Congenital Heart Defects

Congenital heart deffects are cardiac anomalies that involve the heart chambers, valves, and great vessels.

Their cause is unknown in 90% of the cases.

Congenital heart disease is frequently associated with other congenital defects.For example, cataracts, skeletal anomalies and deafness are seeing more commonly in people with congenital heart disease

Predisposing Factors to congenital heart defects

1. Fetal or maternal infection during the first trimester. The most common infection being Rubella and other teratogenic viral infections, like herpes simplex
2. Chromosomal abnormalisties such as trisomy21,18 and trisomy 13.
3. Maternal insulin dependent diabetes mellitus
4. Teratogenic drugs such as thalidomide and alcohol.
5. Heredity and genetic factors
6. Syndromes such as

- *Turners syndrome that is associated with aortic valve stenosis (AVS), coarctation of the aorta (CoA).*
- *Noonan's syndrome that is associated with dysplastic pulmonary valve.*
- *Down syndrome (Trisomy 21) is associated with artrioventricular (AV) canal defect,* Ventricular septal deffect (VSD)*. About 50% of the children with Down syndrome have a congenital heart defect.*

Classification of congenital heart defects

These heart deffects are lassified into three broad categories depending on the presence or absence of cyanosis.

1. Acyanotic and
2. Cyanotic
3. Obstructive

Acyanotic heart deffects (Group I) Left-to-Right Shunt

(A) These are lesions that lead to increase in the volume of blood (load) in the heart.

They are mostly associated with left-to-right shunting.

They include:

1. Atrial septal deffect (AVD)
2. Ventricular septal deffect (VSD)
3. Patent ductus arteriousus (PDA)
4. Regurgitatitant lesions like Atrioventricular valve regurgitation
5. Cardiomyopathies such as myocarditis

They result to a return of oxygenated blood back to the heart

the increase in bloof flow to the lungs leads to signs of respiratory dystress such as tachypnea, nasal flaring and chest retractions, shift if fluid to the interstitial causes pulmonary oedema and cardiac failure

(B) Lesions that increase pressure in the cardiac load mostly due to obstruction in blood flow in the heart.

1. Pulmonic valve stenosis
2. Aortic valve stenosis
3. Coarctation of the aorta
4. Tricuspid or mitral stenosis.

Characteristics of Acyanotic heart defects

These patients present with:

- Frequent upper respiratory tract infections, ie bronchopeneumonia
- Tachypnea
- Absence of cyanosis
- Features of congestive cardiac failure
- Precordial bulge due to cardiomegaly
- Hyperkinetic precordium on palpation
- Tricuspid or mitral delayed diastolic murmur
- Cardiomgaly, plethoric lung fields on a x-ray

Cyanotic Heart defects (Right-to-Left-Shunt)

These defects can be divided to

Those that ***decrease pulmonary blood flow*** include;

- Tetralogy of Fallot,
- Tricuspid atresia
- Transposition of great arteries with
  – DORV and PS
  – VSD with PS
  – Ebstein anomaly

They result from the obstruction in the pulmonary blood flow.

The child may be acyanotic during rest but will get cyanotic ("tet") spells during stress.

Those that ***increase pulmonary blood flow*** include;

- Transposition of great vessels,
- Persistent truncus arteriosus
- Single ventricle and
- Truncus arteriosus.

Thet result in mixing of systemic and pulmonary blood within the heart.

Characteristics of Cyanotic heart defects

- Cyanosis is usually accompanied by polycythemia and finger clubbing
- These patients have a normal Pulmonary Arterial Pressure
- They have a diminished Pulmonary Arterial Pressure with a decreased pulmonary blood flow too as a result of pulmonary stenosis
- They also have increased Pulmonary Arterial Pressure because of:
    - Increased pulmonary blood flow with slight cyanosis
    - Decreased pulmnary blood flow in moderate to severe cyanosis and irreversible pulmonary arterial hypertension

Obstructive Congenital heart defect (Group III)

Right-sided

• Pulmonary stenosis (valvular)

Left-sided

- Coarctation of aorta
- Congenital aortic stenosis
- Vascular rings
- Anomalous origin of coronary arteries
- Congenital mitral stenosis
- Congenital mitral incompetence
- Dextrocardia

Characteristics of Obstructive CHD

Patients with obstructive congenital heart defects present with;

- Cyanosis
- Absence of frequent upper respiratory tract infections
- Absence of precordial bulge
- Forcible or heaving cardiac impulse
- Thrill
- Ejection systolic murmur
- Delayed corresponding second sound.