UNIVERSITY OF ANBAR

الكلية : الطب

القسم او الفرع :طب الاطفال وحديثي الولادة

المرحلة: الخامسة

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اسم المادة بالغة العربية : طب الاطفال وحديثي الولادة

اسم المادة باللغة الإنكليزية : طب الاطفال وحديثي الولادة

اسم المحاضرة الأولى باللغة العربية: جهاز القلب والدوران الجزء الثاني

اسم المحاضرة الأولى باللغة الإنكليزية: Pediatrics cardiology part 2 lec 2

Pediatrics cardiology (lec 2 /1 hr)

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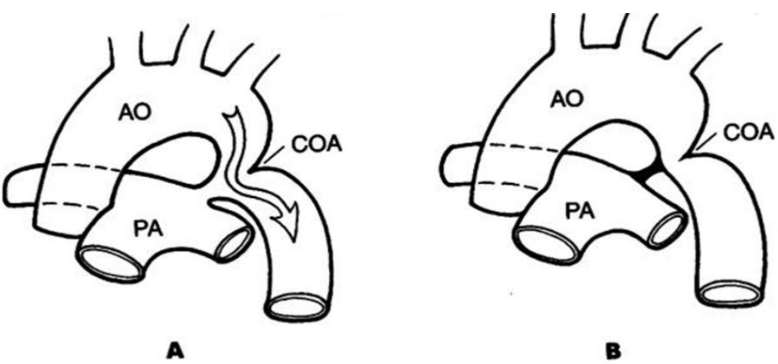
COARCTATION OF THE AORTA

Localized, discrete narrowing of the thoracic aorta.

occurs in 6-8% of congenital heart disease.

More common in males with M:F = 2:1.

More common cardiac lesion in Turner syndrome.



C/F:

In infants, usually presented with congestive heart failure, dyspnoea, poor feeding.

In older children, might be only systemic hypertension

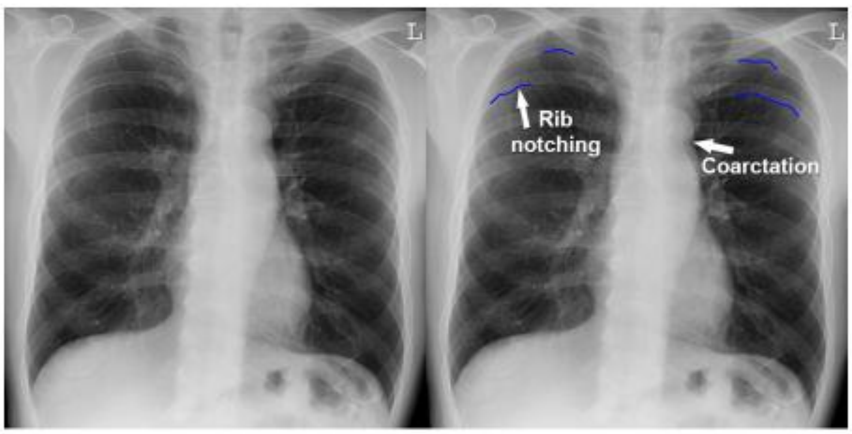
OE: Radiofemoral delay and pressure difference between the upper and lower limb by 20 mmHg (diagnostic)

Diagnostic tests:

Chest x-rays shows rib notching ,post stenotic dilatation of the aorta .

Echo: diagnostic, non-invasive, relatively cheap test of choice.

MRI & CT angiography: radiation exposure, costlier, needs GA, not always available used in difficult cases only.



Rx:

Balloon angioplasty (with or without stent)

Surgery, e.g. by end to end anastomosis.

Prognosis:

The mean age of death for untreated coarctation is about 34 years and the most common cause of death are: heart failure, aortic rupture, infective endocarditis, intracranial hemorrhage (due to B.P. or aneurysm).

PULMONARY STENOSIS

Classification according to severity:

- **Mild**: when the pressure gradient < 50 mmHg
- **Moderate**: when the pressure gradient 50-80 mmHg
- **Severe**: when the pressure gradient > 80 mmHg

C/F:

Most patients are asymptomatic and discovered accidentally.

Symptoms: exertional dypnoea and sometime chest pain.

O/E:

The asymptomatic patient has normal growth (unless it is a severe case).

In symptomatic patients:

S1 is normal.

Non-fixed splitting S2.

Systolic ejection click at pulmonary areas.

Ejection systolic murmur with thrill (i.e. > grade 4), maximally at pulmonary area.

CXR:

Prominent pulmonary conus caused by post-stenotic pulmonary dilatation.

Normal pulmonary vascularity, i.e. there is no oligemia of the lung as expected after pulmonary stenosis because no intracardiac shunt (i.e. all the blood in the heart from the systemic circulation goes to the lung).

ECG: Rt. Axis deviation and RVH & peaked P-wave (RAH).

Echo:

For Dx

Severity determination

Level of stenosis (supra, valvular or sub).

Rx: Mild: just reassurance; follow up.

Moderate to severe: balloon valvuloplasty;

if failed □ surgical valvotomy.

TETRALOGY OF FALLOT

It is the “commonest” cyanotic congenital heart disease in children and adults. It is a combination of:

VSD

Pulmonary stenosis (PS).

Overriding of aorta.

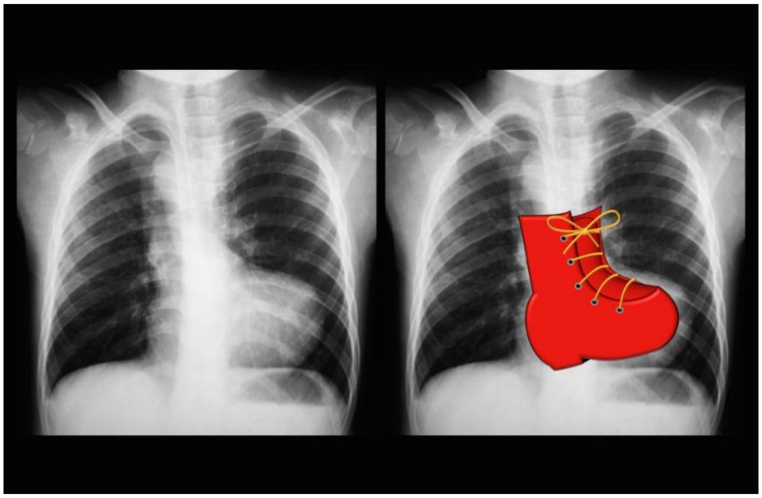
Right ventricular hypertrophy (RVH).

Hemodynamics: The degree of right ventricular outflow obstruction (i.e. P.S.) determines the degree of pulmonary blood flow and the severity of cyanosis.

C/F:

Cyanosis: most patients develop cyanosis in the first 6 month – 1 year of life.

Dyspnoea: occurs on exertion □ they have a limited exercise tolerance.



Squatting: it is the characteristic posture of children with TOF after any physical effort.

O/E:

Cyanosis (central and peripheral).

Clubbing (usually after 3 months of age).

Single S2 & ejection systolic murmur at the left sternal border.

Normal pulse & quiet precordium with RVH.

ECG: Right axis deviation (RAD) & right ventricular hypertrophy (RVH).

CXR:

Normal heart size.

Rounded apex situated above the diaphragm (hypertrophied RV).

Concavity at the main pulmonary artery area.

Reduced pulmonary vascularity (oligemic lung).

Coeur-en-sabot (boot shape)

Echo: It is essential for Dx.

Catheterization: Gives the definitive Dx and as a preoperative step.

Complications:

Hypercyanotic spells

Cerebrovascular accidents (CVA)

Cerebral abscess

Infective endocarditis

Hematological complications include bleeding and thrombosis.

Rx:

Medical:

Iron supplementation & nutritional support.

Phlebotomy: in symptomatic patients with hematocrit more than 65% (as there is a risk for CVA).

Surgical:

#Palliative to □ pulmonary blood flow e.g.: Blalock-Taussig shunt (BT shunt).

#Total surgical repair.

All patients should have complete surgical correction at the end.

Timing of the operation varies greatly from institution to institution, but early surgery is generally preferred.

Most centers prefer primary elective repair by 1 to 2 years of age, even if the patients are asymptomatic, acyanotic, or minimally cyanotic.

However, cyanotic spells, SPO2 <75%, or being symptomatic may speed the surgery

Hypercyanotic spells (Tet spells, Fallot spells, hypoxic spells):

Hypoxic spells are characterized by a paroxysm of hyperpnoea (i.e., rapid and deep respiration), irritability and prolonged crying, increasing cyanosis, and decreasing intensity of the heart murmur.

Occur in infants: peak incidence between 2 and 4 months of age.

Occur in the morning after crying, feeding, or defecation.

May lead to limpness, convulsion, cerebrovascular accident, or even death.

Treatment of the hypoxic spell

Break the vicious circle of the spell:

1 -The infant should be picked up and held in a knee-chest position.

2 -Morphine sulfate, 0.2 mg/kg (SC, IV) as a sedative.

3 -Oxygen is usually administered (little effect).

4 -Acidosis should be treated with sodium bicarbonate ($NaHCO_3$), 1 mEq/kg administered intravenously. The same dose can be repeated in 10 to 15 minutes.

If the hypoxic spells do not fully respond to these measures, the following medications can be tried ;

1 -Vasoconstrictors such as phenylephrine, 0.02 mg/kg administered intravenously, may be effective (by raising systemic arterial pressure).

2 -Ketamine (GA), 1 to 3 mg/kg. It increases the systemic vascular resistance and sedates the infant.

3 -Propranolol (Beta Blocker), 0.01 mg/kg administered by slow intravenous push, reduces the heart rate and may reverse the spell.

4 -Emergency surgical treatment (palliative or corrective)

TRANSPOSITION OF GREAT ARTERIES

In TGA, there is an inverted relationship, aorta & RV, pulmonary artery and LV.

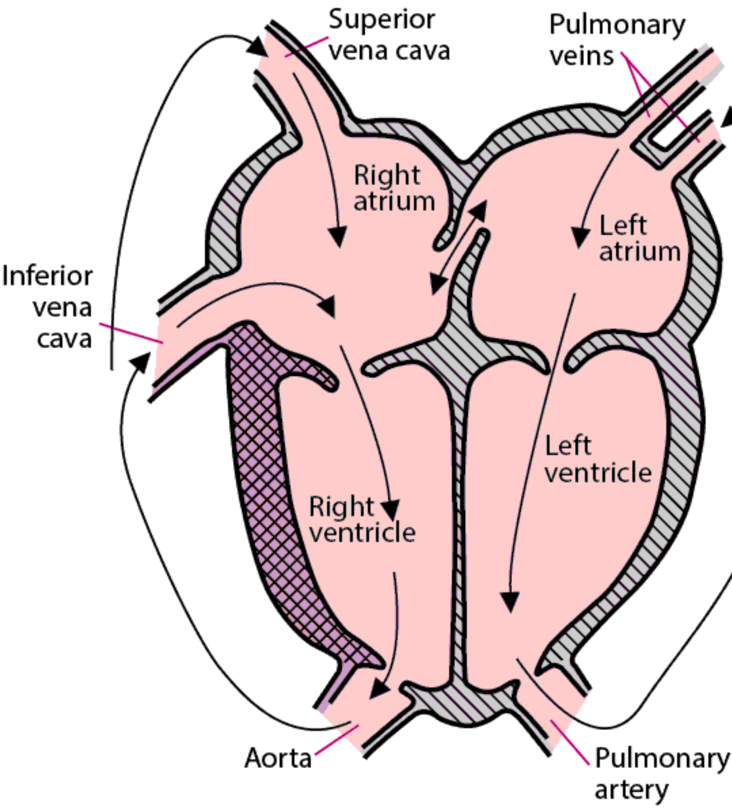
5% of CHD.

M:F is 2:1

More common in infants with DM mothers.

It is the most common cause of cyanotic CD at birth.

Without surgery, 90% will die within the first year of life.



Clinical features: Cyanosis and tachypnea in the first hours or days of life

CXR: Mild cardiomegaly with "egg-on-side" appearence of heart with a narrow base and increase in pulmonary vascularity.

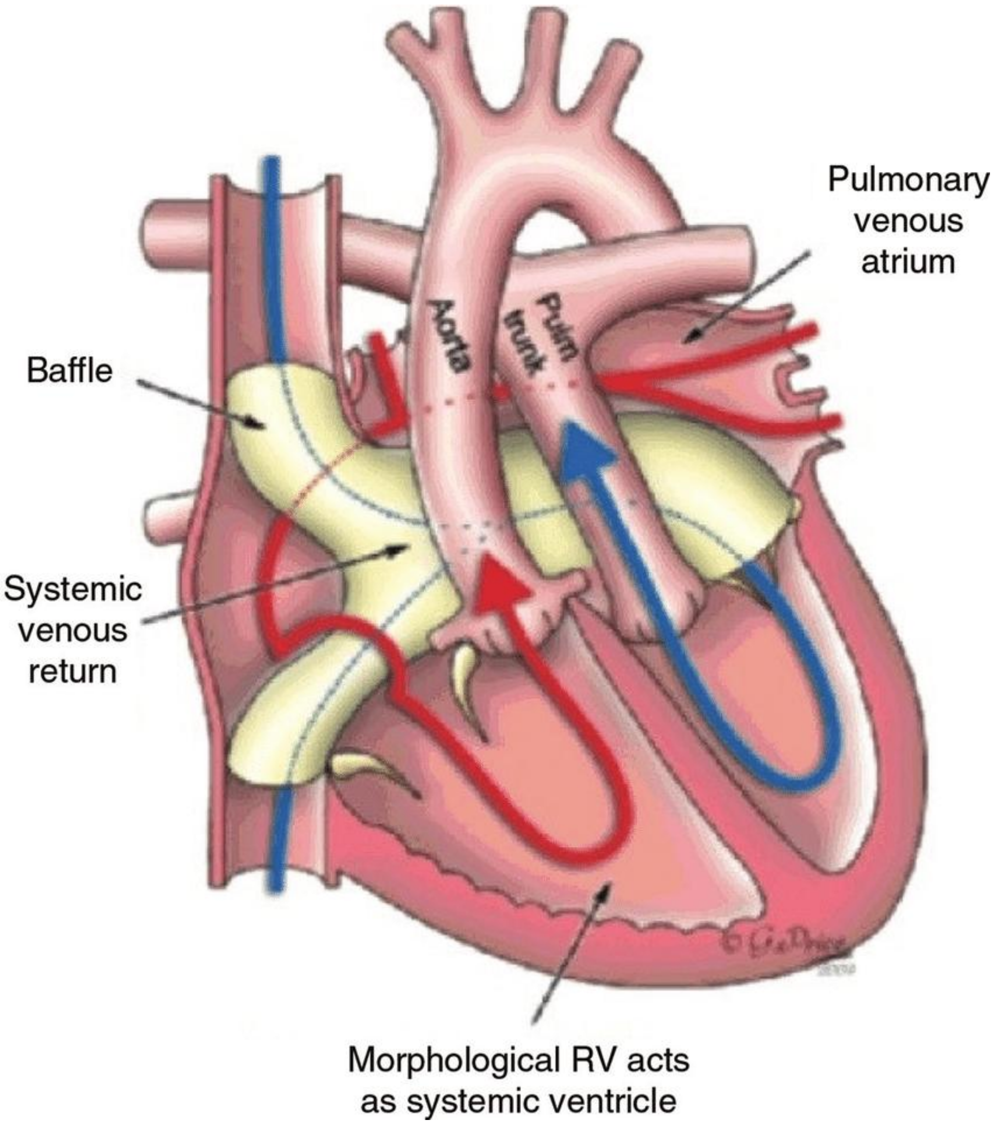
Surgical treatment:

Palliative ; Rashkind Atrial Septostomy: Help mixing of arterial and venous blood

Corrective surgery:

Atrial switch (Physiological)

Arterial switch (anatomical): depends on the anatomy and the surgeon.



Atrial Switch Procedure