Pathology of the Heart
Lecture II

by
Thair Wali Ali

- Patent Ductus Arteriosus PDA
  - During intrauterine life, the ductus arteriosus permits blood flow from the pulmonary artery to the aorta, thereby bypassing the unoxygenated lungs.
  - Shortly after birth, however, the ductus constricts; this occurs in response to increased arterial oxygenation, decreased pulmonary vascular resistance, and declining local levels of prostaglandin E_2.

- In healthy term infants, the ductus is functionally nonpatent within 1 to 2 days after birth; complete, structural obliteration occurs within the first few months of extrauterine life to form the *ligamentum arteriosum.*

- Ductal closure is often delayed (or even absent) in infants with hypoxia (resulting from respiratory distress or heart disease).

- PDAs account for about 7% of cases of congenital heart lesions; 90% of these are isolated defects.

- The remaining occur with other congenital defects, most commonly VSDs.

Clinical Features

- PDAs are high-pressure left-to-right shunts, audible as harsh "machinery-like" murmurs.
- A small PDA generally causes no symptoms.
- Larger bore defects can eventually lead to the Eisenmenger syndrome with cyanosis and CHF.
- The high-pressure shunt also predisposes affected individuals to infective endocarditis.

- Preservation of ductal patency (by administering prostaglandin E) may be critically important for infants with various forms of congenital heart disease wherein the PDA is the only means to provide systemic or pulmonary blood flow (e.g., aortic or pulmonic atresia).

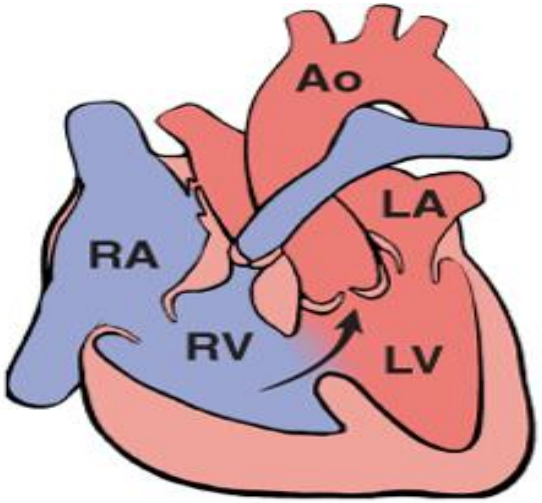
Right-to-Left Shunts

- Poorly oxygenated blood from the right side of the heart is introduced directly into the arterial circulation.

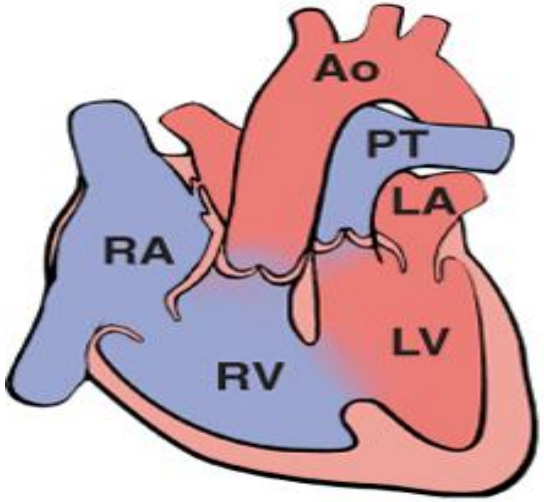
- Cardiac malformations associated with right-to-left shunts are distinguished by *cyanosis at or near the time of birth.*

- Two of the most important conditions associated with cyanotic congenital heart disease are:
  - Tetralogy of Fallot (TOF)
  - Transposition of the great vessels (TGV)

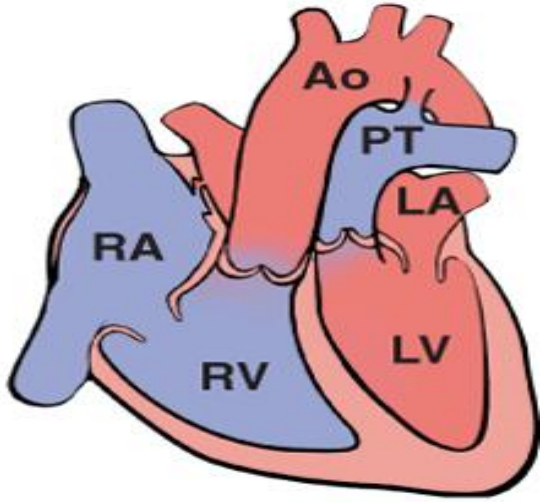
- Clinical findings associated with severe, long-standing cyanosis include:
  - Clubbing of the fingertips (hypertrophic osteoarthropathy)
  - Polycythemia.
  - In addition, right-to-left shunts permit venous emboli to bypass the lungs and directly enter the systemic circulation (paradoxical embolism).



A Classic Tetralogy of Fallot



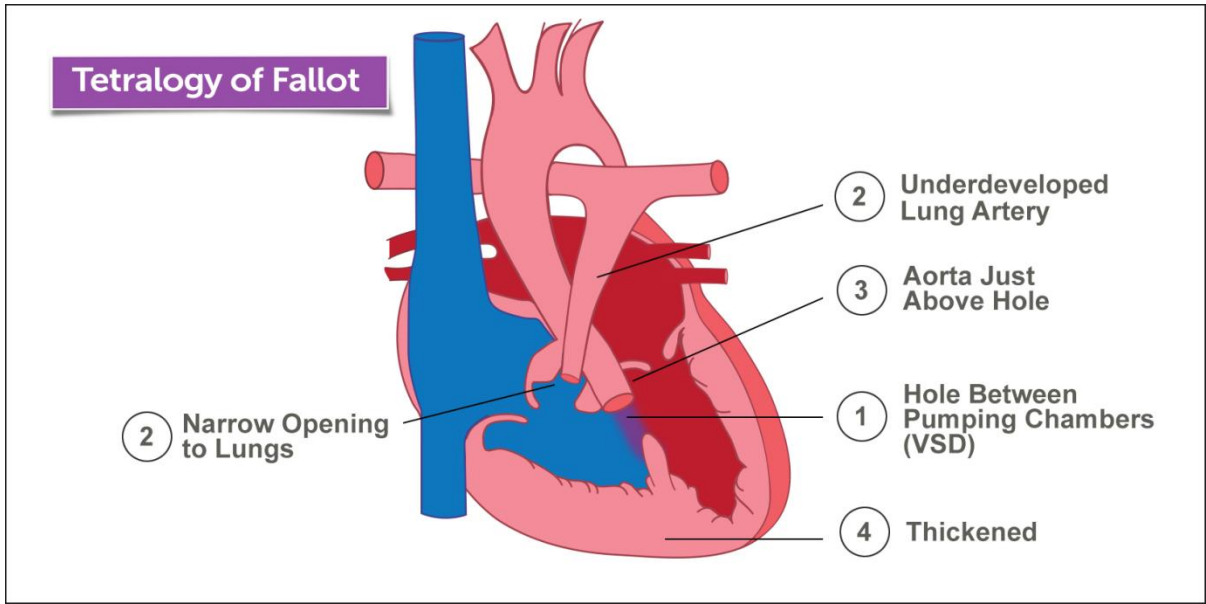
With VSD



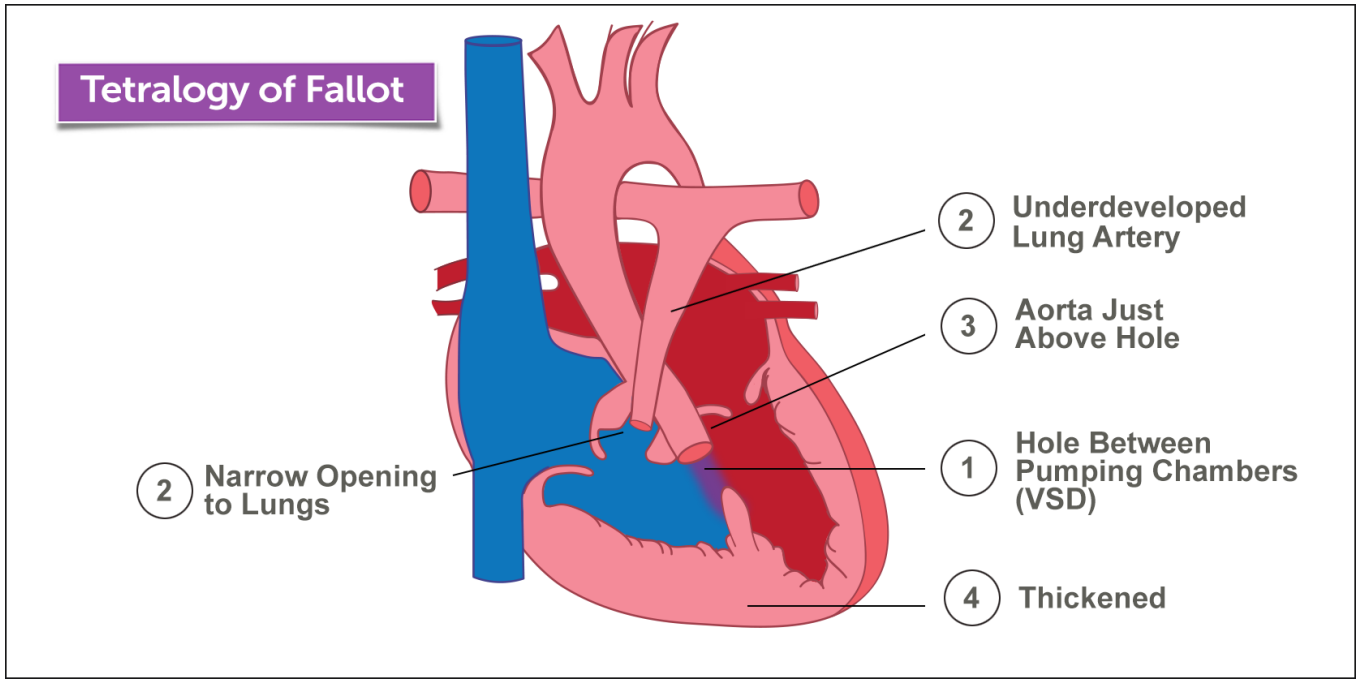
Without VSD

B Complete Transposition

- Tetralogy of Fallot (TOF)
  - 5% of all congenital cardiac malformations
  - is the most common cause of cyanotic congenital heart disease



- Teatures of TOF
  1. VSD,
  2. Obstruction to the right ventricular outflow tract (subpulmonic stenosis),
  3. an aorta that overrides the VSD
  4. right ventricular hypertrophy.



Clinical Features

- The clinical severity largely depends on the degree of the pulmonary outflow obstruction.
  - If it is mild, the condition resembles an isolated VSD, because the high left-sided pressures on the left side cause a left-to-right shunt with no cyanosis.
  - More commonly, marked stenosis causes significant right-to-left shunting and consequent cyanosis early in life.

- As patients with tetralogy grow, the pulmonic orifice does not enlarge, despite an overall increase in the size of the heart.

- Hence, the degree of stenosis typically worsens with time resulting in increasing cyanosis.

- The lungs are protected from hemodynamic overload by the pulmonic stenosis, so that pulmonary hypertension does not develop.

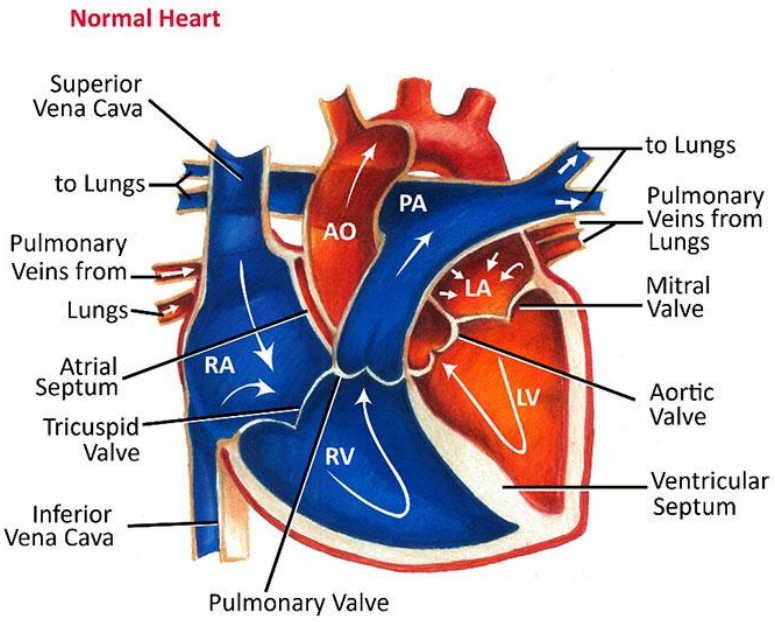
- As with any cyanotic heart disease, patients develop **erythrocytosis** with attendant **hyperviscosity**, and **hypertrophic osteoarthropathy**.

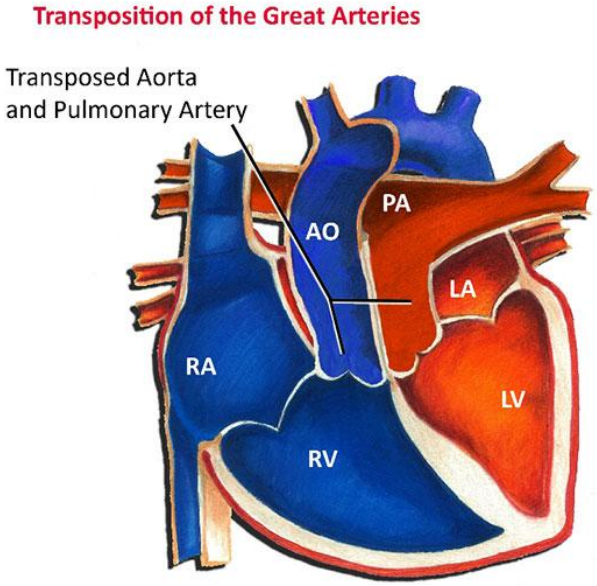
- The right-to-left shunting also increases the risk **for infective endocarditis**, **systemic emboli, and brain abscesses**.

- Surgical correction of this defect is now possible in most instances.

- Transposition of the Great Arteries (TAG)
  - The aorta arises from the right ventricle and the pulmonary artery emanates from the left ventricle

    - The functional outcome is separation of the systemic and pulmonary circulations, a condition incompatible with postnatal life unless a shunt exists for adequate mixing of blood and delivery of oxygenated blood to the aorta.





- Patients with TGA and a VSD (~35%) tend to have a relatively stable shunt.

- Those individuals with only a patent foramen ovale or PDA (~65%) tend to have unstable shunts that can close and often require surgical intervention within the first few days of life.

Obstructive Lesions

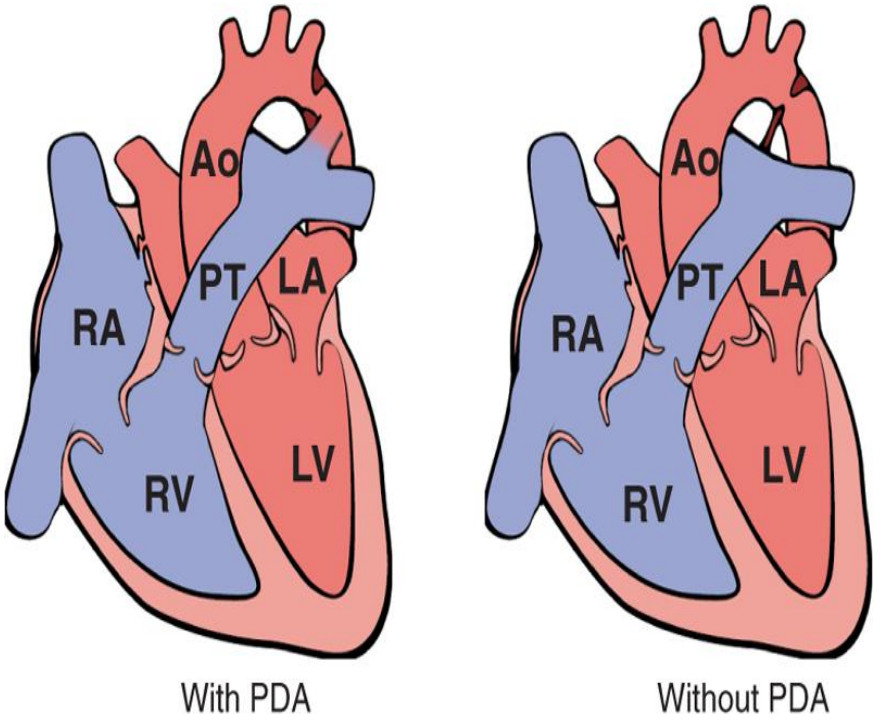
- Congenital obstruction to blood flow can occur at the level of the heart valves or within a great vessel.

- Obstruction can also occur within a chamber, as with subpulmonic stenosis in tetralogy of Fallot.

- Common examples of congenital obstruction include pulmonic valve stenosis, aortic valve stenosis or atresia, and coarctation of the aorta.

Aortic Coarctation

- Males are affected twice as often as females
- Two classic forms have been described:
- an "infantile" form with hypoplasia of the aortic arch proximal to a PDA,
- an "adult" form in which there is a discrete ridgelike infolding of the aorta, just opposite the ligamentum arteriosum distal to the arch vessels.



Coarctation of Aorta

- Coarctation of the aorta may occur as a solitary defect, but in more than 50% of cases, it is accompanied by a bicuspid aortic valve.

Clinical Features

- Clinical manifestations depend almost entirely on the severity of the narrowing and the patency of the ductus arteriosus.

- ***Preductal coarctation of the aorta with a PDA***

  – usually leads to manifestations early in life, hence the older designation of *infantile* coarctation;

  – it may cause signs and symptoms immediately after birth.

  – In such cases, the delivery of poorly oxygenated blood through the ductus arteriosus produces cyanosis localized to the lower half of the body.

  – Femoral pulses are almost always weaker than those of the upper extremeties.

  – Many such infants do not survive the neonatal period without intervention.

- ***Postductal coarctation of the aorta without a PDA***

  - is usually asymptomatic, and the disease may go unrecognized until well into adult life.

  - Typically, there is upper extremity hypertension, due to poor perfusion of the kidneys, but weak pulses and a lower blood pressure in the lower extremities.

- Claudication and coldness of the lower extremeties result from arterial insufficiency.

- Adults tend to show exuberant collateral circulation "around" the coarctation involving markedly enlarged **intercostal and internal mammary arteries**; expansion of the flow through these vessels leads to radiographically visible "**notching**" of the ribs.


Thank
you!!!
...