ACYANOTIC CONGENITAL HEART DEFECTS

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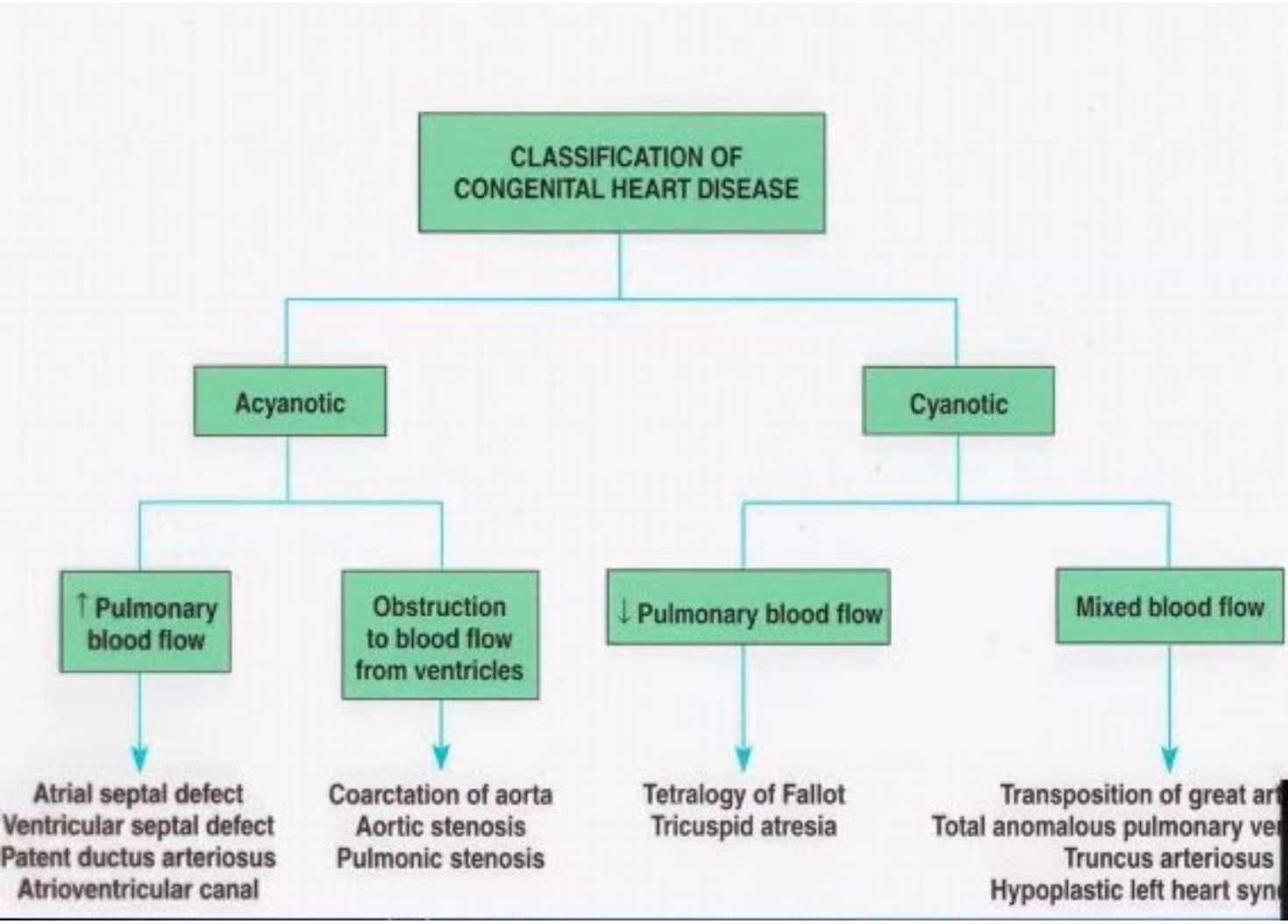
INTRODUCTION:

- Congenital heart diseases are broadly classified into two categories:

1. Cyanotic heart diseases
2. Acyanotic heart diseases

- Cyanotic heart diseases refer to congenital heart defect that occurs due to deoxygenated blood bypassing the lungs and entering systemic circulation(right to left shunt) or mixture of oxygenated and deoxygenated blood entering systemic circulation,hence leading to cyanosis.
- Acyanotic heart diseases refer to congenital heart defect in which infant has no cyanosis because there is no mixing of deoxygenated blood in systemic circulation.



LESIONS RESULTING IN INCREASED VOLUME LOAD

- The most common lesions in this group are those that cause left-to right shunts
- Atrial septal defect
- Ventricular septal defect
- Patent ductus arteriosus.

INCIDENCE

- Ventricular septal defect 30%
- Patent ductus arteriosus 12%
- Atrial septal defect 7%
- Pulmonary stenosis 7%
- Aortic stenosis 5%
- Coarctation of aorta 5%

ATRIAL SEPTAL DEFECT

- Secundum ASD
- Primum ASD
- Siuns Venosus ASD

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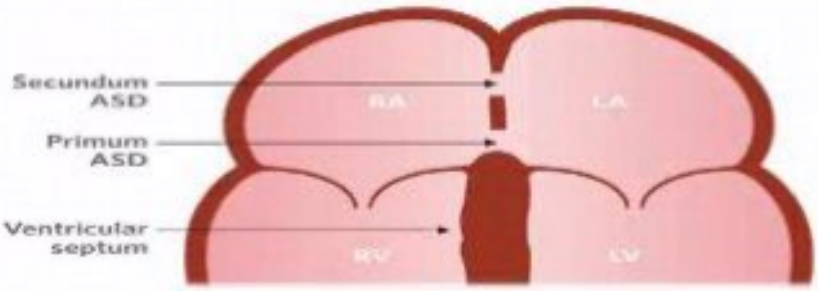
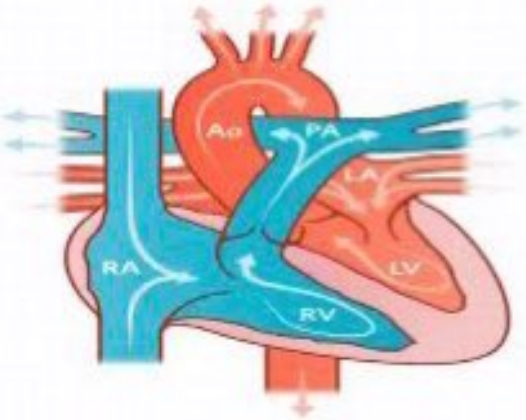
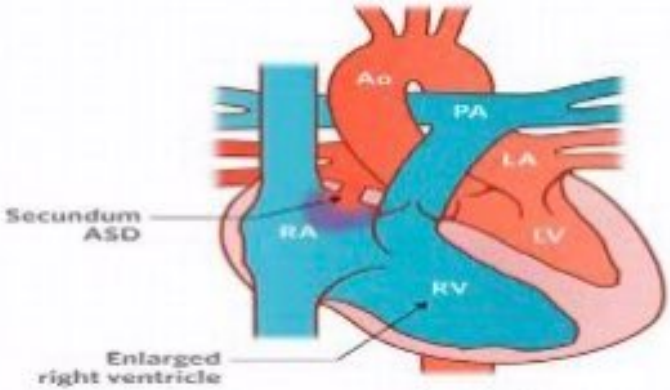
Atrial septal defect (ASD)

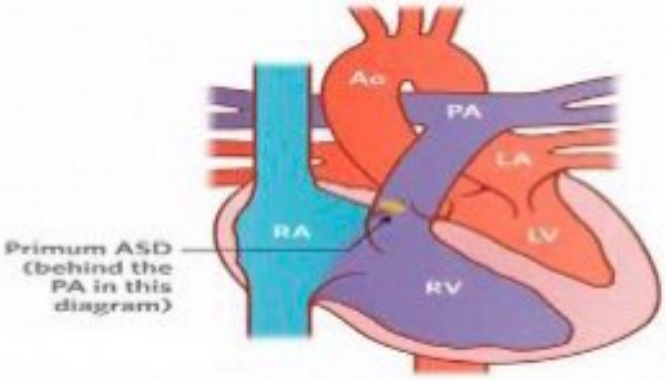




Normal heart and circulation



Secundum atrial septal defect



Primum atrial septal defect

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OSTIUM SECUNDUM DEFECT

- An ostium secundum defect in the region of the fossa ovalis is the most common form of ASD and is associated with structurally normal (AV) valves.
- Most common atrial septal defect(80%)
- May be single or multiple i.e fenestrsted
- Females outnumber males 3 : 1

CLINICAL FEATURES

- Often asymptomatic.
- Subtle faliure to thrive and exercise intolerance.
- Mild left precordial buldge.
- wide Fixed splitting of S2.
- Ejection systolic murmur at left middle and upper sternal border i.e harsh ,medium pitch , sometimes with thrill.

PRIMUM ASD

- It is defect in lower part of atrial septum near the atrioventricular valve.
- 10% of ASDs.
- Asymptomatic.
- Soft systolic murmer at upper left sternal egde.
- Apical pansystolic murmer(Atrioventricular valve regurgitation)
- Fixed split of S2

PRIMUM ASD

- It is defect in lower part of atrial septum near the atrioventricular valve.
- 10% of ASDs.
- Asymptomatic.
- Soft systolic murmer at upper left sternal egde.
- Apical pansystolic murmer(Atrioventricular valve regurgitation)
- Fixed split of S2

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SINUS VENOSUS ASD

- A defect in upper end of atrial septum such that superior vena cava overrides the atrial septum.
- 5% of ASDs.
- Soft systolic murmur at upper left sternal edge.
- Fixed split S2.

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DIAGNOSIS

- Xray chest.
- ECG.
- Echocardiography.
- Cardiac catheterisation.

XRAY CHEST

- Enlargement of the right ventricle and atrium, depending on the size of the shunt.
- pulmonary artery is enlarged, and pulmonary vascularity is increased.
- Cardiac enlargement is often best appreciated on the lateral view

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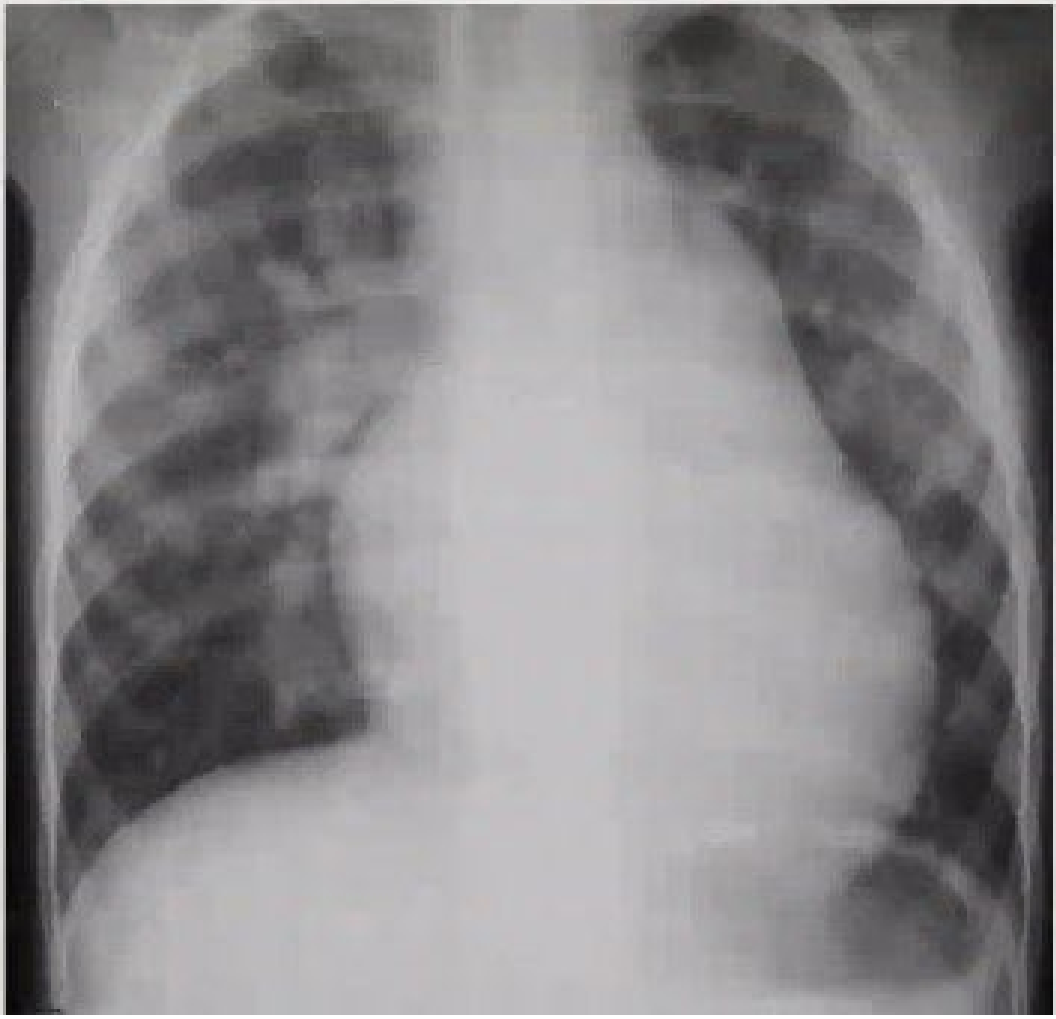

ECG

- right axis deviation
- minor right ventricular conduction delay (rsR' pattern in the right precordial leads)

TREATMENT

- Transcatheter and surgical device closure is indicated in all symptomatic patients
- In all asymptomatic patients with a 2:1 systemic to pulmonary shunt ratio
- Surgery should be performed at age of 1 year prior to school going age

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VENTRICULAR SEPTAL DEFECTS

- Most common cardiac defect.
- Restrictive and non restrictive.
- Restrictive i.e right ventricular pressure is normal usually <5mm
- Non restrictive right and left ventricular pressure is equalized usually large >10mm
- Membranous most common in posterioinferior and anterior to septal leaflet
- muscular midportion of apical region may be single or multiple (swiss cheese type)

CLINICAL FEATURES

- Small VSDs with trivial left-to-right shunts and normal pulmonary arterial pressure asymptomatic.
- Large VSDs are associated with
- dyspnea,
- feeding difficulties,
- poor growth,
- profuse perspiration, recurrent pulmonary infections, cardiac failure in early infancy

EXAMINATION

- Left precordial bulge, apical thrust, systolic thrill at left lower sternal border
- A loud, harsh, or blowing holosystolic murmur at the left lower sternal border
- Loud P2
- Ejection systolic murmur at pulmonary area can also be heard

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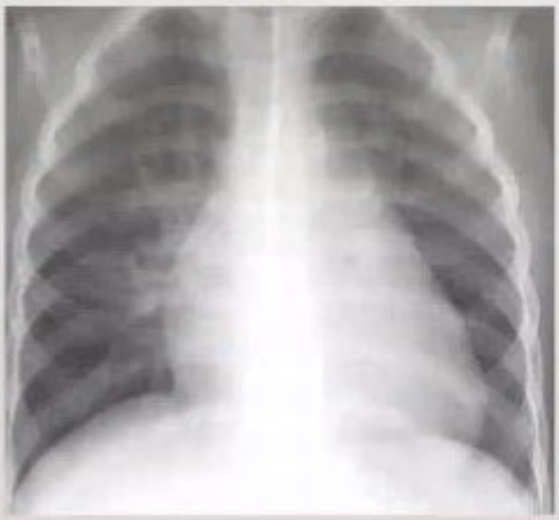
X RAY CHEST



- Increased Pulmonary vascular markings.
- Cardiomegaly.

ECG

- Small defects shows normal ECG or left axis deviation
- The presence of right axis deviation due to right ventricle hypertrophy suggests pulmonary hypertension
- Notched or peaked P waves
- Biventricular hypertrophy with extreme left axis deviation

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TREATMENT

- 30–50% of small defects close spontaneously, most frequently during the 1st 2 yr of life.
- Small muscular VSDs are more likely to close (up to 80%) than membranous VSDs are (up to 35%).
- Small VSDs needs reassurance and no prophylaxis for infective endocaditis is required.
- Large VSD require surgical closure usually at age of 3-5months.

PATENT DUTUS ARTERIOSUS

- Connection between Pulmonary Artery & descending aorta
- Female patients with PDA outnumber males 2 : 1.
- PDA is also associated with maternal rubella infection during early pregnancy
- Common in pre-matures
- PDA persists beyond 1st few weeks of life rarely close spontaneously or with medical managment but early closure is not indicated in pre-mature babies

CLINICAL FEATURES

- Small PDA are asymptomatic
- Large PDAs are associated with growth retardation, congestive cardiac failure, feeding abnormalities

On physical examination

- wide pulse pressure due to runoff of blood in pulmonary artry during diastole with bounding pulses
- Displaced heaving apex beat

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- Palpable systolic thrill in left 2nd intercoastal space radiating towards left clavicle and left sternal border
- A continous machinery murmur heard all over the precordium

ECG

- If left to right shunt is small,ECG is normal.
- If large shunt left ventricular or biventricular hypertrophy present.

TREATMENT

- Irrespective of age, patients with PDA require catheter or surgical closure
- closure of the ductus is indicated in asymptomatic patients, preferably before 1 yr of age

Medical management

- Indomethacin 0.2mg/kg/dose in three doses 8-12 hours apart
- Contraindication to indomethacin includes derranged RFTs, thrombocytopenia, sepsis or Necrotising Enterocolitis.

COARCTATION OF AORTA

- Constriction of aorta of varying degree at any point transverse arch to bifurcation of iliac artries
- 98% are juxtaductal occur below origin of left subclavian artery and origion of ductus arteriousus

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CLINICAL FEATURES

- Diagnosed after infancy as it is usually asymptomatic
- Neonates and infants with severe coarctation and transverse arch hypoplasia shows lower body hypoperfusion, acidosis and severe heart failure signs are delayed till closure of ductus arteriosus
- Before closure of ductus may show differential cyanosis of lower limbs.
- Heart is enlarged with systolic murmer at left sternal border and loud first heart sound.

- In 2nd decade of life may complain of leg pain and weakness after excercise
- Older children present with hypertension.
- Classic sign of coartation of aorta is disparity of blood pressure in arms and legs
- Lower limb puses are weak and may b absent in contrast to upper limb bounding pulses
- Radial and femoral pulses should be palpated for radio-femoral delay

- Radio-femoral delay due to collatrals
- Diffrence in B.P of both arms usually higher in right than left is due to involvement of left subcliavian artery
- Precordial impluse and heart sounds are normal
- Short systolic ejection click may b heared due to bicupid aortic valve

DAIGNOSIS

- Chest X-Ray finding depends upon age and effect of collatrals and hypertension
- Cardiomegaly
- Pulmonary congestion
- Notching of inferior border of ribs due to collaterals

ELECTROCARDIOGRAM

- Normal in younger children.
- Left ventricular hypertrophy in older children
- May show right or biventricular hypertrophy in neonate

TREATMENT

- In neonates prostaglandins E1 is given to reopen ductus and adequate lower limb perfusion
- Older children with heart failure and good perfusion are treated with anticongestive medications
- Mainstay of treatment is surgical repair.

- 3) subclivian flap procedure division of left subclivian artry and incorporate into wall of repaired coarctation
- 4)primery stent placement is under trials
- After surgery amplitude of pulses in lower limbs is increased
- Rebound hypertension is common which is medically managed

PULMONARY STENOSIS

- The pulmonary valve cusps are deformed to various degrees, as a result valves open incompletely during systole.
- The obstruction to outflow from right ventricle to pulmonary artery results in increased right ventricle systolic pressure and wall stress leading to hypertrophy of right ventricle.

CLINICAL FEATURES

- Patients with mild to moderate stenosis are asymptomatic.
- If severe stenosis, signs of right ventricular failure such as hepatomegaly, peripheral edema and exercise intolerance may be present.
- With mild pulmonary stenosis, venous pressure and pulse are normal, heart is not enlarged, low or medium pitched ejection systolic murmer is maximally audible over pulmonic area, S2 is split with soft pulmonic component.

- In moderate pulmonic stenosis venous pressure is slightly elevated, right ventricular lift present, loud ejection systolic murmur in pulmonary area, S2 split with soft pulmonic component.
- In severe pulmonic stenosis venous pressure raised, heart is enlarged, right ventricular lift present ,loud and long ejection systolic murmur audible in pulmonic area radiating to entire precordium, neck, lung fields and back.

CXR

- In mild cases ,only post stenotic dilatation of pulmonary artery present.
- In moderate cases ,heart is mildly enlarged with normal or decreased pulmonary vascularity.
- In severe cases, cardiomegaly, prominence of main pulmonary artery and decreased pulmonary vascular markings.

ECG

- In mild cases, normal or mild right ventricular hypertrophy.
- In moderate cases, right ventricular hypertrophy and prominent spiked P wave.
- In severe cases, gross right ventricular hypertrophy with prominent and spiked P wave.

TREATMENT

- Patients with moderate or severe pulmonic stenosis require relief of obstruction.
- Balloon valvuloplasty is initial treatment of choice.
- Patients with severely thickened valve may require surgical intervention.

AORTIC STENOSIS

- Aortic stenosis is more frequent in males(3:1)
- Most common form is valvular aortic stenosis in which leaflets are thickened and commissures are fused.
- Subvalvular and supravalvular aortic stenosis are less common.

CLINICAL FEATURES

- In less severe form of aortic stenosis, children are asymptomatic and show normal growth and development. Pulses, heart size and apical impulse are normal.
- In severe cases, fatigue, angina, dizziness or syncope may develop. Pulses diminished ,heart enlarged with LV apical thrust, second heart sound diminished.

CXR

- Heart size normal.
- Prominent ascending aorta.

ECG

- Occasionally normal but evidence of left ventricular hypertrophy present.

TREATMENT

- Balloon valvuloplasty in cases of moderate to severe stenosis to prevent LV dysfunction.
- Aortic valve replacement in case of calcified valve.