Pediatrics Notes

Department of Child Health School of Medical Sciences KNUST

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Preface

The Professors, Senior Lecturers and Lecturers in the Department of Child Health, School of Medical Sciences, Kwame Nkrumah University of Science and Technology put together these clinical notes. Members of the Child Health Department include:

Prof Sampson Antwi
Prof Daniel Ansong
Prof Alex Osei-Akoto
Prof Emmanuel O. A. Addo-Yobo
Prof Joslin Alexei Dogbe
Prof (Mrs) Gyikua Plange-Rhule
Dr Samuel Blay Nguah
Dr Emmanuel Ameyaw
Dr Anthony Enimil
Dr (Mrs) Vivian Paintsil
Dr Serwaa Asafo-Agyei
Dr Charles Hammond
Dr (Mrs) Sandra Kwarteng Owusu
Dr Adwoa Pokua Boakye Yiadom
Dr Naana Ayiwa Wireko Brobby
Dr (Mrs) Akua Afriyie Ocran

1 List of Contributors

The following are the contributors to this book:

Prof. Sampson Antwi

MB ChB, FWACP, FGCPS

Prof Sampson Antwi is a Professor of Pediatric Nephrology at the Department of Child Health, School of Medical Sciences, Kwame Nkrumah University of Science and Technology in Ghana. He is currently the Head of Department. He also has vast experience in pediatric HIV care and is a fellow of the International Pediatric Nephrology Association.

Prof. Daniel Ansong

MB ChB, FWACP, FGCPS

Prof Sampson Antwi is a Professor of Pediatric Nephrology at the Department of Child Health, School of Medical Sciences, Kwame Nkrumah University of Science and Technology in Ghana. He is currently the Head of Department. He also has vast experience in pediatric HIV care and is a fellow of the International Pediatric Nephrology Association.

2 Child History & Examination

3 Growth and Development

4 Pediatric Anthropometry

Part I

Neonatology

5 Neonatal History & Examination

6 Neonatal Delivery Pathologies

6.1 The health newborn

- Cries / Breathes normally
- Pink all over
- Well-flexed & moves all limbs spontaneously
- Suckles well at the breast
- Birth weight 2.5 – 4.0kg
- Normal vitals signs

6.2 Occurrences at birth

- The fluid in the alveoli is absorbed and replaced by air. If the transition is not smooth, it results in insufficient oxygen delivery to the vital organs…
- Poor muscle tone
- Respiratory distress or depression
- Slow heart rate
- Low BP
- Cyanosis

6.3 Birth Asphyxia

6.3.1 Definition

> **!** World Health Organisation definition
>
> Birth Asphyxia is the medical condition resulting from deprivation of oxygen in the newborn that lasts long enough during the birth process to cause harm, usually to the brain.

6.3.2 Risk factors

Any condition that will lead to impairment of oxygenation or blood flow to the newborn's brain in the perinatal period. These include:

- Prolonged labour (CPD)
- Placental failure
- Cord around the neck
- Problems with oxygenation of maternal blood / maternal disease
- Anaemia and bleeding in the baby
- Congenital heart disease
- Infections
- Deficient medical skills and or knowledge

6.3.3 Presentation

The asphyxiated baby may have any of the following:

- Poor Apgar Scores
- May not cry at birth
- Floppy/spastic
- Breathing problems
- Unresponsive
- Seizures
- Irritable

6.3.4 The APGAR Score

It is an objective method of quantifying the newborn's condition. And is useful for conveying information about the newborn's overall status and response to resuscitation

Table 6.1: The APGAR Score

	0	1	2
Heart Rate	0	<100	>=100
Respiration	0	Weak or Irregular	Good Cry
Reaction	None	Slight	Good
Colour	Blue or Pale	Pink body limbs blue	All pink
Tone	Limp	Some movement	Active movement, limbs well flexed

8-10 = No Asphyxia
5-7 = Mild Asphyxia
3-4 = Moderate Asphyxia
0-2 = Severe Asphyxia

6.3.5 Management

- Largely supportive
- Newborn resuscitation/oxygenation
- Correction of fluid & electrolyte imbalances including shock
- Control of seizures
- Treatment of any underlying infection
- Look out for birth injuries
- Active cooling found to improve neurological outcome
- Temperature maintenance
- Full Blood Count, Culture & Sensitivity, Blood glucose etc
- Serum electrolytes: Na, K, Ca & Mg
- Start empiric 1st line antibiotics according to protocol: / X'pen & Gentamycin
- Start IV Fluids at 50ml/kg (Plain 10% Dextrose).
- Pass a urethral catheter and monitor the baby's urine output.
- The target temperature of the baby is 36.50C – 37.50C

6.3.6 Hypoxemic Ischaemic Encephalopathy

- The most important consequence of birth asphyxia is
- The outcome ranges from complete recovery to death
- 25 - 30% end up with permanent damage like Cerebral palsy & Mental retardation
- Prognosis dependent on gestational age, management of metabolic & cardiopulmonary complications & the severity of the encephalopathy
- Subsequent competent care and available facilities also influence the outcome

6.4 Birth Injuries

A birth injury can simply be referred to as any form of damage incurred by the baby during the birthing process. Injury may occur as a result of inappropriate or deficient medical skill or attention or may occur despite skilled and competent obstetric care.

Predisposing conditions include:

- Cephalopelvic disproportion (CPD) / Small maternal stature / Primiparity

- Macrosomia
- Shoulder Dystocia
- Prematurity
- Prolonged or precipitous labour
- Abnormal presentation
- Instrumentation
- Handling after delivery

6.4.1 Fracture

Generally, the affected limb looks deformed or swollen, and the baby barely moves it on account of pain

6.4.1.1 Clavicle

This is the most fractured bone during delivery; mostly during delivery of the shoulder in vertex and of the extended arms in the breech. Signs of a fracture may include no free arm movement on the affected side, crepitus and bony irregularity, and absent Moro reflex. It has an excellent prognosis, even though it is commonly missed. Treatment, if any, includes immobilization of the arm and shoulder as shown below.

This is the first type of humeral fracture

Figure 6.1: Humeral Fracture (immobilised)

6.4.1.2 Humerus

The x-ray below shows another commonly fractured bone, the humerus.

Figure 6.2: X-ray of a humeral fracture

6.4.1.3 Femur

Risk factors: big baby, breech presentation, incompetency. The affected thigh looks deformed, swollen and may be reddened. The main mode of management involves splinting the limb from the waist to below the knee.

Figure 6.3: Femoral Fracture

Figure 6.4: Femoral Fracture Splinting

6.5 Nerve injuries

6.5.1 Brachial plexus injuries

The nerves of the brachial plexus may be compressed, stretched or torn in a difficult delivery. Paralysis occurs as a result of nerve compression from either haemorrhage or oedema. Permanent paralysis can occur from the tearing of the nerve or avulsion of the nerve root from the

spinal cord or oedema. Erb's palsy (C5-C6) is the most common type of BPI and is associated with a lack of shoulder motion. The involved extremity lies adducted, prone, and internally rotated. Grasp reflex is usually present and prognosis is generally good. Also described as the Waiter's tip position.

Figure 6.5: Erb's Palsy

6.5.2 Klumpke's paralysis

6.5.3 Facial nerve paralysis

Loss of voluntary muscle movement in the face on account of pressure on the facial nerve during the delivery process. Risk factors include instrumental delivery, poor delivery skills, big baby etc. Usually resolves spontaneously after a few months

6.6 Scalp Injuries

6.6.1 Cephalhematoma

Tearing or disruption of the superficial veins under the periosteum leads to haemorrhage and subsequent swelling. Suture lines confine the cephalhematoma and limit the extent of the bleeding. There could be an underlying linear skull fracture. Prognosis is good with most of them resolving between 2 weeks to 3 months.

6.6.2 Subgaleal Hemorrhage

The subgaleal space is located between the galea aponeurotica & the periosteum. The space extends from the orbital ridges to the nape of the neck and laterally to the ears. Bleeding is caused by damage to the large emissary veins located in the subaponeurotic layer. The bleeding associated with subgaleal haemorrhages can be extensive. Clinically, the baby may present with pallor and lethargy, followed by tachycardia, tachypnea and hypotension. The scalp may appear tight and boggy and complications include anemia, hypovolemic shock and jaundice.

Figure 6.6: Cephalhematoma

Figure 6.7: Subgaleal Bleed

6.7 Visceral injuries

6.7.1 Liver and spleen

Usually results from pressure on the liver during delivery of the head in breech presentations. Risk factors include macrosomia, intrauterine asphyxia, extreme prematurity, and hepatomegaly

6.8 Respiratory Distress

6.8.1 Meconium aspiration syndrome

Fetuses sometimes pass meconium whilst in utero as a result of some form of stress. If the stress has been going on for a while and the fetus has been passing meconium for a few days, the cord, skin and nails may be stained. Occurs when the fetus passes meconium into the surrounding liquor and then aspirates this into the lungs. Tends to happen in term and post-date babies. Distressed fetuses tend to pass meconium either just before or during the delivery

process. The smaller the amniotic fluid volume, and the more meconium the baby passes, the thicker the fluid and the more dangerous it becomes if aspirated.

History of Pregnancy and delivery looking for predisposing factors such as fetal distress, post-maturity, meconium-stained liquor etc. Physical examination looking for signs of meconium-staining on the baby, and evidence of respiratory distress (fast breathing, chest indrawing, cyanosis etc.). Investigations include FBC, Blood C&S, and sometimes a chest X-ray depending on the severity. Management is mainly supportive. Includes antibiotics, respiratory support, supportive treatment, IVFs and nutrition.

6.8.2 Transient tachypnoea of the newborn

Caused by delay in clearance of fetal lung fluid. Typically resolves within 72 hours. Often associated with Caesarean Section Delivery. Severity varies but is often mild with just tachypnoea. Management involves supportive treatment of the respiratory with oxygen.

7 Neonatal Emergencies

7.1 Introduction

The usual newborn cries on delivery, is pink all over, well flexed and moves all limbs spontaneously, suckles well at the breast, breaths normally and weighs between 2.5 – 4.0kg. Neonatal emergencies are not uncommon and encompass a wide range of conditions occurring in the first 28 days of life. The classical mnemonic for these is **THE MISFITS.**

Trauma/Abuse, **H**eart & Lung, **E**ndocrine, **M**etabolic disturbances, **I**nborn errors of metabolism, **S**epsis, **F**ormula, **I**ntestinal, **T**oxins, **T**risomies, and **S**eizures.

7.2 Approach to newborn emergencies

Presenting features of many serious neonatal disorders are nonspecific. The history and physical examination are essential in the overall approach to the patient. Prenatal, perinatal and postnatal history play a huge role in neonatal assessments They guide and inform the health worker on the most appropriate investigations, which would eventually lead to a correct diagnosis. A complete history may unmask the likely cause of symptoms and guide further questioning, for example, sepsis.

Examination on the other hand involves assessing the Airway, Breathing, Circulation, Random Blood Sugar, provision of Oxygen and checking Oxygen saturation. This can be pre- and postductal. Others include temperature checks and other vital signs. The weight, current weight, head circumference, and length. Intravenous access should be obtained for possible further treatment. Appropriate investigations should also be done.

Requisite investigations should also be done accordingly.

7.3 Respiratory Emergencies

This is one of the most common and includes:

1. Primary pulmonary Hypertension,
2. Meconium Aspiration Syndrome,

3. Congenital Pneumonia
4. Birth Asphyxia and
5. Respiratory Distress Syndrome

7.3.1 Respiratory Distress Syndrome

RDS is due mainly to a lack of surfactant in the lungs. Surfactants are essential for reducing the surface area of the lungs, thus helping in breathing. Incidence and severity increase with decreasing gestational age. Other risk factors include prematurity, male gender, multiple gestations, being born to a mother with diabetes mellitus and hypothermia. Signs of RDS include tachypnea, grunting, recessions and cyanosis. Prevention involves preventing preterm births and administering corticosteroids to the mother of gestation between 24 to 34 weeks before delivery. Treatment however involves the administration of surfactant.

Note that this condition is different from *Respiratory Distress* in a newborn. Respiratory distress is a more generalised term used as a single or a combination of signs as a result of increased work of breathing. It can result from pulmonary as well as non-pulmonary causes. These include cardiac, neurological (eg. Asphyxia), haematological (Anemia) and sepsis. It occurs in both term and preterm children.

7.4 Endocrine Emergencies

Neonatal jaundice is the most prominent endocrine disorder in this section. This is appropriately discussed in Section 8.1

7.5 Metabolic Emergencies

7.5.1 Hypoglycemia

Hypoglycemia is common in the stressed neonate and glucose levels should be monitored regularly. In the newborn period, it is defined as a random blood glucose of < 2.6mmol/l. Risk factors include sepsis, Infant of a diabetic mother, prematurity, Intrauterine growth restriction, birth asphyxia and hypothermia. Signs include Lethargy, poor feeding, seizures, and apnea. Neurological damage may result from hypoglycemia in neonates.

Neonatal hypoglycemia is most commonly seen in macrosomic infants and infants of diabetic mothers. For these babies, during pregnancy, maternal glucose crosses the placenta to cause fetal hyperglycaemia. The fetal pancreas responds by increasing insulin production. Following delivery, the hyperglycaemic stimulus is instantly removed but insulin production may take

longer to slow down. This results in an increased risk and incidence of hypoglycemia at the early newborn period.

Management involves initially checking the airway, breathing and circulation. 2ml/kg of IV 10% Dextrose or 5ml/kg of 5% Dextrose may be given PR if IV access is unavailable. Ensure the baby has a normal body temperature (temperature target: 36.50 – 37.50C) as hypothermia prone the baby to hypoglycaemia. Always look for the underlying cause of the hypoglycemia and treat it appropriately.

7.6 Neonatal Sepsis

Sepsis in the neonate kills more than a million babies worldwide every year. It is a clinical syndrome characterized by signs of infection with accompanying bacteremia in the first month of life. It can be categorized into *Early Onset Neonatal Sepsis* (EOS), which refers to the presence of signs of infection accompanied by a positive culture within the first 72 hours of life, and *Late Onset Neonatal Sepsis* (LOS), which signifies the onset of signs of infection with a positive culture after 72 hours of life.

Causative organisms: Early-onset sepsis is typically caused by organisms from the maternal genital tract, whereas late-onset sepsis is caused by organisms in the caregiving environment or community. Common organisms are *Klebsiella pneumoniae,* E. coli, and Coagulase Negative Staphylococcus among others. For many of these organisms, the resistance rate to antibiotics is alarmingly going up.

Antenal risk factors known to be associated with neonatal jaundice include spontaneous rupture of membranes, less than 37 completed weeks of gestation, spontaneous preterm labour, rupture of membranes greater than 18 hours before delivery, maternal chorioamnionitis, maternal fever of 38 degrees Celsius or more, maternal invasive bacterial infection requiring antibiotics, pre-labour rupture of membranes, Group B Streptococcus infection in a previous baby, or current pregnancy, meconium-stained amniotic fluid and foul-smelling liquor.

Postnatally, risk factors for neonatal sepsis include prolonged resuscitation at birth, prematurity, invasive procedures, mechanical ventilation, excessive handling, home delivery, lack of hand washing and inadequate Infection prevention control. Others include overcrowding and prolonged hospital stay.

Signs of neonatal sepsis include

1. Abnormal colour: Pale, cyanotic, mottled appearance, jaundice, grey
2. Temperature instability
3. Abdominal signs: distension, poor feeding, vomiting, diarrhoea
4. Respiratory: Apnea, respiratory distress, gasping (Abnormal breathing)
5. Hypo - or hyperglycemia
6. Cardiovascular: Shock, tachycardia (HR $>$ 180), Bradycardia (HR $<$ 80)

7. Abnormal bleeding
8. Central Nervous System: Excessive crying, irritability, seizures, altered tone, lethargy,

Management of neonatal sepsis should be comprehensive. It should include an initial evaluation for resuscitation of the airway, breathing and circulation. Further, the blood sugar should be measured. Early reversal of the shock state by administering an initial bolus of 10ml/kg of crystalloid or its equivalent should be done in the shock present. Vasopressors or inotropes should be used in septic shock only after appropriate volume resuscitation has been done. The goals of the resuscitation should be Normal Cap refill (less than 2 seconds), normal pulses, warm extremities, and appropriate urine output (greater than 1mL/kg/hr).

Increased successful treatment of neonatal sepsis requires early recognition and urgent administration of appropriate antibiotics.

After resuscitation, ongoing management usually starts with a detailed history to assess risk factors and other presentations. A thorough physical examination will then be performed, looking for and documenting specific signs indicating severity.

Investigations usually include a blood culture. This is considered the gold standard for diagnosis. Ideally, a culture should always be done before the first dose of antibiotics. Other auxiliary investigations include a complete blood count, blood gases, urine culture, and a lumbar puncture. The threshold for performing a lumber puncture in all symptomatic newborns suspected of sepsis should be encouraged. It should however be deferred in neonates considered too unstable to tolerate the procedure, or where there is an absolute contraindication.

Supportive treatment is essential for a good outcome in neonatal sepsis. Antibiotics are not the entire solution to their treatment. Nutrition or breastfeeding should be optimized. The environment should be thermo-neutral and oxygen saturation should be maintained within the normal range (89 – 95%). Intravenous fluids should be used if the infant is hemodynamically unstable. Monitoring of the blood glucose levels should be instituted. Packed red cells and fresh frozen plasma should be used in the event of anaemia or bleeding.

7.7 Gastrointestinal emergencies

Gastrointestinal emergencies in newborns can be broadly divided into the following:

Obstructive: Some of these are Tracheoesophageal fistula, Duodenal Atresia, Hirschsprung's Disease, Biliary Atresia, Pyloric Stenosis, Intestinal Volvulus, Imperforate Anus and Necrotizing enterocolitis.

Abdominal wall defect: The most notable examples here are Omphalocele and Gastroschisis

Figure 7.1: Abdominal distension secondary to intestinal obstruction in a newborn

7.7.1 Gastroschisis

This is an anterior abdominal wall defect, located to the right of the umbilicus, and contains herniated intestines that have no material covering the sac. It occurs in approximately 1 in 10000 births. Rarely, it is associated with other genetic syndromes. However, it may be associated with intestinal atresia, stenosis and malrotation. Other associations include prematurity (50-60%)and cryptochidism (31%). Generally, it has a better prognosis compared to an omphalocele. The prognosis is excellent for small defects. Mortality is expertise and facility-dependent but generally around 5 to 10%. Necrotizing enterocolitis is a well-recognised complication, occurring in as much as 18%.

Figure 7.2: Gastroschisis in a newborn

7.7.2 Omphalocele

Figure 7.3: Omphalocele in a newborn

8 Neonatal Jaundice

8.1 Introduction

Jaundice is the yellowish discoloration of the skin, eyes and mucous membranes, caused by a pigment called bilirubin in the blood. Out of 10 term and 10 preterm newborns, 6 and 8 will develop jaundice respectively, all in the 1st couple of weeks of life. Universally accepted as one of the commonest causes of admission and readmission in the first month of life. At Komfo Anokye Teaching Hospital Mother Baby Unit, monthly admissions average between 300 and 400 and about 15 to 25% of all these admissions are cases of neonatal jaundice. Whereas the developed world describes kernicterus as a rare condition, unfortunately, the same cannot be said for us in developing countries. On average, cases of severe Neonatal Jaundice have ranged from 2.2% to 30.8% of all jaundice cases, with the monthly mortality ranging from 2.8% to 15.2%(REFERENCE). Remember, kernicterus is the only preventable cause of cerebral palsy!

8.2 Bilirubin metabolism

Humans continuously form bilirubin and the liver is the main organ responsible for the metabolism of bilirubin. For every gram of haemoglobin, 35mg of bilirubin is produced. The bilirubin is conjugated by the UGT enzyme, making it water-soluble, which is then released into the bile before being excreted in the stool (and urine). It can also be broken down in the intestine by bacterial enzymes like E. coli. However, at birth, the newborn has several challenges. The liver is immature, and the levels of bilirubin uridine diphosphate glucuronosyltransferase (bilirubin-UGT) enzyme are low. Newborns have -glucuronidase in the intestinal mucosa/brush border, which deconjugates the conjugated bilirubin found in the meconium. The unconjugated bilirubin can now be reabsorbed through the intestinal wall and recycled back into the circulation. This process is known as the “enterohepatic circulation of bilirubin”. The gut is sterile and, subsequently, infants have far fewer bacteria in the gut, and so very little, if any, bilirubin is reduced to urobilin and stercobilin.

Specifically in newborns, more bilirubin is produced, on account of the short life span of Red Blood Cells and high Hemoglobin levels. The liver is immature. They also have fewer bacteria and low intestinal enzymatic activity in the intestine

8.3 Types of bilirubin

There are two types:

8.3.1 Conjugated (Direct) Bilirubin

This is water soluble, excreted in the urine and stool, and not toxic to the brain. However, high amounts could indicate underlying liver disease or injury.

8.3.2 Unconjugated (Indirect) Bilirubin

This is lipid soluble, can cross the blood-brain barrier and is toxic in high amounts to the brain.

In very high concentrations, unconjugated bilirubin, which is lipid soluble, is toxic to the developing brain. Once it crosses the blood-brain barrier and binds to brain tissue and deposits in the developing brain. Since this is an irreversible process, it leads to long-term neurological issues and even death.

8.4 Types of Jaundice

There are two main types of jaundice:

1. Physiological jaundice and
2. Pathological jaundice.

There are three main mechanisms for jaundice:

1. Increased bilirubin production
2. Decreased bilirubin clearance and
3. Increased enterohepatic circulation.

8.4.1 Physiological jaundice

8.4.1.1 Increased bilirubin production

in term newborn infants, bilirubin production is 2 to 3 times higher than in adults. This occurs because newborns have more RBCs and fetal RBCs have a shorter life span than those in adults. Unfortunately, the liver being immature, cannot conjugate and excrete all the bilirubin from the breakdown of all the excess RBCs, thereby resulting in spillover of bilirubin into the blood.

8.4.1.2 Bilirubin clearance or excretion

This is decreased in newborns, mainly due to the low levels of the UGT enzyme in the liver. UGT activity in term infants at day 7 of age is approximately 1% of that of the adult liver and does not reach adult levels until about 14 weeks of age.

8.4.1.3 Enterohepatic circulation

The presence of the ß-glucuronidase results in an increase in the enterohepatic circulation of bilirubin, further increasing the bilirubin load in the infant. This is a diagnosis of exclusion

8.4.2 Pathological jaundice

8.4.2.1 Definition

Neonatal jaundice is said to be pathologic if:

- Jaundice in the 1st 24 - 48 hours of life.
- Rate of SB rise $>$ 0.5 mg/dL (8.5µmol/L) per hour
- Jaundice all over the body (including palms & soles)
- Presence of a danger sign
- History of previous siblings having had jaundice at birth
- Jaundice in a term newborn after 2 weeks of age or in a preterm infant after 3 weeks of age
- Direct (conjugated) bilirubin concentration $>$ 20% of the total

It can be caused by certain pathologic conditions or exaggeration of the mechanisms responsible for physiologic neonatal jaundice. Identification of what is causing the jaundice is useful in guiding management, including counselling of the parents and what to expect for the next pregnancy. Most common cause is increased bilirubin production due to haemolytic disease processes that include the following:

- Isoimmune-mediated haemolysis (e.g., ABO or Rhesus D incompatibility)
- Erythrocyte enzymatic defects, e.g. G6PD deficiency
- Sepsis, especially Urinary Tract Infection
- Polycythaemia
- Birth Injuries resulting in sequestration of blood within a closed space, e.g. cephalohematoma, subgaleal bleed.

8.4.2.2 ABO incompatibility

This is one of the most common causes of isoimmune hemolytic disease during the neonatal period. Infants with blood group A or B, carried by blood group O mother, will have a positive antibody because of maternal anti-A or anti-B transfer into the fetal circulation.

8.4.2.3 Rhesus Incompatibility

Rh incompatibility can occur when an Rh-negative pregnant mother is exposed to Rh-positive fetal red blood cells secondary to feto-maternal haemorrhage during pregnancy/delivery. As a result, the mother's blood gets exposed to the fetal circulation and sensitization occurs leading to maternal antibody production against the foreign Rh antigen. Once produced, maternal Rh (IgG) antibodies may cross freely from the placenta to the fetal circulation, where they form antigen-antibody complexes with Rh- positive fetal RBCs and eventually are destroyed, resulting in a fetal alloimmune-induced hemolytic anaemia and jaundice. The first pregnancy is usually not affected, but more antibodies are produced with each pregnancy making the jaundice worse with each pregnancy.

8.4.2.4 Decreased clearance

Inherited defects in the gene that encodes the UGT liver enzyme (eg, Gilbert Syndrome), decrease bilirubin conjugation (eg Crigglar Najjar). In physiological jaundice, the levels are naturally low, but here, in addition to the low levels the UGT enzyme is either defective, absent or has a reduced function. This reduces hepatic bilirubin metabolism and its clearance thereby increasing the total serum unconjugated bilirubin levels.

8.4.2.5 Increased enterohepatic circulation

The major causes are

- Breastfeeding jaundice
- Breast milk jaundice

- Impaired intestinal motility is caused by functional or anatomic obstruction.
- Congenital hypothyroidism also causes increased enterohepatic circulation on account of reduced gut motility.

8.5 Assessing for Neonatal Jaundice

- Baby should be assessed in natural daylight
- Look for yellow eyes & skin, check the white part of the eyes only if the baby opens the eyes voluntarily.
- You may blanch the skin on the bridge of the nose or the palms/soles of the feet if they turn yellow…
- Remember that the yellowing spreads from head to toe…
- Do not rely on visual inspection alone to estimate the bilirubin level in a baby with jaundice!!! It can be very subjective!!

8.6 Clinical features

The clinical features of neonatal jaundice may include:

- Baby looks yellow! The yellowness appears cephalocaudal.
- May not be as active as he/she used to be
- Lethargic/hypotonic
- Weak cry, irritable
- Poor feeding
- High-pitched cry / poor cry
- Seizures
- Arching of the neck/back

Thus to evaluate a child with jaundice we:

- Determine birth weight, gestation and postnatal age (in hours)
- Assess clinical condition (well or ill)
- Degree of jaundice (visual inspection, SBR etc)
- Look for evidence of kernicterus / BIND

8.7 Management

The general principle of treatment includes

- Encourage frequent exclusive breastfeeding.

- Start Intravenous fluids only when there are signs of dehydration
- Watch out for danger signs
- Pathologic Neonatal jaundice is treated with
  - Phototherapy
  - Exchange Blood Transfusion (EBT)
  - Antibiotics

Be interested in the cause as this will serve as a guide in the management of the baby and direct your counselling as well as impact on subsequent pregnancies Loads of information in the maternal and child health record book, Gravidity and Parity, G6PD status, maternal Blood group & Rhesus status etc

8.7.1 Investigations

This should include but not be restricted to

- Serum Bilirubin (conjugated, unconjugated and total)
- Full Blood Count
- G6PD screening
- Blood Culture & Sensitivity
- Baby's blood group (only necessary if mother's blood group is O)
- Others include Direct Coomb's test, Urine C & S etc

8.7.2 Phototherapy

Phototherapy is the use of visible light to treat high levels of serum bilirubin in the newborn.

Figure 8.1: Phototherapy Unit

The dose of phototherapy is a key factor in how quickly it works. The dose in turn is determined by:

- The wavelength of the light
- The intensity of the light (irradiance)
- The distance between the light and the baby
- The body's surface area is exposed to the light.

Effective phototherapy lowers serum bilirubin levels by converting the lipid-soluble bilirubin into water-soluble forms that can easily be excreted in the stool and urine Phototherapy also prevents the need for an Exchange Blood Transfusion and prevents bilirubin from depositing in the brain. The breakdown of bilirubin begins almost instantaneously when the skin is exposed to light, hence, phototherapy should be started as early as possible.

In initiating phototherapy, always note the time the baby's SBR sample is being taken and estimate the age in hours up until that time. Interpret bilirubin levels according to the baby's postnatal age in hours and manage the bilirubin levels according to the threshold table Start phototherapy if the SBR plots on or above the line appropriate for age (in hours) and gestational age If the SBR plots just underneath the line, repeat the SBR after 6 hours or start phototherapy if a repeat is not feasible. Repeat the SBR at least 24 to 48 hours after initiation of phototherapy. Discontinue phototherapy when the SBR plots below the line.

The side effects of phototherapy include:

- Increase insensible water loss
- Loose stools
- Skin rash
- Bronze baby syndrome
- Hypo- or Hyperthermia
- Interruption of mother-baby bonding

8.7.3 Sunlight Therapy

Works for physiological jaundice, however, one can never tell by looking at a baby what kind of jaundice a baby has Err on the side of caution, at least always have the SBR checked first Remember prolonged exposure to UV rays can be harmful to the developing skin Baby cannot be put in the light for more than 30 minutes in a day Even most of the available literature and studies that recommend sunlight still advice that if the jaundice is severe, the baby must be managed in the hospital!! A serum bilirubin high enough to warrant treatment should be managed in the hospital.

8.7.4 Exchange Blood Transfusion

Provides a means of rapid reduction of circulating bilirubin in the blood. Involves manual removal of the baby's blood and simultaneously replacing it with compatible donor blood.

In addition to reducing bilirubin levels, EBT removes partially hemolyzed RBCs, RBCs coated with antibodies and circulating immunoglobulins.

Complications of exchange blood transfusion include:

- Cardiac & respiratory disorders

Figure 8.2: Exchange Blood Transfusion

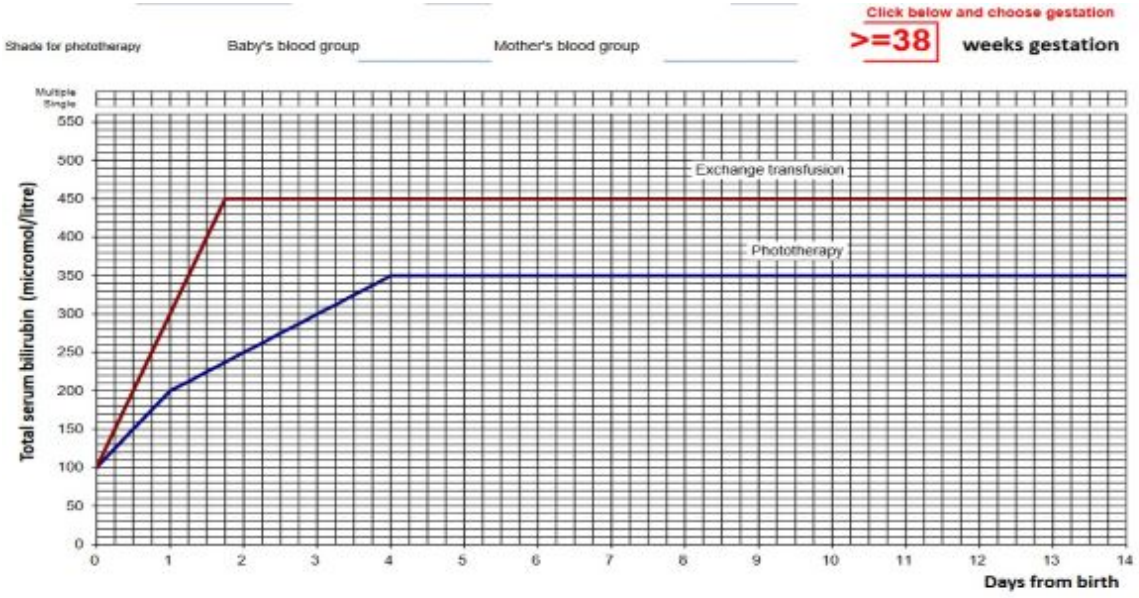

Figure 8.3: Bilirubin Graph (> 38 weeks)

- Shock due to bleeding or inadequate replacement of blood infection
- Catheter-related complications
- Changes in the composition of the blood (high or low potassium, low calcium, low glucose, changes in pH)
- Thrombocytopenia
- And the rare but serious complications of air embolism, portal hypertension, and necrotizing enterocolitis.

8.7.5 Intravenous Immunoglobins

Treatment with intravenous immunoglobulin (IVIG) has been suggested as an alternative therapy to Exchange Blood Transfusion for isoimmune hemolytic jaundice to reduce the need for Exchange Blood Transfusion and duration of phototherapy and hospitalization in isoimmune hemolytic disease of the newborn. It has been proposed that IVIG blocks the binding of the antibody to the antigen. With this blockade, hemolysis no longer occurs.

8.8 Long term complications

The effects of bilirubin toxicity include

- Hearing loss
- Cerebral palsy
- Mental retardation
- Dental complications
- Delayed developmental milestones
- Seizure and visual disorders

8.9 Recommendations

- Always err on the side of caution
- An SBR is always more objective
- Look out for danger signs
- As much breastmilk as possible by any means necessary
- Sunlight therapy is not recommended, if the baby is yellow enough for you to want to put him/her under the sun, then the baby needs to be brought to the hospital!

9 Newborn Delivery and Resusctation

10 Preterm and Low Birth Weight

11 Newborn Feeding

Part II

Pulmonology

12 Respiratory Disorders I

13 Respiratory Disorders II

Part III

Cardiology

14 Anatomy, Physiology & Pathology

14.1 Anatomy

The heart is located in the mediastinum of the chest, bounded anteriorly by the sternum, posteriorly by the spine and laterally by the lungs. Externally, the right ventricle is anterior. Most of the left ventricle, left atrium and right atrium are posterior. Internally, the right and left atria are separated by the tricuspid and mitral valves from the right and left ventricles respectively. The arterial supply of the heart is through the coronary arteries while venous drainage is through the coronary sinus. The aorta and pulmonary arteries arise from the left and right ventricles. The heart has three layers:

1. Endocardium: Inner epithelial layer of the heart
2. Myocardium: Muscular part of the heart
3. Pericardium: Outer layers of the heart. Divided into the visceral and parietal pericardium.

Venous blood enters the right atrium through the inferior and superior vena cavae. It empties in atrial systole into the right ventricle through the tricuspid valve. It then moves on through the pulmonary valve in ventricular systole, to the pulmonary artery and then the lungs. Blood returning from the lungs enters the left atrium through the four pulmonary veins. In atrial systole, it moves into the left ventricle through the mitral valve. Finally, it empties into the aorta through the aortic valve.

14.2 Conduction system

The heart has an inherent electrical system that automatically paces and conducts depolarization throughout it. The parts are:

1. *Sinoatrial (SA) node*: This is the pacemaker of the heart and depolarizes the two atria.
2. *Atrioventricular (AV) node*: Receives impulses from the SA node, and delays a bit before propagating it further.
3. *His-purkinje fibre* system: Responsible for the spread of electrical impulses to the ventricles

14.3 Heart as a pump

There is a difference in the pumping action of the heart in utero and after birth.

1. Fetal
    - Most work is done by the right ventricle
    - The right Ventricle is therefore relatively hypertrophic
    - Only 15% of the cardiac output is pumped into the lungs
2. After birth
    - Gradual transition to Left ventricle dominance
    - Gradual fall in pulmonary pressure (over 6 weeks)
    - The left ventricle does most of the work and becomes more hypertrophic than the right

14.4 Systolic and diastolic functions

Systole: This is the contractile phase of the heart. It starts after the atria is filled with blood. The atria then contract, emptying its content into the ventricles. At this stage, the ventricle also undergoes systole, which further empties the blood into the aorta and pulmonary arteries.

Diastole: This is the relaxation phase where the heart relaxes and lets in blood. It also starts with the atrium and then the ventricles.

Compliance: This describes how easily the heart chamber relaxes in response to the inflow of blood.

14.5 Intracardiac Pressures

The pressures in the heart vary for different ages and individuals. Generally, the pressures in the atria are lower than the ventricles. Also, the peak systolic pressure in the left ventricle is higher than in the right. The diastolic pressure in the left ventricle is however lower than the right ventricle. Also, both systolic and diastolic pressures in the aorta are higher than that in the pulmonary artery.

Systolic pressure in general is generated by the ventricles. In conditions such as coarctation of the aorta, aortic stenosis and pulmonary hypertension, the ventricles increase their workload to generate enough pressure. The diastolic pressure on the other hand is maintained by the closure of the aortic and pulmonary valves. Thus, an incompetent pulmonary or aortic valve leads to a decrease in diastolic pressure in the two vessels respectively.

14.6 Fetal circulation

The heart begins developing in the fetus as the cardiogenic area and primitive blood vessels as early as 18 days old. By 20 days the paired endocardial tubes are formed. By day 35 the heart would have completed its looping with blood already flowing through.

Three main shunts exist in the fetal circulation. These are adaptations to using the placenta rather than the lungs for oxygenation. These shunts are the ductus venosus, ductus arteriosus and patent foramen. At birth, these shunts regress. The ductus venosus regresses quickly to form the ligamentum arteriosus. The foramen ovale closes at birth physiologically but may take years to close anatomically. The patent ductus arteriosus closes soon after birth but can physiologically stay open for 72 hours. Beyond this period it is considered to be pathologic if it continues to stay open. Patent ductus arteriosus are quite common in preterm newborns.

14.7 Pathologic classification

The pathology of pediatric cardiac disorders varies. Broadly, they can be divided into these:

1. ***Congenital heart disorders***: These are cardiac conditions that a child is born with. Thus they are present at birth. They form about 85% of all pediatric heart diseases seen in the Komfo Anokye Teaching Hospital (KATH). It is further divided into:
    - *Acyanotic*: These are congenital heart diseases that are traditionally not known to be associated with cyanosis. Examples include ventricular septal defect (VSD), Atrial Septal Defect (ASD) and Patent Ductus Arteriosus (PDA)
    - *Cyanotic*: These on the other hand are associated with cyanosis and include Tetralogy of Fallot (ToF), Truncus Arteriosus and Tricuspid Atresia.
2. ***Acquired heart disorders***: These are heart conditions that are not present at birth but develop afterwards. They include Infective Endocarditis (IE), Rheumatic Heart Disease (RHD) and Endomyocardial Fibrosis (EMF)
3. ***Rhythm disorders***: This set of disorders can present as either congenital or acquired. They affect the electrical system of the heart leading to an increase in heart rate (tachyarrhythmia), decrease in heart rate (bradyarrhythmia) or even normal heart rate.
4. ***Secondary cardiac disorders***: Some pathologies tend to affect the heart as a complication. Such conditions include some glycogen storage disorders resulting in cardiomyopathy and Rheumatoid arthritis resulting in pericardial effusion.

15 Evaluating Heart Diseases

To fully evaluate a child with a suspected cardiac condition, one needs to go through the regular steps applicable in medicine. These are outlined below:

15.1 History

The history is traditionally divided into:

15.1.1 Prenatal

Prenatally, the history should delve into but not be limited to the following:

1. ***Infections***: Some infections are the well-known TORCHES. They include Toxoplasmosis, HIV, syphilis, parvovirus B19 (fifth disease), varicella (chickenpox) and (Zika), Rubella, Cytomegalovirus, and Herpes simplex virus. Rubella when acquired in the first trimester of pregnancy is very well known to be associated with PDAs.
2. ***Medications:*** The use of some medications, including herbs predisposes to heart disease in newborns. Anticonvulsant such as phenytoin, carbamazepine, and valproic acid are highly teratogenic.(Kalisch-Smith, Ved, and Sparrow 2019)
3. ***Recreational drugs***: Excessive smoking, cocaine, and alcohol use in early pregnancy are all associated with teratogenic effects on the heart.
4. ***Maternal illnesses:*** Maternal medical conditions during pregnancy may be associated with heart diseases in their fetuses. Diabetes mellitus is particularly well known, predisposing to hypertrophic cardiomyopathy, d-TGA, etc. Autoimmune conditions such as Systemic Lupus Erythematosus may also predispose to rhythm disturbances in the fetus and child, even when the mother is not symptomatic.
5. ***Family history of CHD***: The recurrence of CHD in first-degree relatives varies but is almost always higher than the rest of the population. For instance, having a first-degree relation with a cornoventricular defect was associated with a recurrence risk ratio of 24.3 (95% CI,12.2 to 48.7), 7.1 (95% CI, 4.5 to 11.1) for isolated ASD, and 3.4 (95% CI, 2.2 to 5.3) for isolated VSD.(Øyen et al. 2009)

15.1.2 Perinatal

Perinatal history associated with heart disease may include the following:

1. ***Birth weight***: A high birth weight, often associated with a child of a diabetic mother is also associated with an increased incidence of CHDs. Conversely, a low birth weight may also be associated with fetal alcohol syndrome or congenital rubella syndrome, both of which are associated with CHDs.
2. ***Newborn resuscitation***: Some critical CHDs can be similar to neonatal asphyxia in a newborn, thus requiring resuscitation.

15.1.3 After birth

Ascertaining history after birth is the most extensive. Many of these are directed to the features of heart failure. These include:

1. ***Growth failure***: Poor weight gain is a very prominent feature of CHDs in children. Many clinically significant CHDs result in poor feeding, chronic metabolic demand on the patient and poor oxygenation in cyanotic CHDs. All these results in increased caloric demand, resulting in poor growth.
2. ***Cyanotic spells***: Some cyanotic CHDs are associated with recurrent periods where the child has increasing cyanosis, sometimes associated with weakness, fast breathing and even unconsciousness. The presence of these spells may be pointed to a CHD.
3. ***Squatting and exercise intolerance***: A common presentation of heart diseases in children is exercise intolerance. However, for some cyanotic congenital heart diseases, most notably ToF, the added feature is frequent squatting when the child becomes fatigued.
4. ***Delayed milestones***: Growth failure, easy fatiguability and the presence of other genetic syndromes may result in delayed motor milestones in the child.
5. ***Others***: *Fast* and sometimes *difficulty breathing* are also common presentations of CHDs. Some children develop *oedema*. This is predominantly seen in the faces of younger children and the feet of older children. Frequent *lower respiratory infection* is also seen in children with heart diseases, especially those associated with heart failure.
6. ***Uncommon symptoms***: Uncommon presentation of heart disease in children include:
    - *Chest pain* is a rather feared symptom in adults but usually portends another diagnosis rather than heart disease in children.
    - *Syncope* can be observed in children with an arrhythmia, or left or right ventricular obstruction. However, this is still not a common presentation in pediatric heart diseases.
    - Older children report *palpitations*.
    - *Joint swelling* does occur in Rheumatic Heart Disease but again not a common presentation in children with a heart pathology.

15.2 Clinical examination

Clinical examination for a child with a suspected heart disease should always start as a general. One should first look out for life-threatening signs and intervene quickly. Subsequent steps could include:

1. ***Nutritional status*** is very important as many children with chronic heart conditions with significant heart failure present with malnutrition. The growth pattern of the patient should always be evaluated.
2. ***Dysmorphism*** is very critical in pediatric heart diseases. As much as 23% of all children with CHD will have a chromosomal abnormality.(Wang et al. 2023) There are many genetic syndromes with well-documented recognisable heart defects. Below are just a few adapted from Ko (2015):

   Table 15.1: Common genetic syndromes associated with congenital heart diseases

Genetic syndrome	% with CHD	Cardiac anomalies
Down Syndrome	40 to 50	Atrial Septal Defect, Ventricular Septal Defect, Atrioventricular Canal Defect, Patent Ductus Arteriosus, Tetralogy of Fallot
Turner syndrome	25 to 45	Coarctation of the Aorta. Bicuspid Aortic Valve, Aortic Stenosis, Hypoplastic left heart syndrome
DiGeorge syndrome	70 to 75	Aortic arch anomalies, Truncus arteriosus, Tetralogy of Fallot
Williams syndrome	75 to 80	Supravalvar Aortic Stenosis, Peripheral Pulmonary Stenosis
Noonan syndrome	70 to 80	Pulmonary Stenosis, Hypertrophic Cardiomyopathy, Atrial Septal Defect
Kabuki syndrome	31 to 55	Coarctation of the Aorta, Atrial Septal Defect, Aortic Stenosis, Mitral Stenosis, Hypoplastic left heart syndrome
Alagille syndrome	90	Peripheral Pulmonary Stenosis, Pulmonary Stenosis, Tetralogy of Fallot

3. ***Colour***: The skin colour of a child with a CHD could hold signs of its presence. *Cyanosis*, the blueish duskiness of the skin and mucous membranes can be seen in children with a cyanotic congenital heart disease. In black skin, this may be difficult and can only be observed in the mouth and tongue (Figure 15.1). Mild cyanosis is often not visible and may require the use of pulse oximetry. *Pallor* can be observed in patients with CHDs. or in other AHDs such as infective endocarditis. *Jaundice* can be observed in patients with Infective Endocarditis or those with hepatic injury secondary to chronic heart failure.

Figure 15.1: Cyanosis in the tongue of a child

4. ***Clubbing***: All four stages of digital clubbing are seen in children with cyanotic CHD or Infective endocarditis. (Figure 15.2) Note that some cases of finger clubbing may be familial.

5. ***Respiratory signs***: Respiratory signs commonly associated with heart diseases in children are tachypnoea, dyspnoea, chest recessions and increased work of breathing. These are especially true when there is associated heart failure and worsens with exercise or breastfeeding the the breastfeeding infant.

15.3 Investigation

Chest x-ray showing a globular heart

Figure 15.2: Finger clubbing

Figure 15.3: Chest x-ray showing cardiomegaly and lung shadowing

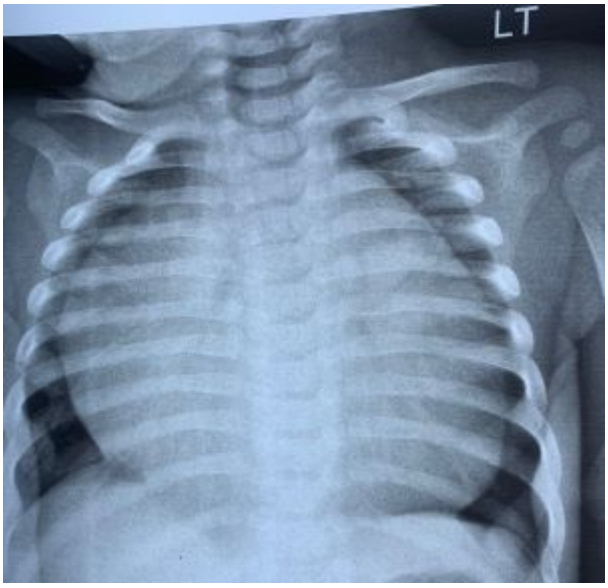

Figure 15.4: Chest x-ray showing a globular heart

Figure 15.5: Chest bulge and Harrison's sulcus in child

16 Heart Failure

16.1 Definition

The inability of the heat to provide enough output to the body.

16.2 Causes

Varies especially in children. They can occur in both structurally normal hearts and in congenital cardiac malformations. There are 3 main groups of causes:

1. Ventricular dysfunction: This is where there is a dysfunction of the ventricles. It is usually a systolic dysfunction but may also be diastolic. Examples are:

    - Cardiomyopathy (dilated, restrictive and hypertrophic)
    - Myocarditis
    - Arrhythmias
    - Coronary artery anomalies
    - Post-op cardiac dysfunction

2. Volume overload: This occurs in conditions associated with increased volume (preload) in the heart especially the ventricles. The ventricle must therefore eject an increased blood volume, leading to tachycardia. It may or may not be associated with ventricular dysfunction. Examples include:

    - Ventricular septal defect (left to right shunt)
    - Atrial septal defect
    - Patent ductus arteriosus
    - Aortic regurgitation (left ventricle)
    - Mitral regurgitation (Left atrium)

3. Pressure overload: This is when heart failure is caused by an increased pressure (afterload) in the heart. Ventricles must therefore contract against higher pressures. It may or may not be associated with ventricular dysfunction. These include:

    - Hypertension

- Aortic valve stenosis
- Pulmonary stenosis
- Coarctation of the aorta

In all these, the result is decreased cardiac output and pulmonary oedema.

16.3 Classification

The symptoms of heart failure vary significantly with infants and young children having different presentations compared to older children. The classification of heart failure there is not uniform. The most well-known classification is the NYHA classification which is appropriate for older children. It is shown below:

Table 16.1: NYHA Classification

Class	Patient Symptoms
Class I (Mild)	No limitation on physical activity. Ordinary physical activity does not cause undue fatigue, palpitation or dyspnoea
Class II (Mild)	Slight limitation of physical activity. Comfortable at rest but ordinary physical activity results in fatigue, palpitation or dyspnoea
Class III (Moderate)	Marked limitation of physical activity. Comfortable at rest but less than ordinary physical activity causes fatigue, palpitation or dyspnoea
Class IV (Severe)	Unable to carry out any physical activity without discomfort. Symptoms of cardiac insufficiency at rest. If any physical activity is undertaken discomfort is increased

On the other hand, the Ross classification shown below is more suited for infants and young children.

Table 16.2: Modified Ross Classification

Class	Symptoms
Class I	Asymptomatic
Class II	Mild tachypnoea or diaphoresis in feeding in infants Dyspnoea on exertion in older children
Class III	Marked tachypnoea or sweating with feeding in infants Marked dyspnoea on exertion Prolonged feeding times with growth failure
Class IV	Symptoms such as tachycardia, retraction, grunting, or diaphoresis at rest

16.4 Pathophysiology

A schematic drawing of the various processes involved is shown below:

16.5 Signs and symptoms

The symptoms of heart failure are variable and age-dependent. For infants, the symptoms include poor feeding, sweating with breastfeeding, prolonged feeding time, tachypnoea, poor weight gain and dyspnoea. For young children symptoms include recurrent respiratory tract infection, recurrent wheezing, fatigue, exercise intolerance, facial and recurrent cough. Older children have symptoms that more resemble those of adults. These include tachypnoea, tachycardia, recurrent wheezing, pedal swelling, palpitations, and vomiting.

Signs of heart failure also vary with age. These include for infants, failure to thrive, tachycardia, tachypnoea, hepatomegaly, displaced apex (cardiomegaly), S3 gallop, oedema (pedal in older children and facial or abdominal distension in older children).

16.6 Investigation

The investigations required are generally towards the likely underlying pathology. Some of them would include:

Chest x-ray: This may show cardiomegaly, increased pulmonary lung markings, pulmonary oedema, pleural effusion and heart shape.

Electrocardiogram: This helps to identify chamber enlargement and dysrhythmias that may be the cause or consequent to the heart failure

Echocardiogram: This identifies and quantifies the function of the ventricle as well as the chamber sizes

Blood test: The complete blood count helps to identify anaemia or polycethemia. The serum urea and creatinine identify possible renal dysfunction. Other tests include BNP (Brain Naturetic Peptide) and Troponin both of which are elevated in heart failure.

Other investigatory modalities include Magnetic Resonance Imaging, Cardiac catheterization,

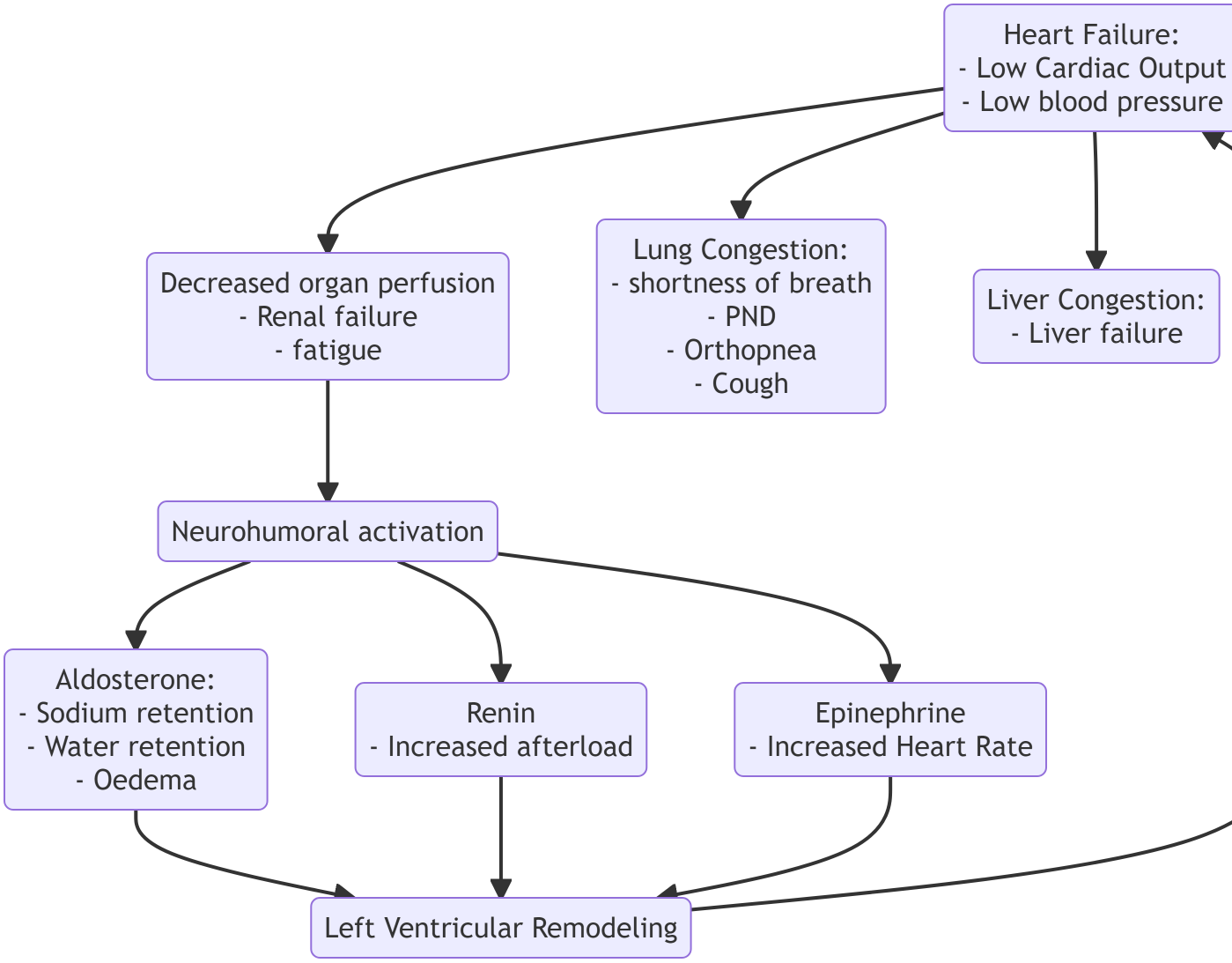

Figure 16.1: Pathophysiology of heart failure

16.7 Treatment of Heart Failure

This is done with some goals:

1. Improve the quality of life
2. Arrest and possibly reverse the heart failure
3. Sustain till other definitive therapeutic interventions are employed, including surgery.

The treatment for heart failure is dependent on the pathophysiology, clinical features and stage of the disease.

16.7.1 Non-pharmacological treatment

This includes Fluid restriction (in case of congestion) and fluid overload, intubation and/or mechanical ventilation to help support breathing and reduce the workload on the heart and patient. Heart transplantation is the last option in some cases of heart failure.

16.7.2 Pharmacological treatment

Treatment depends on the clinical presentation and cause of the heart failure. There are 2 main groups to be considered:

16.7.3 Acute decompensated heart failure

Table 16.3: Drugs used in acute decompensated heart failure

Drug	Action
Diuretics	Notable here is furosemide. The aim is to help decongest the lungs, reduce preload by vasodilatation and improve heart failure symptoms.
Inotropes	These include adrenaline, noradrenaline, dopamine and dobutamine. They help improve the contractility of the heart, increase heart rate, and increase peripheral vascular resistance, thus maintaining the blood pressure and cardiac output. They are usually Intravenous medications.

16.7.4 Chronic heart failure

These are usually oral medications given to treat heart failure on an outpatient basis

Table 16.4: Drugs for chronic heart failure treatment

Group	Action
Diuretics	These are given to decongest the lungs, liver and other edematous organs. The most commonly used is furosemide.
Aldosterone antagonists	These counteract the aldosterone effect of water and sodium retention. They decrease afterload while helping in reversing cardiac remodelling.
ACE-I/ARB	Angiotensin-converting enzyme inhibitors and Angiotensin II receptor blockers counteract the renin effects of increasing afterload. They thus decrease the afterload and help reverse and prevent cardiac remodelling
Digoxin	This is probably the oldest anti-heart failure medication. It has negative chronotropic and positive inotropic effects. Thus increasing contractility and reducing heart rate.

16.8 Complications

Complications of heart failure include renal failure, hepatic failure, pulmonary hypertension, arrhythmia, and thromboembolic effects.

17 Atrial Septal Defect

17.1

17.2 Introducion

1. Defect in the inter-atrial septum
2. 5-10% of all CHD
3. Types
    - Secundum ASD (most common, 50-70%)
    - Primum ASD (30%)
    - Sinus venosus ASD
    - Coronary sinus ASD

17.3 Pathophysiology

- Left to right shunting and thus acyanotic
- leads to volume overload of the right atrium, ventricle, pulmonary artery and pulmonary oedema
- Consequent dilatation of the right atrium and ventricles
- Minimal pressure transmitted so no significant pressure overload
- Consequently, pulmonary oedema is usually insignificant
- Rarely have overt heart failure
- However, long-standing liaison or a very big lesion with a pulmonary-to-systemic flow ratio of 2 or more will lead to heart failure and pulmonary hypertension after about 15 to 20 years No Reversal of shunt

17.4 Clinical presentatoin

- Usually asymptomatic except for big lesion with high Qp: Qs
- They often have slender bodies

- Auscultation reveals a widely fixed split-second heart sound and a grade 2/6 to 3/6 ejection systolic murmur at the upper sternal border
- Many are almost silent, especially the small lesions which are often detected during an echocardiogram for another reason

17.5 Investigations

- Bedside SpO2 is usually normal and hence an acyanotic heart disease
- In older patients, a chest x-ray may show
  - Cardiomegaly
  - Prominent pulmonary artery
  - Increased vascular markings
- The electrocardiogram may show
  - Right axis deviation due to the right ventricular dilatation
  - Right atrial enlargement
- An echocardiogram is diagnostic as it visualises the defect, and quantifies the shunt and other chamber sizes.
- Cardiac catheterization is often done in long-standing cases to detect complications that may have arisen.

17.6 Natural history

- Most ASDs will close spontaneously by 4 years, with smaller ones having a higher closure rate than bigger ones. A long-standing large defect however leads to chronic heart failure and pulmonary hypertension in early adulthood.
- Arrhythmias may arise because of the dilated right atrium.
- Though there are reported cases of paradoxical strokes in patients with ASDs, it remains an uncommon occurrence.
- Infective endocarditis is also rare in ASDs.

17.7 Treatment

There is no need for exercise restriction or prophylaxis for endocarditis. If there is no sign of heart failure, a device closure is often done after infancy or a surgical closure at 2-4 years of age. However, if there is heart failure, Medical treatment for heart failure is immediately instituted. Then a planned device closure or surgical closure can be done within the first year of life.

17.8 Prognosis

Prognosis is generally good with many living into adulthood even without corrective surgery. Post-surgical mortality i currently less the 0.5%. The patient will need very little long-term follow-up after the corrective surgery.

18 Ventricular Septal Defect

18.1 Introduction

This is the most common Congenital Heart Disease (CHD), being seen in about 15-20% of all CHDs. VSD occurs in different anatomical locations. The most common is the perimembranous. Others are inlet, outlet, muscular and infundibular. They also occur in different shapes and sizes as well.

18.2 Pathophysiology

Typically VSD without the presence of another congenital heart malformation results in a left to right ventricle shunting lesion because of the pressure difference. Thus, a VSD is usually an acyanotic congenital heart lesion. This leads to a volume overload of the pulmonary artery, lungs, left atrium and left ventricle. These chambers subsequently dilated. Pulmonary edema develops from lung congestion leading to signs of heart failure.

Secondly, depending on the size of the defect the pressure in the left ventricle will get transmitted to the right. How much pressure is transmitted depends on the size of the defect with bigger defects transmitting more than smaller ones. This can result in increased right ventricular pressure, and hypertrophy. It also worsens the pulmonary oedema already mentioned above.

Persistent pressure and volume overload cause remodelling of the pulmonary vasculature, resulting in permanent changes and pulmonary hypertension. When the pulmonary pressure rises significantly higher than the systemic pressure, a reversal of the shunt results, leading to decreased oxygen saturation.

18.3 Clinical presentation

The clinical presentation of VSDs is variable and depends on the size and position.

18.3.1 Position

A perimembranous VSD of comparable size may exhibit more signs of heart failure compared to one that is mid-muscular or apical.

18.3.2 Size

Generally, the sizes of VSDs determine the extent of the volume and pressure overload of the right heart and lungs. Larger ones result in relatively higher pressure and volume.

Small

Small VSDs are usually asymptomatic with no volume or pressure overload of the lung, pulmonary artery and right heart.

Moderate

Some volume and pressure overload usually accompanies moderate-sized VSDs. They are thus often accompanied by some heart failure and recurrent lower respiratory infections.

Large

Large defects are accompanied by severe volume and pressure overload. They present with persistent heart failure and failure to thrive, exercise intolerance. when longstanding, they often end up with significant pulmonary hypertension.

18.4 Investigations

18.5 Management

18.6 Prognosis

19 Patent Ductus Arteriosus

20 Coarctation of the Aorta

21 Tetralogy of Fallot

21.1 Definition

21.2 Incidence/prevalence

21.3 Aetiology

21.4 Pathogenesis

21.5 Signs and symptoms

21.6 Investigations

21.7 Treatment

21.8 Complications

21.9 Prognosis

21.10 Differential diagnosis

21.11 Prevention

Figure 21.1: Boot shaped heart of Tetralogy of Fallot

22 Rheumatic Heart Disease

23 Infective Endocarditis

24 Endomyocardial Fibrosis

Figure 24.1: Endomyocardial Fibrosis showing classical Egg-on-Stick appearance

25 Miscellaneous Conditions

Part IV

Infectious Diseases

26 Immunodeficiencies

27 HIV

28 Bacterial Sepsis & UTI

29 Tuberculosis

30 Immunization

31 Viral Infections

Part V

Oncology

32 General Principles

33 Oncological Emergencies

34 Leukemia

35 Lymphoma

36 Retinoblastoma

37 Wilm's Tumor

Part VI

Nephrology

38 Hypertension

38.1 The Concept of Blood Pressure

Blood pressure is the force exerted by the blood against any unit area of the vessel wall. Physiologically,

$$BP = CO \times TPR = SV \times HR \times TPR$$

Where:

- HR is the Heart Rate
- BP is the Blood Pressure
- TPR is the Total Peripheral Resistance
- CO is the Cardiac Output
- SV is the stroke volume

38.2 Ways of measuring blood pressure

1. **Direct intra-arterial** measurements by placing a catheter into the vessel and measuring the pressure "in line" with the vessel (end-on-pressure). This method is used by physiologists and Intensivists. The principle is employed in the measurements of central venous pressure and intracranial pressure in clinical practice.
2. **The auscultatory method** is done with the use of a sphygmomanometer (either mercury or aneroid) and a stethoscope. This is the gold standard in clinical practice. Korotkoff sounds 1 and 5 sounds are measured for systolic and diastolic bleed pressures respectively. Values obtained are generally lower than direct & oscillometric measurements.
3. **The palpation method** (flush technique) is performed with the use of a sphygmomanometer and palpating finger. Largely unreliable. Only systolic blood pressure can be measured with this technique. The palpated pulse is generally lower than Korotkoff sound 1 by 10mmHg.
4. **The oscillometric method** uses a sphygmomanometer and a monitor e.g. digital blood pressure devices and Dynamap. Here, pulsatile blood flow through arterial wall oscillations is transmitted to the cuff encircling the extremity. Korotkoff sound 1 is recorded at the point of rapid increase in oscillation amplitude. Korotkoff sound 5 is recorded as

the point of a sudden decrease in oscillation amplitude. Values obtained by oscillometric measurements are generally higher than auscultatory.

5. **Doppler ultrasound technique**: Here a Doppler ultrasound is held over the pulse to magnify the sound so that it is audible without a stethoscope. The sound detected may be 5mmHg higher than Korotkoff sound 1.
6. **Ambulatory blood pressure measurements**. Here, multiple measurements are recorded over time (e.g. 24 hours) with digital devices attached to the limb whilst the patient engages in normal activities outside the hospital. Results are analysed on a computer or paper tracer built into the device using the mean of the readings. It provides a truer picture of blood pressure trends useful in diagnosing "white coat hypertension" and nocturnal hypertension (absence of a normal physiological drop in blood pressure during sleep).

38.3 Definition of Hypertension in children

In adults, the epidemiological definition is based on the risk of adverse events (e.g. Stroke) being>140/90mmHg. **In children**, hypertension is defined statistically based on normative data: 95th centile for age, height, and gender (Refer to height centile chart and blood pressure levels). By this statistical definition, 5% of children will be classified as hypertensives. Other definitions include:

- **Normal blood pressure**: < 90th centile for age, height, and sex.
- **Pre-Hypertension**: 90th – <95th centile for age, height, and sex
- **Stage 1 Hypertension**: 95th - 99th + 5 mmHg
- **Stage 2 Hypertension**: *> 99th centile + 5mmHg*

A sample of the blood pressure chart is shown below.

Blood Pressure Levels for Boys by Age and Height Percentile (Continued)

Age (Year)	BP Percentile ↓	Systolic BP (mmHg) ← Percentile of Height →							Diastolic BP (mmHg) ← Percentile of Height →						
		5th	10th	25th	50th	75th	90th	95th	5th	10th	25th	50th	75th	90th	95th
11	50th	99	100	102	104	105	107	107	59	59	60	61	62	63	63
	90th	113	114	115	117	119	120	121	74	74	75	76	77	78	78
	95th	117	118	119	121	123	124	125	78	78	79	80	81	82	82
	99th	124	125	127	129	130	132	132	86	86	87	88	89	90	90
12	50th	101	102	104	106	108	109	110	59	60	61	62	63	63	64
	90th	115	116	118	120	121	123	123	74	75	75	76	77	78	79
	95th	119	120	122	123	125	127	127	78	79	80	81	82	82	83
	99th	126	127	129	131	133	134	135	86	87	88	89	90	90	91
13	50th	104	105	106	108	110	111	112	60	60	61	62	63	64	64
	90th	117	118	120	122	124	125	126	75	75	76	77	78	79	79
	95th	121	122	124	126	128	129	130	79	79	80	81	82	83	83
	99th	128	130	131	133	135	136	137	87	87	88	89	90	91	91

Figure 38.1: Blood Pressure Centile Chart

38.4 Plotting the blood pressure centile

1. Measure the child's height
2. Determine the height centile. If the height centile falls between 2 centiles, use the closest centile. Otherwise, use the lower height centile.
3. Determine the blood pressure centile.
4. Classify blood pressure using the definitions above.

38.5 Hypertensive emergency

This is an acutely elevated blood pressure with evidence of threatening end-organ damage involving the following organs:

- Brain (severe headache, visual changes, cranial nerve palsy, papilloedema)
- Heart (acute chest pain and tightness, shortness of breath)
- Kidney (decreased urine output acutely, proteinuria and haematuria on dipstick)

It is thus a symptomatic, severe Hypertension.

38.6 Hypertensive Urgency

This is severe hypertension without evidence of end-organ damage or symptoms. The blood pressure should nevertheless be treated urgently but not aggressively like in a hypertensive emergency to prevent progression into a hypertensive emergency. If possible, the patient should be managed as in-patient.

38.7 Rules of blood pressure measurement

1. Select the right cuff size
    - The length of the inflation bladder should be at least 80% of the mid-arm circumference.
    - The width of the inflation bladder is at least 40th of the mid-arm circumference.
2. The child should rest for at least 5 minutes in a comfortable environment and position.
3. Arm resting and supported at heart level (The reference level. Values outside this reference level are higher). The lower edge of the cuff is 2cm above the cubital fossa.
4. Bladder tubings should lie over the brachial artery.
5. Bell of the stethoscope is used
6. Korotkoff sounds 1 and 5 are used for systolic and diastolic respectively.

7. Multiple measurements are made (preferably at different settings) and the lowest reading is taken. For research purposes, 3 measurements are taken and an average of the last 2 used.

Blood pressure readings obtained in the legs are 10-20mmHg higher than the arm pressure in any individual. Arm blood pressure higher than leg blood pressure occurs in aortic coarctation distal to ductus arteriosus.

38.8 When to suspect hypertension

Suspect hypertension in any child with any of the following conditions:

- Alteration in consciousness including aggressive behavior and convulsion
- Oedematous
- Known kidney disease or evidence of abnormal urinalysis
- Heart failure
- Obesity
- Failure to thrive
- Stroke or other palsies including cranial nerve palsy
- History of Low Birth Weight (small number of nephrons)
- Unexplained anaemia, or blurred vision
- Neurofibromatosis
- Other syndromes like Turner & Williams

38.9 Aetiology of hypertension

Generally, childhood Hypertension is considered to be of secondary cause until proven otherwise. This is particularly so among the very young and the severely hypertensive. The majority (~80%) are of renal origin. However, the number of children with essential Hypertension is on the rise, particularly among obese adolescents and those with a positive family history.

Broadly, aetiology can be categorized into:

- Renal disease
- Vascular disorders
- Endocrine causes
- Neurologic causes
- Renal tumours
- Catecholamine-secreting tumours
- Drug-induced
- Miscellaneous causes

However, since these are often age-specific categorizations are done by age as below:

38.9.1 Neonate to one-year

Congenital

- Congenital lesions of the vasculature
  - Renal Artery Stenosis
  - Aortic coarctation
- Congenital lesions of renal parenchyma
  - Polycystic Kidney disease
  - Dysplastic kidneys
  - Obstructive uropathy
- Congenital Adrenal Hyperplasia
  - 11- hydroxylase deficiency
  - 17- hydroxylase def

Acquired

- Renal artery or vein thrombosis secondary to umbilical artery or vein catheterisation
- Bronchopulmonary dysplasia
- Medications
  - Theophylline/caffeine
  - Phenylephrine and Ephedrine Nasal Drops in cold medications
  - Steroids
  - Vitamin D intoxication
- Total Parental Nutrition (high Ca2+)
- Maternal drug use: Cocaine, heroin

38.9.2 One- to five years

- Renal Artery Stenosis
- Glomerulonephritis
- Renal vein thrombosis
- Wilms tumour
- Neuroblastoma
- Phaeochromocytoma
- Cystic kidney disease
- Monogenic Hypertension (e.g. Liddle's syndrome)

38.9.3 Five- to ten-years

- Glomerulonephritis
- Renal scars from reflux nephropathies or Urinary Tract Infections
- Renal Artery Stenosis
- Cystic renal disease
- Endocrine tumours
- Essential Hypertension
- Obesity

38.9.4 Ten- to twenty-years

- Obesity
- Essential hypertension
- Reflux nephropathies with repeated Urinary Tract Infections
- Glomerulonephritis
- Renal Artery Stenosis
- Endocrine tumours
- Hyperthyroidism
- Drugs (Oral Contraceptive Pill, illicit drugs)

38.10 Evaluation of the Hypertensive Child

- Patient's history
- Symptoms of renal disease (haematuria, oliguria, evidence of bodily swelling, polyuria, enuresis)
- Symptoms of vasculitis or rheumatology (Joint swelling & rash)
- Past medical history (umbilical artery/vein catheterisation, previous renal disease e.g. Previous swelling)
- Drug History (steroids, Oral Contraceptive Pill, amphetamines, other illicit drugs)
- Birth History: Low Birth Weight
- Family History of Hypertension

Clues on physical examination include:

- Coarctation of the Aorta & Takayasu:
  - Femoral artery delay or imperceptible
  - Blood pressure discrepancy between arm & leg →COA, Takayasu arteritis
- Neurofibromatosis
  - Caf au lait spots

- RAS, Takayasu arteritis
  - Abdominal bruit
- Congenital adrenal hyperplasia
  - Ambiguous genitalia
- Dysmorphism suggestive of Turner or William syndromes
- Signs of Chronic Renal Failure: Growth failure (stunted), renal rickets, anaemia, oedema
- Bedside urine dipstick positive for protein and blood (± oedema)

38.11 Investigations

The rationale is 2-fold:

1. To define aetiology
2. To assess the presence of end-organ damage

Some of the investigations include:

- Full blood count
- Urine dipstick, microscopy and culture
- BUE, Serum Creatinine, Ca, Mg, PO4, blood gases
- Uric acid
- KUB ultrasound and Doppler studies to rule out Renal Artery Stenosis
- Chest X-ray for cardiomegaly
- Echocardiogram for Left Ventricular Hypertrophy (end organ damage)
- Fundoscopy
- Plasma Renin Activity (PRA) for RAS & renin secreting tumours
- Pre/post captopril nuclear scan
- MRA or CT Angiogram
- DMSA scan for renal scars
- Urine HVA & VMA for catechol amine secreting tumours/MIBG scintigraphy

38.12 Uric Acid and hypertension

Uric acid is increasingly being implicated in the pathogenesis of Hypertension in both adults and children. It is believed to cause endothelial dysfunction leading to microvascular and inflammatory injury to the kidneys. There are also reduced levels of endothelial-derived nitric oxide and associated elevation of the Renin-Aldosterone-Angiotensin System. Elevated uric acid levels in hypertensive individuals are associated with adverse outcomes like stroke. Allopurinol treatment is advocated for such individuals.

38.13 Complication of Hypertension

Some complications of Hypertension are listed below:

- Hypertensive encephalopathy
- Left Ventricular Failure
- Stroke
- Subarachnoid haemorrhage
- Secondary renal damage
- Retinopathy

38.14 Treatment of hypertension

38.14.1 Non-drug treatment

- Reducing salt intake
- Weight reduction for obesity-related hypertension
- Intake of more vegetables on account of potassium richness

38.14.2 Drug Treatment

Principles of anti-hypertensive therapy:

- Long-acting (once-daily medication)
- Maximise treatment dosage before adding on
- Agents used will come from the "ABCD" group:
  - **A**CE inhibitor and ARBs (Avoid if RAS suspected or in hypovolaemia)
  - **B**eta-blocker
  - **C**alcium channel blocker
  - **D**iuretic
  - **E**very other drug (methyl dopa, alpha-blockers, vasodilators like hydralazine

Generally, **A & B** drugs are not combined for Blood pressure control. Rather: **A + C + D** or **B + C + D**

38.15 Hypertensive encephalopathy

Hypertension with changes in mental status and/or seizures. Other manifestations are:

- Facial palsy
- Visual changes→blindness
- Coma

Pathophysiology: Disruption of the normal autoregulatory mechanisms of cerebral blood flow. The inability of cerebral vasculature to constrict appropriately in response to the abrupt increase in cerebral blood flow leads to cerebral hyperperfusion. Generally, short-acting antihypertensives are preferred in the initial instance of treatment so that any potentially harmful drop in blood pressure (which could lead to Posterior Reversible Encephalopathy Syndrome {PRES}) could be reversed. Subsequently, long-acting agents could be used Sublingual nifedipine could cause a precipitous drop in blood pressure so it is best avoided or should be used with extreme caution

Treatment outline:

- Use anti-hypertensive drugs
- Blood pressure should be brought down slowly to a desirable level (?stage I) by 48hrs (though not to normal levels) as follows:
  - 1/3 of total blood pressure reduction in 1st 12-hrs
  - Next one-third of the subsequent 12-hrs
  - Final one-third over 24-hrs
- Alternatively, by a quarter within 6 hours, and the rest in the next 24-36hrs

Commonly preferred drugs include Labetalol infusion, Na nitroprusside infusion, and IV hydralazine infusion. After achieving the desired blood pressure target, oral antihypertensives are then started

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