INDEX

NEONATOLOGY INDEX

I. CARE OF THE NEWBORN...

TYPICAL CONCERNS WITH THE NEWBORN INCLUDE:

- Asphyxia and resuscitation
- Low birth weight
- Prematurity
- IUGR
- Disorders of adaptation to extra-uterine life
  - Temperature instability and cold stress
  - Respiratory distress
  - Persistent pulmonary hypertension
  - Gastro-intestinal disorders
  - Neonatal jaundice
  - Haematological disorders
  - Fluid and metabolic disorders
- Birth trauma
- Fetal hypoxia or birth asphyxia
  - HIE
  - Neonatal seizures
  - Intraventricular haemorrhage
- Infection
- Congenital disorders
  - Maldevelopment
  - Inherited disease

COMPLICATIONS OF PREMATURITY

- Respiratory problems (immaturity of higher control centers leading to periodic apnoea and inadequate surfactant leading to HMD)
- Temperature instability (small glycogen and fat stores, large body surface area, poor muscle tone, inability to shiver)
- Hypoglycaemia (poor fat and glycogen stores)
- Hepatic immaturity (bleeding tendency and jaundice)
- Oedema (often seen in preterm infants)
- Feeding difficulties (adequate coordination for sucking and swallowing occurs a 35 weeks along with slow gastric emptying increases risk of aspiration)
- Intraventricular haemorrhage (constant hazard due to the rich network of unsupported capillaries in the germinal matrix. Fetal hypoxia, birth asphyxia, fluctuations in BP and an unstable metabolic status increase the risk)
- Ischaemic brain injury
- Immaturity of the immune system (predisposition to infections with gram – organisms. These babies also present with atypical signs possibly delaying dx.
- Renal immaturity (cannot concentrate urine)
- Anaemia is a common problem The early form arises from exaggerated physiological factors and sluggish erythropoietic response. Late anaemia occurs with rapid growth and depletion of iron and folate stores.

These complications are especially common in high risk pregnancies:

FACTORS IDENTIFYING THE HIGH-RISK PREGNANCY		
MATERNAL	LABOUR AND DELIVERY	FETAL
Obstetric - Elderly primigravida - Anaemia - Poor weight gain/obesity - Previous abruption - Previous assisted delivery - Poor obstetric history (stillbirth or >2 abortions) - Previous LBW - Medical disorders - Pregnancy induced HPT **Social** - Age <16 or >35 - Low SES - Alcohol consumption - Smoking - Child with CP	- Maternal HPT - Maternal hypotension - Maternal sedation - PROM - Prolonged first or second stage of labour - C/S - Breech - Cord compression - Precipitate delivery - Preterm labour - Assisted delivery	- Oligohydramnios - Polyhydramnios - Multiple pregnancy - Fetal distress (acidosis, meconium-stained liquor, abnormal FHR) - Growth retardation - Post-maturity - Malformations

INITIAL MANAGEMENT AND RESUSCITATION OF THE NEWBORN

- The temperature of the delivery room must be between 23 and 28 degrees
- On delivery of the head the mouth is suctioned gently if the liquor is meconeum stained
- The cord is probably best clamped after the infant has uttered its first cry. On the other hand early clamping facilitates rapid resuscitation of the asphyxiated newborn
- Do APGAR
- Look for gross abnormalities
- Dry and wrap baby in a warm towel
- Give Vitamin K 1mg imi
- Give erythromycin eye ointment
- Identify baby with tags

FACTORS IDENTIFYING THE NEONATE AT RISK	
HIGH RISK	MEDIUM RISK
- Pre-term or post-mature - SGA/LGA - LBW/HBW - Neurological depression after resus - Metabolic problems after birth - Any congenital abnormality	- Birth weight 1.6-2.49kg - Clinically stable after resus - Birth trauma - Abnormal CNS signs - Cold exposure - Low blood sugar - Jaundice - Anaemia - Multiple births

BIRTH ASPHYXIA AND RESUSCITATION

RESUSCITATION OF THE NEWBORN – see algorithm (next page)

THE INFANT WHO DOES NOT RESPOND TO RESUSCITATION:

POSSIBLE CAUSES (DOPES)

- D: Displacement of the tube (can be in right main bronchus or in esophagus)
- O: Obstruction of the tube (kink, mucous, blood etc.)
- P: Pneumothorax
- E: Equipment failure (ventilator, connections etc.)
- S: Stomach distention

Table 1 The APGAR score

SIGN	SCORE 0	SCORE 1	SCORE 2
Heart Rate	Absent	< 100/min	> 100/min
Respiration	Absent	Weak	Good Cry
Muscle Tone	Flaccid	Some Flexion	Well Flexed
Reflexes	No Response	Grimace	Cough/ Sneeze
Colour	Pale/Blue	Blue Extremities	Completely Pink

CONSEQUENCS OF SEVERE HYPOXIA

Hypoxia, hypercapnia and acidaemia cause tissue injury. Clinical manifestations of the cerebral insult are those of hypoxic ischaemic encephalopathy (HIE). Acidaemia itself affects the myocardium, with a consequent drop in cardiac output. The combination of hypoxia, acidaemia and hypotension will affect the lungs, kidneys, gut and liver Metabolic disturbances such as hypoglycaemia, hperglycaemia, hypercalcaemia and SIADH may occur. There may also be clotting disturbances resulting in DIC. Neonates with severe asphyxia who have not established sustained respiration after 20 minutes almost always develop signs of HIE with a poor long-term outcome.

CARE FOLLOWING RESUSCITATION

- Temperature
- Blood sugar
- Maintain a clear airway (suction if needed)
- Oxygen if needed
- Tube feeding in cases of pneumonia or subtle convulsions
- Persistent pulmonary hypertension characterized by persistent cyanosis in the presence of metabolic acidosis
- Enlarging kidneys, decreased urine output and the presence of blood or protein in the urine suggests renal involvement

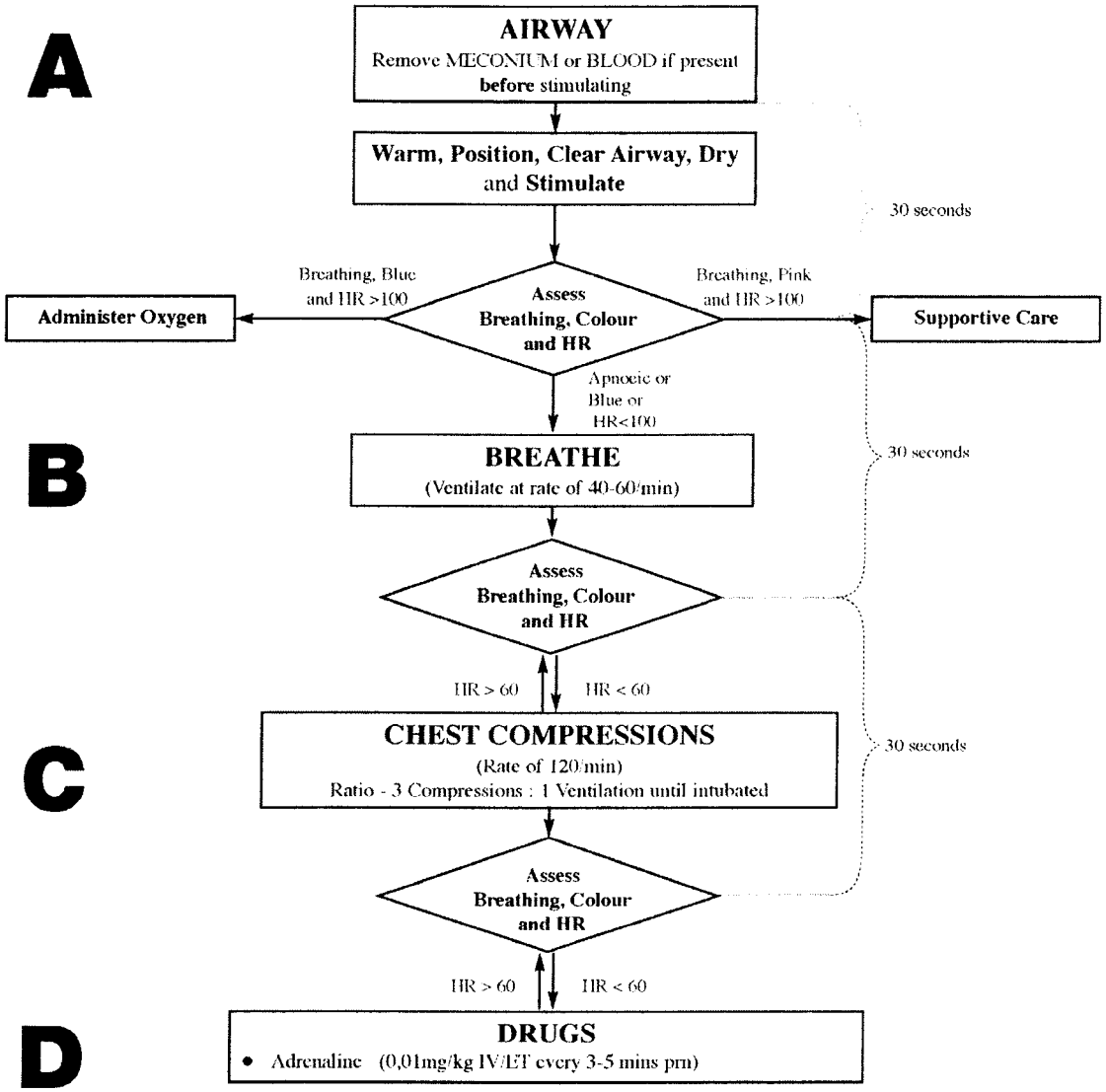
TRANSPORT OF THE HIGH RISK NEONATE

- Contact referral centre
- Competent health care worker must accompany baby
- Ensure clear airway
- Ensure that oxygen and suction is available
- Prevent heat loss during transfer
- Monitor vitals on route
- Monitor IV fluids on route
- Ensure that medical records are sent with the patient

Newly-Born Life Support Algorithm

2004

SAPA



ONLY IF SPECIFICALLY INDICATED:

- **Naloxone** (0,1mg/kg IV/ET/IM/SC if narcotic use suspected)
- **Normal Saline** (10ml/kg IV over 5-10 mins if perfusion poor)
- **Bicarbonate** (1mEq/kg IV(diluted) only for persistent metabolic acidosis in **prolonged** resuscitation)
- **Dextrose** (0,2g/kg IV(diluted) only if hypoglycaemic)

The algorithm follows the assumption that the previous step was unsuccessful and the newly-born is deteriorating

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POST HYPOXIC DAMAGE

HYPOXIC ISCHAEMIC ENCEPHALOPATHY (HIE)

This results from significant hypoxia of the fetus or newborn. Hypoxia is due to the failure of gas exchange at placental level in the fetus and pulmonary level in the newborn. Perinatal hypoxia is predominantly an antenatal event, with no more than 10 percent occurring postpartum. HIE is probably the major cause of CP in the developing world.

The systemic response to hypoxia, hypercarbia and mixed acidosis is maintenance of cerebral blood flow at the expense of other organs. It follows that if an episode of hypoxia is sufficiently prolonged and severe, other organs such as the heart will be affected. With further decreased cardiac output, hypotension occurs and perfusion of the brain, kidney, lung and gut is compromised. The clinical effects are those of ischaemia of these organs.

CLINICAL FEATURES

- Hypoxia during labour may be followed by a lucid interval of 12-18 hours before convulsions occur, which at times are subtle and expressed as apnoeic or cyanotic attacks.
- The respiration is often irregular with a Cheyne-Stokes pattern suggesting diffuse bilateral hemisphere pathology.
- Severe insult; brain stem signs may be present (fixed dilated pupils, abnormal/absent eye movements)
- Motor weakness
- Full fontanelle as a result of cerebral oedema

CLINICAL GRADING OF HIE		
MILD	MODERATE	SEVERE
Manifests neurological signs such as feeding and tone disturbances for 24-28 hours	- Convulsions - Signs lasting for 4-5 days	- Severe physical signs - Signs persist for >7days

MANAGEMENT

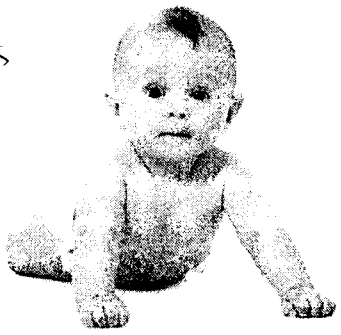
The most important aspect is prevention. Identify high risk babies and take the necessary steps as precautions. Once hypoxic brain damage has occurred:

- Raise head 30 degrees
- Control seizures
- Keep infants core temperature at 35 degrees for 48 hours. This reduces the risk of progressive brain damage
- Maintain blood sugar above 2.5
- Maintain BP with fluids and dopamine if needed
- Adequate resus to prevent acidosis
- Monitor for clotting problems
- Feeding is by NG tube (not exceeding 80ml/kg/day for the first 2-3 days)
- Respiratory support for a limited period may be considered for the infant with severe HIE
- Infection. Early neonatal meningitis may be clinically indistinguishable from HIE. If in any doubt, do LP. Routine antibiotics not recommended.

COMPLICATIONS

- Pneumonia
- Hypoglycaemia
- SIADH
- Developmental delay, CP etc.

due to convulsions



PROGNOSIS

Depends on the duration and severity of the cerebral insult.

CONVULSIONS

Neonatal seizures result from an insult to the brain. In themselves they are also injurious to the neurons. Furthermore, the concurrent respiratory disturbance causes hypoxia and hypercarbia.

- SUBTLE CONVULSIONS – Very common. Signs include
  - deviation of the eyes
  - repetitive blinking or fluttering of the eyelids
  - drooling, sucking
  - cycling movements of the lower limbs
  - rowing movements of the upper limbs
  - tonic posturing of a limb
  - apnoea attacks
  - cyanotic episodes
  - abnormal cry
  - stertorous respirations
- TONIC SEIZURES – indicate severe encephalopathy
- CLONIC CONVULSIONS – May be focal or multifocal. May also present as myoclonic jerks.
- JITTERINESS – Must be distinguished from convulsions; it is not accompanied by loss of consciousness nor abnormal eye movements and stops as soon as the limbs are held. However, I easily recommences with stimulation. Bradycardia, pallor or cyanosis do not occur.

CAUSES OF SEIZURES

- Intrapartum asphyxia
- Intracranial bleeds
- Hypoglycaemia
- Meningitis
- Low Na, Ca or Mg
- Inborn error of metabolism
- Narcotic/alcohol withdrawel
- Hypothermia

MANAGEMENT

- Stop convulsions ASAP
- Supportive care
- Keep airway clear
- Nurse baby prone or on the side
- Oxygen for cyanosis
- Monitor vitals and sugar
- When seizures have ceased for 24-48 hours, the Anticonvulsant dosage may be reduced gradually over days.

ANTICONVULSANT DRUG USAGE

DRUG	DOSAGE
SEIZURE CONTROL	
- Diazepam	0.5mg/kg/dose PR
Or	
- Phenobarbitone	15mg/kg imi/ivi
MAINTENANCE	
- Phenobarbitone	3-5mg/kg/day in 3 divided doses PO
Or	
- Phenytoin	3-5mg/kg/day in 3 divided doses PO

PROGNOSIS

- Determined by the nature of the underlying neurological disease
- Babies with convulsions due to HIE have only a 50% chance of normal development
- Hypoglycaemia-associated convulsions have a similar outcome
- Convulsions due to intracranial infection are associated with permanent damage in 20-50% of cases
- Severe IVH causes 65-100% morbidity with a mortality of 50-65%!

FOLLOW-UP

- *Assess neurological status at regular intervals (deficits usually manifest at 9-12 months)*
- *Minor problems are often not detected during the preschool period but arise later as attention and learning difficulties*
- *Involve other disciplines as needed*

INTRAVENTRICULAR HAEMORRHAGE (IVH)

The classic haemorrhage of the preterm infant occurs within the first 72 hours of life, often in association with respiratory distress. It commences as a haemorrhage into the germinal matrix and then may burst into the ventricles. The delicate vessels of the germinal matrix form a large unsupported network of capillaries which ruptures easily. As the fetus matures the germinal matrix becomes less vascular. This form of haemorrhage is therefore rare in the term baby.

PATHOGENESIS

Not fully clarified. Hypoxia and ischaemia are major factors. Raised central venous pressure ocuring during resuscitation may contribute. There is also marked fibrinolysis in the newborn promoting spread of the haemorrhage.

CLINICAL PRESENTATION

- Depends on the size and rate of bleeding
- Loss of consciousness
- Apnoea
- Convulsions
- Full fontanelle
- Anaemia
- Altered muscle tone
- Behavioral disturbances
- Progressive head enlargement
- Onset may be gradual or sudden

DIAGNOSIS –US/CT

GRADING:

Grade 1 and 2 has a good chance of full recovery!

1. Bleed in germinal layer only
2. Extension of the bleed into the ventricles
3. Ventricles dilated with blood – high rate of obstructive hydrocephalus
4. An associated periventricular venous infarct
    - Unilateral – hemiplegia
    - Bilateral – death

MANAGEMENT

Intervention is not indicated for those with severe haemorrhage as the mortality is in the region of 90% and the remaining patients have severe morbidity. Blood transfusions, anticonvulsants and eventual shunting may be indicated.

PERIVENTRICULAR LEUCOMALACIA (PLV)

This occurs when ischaemia is prolonged or severe in a preterm infant. This degenerative process may resolve or progress to multiple small cysts, clinical features are determined by the site and extent of the injury. The diagnosis is made by intracranial U/S.

PARASAGITTAL CEREBRAL DAMAGE

This typical cerebral lesion occurs in the term infant. There is necrosis of the cerebral cortex and subcortical white matter with characteristic bilateral symmetrical distribution involving leucomalacia of the parasagittal and supero-medial aspects of the cerebral hemispheres. The 'watershed' infarct which follows cerebral hypoperfusion emphasizes the ischaemic nature of the lesion. Clinically, spastic motor deficit, seizures and intellectual impairment occur.

FOCAL ISCHAEMIC CEREBRAL INJURY

Occur as a result of generalized cerebral hypoperfusion with the middle cerebral arteries being most frequently involved. Infarction occurs with subsequent cystic development which may or may not communicate with the lateral ventricles.

2. NEONATOLOGY EXAMINATION...

1st 28 days of life!!!

IMPORTANT BUT TERRIBLY CONFUSING TERMINOLOGY

- LOW BIRTH WEIGHT BABY – Baby with birth weight of <2.5kg, irrespective of gestational age
- PRETERM BABY – This is a baby delivered before 37 completed weeks of gestation (<259days)
- TERM BABY – A baby delivered from 38-42 weeks gestation
- POST-TERM BABY – A baby delivered after 42 weeks gestation
- APPROPRIATE FOR DATES BABY – Also appropriate for gestation (gestational age) baby. This is a baby whose weight falls between the 10th and 90th percentiles for gestational age (on a population specific chart)
- SMALL FOR DATES BABY – Birth weight <10th percentile
- LARGE FOR DATES BABY – Birth weight >90th percentile

SYMMETRICAL IUGR -CAUSES

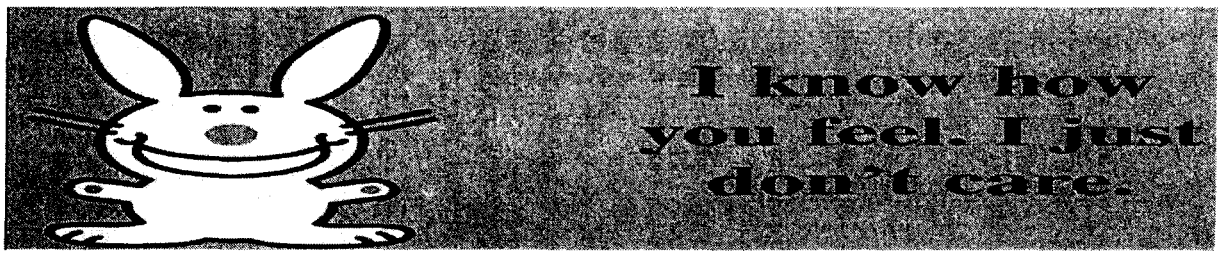
- Low maternal weight
- Genetic abn
- Chromosomal defects
- Teratogenic agents
- Cong infx

ASYMMETRICAL IUGR

- Due to uteroplacental factors later in pregnancy
- PET
- Smoking
- Placental infarcts etc.

- INTRAUTERINE GROWTH RETARDATION (IUGR) - The term IUGR denotes an abnormal situation with reduction of growth, a downward inflexion from the normally steady progression. This term is highly misleading, because in any normal population of fetuses, some 10% will by definition have a weight below the 10th percentile. Therefore, IUGR is better restricted to those fetuses where there is definite evidence that growth has faltered. A fetus whose weight has fallen from the 90th percentile to the 30th in a short time is almost certainly in greater peril than a fetus who has maintained a position on the 5th percentile.
- FETAL MALNUTRION – Fetal malnutrition (FM) is defined as failure to acquire adequate quantum of fat and muscle mass during intrauterine growth. In FM, the subcutaneous tissues and underlying muscles are diminished and the skin of arms, legs, elbows, knees and interscapular regions is very loose. In severe FM, the neonate may look "emaciated" or "marasmic" as the skin appears "several sizes" too large for the baby. The decreased subcutaneous fat and muscle are evident by more quantitative measures such as upper arm circumference, triceps and interscapular skin fold measurements with estimate of arm muscle area. Buccal and buttock fat pads are reduced and the scalp hair may be coarse, patchy, or "straight and starring" as in marasmus or even have a "Flag-Sign" as in severe protein-calorie malnutrition (Kwashiorkor). Fetal malnutrition is therefore, also a clinical diagnosis. Babies who show evidence of muscular wasting should therefore be labeled appropriately.
- POSTMATURITY – Develops as a result of prolonged pregnancy (pathophysiology = placental insufficiency) and is characterized by the following clinical signs post delivery: Loss of subcutaneous fat, dry cracked skin, absence of vernix and lanugo, meconium stained liquor and skin, long nails, abundance of scalp hair, scaphoid abdominal shape, and... wait for it... an attentive, apprehensive facial expression..?

???CONFUSED...?



BACK TO BUSSINESS - SUGGESTED EXAMINATION ROUTINE:

1. GENERAL
2. HEAD (SKULL,FACE,NECK)
3. CHEST (GENERAL, HEART, LUNG)
4. ABDOMEN (INCLUDING UMBILICUS)
5. NAPPY AREA
6. LIMBS
7. BACK
8. NEUROLOGICAL
9. GESTATIONAL AGE DETERMINATION

You can do the examination in any order as long as you don't leave anything out..

Outline only – details discussed later

GENERAL EXAMINATION -

1. **GENERAL APPEARANCE AND APGAR**
   - APGAR – resus if needed
   - Low birth weight (<2.5kg) – preterm vs small for dates etc.
   - ?Acutely ill (resp distress etc)
   - Posture and movement (Normal = flexion + symmetrical)
   - Colour: Jaundice – NB abn in 1st 24hrs!
     Cyanosis – peripheral vs central
     Polycythaemia – Hb>22/Hct>65%
     Pallor – look at mucous membranes
     Redness – (polycythaemia, overheating, cold injury)

2. **MEASUREMENTS**
   - Temperature – axillary 36-37degrees
   - RR – normal = 40-60bpm
   - HR – normal 120-160bpm
   - Glucose >2.2mmol/L
   - Weight – Average: Male – 3.4kg, female – 3kg
   - Length - Normal = +/- 53cm
   - Head circumference – Normal = +/- 35cm measured above eyebrows and around occiput
   - BP – best method is doppler, other include flush and palpation methods

Weight	Systolic	Diastolic
1-2kg	50	30
2-3kg	60	35
>3kg	70	40

3. **NUTRITIONAL STATUS**
   - Ponderal index = [weight(gram])/[length(cm)^3] times 100 (Some authorities classify normal **ponderal index** as 2.32-2.85g/cm3)
   - Clinical Assessment of Nutritional Status [CANS] of the fetus using the CANSCORE

1. *Hair*
   Large amount, smooth, silky, easily groomed(4).
   Thinner, some straight, "staring" hair(3).
   Still thinner, more straight, "staring" hair which does not respond to brushing(2).
   Straight "staring" hair with depigmented stripe (flag sign)(1).
2. *Cheeks*
   Progression from full buccal pads and round face(4), to significantly reduced bucal fat with narrow, flat face(1).
3. *Neck and Chin*
   Double or triple chin fat fold, neck not evident (4); to thin chin. No fat fold, neck with loose, wrinkled skin, very evident (1).
4. *Arms* - Full, round, cannot elicit "accordion" folds or lift folds of skin from elbow or tricep area (4); to a striking "accordion" folding of lower arm, elicited when examiner's thumb and fingers of the left hand grasp the arm just below the elbow of the baby and thumb and fingers of the examiners right hand circling the wrist of the baby are moved towards each other; skin is loose and easily grapsed and pulled away from the elbow.
5. *Legs*
   Like arms.
6. *Back*
   Difficult to grasp and lift skin in the interscapular area(4); to skin loose, easily lifted in a thin fold from the interscapular area(1).
7. *Buttocks*
   Full round gluteal fat pads (4); to virtually no evident gluteal fat and skin of the buttocks and upper posterior high loose and deeply wrinkled(1).
8. *Chest*
   Full, round, ribs not seen(4); to progressively prominence of the ribs with obvious loss of intercostal tissues(1).
9. *Abdomen*
   Full, round, no loose skin(4); to distended or scaphoid, but with very loose skin, easily lifted, wrinkled and "accordion" folds demonstrable.

HYPOTHERMIA

- In LBW babies increases mortality by 25%
- Most common cause is low ambient temp
- Hypoxia also interferes with heat production
- Hypotonia diminishes metabolism in muscle
- LBW baby has no brown fat stores
- Sepsis may also interfere with metabolism
- Adverse effects include; MA, hypoglycaemia, decreased surfactant production, rise in FFA
- Mx: Prevention; dry and wrap baby at birh, skin to skin contact, radiant heaters, space blankets etc.
- NEONATAL COLD INJURY – Temp 32 degrees or less. Present as multi-organ dysfx. Rx as above and manage complications

OVERHEATING

Babies in incubators at high risk. Compl include increased water loss with dehydration and increased Na. Apnoea, heat stroke and death.

NEONATAL HYPOGLYCAEMIA

Causes

1. decreased carbohydrate stores (premature, IUGR)
2. infant of a diabetic mother (IDM): maternal hyperglycemia —> fetal hyperglycemia and hyperinsulinism—> hypoglycemia in the newborn infant
3. sepsis
4. endocrine: hyperinsulinism due to islet cell hyperplasia (e.g. Beckwith Wiedeman syndrome), panhypopituitarism, suppression of hypothalamo-pituitary axis (HPA)
5. inborn errors of metabolism: fatty acid oxidation defects, galactosemia

Clinical Findings

- signs often non-specific and subtle: lethargy, poor feeding, irritability, tremors, apnea, cyanosis, seizures

Management

- obtain critical sample (blood taken during hypoglycemic episode) send for glucose, insulin, cortisol, growth hormone (GH), ß-hydroxybutyrate, lactate, ammonia, free fatty acids (FFA's), acid-base status
- provide glucose IV (e.g. D25W)
- hyperinsulinism: treat with diazoxide

BECKWITH- WIEDEMANN SYNDROME

- an overgrowth disorder present at birth characterized by an increased risk of childhood cancer
- Macroglossia
- Macrosomia
- Umbilical hernia/omhalocoele
- Ear pits/sinuses
- Neonatal hypoglycaemia
- Other congenital abnormalities

In the 1960s, Dr. J. Bruce Beckwith, an American pathologist and Dr. Hans-Rudolf Wiedemann, a German pediatrician, independently reported cases of a proposed new syndrome.[18][19] Originally termed EMG syndrome (for exomphalos, macroglossia, and gigantism), this syndrome over time became known as Beckwith-Wiedemann syndrome or Wiedemann Beckwith syndrome.

4. **OEDEMA** - Periorbital, dorsum of hands, anterior aspect of the distal lower limbs

5. DEHYDRATION –signs

6. LYMPHADENOPATHY – congenital infx

Body weight % lost	Clinical state	Signs
<5%	Not unwell	Thirst, dry mucous membranes
5-10%	Apathetic	Sunken eyes and fontanelles, oliguria
>10%	Usually shocked	Signs of shock

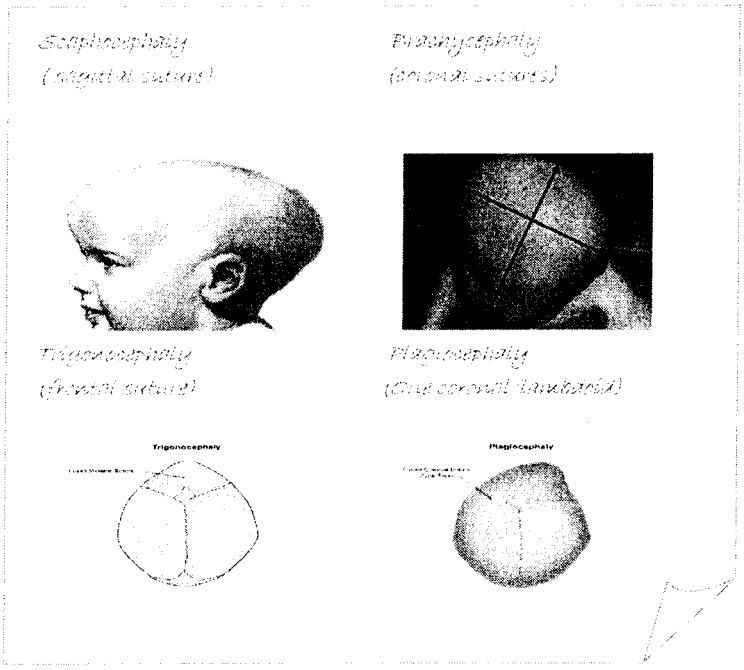
7. **SKIN – see atlas for pictures...**
   - Normal – light pink soft and friable
   - Preterm – very thin and transparent
   - Vernix caseosa – White, greasy, secreted by sebaceous glands, normal in term baby
   - Lanuga – Fine downy hair, most common on back and face + preterm babies. They usually fall out during 1s few months
   - Milia – small white pinpoint papules found on nose and chin shortly after birth due to keratin plugs in the sebaceous glands.
   - Skin peeling – Most common in post term and IUGR's. Severe = congenital syphilis
   - Erythema toxicum – common, benign rash Appears in 1st few days. Multiple small red areas with central whitish yellow papule
   - Mongolian spots – occurs over buttocks, back or shoulders. Benign, most common in blacks
   - Stork mark – flat, pinkish capillary haemangioma between the eyebrows, on the forehead and eyelids and on the nape of the neck. It fades gradually over the first 2 years
   - Port wine stain – capillary haemangioma which appears flat and pale pink and may be easily overlooked in the infant. It occurs in the distribution of the 5th cranial nerve and does not cross the midline. It darkens with time to form a flat purple patch of skin. It does not fade. Treatment is difficult and includes laser therapy and simply cover the mark with cosmetics.
   - Sturge-Weber syndrome is a rare association if a unilateral port wine stain of the face and an intracranial haemangioma of the pia aracchnoid on the same side. Affected children may present with seizures, hemiplegia etc.
   - Strawberry mark – **only appears a few days after birh..** Soft, raised, bright red capillary haemangioma. Sometimes it involves deeper tissues and is combined with a cavernous haemangioma which gives it a blue tinge.

THE SKULL...

INSPECTION

SIZE – Impression: Macro,micro and anencephaly. Hydrocephaly discussed on next page
SHAPE – soft skull bones attached by fibrous tissue allows for all kinds of skull shapes!
- Moulding
- Craniostenosis (early, permanent closure of sutures)

Midline defects (encephalocoele/ meningocoele)
Swellings (see palpation)
Indentations (amniotic bands, depressed skull fracture)
Fontanelles – sunken, bulging (see palp)
Hair – term(coarse and silky), preterm (fine, downy). Low posterior hairline – look for congenital abnormalities...
Scalp – Aplasia cutis (absent area of skin, usually in midline, associated with congenital abnormalities). Petechiae – if confined to scalp and face probably as a result of shoulder dystocia or cord wrapped around neck, if generalized think neonatal thrombocytopaenia....

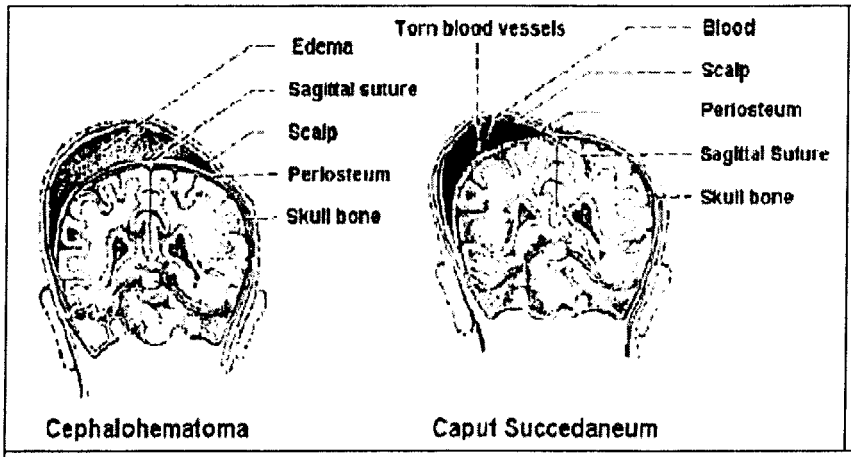


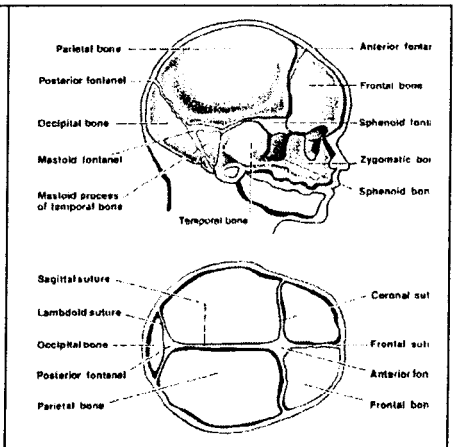
PALPATION

Head circumference
Scalp swellings:
- Cephalhaematoma – haematoma between skull and periosteum, does not cross suture lines, takes 6-8 weeks to reabsorb. May mimic a depressed skull fracture (hard edges, soft centre)
- Vacuum (suction) haematoma
- Caput succedaneum – Oedematours scalp swelling, disappears within days, may cross suture lines
- Subaponeurotic haemorrhage – extensive haemorrhage below the epicranial aponeurosis (often after assisted delivery), may cross suture lines, associated blue swellings of eyelids or behind ears may occur. Baby can be shocked.

Complications of a cephalhaematoma:
- Anaemia
- Jaundice
- Infection
- Calcification

SWELLINGS OF THE HEAD				
	CAPUT SUCCEDANEUM	*VACUUM EXTRACTION HAEMATOMA*	*CEPHALHAEMATOMA*	*SUBAPONEUROTIC HAEMORRHAGE*
SITE	Diffuse over presenting part	Localized at site of vacuum application. Skin and subcutaneous tissue involved	Localized, usually over parietal bones, under periosteum. Extension limited by periosteal adhesion of sutures	Diffuse over whole head underneath cranial aponeurosis
CAUSE	Oedema and bruising of presenting part	Oedema +/- hemorrhage at vacuum site	Haemorrhage often due to CPD	Diffuse hemorrhage; sometimes follows vacuum extraction or poorly applied forceps
ONSET	Present at birth	Present at birth	Often only detected 6-12 hours after birth. Becomes progressively larger over 1-2 days	May be present at birth; swelling often increases during first 2 days
DISTINGUISHING FEATURES	Diffuse. Petechiae over swelling	Usually well defined. Localized abrasions at periphery of swelling. Overlying skin may be purple	Well defined. Does not cross suture lines. May be bilateral, but then a groove is present between the 2 swellings. The skin is normal	Diffuse and sometimes massive haemorrhage. Crosses suture lines. Bluish discoloration of upper eyelids or behind ears. Skin is normal
COURSE	Disappears within 48 hours	Subsides within 1 week	Persists 6-8 weeks. Centre may become fluctuant	Gradual reabsorption of blood
COMPLICATIONS	Nil	Anaemia, infection, jaundice	Anaemia, jaundice, infection if aspirated. Rarely, underlying skull fracture	Severe anaemia, shock, jaundice
TREATMENT	Nil	Local antiseptic for abrasions. Treat complications	Usually nil. Observe for complications	Vitamin K, may need urgent blood transfusion





HYDROCEPHALY:

Def: Condition of excessive accumulation of intracranial CSF as a result of disturbance of the formation, flow or reabsorption of CSF.

CLINICAL PICTURE: The head circumference may be normal at birth and percentile lines may be crossed during F/U visits. A correlation exists between length and head circumference, which may be used in the cases of premature or unusually large babies. Brain tissue is more compliant than the skull and the ventricles are therefore already large before the head circumference starts to increase. A full fontanelle, splayed suture lines, a sunset sign and poor growth only occur later on. Diagnosed with sonar, CT and sometimes MRI. Treatment involves a ventriculo peritioneal shunt. If this is done at an early stage, before irreversible brain damage has taken plance, normal neurological development takes place.

CONGENITAL CAUSES OF HYDROCEPHALUS:

- AQUEDUCT STENOSIS: 66% of cases of congenital hydrocephalus. It is caused by forking of the aqueduct, a septum or gliosis with obliteration of the aqueduct.
- CHIARI MALFORMATION: It is a condition of hypoplasia of the posterior cranial fossa with herniation of the cerebellar tonsils through the foramen magnum. The superior cerebellar vermis herniates upwards through an incompletely formed tentorial hiatus, resulting in kinking of the aqueduct.
- DANDY WALKER SYNDROME: It is associated with cystic dilatation of the fourth ventricle and hypoplasia of the cerebellar vermis. There is outflow obstruction of the fourth ventricle with or without accompanying aqueduct stenosis.
- ARACHNOID CYSTS: Intracranial arachnoid cysts may exert pressure on the foramen of Monro or on the aqueduct, resulting in obstruction of the CSF circulation.
- VENA OF GALEN VASCULAR MALFORMATIONS: This leads to increased venous pressure in the superior sagittal sinus, resulting in resistance to CSF re-absorption.

PALPATION CONTINUED.

Fontanelles:

- Anterior; diamond shaped, 2.5 by 2.5cm, closes at 8-18months
- Posterior; triangular, 0.5cm diameter, closes at 6weeks
- 3rd fontanelle – between ant and post fontanelles, 3cm in front of posterior fontanelle. Associated with Downs but may be normal ☺
- Bulging – think raised ICP
- Sunken – think dehydration

Sutures:

- Splaying; Think raised ICP (normal may be up to 1cm for sagittal suture!)
- Mobility; Normal in newborn
- Moulding; Overriding of adjacent skull bones. Can be easily palpated.
- Craniostenosis/craniosynostosis;
- Craniotabes – Refers to areas of softening of the skull bone. It presemts mostly in the temporal and parietal areas adjacent to the suture lines. It occurs as a result of defective ossification and is present in most preterm babies and in 10-30% of normal term babies. During the neonatal period craniotabes is not usually pathological and usually disappears within a few weeks. In older babies causes include; rickets, raised ICP, hypothyroidism, osteogenesis imperfect and cleidocranial dysostosis.

CLEIDOCRANIAL DYSOSTOSIS

- Gene mutation on short arm of chromosome 6 (autosomal dominant)
- Partly or completely absent clavicles
- Underdeveloped bones and joints, fontanelles fail to close, Frontal bossing, hypertelorism

PERCUSSION – Cracked pot sign is of little importance in the newborn due to open sutures and fontanelles, therefore NOT an indication of raised ICP in the newborn!

AUSCULTATION – murmer over skull may indicate a intracranial AV-malformation

TRANSILLUMINATION – May indicate hydranencephaly, subdural effusions or cysts

FACE AND NECK

Now is a good time for a tea break...

EXAMINATION OF FACE

- Shape
- Skin (see general exam)
- Ears
- Eyes
- Nose
- Mouth
- Jaw

EARS:

- PINNA- term baby's ears are firm because of cartilage. If absent consider Goldenhar syndrome
- ABNORMAL EARS – low set or rotated ears may be normal but congenital abnormalities should be sought
- PRE-AURICULAR SINUS – (blind ending hole situated in front of the ear and may be associated with renal problems. Excision indicated for recurrent infection
- PRE-AURICULAR SKIN TAGS – single or multiple. Isolated or associated with cong abn e.g. Goldenar, Treacher-Collins, renal problems

Abnormal shape may be due to; familial traits, pressure effects, congenital abn or facial palsy.

Diff asymmetrical face when crying

- UMNlesion (forehead spared)
- LMN lesion (forehead involved)
- Cayler syndrome (congenital absence of depressor anguli oris on 1 side

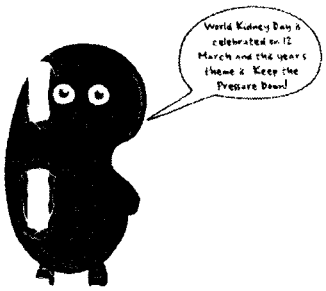
Abnormal ears – always check the kidneys!

GOLDENHAR SYNDROME

- Documented in 1952
- Cause unknown, thought to be multifactorial
- Chief markers are incomplete development of the ear, nose, soft palate,lip and mandible.
- Other problems include severe scoliosis, hearing loss. coloboma

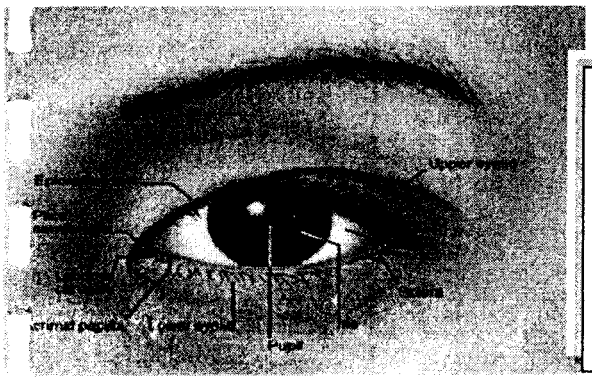
TREACHER-COLLINS

- Gene defect chromosome 5. (AD)
- Features; downward slanting eyes, micrognathia, conductive hearing loss, underdeveloped zygoma, malformed or absent ears
- Coloboma



EYES

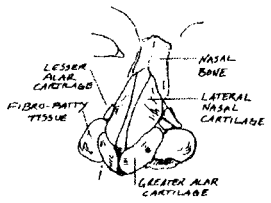
- Eyes can open from 25 weeks gestation
- Eyelid swelling may be the result of birth trauma or local infection
- Subaponeurotic haemorrhage – reddish-purple eyelids
- Ptosis (local infx or injury, 3rd cranial nerve fallout, Horners, myasthenia etc)
- Coloboma – This is a split in the eyelid, associated with Goldenhar and Treacher-collins
- Palpebral fissures; Mongoloid (as in Down therefore lateral and upwards extension) and anti-mongoloid with lateral and downward extension (associated with Treacher Collins)
- Epicanthic folds – these are abnormal, vertical, sickle cell shaped skin folds extending downwards over the inner canthus of the eye. May be familial and normal but are commonly associated with chromosomal abnormalities such as Downs. NB- A broad flat nasal bridge may give the impression of epicanthic folds...
- Tears; seldom noticed in neonatal period, may indicate nasolacrimal duct obstruction
- Conjunctivitis; inflamed eye – immediate antibiotic treatment needed
- Hypertelorism ; Normal distance between the midpoint of the two pupils in the term baby is 3-5cm.
- Vision/eye movements; Transient strabismus is normal up to 3months of age. Nystagmus at any age necessitates ophthalmological referral
- The oculo-cephalic reflex (doll's eye reflex) – normal in 1st few weeks of life
- Proptosis – Bilateral think familial or hyperthyroidism. Unilateral think retrobulbar haemorrhage
- The sclera – Bluish sclera at birth may be normal and is not necessarily a sign of osteogenesis imperfect.
- Subconjunctival haemorrhage – This is often observed after difficult deliveries, no treatment is needed and it resolves spontaneously within one month
- The cornea – An opaque cornea may be an indication of congenital glaucoma, an abnormal anterior chamber or keratitis
- The iris – Often blue at birth until 3-6 months of age and is usually not an indication of the future eye colour. The small grey-white spots around the iris in babies with Down's are called Brushfield spots.
- The setting sun sign (white sclera visible between upper eyelid and iris) – Indication of raised ICP or kernicterus
- The pupil – The normal pupil reacts to light, unequal size think 3rd nerve palsy, Horner etc.
- The lens – Congenital cataract
- Fundoscopy – red reflex is normal. White reflex, may indicate retino-blastoma or cataract – refer to Ophthalmologist.



Causes of congenital cataract:

- Congenital rubella
- Toxoplasmosis
- Galactosaemia
- Hereditary

NOSE



- The newborn is an obligate nose
- breather! Any narrowing or obstruction may result in breathing and feeding problems
- In babies with respiratory distress, rather put the NG tube through the mouth
- Nasal hypoplasia refers to a severely flattened nose with virtually no nasal bridge. Causes include warfarin intake by mother in 1st trimester or Conradi

CONRADI SYNDROME

- X-linked dominant
- Growth deficiency
- Flat nasal bridge
- Flat face
- Cataracts
- Short limbs
- Bald spots, flaky skin

- Millia; already discussed
- Choanal atresia; this means that the nasal passages are not patent. 1 or both of the posterior nasal openings may be occluded. Ensure that both nasal passages are patent by listening over each nostril while occluding the other with the baby's mouth closed. If a nasogastric tube can be inserted, it may be assumed that the nostril is patent. Choanal atresia is characerised by noisy breathing, cyanosis and apnoea in the quiet baby (mouth closed) as apposed to the pink colour of the same baby crying with the mouth open.
- Ala nasae; these are the wings of the nose. Nasal flaring is a sign of respiratory distress
- Nasal septum – should be central, may displace with pressure

Nasal discharge, may indicate congenital syphilis

Philtrum; shallow or absent may indicate FAS

Nasolachrymal duct; if blocked present with excessive tearing, may become secondarily infected.

THE MOUTH

LIPS

- Sucking blisters; may be present at birth or within a few days
- Thin upper lip; FAS
- Cleft lip, look for cleft palate
- Fish mouth; corners of mouth inverted common in Potter facies (accompanied renal agenesis)
- Round mouth; found in ichthyosis (tight parchment like skin)

GUMS

- Retention cyst; often bilateral and of little importance. Regresses spontaneously
- Epulis; rare outgrowth of gums, surgical removal is necessary
- Teeth; A baby may be born with natal teeth, these teeth may be left alone until their presence results in problems e.g. tongue lacerations, injury to moms nipple, loose with risk of aspiration

PALATE

- Epsteins pearls; epithelial inclusions that look like small grayish-white papules. They may occur singly or in groups in the midline of the palate, frequently at the junction of the hard and soft palates. They are of no clinical importance
- Cleft palate; Always palpate the palate as a submucosal cleft may be missed otherwise. Look for associated congenital abnormalities. Provide emotional support and discuss further management with parents.

TONGUE

- look for central cyanosis,
- macroglossia (hypothyroidism, Beckwith syndrome, storage diseases and tumours of the tongue. In Downs syndrome the tongue appears large due to the small mouth
- Fasciculations of the tongue = LMN lesion
- Frenulum of the tongue; if severely thickened with functional impairment consider surgery

ORAL CAVITY

- Milk crusting; easily removed, no pinpoint bleeding
- Thrush (Candida Albicans); small white spots on tongue, gums, palate and buccal mucosa, red inflamed mucosa
- Drooling; a newborn who drools continuously should have a high gastro-intestinal obstruction such as oesophageal atresia excluded. The baby with respiratory distress who constantly foams at the mouth probably has a trachea-oesophageal fistula

JAW

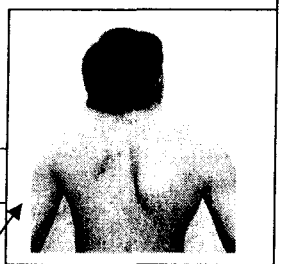
- Micrognathia; May form part of Pierre Robin syndrome. Feeding and breathing problems may result
- Skew jaw; due to intrauterine compression, should resolve within 1st few months

EXAMINATION OF THE NECK

- Excessive skin folds may be present in Downs
- Skew neck; intrauterine compression, resolves spontaneously
- Webbed neck (Turner, Noonan, Klippel-Feil#)
- Midline swellings (enlarged thyroid, thyro-glossal cyst, dermoid cyst)
- Laterally located swellings (cystic hygroma, branchial cyst, sternomastoid swelling)
- Trachea; difficult to palpate in newborn. Displacement as for adults
- Mobility of neck; neck is normally very mobile, stiff neck may indicate fusion of cervical vertebrae as in Klippel Feil syndrome
- Clavicles; absent or hypoplastic (cleidocranial dysostosis), exclude fractures.

NEONATAL TORTICOLLIS = sternomastoid 'tumour'

- These neonates present with a torticollis within the first few weeks after birth (rarely at birth). On examination, a firm mass may be palpable on the side of the tilt in the SCM muscle.
- The aetiology is unknown but may have been caused by trauma sustained intrauterine or at birth.
- Confirm the diagnosis and exclude secondary causes (congenital cervical anomalies, ocular disorders, cervical adenitis, acute fascitits)
- Prevent the condition from worsening as neglect could result in plagiocephaly, hemiatrophy of the face and diplopia.
- Management includes physio. It is recommended that the baby be carried on the arm in the 'anti-reflux' position with the tumor on the side of the forearm because in doing so the affected muscle is continuously stretched
- 80-85% resolve in 18months
- Surgery indicated for the rest (tenotomy)



THYROGLOSSAL CYST

- Unilocular, midline, subplatysmal, clear, transilluminable cyst, communicates with caecum of tongue
- Originates along the embryonal course of the thyroid (from the foramen caecum in the tongue up to the pretracheal position of the thyroid
- Moves when the tongue is stuck out
- Complications; infection, fistuli, scars
- Surgery is indicated

DERMOID CYST

- Developemental cyst at lines of fusion
- Firm round mass at lateral eyebrow region (angular dermoid), bridge of nose or in the neck or genital regions.
- Dermoid cysts contain mature ectodermal structures other than bone or nerve. They are filled with keratinous material.
- If in midline (e.g. bridge of nose, always exclude intracranial extension)
- Angular dermoids may be removed by direct excision – beware the frontal branch of the facial nerve!!!!!

#KLIPPEL – FEIL

- characterized by the congenital fusion of any 2 of the 7 cervical vertebrae
- short neck
- low hairline
- restricted mobility of upper spine
- other congenital abnormalities e.g. scoliosis, spina bifida etc.
- management includes surgery by orthopaedic surgeon

The 18th Dynasty Egyptian pharaoh Tutankhamun is believed by some to have suffered from Klippel–Feil syndrome

EXAMINATION OF THE CHEST

INSPECTION

SIZE – The chest circumference of the term baby is approximately the same or slightly smaller than the head circumference, where as the preterm infant has a proportionally larger head.

SHAPE

- Excessive intrauterine pressure as in oligohydramnios
- Asphyxiating thoracic dystrophy; this is characterized by short broad ribs and a rigid chest. These babies usually present with respiratory distress.
- Absence of the pectoralis major muscle = **Poland syndrome**
- Pectus excavatum; indentation of the sternum and may be associated with other conditions e.g. **Pierre Robin syndrome.**
- Prominent chest; diaphragmatic hernia,

BREAST TISSUE

- Normally situated in the mid-clavicular line
- Additional nipple; small pigmented area or a perfect miniature nipple in mammary line.
- Wide spaced nipples = Turner syndrome
- Used to determine gestational age (discussed later)
- Breast enlargement; physiological (mothers oestrogen), mastitis, abscess

PIERRE ROBIN SYNDROME

- Not a single gene defect, rather a chain of congenital malformations
- Micrognathia
- Glossoptosis
- Upper airway obstruction
- Cleft lip/palate

- Cyanosis – see cardiology section
- Nasal flaring, nasal passages
- Pattern of respiration
- Respiratory rate
- Retraction

} see resp

PALPATION

Of limited value, palpate trachea.

PERCUSSION

See resp section

AUSCULTATION

See resp section

moves outwards.

PARADOXICAL BREATHING

As a result of diaphragmatic paralysis. During inspiration the paralyzed side of the diaphragm moves upwards instead of downwards and the normal accompanying bulging of the abdomen during inspiration is not noted.

PERIODIC BREATHING

During active REM sleep, periodic breathing is often observed. Characteristics of periodic breathing include;

- Periods of regular breathing between 30 to 40 times per minute
- Periods of regular breathing alternating with periods of absent breathing efforts lasting 5-10 seconds
- Periods of more rapid respiration between 50-60 times per minute lasting about 10-15 seconds.

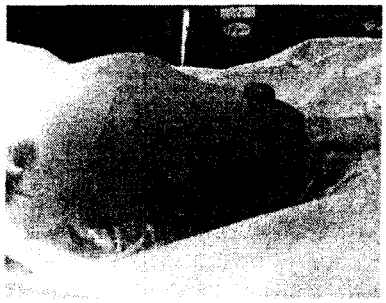
This pattern of breathing is found in REM sleep as well as in premature babies. You must differentiate apnoea from periodic breathing (as apnoea needs immediate intervention) – see table

EXAMINATION OF THE HEART – SEE CARDIOLOGY SECTION

ABDOMEN

INSPECTION

- SIZE – Abdo circum = 1cm less than chest circumference
- SHAPE – Usually prominent (poorly developed abdominal muscles, abdominal bladder, large liver)
  - ABDOMINAL DISTENTION:
    - Excessive air (bowel obstruction, NEC, ileus, bowel perforation etc.)
    - Organomegaly (liver, spleen, kidney)
    - Ascites (Rh incompatibility, renal abnormalities, obstructive uropathies, intestinal obstruction, cardiac failure, etc.)
  - SCAPHOID ABDOMEN (consider diaphragmatic hernia)
  - PRUNE BELLY SYNDROME
  - BOWEL PERISTALSIS (clearly visible peristalsis may suggest pyloric stenosis)
- SKIN -
  - TRANSPARENCY AND ABDOMINAL VEINS. The more preterm the baby, the thinner and more transparent is the skin. The abdominal veins are easily visible. The normal direction of blood flow is away from the umbilicus
  - SKIN LESIONS (discussed elsewhere)



PRUNE BELLY SYNDROME

- = Eagle Barret syndrome
- Absent abdominal wall muscles
- Hydronephrosis/hydroureter
- Undescended testicles

THE UMBILICUS

UMBILICAL STUMP: The stump consists of two parts; the funicular (the skin portion at the base of the umbilicus) and the amniotic parts (bloodless structure, jelly-like, with a blue-white glistening appearance for the first few hours after birth). The amniotic part subsequently dries and changes into a dull brown-black structure. A sharp line of demarcation develops between the skin and amniotic part. Mummification is followed by separation of the two parts usually between 5-10 days after birth. The area of separation has a wet granulating surface for a few days until it heals fully. Contraction of the disappearing umbilical vessels results in the funicular part of the umbilical stump invaginating to form the naval. Sometimes the umbilical stump is so long that full invagination and involution does not occur. This is of no medical importance.

Normal cord – 2 arteries, 1 vein (in case of 1 artery only look for congenital abnormalities)
Thick cord – associated with DM
Thin cord – Post maturity
Green cord – Meconium stained
Omphalitis – Signs (moist, pus, offensive smell, inflamed)
Moist umbilicus post separation; diff

- UMBILICAL GRANULOMA – Reddish brown growth at the base of the umbilicus. Excessive granulation tissue, usually following omphalitis. Treatment; silver nitrate stick (protect surrounding skin with Vaseline)
- UMBILICAL POLYP – This is a mucosal remnant of the omphalo-mesenteric duct. It is visible as a bright red moist nodule that secretes mucus. Excision is required.
- PATENT URACHUS – the urachus is the embryological connection between the bladder and the umbilicus. A patent urachus causes urine to leak from the umbilicus
- PATENT VITELLO-INTESTINAL DUCT – The persistence of the embryonic connection between the yolk sac and the small intestine, the patent vitello-intestinal duct, presents with meconeum/faeces at the umbilicus.

- ABNORMALITIES OF THE ABDOMINAL WALL
  - UMBILICAL HERNIA – More common in black, preterm, Downs, hypothyroidism and Beckwith syndrome babies. Usually present at birth and becomes more obvious during crying. Usually a soft, reducible swelling and is covered with skin and peritoneum. The size of the hernia may differ widely, depending on the hernia orifice which may vary from 1-5cm. The hernia sac contains omentum with or without bowel loops. The prognosis of an umbilical hernia is usually very good and most resolve spontaneously towards the end of the first year of life or shortly thereafter. Surgery should be considered if the possibility exists that the hernia may become strangulatd or if it does not regress within 3 years.

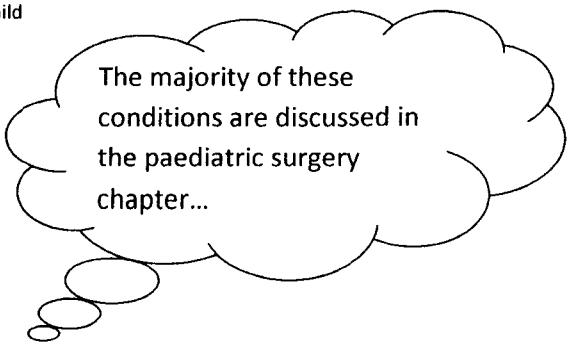
- EXOMPHALOS = OMPHALOCOELE – Herniation of the abdominal contents through the abdominal muscles into the base of the umbilical cord. The wall of the hernia sac consists only of a layer of amniotic membrane without any skin coverage. An exomphalos develops as a result of incomplete closure of the fetal abdominal rectus muscles, with the result that the midgut remains in the umbilical cord instead of in the abdomen. **This condition is associated with other congenital abnormalities.**
- GASTROSCHISIS – The abdominal contents herniated through a para-umbilical defect in the abdominal wall. No hernia sac is present. The bowel loops protruding through the abdominal wall lie uncovered outside the abdomen.
- DIVARICATION OF THE RECTUS SHEATH – This is an elongated bulging of the abdomen in the midline between the umbilicus and the xiphisternum. It is easily noticed when the baby tenses its abdominal muscles e.g. when crying. The newborn baby's abdominal rectus muscles are not properly developed at this stage, hence the resultant bulging with increased abdominal pressure. It is completely normal at this stage and disappears in time.

PALPATION

Basically as for older kids. Liver palpable 2cm below costal margin, spleen may normally be palpable. The kidneys may also be palpable in the first few days of life. The left kidney is often more easily palpable than the right kidney. The bladder is an abdominal organ in the newborn. A bladder which is palpable directly after micturition or remains persistently palpable needs to be investigated.

PERCUSSION AND AUSCULTATION –same as older child

THE NAPPY AREA



INGUINAL AREA

- Inguinal lymph nodes
- Femoral pulses
- Possible inguinal hernia
- Undescended testicle

INGUINAL HERNIA - Actively exclude inguinal hernias, especially in preterm boys. These are almost always indirect and may be uni- or bilateral. Pathology: The pathology is a patent processus vaginalis between the internal opening of the inguinal canal and the scrotum. Inguinal hernias should be repaired as soon as possible (first elective list) as there is a danger of incarceration

THE SKIN

- Mongolian spot – discussed elsewhere
- Moniliasis – Usually secondary to thrush in the mouth
- Ammoniacal dermatitis – Usually found in the baby who is already a few weeks old. Some intestinal bacteria can change the urinary urea in the wet nappy to ammonia. This irritates the nappy area and results in dermatitis. The rash is red and erythematous, and ulcerative lesions may develop in the affected parts. The skin folds are usually not involved.
- Contact dermatitis – This erythematous rash is found particularly over the buttocks, peri-anal area, proximal thighs and lower abdomen. It does not usually involve the skin folds. The rash may be caused by the irritating effect of washing powders, soaps and certain topical medications.
- Congenital syphilis – A macular copper coloured rash may be present on, among others, the buttocks (discussed elsewhere)
- Seborrhoeic dermatitis - The whole nappy area including the skin folds may manifest a dry, sometimes oily erythematous flaking. The scalp, neck, ears and chest are also frequently affected. Treatment is required with antifungal, keratolytic, anti-inflammatory and/or corticosteroid therapy.

GENITALIA

- Gender and intersex – discussed in endocrine chapter
- Gestational characteristics – discussed later in this chapter
- Abnormalities (e.g. hypospadia etc.)
- Discharges – a white vaginal discharge is often observed during the first few days of life. It contains cells and secretions from the vagina and uterus. It resolves spontaneously and no treatment is required.
- Micturition – virtually all babies, boys or girls will pass urine within the first 24 hours of life. If this does not happen, you should be concerned and investigate further. The baby should be able to pass urine comfortably with a good stream. In contrast, sustained dribbling of urine is of concern and may indicate posterior urethral valves or a spinal cord lesion e.g. meningomyelocoele.

- Anorectal atresia – In this type, the anus and parts of the rectum are absent. Associated fistulas through which meconium is passed to the bladder, urethra, vagina and the perineum are occasionally found.
- Anal sphincter tone – A myelomeningocoele may give rise to a lowered anal sphincter tone with the appearance of a gaping or wide-open anus. Associated abnormal function of the bladder sphincter may occur with dribbling of urine.
- Anal skin tags – these usually shrink within weeks.
- Rectal examination – Not routinely done!!

THE STOOLS

- Normal – most newborns pass meconium within 24 hours after birth
- Meconium – dark black-green, tar-like. Passed in first 2-3 days of life. Odorless and sterile.
- Transitional stools – Meconium stools are followed by these stools which are softer and yellow green-brown. These contain mucous and remnants of milk.
- Milk stools – Normal milk stools appear by the third to fourth day of life. Their characteristics depend on the type of feed that the baby receives. Breast-fed babies stools appear bright yellow, occasionally watery, odorless, slightly sour. Artificially fed babies stools are light yellow to grey-green, firmer with a more offensive smell.
- Bloody stools – this is always a cause for concern. Neonatal causes include NEC, bleeding diathesis, intestinal infections and intestinal pathology
- Pale stools – Obstructive jaundice – see surgical conditions
- Hunger stools – These appear as dry, green mucoid stains or stools on the nappy. It is an indication that the baby is receiving inadequate food.
- Steatorrhoea – Fat malabsorption. Exclude cystic fibrosis
- Diarrhoea – Infective/non-infective
- Constipation – hypothyroidism, dehydration etc. Breast fed infants usually do not get constipated.

THE LIMBS...

MOVEMENT

Asymmetry may indicate injury to a particular limb. Especially take note of this when testing the Moro reflex..

POSITION

- Determined by the gestational age
- Term: fully flexed
- Preterm: incomplete flexion
- Must be symmetrical
- Decerebrate: Arms and legs in extension and turned inward with the hands fisted. This indicates brainstem pathology

SWELLING (localized or diffuse)

- Tissued drip
- Oedema
- Haematoma with or without underlying fracture
- Possible infection

KLIPPEL-TRENAUMAY SYNDROME

- Hemihypertrophy
- AV fistuli
- Cutaneous haemangioma

IN ENTATION

A result of amniotic bands. Auto-utation may occur in severe cases

LE GTH

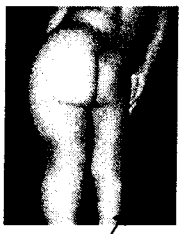
Compared to the older child an adult, the newborn baby has a relatively la head and short limbs.

GENERAL EXAMINATION OF THE LIMBS

- Position
- Movement
- Swelling
- Difference in size
- Indentation
- Length
- Completeness

HEMIMELIA

HEMIHYPERTROPHY



SIZE – always compare the size of limbs

- Limb enlargement; causes:
  - Neurofibromatosis
  - Lymphangiomata
  - Haemangiomata
  - Wilms tumors (may be associated with hemihypertrophy)
  - Klippel-Trenaumay syndrome

Pl COMELIA

INCOMPLETE LIMBS

- Ectromelia – absence of entire limb
- Hemimelia – this indicates that the distant part of the limb is absent
- Phocomelia – the hands and feet are attached to the trunk with a short arm or leg stump.

THE UPPER LIMBS

- Position and movement
- Clavicle
- Joints
- Swellings
- Skin
- Hands
- Palms
- Pulses

POSITION AND MOVEMENT

ASYMMETRICAL LIMBS

Fractures, dislocation, septic arthritis, osteitis, periostitis of congenital syphilis and nerve injury

- Erbs palsy 'waiters tip position' –
- Klumpke's palsy – injury to lower nerves of brachial plexus. Very rare! The palmar grasp reflex disappears and the hand adopts the claw-hand position

CLAVICLE – absent or hypoplastic, think cleidocranial dysostosis

JOINTS - look for contractures and signs of septic arthritis

THE PALM

- Simian crease (single palmer crease) is associated with Down's syndrome but may be normal!
- Peeling palmar skin lesions usually indicate congenital syphilis. The same is said for the soles of the feet

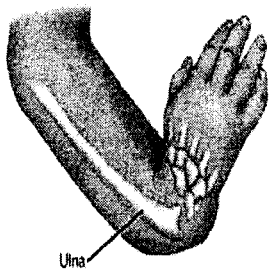
THE PULSES – The femoral pulses in the newborn should be easily palpable. If the brachial and even the radial arterial pulses are easily palpable and bounding a PDA must be considered.

THE HANDS

- Colour – look for peripheral caynosis
- Oedema – must be investigated
- Position – fisting may indicate CP
  - Radial club hand – associated with Fanconi's, VATER syndrome and haematoligical abnormalities
  - Lobster claw hand – AD inheritance

ERB'S PALSY

RADIAL CLUB HAND



Radial clubhand; note absence of radius

LOBSTER CLAW HAND

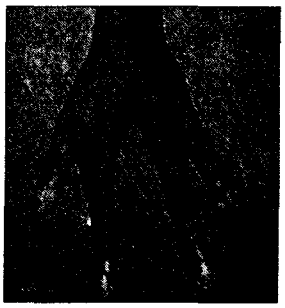


Figure 1: Lobster claw deformity of left hand

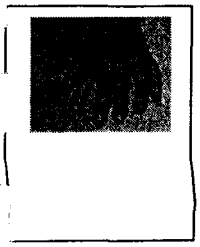
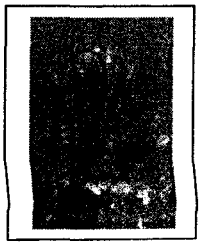
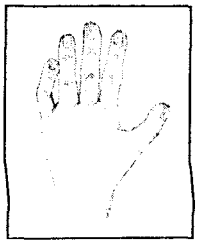
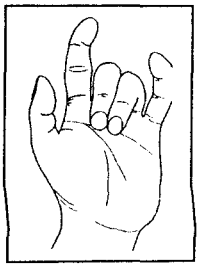
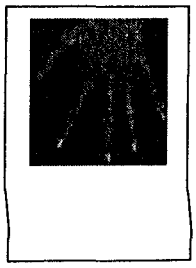
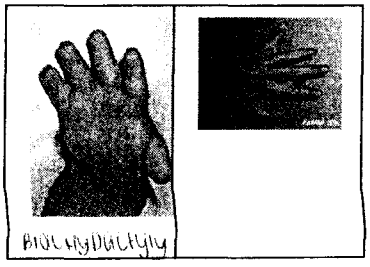
VATER SYNDROME/ VACTERL...

- Vetebral anomalies
- Anal anomalies
- Cardiac
- Trachea
- Esophagus
- Renal and radius
- Limbs (the radius may be absent)

FANCONI'S syndrome

- disease of the proximal renal tubules
- glucose, amino acids, uric acid, phosphate and bicarbonate are passed into the urine, instead of being reabsorbed

- Nails – In the term baby the nails reach the fingertips. The post-term baby's nails are often longer than the fingertips and the baby may scratch itself. Preterm babies nails are usually shorter and do not reach the fingertips. Nail hypoplasia may indicate fetal hydantoin syndrome or Turner's syndrome
- Fingers
  - Polydactyly – this refers to more than 5 fingers per hand. The extra finger is usually a rudimentary tag with a thin pedicle attached to the little finger. The thin pedicle can be ligated using black silk. If the pedicle has a broad base or contains a bone it must be surgically removed.
  - Syndactyly – this refers to the partial or complete fusion of two or more fingers
  - Clinodactyly – this refers to incurving of the little finger and is often present in Down's
  - Camptodactyly – this is a flexion deformity of the finger. The fingers curve over one another. It is found in Trisomy 13 and 8.
  - Arachnodactyly – this refers to long fingers as in Marfans
  - Brachydactyly – this refers to short fingers as in Downs and others
  - Webbed fingers – this refers to two or more fingers being partially attached to one another by a skin fold. It may be found in among others, Poland syndrome (which is characterized by the absence of one pectoralis muscle).
  - Amniotic band syndrome – in this syndrome indentation or even amputation of the arms, hands and legs may occur. It may even look as if a thread of cotton has been wound around the fingers

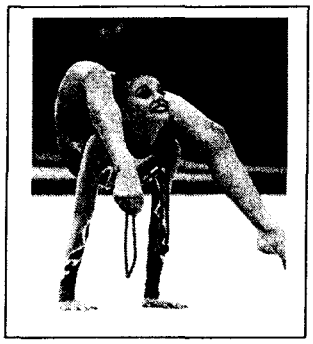







THE LOWER LIMBS

- Position and movement
  - Normal – flexion
  - Frog position – premature babies and babies with decreased muscle tone
  - Breech position – see picture!
- Shape
  - The newborns legs often appear slightly bowed and rotated outwards (normal).
  - Abnormal bowing of the legs, such as posterior bending of the tibia, may be due to intra-uterine pressure
  - Conditions such as osteogenesis imperfecta and certain types of dwarfism may also cause abnormally bent legs
- Joints
  - The knee; genu recurvatum (hyperextension of the knee). This usually indicates intra-uterine breech with extended legs. Dislocation of the knee may occur which requires further orthopaedic attention
  - The hips; always examine the newborns hips for congenital hip dysplasia using either Barlow's or Ortolani's tests (see orthopaedic section)
- Skin – discussed elsewhere
- Pulses – discussed elsewhere
- Feet
  - Study orthopaedic chapter
  - Sandal gap – associated with Down's but may be normal
  - Plantar skin creases – In the Down's baby a prominent deep plantar skin crease is often observed opposite the first toe space. Horizontal creases increase with maturity (being almost absent in the premature baby).
    - Skin lesions on the foot-soles often suggest congenital syphilis

EXAMINATION OF THE BACK

- Curvature
- The overlying skin
- Spina Bifida
- Sinus or Dimple



CURVATURE

At birth the spinal column of the newborn appears C-shaped with no fixed curves. At approximately **3 months** of age, when the baby develops head control, the normal cervical curve of the spinal column develops. The normal lumbar curve develops at approximately **12 months** of age when the baby begins to walk

Abnormal curvature: Look for kyphosis and scoliosis (discussed in orthopaedic chapter)

SPINAL DYSRAPHISM *(10% of life births!!!)*

- Includes a variety of congenital defects of the spinal cord as a result of abnormal closure of the neural tube

SPINA BIFIDA CYSTICA/APERTA

- A noticeable lesion occurs, usually in the dorso-lumbar area

  - Meningocoele: The cyst contains CSF only. The surgery is relatively simple and the prognosis is good. The patients are usually neurologically intact. If however it is leaking CSF, urgent surgery (within 24hrs) is indicated to prevent infection.
  - Myelocoele: The contents of the sac contains neuroneal elements
  - Rachischisis/Aperta: The whole spinal cord is folded open on the skin surface and no dural covering exists.

The mother presents with polyhydramnios and Increased AFP, twice the average for gestational age. The lesion may be demonstrated early in pregnancy with ultrasound.

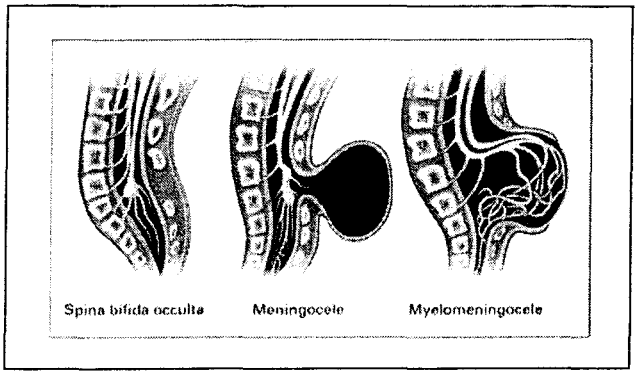
The degree of neurological fallout depends on the height of the lesion; almost all are incontinent with regards to urine and faeces. Approximately 90% have hydrocephalus and need a VP shunt. Urological and orthopaedic problems need long-term follow-up.

SPINA BIFIDA OCCULTA

In this group there are subtle cutaneous stigmata indicative of an underlying malformation of the spinal cord or cauda equine. If recognized early and treated accordingly, these patients, who often are initially neurologically intact or who have only slight neurological deficit, can be saved from progressive neurological deterioration.

Cutaneous stigmata include:

- Sub-cutaneous lipoma over the vertebral column
- A tuft of hair
- Area of pigmentation
- Vascular stain
- Dermal sinus over the vertebral column



Lesions occurring in this group include:

- DIASTTEMATOMYELIA: It is a split spinal cord caused by a bony spur which develops from the posterior aspect of the vertebral body. Neurological fallout increases as the skeleton grows and the cord gives the impression of moving upwards.
- LIPOMYELOMENINGOCOELE: Is associated with a sub-cutaneous lipoma which stretches through a defect in the dura and implants on the dorsal surface of the cord or in between the roots of the cauda equine.
- DERMAL SINUS: A tract stretches from a sinus on the skin surface to the cord. It may be responsible for repeated attacks of meningitis and may occur on bony seams from the cranium to the sacrum near the midline. It may be associated with an intradural dermoid tumour.
- SHORT FILUM TERMINALE: May be isolated or associated with any of the above-mentioned conditions. Cutaneous stigmata are always present. Patient presents with neurological fallout (even enuresis) as the child grows taller and the cord is stretched. This may also occur post myelominingocoele repair.

FOR COMPLETE NEUROLOGICAL EXAMINATION OF THE NEWBORN SEE THE NEUROLOGY SECTION.

DETERMINATION OF GESTATIONAL AGE:

RELIABLE DETERMINATION OF GESTATIONAL AGE IS ESSENTIAL BECAUSE:

- It helps to determine if the baby is term, pre-term or post-term
- It helps to determine if the baby is small, normal or large for dates
- Depending on the category the baby falls under, specific complications can be anticipated
- In the preterm baby the initial accurate gestation determination is essential for its corrected age, which is important for both the neurological and physical follow-up evaluations during the first two years of life. Corrected age is the chronological age minus the number of weeks preterm that the baby is born before 38 weeks gestation.

ANTENATAL DETERMINATION OF GESTATIONAL AGE

- LNMP
- EUS

POSTNATAL DETERMINATION OF GESTATIONAL AGE

THE NEW BALLARD METHOD OF GESTATIONAL AGE DETERMINATION

The new Ballard method of gestational age determination is an expanded and refined version of the former Ballard method. The new method makes allowance for the reliable gestational assessment of newborn babies with a gestation from 20 weeks onwards. It depends on the evaluation of 6 neuromuscular and 6 external physical criteria.

TECHNIQUE

The determination should preferably be done within 96 hours (and in the case of the <1kg baby within 12 hours) after birth in the quiet, non-crying baby in the supine position and with the clothes removed. Use the maturational score forms in which specific attributes of maturity are awarded points in each of the twelve categories. With the assistance of the accompanying table the combined maturational score is used to calculate the corresponding gestational age.

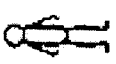
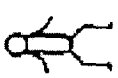
EXTERNAL CRITERIA:

- SKIN: Note and palpate the skin of the abdomen in the non-crying baby between your thumb and index finger for texture, colour and transparency
- LANUGO: Note the presence and appearance of lanugo on the back
- FOOT-SOLE: Measure the length of the foot from the heel to the tip of the toe. Note the appearance and distribution of the skin creases from the toes to the heel with the sole stretched.
- BREASTS: Note the development of the nipple and areola. Pick up the breast tissue between your thumb and forefinger and determine the diameter.
- EYE/EAR: Note whether the eyelids have already separated. Inspect, palpate and fold the pinna over. Note the shape, curvature, amount of cartilage and the recoil of the pinna after folding.
- GENITALIA: Boy; inspect and palpate the scrotum for transverse folds and testes
  Girl; inspect the clitoris, labia majora and minora for size and appearance with the legs in abduction

NEUROMUSCULAR CHARACTERISTICS

- POSTURE: Evaluate the extent of limb flexion and extension in the supine position
- SQUARE WINDOW: Flex the wrist between your thumb and index finger. Determine the angle formed between the hypothenar eminence of the hand and the ventral forearm
- ARM RECOIL: Flex the arms at the elbows for five seconds. Then extend the arms fully by pulling on the hands. Let go suddenly and note to what extent the arms flex.
- POPLITEAL ANGLE: With the baby on its back and the pelvis flat on the examination surface, hold the thigh in the knee-chest position with your index finger while your thumb supports the knee. Straighten the leg now as far as possible by pressing lightly on the top of the ankle with the other index finger. Measure the popliteal angle of the knee.
- SCARF SIGN: Take the baby's hand and attempt to place it around the neck with the hand as far posteriorly as possible over the opposite shoulder. Assist the movement by pushing the elbow over the body. Note to what extent the elbow moves over the body.
- HEEL-TO-EAR TEST: Attempt to pull the foot as close as possible to the head without forcing it. The knee may slide down next to the body. Note the distance between the foot and the head, and the degree of knee extension.

Neuromuscular Maturity

	-1	0	1	2	3	4	5
Posture							
Square Window (wrist)	 >80°	 90°	 60°	 45°	 30°	 0°	
Arm Recoil		 180°	 140°-180°	 110° 140°	 90-110°	 <90°	
Popliteal Angle	 180°	 160°	 140°	 120°	 100°	 90°	 <90°
Scarf Sign							
Heel to Ear							

Physical Maturity

Skin	sticky friable transparent	gelatinous red, translucent	smooth pink, visible veins	superficial peeling &/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 40-50mm -1 <40mm -2	>50mm no crease	faint red marks	anterior transverse crease only	creases ant. 2/3	creases over entire sole	
Breast	imperceptible	barely perceptible	flat areola no bud	stippled areola 1-2mm bud	raised areola 3-4mm bud	full areola 5-10mm bud	
Eye/Ear	lids fused loosely-1 tightly-2	lids open pinna flat stays folded	sl. curved pinna; soft; slow recoil	well-curved pinna; soft but ready recoil	formed & 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	prominent clitoris small labia minora	prominent clitoris enlarging minora	majora & minora equally prominent	majora large minora small	majora cover clitoris & minora	

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

3. NEONATAL INFECTIONS – please study in conjunction with chapter on infectious diseases!

The reported prevalence of infection of the newborn varies between 7 and 30% Factors which predispose to infection include:

- Maternal infection
- LBW
- Obstetric or resuscitation procedures
- Anatomic: long cord stump, delicate or cracked skin
- Immature host defense mechanisms
- Nursery environment: crowding, understaffing, poor hand washing

Amniotic fluid infection syndrome (chorioamnionitis) is a common condition which is particularly prevalent in poor socio-economic conditions and is associated with preterm labour and prelabour rupture of membranes.

SUPERFICIAL INFECTIONS

- Common sites; skin, umbilicus, eye, mouth, perineum
- Usually treated with local antibiotics
- Oral thrush – treat with nystatin suspension
- Perineal candida – nystatin cream

SEPTICAEMIA

The diagnosis of infection may be very difficult in the newborn because of subtle and non-specific presentations. Suspect septicaemia if 3 or more of the following are present

- Any predisposing factors
- Unstable temperature
- Lethargy
- Poor colour
- Apnoea
- Feeding difficulties
- Vomiting
- Abdominal distension
- Sclerema
- Superficial sepsis

A combination of the following signs is strongly indicative of serious infection:

- Purpura
- Anaemia
- Jaundice
- Hepatomegaly
- Splenomegaly
- Full fontanelle
- Swollen joint

4. FAILURE TO THRIVE

This implies that the baby fails to gain weight adequately. Following initial weight loss for a few days, the healthy term baby should regain birth weight by the 10th day and the preterm baby by the 14th day. Thereafter expected weight gain in a term infant is a minimum of 10g per week.

CAUSES:

- Incorrect feeding (breastfeeding difficulties/inadequate formula)
- Metabolic acidosis may develop in artificially fed healthy preterm babies, particularly during rapid growth when hydrogen ions are produced which may not be excreted by the immature kidney. This may lead to weight loss
- Anaemia
- Occult infections
- Subclinical cold stress (increased calorie demand from raised metabolism)
- Congenital heart lesion, GIT anomalies, chronic chest conditions, metabolic diseases, endocrine disorders and brain damage.
- Psychological stress in baby (e.g. mom with depression)

VOMITING

- Very common in first few days of life
- Regurgitation of the first few feeds is not serious (usually the result of irritation of the gastric mucosa by swallowed blood or meconium)
- May be the first sign of serious disease, especially if associated with other symptoms
- Meningitis, septicaemia, UTI and NEC must be considered
- Intestinal obstruction must be excluded
- Intracranial injury, congenital abnormalities and infection may also cause persistent vomiting
- Rare causes include metabolic problems (discussed elsewhere)

ABDOMINAL DISTENTION

- Common physical sign in the newborn
- May however be a sign of serious illness
  - Gas (accumulation of air from obstruction or ileus)
  - NEC
  - Meconium ileus (this is a pointer to cystic fibrosis)
  - Paralytic ileus (result of hypoxia, shock, electrolyte imbalance or infection)
  - Abdominal masses
  - Ascites (cardiac failure, nephritic syndrome)
  - Sepsis
- Exclude surgical causes (always consult with paediatric surgeon), pass a NG tube and put up an IV line

NECROTIZING ENTEROCOLITIS (NEC)

- intestinal inflammation associated with focal or diffuse ulceration and necrosis primarily affecting terminal ileum and colon
- affects 1-5% of all newborns admitted to ICU

Etiology

- multifactorial associations
  - prematurity —> immature defenses
  - perinatal asphyxia leading to bowel ischemia
  - introduction of formula/breast milk provides substrate for bacterial overgrowth
  - bacterial invasion of bowel wall with gas production (pneumatosis intestinalis)
  - infection: C. *difficile* toxin, coagulase negative Staph in NICU
  - tissue necrosis and perforation results

Clinical Features

- distended abdomen and signs of obstruction (vomiting)
- increased amount + bile stained gastric aspirate/vomitus
- frank or occult blood in stool
- feeding intolerance
- diminished bowel sounds
- signs of bowel perforation - sepsis, shock, peritonitis, DIC

Investigations

- abdominal x-ray: intramural air ("train tracks"), free air, fixed loops, thickened bowel wall
- high WBC, low platelets, electrolyte imbalances, acidosis, hypoxia, hypercarbia

Treatment

- NPO, vigorous IV fluid resuscitation, NG decompression
- TPN
- antibiotics for infection (triple therapy given empirically)
- serial abdominal x-rays detect early perforation
- surgical resection of necrotic bowel and surgery for complications (e.g. perforation, strictures)

STUDY THIS PAGE IN CONJUNCTION WITH THE GIT CHAPTER...

5. GASTRO-INTESTINAL DISORDERS OF THE NEWBORN

DIARRHOEA

- Usually dietary in origin
- Lactose intolerance (self limiting)
- Infective (see GIT chapter)

Treatment:

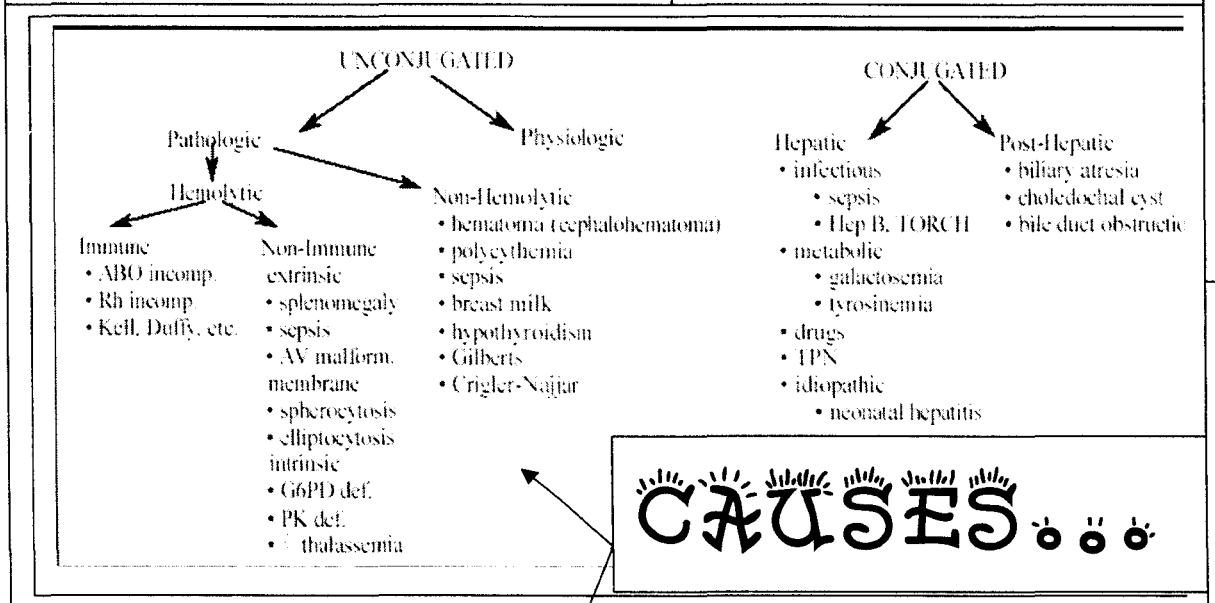
- Fluid replacement and electrolyte balance is critical in the newborn
- Mild: ORS
- Severe: Crystalloid at 20ml/kg for rapid correction
- Calculate fluid requirements
- Antibiotics for very ill babies
- Supportive measures

6. JAUNDICE IN THE NEWBORN

JAUNDICE...

- very common - 50% of term newborns develop visible jaundice
- jaundice visible at serum bilirubin levels of 85-120 umol/L look at sclera, mucous membranes, palmar creases, tip of nose
- jaundice more severe/prolonged (due to increased retention of bilirubin in the circulation) if following factors present
  - prematurity
  - acidosis
  - hypoalbuminemia
  - dehydration

JAUNDICE APPEARING IN THE 1ST 24HRS IS NEVER PHYSIOLOGICAL and strongly suggests excessive haemolysis or sepsis



GOLDEN RULES OF PHYSIOLOGICAL JAUNDICE

- Not apparent in 1st 24hrs
- Infant remains well
- S-bilirubin does not reach treatment values
- Jaundice must fade by 14days
- pathophysiology
  - increased hematocrit and decreased RBC lifespan
  - immature glucuronyl transferase enzyme system (slow conjugation of bilirubin)
  - increased enterohepatic circulation
- term infants: onset day 2-3 of life, resolution by day 7 of life
- premature infants: higher peak and longer duration
- risk factors
  - polycythemia
  - prematurity
  - infant of diabetic mother (IDM)
  - ethnic group (i.e. Native)
  - cephalohematoma
  - breast feeding

Breast Feeding Jaundice

- common
- due to lack of milk production and subsequent dehydration

Breast Milk Jaundice

- rare (1 in 200 breast-fed infants)
- due to substance in breast milk that inhibits glucuronyl transfersase
- onset day 4 to 7 of life, peak at 2nd to 3rd week of life

<24 hours	24-72 hours	72-96 hours	Prolonged (>1 week)
ALWAYS PATHOLOGICAL • Haemolysis (Rh or ABO) • Sepsis (GBS, congenital infections)	• Physiological • Polycythaemia • Dehydration • Hemolytic - G6PD deficiency - Pyruvate kinase deficiency - Spherocytosis - Large haematoma • Sepsis/congenital infection	• Physiological • Breast feeding jaundice • Sepsis	• Breast milk jaundice • Prolonged physiologic jaundice in preterm • Hypothyroidism • Neonatal hepatitis • Conjugation dysfunction - Gilbert - Crigler-Najjar • Inborn errors of metabolism • Obstruction e.g. biliary atresia

GILBERT'S DISEASE

- Inherited metabolic disorder
- Prevalence 1-2%!
- Decreased UDP-glucuronosyltransferase activity
- May be diagnosed later in life with fasting or intercurrent illness (also with drinking)
- Benign, no intervention needed.

Rh incompatibility is usually picked up prior to delivery. If not it may cause anemia, jaundice, HSM and cardiac failure at birth or in the 1st 24hrs of life.

ABO incompatibility is the most common cause of isoimmunization in the newborn. Potential ABO incompatibility exits with a type O mother and a type A or B infant. The direct Coombs test is usually positive. Bilirubin levels usually do not rise very high.

CRIGLER-NAJJAR DISEASE

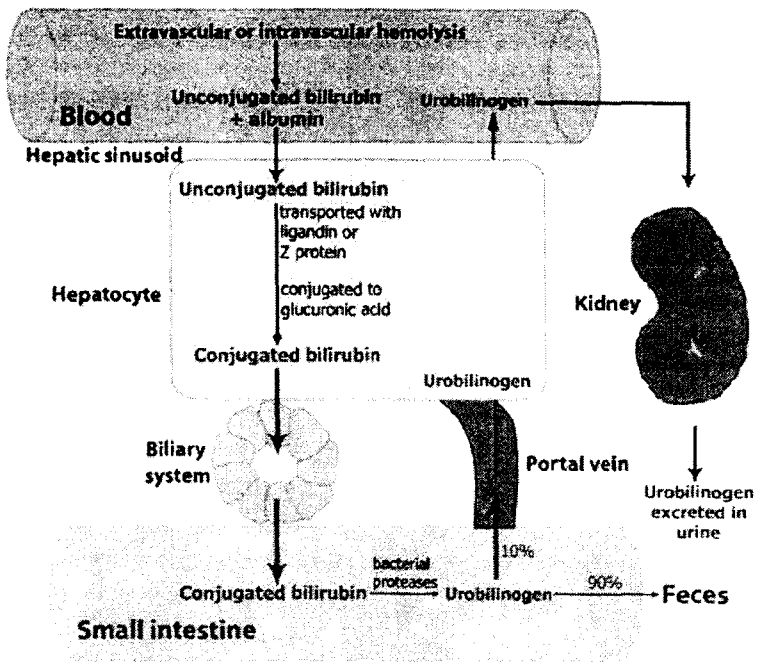
- Inherited disorder presenting in the first days of life with jaundice +/- CNS signs.
- Caused by a mutation resulting in abolition of bilirubin UDP- glucuronosyltransferase
- Tx: Liver transplant before kernicterus develops. Phototherapy whilst awaiting transplant

KERNICTERUS — yellow discolouration of Basal Ganglia.

- unconjugated bilirubin concentrations exceed albumin binding capacity and bilirubin enters and is deposited in the brain resulting in damage
- incidence increases as serum bilirubin levels increase above 20mg/dl
- can occur at lower levels in presence of sepsis, meningitis, hemolysis, hypoxia, hypothermia,hypoglycemia and prematurity
- early manifestations: lethargy, hypotonia, poor feeding, high-pitched cry and emesis
- later signs: bulging fontanel, opisthotonic posturing, pulmonary hemorrhage, fever, hypertonicity, seizures

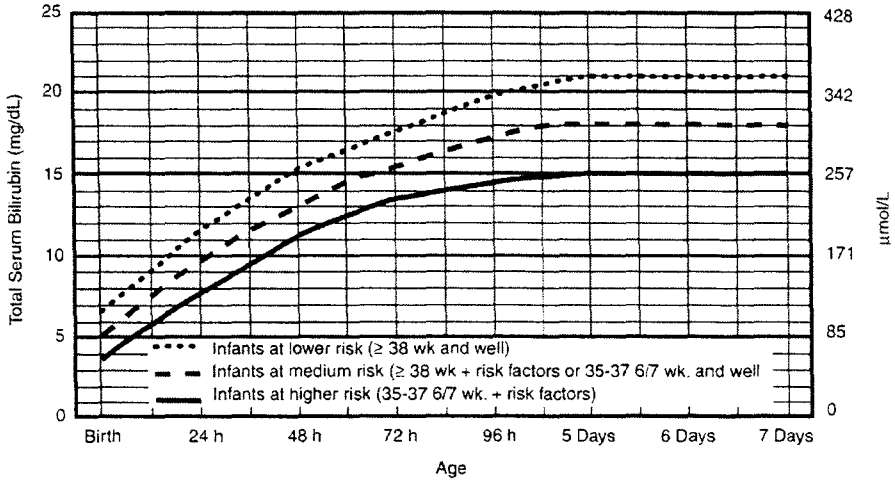
COMPLICATIONS: sensorineural deafness, choreoathetoid cerebral palsy (CP), enamel dysplasia , MR

TREATMENT: exchange transfusion



TREATMENT OF UNCONJUGATED HYPERBILIRUBINEMIA

- Treat to prevent kernicterus
- breast feeding does not need to be discontinued
- treat underlying causes: e.g. sepsis
- phototherapy
  - insoluble unconjugated bilirubin is converted to excretable form via photoisomerization
  - serum bilirubin should be monitored during and immediately after therapy (risk of rebound)
  - contraindicated in conjugated hyperbilirubinemia: results in "bronzed" baby
  - side effects: hypernatremic dehydration, eye damage
- exchange transfusion
  - prevents toxic effects of bilirubin by removal from body
  - indications: depend on level and rate of rise of bilirubin
  - most commonly performed for hemolytic disease



- Use total bilirubin. Do not subtract direct reacting or conjugated bilirubin.
- Risk factors = isoimmune hemolytic disease, G6PD deficiency, asphyxia, significant lethargy, temperature instability, sepsis, acidosis, or albumin < 3.0g/dL (if measured)
- For well infants 35-37 6/7 wk can adjust TSB levels for intervention around the medium risk line. It is an option to intervene at lower TSB levels for infants closer to 35 wks and at higher TSB levels for those closer to 37 6/7 wk.
- It is an option to provide conventional phototherapy in hospital or at home at TSB levels 2-3 mg/dL (35-50mmol/L) below those shown but home phototherapy should not be used in any infant with risk factors.

RESPIRATORY DISTRESS IN THE NEWBORN

RESPIRATORY DISTRESS SYNDROME (RDS)

= HYALINE MEMBRANE DISEASE

Pathophysiology - surfactant deficiency —> poor lung compliance due to high alveolar surface tension and atelectasis —> respiratory distress —> hypoxia + acidosis. Surfactant decreases alveolar surface tension, improves lung compliance and maintains functional residual capacity

Risk Factors
- premature babies: rare at term, risk is inversely proportional to birth weight and GA
- infants of diabetic mothers (IDM): insulin inhibits the cortisol surge necessary for surfactant synthesis
- C-section
- asphyxia, acidosis
- males > females

Clinical Features
- onset within first few hours of life, worsens over next 24-72 hours, with symptoms of respiratory distress. Improvement usual on day 3-7 (increased surfactant)
- infants may develop respiratory failure and require ventilation

CXR: decreased aeration and lung volumes, reticulogranular pattern throughout lung fields with air bronchograms, atelectasis; may resemble pneumonia. The "ground glass" apperance of lungs is pathognomonic of RDS

Prevention
- steroid therapy (60% reduction in RDS) for mothers prior to delivery of premature infants
- monitor lecithin:sphingomyelin (L/S) ratio

Treatment – supportive + surfactant
- O 2 , assist ventilation with PEEP or CPAP, nutrition
- administer fluids cautiously to avoid pulmonary edema

Prognosis - in severe prematurity and/or prolonged ventilation, increased risk of bronchopulmonary dysplasia (BPD)

Complications
- patent ductus arteriosus (PDA)
- bronchopulmonary dysplasia (BPD)
- retinopathy of prematurity
- pulmonary air leaks (pneumothorax)
- intracerebral/intraventricular hemorrhage (ICH/IVH)

CONGENITAL PNEUMONIA - Intrauterine infection (usually GBS or E.coli). Increased risk with PROM. High index of suspicion as baby may die within hours of birth!!! Mx – supportive and antibiotics

DIFFERENTIAL DIAGNOSIS (CHAMPION)

- CARDIAC –*cong heart disease, PPHN*
- HAEMATOLOGICAL – *blood loss, polycythaemia*
- ANATOMICAL – *Tracheo-oesophageal fistula, cong diaghragmatic hernia, upper airway obstruction*
- METABOLIC – *Hypoglycaemia, inborn errors of metabolism*
- PULMONARY – *RDS, TTN, Meconium aspiration, Pleural effusion, pneumothorax, PPHN, cong lung abnormalities*
- INFECTIOUS – *sepsis, pneumonia*
- O O O O OROS!!!
- NEUROLOGICAL – *CNS damagae (trauma, haemorrhage), drug withdrawel syndromes*

MECONIUM ASPIRATION SYNDROME (MAS)

- 10-15% of all infants are meconium stained at birth, ~5% of meconium stained infants get MAS
- usually associated with fetal distress in utero, or post-term infant
- higher incidence of MAS with thick meconium
- respiratory distress within hours of birth
- tachypnea, hypercarbia, small airway obstruction, chemical pneumonitis

CXR: hyperinflation, streaky atelectasis, patchy infiltrates

complications: hypoxemia, acidosis, PPHN, pneumothorax, repiratory failure, death

treatment: supportive care and ventilation, may benefit from surfactant replacement (surfactant function is inhibited by meconium)

prevention: careful in utero monitoring, suction naso/oropharynx at perineum, then intubate and suction below cords at birth

S/I

- FBC, R-GLUC, ABG, B/C
- CXR

PRESENTATION

- RR>60
- HR>160
- Central cyanosis
- Nasal flaring
- Grunting
- Intercostal retractions
- Decreased air entry/crackles on auscultation

PERSISTENT PULMONARY HYPERTENSION (PPHN)

Severe hypoxemia due to persistence of fetal circulation - R to L shunt through PDA, foramen ovale, intrapulmonary channels —> decreased pulmonary blood flow and hypoxemia —> further pulmonary vasoconstriction

Risk factors
- asphyxia, MAS, RDS, sepsis, structural abnormalities (e.g. diaphragmatic hernia)

Treatment
- O 2 given early and tapered slowly, minimize stress and hypoxia
- high frequency oscillation, inotropes (to make systemic pressure greater than pulmonary pressure), alkalinization, extracorporeal membrane oxygenation (ECMO)

TRANSIENT TACHYPNEA OF THE NEWBORN (TTN)

also known as
- "wet lung syndrome"
- respiratory distress syndrome type II

Pathophysiology - delayed resorption of fetal lung fluid —> accumulation of fluid in peribronchial lymphatics and vascular spaces —> tachypnea

Risk Factors
- full term or slightly premature infant
- no labour/short labour (?lack of catecholamine release)
- C-section (lungs are not compressed during passage through pelvic floor)

Clinical Features
- tachypnea within the first few hours of life, mild retractions, grunting, without signs of severe respiratory distress
- usually resolves in 24-72 hours

CXR: fluid in fissures, increased vascularity, slight cardiomegaly
Treatment - supportive: O 2 , nutrition, careful fluid administration

BRONCHOPULMONARY DYSPLASIA (BPD)

Chronic lung disease after prolonged intubation/ventilation with high pressures and high O 2 concentrations
- May have cardiac component (CHF)

Treatment: gradual weaning from ventilator, nutrition, avoid stress, dexamethasone may help decrease inflammation and encourage weaning, diuretics, branchodilators

APPLICATION OF KMC

- FROM BIRTH – An infant who does not require resuscitation may be placed in the kangaroo position at birth and is not separated from the mother
- INTERMITTENT – VLBW babies and those who require medical intervention may have KMC intermittently in the neonatal unit. Very ill infants who require assisted ventilation also benefit, provided the nursing staff monitor vital functions.
- CONTINUOUS – Commence this once an infant on intermittent KMC is stable, feeding well and gaining weight. It is best conducted in a ward with sleeping, ablution and recreation facilities and the area also serves as a venue for training mothers and staff. A nurse skilled in KMC provides supervision.
- Encourage the mother to move about with her infant. As her confidence and competence grow she and baby can prepare for discharge.
- THE DYING CHILD – When medical intervention is no longer effective the close contact provided by KMC gives a dying infant comfort and aids the grieving process.
- TRANSIT KMC – If a transport incubator is unavailable, use this method.

F/U after discharge:

Infants less than 1.6kg should be assessed daily. Those under 2kg should be seen every 2-3 days for the next 2 weeks.

ADVANTAGES:

- Cost effective
- Can be applied to infants of all weights and gestational ages
- Temperature regulation is stable
- HR and RR is more regular
- Growth is more rapid
- Fewer infections
- BF is enhanced
- Deep sleep is enhanced
- Bonding
- Mom gets more comfortable and competent with handling the baby sooner

KMC...

This remarkably simple innovation has improved the care and outcome of preterm babies particularly in underdeveloped regions. It has 4 components:

1. POSITION – Remove baby's clothes except for a cap and nappy. Place the infant upright against mother's skin between her breasts and ensure that the airway is not obstructed. A tight –fitting cotton garment will secure baby and enable mother to walk about. Baby should remain skin-to-skin as long as possible.
2. NUTRITION – If the infant is able to suckle, the breasts are always available for demand feeding. For the baby too small to breastfeed, give expressed breast milk via a feeding tube.
3. SUPPORT – The encouragement and assistance of staff is essential for success, particularly in the beginning. Family members need to know the significance of KMC as their support is needed throughout, particularly when mother and baby come home.
4. DISCHARGE – Mothers act as mobile incubators to provide temperature control, hence infants may go home earlier than usual. A baby who is stable, feeding well (ideally fully breastfed) and growing, may be discharged home regardless of weight or gestational age. However, KMC must be continued up to at least 2kg.

WRITTEN POLICY – Must be drawn up by staff as their commitment to breastfeeding. It must ensure that clinical and managerial practices do not interfere with breastfeeding

STAFF TRAINING – An important reason for the discontinuation of breastfeeding is that mothers receive conflicting advice from professional staff. It is recommended that those who are in clinical contact with mothers should receive at least 18 hours of training to ensure that information given to mothers is factual and consistent

INFORMATION DURING PREGNANCY – The mothers decision to breastfeed ought to be made before the birth of her infant and with adequate and factual information.

CODE OF MARKETING BREASTMILK SUBSTITUTES - Companies that manufacture infant formulas should not sponsor information leaflets or posters for mothers. Any display o this nature undermines breastfeeding.

BREASTFEEDING SOON AFTER BIRTH – Early feeding has a profound effect on the establishment and maintenance of lactation. It promotes the production of prolactin and oxytocin and consequently a healthy baby should be put skin-to-skin on the mother's chest at birth. This is also valid for C/S where possible.

ROOMING IN – Nurseries are for sick babies only! Do not separate the mother from the infant unless there is a medical indication. To initiate lactation, frequent feeding is essential and a mother needs to learn subtle hunger cues (sucking, rooting, mouth and tongue movements, soft sounds etc.). Crying is the last sign and by this time baby is often frantic and needs a lot more time, effort and patience to get on the breast.

POSITIONING – There are many ways to hold baby for feeding and each mother and infant should be assisted to find the most comfortable arrangement. Whatever the position, ensure that baby's head is not turned to one or other side but is aligned with the body to face mother. The classical position is popular for large babies. Here the infant lies across mothers chest with the head in the crook of her arm. Small infants and twins may be tucked under mother's axilla with the body on her forearm and head in her hand (rugby ball grip)

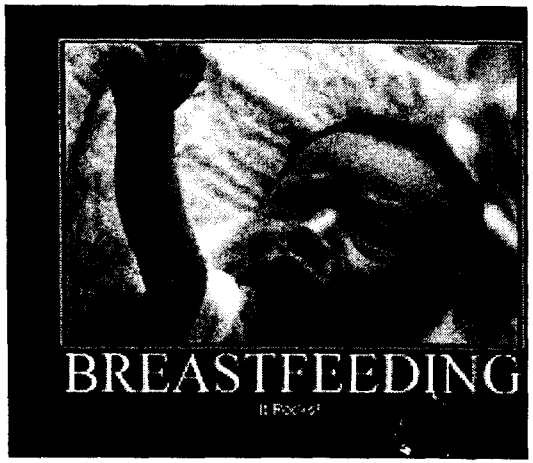
Only a third of woman who start breastfeeding in hospital are breastfeeding 6 months later. This is as a result of feeding problems

10 STEPS TO SUCCESSFUL BREASTFEEDING

1. Each maternity service is to have a written breastfeeding policy
2. Staff are to be trained appropriately to implement the policy
3. Pregnant woman are to know the benefits and technique of breastfeeding
4. Mothers are assisted to breastfeed within half an hour of the delivery
5. Mothers and babies stay together 24 hours a day
6. Mothers are shown how to breastfeed and maintain lactation even if separated from baby
7. Babies are breastfed on demand
8. Babies are exclusively breastfed for 6 months
9. Artificial teats and dummies are not used
10. Breastfeeding support groups are encouraged

LATCHING – Do not put the breast into baby's mouth, put baby ON the breast. When the infant's mouth opens in response to the rooting reflex place it over the areola. Beneath this are the milk sinuses which, when compressed by suckling, cause milk to flow into the mouth. Do not permit the baby to suck on the nipple – it is hard work for baby and cracks mom's nipples. Do not wash nipples before feeding. Apply colostrums to cracked nipples (superior to creams and lotions). Demand feeding is best.

EXPRESSING BREASTMILK – Hand expression; gently and rhythmically compress areola (should be painless). Breast pump; this is faster but strict hygiene is important. The more the breasts are emptied the more milk they produce. The colour and consistency of the milk may vary from grey, blue, yellow, thick or watery.



MILK STORAGE – Label!! Collect in clean glass, plastic or steal.

BREAST ENGORGEMENT - Can be corrected by correct positioning and frequent feeding. It usually starts on day 3-5 and is characterized by a low grade fever and painful, swollen, red, tender breasts. Treat with gently massage and warm compresses before a feed. Cold compresses (or cabbage leaves may be used between feeds)

MASTITIS – A tender spot on the breast without fever is indicative of a plugged milk duct whereas the dx is mastitis in the presence of a fever. Mx; continue breastfeeding, apply warm packs, Cloxacillin.

BREAST ABSCESS – Red, tender, fluctuating mass in the breast that requires surgical drainage, antibiotics and rest. BF can be continued.

INDADEQUATE BREASTFEEDING (5%)– Mothers sometimes stop BF because they feel their milk is insufficient. This is most unlikely if the infant has 6 wet nappies a day, gains weight and is not dehydrated.

BREASTFEEDING SUPPORT GROUPS – Feeding counselors must be sympathetic and approachable. All maternity units must have a list of BF support groups. A reason for not BF is often lack of time and support rather than lack of milk.

HISTORY TAKING, PHYSICAL EXAMINATION, AND EVALUATION OF THE SICK CHILD

KIM – JBURG 2011

CHAPTER 1 – HISTORY TAKING, PHYSICAL EXAMINATION, AND EVALUATION OF THE SICK CHILD

1. As per usual:
- Name, age, birth date, gender, race
- Present illness
- Main Complaint
- Systematic Review
- Rx & response Change in condition

2. Paediatric History:
Pregnancy & mother s health during pregnancy
Events of labour & delivery
Condition of baby in neonatal period
Growth & development
Immunizations
Diet & feeding history
Previous diseases
Previous operations &

3. Perinatal Health:
Prenatal History
Pregnancy
Previous pregnancies
Include: infections, illnesses, vaginal bleeds, toxaemia
Serology tests
Blood group of the

appropriate to age
If school-age – grades & marks achieved

Taking a complete History

Neonatal History
Cyanosis, difficulty in establishing feeds, convulsions, blood transfusions, nursing in incubator, length of stay
Jaundice + age of onset, duration & Rx

5. Immunization Hx:
From RTHC card
List which were given, adverse reactions
Dates & number of immunizations

6. Diet & Feeding:
BF vs FF
Vitamins, Iron & introduction of solid foods
If feeding difficulty or nutritional problem:
Date of onset of problem
Method of feeding
Types of formula
Interval between feeds
Weight changes
24hr dietary recall

8. Family Hx:
Very NB in paeds

7. Previous diseases & hospitalization:
Onset, Symptoms, Dx, Rx, Course, Cx
Results of any surgery
Include accidents, injuries & poisonings

9. Social Hx:
Occupation of parents
Housing
Access to clean water
School & play facilities
Caretaker

THE PAEDIATRIC PHYSICAL EXAMINATION

1. Age – dependent vital signs:

AGE	PULSE RATE	RR	BP
Day 1	120 – 140	40 – 50	60/40
1 yr	80 -140	30 - 40	80/55
2 – 5 yrs	70 – 115	20 – 30	90/60
School - going	70 - 115	15 - 20	100/65

2. Dentition & Closure of fontanelles

TEETH		FONTANELLES	
6-8 months	Lower incisors	Posterior	3 months
7-9 months	Upper incisors	Anterior	9-18 months
8-11 months	Lateral incisors		
10-16 months	First molars		
16-20 months	Cuspids		
20-30 months	Posterior molars		

3. Measurements:

Weight

Once a month for 1st 6 months
Once every 3 months for next 6 months
Twice a yr for next 3 years
Then yearly
Compare weight with weight–for–age charts

Height

Length is measured at each visit
Birth to 2 years – horizontal board with fixed vertical headpiece and sliding foot piece
Height – with rule fixed to wall and sliding headpiece

Head circumference

Routinely up to 2 years
Use greatest fronto-occipital circumference

Fontanelles

Ant: 2.5cm x 2.5cm @ 3 months, closes 9-18 months
Post: smaller & closes by 3 months

Upper & lower body segment ratio

Lower: from upper level of pubic ramus to base of heel
Upper: Height minus lower measurement
Limbs grow faster than trunk from fetal life to mid – puberty
Ratios: At birth = 1.7:1
At 10 yrs = 1.0:1
At 14 yrs = 0.9:1

Temperature

> 38°C = fever
< 35.5°C = hypothermia
Axillary or groin = 1°C lower than rectal / 0.5°C lower than oral

1. Genearal Appearance

Well, ill, comfortable, uncomfortable, breathing easily or distress
Describe facies
Well or acute distress, chronically ill, alert, comatose, delirious, lethargic, dull, bright, responsive, hostile, co-operative
Note interaction between child & caregiver

2. Skin

Lesions: note distribution, colour & character
Rashes: petechiae or purpura (better seen if skin is stretched & teated for blanching
Vasculitis: palpable purpura & don't blanch
Pellagroid lesions: in malnutrition.
Acute, bullous lesions of kwashiorkor can be mistaken for second degree burns.
Cyanosis: pulmonary or congenital heart disease
Jaundice: sclerae, skin, mucous membranes. Visible in newborn when total serum bili >90umol/l, older child >40umol/l. Cellular or obstructive liver disease, or haemolysis.
Pallor: nail bed, conjunctiva, oral mucosa or tongue. Due to hypoproteinaemia, low Hb, or shock.
Tissue turgor: for dehydration, (may not be decreased in obese patient)
Malnutrition: little subcutaneous tissue. Chronic disease: skin feels thin & loose.
Hair: paler, lustreless, red or grey, easily broken, thinner, lacking crinkle in malnutrition
Clubbing: look at fingers in profile (best sign in children) Also look for koilonychias, brittleness, or discolouration.
Hands: single palmar creases, missing digits, clinodactyly (incurving of little finger) Terminal thickening og radius at wrist – rickets
Rheumatic nodules: elbow, knee, wrist, ankle, over occiput
Muscle wasting: reflects protein deficiency. Muscle power decreased

3. Head

Shape, bossing, fontanelles (open, closed, prematurely or normally)
Premature closure of saggital suture: boat-shaped scaphocephalic skull. Closure of all sutures: small skull with proptosis
Ant fontanelle: closes prematurely in microcephaly & craniostenosis, open longer than normal in rickets, hydrocephalus & cretinism
Bulging: crying or straining, in relaxed child: raised ICP due to meningitis, encephalitis, brain tumour, subdural haematoma
Depressed: dehydration
Transillumination of skull: demonstrates abnormal collections of fluid, which may lie away form the fontanelle – hydranencephaly
Auscultation of skull: bruit – av fistula or vascular cerebral tumour
(Does it conduct from the neck?)

4. Face:
Look for features of cretinism, hypertelorism, mongoloid lanting of the eyes in Down Syndrome, depressed or abnormal nasal bridge, any tics or habit spasms

5. Eyes:
Squint, infections, cataracts, conjunctival haemorrhages, dryness, keratomalacia, Biltot's spots, corneal scars (Vitamin A deficiency)

6. Nose:
Discharge: mucopurulent or blood-stained watery discharge of congenital syphilis
Ascertain patency of nasal passages

7. Mouth & Throat:
Examine lips, teeth, gums, tongue & palate – excluded cleft palate
Look for Koplik's spot on buccal mucosa in measles, herpes ulcers of tongue, gingivitis
Tonsillar exudate occurs in infectious mononucleosis and moniliasis, rarely in diphtheria
Retropharyngeal abscess – swelling of posterior pharyngeal wall (always unilateral)
Postnasal drip & pharyngeal hyperplasia – in nasal allergy or infection

8. Ears:
Abnormalities of size, shape & position of pinnae, discharge from ear
Direction of ear canal in newborns & infants: upward
Older children: downward & forward

9. Neck
Neck stiffness: Kernig & Brudinski
Free movement of neck, limited by inflamed lymph glands, muscular spasm (trauma), joint disease, bony disease, apical lobar pneumonia
Feel for lymph nodes: submental , tonsillar, cervical (deep & superficial), supraclavicular

10. Chest:
Premature: rib cage is thin & chest may collapse with each inspiration
Infancy: chest almost round, AP diameter = transverse diameter
Funnel-shaped chest: congenital anomaly
Pigeon chest – anomaly / rickets
Look for swellings at costo-chondral joints – rachitic rosary
Harrison's sulcus - unless marked, it is not diagnostic of any disease (occurs in rickets, premature &chronic pulmonary disease)
Normal respiratory activity = abuominal movement until 6/7 yrs
Note any asymmetry
Precordium may bulge in cardiomegally
Pneumothorax/ localised disease – chest may flatten
Less movement of chest on side involved with pneumonia, hydro- or pneumothorax, obstructive foreign body or atelectasis

11. Cardiology, Respiratory & GIT systems
Refer to specific chapters

IMCI

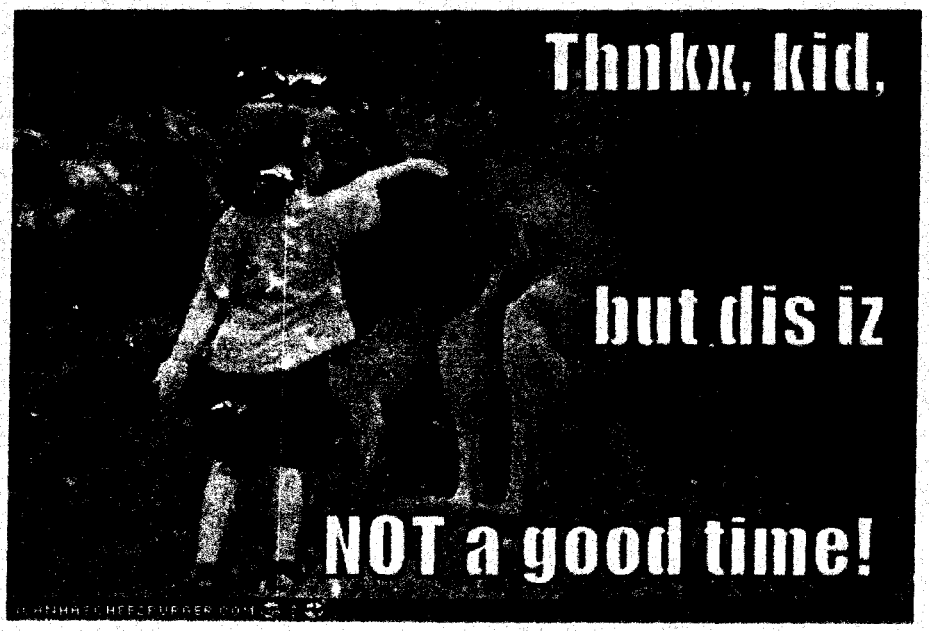
- Check for danger signs: inability to drink, vomiting everything, lethargy or coma, convulsions
- Ask about main symptoms and check fpr relevant danger signs:

- of respiratory illness: tachypnoea, indrawing, stridor, wheeze in a calm child
- of diarrhoea: delayed capillary refilling, not able to drink, lethargy
- of fever: bulging fontanelle, stiff neck, measles rash

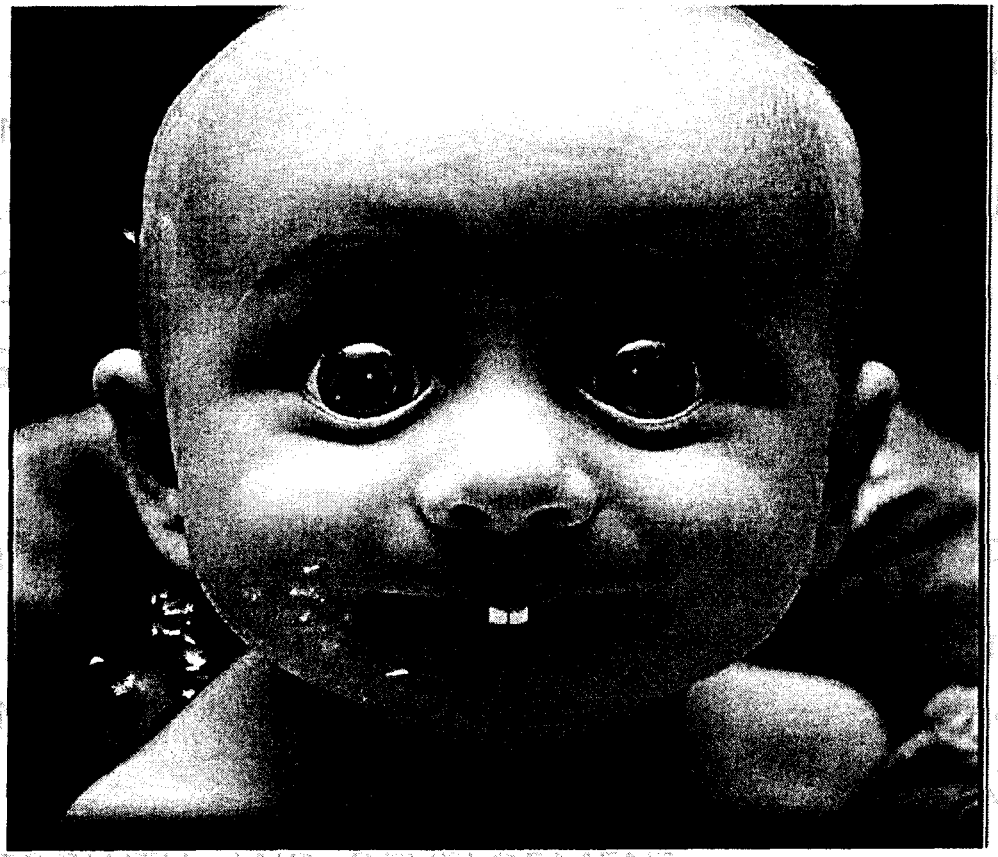
- Check foe malnutrition & anaemia
- Check immunixation status
- Assess other problems: HIV

GROWTH & DEVELOPMENT

ANDRE & OLLIE



GROWTH AND DEVELOPMENT



Chapter 2
Growth and Development

Signs and symptoms suggesting disorders of growth and development

1) Abnormal size at birth
   - Small for gestational age
   - Large for gestational age
   - Microcephaly
2) Abnormalities of growth
   - Short and thin (nutrition / disease)
   - Short and fat (syndromes / hormones)
   - Abnormal slow or fast growth
3) Global abnormalities of development
   - Cerebral palsy
   - Autism
   - Degenerative and metabolic disorders
4) Specific abnormalities of development
   - Abnormal fast or slow puberty development
   - Specific learning disorders
   - ADHD

Side note: Normal development and growth can only take place if the biological, emotional and social needs of the child are being met.
Children are able to "catch – up" on most delays if abnormalities are corrected.

Definitions and terminology

1) Growth: Increase in size, composition and distribution of tissues change in proportions, shape and function
2) Velocity: Speed at which changes take place, measured over time
3) Growth spurt: Increase in velocity of growth
4) Growth lag: Decrease in rate of velocity from what is expected to be normal.
5) Catch-up growth: Return towards size that would be expected if growth lag had not taken place.
6) Development: Increase in complexity of structures and their functions which take place in the same time period and often in parallel fashion. (product of interaction between maturation and learning)
7) Milestones: Usual age at which the ability to perform a specific activity is achieved
8) Chronological age: Calculated from date of birth
9) Corrected age: Chronological age is adjusted for prematurity
   For 1 year to assess growth
   For 2 years to assess development

Factors affecting growth and development

1) Hereditary and constitutional factors
2) Intra – uterine period
3) Postnatal period
4) Nutritional status and habits
5) Health status
6) Socio – economic status
7) Cultural factors

Development assessment

Done on grounds of 4 parameters:
- Gross motor
- Fine motor
- Language
- Personal – social aspects

1) Gross motor / Locomotor

Progression of abilities which ultimately enable the child to assume an upright posture and perform shilled activities while maintaining posture and equilibrium.
The primitive reflexes must be suppressed before voluntary movements can commence

2) Fine motor, manipulation and adaptive behaviour

Series of skills which develop through visually guided ability. Adaptive behaviour helps to place skills in context of environment inroder to initiate new experiences and learn from previous ones.

3) Language and communication

Formulation of thought and then transmission of that though through meaningful symbols or sounds

4) Personal and social aspects

Personal development is assessed on culturally monitored skills of daily living. Social development is behaviour which is in accordance with social experiences acquired through socialisation.

The Tanner Stages

Because the onset and progression of puberty are so variable, Tanner has proposed a scale, now uniformly accepted, to describe the onset and progression of pubertal changes (Fig. 9-24). Boys and girls are rated on a 5 point scale. Boys are rated for genital development and pubic hair growth, and girls are rated for breast development and pubic hair growth.

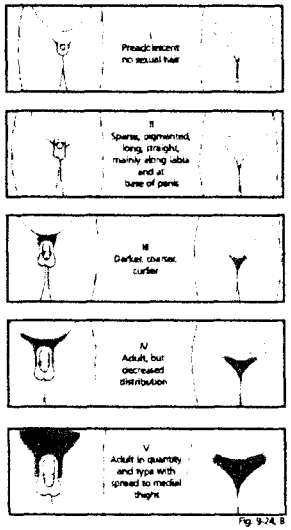


Fig. 9-24, B

Pubic hair growth in females is staged as follows (Fig 9-24, B):

- **Stage I (Preadolescent)** - Vellos hair develops over the pubes in a manner not greater than that over the anterior wall. There is no sexual hair.
- **Stage II** - Sparse, long, pigmented, downy hair, which is straight or only slightly curled, appears. These hairs are seen mainly along the labia. This stage is difficult to quantitate on black and white photographs, particularly when pictures are of fair-haired subjects.
- **Stage III** - Considerably darker, coarser, and curlier sexual hair appears. The hair has now spread sparsely over the junction of the pubes.
- **Stage IV** - The hair distribution is adult in type but decreased in total quantity. There is no spread to the medial surface of the thighs.
- **Stage V** - Hair is adult in quantity and type and appears to have an inverse triangle of the classically feminine type. There is spread to the medial surface of the thighs but not above the base of the inverse triangle.

The stages in male pubic hair development are as follows (Fig. 9-24, B):

- **Stage I (Preadolescent)** - Vellos hair appears over the pubes with a degree of development similar to that over the abdominal wall. There is no androgen-sensitive pubic hair.
- **Stage II** - There is sparse development of long pigmented downy hair, which is only slightly curled or straight. The hair is seen chiefly at the base of penis. This stage may be difficult to evaluate on a photograph, especially if the subject has fair hair.
- **Stage III** - The pubic hair is considerably darker, coarser, and curlier. The distribution is now spread over the junction of the pubes, and at this point that hair may be recognized easily on black and white photographs.
- **Stage IV** - The hair distribution is now adult in type but still is considerably less that seen in adults. There is no spread to the medial surface of the thighs.
- **Stage V** - Hair distribution is adult in quantity and type and is described in the inverse triangle. There can be spread to the medial surface of the thighs.

In young women, the Tanner stages for breast development are as follows (Fig. 9-24, C):

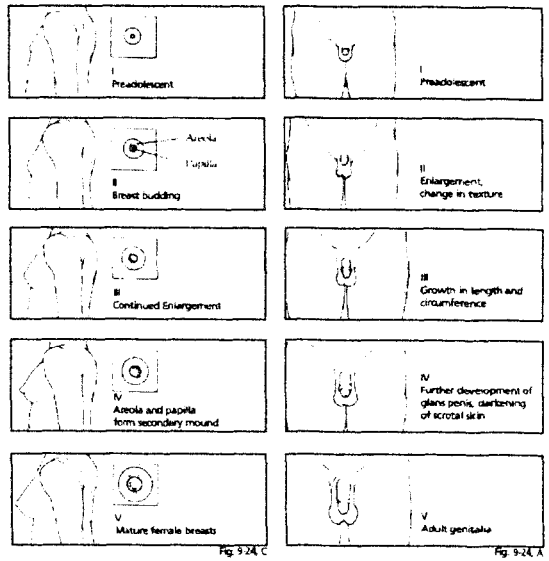


Fig. 9-24, C

Fig. 9-24, A

- **Stage I (Preadolescent)** - Only the papilla is elevated above the level of the chest wall.
- **Stage II - (Breast Budding)** - Elevation of the breasts and papillae may occur as small mounds along with some increased diameter of the areolae.
- **Stage III** - The breasts and areolae continue to enlarge, although they show no separation of contour.
- **Stage IV** - The areolae and papillae elevate above the level of the breasts and form secondary mounds with further development of the overall breast tissue.
- **Stage V** - Mature female breasts have developed. The papillae may extend slightly above the contour of the breasts as the result of the recession of the aerolae.

The stages for male genitalia development are as follows: (Fig. 9-24, A):

- **Stage I (Preadolescent)**- The testes, scrotal sac, and penis have a size and proportion similar to those seen in early childhood.
- **Stage II** - There is enlargement of the scrotum and testes and a change in the texture of the scrotal skin. The scrotal skin may also be reddened, a finding not obvious when viewed on a black and white photograph.
- **Stage III** - Further growth of the penis has occurred, initially in length, although with some increase in circumference. There also is increased growth of the testes and scrotum.
- **Stage IV** - The penis is significantly enlarged in length and circumference, with further development of the glans penis. The testes and scrotum continue to enlarge, and there is distinct darkening of the scrotal skin. This is difficult to evaluate on a black-and-white photograph.
- **Stage V** - The genitalia are adult with regard to size and shape.

Source:

Reprinted with permission from Feingold, David. "Pediatric Endocrinology" In *Atlas of Pediatric Physical Diagnosis, Second Edition*, Philadelphia: W.B. Saunders. 1992, 9.16-19

Child development stages

Child development stages describe theoretical milestones of child development. Many stage models of development have been proposed, used as working concepts and in some cases asserted as nativist theories.

This article puts forward a general model based on the most widely accepted developmental stages. However, it is important to understand that there is wide variation in terms of what is considered "normal," driven by a wide variety of genetic, cognitive, physical, family, cultural, nutritional, educational, and environmental factors. Many children will reach some or most of these milestones at different times from the norm.

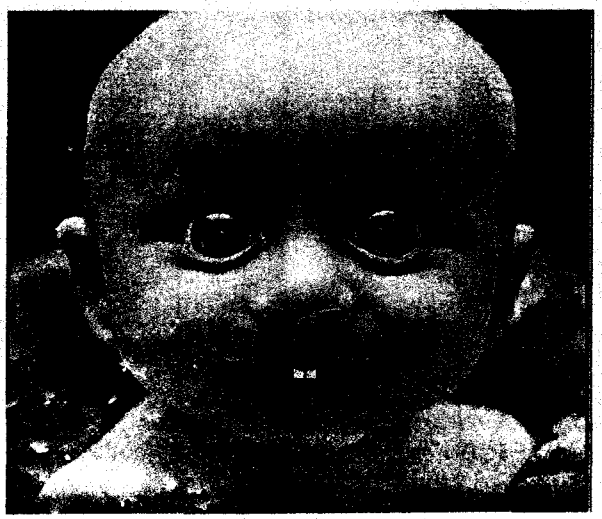
Overview of motor, speech, vision and hearing development

Developmental Milestones[1]

Age	Motor	Speech	Vision and hearing	Additional Notes
4–6 weeks				Smiles at parent
6–8 weeks		Vocalizes		
12–20 weeks			Hand regard: following the hand with the eyes.[2]	Serves to practice emerging visual skills.[3] Also observed in blind children.[2]
3 months	Prone:head held up for prolonged periods. No grasp reflex	Makes vowel noises	Follows dangling toy from side to side. Turns head round to sound	Squeals with delight appropriately. Discriminates smile.
5 months	Holds head steady. Goes for objects and gets them. Objects taken to mouth	Enjoys vocal play		
6 months	Transfers objects from one hand to the other. Pulls self up to sit and sits erect with supports. Rolls over prone to supine. Palmar grasp of cube	Double syllable sounds such as 'mumum' and 'dada'	Localises sound 45 cm lateral to either ear	May show 'stranger shyness'
9–10 months	Wiggles and crawls. Sits unsupported. Picks up objects with pincer grasp	Babbles tunefully	Looks for toys dropped	Apprehensive about strangers
1 year	Stands holding furniture. Stands alone for a second or two, then collapses with a bump	Babbles 2 or 3 words repeatedly	Drops toys, and watches where they go	Cooperates with dressing, waves goodbye, understands simple commands
18 months	Can walk alone. Picks up toy without falling over. Gets up/down stairs holding onto rail. Begins to jump with both feet. Can build a tower of 3 or 4 cubes and throw a ball	'Jargon'. Many intelligible words		Demands constant mothering. Drinks from a cup with both hands. Feeds self with a spoon. Most children with autism are diagnosed at this age.
2 years	Able to run. Walks up and down stairs 2 feet per step. Builds tower of 6 cubes	Joins 2–3 words in sentences		Parallel play. Dry by day
3 years	Goes up stairs 1-foot per step and downstairs 2 feet per step. Copies circle, imitates cross and draws man on request. Builds tower of 9 cubes	Constantly asks questions. Speaks in sentences.		Cooperative play. Undresses with assistance. Imaginary companions
4 years	Goes down stairs one foot per step, skips on one foot. Imitates gate with cubes, copies a cross	Questioning at its height. Many infantile substitutions in speech		Dresses and undresses with assistance. Attends to own toilet needs

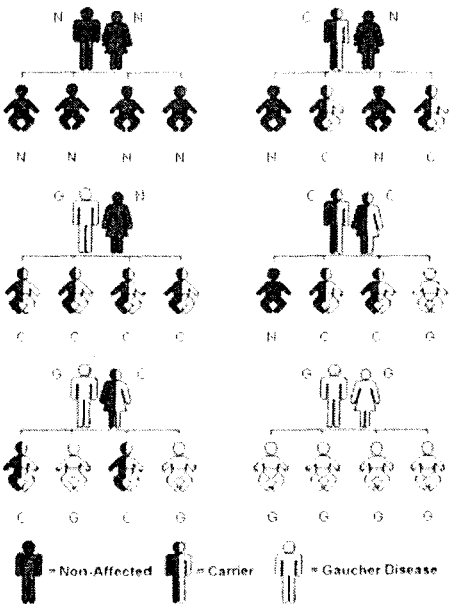
GENETIC & CONGENITAL DISORDERS

ANDRE & OLLIE



GENETIC AND CONGENITAL DISORDERS

- ABNORMAL LOOKING CHILD
- MONOGENIC INHERITANCE
- AUTOSOMAL DOMINANT
- AUTOSOMAL RECESSIVE
- X-LINKED INHERITANCE
- POLYGENIC OR MULTIFACTORIAL DISORDERS
- CHROMOSOMAL DISORDERS
- NON GENETIC CONGENITAL ABNORMALITIES
- INTELLECTUAL DISABILITY
- PREVENTION



Chapter 3: Genetic and congenital disorders

- Down syndrome (1 in 700)
- Spina Bifida (1 in 800)
- Facial cleft/ club feet (1 in 1000)
- Cardiac defects (1 in 200)
- Hydrocephalus (1 in 800)
- Microcephaly (1 in 600)
- Albinism(1 in 3000)
- Cysticc

1. Abnormal looking Child (FLK)

- Malformation- primary abnormal development e.g. genetic or teratogenic
- Deformation-external factors e.g. oligohydraminos leading to pulmonary hypoplasia/arthrogyphosis
- Disruption- interruption by extrinsic factor e.g. Amniotic bands

Process when confronted with an FLK

1. Collect biographical detail
2. Take parental history
3. Draw a pedigree
4. Pregnancy and birth history
5. History of the child
6. Head to toe examination
7. Try to establish an association (common factor such as embryological timing) or sequence (e.g. Potter sequence)

2. Monogenic (unifactorial) inheritance

- 10 in 1000 births
- PLEASE NOTE!!! Genetics can be very complicated: Some exceptions such as retinitis pigmentosa which may be inherited either dominantly, recessively or x- linked. Other exceptions are osteogensis imperfect subtypes and polycystic kidney disease (AD or AR). Weird modes of inheritance such as variable expressivity or non-penetrance, chimerism and imprinting all can affect certain conditions.
    - the disease.

3. Autosomal dominant (Table 3-1)

- 50: 50 risk for each pregnancy, although depending on who gave you the gene (mom or dad) it can be worse/ start later e.g. Angelman syndrome or Beckwith-Weideman syndrome
- Wide spectrum: Polydactyly(mild) to achondroplasia(sever but not lethal) to lethal.
- NEW MUTATIONS COMMON i.e. with an older father the child might have an AD condition although neither parents have the gene. Other siblings are then not at high risk for the gene,but the affected child will have the same chance of passing it to their offspring (50:50) Good example is neurofibromatosis. While something like huntington's has a very low rate of mutation
- **Example of variable expressivity: Waardenburg syndrome**
    - Some siblings have profound deafness, white forlock and heterochromia irides while other may only have the white forlock.
- **Example of Penetrance: Post axial polydactyly**
    - 1 in 100 black indiviuals in africa
    - Some members have gene, but not disease i.e. only seen in 70 out of 100 parents who MUST have the gene (70% penetrance) for the child to have
- **Example of high penetrance: Neurofibromatosis type 1**
    - 1 in 3000 across all populations
    - NF1 locus on Ch 17
    - High penetrance ,most will have some symptoms
    - Offspring of affected females more severly affected
    - 50% of cases are new mutations
    - Café au lait patches
    - Neurofibromas (duh)
    - Lisch nodules
    - Kyphosis etc. may be associated
    - Intellectual disability, seizures, malignant changes in 5 to 10 %

4. Autosomal recessive (table 3-2)

- More than 647 identified, very common, each person may carry 4 to 5 recessive genes.
- Where both parents are heterozygous, risk is 1 in 4 for homozygous child. 1 in 2 for heterozygous child etc.
- First cousins have a 1 in 16 risk for a sever recessive disorder even higher for example if an uncle and a neice marry (common in certain African cultures)
- Only affected if homozygous
- May also be affected if they have 2 deleterious genes (compound heterozygote)
- If disorder is heterogenous (caused by genes at different loci) then two affected people will only have unaffected children even though the children will be heterozygous at both loci e.g occulocutanoeus albinism
- Not dangerous in single dose (heterozygous),
- Heterozygous may be benefcial in some cases e.g sickle cell Hb gives protection against falciparum malaria. Cystic fibrosis gene may protect against cholera.

• **Occulocutaneous albinism (different races affected differently)**
• most common recessive condition in black south Africans 1 in 4000
• 1 in 16000 white
• **Cystic fibrosis**
• 1 in 200 white babies,but very rare in black children
• **B- Thalassaemia**
• Common in indian or greek
• **Tay-Sachs**
• Common in Ashkenazi Jews

5. X-linked inheritance (Table 3-3)

- 190 conditions, a few are dominant, most are recessive
- Female carriers <u>may/may not</u> be affected due to imprinting (Although women have two X-chromosomes, only 1 is actively involved in somatic activity) e.g. a mild decrease in factor VIII in a female carrier of Heamophilia A
- Female carriers have a 1 in 2 chance of producing an affected son, 1 in 2 for daughter carrier
- Affected males have unaffected sons (gave the Y-chromosome)
- All daughters who of affected males will be carriers
- NEW MUTATIONS common like in Duchenne's or haemophilia
    - Important to find out if the mother or father produced this mutation for future pregnancies

Condition	Genetic Properties	Features
Duchenne's Muscular dystrophy	• 1 in 3000 boys • Big area of distal short arm of X (Xp) • Deletion can be detected in 60%- important to determine carrier state of sister • 1/3 caused by new mutation	• Please refer to Neuro and to table 3-3
Haemophilia A	• 1 in 5000 boys • Deficiency of Factor VIII • Mother is mostly a carrier, therefore sisters are carriers • New mutations can occur occasionally	• Please refer to haematology

6. Polygenic or multifactorial disorders

- Require gene + other factors for disease
- More common than monogenic disorders, 2 % of births
- Recurrence risk for a couple is small (generally 3 to 5%), but if there is a lot of affected family members the risk increases
- Certain multifactorial disorders have different modes of inheritance e.g if cleft lip occurs with sparse hair and ectrodactyly then the correct diagnosis is actually ectrodactyly-ectodermal dysplasia-clefting syndrome (EEC) which is dominantly inherited.
- <u>Neural Tube Defect</u>
    - One of the Most common polygenic defect (1 in 800 in SA)and includes:
        - Anencephaly
        - Encephalocoele
        - Spina bifida
        - Myelomenigoceoele
        - Sacral agenesis
        - Associated with hydrocephalus due to Arnold-Chiari malformations
        - Can be associated with mental defects etc.
        - Lower limb paralysis +/- incontinence
    - Folate supplementation 3 months before and 3 months after conception reduces risk
    - Can be detected by AFP in amniotic fluid (can also have a high maternal AFP in mother at 16 weeks)+ Ultrasound diagnosis
- <u>Cleft lip +/- Palate</u>
    - 1 in 1000 white
    - 1 in 5000 black
    - Genetic/family component
    - Drugs play a role such as
        - anticonvulsants (phenytoin, phenobarb, primidone)
        - retinoic acid VIT A
        - barbiturates
        - valium = diazepam
        - +/-caffeine and alcohol

DRUGS causing CLEFT palate

- **Congenital Heart Defects**
  - Can be a component of several factors including:
    - Rubella embryopathy
    - Uncontrolled maternal diabetes
    - Increased alcohol consumption
    - Lithium
    - Anti-epiletics
  - It is a matter of genetic predisposition with other factors such as timing, dosage of drugs etc. which determine the defect
- **Others include**: congenital hip dislocation, diabetes, isolated hydrocephalus, Talipes equinovarus

7. Chromosomal disorders

- Very common, 6 in 1000
- Balanced translocations may have little or no effect
- Most are sporadic (little chance of recurrence) but some are inheritable

Indications for chromosomal analysis (especially in presence of multiple dysmorphic features)

1. Mental retardation of unknown origin
2. Multiple congenital abnormalities with no known teratogen exposure
3. Recurrent spontaneous miscarriage/stillbirths/unexplained neonatal deaths
4. Ambiguous genitalia
5. Hypogonadism / primary amenorrhoea
6. Family history of chromosomal translocation

Down Syndrom

- 1 in 700
- Small brachycephalic head
- Third fontanelle
- Facial dysmorphism
- Open mouth and protuberant tongue
- Epicanthic folds
- Upslantiong palpebral fissure
- Short fingers
- Single palmar crease
- Abnormal dermatoglyphics
- Clinodactyly
- Single interphalangeal crease of 5th finger
- Sandal gap
- Short stature
- Hypotonia

- Cardiac,skeletal, git defects
- Increased risk of haematological and endocrine diseases
- Heart defects lead to death in 20% by the first year
- Rest have Iq's ranging from 30 to 70
- 3 types
  - Trisomy 21 due to non-dysjunction
  - Translocation- new mutation
  - Mosaic- some cells have the extra 21 and others don't
- Risk of sporadic down's syndrome correlates with maternal age
  - Risk triples between 30 and 35 (1 in 300)
  - Triples again between 35 and 40 (1 in 100)
  - 45 (1 in 30)

True Hermaphrodites

- Both testicular and ovarian tissue with ambiguous genitalia
- 46 XX karyotype
- More common in SA than other areas
- Surgery for psychological/sexual function etc.
- Must exclude congenital adrenal hyperplasia which is an autosomal dominant condition

8. Non Genetic congenital abnormalities

- Teratogens include
  - Prenatal infections
  - Radiation
  - Drugs
  - Dietary substances/ toxins etc.
- **Fetal alcohol syndrome**
  - Large range of abnormalities, most sensitive during first month of pregnancy
  - Small for gestational age
  - Abnormal facies
    - Small palpebral fissure
    - Short upturned nose
    - Small palpebral fissure
    - Smooth upper lip with absent philtrum
    - Small mouth with angles turned down
    - Microcephally
    - Mental retardation
    - Heart defects
- **Congenital Rubella Syndrome**
  - Most common viral teratogen
  - Cataracts
  - Deafness
  - Heart defects

- Microcephally with mental retardation
- The extent of the damage is related to the stage of embryological development.

- **CMV/Toxoplasmosis**
  - Infection early in pregnancy is significant
  - Microcephaly and mental retardation
- **Other viruses** e.g. Hepatitis can cause biliary atresia. AND don't forget HIV!
- **Teratogenic drugs**

o Thalidomide	• Off market
o Warfarin	• Bony abnormalities,high perinatal loss, warfarin syndrome
o Anticovulsants	• Congenital heart disease, clefting, neural tube defects, MR
o Lithium	• Heart defects
o Isotretinoin	• Abnormal external appearance, heart, brain
o Radiation	o High doses usually result in miscarriage. Otherwise increase in leukemias,MR, malformations by about 1 in 1000
o Cigarrette smoking	o SGA

9. Intellectual disability

- Common 1 to 2 % have an IQ less than 80
- Mono/polygenic inherited condition 22%
- Chromosomal 15%
- Environmental and infection 15%
- Unkwon 43%

10. Prevention

- **Primary**
  - Genetic counselling
  - Behaviour/medication modification
  - Good ANC etc
  - Innoculations,folate etc more than 3 months before planning preganancy
  - Consider age
  - Consider family history
- **Secondary**
  - Offer TOP once prenatal investigations have confirmed

Procedure	Advantage	disadvantage
Chorionic villus sampling	• 9-11 weeks • Early diagnosis • Earlier top	• Invasive • 5% miscarriage risk • Cannot detect neural tube defects
Amniocentesis	• Chromosomal and AFP (neural tube defects) • Low risk of miscarriage 1%	• 13- 22 weeks • Possibility of late termination
ultrasound	• None invasive	• Operator dependant, high resolution equipment needed • Chromosomal analysis not possible
cordocentesis	• Confirmation of suspected abnormality	• High risk of miscarriage 10%

Tertiary

- Newborns are screened for phenylketonuria and hypothyroidism etc.

Indications for prenatal diagnosis

1. Advanced maternal age (35)
2. Chromosome disorders e.g. Down's
3. Neural tube malformations
4. Metabolic disorders e.g Tay Sachs (especially in whole population screening)

5. X-linked disorders
6. Abnormal maternal serum screen (Triple test at 15 – 20 weeks AFP,HCG, estriol E3)
7. Abnormal fetus scan (some cases)

Aims of Genetic counselling

- Comprehend medical facts,diagnosis,prognosis and management
- Appreciate genetics and recurrence risk
- Understand options for dealing with recurrence risk
- Choose appropriate course of action and carry out their choice
- Make the best possible adjustment to the disorder

Table 3.1 Common dominantly inherited disorders

Disorder	Clinical features
Achondroplasia	Short-limbed dwarfism
Apert's syndrome	Craniostenosis and syndactyly
Cataracts (some)	Lens opacities — usually bilateral
Ectodermal dysplasia	Hypohidrosis, transparent skin, abnormal and sparse hair, adontia/hypodontia
Haemophilia C (PTC deficiency)	Bleeding disorder
Hypercholesterolaemia	Raised cholesterol levels, myocardial infarction
Huntington's disease	Progressive chorea, dementia, family history
Marfan's syndrome	Tall structure, arachnodactyly, lens dislocation, dilatation ascending aorta
Myotonic dystrophy	Muscle weakness, myotonia, cardiac arrythmias
Neurofibromatosis	Pigmented spots (6 or more), neurofibroma, Lisch nodules
Osteogenesis imperfecta	Skeletal fractures, brittle bones, blue sclerae
Polycystic kidneys (adult type)	Progressive cystic renal enlargement with renal insufficiency
Polydactyly	Extra digits
Porphyria (variegate)	Drug sensitivity, recurrent abdominal pain, CNS signs
Retinitis pigmentosa	Pigmented retinae, night blindness, constricted visual fields
Spherocytosis	Haemolytic anaemia
Tuberous sclerosis	Adenoma sebaceum, seizures, mental retardation
Waardenburg syndrome	Deafness, heterochromia, white forelock

Table 3.2 Common recessive disorders

Disorder	Clinical features
Adrenogenital syndrome	Ambiguous genitalia, abnormal steroid production
Albinism (oculocutaneous)	Hypopigmentation of skin, hair and eyes, visual defects
Cystic fibrosis	Malabsorption, failure to thrive, recurrent chest infections
Deafness (some)	Severe bilateral congenital deafness
Galactoseamia	Cataracts, jaundice, vomiting, lethargy in early infancy
Microcephaly (some)	Head circumference < 3rd percentile, mental retardation
Mucopolysaccharidoses	Coarse features, growth and (some) mental retardation, stiff joi
Retinitis pigmentosa (some)	Pigmented retinae, late-onset blindness
Sickle-cell anaemia	Chronic haemolytic anaemia, intermittent pain crises
Spinal muscular atrophy	Hypotonia, weakness, absent deep tendon reflexes
Tay-Sachs disease	Mental retardation, seizures, cherry red spot on macula
Thalassaemia(s)	Microcytic anaemia, jaundice, hepatosplenomegaly, stunted gro

Table 3.3 Common X-linked recessive disorders

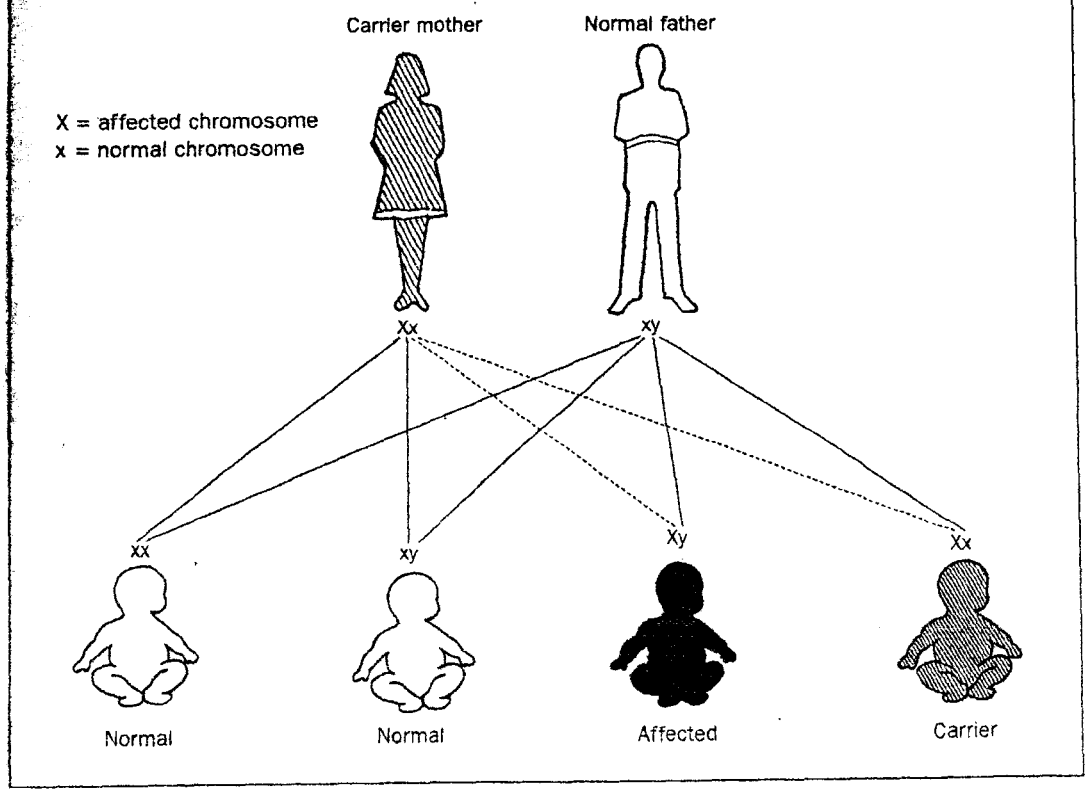
Disorder	Clinical features
Duchenne's muscular dystrophy	Pelvic muscle weakness and deterioration, pseudohypertrophy of calves
Fragile X syndrome	Intellectual disability, large testicles, long facies
Glucose-6-phosphate dehydrogenase deficiency	Haemolytic anaemia, jaundice, haemoglobinuria after exposure to haemolytic agents
Haemophilia A and B	Bleeding diathesis, factor VIII(a) or factor IX(b) deficienc
Hydrocephalus (aqueduct stenosis)	Macrocephaly, dilated cerebral ventricles
Incontinentia pigmenti	Bizarre hyperpigmentation patterns, dental anomalies, hair loss
Microphthalmia	Small eyes
Mucopolysaccharidosis (Hunter)	Mental and growth retardation, coarse facies, stiff joints
Ocular albinism	Hypopigmentation of the fundus
Retinitis pigmentosa (some)	Retinal pigmentation and degeneration, night blindness constricted visual fields
Vitamin D-resistant rickets (familial)*	Hypophosphataemia, rickets

*X-linked dominant

Table 3.5 Common chromosome disorders

Disorder	Clinical features
Trisomy 21 (Down syndrome)	Hypotonia, epicanthic folds, heart defects, intellectual disability
Trisomy 18 (Edward syndrome)	Prominent occiput, low-set malformed ears, clenched hands with overlapping fingers, short sternum, rocker-bottom feet, heart defects, profound develop mental delay (90% die in first year)
Trisomy 13 (Patau syndrome)	Cleft lip and palate, polydactyly, scalp defects, clenched fists, microphthalamia, heart defects, severe intellectual disability (50% die by 1 month)
45X (Turner syndrome)	Short female, broad chest, ovarian dysgenesis, peripheral lymphoedema at birth, webbed neck, coarctation of aorta, usually normal intelligence and lifespan
47,XXY (Klinefelter syndrome)	Tall male, hypogenitalism, mild intellectual disability, behavioural problems.
47XYY syndrome	Tall stature, mild intellectual disability
Fragile X/associated mental retardation (Martin-Bell syndrome)	Enlarged testes, mild to moderate intellectual disability, long facies, large ears, (20–30% female heterozygotes, mildly intellectually disabled)

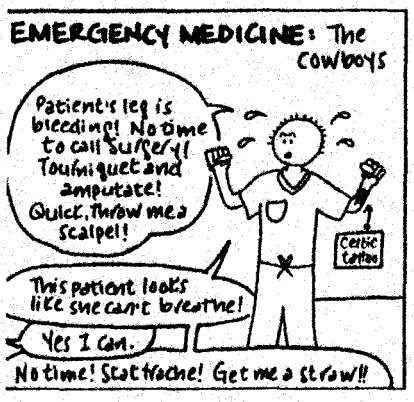
Figure 3.4 X-linked recessive inheritance

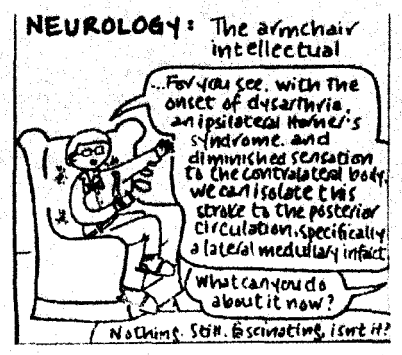


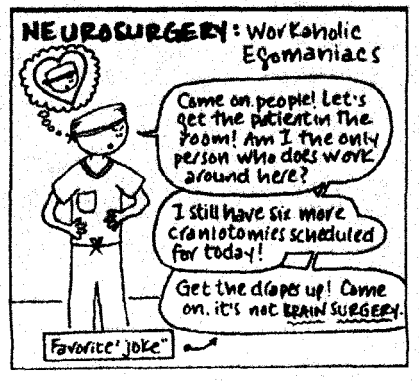
CARDIOLOGY

KOBUS - PE

CARDIAC EXAMINATION
CONGENITAL HEART DISEASE & VALVULOPATHIES
HEART FAILURE
INFECTIVE ENDOCARDITIS
RHEUMATIC FEVER
CARDIOMYOPATHIES
PERICARDIAL DISEASES
MYOCARDITIS
ARTERITIS
DYSRHYTHMIAS







CARDIOLOGY

Cardiac Examination
Congenital Heart Disease & Valvulopathies
Heart Failure
Infective Endocarditis
Rheumatic Fever
Cardiomyopathies
Pericardial diseases
Myocarditis
Arteritis
Dysrhythmias

This shouldn't take you more than 4 minutes, so please practice it tomorrow!

Cardiac Exam

1-General
2-Arm
3-Leg
4-Neck & Face
5-Thorax
6-Auscultate back
7-Auscultate for crackles
8-Abdomen
9-Weigh the child

Completely expose the patients thorax and abdomen

1) General: General impression
Acute/Chronic/Acute-on-chronic
Any IV lines/O2 face mask/catheter/parenteral feeding?
Resp distress? (nasal flearing/tachypnoea) (think HF)
Sweating/Puffy eyes/blue lips? (think HF)
Vitals

Please don't memorize this. Say what you see.

2) Arm: Palpate Radial pulse (brachial pulse in small children)

Comment on:
-Rate, -Rhythm → Peripheral pulse palpation
-Volume, -Character → Central pulse palpation

Palpate both R&L pulses (radio-radio delay: think coarctation)
Waterhammer pulse (Think AI), Absent/Asymmetrical? (Think takayasu)
Nails: Clubbing (think Cyanotic heart lesion)
Cianosis (think Cyanotic heart lesion)
Splinter haemorrhages (think IE)
Cap filing time (>3s, think dehydration/shock)
Feel extensor surface of hand & elbow for subcutaneous nodules (think Rheumatic Fever)
Elbow swollen/tender/decreased ROM (think Rheumatic Fever)

NB: ALWAYS COMPARE LEFT & RIGHT

3) Leg: Palpate Dorsalis Pedis/Tib Posterior
Nails: As above
Push finger against Tibia for 10 seconds, oedema? NB: Is it bilateral? (Think HF)
Feel skin over Tibia for nodules
Knee swollen/tender/decreased ROM
Palpate radial&femoral pulses (radio-femoral delay: think coarctation)
AGAIN: DON'T FORGET TO COMPARE L&R

4) Neck&Face: Distended neck veins (think RHF,SVC obstruction, constrictive pericarditis, rarely tamponade)
JVP
Supraclavicular pulsations (think coarctation)
Auscultate neck when listening to heart
Face oedematous, especially eyes? (HF)
Conjunctiva and mucus membranes yellow? (Jaundice – Hepatic impairment)
Sclera pale? (Anaemia)
Blue lips (peripheral cyanosis) (HF)
Blue tongue (central cyanosis) (HF)
Dental Caries (IE)

apex beat

Please comment on:
1)Position. If lateral displaced:
-chest deformity
-mediastinal shift 2° to pleural effusion
-tension pneumothorax
-pneumonectomy/lung collapse
If Infero-lateral displaced:
-L ventricular dilatation
2)Character. Heaving = AS or MI
= L ventricular hypertrophy
Tapping = MS
Double impulse = HOCM
Impalpable? Think of hyperinflation, large pleural/pericardial effusion. dextrocardia and fat kids.

5) Thorax

Inspection: Scars
Visible pulsations
Chest deformity

Pectus Excavatum
Pectus Carinatum
Barrel chest---------Hyperinflation

Palpation: Trachea central
Lay whole hand flat over precordium – general impression + thrills
Locate apex beat. Normally in 5th ICS 1cm lat to midclavicular line

@ Apex = AS
L sternal edge = VSD

Palpate for thrills @ apex + both sides of sternum –feels like purring cat!
Feel for parasternal heaving----------R ventricular hypertrophy
Pulsation below xiphisternum----------R ventricular hypertrophy
Palpable P2 (2nd L ICS)----------Pulm Hypertension

Percussion: Heart borders. Decreased cardiac dullness→hyperinflation
Not reliable clinical test, doesn't tell you much

Auscultation: Pt @ 45°
Low pitched
High pitched
First listen with bell, then diaphragm
ID: S1 & S2
-Intensity
-Character
-Splitting
-Fixed
S3/S4?
Murmurs?
Opening snap?
Ejection clicks?
Midsystolic click?
Pericardial rub?-----@ diaphragm (think pericarditis)

MS. Early diastole. Best @ apex.

S1---------S2--OS-----S1

S. Early systole. 2nd R S.

S1--EC------S2

Mitral valve prolapse. @ apex. Mid systolic.

S1------MC-------S2

So where do you listen?

N
E
C
K

2nd R ICS
A-valve

2nd L ICS
P-valve

4th ICS parasternal
T-valve

Base

5th ICS miclav
M-valve

A bit more on heart sounds

S1 – closure of mitral & tricuspid valve
LOUD – MS, Tachycardia (short PR interval)
SOFT – HF, effusion, valve incompetence
VARIABLE – extrasystoles, AF

S2 – P2 = closure of pulmonary valve
A2 = closure of aortic valve
LOUD – systemic HT, pulm HT
SOFT – decreased BP, effusion, valve incompetence
SPLIT – Widens on inspiration:
Enhanced physiological
RBBB, Pulm HT, Pulm Sten, VSD

Fixed splitting = ASD

Widens on expiration (Reverse splitting):
AS, LBBB

S3 – gallop rhythm only if HR > 100
Soon after S2, early diastloe
PHYSIOLOGICAL – Hyperdynamic circulation
PATHOLOGICAL – Large poor contracting L ventricle
Thus HF

S4 – Atria contracting against a stiff ventricle
Best heard with bell at apex
Just before S1, late diastole
PATHOLOGICAL & SOFT – HT, AS, L vent hypertrophy

When you mention a murmur – comment on 9 things:

1) Timing (systolic/diastolic)
2) Location (where it is heard the loudest)
3) Radiation (where else can you hear it)
4) Duration (pan/early/late)
5) Intensity:
   1=soft, soft (optimal conditions needed)
   2=soft but audible
   3=louder, no thrill
   4=loud, palpable thrill
   5=louder, stet still needed, palpable thrill
   6=no stet needed
6) Shape/Character (crescendo/decrescendo)
7) Pitch (low/high)
8) Variation with maneuvers
9) Added sounds (snaps/clicks)

Accentuating maneuvers:
R heart – Inspiration & squatting
L heart – Expiration & vulsulva

Innocent murmur
50-80% of children have a Murmur @ some point.
Most murmurs are functional (innocent), without structural abnormalities.

Hallmarks of innocent murmur:
>No Sx's, normal CVS exam, systolic, no radiation, variation with posture, louder during fever, grade 3 or less.

4 Innocent heart murmurs:

Type	Description	Diff Dx
Still's murmur	L lower sternal border Vibratory	Small VSD, subaortic stenosis
Ejection pulmonary	L upper sternal border Soft	ASD, PS
Venous hum	Infraclavicular, R>L Continuous	PDA
Supraclavicular arterial bruit	Above clavicles Low intensity	AS, bicuspid aortic valve

Clinical pearl:
-In multiple murmurs, which 1 is dominated by clinical sg's?
-Severity of murmur is determined by duration and not loudness

6) Auscultate the patients back---listen for bruits over ribs (think of coarctation)

Other Sg's of aortic carctation:
- Collaterals sometimes felt around scapula
- Prominent pulsation in supraclavicular notch
- Bruit over site of coarctation (btwn scapula)
- Upper limb hypertension
- R arm BP > L arm BP (depending on site of coarctation)
- Decreased BP in legs
- Radio-radio delay
- Radio-femoral delay

7)Auscultate for crackles (pulm oedema – think of HF)

8)Abdomen

Inspection: Visible pulsations
Ascites → Fullness of flanks, Smiling umbilicus

Spleen vs. Kidney
- Cant get fingers btwn ribcage & spleen
- S enlarge to R iliac fossa, K down
- S = notch
- S moves on early inspiration, K on late inspiration
- S not ballotable
- If S enlarged = Traube's space dull on percussion

Palpation: Heat up you hands, put on stomach, win trust
First palpate superficial, then deep
Begin under umbilicus for liver and spleen, any tenderness?
Liver----child < 5 yrs: 1-2cm below ribs in midclavicular line = normal
-----in congestion (HF):liver enlarged,smooth edge & surface,tender
Spleen-----superficial organ, moves on inspiration
-----enlarges 2° to liver congestion
Palpate over abdominal aorta-----prominent pulsation below umbilicus
-----think of aneurism
Palpable thrill over liver (think tricuspid incompetence)

Normal liver position:
*Age 0-6 months = 1-2 fingers below rib cage
*Age 6-24 months = 0-1 finger below rib cage
* Age 2+= usually not palpable (but remember, a palpable liver might still be normal btwn 2-5 yrs 1-2cm in midclavicular line)

Percussion: Percuss liver----hold fingers parallel to expected enlargement
----lung pathology can push liver down, so a normal liver can be palpated as "large".Upper border: below 6th rib MC line
Lower border:1-2cm below ribs
Fluid thrill, shifting dullness, puddle's Sg in older kids (percussion with pt on all fours)

Auscultation: Listen over abdo aorta----murmurs/bruits
Bruit over liver (think tricuspid incompetence)
Bruit over renal arteries (think renal artery stenosis)
Litman scratch test (older kids)----hepatomegally

Sx's & Sg's suggestive of cardiac disease:
- Central cyanosis not responding to O2
- Pallor & sweating despite a normal Hb
- Failure to feed and shortness of breath after feeding
- Tachypnoea & resp distress
- Failure to thrive despite good nutrition
- Sudden gain in weight or development of oedema
- Unexplained hepatomegally
- Heart murmur
- Abnormal pulse & Apnoea/syncope attacks

9) Always weigh the child at the end of the exam

Some clinical Sg's not yet discussed:

Age-dependant range of HR, RR & BP

Age	HR	RR	BP
Day 1	120-140	40-50	60/40
1 Year	80-140	30-40	80/55
2-5 Years	70-115	20-30	90/60
School going child	70-115	15-20	100/65

Estimated weight = (age x 2) + 9
Estimated SBP = (age x 2) +70

Causes of Tachycardia:

Cardiac	Non-Cardiac
1)Any shunt/septal defect	1)Anaemia
2)AS & MS	2)Dehydration
3)Myocarditis	3)Shock
4)Cardiomyopathy	4)Infection/sepsis
5)HF	5)Hyperthyroidism

Causes of Bradycardia:

Cardiac	Non-Cardiac
1.Heart block (congen)	1.B-blockers
2.Myocarditis	2.Digoxin
3.Terminal HF	3.Hypothyroidism
	4.Typhoid
	5.Raised ICP

The PULSE & BP

Absent pulses:
-Takayashu's arteritis
-Coarctation (↓ femoral pulse)

Irregular Pulse:
*Sinus arrhythmia
*Extrasystoles
*AF (rare in kids)
*Aflutter with variable response
*Heart block with variable response

Pulse Types:

1.Waterhammer/Collapsing pulse
Def: Forceful impulse but immediate collapse
-AI

2.Pulses Paradoxys
Def: ↓ in SBP > 10 mmHg on inspiration
-Severe airway obstruction, eg.asthma
-Pericardial tamponade/effusion

3.Pulses Alternans
Def: regular rhythm, irregular volume
-Advanced HF

4.Pulses Bisferiens
Def: 2 systolic peaks separated by midsystolic dip
-AS + AI

5.Bigeminus
Def: Ectopic after every normal beat

CYANOSIS

Remember:
Cyanosis = present when 3-5 g/l haemoglobin is unbound to O2-----so you turn blue when the unbound Hb concentration ↑.
Central = Tongue
Peripheral = Hands, Feet, Lips

NB: If pt anaemic, he wont look cyanosed, although low saturation, because remember: Cyanosis = unbound haemoglobin.

Peripheral cyanosis 2° to :
>Shock
>Dehydration
>HF
>Sepsis
>Peripheral vasospasm
>Cold

Central cyanosis
1)V/Q mismatch.
Eg. Pulm oedema, Pneumonia
Responds to O2---Physiological
2)Shunt
Eg. R to L shunts
Doesn't respond to O2---Anatomical
3)Haemoglobinopathies
Eg. Methaemoglobinaemia
Sulfhaemoglobinaemia
4)↓FiO2

Cyanosis cardiac in origin:
1.STOP O2 – Provoke closure of ductus arteriosus.
2.Maintain temp
3.5% glucose IV
4.NaHCO3 IV (Correct ↓ Ph)
5.Oral prostaglandin E2

Central cyanosis due to cardiac abn distinguished from resp cause by failure of PaO2 to rise above 15kPa after breathing 100% O2 for 10 mins.

CLUBBING

Clubbed or not Clubbed ????
(almost like "to be or not to be", but not quite the same)

Examine the ring finger
1. Shamroffs diamond disappears
2. Interphalangeal depth ratio > 1
3. Lovi Bond angle > 180°
4. Soft spongy nail bed
5. See-saw sg

Causes:

CVS
- -Cyanotic heart disease
- -Infective endocarditis
- -Atrial myxoma (rare in kids)

Resp
- -Chronic suppurative lung diseases
  - >Bronchiectasis
  - >Cystic fibrosis
  - >Lung assess
  - >Empyema
- -Interstitial lung disease

GIT
- -Inflam Bowel diseases
  - >Chrons/ Ulcerative colitis
- -Billiary atresia
- -Cirrhosis/Liver abscess

Unilateral – AV malformations
Just toe nails – PDA
Congenital

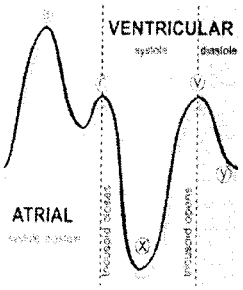
OEDEMA

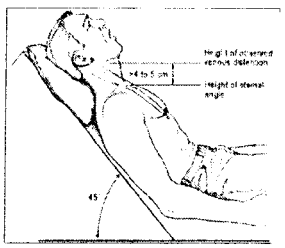
1. ↑ **HP**: RHF (DVT in adults)
2. ↓ **COP**: ↓ plasma proteins
   - *Cirrhosis
   - *Malnutrition
   - *Nephrotic syndrome
   - *Protein losing enteropathy
3. **Lymphnode obstruction**
   Remember in adults = 1° - Milroy syndrome
   2° - Infection
   Malignancy, radiation
4. ↑ **vascular permeability**
   *Infection

Anasarca = Severe HF, Renal pathology (esp. nephrotic syndrome)

JVP

Only do in older children





Features of the JVP
A venous pulse is not usually palpable.
Pressing at the base of the vein will make the vein visible as it continues to fill and distend above the point of pressure NB do not do this in exams.
Hepatojugular reflex aids identfication of JVP - probably by forcing blood out of liver into IVC and therefore into right atrium increasing its pressure.
JVP alters with changes in posture.

How to find the JVP
Sit patient at *45°* and turn head slightly away from you.
Look for JVP in *internal jugular vein* (not external jugular vein) medial to the clavicular head of sternocleidomastoid; the vein passes behind the angle of the jaw in direction of earlobe.
Measure JVP in cm above the sternal notch - a vertical not diagonal distance - if larger than 4cm the JVP is raised.

Abnormalities of the JVP

1) Raised JVP with normal waveform
- >right heart failure
- >fluid overload
- >bradycardia

2) Raised JVP with absent pulsation
- >SVC obstruction - full dilated jugular veins, no pulsation, oedematous face and neck

3) Large a wave
- >tricuspid stenosis - atria contracts against stiff tricuspid and so pressure in atria rises higher than normal
- >pulmonary hypertension - there are generally higher pressures on the right side of the heart
- >pulmonary stenosis

4) Extra-large a wave = Cannon wave
Occurs when atrium contracts against closed tricuspid eg
- >complete heart block
- >atrial flutter
- >single chamber pacing
- >nodal rhythm (AV node is in charge)
- >ventricular extra-systole
- >ventricular tachycardia

ie any condition in which the atria and the ventricles are not conducting in appropriate rhythm

5) Absent a wave
- >atrial fibrillation

6) Systolic waves = combined c-v waves = big v waves
- >tricuspid regurgitation (c-v wave because the pressure in the right atrium is raised throughout ventricular systole - tip is to watch for earlobe movement!)

7) The slow y descent occurs in tricuspid stenosis (if the HR is so low as to allow the length of descent to be appreciated!)

8) Paradoxical JVP = *Kussmaul's sign*
Normally the JVP should rise on expiration and fall on inspiration.
When the JVP rises on inspiration it indicates
- >pericardial effusion
- >constrictive pericarditis
- >pericardial tamponade

Congenital Heart Disease (CHD)

See pictures @ end of chapter when u study this

- 6-8/1000 live births

Clinical Picture

***HF**
***Shock**
***Sweating**
***Murmur**
***Cyanosis**
***Shortness of breath**
***Feeding problems**
***Failure to thrive**

Causes

- 85-90% cause is unknown
- Known causes:

Maternal factors =
>DM
>Medications (phenytoin), alcohol-----ASD/VSD
>Infections (rubella-----PDA/PS, CMV)

Infant factors =
>Prematurity-----PDA
>Chromosomal abnormalities
Downs syndrome (tris 21)------AVSD
Turner syndrome (tris 13)------AS/Coarctation
Williams syndrome (deletion of 26 genes frm long arm of chromosome 7)------supravalvular AS
>Positive Family Hx

Although there are > 100 different cardiac malformations, a small nr account for the majority of cases. These are classified into:

Cyanotic

↑ PBF
*TGA
*Truncus a.
*TAPVC
*HLHS

Hallmarks:
>Cyanosis
>Chest deformity
>Cardiomeg
>CCF
>Cweating (sweating)
>SOB
The 5 C's and the S

↓PBF
*TOF
*Ebstein
*Tricuspid atresia
*Critical PS

Hallmarks:
>Cyanosis
>No cardiomeg
>No CCF

Acyanotic

Normal PBF
*AS
*Coarct.
*PS/TR/MR

↑PBF
*ASD
*VSD
*AVSD
*PDA

All of the acyanotic heart lesions might present with CCF, exept **PS** and **ASD**

Abbreviations:

PBF – Pulmonary blood flow
TGA – Transposition of great arteries
Truncus arteriosus
TAPVC – Total anomalous pulmonary venous connection
TOF – Tetralogy of Fallot
Ebstein anomaly
PS – Pulmonary stenosis
AS – Aortic stenosis
Coarct – Coarctation
ASD – Atrial septal defect
VSD – Ventricular septal defect
AVSD – Atrioventricular septal defect
PDA – Patent ductus arteriosus
HLHS – Hypoplastic left heart syndrome

The Top 7 Congenital cardiac Anomalies:

VSD	38%
PDA	20%
Coarct	10%
TOF	6%
AS	4%
ASD	3%
Isolated PS	3%

--Systemic venous return re-enters systemic circulation directly
--O_2 sats < 75%
--Differentiate btwn cardiac and other causes of cyanosis with hyperoxic test

Cyanotic Congenital Heart Disease

1) Transposition of the Great Arteries (TGA)

*Parallel pulmonary & systemic circulation:
Systemic: Body→ RA→RV→Aorta→Body
Pulmonary: Lungs→LA→LV→Pulm a.→ Lungs
*Newborns present with progressive cyanosis unresponsive to O_2 as the ductus arteriosus closes and mixing btwn the 2 circulations diminishes; severe hypoxaemia, acidosis, and death can occur rapidly.
*If VSD present, cyanosis is not prominent, infant presents with CCF after few weeks of life.
*Murmur: None if no VSD

S/I:
ECG = Right axis deviation/RVH
CXR = egg-shaped heart with narrow mediastinum "egg-on-a-string" + plethora

Mx:
>>Prostaglandin E1 infusion to keep ductus open until septotomy or surgery
>>Balloon arterial septostomy with catheter
>>Surgery: arterial switch procedure

Infants without VSD must be repaired within 2 weeks to avoid weak LV muscle

2) Persistent Truncus Arteriosus

*The embryological structure known as the truncus arteriosus never properly divided into the pulmonary artery and aorta
*Anatomical changes associated with this disorder:
>Single artery arising frm the 2 ventricles, which gives rise to both pulmonary artery and aorta
>Right sided aortic arch
>Large VSD
>Pulm HT
>Complete mixing at level of great vessels
*Cyanosis, CCF, Cardiomeg, Widened pulse pressure, Collapsing pulse
*Murmur: Systolic ejection click & long systolic murmur @ L sternal border

S/I:
ECG = Normal to Right axis deviation
Biventricular hypertrophy
CXR = Plethora

Mx:
>>Neonatal Surgical repair

3) Total anomalous pulmonary venous connection (TAPVC)

*All 4 pulmonary veins are malpositioned and make anomalous connections to the systemic venous circulation
*A Patent foramen ovale & ASD must be present, otherwise the condition is fatal
*4 Variants:
1)Supracardiac (50%)=bld drains to one of innominate veins or SVC
2)Cardiac (20%)=bld drains into coronary sinus or R atrium
3)Infradiaphragmatic(20%)=bld drains into portal or hepatic veins
4)Mixed(10%)
*Cyanosis, CCF, Cardiomeg, tachypnoea, dyspnoea, S3, R vent heave
*Murmur: Ejection systolic @ 2nd L ICS, fixed split S2

S/I:
ECG = Right axis, RVH
CXR = Snowman sg
Plethora

Mx:
>> Prostaglandin E1 infusion to keep ductus open
>>Surgical redirection

4) Hypoplastic left heart syndrome (HLHS)

*A spectrum of hypoplasia of left ventricle, atretic mitral and/or aortic valves, small ascending aorta, coarctation, with resultant systemic hypoperfusion
*Most common cause of death frm congenital heart disease in 1st month of life
*Cyanosis, CCF, Cardiomeg
*Persistent with circulatory shock and metabolic acidosis on closure of the ductus
*Murmur: Precordial systolic murmur. Ejection systolic click. S3.

S/I:
ECG = Right Axis, RVH
CXR = Plethora

Mx:
>>Intubate & correct metablic acidosis
>>IV infusion of Prostaglandin E1
>>Surgical correction: Norwood procedure
>>Transplantation
>>Palliative

Cyanotic ↓ PBF

5) Tetralogy of Fallot

*Embryologically a single defect with hypoplasia of the conus causing:
--VSD
--Overriding aorta
--Right ventricular outflow tract obstruction (RVOTO)
--RVH
*Direction & degree of shunt are functions of the relative outflow tact obstruction
*Infants may initially have a L to R shunt & therefore are not cyanotic, but the RVOTO is progressive, resulting in increasing R to L shunting with hypoxaemia and cyanosis
*Hypoxic spells:
*1° pathophysiology is hypoxia, leading to ↑ PVR, & ↓ SVR, occurring in exertional states (e.g.crying)
*paroxysm of rapid & deep breathing, irritability, crying
*peak incidence @ 2-4 months
*if severe, may lead to seizures, LOC, death
*Mx:O_2, knee-chest position, fluid bolus, morphine, Propanolol, correct metabolic acidosis
Murmur: Single loud S2 (severe PS). Ejection systolic L sternal border

S/I:
ECG = Right Axis, RVH
CXR = Oligaemia
Boot shaped heart
Right aortic arch

Mx:
>>Surgical repair including closure of VSD & widening of RVOTO

6) Ebstein Anomaly of the tricuspid valve
*Tricuspid valve is dislaced into body of R ventricle & is incompetent
*Part of the right ventricle is above the valve (atrialized)
*A R to L shunt occurs through the foramen ovale
*Cyanosis, No cardiomeg, No CCF
*Murmur: Pansystolic murmur & diastolic scratch @ xiphisternum

S/I:
ECG = WPW picture (50% of pts)
RA enlargement (tall P's)
1st ° AV block (prolonged PR)
Atypical RBBB
Might present with AV nodal reentry tachycardia (antidromic)
CXR = Oligaemia

Mx:
Pharmacological:
If reentry tachycardia---procainamide
Bblockers, Ca^{2+}blockers & digoxin contraindicated→promote conduction through accessory pathway.
If AF with pre-excitation---procainamide

7) Tricuspid atresia
*Complete absence of tricuspid valve
*Hypoplastic/absent right ventricle
*ASD & VSD present
*Cyanosis, poor feeding, no cardiomeg
*Murmur: No murmur or soft systolic over precordium, Fixed S2
Depending on VSD size→Holosystolic murmur

S/I:
ECG = Left Axis, LVH, P pulmonale
CXR = Oligaemic

Mx:
>>PGE1 to maintain PDA
>>Blalock-Taussig shunt btwn subclavian & pulmonary artery
>>Cavopulmonary anastomosis
>>Fontan procedure to redirect IVC and hepatic vein flow into plm circulation

8) Critical Pulmonary Stenosis (trilogy)
*Critical stenosis→pressure in R atrium & R ventricle greater than left
*R to L shunt occur through foramen ovale or ASD
*Cyanosis, No cardiomeg, dizziness, distended neck veins
*Murmur: Ejection systolic @ 2nd LICS. Soft S2

S/I:
ECG = Right Axis, RVH, P pulmonale
CXR = Oligaemic

Mx:
>>Balloon valvuloplasty
>>Valve replaicement

9) Eisenmenger's Syndrome
*Obstruction due to pulmonary arteriolar vascular disease causing severe pulm HT.
*L to R shunt caused by VSD causes increased bld flow through pulmonary vessels, causing pulm HT, which in turn causes increased pressure in the R side of the heart and reversal of the shunt
*If pulm HT complicates an ASD or PDA, there will also be reversal of the shunt through these defects
*Progressive cyanosis, no cardiomeg
*Murmur: Loud P2. Depending on septal defect.

S/I:
ECG = Right axis, RVH
CXR = Oligaemic, R atrial & ventricle enlargement

Mx:
>>Pulmonary vasodilator Rx
>>Heart-lung transplantation
>>Bilateral lung transplantation

Cx's of cyanotic heart lesions:
- Polycythemia
- Pulm HT
- Stroke
- CCF
- IE
- Arrhythmias

Apart from a CXR & ECG, always do an ECHO, FBC, LFT & ABG as part of your work-up if you suspect a cyanotic heart lesion

Acyanotic Congenital Heart Disease

↑ PBF (L to R shunt lesions)
*Extra bld is displaced through a communication frm the L to the R side of the heart, resulting in ↑ PBF.
*Shunt volume dependant on:
→size of defect
→pressure gradient
→peripheral outflow resistance
*Untreated shunts can result in pulm vascular disease, RVH & R to L shunts
*Eg.ASD/VSD/AVSD/PDA

Normal PBF (Obstructive lesions)
*Eg.AS/Coarct/PS/TR/MR

1) Atrial Septal Defect (ASD)
*3 Types
→Ostium primum – common in Down Syndrome
→Ostium secundum – most common type (50-70%)
→Sinus venosus – defect located @ entry of SVC into RA
*Often asymptomatic in childhood
*Murmur: Ejection systolic @ 2nd L ICS (2-3/6)
Mid diastolic @ 4th L ICS
Widely split fixed S2

S/I:
ECG = Right Axis. RsR in V1 (Partial RBBB)
Mild RVH
CXR = RA & RV enlargement
Plethora

Mx:
>>Natural hx→80-100% spontaneous closure if ASD<8mm
>>If remains patent→CCF & Pulm HT in adulthood
>>Elective surgery/catheter closure @2-5 yrs

2) Ventricular Septal Defect (VSD)
*Most common congenital heart defect (38%)
*Small VSD (majority)
→asymptomatic, normal growth & development
→murmur: early systolic/holosystolic @ LLSB
*Moderate to large
→delayed growth & development, ↓exercise tolerance, recurrent URTI's, "asthma" episodes
→murmur: holosystolic @ LLSB, ± thrill, mid-diastolic rumble @ apex. Loud S2. Intensity of murmur inversely related to size of VSD.
→2° pulm HT & CCF by 2 months

S/I:
ECG = Small VSD → Normal
Large VSD → LVH/RVH/LAH
CXR = Small VSD → Normal
Large VSD → Cardiomeg
Plethora

Mx:
>>Rx CCF & Surgical closure

3) Patent Ductus Arteriosus
*Patent vessel btwn descending aorta & pulm a
*Functional closure within first 1-15hrs of life, anatomical closure within first days of life
*Common in prem babies
*May be asymptomatic or have apneic or bradycardia spells, poor feeding
*Associated ↑HR, bounding pulses, hyperactive precordium, wide pulse pressure (collapsing pulse)
*Murmur: continuous "machinery" murmur, best heard @ L infraclavicular area, Loud S2, Ejection @ 2nd LICS in infants

S/I:
ECG = Normal or biventricular enlargement
CXR = Cardiomeg
Plethora

Mx:
>>Spontaneous closure common in prem babies, less common in term infants
>>Indomethacin, Surgical ligation, catheter closure
>>High risk for IE, antibiotic prophylaxis required until 6 months after closure

4) Atriaventricular Septal Defect (AVSD) a.k.a. Endocardial Cushion Defect
*Common form of this abnormality = Ostium primum + cleft in mitral valve → resulting in a L to R shunt btwn L ventricle & R atrium
*Commonly associated with Downs Syndrome
*Depending on size of defect→CCF early in life
*Murmur: Pansystolic @ apex, Ejection @ 2nd L ICS, Mid diastolic @ 4th L ICS, Loud S2, fixed split

S/I:
ECG = RsR in V1 (Partial RBBB)
CXR = Cardiomeg
Plethora

Mx:
>>Depending on size, complete surgical repair before 3 months of age

5) Coarctation of the Aorta

*Narrowing @ aorta almost always @ level of Ductus
*Commonly associated with bicuspid aortic valve (50%)
*If severe, pt present with shock in neonatal period when the ducts closes
*Often asymptomatic with upper extremity systolic BP of 140-145mmHg
*If associated with other lesions (PDA/VSD)→CCF
*Other Sg's of aortic carctation:

- Collaterals sometimes felt around scapula
- Prominent pulsation in supraclavicular notch
- Bruit over site of coarctation (btwn scapula)
- Upper limb hypertension
- R arm BP > L arm BP (depending on site of coarctation)
- Decreased BP in legs
- Radio-radio delay
- Radio-femoral delay

*Murmur: Ejection systolic @ back, mid diastolic @ apex

S/I:
ECG = RVH in infancy, LVH later in childhood
CXR = Large proximal aorta
3 sg in older children

Mx:
>>Balloon arterioplasty
>>Surgical correction
>>Cx→Hypertension

6) Aortic stenosis

*Aetiology
--congenital: abnormal valve (bicuspid)
--acquired: rheumatic heart disease
*Obstruction→Pressure overload→↑in LVEDP→concentric LVH→Subendocardial ischaemia→LVHF/arrhythmias/chest pain
*Not all LV outflow obstruction = AS
-**Supravalular obstruction**→congenital fibrous diaphragm above aortic valve + mental retardation + hypercalcaemia = Williams syndrome
-**Subvalvular diaphragm**→congenital fibrous ring
-**HOCM**→septal muscle hypertrophy
*Sx's: asymptomatic, exertional chest pain, syncope, dyspnoea (CCF)
*Sg's: small volume pulse, older children→pulses parvus et tardus (slow upstroke), heaving apex
*Murmur: ejection systolic/mid systolic 2nd R ICS, radiating to neck, musical quality @ apex (Gallavardin phenomena), reverse splitting, soft S2, S4 (early in disease—LV hypertrophy), S3 (later in disease—LV dilate)
*Cx: sudden LV failure, arrhythmias, IE, AV block

S/I:
ECG = LVH
CXR = Large proximal aorta

Mx:
>>Surgical/Balloon valvuloplasty
>>Valve replacement
>>Exercise restriction
>>IE prophylaxis

7) Pulmonary Stenosis

*Aetiology
--Congenital: abnormal valve
--Acquired: Rheumatic heart disease
*Obstruction→Pressure overload→RVH→RHF
*Again, not all RV outflow obstruction = PS (90% is Valvular though)
-supravalvular
-subvalvular
*Usually part of congenital heart disease (eg.TOF), or in association with other syndromes (eg.congenital rubella, Noonan syndrome)
*Critical PS = inadequate pulmonary bld flow, dependant on ductus for oxygenation, progressive hypoxia, cyanosis
*Sx's: asymptomatic to CCF
*Murmur: systolic @ 2nd L ICS, pulm ejection click, normal/loud/soft S2, right S4

S/I:
ECG = RVH, Right axis
CXR = Dilated post-stenotic pulm artery, RV enlargement, normal lung vascularity

Mx:
>>Balloon valvuloplasty

8)Mitral valve incomtetence

*Chronic MR→gradually ↑flow across MV during systole→progressive LAE→↓fraction of SV flows forward→LV dilate→CCF
*MR causes LV dilatation→causes annulus dilatation→worsens MR!!!
*Aetiology:
→Annulus = dilated cardiomyopathy, myocarditis, CCF
→Leaflets = congenital, IE, RF
→Chordae = IE
→Papillary muscles = HOCM, aneurysm, infarct
*Sx's: Few sx initially due to gradual LAE, later dyspnoea, orthopnoea, lethargy, palpitations
NB = Hx of Rheumatic Fever!!
*Sg's: Apex displaced (LV hypertrophy, later dilatation)
Soft/absent S1, S3 usually present, pansystolic murmur radiating to back & axilla

S/I:
ECG = LAE, LVH
CXR = LAE, LVH, Pulm HT
ECHO = Aetiology, severity, LV Fx, EF

Mx:
Asymptomatic = Serial ECHO's, IE prophylaxis
Symptomatic = Digoxin & Diuretics
Surgery = For acute MR, Persistent Sx's despite medical Rx, LV DysFx.

Cx: CCF, Pulm HT, Pulm oedema, AF, IE

9)Mitral Valve Prolapse (Barlow syndrome)

*3-5% of population, leaflets displace into LA during systole
*Aetiology
→Connective tissue disease (eg.Marfan)
→Alone, my be associated with pectus excavatum, straight back syndrome, other skeletal abnormalities
→Myxomatous degeneration of chordae & leaflets
*Sx's: chest pain (prolonged, non exertional, stabbing)
Other sx'x of MR
*Sg's: click-murmur syndrome
Mid-systolic click
Mid to late systolic murmur (regurge after prolapse)

S/I:
ECG = non-specific T changes
ECHO = systolic prolapse of MV leaflet into LA
CXR = same as MR

Mx:
Asymptomatic = good prognosis
Symptomatic = as for MR

Cx: Same as MR (depends on MR severity)

10)Mitral stenosis

*MS→LV inlet obstruction→LAE→↑LA pressure→Pulm HT→↑in RV pressure→RHF
*Rare in children, but has been described in kids of 8yrs
*Aetiology = Rheumatic heart disease most common
*Sx: poor effort tolerance, dyspnoea, coughing
*Sg: Sg's of RHF, giant a-waves (pulm HT), tapping apex (not displaced), palpable S1, palpable S2 (pulm HT), L parasternal heave, loud S1, Loud P2, opening snap, mid diastolic rumble @ apex, pulm regurge (graham steel)

S/I:
ECG = Pmitrale
CXR = LA enlargement (double contour, spraying of carina, pulm congestion)
ECHO = Thickened valve, leaflet fusion, LAE, EF

Mx: Rx AF, IE prophylaxis, Rx CCF, surgery
Cx: IE, CCF, EF, Emboli

11)Aortic incompetence

*AR→bld flow back into LV→volume overload→LV dilatation→↑SV→↑SBP & ↓DBP
*Aetiology = Supravalvular (aortic root disease with dilatation)
→ connective tissue diseases
→ dissecting aortic aneurysm
Valvular
→Congenital abnormalities (bicuspid AV)
→Connective tissue diseases
→RF/IE
*Sx'x: dyspnoea, fatigue, palpitations
*Sg's: (chronic AR: Not all the sg's as in adults are seen)
→Distended neck veins during systole (corrigan's pulse)
→Bounding/waterhammer pulse
→pistol shot over femoral arteries
→Systolic-diastolic femoral murmur (Duroziez's murmur)
→Heaving apex (hyperdynamic), soft S1, soft/absent S2, S3 in severe disease, ejection systolic 2nd R ICS, Austin flint (diastolic rumble) @ apex.

S/I:
ECG = LVH, LAE
CXR = LV enlargement, LAE, aortic root dilatation
ECHO = regurge

Mx: restrict activities, rx CCF, rx arrhythmias, surgery if severe, IE prophylaxis

Cx: CCF, IE, arrhythmias

Heart Failure

What is Heart Failure?-----"a Clinical syndrome of effort intolerance due to a cardiac abnormality, usually accompanied by neurhormonal adaptations resulting in sodium and water retention" (Parker,M.1989)

HF is rarely seen in paediatric practice, & is usually encountered in babies. Clinical features are different frm those in adults, i.e. babies do not climb stairs or need extra pillows @ night. Feeding is the only exertion they undertake, & not being ambulant bipeds @ this time of life, their ankles do not swell up.

Sx's: **1**

Infant: fatiguability, feeding difficulties, exertional dyspnoea, resp distress, vomiting, lethargy, cyanosis, sweating, sudden weight gain.
Child: fatiguability, ↓appetite, failure to thrive, resp distress, syncope, frequent URTI's or "asthma" episodes.
Orthopnea, PND, oedema uncommon in kids

Sg's:
RHF = distended neck veins, hepatomeg, splenomeg, ascites, oedema (puffy eyes), S3 (gallop if ↑HR), ↑HR
LHF = pulm oedema (crackles), pleural effusion, resp distress, cyanosis, ↑RR, ↑HR, S3, pulses alternans
RHF&LHF = Failure to thrive, dysmorphic features associated with congenital syndromes

4 Key features: ↑HR, ↑RR, hepatomeg, cardiomeg

S/I: **2**

CXR---
Enlarged/abnormal cardiac shadow
Lung fields
--- ↑ pulm vascular markings (plethora)
Eg. L to R shunt (large VSD)
--- ↓ pulm vascular markings (oligaemia)
Eg.PS, (decreased pulm bld low)
---Sg's of pulm oedema
---Pleural effusion
ECG---
Rate/rhythm, Axis, Hypertrophy of either ventricles
ECHO--- Anatomical abnormality, EF, monitor progression
UCE/LFT/Hb

APPROACH

1)Sx's/Sg's
---Infant
---Child
2)S/I
---CXR
---ECG
---ECHO
---BLDS
3)Aetiology
4)Mx
---Rx aetology
---General
---Drugs
5)Cx's
6)Ability to feed & nutritional status
7)Daily weigh the pt. Is the diuretic effective?

Aetiology: **3**

>CHD (Cyanotic & Acyanotic)
>Cardiomyopathy
>Myocarditis
>Arrhythmias (esp.Tachydysrhythmias)
>Anaemia
>Cor pulmonale

Mx: **4**

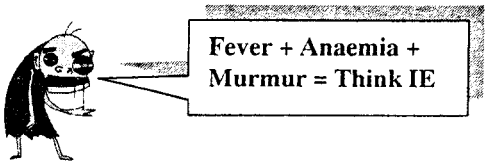
1)Correct underlying cause
2)General:
*Nurse baby @ 60°
*O_2 by facemask
*Restrict fluid intake (60ml/kg/day)
*increase caloric intake
3)Drugs:
Diuretics = **Furosemide** most effective diuretic for Acute HF. Initially IV, oral later.
Dose: 1-6mg/kg/day (0.5-1mg/kg/dose)
NB: Replace K^+. 1-2 mmol/kg/day.
Spironolactone (K^+-sparing diuretic)
Dose: 2-3mg/kg/day in 2 divided doses
Inotropic agents = **Digoxin** (+inotrope/↑ parasymp activity/↓ symp activity)
Dose: *preterm:* 0.04-0.05 mg/kg (TDD) in 4 divided doses
Maintenance = ¼ of TDD
2months – 2years: 0.06-0.08 mg/kg (TDD) in 4 doses
Maintenance = ¼ of TDD
>2 yrs: 0.04-0.06 mg/kg (TDD) in 4 divided doses
Maintenance = 0.01mg/kg/day
Dopamine (in severe CCF)
Dose: 3-10ug/kg/min
Afterload reduction = ACEI, eg **Captopril** (esp.in dilated cardiomyopathy)
Dose: 0.5-6mg/kg/day in 4 divided doses

"All infants presenting with HF, Rx is **digoxin** & **diuretics**, regardless of the dx"- Coovadia
TDD = Total digitalizing dose

Cx: **5**

>Impaired liver Fx
>Impaired renal Fx
>Thromboembolism
>Arrhythmias

In older children (>13yrs), Framingham criteria can be used to Dx



Infective Endocarditis

-Infection of encocardium, usually caused by organisms lodging on abnormal valves
-Uncommon disease: 1/4600 hospital in-patients, usually children 7-10 yrs
-Congenital heart disease has replaced RF as the main cause of susceptibility to endocarditis
-Pathogenesis = [Bacteraemia + Abnormal cardiac endothelium]
Portal of entry (oropharynx/nosocomial infection)→Bacteraemia→Turbulent flow over diseased valves→Deposition of bacteria→Vegetation (clump of fibrin, plts, wbc, bacteria)→Endocarditis→Septic emboli
-Valve involvement = MV>>AV>>TV>>PV (Sub-acute & acute classification not used anymore)

1) Sx's:
Fever, flu-like sx's, cough, chills, dyspnoea, night sweats, rigors
Poor dental hygiene? Recent surgery/dental procedure?

APPROACH:

1) **Sx's**
2) **Sg's**
3) **S/I**
4) **Dx**
5) **Mx**
6) **Prevent Cx**
7) **Prophylaxis**

2) Sg's:
General – Clubbing
Anaemia
Cardiac – Murmur
CCF
Pyrexia
Anaemia
Arthralgia
Skin lesions: Oslers nodes – 'ouch', raised, painful 3-15mm on soles/palms
Janeway lesions – 'pain away', painless 1-2cm on toes/palms/fingers/toes
Splinter haemorrhages – proximal nailbed
Petechia – mucous membranes/skin
Eye – Roth spots (emboli)
Splenomegally
CNS – emboli – focal sg's
Renal – Haematuria
Embolic Phenomena – emboli – abscesses in organs (brain, heart, kidney, spleen)

Full-blown picture rarely seen in kids
Fever, anaemia, murmur earliest & most common in kids

3) S/I:
Bld cultures – 3 specimens @different Times
Bld – FBC = anaemia/↓Plts
CRP/ESR/PCT = ↑
UCE/LFT/CMP
Urine analysis – haematuria
ECG – dyssrhythmias, ST↑ AvR
CXR – cardiomegally
ECHO – vegetations (if>2mm)

4) Dx:
*High index of **suspicion** in pt's with fever of unknown origin, & fever in rheumatic heart disease or congenital heart disease
*Positive bld cultures confirms dx** (can be negative). When in doubt = Rx
*Modified Duke criteria**

Modified Duke Criteria
2 Major/1major + 3minor/5minor

Major
+ve bld cultures: *typical organisms in 2 cultures
*3 +ve cultures>12hrs apart
Endocardium involvement: *+ve ECHO
*new valvular regurge
Single +ve culture for **coxiella burnetti**

Minor
Predisposition (Rheumatic/Congenital)
Fever
Vascular phenomena (petechia/splinter)
Immunologic phenomena (Osler/Janeway/Roth/Glomerulonephritis)
+ve bld culture that do not meet major criteria
+ve ECHO that do not meet major criteria

ORGANISMS
Staph Aureus – commonest in SA children (break in skin, eg.lesions/sepsis/IV's)
Strep Viridans – dental extractions/oral surgery, prosthetic valves
Staph Epidermidis
Enterococcus
Gram –ve HACEK organisms: Haemophilus, Actinobacillus, Cardiobacterium, Eikenella, Kingella
Candida

CULTURE –VE IE:
Cause = prior antibiotics
Organisms failing to grow in culture: coxiella burnetti, Chlamydia, bartonella, legionella

6) Cx
>CCF
>Systemic emboli
>Intracardia abscess formation
>Glomerulonephritis 2° to immune complex deposits→RF

5) Mx
Antibiotics: Depend on organism sensitivity. Empirical Rx = penicillin + genta. Rx for 6 weeks.
Surgery: HF due to valve damage, Antibiotic failure, valve perforation, mycotic aneurisms

7) **Prohylaxis** (oral/dental surgery)= Amox 50mg/kg 1 hour before, then 25mg/kg 6hrs later

NB: Sx's & Sg's appear 2-3 weeks after the throat infection.

Rheumatic Fever

Def: Inlam disease, due to an autoimmune reaction, triggered by molecular mimicry btwn the cell wall M proteins of the infecting strep pyogenes and cardiac myosin & laminin

Epidemiology: children & young adults (5-15)
Aetiology: 3% of untreated group A b-haemolytic streptococ pharyngitis develop acute rheumatic fever
Path: Affects the **heart, skin, joints, CNS**
All 3 layers of the heart might be affected
Rheumatic carditis→**Aschoff** nodules (granulomatous lesion with central necrotic area)
Small valvular vegetations may develop
Synovial membranes acutely inflamed→subcutaneous nodules

S/I:
→**Throat swab**: all children complaining of a sore throat
→**Bld**: ASOT – increased titre
Dnase B – increased titre
WCC – increased
CRP/ESR – increased
→**CXR**: cardiomeg/pulm oedema
→**ECG**: AV block/features of pericarditis
→**ECHO**: cardiac dilatation/valve abnormalities

Hx:
>Throat infection?
>Previous RF?
>Fever?
>Painful joints?
>Rash?

APPROACH
1) **Hx & Exam**
2) **S/I**
3) **Dx→Jones Criteria**
4) **Mx**
5) **Cx**

Modified Jones Criteria (1992)
Evidence of recent strep infect + 2 major/1major + 2 minor

Evidence of strep infect = recent pharyngitis, hx of scarlet fever, +ve throat swab, ASOT increased, increased Dnase B titre

Major =
1)Polyarthritis: Flitting arthritis, painful, swollen, tender (the large joints)
2)Erythema Marginatum: Transient erythematous rash, mostly trunk
3)Subcutaneous Nodules: Painless, pea-sized, hard, mobile, over extensor surfaces, spinous processes, occiput, rarely scalp
4)Chorea: (Sydenhams chorea/St. Vitus dance) Spasmodic/unintentional/choreiform, develop late. Mostly girls btwn 7-14.
5)Carditis: (1 or more of following)
1 New/changing murmur→endocarditis
2 Cardiomeg→if associated with soft heart sounds & ↑HR, suspect myocarditis
3 Friction rub/pericardial effusion→pericarditis
all CCF→pancarditis(involvement of endo-,myo-,and pericardium)

Minor =
1)Previous rheumatic fever
2)Fever
3)Arthralgia
4)Prolonged PR interval
5)↑WCC/CRP/ESR

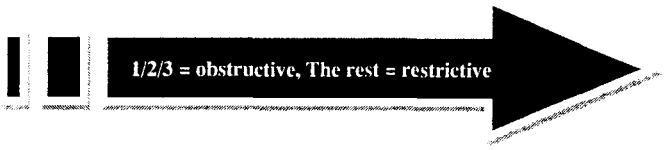
Mx:
1)Carditis:
Bedrest
Aspirin 60mg/kg/day (until no sx's/sg's)
Corticosteroids (prednisone 2mg/kg/day until no sx's)
Mx CCF
2)Anti strep Rx
a)immediate→Benzathine pen, 1.2 mil IU IMI stat, then Phenoxymethyl pen, 500mg bd x 10 days
b)prophylaxis→Phenoxymethyl pen, 500mg/day x 5 years after last attack
3)Immobilize joints in severe arthritis
4)Chorea→Haloperidol (0.025-0.05mg/kg/day), or phenobarb (3-5mg/kg/day)

Cx's:
*60% with carditis = chronic rheumatic heart disease
MV(70%), AV(40%), TV(10%), PV(2%)
*Recurrence = precipitated by other strep infections

Mx of Chronic Rheumatic Heart Disease
(thus valvulopathy secondary to RF)
>>Prevent recurrent attacks
>>Surgical intervention/balloon valvuloplasty

RF can be prevented by treating streptococ throat infections with penicillin within 1 week of the onset of Sx's

Cardiomyopathy



1)Dilated Cardiomyopathy
2)HOCM
3)ASH
4)Abnormal substance deposition
5)Endomyocardial Fibrosis
6)EFE
7)BeriBeri...
8)Chagas...
9)Inherited disorders...
10)Muscle & nerve
11)AIDS

→Variety of non-inflammatory conditions causing myocardial dysfunction resulting in CCF
→Dx of cardiomyopathy only made once CCF has been treated & all other causes for CCF have been excluded (valvulopathy, congenital heart disease etc)
→Common in Africans, NB cause of CCF

Dilated Cardiomyopathy (Most common type)

Aetiology: Idiopathic
Collagen vascular disease
Infectious (post viral (Coxsackie), HIV)
Metabolic (Uraemia, nutritional deficiency)
Clin pic: CCF, Systemic/Pulm emboli, *arrhythmias*, sudden death. Pansystolic murmurs (MR/TR) might be present due to dilatation of valve rings.
S/I:
ECG = Poor Rwave progression
Non-specific ST-T wave abnormalities
L-axis, LVH, Biatrial enlargement, arrhythmias (ectopics, AF, WPW)
CXR = Global cardiomeg with pulm congestion
ECHO = 4-chamber enlargement, ↓EF, valve incompetence

Diff Dx: Acute myocarditis, pericarditis, RF.
Mx = Rx underlying cause
Rx CCF, Arrhythmias
Anticoagulate when EF<20%, or AF, or Hx of emboli
Immunize – All up to date + influenze
Surgery→Cardiac transplant

Cardiomyopathy in disorders associated with deposition of abnormal substances

→mucopolysaccharides in Hurler's & Hunter's syndrome
→glycogen in Pompe'sType 2 glycogen storage disease

Endomyocardial Fibrosis

>Common cause of CCF in Central Africa
>Mitral/Tricuspid valve incompetence →striking clin feature
>Fibrosis of endocardium and valves→pathological feature
>Sometimes = Cavity of RV completely obliterated

Cardiomyopathy associated with Chagas disease

>South & Central America
>Parasite = Trypanosoma cruzi
>Infection in 1st year of life→acute interstitial myocarditis→fibrosis

Cardiomyopathy in AIDS

>CCF
>Damage to conductive system
>Pericardial effusion,
>Arteriopathy involving small/medium sized vessels

Hypertrophic Obstructive Cardiomyopathy (HOCM)

Hypertrophy of myocardium, especially the ventricular septum & papillary muscles, causing obstruction of the outflow tract & distortion of valves
Aetiology: Familial
Clin pic: Asymptomatic, Dyspnoea, CCF, arrhythmias, sudden death, pulses bisferiens, heaving apex, normal/reversed S2, S4
S/I:
ECG = LVH, prominent Q, tall R in V1, left axis, BBB
CXR = ± cardiomeg
ECHO = LVH, LAE, MR, diastolic dysfx

Mx = Avoid extremes of exertion
IE prophylaxis
Medical → Bblocker + CCB. Not digoxin (↑myocardial contractility)
Surgical→myomectomy/ethanol ablation
Rx arrhythias

Asymmetrical Septal Hypertrophy (ASH)

--Infants born to diabetic mothers particularly if uncontrolled –
→Baby asymptomatic = Rx as for HOCM
→Usually resolves in 6 months

Primary Endocardial Fibroelastosis (EFE)

>Cause = unknown
>Greatly thickened endocardium(mostly LV)----restricting contraction of heart
>Presents within 1st 2yrs of life→CCF
>ECG = LVH!!!
>Heart size does not diminish with Rx

Cardiomyopathy associated with BeriBeri

>BeriBeri (due to thiamine deficiency)→high cardiac output failure.
>CNS Sg's = peripheral neuropathy, nystagmus, encephalopathy
>Rx = Thiamine, 50-100mg/day/IM

Cardiomyopathy associated with inherited disorders of muscle & nerves

→Duchenne's muscular dystrophy
→Myotonia atrophica
→Friereich's ataxia

Cardiomyopathy of unknown origin

>Common
>CCF

Pericardial Diseases →

1. **Acute pericarditis**
2. **Chronic pericarditis**
3. **Recurrent pericarditis**
4. **Constrictive pericarditis**
5. **Pericardial effusion & Cardiac tamponade**

1.Acute Pericarditis – Inflam of pericardium
Can be **Dry**, **Fibrinous** or **Effusive**, independent frm its aetiology

Sx: Lower **chest pain/epigastric pain** (positional-better when sitting up, worse when lying down), **fever, poor feeding, dyspnoea, non-productive cough**
Sg:Pericardial friction rub→50% of cases (best heard at left sternal border, with child sitting up), **pleural effusion** may be present

APPROACH
1)Sx's & Sg's
2)S/I
3)Mx
4)Cx

Mx
1)Rx **underlying cause**
2)Analgesia – **NSAIDS**
3)**Steroids**

Cx =
T – Tamponade
R – Recurrence
R – Residual constrictive pericarditis
A – Arrhythmias
P – Pericardial effusion

ECG = ST elevation (saddle shape) > ST resolution > T flattening > T inversion > normal T. ST depression in AvR. PR segment depression in ant/lat/inf leads.
BLDS = FBC, ESR, CRP, LDH (infect markers)
TropI,CK-MB (cardiac markers)
Viral serology, bld cultures, autoantibodies if indicated.
CXR = Normal/cardiomeg (pericardial effusion), pulmonary/mediastinal pathology.
ECHO = If pericardial effusion suspected
CT/MRI = previous tests inconclusive
PERICARDIAL BIOPSY = specific aetiology

CLINICAL PEARL =
TRIAD:
Chest Pain
ECG changes
Friction rub

Aetiology (I AM MR.TIN)
Infectious→
Viral (Coxsackie,Echo,Mumps,EBV,CMV,HIV,Parvo)
Bacterial (Pneumo,meningo,hemophilus,TB)
Fungal (candida,histoplasma)
Autoimmune→ SLE,RA,RF,Reiter's
Metabolic→ Renal failure(uraemia),DKA
Trauma→ Penetrating,foreign body,cardiac surgery
Idiopathic
Neoplasia (primary/secondary)

Most common in kids =

2.Chronic Pericarditis – pericardial inflammation (> 3 months)

Effusive | Adhesive | **3.Constrictive**

Chronic pericarditis→fibrosed thickened pericardium→impair ventricular filling→reduce ventricular Fx.
NB: Constrictive pericarditis = treatable
Constrictive cardiomyopathy = most cases not treatable
Both have similar clinical picture→Thus work the patient up properly!!

It is NB to differentiate btwn chronic inflammatory effusion frm non-inflammatory hydropericardium (CCF)→CCF vs. Pericardial pathology

The diagnostic workup is the same as for acute pericarditis. Detection of curable causes (TB, toxoplasmosis, myxedema, autoimmune, systemic diseases) allows for specific therapy. Symptomatic treatment is same as for acute pericarditis.

Approach
1)Sx/Sg
2)**Confirm Dx & find cause**
>ECG
>BLDS
>CXR
>ECHO
>CT/MRI
3)Mx – Rx cause
Surgical – **resection of pericardium**
Diuretics & salt restriction

Sx = Abdo pain/Fatigue/Dyspnoea
Sg = Mimics CCF→ascites/hepatosplenomeg/ Oedema/ ↑JVP/Kussmauls sg/pulses paradoxus/AF

ECG = small QRS, inverted T, AF
CXR = small heart, pericardial calcification
CT/MRI = if no calcification on CXR—do CT—if still none→cardiomyopathy?
Endomyocardial biopsy = pericarditis vs cardiomyopathy
ECHO = Thickened/calcified wall. ↓EF
BLDS = as for acute pericarditis

Causes = Same as acute pericarditis

CLINICAL PEARL: Diff for Sg's of RHF:
CCF
Cor Pulmonale→Sg's of lung pathology? Hx? CXR? Lung Fx studies
Constrictive pericarditis→Kussmaels Sg? Calcifications on CXR/CT. Normal size heart on CXR
Restrictive cardiomyopathy→none of above, biopsy?
Large Pericardial effusion→Kussmaels Sg? Globular heart on CXR.
Isolated RHF→rare (heart fails in sincitium)

4.Recurrent Pericarditis

Mechanisms suggested to explain recurrence include:
1)Insufficient dose/treatment duration
2)Early corticosteroid rx causing augmented viral DNA/RNA replication in pericardial tissue leading to increased viral antigen exposure
3)Reinfection
4)Exacerbation of the connective tissue disease

Evidence of an immunopathological process includes:
>Presence of anti-heart antibodies
>Quick response to steroid rx
>Sx/sg suggestive of auto-immune disease, eg Lupus.

Re-evaluate the pt
Consider:
1---autoimmune
2---re-infection

5.Pericardial effusion – accumulation of fluid in pericardial sack, often complicate acute pericarditis

Aetiology:
Transudate = CCF, low albumin autoimmune
Exudate = Infective

APPROACH
1)Sx/Sg------NB: Monitor Vitals!!
2)Confirm Dx & find cause
***ECG *BLDS *CXR *ECHO----Procedure of choice**
***Dx tap: transudate/exudates**
3)Mx – Rx cause
Most resolve spontaneously
Small – serial ECHO, NSAIDS
Large – pericardiocentesis
4)Monitor pt (CXR/ECHO/Rx response)
5)Cx - TAMPONADE

Sx = similar to acute pericarditis, cough (phrenic irritation)
Sg = raised JVP, soft S1/S2, impalpable apex
Ewart's Sg→effusion compress base of left lung--dull to percuss below angle of left scapula

ECG = Lowvoltage QRS,Electrical alternans, Flat T
CXR = Enlarged, Globular heart
ECHO = Echo-free zone around heart
BLDS = As for acute pericarditis
Dx tap = MCS, WCC, ANA, glucose, total protein, albumin

6.Cardiac Tamponade – Cx of effusion. CLINICAL DX!! = MEDICAL EMERGENCY

Pathophys: ↑ pericardial pressure→↓ venous return→↓ diastolic ventricular filling→↓CO→↓BP+venous congestion
MAIN PROBLEM = CIRCULATORY COLLAPSE→DEATH

Sx = Resp distress, flushed, syncope, sweating
Sg = Tachycardia
Becks triad = Distended neck veins
Muffled heart sounds
Hypotension
Pulses paradoxus
Kussmauls Sg
Impalpable apex
Electrical alternans (ECG)
Pulses alternans

APPROACH:
1)Pick it up!!
2)ABC
3)Tap it
4)Find cause
5)Further Mx

Do emergency pericardiocentesis
Preferably guided by ultrasound, but do not wait for sonar machine!!! Just do it!!
(use CVP set, so fluid can be tapped through it) When the needle is in the pericardial sac, it will move as the heart contracts (you are going to think that you sticked it into the ventricle)----do not freak out. The bld/fluid aspirated should not clot. Confirm placement by US when finished. Do not tap drv.

Bld already drawn should be sent away for same tests as an effusion.. Sent pericardial fluid away for same tests.

Airway = Intubate & ventilate if resp failure. Maintain airway
Breathing = 100% O2
Monitor SATS, RR
Circulation = 2xIV. 20ml/kg fluid bolus R/L to temporarily ↑ CO. Draw same blds as for effusion. Monitor BP, HR, ECG, Temp, Urine output

Rx underlying cause
Contact Cardiothoracic surgeon ASAP. Pericardiotomy if effusion recurrent

Constrictive Pericarditis Vs Cardiac Tamponade

	Constrictive pericarditis	Cardiac tamponade
Kussmael	+	JVP to ↑ to see
Pulses Paradoxus	33% of cases	Always
BP	↓	↓↓↓↓↓
ECHO	No effusion	Effusion

Myocarditis
→Inflammation of myocardium.
→Rare condition

Approach:
1)Sx/Sg
2)S/I
3)Mx
4)Cx

Sx's:
Newborn & infant→Failure to thrive/Poor weight gain/Feeding difficulties/Resp failure/Fever
Child > 2→ Abdo pain/Chest pain/fatigue/swelling of legs, feet,face/ Palpitations

Sg's:
CCF/Tachycardia/Tachypnoea/Decreased urine Output/Dysrhythmias/ Sg's of systemic infection, including skin rash & arthralgia/Loud S3

Cx: Dilated Cardiomyopathy
CCF
Dysrhythmias

Mx: Aim→Control CCF, thus digoxin & diuretic
Rx identifiable cause
Anti-inflammatory: corticosteroids & IV immunoglobulin

S/I: CXR (cardiomeg)
ECHO (valvulopathy secondary to annular dilatation)
Blds (Cultures/infective markers/UCE/LFT/Viral isolation)
ECG (heart block/inverted T in lead 1,V5,V6)
Endomyocardial biopsy (most accurate way to make Dx)

Causes:
>Coxsackie
>Influenza
>Adenovirus
>Polio
>Rubella
>Lyme disease
>Allergic reaction to certain medications
>Autoimmune conditions

how normal are you on a scale of -10 to 0?

In this chapter we will only focus on
Kawasaki disease & Takayasu's disease

Arteritis

Before we start, lets just zoom out a bit and see where this fits in.

Vasculitis refers to a heterogenous group of disorders that are characterized by inflammatory destruction of blood vessels. Both arteries and veins are affected. Vasculitis is primary due to leucocyte migration and resultant destruction. Although both occur in vasculitis, inflammation of veins (phlebitis) or arteries (arteritis) on their own are separate entities.

Classification:

Large vessel vasculitis	Medium vessel vasculitis	Small vessel vasculitis
Takayasu arteritis	Polyarteritis Nodosa (PAN)	Wegener's granulomatosis
Giant cell (temporal) arteritis	Kawasaki disease	Churg-Strauss arteritis
	Isolated CNS vasculitis	Microscopic polyarteritis
		Hypersensitivity vasculitis
		Henoch-Schonlein purpura
		Essential cryoglobulinemic vasculitis
		Vasculitis secondary to connective tissue disorders

DIFF Dx for Kawasaki:
→Group A strep infect
→Staph toxic syndromes
→Measles
→Drug reactions
→Rickettsial infect
→Infectious mononucleosis
→Other rheumatic diseases
→Stevens-Johnson

Kawasaki Disease a.k.a.mucocutaneous lymphnode syndrmoe
*Acute vasculitis of **unknown aetiology**
*Systemic necrotizing medium-sized vessel vasculitis
*Common cause of acquired heart disease in children
*Peak age<5 yrs. Orientals>Blacks>Caucasians. M>F

Approach:
1)Clin pic + diagnostic criteria
2)S/I
3)Mx
4)Cx

Diagnostic Criteria:
Fever persisting > 5 days AND 4 of the following:
→ bilateral nonpurulent **conjuncticitis**
→ **red fissured lips, strawberry tongue, erythema of oropharynx**
→ changes of the **peripheral extremeties**
----acute phase:erythema, oedema of hands and feet, groin peeling
-----subacute: peeling from tips of fingers and toes
→ **polymorphous rash**
→ **cervical lymphadenopathy** > 1.5 cm in diameter

Always exclude **scarlet fever** and **measles**

Atypical Kawasaki disease= < 5/6 diagnostic criteria but coronary artery involvement.

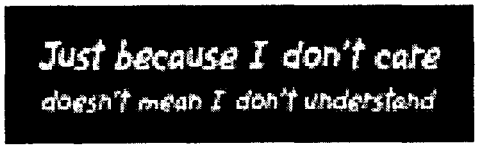
S/I:
Blds = FBC: normocytic anaemia, later thrombocytosis
ESR & CRP: ↑
LFT: evidence of hepatic inflammation & ↓ serum albumin
ECG = arrhythmias
US/CT = gallbladder enlargement
ECHO = coronary artery aneurysms, confirm with ANGIO
URINE = proteinuria
LP = aseptic meningitis

Associated features:
Acute phase (as long as fever persists, about 10 days)
→most of Dx criteria present
→irritability, aseptic meningitis, myocarditis, pericarditis, CCF, diarrhea, pancreatitis, urethritis, gallbladder enlargement
Subacute phase (resolution of fever, peeling of skin, 11-21 days)
→arthritis
Convalescent phase (lasts until ESR & Plts normalize > 21 days)
→Coronary artery aneurysm, aneurysm rupture, MI, CCF, arthritis may persist

Cx:
*Aneurysms of the proximal portions of the coronary arteries (in 20-25% of untreated children, 4-8% if treated within 10days of fever)
*50% of aneurysms regress within 2 yrs
*20% develop stenosis with risk of MI
*Risk factors for coronary disease = male, age<1 or >9 yrs, fever > 10 days, thrombocytosis, leucocytosis
*Endothelial dysfunction with early onset atherosclerosis

Mx
→IV immunoglobulin
→Salicylate therapy: Aspirin high dose until fever subsides. Except for Kawasaki and a few others, aspirin is not normally given to children < 12, due to its association with Reye's syndrome.
→Corticosteroids
→Pt's not responding to above meds: cyclophosphamide & plasma exchange
→Follow up with 2D-echocardiograms

So, how do you feel today? Well, today I feel like shouting this from bridge C on level 9, as loud as I can ...



But this is how I really feel...

Takayasu's disease a.k.a. pulseless disease
*large vessel vasculitis of unknown aetiology
*Mostly affect young women in 2nd & 3rd decade
*Has been reported in children of 6 months & adults of every age
*Characterized by granulomatous inflammation of aorta & its major branches, leading to stenosis (90% of pt's), thrombosis & aneurysm formation.
*Lesions in TA are segmental with a patchy distribution

Approach:

1) **Clinical picture**
2) **S/I**
3) **Mx**
4) **Cx**

Sx:
Systemic = Fever/Fatigue/Weight loss/Myalgia/Arthralgia/Seizures
Skin rash (Erythema nodosum, pyoderma gangrenosum)/Headaches/Dizziness/syncope
Related to ischaemia = Visual disturbances (blurred/diplopia)
Ischemic stroke/TIA
Carotidynia
Abdo pain
Claudication (rare in children)
Sg: Assymetrical pulses (late sg is absent radial pulses)
>30mmHg Bp difference btwn arms
Poststenotic dilatations producing what appear to be bounding pulses
Hypertension (renal artery stenosis→ ↑ renin production)
Subclavian bruits
Fundoscopic→ Retinal haemorrhages/Cotton-wool exudates/Venous dilatation & beading/Microaneurysms/Optic atrophy
Reported skin lesions→ erythema nodosum-like lesions, pyoderma gangrenosum, leukocytoplastic vasculitis, panniculitis

S/I:
BLDS =
FBC – normocytic normochromic anaemia (50% of pt's)
Thrombocytosis
CRP/ESR - ↑
LFT - ↑ transaminases & hypoalbuminaemia
Antiendothelial antibodies - ↑
Antinuclaer antibodies – negative
Rheumatoid factor - ↑ in 15% of pt's
Immunoglobulins (G,M,A) - ↑
Arteriography (invasive/MRI) =
NB for making Dx
Stenosis – 90%
Aneurysm – 27%
Gallium-67 radionuclide scan =
Increased uptake in aorta & branches
CXR = widening of ascending aorta, irregular descending aorta, aortic calcifications, rib notching
ECHO =
Evaluate aortic valve Fx

Mx:
Medical: Goal→Control active inflammation, normalize clinical & laboratory parameters while preventing further vascular damage, Mx symptoms (eg.hypertension & seizures)
→Daily high-dose corticosteroid until all evidence of active inflammation has resolved (prednisone 1-2mg/kg/day for 4-6 weeks)
→ Pts not responding: weekly methylprednisone infusion (risk of steroid-induced toxicity), or low-dose weekly methotrexate, or cyclosporine, or mycophenolate mofetil, or ifliximab.
Surgical: Following the acute phase, pt's with fibrotic changes require surgery for symptomatic stenotic occlusions, either by percutaneous angioplasty/stenting, or by resection and grafting.

Classification:
Type 1 = disease of aortic arch & its branches
Type 2 = disease of thoracic & abdominal aorta
Type 3 = Extensive disease

Cx:
>CCF due to AI, myocarditis, hypertension
>aortic aneurysms, thrombosis, rupture
>Ischaemic stroke
>MI
>Hypertension

Diff Dx for TA
→ Acute lymphoblastic leukaemia
→Behcet Syndrome
→Hodgkin Disease
→PAN
→RF
→Fever of unknown origin

Dysrhythmias in children (this is not a course on ECG's, but just a short summary of common childhood dysrhythmias. Please refer to "ECG made easy" and "Principles and Practice of Medicine – Davidsons" if you don't understand)

Sinus arrhythmia→Phasic variations with normal respiration. In almost all normal children.

Premature atrial contraction (PAC)→May be normal variant or can be caused by electrolyte disturbances, hyperthyroidism, cardiac surgery, digitalis toxicity.

Premature ventricular contraction (PVC)→Common in adolescents. Benign if single, uniform, disappear with exercise, no associated structural lesions. If not benign, may degenerate into more severe dysrhythmias.

Supraventricular Tachycardia (SVT)→Most frequent sustained dysrhythmia in children. Can be life-threatening if ↓↓in CO. Caused by re-entry via accessory pathway (AV node most common site). Characterized by a heart rate > 210 bpm. Rx: vagal maneuver (press eye, vulsalva, ice on face), adenosine (50mcg/kg IV stat, increasing by 50mcg/kg increments every 2 minutes up to a max of 250 mcg/kg) , dogoxin (except in WPW), b-blocker, DC cardioversion (2-4 J/Kg) if medical Rx fail or haemodynamic unstable.

Complete Heart block→Congenital heart block can be caused by maternal RHO antibody formed in mother with CVD. Clinical symptoms related to level of block. The lower the block, the greater the symptoms of inadequate CO. Symptomatic pt's need a pacemaker.

Atrial flutter→May be present at birth---leading to CCF. Rx: digoxin. Flutter resistant to medical Rx: DC cardioversion.

PULMONOLOGY

KOBUS - PE

RESPIRATORY EXAMINATION
UPPER RESPIRATORY TRACT INFECTIONS (PLS SEE ENT)
LOWER RESPIRATORY TRACT INFECTIONS
→LARYNGOTRACHEO-BRONCHITIS
→TRACHEOBRONCHITIS
→BRONCHIOLITIS
AIRWAY OBSTRUCTION
→EXTRATHORACIC
→INTRATHORACIC
SUPPURATIVE LUNG DISEASE
DISEASE OF THE PLEURAL CAVITY

PULMONOLOGY

Respiratory Examination
Upper respiratory tract infections (pls refer to ENT chapter)
Lower respiratory tract infections:
→Laryngotracheo-bronchitis
→Tracheobronchitis
→Bronchiolitis
→Pneumonia
→Pertussis
→TB (pls refer to TB chapter)
Airway obstruction
→Extrathoracic
→Intrathoracic
Suppurative lung disease
Disease of pleural cavity

Respiratory Exam

1-General Inspection
2-Clothes
3-Hands
4-Face →Nose
→Ears
→Mouth/throat
5-Neck
6-Chest
7-Finishing off

1) General Inspection: →Age of child
→Alert/Awake? Is he running around? Fat/Skinny/Sweating
→Who is he with?
→O2 = how much/what method (mask/nasal prongs)
→IV = what fluids/how much/central line
→Catheter
→Sg's of resp distress? Any audible cough/wheeze/stridor?
→Acute vs Chronic

2) Clothes: Next it is best to take the child's clothes off. Be sensible. If the child is < 2, then ask the parents to help. If the child is a bit older, ask them to take their own clothes off. In older kids – be wary of privacy.

3) Hands: Clubbing (think CF/chronic supporative lung disease)
Tremor (think B2 agonist, eg.salbutamol, fenoterol)
Check capillary refill
Feel temp of hands. Cyanosis?
Check radial/brachial pulse (radial difficult in small kids)
Check resp rate @ same time

It is often a good idea to leave the throat for last, as "gagging" might upset the child, making your examination a bit more difficult.

4) Face:

Nose: Watery/red?
Nasal polyps (think CF)

Ears: TAKE THE TEMPERATURE
ALWAYS look inside! ALWAYS!! Bulging tympanic membrane/inflamed/light reflex lost/perforation?
In infection, wax is less likely, as the high temperature often melts it. If your view is obstructed by wax→ not necessary a bad sign.

Mouth & throat: Look @ lips & under tongue for cyanosis
With torch & tongue depressor, examine oropharynx for sg's of infection. Tonsils enlarged/membrane/pus? Infection in laryngopharynx will generally not have any throat sg's.

NB: If you suspect acute epiglottitis→Do NOT examine the throat!! The child will cry→ Upper airway obstruction→ DEATH. Call an ENT & Anesthesiologist ASAP.

5) Neck: Lymphnodes (examine in the same way as in an adult)
Trachea central? In the OSCE, just say you would check it→unpleasant and will upset the child.
Tracheal Tug? (trachea is pulled posteriorly & superiorly during inspiration, and results from recruitment of accessory muscles in laboured breathing)→think resp distress

6) Chest:

Remember: hyperinflation = obstructive lung diseases = Asthma, Bronchiectasis & Cystic fibrosis

Inspection:
Any scars (eg.surgery for meconium ileus in CF)
Rashes
Hickman line (Broviac line)
Subcostal/Intercostal recession (think resp distress)
Hoover sg - Hyperinflation→flattened diaphragm contracts inwards instead of downwards→paradoxically pulling the inferior ribs inward
Harrison's sulcus = 2 symmetrical sulci, horizontal, at the lower margin of the anterior thorax, at the attachment of the diaphragm. A sg of prolonged resp distress in children. Most commonly present in children with asthma who have required an in increased respiratory effort over several months. Also present in Rickets where there is insufficient calcium for bone mineralization→the soft ribs are distorted by the pull of the diaphragm.
Chest deformity

Pectus Excavatum
Pectus Carinatum
Barrel chest------Hyperinflation

Palpation:
Chest expansion→In young children, you only need to check it once, usually in the front. In older children with a larger thorax, you should check it 4 times: twice in front, and twice at the back (under axilla, and at the bottom of the thorax)
Measure chest expansion with tape→measure at full inspiration and full expiration
Tactile fremitus →poor sg in consolidation, atelectasis or space occupying lesion, but ↓ fremitus in pleural Effusion is often detectable.
Apex displaced/impalpable?

Percussion: Difficult in small children (<2). Always compare left and right. Stonydull/dull/resonant/hyperresonant?
↓cardiac dullness/liver displaced down? (think hyperinflation)

Auscultation: NB: Compare sides and listen to all the lobes, including under the axilla and to the apices above the clavicles.

Bronchial/vesicular breathing/crackles/wheeze/aegophany/vocal resonance/whispering pectoriloquy

7) Finishing off: →Feel for the liver. If lower than expected, if might be displaced by hyperinflated lungs.
Normal liver position:
*Age 0-6 months = 1-2 fingers below rib cage
*Age 6-24 months = 0-1 finger below rib cage
*Age 2+= usually not palpable (but remember, a palpable liver might still be normal btwn 2-5 yrs
1-2cm in midclavicular line)
→Do a peak flow test
→Check O2 sats & BP

Remember to look in the ears and throat if you left it out earlier!

Danger Sg's (IMCI) that indicate a high risk of death from resp illness include:
*<2 months of age
*↓ LOC
*Stridor when calm
*Severe malnutrition
*Associated symptomatic HIV/AIDS

Clinical Sg's and common abnormalities

Sg's of resp distress:
→Tachypnoea
→Tracheal Tug
→The use of accessory muscles
→Decreased saturation & cyanosis
→Gasping
→Nasal flaring
→Tripoding

Tachypnoea:
Think of = Infection
V/Q mismatch (impaired gas exchange)
-airway obstruction (upper or lower)
-pulm oedema/consolidation
Shunt
Haemoglobinopathies
↓FiO2
Anxiety
Metabolic acidosis
Toxins, eg salicylates

↓RR:
Think of = Drugs, eg opiods
↑ ICP
Hypercapnia

Tracheal deviation
To side of lung lesion
1-lung collapse
2-upper lobe fibrosis
3-pneumonectomy

Away from side of lesion
4-tension pneumo
5-massive pleural effusion

Upper mediastinal mass
6-lymphoma

Age-dependant range of HR, RR & BP

Age	HR	RR	BP
Day 1	120-140	40-50	60/40
1 Year	80-140	30-40	80/55
2-5 Years	70-115	20-30	90/60
School going child	70-115	15-20	100/65

BP and the lungs
Pulses Paradoxys
(↓in SBP>10mmHg during insp)
→Severe airway obstruction
→Cardiac tamponade/large pericardial effusion

Breathing patterns:
Cheyne-Stokes: newborn, especially prem baby.
Pathological: brainstem stroke, severe CCF
Kussmael: Acute renal failure, metabolic acidosis, Salicylate/methanol poisoning

Percussion:

Stony Dull	Dull	Resonant	Hyperresonant
-pleural effusion	-consolidation	-normal	-hyperinflation
-haemothorax	-atelectasis		-pneumothorax
	-severe pulm fibrosis		

Sounds on auscultation:

→ Insp > Exp = Vesicular
Normal
No "gap" btwn insp & exp

→ Ins = Exp = Bronchial
Normal over trachea/bronchi
Abnormal over rest of lung
"gap" btwn insp & exp

Vesicular→ normal, ↓in effusion, pneumo, obstructive lung disease
Bronchial→consolidation, pulm fibrosis, @ top of effusion
Aegophony→pt say "ee", you hear "aa"
Consolidation
Amphoric breathing→sounds like 'blowing over a bottle'
Heard over cavity
Crackles→Early inspiration = small airway disease, eg.bronchiolitis
Middle inspiration = pulm oedema, consolidation
Late inspiration = Pulm oedema, fibrosis, consolidation
Biphasic = Bronchiectasis, oedema, consolidation
Pleural friction rub→Inflamed parietal & visceral pleura move over each other. Deep breathing, end of insp, beginning of exp.
Pneumonia, PE, Pulm vasculitis
Whispering pectoriloquy→↑ in vocal resonance to the extent that when a pt whispers, his voice may be heard clearly.
Consolidation.
Stridor→upper airway obstruction.(glottis to carina)
Usually inspiratory, associated with chest wall recession.
Wheezing→small airway obstruction.
More marked on expiration.

Causes of clubbing:
CVS
-Cyanotic heart disease
-Infective endocarditis
-Atrial myxoma (rare in kids)

Resp
-Chronic suppurative lung diseases
>Bronchiectasis
>Cystic fibrosis
>Lung assess
>Empyema
-Interstitial lung disease

GIT
-Inflam Bowel diseases
>Chrons/ Ulcerative colitis
-Billiary atresia
-Cirrhosis/Liver abscess

Unilateral – AV malformations
Just toe nails – PDA
Congenital

Sg's of hyperinflation
→hyperresonant
→↓cardiac dullness
→liver displaced ↓
→hoover sg
→harrison sulcus

Impalpable Apex beat:
→hyperinflation
→large pleural/ pericardial effusion
→dextrocardia
→fat kids

Sg's of consolidation:
-bronchial breathing
-crackles/wheeze
-aegophany
-vocal resonance/fremitus
-whispering pectoriloquy

The "Coughers"

Cough- reflex, involuntary explosive expiration→primary defence mechanism of resp tract.

Barking = subglottic pathology eg.croup
Paroxysmal prolonged bouts ending in sharp intake = whooping cough
Brassy = tracheal compression
Cough soon after put to bed = upper airway pathology with postnasal drip
Cough early morning or after exercise = asthma
Productive cough = bronchiectasis

Can be acute or chronic (>21days)

Type of cough	Cause	Diagnostic Clue
Acute	Viral resp infection	Coryzal Sx's
	Bronchiolitis	Wheeze in < 1 year
	Pneumonia	Fever & dyspnoea
	Foreign body	Sudden onset
Chronic	Allegic rhinitis	Enlarged tonsils, adenoids
	Pharyngitis & sinusitis	Postnasal drip
	Asthma	Wheeze, night cough
	Bronchitis	Wheeze (more on exp)
	Aspiration	Wheeze
	TB	Weight loss
	Bronchiectasis	Crackles, wheeze, clubbing
	Obliterative bronchiolitis	Wheeze
	CF	Chronic diarhoea, clubbing,wheeze
Psychogenic		Honking cough, ceases with sleep

Neonates are obligate nose breathers until 5 months of age, although 40% of term babies will convert to oral breathing if nasal obstruction occurs.

Airway Obstruction

Divided into upper (extrathoracic) & lower (intrathoracic) obstruction.

STRIDOR	**WHEEZE**
Inspiratory	Expiratory
Recession marked	Recession less marked
Tachypnoea moderate	Tachypnoea marked
CXR normal	CXR hyperinfl. Barrel chest

The "Stridors"

Causes of Extrathoracic airway obstruction

Acute:

Infectious =
Laryngo-tracheo-bronchitis (croup)
Epiglottitis
Bacterial tracheitis
Retropharyngeal abscess

Trauma =
Intubation/thermal injury

Mechanical =
Foreign body

Allergic =
Angioneurotic oedema

Chronic:

Infectious =
Laryngeal papillomatosis
Tonsil/adenoid hypertrophy

Trauma =
Subglottic stenosis

Congenital =
Laryngomalacia
Laryngeal cysts

And the "Wheezers"

Causes of Intrathoracic airway obstruction

Large airways	**Small Airways**
Foreign body	Acute viral bronchiolitis
TB gland obstruction	Asthma
Congenital	Pneumonia
-Anomalous artery	CCF
-Bronchogenic cyst	Aspiration
-Tracheomalacia	CF/Bronchiectasis

Now as we can see, **not all that wheeze = asthma**→a common mistake made by many GP's. When I was 3, a GP treated me for asthma for 2 months. 4 months later he received an envelope from my cardiothoracic surgeon (who by the way had to perform a left pulmonectomy on me), containing my favorite 'leggo'-dragon's head. Thanx doc.

Physical Sg's in chest diseases (classical sg's seen in older children)

	Broncho-pneumonia	Lobar pneumonia	cavitation	collapse	fibrosis	hyperinflation	effusion	pneumothorax	bronchiectasis
Mediastinal Shift	None	None	None	Towards	Towards	None	Away	Away	None
Vocal fremitus	Normal	↑	Might ↑	↓	↓	↓	↓	↓	Normal
Vocal resonance	Normal	↑	Might ↑	↑	↑	↓	↓	↑↑	Normal
Percussion	Resonant	Dull	Hyperresonant- If air Dull- If fluid	Dull	Dull	Hyperresonant	Stony Dull	Hyperresonant	Resonant
Breath sounds	Vesicular	Bronchial	Amphoric	↓ or bronchial	↓ or bronchial	↓	↓↓↓	↓↓↓	Vesicular
Adventitious sounds	Crackles	Crackles Whispering pectoriloque Aegophony	Crackles	None	None	Might have wheeze	Crackles above		Biphasic crackles

LOWER RESPIRATORY TRACT INFECTION

*normal for child <3yr to develop 6-8 resp tract infections/year
*majority of these infections = URTI (see ENT chapter)
*factors influencing incidence:
→Poor nutritional status
→Poor SES
→Parental smoking
→Parasitic infection
→Structural abnormalities
→Immunization (25% of deaths frm resp infect can be prevented)
→HIV incidence

→ ***LTB**
***Bronchitis**
***Bronchioltits**
***Pneumonia**

Do not get confused:
When we talk about URTI & LRTI, the anatomical division point is the vocal cords. When we talk about upper airway and lower airway obstruction, the anatomical division point is the carina.
Thus: Croup is due to a LRTI but presents as an upper airway obstruction (stridor)

Laryngotracheo-bronchitis (LTB) a.k.a. croup

*Most commonly kids btwn 6 months-2 yrs. Stridor in kids> 3 yrs, look for another cause.
*Aet: Almost always viral→parainfluenza viruses, measles, Herpes simplex

Clin pic
2-3 days post URTI
Sx's→Stridor
→Bark-like cough
→Hoarse voice

No High Temp
Children do not appear toxic

Sg's: →Stridor (insp, later exp)
→Pulses paradoxus (worsening upper airway obstruction)
→Uses abo muscles to overcome obstruction during exp
→Hypoxic, ↑HR, restless, confusion

Approach:
1)Clin pic
2)Grading
3)Mx

Mx: According to grading system of Klein
*adrenalin nebs=1:1000 solution 1ml + 1ml saline, can repeat every 20 mins.
*Parenteral steroids (dexa 0.6mg/kg stat) or nebulized steroids for grade 2 or more, C/I in measles & herpes LTB.
*Have a low threshold for intubation→prognosis excellent

Grading system of Klein

GRADE	CRITERIA	Rx
1	Insp stridor	Observe
2	Insp + Exp stridor	Nebulize adrenaline
3	Insp + Exp stridor + Pulses Paradoxus	Continuous adrenaline nebs Intubate if no improvement
4	Impending apnoea	Intubate

Acute Bacterial Epiglottitis

*Although this is an URTI, it will be discussed here, as this must be distinguished frm LTB.
*Uncommon cause of upper airway obstruction, but ↑↑↑ mortality if not recognized.
*Kids 2-5yrs. Mostly H.Influenza B.

MEDICAL EMERGENCY

Clin pic
Sx's: Dysphagia, drooling, high temp, toxic looking (septic), stridor
Aireay patency protected by sitting in "tripod" position, muffled Voice
Sg's: "cherry red" swollen epiglottis
Later xray=swollen epiglottis ('hitch-hikers' thumb)
Be careful: do not do an ENT exam, unless an ENT or anaesthesiologist is present.

Approach:
1)Clin pic
2)S/I
-later xray
-bld culture
-throat swab
3)Mx

Mx:
Nasotracheal intubation under GA. If airway obstructs before intubation→apply positive pressure ventilation with ambu bag.
Ab's: Amoxycillin (200mg/kg/day) IV and chloramphenicol (100mg/kg/day) IV started immediately after intubation.
Oedema subsides quickly, can normally be extubated after 36-48 hrs

S/I: Don't do bld cultures or throat swab before child is intubated. He might cry→complete airway obstruction

Features	Croup	Epiglottitis
Appearance	Well	Toxic
Cough	Barking	Slight/absent
Voice	Hoarse	Muffled
Drooling	No	Yes
Able to drink	Yes	No

Acute Viral Bronchiolitis

*infection of bronchioli, peak incidence: 3-4 months of age, but can occur up to age 2
*mostly autumn & winter
*aet: mostly RSV, adenovirus
*Dx = clinical, most infants = full recovery within 2 weeks

Mx: supportive→ Rx hypoxia & maintain hydration
*nebulize with adrenaline/B2 agonist/ipratropium bromide (30% will improve), no improvement→stop
*Ab's only if: WCC>15; temp>38.5; CXR suggests infection

MILD *feeding well *RR<40/min *sats>92 (on air)	Mx @ home
MODERATE *Difficulty breathing *RR>40/min *Marked recession *sats<92	Admit to hosp O2 via nasal canula/headbox IV/NG fluids
SEVERE *recurrent apnoea *RR>60/min *Severe recession *Sats<90	Admit to ICU High [O2] Intubate & ventilate if Resp failure/recurrent

Clin pic:
Sx's: Hx of URTI, Coryzal sx's, cough, difficulty breathing, feeding problems, low-grade temp
Sg's: Tachypnoea, subcostal/intercostals recession, chest hyperinflation, bilateral fine crackles, wheeze

Infants @ risk of developing severe disease:
*Prem babies
*Babies<6 weeks
*Babies with chronic lung disease eg.CF
*Infants with congenital heart disease

Approach:
1)Clin pic
2)S/I
3)Mx
4)Cx

S/I:
>CXR (hyperinflation)
>Viral isolation – nasopharyngeal aspirate (little practical importance)
>O2 SATS

Cx:
*recurrent LRTI's
*bronchiolitis obliterans: pt remain symptomatic for months after attack

Tracheobronchitis

Def: Clinical syndrome produced by inflammation of trachea and bronchi. (rarely children<2, if < 2= most likely bronchiolitis)
Aet: 90% = viral→Adeno, rhino, influenza, parainfluenza. Secondary bacterial→S pneumonia, M catarrhalis, H influenza, Chlamydia, mycoplasma.

Clin Pic:
Bronchitis begins as a resp tract infection that manifest as the common cold: coryza, slight fever, sore throat, back & muscle pain. Cough = dry initially, productive later, harsh or raspy sounding.
Sg's: No sg's of consolidation. Wheeze might be present.

Approach:
1)Clin pic
2) S/I
3)Mx

Mx
→Maintain hydration
→Maintain oxygenation
→Antipyretic & analgesics
→Avoid 2nd hand smoke inhalation
→Admit pt if danger sg's are present, poor saturation or resp distress-------O2 & ? bacterial infect.
→Refer to pulmonologist if Sx's persist > 2 weeks.

S/I
CXR→Normal
Blds: CRP & WCC (only if hospitalized)
Sputum culture (If bacterial)

Infections causing mainly Lower resp disease

Bordetella pertussis & parapertussis
Strep pneumonia, H influenza, Staph aureus, TB, PCP
Atypical organisms: Chlamydia pneumonia, Mycoplasma pneumonia
RSV, influenza, adenovirus
Aspergillus, Histoplasma

See Infectious Disease chapter

Pneumonia

*Def: Inflammation of lung parenchyma with consolidation of alveoli.
*Incidence: Greatest in 1st yr of life
*NB→if cough/wheeze persist>14 days after Dx was made & Rx given→exclude TB, foreign body, Cx of pneumonia, or wrong drug.

Approach:
1)Clin pic
2)Classification
3)S/I
4)Mx
5)Cx

Sg's of resp failure =
Features of hypoxia: severe tachypnoea, chest indrawing, restlessness, grunting, tachycardia, central cyanosis
Features of hypercarbia: bounding pulses and peripheral vasodilatation

Clin Pic
Neonates: lethargy, poor feeding, ↑temp, apnoea, tachypnoea
Older children: runny nose + sore throat, cough, fever, tachypnoea
More serious pneumonia: tachypnoea, chest indrawing, poor feeding + sg's of resp failure

Classical sg's of consolidation is only seen in older children:
-bronchial breathing
-crackles/wheeze
-aegophany
-vocal resonance/fremitus
-whispering pectoriloquy

Classification

Acute Resp tract infections: WHO classification & Mx		
Severity	**Criteria**	**Mx**
1) No Pneumonia	Cough No tachypnoea	Supportive measures Antipyretic No Antibiotics
2) Pneumonia	Cough Tachypnoea No rib/sternal retraction	Supportive measures Antipyretic Antibiotics
3) Severe Pneumonia	Cough Tachypnoea Rib/Sternal retraction	Supportive measures Antibiotics Refer to Hosp
4) Very severe pneumonia	Cough Tachypnoea Chest wall retraction Unable to drink Cyanosis	Supportive measures O2 Antibiotic Refer ASAP to Hosp

Mx
Mainstay = Ab's & O2
Ab's:
Amoxycillin (200mg/kg/day in 4 devided doses) x 10d or **Co-trimoxazole**. Add cloxacillin if staph suspected. For **severe pneumonia** = parenteral penicillin for 2 days, then amoxicillin. For **very severe pneumonia** = penicillin + aminoglycoside/chloramph-enicol. Atypical organisms = erythromycin. Stop Ab's when child is fever free for 3days. Most pt's respond within 5-7days. Lung abscess/empyema = Rx for 21-42 days.
O2: Severe pneumonia/very severe pneumonia→ nasal prongs (0.5-2L/min). Do ABG if child doesn't respond clinically. MONITOR SATS
Bld transfusion: Maintain Hct>30 in very severe pneumonia.
Hydration: PO/NG. IV if distressed (50-80ml/kg/day)
Fever: t>38 °C = paracetamol (30mg/kg/d in 4 divided doses)
Lower Airway obstruction: B2 agonist/anticholinergic.
Nutritional support: Pt need extra 60kcal/kg/d.
Other supportive measures: Vit A in measles pneumonia & HIV. Nasal passage kept clear with saline nasal drops.

Rx with no proven benefit in uncomplicated pneumonia:
*mucolytics
*chest physio
*postural drainage
*nebulized rx

S/I
CXR: Widespread, poorly demarcated, alveolar opacities with air bronchograms. Classical lobar/segmental opacification with air bronchograms less common.
Viral pneumonia→perihilar streaking, interstitial changes, air trapping.
Mucous plugging in viral pneumonia→results in lobar collapse (easily confused with bacterial pneumonia)
Staph pneumonia→areas of breakdown, lung abscesses, empyema, pyopneumothorax
Klebsiella, anaerobes, H.influenza,TB→cavitating/expansile pneumonia
Anaerobes, S.aureus,H.influenza,TB→pleural effusion/empyema
Blds:
↑WCC (leucocytosis)/↑CRP = bacterial
Bld cultures = in only 25% of cases organism is cultured
Throat swab/nasopharyngeal aspirate = not specific (contamination by URT organisms)
Sputum = TB/PCP
Tuberculin skin test

Clues to bacterial infection =
Polymorphonucleur leucocytosis, lobar consolidation & pleural effusion

Common Causes of Pneumonia

Neonates	Group B streptococ E.Coli CMV, Herpes virus
1-3 months	H.influenza S.aureus S.pneumonia CMV, Influenza, parainfluenza
3 months -5 yrs	H.influenza S.aureus S.pneumonia RSV, Adenovirus, Influenza, TB
> 5 yrs	S.pneumonia H.influenza Influenza virus, TB
Immune-compromised	Gram-negative S.aureus Opportunistic organisms PCP TB
Hospital Acquired Pneumonia	Gram-negative S.aureus Methicillin-resistant

Cx:
*para-pneumonic effusion-common
*empyema
*lobar collapse
*pneumothorax
*lung abscess
*hepatitis, pericarditis, myocarditis, meningoencephalitis

Failure to respond
*Wrong drug/Wrong bug (esp.TB/opportunistic in HIV)
*Empyema/Abscess/Effusion
*Foreign body/bronchiectasis

Prognosis = Good, except adenovirus/measles pneumonia = persistent airway obstruction/↓ lung Fx

Other causes of Pneumonia

Pulmonary TB (see TB chapter)
Hydrocarbon Inhalation
Atypical Pneumonia
→Mycoplasma
→Chlamydia
Eosinophilic Pneumonia (Loeffler)
Aspiration
→Prem babies
→CNS defects
→Gastro-oesophageal defects
→Anatomical defects
→Tracheo-oesophageal fistula
→Cleft palate
Infectious diseases
→Pertussis
→Measles with 2° bacterial infection
→Legionnaires disease
→HIV:
Infectious = Bacterial
PCP
TB
CMV
Non-Infectious = Lymphocytic Interstitial Pneumonitis
Lymphoma
Kaposi's sarcoma

Hydrocarbon Inhalation
*paraffin,petrol, & other volatile hydrocarbons
*20-40% = develop chemical pneumonitis
*resp distress within 30 minutes, worsens up to 48hr later
*2° bacterial infect common
*CNS sg's if large volume ingested
*Mx: O2 & monitor. Prophylactic Ab's doesn't improve outcome
*induced vomiting contraindicated
*Ab's only if 2° infect suspected

Pulmonary eosinophilia (PIE)
*Pulm infiltrates + eosinophilia on bld smear
*Simple PIE (loeffler) = Sx's<1 month. Hx of allergy
*Prolonged PIE = Sx's 2-6 months.
*Tropical PIE = eosinophilic lung granulomas contain degenerating microfilaria. Cause likely nematode (eg.ascaris), but also amoebiasis, trichinosis
*rare causes: PAN, Hodgkin's, asthma, drugs eg.penicillin.
*cough/wheeze/crackles
*Mx = Rx Sx's, broad spectrum vermicide (eg.albendazole)

Bacterial
*similar organisms to non-HIV Pt. Clinical pic more aggressive, CXR more atypical.
*higher rate of Cx
*Ab's needed for longer period

CMV
*Clin pic similar to PCP
*Clinically impossible to distinguish btwn CMV & PCP
*Dx = positive IgM, CMV DNA PCR, urine culture, lung biopsy
*Rx = Ganciclovir

PCP
*young, well-nourished children
*insidious onset progressive tachypnoea, fever, non-productive cough
*few clinical findings
*Resp distress + hypoxia = out of proportion to clinical findings
*CXR = bilateral perihilar interstitial infiltrates
*↑LDH
*Dx = ELISA/PCR on sputum/NGA/gastric aspirate
Rx = Co-trimoxazole (Bactrim)

Kaposi's
*trachea, bronchi, lung parenchyma, pleura, skin, lymph nodes
*suspect if oral haemorrhagic lesions present
*Dx = biopsy
*Rx = chemotherapy
*outcome = poor

Lymphocytic interstitial pneumonitis (LIP)
*common btwn 2-5 yrs
*associated with EBV
*slow progressive dyspnoea/cough, sg's of chronic lung disease, clubbing, chest deformity
*lymphadenopathy, parotid enlargement, hepatomeg
*CXR = reticulonodular/diffuse shadowing (similar to military TB)
*Dx = biopsy
*Rx = prednisolone

Lymphomas (B & T cell)
*Outcome poor
*survival<1yr without ARV

Recurrent & Persisting Pneuonia

Recurrent pneumonia→>2 episodes within 18 months
Persisting pneumonia→pneumonia>30 days

APPROACH: (Coovadia)
Hx, exam, CXR
↓
Cause normally apparent
↓
If not: exclude TB, HIV disease, asthma, foreign body
↓
Still no identifiable cause: refer to pulmonologist

Rule of thumb
No fever→No antibiotics
(except in immunocompromised pt's)

Causes of recurrent/persisting pneumonia
Diffuse (pneumonia not limited to 1 lobe)
Allergic
→undiagnosed asthma
→pulm infiltrates with eosinophilia
Inflammation
→airway damage by viral/bacterial infect
Recurrent aspiration
→sucking/swallowing abnormality
→tracheo-oesophageal fistula
→gastro-oesophageal reflux
Muco-ciliary dysfunction
→CF
→Immontile cilia syndrome
Immunodeficiencies
Acquired
→HIV & malnutrition
Congenital
→T-lymphocyte deficiencies
→Agamma-globulinaemia
→Complement deficiencies
→Neutrophil abnormalities
Passive smoking

Localized (pneumonia occurs in same lobe)
Foreign body
Extrinsic compression eg.lymph nodes
Bronchiectasis
Congenital lung lesions

Extrathoracic Airway Obstruction

Laryngeal papillomatosis
*HPV (pt born via NVD→mother had HPV warts)
*Airway obstruction can occur if viral resp infect is superimposed
*Mx = Removal by laser
*Can reoccur after removal

The "Stridors"

Causes of Extrathoracic airway obstruction

Acute:
Infectious =
Laryngo-tracheo-bronchitis (croup)
Epiglottitis
Bacterial tracheitis
Retropharyngeal abscess

Trauma =
Intubation/thermal injury

Mechanical =
Foreign body

Allergic =
Angioneurotic oedema

Chronic:
Infectious =
Laryngeal papillomatosis
Tonsil/adenoid hypertrophy

Trauma =
Subglottic stenosis

Congenital =
Laryngomalacia
Laryngeal cysts

Bacterial tracheitis
*Infect of trachea by S.aureus, H.influenza, C.diphtheria
*High grade fever, coughs up large amounts of tenacious, yellow sputum
*Mx = amoxicillin & cloxacillin IV, intubate if resp failure
*Suspect diphtheria if pt not immunized

Retropharyngeal abscess
*Abscess formation in lymph nodes in prevertebral space→can lead to airway obstruction
*Child is toxic, has dysphagia, hyperextension of neck, noisy breathing, enlarged submandibular lymph nodes, on exam of throat = large retropharyngeal mass noted.
*Mx = broad-spectrum ab's & surgical drainage under GA

Intubation injury (subglottic oedema)
*Cricoid→narrowest part in child's airway
*Cricoid injured during intubation (too large tube)→Oedema→stridor
*Always ensure that a small air leak with ventilation is present→if not, re-intubate with smaller tube

Foreign body
*Sudden stridor
*Life threatening airway obstruction→Heimlich/place child over knee & give few hard thumbs on back→not successful, intubate ASAP and force FB into one of bronchi→remove FB via bronchoscopy
*Oesophageal FB→displace tracheo-oesophageal membrane anteriorly into lumen of trachea→remove via gastroscopy

Subglottic stenosis
*Pt's intubated with too large tube/prolonged ventilation→necrosis of mucosa in subglottic region→heals with fibrosis→subglottic stenosis
*Dx = bronchoscopy
*Mx = Complex surgical correction

Laryngomalacia
*Prominent aryepiglottal tissue→sucked into laryngeal opening during inspiration
*Most common cause of extrathoracic airway obstruction in 1st month of life
*Stridor less prominent if nursed prone
*Stridor more prominent during fever & activity
*Dx = laryngoscopy
*No surgical intervention required
*Spontaneous recovery within 1 yr

Intrathoracic Airway Obstruction

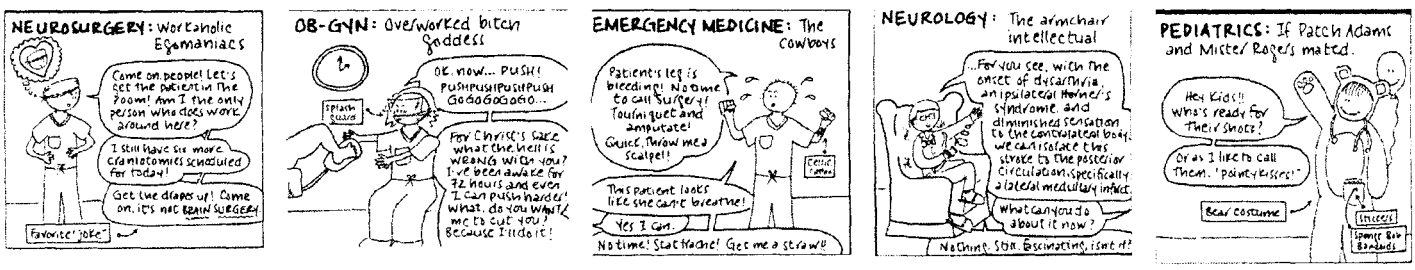
And the "Wheezers"

Causes of Intrathoracic airway obstruction

Large airways	Small Airways
Foreign body	Acute viral bronchiolitis
TB gland obstruction	Asthma
Congenital	Pneumonia
-Anomalous artery	CCF
-Bronchogenic cyst	Aspiration
-Tracheomalacia	CF/Bronchiectasis

Foreign Body Inhalation
*typical Hx = healthy child who choked while playing→started coughing→turned blue→short of breath for a period→wheeze
*classical triad = unilateral wheeze
decreased unilateral air entry (most common sg)
lung collapse
*normal clin exam = does not exclude dx
*always think of FB in the following clin pic's;
→stridor not responding to Rx
→asthma not responding to bronchodilators
→pneumonia not responding to Rx
→recurrent pneumonia in same lobe
→resp failure of sudden onset
→unexplained lobar collapse
→localized bronchiectasis
*Radiology: only 10-30% of FB's are radio-opaque
Indirect evidence of FB: Lobar collapse
Lobar hyperinflation
Decreased vasculature as a result of hypoxic vasoconstriction
*rigid bronchoscopy is essential for Dx & Rx

CCF (see cardiology chapter)
*Children <2 with LHF, might present with experatory wheeze which results frm bronchial mucosal oedema causing increased airway resistance.
*Narrowing of the left main bronchus by an enlarged left atrium may contribute to the expiratory wheeze.
*Alveolar opacification as a result of lung oedema is difficult to differentiate frm pneumonia.



Asthma

Def: Chronic inflammation
Of the bronchial mucosa
Resulting in bronchial hyper-reactivity
Associated with mucosal oedema, mucosal secretions & bronchoconstriction (when triggered)
Leading to airway narrowing & clin pic
*common btwn 2-5 yrs

Risk factors for asthma:
>Fam Hx
>Co-existent atopy
>Male sex
>Parenteral smoking
>Bronchiolitis in infancy

Trigger factors:
Smoking
Temp changes
URTI's
Allergens
Exercise
Emotion

There is a colour code for asthma drug inhalers:
'Preventers',eg.inhaled steroid is mostly BROWN (sometimes purple, green, orange)
'Relievers',eg.B2 agonists are BLUE

Clin pic
*cough, dyspnoea, wheeze (sx's severity might ↑ at night)
*sg's of hyperinflation, pectus carinatum, Harrison sulcus, wheeze

Dx
*>15% increase in FEV1 after B2 agonist administered
*therapeutic trail with B2 agonist (asthma diary kept at home)

Approach
1)Clin pic
2)Diff Dx
3)Dx
4)Classification
5)Mx
→**acute**
→**maintenance**
6)Education
*Emotional support, avoid allergens, exercise program, monitor resp Fx with peak flow meter, NB = compliance

Diff Dx

Foreign bodies in the airways
(unilat wheezing might be noted)

Supraglottic
→Retropharyngeal abscess
→Tonsillar abscess
→Epiglottitis

Laryngeal causes
→Croup
→Stenosis
→Tetany
→Vocal cord paralysis
→Angioedema of larynx

Tracheal causes
→Tracheomalacia
→Tracheitis
→Lymp node compression

Bronchial causes
→Bronchiolitis
→Bronchitis
→Bronchiectasis
→Lymph node compression

Pulmonary causes
→Pneumonia
→CF
→TB
→Pertussis
→Atelectasis
→Loeffler's syndrome

Other
→CCF/Gastro-oesoph. reflux
→Hyperventilation

Acute Asthma Attack → O2 4L/min + B2 agonist nebs for 20mins x 2 or subcutaneous adrenalin
ASSESS RESPONSE

Criteria	Responder	Non-responder
PEFR	>80%	<80%
RR	<40	>40; HR>140
Retraction	absent	present
Speech	normal	impaired
Feeding	normal	impaired

Responder → Review current Rx, exclude possible precipitants, follow-up

Non-responder → Intensify Rx:
*O2 (high flow)
*B2 agonist
*Ipratropium bromide
*Steroids (oral/IV)
*Maintain hydration

Assess Response

Features of life threatening asthma:
↓LOC
Silent chest
Poor resp effort
Sats<85%;cyanosis
Exhaustion

Mx = Intubate, continuous B2 agonist nebs, IV salbutamol/aminophyline

→ Admit to ICU

Non-responder = Acute severe asthma
Monitor ABG, ECG, PEFR(6hrly)

No features of life threatening asthma:
Continuous B2 & ipratropium bromide nebs.

No improvement → Improvement → Admit to ward

Classify severity at presentation

	Intermittent	Persistent		
		Mild	Moderate	Severe
Category	1	2	3	4
Daytime sx's	< 2/week	2-4/week	>4/week	Continuous
Night-time sx's	<1/month	2-4/week	>4/week	Frequent
PEF	>80%	>80%	60-80%	<60%

Maintenance

All Categories: Short-acting B2 agonist as needed
Environmental control
Education/self Mx

Step 1 (intermittent): Short-acting B2 agonist as needed

Step 2 (mild): low dose inhaled corticosteroid

Step 3 (moderate): medium dose inhaled corticosteroid
LABA/sustained release theophylline
Consider adding Leukotriene receptor antagonist

Step 4 (severe): high dose inhaled corticosteroid
LABA/sustained release theophylline
Leukotriene receptor antagonist

Step 5 (severe & still uncontrolled): As step 4 + alternate day oral steroids

A rescue course of oral prednisolone might be needed at any step:1-2mg/kg/d for 7-14 days to achieve control.
Step down once control is achieved
Control = <2 daytime sx's/week, no night-time sx's, no limitation of activity, reliever ≤2/week, normal lung Fx, no exacerbations

Medications used:

Controllers (preventers)		Relievers
Anti-inflam action	**Sustained bronchodilation**	**Quick relief (bronchodilation)**
Inhaled corticosteroids Beclomethasone Budesonide	**Laong-acting B2 agonist** Salmeterol Formoterol	**Short-acting B2 agon.** Salbutamol Fenoterol Terbutaline
Leukotriene antagonists Montelukast	**Phosphodiesterase-inhibitors** Theophyline Aminophyline	**Anti-cholinergics** Ipratropium bromide
Oral corticosteroids Prednisone Prednisolone Methylprednisone		

Asthma continue...

Administration of medication by inhalation

	Advantages	Disadvantages	Age group
Metered dose inhaler	Small & portable	Coordination important	Suitable only for competent older children
Metered dose inhaler + spacer	Coordination unimportant Any age-group	Bulky	Any age
Dry powder inhaler	Coordination unimportant Small & easy to use	Requires rapid inspiration	Children>5yrs
Nebulizer	Usable at all ages Effective in severe attack	Expensive, noisy, Rx takes>5mins, frightens some infants	Any age

Mode of action, indications & side effects of bronchodilators

Reliever	Mode of action	Use	Side effects
Short-acting B2 agonist: Salbutamol Fenoterol Terbutaline	Smooth muscle relaxation	Relief of bronchospasm	Tachycardia Hypokalaemia Restlessness
Long-acting B2 agonist: Salmeterol Formoterol	Smooth muscle relaxation	Step 2 Nocturnal asthma, exercise induced, alternative to high-dose steroid	
Phosphodiesterase inhibitor: Theophylline (oral) Aminophylline (IV)	↑cAMP	Step 3/4 Oral for nocturnal asthma, IV in acute severe asthma	Restlessness Diuresis Cardiac arrhythmias
Anticholinergics: Ipratropium bromide	Inhibit cholinergic bronchoconstriction	Add on to B2 agonist in acute attack, 1st line bronchodilator in infants	Dry mouth Urinary retention

Medications for Pediatric Asthma

MEDICATION	DOSAGE
Short-course Systemic Steroids	
Prednisolone (5 mg/5 mL or 15 mg/5 mL)	1 mg/kg/day to 2 mg/kg/day orally; maximum 60 mg/day
Rescue Medications	
Albuterol ampules* (.063 mg/3mL; 1.25 mg/3mL; 2.5 mg/3mL)	0.63 mg/3ml to 2.5 mg/3mL saline every 4 to 6 hours as needed (may be dosed 2.5 mg every 20 minutes x3 doses OR 0.15 mg/kg to 0.3 mg/kg up to 10 mg every 1 to 4 hours as needed OR up to 0.5 mg/kg/hr continuous nebulization for acute exacerbations)
Levalbuterol (R-albuterol)* (.63 mg/3 mL; 1.25 mg/3mL) (.31 mg/3 mL)	0.63 mg/3ml to 1.25 mg/3mL saline every 4 to 6 hours as needed (may be dosed 1.25 mg every 20 minutes for 3 doses then 0.075 mg/kg to 0.15 mg/kg up to 5 mg every 1 to 4 hours as needed)
Ipratropium (0.25 mg/mL saline*) (severe exacerbation only, is not to be used as first-line therapy)	0.25 mg to 0.5 mg every 20 minutes x 3 then as needed (may mix with albuterol in nebulizer)
Ipratropium with albuterol nebulizer solution: (0.5 mg ipratropium bromide and 2.5 mg albuterol)	1.5 mL every 20 minutes x 3 doses then as needed for up to 3 hours
Maintenance Medications	
Cromolyn sodium	1 ampule 3 to 4 times per day
Montelukast	4 mg orally daily (age 2-5); 5 mg orally daily (age 6-14)
Budesonide ampules*	0.25 mg, 0.5 mg, 1 ampule 1-2x per day
Inhaled corticosteroids via metered-dose inhaler with or without the use of a spacer	Beclomethasone, budesonide, budesonide/formoterol, ciclesonide, fluticasone, fluticasone/salmeterol, mometasone (dose varies per medication)

Estimated Comparative Daily Dosages for Inhaled Corticosteroids in Adults and Children

Drug Name Generic	Brand	Low Daily Dose (mcg) Adult	Low Daily Dose (mcg) Child*	Medium Daily Dose (mcg) Adult	Medium Daily Dose (mcg) Child*	High Daily Dose (mcg) Adult	High Daily Dose (mcg) Child*
Beclomethasone HFA 40 or 80 mcg/puff	QVAR	80-240	80-160	240-480	160-320	>480	>320
Budesonide DPI 200 mcg/inhalation	Pulmicort Turbuhaler	200-600	200-400	600-1200	400-800	>1200	>800
Budesonide inhalation suspension for nebulization (child dose)	Pulmicort Respules	-	0.5 mg	-	1.0 mg	-	2.0 mg
Flunisolide 250 mcg/puff	AeroBid, AeroBid-M	500-1000	500-750	1000-2000	1000-1250	>2000	>1250
Fluticasone MDI 44, 110, or 220 mcg/puff	Flovent	88-264	88-176	264-660	176-440	>660	>440
Fluticasone DPI 50, 100, or 250 mcg/ inhalation	Flovent Rotadisk	100-300	100-200	300-600	200-400	>600	>400
Triamcinolone acetonide 100 mcg/puff	Azmacort	400-1000	400-800	1000-2000	800-1200	>2000	>1200

*Children 12 years of age

HFA = hydrofluoroalkane; DPI = dry-powder inhaler; MDI = metered-dose inhaler

Source: Executive Summary of the NAEPP Expert Panel Report. Guidelines for the Diagnosis and Management of Asthma—Update on Selected Topics 2002. NIH publication 02-5075; 2002.

Classical presentation:
Recurrent chest infections
Large offensive stools
Failure to thrive

sweat test !!

Cystic Fibrosis

*Most common fatal autosomal recessive disease in young Caucasians, with an incidence of 1 in 2500 live births (1 in 25 people are carriers)

*There is a mutation in a gene that codes for a chloride channel that's responsible for chloride(& water) movement across epithelial membranes. The gene's name = CFTR (cystic fibrosis transmembrane conductance regulator) situated on the long arm of chromosome 7. There are many identified mutations→most common = ΔF508, accounts for 80% of cases. Chloride channel normally allows chloride movement out of the cell into the lumen (airway & pancreas). Mutation→↓chloride level→↑re-absorption of sodium frm fluid in lumen→↓excretion of water→relatively dehydrated epithelium→viscous mucous plugs→predispose to recurrent bacterial infection→bronchiectasis (result of recurrent P.auruginosa & S.aureus infections)

*Note – in sweat glands, CFTR plays different role in ion regulation→allowing reabsorption of chloride ions frm the sweat = hence the sweat test for CF.

Clin Pic:

Newborn: →Malabsorption
→Recurrent/persistent chest infect
→Failure to thrive
→Meconium ileus (10-20%)
→Extended period of neonatal jaundice

Age 0-2: →All of above + steatorrhoea

Age 2-8: →Bronchiectasis
→Rectal prolapse
→Nasal Polyps
→Sinusitis

Age 8+: →Cor pulmonale
→Diabete Mellitus
→Cirrhosis & portal HT
→Distal intestinal obstruction
→Pneumothorax
→Haemoptysis
→Infertility in ♂ (no vas deference)
→Clubbing
→Osteoporosis

NB Sg's =
*Hyperinflation
*Insp crackles
*Exp wheeze
*Hepatomeg
*Clubbing
*Nasal polyps
*Cyanosis
*Sg's of resp infect

Diagnostic tests:

Sweat test→this uses electrodes to provoke sweating. Two electrodes are placed on the skin, and a small current passed through them. Filter paper is used to collect a sample of sweat. The concentration of both sodium & chloride ions is measured:
*Child>60mmol/l Cl→abnormal
*Adult>90mmol/L Cl→abnormal

Elastase in faeces→low levels = diagnostic for pancreatic insufficiency

Screening in newborns = Guthrie test→Immunoreactive Trypsin (IRT) raised in CF babies.

Other tests (not for Dx, but indicate abnormality) =
*vitamins A.D.E.K→↓
*CXR→hyperinflation,bronchiectasis
*Abdo US→fatty liver, cirrhosis, chronic pancreatitis
*spirometry→obstructive pattern

Approach:
1)Clin Pic
2)Dx
3)Mx

Prognosis
*median survival in UK in 1990 = 40yrs
*95% die frm resp complications
*resp failure = main cause of death (usually during acute exacerbation of resistant infection)

Mx→ multi-disciplinary team (aim: minimize progression, maintain adequate nutrition)
1)Nutrition:
→oral pancreatic enzymes
→PPI's
→restrict fat intake
→NG/gastrostomy tube if needed
→high protein/calorie diet
2)Antibiotics
→flucloxacillin (prophylaxis)
→Nebulize pt btwn exacerbations (saline & tobramicin)
→pseudomonas infection difficult to treat---culture & determine Ab sensitivity

Rx aggressively with IV antibiotics

3)Physiotherapy & postural drainage
4)Mucolytics (nebulized DNase)→↓sputum viscosity→help with mucociliary clearance
5)Exercise
6)Ursodeoxycholic acid (UDCA)→prevent liver impairment by ↓cholestasis & ↑bile acid reabsorption
7)Lung transplant & Genetic counselling

The Future

Future treatments could involve gene transfer therapy, whereby the affected CTFR gene could be replaced by a fully functioning version through adenovirus therapy. CF is a particularly good candidate for such therapy, as the bronchial mucosa is easier accessible via inhalation.

Suppurative Lung Disease

2 Main causes of chronic suppurative lung disease = *bronchiectasis* & *lung abscess*

Bronchiectasis
→Permanent destruction of bronchial walls & lung tissue due to chronic infection
→3 mechanisms: Bronchial lumen obstruction (eg.foreign body/TB lymph nodes)
Parenchymal destruction (frm necrotizing pneumonia→staph, klebsiella, anaerobes, TB)
Repeated resp tract infections
Dyskinesia →rare cause of bronchiectasis(eg.kartagener's syndrome)
Clin pic: Repeated LRTI's, Productive cough, Halitosis, Clubbing, Growth restricted, Crackles (biphasisc), Wheeze, Pulm HT, Cor pulmonale,
S/I: CXR(honey-comb appearance & fibrosis), CT. [NB = clinical dx!]
Mx = prevent by immunization of children, Rx pneumonia & FB & TB correctly
Physiotherapy & postural drainage
Surgery = if disease is unilateral, no Pulm HT, lung Fx not so compromised that the child will be a respiratory invalid after the surgery

Lung Abscess
*mostly follow S.aureus, H.influenza, Klebsiella pneumonia, TB, anaerobes & sometimes S.pneumonia.

Clin pic: Child is toxic, high fever, foul-smelling sputum, poor response to Ab's, amphoric breathing over cavity

S/I: CXR (cavity with fluid level)
Must be differentiated from a loculated pyopneumothorax, diaphragmatic hernia, echinococcus cyst.

Mx = IV Ab's (Pen, Clox, & aminoglycoside)
If abscess does not drain→exclude obstruction of bronchus & transthoracic drainage of abscess

(RX) 21-42d

Pleural Effusion
Pneumothorax

Diff for opaque CXR:
>Pleural effusion
>Total lung collapse
>Pneumonectoy
>Diaphragmatic paralysis
>Diaphragmatic hernia

Diseases of the Pleural Cavity

Pleural Effusion

*Fluid accumulation in pleural space
*Mechanism: ↑hydrostatic pressure, ↓ COP → Transudate
↑microvascular permeability, Lymphatic obstruction → Exudate

stony dull

Work-up
CXR→Confirm Effusion
Diagnostic Tap→Done over maximal dullness
Note appearance of fluid
Send away for: total protein, LDH, cytology (+diff), MSC (AFB+ADA+TB PCR), glucose & Ph.
CT chest with pleural contrast→benign vs. malignant disease
Pleural biopsy for TB culture + histology

Clin Pic
Sx's: Dyspnoea, Sx's of LRTI
Sg's:↓/absent breath sounds
Crackles above effusion
Stony dull on percussion
Tachypnoea, ?Resp distress, fever, lymphadenopathy (?TB)

Approach:
1)Clin Pic
2)Work-up
3)Mx
4)Cx

Lights Criteria→For an Exudate
Fluid Protein:Serum protein > 0.5
Fluid LDH:Serum LDH > 0.6
Fluid LDH > ⅔ of upper limit of normal serum LDH

Cx:
Pleural thickening
Calcification
Haemorrhagic change
Atelectasis
Empyema

Mx: Rx the cause
Maintain hydration
Optimize nutrition

TB effusion→Rx as for PTB + steroids might ↓ fluid volume, but don't affect long term change

Causes:

Transudate	Exudate
Common	**Common**
Fluid overload	Para-pneumonic
LHF	TB
Hepatic cirrhosis	Rheumatoid disease
Liver failure (↓protein)	Malignancy
Nephrotic syndrome	
Nephritic syndrome	
Renal failure	
Uncommon	**Uncommon**
Constrictive pericarditis	Empyema
PE	Subphrenic abscess
	Post Cardiac surgery
	Drug induced (amiodarone, bromocriptine, methotrexate, phenytoin, nitrofurantoin)
	PE

Clinical features of empyema
Systemic: pyrexia, rigors, malaise, polymorphonucleur leucocytosis, ↑CRP
Local: pleural pain, dyspnoea, cough, copious purulent sputum if empyema ruptures into a bronchus (bronchopleural fistula) + clinical Sg's of fluid in pleural space

Large pleural effusion:
TB & Malignancy
Bilateral effusion:
Think transudate & Rheumatoid

Pneumothorax

*Air accumulation in pleural space
*Spontaneous pneumothoraces occur during **staph pneumonia**, an acute **asthma** attack, **pneumatocele rupture**, or following **hydrocarbon inhalation pneumonia**. Traumatic causes including chest wall **trauma** & **mechanical ventilation**.

Clin Pic
Sudden onset dyspnoea, unilateral pleuritic chest pain, hyper-resonant on percussion and ↓air entry on auscultation, tachypnoea
Tension pneumo = Tachypnoea, tachycardia, hypotension, cyanosis, tracheal displacement away frm the silent hemithorax.

Approach:
1)Clin Pic
2)S/I
3)Mx

S/I: CXR

Mx
ABC
IC drain insertion, needle thoracostomy in tension pneumo followed by IC drain.

RENAL DISORDERS

ALEX

UTI'S
HAEMORRHAGIC CYSTITIS
GLOMERULONEPHRITIS
NEPHROTIC SYNDROME
HUS
HYPERTENSION IN CHILDREN
INHERITED & CONGENITAL RENAL DISORDERS
ACUTE RENAL FAILURE
CHRONIC RENAL FAILURE

RENAL

- UTI'S
- HAEMORRHAGIC CYSTITIS
- GLOMERULONEPHRITIS
- NEPHROTIC SYNDROME
- HUS
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- INHERITED & CONGENITAL RENAL DISORDERS
- ACUTE RENAL FAILURE
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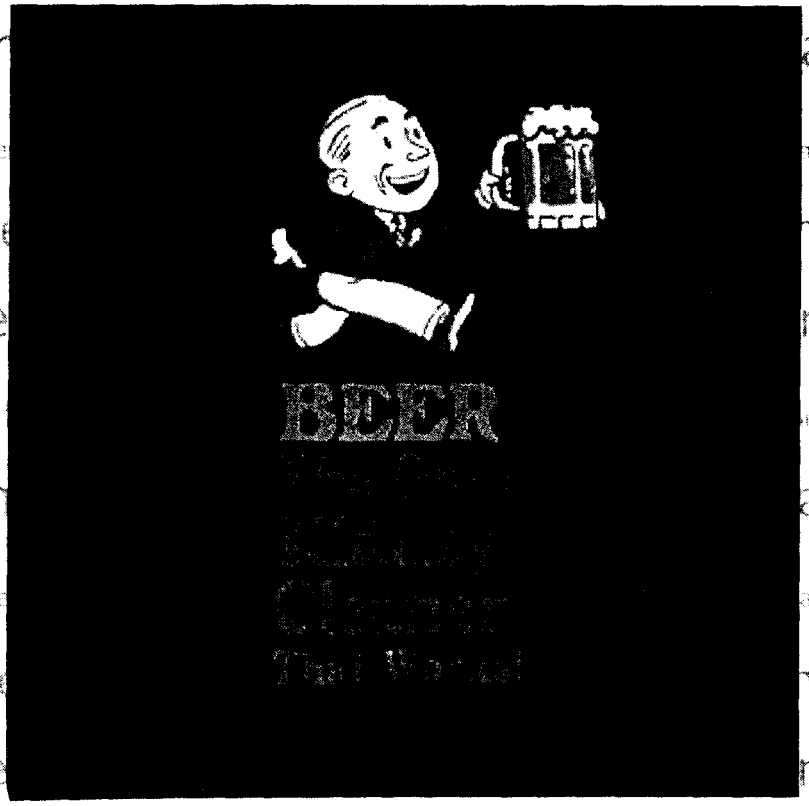


Diagnosis:

History: Male (<6months); Female (<12 months); uncircumcised male; fever (>38°C or >2days); prior UTI; malnourished child.

- Examination: Ill appearance of child
  - Infancy: lethargy; failure to feed; unstable temperature; apnoea; poor colour; jaundice; anaemia; purpura; & H/S-megaly.
  - Children: usually urgency as an isolated complaint.
  - Consider Pyelonephritis: **FFFAD**: Frequency; Fever; Flank pain; Abdominal Pain; Dysuria.

Special Investigations:

- Urine Dipstick: Protein: 1+/2+
  - Leucocytes: >trace
  - Nitrites: >trace
- Microscopy: White Cell Casts >10/HPF
- Culture:
  - >10^5 organisms/ml if clean catch MSU sample.
  - >10^3 organisms on catheter sample.
  - Any growth on supra-pubic bladder aspiration.
- Sensitivity: in order to choose the appropriate drug that the organism is sensitive to.
- Further Investigation depends on Age:

Approach to UTI's in Children:

1) **Diagnosis**
2) **Special Investigations & cause**
3) **Management**
4) **Complications**
5) **Counselling of mother**

Child <2 years:

- U/S: Kidneys, Ureters & Bladder.
- Voiding Cysto-urethrogram (VCUG)

Child >2 years:

- U/S: Kidneys, Ureters & Bladder.
- VCUG only if: ultrasound is suspect; temperature >38°C; flank pain; vomiting.

Management:

- Admit if:
  - Infant
  - Severe systemic illness + dehydrated child.
  - Immuno-compromised child.
- Initial Therapy: prior to Sensitivity results coming out includes:
  - Antibiotics: gram+ & gram- cover: given intravenously if very ill then changed to oral treatment when apyrexial, or started orally and treated until sensitivity results out.

Complications:

- Recurrent Infections: once treated initially, prophylactic therapy given for 6months at the following drugs but in lower doses: Macrodantin, Nalidixic acid, Cefaclor or Co-trimaoxazole.
- Complex UTI: reflux of urine: where bowel bacteria ascend the urethra, bladder, & then flow of urine interrupted, urine reflux occurs, with ascent of organisms up the ureters and into the calyces &parenchyma of the kidney. To exclude structural/functional derangements of the kidney: U/S, IVP, VCUG in first few months after UTI. Technetium scan if any doubt (good to visualise renal scarring)

Causative Organisms:

- **E.coli** predominantly
- **Klebsiella**
- Proteus
- Enterococci
- Staphylococci
- Pseudomonas
- Acinetobacter
- Serratia

PROCEDURE: HOW TO DO A SUPRAPUBIC BLADDER ASPIRATION:

Indicated if it is difficult to obtain an uncontaminated urine sample. In children the bladder is more of an abdominal organ. Use U/S guidance, but if not available, try palpate the bladder, insert 22guage needle (with 20ml syringe connected) 1-2cm above Symphysis pubis with (out) local anaesthesia. Insert needle in direction of head at 10-20° off the perpendicular & pull plunger to aspirate urine while advancing. If failed first attempt, repeat procedure but not more than three times. Remove needle once sample obtained & apply gentle pressure with swab at insertion site.

Approach to Schistsomiasis in Children:

1) **Diagnosis**
2) **Special Investigations & cause**
3) **Management**
4) **Complications**
5) **Counselling of mother**

Diagnosis:

- History: blood in the urine, painful micturition, may complain of Swimmer's Itch (transient urticaria); recent history of exposure in highly endemic area.
- Examination: Dysuria; Frequency; Haematuria.

Special Investigations:

- Urine Dipstick: Blood & Protein elevated.
- Urine MCS: terminal urine sample: the timing of urine collection is also NB: increased ovum secretion post-exercise & in the middle of the day. Microscopy will show a terminal spike indicating S.haematobium.
- Bloods:
  - FBC: eosinophilia (acute phase)
  - Serology: Schistosoma ELISA.
  - U&E: usually no changes in urea/creatinine/GFR.
- Radiology: IVP: calcifications of bladder wall; dilated ureters; filling defects; hydro-nephrosis & small bladder capacity.
- Cystoscopy; definite diagnosis but only done if unsure of diagnosis or if complications are apparent
- Causative Microorganism: Schistosoma Haematobium

anagement:

- Medical Management: Praziquantel
- Prophylaxis/Prevention: improve housing & sanitation; snail adication; be aware when entering a high endemic area.

Complications:

- Hydronephrosis
- Iron-deficiency anaemia
- Acute urinary retention
- Renal Failure due to obstructive uropathy
- Strangury: painful, frequent urination of small volumes expelled slowly in spite of straining & urgency, ending with a feeling of incomplete voiding.

Haemorrhagic Cystitis:

Causes:

- n occur with acute bacterial or viral (adenovirus) infections.
- Daily Cyclophosphamide per os (15% of cases).

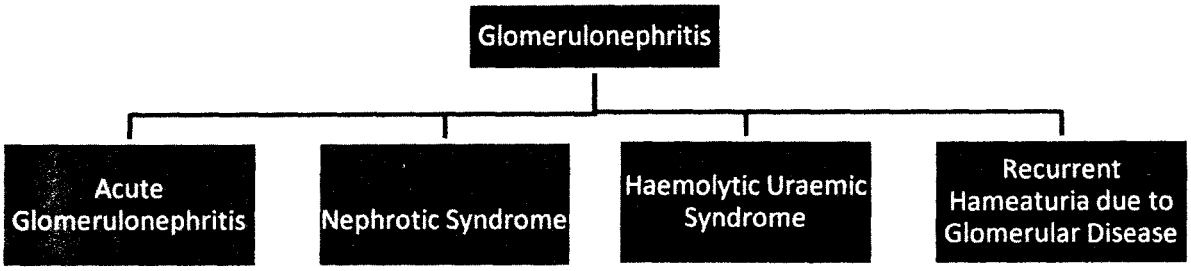
Clinical Features:

- Gross Haematuria
- Frequency.
- Urgency
- Dysuria
- Supra-pubic pain.

Special Investigations: U-dipstick; Urine MC&S; U/S (Kidneys/Ureters/Bladder); +/- VCUG.

Management:

- Treatment as for UTI if suspected bacterial cause.
- Cyclophosphamide: treatment with MESNA disulfide inactivates the cyclophosphamide metabolites that cause the bladder damage, and together with good hydration will prevent the haemorrhagic cystitis. Usually used for when cyclophosphamide is given IV, maybe difficult/impractical if taken orally.
- Viral organism: usually the symptoms resolve within a few days.



Acute Glomerulonephritis:

- Definition: acute inflammation of the glomerulus causing a decreased GFR (Glomerular Filtration rate).
- Causes of Acute Glomerulonephritis:

Cause	Organism
Bacteria	**Streptococcus (PSGN)**; Staphylococci, S.typhi, T.pallidum, Pneumoccoci
Viral	Hep B; Echovirus; EBV; VZV; **HIV**
Protozoa	Malaria
Collagen Vascular Disease	Henoch-Schönlein Purpura; SLE; Polyarteritis Nodosa; Systemic Sclerosis
Genetic	Alport's Syndrome
Drugs	Methicillin
Miscellaneous	Sickle cell disease; Sarcoidosis; Irradiation

The most common cause in the South African setting is Post-Streptococcal disease (PSGN).

- Pathogenesis:

TRIGGER: behind the PSGN (Post-Streptococcal Glomerulonephritis) is a Group A-β-haemolytic streptococcal infection of the skin or the throat.

↓

Immune Response: formation of immune complexes into circulation that are trapped in the glomeruli.

↓

Complement Cascade Activation: by the immune complexes, results in inflammation & polymorphonuclear leucocyte attraction.

↓

Pathology: Endothelial cells swell, fibrin deposited & capillary lumens occluded.

↓

Proteolytic enzymes damage the basement membrane resulting in blood cells & plasma constituents to escape in the urine.

® PSGN according to infected site:

	Skin	Throat
Country	Tropical/Subtropical (Blacks>Caucasians)	Temperate
Season	Summer	Winter/Spring
Age of Onset	Pre-School and School going child	Primarily school-going children
Sex Distribution	M=F	M>F
Risk of Developing Nephritis	High	Low
Period from infection to nephritis	21 days	10 days
Antibodies	Anti-DNase B Anti-hyalouronidase	Anti- NADase ASOT

Clinical Features:

- Well Nourished children(mount a good immune response)
- Abrupt onset, most commonly presenting with haematuria, proteinuria, casts in urine (red cell casts=glomerulonephritis) & oedema.
- Hospital admission required if: (i) Oliguria.
  (ii) hypertension: complicated with encephalopathy: headache, restless, drowsy, vomits, blurred vision, and convulsion.
  (iii) severe oedema: usually peri-orbital.

Special Investigations:

- Urine Testing: Red to brown colour; decreased GFR; oliguria.
- Bloods: Urea; Creatinine; Electrolytes (NB K+ level); acid-base study; FBC (mild leucocytosis); ECG (monitor hyperkalaemia).
- Streptococcal Antibodies: Anti-DNase B; Anti-hyaluronidase; Anti- NADase; ASOT.
- C3 : decreased.

Management:

- **Admission if**: Oliguria; Hypertensive or severely oedematous.
- **Fluid Management & Food Intake**
  :Oliguria (<300ml/day): restrict fluids to 300-400ml/day + previous days urine output.
  :High calorie diet (400 kcal/day), but protein restricted if urea is high.
  :Lactose>Glucose & Dextrose (more readily palatable).
  : Start normal diet once kidney function restored.
- **Regular Monitoring:** Fluid intake/output; weight; urine testing; 3-hourly BP; and serum biochemistry (K+).
- **Hyperkalaemia:** treatment vital if ECG changes occur or if serum K+ >7,0mmol/l. Treatment involves:
  - Calcium gluconate (10%) 0,5ml/kg over 2-4 mins with ECG running. Sodium Bicarbonate (decreses risk of arrythmia & reverses acidaemia) 2,5mmol/kg once, &not repeated if patient is hypertensive due to sodium load. Glucose (50%) 1ml/kg IVI & then Glucose (30%) IVI at rate rerquired for patient's fluid balance. Insulin added at 1unit per 3g glucose: requires blood sugar monitoring. Works within 1-2 hours.
  - If K+ <6,0mmol/l with normal ECG, omit calcium gluconate and give oral ion exchange resin or by retention enema.
  - If K+ <6,0mmol/l after 2-3hours, repeat step 1 and plan for peritoneal dialysis.

Management Continued:

- **Drugs:** avoid drugs exreted by the kidneys (especially digoxin)
  - # Diuresis: Furosemide: 1ml/kg IVI: pulmonary oedema & circulatory congestion.
  - # Penicillin: after skin/throat swab taken for MC&S.
  - # Convulsions: Diazepam.
  - #Hypertension: anti-hypertensive.
- **Circulatory Congestion & Pulmonary Oedema Steps:**
  1) Oxygen
  2) IVI Furosemide
  3) Rotating tourniquet
  4) Venesection
  5) Peritoneal dialysis
  6) Artificial ventilation
- Indications for dialysis:
  - Deteriorating clinical condition: uraemic Syndrome (severe encephalopathy; bleeding tendency, blood effusions; colitis).
  - Fluid overload unresponsive to diuretic therapy.
  - Unresponsive hyperkalaemis to treatment.

Unusual Complications:

- Rapidly progressing Glomerulonephritis: >1% of PSGN cases, where there is prolonged oligo-anuria, progressing to chronic renal failure & death. On histology there is crescent formation of glomeruli. No good treatment: Quadruple thrapy advised: cyclophosphamide, steroids, anti-coagulants & anti-platelet drugs.
- Nephrotic Syndrome.

Long Term Sequelae:

- Majority have no long term sequelae.
- <1% get chronic nephritis & hypertension.

Recurrent Haematuria due to Glomerular Disease: Macroscopic haematuria during a mild URTI or after exercise: the following are the usual causes:

- Berger's Disease (IgA Nephropathy)
- Benign Familial Haematuria
- Glomerulonephritis: Minimal change, Mesangial proliferative
- Alport's Syndrome: Family History, Male; Eye signs; deafness; progressive renal impairment.

Nephrotic Syndrome:

- Definition: heterogeneous group of glomerular disorders resulting in heavy proteinuria (2g/m2/day or U-dipstick 3+ or 4+). Increased protein loss = hypoalbuminaemia &/or oedema.
- Clinical Features:

Signs & Symptoms	• Oedema: usually peri-orbital &worse in the morning. In black children may become profuse- Anasarca. • Usually there are no other signs of renal disease, and the child often does not appear ill.
Special Investigations	• Urine dipstick: 3+/4+ protein. • U-MCS: hyaline casts. • Bloods: raised serum lipids & α2-globulin
Complications	• Predisposition to infection: pneumococcal/gram negative primary peritonitis. • Vascular thrombosis • Circulatory Failure: cold peripheries; cyanotic extremities; pallor; hypotension and a thread pulse.

- Causes of Nephrotic Syndrome: the actual cause in the majority of cases is unknown, but the following are some of the most common:

Infections	**P.malariae**; **S.mansonii**; T.pallidum; Streptococci; **Hepatitis B**; Infective endocarditis; hydatid disease.
Toxic Causes	Heavy metals (mercury, lead); drugs (penicillamine, captopril)
Allergy	Bees; Pollen
Vasculitis	Henoch-Schönlein Purpura; SLE; Polyarteritis Nodosa; dermatomyositis
Malignancies	Lymphoma
Miscellaneous	Renal vein thrombosis; constrictive pericarditis; diabetes; amyloidosis; Alport's syndrome; Haemolytic-Uraemic Syndrome

- Management:
  - **Supportive:** low salt; high protein diet.
  - **Diuretics:** for severe oedema. Beware of furosemide: can complicate with hypotension & volume depletion if very hypo-albuminaemic child. Use a K+ sparing diuretic.
  - **Albumin (20%) IVI at 1g/kg over 6 hours**: if severely oedematous and oliguric with severe intravascular volume depletion.
  - **Steroids:** but contra-indicated if cause is Hepatitis B.
  - **Cyclophosphamide:** Frequent relapse/steroid dependant.
  - If unresponsive to steroids: needs renal biopsy & Steroid + Cyclophosphamide.
  - **Adjunctive therapies**: anti-hypertensives; anti-coagulants; Lipid lowering drugs & vitamin supplementation.
  - **New Drugs**: Tacrolimus: improved prognosis in black children.

Initial Episode:
Prednisone 60mg/d/m2 (max 60mg/day) in divided doses over 4 weeks. Then 40mg/m2 as single dose on alternate days for 4 weeks. Reduce to 15mg/m2/month over 2,5 months

First 2 Relapses:
Prednisone 60mg/d/m2 (max 60mg/day) until remission, follow with 40mg/m2 (max 60mg/day) on alternate days for 4 weeks.

Frequent Relapses:
Maintenance Prednisone 0,1-0,5mg/kg/alternate day for 3-6months, then reduce dose

Relapse on Prednisone: .1mg/kg/alternate days

Cyclophosphamide 3mg/kg/day po for 8 weeks

Post-cyclophosphamide relapses or steroid resistance: Do a **BIOPSY**.

However, the disease differs depending on geographical environmental & genetic factors.

	Non-African Children	African Children	Malaria Areas	Non-malaria areas
Incidence /Prognosis	Rare in non-tropical areas with good prognosis if properly managed	Infective causes primarily. Malaria; Hep B (assoc. with membranous nephropathy); Bilharzia.	Higher incidence of nephrotic syndrome in malaria areas.	Primarily Black Southern African children, where there is 'obvious' structural lesions of the glomerulus.
Histology	Minimal change disease (80% of all paeds nephrosis cases)	Renal biopsy required for management & prognosis.	Structural glomerular disease; especially diffuse proliferative glomerulonephritis.	• Hep B : associated with membranous nephropathy • Other: Focal segmented glomerulosclerosis (now the most common)
Clinical Presentation	• Male; <5years • Highly Selective Proteinuria • Normal Renal function	In SA: whites & Indians have minimal change disease, but varies in blacks.	• M=F; 5-8years • Poorly Selective Proteinuria.	• Preschool & 8-11 year olds. • Proteinuria • May be associated with hypertension & haematuria.
Treatment	Respond well to steroid therapy	Respond well to steroid therapy.	Poor response to anti-malarials, steroids & immune-suppressants.	Poor responsiveness to steroids & cyclophosphamide.
Complications	Frequent relapses (2 in 6months or 4 over 12month period).Managed on variety of drugs.		Death due to progressive renal failure common.	

- Indications for Renal Biopsy:
    1. Steroid resistant/dependant or frequent relapse of nephrotic syndrome & more aggressive immunosuppressant therapy is being contemplated.
    2. Persistent Hyper-Complimentaemia with proteinuria or worsening renal function.
    3. Congenital or infantile nephrotic syndrome.

Haemolytic Uraemic Syndrome:

Triad	1. Haemolytic Anaemia (fragmented cells on peripheral blood smear). 2. Thrombocytopenia. 3. Acute Renal damage manifesting as Uraemia.
Epidemiology & Aetiology	• Primarily in Infancy & childhood. • Causes: o Post-Diarrhoea (90%): Infection: **E.coli 0157:H7 & S.dysenteriae type 1: shiga like toxin producing bacteria.** o Non-Diarrhoea Related: S.pneumoniae, Glomerulonephritis, Maignancy, Collagen Vascular Diseases, Drug-induced, Post-transplant & idiopathic. o Inherited: Autosomal dominant/recessive. o Sporadic
Pathology	Damage to endothelial cells in the glomeruli (especially shiga-like toxins). These cells release cytokines, leading to blockage of small blood vessels. The endothelial cells become swollen & dislodged from basement membrane. The sub-endothelial area now exposes collagen with resultant platelet activation, and the space fills with debris: platelets, fibrin & lipids. Swollen Endothelial Cells + Debris in Sub-endothelial Space + Thrombi = Reduced Capillary diameter = Reduced GFR.
Clinical Features	• Features preceding renal failure: bloody diarrhoea, vomiting & URTI. Shigella will have marked abdominal pain/tenderness. • Signs of Acute Renal Failure with severe pallor & oliguria/anuria; convulsions; purpura +/- frnak bleeding. • GIT signs: may become severe or persistent in some cases. • Factors suggesting Poor Outcome in HUS: o Age of <2 years o Prolonged oligo-/anuria o Neurological damage o High WBC count (>20x 10^9/ℓ) o S. Dysenteriae type 1
Complications	**MHICE: M**etabolic disturbances; **H**aemorrhaging; **I**nfection; **C**ardiac Failure; **E**ncephalopathy
Management	• Slow continuous form of dialysis (haemodiafiltration): modality of choice: recover within ten days; if unavailable, the performance of a peritoneal dialysis is just as effective. • Plasmapharesis: if recurrent. • Control: convulsions, hypertension, fluids, electrolytes (replenish Na+ loss in Shigella), energy requirements. • Dialysis: indicated in volume overload or hyperkalaemia not responding to treatment or clinical syndrome of uraemia. • Blood Transfusion: may aggravate/induce hypertension (only if HB <6g/dℓ). FFP's may be helpful.
Diff Diagnosis	DIC; Thrombotic thrombocytopenic purpura

Hypertension in Childhood:

Measurement of BP

- 2-3 year old: taken by auscultation. Cuff size should cover 2/3's of upper arm &inflatable bladder must encircle arm girth.
- Infancy: can use alternate techniques such as Doppler U/S or Flush Method.
- If BP sufficient with pos+ clinical features, one measurement is enough, but if there is doubt, BP should be measured on three seperate occasions.

Defintion of Hypertension in Children:

Rule of thumb:

SBP= 100+2,5xage(years)

DBP= 70+1,5xage(years)

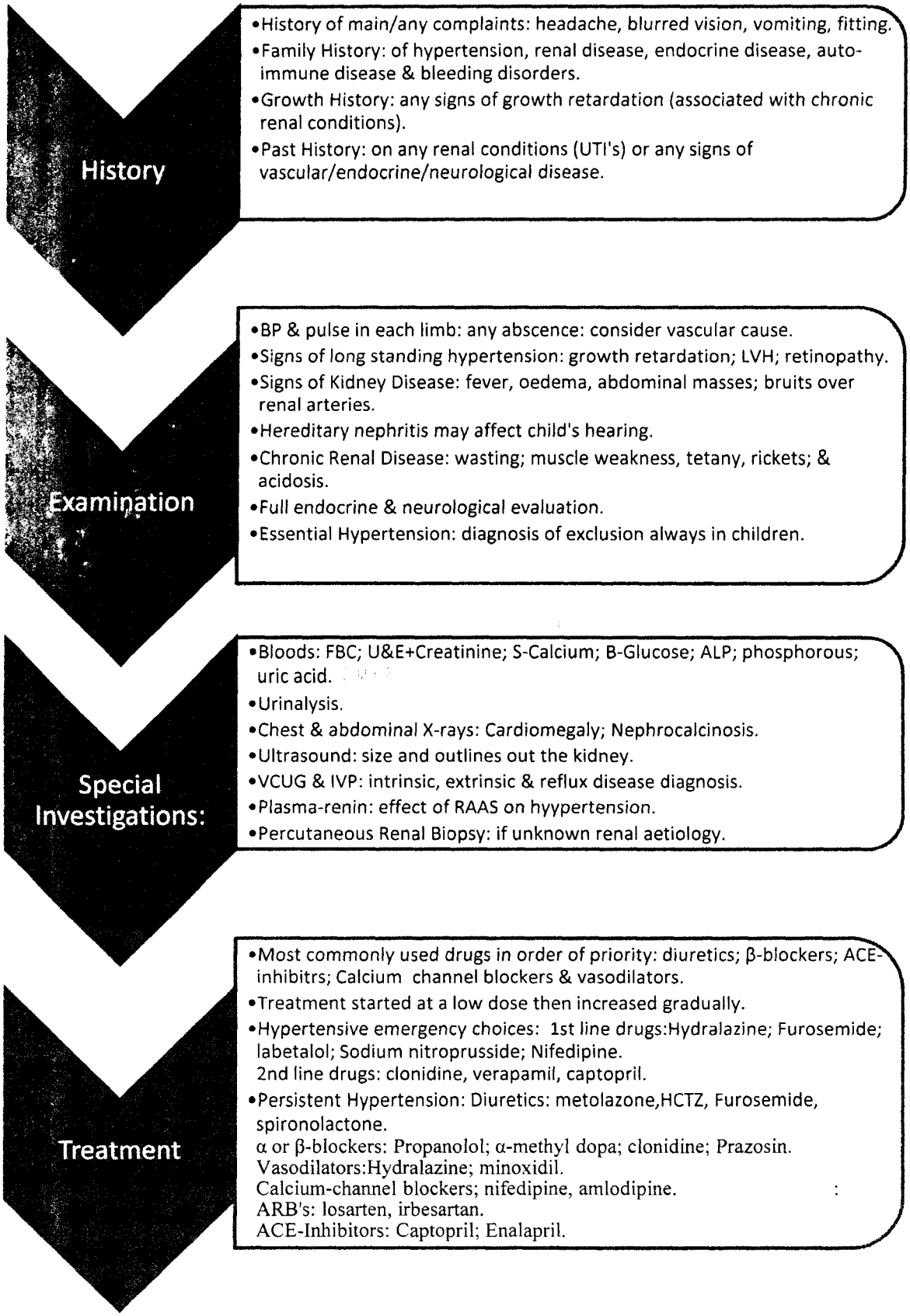
- Newborn: 1st 12hrs: Prem (65/45); Term (80/50)
- Newborn:1st week: Prem (80/50); Term (100/70)
- 6 weeks-6 years: >115/80
- 8years: >120/82
- 9 years: >125/84
- 10 years: >130/86
- 12 years: >135/88
- 14 years: >140/90
- 16 years: >145/92
- 18 years: >150/94

S° Causes of Hypertension:

80% of which are renal diseases

- **Renal Diseases (80%):** Glomerulonephritis; Interstitial nephritis; Reflux/Onstructive nephropathy; Congenital abnormalities; Renin secreting tumours.
- **Vascular Diseases:** coarctation of the aorta; Renal artery stenosis/thrombosis.
- **Endocrine Disorders:** Adrenal disorders; Hyper-throidism/parathyroidism; Neuroblastoma; Diabetes.
- **Miscellaneous:** Polio; excessive liquorice ingestion; Neurofibromatosis.

Approach to Hypertensive Child:



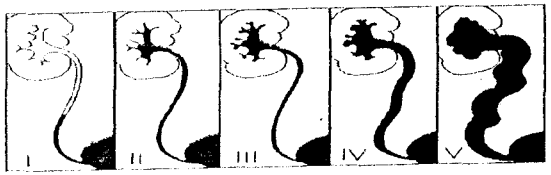
Inherited & Congenital Renal Disorders:

Posterior Urethral Valves:

- Definition: abnormal mucosal membrane that usually act as a valve to obstruct urine flow; usually obstructing the urethra just distal to the veramontanum.
- Pathology: proximally dilated urethra, bladder trabeculations form & formation of bilateral hydro-uretero-nephrosis.
- Presentation: most common cause of obstructive uropathy: usually associated with a dilated bladder. The greater the obstruction, the earlier the patient presents, usually up to 2/3's present before 1 year. Often associated with UTI's in male children.
    - Neonate/Infants: UTI's; Acute Renal Failure; Failure to thrive; Respiratory distress (associated with bilateral hydro-ureteronephrosis); palpable bladder; urinary ascites.
    - Older Child: Recurrent UTI; Overflow incontinence; chronic renal failure.
- Special Investigations: VCUG (Voiding Cysto-urethrogram) or cystoscopy to confirm diagnosis.
- Management:
    - Resuscitation: fluids, electrolytes. Antibiotics if UTI.
    - Urethral catheter: relieve obstruction.
    - Refer to UROLOGY for ultrasound/VCUG/Cystoscopic repair.

Vesico-Ureteric Reflux (VUR):

- Definition: The backflow of urine from the bladder into the ureters, that is usually prevented by a flap-valve mechanism at the junction between the bladder and the ureters. If this "flap" is not properly developed, reflux will occur.
- History:
    - Usually presents with Urinary Tract Infection.
    - Family History: there is a definite genetic predisposition.
    - Social History: occurs less in black children.
- Examination: Ill appearance of child
    - Infancy: lethargy; failure to feed; unstable temperature; apnoea; poor colour; jaundice; anaemia; purpura; & H/S-megaly.
    - Children: usually urgency as an isolated complaint.
- Special Investigations:
    - U/S: may be helpful to exclude urinary obstruction if child is acutely ill.
    - Cystography: investigation of choice.
    - Technetium scanning: recommended after the acute phase of the illness to detect renal scarring.
- Grading: Grd 1: ureters only; Grd 2: ureters,pelvis & calyces affected without dilatation.



Grade 3: mild/moderate dilatation /tortuosity of ureters &mild dilatation of calyces/pelvis.
Grade 4: grade 3 + obliteration of the sharp angle of fornices.
Grade 5: Gross dialatation & tortuosity.

Management:

- Treatment of the UTI: then continuous prophylactic treatment with low dose macrodantin/ trimethoprim/ cefaclor (especially effective in young infants preventing UTI's and their complications).
- Indications for Surgery:
    - Failure of Medical treatment of UTI.
    - Poor compliance to medical treatment.
    - Very severe reflux that is unlikely to resolve spontaneously.
    - Persistent VUR in an adolescent female.
- Types of Surgery:
    - Endoscopic: minimally invasive but only indicated in mild to moderate cases.
    - Open Surgery: re-implantation of the ureters into the bladder: excellent results (>90% success rate).

Approach to Acute Renal Failure (ARF):

1. Diagnosis
2. Special Investigations/Aetiology
3. Management
4. Complications
5. Pathology
6. Counselling of Mother

History: history is important in finding out the cause behind the RF.

Signs:

- Hypertension: raised BP.
  Oedema: Fluid overload.
- Tachypnoea: child is hyper-ventilating.

Special Investigations:

Urinalysis:

- U-output: falls below 180ml/m2/day or <0,5ml/kg/hour: Oliguria.
- U-sodium & U-creatinine.

Bloods:

- U&E + Creatinine: raised s-creatinine & b-urea (Azotaemia); hyperkalaemia; Hyponatraemia; Hypocalcaemia.
- FBC: normocytic normochromic anaemia.
- Blood Gas: Acidosis.

Ultrasound: to exclude any obstruction of the urinary system.

Complications:

- Parenchymal Kidney damage: if left untreated.
- Metabolic acidosis
- Hyperkalaemia: cardiac complications.
- Pulmonary oedema

Management:

- Pre-Renal Failure:
  - primary concern is to replace lost body fluids: Normal Saline/Plasma/Blood 20mℓ/kg over 30-60mins.
  - If Gastro-enteritis is the cause: us IVI half-Darrow's dextrose.
  - Then require dieresis:
    Furosemide 2mg/kg IVI
    Dopamine 1µg/kg/min
    Wait 60 mins for dieresis.
  - Reassess hourly, monitor with CVP.
  - No Response, more than likely renal parenchymal damage: reduce fluids.
- Post-Renal Failure: obstruction of urinary system. Relieve this by urinary catheterization.

Possible Pathophysiology of ARF:

i. Hypoperfusion of Kidney: vascular occlusion.
ii. Occlusion of smaller blood vessels: afferent & efferent arterioles usually affected.
iii. Glomerular abnormality
iv. Tubular dysfunction/damage
v. Obstruction of Urinary tract

Site	Cause	Causative Agent
Pre-Renal	**Dehydration**	Gastro-Enteritis (GE)
	Shock	GE; haemorrhage; burns
	Cardiac Failure	Septicaemia
	Hypoproteinaemia	Nephrotic Syndrome
	Vascular Occlusion	Renal artery occlusion
Renal	**Acute Glomerulonephritis**	**PSGN**; Non-PSGN causes
	Haemolytic Uraemic Syndrome	Dehydration/Shock/Nephrotoxins
	Tubular Necrosis	Septicaemia; Ischaemia; Intravascular coagulation; dehydration
	Cortical Necrosis	
	Pyelonephritis	
	Interstitial Nephritis	Drugs:Methicillin/ Sulphonamides
	Congenital Renal Abnormalities	Polycystic Kidneys; Renal dysplasia
Post-Renal	Obstructive Uropathy	Congenital(urethral valves) vs Acquired (Infection/drugs)
	Vesico-Ureteric Reflux	

Tests to distinguish between Pre-Renal & Renal Failure

	Pre-Renal	Renal
Fractional Excretion of Sodium:	<1%	>3%
U-sodium (mmol/ℓ)	<20	>25
Urine/plasma ratios of:		
Osmolality	>1,2	<1,1
Urea	>8	<8

Fractional Excretion of Sodium= (U-Sodium/Plasma-Sodium) x (Plasma Creatinine/U-creatinine) x 100

Approach to Chronic Renal Failure:

1. Diagnosis
2. Special Investigations/Aetiology
3. Management
4. Counselling of Mother

Diagnosis:

History: patient presents usually with a history of weakness, poor growth, loss of weight, polyuria, polydipsia, & vomiting.
There may be no signs or symptoms at presentation of any prior renal disease.
Examination: the patient is usually pale/anaemic, hypertensive, wasted, hypotonic, neurologically disturbed. May also present with signs of tetany & rickets.

Special Investigations:

- Bloods:
  - U&E: hypokalaemia; hyperphosphataemia; hypocalcaemia.
  - Parathyroid hormone: raised.
  - FBC: anaemia.
- Urinalysis: GFR chronically <30% of expected.
- Blood Gas: acidosis.
- Ultrasound: Chronic renal failure kidneys are usually smaller in size.

Aetiology:

- Glomerulonephritis.
- Nephrotic Syndrome(focal segmental glomerulosclerosis)
- Pyelonephritis.
- Congenital Renal Abnormaities.

Management of Chronic Renal Failure

Water & Electrolyte Mx:

Water: balance input &output

Na+: restrict if hypertensive

-K+: Hyperkalaemia: restrict intake; ayexalate 1g/kg/dose & oral bicarbonate

Nutrition:

High energy intake (fats & CHO); Moderate protein intake; water soluble vitamins, iron & zinc supplementations

Acidosis:

If sodium bicarbonate <20 mmol/ℓ: oral citrate solution or bicarbonate.

Infection:

Use appropriate antibiotic, and adjust dose if excreted by kidney.

TREATMENT OF CHOICE:

Chronic renal failure is an irreversible renal disease leading to insufficient functioning of the renal tissue; thus the following are the gold standards in management of the condition:

- Dialysis: when symptoms become unmanageable and while awaiting transplantation.
- Transplantation

Renal Osteodystrophy:

- Hyperphosphataemia: lower phosphate intake; Calcium carbonate (phosphate binder)
- Hypocalcaemia: oral calcium supplementation.
- Hyperparathyroidism: Dihydroxy-Vitamin D supplementation

Anaemia:

- Hb <10g/dℓ: iron+folate supplements. Consider EPO.
- Hb <6g/dℓ: Packed Red cell transfusion @ 10ml/kg slowly.

Hypertension:

Emergency HT:
Nifedipine/Diazoxide, or Furosemide if fluid overload present.