Pathology of the heart lecture I

by
Thair Wali Ali

Congenital heart disease

- Congenital heart diseases are abnormalities of the heart or great vessels that are **present at birth**.

- Most such disorders arise from faulty embryogenesis during gestational **weeks 3 through 8**, when major cardiovascular structures develop.

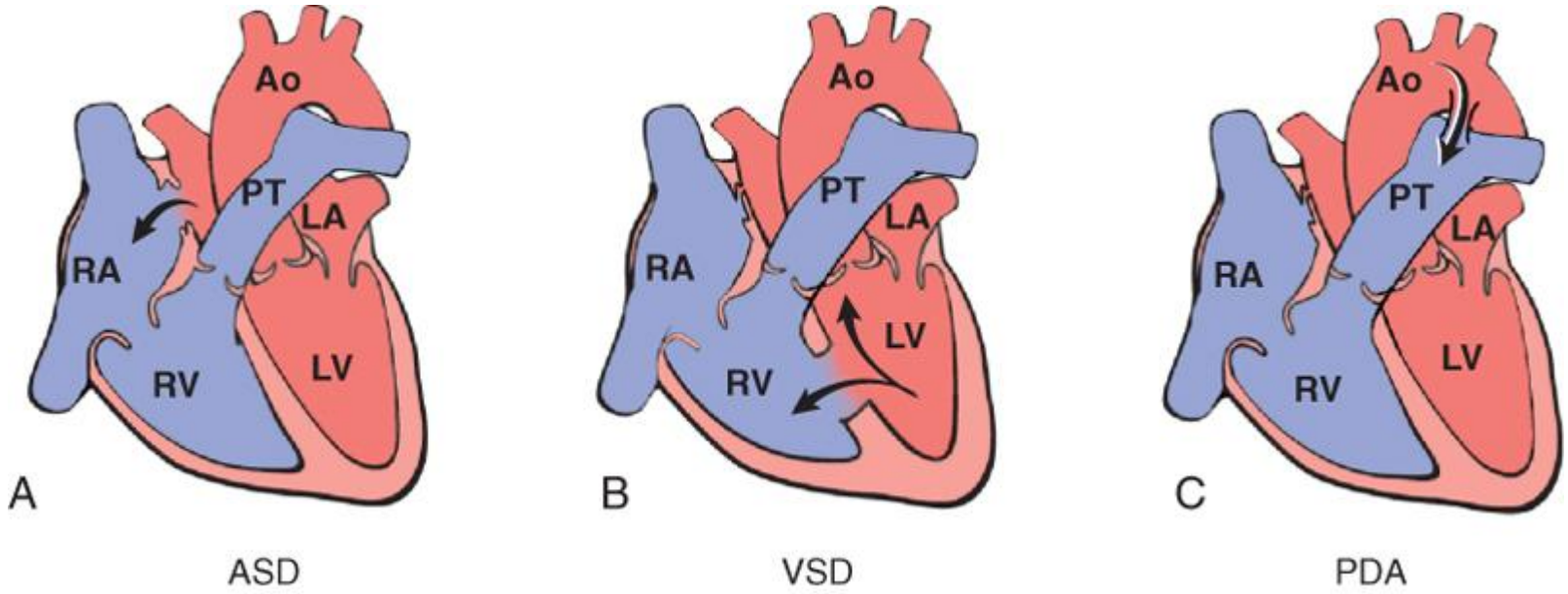
Congenital heart diseases can be subdivided into three major groups:

1. Malformations causing a ***left-to-right*** *shunt*
2. Malformations causing a ***right-to-left*** *shunt* (cyanotic congenital heart diseases)
3. Malformations causing ***obstruction***

Left-to-Right Shunts

- *left-to-right shunts* increase pulmonary blood flow and **are not associated** (at least initially) with cyanosis.
- They expose the low-pressure, low-resistance pulmonary circulation to increased pressure and volume, resulting in **right ventricular hypertrophy and-eventually-right-sided failure**.

- They represent the most common type of congenital cardiac malformation
- They include
  1. atrial septal defects
  2. ventricular septal defects
  3. *patent ductus arteriosus*.



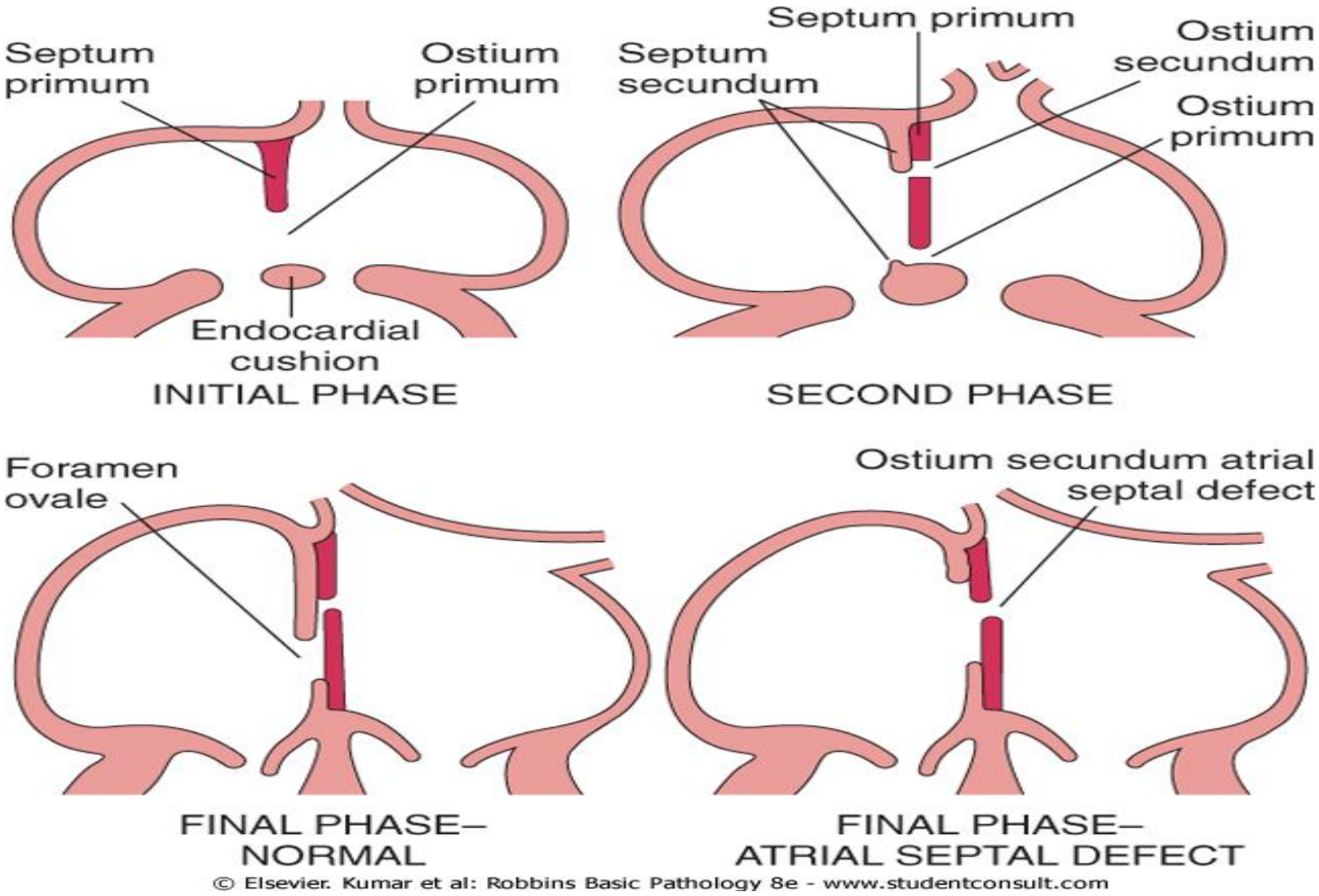
- Cyanosis is not an early feature of these defects,
  - but it can occur late, after prolonged left-to-right shunting has produced pulmonary hypertension sufficient to yield right-sided pressures that exceed those on the left and thus result in a reversal of blood flow through the shunt.

- Such reversal of flow and shunting of unoxygenated blood to the systemic circulation is called ***Eisenmenger syndrome***.

- Once significant pulmonary hypertension develops, the structural defects of congenital heart disease are considered irreversible.

- This is the rationale for early intervention, either surgical or nonsurgical.

Atrial Septal Defects



- Types of ASD:
  - ***ostium secundum ASD:***
    - The most common (90%)
    - occurs when the septum secundum does not enlarge sufficiently to cover the ostium secundum.
  - ***Ostium primum ASDs***
    - less common (5% of cases);
    - occur if the septum primum and endocardial cushion fail to fuse
    - are often associated with abnormalities in other structures derived from the endocardial cushion (e.g., mitral and tricuspid valves).

- **The *sinus venosus***
  - 5% of cases
  - located near the entrance of the superior vena cava
  - associated with frameshift mutations in the NKX2.5 transcription factor.

Clinical Features

- ASD are less common than VSD, but they are the most common defects to be first diagnosed in adults (which are less likely to spontaneously close).

- ASDs initially cause asymptomatic left-to-right shunts. Later cause pulmonary hypertension (less than 10% of patients with uncorrected ASD).

- Ostium primum defects are more likely to be associated with evidence of CHF, in part because of the high frequency of associated mitral insufficiency.
- The objective of surgical closure is to prevent complications:
  - ❖heart failure
  - ❖paradoxical embolization
  - ❖pulmonary hypertension

Ventricular Septal Defects
VSD

- The ventricular septum is normally formed by the fusion of an intraventricular muscular ridge that grows upward from the apex of the heart with a thinner membranous partition that grows downward from the endocardial cushion.

- The basal (membranous) region is the last part of the septum to develop and is the site of approximately 90% of VSDs.

- Although more common at birth than ASDs, most VSDs close spontaneously in childhood, so that the overall incidence in adults is lower than that of ASDs.

- 30% of VSDs occur in isolation; more commonly, they are associated with other cardiac malformations.

Clinical Features

- Small VSDs may be asymptomatic, and those in the muscular portion of the septum may close spontaneously during infancy or childhood.

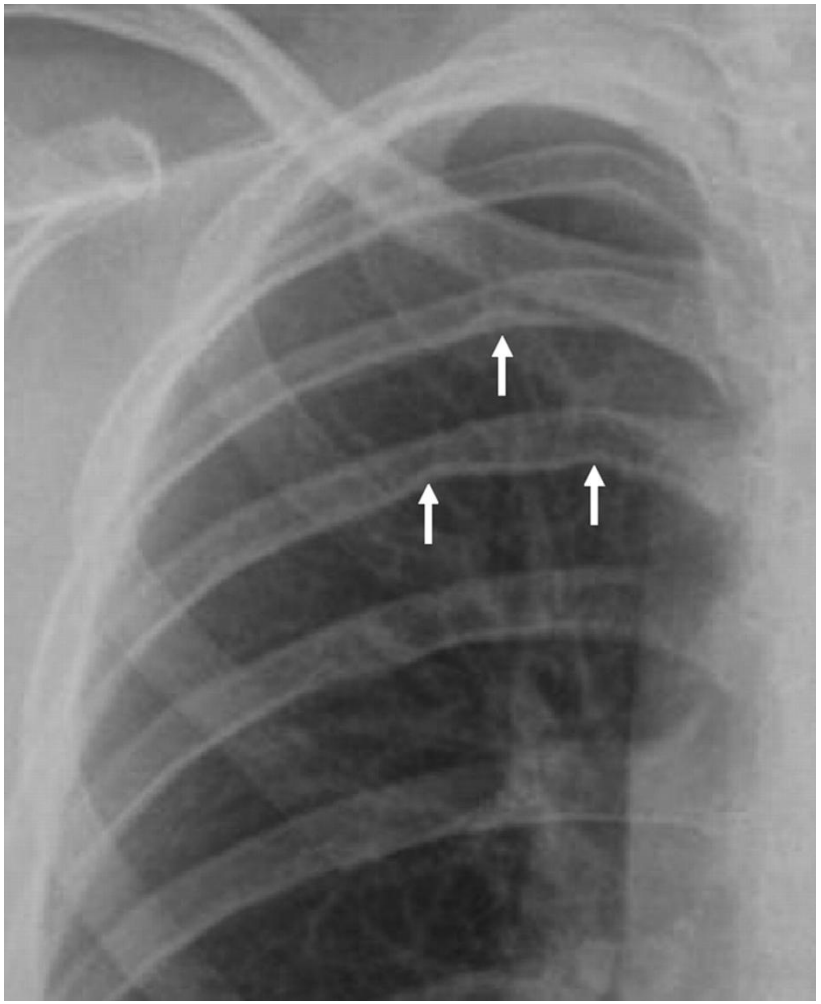
- Larger defects, however, cause a severe left-to-right shunt, often complicated by pulmonary hypertension and CHF.

- Progressive pulmonary hypertension, with resultant reversal of the shunt and cyanosis, occurs earlier and more frequently in patients with VSDs than in those with ASDs; hence, early surgical correction is indicated for such lesions.

- Small- or medium-sized defects that produce jet lesions in the right ventricle are also prone to superimposed infective endocarditis.

- 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.

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