Coarctation of Aorta

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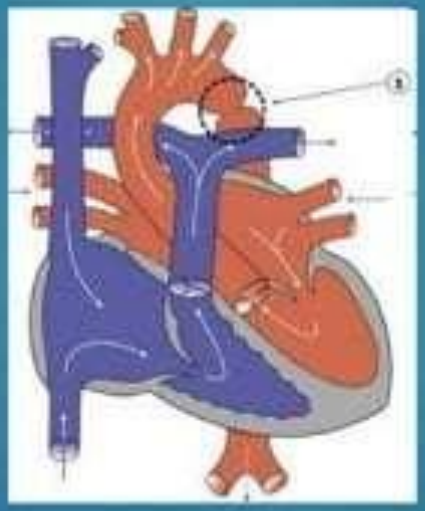

Definition

- *Coarctation of Aorta consists of a discrete narrowing in the proximal thoracic aorta, just opposite to the insertion of the ductus arteriosus.*
- *It may be long segment stenosis*

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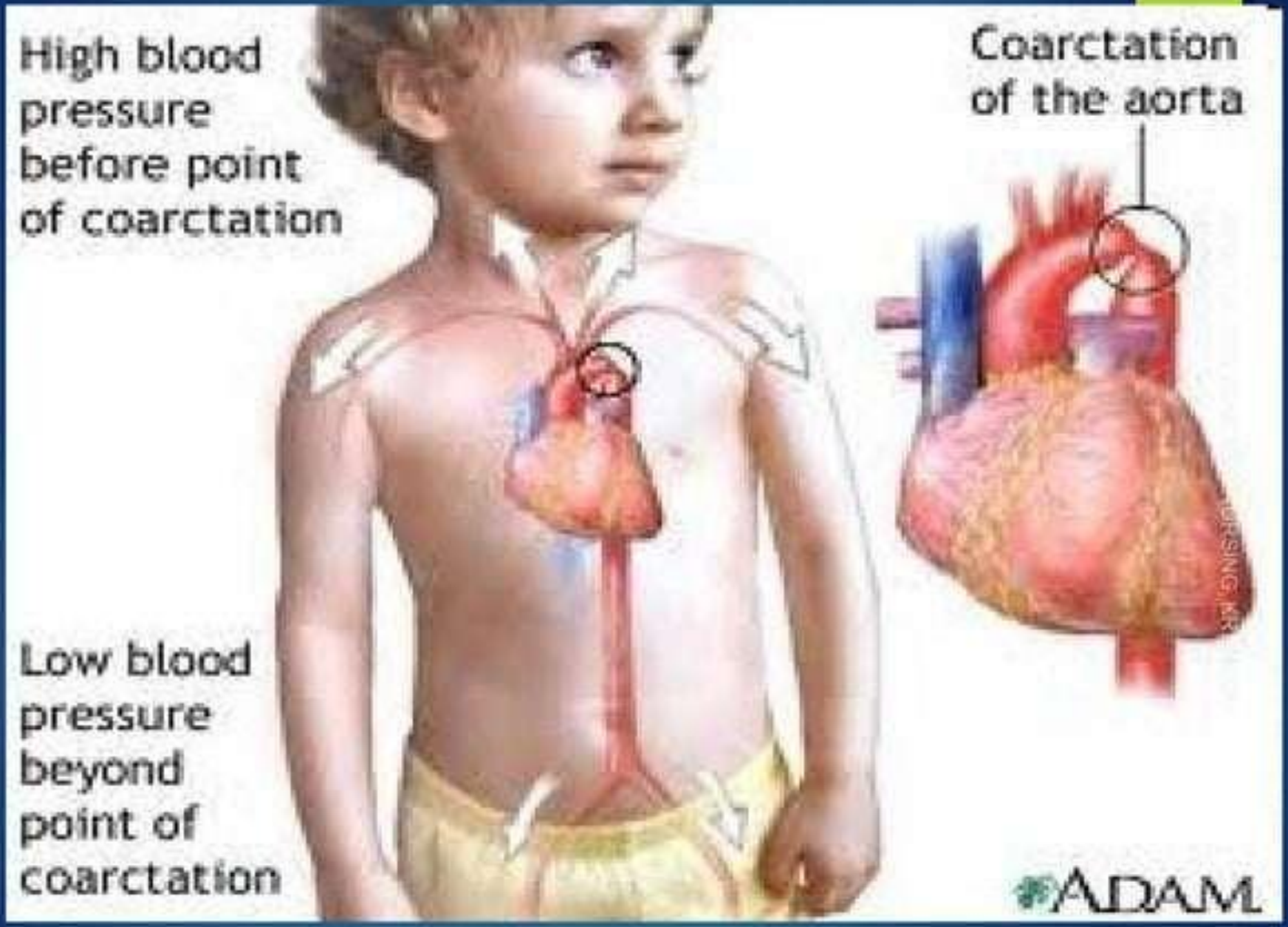
High blood pressure before point of coarctation
Coarctation of the aorta
Low blood pressure beyond point of coarctation
ADAM

Types

1. Simple coarctation :

it can be with or without the presence of PDA

2. Complex Coarctation:

coexist with VSD, ASD, TGA, aortic stenosis, AVSD

3. Other associated anomalies:

variations in brachiocephalic artery anatomy

Development of collateral arterial circulation

berry aneurysms of the circle of Willis

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Etiology

1. *Abnormal migration of ductus smooth muscle cells into the periductal aorta, with subsequent constriction & narrowing of the aortic lumen*
2. *Develop as a result of hemodynamic disturbances that reduce the volume of blood flow through the fetal aortic arch and isthmus*

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Epidemiology

- Common with 6-8% of CHD"S
- Male > female- 2:1
- Present in 35% of female with turners syndrome (X0)

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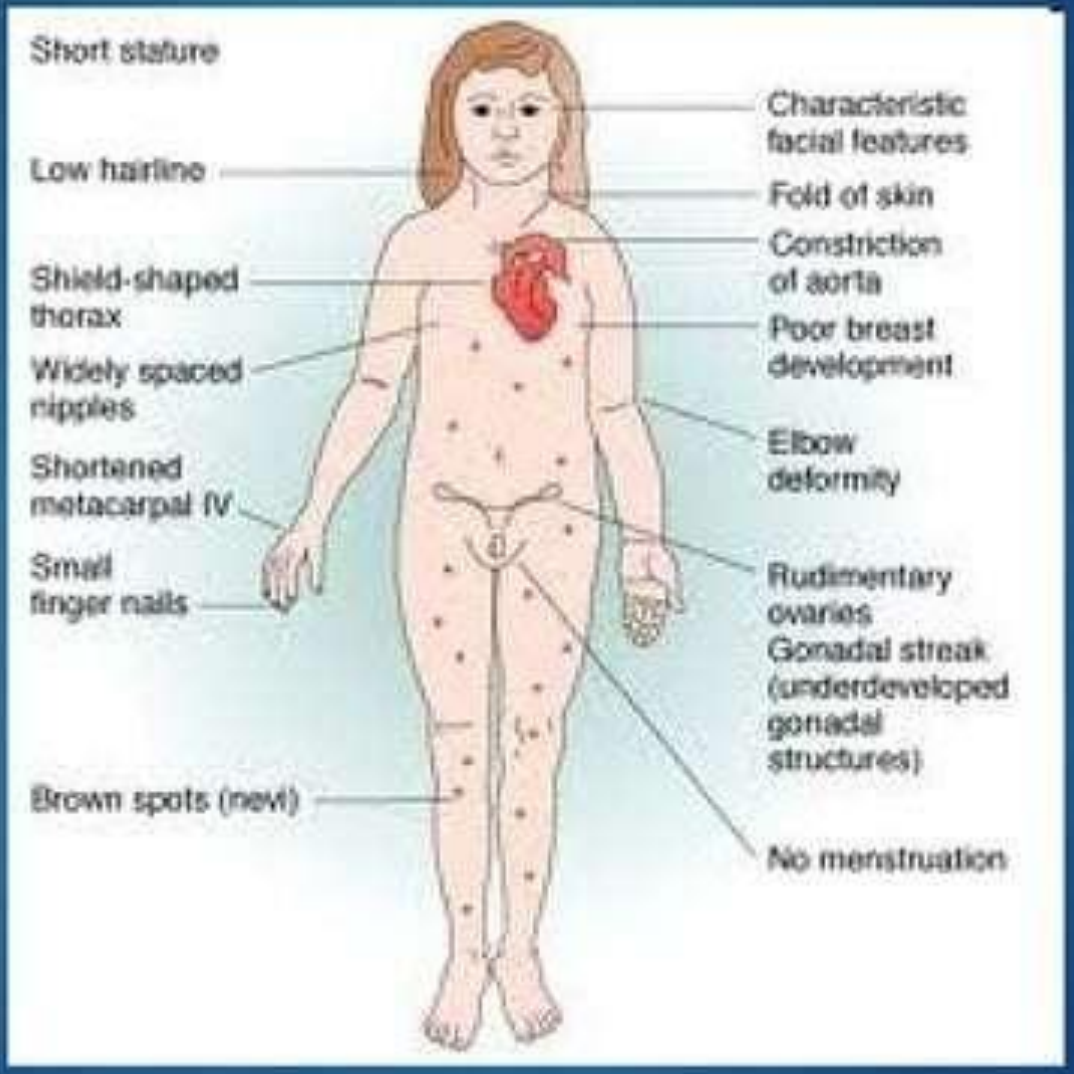
Turners syndrome



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Pathophysiology

Blood from right ventricle

↓

Pulmonary artery

↓ (with PDA)

Descending arch of aorta

↓

Supplied to the body

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If PDA closes

Narrowing or stenosis of aorta

↓

Left ventricular overload or hypertrophy

↓

Left atrial overload

↓

Pulmonary congestion

↓

Pulmonary artery hypertension

↓

Right ventricular hypertrophy

↓

Congestive cardiac failure

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Clinical features

1) Asymptomatic with minor defect
2) Feeding difficulty, poor weight gain
3) Dyspnoea on exertion
4) Failure to thrive
5) Pitting edema
6) Gallop rhythm
7) Heart murmur
8) Fatigue, weakness
9) Frequent epistaxis

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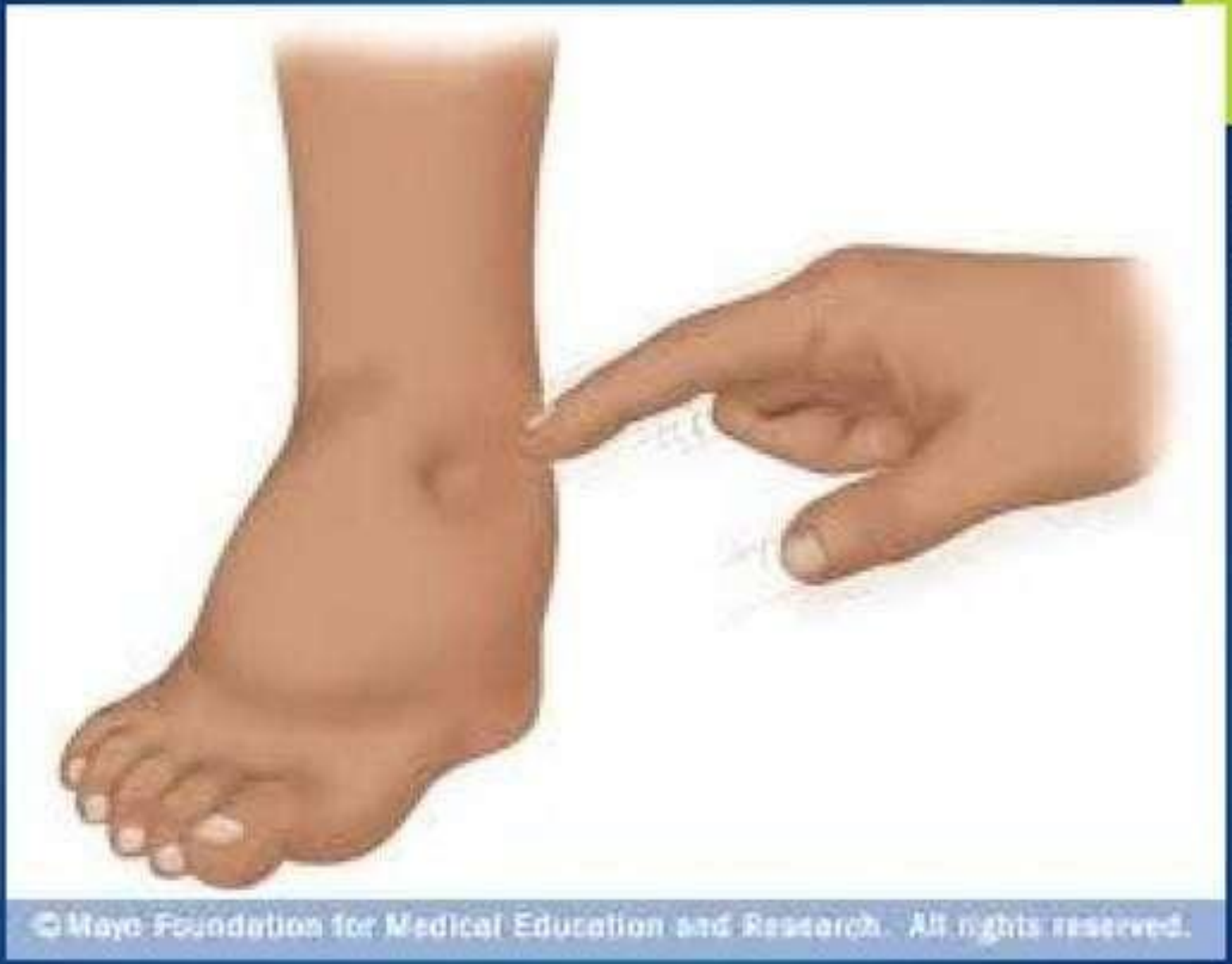

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10) Cramps and intermittent claudication
11) Headache
12) Over growth of upper limbs and chest
13) Dilated and tortuous collaterals may be seen over the inter scapular area in older children called as ' SUZMAN SIGN'.

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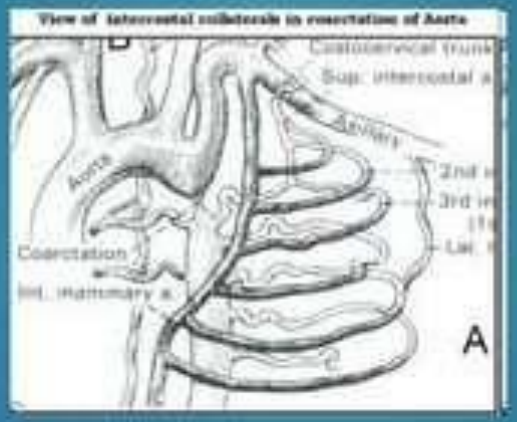
Collaterals- Suzman sign



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14) *Tachycardia, Tachypnoea*

15) *Diaphoresis*

16) *Hepatomegaly*

17) *BP & pulse of upper and lower limb vary*

Upper limb may have full noting, bounding pulse where as lower extremities pulse may not be palpable.

BP will have the variation of 20mm of Hg

from upper to lower limb.

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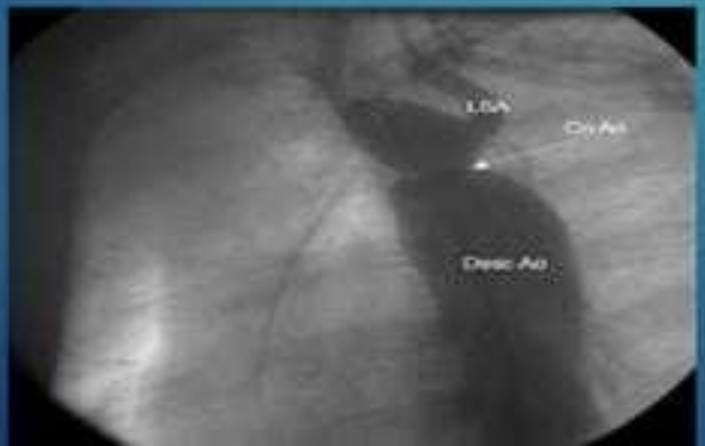
Diagnosis

A. ECG
B. Echocardiography: stenosis, lesions
C. X-ray: ventricular enlargement, 'E' or '3' sign on barium swallow, due to precoarctational, post coarctational dilatation & middle narrowed coarctation.



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Management

Medical :

- Ionotropic support and diuretic therapy
- Prostaglandin infusion

Surgical :

- End to end anastomosis
- Prosthetic patch aortoplasty
- Subclavian flap aortoplasty
- Percutaneous balloon angioplasty

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- *Following surgery re-coarctation, systemic HTN can arise. Which could be corrected with balloon angioplasty.*

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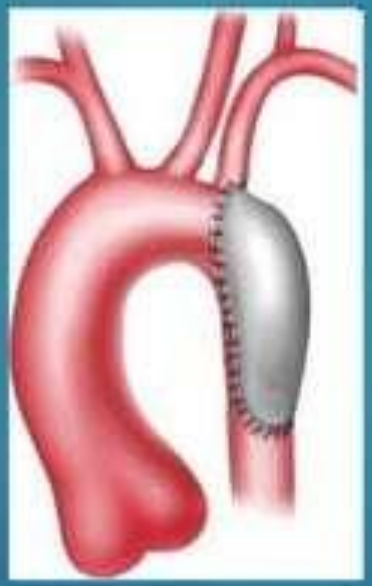
Patch aortoplasty



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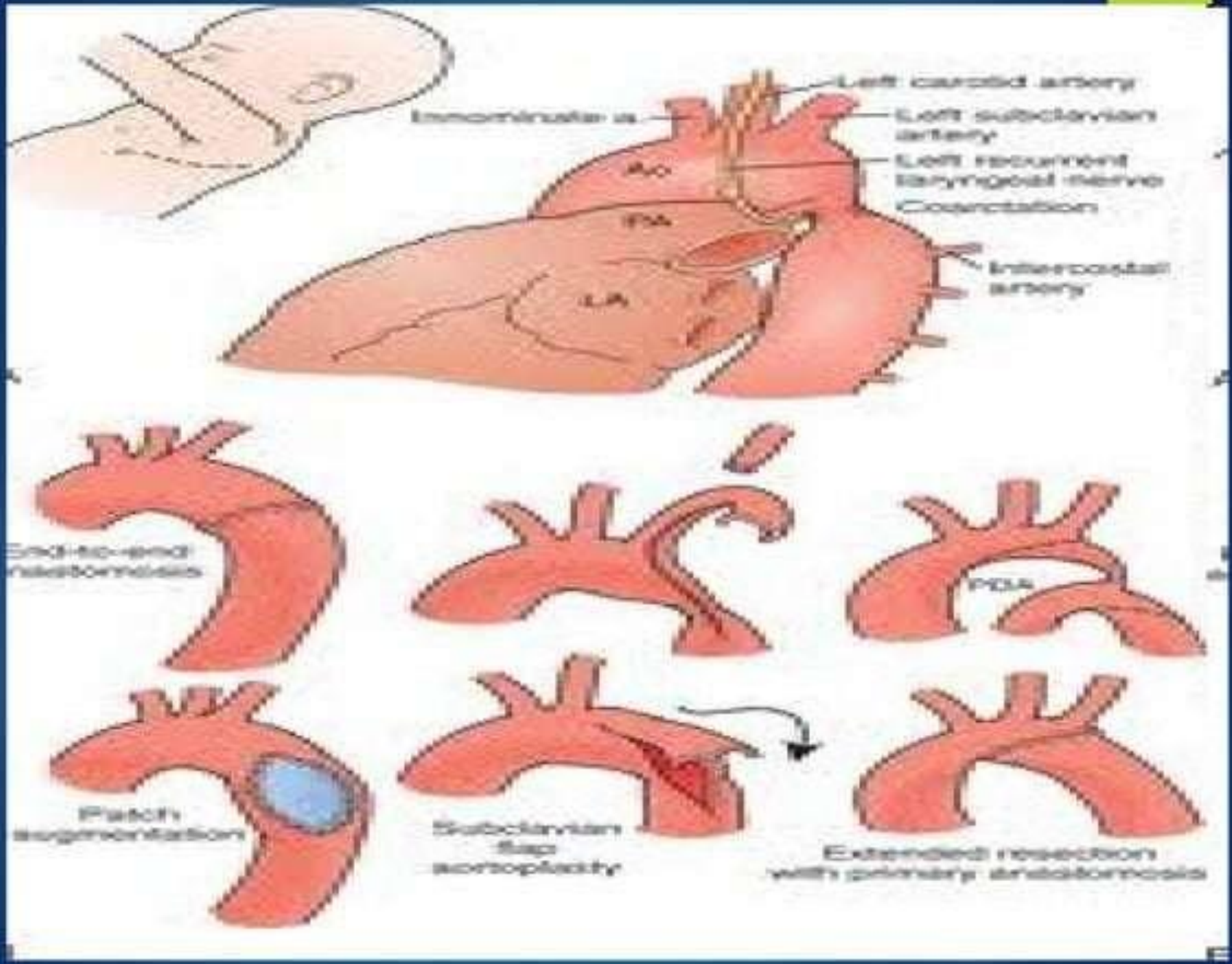
Left carotid artery
Left subclavian artery
Left recurrent laryngeal nerve
Coarctation
Intercostal artery
Ao
PA
LA
PDA
Patch augmentation
Subclavian flap aortoplasty
Extended resection with primary anastomosis

Complications

- CCF
- Aortic rupture
- Aortic aneurysm
- CVA
- Rupture of berry intracranial aneurysm
- Rupture of an intercostal aneurysm
- Dissection of aorta

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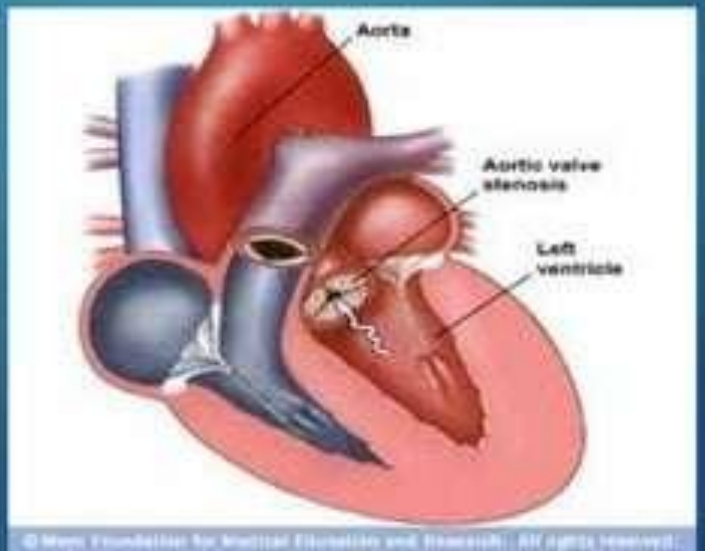
Aortic stenosis

- *Obstruction to the left ventricular outflow tract which may be at the level of the aortic valve, above the valve (supravalvular) or below the valve (infravalvular)*
- *Constitutes 8% of CHD*



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Hemodynamics

Blood from the left ventricle

↓through stenosed valve

Aorta

↓

But stenosed valve of the aorta creates more pressure in the left ventricle

↓

Left ventricular hypertrophy

↓

MI & pulmonary edema

CCF, & Infective endocarditis

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Clinical features

During infusion:

- Decreased cardiac output
- Feeble peripheral pulses
- Pale look
- Dusky & Cool skin

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In older children:

- Chest infections
- Chest pain or angina
- Dyspnea on exertion
- Fatigue
- Narrow pulse pressure
- Fainting episodes (syncope)
- Exercise intolerance

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Diagnosis

- Pulse is low volume & there is a thrill in the suprasternal notch
- Auscultation reveals a harsh, low-pitched systolic ejection murmur, maximal at the second right intercostal space.
- Chest X-ray- dilated aorta, enlargement of left ventricle
- ECG-T wave inversion
- Echo- identifies level of obstruction

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Prevention & Treatment

- *Restriction of physical activities like athletics, outdoor games, strenuous activities and competitive sports. Because it can be a deadliest disease causing death within no time.*

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- *Balloon dilatation or balloon aortic valvuloplasty is a non-operative relief of the lesion. Done through femoral catheterization. Only contraindication is aortic regurgitation.*

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Surgery

- Aortic valvulotomy
- Aortic valve replacement

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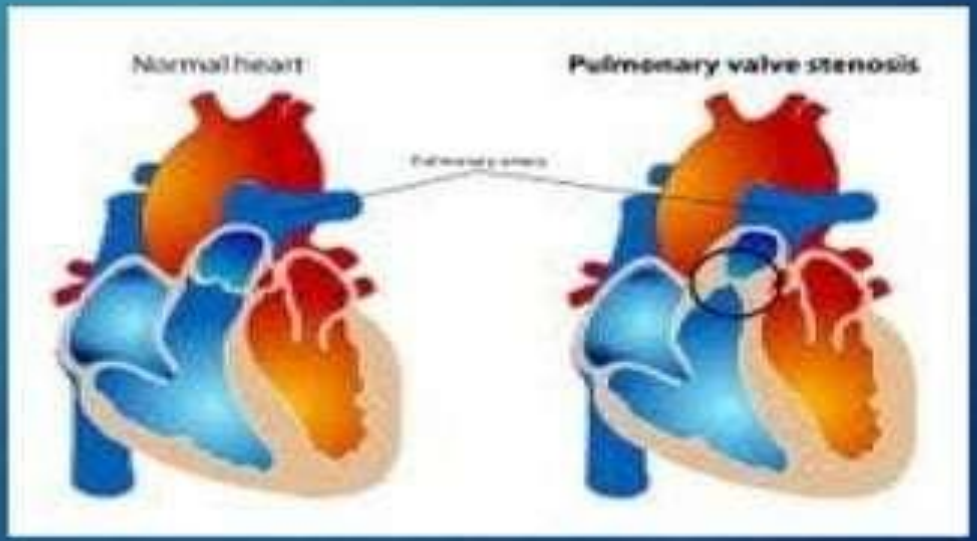
Pulmonic stenosis

- There is obstruction to flow of blood from the right ventricle to lungs.
- Accounts for 2% of CHD.
- 90% are at the level of the valve remaining supravalvular and subvalvular (infundibular)



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Hemodynamics

Blood from the right ventricle

↓ through stenosed valve

Pulmonary artery

↓

Because of obstruction to the output and to maintain the cardiac output right ventricle undergoes hypertrophy

↓

Rt. Sided CCF

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Clinical features

- Child may be generally asymptomatic but may have decreased exercise tolerance with fatigue and dyspnea.
- With severe obstruction :

Dyspnea

Cyanosis,

Precordial pain

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- *On physical examination: the patients ate characteristically described as having a round face and hypertelorism. Port-wine angiomatous malformation over the skin. Turner's phenotype without chromosomal abnormalities is associated with pulmonic stenosis.*

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Diagnosis

- Auscultation reveals a systolic ejection murmur over pulmonic area
- Chest X-ray- enlargement of right ventricle and main pulmonary artery
- ECG- rt. Ventricular hypertrophy
- 2D Echo -Level of obstruction

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Treatment

- At cardiac catheterization if the right ventricular pressure is 75% or more of the systemic systolic pressure child can be operated.
- Balloon pulmonary valvuloplasty

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