CONGENITAL HEART DISEASE.

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Epidemiology

Prevalence:0.5-0.8% of live births (8/1000).Leading cause ofdeath in children with CHD. ■

Etiology:Unknown,multifactorial inheritance,genetic factors implicated,high incidence in first degree relatives. ■

3% have a single gene defect,13% have associated chromosomal abnormalities. ■

2-4% are associated with environmental or maternal conditions & teratogenic influences. ■

Gender differences:ASD,VSD,PDA & Pulmonic stenosis more common in girls,left sided lesions in boys. ■

Classification

Acyanotic:according to the predominant physiologic load placed on the heart. ■

Volume load:L-R shunts-ASD,VSD,PDA. ■

Pressure load:Ventricular outflow obstruction ■

Pulmonary,aortic valve lesions,aortic coarctation & pulmonary stenosis. ■

Cyanotic:based on pathophysiology. ■

Decreased pulmonary bloodflow:TOF,Pulmonary atresia,Tricuspid atresia,Single ventricle with pulmonic stenosis. ■

Increased pulmonary blood flow:Transposition of great vessels,Truncus arteriosus. ■

ATRIAL SEPTAL DEFECT.

Sinus venosus defect:high in the septum. ■

Ostium secundum defect:midseptum. ■

Ostium primum defect:low in the septum. ■

Pathophysiology:L-R shunt-increased flow across Rt heart-RV & PA enlargement. ■

Clinical features:asymptomatic,slow wt gain,frequent LRTI. ■

Diagnosis:Rt ventricular heave,systolic murmur,fixed wide split S2. ■

Investigations:

CXR:enlarged heart & PA,increased vascularity. ■

ECG:Rt axis in secundum defect,hallmark of primum defect is extreme Lt axis,RVH. ■

ECHO:RVH,valve anatomy,flow direction. ■

Treatment:closure during cardiac cathetrization,surgical closure. ■

VENTRICULAR SEPTAL DEFECT.

Most common CHD (26%),may be single or multiple. ■

Pathophysiology:Lt-Rt shunt as long as pulmonary vascular resistance is lower than systemic resistance,if reverse shunt reverses ■
Large defects lead to pul.hypertension-Eissenmenger syndrome. ■
Clinical features: depend on size,asymptomatic,growth failure,recurrent LRTI,congestive heart failure,SOB,cyanosis ■
Diagnosis:pansystolic murmur,loud p2. ■

Investigations

CXR:cardiomegaly,enlarged LA&LV. ■
ECG:extreme lt axis is charecteristic,biventricular hypertrophy. ■
ECHO:chamber size & pressures. ■
Cardiac catheter:O2 content,PA pressure,size & no of defects. ■
Treatment:Endocarditis prophylaxis,digoxin,diuretics. ■
Surgical closure before pulmonary vascular changes become irreversible. ■

PATENT DUCTUS ARTERIOSUS.

Connection between PA & descending aorta ■
10% of CHD. ■
Pathophysiology:Lt-Rt shunt,reverses if pulmonary resistance increases-RV enlargement.If PDA is large Eissenmenger syndrome can develop. ■
Clinical features:depend on size & direction of flow,slow growth,LRTI,SOB,cyanosis. ■
Diagnosis:bounding pulse,continous murmur,loud S2. ■

Investigations

CXR:cardiomegaly,increased pul vascularity. ■
ECG:Lt or biventricular hypertrophy. ■
ECHO:2D visualises PDA,doppler shows turbulance. ■
Cardiac catheter:PA pressures & O2 sats. ■
Treatment:Endocardial prophylaxis as long as patent,Indomethacin. ■
Surgical:ligation is curative. ■