ear affected by conductive deafness because there is masking in the intact ear from combination of transmission of sound through air & bone conduction on the intact ear.

Rinne`s test: tuning fork over the mastoid process, no perception of sound on the ear affected by nerve deafness.

Audiogram is confirmatory



Management

Augmentin syrup 156, 312, 457mg, tab 612 mg, 50mg/Kg ÷3 X 5 days.

Otal ear drops.

Cetal drops 100, syrup 250, suppository 120, maximum daily dose = 1200 mg/day.

Marcofen pediatric syrup or suppository 100 mg, 1 X 2 for baby > 6 month age.

Complications

- Chronic otitis media.
- Purulent otitis media.
- Perforation.
- Mastoiditis.
- Brain abscess.

PYREXIA OF UNKNOWN ORIGIN

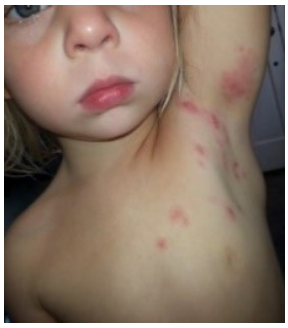
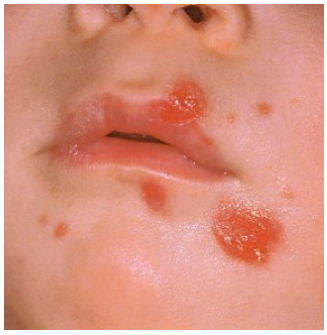
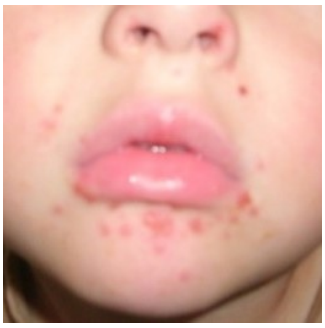
Fever ẃ lasts for > 2 wks ēout reaching a clinical or laboratory diagnosis. If fever is more by night you can think in TB, typhoid or brucellosis. If fever is associated ē bradycardia, think in typhoid or yellow fever.

Investigations

- CBC: may show Leukemia, or marked ⇧ of eosinophilic count in case of parasite infect.
- Antibodies: for parasitic infestation as; Fasciola Hepaticus, Bilharziasis, Amoebiasis, Toxocariasis, Hydated, Toxoplasmosis, Leishmaniasis, Trichenella.

- Antinuclear antibodies: for juvenile Rh^{ed} arthritis.
- Double strand DNA: for SLE.
- Blood film: for malaria.
- U/S abdomen: for liver abscess, abscess, tumors as neuroblastoma.
- Urine catecholamine VMA, HVA: for neuroblastoma.
- CXR: for lung abscess.

IMPETIGO

Bacterial skin infection. Often called school sores, mostly affects children. Is quite contagious. Commonly staph Aureus or Group A Strep Pyogenes. Classically presented ē ruptured vesicles ē honey-colored crusting, may be bullous, more common in pre-existing ting skin disease, rapid spread. Commonly starts around face/mouth.

Management

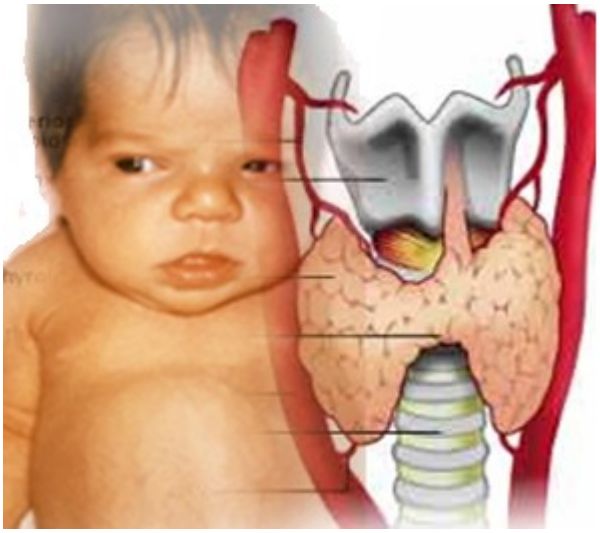
During the infectious stage (while the impetigo is oozing or crusted); cover the affected areas, avoid close contact ē others, affected children must stay away from school until crusts have dried out, use of separate towels & flannels. Change & launder clothes & linen daily. Topical Fusidic Acid or oral Flucloxacillin; Flummox syrup 250 mg, capsule 500 mg, amp 1000 mg (in severe cases), dose 50-100 mg/Kg ÷ 3 for 5 days. Ectometherine ointment after warm bath & good rubbing of skin.

CHAPTER VIII

ENDOCRINAL DISORDERS

- Congenital Hypothyroidism
- Hypopituitarism
- Syndrome Inappropriate Antidiuretic Hormone Secretion
- Cushing Syndrome
- Neuroblastoma

CONGENITAL HYPOTHYROIDISM



Screening program is applied in Egypt for early detection of such cases by testing blood sample (heel prick) on 7th- 10th day of life, if TSH is high, confirmatory test by T_3 & T_4 to be done. Hypothyroidism should be excluded in every infant.

Incidence: 1/3000 live births

Clinical picture

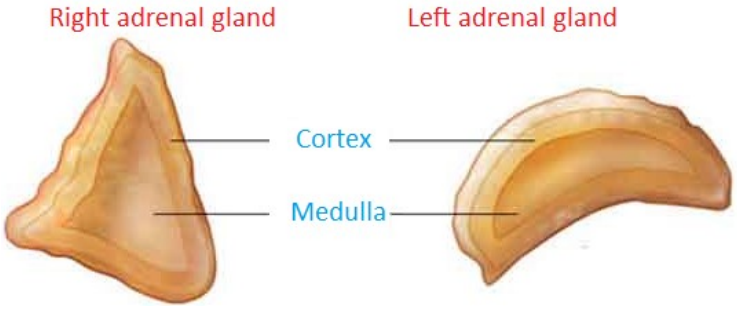
- Usually present ē unconjugated hyperbilirubinemia, but may be conjugated & associated ē NHS.
- Prolonged jaundice.
- Lethargy & Reluctant to feed.
- Physically & mentally constipated.
- Big tongue.
- Cold skin.
- Delayed milestone.

Investigations: TSH, T_3, T_4, & Bone age.

Management: Eltroxin 50, 100 ug tab., 4 ug/Kg/day, ē monitoring TSH.

CONGENITAL ADRENAL HYPERPLASIA

Chromosome 6, Gene CYP 21 A 2, Location 6 p 21.33, described 1960.



Introduction

Each adrenal gland has 2 parts:

*Adrenal Medulla (inner area), secretes catecholamines ẃ mediate stress response (help prepare a person for emergencies). The catecholamines include; Norepinephrine, Epinephrine & Dopamine.

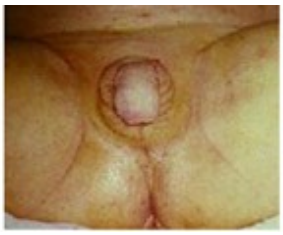
*Adrenal Cortex (outer area, encloses Adrenal Medulla.), secretes steroid hormones. The steroid hormones include; *Mineralocorticoids*: are essential to maintain sodium & fluid balance. & *Glucocorticoids*: exert a widespread effect on metabolism of carbohydrates & proteins. & Sex hormones (secondary source). Each of steroid hormones secreted from special zone of adrenal cortex, the Zona Glomerulosa produces predominantly mineraloco-rticoids. Zona Fasciculata, produces predominantly glucocorticoids. Zona Reticularies, produces predominantly androgens.

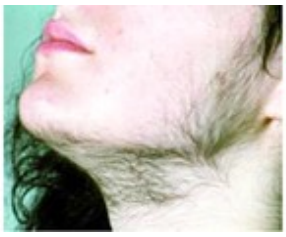
CAH is familial disorder of adrenal steroid biosynthesis. The defect is expressed as adrenal enzyme deficiency. 5 major enzyme deficiencies are clinically important: 21 Hydroxylase, 11-b-Hydroxylase, 17-a-Hydroxylase, 3-b-Hsteroid Hydrogenese & 20, 22 Desmolase deficiency. The enzyme deficiency causes reduction in end-products & accumulation of hormone precursors & ⇧ ACTH. The clinical picture reflects the effects of inadequate production of cortisol & aldosterone & ⇧production of androgens & steroid metabolites.

21 Hydroxylase deficiency: most common type, accounts > 80% of cases.

Incidence: 1: 5000- 15000 live births. The gene is located on the short arm chromosome 6 near the C 4 locus in close association ē HLA genes. Heterozygous carriers can detected by ACTH stimulation test. It is characterized by ⇩ production of cortisol & aldosterone & ⇧ production of progesterone; 17-OH-progesterone & sex steroids. The urinary steroid metabolites (17-ketosteroids & pregnanetriol) ⇧ above normal levels. ⇩ secretion of aldosterone results in salt loss ē hyponatremia & hyperkalemia; plasma renin activity is therefore ⇧. In partial enzyme deficiencies, the aldosterone deficiency is not expressed, pt remain normonatremic & normokalemic. The ⇧ androgens causes virilisation of girls, or ambiguous genitalia &dark scrotum in boys. There are 2 forms, classic early virilisation type ē or ēout salt-losing crisis & non-classic type ē late onset virilisation. Male babies ē non salt-losing (non-classic type) remains asymptomatic till childhood when showing sexual precocity signs. Because members of the same family may have classic, or nonclassic & asymptomatic forms, the disorder may be due to allelic variations of the same enzyme.

Increase androgen production result in Ambiguous genitalia in newborn Girl classic form





Women with excess hair growth. non classic form

Clinical Picture

Commonly presented ē:

- Ambiguous genitalia, labial fusion, clitromegaly, or penile, testicular enlargement & will developed muscles in boys, early appearance of pubic, armpit hair, hyperpigmentation of genitalia.
- Advanced bone age.
- Poor feeding.
- Diarrhea.
- Dehydration.
- Electrolyte disturbances.
- Arrhythmia.

Investigations

- Chromosomal studies: a Karyotype is essential in the evaluation of infant ē ambiguous genitalia in order to establish the chromosomal sex.
- Neonatal screening: by measuring 17-OH-progesterone from heel blood samples collected on filter paper. This approach allow early identification of newborns ē CAH & prevent salt wasting crisis in boys who are unrecognized at birth. It also identifies the completely virilised girls ē ambiguous genitalia who may be mistaken for boys ē cryptorchidism.
- Prenatal diagnosis is possible through biochemical & genetic tests.
- ⇧ ACTH.
- ⇧ 17-OH- progesterone, or 11 β hydroxyl deoxy progesterone in blood & urine (⇧ 20 times).
- Advanced bone age (from excess androgen).

Management

Replacement therapy: (4S) Steroids, Sugar, Salt, & Na bicarbonate for metabolic acid osis. Correction of dehydration, acidosis & electrolyte imbalances. Prednisolone (is 5 time potent than hydrocortisone), 1mg/Kg/day. Florinef (9 α fludrocortisone) 0.1mg tab 1X1 daily in case of Aldosterone deficiency

11- B - Hydroxylase Deficiency

Accounts for 5-10 % of cases of CAH. Gene is located on the long arm of chromosome 8. Characterized by ⇩plasma renin activity & ⇧high serum 11-deoxycorticosterone & 11-deoxycortisol concentrations ē ⇧of its urinary metabolites (compound-S).Because of the strong mineralocorticoid activity of deoxycorticosterone, the condition is characterized by salt retention, ⇧ BP & hypokalemic alkalosis. ⇧ Plasma androgens may cause virilisation of female fetus.

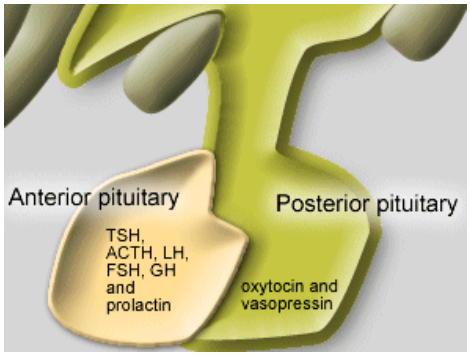
17- α – Hydroxylase deficiency

Genetic defect is on chromosome 10. Presents ē similar features of those of 11-Hydroxylase deficiency except that androgens are low, so no virilisation in girls & genitalia is ambiguous in boys.

3- B- Hydroxysteroid dehydrogenase deficiency

Very rare disorder that result in accumulation of DHEA, ẃ is converted to testostero ne in peripheral tissues. It can cause virilisation of female fetus & leads to ambiguous genitalia in the newborn.

HYPOPITUITARISM



50% of cases are associated ē NHS, may be due to hypothalamic dysfunction or deficiency of anterior/posterior pituitary function.

Clinical picture

•NN jaundice. •Short stature. •Normal mentality. •Small lips. •Smooth skin. •Silky hair •Hypoglycemia especially after protein diet. •May associated ē headache, vomiting (tumor) in older children.

Investigations

- Growth hormone level in blood is low.
- X ray skull (tumor).
- C-T scan brain .

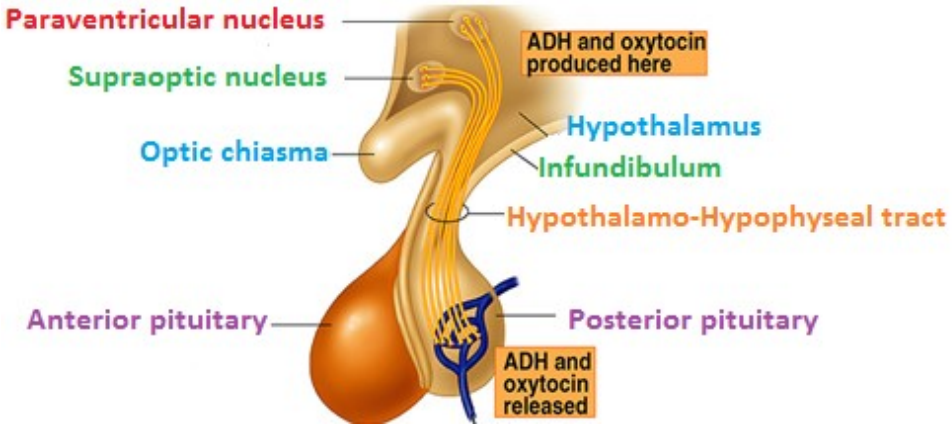
Exercise test (older children): severe exercise for 10 min & measuring the G.H. before & after will show no increase (normally is associated ē rise in G.H. 15 mu/l) .

Management

Surgical.

Medical: genotropin 4, 16 units/vial, dose 0.1 unit/Kg/day SC, Rx should be continued for several yrs until closure of epiphysis occurs.

SYNDROME INAPPROPRIATE ANTIDIURETIC HORMONE



The antidiuretic hormone, also called vasopressin as it has potent vasoconstrictive effect, formed in the supraoptic & Para ventricular nuclei of the hypothalamus transported to the posterior lobe of the pituitary gland & stored. Act on the distal tubule & collecting duct in the kidney, promte water reabsorption & cause ⇧ of the extracellular fluid volume & ⇩ of urinary output, its release is stimulated by:-

- ⇩ of BP • ⇩ of blood volume • ⇧ of plasma osmolality • Fear, pain & anxiety.

Causes

- Positive pressure ventilation.
- Chronically ill or malnourished children.
- Mal ignancies, lymphomas, sarcomas.
- Meningitis, cerebral Hge or tumors, multiple sclerosis.

- Pneumonia, chronic obstru- ctive pulm. diseases, bronchial asthma, TB, resp. failure.
- Jullian Barre syndrome.
- Bone marrow transplantation, stem cell transplant.
- Drugs as; ant malignant, antidepressant, antipsychotic, chloropropamide, loop diuretics 'commonly thiazide'.
- Idiopathic.

Diagnosis

IADHS is the most frequent cause of hyponatremia in hospitalized pts.

Symptoms of hyponatremia; nausea, vomiting, cramps, agitation, changes in mental status, seizures, bulbar palsy, hypothermia, coma.

#⇧ Intravascular volume from water retention.

⇧ CVP.

⇩ Urine output,

⇩ Serum sodium to < 135 mEq/l (delusional hyponatremia) & ⇩ of serum chloride.

⇩ Serum osmolality to < 275 mOsm/l.

#⇧ Urine sodium (concentrated urine).

⇧Urine specific gravity to > 1.020

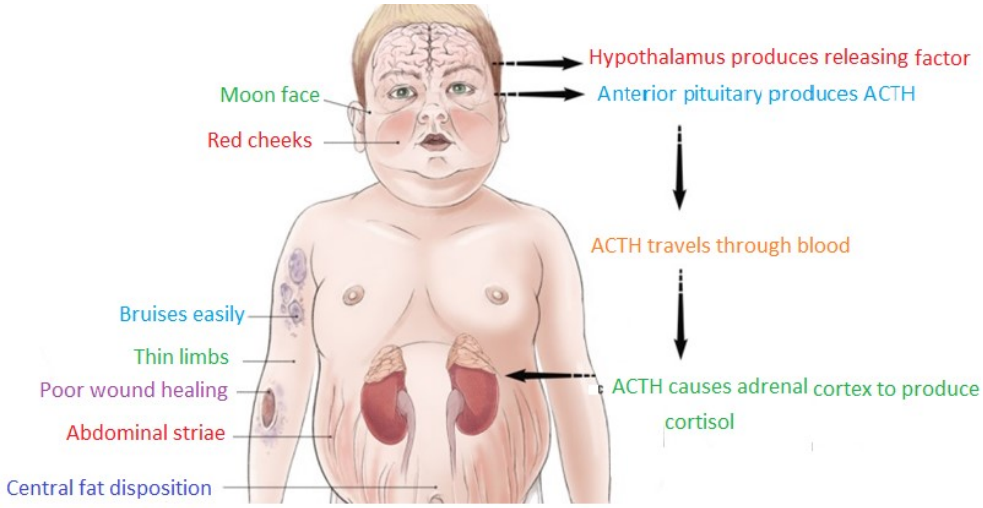
⇧ Urine osmolality to > 100 mOsm/l.

Normal renal function.

Management

- Treatment of underlying cause.
- Normalize serum sodium over 24-48 hours.
- Fluid restriction.
- Loop diuretics.

CUSHING SYNDROME



Excess production of cortisol due to excess release of ACTR factor from hypothalamus, or adrenal hyperplasia or 2^{ry} to ⇧ ACTH of pituitary or excess intake of cortisone.

Clinical picture

Short stature, obesity, moon & plethoric face, hirsutism, skin striae, easy bruising.

Investigations

- CBC: polycythemia
- Cortisol level in blood: marked ⇧
- U/S or CT scan of adrenals.
- C-T scan brain (for pituitary).
- X ray skull (pituitary).
- Loss of normal diurnal variation of cortisol (highest at 9 am, lowest at 9 pm).
- Dexamethasone suppression test: 1 mg Decadrone IM before sleep, measuring cortisol level in morning, no suppression means adrenal hyperplasia.

Management: surgical.

Metyrapon block synthesis of cortisol.

CHAPTER IX

GASTROENTEROLOGY & METABOLISM

- Vomiting
- Gastro oesophageal reflux
- Hirschsprung Disease
- Diarrhoea
- Marasmus
- Kwashiorkor Disease
- Inborn Errors of Metabolism
- Rickets

VOMITING

Vomiting in neonatal period

Frothy, mucoid vomiting: caused by oesophageal atresia or TOF. Presented ē cough, cyanosis during feeding, continuous drooling frothy material.

Bloody vomiting: result from cracked nipple, swallowed maternal blood, vitamin K deficiency, hemorrhagic disease neonate, trauma from oro or nasogastric tube.

Milk vomiting: feeding problem, NEC, sepsis, UTI, IC Hge, adrenogenital syndrome, or digitalis toxicity.

Bile stained vomit: yellow, green, sometimes consist of milk result from; intestinal obstruction, atresia, volvulus, Hirschsprung disease, NEC. Usually associated ē abdominal distension, constipation or infrequent stool, visible peristaltic intestinal movements, forceful expulsion of milk through nose, mouth.

Vomiting in post natal period

⅄Congenital hypertrophic pyloric stenosis ⅄Intestinal intussusception, maximum age 3-12 months ⅄Gastro oesophageal reflux ⅄Adrenogenital syndrome.

Investigations: # Plain X Ray erect & supine # Sonar abdomen.

MALLORY WEISS SYNDROME

Associated ē severe vomiting ẃ result in tear of mucosa of gastroeosophageal junction, causing severe hematemesis.

GASTRO OESOPHAGEAL REFLUX

Common in the first 6 months of life, commonly resolve spontaneously within few months, result from low pressure of esophageal sphincter & unorganized motility.

Diagnosis

Barium swallow: to R/O oesophageal stenosis, trachioesophageal fistula or hiatus hern-

ia #24 hours monitoring of lower oesophageal PH.

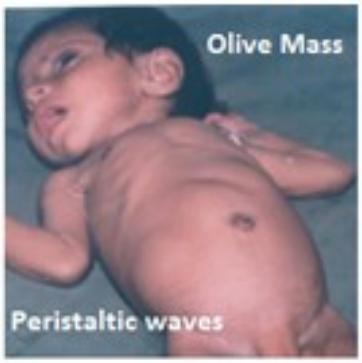
Complications

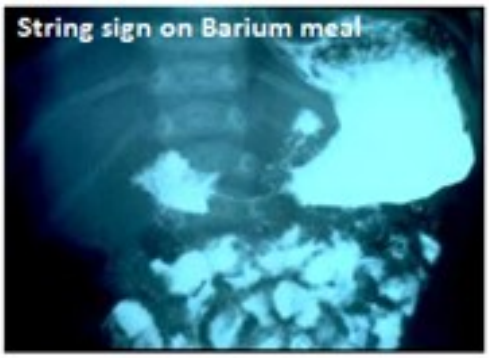
- Recurrent pneumonia.
- Failure to thrive.

Management

- Proper feeding, eructation, prone position, head to one side, elevation of head of bed 30^0, nursing baby while head in sitting position.
- Thickening of feeds ē rice or cereals.
- Special milk formula (antiregurgitation formula), Frisovom milk formula, S 26 (AR).
- Surgery in severe cases.

CONGENITAL HYPERTROPHIC PYLORIC STENOSIS





Incidence

1/5000 babies, more in boys, start to appear on 2nd- 3rd wk of life. It is one of most common GIT disorders in early infancy. Hypertrophy of the circular muscles of pylorus results in constriction & obstruction of gastric outlet.

Diagnosis

- Projectile vomiting after feeding.
- Dehydration & metabolic alkalosis.

- Peristaltic movements from left to right, small olive shaped mass in epigastrium or under the liver.
- Feeding test: diagnostic.
- X ray abdomen: erect/supine-double bubble appearance.
- Barium meal: string sign.
- Abdominal sonar: diagnostic.

Management

- IVFs, correction of dehydration & metabolic alkalosis.
- Surgical operation (Rammstdt operation).

HIRSCHSPRUNG DISEASE

Incidence

1/ 5000 live birth. The most common chromosomal abnormality associated ē HSCR is Down syndrome ẃ occurs in 2-10 % of individuals ē HSCR.

Clinical Picture

- Constipation. •Abdominal distension. •Projectile vomiting. •Bile stained vomitus.

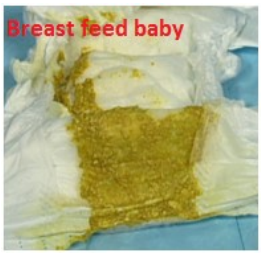
Diagnosis

- PR exam: small empty rectum, hard stool above & explosive passage of stool.
- Plain X ray: multiple fluid levels.
- Barium enema: characteristic transitional zone, funnel shaped separate normal mu-

cosa (above) from the aganglionic segment.

- Monomeric studies: using special balloon.
- Rectal suction biopsy is diagnostic. Management: surgical.

DIARRHEA



Breast feed baby stool is never formed, is yellowish, like yoghurt, in 1st few days he may pass stool every hour, may be mixed ē mucous or fluid, may be green in color, gradually he may pass stool every 2-3 days. While artificially feed baby stool is will formed, ē no mucous or fluid.

Types & Causes

Watery diarrhea

Rota viral infection; commonest cause in infants & children, preceded by URTI or otitis media in 50% of cases, vomiting start first & stop, then watery stool lasts for 3-5 days, can spread as epidemic in pediatric word. E. Coli. Cholera.

Bloody or Mucoid diarrhea

Enteropathogenic E coli, Salmonella, Shigella, Campylobacter, Cryptosporidium, Amoebiasis, Giardiasis. Diagnosed by stool examination & culture.

CHRONIC DIARRHEA

Lactose intolerance: frothy acidic stool (normal stool pH is 5-7), ⇩ to 4 or less. The presence of reducing substances in stool (+ve clinetest).

Treated by isomil (Soya milk) ẃ is lactose free.

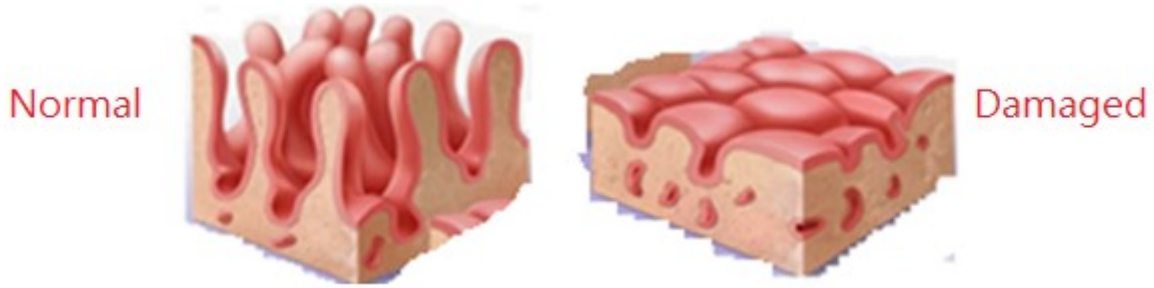
Glucose/Fructose intolerance: managed by Galactomil 18 for glucose intolerance or Galactomil 19 for fructose intolerance.

Cow milk protein allergy: diagnosed by on/off cow milk, ⇧ of eosinophilic count, rise in immunoglobulin E. Treated by isomil milk, improves within 1 year.

Irritable bowel syndrome: undigested food particles, may associated ē food allergy, improves within 3-4 yrs.

Giardiasis: treated by Flagyl syrup 125 mg/tsp, 15 mg /Kg/day÷3.

Coeliac disease:



An autoimmune disease caused by consumption of gluten containing products (e.g. wheat) in predisposed individuals. Characterizes by inflammation of the small intestine resul-ting in atrophy of the intestinal villi & reduced resorption of nutrients. The symptoms of the disease are diarrhea, vomiting, abdominal pain, wt loss, growth retardation & anemia

Diagnosis

Depend upon the following criteria:-

1- Presence of symptoms.

2-Detection of antibodies against Endomysium (EmA), tissue Trans-Glutaminas (tTG) or Gliadin.

3- a +ve small intestine biopsy (loss of normal ratio of villi to crypts ẃ is normally 1:4) using a crossly, Watson capsule, this changes in intestine can also seen in case of Crohn`s

disease, kwashiorkor, Tropical sprue, Bilharziasis & Lymphoma.

4- Disappearance of symptoms & fall in antibody titer on free gluten diet.

Treatment: Glutein free diet for 2 yrs, then biopsy, monitoring antibodies for α gliadin, doing glutein challenge test, 10 gm PO & monitoring antibodies & taking biopsy.

Celiac disease & relation to Autism

A significant number of sera samples from children ē autism exhibited elevated Anti-Gliadin. So it`s recommended to do the Anti-Gliadin test for pt ē autism.

Comparison between Anti-Gliadin & Anti-Ttg (IgA & IgG)

		Diagnostic accuracy			Sensitivity
		0-2 Years	2-4 Years	> 4 years	
IgA	Anti Gliadin	89%	87%	89%	84%
	Anti- tTG	91%	89%	96%	97%
IgG	Anti Gliadin	91%	89%	92%	96%
	Anti- tTG	72%	76%	67%	65%

Cystic fibrosis: common AR in Europe.

Incidence: 1/2000 baby.

Clinical picture: presented ē chronic diarrhea, steatorrhoea, pancreatic insufficiency, meconium ileus, recurrent RTI, bronchiectasis.

Diagnosis

- +ve sweat chlorid test, > 60 (pilocarpine iontophoresis). False +ve sweat chloride test seen in glycogen storage dis., adrenal insufficiency, diabetes insipidus & G6PDD.
- Pancreatic enzymes.
- Chest X ray show characteristic foamy appearance in case of bronchiectasis.

Management

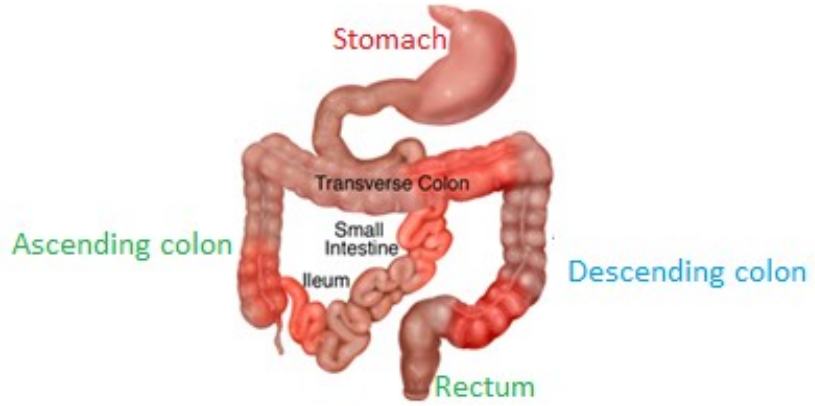
include (4 P) Pancreatic enzymes,

Postural drainage,

Penicillin &

Physiotherapy.

Crohn`s disease



In children presented ē bloody offensive stool, frequent diarrhea, failure to thrive.

Diarrhea investigations

- CBC.
- Stool analysis & culture.
- Serum electrolytes.
- Reducing substances in stool.
- Fat in stool in case of chronic diarrhea; normally fat is < 30% & more than that mea-ns steatorrhoea.

Diarrhea Management

- Diet regime: boiled rice, rice budding, carrot, smashed potato, apple, lemon juice, jelly, tea & toast.
- Rehydration: ē the presence of dehydration, calculate deficit; mild 2-5%, moderate 5-10%, severe >10% of body weight.
- Compensate for every stool motion by one cup of extra fluid.

- Daily fluid requirement: calculated as follow:

If BW < 10 Kg = BW X 100 ml.

If BW 10-20 Kg = 1000 + 50 ml for each Kg >10.

If BW 20-30 Kg = 1500 + 25 ml for each Kg> 20.

- Electrolytes: Kcl amp 20%, 1 mg/Kg, after passing urine +Ca gluconate amp 10% 1 ml/Kg. The daily requirement of Na or CL is equal to 2 meq/Kg/day.
- Calories: form 0-10 yrs. age baby require 100 cal/Kg/day (the BMR for NN is equal to 45 cal/Kg/day while for adult is equal to 20 cal/Kg/day).
- Specific treatment directed to the cause.

Composition of intravenous fluids

*Normal saline: Na^+ 154 meq/L, CL^+ 154 meq/L,

*Ringer solution: Na^+ 150 meq/L, CL^+ 150 meq/L, K^+ 4 meq/L, Ca^+ 4 meq/L.

*Ringer lactate: Na^+ 30meq/L, CL^+ 130meq/L, K^+ 4meq/L, Ca^+ 4 meq/L, HCo_3 30meq/L.

CHOLERA

Caused by bacteria Vibrio Cholera. Mode of infection through contaminated water, food, poor hygiene & poor sanitation of environment. Incubation period: 1-2 days.

Clinical picture

- Effortless, painless, severe watery diarrhea (rice water stool) up to the loss of 1 L/hour.
- Vomiting in severe cases. • Rapid development of marked dehydration, metabolic acid-osis, hypokalemia & hypotension. • Electrolyte imbalance. • May be fatal within hours.

Investigations: • Serum electrolytes • Stool culture, agglutination • Blood gases.

Management

- Rapid correction of; dehydration, electrolyte imbalance. Using normal saline or ringer lactate, Kcl 15%. $NaHco_3$ in case of metabolic acidosis, saline ēout alkali leads to pulm. edema especially if pt is oliguric & excess fluids given.
- Septrin syrup 40 mg TMS + 200 mg SMZ, tab 80 + 400 mg, dose TMS 4-8 mg/Kg÷2 for 3 days. or
- Erythromycin syrup 200 mg, tab 500 mg, 30 mg/Kg/day ÷ 3 for 3 days or
- Quinolone group (e.g. Ciprofloxacin tab.) 250, 500 mg, 1 X 2 X 5 days for adults, amp 200 mg/100 ml for IV infusion.
- Isolation & notification of authorities.

BOTULISM

Ingestion of toxins clostridium botulism in canned or contaminated food stuffs. There is marked affinity of toxin to neurons & nervous system.

Clinical picture

Nausea & vomiting, then constipation, flaccid paralysis, cranial nerve palsy, respiratory failure & may progress to death (but mostly it carry good prognosis).

Investigations

Stool culture for clostridium botulinium.

Blood for clostridium toxins.

Management

- IVFs .
- Induction of diuresis (forced alkaline diuresis); normal saline 20 ml/Kg over 1-2 hours, followed by Lasix 1 mg/Kg IV.

- Laxative: Mg SO_4 sachets.
- Botulinium immunoglobulins.
- Intubation needed in 50% of cases.

TYPHOID FEVER

Caused by salmonella typhi, paratyphi A, B, C. Incubation period 1-2 weeks. Mode of transmission include; fecal oral, flies, poor sanitation, restaurant, food dealers.

Clinical picture

Infants: fever, vomiting, diarrhea. Children: fever more by night, malaise, headache, abdominal pain, bradycardia, rose spots by 5th day in 50% cases as macular, maculopapular rash in anterior abdominal wall, distended abdomen, hepatosplenomegaly, pulmonary rales. Diagnose highly suggested clinically by presence of fever, bradycardia, distended abdomen, rose spots on 5th day & pulmonary rales.

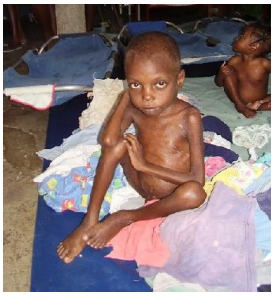
Investigations

- CBC: leucopenia (typhoid, brucellosis, TB, viral infection).
- Widal test: TO antigen titer is 1/160 or more (normally is 1/80).
- Blood culture. • Urine & Stool culture. • PCR for typhoid.

Management

Claforan "3rd gen. Cephalosporin" amp 500 mg/12 hrs. IV/IM, dose 50 mg/Kg/ day÷2, give 1st two doses & continue ē oral. OR Septrin syrup 40 TMS +200 SMZ, tablet 80+400, dose TMS 4-8 mg/Kg ÷2. Discharge after 3 consecutive stool cultures are negative. Chronic carriers are treated for 4 weeks & cholecystectomy may be needed & is curable. Vaccination for typhoid is given to travellers to endemic areas, food dealers in restaurant & streets & for health care workers.

MARASMUS



Calories malnutrition result from poverty, diminished intake, improper feeding, or ē chronic diarrhea.

Clinical picture

Marked reduction of weight, loss of SC fat, start in the thigh, spread to abdomen & face. Senile face, subnormal body temperature, bradycardia, multiple vitamins & minerals deficiency, ⇧ susceptibility to infection.

Management

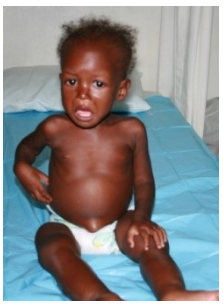
IVFs: Glucose + Saline + 2 ml Kcl, start ē 60 ml/Kg then ⇧ gradually 20 ml/day to a maximum of 120 ml/Kg/day.

Continue breast feeding or use acidified skimmed milk.

Multivitamins ē no iron, give daily requirements.

Plasma or 20 % human albumin.

KWASHIORKOR DISEASE



Protein malnutrition as a result of diminished intake, or diminished synthesis of proteins (liver disease), excessive loss of proteins (nephrosis), presence of aflatoxin in stored fo-

od (way of storage of cereals in many underdeveloped African countries).

Clinical picture

Edema, dermatitis, pigmentation of skin, hepatomegaly, weakness, multiple vitamins deficiencies, ⇧ susceptibility to infection.

Investigations

- CBC: anemia.
- Serum electrolytes: imbalance of Na+, k+, Mg+, Ph+.
- Serum proteins: hypoproteinemia.

Management

Day 1: glucose & saline 60 ml/Kg/day, correction of any electrolyte disturbances.

Day 2-7: start skimmed or ½ strength milk formula or ½ strength isomil, 60 ml/Kg/day, ⇧ 20 ml/Kg/day, up to a maximum of 120 ml/Kg/day & continue this regime for 2 weeks.

From the 4th week: start full strength isomil milk formula.

From the 5th week: start gradually introducing mixed, protein rich food ē high calories.

INBORN ERRORS OF METABOLISM

Incidence: 1/5000 live births. Suspected in baby or child ē: hepatic dysfunction, failure to thrive, unexplained neurological deterioration, metabolic acidosis, hypoglycemia, inappropriate ketosis, hypotonia, or cardiomyopathy.

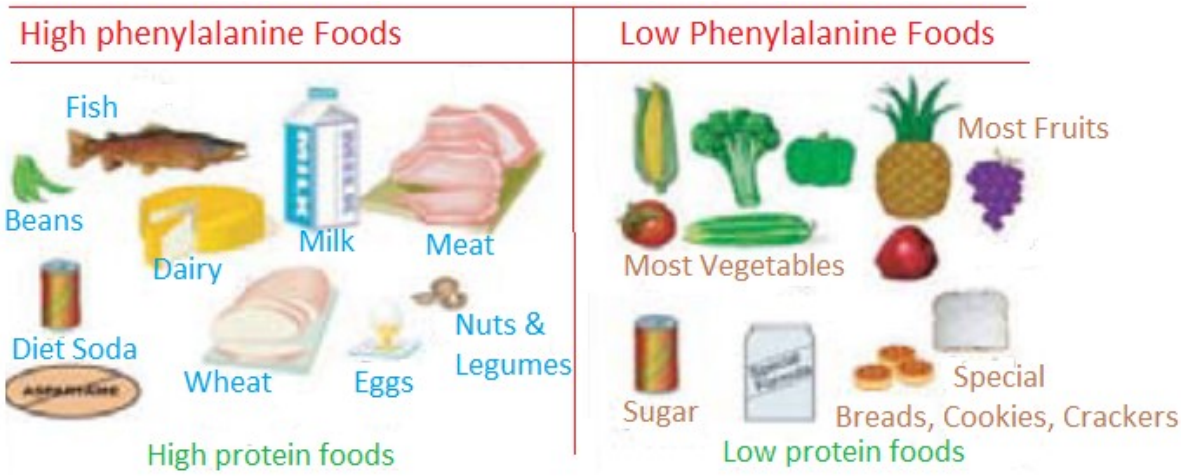
Investigations

- Plasma amino acids, ammonia, glucose & carnitine.
- Blood pH, serum electrolytes, lactate & pyruvate.
- LFTs.
- Urine for; smell, pH, ketones, reducing substance, amino acids & organic acid.

PHENYLKETONURIA

Chromosome 12, Gene PAH, Location 12 q 23.2

Discovered by Norwegian physician Dr. Ivan Folling, in 1934.



Symptoms can minimize by adherence to a strict phenylalanine free diet

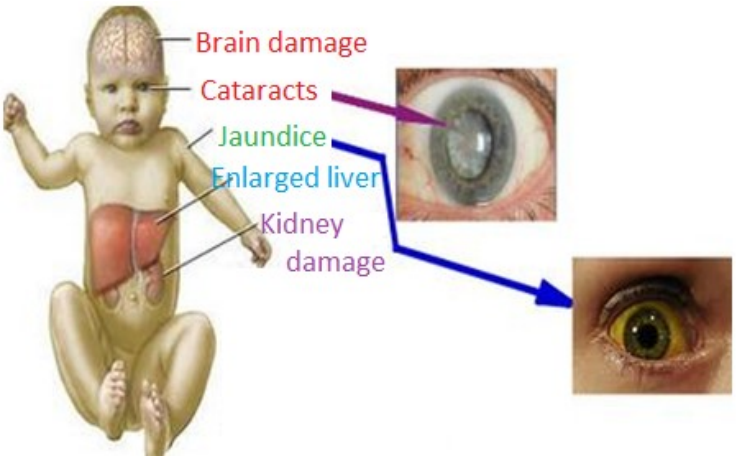
Incidence

1/10.000 Births. Deficiency of enzyme phenylalanine hydroxylase.

Clinical picture

▲Delayed milestones ▲Vomiting ▲Frequent diarrhea ▲Musty odor of skin, hair & urine ▲Skin problems &sensitivity to light ▲Irritability ▲Brain damage.

GALACTOSAEMIA



Incidence

1/50.000 live births. Deficiency of galactose 1,6 Phosphate uridyl transferase.

Diagnosis

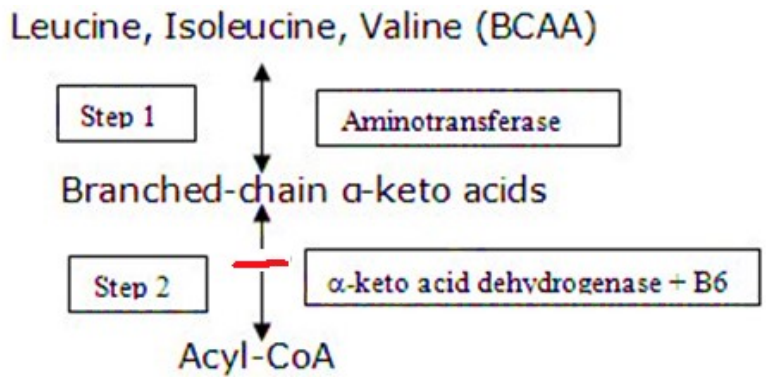
•Prolonged neonatal jaundice. •Hypoglycemia. •Vomiting in the first two weeks of life. •Liver cirrhosis. •Cataract. •Marked rise of galactose, G-1-phosphate in urine & blood. •Reducing substances in urine "galactose, galactose-1-phosphate". •U/S abdomen for liver cirrhosis. •LFTs for rise in bilirubin & liver enzymes.

Management: Galactose free milk, soya milk (isomil milk).

MAPLE SYRUP URINE

Chromosome 19, Gene BCKDHA, Location 19 q 13.2

Described by American pediatrician Dr. Menkes et. Al, in 1954



Incidence

1/180.000 Births. Disorder of branched chain amino acids metabolism; Leucine, Isoleucine & Valine. Accumulation of these 3 amino acids & their corresponding ketoacids leads to encephalopathy & progressive neurodegeneration.

Clinical picture

- Distinctive sweet odor of urine, smell is also present & sometimes stronger in ear wax
- Lethargy.
- Seizures.
- Brain Damage.
- Coma.

Investigations

- Urine Amino Acid Test:

Valine: normal level in children 17-37 & in adults: 19-74 (micro mol/L).

Leucine: normal level in children 9-23 & in adults 20-77 (micro mol/L).

Isoleucine: normal level in children 3-15 & in adults 4-23 (micro mol/L).

- Plasma Amino Acid Test for Valine, Leucine, Isoleucine(Chromatography), baby should not eat for 4 hrs before the test.

α-1- ANTITRYPSIN DEFICIENCY

Occur in 10% of neonates ē liver disease. Presented in NN period ē fulminate liver disease. In children presented ē liver cirrhosis. In adults presented ē obstructive lung disease, emphysema, chronic bronchitis, asthma, & repeated lung infection.

Diagnosis: serum α-1 Antitrypsin concentration.

Management: no specific treatment.

TYROSINAEMIA

1/100.000 live birth. Body doesn't have enzyme Fumaryl Acetoacetate Hydrolase (FAH) to metabolize tyrosine, lead to accumulation of toxic metabolic products in va-rious body tissues. Death from hepatic failure frequently occur between 3-9 months of age, unless a liver transplantation performed.

Diagnosis

- Detection of enzyme level in blood.
- Presence of serum tyrosine, methionine, α fetoprotein.
- Urine succenylacetone.

HEREDITARY FRUCTOSEMIA

Deficiency of fructose 1 phosphate Aldolase.

Diagnosis: •Enzyme activities. •Liver biopsy.

GLYCOGEN STORAGE DISEASE

Genetic enzyme deficiencies. Accumulation of glycogen within the cells.

Diagnosis: •Liver biopsy. •LFTs.

RICKETS

Failure of mineralization of osteoid tissue of growing bones due to Vit. D deficiency. The daily requirement Vit. D is 400 unit & of Calcium is 400- 800-1200 mg for neonate, child, adult respectively. The diet contains Vit. D3 ẃ changed by liver into 25-hydroxycalciferol ẃ in turn changed into 1, 25 dihydroxycalciferol (Vit. D2) in kidney, ẃ is the active form Vit. D, causing absorption of Ca+ & Ph+. The Vit. D3 in skin change by effect of ultra violet rays (sun rise & set) to Vit. D2, ẃ represent 10% of Vit. D2 in body. ⇩ Ca+ ⇨ stimulation of PTH ⇨ mobilization of Ca+, Ph+ from bones.

Clinical picture

- Delay of; teething, walking & closure of fontanels.
- Bossing of head, flat bones of skull (box like appearance), craniotabes (firm pressure over occiput or posterior parietal bone gives feeling of ping bong ball sensation).

- Broadening of wrist, ankles.
- Green stick fracture long bone, bowing of legs.
- Skeletal deformities, rachitic rosary at costochondrial junction.

Differential diagnosis

- Vitamin D resistant rickets (Hypophosphatemic rickets, is X- linked dominant, occur in 1/20.000 live births, characterized by renal defect in reabsorption of phosphorous. Respond to high dose of vitamin D.
- Renal tubular acidosis type I, II. Characterized by; weakness, hypotonia, nausea, vomiting , failure to thrive, constipation, polyuria. Diagnosed by blood gases analysis.

Complications

- Anaemia
- Infection
- HF
- Chest deformities
- Contracted pelvis & difficult labor.

Investigations

- Normal serum calcium.
- ⇩ Serum phosphorus.
- ⇧ serum alkaline phosphatase (formed by osteoblasts, liberated into circulation when it is unable to form new bones).
- Normal parathyroid hormone.
- X ray bones of wrist, ankles shows active, healing, or healed rickets stage.

Management

- Ample amount of milk.
- Egg yolk to baby from the 5th month of life, very rich in Vit D.
- D Ca B_{12} syrup (contain 1000 U Vit D + 50mg Ca + 10 ug Vit. B_{12}/5 ml), dose 1tsp X 2/day for 2 months. Do X ray every month + Ca+ level to keep it < 10.5 mg/dl. OR
- D Ca B_{12} amp (contain 20.000 U Vit D + 50 mg Ca + 10 ug Vit B_{12}), IM weekly for 2 months ē the same precautions as monitoring X ray, serum Ca level every month. OR
- One Alpha drops 1 drop daily, ē monitoring as above.

CHAPTER X

UROLOGY

- Urinary Tract Infection
- Vesico Ureteric Reflux
- Nocturnal Enuresis
- Acute Nephritic Syndrome
- Post Streptococcal Glomerulonephritis
- Haematuria
- Nephrotic Syndrome
- Renal Failure Neonate

URINARY TRACT INFECTION

In children 90% of cases of UTI are caused by Ecoli, while 3% caused by streptococci. UTI is commoner in females. The child kidney is 30 gm weight & measure 2.5 X 1.5 X 0.5 cm, lies opposite 1st, 2nd lumbar vertebrae, moves up & down ē respiration.

Clinical picture

Wide variety of presentation include; • PUO. •Renal colic. •Jaundice. •Septicaemia. •Failure to thrive.

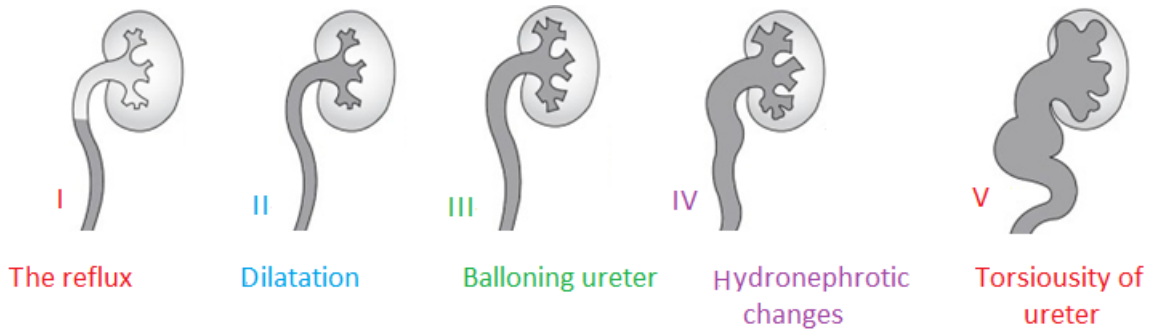
Investigations

- Urine analysis.
- Urine bacterial cell count: > 100.000 is diagnostic.
- Urine culture, sensitivity.
- Plain X ray UT.
- Renal U/S.
- IV pyelography ē micturating cystogram.
- Blood culture, sensitivity.
- Renal function tests: urea, creatinine, eGFR.

Management

- Septrin syrup 40 mg TMS + 200 mg SMZ, tab 80 mg + 400 mg, dose TMS 4-8 mg/Kg ÷ 2, then daily prophylactic for 6 months.
- Quinolone group antibiotics used for adults, tab 1 X 2 X 5 days.
- Coli urinal granules, 1 spoon + ½ cup water X 3 after meals.
- Rwatinex capsule 1X3 before meals.
- Excess water intake (water is the best diuretic).

VESICO URETERIC REFLUX



For every child < 5 years ē UTI do IVP ē micturating cystourethrogram.

Vesico ureteric reflux include 5 stages:-

I -The reflux.

II -Dilatation ureter.

III –Ballooning of ureter.

IV -Hydronephrotic changes.

V -Torsioucity of ureter.

Investigations: •Urine analysis. •IVP ē micturating cystourethrogram. •U/S of UT.

Management

•The hydronephrotic changes improves gradually, no formation of scars after age 2 yrs.

•Septrin syrup 40mg TMS + 200mg SMZ, dose TMS 2-4mg/ Kg as daily prophylactic, regular follow up by urine analysis & U/S. Septrin act better on alkaline urine, so give milk ē it or effervescent as Citro Cal or Epimag.

NOCTURNAL ENURESIS

90% of children become dry at night by the age of 1 ½ - 5 years, while 10% continue for more further years. 95% of cases are due to psychological problem (divorce, separation, over protection, death of one of parent, deprivation, fear, anxiety, shiness) or faulty training, MR, we must at first R/O organic cause as DM, renal anomaly as ectopic ureter in

vagina, UTI, diabetes insipidus or chronic renal failure.

Investigations

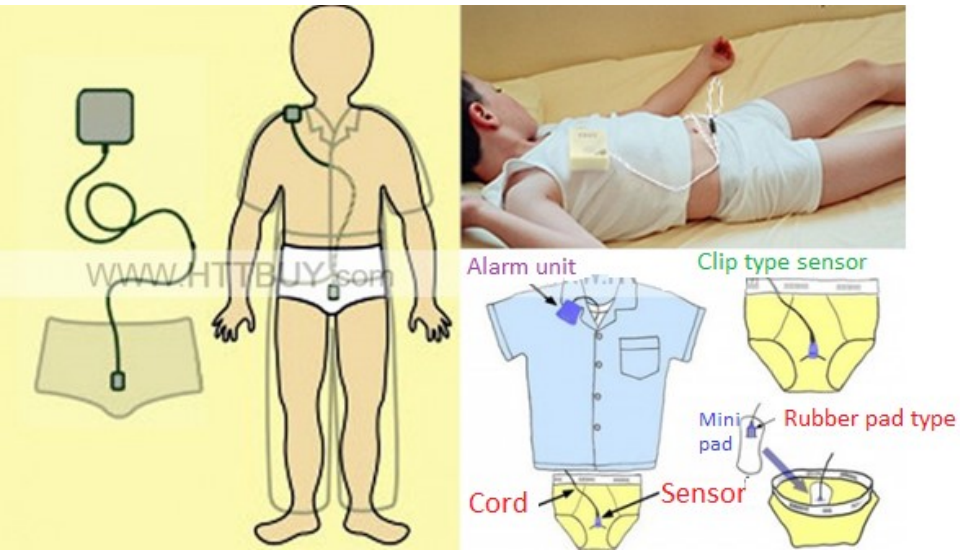
•Urine; for specific gravity, sugar, pus cells •Blood sugar •U/S of UT •Renal function tests; urea, creatinine.

Management

•Minirin 0.1 mg tab. one tab. daily before sleep for one wk, if child improve we give ½ tab daily for one month then stop medication. If child not improving, we double the dosage to 2tab daily for one wk & we follow the same previous regime.

•Electric buzzer.

•Reassurance, explanation & Psychological treatment.



ACUTE NEPHRITIC SYNDROME

Is the clinical correlate of acute glomerular inflammation. Characterized by sudden onset (i.e. over days to wks) of ARF & oliguria (<400 ml of urine/day) associated é hypertension, oedema & presence of active urinary sediments.

Etiology

Acute nephritic syndrome can result from renal-limited primary glomerulopathy or from

secondary glomerulopathy complicating systemic disease. In general, rapid diagnosis & prompt Rx are critical to avoid the development of irreversible renal failure. Immune complex glomerulonephritis may result from:-

1. Idiopathic.

2. Represent a response to a known antigenic stimulus (e.g. post infectious).

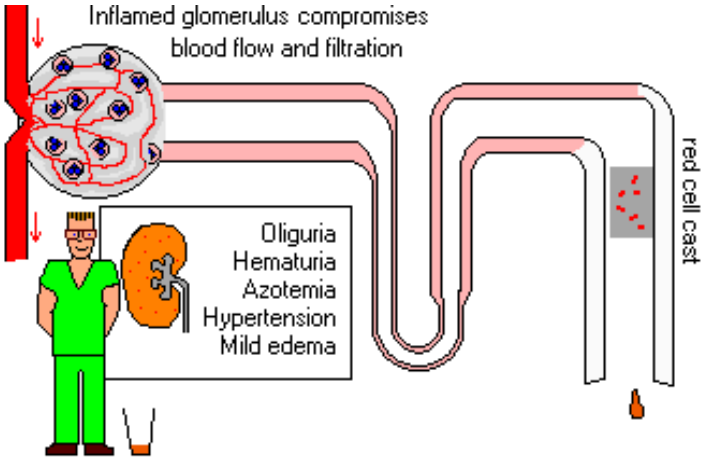
3. Part of multisystem immune complex disorder (e.g. lupus nephritis, Henoch-Schonlein purpura, cryoglobulinemia, bacterial endocarditis).

Clinical features

ECFV expansion, oedema & ⇧ BP develop because of impaired GFR & enhanced tubular reabsorption of salt & water. Pt may present é CHF & pulmonary oedema, also picture of ARF may also occur. Evidence of underlying cause can detected (e.g fever, skin lesions, or joint swelling).

Laboratory tests

- Urine analysis: as a result of injury to glomerular capillary wall, urine typically revea- ls RBCs casts, dysmorphic RBCs, leukocytes & subnephrotic proteinuria of <3.5 gm /24 hrs (nephritic urinary sediment). Hematuria is often macroscopic.
- Renal function test: ⇧ serum creatinine.
- Serology: immunologic assays suggesting the underlying disease.



POST-STREPTOCOCCAL GLOMERULONEPHRITIS

Etiology & Epidemiology

This is the prototypical post-infectious GN & leading cause of acute nephritic syndrome. Most cases are sporadic, though the disease can occur as an epidemic. GN develops, on average, 10 days after pharyngitis or 2 weeks after a skin infection (impetigo) é a nephrogenic strain of *group A β-hemolytic streptococcus*. Immunity to these strains is type-specific, long-lasting. Repeated infection & nephritis are rare. Epidemic post-streptococcal GN is most commonly encountered in children of 2-6 years of age é pharyngitis during the winter months. This entity appears to be ⇩ in frequency, possibly due to more widespread & prompt use of antibiotics. Post-streptococcal GN in association é cutaneous infections usually occurs in a setting of poor personal hygiene or strept super infection of another skin disease.

Clinical Picture

The classic presentation of post-streptococcal GN is full-blown nephritic syndrome é oliguric ARF; however, most pts have milder disease. Indeed, subclinical cases outnumber overt cases by 4-10 fold during epidemics. Pt é overt disease present é gross hematuria (red or "smoky"), headache & generalized symptoms as anorexia, nausea, vomiting & malaise. Swelling of renal capsule can cause flank or back pain.

Physical examination: ⁂ Hypervolemia. ⁂ Oedema. ⁂ Hypertension.

Laboratory findings

****Urinalysis:*** urinary sediment is nephritic é dysmorphic RBCs, RBCs casts, leukocytes, occasionally leukocyte casts & subnephrotic proteinuria & <5% of pts develop nephrotic range proteinuria.

****Renal function test:*** serum creatinine often mildly ⇧ at presentation.

*__Serology:__ most pts (>90%) have circulating antibodies against streptococcal exo-enzymes such as ASO, DNAase.

Course & prognosis

Post-streptococcal GN is typically an acute disease é spontaneous recovery occurring in almost all pts, even those who develop renal insufficiency during the acute episode. Resolution of the clinical manifestations of post-streptococcal GN is generally quite rapid, assuming concurrent resolution of infection. Diuresis typically begins within one wk & the serum creatinine returns to the previous baseline by 3-4 weeks. Hematuria usually resolves within 3-6 months. Proteinuria also falls during recovery, but at much slower rate. Generally, long-term prognosis is good.

Complications

•CHF & Pulm oedema. •ARF. •Sever hypertension é hypertensive encephalopathy.

Treatment

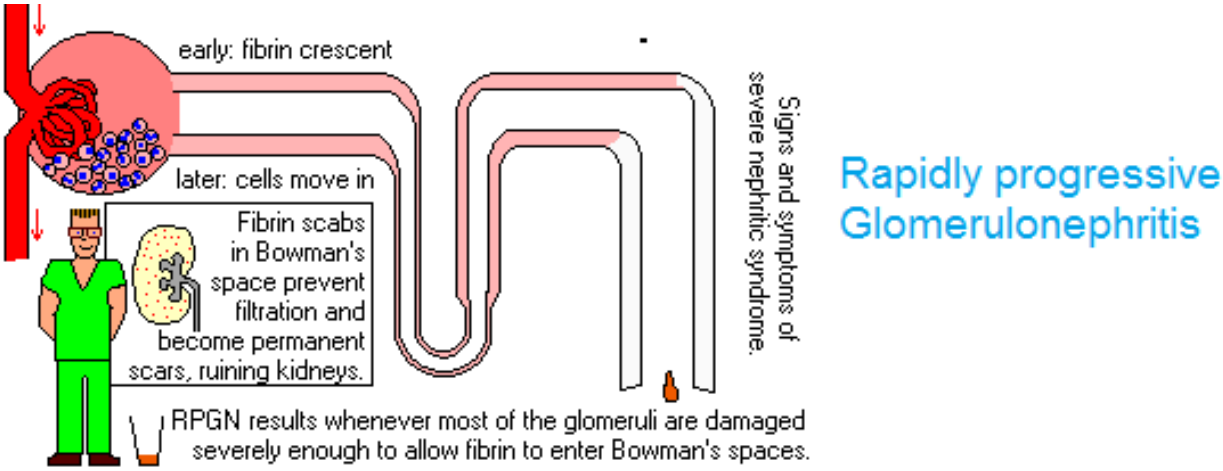
1. Eliminating the streptococcal infection: é antibiotics.

2. Supportive therapy until spontaneous resolution occurs including:-

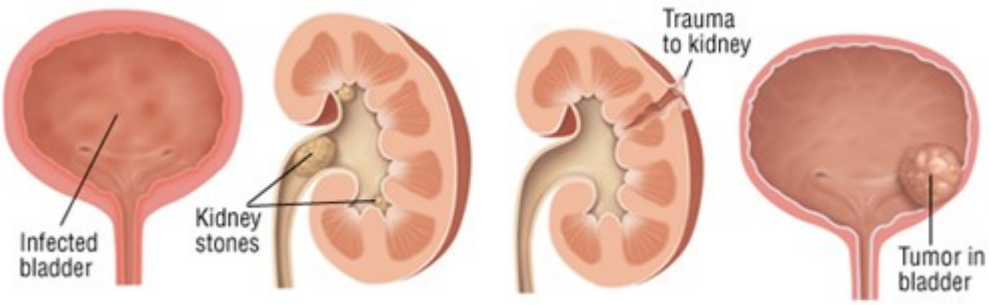
a) Bed rest. b) Salt restriction. c) Diuretics are employed to control ECFV.

d) Anti-hypertensive drugs to control high BP.

3. Dialysis: some pts may need it.



HEMATURIA



May be gross (macroscopic), or microscopic (presence of at least 5 RBCs per HPF of centrifuged urine &/or 5 RBCs per microliter in uncentrifuged specimen.

Common causes in children are:

1- IgA nephropathy.

2- Benign familial hematuria.

3- Alport syndrome (nerve deafness + hereditary nephritis).

4- Glomerulonephritis.

Investigations

- Plain X ray urinary tract.
- Coagulation profile (BT, CT, PT, and APTT).
- CBC & Blood film for sickling.
- Hb electrophoresis (sickle cell anemia).
- Complement C_3, C_4 (glomerulonephritis).
- Ant double stranded DNA (SLE).
- Urine analysis (Biliharziasis).
- Blood urea & serum creatinine (hemolytic uremic syndrome).
- IVP (polycystic kidney).
- U/S abdomen, renal biopsy (glomerulonephritis).

NEPHROTIC SYNDROME

Temporary disturbance in kidney function, more in boys & in age group 2-5 years. The prognosis is poor in case of age <1 year, or é severe degree of hypoproteinemia, or é marked ⇧ of serum creatinine & é prolongation of PT. The disease characterized by significant proteinuria of >3.5 gm/ 1.73 m^2/24 hours (>3-3.5 gm/ 24 hours).

Etiology

1. Multisystem diseases account for 50-70% of adult nephrotic syndrome as seen é; DM, collagen vascular diseases & amyloidosis.

2. Neoplasms: including; leukemia, lymphoma & solid tumors.

3. Infections: including; viral, bacterial, protozoal & helminthic.

4. Primary glomerulopathies (idiopathic): account for 30-50 % of adult nephrotic syndrome. In children 90% of cases are primary glomerulopathies.

Clinical picture

1. Proteinuria & hypoalbuminemia: in general, the greater the proteinuria, the lower the serum albumin level. Hypoalbuminemia is compounded further by ⇧ renal catabolism & inadequate hepatic synthesis of albumin. The proteinuria is believed to be due to ⇧ permeability of the glomerular basement membrane to proteins.

2. Oedema: common sites include; feet, face, periorbital areas & scrotum. Hypoalbuminemia & primary water & salt retention by kidney are the postulated mechanisms for oedema formation.

3. Hyperlipidemia: a consequence of ⇧ hepatic lipoprotein synthesis & ⇩ clearance. Hyperlipidemia may accelerate atherosclerosis & progression of renal disease.

4. Hypercoagulability: is multifactorial: some of the mechanisms are loss of anti-thrombin III in the urine, ⇧ fibrinogen production by the liver & ⇧ platelet aggregation. Sponta-

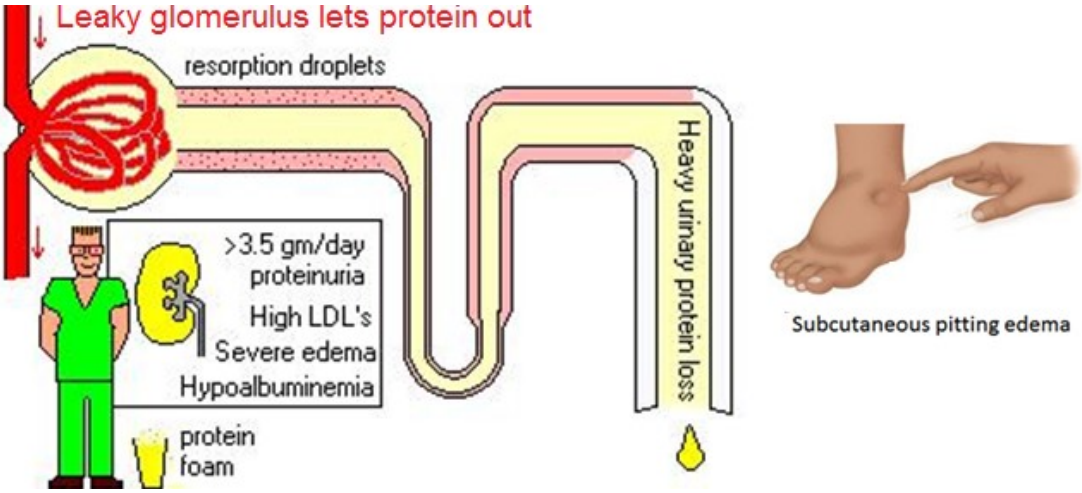
neous peripheral arterial or venous thrombosis, renal vein thrombosis & pulm. embolism may occur. The clinical features that suggest acute renal vein thrombosis include:-

•Sudden onset of flank or abdominal pain, gross hematuria.

•Left sided varicocele (as the left testicular vein drains into the renal vein).

•⇧ proteinuria & an acute decline in GFR. The chronic renal vein thrombosis is usually asymptomatic.

5. Other complications: •Protein malnutrition. •Iron-resistant microcytic hypochromic anemia due to transferrin loss. •Hypocalcaemia as a consequence of Vit D deficiency due to enhanced urinary excretion of cholecalciferol-binding protein.•⇧ Susceptibility to infection from urinary loss & ⇧ catabolism of immunoglobulin.



Investigations

⅄Confirming significant proteinuria: proteinuria 40mg/Kg/day, or >2gm/day or measuring urinary protein by dipstick (+3 diagnostic). ⅄Serum proteins: < 2½ gm/dL ⅄⇩ Serum sodium. ⅄⇧ Serum cholesterol. ⅄Urine analysis: microscopic hematuria ⅄⇧ Blood urea & creatinine. ⅄Prolonged PT. ⅄Immunoglobulins electrophoresis (myeloma screen), complement (C3, C4) & autoantibodies (ANA, ANCA, anti-dsDNA & anti-GBM). ⅄Renal U/S. ⅄Renal biopsy to identify the underlying histopathologic anomaly: include minimal

change (80% of cases in children <10 yrs). Membranous glomerulopathy (60-70% of cases in adults). Focal segmental glomerulosclerosis.

Treatment

Fluid balance, hypovolemia & BP. A 'no added salt' diet is appropriate measure. If hypovolemia is present it should be promptly corrected é administration of 10-20 ml/kg of 4.5% albumin. Diuretics used in some cases to help control oedema until remission begins, e.g. Furosemide at 2 mg/kg/24 hour. The use of diuretics should be reviewed on a daily basis & the pt's electrolytes should be checked regularly. 20% albumin in combination é diuretics is used in centers to relieve severe symptomatic edema: 0.5-1.0 gm/kg of 20% Albumin can be given slowly over 4-6 hours & 0.5-1 mg/kg of Furosemide given at the end or midway through the infusion. Rapid administration should be avoided to prevent intravascular volume overload. 20% Albumin should never be used to correct low serum albumin.

Measures to control proteinuria/hypertension: ACEIs: ⇩ proteinuria by ⇩ GF presssure. Controlling hypertension by keeping BP < 130/80.

Measures to control infection: streptococcal pneumonia & G-ve organisms are the commonest pathogens causing possible peritonitis, septicemia, or cellulitis. Prophylactic oral Phenoxy Methyl Penicillin (12.5 mg/kg twice daily) is recommended while the pt is edematous & any suspected infection to be treated é broad spectrum antibiotics while awaiting culture.

Measures to guard against thromboembolism: anticoagulant indicated for pt é deep venous or arterial thrombosis or pulmonary embolism. Heparin may not be effective because of urinary loss of ant thrombin III.

Measures to control hyperlipidaemia: may need lipid lowering agents. Diet.

Mobilization: Bed rest may ⇧the risk of venous thrombosis, so pt is encouraged to mobileze as normal.

Diet: as mentioned above 'no added salt', diet is advisable in view of the salt & water overload. No evidence for use of a high protein diet. Encouraged to have a normal healthy diet.

No response: add Prednisolone orally 1 mg/Kg/day ÷ 3 (tab 5 mg) for 4 weeks or until urine return to normal, then do weaning gradually over 2 weeks + Aldactone 25mg tab., 1 mg/Kg/day + Lasix 40 mg tab (1 mg/Kg/day). 90% respond to these regimes, while 20% need renal biopsy & may need course of Cyclophosphamide.

RENAL FAILURE NEONATE

Common in SCU as a result of hypoxia, hypotension, ischemia, dehydration, sepsis, drug induced as penicillin, Sulpha, Lasix, Quinolone, NSAIDs, & Gentamicin.

Types

Pre renal failure: due to severe illness, dehydration, Hemorrhage, age, shock. The prerenal failure will respond to forced alkaline diuresis in the form of 20 ml of normal Saline/Kg over 2 hours then Lasix 1 mg/Kg as IV bolus, diuresis takes place within 30 min, 24 hours urine for sodium is <10, urine/plasma osmolality is >2.

Renal failure: drug intoxication resulting in interstitial nephritis, ATN, in such conditions we must stop all medication & start prednisolone 1 mg/Kg/day for one week, may be due to chronic glomerulonephritis or cong. anomalies. Renal failure will not respond to forced alkaline diuresis, 24 hours urine Na is >40, urine/plasma osmolality is <1.1

Post renal failure: as a result of stones, tumor & obstruction to the flow of urine.

Investigations

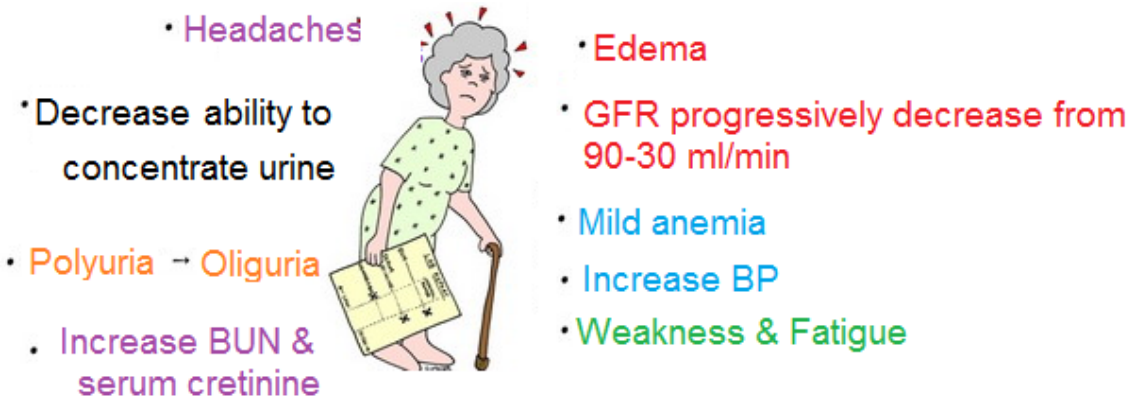
- Sudden ⇧ of BUN.

- Serum creatinine >5, but in NN is not good indicator as it reflect maternal level as it freely cross placenta, unless it coming up.
- eGFR < 50%.
- Potassium > 6.0 meq/L
- Anemia due to ⇩ of erythropoietin.
- Thrombocytopenia. • ⇧ of Ph^{+} & ⇩ of Ca^{+}.
- U/S: infant kidney 25 gm, diameters 2.5 X 1.5 X 0.5 cm. Adult kidney 300 gm, 10 X 5 X 2.5 cm.
- X Ray Bones: changes of bones due to renal osteodystrophy, inability of kidney to form 1, 25 dihydroxycholicalceferol.
- Oliguria in infants if urine <1 ml/Kg/hr, or sudden hematuria, followed by anuria (urine < 200 ml/day)

Management

- Fluid chart: fluid intake = urine output + 500 ml insensible water loss.
- No salt, salt free diet.
- Proteins: 20 gm/day.
- Douputamine: used in case of perfusion permanently poor as in case of, shock, hypotesion, renal failure. Dose calculated according to the following formula 'Pt BW X 3' = number of mg of Douputamine to be collected from the vial 250 mg, by insulin syringe, add 50 ml of glucose 5%, give 2 ml/hr ẃ is equal to 2 ug/Kg/min, can be doubled to 5 ug/Kg/minute, this dose will ⇧ the blood supply to internal organs. A higher dose "6-10 ug/Kg/minute", cause the same previous effect + positive inotropic, chronotropic effect on heart. Much higher dose "11-15 ug/Kg/min cause the same previous effect + peripheral vasoconstriction.

- Ca gluconate 10% strictly IV over 30 minutes, amp. 10 ml, dose 1 ml/Kg=100 mg/Kg, after dilution ē G 5% in ratio 1:4 ē monitoring of heart rate.
- Metabolic acidosis: if, PH is < 7.2, corrected by $NaHco_3$ dose calculated according to the formula "base deficit X B W ÷ 2" =meq $NaHco_3$ to be given, amp of $NaHco_3$ 20 ml contain 17.5meq , or approximately 1-2 meq/Kg, ½ IV & ½ in the running drip.
- Hyperkalemia: corrected by G 50% 1 ml/Kg +insulin 1 U/5 gm glucose.
- Prednisolone: 1 mg/Kg/day for one week.
- Anaemia: corrected by blood transfusion.
- Thrombocytopenia: corrected by platelet transfusion 1 U/Kg.
- No response: renal dialysis & avoid use of heparin during dialysis to avoid heparin induced thrombocytopenia.
- Stop all the antibiotics unless it is absolutely indicated, using the minimum dose as all are metabolized by kidney & liver.
- Zantac/Contrloc used very carefully as they affect kidney & liver, amp 50mg IV/12 hour.



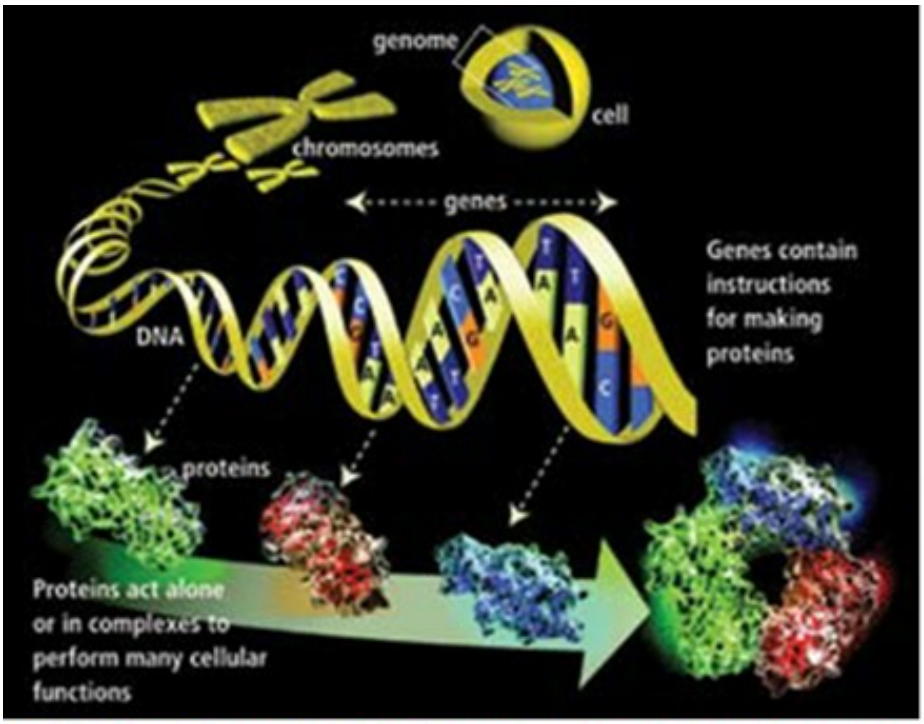
CHAPTER XI

GENETICS

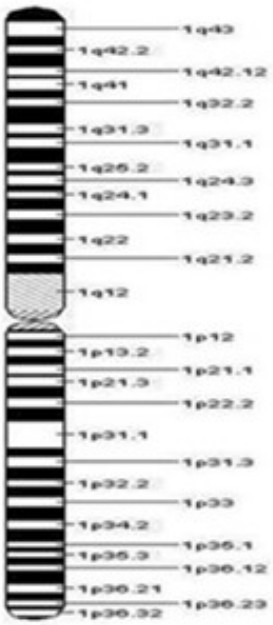
- Introduction
- Genes & Human disease
- Chromosomal Anomaly Disorders
- Autosomal Dominant Disorders
- Autosomal Recessive Disorders
- X - Linked Dominant Disorders
- X - Linked Recessive Disorders

Introduction

Globally, at least 7.6 million children are born annually é severe gen-etic or cong. malformation; 90% of them are born in mid & low income countries. Precise prevale- nce data are difficult to collect, especially in developing countries owing to great diversity of condition & also because many cases remain undiagnosed. The genetic & congenital disorders are the 2nd most common cause of infant & childhood mortal-ity & occurs é a prevalence of 25-60 per 1000 births. The higher prevalence of gene- tic diseases in a particular community may, however, be due to some social or cult- ural factors, such as consanguineous marriages, ŵ results in higher rate of autosom- al recessive conditions including cong. malformations, stillbirths, or mental retardati-ation. Furthermore, maternal age >35 yrs is associated é higher frequencies of chro- mosomal abnormalities in the offspring, also environmental pollution, wars, chemical weapons & radiations are very important as it was seen in gulf area after gulf war & the marked rise of cong. malformations especially of bones.



Each strand of D.N.A. is really a long string of genes



Chromosome 1

Chromosome

Human body is made-up of trillions of cells, each specializing in particular function & each working together in a complex symphony of interactions é the exception of RBCs, ẃ contain no nucleus & no nuclear deoxyribonucleic acid. Chromosomes are subcellular structures that exist in the nucleus of each cell that makes up the human body & responsible for transferring genetic information from one generation to another. There are 23 pairs of chromosomes existing in the human cell -22 Autosome + Sex Chromosome known as XY (male) & XX (female). Chromosomes are very long thin strands of DNA coiled up like a ball of string, for example: Chromosome 1 contain > 3000 genes & 240 million base pairs, of ẃ 90% have been determined. Chromosome 2 contains 〉2500 genes & 200 million base pairs, of ẃ 5% have been determined. X chromosome contains 〉 1400 genes & 150 million base pairs, of ẃ 95% have been determined. Y chromosome contains 〉 200 genes & 50 million base pairs of ẃ 50 % have been determined.

Genes

Are instruction manuals for our bodies. They are the directions for building all the proteins that make our body function. Scientists now think there are 30.000 different human genes in each cell. Over 90% of the genetic code is identical in all of us; there are small variations in DNA between people. It is these small variations that make us all different from each other. Currently estimate that 10.000 of human diseases are known to be monogenic.

CONGENITAL ANOMALIES

Incidence: 1/100 born baby.

Chromosomal anomalies: 50%. Unknown: 30%. AD: 15%. AR: 2.5%. Drug induced: 2.5%.

2-3% of populations are mentally retarded. The cause identified in 50-60 % of cases.

The commonest causes of congenital anomalies (genetic base) are; Down syndrome 17%. Fragile X syndrome 3%. Inborn errors of metabolism: 2%

CHROMOSOMAL ANOMALIES

Any change in the normal structure or number of chromosomes often results in phy-sical or mental abnormalities.

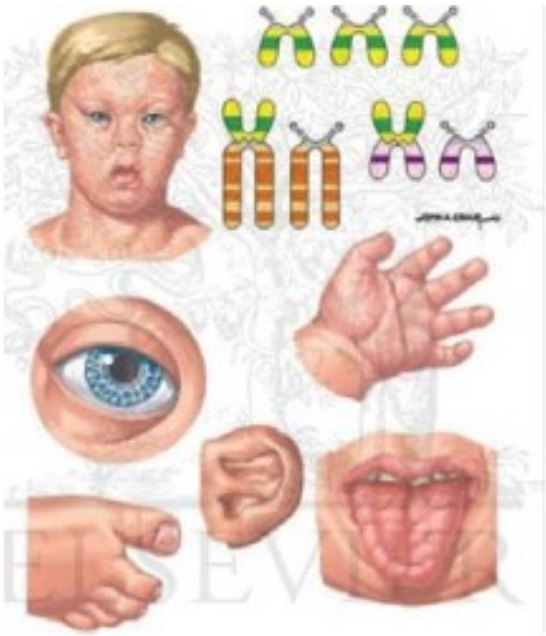
DOWN SYNDROME

Extra copy of chromosome 21, 47 XX + 21, Location 21 q 22.3

Named after British physician, John Langdon Down, in 1866.

Abnormality	AFP	hCG	µE3
Neural Tube Defects	↑	—	—
Trisomy 21	↓	↑	↓
Trisomy 18	↓	↓	↓

AFP (Alpha Feto Protein.
hCG(human Chorionic Gonadotrophine)
uE3 (unconjugated Estriol).



Incidence

1/700 Life Births, incidence ⇧ ē age. Presence of extra copy of chromosome 21(47

XX + 21). The commonest chromosomal anomaly, include three types,

Non-disjunction (95%),

Translocation (4%) &

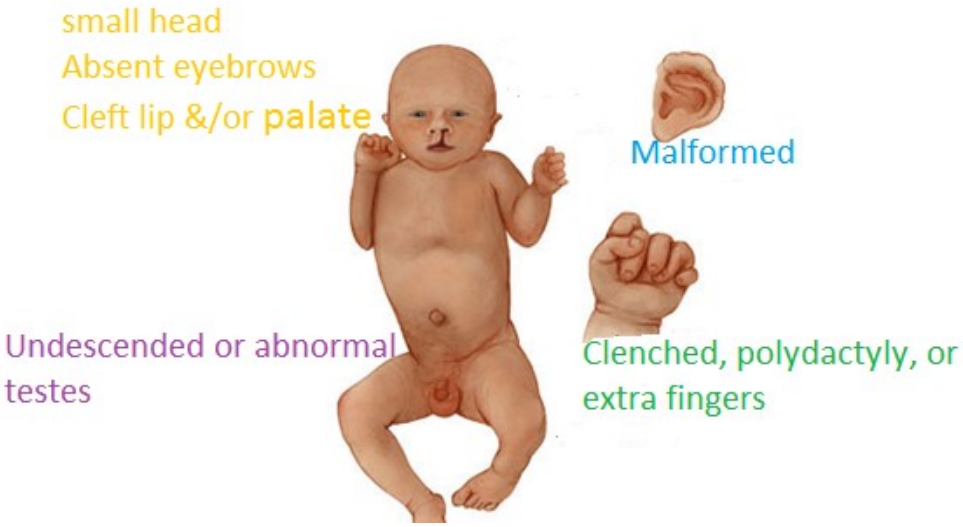
Mosaicism (1%)

•MR •Delayed milestones. •Upper slanting of eyes. •Epicanthus fold. •Brush field spots in iris. •Depressed nasal bridge. •Big tongue. •Flat occiput. •Single palmar & plantar creases. • Rudimentary 5th Finger. •Gape between 1st & 2nd toes. •CHD (50% VSD). •Hypotthyroidis ism (30% of cases). •Leukemia (10 times more common). •Intestinal obstruction. •Hirschsprung disease. •Usually associated ē infertility.

TRISOMY 13 (Patau Syndrome)

Three copies of genetic material for Chromosome 13

First described by German born American geneticist, Klaus Patau in 1960



Incidence: 1/5000 Births.

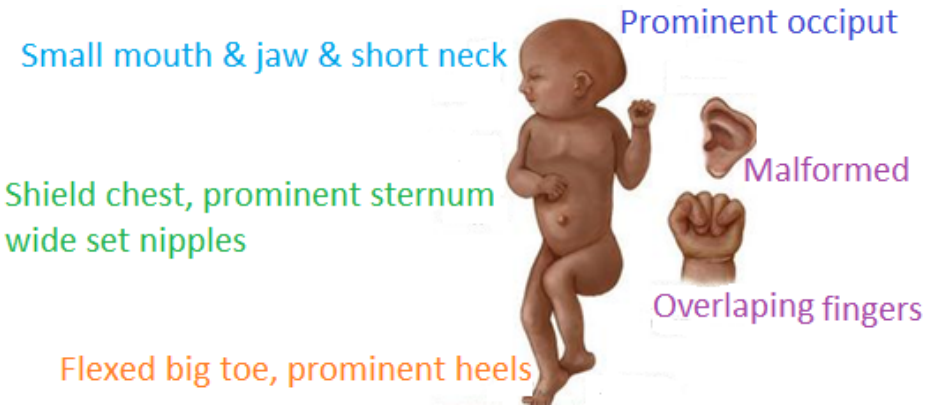
•MR. •Microcephaly. •Microophthalmia & Visual impairment. •Clenched hands ē extra-finger &/or toes. •Cleft lip ē or ēout cleft palate. •Hypotonia. •Brain anomalies. •Heart defect (VSD/ASD). •Renal anomalies. •NTD. •Undescended testes. •Deafness.

Survival beyond the first year is uncommon.

TRISOMY 18

Extra copy of Chromosome 18, 47 XY +18, Location 18 p11.1-q12.1

Described by British medical geneticist R. Edward, in 1960 (Edward syndrome)



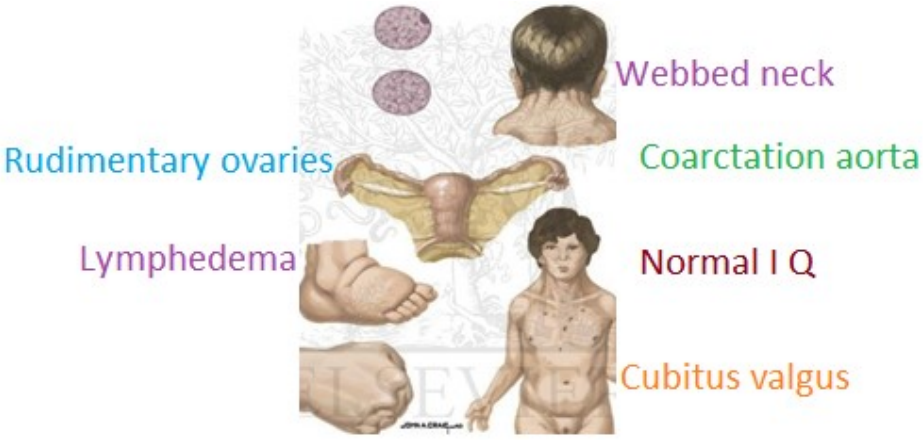
Incidence: the most common chromosomal abnormality after Down syndrome.

•Abnormal- shaped skull & short neck. •Small jaw & mouth. •Severe intellectual & physical defects. •Delayed milestones. •Dysplastic malformed low set ears. •Shield chest, prominent sternum ē wide set nipples. • Clenched fists ē overlapping of fingers. • Dysplastic malformed low set ears, shield chest, prominent sternum ē wide set nipple • Clenched fists, overlapping of fingers. •CHD.

TURNER SYNDROME

One of X Chromosome is missing , Gene SHOX

Named after Henery Turner, Endocrinologist, USA 1938

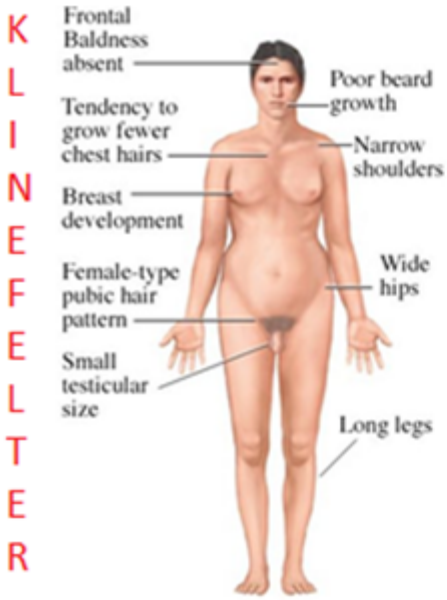


Incidence: 1/2500 Live female birth. The most common female sex chromosome abnormality (XO).

•Normal female external genitalia, uterus & fallopian tubes. Rudimentary ovaries & primary amenorrhea (infertility). •Short stature & webbed neck. •Cubitus valgus. •Shield shaped thorax & widely spaced nipples. •Low posterior hairline & brown spots (Nevi) of the skin •Lymphedema of hands & feet. •Low seat ears. •Small finger nails. •Coarctation of aorta. •Normal IQ.

KLINEFELTER SYNDROME

47XXY karyo type (mosaic 47 XXY /46 XY). Described by American Endocrinologist, Harry Fitch Klenefelter, in 1942.



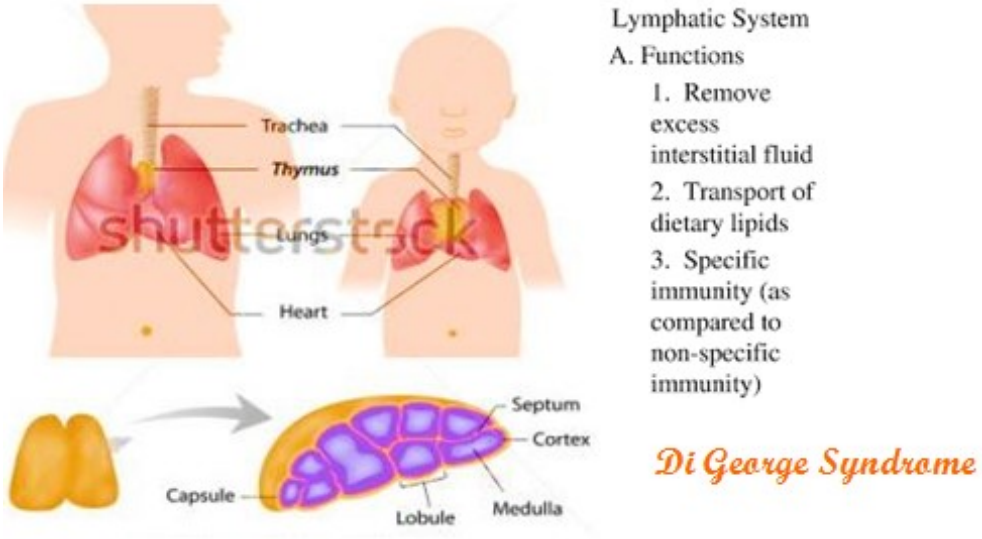
- Lower IQ than sibs
- Tall stature
- Poor muscle tone
- Reduced 2ry sexual characteristics
- Gynaecomastia (male breast)
- Small testes / infertility

Incidence 1/500 Births.

•Become more evident during puberty. •Tall Stature. •Long Arms & Legs. •Gynecomastia. •Female type pubic hair pattern. •Wide hips. •Small firm testes, azoospermia & infertility. •Elevated level of gonadotrophins.

DI GEORGE SYNDROME

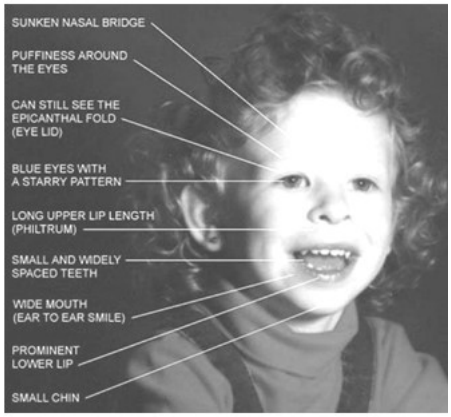
Deletion from chromosome 22, location 22 q 11.2. Described in 1968 by Italian American pediatric endocrinologist, A. DiGeorge.



Incidence: 1/4000 Births.

•The lost gene is normally required for development of thymus, immune system & T cells •Poor immune system function. •Recurrent infection. •CHD (Truncus Arteriosus). •Cleft palate. •Hypoparathyroidism. •Hypocalcaemia.

WILLIAMS SYNDROME



Deletion of about 26 genes from chromosome 7, Location 7 q 11.23- Identified by Newz-eland Cardiologist, John Williams, in 1961. Incidence 1/8000 Births.

•Unusually cheerful demeanor & ease ē strangers. •Elfin like facial features, wide mouth ,full lips. •Delayed milestones. •MR. • Widely spaced teeth, long phylum, small chin. •Flattened nasal bridge & small upturned nose. •Hypercalcaemia & Supravalvular aortic stenosis. •Hyperacusis (sensitive hearing). •Hypothyroidism. •Early puberty.

ANGELMAN SYNDROME

Deletion of Chromosome 15, Location 15 q 11.2- q 13

Named after British, Pediatrician, Harry Angelman, in 1965

Incidence: 1/10.000 - 20.000 Births

•Primarily affects the nervous system. •Hyperactivity & short attention span. •Happy, excitable, demeanor ē frequent smiling, laughter. •Sleep disturbances. •Hand flapping movement. •Seizures. •EEG gives characteristic pattern (large amplitude slow spike).

PIERRE ROBIN SYNDROME

Chromosome 17, Location 17 q 24.3-q 25.1

Named after the French Dental surgeon Pierre Robin, in 1860

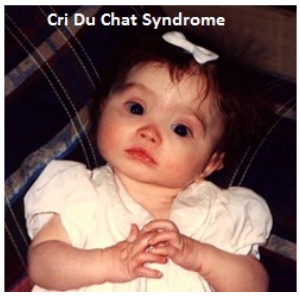
Incidence: 1/10.000 Births.

•Microcephaly. •Microophthalmia. •Microganthia. •Glossoptosis, backward displacement of tongue ẃ may cause closure of throat & obstruction to the air passage during respiration. •Most of cases suffer from cleft palate, but none of them has cleft lip.

CRI DU CHAT SYNDROME

Deletion of short arm of chromosome 5, Location 5 p 15.2

Described by French, Geneticist, Jerome Lejeune, 1963



Incidence: 1/50.000 Births.

•MR. •Delayed milestones. •Hypotonia. •Microcephaly & round face. •Microganthia. •Microophthalmia. •Hypertelorism. •Epicanthus fold. •Low set ears. •High-pitched cat like cry is characteristic. •Renal or cardiac anomalies.

PRADER WILLI SYNDROME

Deletion of long arm of chromosome 15, Location 15 q 11.2

Described by, Swiss Endocrinologists, Andrea Prader, and Heinrich Willi, in 1956

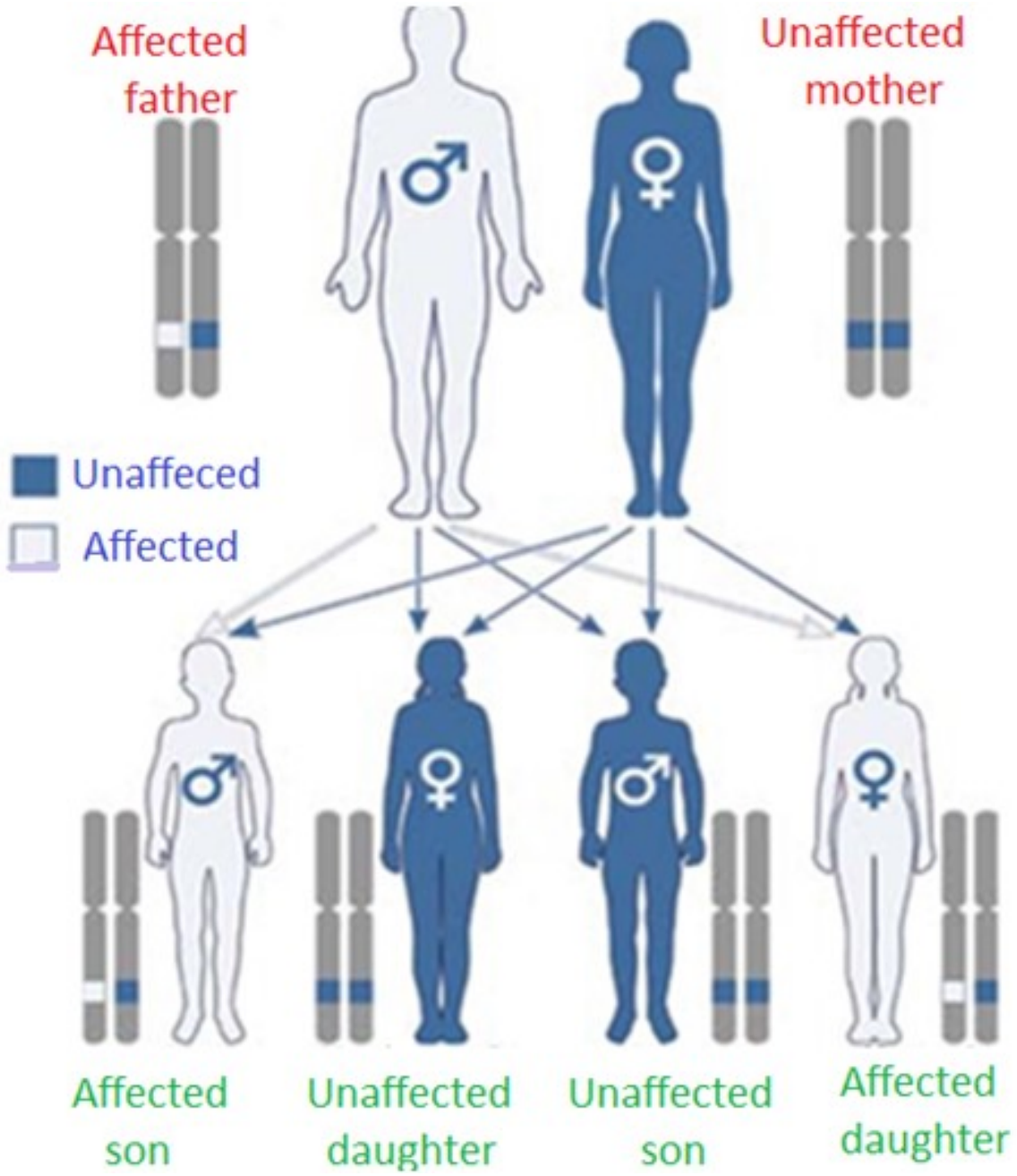
Incidence: 1/10.000 Births.

•MR • Almond shaped eyes •Short stature • Small hands & feet •Unstoppable appetite •Morbid obesity •High tendency for type II DM •Hypotonia •Hypogonadism & Infertility.

AUTOSOMAL DOMINANT DISORDERS

An affected person usually has at least one affected parent. Disorder affects either sex & can be transmitted by either sex. Affected person has 50 % chance of passing the defect on to their children.

Each child has 50% chance to have the disease.

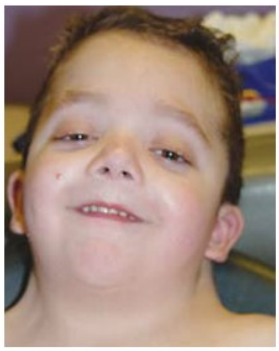


NOONAN SYNDROME

Chromosome 12, Gene PTPN11, Location 12 q 24

Named after, Jacquline Noonan, Pediatric Cardiologist, USA, in 1968.

It`s believed to be the male version type of Turner syndrome.



Incidence: 1/1000 Births.

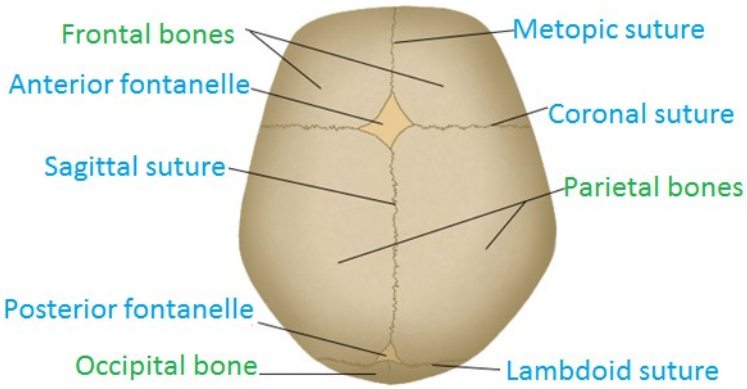
⊙Delayed milestones. ⊙Short stature. ⊙Hypotonia. ⊙Webbed neck. ⊙Microganthia. ⊙Low set ears. ⊙Cubitus valgus. ⊙Strabismus. ⊙High hairline at front of head. ⊙Cryptorchidism. ⊙Congenital heart disease (V.S.D. or A.S.D.)

CRANIOSYNOSTOSIS

Chromosome 7, Location 7p 21

Cranio = cranium, syn = together, ostosis = related to bones.

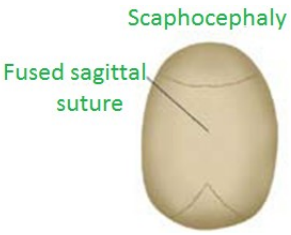
Normal skull of the Newborn



Craniosynostosis is premature closure of ≥ 1 cranial sutures, too soon in utero, disturbing the normal growth pattern of skull & facial bones.

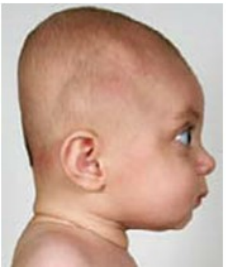
Incidence: 1/2000 Births. It includes the following:-

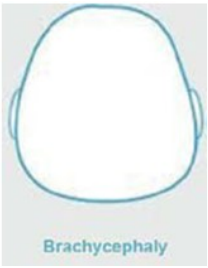
SCAPHOCEPHALY



Fusion of sagittal suture lead to elongated head & prominent forehead.

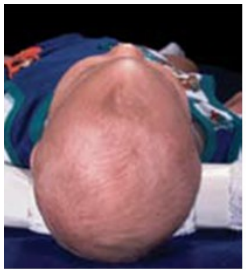
BRACHYCEPHALY

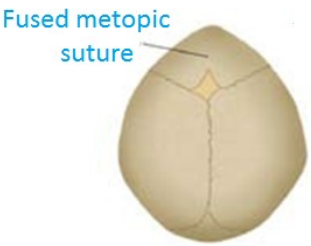




Fusion of coronal suture leads to flattening of the head.

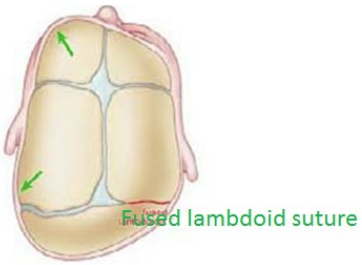
TRIGONOCEPHALY





Fusion of metopic suture lead to triangular shaped head & eyes appeared very close.

PLAGIOCEPHALY



Fusion of lambdoid suture lead to asymmetry of the head & ears are not aligned.

CHARCOT MARIE TOOTH DISEASE

Genes TRPV 4 .MPZ or CMTA 1, Location 1 q 23.3

Named after, Jean Martin Charcot, Pierre Marie, Hiward Tooth, England 1886

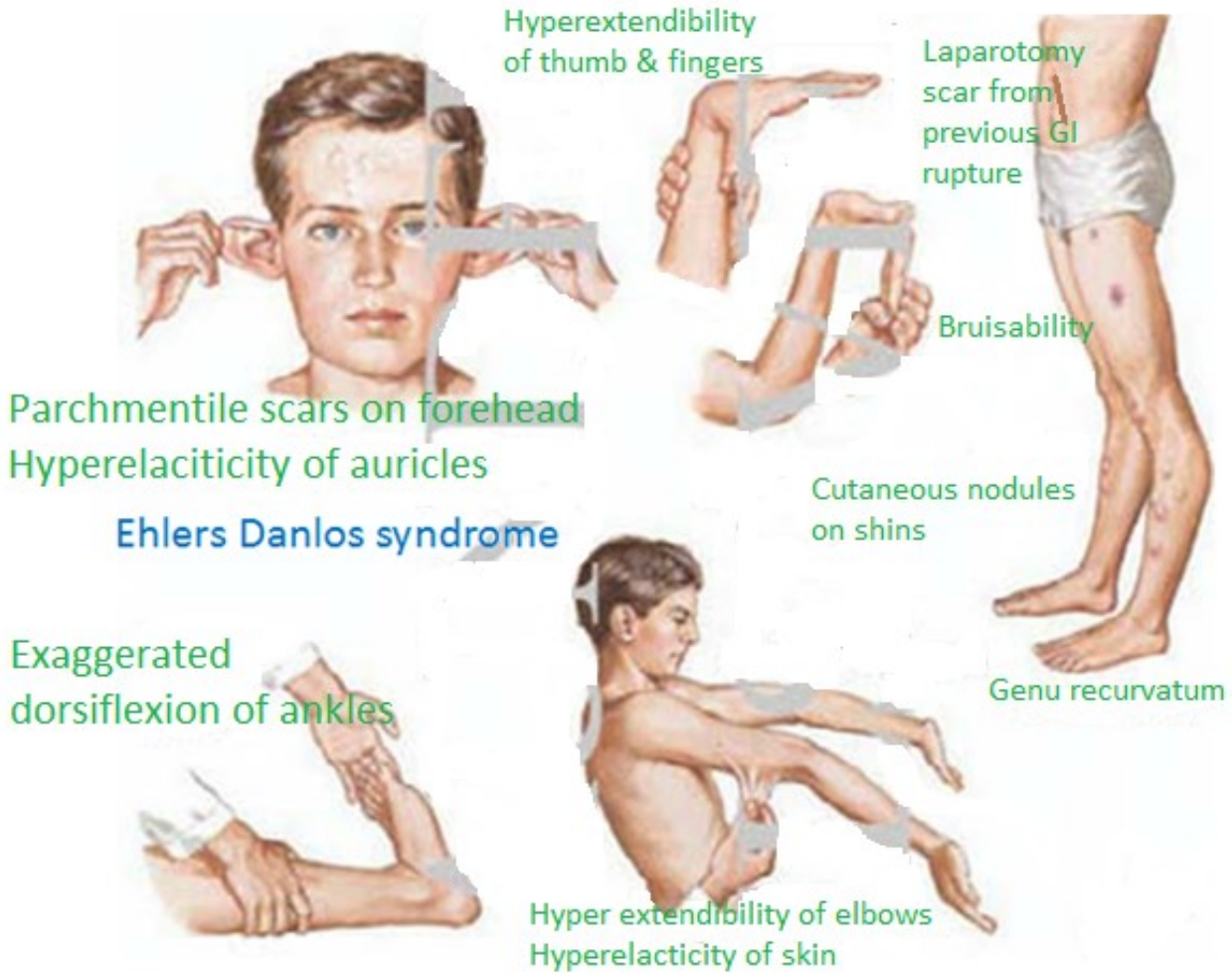
Incidence: 1/2500 people.

⊙Hereditary motor & sensory neuropathy. ⊙Begin between childhood & young adulthood. ⊙Muscle weakness, atrophy, loss of sensation & ⇩ reflexes. ⊙Affecting first the legs, progressively spread to affect different parts of body, including diaphragm & vocal cord. ⊙Foot drop, abnormal gait, skeletal deformities. ⊙Episodic painful muscular contractions.

EHLERS DANLOS SYNDROME

Chromosome 2, Gene COL 5 A 2, Location 2 q 32.2

Named after, Edvard Ehler, Dermatologist (Denmark) & Henry Danlos, physician (France), in 1908.



Incidence: 1 / 5000 Births.

⦿Connective tissue disorder, elasticity of skin.

⦿Hyperflexability of joints, hypotonia, skeletal deformities.

⦿Cong. heart defect (aortic aneurysm).

⦿Blue sclera, retinal detachment.

⦿High arched palate.

MARFAN SYNDROME

Chromosome 15 Gene FBN 1, Location 15 Q 21.1

Described by, Dr.Bernard Marfan, French Pediatrician, 1896

Incidence: 1/5000 Births. Connective tissue disorder.

- Tall & slender, arm span exceeds body height.
- Skeletal deformities; either scoliosis or kyphosis.
- Sunken chest, pectus excavatum, or pectus carinatum.
- Arachnodactyly. Dislocated lens in one or both eyes.
- Congenital defect in heart (Aortic dilatation).

BECKWITH-WIEDEMANN SYNDROME

Chromosome 11p15 Described by Drs. Beckwith & Wiedemann in 1969.

Incidence: 1/14.000 births.

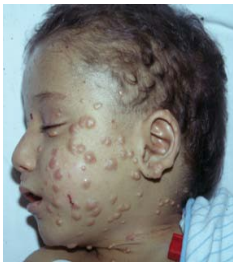
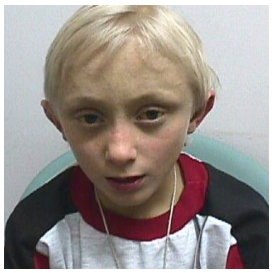
- Pediatric overgrowth syndrome.
- LGA.
- Microcephaly.
- Macroglossia.
- Enlargem-

ent of some organs & tissues. ✻Abdominal wall defect (omphalocele, umbilical hernia). ✻Creases in ear lobes & may be low set ears. ✻Cryptorchidism. ✻Poor feeding. ✻Hypoglycemia. ✻Seizures. ✻⇧risk of childhood cancer.

ALAGILLE SYNDROME

Chromosome 20, Gene JAG 1, Location 20 p 12

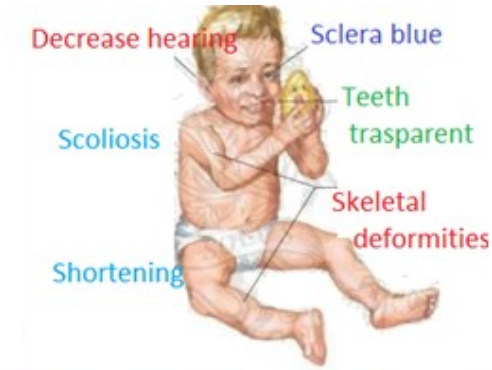
Described by Dr. Alagille et al, France in 1975

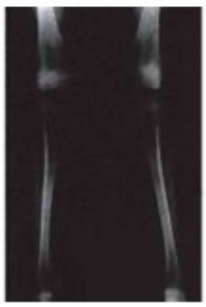



Incidence: 1 / 10.000 Births.

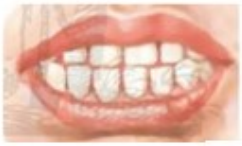
•Abnormalities in the bile ducts of liver. •Cholestasis, jaundice. •Deep-set eyes & prominent forehead. •Deposition of cholesterol in the skin (xanthomas). •CHD (pulmonary stenosis). •Renal anomalies.

OSTEOGENESIS IMPERFECTA





Thin & osteoporotic bones, fractures, deformities

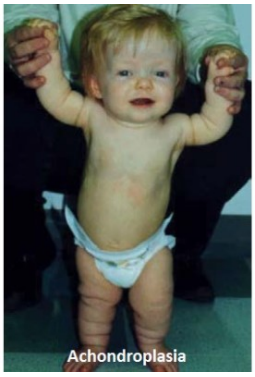


Teeth normal or soft & transparent

Incidence: 1/20.000 Births ▲ Extremely fragile bones ▲Skeletal deformities ▲Pathological fracture of bones ▲Deafness & Blue Sclera.

ACHONDROPLASIA

Chromosome 4, Gene FGFR 3, Location 4 p 16.3



Incidence: 1/25000 Births.

◊ The problem in converting cartilage to bones, particularly in long bone.

◊Short -limbed dwarfism & Hypotonia..

◊Average adult height about 4 feet (120 cm).

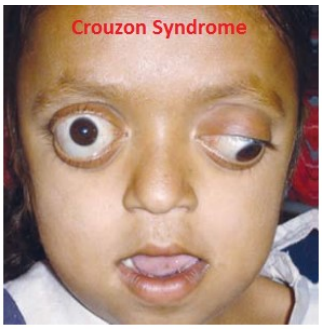
◊ Normal sized torso, large head ē prominent forehead.

◊ Redundant skin folds in arms & legs.

CROUZON SYNDROME

Chromosome 10, Gene FGFR 2, Location 10 q 26.13

Named after, Dr. Octave Crouzon a French Physician in 1912.



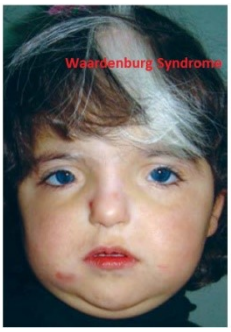
Incidence: 1/25000 Births.

◈Premature ossification of ≥ 1 cranial sutures, most often coronal & sagittal. ◈Exophthalmos. ◈Hypertelorism. ◈Strabismus (eyes do not point to the same direction). ◈MR. ◈Dental deformities.

WAARDENBURG SYNDROME

Chromosome 2, Gene PAX 3, Location 2 q 36.1

Named after Dutch ophthalmologist Dr. Petrus Johannes Waardenburg, 1951



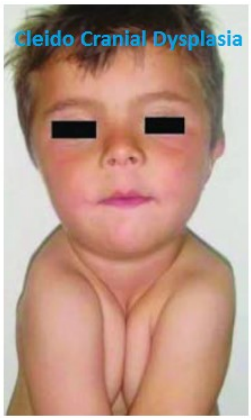
Incidence: 1 / 42000 people.

- Hypopigmentation of skin and hair or distinctive hair colouring (such as patch of white hair or hair prematurely turns grey).
- Medial eyebrow flare (synophrys).
- Two differently coloured eyes or brilliant blue iris.
- Hearing loss with varying degrees affect one or both ears.

CLEIDO CRANIAL DYSPLASIA

Chromosome 6, Gene RUNX 2, Location 6 p 21.1

In Greek; Cleido, Collarbone. Cranial, Head, Dysplasia, Abnormal form.



Incidence: 1 / 200.000 Births.

- Affects the development of bones &teeth.

- Shorter than other members of theirfamily.
- Incomplete or absent clavicles.
- Deformities of vertebral column.
- Osteopenia, may develop osteoporosis.
- Under mineralization of skull bones.
- Delayed closure of fontanels may remain open into adult age.
- Boozing of head, high arched palate & defective tooth.

APERT SYNDROME

Chromosome 10, Gene FGFR 2, Location 10 q 26.13

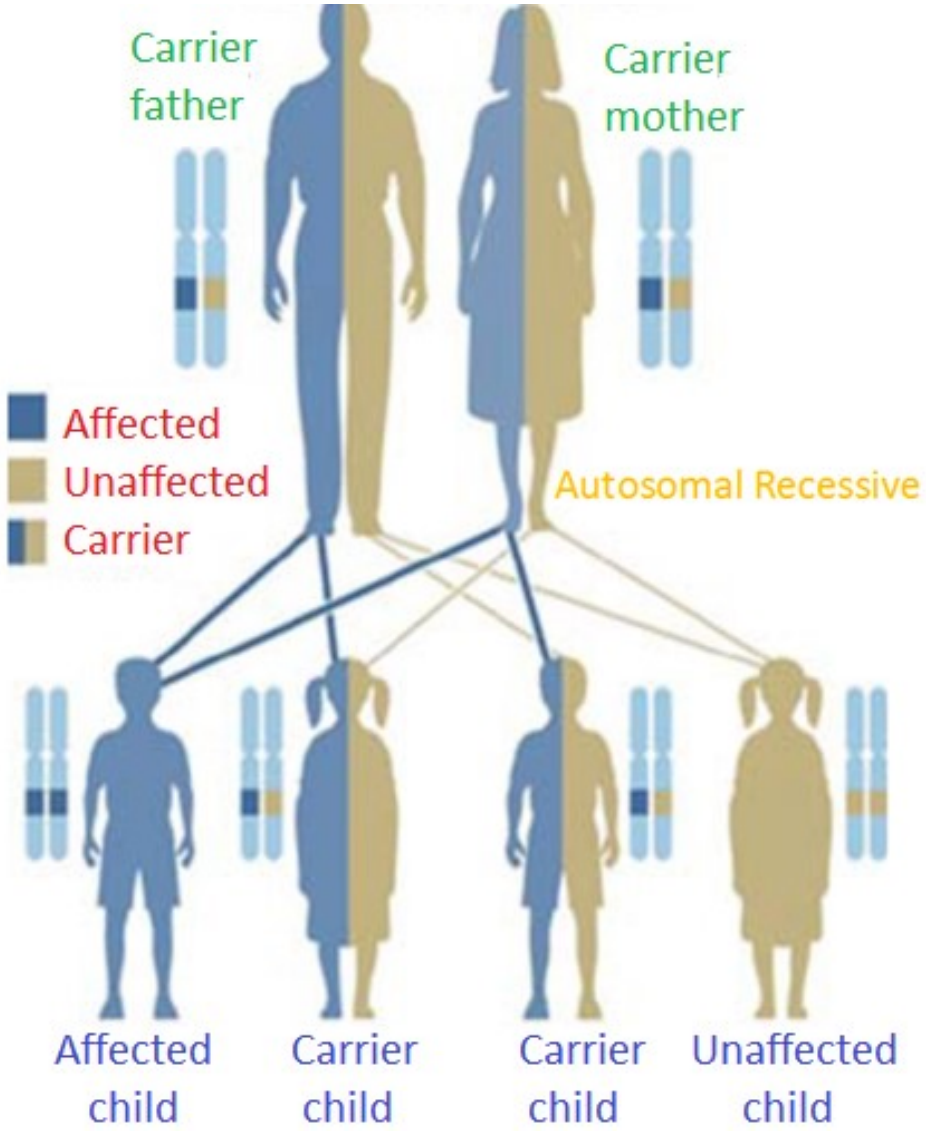
Recorded by, Dr. Eugan Apert, France, In 1906.

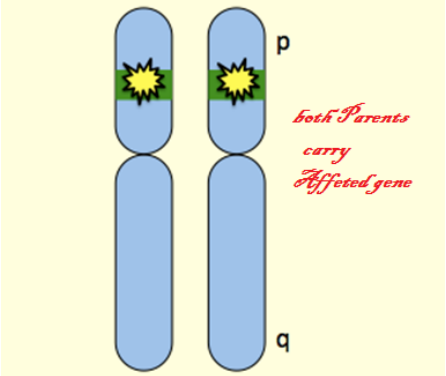
Incidence: 1/200.000 Births.

- Premature closure of ≥ 1 of the cranial sutures.
- Abnormal facial features.
- MR.
- Bossing of head.
- Exophthalmos & hypertelorism.
- Depressed nasal bridge.
- Deformed high arched palate & dental deformities.
- Syndactyly.

AUTOSOMAL RECESSIVE

Both parents are carrier (heterozygous) for the mutant gene & are asymptomatic. The Chance of inheriting the disorder to their siblings is 25%, carrier 50% & unaffected 25%.





FAMILIAL MEDITERRANEAN FEVER

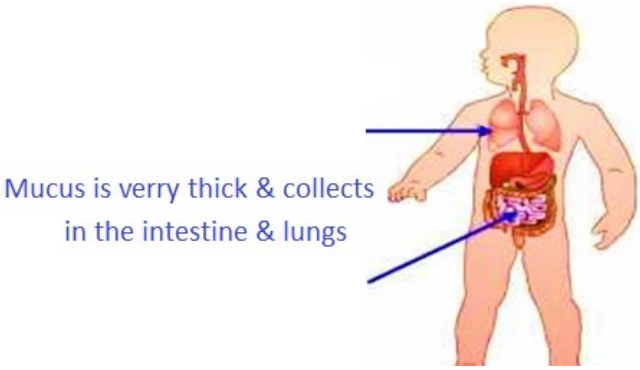
Chromosome 16, Gene FMF, Location 16 p 13.3

Incidence: 1/ 500 Armenian and Sephardi Jews. 1/2600 in Arabs.

•First episode usually occurs in childhood or teenage yrs •Painful episodes last for 12 -72 hrs in the form of abdominal pain, joint pain, chest pain •Fever •Skin lesions •No specific diagnostic test except chromosomal study •Colchicine ⇩ the inflammation.

CYSTIC FIBROSIS

Chromosome 7, Gene CFTR, Location 7 q 31.2 (1930s)

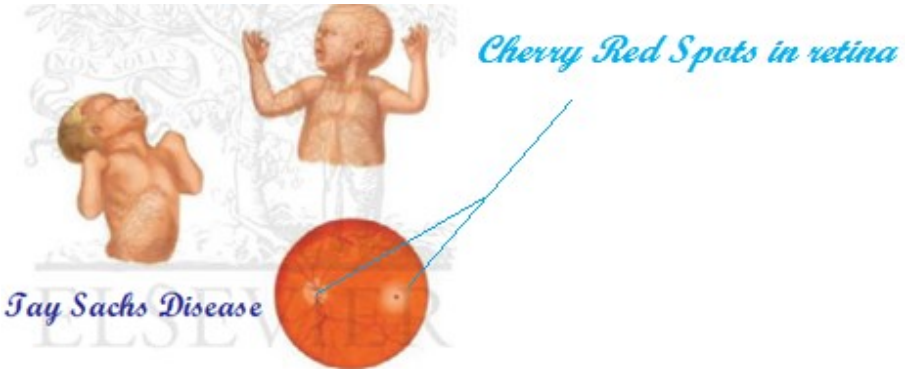


Incidence: *commonest fatal AR disease in Europe. 1/3000 Births, in USA & Europe.* ✳Abnormal transportation of chloride & sodium across epithelium leading to thick viscous secretions. ✳Salty tasting of skin. ✳Recurrent chest infection. ✳Meconium ileus ✳Chronic diarrhea. ✳Cong. absence of vas deferens & infertility. ✳Nasal polyposis. ✳Rectal prolapse. ✳Cholelithiasis. ✳Pancreatitis. ✳Liver Cirrhosis.

TAY SACHS DISEASE

Chromosome 15, Gene HEXA, Location 15 .q 23

Named after Dr. Warner Tay, British ophthalmologist 1881 and Dr. Bernard Sachs, American neurologist, in 1887.



Incidence: 1/3500 in Jewish & 1/320.000 in general population.

•Metabolic disorder. •Deficiency of enzyme B-Hexosaminidase A, results in accumulation of fatty acid derivative (ganglioside) in neurons. •Infantile type is the commonest & severest form, tend to get worse very quickly & child usually dies by age 4-5 years •Delayed milestones. •Paralysis. •Convulsions. •Blindness, Cherry Red Spots in Eyes. •Deafness. •Dementia. •Psychosis.

WERDNIG HOFFMAN DISEASE

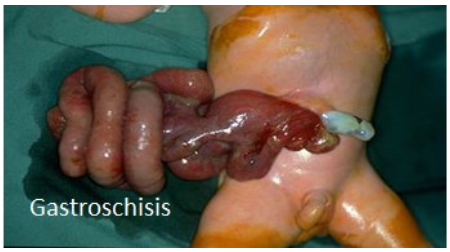
Chromosome 5, Gene SMN 1, Location 5 q 13.2. Described by Dr.Guido Werdnig, Australian neurologist & Dr.Jehann Hoffman, German neurologist in 1891.

Incidence: 1/10.000 Births.

The most common genetic cause of infant death. Degenerative disease of nerve cells of lower brain steam & anterior horn cells of the spinal cord. The infantile type is the severest form, represent 80% of cases. Muscle weakness. Floppy infant, Frog leg position. Dysphagia. Accumulation of secretions in lungs & respiratory distress. Delayed milestones. Areflexia. Skeletal deformities. Fasciculations (twitching of the tongue). Baby usually dies by the age of 20 months.

GASTROSCHISIS

Chromosome 3, Location 3 Q 27.3



Incidence: 1/10.000 Births.

Anterior abdominal wall defect. Usually <4 cm. to the right side of the umbilicus Protrusion of abdominal content. No covering sac.

Cover ē sterile gauze soaked in saline & transfer to surgery.

ALBINISM

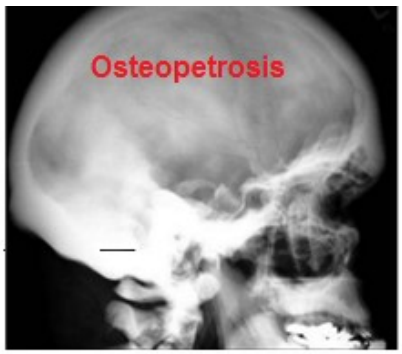
Chromosome 11 , Gene TYR , Location 11q14.3

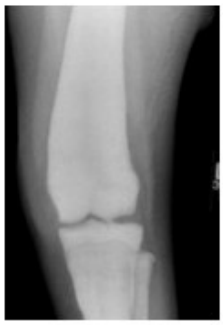
Incidence: 1/17000/ Births.

- Defect in melanin production that results from absence or defect of tyrosinase (a copper containing enzyme) involved in production of melanin.
- Little or no color in the skin, hair & eyes.
- Photophobia.
- Functional blindness.

OSTEOPETROSIS

Diffuse thickening in the region of occiput, cervical vertebrae, maxilla





Densely sclerotic bone
Lack of trabeculation,
Block femoral metaphysis

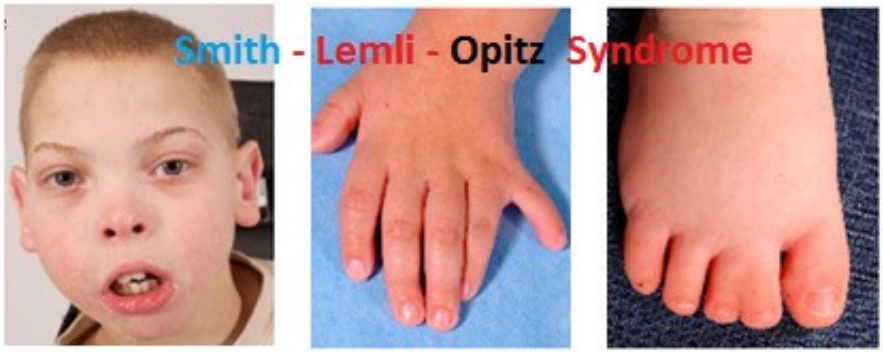
Osteopetrosis, literally "stone bone", also known as marble bone disease, it is an extremely rare hereditary disorder. There is defective osteoclast function & overgrowth of bones; ẃ become thick, dense & sclerotic. However, their increased size does not improve their strength. Instead, their disordered architecture, result in weak & brittle bones that result in multiple fractures ē poor healing. There are 2 types; Infantile osteopetrosis ẃ is AR & Benign adult osteopetrosis ẃ is AD. The disease is characterized by excessive density of bones, obliteration of marrow cavities, development of secondary anemia, defect in osteoclastic activity, failure in remodeling of developing bone. There is an excessive bone formation ẃ is mechanically weak so the fractures are common, & osteomyelitis is common complication of tooth extraction.

No specific treatment.

SMITH-LEMLI-OPITZ SYNDROME

Chromosome 11, Gene DHCR7, Location 11 q 13.4

Described by Belgian Pediatricians Smith Lemli, Luc Lemli & John Optiz, 1964



Incidence 1/20.000/ Births (Caucasian).

Delayed milestones MR Microcephaly Cleft palate Defective dentation.

Syndactyly & Polydactyly Hypospadias Cryptorchidism.

HURLER SYNDROME

Chromosome 4, Gene IDUA, Location 4 p 16.3

Named after, Australian Pediatrician Gertrud Hurler in 1919.

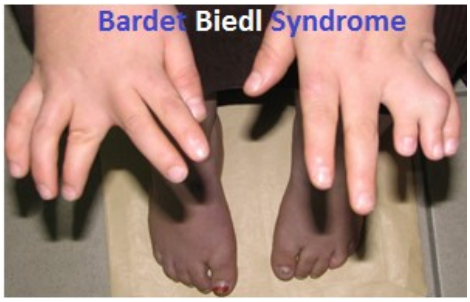
Incidences 1 /100.000 Births.

IEM (Mucopolysaccaroidosis type 1). Deficiency of α -1-Iduronidase enzyme. Coarse facial features, low flat nasal root, macroglossia, widely spaced teeth. MR. Delayed milestones. Short stature. Clouding of cornea. Hepatosplenomegaly. Progressive deterioration ē death occurring by age of 10 years.

BARDET BIEDL SYNDROME

Chromosome 11 , Gene BBS1, Location 11q13.2

Named after Dr. Bardet G.(France) in 1920 and Dr. Biedl A.(Dutch)in 1922



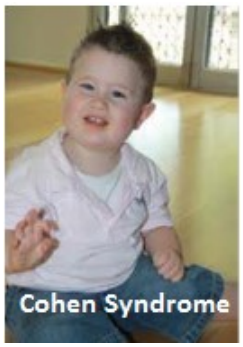
Incidence: 1/150.000 Births in USA & Europe. 1/14.000 Births in Middle East.

⦿MR. ⦿Obesity. ⦿Polydactyl &Syndactyly. ⦿Hypogenitalism. ⦿Retinitis pigmentosa. ⦿Visual impairment.

COHEN SYNDROME

Chromosome 8, Gene COH 1, Location 8 q 22.2

Named after American geneticist Michael Cohen in 1973



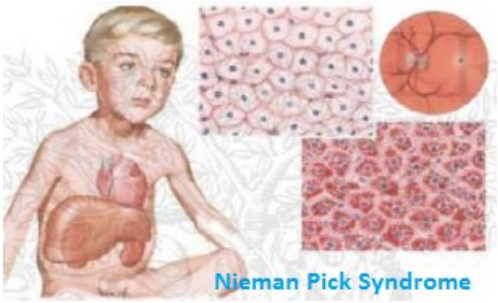
Incidence: very rare <1/200.000 people. Diagnosis is generally raised at school age.

⦿MR. ⦿Square head, short neck & full cheeks. ⦿Microophthalmia, epicanthus fold, downward slanting of eyes & long eyelashes. ⦿Chorioretinal dystrophy & visual impairment. ⦿Upper lip is thin & does not cover the front teeth (giving an open mouth expression) ē prominent upper central incisors. ⦿Thick hair. ⦿Mild cutaneous syndactyly. ⦿Obesity in late childhood or adolescency.

NIEMANN PICK SYNDROME

Chromosome 11, Gene SMPD 1, Location 11 p 15.4

Named after Dr. Albert Nieman and Dr. Ludwig Pick, in 1920.



Incidence: 1/250.000 whole world. 1/40.000 Ashkenazi Jews.

⦿Lipid storage disease. ⦿Harmful accumulation of lipids in liver, spleen, lungs, bone marrow & brain (Sphingolipidosis). ⦿Failure to thrive. ⦿Prolonged jaundice. ⦿Hepatosplenomegaly. ⦿Progressive deterioration of CNS. ⦿Seizures. ⦿Ophthalmoplegia.

⦿The 4 D "Dysphagia, Dysarthria, Dystonia, Dementia".

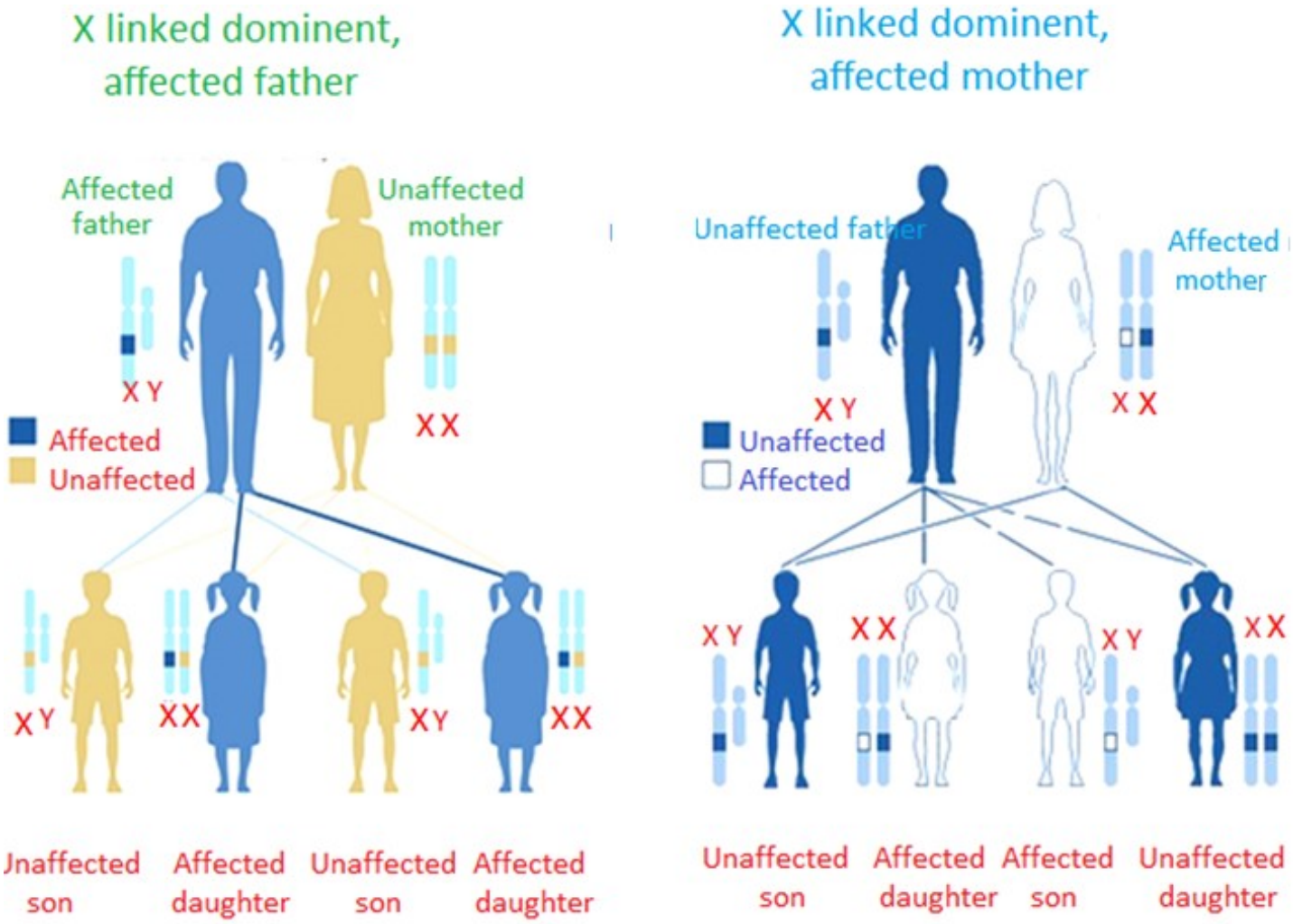
COCKAYNE SYNDROME

Chromosome 5, Gene ERCC 8, Location 5 q 12.1

Named after British Dr. Alfred Cockayne in 1936

Incidence: 1/250.000 Births. ◇Defect in DNA repair mechanism. ◇Affect any or all organs of the body. ◇Progressive disease. ◇Typically become apparent after age 1 year. ◇Delayed milestones. ◇Senile appearance. ◇Sensitivity to sunlight's & sun exposure can cause sunburn. ◇Deafness. ◇Pigmentary retinopathy & eye anomalies.

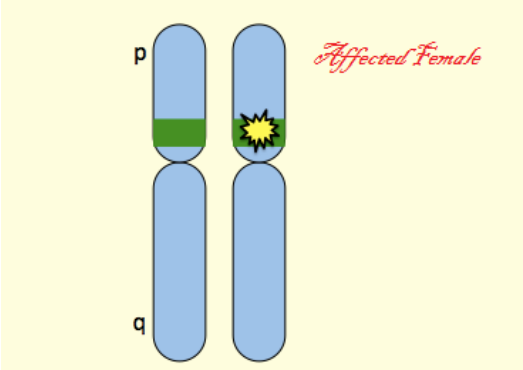
X - LINKED DOMINANT DISORDERS



Characteristic of X- Linked inheritance is that fathers cannot pass X- linked traits to their sons. (No Male- To -Male Transmission).

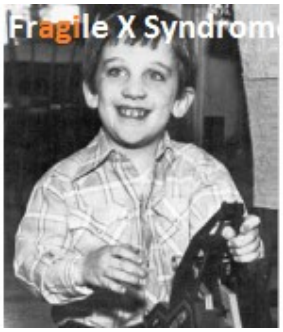
If Father affected: all of his daughters will be affected but none of his sons will affected.

If Mother affected: 50% chance of sibling to be affected regardless of sex.



FRAGILE X SYNDROME

X chromosome, Gene FMR 1, Location X q 27.3



Incidence: 1/5000 children.

◊ The most common form of inherited MR.

◊Baby often LGA & HC > 90th centile.

◊Long face & Prominent & elongated ears.

◊Macroorchaidsm.

RETT SYNDROME

X-Chromosome, Gene MECP 2, Location X q 28

Described by, Andreas Rett, Austrian, pediatrician in 1966.

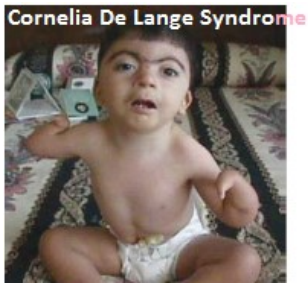
Incidence: 1/10.000 Births.

◊Disorder of the CNS. ◊Affect especially areas of expression. ◊Affect language: severe language development problems. ◊Affect hand use, Apraxia, inability to perform motor function & hand motion abnormalities. ◊Delayed milestones start to appear by age 6-18 months. ◊Autistic like behavior, diminished eye contact loss of normal sleep pattern. ◊Excessive salivation & drooling. ◊Floppy arms & legs, shaky, unsteady stiff gait or toe walking. ◊Breathing problems, ẃ became worse ē stress but normal during sleep.

CORNELIA DE LANGE SYNDROME

X chromosome, Gene DXS 423 E, Location X p 11.22

Named after Cornelia De Lange, Dutch Pediatrician in 1933.



Incidence 1/10.000 live births.

☞Delayed milestones. ☞MR. ☞Hirsutism ē low anterior & posterior hairlines. ☞Arched eyebrows & long eyelashes. ☞Small upturned nose. ☞Crescent shaped mouth & small widely spaced teeth. ☞Low set ears. ☞Limb deformities.

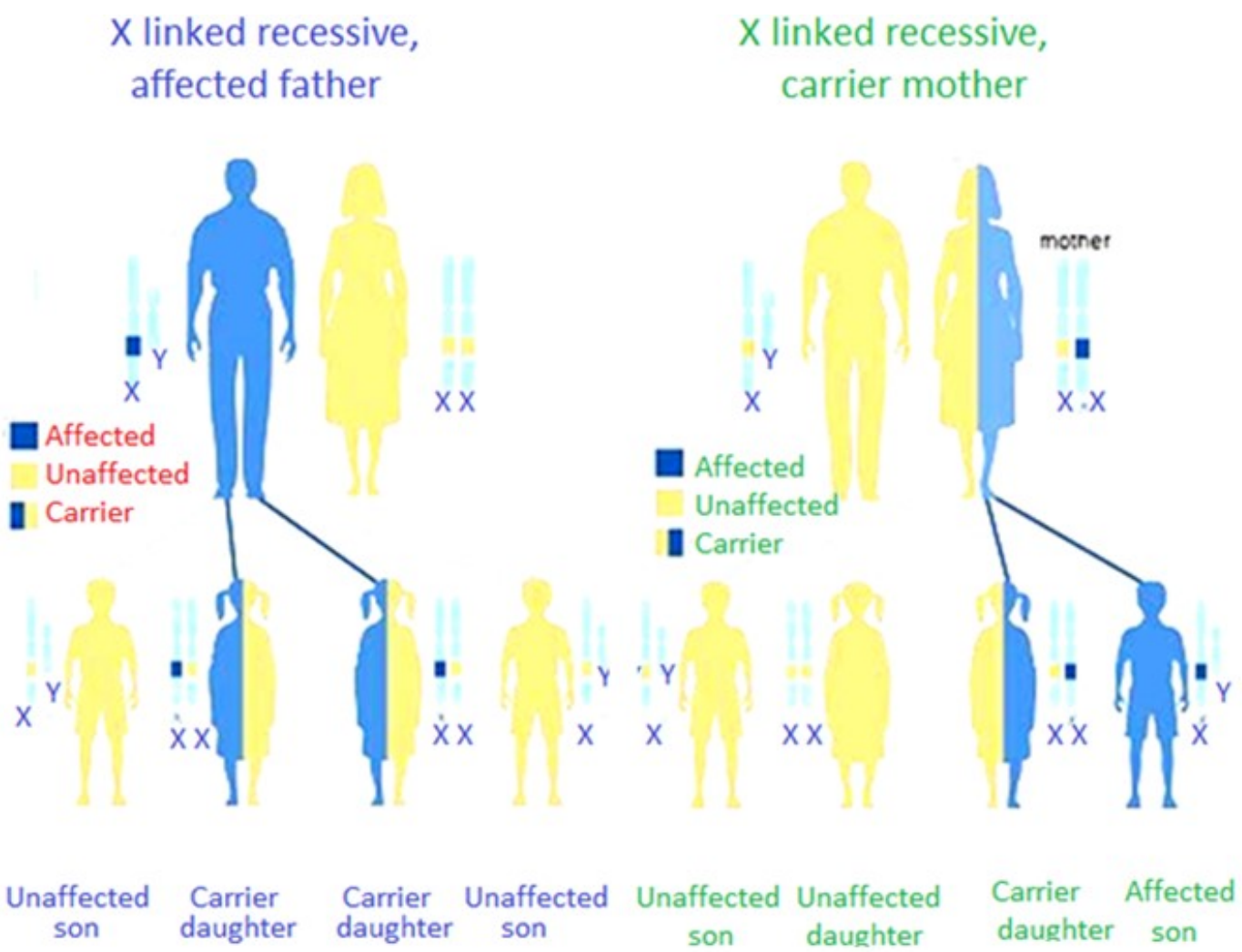
HYPOPHOSPHATEMIC RICKETS

X Chromosome, Gene, PHEX, Location X p 22.11

Vitamin D resistant Rickets. Incidence: 1/20.000 Births.

✻Delayed physical milestones: teething, walking. ✻Delayed closure of fontanels, craniotabes, bossing of head. ✻Broadening of wrist & ankles. ✻Rachitic rosary at costochondrial junction. ✻Green stick fracture of bones. ✻Skeletal deformities, bowing of legs. ✻⇩Serum Phosphors, ⇧ Serum Alkaline Phosphatase. ✻Normal serum calcium, 1- 25 dihydroxyvitamin D & PTH. ✻Give oral Phosphate & Calcitriol.

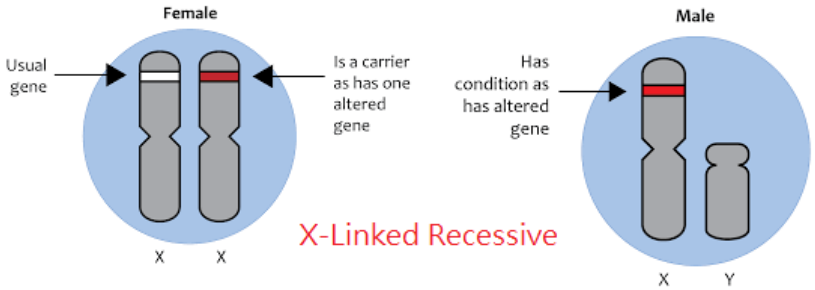
X LINKED RECESSIVE DISORDERS



A characteristic of X-Linked inheritance is that fathers cannot pass X- linked traits to their sons. No Male- to -Male Transmission.

If Father affected none of his sons will affected & all of his daughters will be carriers.

If Mother carrier there will be 50% chance for sons to be affected & 50% chance for daughter to be a carrier.



COLOR BLINDNESS

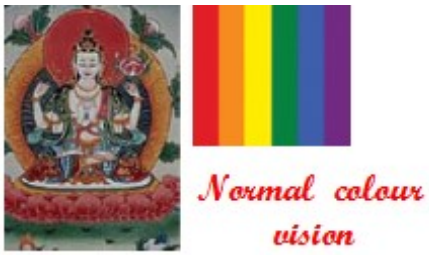
X - Chromosome Gene, OFN1LW, Location Xq28

Described by, English Chemist, John Dalton in 1798

Incidence 5% in males & 0.5% in females.

•Inability or ⇩ ability to see color, or perceive color differences under normal lighting conditions. •People usually have 3 types of cone cells in the eye, each type senses either red, green or blue light (the 3 basic colors) & most of cone cells are found in macula, which is the central part of retina.

NORMAL COLOR VISION



GREEN COLOR BLINDNESS (DEUTERANOPIA)

The commonest 95%

BLUE COLOR BLINDNESS (TRITANOPIA)

RED COLOR BLINDNESS (PROTANOPIA)

TOTAL COLOR BLINDNESS (MONOCHROMACY)

Only as if it were on a black & white television as 2-3 cone pigments are missing .

DUCHENNE MUSCULAR DYSTROPHY

X Chromosome, Gene DMD, Location X p21-21.1

Gower's sign, climbing his legs when in standing up.

Incidence: 1/4000 boys.

- Muscular degeneration, progressive muscular weakness of legs, pelvis, loss of muscle mass, weakness spread to arms, neck & other areas of the body.
- The sensation is intact
- By age 12 years most pts are wheel chair dependent associated ē skeletal deformities.
- Intellectual impairment.
- Average life expectancy is around 25 years.

Investigations:

•Muscle biopsy. •DNA studies. •Rise of serum creatine kinase, creatine phosphokinase & serum aldolase. •Nerve conduction velocity (EMG).

ALPORT SYNDROME

X Chromosome, Gene COL 4 A 5, location X q 22.3

Identified by British Physician.Ceciel Alport in 1927.

Incidence: 1/5000 Births.

•Hereditary nephritis. •Hematuria. •Sensory neural deafness. •Visual deterioration.

• Retinopathy.

WISKOTT ALDRICH SYNDROME

X Chromosome, Gene WAS, Location X p 11.22-p 11.23

Named after Pediatricians A Wiskott (Germany) &R.A. Aldrich(USA), in 1954

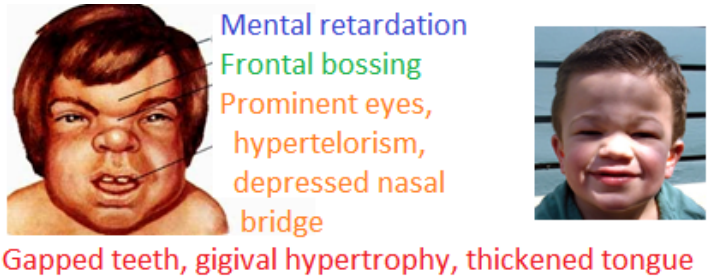
Incidence 1-10/million male is worldwide.

•Thrombocytopenia. •Immune deficiency. •Repeated infection. •Eczema.

HUNTER DISEASE

X Chromosome , Gene IDS , Location X q 23

Named after Charles A. Hunter, Canada in 1917



Incidence: 1/162.000 Births.

•Mucopolysaccharidosis Type II, due to deficiency of enzyme iduronate 2 sulfase required for the degradation of specific glycosamilycans, its absence results in harmful accumulation of these substances in cells throughout the body. •The disease is progressive & life-limiting disease, start to appear after the first year of life. •Delayed milestones •MR. •Large head, prominent forehead, fattened nasal bridge & big tongue. •Deafness. •Visual disturbances. •Joint stiffness, carpal tunnel sy. •Frequent infection of ears & respiratory tract. •Hepatosplenomegaly. •Aggressive behavior.

LESCH NYHAN SYNDROME

X Chromosome , Gene HPRT 1 , Location X q 26.2-q 26.3

Recognized in USA by medical student, Michel Lesch & his mentor pediatrician Bill Nyhan who published their findings in 1964.

Incidence: 1/380.000 Births.

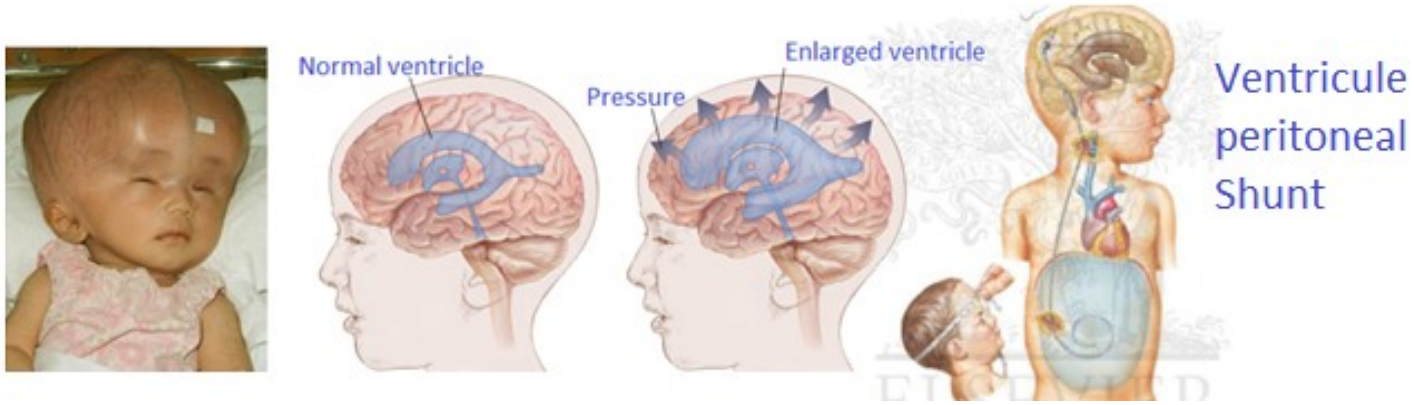
•Inborn error of metabolism result from deficiency of enzyme Guanine Hypoxanthine Phosphoribosyl transferase, causing accumulation of uric acid. •Delayed milestones. •MR. •Self-destructive behavior resulting in self-mutilation through biting finger tips & lips (of unknown cause). •Hyperuricemia & severe gout. •Uric acid urinary stones. •Repetitive movement of arms & legs similar to those seen in Huntington- n's disease.

MULTIFACTORIAL DISORDERS

Conditions or diseases arising from combination of GENETIC & NON- GENETIC causes, including ENVIRONMENTAL FACTORS.

HYDROCEPHALUS

Chromosome 3, Gene Location 3 q 22 - q 24



Incidence 4/1000 Live Births.

Congenital stenosis/obstruction of aqueduct of Sylvia's, foramen of Oval, or foramen of Magendi. Normally choroid plexus produce 750 ml CSF daily. The defect may be in excess production of CSF, or in its reabsorption, or 2^{ry} to IC Hge, or choroid plexus papilloma, or tumor or meningitis. Early detection through U/S ŵ is routinely supposed to be done during the 1st &2nd TM of pregnancy, will visualize such cong. cases, through measuring diameters of BPD, lateral ventricle & thickness of brain cortex, also sonar visualize presence or absence of other apparent congenital anomalies, allowing time for remeasure-

ment of the previously mentioned data, for decision to be taken about termination of pregnancy ẃ is affected by many factors.

Clinical picture

Baby present é progressive ⇧ in HC, bossing of head, wide fontanels, wide separation of sutures, sun-sitting eyes, cracked pott sign, & may result in MR.

Investigations

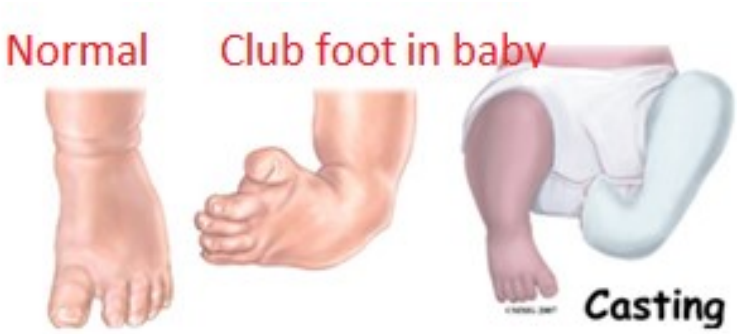
Transillumination using fibroptic light, X ray skull & CT scan brain, also TORSCH screening to R/O IUI.

Management

Ventriculoperitoneal shunt (common operation), or ventriculoatrial shunt operation.

CONGENITAL TALIPES EQUINOVARUS

Chromosome 5, Gene PITX 1, Location 5 q 31.1



Incidence: 1/500 Births (50% of cases are bilateral).

The most common birth defect. More common in males. Normally the neonate can touch leg ē small toe & straight line can pass from the heel of feet to 2nd toe.

May be Positional, in ẃ no restriction of passive movements of ankle joint.

May be structural, in ẃ you elect very restricted passive movements of ankle joint.

Management: through casting.

HARE LIP AND/OR CLEFT PALATE

Chromosome 1, Gene IRF 6, Location 1 q 32 - q 41

Incidence: 1/700 Births.

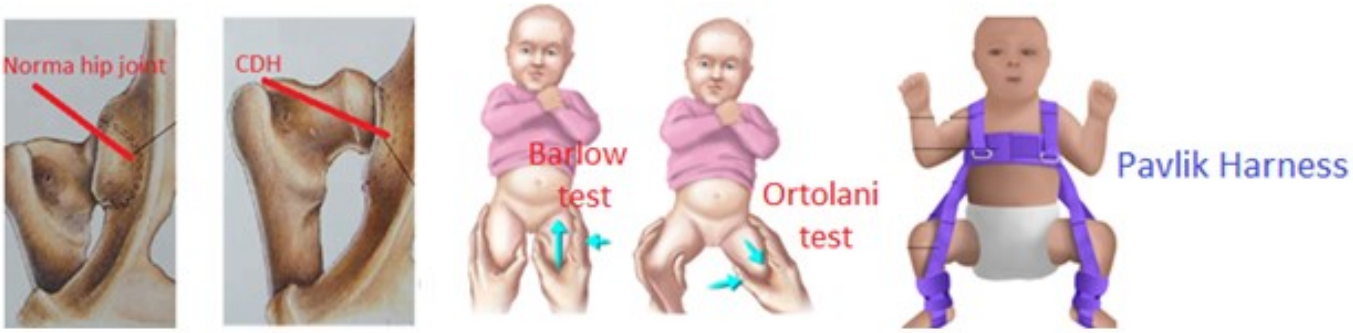
Hare Lip results from failure of fusion of medial nasal & maxillary processes at 6-8 wks of gestation. Cleft palate results from failure of primary palatal processes to fuse at 7-12 wks gestation. Each one may be unilateral or bilateral & may be combined

Management

First operation when baby 10 pounds & thrive for correction of hare lip. Second operation at age one year before starting speech for correction of cleft palate. Small hare lip may treated by T-shaped strapping ẃ may be effective. Baby susceptible to aspiration pneumonia. Showing the parents a picture of such condition before & after operation is helpful for reassurance. The condition needs team approach & speech therapist.

CONGENITAL HIP DISLOCATION

Chromosome 13, Gene Location 13 q 22



Incidence: 1/1000 Babies.

Common in females & breech presentation. Early diagnosis & Rx is important.

Differentiate it from sublaxation of hip where we not feel or hear click, clunk, or jerky movement, in such condition the femur head is within the acetabulum & no asymmetry of both inguinal ligaments, only we detect excessive movement of the head of femur within the joint. CDH may be unilateral or bilateral, if unilateral it characterized by asymmetry of both inguinal ligaments, incomplete abduction of thigh in the affected side & difference in the length of both lower limbs.

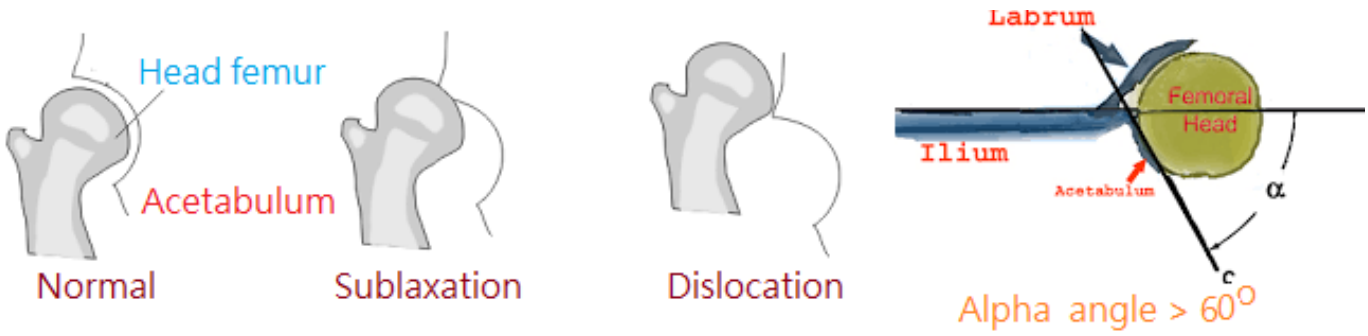
Diagnosis

Barlow test: fixation of one side in abduction position & moving the other é pushing the lesser trochanter to the outside using the big finger to feel a click + clunk + jerky movement.

Ortolani test: bilateral abduction of both thighs together é pushing of the greater trochanter using middle finger to feel a click.

X ray of Hip joint: not conclusive before age of four months when the ossific center of femoral epiphysis appears.

U/S of hip joint: is conclusive test by detection of α, & ß angles during the static & dynamic state of hip joint.

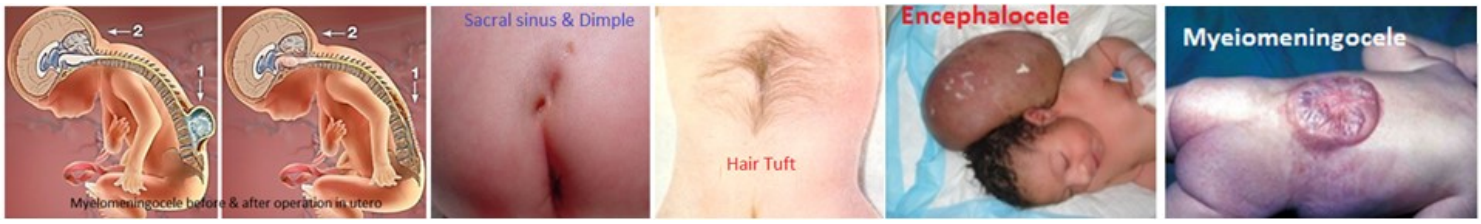


Management

Fixation from age of 2 wks, for a period of 2-3 months, using Pavlik harness, Van Rosen cast, or Aberden cast. This instability worsens as the child grows if not treated.

NEURAL TUBE DEFECT

Chromosome 6, Location 6 q (T Locus)



Incidence: 1/2000 Births. Commonly arise from the lumbosacral region, diagnosed prenatally by U/S & the rise of α fetoproteins in amniotic fluid.

Types

Spina Bifida Occulta: mid defect of vertebral Bodies, presence of dimple, dermal sinus, lipoma, or tuft of hair in lumbosacral region, discoloration of skin. Occasionally associated é Diastematomyelia, Syringomyelia & Tethered Cord. X-ray shows failure of closure of vertebral bodies & most cases develop hydrocephalus later.

Spina Bifida Manifesta: include:-

Meningocele: bone defect, herniation of meninges & intact overlying skin.

Myelolomeningocele: both meninges & spinal cord protrude through the skin defect.

Encephalocele: bone defect of the skull é herniation of meninges & brain tissues.

Clinical picture: usually associated é severe physical & mental disabilities, paralysis of both lower limbs, quadriplegia & incontinence of urine & stool.

Prenatally diagnosis: ⇧ α Fetoproteins in amniotic fluid & U/S.

Management: prophylactic Folic acid (Folate) is necessary for RBC production & neural tube formation in embryo, repeated studies have shown that woman who get 400 mcg daily prior to conception & during early pregnancy reduce the risk that their baby will be born é a severe NTD (spina bifida, anencephaly, encephalocele), all these defects occur during the 1st 28 days of pregnancy usually before woman even know she`s pregnant.

Surgical correction within the 1st few days, selection of cases depend upon the number of affected vertebrae, presence or absence of paralysis of LL, loss of sensation of LL, incont- inence of urine or stool, presence of associated congenital anomalies (VACTERL) in addition to the religious, economic factors.

VACTERL association: should be considered & excluded. It is association of congeni- tal malformations typically characterized by the presence of 3 of the following & occur in 1/10.000 live births:-

- V: Vertebral: NTD
- A: Anal: imperforate Anus.
- C: Cardiac: VSD, ASD, PDA, F_4, CoA
- T: Tracheal: TOF
- E: Oesophageal atresia.
- R: Renal: agenesis, ureteral abnormalities, Hypospadias.
- L: Limb: Polydactyly, wrist/Knee anomalies.

VACTERL association: 20%

- Vertebral: 17%
- Anal 12%
- Cardiac 20%
- Renal 16%
- Limb 5%

- CHARGE association: Cloboma, Heart defect, Atresia choanae, developmental Retardation, Genital hypoplasia, Ear deformity.
- Schisis association : Omphalocele, Neural Tube Defect, Cleft Lip & Palate And Genital Hypoplasia.

ANENCEPHALY

Infant fail to develop a normal head & brain structure, as result of failure of closure of the neural tube at the 3rd - 4th week of gestation.

Incidence: 1 in 670 births. Couples that have had a previous child é a NTD have 1 in 40 chances of recurrence. More distant (2nd degree) relatives to an individual as nices, or nephews would have 1 in 200 risk of a NTD. 3rd degree relatives such as cousins have a 1 in 400 risk. 4th degree would have a risk similar to general population. Anencephaly is seen 5 times more often in females than males.

Causes: *Folic Acid deficiency *Hypervitaminosis A *Undiagnosed diabetes *Environ-emental/chemical exposure.

At Risk: •Women taking anticonvulsant medication •Woman é undiagnosed or uncontrolled DM •Any woman é a family history of NTD.

Suggested screening test: it is common to screen women`s blood for:-

*AFP (protein produced by the fetus that is excreted into the amniotic fluid). Abnormal level may indicate brain defect or NTD.

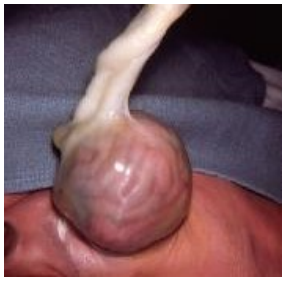
*Amniocentesis can help detect NTD by measuring AFP.

Prevention: tacking 4-5 mg of Folic Acid daily for 2-3 months before conception for all women at risk or having a child é NTD.

Management: there is no cure or standard treatment for it.

EXOMPHALOS

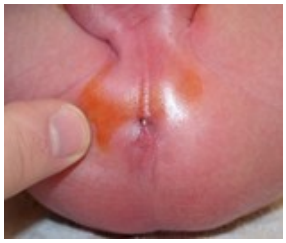
Chromosome 2, Gene Location 2 q 31.1



Incidence: 1/5000 live births

Abdominal wall defect at the base of umbilical cord, infant born ē sac protruding through the defect ẃ contain small intestine, liver, large intestine, sac attached in its center to the umbilical cord & it turned black in color after a while. Considered surgical emergency, primary consideration is infection &drying of the contents. Cover ē warm, moist, sterile dressing. Nasogastric tube to keep stomach empty. Surgical intervention as soon as infant is suitable.

IMPERFORATE ANUS



Incidence: 1 / 5000 live births.

The baby develops this defect or abnormality during the 5th-7th week of the mother's pregnancy. Malformation of the anorectal region, include wide spectrum of abnormalit-ies, either supra, infra, or intermediate to the levator ani muscles (pelvic floor) ẃ is taken as landmark. The rectum may end by blind pouch that does not connect ē the colon or may have opening to bladder, urethra, or vagina.

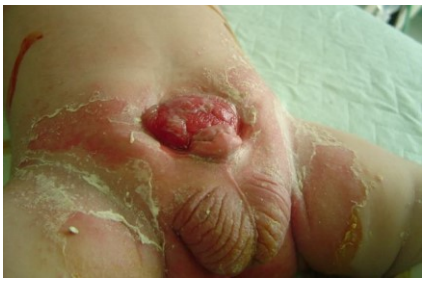
Clinical picture

- Absence or misplaced anal opening, or anal opening very near to the vaginal opening in the female.
- No passage of stool within 24-48hrs after birth or may passed through vagina or urethra
- Abdominal distension.

Investigations: coin shadow X ray, to localize the site of obliteration.

Management: surgical correction.

BLADDER EXSTROPHY



Incidence: 1:30000-50000 live births é a 2:1 male/female ratio. There is no clear evidence for genetic predisposition of this anomaly, however, it can reoccur in the same family by a ratio of 1:100 & there is 500 times greater chance of having bladder exstrophy in the offspring of individuals having bladder exstrophy or epispadias than in the general population. Bladder exstrophy was thought to occur as a result of an abnormal over-development of the cloacal membrane. The abnormality occurs in a range of spectrum, from epispadias to cloacal exstrophy. It is a severe anomaly & predominantly affects males. Characterized by the exposure & protrusion of the mucosal surface of the posterior wall of the bladder to the exterior.

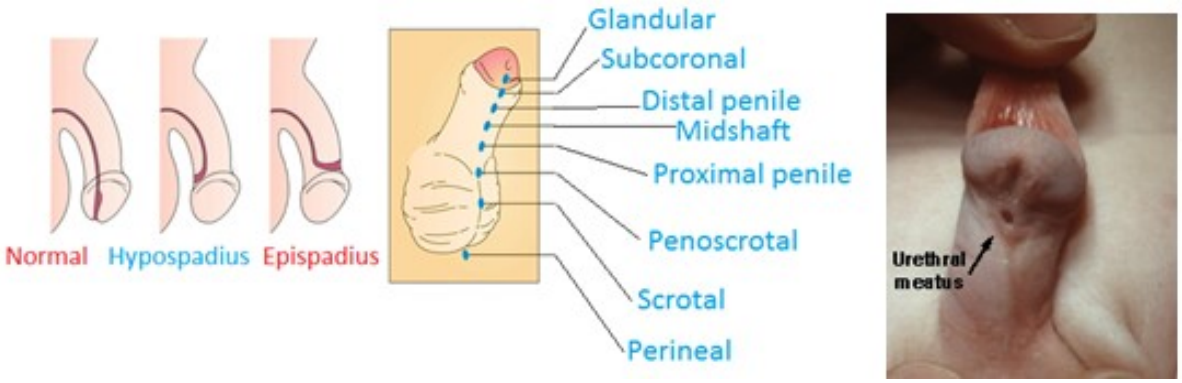
Clinical picture: clinically it is associated é urinary drainage to the exterior & epispadias & may be associated é other anomalies as NTD, vesicoureteric reflux, inguinal hernias, undescended tes-tes, rectal prolapse & foreshortened penis in males & stenosed vagina é

bifid clitoris in females. The main problems associated é this abnormality are: skin irritation from continuous urine leakage & ascending UT infection.

Management

Surgical reconstruction usually done 24-48 hrs after birth, in a single or multiple stages.

HYPOSPADIAS



Incidence

1/300 live male. Varies in severity, in most cases, the penis opening is located near the tip of the penis in the glans (bulb), in more severe form of hypospadias occur when the penile opening is mid shaft or base of the penis & occasionally in the scrotum or perineum (underneath the scrotum), this anomaly is often associated é chordae - a tight fibrous band that results in a downward curvature of the penis seen é penile erection.

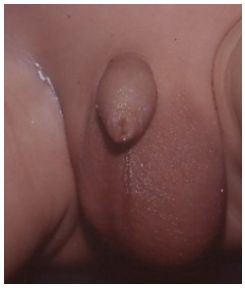
Clinical picture

The meatus occurs on the under surface of the penis, usually associated é three features: ventral meatus, ventral curvature (chordee), dorsal “hood” deficient foreskin ventrally & the child has to sit down to void.

Management

Infant é hypospadias should not be circumcised, surgery is usually completed before the child starts school & today most urologists recommend repair before 18 months of age.

CONGENITAL HYDROCELE

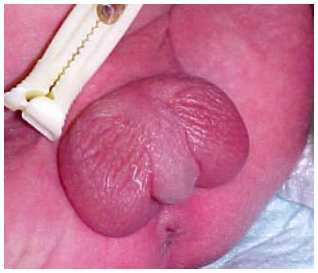


Accumulation of liquid in scrotum between visceral &parietal layers of tunica vaginalis, due to persistence or delayed closure of processus vaginalis of testis. Clear serous fluid, trans illuminating mass in one or both sides, fluid is squeezed into scrotum during birth & known as hydrocele ẃ usually disappears within few months. Presence of frequent erections are common. May be communicating, or hydrocele of the cord, or abdomenoscrotal hydrocele.

Diagnosis: •Cystic swelling. •Non tender. •Trans illuminating. •Can get above swelling & testis not palpated. •Often associated ē inguinal hernia.

Management: surgery indicated if spontaneous resolution not occurs by age 1 year.

DISORDER OF SEX DEVELOPMENT



Incidence: 1/4000 infants. Hermaphroditism can be caused by translocation of a segment of the Y chromosome containing the SRY gene (480000; Yp11.3) to the X chromosome.

Types

•46,XXDSD (Virilzed Female): female genotype 2 ovaries, external genitalia show vaiable degree of virilization.

•46,XYDSD(Undervirilzed Male): male genotype 2testes,external genitalia show variable degree of feminization.

•45,X/46, XY mosaicism (True Hermaphroditism): range from normal male to normal female phenotype, both ovaries & testes present. Complete Gonadal Dysgenesis: either pure CGD (testes or ovaries) or mixed CGD (testes & streak gonad).

Causes

•Congenital adrenal hyperplasia. •Placental aromatase deficiency. •Ovarian tumour. •Maternal hyperandrogenic condition. •Enzyme defect in testosteron synthesis. •Defect in testosteron metabolism. •Endorgan resistance to testosteron. •Maternal drug ingestion: Spironolacton, Phenytoin. •Complete gonadal dysgenesis.

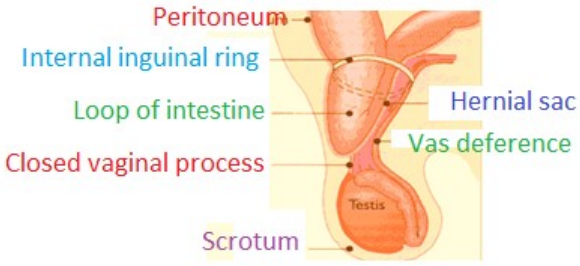
Investigations

- Ultrasonography/MRI for sex determination.
- Chromosomal study.
- Gonadal biopsy
- Hormonalstudies: testestrone, DHT, FSH, LH, 17-hydoxypregnenolone ,11dexycort icosterone, plasma renin activity, serum electrolytes.

CONGENITAL UMBILICAL HERNIA

- Seen in 10% of infants. Affecting boys more than girls, more common in black child.
- Resolve ēout any Rx by age 2-3 years.
- Obstruction or strangulation is rare.
- Surgery necessary only if not closed after 4 years.

CONGENITAL INDIRECT INGUINAL HERNIA



Incidence: 30% of PT infants <1000 gm & 5% of PT infants <1500 gm. Protrusion of whole or part of intestine through processes vaginalis. More common in males. In females the ovary is often in the sac. May be: reducible, irreducible, obstructed, strangulated, or inflamed.

Management: inguinal hernia repair is the most common operation performed in the PT infants.

CONGENITAL CRYPTORCHIDISM

Incidence: 5% in FT boys & 1% in 1 year old boys.

- Most common genital problem in paediatrics. Commoner in PT & SGA.
- Failure of intra abdominal testis to descend into scrotum.
- The testis either absent, ectopic, retractile (can manipulated into srotum).
- May be unilateral (80%), or bilateral (25%).
- Testis often palpable within inguinal canal.
- Non palpable testis occur in 20% & 20-40% of non palpable testis, the testis are absent on surgical exploration.
- 90% of untreated males ē bila-teral absence develop azospermia & their risk of neoplasia is 3%.
- Commonly asso ciated ē Prader Willi, Kallmann & Laurence Moon Biedl syndrome.

Diagnosis: •Clinical examination. •Ultrasonography abdomen & pelvis.

Management: surgical consultation at age 6 months. Operation usually by age 1 year, orchidopexy if testis is felt in inguinal canal or below (not retractile), orchidectomy if atrophic intraabdominal testis detected especially after puberty for high risk of malignancy.

AUTISM

Chromosome 2, Gene Location 2 q 31.1

Incidence: 1/100 Children. 4 times more prevalent in boys. No known racial, ethnic, or social boundaries. Very complex, often baffling developmental disability. First described by Leo Kanner in 1943 as early infantile autism "Auto"- Children are "locked within themselves". For next 30 years, considered to be an emotional disturbance. Very likely neurological in origin- not emotional, not the refrigerator mom. Autism impacts normal development of the brain in areas of social interaction & communication.

☆Difficult to communicate ē others & relate to the outside world.

☆Unusual responses to people.

☆Lack of expression & communication ē others.

☆Attachment to objects.

☆Resistance to change in routine & never say "i" .

☆Lack of emotion ē others - masked face.

☆May exhibit repeated body movements: hand flapping, roc-king, abnormal movements & habits.

☆Compulsive behavior.

☆ Echolalia & language problems.

☆⇧ tolerance to painful stimuli.

☆Occasionally, aggressive &/or self-injurious behavior may be present.

☆Epilepsy is common to occur.

Locked within themselves " young boy ē autism who has arranged his toys in a raw (compulsive behaviour)

HYPERACTIVE CHILD

First called minimal brain damage. Afterwards called minimal brain dysfunction. Next called ADD. Finally called ADHD (Attention Deficit Hyperactivity Disorder). Its prevalence rate is about 6-10%. Only affects children, Symptoms present before age 7 years & disappear in adolescences. The male to female ratio is 3:1. It is a brain-based disorder ẃ is associated ē lifelong impairment in functioning. Environmental factors can contribute to the expression, severity, course & comorbid conditions. Long-term developmental outcomes for individuals ē ADHD can include serious substance abuse, chronic criminality, depression & suicide.

Clinical picture

Significant impairment in social or school functioning. Some symptoms that cause impairment are present in 2 or more settings (e.g. school, home, recreational settings). Not due to another disorder (e.g., Autism, Mood Disorder, or Anxiety). The following are required for diagnosis of ADHD:-

<u>Inattention symptoms (at least 6 symptoms required)</u>

- Fails to give close attention to details or makes careless mistakes in schoolwork, etc.
- Difficulty sustaining attention.
- Does not seem to listen when spoken to directly.
- Does not follow through on instructions & fails to finish schoolwork, etc.
- Difficulty organizing tasks & activities.
- Avoids tasks requiring sustained mental effort.
- Loses things necessary for tasks or activities.
- Easily distracted by extraneous stimuli.
- Forgetful in daily activities.

Hyperactivity-Impulsivity symptoms (at least 6 symptoms required)

- Difficulty playing or engaging in activities quietly.
- Always "on the go" or acts as if "driven by a motor".
- Talks excessively.
- Blurts out answers.
- Difficulty waiting in lines or awaiting turn.
- Interrupts or intrudes on others.
- Runs about or climbs inappropriately.
- Fidgets ē hands or feet or squirms in seat.
- Leaves seat in classroom or in other situations in ẃ remaining seated is expected.

Management

- Stimulant medications "1st line"; Ritalin, Concerta, Metadate, Given in short-acting (4-6 hours) or long-acting (6-12 hours) doses.

The side effects include; difficulty sleeping, lack of appetite, fatigue, headache, stomach-ache, possible occurrences of motor tics

- Alternative non-stimulant medications; Strattera, Antidepressants such as Zoloft or Cel-exa. Affect dopamine & norepinephrine levels in the brain.

Side Effects: difficulty sleeping, lack of appetite,

Stimulant medications & behavior therapy are currently the only established evidence based treatments for ADHD. Combined behavioral-pharmacological treatment has the greatest impact on functional outcomes, is preferred by parents & teachers & is most likely to result in normalization of behavior .

CHAPTER XII

- Vaccination
- Weaning
- Infant Development
- 12 Reasons Babies Cry
- 6 Serious Baby Symptoms
- Child Abuse
- Poisoning

VACCINATION

Immunization is a procedure designed to ⇧ concentration of antibodies &/or effector T-cells ẃ are reactive against infection through administration of antigenic material (vaccine) to stimulate an individual`s immune system to develop adaptive immunity to a pathogen, so the vaccine is biological preparation, improves immunity to particular disease, typically contains agent resembles disease-causing microorganism. It is often made of weakened or killed microbe or its toxins, or one of its surface proteins, stimulate immune system to recognize the microbe as foreign body, destroy, remember it. Immunization is one of the most powerful, cost-effective of all health intervention, prevent debilitating illness, disability. The rights of medication administration should applied to the vaccine, these rights of medications include; right pt., right vaccine, to be given in the right time, giving the right dosage, through right route, in the right site & to be associated ē right documentation.

Types

- Live bacteria (BCG).
- Live virus (OPV, MMR).
- Killed bacteria (Pertussis, S. Typhi).
- Killed virus (IPV, Rabies, HAV).
- Toxoid (DT, TT).
- Capsular polysaccharide (HiB, Pneumo, Meningo).
- Viral subunit (HBsAg).
- Bacterial subunit (acellular Pertussis).

Obligatory vaccination schedule

BCG: 0.1 ml SC, at first month of age & 5 years after doing tuberculin test.

Polio: 2 drops, at 2, 4, 6, 18 months, 5 years.

DPT: 0.5 ml IM, in shoulder muscle, at age 2, 4, 6, 18 months, & at 5 yrs age boost-er dose of DT (diphtheria tetanus).

Hepatits B: 0.5 ml IM at the shoulder muscle at age 2, 4, 6 months.

Measles: 0.5 ml IM at the gluteal muscle, at age 9 months.

MMR: 0.5 ml IM at shoulder muscle, at age 12 months.

Additional vaccination schedule

- Chicken pox: at age 1& 5 year.
- Meningococcal: at age 2 & 5 year.
- Haemophilus influenza bacteria: at age 2, 4, 6 & 18 months.
- Pneumococcal: at age 2, 4, 6, 18 months.
- Rota virus: at age 2 & 4 month.
- Hepatitis A: at age 12, 24 months.

Recommended vaccines protect against 12 diseases

•TB •Polio •Diphtheria •Measles •Pertussis •Mumps •Tetanus •Rubella •Hepatitis B •Chicken pox) •Meningococcal disease •Haemophilic influenza type b.

Tuberculosis Vaccine

BCG: 0.1 ml given ID over right deltoid muscle, or by multiple puncture technique, given within 1 month from birth, scar formation 2-6 weeks after injection, small spot may app-ear at site of injection, can grow into circle up to 7 mm diameter, may become crusty where fluid has dried on surface, can be painful, bruised for few days other side effects

rare, severe anaphylactic reaction very rare, vaccine not given to immunocompromised or HIV infection because as it may cause disseminated or life threatening infection.

Poliomyelitis Vaccine

Eliminated from Egypt since 2005, 2 types; one given PO (OPV) other given IM (IPV) 0.5 ml, also available as combination vaccine containing (IPV+DPT) or (IPV+DPT+HB), given at birth & at 2, 4, 6, 12, 18 months of age. It is one of the safest vaccines, recommended to give children < 5 years extra doses should there be any national campaign, side effects of OPV is minimal, may lead to GIT upset like diarrhoea, vomiting, does not cause fever, extremely rare side effect is vaccine associated paralysis in 1/million doses, the injectable form IPV may lead to mild fever lasts for 1-2 days, local pain, swelling, redness, tiredness in 20% of cases.

DPT Vaccine

0.5ml IM in shoulder muscle at ages 2, 4, 6, 18 months & 5 years, side effects, local pain, swelling, redness, difficulty in walking in 50% cases persist for 1-3 days, systemic side effects as fever, excessive crying, anorexia, vomiting, irritability 25% of cases. Pertussis vaccine not to given & replaced by DT vaccine to children ē any of the following; previous history of neurological disease, family history of neurological disease, severe reaction to previous vaccination, or in case of over age 18 months (as reactions are more frequent).

MMR Vaccine

0.5 ml IM at age 1, 5 year, most children are perfectly well after vaccination, few develop local pain, mild fever, faint rash 7-10 days later lasts for few days, mild swollen face.

Hepatitis B Vaccine

0.5 ml IM at 0, 1, 6 month, 20% of vaccines show local reaction at injection site. Fev-er, headache, sore throat, nausea, diarrhea, anorexia, are rare.

Hemophilic influenza type b Vaccine

0.5 ml IM at age 2, 4, 6, 12 months, 25% of cases experience pain, redness, swelling at site of injection, mild fever, no evidence linking the Hib vaccine to autism.

Meningococcal meningitis Vaccine

0.5 ml IM at age 1, 5 year, 50% of cases develop pain, redness at site of injection, lasts 1-2 days, less number have mild fever.

Varicella vaccine/chicken pox

0.5 ml SC at age 1, 5 year, local reaction at injection site, mild fever in 1% of cases, mild rash in 5% cases appear after 5-12 days.

Influenza vaccine

0.25 ml IM at age 6 month, repeated every year in autumn.

Desensitization

-0.1 ML 1/20 concentration + 0.05 ml adrenaline subcutaneous.

-0.1 ML 1/10 concentration + 0.05 ml adrenaline SC, after 30 minute.

-0.01 ML full concentration + 0.05 ml adrenaline SC, after 30 minute.

-0.1 ML full concentration + 0.05 ml adrenaline SC, after 30 minute.

-1/2 ML full concentration + 0.05 ml adrenaline SC, after 30 minute.

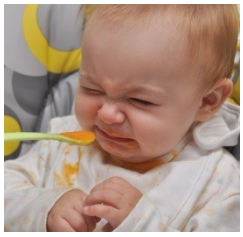
-Full dose IM, after 30 minutes.

(ē presence of adrenaline, decadrone ready for use in case of emergency)

Vaccine	Birth	1 month	2 months	4 months	6 months	12 months	15 months	18 months	24 months	4 years	5 years	6 years	
HepB (prevents hepatitis B)	1st	2nd			3rd								1/2 ml IM
DTaP (prevents diphtheria, tetanus, pertussis)			1st	2nd	3rd		4th			5th			1/2 ml IM
Hib (prevents haemophilus influenzae type b)			1st	2nd	3rd	4th							1/ ml IM
IPV (prevents polio)			1st	2nd	3rd					4th			3 drops PO
RV (prevents rotavirus)			1st	2nd	3rd								1 ml PO
PCV (prevents pneumococcus)			1st	2nd	3rd	4th							1/2 ml SC/IM
Flu (prevents influenza)	Ask your baby's provider if your child needs other flu vaccines, like H1N1.				yearly								1/2 ml IM
MMR (prevents measles, mumps, rubella)						1st				2nd			1/2 ml IM
Varicella (prevents varicella, also called chickenpox)						1st				2nd			1/2 ml SC
HepA (prevents hepatitis A)						1st and 2nd at least 6 months apart							1/2 ml IM

BCG 0.1 ml ID during the 1st month

WEANING



May be difficult task for the mother to perform, but usually has happy end. Weaning is the process of gradually introducing infant to what will be its adult diet & withdrawing the supply of its mother`s milk. The American Academy of Pediatrics recommends feeding a baby breast milk for the first 6 months of life & continuing breast feeding until the child is at least one year old & for as long after that as the mother &child both wish to continue. In general, babies under 6 months have kidneys &guts that are not mature enough to cope é a more diverse diet, research shows that babies need nothing but breast milk or infant formula for the first six months of life, this gives a baby`s digestive system time to develop so that they cope fully é solid foods, & that early weaning can ⇧the risk of infections & the development of allergies like eczema & asthma. It is recommended also to start weaning when;-Baby is able to stay in sitting position & are able to hold their head steadily - Coordinate their eyes, hands & mouth, can look at food, grab it, & put it in their mouths all by themselves.- Able to swallow their food. The process of weaning is slowly over months, slowly tapering off how long & how often mother breast feed, over the course of months, this will cause breast milk to gradually diminish & prevent discomfort caused by engorgement, mother to try first to drops the mid-day breast-feeding session, once mother successfully dropped one feeding she can start working on dropping another, but not to give the child cow`s milk but iron- fortified formula during the first year of life.

Foods to introduce to baby from age 6 month: as a role we start gradually, item by item separately, in a small amount ẃ gradually increased, watching for any sign of indigestion or allergy. Starting by group 1.& gradually moving to groups II & III.

Group I: mashed or soft cooked fruit & vegetables like; potato, sweet potato, carrot, apple, pear, banana, avocado. Make into purees & given by spoon.

Group II: baby rice, baby cereals mixed é fortified milk, yoghurt (skimmed milk).

Group III: soft cooked meat such as chicken, mashed fish, mashed hard-boiled eggs. Gradually the mother will be able to ⇧ the amount & variety of food the baby eats until he can eventually eat the same as the rest of the family, in smaller amount.

From the 8 month the baby will move towards eating three meals a day.

Foods ẃ not to be given to baby below 1 year

Salt, honey, sugar, whole nuts, tea, coffee, soft boiled or raw eggs, liver paste, soft, coked, cheeses, certain fish ẃ may contains traces of mercury (shark, sward fish).

Allergic baby

If the child already has an allergy or an allergic condition as asthma or eczema, then he has more of a chance being allergic to peanuts, we try foods that are most likely causing reaction one at a time starting é small amount e.g. grains, as wheat (& others that contain gluten), fish, citrus fruits, nut butters, egg & Cow`s milk.

When is it time to give up the Pacifier

12 months is a good time for weaning the child from the pacifier because by that time this marks the beginning of a dramatic speech development phase. If the child often has a pacifier in his mouth, he may be less likely to babble & practice talking, also for normal development of tongue, lip, & avoidance of pushing upper teeth towards the lip.

INFANT DEVELOPMENT

The critical period of child is the first 1000 days of life"the golden interval". Ensuring normal growth & development of the child physical, mental, psychological & social milestones are important to look for. Many factors affect child growth & development, starting from time of pregnancy, include; mother health, medications, genetic factors, drug abuse, IUI, availability of medical facilities, mother nutrition & education, home atmosphere, sanitary environment, water supply, environmental pollution & traditional practices…….

Age One Month


Social: the baby start to develop social smile, when mother`s & baby`s eyes meet, the baby may give mother a smile. The baby prefers human faces to all other patter-ns. Baby smile at faces when closed to him or stairs at them. Baby respond to comforting voice é facial movement & by alteration of breathing. The baby has erotic feeding schedule. Demanding cries. He/She sleep most of the time when not being handled or fed, sleep off & on random times for 12-18 hrs/day.

Motor: baby lies in a more relaxed & less flexed posture. No head control when in sitting position. Raise his head slightly when in prone position. Moves head from side to side when in prone position or in back position. Hands stay clenched. Makes jerky, quivering arm thrusts. Brings hands within range of eyes & mouth. Strong grasp reflx.

Language: many researchers believe the work of understanding language begins while a

baby is still in utero, baby begins to make sounds cries first, then vocalizes; oohs & aahs & makes cooing sounds in the first month or two.

Vision: see best at 20-30 cm. Has blurred vision & see only black & white pattern. Follow briefly dangling object at distance 20-30 cm when moving in front of his eyes in range 45 degrees. Eyes wonder & occasionally cross. Baby will close his eyes in the presence of sudden bright light. Presence of red eye reflex.

Hearing: respond to familiar sound & voices by either turning head toward it, blinking, deep breath, or jerky movement of limbs.

Smelling&Taste: recognize the scent of his own mother`s breast milk. Prefere sweet rather than bitter taste.

Sensation: prefers sot sensatios & dislikes rough or abrupt handling.

Growth parameters

Head circumference: average HC is 35cm at birth & ⇧½ inch/month (1.27cm), during the first six months of life.

Weight: average 3.5kg, ⇧ 1.5-2 pounds/month (700-900gm), during the first five months of life (baby double his birth weight by the end of fifth month of life).

Height: average 50 cm. at birth, ⇧ 1 inch/month (2.54 cm), during the first six months of Life (about 65 cm by the end of six month of life).

Baby at age 3 Months

Social: baby smiles responsively. Shows emotions. Showing his preference to his mother, able to recognize his mother & other care givers. Enjoy playing é other people & may cry when playing stop. Begin to react & relate to the world around them. Interacting é people & may imitates some movemental facial expression from the person playing é him. Crying is no longer the baby`s primary method of communication. The baby enjoys being around other babies. He/She sleep 6-7hrs at a time ẃ translates into a good night`s sleep for his mother & during the day baby will take a few naps of about 1.5-2hr each day.

Motor: lies on tommy é propped up on forearms é the head up & looking. When head upright, head has reasonable control, able to hold his/her head about 45^0. When pulled up to sit, head does not flop back. Kicks are getting stronger. Pushes down on legs when feet are placed on a firm surface. Grasp reflex disappear as many of other primitive reflexes. Play ē his own fingers. Sucks fingers & fist. Grasp clothing & hair of others wh-om come nearer to him. Hands are kept open most of the time. Will not pick up a toy. Briefly wave a rattle put in his hand & go straight into the mouth. He/She can roll over one way.

Language: begins to babble, squel, gurgle. Begins to laugh. Makes lauder sounds. Imitate some sounds.

Vision: eyes are bright & alert. Acuity & Field of vision improving. Excited to see his mother. Excited to see food coming. Focus on moving objects in front of his eyes. Recognize familiar objects & people at a distance 40-50 cm. Watches faces intently. Gaze intently at his/her own reflection in a crib mirror. Prefer to look at brightly coloured toys.

Hearing: identify, like, smile & turn head at the sound of his mother. Searching for voice of her mother. Love listening to all kinds of music. Quieted when hearing an unexpected sound.

6 Months

Social: often seems happy. Knows familiar faces. Start to be conservative towards strangers. First sign of fear when baby is é strange people or in new situations. Shows curiosity about things nearby. Likes to play é others, especially parents, peekaboo (the person hides his face, pops back into the baby`s view & says I see you). Sleep longer at night 6-8 hour consistently & takes naps 2-3 times a day, each lasts for 1 -3 hr.

Motor: now baby show complete head control. Fully sitting éout any support from mom or dad. When standing, support weight on legs. Rolls over in both directions (front to back & back to front). Begins to pass things from one hand to another. Still everything goes into his mouth & starts to reach for a toy.

Language: more babbling é new vowel consonant combinations. Makes sounds to show joy & displeasure.

Vision: able to see at longer distances. Acuity & field of vision improving. Likes to lo-ok at self in a mirror. Looks around at things nearby. Eyes may change from their birth color.

Hearing: turns decisively to a side to locate a noise. Respond to own name. Respond to sound by making sounds.

Growth

Head circumference: is about 45 cm ⇧ about ¼ inch/month (0.6 cm.) to reach about 49 cm by the end of the first year.

Weight: is about 7-8 kg & gain about 1 pound/month (450 gm) (=15 gm/day) to reach ab-

out 11 kg +/- 1 kg by the end of the first year.

Height: is about 65 cm & gain about ½ inch/month (1.27 cm) to reach about 75 cm by the end of the first year of life.

Teething: the first teeth start to appear (lower central incisor).

9 Months

Social: respond to his/her own name & simple commands. Understand the meaning of "NO". Pay attention to conversation. Show interest in & dislike of foods. Has his favorite toy. Looks for things he sees you hide. May be afraid of being left alone. Imitates speech sounds.

Physical: get on hands & feet & rock back & front. Crawl backwards first, then forw- ard. Begin to pull up to stand. Hold an object in each hand. Bang toy on table & bangs objects together. Puts hands forward when the head is pointed to the ground (parachute reflex) to protect self from falling. Flying baby stunt is an example of the parachute reflex seen in the picture above.

12 Months

Social: uses simple gestures, as shaking head for “no” or waving ”bye bye”. Pays increasing attention to speech. Responds to “no”. Cries or shows emotion when told “no”. Can express emotions ranging from happy to sad. Identify by use everyday objects; toothbrush, cup, hair-brush, or toy telephone. Have favorite things & people. Explore things in different ways, like shacking, banging, or throwing. Find hidden things easily & search for his toy if it falls down. Tests parental response to his actions during feeding (what do you do when he refuse). Hands his mother a book when he wants to hear a story. Puts out his arm or leg to help his mother during dressing. Play “ball“ receiving & returning a rolled ball. Follow a simple directions like, “pick up the toy”/& simple verbal requests. Pokes up é index (pointer) finger. Able to sleep up to 12 hrs at night éout a feeding.

Physical: crawls forward on belly by pulling é arms & pushing é legs. Gets from sitting to crawling to prone position. Pull up to standing position. Sit back down from standing position. Walk holding on to furniture or é assistance. Stands momentary éout support. Cruises or take a few steps unassissted. Use pincer grasp, pick up food & small objects é thumb & index finger. Finger feedhimself. Takes objects out of con-tainer & put things in it. Turn pages in a book, but often several at a time.

Language: saying Da-Da & Ma-Ma & exclamations like“Uh-Oh“. Saying two wards other than Da Da, Ma Ma. Tries to say words you say.

Vision: follow a fast moving object. Looks at the right picture or when it is named.

Growth: his birth weight is trippled. Grow ½ inch of height each month. ⇧ head size by about ¼ inch/ month. Four to six teeth erupted (lower & upper central incisors).

18 Months

Walk alone & dance to music. Build a tower out of blocks. Climb stairs while holding in. Drink well from cub. Begin to feed self é spoon. Scribble é crayon or pencil. Take afternoon nap. Saying 10-15 words. Saying two word sentences, as “mommy up”. Have first molar teeth appear.

24 Months

Run well, stand momentarily on one foot, kick & catch ball. Can go up-stairs one foot be-s-ide the other in each step. Turns pages in a book, one at a time. Turn door knobs. Wearing the shoes. Begin to ride a tricycle. Saying about 50 words, beginning to put 3 words together (me want ball). Identify his body parts & naming pictures. Appetite ⇩ greatly. Begins to have bladder & bowel control.

36 Months

Make walk up & down the stairs é alternating feet (éout holding the rail)

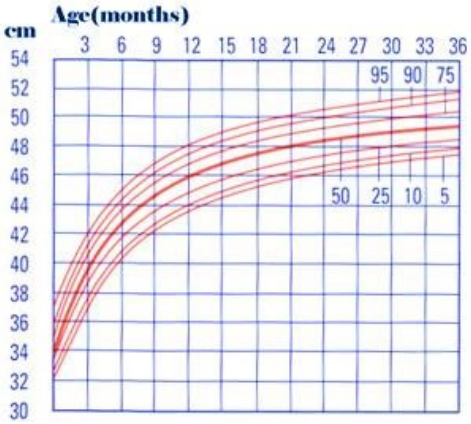
Briefly balance & hope on one foot

Can constructe a block tower of >6 cubes

Can dress self

Make walk up &down the stairs é alternating feet. Can briefly balance & hope on one foot. Can construct a block tower of >6 cubes. Can dress self. Vocabulary many hundreds of wards. Like to listen to stories & ask for it. Counts 3 objects. Follows instructions formed of 2 or 3 steps. Answer simple questions & frequently asks questions. Can copy a circle é a pencil or crayon. All 20 teeth appear. May have daytime control over bowel & bladder functions (may have night time control as well).

Head Circumference



Body Segments: Upper segment is longer than lower segment since birth up to age of 12 years. Upper segment is equal to lower segment after age 12 years.

Teething: at age 2 yrs there are 20 milk teeth, not replaced if it falls. Child start to change milk teeth to permanent one at age 4 years, each year he change 4 teeth. At age 6 years the cusped teeth start to appear (has no milk teeth before).

THE 9 MAJOR PHYSICAL MILESTONES

1-Eye contact (6 -8 weeks): between the baby & his mother. Is the first milestone to be noticed by the mother? It means that baby pay attention to his mother & following her é his eyes, means that his neurological growth & ability to communicate are on track. Also he is demonstrating that his brain is registering a familiar face, in a sense, he is saying "Hey, I Know who you are".

2-Smiling (8 weeks): an infant can`t produces what is called a social smile until about 8 wks. It takes that long for his nervous system & vision to develop enough to see & produce a smile in response. Smiling is a baby`s first social skill- he`s picking up on how relationships' work- as well as a signal of emotional growth.

3-Rolling over (2 or 3 months): but flipping from back to front often takes until around 5 months because it requires core coordination & muscular strength.

4-Grabbing (3 or 4 months): being able to grab things means he can engage more in play whether by himself or é others. Baby begins to gauge where things are in space & they can plan an action, such as grabbing a pacifier.

5-Hugging (5 months): the baby will quickly learn to hug Mom, Dad & other people he/she`s comfortable around, as well as her stuffed gorilla, the cat & anything else he/she adores.

6-Sitting up (8 months): once the baby has enough balance, arm strength & head, neck & lower body control, he well be able to sit up & take in a whole new world. At this point, his improving eyesight will allow him to see objects outside his direct line of vision & he will try to pull himself up to get a better look. At first, he will not be able to sit up for long on her own & may need to put his hand for balance, to motivate him to sit well, dangle or set his favorite toy in front of him, then slowly move it from side to side to encourage

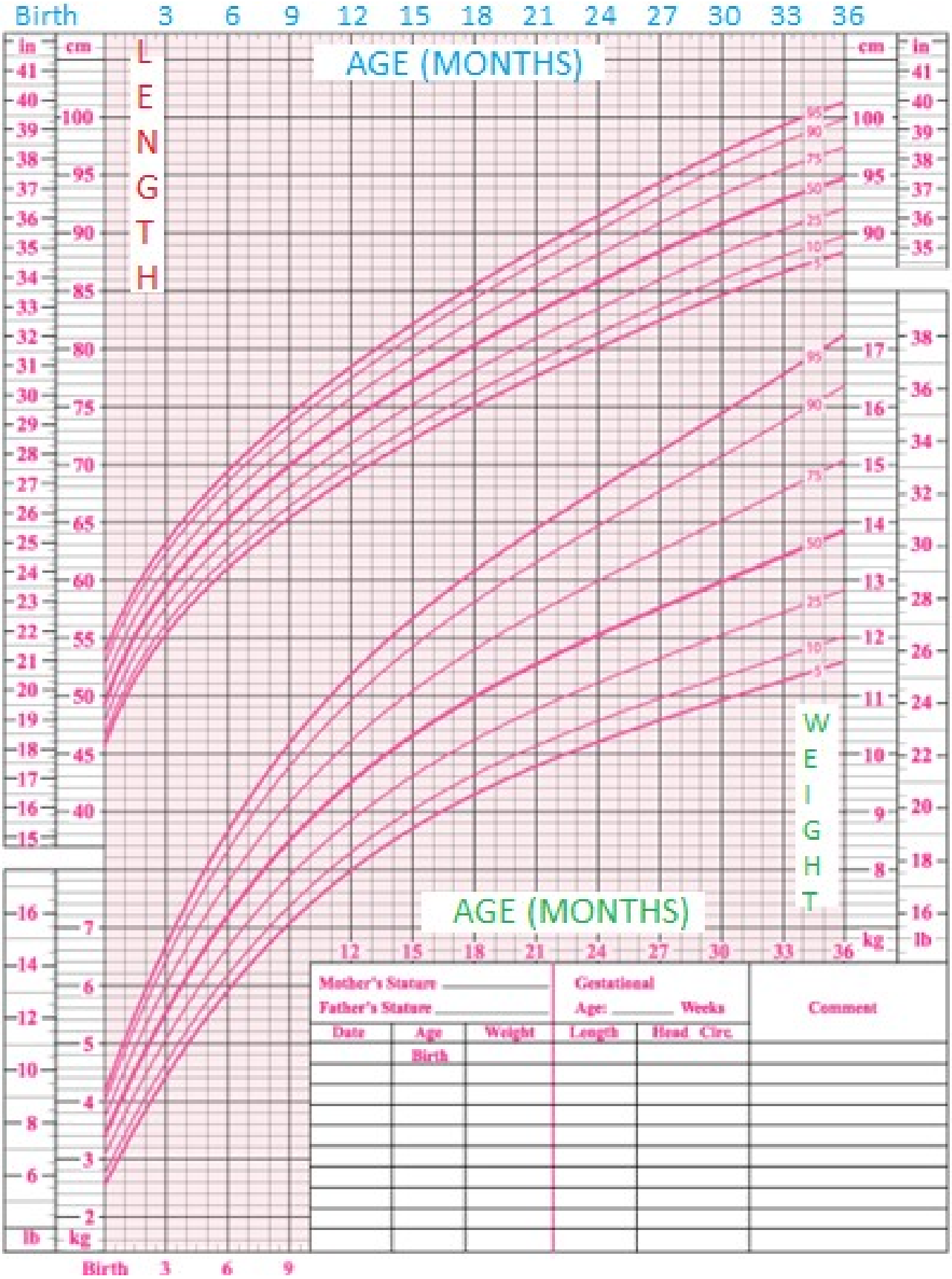
him to reach for the toy & rely solely on his torso & legs for balance, he well be sitting éout help in no time !

7-Crawling (6 to 10 months): after sitting up, he will start to test his arms if they can support him or not, so he will start the typical hands & knees crawl.

8-Pulling up (8 months): at around 8 months, her torso & leg muscles will be strong enough for her to stand up on her own. At first she will look for things to pull up on, the side of the crib, the arm of the sofa, or mother leg.

9 - Walking (10 to 18 months): first step represent a huge developmental leap, walking requires muscle strength, coordination, balance & a certain level of emotional maturity.

THE PERCENTILE CHART FOR WEIGHT & HEIGHT



12 REASONS BABIES CRY

1.Hunger: baby signs of hunger will help the mother to start baby`s feeding before the crying stage, some signs include; fussing, smacking of lips, or rooting.

2. Dirty dia-per: instruction to the mother to keep him always dry.

3. Needs sleep: may be over tired, mother have to carry him & speak to him é soft voice.

4. Wants to be he-ld: babies need a lot of cuddling. They like to see their parents faces 7 hear their voices & listen to their heart beats & they can even detect their unique smell. Crying can be their way of asking to be held close. Mother may wonder if she will spoil her baby by holding him so much is a common is a common question raised by parents?, but during the first few months of life that is not possible.

5. Tummy troubles: gas, colic & more can leads to a lot of crying, putting the baby on his back, holding his feet & moving his legs in a gentle bicycling motion may be helpful when done by mother. Other possible causes of babies tummy troubles including ; reflux, milk allergy, lacto-se intolerance, constipation & intestinal blockage.

6. Needs to burp: babies swallow air when they breast feed or suck from a bottle & the air is not released, so he may need to be burped.

7. Too cold or too hot: as a role, they are comfortable wearing one more layer than the mother need to be comfortable.

8. Something small: some babies are extra-sensitive to things like scratchy clothing tags or fabric & they can be very picky, may be the light is too bright or the TV is annoying, he may be need soft music instead, or may be the pacifier tastes gross & need washing, or may be the tag or outfit is itchy.

9. Teething: can be painful as each new tooth pushes through tend-er young gums, some babies suffer more than others, but all are likely to be fussy & tearful at some point along the way. The first tooth breaks through between 4 & 7 months. Applica- tion of local massage to the erupting teeth 3-4 times daily using medicated analgesic & anti-inflammatory cream will help to sooth the effect of teething.

10. Wants less stimulation: too much light or noise around, crying can baby`s way of saying, “I have had enough”.

11. Wants more stimulation: he opposite to the above baby will cry when he thinks he`s alone, or if mother put him on the floor é his toys while she work on the compu- ter, he fusses, while he`s happiest when the mother pop him in a baby carrier while she wash dishes, do laundry, another house works. He`s also especially peaceful in stores & other public places because he`s so interest- ed in & curious about the surr-ounding world.

12. Not feeling well: less active, refuse feeding, feverish. The cry of a sick baby tends to be distinct from one caused by hunger or frustration. Such cry, ẃ awaken baby fr-om sleep, may be the resultant of otitis media, meningitis or intestinal obstruction ??

6 SERIOUS BABY SYMPTOMS

1. Blue lips or tongue: cyanosed baby not getting enough O_2 ẃ may be cardiac or pulmonary in origin, clinical assessment, X- ray chest & heart, ECHO & ECG.

2. Strained breathing: difficult breathing (dyspnea), fast breathing (tachypnea), flari ng of nose, intercostal & subcostal recession are signs of respiratory distress. Do X-ray chest & heart, ABG, CBC, …..

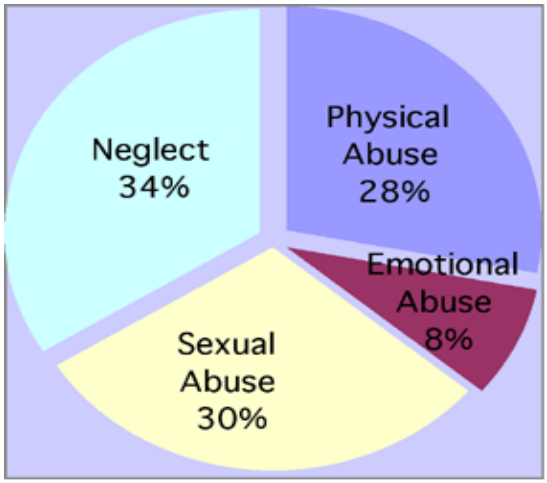
3. Fever: rectal temp. > 38 ^{o}C, is indicative of infection, may be viral, bacterial, fungal or parasitic, clinical assessment may detect the site of infection, upper/lower RTI, tonsillitis, otitis media, prodromal stage of one of the common viral infections as measles, rubella, chicken pox. Confirmation by specific tests, CBC, CRP, cultures & other investigations may needed as CSF & blood film…

4. Yellowing of the skin: if baby is getting yellower, after birth, he may need full investigations to R/O pathological jaundice. conjugated or unconjugated hyperbilIrubinaemia, serum bilirubin (total & direct, Hb, Hct, reticulocytic count, Rh grouping, coomb`s test, LFT, hepatitis screening tests (antigens & antibodies).

5. Dehydration: baby lethargic é dry tongue & mouth, sunken eyes, depressed fontanels & not making wet diapers (may associated é vo-miting, diarrhea, insufficient fluid intake, or neglected baby), such baby need evaluation for the degree of dehydration, electrolytes in blood for determination of type of dehydration, may need ABGs for presence or absence of metabolic acidosis, stool analysis. Management includes rehydration using either the oral route or in severe cases through IVF, at first to replace the deficit within 2-4 hours, then giving the maintenance according to the body weight & specific Rx according to the cause.

6-Vomiting: may be associated é diarrhea, gastroenteritis, URTI, tonsillitis, food allergy, UTI, gastrointestinal anomaly. Bright green or coffee grounds vomit denotes intestinal obstruction or bleeding. Careful abdominal examination for; intestinal sounds, abdominal tenderness, distension & signs of dehydration. Investigations: severe vomiting may associated é electrolyte imbalance, metabolic alkalosis. Investigations may include X-ray abdomen (erect & supine), electrolytes, ABGs, coagulopathy studies, urine & stool analysis.

CHILD ABUSE



Child abuse represent about 2% of attendance to casualty. Neglect seen in about 34 %. Sexual abuse occur in 30% of children < 5 yrs & it is more in girls. Physical abuse represent 28% of children. While emotional abuse seen in about 8% of cases.

Neglect: the failure of a parent, guardian, or other caregiver to provide for a child's basic needs. This can also include failure to protect them from a known risk of harm or danger. Neglected child presented ē; frequently absent from school, begs or steals food or money, lacks needed medical or dental care, immunizations, glasses…, is consistently dirty & has severe body odor, lacks sufficient clothing for the weather,

abuses alcohol or drugs, states that there is no one at home to provide care.

Physical Abuse

The non- accidental physical injury of a child. The child may presented ē unexplained burns, bites, bruises, broken bones, or black eyes, or has fading bruises or other marks noticeable after an absence from school. The child seems frightened of the parents & protests or cries when it is time to go home. Shrinks at the approach of adults. Reports injury by a parent or another adult caregiver.

Sexual Abuse

Anything done ē a child for the sexual gratification of an adult or older child. The child may presented ē difficulty walking or sitting, or suddenly refuses to change for gym or to participate in physical activity. He/She may reports nightmares or bed-wetting. The child may presented ē a sudden change in appetite, or demonstrates bizarre, sophisticated, or unusual sexual knowledge or behavior. Girl may becomes pregnant or contracts a STD.

Emotional Abuse

A pattern of behavior that impairs a child's emotional development or sense of self-worth. Shows extremes in behavior, delayed in physical or emotional development, has attempted suicide, Lack of attachment to the parent.

Diagnosis of child abuse

- Type of family; young parents, single, divorce, separation, limited mother abilities, addiction, criminal records.
- Characteristics of abused child; delayed presentation, inadequate explanation, frequent attendance, children never lie about sexual abuse, characteristic features & attitude of the child. Evidence of neglect & poor hygiene. Failure to thrive. Body injuries ẃ are multiple, of different ages, regular & similar. Child may present ē abdominal pain, UTI, puritis

ani, anal fissure, constipation or vaginal discharge or leeding. May presented ē changes in behavior or school performance.

Management of child abuse

#Never accuse parent or mentioning that child abused.

Admit for further investigations: coagulopathy profile, X ray bone.

#Photograph on admission.

#Examination by expert paediatrician or forensic medicine doctor: if sexual abuse suspected, forensic samples may taken, swabs from vagina, anus, throat, urine analysis.

#Inform local authority & social service.

Team approach: paediatrician, paediatric surgeon, social worker.

Foster care may required.

HARMFUL TRADITIONAL PRACTICES

Female genital mutilation

Harmful traditional practices are many. In this chapter only those ē reproductive health relevance are considered. These are Female genital mutilation (FGM), early marriage & abduction. FGM is practiced all over the world but more in Africa & Asia. In Africa alone >100 million women in 26 countries are subjected to genital mutilation. It is believed that FGM was practiced in the Ancient Egypt. History also shows the existence of FGM in the Pre-Islamic Arabia, ancient Rome & Tsarist Russia. In England, in the 19th & 20th century, FGM was practiced for psychological disorders. While practiced mainly in Muslim countries, FGM did not originate ē the rise of Islam. Both Christians & Muslims in African countries have their daughters mutilated. It is a misbelief that female mutilation is an Islamic practice. Mutilation: "to cut off or damage an important part of body" FGM - involved removal of parts or the whole of external genitalia of female.

Types

1- Clitoridectomy: it has two forms:-

•Dissection & removal of foreskin of clitoris.

•Removal of the whole of clitoris. Is the type is is more practiced in Egypt.

2- Total excision of clitoris, partial or total removal of the labia minora ēout closure of the vulva. The most common type in Africa done in more than 20 countries.

3- Excision + Stitching (Infibulation): Clitoris, labia minora, & inner walls of labia major are removed. The two labia are joined to seal except for urine & menses. Defibulation is done at marriage, during labor, or if there is absolute need for pelvic examination. Infibulation is done in Djibouti, Eastern Red Sea, Ethiopia & Somalia.

Condition under ẃ FGM is practiced: it is usually done by untrained person & under unhygienic condition ē unsterile instruments.

Immediate consequence includes:

Hemorrhage & Shock, Pain, Infection & Septicemia, Tetanus, Retention of urine, Injury to surrounding tissues, Delayed healing.

Long Term effects include:

Gynecological Problems. Ugly scar formation. Labial fusion. Narrowed vaginal opening. PID. Infertility. Dysmenorrhea. Hematocolpos (blood in uterine cavity). UTI. ascending infection. Renal failure. Sexual problem as; dyspareunia, vaginal laceration. Problems in childbirth & neonate. & Fistula formation.

Early Marriage

It is deep-rooted, widespread among many African countries, Christians & Muslims. It is parent -centered marriage between two families. It has devastating effect on child, family & community. Include; Promissory Marriage & Child marriage (usually the girl is under

the age of 10 years).

Consequence of early marriage

Sexual abuse, vaginal & perinea tear, Early pregnancy - child bearing & unwanted pregnancy, Maternal morbidity (fistula) & mortality (hemorrhage, obstructed labor etc.). The social Impact of early marriage includes; Denied education & own choices, Illiterate mother more often raise illiterate child, Urban migration, Many children (early pregnancy). Psychological trauma from the 1st sexual experience.

POISONING

Poisoning is when a person is exposed to a substance that can damage their health or endanger their life. Most cases of poisoning happen at home and children under five have the highest risk of accidental poisoning.

Signs and symptoms of poisoning

The will depend on the type of poison and the amount taken in, but general things to look out for include:

- vomiting
- stomach pains
- confusion
- drowsiness and fainting fits

If a child suddenly develops these symptoms, they may have been poisoned, particularly if they're drowsy and confused.

Management

General measures

- Stomach wash: pt lies on left side, stomach wash tube, give 50 ml water or saline then put the top of tube down to get rid of content, repeat 20 -25 times, leave in stoomach activated charcoal as Neocarbortina or Ultracarbon tablet 250 mg, 1 gm/Kg, swallowed or dissolved in water, maximum dose 10 gm, at home use cup of milk +2 egg white + 2 tsp starch +2 tsp flower.
- Diuretic: Lasix tab. 40 mg, amp. 20 mg, 1 mg/ Kg to induce diuresis.
- Antihistaminic: in case of drug overdose.
- Laxative: osmotic laxative, Mg SO4 sachet 5 grams, 250 mg/Kg.

ASPIRIN POISONING

Dangers of aspirin poisoning are convulsions, metabolic acidosis, tachypnea, fever.

Investigations: ⅄ABGs. ⅄Salicylate level in blood if available.

Management: •Stomach wash or induction of vomiting during the first 24 hrs of ingestion. •IVFs: normal saline 20 ml/Kg over 1 hr, then maintenance. •Diuretic: Lasix 20 mg amp. 1 mg/Kg IV. • After pt. pass urine add to running fluid Kcl 20% 1 ml/Kg to alkaline urine & induce forced alkaline diuresis. •Metabolic acidosis: $NaHco_3$, 1 ml/Kg.

ORGANOPHOSPHROS COMPOUNDS

Clinical picture: the danger of organophosphros are tremors, convulsions,coma, sweating , diarrhea, vomiting, meiosis.

Management: ☆Stomach wash or induction of vomiting during the first 24 hrs of ingestion. ☆ Atropine 1 mg/1 ml amp. 0.01 mg/Kg IM, can be repeated after 15 min.

LEAD POISONING

Effect of lead

Inhibits both hem & globulin synthesis. Interfere é the breakdown of RNA by inhibiting the enzyme pyrimidine 5 nucleotidase?, accumulation of denatured RNA in RBCs giving rise to basophilic stippling. Lead poisoning cause hypochromic/haemolytic anemia é bone marrow ring sideroblast & ⇧ of the free erythrocyte protoporphyrin & blood lead level.

Causes

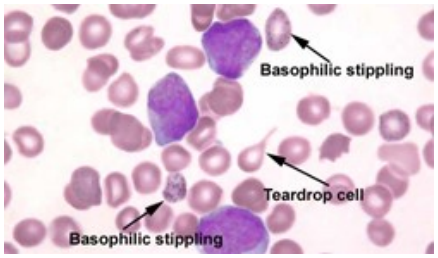
Lead is everywhere in the environment due to industrialization, environmental pollution through; leaded gasoline & the car exhaust ẃ contaminate air, agricultures, water surfaces, animals & even clothes through dust house, or lead taken by inhalation or ingestion or drinking of contaminated water through lead melting factories or polluted air

or through using lead pipes in houses, also through traditional practices seen especially in Bedouin communities, Africa & Islamic countries, as using alkohl as cosmetic for eyes, putting it on the umbilical cord of babies, or as cosmetic for eyes of lactating women as lead was found to be excreted in breast milk or through toys containing lead, or kitchen utensils made from or its component contain lead as é ceramic glazes, or from herbal medicine, pesticides, wall paints or wall paper containing lead through Pica.

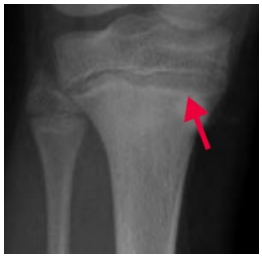
Clinical picture

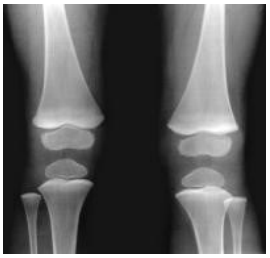
Hypochromic/haemolytic anaemia, reduction in IQ & attention span, poor school performance, behavioral problems (e.g. Hyperactivity), impaired growth & hearing loss, lead encephalopathy, seizures, coma & even death. Abdominal or joint pain.

Investigations



Blood film





Lead lines on metaphysis of long bones on X ray

- **CBC:** Hypochromic anaemia.
- **Basophilic stippling.**
- **Blood lead level;** normal level is <10µg/dl.

⅄⇧ of lead level >10 µg/dl may cause im-paired cognitive development in children. ⅄⇧ of lead level >45 µg/dl cause gastointestinal symptoms.

⅄⇧ of lead level >70 µg/dl carries high risk of acute CNS symptoms

⅄⇧ of lead level >100 µg/dl may be life-threatening.

- **Erythrocyte protoporphyrin:** >200
- **X ray bones:** may show transverse line in tubular growing bones.

Management

Current British paediatric opinion generally accept that a conc. of 37ug/100 ml or above is evidence of excessive exposure. At least 2 tests are required before initiation of Rx including:- ①An indication of the internal accumulation of lead.

②An indication of adverse metabolic effects.

Blood lead 37-49 ug/100 ml & normal-moderate ⇧ of EPP

Adequate dietary intake particularly calcium, iron & zinc must be assured. Follow-up every 3 months for at least one year until blood lead steadily ⇩ & stabilises at or near the normal range.

Blood lead 37-49 ug/100 ml & EPP markedly elevated (>500 ug/dl)

Do Ca-EDTA mobilization test: by giving a single IM injection of CaEDTA, if >1ug of lead for each mg of CaEDTA administered excreted in 24 hrs collection of urine, this provides evidence that there is excessive lead in the body & that the test is +ve. This test should not used in pt é symptoms of plumbism. In such case of +ve CaEDTA mobilization test, give pt D-Penicillamine 20 mg/kg/day for 2 doses, PO, for 2 days.

Blood Lead 50-69ug/100 ml & EPP <250 ug/dl & +ve Ca-EDTA mobilization test give Ca-EDTA 50 mg/kg/day, 3-5 day course of deep IM injections. Do urine analysis & serum creatinine every 48 hrs during the course of Rx to ensure no side effects. **N.B.:** Ca-EDTA is a non-metabolisable drug, excreted exclusively by kidneys.

Blood Lead 70 ug/100ml or more & EPP >250 ug/dl

Give Ca-EDTA 50 mg/kg/day, 3-5 day course of deep IM inj + BAL 500 mg/M^2/day course, deeply IM. BAL is contraindicated in pt é acute hepatocellular injury or males é G6PDD. BAL often causes vomiting & pt. receiving it should placed on parenteral fluids or clear liquids orally. In general the chelating agents should not be given for more than a week at

a time, but several courses may be given at short intervals over a period of 1-2 months.

Acute lead encephalopathy

In emergencies in ẃ lead tests are not immediately available & where acute lead encephalopathy is a diagnostic possibility, the find-ing of strongly +ve qualitative urinary coproporphyrin (CP-U test), of many stippled erythrocytes in bone marrow & of glycosuria & hypophosphatemia indicate presumptive plumbism. In such condition the following management should be instituted:-

- BAL 500 mg/M^2/day 4 hourly by deep IM for 2-3 days only. After the first dose of BAL use BAL & Ca-EDTA at a separate IM sites. The recommended dose of Ca-EDTA is 1500 mg/M^2/day, 4 hourly for 2 days then 1000 mg/M^2/day, 8 hourly for 3 days.
- Control of seizures - Diazepam to start é then long-term anticonvulsant therapy é Phenobarbital.
- Reducing cerebral oedema: careful calculation of the minimal fluid requirement. Mannitol may be needed to establish adequate urine flow.
- For young children é Pica, they should be followed at least until school age in order to prevent recurrences & to assess the degree of residual brain damage; ẃ may not become evident until several yrs after an acute episode.

IRON POISONING

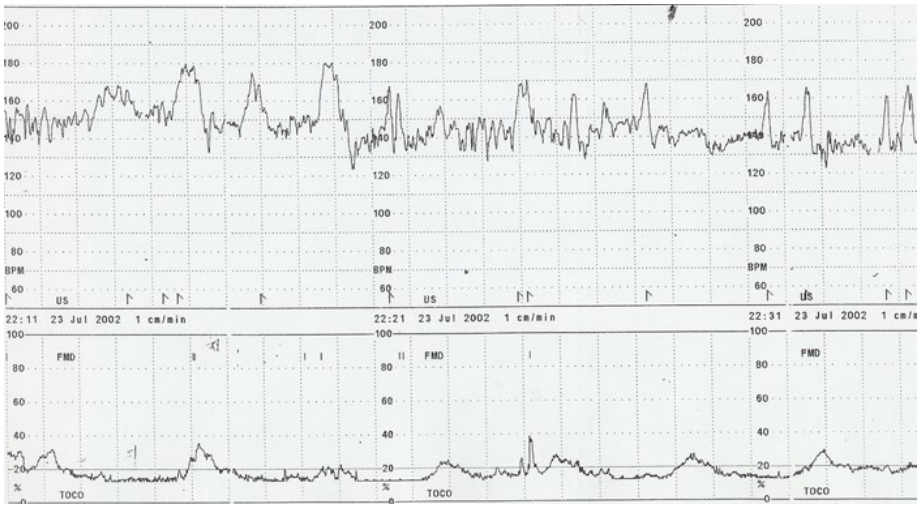
Diagnosis: history taking, &Check for serum level of iron.

Management

▲ Stomach wash or induction of vomiting during the first 12 hours of ingestion, leave in stomach in the last wash 5 gm Desferroxamine. ▲DesferroxamIne (desferal) IV, 5 mg/Kg /hr maximum 80 mg/Kg/day or 20 mg/Kg/6 hrs IM, for 24 hr.

Multiple Choice Questions

1- A mother presents to her local hospital at 39 wks complaining of mild lower abdominal pain & reduced fetal movements since the night before. Her pulse & BP are normal & the fundal height is consistent ē the gestational age. She is not yet in labor. A cardiotocograph is performed & shows the following:



Which of the following statements is true?

A) This is non-reassuring CTG & doctors should perform a fetal scalp pH meas- urement.

B) The baseline is 160 bpm ē prolonged decelerations.

C) The baseline is 130 bpm ē decelerations.

D) This is a reassuring CTG & the mother should be allowed home after a few hours.

E) The CTG shows very little beat-to-beat variability & should be repeated in 30 min.

2- Which of the following statements is true of the fetal circulation?

A) 100% of the cardiac output goes to the lungs via the pulmonary artery.

B) The arterial duct (ductus arteriosus) helps send oxygenated blood to the brain.

C) Blood shunts across the duct ‘left to right’ (from aorta to the pulmonary artery).

D) The foramen ovale typically closes by 36 weeks' gestation.

E) The umbilical vein carries well-oxygenated blood.

3- Which one of the following factors does NOT ⇩ the perinatal mortality rate?

A) Improved antenatal care.

B) Lower social class.

C) Improved maternal health.

D) Prenatal care and counselling.

E) Maternal education.

4- Which one of the following does NOT increase the rate of perinatal loss?

A Female sex of fetus.

B Low birth weight.

C Multiple pregnancy.

D Maternal age <20 years.

E Maternal age >40 years.

5- Which one of the following pregnancies carries the highest risk of adverse outcome?

A) Singleton pregnancy.

B) Dichorionic diamniotic (DCDA) twins.

C) Monochorionic diamniotic (MCDA) twins.

D) Monochorionic monoamniotic (MCMA) twins.

E) Disappearing twin.

6- Which one of the following is true of congenital abnormalities seen in the newborn?

A) Deformations are typically due to an insult in the first trimester whereas malformations occur due to insults in the second or third trimester.

B) Congenital anomaly registers allow researchers to identify clusters of abnormality &

may allow earlier identification of a teratogenic source.

C) Deformities of skull bones are extremely rare & need urgent neurosurgical referral.

D) An abnormal uterus or breech position does not cause limb abnormalities to occur.

E) Talipes equinovarus responds best to early corrective surgery.

7- With regard to maternal medical disorders, ẃ of the following statements are FALSE?

A) Mothers ē epilepsy should have their medication reviewed prior to pregnancy.

B) Congenital myotonic dystrophy is likely to be more severe in each generation.

C) Infants of mothers with insulin-dependent diabetes should be put on an insulin infusion soon after birth to avoid neonatal ketoacidosis.

D) Pre-eclampsia is more likely to occur if mother has a new partner.

E) Maternal systemic lupus erythematosis is unlikely to have any effects on the fetus.

8- Which of the following diseases in the newborn is NOT a manifestation of maternal IgG antibody crossing the placenta?

A) Myaesthenia gravis.

B) Malar rash (systemic lupus erythrematosis).

C) HIV infection.

D) Thyrotoxicosis.

E) Hemolytic disease of the newborn (rhesus).

9- Which of the following statements is FALSE regarding normal birth?

A) As the uterus contracts, blood flow through the uterine artery is interrupted.

B) Fetal blood oxygen levels can be 30–45 mmHg without causing brain damage.

C) The fetal brain can use alternative fuels such as ketones if the supply of glucose is

interrupted.

D) The umbilical arterial pH is the same or higher than the venous pH.

E) Most babies are cyanosed at birth

10- Which of the following statements is true of the Apgar score?

A) APGAR stands for Airway, Pulse, Gasp, Alertness, Reflexes

B) Umbilical cord gases are not as reliable as Apgar scores when it comes to assessing the severity of asphyxia.

C) The Apgar score has excellent consistency when administered by different operators.

D) An Apgar score of <7 at 1 minute means the baby requires immediate advanced resuscitation.

E) An Apgar score of <5 at 10 minutes is associated ē a poor outcome.

11- A baby is born at 29 wks following a large maternal APHge. You arrive in the delivery room just as the baby is being transferred to the resuscitation platform.

Which of the following is a CORRECT action that you should undertake?

A) Dry the baby & wrap in several warm towels, then assess the baby`s condition.

B) Don't worry about universal precautions, as resuscitation cannot be delayed.

C) Ask the obstetrician to administer intramuscular betamethasone to the mother.

D) Administer IM naloxone (200 µg) to the baby's thigh & massage vigorously.

E) Place the baby in a sterile plastic bag up to the neck, cover the head ē a hat & place the baby under a radiant warmer.

12- A full-term baby is born by forceps delivery following a prolonged 2nd stage. At the newborn examination a few hours later, the baby is noted to have an asymmetrical cry. Which one of the following actions should NOT be undertaken?

A) Arrange an urgent MRI to check for intracerebral bleeding or unilateral infarction.

B) Let the baby attempt breast feeding.

C) Inform the obstetrician so that a risk management review can take place.

D) Prescribe artificial tears to be administered to the baby's eye.

E) Consider diagnosis of asymmetric crying facies sy. if there is no evidence of bruising.

13- Which one of the following is NOT a reported benefit of breast feeding?

A) Promotes mother–infant bonding.

B) Decreased intelligence quotient (IQ).

C) Decreased risk of asthma and eczema in infants predisposed to these conditions.

D) Reduced likelihood and severity of cows' milk protein allergy.

E) Decreased incidence of infant obesity.

14- Which of the following statements is FALSE with regard to breastfeeding versus formula feeding?

A) Lactation helps the mother lose weight acquired in pregnancy.

B) Breastfeeding is more convenient and cheaper.

C) Lactational amenorrhoea is a form of contraception.

D) Oxytocin release during breastfeeding contracts the uterus and helps its involution.

E) Breastfeeding increases the risk of premenopausal breast cancer.

15- Which one of the following statements is FALSE ē regard to the composition of human milk versus cows' milk?

A) Human milk contains more lactoalbumin.

B) Human milk contains more vitamins A, C & E.

C) Human milk contains more calcium & phosphate.

D) Human milk contains more lactoferrin.

E) Human milk contains more IgA.

16- Which one of the following practices may INCREASE the risk of complications arising when using parenteral nutrition?

A Prophylactic antifungals.

B Prophylactic antibiotics.

C Prophylactic insulin infusion.

D Administration via peripherally inserted central catheters (PICC).

E Biochemical surveillance in infants receiving parenteral nutrition.

17- A 35-wk PT female infant is now 4 hrs old and self-ventilating in air. She was born via emergency LSCS for breech position after preterm labor. Her mother was given 1.2 g IV benzylpenicillin 3 hrs before delivery. Apgars were 91 & 95 respectively, birth weight was 2600 g. In the last hour the baby has had 4 episodes where she stopped breathing for >20 sec., her HR dropping to <80 bpm & her O_2 saturations to <70%. She requires firm stimulation by the nursing staff to resolve these episodes. Between episodes her oxygen saturations on pulse oximetry are 96% & her examination is normal. She is not currently on any medications.

Which one of the following options is the most appropriate next step in the management of her apnoea?

A) Place her in 30% oxygen.

B) Commence caffeine for apnoea of prematurity.

C) Perform a sepsis work-up and commence IV antibiotics.

D) Start mask continuous positive airway pressure (CPAP).

E) Intubate & ventilate the baby.

18- A 31-wk female infant weighing 1300 g is born via spontaneous vaginal delivery after her mother unexpectedly presented 6 hrs earlier ē preterm labor. Her mother rec-eived IV antibiotics & IM betamethasone prior to delivery. She is born in good condition (Apgars 61 & 95) & requires 30 s of positive pressure ventilation during resuscitation. She develops early respiratory distress (RR is 60/min, slight intercostal recession, nasal flaring & audible expiratory grunt when handled). She is admitted to the intensive care nursery & is maintaining oxygen saturations of 91% in 28% cot oxygen.

Which one of the following options is the most appropriate next step in the manage-ment of this baby?

A) Insert an umbilical artery catheter to ensure her PaCo2 is < 65 mmHg.

B) Commence 10% dextrose.

C) Get a chest radiograph.

D) Take blood cultures & commence IV antibiotics.

E) Commence nasal CPAP.

19- You are asked to review a newborn term male infant because his mother is a hepatitis B carrier. The baby is now 2 hrs of age & is attempting a breast feed ē his mother. The mother's serology is as follows:-

HBSAg positive.

HBeAg negative.

anti-HBeAb positive.

Which of the following options is the most appropriate next step in the management of this baby?

A) No management required because of low risk of transmission.

B) Cease the breastfeed immediately and allow only formula feeding.

C) Arrange for Hep B immunoglobulin to be administered within the next 24 hours.

D) Arrange Hep B vaccine to be administered within the next 72 hours.

E) Arrange for both HB immunoglobulin & HB vaccine to be administered within the next 10 hours.

20- Regarding preterm delivery at the borderline of viability, which of the following statements is true?

A) Below 26 wks gestation, boys have a better prognosis than girls.

B) For babies born at 24 wks' gestation there is a 10–15% chance of survival.

C) Ethically, when it comes to making decisions about whether to offer intensive care to an EPT baby, the views of the parents supersede all other considerations.

D) There is no evidence to support the use of adrenaline in babies born in very poor condition at <25 wks.

E) Babies born at 22 wks are classified as 'stillbirths'.

21- A baby is about to be delivered at 26 wks gestation. The mother received 2 doses of betamethasone 24 hrs ago.

Which one of the following is good practice in the first 'golden hour' after delivery?

A) All babies at this gestation should be intubated electively at birth so that they can receive surfactant.

B) Baby should be born into a plastic bag or wrap so that he can maintain body temp.

C) A saturation probe should be attached to the baby's left hand & oxygen given until the sat.O_2 is >96%.

D) The baby should be dried, weighed & then placed under a radiant heater.

E) Once the baby is transferred to NICU he/she should be washed, weighed & placed in an incubator humidified to >80% humidity.

22- Which of the following is true of small for gestational age babies?

A) SGA is usually defined as birth weight < 3rd centile.

B) Asymmetrical IUGR usually reflects a fetal disease process.

C) Small mothers tend to have small babies.

D) IUGR babies have more respiratory distress at birth.

E) The long-term prognosis for IUGR babies is better than for macrosomic babies.

23- Which of these statements is true?

A) Very low birth weight is defined as BW <1000 g.

B) Extremely low birth weight is defined as BW <750 g.

C) Preterm IUGR babies should be fed 150–165 mL/kg of high-calorie milk from birth to prevent hypoglycemia.

D) Babies ē congenital CMV infection are often SGA.

E) Down's syndrome babies have a higher than expected birth weight but tend not to grow as tall as their peers during childhood.

24- With regard to prophylactic surfactant (given within 15 min of birth) administered to intubated infants born <30 wks gestation, ẃ of the following statements is true?

A) There is a ⇩ incidence of PDA.

B) There is an ⇧ incidence of bronchopulmonary dysplasia or death.

C) There is a ⇩ incidence of pneumothorax.

D) There is an ⇧ incidence of pulmonary interstitial emphysema.

E) There is ⇧ incidence of mortality.

25- A 32-wk male infant is born via spontaneous vaginal delivery after preterm labor. He is born in good condition ē Apgars 91& 95 & does not require any resuscitation but does have RR of 60/m, slight intercostal recession, nasal flaring & an audible expiratory grunt when handled. He is admitted to the intensive care nursery, has a blood culture collected & is commenced on antibiotics &10% dextrose at 60 ml/kg/D. He is maintaining oxygen saturations of 92% in 25% cot oxygen.

Which one of the following would be the MAJOR BENEFIT of commencing him on nasal CPAP versus leaving him in cot oxygen?

A) Decreased chance that he will need to be intubated and ventilated.

B) Decreased chance of him having a spontaneous pneumothorax.

C) Decreased total length of stay in hospital.

D) Decreased chance of developing retinopathy of prematurity.

E) Decreased chance of him developing chronic neonatal lung disease.

26- A 32-wk male infant weighing 1750 g has just been born via spontaneous vaginal delivery after his mother unexpectedly presented 3 hrs earlier ē preterm labor. She was given 1.2 g of benzylpenicillin IV & 11.4 mg of betamethasone IM 2 hrs before delivery. He was born in good condition (Apgars 91 & 95) & does not require any resuscitation but does have a RR 60 bpm, slight intercostal recession, nasal flaring & audible expirato- ry grunt when handled. He is admitted to the intensive care nursery & is maintaining oxygen saturations of 92% in 25% cot oxygen.

Which one of the following options is the most important next step in management?

A) Insert an umbilical artery catheter to ensure his $PaCo_2$ is < 65 mmHg.

B) Commence 10% dextrose at 60 mL/kg per day.

C) Get a CXR to exclude infection or pneumothorax.

D) Take blood cultures & commence IV antibiotics.

E) Commence nasal CPAP.

27- You are called to the delivery suite for a 39 wk baby about to be born through thick meconium. You arrive in time to prepare a 3.5 Fr neonatal ETT, check your laryngo-scope is working, ensure you have suction & a device to aspirate meconium with. The baby is then born & the nurse immediately starts suctioning the baby's mouth while the obstetrician is clamping & cutting the cord. The baby starts crying & the midwife brings over a screaming, vigorous baby who is now 30 seconds old.

Which one of the following is your next step in management?

A) Intubate the baby and suction the ETT in case there is meconium below the cords.

B) Dry the baby down, remove the wet wraps & give suction only if required.

C) Give immediate deep oral suction.

D) Visualize cords ē laryngoscope, only intubate if you see meconium below the cords.

E) Intubate the baby only if the 1 min Apgar is less than 6.

28- A 31-wk preterm female infant is now 24 hrs old & self-ventilating in air. She was born via emergency LSCS for maternal pre-eclampsia (her mother was given 2 doses of betamethasone 24 hrs before delivery). In the last hr the baby has had 2 episodes where she stops breathing for >20 sec, her HR drops to <80 bpm & her oxygen saturati- ons to <70%. She requires firm stimulation by the nursing staff to resolve these episo-des.

Between episodes she looks well, her oxygen saturations on pulse oximetry are 96% & her examination is normal. Routine FBC collected this morning is normal & her CRP is <1 mg/mL. She is currently on IV amoxicillin & gentamicin. Which one of the following is the most appropriate next step in the management of her apnoea?

A) Place her in 30% oxygen.

B) Commence caffeine.

C) Perform a sepsis work-up and add IV vancomycin.

D) Start continuous positive airway pressure (CPAP).

E) Intubate & ventilate the baby.

29- A 28-wk gestation female infant, birth weight 800 g, was intubated at birth for resuscitation & given endotracheal surfactant. She is now 2 hrs old & is being ventil-ated using the assist/control (SIPPV) mode ē volume guarantee. The back-up rate is set at 35 breaths/min, the inspiratory time is 0.35 s, the volume is set at 3.2 mL (4 mL/kg) & the maximum PIP is set at 25 cmH2O. The current PIP is 16 cmH2O, the PEEP is 5 cmH2O & the baby is breathing at a rate of 55 breath/min. If the infant has a spont-aneous pneumothorax, which one of the following is most likely to occur?

A) Inspiratory time will increase.

B) Peak inspiratory pressure will increase.

C) Tidal volume will increase.

D) PEEP will increase.

E) Ventilator rate will increase.

30- A 29-wk gestation infant, intubated since birth for moderate RDS, has been stead-ily improving & is now 12 hrs old. The baby was given a loading dose of caffeine at 8 hrs of

age. The baby is vigorous, well perfused & passing urine. BP is within normal limits. The level of ventilatory support has been progressively weaned & is currently as follows:

Mode: synchronized intermittent positive pressure ventilation (assist control);

FiO_2: 0.21;

PIP: 12 cmH2O;

PEEP: 5 cmH2O;

RR (ventilator): 30 breaths/min;

IT: 0.35 s;

Baby's spontaneous breathing rate: 50 breaths/min

The most recent arterial blood gas is as follows:

pH	7.43	[7.34–7.43];
PaCo2	31 mmHg	[31–42];
Pao2	68 mmHg	[45–60];
Bicarbonate	23 mmol/L	[20–26];
Base excess	1.6	[-5.0–5.0]

Which one of the following would be the most appropriate next step?

A) Extubate to nasal CPAP.

B) Decrease PEEP by 1 cmH2O.

C) Reduce IT to 0.3 s.

D) Increase PIP to 13 cmH2O.

E) Decrease set ventilator rate to 20 breaths/min.

31- A 26-wk gestation male infant, intubated & ventilated since delivery, is 6 hrs old & has received 2 doses of surfactant. His CXR has a ground glass appearance ē air bronch-

ograms. The baby RR is 65/min. The ventilation settings are as follows:

Mode: assist control (SIPPV);

FiO_2: 0.3

PIP 18 cmH2O

PEEP 6 cmH2O

RR: 40 breaths/min;

IT 0.35 s.

The arterial blood gas is as follows:

pH: 7.19 [7.34–7.43]

Paco2: 62 mmHg [31–42]

Pao2: 56 mmHg [45–60]

Bicarbonate: 19 mmol/L [20–26];

Base excess: -4.8 [-5.0–5.0]

Which one of the following alterations to the ventilator settings would be the most appropriate to improve ventilation

A) Convert to SIMV.

B) Increase inspiratory time to 0.6 s.

C) Increase PEEP to 7 cmH2O.

D) Increase PIP to 20 cmH2O.

E) Increase rate to 60 breaths/min.

32- Which of the following statements is true of the cardiovascular system?

A) Blood pressure = stroke volume X heart rate.

B) Left-to-right shunting across the duct is normal in the fetus

C) The fetal heart does not start pumping blood until 18–20 wks, because the placenta performs this function.

D) Contractility of the heart is independent of preload and afterload.

E) The ductus arteriosus can be kept open by the use of ibuprofen or indomethacin, but these have some side effects.

33- A baby is found to have a loud (3/6) 'rasping' systolic murmur at the lower left sternal edge on day 2 of life. The pulses are normal & the baby does not look cyan-osed. Which one of the following is a LIKELY diagnosis?

A) ASD.

B) Pulmonary atresia ē VSD.

C) VSD.

D) PDA.

E) Normal heart ē physiological murmur.

34- A baby is born at 37 wks gestation ē antenatally diagnosed transposition of the great arteries. The mother has poorly controlled insulin-dependent diabetes. The baby is dried & wrapped & transferred to the neonatal unit where a preductal SAo2 is measured at 65%. Which of the following actions should not be performed within the next 30 min?

A) Transfer the baby immediately to the nearest specialist cardiac center.

B) Check a blood glucose and temperature.

C) Monitor pre & postductal oxygen saturations

D) Start a prostaglandin E1 infusion.

E) Perform an arterial blood gas measurement.

35- A 28-wk gestation preterm baby weighing 800 g develops a new murmur on day 5 of life. This is loudest at the left sternal edge but sounds continuous at the left clavicle Which one of the following is an appropriate further action?

A) Measure the systolic & diastolic blood pressure.

B) Measure pre & postductal saturations as there is likely to be a significant drop between them due to ductal shunting.

C) Prescribe aspirin 75 mg once daily for 3 days

D) Perform an ECG.

E) Stop all enteral feeding.

36- A 31-wk female infant is born via elective CS for IUGR & reversed end diastolic flow; birth weight is 1010 g. Which one of the following management options will be most effective in decreasing the risk of her developing NEC?

A) Oral antibiotics.

B) Prebiotics.

C) Feeding with breast milk only.

D) Probiotics.

E) Oral immunoglobulin.

37- You are asked to review a 5-wk old ex 31-wk male infant who is has started to have increasing frequency of vomiting after feeds. The vomit appears to be undigested milk only & is not green in color. He is currently having alternate suck & nasogastric feeds ē formula & appears to vomit after both types of feeds. On examination he appears well & his abdomen is soft & non-tender. He has a normal urine output & is passing normal stools. ẃ one of the following is the most appropriate examination to identify a cause of

the vomit?

A) pH probe.

B) Upper GI contrast study.

C) Lower GI contrast study.

D) Plain abdominal radiograph.

E) Ultrasonography of the abdomen.

38- A term baby is referred from a peripheral hospital with failure to pass meconium in the first 3 days of life. A lower gastrograffin enema is performed & results in the baby successfully passing meconium. No other abnormalities are detected on routine examination. The baby is sent back to the peripheral hospital, breast feeding & stooling normally. Which one of the following is the MOST IMPORTANT investigation to arrange in the long-term follow-up of this baby?

A) Neonatal screening test.

B) Immunoreactive trypsinogen.

C) Rectal biopsy.

D) Sweat test.

E) Thyroid function test

39- Adequate renal function & normal amniotic fluid production is NOT required in the fetus for which one of the following functions?

A) Providing space for growth and movement.

B) Excretion of waste products.

C) Protection against infection.

D) Maintenance of temperature.

E) Development of the respiratory system.

40- A 38-wk male infant born via elective LSCS is admitted to the neonatal intensive care ē respiratory distress. He is started on CPAP & 60 mL/kg per day of 10% dextrose. At 12

hrs of age he is on 8 cm of CPAP in 25% oxygen ē oxygen saturations in the high 90s, he has a RR of 50 & appears comfortable, his HR is 154 & his BP is 55/35 mmHg. He passed urine shortly after birth but none since. A capillary blood gas is performed. Results:

pH: 7.37 [7.35–7.5];

Pco2: 38 mmHg [27–40];

Sodium: 127 mmol/L [130–149];

Chloride: 98 mmol/L [105–115];

Potassium: 5.5 mmol/L; [4.0–7.0];

Glucose: 3.7 mmol/L [3.6–7.7].

Which one of the following is the MOST appropriate next step in management?

A) Insert a urinary catheter.

B) Continue current management.

C) Restrict total fluid intake.

D) Add sodium chloride to maintenance fluids.

E) Increase the dextrose concentration.

41- A 16-day-old ex 32 wk male infant, birth weight 1750 g, is self-ventilating in air in the special care nursery. He has been on full nasogastric feeds of breast milk only & is not on any other medications. On routine observations he is noted to have an axillary temp of 37.6 °C, his pulse rate is 180 bpm & his RR is 65/min. He appears to be well wrapped in the incubator. As part of a septic work-up for infection, the junior registrar asks for a bag

urine to be collected. The microscopy results of the bag urine are as follows: WBCs count: 30.000/mL; RBCs count: 20.000/mL; Epithelial cells: 30.000/mL. Comment: bacteria seen.

Which one of the following is the MOST appropriate next step in management?

A) Collect a supra-pubic aspirate.

B) Start oral antibiotics.

C) Unwrap the baby.

D) Repeat the bag urine.

E) Perform a renal ultrasound scan.

42- Which one of the following is the most important cause of jaundice presenting in the first 24 h of life to EXCLUDE ?

A) Prematurity.

B) Hemolysis

C) Breast feeding.

D) Physiological jaundice.

E) Early-onset sepsis.

43- Which one of the following is the major contributor to the development of physiological jaundice?

A) Breastfeeding.

B) Decreased hepatic bilirubin excretion.

C) Immature hepatic enzymes.

D) Enterohepatic circulation.

E) Increased bilirubin production.

44- Which one of the following is the most common cause of late-onset hemorrhagic disease (vitamin K deficiency bleeding)?

A Neonatal septicemia.

B Complete oral (instead of IM) vit K administration.

C Formula feeding.

D Cystic fibrosis.

E Hypoxic ischemic encephalopathy.

45- Which one of the following is most likely to lead to a decrease in the need for early top-up blood transfusion for extremely preterm babys?

A) Iron supplementation.

B) Erythropoietin.

C) Vitamin E supplementation.

D) Decreased frequency of blood sampling.

E) Folate supplementation.

46- Which one of the following is LEAST likely to present as hydrops fetalis?

A) α-Thalassemia.

B) β-Thalassemia.

C) ABO incompatibility.

D) Rhesus isoimmunisation.

E) Twin-to-twin transfusion.

47- A female infant is born ē an omphalocele that was diagnosed antenatally. The infant was delivered vaginally at 38 wks' gestation. Birth weight was 4500 g. The omphalocele was covered ē a clear polythene film after birth & the baby has been admitted to the

nursery for IV 10% dextrose while awaiting surgical consult. After insertion of the peripheral IV cannula a venous plasma glucose is collected & is 20mg/L. The baby's core temp is measured at 36.0 °C.

Which one of the following is the most likely cause for the plasma glucose reading?

A) Defect in fatty acid oxidation.

B) Abnormal insulin secretion.

C) Cold stress.

D) Delayed serum cortisol response.

E) Impaired glycogenesis.

48- A baby is found to be irritable & jittery on the postnatal ward. The midwife collects a blood glucose level & the result is 28 mg/L. The mother is adamant that she wants to breast feed the baby.

Which one of the following is the most appropriate next step?

A) IV 10% dextrose at 60 mL/kg/day.

B) Attempt to breast feed.

C) NGT expressed breast milk.

D) NGT formula feed.

E) IM glucagon.

49- Which one of the following conditions is LEAST likely to benefit from inclusion in a neonatal screening program?

A) Hyperthyroidism.

B) Phenylketonuria.

C) Cystic fibrosis.

D) Galactosaemia.

E) Medium-chain acyl coenzyme A dehydrogenase deficiency.

50- You are asked to counsel a woman who is planning to have another baby after her first baby was born ē spina bifida. Which one of the following pre-conception management options is most likely to reduce the risk in any subsequent pregnancy?

A) Sodium valproate.

B) Thiamine.

C) Folic acid.

D) Ultrasound screening.

E) Vitamin B12.

51- Which one of the following is the most important risk factor in the aetiology of intraventricular haemorrhage?

A) Coagulation disorder.

B) Intermittent positive-pressure ventilation.

C) Pneumothorax.

D) Extreme prematurity.

E) Hypoxic–ischaemic encephalopathy.

52- Which one of the following is the most useful tool in the prediction of neurodevelopmental outcome for a baby ē moderate encephalopathy secondary to hypoxic–ischaemic encephalopathy?

A) Doppler assessment of the cerebral arteries.

B) Bedside amplitude integrated EEG.

C) MRI.

D) Neurological examination.

E) Cranial ultrasound.

53- You are asked to urgently review a term 3.5 kg infant who is now 25 hrs old. The nurse on the postnatal ward is concerned that the infant may be having seizures. The infant appeared to be startled & then had jerking of the upper limbs that lasted for 10 seconds. He was born vaginally via a Ventouse extraction after a prolonged second stage of labour. His Apgar scores were 6 at 1 minute and 9 at 10 minutes. He required suction & 30 seconds of positive pressure before regular respirations were established. He quickly became vigorous & required no further management. He did not require admission to the nursery. A blood gas analysis from the umbilical cord at the time of delivery revealed a pH of 7.25 [7.35–7.45], Pco2 of 50 mmHg [31–42]. On arrival you find a sleeping, well-looking infant who starts to have rhythmic jerking of both arms & legs when you begin examining him. When you touch the arms of the baby, the jerking appears to stop. You fail to find any abnormality on examination & arrange collection of the following investigations:

Sodium: 134 mmol/L [135–145];

Potassium: 4.3 mmol/L [3.7–6.0];

Calcium: 1.8 mmol/L [1.90–2.70];

Glucose: 2.9 mmol/L [3.0–8.0];

Hb: 154 g/L [145–225];

WBCs count: 22.0 X 109/L [5.0–25.0];

Platelets: 267 X 109/L [150–400]

Which one of the following is the most likely cause of this infant's seizures?

A) Hypoglycemia.

B) Subdural hemorrhage.

C) Hypocalcaemia.

D) Hypoxic–ischemic encephalopathy.

E) Myoclonic jerks.

54- Which one of the following is the most common cause of sensorineural deafness in newborn infants?

A) Prematurity. B) Autosomal dominant inheritance.

C) Autosomal recessive inheritance. D) Congenital rubella.

E) Hyperbilirubinemia.

55- Which one of the following is true of thermoregulation in the newborn?

A) The normal newborn baby has core temp. 36–36.5 °C.

B) Preterm babies have little subcutaneous fat, but can autoregulate their temp. well using thermogenesis from brown fat.

C) Babies should be placed in a thermoneutral environment to promote energy conservation & growth.

D) Shivering is an effective treatment for hypoxic–ischaemic brain injury.

E) Babies placed naked against their mother's chest will lose heat by radiation & conduction.

56- A 20 yr old primigravid woman presents at 27 wks gestation to a small peripheral hospital that delivers only 100 babies/year. The hospital does not have a neonatal nursery. She has ruptured her membranes & on speculum exam there is pooling of fluid &

her cervix is long & closed. She is having some irregular painful contractions (1 in 10). Which one of the following is the most appropriate next step in her management?

A) Reassure her that she is not in labor.

B) Await delivery & then arrange retrieval of the baby.

C) Transfer baby in utero to facility ē neonatal intensive care.

D) Administer IM steroids & IV antibiotics to the mother.

E) Commence tocolytic medications.

57- You are about to transfer ventilated 34 wk male infant ē RDS by aeroplane to NICU. He has been given one dose of surfactant & is currently ventilated on SIMV ē a back-up rate of 40, pressures 22/6, & Fio_2 25%. His oxygen saturations on pulse oximetry is 95%. His $Paco_2$ prior to departure is 42 mmHg. The aeroplane has a pressurized cabin (equivalent to 5000 feet or 1500 m). As the aircraft climbs to 20 000 feet (6000 m) you notice his oxygen saturations slowly drop into the mid 80s.

Which one of the following is the most appropriate next step?

A) ⇧ inspired O_2 conc.

B) ⇧ PIP.

C) ⇧ PEEP.

D) ⇧ RR.

E) Ask the pilot to fly at 5000 feet (1500 m).

58- All but which one of the following factors increases the risk of adverse neurodevelopmental outcome in extremely preterm infants?

A) Grade IV intraventricular hemorrhage.

B) Inadequate antenatal steroids.

C) Female sex.

D) Neonatal meningitis.

E) Chronic neonatal lung disease.

59- Which one of the following patients is at highest risk of adverse neurodevelopmental outcome?

A) 23-wk male infant.

B) 24-wk female infant.

C) 26-wk female infant ē grade 1 intraventricular hemorrhage.

D) 27-wk male & female dichorionic diamniotic twins.

E) 28-wk infant ē intrauterine growth retardation.

60- Which one of the following factors is most likely to lead to inadequate mother infant bonding?

A) Failure to attend antenatal classes.

B) Elective caesarean section.

C) Parental decision to formula feed.

D) Postnatal depression.

E) Antenatal counselling after fetal abnormality diagnosed.

61- All but which one of the following strategies will help promote parent–infant attachment for extremely preterm infants?

A) Restrictive visiting policy in the neonatal intensive care.

B) Allowing parents to touch baby.

C) Kangaroo cuddling.

D) Syringe-feeding baby.

E) Changing nappies.

62- Which one of the following actions is NOT appropriate when caring for parents immediately after a neonatal death?

A) Providing a single room.

B) Social work involvement.

C) Discussing autopsy.

D) Allowing the mother to spend time with her dead baby.

E) Discussion about planning for future pregnancies.

63- You are called to urgently review a 4-hour-old 25-wk infant who is ventilated for RDS. Prior to this episode she was in 30% oxygen on SIPPV + VG & needed pressures of 20/6 to obtain a tidal volume of 4 mL/kg. She is now in 100% oxygen & the ventila-tor is alarming due to the maximum pressure limit being reached (currently set at 25 cmH2O). On examination you notice that the right chest is full & there is ⇩ air entry on the right compared to the left. The trachea is deviated to the left. His pulse rate is now 80 bpm & oxygen saturations are in the 70s.

Which one of the following is the appropriate next step in management?

A) Arrange an urgent chest radiograph.

B) Increase the maximum pressure limit on the ventilator to 30 cmH2O.

C) Perform emergency thoracocentesis on the right.

D) Re-intubate the baby.

E) Increase the PEEP to 8 cmH2O.

64- You receive an urgent call to the operating theatre & arrive to find your resident resuscitating a 34 wk male infant born by emergency CS for a large APHge. The baby is now 5 minutes old & the resident informs you that 1 minute ago they administered adrenaline via an ETT (size 3.5, inserted 9 cm). The baby is receiving appropriate chest compressions from the neonatal nurse & ventilation in 100% oxygen from the resident , but there are still no signs of life & a heart rate has never been recorded. You confirm that the ETT is correctly placed.

Which one of the following is your next step in management?

A) Cease resuscitation attempts.

B) Seek parental advice about continuing resuscitation.

C) Call your consultant.

D) Insert an emergency umbilical venous catheter to give 2nd dose IV adrenaline

E) Give a second dose of adrenaline (epinephrine) via the ETT.

65- You come on for night duty & are asked to review the radiograph of a 24 wk infant weighing 730 g who has just had a 3.5 Fr umbilical catheter inserted to 10 cm. On the X ray you note that the catheter is just left of the midline & the tip is at the level of T12.

Which one of the following is the most appropriate next step in management?

A) Advise the nursing staff that the catheter is in a good position.

B) Advance the catheter so that it sits above the vertebral level of T10.

C) Withdraw the catheter so that it sits at the vertebral level of L4.

D) Remove the catheter.

E) Withdraw the catheter so that it sits at the vertebral level of L1.

66-A 2-wk-old male newborn is brought to the office by his mother for routine well-child examination. The baby is breast-fed & his mother follows a vegan diet. The mother says she plans to breast-feed until the baby reaches 6 months of age & then she will introduce him to solid foods that conform to her vegan diet. On the basis of this information, the baby is at greatest risk for deficiency of ẃ of the following vitamins?

(A) B1

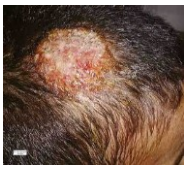
(B) B6

(C)B12

(D) Vit C

(E) Vit k

67-



A 4-yr-old boy is brought to the clinic by his mother because he has an area of sores on his scalp that she noticed while cutting his hair. Weight is 19.1 kg. Vital signs are within normal limits. Physical examination shows a grouping of pustules in the right parietal region. The area surrounding this grouping is boggy & very tender, & annular hair loss is noted in that region. The cervical lymph nodes are diffusely tender & enlarged. No other abnormalities are noted. Which of the following is the most likely diagnosis?

(A) Alopecia areata.

(B) Atopic dermatitis.

(C) Herpes simplex virus.

(D) Kerion.

(E) Mastocytosis.

68- A 5-day-old neonate is brought to the emergency department by ambulance 20 min. after he had sudden onset of irritability, diaphoresis & profound dyspnea. The baby has not had fever or other symptoms of systemic illness. He was delivered vaginally at term ēout complications. Temp. is 37.0°C, HR is 200/min & RR is 50/min. On physical examination, auscultation of the chest shows a grade 2/6 systolic ejection murmur that is heard best at the LUSB & radiates to the left interscapular area. Palpation of the abdomen shows enlargement of the liver. Femoral pulses are absent bilaterally & the lower extremities appear somewhat cyanotic compared ē the upper extremities. No other abnormalities are noted.

Which of the following is the most likely diagnosis?

(A) Coarctation of the aorta.

(B) Diaphragmatic hernia.

(C) Group B streptococcal sepsis.

(D) Patent ductus arteriosus.

(E) Tetralogy of Fallot Content.

69- A 16-yr-old boy is brought to the office because he has had decreasing performance in school over the past 6 months. During this time, the pt has become more irritable & irresponsible, has changed his group of friends & has had decreasing personal hygiene. He was previously a high-achieving student, but his grades have slipped to the extent that he is failing several courses.

Which of the following disorders is the most likely cause of this patient's symptoms?

(A) Bipolar.

(B) Conduct.

(C) Major depressive.

(D) Persistent depressive (dysthymic).

(E) Substance use (substance abuse).

70- A 12-yr-old boy is brought to the office because he has had recurrent episodes of epistaxis during the past 2 months. The patient has no history of bruising or bleeding. While he is in the examination room, an episode of epistaxis occurs. On physical examination, anterior bleeding is visible within the Kiesselbach plexus.

Which of the following is the most appropriate initial step in management?

(A) Administration of clotting factors.

(B) Anterior nasal packing.

(C) Application of direct pressure.

(D) Cautery of the bleeding area.

(E) Referral of the patient to a pediatric otolaryngologist.

71- A 4-yr-old boy is brought to the office by his parents because he has had intermittent non bloody diarrhea & constipation along ē abdominal pain & bloating during the past 3 wks. When the patient's symptoms first began, his stools were watery, but they are currently greasy & foul smelling. He has not had fever or vomiting. The parents say that other children at the patient's day care have had similar symptoms. Vital signs are within normal limits. Physical examination shows vague tenderness to palpation of the abdomen ēout rebound or guarding.

Which of the following studies is the most appropriate next step?

(A) Air contrast enema.

(B) Colonoscopy.

(C) Enzyme immunoassay of stool.

(D) Esophagogastroduodenoscopy.

(E) Fecal leukocyte test.

72- A 6-yr-old boy is brought to the office because he has had pain in his right knee for the past 2 days. The patient walks ē a mild limp because of the pain. He has no history of trauma to the knee. He had cold symptoms 1 wk ago but is otherwise healthy. Vital signs are within normal limits. Physical examination shows full range of motion of the right knee ē limited internal rotation of the right hip. On laboratory studies, ESR is slightly elevated & WBCs count is within normal limits. Anteroposterior & frog-leg X-ray studies of the right hip & knee show no abnormalities.

Which of the following is the most likely cause of the pain in this patient's knee?

(A) Developmental dysplasia of the hip.

(B) Legg-Calvé-Perthes disease.

(C) Septic joint.

(D) Slipped capital femoral epiphysis.

(E) Transient synovitis

73- A 3-yr-old boy is brought to the office by his parents as a new patient for well-child examination. Height is in the 3rd percentile & weight is in the 1st percentile. During the interview, the parents say that the patient has been treated multiple times since infa-ncy because of sinus infections & pneumonia. They also note that his stools are gene-rally loose, greasy & mucousy. During physical examination, the patient coughs frequ-ently. No other abnormalities are noted.

Which of the following studies is most effective to determine the diagnosis?

(A) Bronchoscopy.

(B) CT scan of the sinuses.

(C) Culture of aspirate from the trachea.

(D) Measurement of serum immunoglobulin levels.

(E) Sweat chloride test.

74- A 2-yr-old girl is brought to the office by her parents after blood was noticed in her urine. The parents say the pt has had intermittent abdominal pain during the past 2 months but has been otherwise well. On physical examination, the abdomen is sligh-tly distended & a mass is palpated in the right upper quadrant. Results of urinalysis are positive for blood & protein. Which of the following is the most likely diagnosis?

(A) Cystic nephroma.

(B) Cystitis.

(C) Mesoblastic nephroma.

(D) Neuroblastoma.

(E) Wilms tumor.

75- A 4-yr-old girl is brought to the office by her parents for well-child examination. The parents say the pt has been healthy, but they have noticed that she tires more quickly than her peers when they are playing. She is in the 10th percentile for height & the 40th percentile for weight. BP is 132/82 mmHg in the left arm & 128/80 mmHg in the right arm. Repeat measurements are in the same range.

Which of the following is the most appropriate next step?

(A) Counsel the parents regarding diet and exercise.

(B) Measure blood pressure in the lower extremities.

(C) Order urinalysis.

(D) Recheck blood pressure in two weeks.

(E) Refer the patient to a nephrologist.

76- A male neonate is delivered vaginally at term, & neonatal examination & testing confirms the diagnosis of sickle cell disease. On the basis of this finding, the most appropriate initial step is administration of ẃ of the following?

(A) Erythromycin.

(B) Hydroxyurea.

(C) Oxygen.

(D) Penicillin.

(E) Pneumococcal & meningococcal vaccines.

77- A 5-day-old neonate is brought to the office by his mother for initial examination after uncomplicated pregnancy, delivery & nursery stay. The mother has hypoth-yroidism that is well controlled ē levothyroxine & she is worried that the baby might have congenital hypothyroidism. If this condition is present in this baby, which of the following findings is most likely to be noted on physical examination?

(A) Bradycardia.

(B) Enlarged fontanelle.

(C) Hypothermia.

(D) Umbilical hernia.

(E) No abnormalities.

78- A 12-yr-old girl is brought to the office for well-child examination. During the past 2 months, the pt has had fatigue, occasional vague abdominal pain & unintentional weight loss of 3.18 kg. Results of lipid panel include the following:

total cholesterol 215 mg/dL (N< 170 mg/dL)

LDL 134 mg/dL (N< 110 mg/dL)

HDL 41 mg/dL (N< 35 mg/dL)

Triglycerides 470 mg/dL (N< 37-140 mg/dL)

Which of the following laboratory studies is the most appropriate next step?

(A) Complete blood cell count with differential.

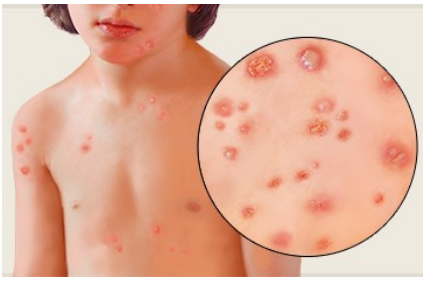
(B) Liver function studies.

(C) Measurement of serum amylase level.

(D) Measurement of serum glucose level.

(E) Thyroid function studies.

79-



This nine-year-old boy has been unwell for the past few days & developed a rash affecting his trunk & limbs yesterday. He has no underlying medical problems & is feeling better today. His mother has used a cream recommended by the pharmacist. Which is the SINGLE MOST appropriate management option?

A) No additional treatment.

B) Oral acyclovir.

C) Topical acyclovir.

D) Topical fusidic acid.

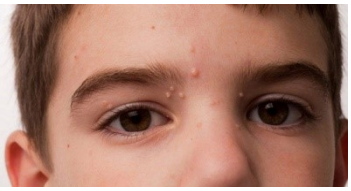
E) Topical mupirocin.

80- A 3-yr-old girl has recurrent UTI & the pediatrician has recommended trimethoprim prophylaxis at a dose of 2 mg/kg at night. She weighs 12.5 kg & trimethoprim suspension is available as 50 mg/5 mls.

What volume of suspension (in mls) should the child's mother give her every evening?

A) 1 ml. B) 2 ml. C) 2.5 ml. D) 3 ml. D) 5 ml.

81-



The parents of a 9 year-old child are concerned about the multiple skin lesions that have spread on his face over the past 12 wks.

Which is the SINGLE MOST appropriate management option?

A) No treatment necessary.

B) Oral Flucloxacillin.

C) Topical acyclovir.

D) Topical fusidic acid.

E) Topical hydrocortisone 1%

82- Specialist referral is MOST APPROPRIATE for which TWO of the following children?

A) A four-week-old boy whose mother reports he does not smile.

B) A four-month-old girl who cannot grasp an object when it is placed in her hand.

C) A four-month-old boy who cannot sit unsupported.

D) A two-year-old girl who cannot hop.

E) A three-year-old boy who cannot combine words into a simple sentence.

83- A 7-year-old girl has a fever associated ē a sore throat & loss of appetite. She has small red ulcers in her mouth & itchy spots on the palms of hands & soles of her feet. Which is the SINGLE MOST likely virus causing her symptoms?

A) Coxsackie A.

B) Herpes simplex.

C) Measles.

D) Parvovirus B19.

E) Varicella-zoster.

84- Rheumatic fever

a. Is most common in the third decade.

b. Is more common in areas of social deprivation.

c. Causes erosive arthritis.

d. Relapse rate may be reduced by prophylactic antibiotics.

e. Is more common following streptococcal pharyngitis than streptococcal cellulitis.

85- Travellers' diarrhea

a. The single most common causative organism is entero-invasive E. coli.

b. Has an incubation period of at least 48 hours.

c. May be due to Aeromonas ssp.

d. May be due to Cryptosporidium.

e. Should be treated ē antibiotics.

86- Chicken pox

a. Has an incubation period of 3-5 days.

b. Rash is preceded by Koplik's spots in the mouth.

c. Fever settles when the rash appears.

d. Should be treated by topical acyclovir.

e. May follow from close contact ē a case of shingles.

87- The paralysis of polio virus infection

a. Is upper motor neurone type.

b. Is asymmetrical.

c. Usually affects the lower limbs more severely than the upper limbs.

d. Is more severe if strenuous physical exercise occurred in the incubation period.

e. May be caused by polio vaccination.

88- - Diphtheria

a. "Bull neck" is diagnostic.

b. It is caused by Gram positive bacilli.

c. Toxin absorption is greatest in pharyngeal disease.

d. Palatal paralysis is a recognized complication.

e. Complete heart block is a manifestation of toxin-induced myocarditis.

89- A 2½ yr old boy appears ē intermittent loose stools for the past 1 month. Stools typically occur during the day & not overnight. The boy is otherwise healthy. The growth & development are normal. The most likely diagnosis is:

A) Chronic enteritis.

B) Rotavirus enteritis.

C) Salmonella enteritis.

D) Food allergy.

E) Toddler's diarrhea.

90- A 5-yr-old boy appears ē an abrupt onset of ⇧ urinary frequency for the last 3 days. He has been voiding every 10-15 minutes during the day. The mother denies history of fever, abdominal pain, dysuria, nocturia, or daytime incontinence. The urinalysis result is normal. The next step in management is:

A) Trimethoprim-sulfamethoxazole.

B) Restriction of fluid intake.

C) Renal ultrasonography.

D) A 24-hour urine sample for calcium.

E) Reassurance.

9I- A 15 yr old boy appears ē swelling, tenderness & increased prominence of the right tibial tubercle. He participated in high school sports. The following therapy is not beneficial in this condition:

A) NSAIDs.

B) Rest.

C) Restriction of activities.

D) Knee immobilizer.

E) Isometric exercise program.

92- The preferred intravenous therapy for an infection in animal bite wounds is:

A) Cefotaxime.

B) Ceftriaxone.

C) Ciprofloxacin.

D) Penicillin.

E) Ampicillin & Sulbactum.

93- Tonsillectomy is indicated in all of the following conditions except:

A) A pt who experienced 6 tonsillar infections that were treated ē antibiotics in the preceding year.

B) A pt who experienced 5 tonsillar infections that were treated ē antibiotics in each of the preceding 2 years.

C) A pt who experienced 3 tonsillar infections that were treated ē antibiotics in each of the preceding 3 years.

D) A pt who experienced 4 tonsillar infections that were treated ē antibiotics in each of the preceding 3 years.

E) A pt who experienced 7 tonsillar infections that were treated ē antibiotics in the preceding year

94 - A palpable left parasternal impulse suggests ẃ abnormality?

A) Right ventricular hypertrophy.

B) Aortic stenosisc.

C) Aortic regurgitationd).

D) Left ventricular hypertrophy.

e) Right ventricular hypertrophy.

95- Which of these combinations of clinical features is most suggestive of mixed mitral valve disease ē a predominance of mitral regurgitation?

A) Displaced apex beat; soft first heart sound; pan-systolic murmur; short mid-diastolic

murmur.

B) Irregularly, irregular pulse; displaced apex beat; ejection systolic murmur; short mid-diastolic murmur.

C) Displaced apex beat; normal first heart sound; pan-systolic murmur; long mid-diastolic murmur.

D) Tapping apex beat; loud first heart sound; pan-systolic murmur; long mid-diastolic murmur.

96- A 14 yrs old girl on exposure to cold has pallor of extremities followed by pain & cyanosis. In later ages of life she is prone to develop?

A. SLE.

B. Scleroderma.

C Rheumatoid arthritis.

D. Histiocytosis.

97- Which is the most reliable way to assess for clubbing?

a) Inspect the nail-bed angle from above.

B) Assessing the fluctuancy of the nail-bed.

C) Schamroth's sign.

D) Inspect the nail-bed angle from the side.

98- Bronchial breathing is characterized by:

A) Inspiratory component louder & longer ē a gap between expiration &inspiration.

B) Expiratory component louder & longer ē a gap between inspiration & expiration.

C) Inspiratory component louder& longer ē a gap between inspiration & expiration.

D) Expiratory component louder & longer ē a gap between expiration &inspiration.

99- Which of the following is true about Cushing's Syndrome?

A) It is due to a deficiency of cortisol hormone.

B) Enlarged extremities are commonly seen.

C) Osteoporosis is not a feature.

D) A moon face & a buffalo hump are characteristic of the disease.

100- Which of the following is true about Addison`s Disease?

A) It is due to a deficiency of prolactin hormone.

B) Leads to generalized pigmentation.

C) It is a recognized cause of hypertension.

D) Diabetes is a complication.

MCQ ANSWERS

1 (D) Correct. This is a reassuring CTG as there is a normal baseline (140 bpm), good beat-to-beat variability (>5 beats), accelerations are present & there are no decelerations. Provided other observations are normal & the mother is well, she can be allowed to go home, ē advice to return if concerned. A 'kick chart' to record the number of movements each day may be useful. The Fetal scalp pH is not appropriate if the mother is not in labor.

2 (E) Correct. The fetus depends entirely on placenta to meet O2 needs. Unusually for a vein, the umbilical vein carries oxygenated blood from the placenta. Umbilical cord pH is usually higher in the vein than in the artery. The foramen ovale only closes after birth. In fact < 10% of the COP goes to the fetal lung.

Anatomy & Physiology of Fetal Circulation: the umbilical cord contain 2 umbilical arteries ẃ return non-oxygenated blood, fecal waste, CO2 to placenta. 1 umbilical vein ẃ brings oxygenated blood & nutrients to the fetus. Oxygenated blood flows from the placenta to the fetus via the **umbilical vein.** After reaching fetus the blood flows through **inferior vena cava,** then blood continues to travel to **ductus venosous** where small amount of blood routed to growing liver. Increased blood flow leads to large liver in newborns, blood continues to travel up the inferior vena cava, empties into the **right atrium**. Then passes to **left atrium** through **foramen ovale,** ẃ completely bypasses the non-functioning lungs, Blood continues journey to **left ventricle** where it is pumped into the **aorta.** Blood is circulated to the upper extremeities & then returns to the **right atrium.** From the right atrium, the blood goes to the **right ventricle** then to the **pulmonary arteries,** where a small amount goes to the maturing lungs. Rest of blood is shunted away from lungs by **ductous ateriosus** back to aorta. Blood travels back from aorta to the two **umbilical arteries** to the **placenta.** The placenta will resupply the blood with oxygen.

3 (B) Correct. Lower social class significantly increases the risk of perinatal mortality.

4 (A) Correct. Females have a slightly lower rate of perinatal mortality. All the other factors are known to ⇧ the perinatal mortality rate.

5 (D) Correct. Pregnancies that share the placenta & amnion are at highest risk of adverse outcome because of risk of twin-to-twin transfusion syndrome & cord entanglement.

6 (B) Correct. It is important to monitor congenital abnormalities to understand their aetiology. Preconception folic acid supplementation to prevent spina bifida shows the benefit of recognizing high-risk groups. Deformities occur to an already formed organ or structure. They therefore occur later, unlike malformations ẃ are due to abnormal tissue development & often occur in very early pregnancy when there is rapid organogenesis.

7 (C) Correct. Infants of diabetic mothers are already producing inappropriately high amounts of insulin in response to in utero exposure to maternal hyperglycemia. They need early, regular feeding or IV dextrose, not more insulin. It is important that mothers ē epilepsy are put on the least teratogenic antiepileptic that will maintain good control of their disorder *before* they become pregnant.

8 (C) Correct. HIV infection is due to transmission of HIV virus, not the antibody. However the presence of maternal anti-HIV IgG can lead to diagnostic difficulty. It is therefore necessary to confirm HIV infection in the newborn by the presence of viral DNA or RNA using a polymerase chain reaction (PCR) method. In Myaesthenia gravis maternal IgG autoantibodies can cross the placenta causing transient disease in the newborn. The same in SLE, Thyrotoxicosis & Rhesus.

9 (D) Correct. The umbilical artery caries used blood away from the fetus & so the pH can never be higher than in the vein. Fetal blood oxygen levels 30–45 mmHg is a normal level

for fetal oxygen concentration. FHb concentration is higher & the FHb dissociation curve is different from that of the adult, allowing the fetus to cope ē lower pO_2 than the adult. The fresh blood is delivered almost directly to the fetal brain. Most babies are cyanosed at birth & it can take up to 10 minutes before oxygen saturations are greater than 90%.

10 (E) Correct. This is one of several criteria for considering therapeutic hypothermia if a baby has HIE, as the outcome may otherwise be poor. Apgar score is named after Virginia Apgar, an American anesthetist. Paired umbilical cord gases are the gold standard for assessing the severity of a perinatal insult. Most babies ē an Apgar of <7 at 1 minute will recover quickly without extensive resuscitation, provided they have an open airway.

11 (E) Correct. This has been shown to be effective thermal care for a newborn preterm baby. Keep the bag/wrap in place until the baby is inside a warm, humidified incubator.

12 (A) Correct. This is likely to be a lower motor facial nerve injury (7th cranial nerve). This can be distinguished from an upper motor nerve injury (ẃ would require imaging) by the involvement of the upper part of the face, including the eyelids. Allow breast feeding as the nerves that control the tongue & gag reflex are not affected & oral feeding is safe.

13 (B) Correct. There is considerable debate as to whether babies who have been breast fed subsequently have a higher IQ than those given formula milk in the early months of life, but there is no doubt that breastfeeding does not decrease the IQ.

14 (E) Correct. Breast feeding confers some health advantages on the mother, as there appears to be some protection against ovarian & premenopausal breast cancer as well as osteoporosis.

15 (C) Correct. Cows' milk contains more calcium & phosphate than human milk, but their absorption is much lower. Human milk contains twice as much lactalbumin as cows' milk (and is immunologically different), but no lactoglobulin.

16 (C) Correct. Although some infants may develop hyperglycemia whilst receiving parenteral nutrition, some will not and prophylactic insulin infusions could lead to significant hypoglycemia in some infants.

17 (C) Correct. This baby is at risk of sepsis because of maternal PPROM & PTL. In spite of the intrapartum antibiotics the baby is now symptomatic. The other measures may be useful in the acute management of apnoea, but the priority should be to treat for possible infection.

18 (D) Correct. The most important management option is to treat her for possible early-onset infection. If she does have early-onset infection, a delay in commencing IV antibiotics may have very serious consequences. All the other options may also be reasonable but should be done in conjunction or after the commencement of IV antibiotics. The CXR is important, the priority should be to commence IV antibiotics.

19 (E) Correct. All infants born to hepatitis B carriers should be given HB immunoglobulin & HB immunization within 12 hours of birth.

20 (D) Correct. There is little evidence to support its use. If it is required then the prognosis for intact survival is poor. Survival to discharge is about 45% at 24 wks, if the baby is born alive. The views of the parents are important but not paramount. Doctors & health professionals must act in the 'best interests' of the baby.

21 (B) Correct. Plastic wraps (and a hat to cover the head) have been shown to improve thermoregulation, but the baby must also be placed under a radiant warmer. As regard intubation, it is often the unit protocol, but increasingly in some centers even the most preterm babies can be managed on nasal CPAP from birth. Only about 40–50% subsequently need intubation. This strategy has been shown to dramatically reduce the incidence of chronic lung disease in some centers.

22 (C) Correct. Babies reflect their mother's centile at birth & tend to graduated towards their mid parental centile during the 1st year of life. SGA is usually defined as birth weight <10th centile. Some of these babies will be normal and just genetically programmed to be small. IUGR refers to a fetus who has not grown as expected.

23 (D) Correct. SGA babies are 9 times more likely to have had a congenital infection. VLBW is defined as birth weight <1500 g (sometimes expressed as ≤1499 g). ELBW is defined as <1000 g.

24 (C) Correct. Many studies found that prophylactic intratracheal administration of natural surfactant to intubated infants <30 wks gestation decreased the risk of pneumothorax. There was no difference in the risk of PDA.

25 (A) Correct: This baby has early RDS. Early application of CPAP reduces subsequent use of IPPV & thus may be useful in preventing the adverse effects of this treatment. Whether or not CPAP causes pneumothorax is controversial, ē some studies finding an increased incidence in groups randomized to CPAP & other studies finding no difference.

26 (D) Correct. This baby is at significant risk of congenital sepsis & therefore anti-biotics should be commenced. Options B & C should be done as well but the most important task is the commencement of antibiotics. At this point he does not require monitoring of arterial blood gases so an umbilical artery catheter is not required, but this would need to be reassessed if the oxygen requirement significantly increases or his condition deteriorates. Some units would collect a capillary or venous gas to ensure that pH and Pco_2 are within acceptable limits. Many units would commence CPAP on this baby, but it would also be reasonable to leave him in cot oxygen.

27 (B) Correct. This is basic neonatal resuscitation. Intubation may become an issue if the baby develops severe respiratory distress.Deep suction or having a look ē a laryngoscope

cope is only likely to cause reflex bradycardia & apnoea in this baby, necessitating further resuscitation. Resuscitation measures should never be based on Apgar scores alone.

28 (B) Correct. This is the normal age in hours for apnoea of prematurity to present. Resuscitation measures should never be based on Apgar scores alone. This baby is at low risk of infection & is already on appropriate antibiotics, so the addition of vancomycin is unwarranted. CPAP is useful in apnoea of prematurity & may be necessary if the caffeine does not decrease her rate of apnoea. Intubation & ventilation would only be required if severe & frequent apnoea continue occur after commencing caffeine & the commencem ent of CPAP.

29 (B) Correct. If a pneumothorax occurs when on volume guarantee, the initial change that you might see is a sudden increase in the PIP as the ventilator tries to compensate for the decreased tidal volume. The other settings will be unchanged (unless someone physically changes them). The inspiratory time unchanged as this is fixed in this mode of ventilation. The tidal volume unchanged as the baby is on volume guarantee. The PEEP unchanged as this is fixed in this mode of ventilation. The ventilator rate is fixed but the baby may initiate more spontaneous breathing & the overall rate could possibly increase or may decrease back to the back-up rate as the baby tires.

30 (A) Correct. This baby is overventilated, as indicated by the low Paco2, & is stable enough to be extubated. Caffeine loading significantly ⇧ the chances of successful extub-ation. Options B, C & D will all ⇧ the tidal volume, further decreesing the Paco2.

31 (D) Correct. This baby has a respiratory acidosis & ventilation needs to be increased to remove more CO2. Changing to SIMV will ⇩ the number of supported breaths from 65 to 40, worsening the respiratory acidosis. Increasing the inspiratory time to 0.6 s will cause Co_2 retention & will worsen the respiratory acidosis. Increasing the PEEP will initially ⇩

the tidal volume (⇩ CO_2 clearance) but if it allows further recruittment of alveolar segments it may lead to an improvement in oxygenation & CO_2 clearance. Increasing the rate to 60 will have no impact at all as the baby is breathing at a rate of 65 in SIPPV mode of ventilation & is therefore already receiving 65 supported breaths per minute

32 (D) Correct. Unlike stroke volume, contractility is independent of preload & afterload & is an innate property of myofibrils. It can be influenced by inotropes (e.g. dobutamine). BP = blood flow X peripheral resistance while the COP = stroke volume X heart rate. The ductus arteriosus carries relatively more oxygenated blood from the pulmonary artery to the aorta in the fetus thereby bypassing the lungs. The fetal heart develops in very early fetal life (18 days) & begins pumping blood by day 21. The placenta does not 'pump' blood to the fetus. Ibuprofen or Indomethacin are used

to close a PDA. Keeping a duct open requires prostaglandin (E_1 or E_2).

33 (C) Correct. The high-velocity turbulence creates a "rasping noise" systolic murmur. An ASD usually is associated ē no murmur or a soft mitral flow murmur. A baby ē pulmonary atresia & VSD would be expected to be cyanosed. A PDA will tend to have a systolic & diastolic component to the murmur & the pulses will be 'tapping' or 'collapsing' due to the low diastolic BP. This loud systolic murmur grade 3/6 is not physiological (Physiological = continious musical murmur ẃ change ē changing position of the baby).

34 (A) Correct. The baby must first be commenced on a prostaglandin E1 infusion prior to transfer. The baby should ideally have been born in a tertiary center, in view of the antenatal diagnosis. This baby is at high risk of hypoglycemia because of the maternal insulin-dependent diabetes. TGA is 20 times more common in infants of diabetic mothers. Measurement of postductal saturation is useful, although in TGA the values are likely to be similar to the preductal (very occasionally they can be higher, because of the switched

anatomy). High-dose oxygen is likely to be useless and may promote duct closure. Start a prostaglandin E1 infusion is the most important measure, a moderate to high dose should be used to ensure the duct is open. Be aware this may cause secondary apnoea. Performing an ABG measurement is important as this baby may develop severe metabolic acidosis secondary to hypoxia. This should be measured & treated ē bicarbonate pending the creation of an atrial septostomy. The definitive treatment of cardiac surgery (switch operation) is then performed within the first few weeks of life.

35 (A) Correct. A wide pulse pressure (low diastolic BP) is almost diagnostic of a PDA in the presence of these symptoms. If there is shunting across the PDA it will be left to right & there will be no drop in postductal saturations (measured in the feet). Although aspirin (acetylsalicylic acid) is a cyclooxygenase inhibitor (like ibuprofen & indomethacin), aspirin is contraindicated in children <12yrs old because of the risk of Reye`s syndrome.

An ECG is unlikely to add any important information at this stage. An echocardiogram would be much more useful. Although a PDA is a risk factor for NEC it is not an absolute indication to stop enteral feeding. This is a controversial area: some clinicians would continue feeding if tolerated, others would reduce to minimal enteral feeding until the PDA is treated & closed. It is certainly not a time for increasing the milk load on the gut.

36 (D) Correct. The relative risk of developing NEC is 0.35 in infants treated ē probiotics compared to placebo. Oral antibiotics do decrease the risk of NEC but are not recommended because of the possibility of antimicrobial resistance. Prebiotics may be beneficial in reducing the risk of NEC but further studies are still required. The use of breast milk is also recommended but does not have as much effect in reducing the risk as probiotics. . Oral immunoglobulin appears to have no benefit in decreasing the risk of NEC.

37 (E) Correct. This baby appears to be developing pyloric stenosis & this can be confi-

rmed on abdominal US by finding the pylorus ≥3 mm thick. Almost all babies will have some gastroesophageal reflux & a pH probe is unlikely to add extra information. The vomit is not bile stained, so a malrotation is unlikely. This baby has no symptoms of lower GI obstruction. Lower GI contrast study is a useful investigation in cases of abdominal distension & failure to pass stool. A plain abdominal radiograph can be useful to establish if the nasogastric tube is correctly placed, but in this child the vomiting occurs after both suck & tube feeds.

38 (D) Correct. This baby has a meconium ileus; 80–90% of these babies will have CF. A sweat test performed after 4 wks of age is the gold standard for the diagnosis of CF. The CF can cause an elevated immunoreactive trypsinogen, but a sweat test performed after 4 wks of age is still the gold standard for the diagnosis of cystic fibrosis.

39 (B) Correct. Urine production is the major contributor to amniotic fluid, but in the fetus the placenta is the organ responsible for the removal of waste products. Inadequate amniotic fluid volume (oligohydramnios) can lead to restricted fetal movement & result in contractures. Amniotic fluid contains a number of factors ẃ help prevent against infection. Amniotic fluid helps to insulate the fetus & maintain temperature. Normal amniotic fluid volume is essential for the development of the lungs. Preterm PRM (particularly before 24 wk gestation) can cause pulmonary hypoplasia.

40 (C) Correct. This baby has hyponatraemia secondary to excessive IV dextrose administration. Fluids need to be restricted until the urine output improves. The addition of sodium chloride will lead to excessive fluid retention & may cause oedema & a deterioration in respiratory status.

41 (A) Correct. While the most likely cause of this baby's temperature is environmental (i.e. he is overwrapped), the bag urine result is abnormal. This may be due to contam-

ination & a clean specimen is required to exclude a urinary tract infection. US is only required if a urinary tract infection is proven on recollection of a clean specimen.

42 (B) Correct. Hemolytic jaundice is the most important cause of jaundice presenting on the first day of life & needs to be excluded. Jaundice of prematurity normally presents after 24 hrs of age. breast feeding does not cause jaundice in the first 24 hrs of life. Physiological jaundice normally presents after 24 hrs of age. Early-onset sepsis can cause jaundice the most common cause in the first 24 hrs.

43 (E) Correct. The major cause of physiological jaundice is increased bilirubin production due to ⇧ Hb levels at birth & a shortened RBCs lifespan. Decreased hepatic bilirubin excretion & Immature hepatic enzymes play a minor role. Enterohepatic circulation.have a significant contribution but not the major contributor.

44 (D) Correct. Infants ē undiagnosed CF can have malabsorption of the fat soluble vit (A, E, D & K) & this would be the most likely cause of late-onset hemorrhagic disease in this list. Septicemia can cause DIC & bleeding, but this is normally around the time of the acute episode. Oral vit K should be just as effective as IM if administered properly. Three doses (birth, 4-7 days & 1 month) are required & failure to complete the course is now the most common cause of late-onset hemorrhagic disease in exclusively breast fed infants. Formula is supplemented ē vit K. HIE can cause DIC & bleeding, but this is normally around the time of the acute episode.

45 (A) Correct. In preterm infants, Iron stores are low & exhausted quickly because of the infant's rapid growth rate. However routine administration of iron is controversial & not given in all units

46 (B) Correct. Classic β-thalassaemia major does not affect neonates because most of the Hb is in the fetal (HbF) form. Whilst it is unusual for ABO incompatibility to cause

hydrops fetalis, it has become one of the more common causes due to the decreasing incidence of rhesus isoimmunization. Rhesus isoimmunisation is one of the most common identifiable causes of hydrops fetalis however the incidence has decreased since the introduction of Anti D immunoglobulin. Twin-to-twin transfusion can cause severe anemia in the donor twin, leading to the development of hydrops fetalis.

47 (B) Correct. This baby most likely has Beckwith–Wiedemann syndrome. There is often hyperinsulinaemia due to beta-cell hyperplasia.(To transfere mmol to mg multiply 18).

48 (A) Correct. The baby should be commenced on a dextrose infusion. This baby's blood glucose level is too low to provide milk alone (either breast or formula). It may be reasonable to also attempt breastfeed but this should not be done in isolation. Glucagon may be required if simple measures fail to increase the blood glucose level.

49 (A) Correct. Babies ē hyperthyroidism are most likely to present in the first few days after birth before a neonatal screening test is performed. Results are often 'batched' & results may not be available until the baby is at least few wks old. Babies who are at risk of hyperthyroidism (secondary to maternal antibodies) should have thyroid function studies done after birth & be closely observed for signs & symptoms of thyrotoxicosis.

50 (C) Correct. Preconceptual administration of folic acid decreases the incidence of neural tube defects in at-risk women by as much as 75%. The use of sodium valproate is known to increase the risk of neural tube defects.

51 (D) Correct. Prematurity is the most important risk factor, being very rare in term infants. A,B,C,D increases the risk of IVH but is not the most important factor.

52 (C) Correct. The change that best predicts a bad outcome is abnormality in signal intensity in the posterior limb of the internal capsule (PLIC) & basal ganglia ē 90% sensitivity & 100% specificity, & positive predictive value of 100%. Cranial ultrasound

findings may be normal even in severe cases of HIE.

53 (E) Correct. This baby has the classic finding of benign myoclonic jerks. The calcium level is slightly low but not at a level that you would expect to cause seizures. The clinical picture & cord pH are not consistent ē HIE.

54 (C) Correct. Inherited causes now account for 50% of all cases of severe sensorineural hearing impairment; 80% are due to single-gene autosomal recessive disorders & 15% to autosomal dominant disorders

55 (C) Correct. Normal temperature is 37 °C. Such babies also lack brown fat. Cooling is effective in term babies in preventing some secondary brain injury. Babies receiving therapeutic hypothermia may shiver, but this is a side effect of treatment. Skin-to-skin contact prevents heat loss, although there is a risk of evaporative heat loss if the baby's back is not covered ē a warm towel or blanket

56 (D) Correct. The most important step in management is to give IM steroids to accelerate lung maturation & IV antibiotics because of the risk of ascending infection. Then transfer should be arranged as this woman has PRM & possibly early preterm labor. She is not actively contracting & her cervix is still closed, but she may progress quickly to established labor. If it is safe to transfer, then this should be done *in utero*, but steroids & antibiotics should be administered first. Tocolytic medication may be very useful to delay labour while transport arrangements are being made, but steroids & antibiotics should be administered first

57 (A) Correct. As the altitude increases there is a decrease in the atmospheric press-ure (from 760 mmHg at sea level to 640 mmHg at 5000 feet). This decreases the amount of oxygen that is available. Although increasing the PIP and PEEP will also improve oxygenation, the most appropriate step is to simply increase the percentage of inspired oxygen.

The CO_2 level is satisfactory & respiratory rate does not need to be changed. Decreasing the actual altitude to 5000 feet will make no difference if the cabin is already pressurized to that level.

58 (C) Correct. Males are at higher risk of adverse outcome than females.

59 (A) Correct. Gestation is the biggest independent factor in predicting adverse neurodevelopmental outcome.

60 (D) Correct. Postnatal depression is the most common cause of abnormal mother infant bonding.

61 (A) Correct. Both parents should be encouraged to visit or telephone to see how their baby is going whenever possible. Most neonatal units will have an 'open-door' policy ẃ allows parent involvement 24 hour/day.

62 (E) Correct. This is not appropriate in the immediate period after a baby's death & should be delayed until a follow-up appointment is arranged (6–8 wks afterwards).

63 (C) Correct. This baby has a right tension pneumothorax & is now in extremis. Urgent action to resolve the pneumothorax needs to be undertaken immediately.

64 (D) Correct. Ideally all adrenaline should be given IV but it is not unreasonable to give the first dose via the ETT while arrangements are being made to insert an emergency UVC. The second dose should be given via the UVC & no further time should be wasted giving another dose down the ETT. Emergency treatment is the priority until extra help arrives, but most guidelines recommend ceasing resuscitation attempts after 10 minutes if there have been no signs of life.

65 (C) Correct. The catheter is not in good position as it is too close to the origin of renal arteries (L1). The catheter simply needs to be repositioned to approximately L4.

66 (C) Correct. B12. Because vit. B12, or cyanocobalamin, is mainly found in animal

products, individuals who follow a strict vegan diet commonly have deficiency of this nutrient. In breastfeeding mothers who follow a vegan diet, the breast milk supply is likely to reflect this deficiency. Because the baby described is breast-fed & his mother follows a vegan diet, he is at greatest risk for deficiency of vit. B12. Option (A), B1, or thiamine, is incorrect because this nutrient is readily available in many vegetables, cereals, & fruits that are included in a vegan diet. Option (B), B6, or pyridoxine, is incurrect because this vit. commonly found in cereals that would likely be consumed by vegans. Similarly, option (D), is incorrect because a vegan diet is rich in fruits & vegetables that contain vit. C. Option (E), K, is incorrect because a vegan diet is rich in leafy greens, beans, & soybeans, all of ẃ are good sources of vit. K.

67 (D) The correct. kerion, because the clinical presentation of inflammatory pustules ē a surrounding area that is boggy & tender is most consistent ē this condition. **Kerion** is the result of the host's response to a fungal ringworm infection of the hair follicles of the scalp. Option (A), alopecia areata, is incorrect because although this condition does cause annular patches of hair loss, it does not cause the surrounding area to be boggy, tender, or inflamed. Option (B), atopic dermatitis, is incorrect because although this condition often develops on the scalp, it does not cause alopecia or tenderness. Option (C), herpes simplex virus, is incorrect because the lesions caused by this agent are vesicles, not pustules. Also, herpes simplex virus is not likely to cause localized alopecia, as noted in the patient described. Option (E), mastocytosis, is incorrect because mastocytomas are not pustules; they are firm, fixed, nodular lesions that may be blistering and vesicular. In addition, mastocytomas are often accompanied by pruritus but not tenderness.

68 (A) Correct. Coarctation of the aorta, because this condition is most likely to manifest as a murmur ē diminished or absent femoral pulses, as noted in the pt described. Option

(B), diaphragmatic hernia, is incorrect because this congenital defect most commonly manifests as severe RD within the first 24 hrs of life. Diaphragmatic hernia in neonates is often accompanied by scaphoid abdomen, but heart murmur & localized perfusion abnormalities are unlikely. Option (C), group B streptococcal sepsis, is incorrect because this condition in neonates is characterized by generalized findings such as temperature instability, poor fee-ding, lethargy, mottled skin, or overall perfusion concerns. Localized perfusion issues and heart murmur or femoral pulse abnormalities are not associated ē this condition. Option (D), PDA, is not correct because the characteristic murmur of this condition is continuous & machinelike. If a PDA does not close spontaneously within the first week, it might eventually present as slowly progressive heart failure, but it would not present as acute localized perfusion issues during the first five days of life. Option (E), F_4, is incorrect because this condition is most likely to present as severe immediate cyanosis after birth or as acute severe systemic cyanosis ē closure of a PDA within the first week. Localized perfusion issues & diminished femoral pulses are not characteristic of tetralogy of Fallot.

69-(E) Correct answer. substance use (substance abuse), ẃ is manifest by a change in personality traits, decreased school performance, neglected hygiene & a change in peer group preferences. Option (A), bipolar, is incorrect because the pt described has not experienced the periods of manic behavior that are characteristic of this disorder.Option (B), conduct, is incorrect because the patient described does not have persistent antisocial behaviors such as lying, stealing, fire-setting, truancy, cruelty to animals & property destruction. Option (C), major depressive, is incorrect because although this disorder includes some of the signs & symptoms as in the clinical scenario, complete loss of interest in friends is more characteristic than a change in peer group. Option (D), persistent depre-

ssive (dysthymic), is incorrect because the criteria for this condition include a one-year history of poor appetite, sleep problems, decreased energy & decreased self-esteem. An acute change or a change in peer groups is not characteristic of dysthymic disorder.

70 (C) Correct answer. Application of direct pressure, because this intervention facilitates hemostasis of the anterior nosebleed. Option (A), administration of clotting factors, is incorrect because this is appropriate for a life threatening bleed in a pt ē hemophilia but not for acute epistaxis. Option (B), anterior nasal packing, is incorrect because although it is a subsequent intervention for hemostasis in any acute anterior nosebleed, it is not the initial step. Option (D), cautery of the bleeding area, is incorrect because this intervention is appropriate to prevent recurrence after acute bleeding is controlled; it is not an appropriate initial intervention for acute epistaxis. Option (E), referral of the pt to a pediatric otolaryngologist, is incorrect because it is an option for long-term management of epistaxis but not an appropriate initial intervention.

71 (C) Correct answer. Enzyme immunoassay of stool, because the clinical scenario described is characteristic of giardiasis & the most appropriate next step is a stool study that would confirm the causative organism of Giardia lamblia. Option (A), air contrast enema, is incorrect because it is an appropriate diagnostic & therapeutic intervention for intussusception but not effective in identifying Giardiasis. Option (B), colonoscopy, is incorrect because this study is used to identify disease states that are specific to the colon & is not the test of choice for infections of the digestive system such as giardiasis. Option (D), esophagogastroduodenoscopy, is incorrect because it is indicated for clinical findings suggestive of upper GIT disease but is not appropriate to evaluate conditions such as giardiasis. Option (E), fecal leukocyte test, is incorrect although it is useful in identifying disease states producing inflammation, it is not effective in correctly diagn-

osing giardiasis.

72 (E) Correct answer. Transient synovitis, because the clinical presentation of pain in the knee, mild limp, limited internal rotation of the hip, & slightly elevated ESR ē normal WBCs count is most consistent ē this condition. Option (A), developmental dysplasia of the hip, is incorrect because although the presentation of this condition is similar to the clinical scenario described, X-ray studies of the hip & knee in a patient ē this condition would show either an abnormality of the femoral head or a chronic dislocation. Also, this is a diagnosis that is more likely to be made during infancy on well-child examination. Option (B), Legg Calvé-Perthes disease, is incorrect because although the clinical presentation of this condition is the same as that described, results of laboratory studies would be within normal limits. Option (C), septic joint, is incorrect because pts ē this condition typically have fever & rapid onset of symptoms as well as elevation of both ESR & WBCs count. Option (D), slipped capital femoral epiphysis, is incorrect because this condition usually develops at an age older than the patient described & is characterized by the finding of a slip on X-ray studies of the hip.

73 (E) Correct answer. Sweat chloride test, because it is the most effective study to confirm the presence of cystic fibrosis (CF). Option (A), bronchoscopy, is incorrect because although this study might identify Pseudomonas aeruginosa via lavage in a pt ē CF, it is not diagnostic for the condition. Option (B), CT scan of sinuses, is incorrect because although sinus disease may be pre-sent in the pt described, this study is not effective to provide a definitive diagnosis. Option (C), culture of aspirate from the trachea, is incorrect because although it does identify pathogens, it does not identify the underlying condition. Option (D), measurement of serum immunoglobulin, is incorrect because these levels might be elevated in pt ē CF, but these findings do not provide a definitive diagnosis.

74 (E) Correct answer. Wilms tumor, because the clinical presentation of hematuria, abdominal pain of 2 months' duration, & a palpable mass in the right upper quadrant of the abdomen is characteristic of this tumor. Option (A), cystic nephroma, is incorrect because this tumor typically presents as an asymptomatic benign mass in the kidney that is found incidentally. Option (B), cystitis, is incorrect because this condition does not include a retroperitoneal mass. Option (C), mesoblastic nephroma, is incorrect because this condition is exceedingly rare; in > 90% of cases, it presents before 1 yr of age. Option (D), neuroblastoma, is incorrect because although it can present ē intra-abdominal mass, proteinuria & hematuria are rarely associated ē this condition. In addition, neuroblastomas also present more often in children younger than 1 yr of age, making this diagnosis less likely in the pt described.

75 (B) Correct answer. Measure BP in the lower extremities. This component of physical exam excludes coarctation of the aorta as a cause of secondary hypertension. Option (A), counsel the parents regarding diet & exercise, is incorrect because although it is appropriate in the setting of primary hypertension, it does not exclude secondary hypertension & is not the appropriate next step in the scenario described. Option (C), order urinalysis, is incorrect because this study is appropriate in evaluation of 2ry hypertension but is not part of the thorough initial physical examination. Option (D), recheck BP in 2 wks, is incorrect because this is the appropriate next step for a pt who is not experiencing symptoms. Option (E), refer the pt to a nephrologist, is incorrect because the hypertension descrIbed has not been determined to be renally mediated.

76 () Correct answer. Penicillin, because administration of this medication in children ē SCD who are younger than 5 yrs of age decreases the risk of serious bacterial infections such as pneumonia. Option (A), erythromycin, is incorrect because this medication is not

a recommended prophylactic antibiotic for neonates. Option (B), hydroxyurea, is incorrect because although it is a medication used to treat sickle cell crisis, it is not indicated as an initial therapy in the pt described. Option (C), oxygen, is incorrect because the clinical scenario described includes no indications for administration of oxygen. Option (E), pneumococcal & meningococcal vaccines, is incorrect because immunization ē these vaccines is not appropriate for the pt described until 2 & 9 months of age, respectively.

77 (E) Correct answer. No abnormalities, because congenital hypothyroidism is a condition that most commonly demonstrates no physical findings early in the course of the disease. Options (A) bradycardia, (B) enlarged fontanelle, (C), hypothermia, & (D), umbelical hernia, are all incorrect because these physical findings are not signs of congenital hypothyroidism in neonates.

78 (D) Correct answer. Measurement of serum glucose level, because the clinical scenario describes classic signs & symptoms of onset of DM, ẃ is confirmed through this laboratory study. Option (A), CBC count ē differential & Option (B), LFTs, are incorrect because the results of these studies would be within normal limits in the pt described & would not be useful in confirming or ruling out the diagnosis of type 1 DM. Option (C), measurement of serum amylase level, is incorrect because although ⇧ levels of triglycerides can lead to inflammation of the pancreas, that is not the cause of the presentation described. Option (E), thyroid function studies, is incorrect because although the pt has fatigue, ẃ is consistent ē possible hypothyroidism, these studies are not effective in determining the diagnosis of onset of type 1 DM.

79 (A) Correect answer: (No additional treatment). The boy has chickenpox & no additional treatment is required as he is recovering & has no underlying medical problems.

80 (C) Correct answer. 2.5 mls. The child weighs 12.5 kg & the dose is 2 mg/kg, so she

needs a once daily dose of 25 mg. If trimethoprim suspension contains 50 mg in 5 mls it will contain 25 mg in 2.5 mls.

81 (A) Correct answer. (No treatment necessary). This child has molluscum contagiosum, a self-limiting illness, for ẃ the listed alternative options are inappropriate.

Knowing when treatment is not required is an important element of general practice.

82 (B &E) Correct answers: A 4-month-old girl who cannot grasp an object when placed in her hand & A 3-year-old boy who cannot combine words into a simple sentence.

83 (A) Correct answer. (Coxsackie A). This child has hand foot & mouth disease ẃ is caused by various strains of coxsackievirus (usually A) or enterovirus.

84- Correct answer: A) False. B) True. C) False. D) True. E) True.

The peak age of onset is 5-15 yrs. Rheumatic fever is rare before the age of 4 yrs. Incidence has fallen significantly since the 1940's. In developed countries the incidence is < 0.1 /1000/yr, while in developing countries is nearer 1/1000/yr. Incidence is higher where there is poverty or overcrowding presumably because of ⇧ transmission & because of undertreatment of streptococcal pharyngitis. The arthritis usually leaves the joints undamaged. Typically it is a migratory arthritis of large joints ẃ settles after 1-4 wks. Relapse of rheumatic fever is common (5% per yr). It occurs particularly after rheumatic carditis. Relapse can be prevented by penicillin V 250mg once or twice daily. Prophylaxis should be continued until age 20 or for at least 5 yrs after the last attack. Compliance ē this oral regimen has been reported to be as low as 10%. Benzathine penicillin 0.9 -1.2 million units IM every 4 wks is an alternative. Streptococcal skin infections are rarely, if at all, complicated by rheumatic fever. Treatment of acute rheumatic fever consists of penicillin & bed rest. The role of salicylates & steroids is controversial. Emergency valve replacement should be considered if there is progressive Ht failure & surgical skills are available.

85- Correct answer: A) False. B) False. C) True. D) True. E) True.

Causes of travellars diarrhea include: Enterotoxigenic E.coli (c.40% of cases), Shigella, Enteroinvasive E.coli, Campylobacter jejuni, Salmonella, Plesiomonas, Aeromonas, Protozoa, Viruses, No pathogen found (20% of cases). The diarrhea usually begins in the first few days. Antibiotics such as ciprofloxacin, especially if started early, reduce the duration of diarrhea & have a good safety profile in adults. Non-gastrointestinal pathogens such as Plasmodium falciparum can also cause diarrhea.

86- Correct answer: A) False. B) False. C) False. D) False. E) True.

The incubation period is 11-20 days, usually 2 weeks. Koplik's spots precede the maculopapular eruption of measles. The fever generally lasts about a week and settles as the last crop of spots appears. The use of acyclovir in uncomplicated chicken pox is controversial. Systemic acyclovir should be used in complicated chicken pox, particularly in adults. Recognized complications of chicken pox include impetigo, pneumonitis, encephalitis & thrombocytopenia. Non-immunes can acquire chicken pox from people ē either chicken pox or shingles. Shingles occurs when immunity is lowered in a person who has suffered from chicken pox in the past.

87- Correct answer: A) False. B) True. C) True. D) True. E) True.

Poliomyelitis is caused by 3 types of polio virus. Type 1 polio virus is the most virulent & prior to immunization was responsible for major epidemics. Polio virus is an enterovirus & is spread by feacal-oral route. Other factors associated ē paralysis are: pregnancy, tonsillectomy. The incubation period is 3-21 days (usually 7-14 days). Clinical features: there is a flu-like prodromal illness. The symptoms may resolve at this stage (abortive infection) or may progress to aseptic meningitis. The meningitis may be complicated by flaccid paralysis ẃ may be bulbar or spinal. Occasionally in children paralysis may be the first sym-

ptom of infection. Diagnosis: 1. Characteristic clinical features (NB no sensory deficit). 2. CSF leucocytosis (neutrophils may predominate initi-ally), raised CSF protein. 3. Isolation of virus from the stool or from a throat swab. Treatment is supportive & most efforts sh-ould be directed towards prevention by immunization: the Sabin vaccine, oral polio vacc-ine, is a live attenuated vaccine inco-rporating antigens from the 3 virus types.

88- Correct answer: A) False. B) True. C) True. D) True. E) True.

Corynebacterium diptheriae is a Gram positive, toxin-producing bacillus. the toxin is responsible for the manifestations of the disease. Of the 3 strains, gravis & intermedius produce more severe infection & mitis milder infection. Classically there is a grey phary-ngeal membrane. The "Bull neck" appearance is due to soft tissue oedema & cervical lymphadenopathy. Other causes of cervical lymphadenopathy may cause a superficially similar appearance. Management: Penicillin 200 mg/kg/D÷ 4 IV for 10 days or Erythrom-ycin 500 mg X 4, 50 mg/Kg/day ÷ 4 for 10 days. in case of penicillin allergy – Diphtheria antitoxin 200.000-80.000 units/D until complete until disappearance of the membrane, after doing allergy test by diluting to conc of 1:100 - 1:1000, 0.1 ml, ID, observe for 1 hr for any local reaction, if sensitive use adsorbed purified toxoid, if not available do desensitization as follow;

0.1 mL 1/20 conc. + 0.05 ml adrenaline SC. After 30 min

0.1 mL 1/10 conc. + 0.05 ml adrenaline SC. After 30 min

0.01 ml full conc. + 0.05 ml adrenaline SC. After 30 min

0.1 mL full conc + 0.05 ml adrenaline SC. After 30 min

1/2 mL full conc + 0.05 ml adrenaline SC. After 30 min

Full dose IM ē the presence of adrenaline, Decadrone ready for use in case of need.

- Isolation until 3 swabs/D are -ve. - Notification.

89- Correct answer E: Toddler's diarrhea is common between 1 to 3 years of age. These healthy children eat carbohydrate-containing beverages & eat snacks throughout the day. Therefore, carbohydrate-containing beverages should be reduced, fat containing foods should be increased & the fluid volume may be reduced.

90- Correct answer E: Reassurance; the condition is self-limited & symptoms resolve within 2-3 months. Rarely, anticholinergic therapy is effective. This boy is diagnosed ē pollakiuria or daytime frequency syndrome of childhood. This may be due to stress for an unknown reason or before the onset of kindergarten.

91- Correct answer A: NSAIDs & other anti-inflammatory drugs are not useful in Osgood-Schlatter disease.

92- Correct answer E: Ampicillin & Sulbactum are the preferred intravenous therapies for an infection in animal & human bite wounds. Ticarcillin-clavulanate is an alternative Rx.

93- Correct answer A: Tonsillectomy is indicated in a pt ē 7 or more tonsillar infections that were treated ē antibiotics in the preceding yr, 5 or more tonsillar infections that were treated in each of the preceding 2 yrs, or 3 or more tonsillar infections that were treated in each of the preceding 3 yrs.

94- Correct answer A: A left parasternal impulse is due to right ventricular or left atrial hypertrophy. Left ventricular hypertrophy causes a sustained, heaving, minimally displaced apex beat.

95- Correct answer A: Displaced apex beat; soft first heart sound; pan-systolic murmur; short mid-diastolic murmur. In mixed mitral valve disease, the first heart sound & the apex beat help to determine ẃ component is more prominent. A soft first heart sound & a displaced, thrusting apex beat suggest that regurgitation predominates. A loud first heart sound & a tapping apex beat suggest that mitral stenosis predominates.

96- Correct answer B. Scleroderma. "Raynaud's phenomenon, defined as episodic vasoconstriction in the fingers & toes, develops in virtually every pt ē systemic sclerosis. In some, episodes may also affect the tip of the nose & earlobes. Attacks are triggered by; a) Exposure to cold. b) Decrease in temp. c) Emotional stress. d) Using vibration tools.

97- Correct answer D: Clubbing can be quite tricky. Inspecting for loss of the nail-bed angle is the most reliable method & this is best done by looking at the nail-bed angle from the side.

98- Correct answer B: Expiratory component louder & longer ē a gap between inspiration & expiration. These are tricky to remember. Option a describes vesicular breath sounds.

99- Correct answer D: Cushing's Syndrome is due to excess glucocorticoids including cortisol not a deficiency. Enlarged extremities are seen ē acromegaly. Osteoporosis is a feature of Cushing's Syndrome along ē the typical moon facies & buffalo hump over the posterior aspect of the neck,

100- Correct answer B: Leads to generalized pigmentation. Addison's disease is due to a deficiency of glucocorticoids such as cortisol. It leads to dehydration & subsequent hypotension (& postural hypotension). DM is a complication of cortisol excess not deficiency.

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