

# CYANOTIC CONGENITAL HEART DISEASES

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# DEFINITION

- Congenital heart diseases are defined as structural and functional problems of the heart that are present at birth.
- The incidence of congenital heart disease world wide is estimated at 8-12 per 1000 live births

# Congenital heart diseases

- There are more children born with congenital heart disease in areas with a high fertility rate such as in Africa
- Hence the burden of caring for this children is higher too in these regions.

# Congenital heart disease

- Cardiac defects represent the greatest overall burden of mortality and morbidity amongst children with congenital anomalies , and, together with neural tube defects and cleft lip and palate, account for 21 million disability adjusted life years world wide .
- One DALYS is equal to one healthy year of life lost due to disability or premature death

# Classification

- These congenital heart diseases present as different anomalies but can be broadly classified into cyanotic or acyanotic
- Cyanotic heart disease
- In cyanotic heart disease, the malformation allows into the arterial system, blood that is not fully oxygenated. The child there fore presents with a bluish discoloration of the skin and mucous membrane.

# Cyanotic heart disease

- The cyanotic examples include:
- Truncus arteriosus,
- Transposition of the great artery,
- Tetralogy of fallot,
- Tricuspid atresia
- Left hypoplastic heart syndrome,
- total anomalous pulmonary venous return,
- double outlet left ventricle,
- Ebstein anomaly,
- single ventricle

# Etiology

## **Etiology of congenital heart disease**

- In most cases the cause of congenital heart disease is unknown (90%).
- It is sporadic and some of the genetic risk factors have been identified as trisomy 21, 13, 18, di George and Turner's syndrome.

# ETIOLOGY

- The environmental risk factors include maternal alcohol
- Drug use
- intrauterine viral infection
- maternal systemic disease especially in the first trimester when the heart is being formed.



# CLINICAL PRESENTATION

- Despite the large number of the different types of congenital heart disease there is a limited number of physiological disturbances that can be produced in infancy

# CLINICAL PRESENTATION

- firstly It may present with cyanosis, heart failure, shock and arrhythmias, this constitutes an emergency, especially in the neonatal period.
- Examples of congenital heart disease that present this way are transposition of the great arteries, Truncus arteiosus, and hypoplastic heart disease

# CLINICAL PRESENTATION

- Secondly it may present as asymptomatic heart murmur example include: tetralogy of fallot
- Thirdly it may present as different respiratory infections and growth failure.
- As the child grows older and if uncorrected they become fatigued on exertion , and present with palpitations syncope edema, dyspnea, chest pains, and growth failure

# Diagnosis

- This can be done prenatal and/or after birth.
- It consists of a careful history, physical examination and cardiovascular examination, chest x-ray, electrocardiography, electrocardiography, cardiac catheterization, and cardiac MRI.

# Diagnosis

- Prenatal diagnosis: by the 22<sup>nd</sup> week of gestation, the heart is fully formed. In advanced nations, a prenatal fetal echocardiography is routinely done to diagnosis cardiac disease.

# Diagnosis

- The overall antenatal detection rate of major congenital heart disease has been found to be between 75% to 83%.
- Thus by the time of delivery plans have already been made for the appropriate surgery.

# Diagnosis

- Physical examination:
- This attempts to assess the physical findings looking out for
- tachycardia,
- dyspnea, edema,
- malnutrition,
- murmur,
- Cyanosis
- high blood pressures.
- The sensitivity of picking cardiac disease using history and physical examination even by a cardiologist is estimated at only 50 %.

# Diagnosis

- Pulse oximetry
- For daily diagnosis of critical congenital heart disease, pulse oximetry is an effective, non-invasive method.
- The Sensitivity, specificity, positive and negative predictive value were 77.78%, 99.90%, 25.93% and 99.99%, respectively



# DIAGNOSIS

- 2- Dimensional Echocardiogram:
- It is the mainstay of diagnosis, it is non invasive, relatively cheap and it has a sensitivity of 95% and specificity of 100%
- Newer technologies such as 3- Dimensional echocardiography are now available.

# DIAGNOSIS

- Cardiac catheterization and Cardiac MRI are reserved for the complex congenital heart disease diagnosis, which constitute 5% of all lesions.
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# TREATMENT

- Truncus Arteriosus
- Where there is only one artery arising from the heart and it branches to form the aorta and the pulmonary artery, its found in association with a Ventricular septal defect.

# Treatment

- Truncus arteriosus: VSD closure, repositioning of the Aorta and building a conduit to the pulmonary trunk and placing it in the right ventricular outlet should be done before six months of life.
- Avoids the complications of pulmonary hypertension,
- Ideally this should be completed within four to six weeks of life

# TREATMENT

- Transposition of the great artery:
- There are two great arteries, the aorta and the pulmonary.
- In transposition of the great artery, the arteries are switched, with the aorta arising from the right ventricle and the pulmonary from the left ventricle.
- This leads to the pumping of deoxygenated blood into the body system by the right ventricle into the aorta, while oxygenated blood is pumped back into the lungs. This defect is associated with ventricular septal defect, pulmonary valve stenosis

# TREATMENT

- TGA: an arterial switch repair (Jatene) procedure is the surgical choice within two weeks of life.

# TETRALOGY OF FALLOT

- Tetralogy of fallot:
- It has four components,
  - i) ventricular septal defect that allows blood to be shunted to the left side of the heart without getting oxygenated at the lungs
  - ii) a stenosis at or below or above the pulmonary valve
  - iii) muscular right ventricle
  - iv) the aorta sits above the VSD. All this leads to a cyanosed at birth or later in infancy

# TREATMENT

- Tetralogy of fallot:
- Staged repair between 6 to 18 month gives best outcomes.
- Surgery may range from an initial blalock taussing shunt to improve pulmonary blood flow for children with poorly developed pulmonary arteries and to allow for arterial growth, then a later full repair by 18 months.
- In children with well developed pulmonary arteries a complete repair can be done in one operation.
- Surgical risk for total correction is at 5%.



# TREATMENT

- Left hypoplastic left heart syndrome: There is variable success in the management of surgical therapy of hypoplastic left heart syndrome.
- Management includes palliation as with Norwood procedure from the neonatal period, heart transplant and supportive care for some.