Heart and Circulatory System I

Daphne T. Hsu, MD

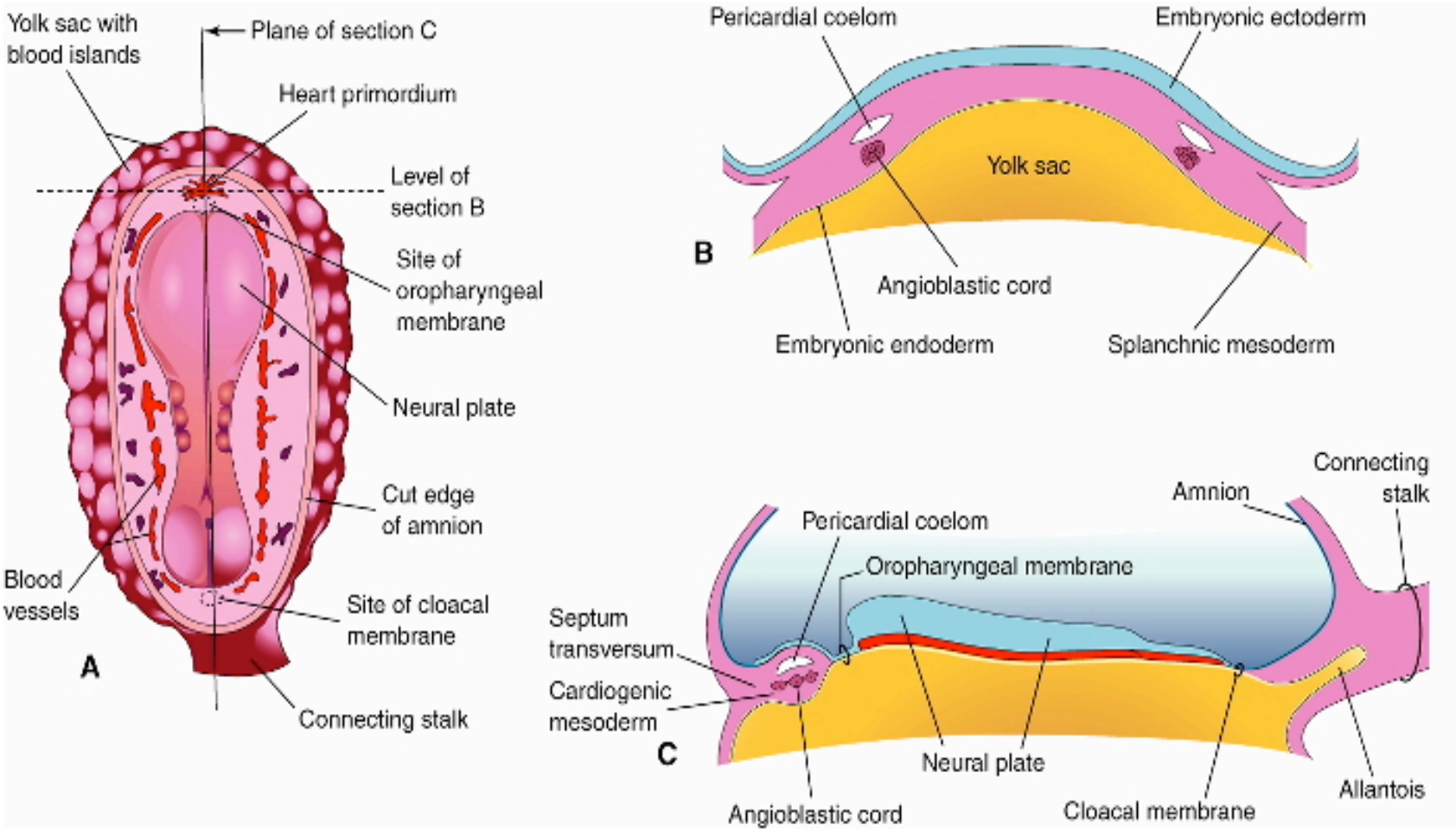
Professor of Clinical Pediatrics

dh17@columbia.edu

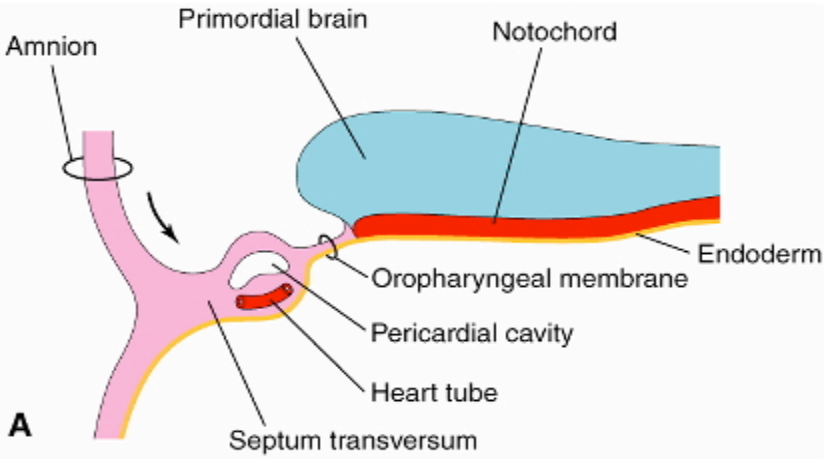
Outline

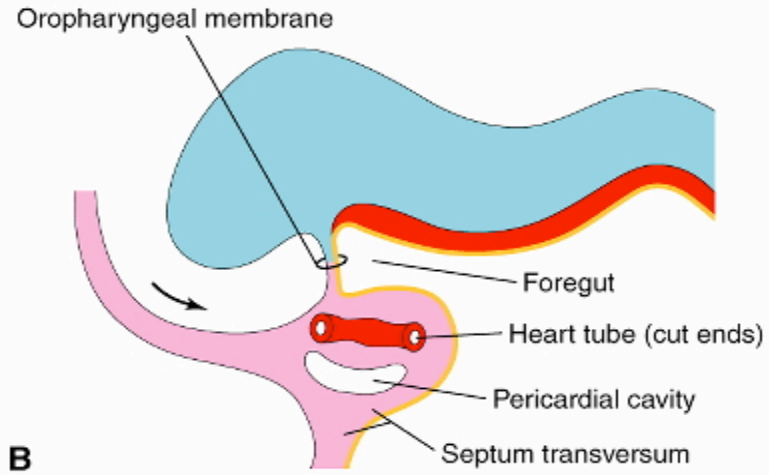
- Vasculogenesis
- Embryonic Folding
- Formation of the Primary Heart Tube
- Looping
- Atrial Septation
- Primitive Ventricular Septum
- Atrioventricular Canal/Endocardial Cushions
- Conotruncal Septation
- Ventricular septation
- Congenital Heart Defects

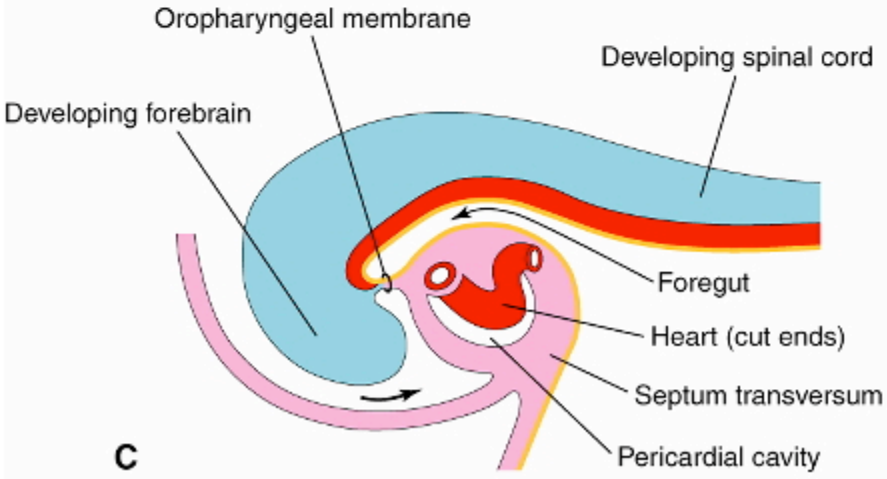
CARDIOVASCULAR SYSTEM: EARLY DEVELOPMENT: WEEK 3



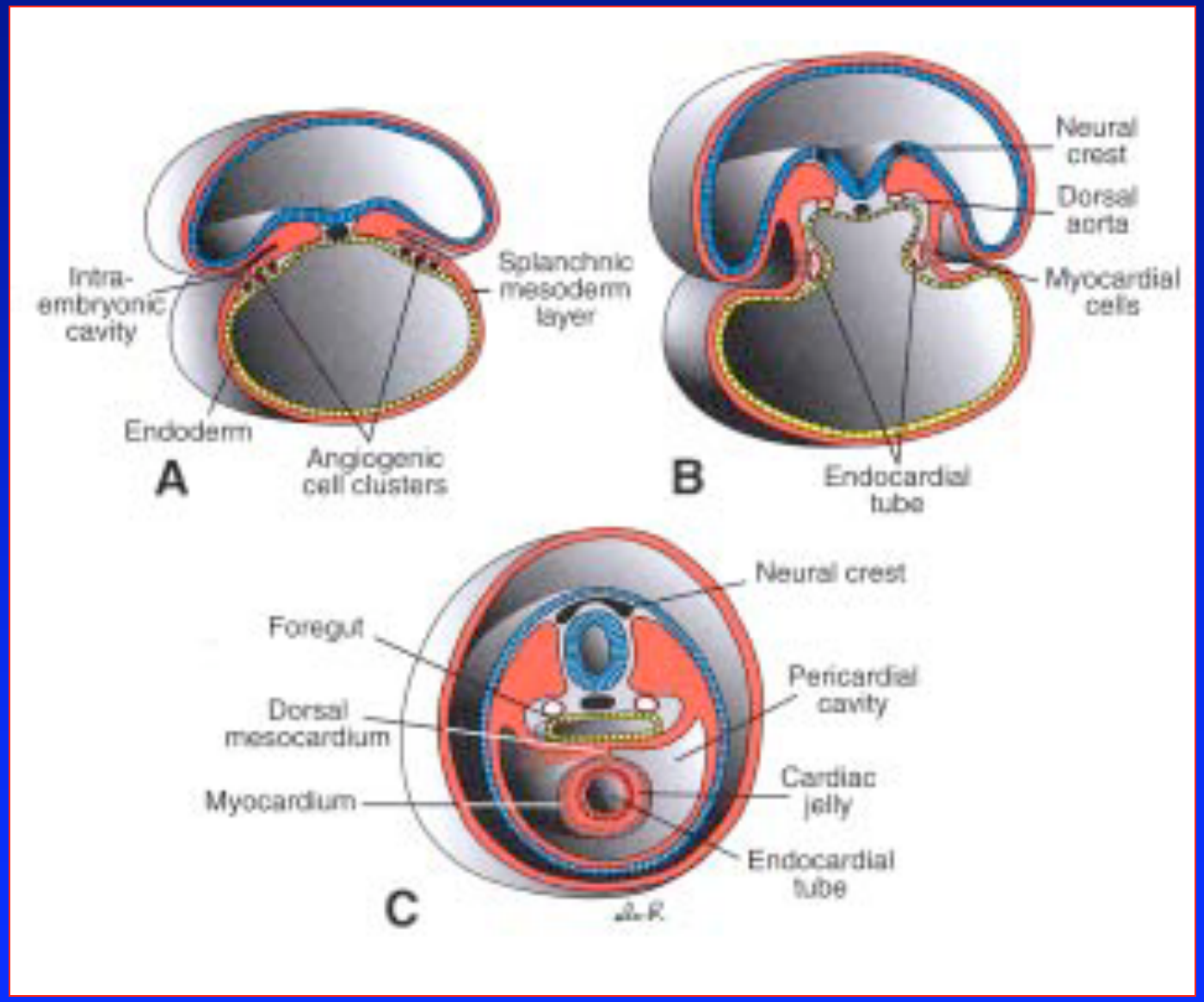
EMBRYONIC FOLDING: WEEK 4



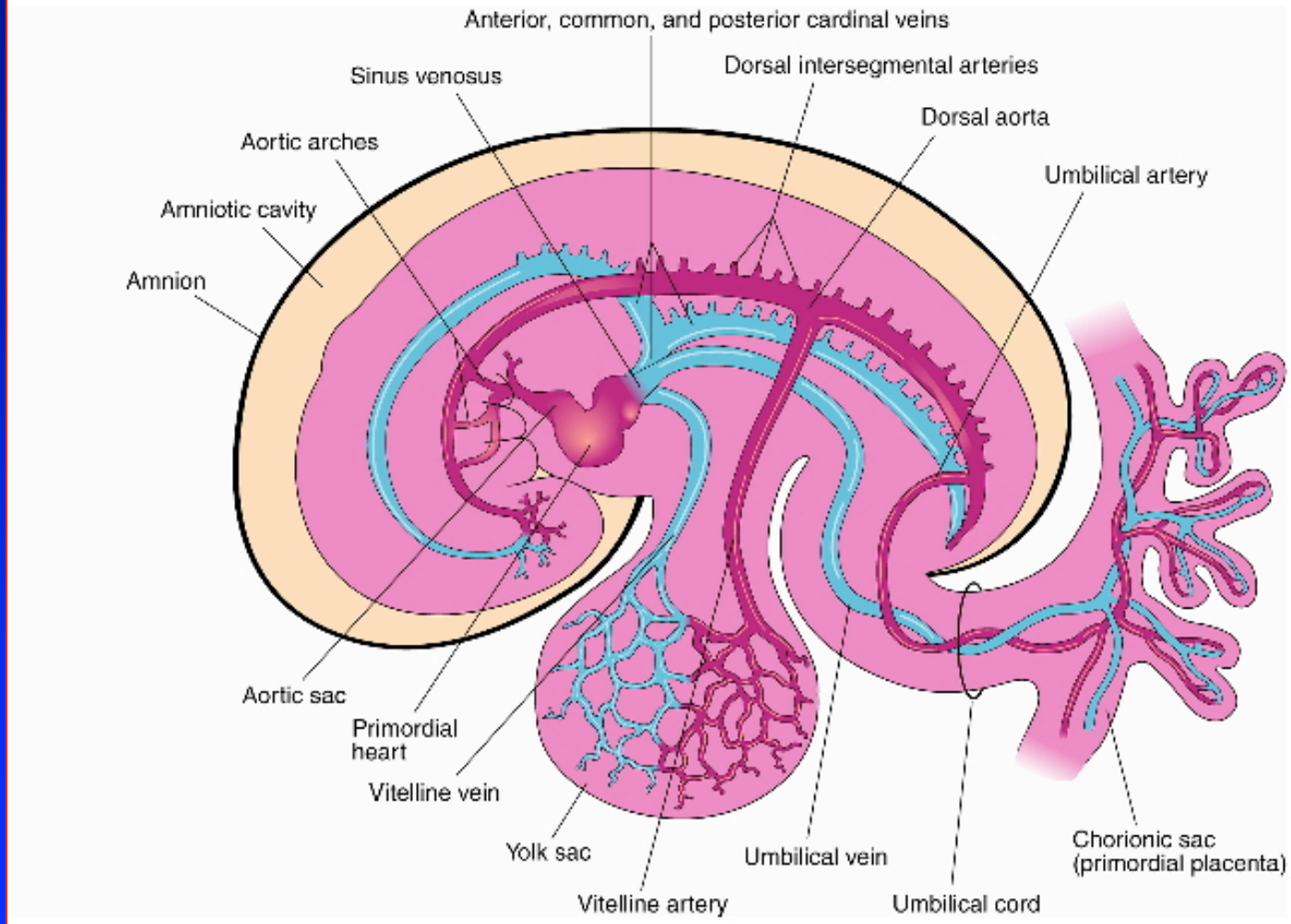




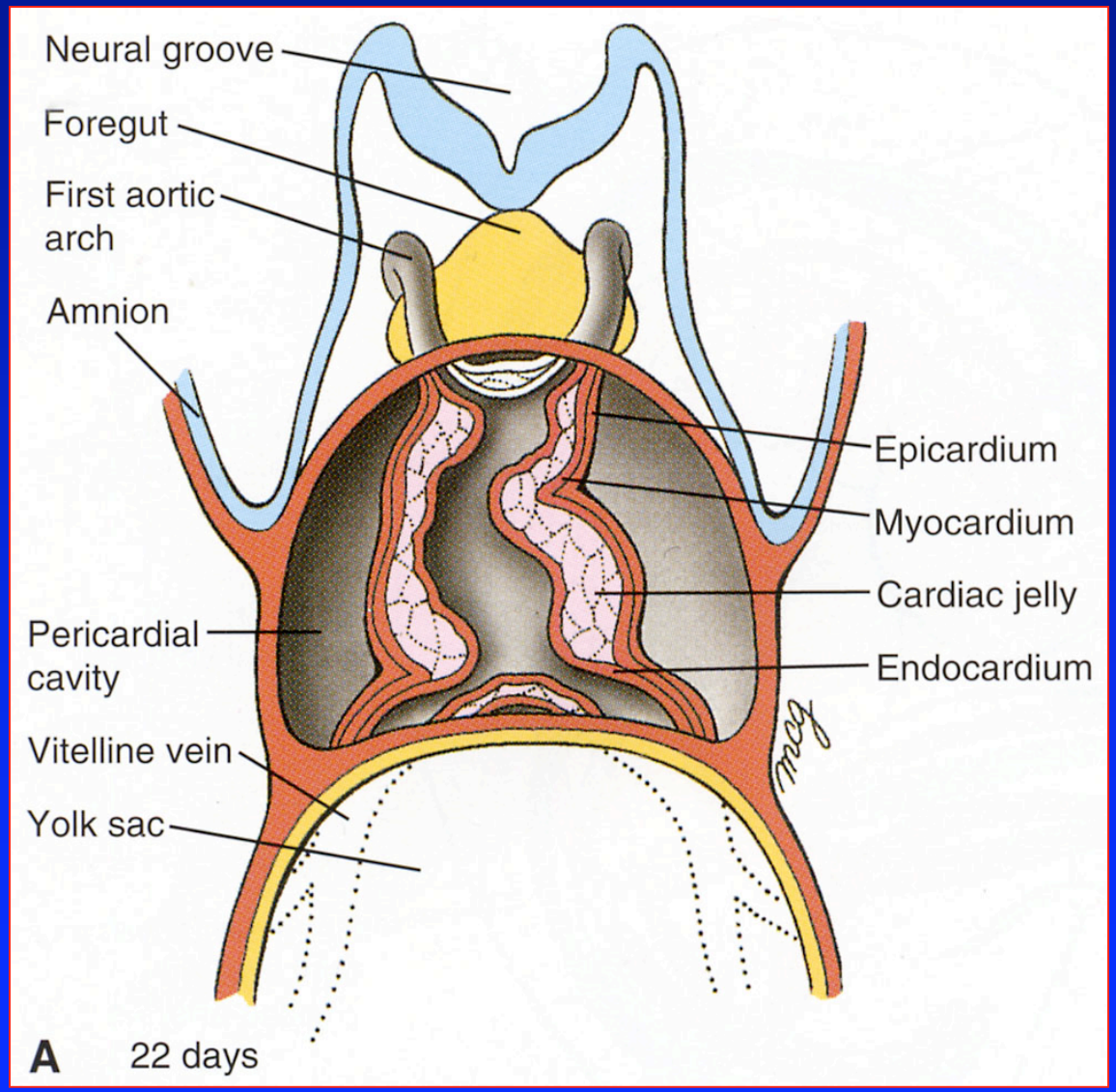
Formation of Heart Tube (17-22 days)



Heart Development: 26 days



PRIMITIVE HEART TUBE: WEEK 4



Cardiac Loop (8-16 somites)

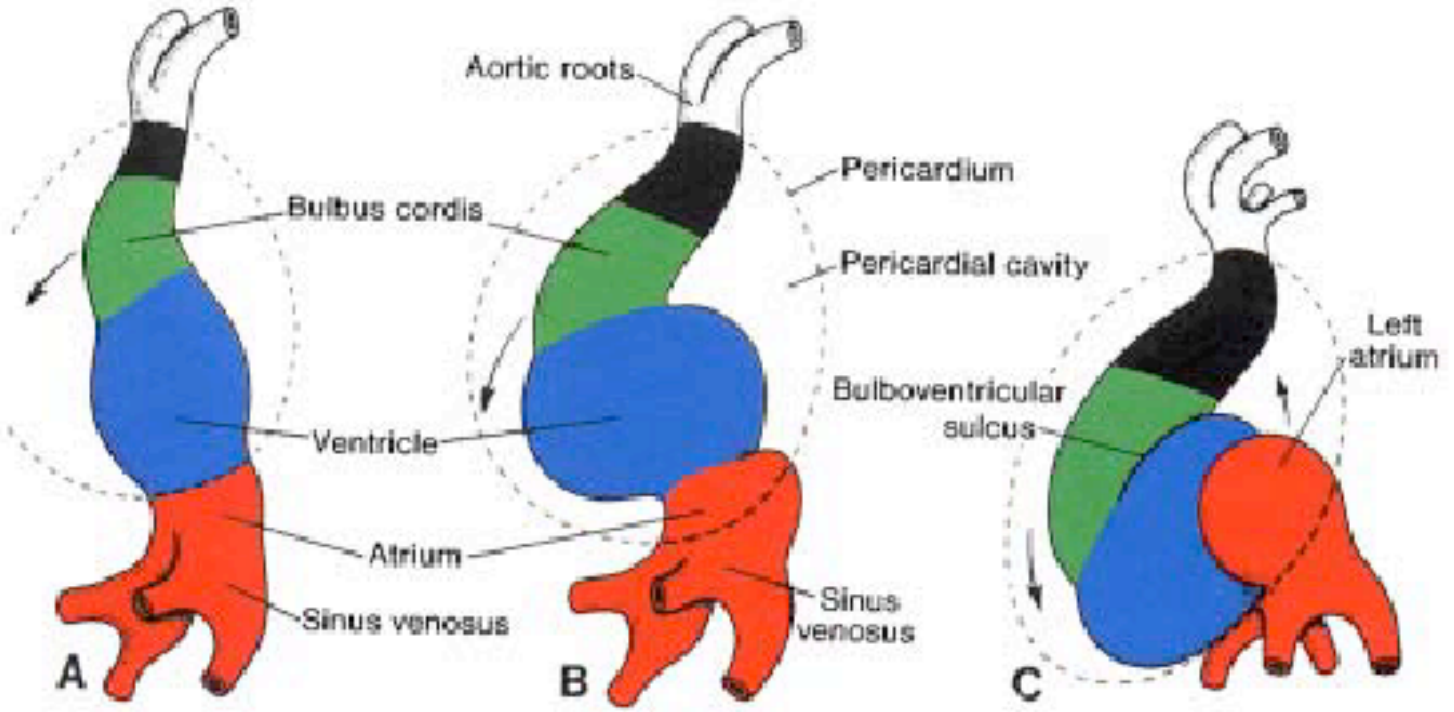
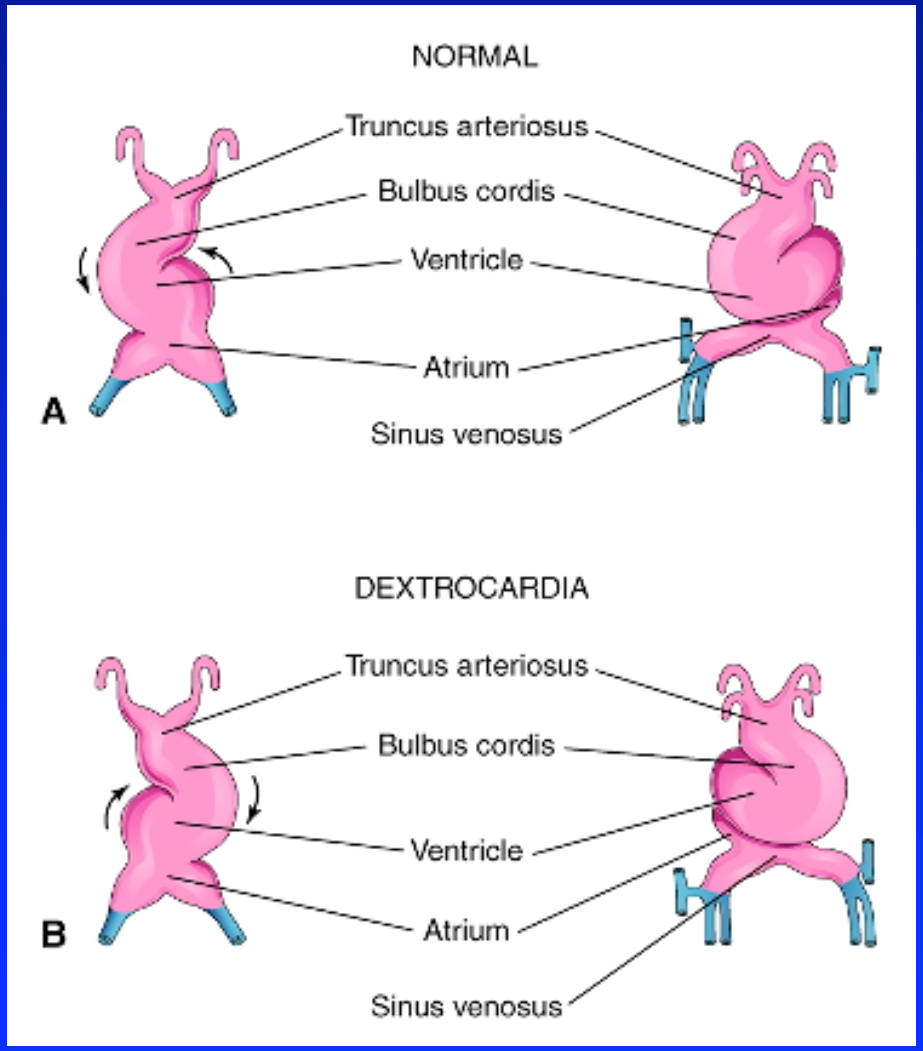


Fig. 8-7. Formation of the cardiac loop. (A) At 8 somites, (B) 11 somites, and (C) 16 somites. Broken line indicates pericardium. Note how the atrium gradually assumes an intrapericardial position.(Modified from Kramer (2).

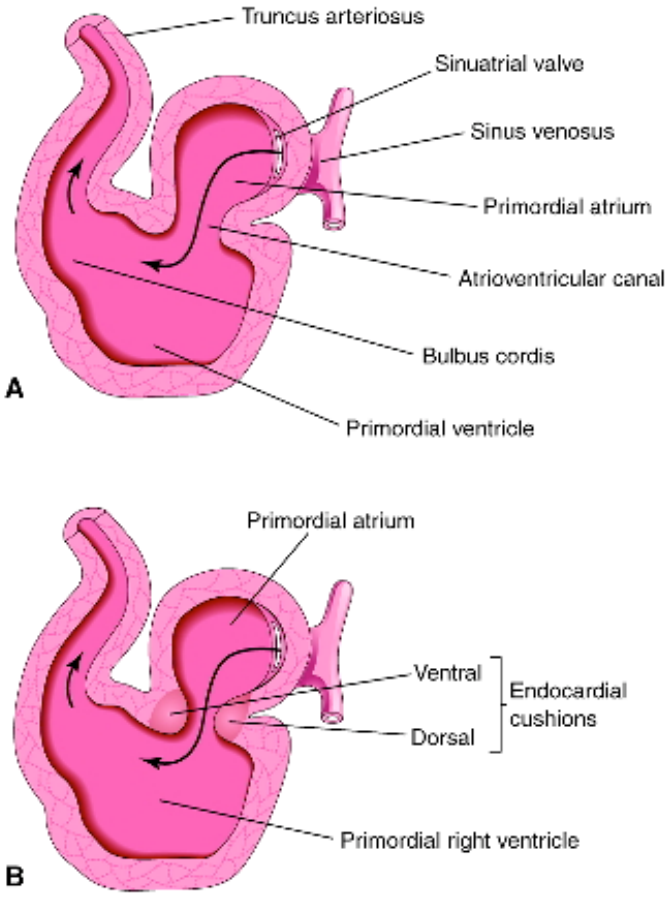
VENTRICULAR LOOPING
END WEEK 4

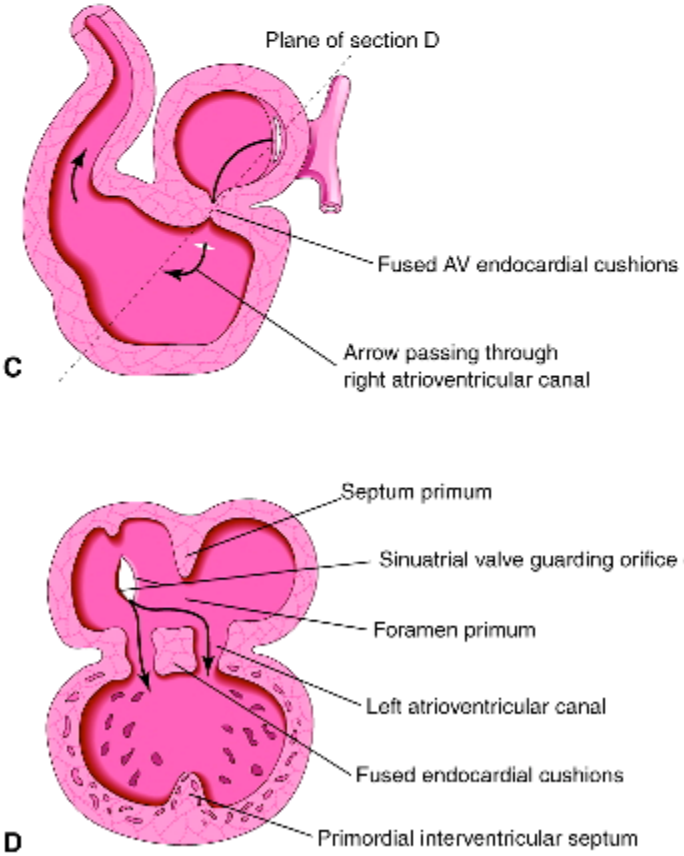


NORMAL : Loop to the RIGHT: Levocardia!

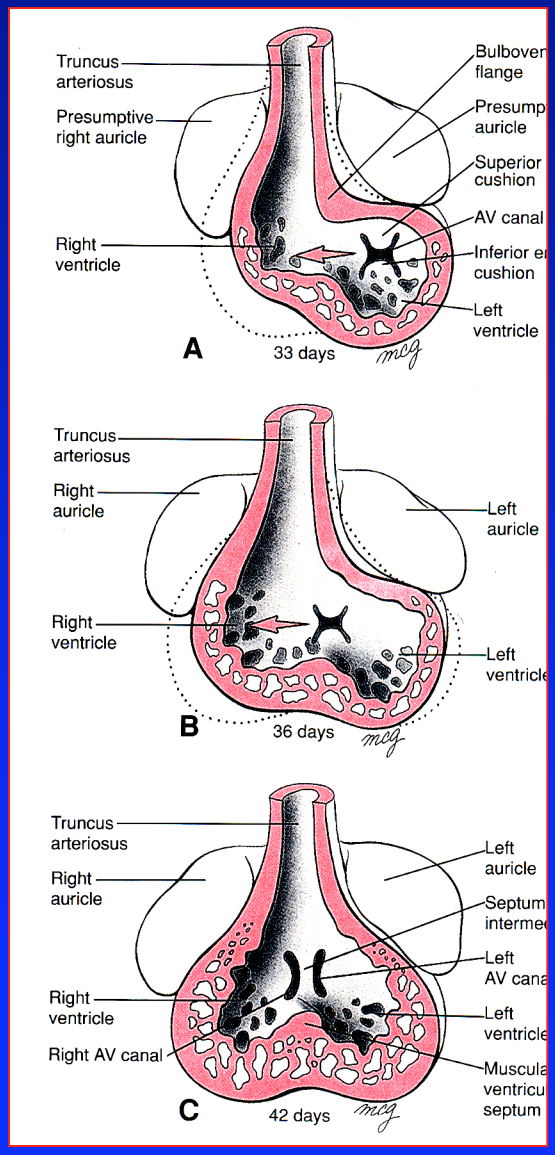
ABNORMAL: Loop to the LEFT: Dextrocardia!

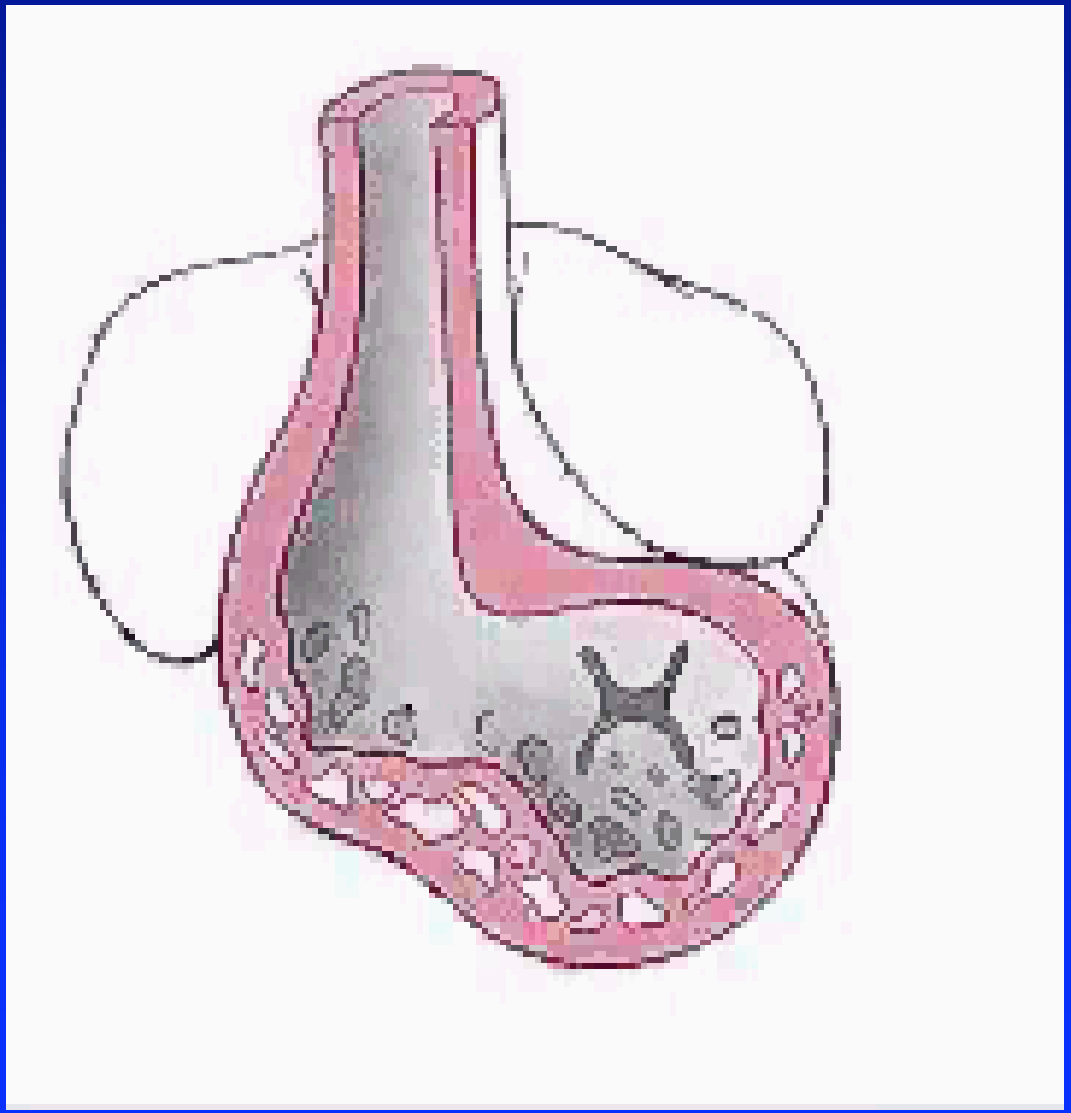
FROM TUBE TO FOUR CHAMBERS
INTERNAL VIEW



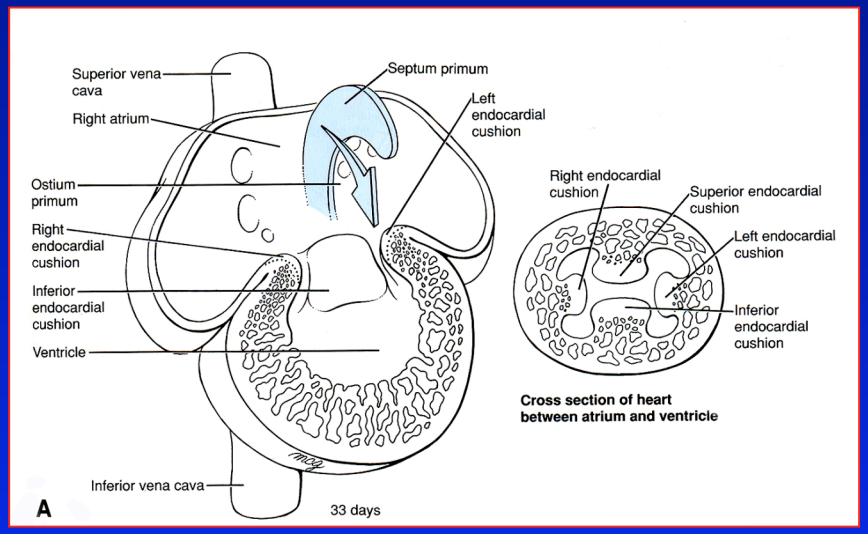


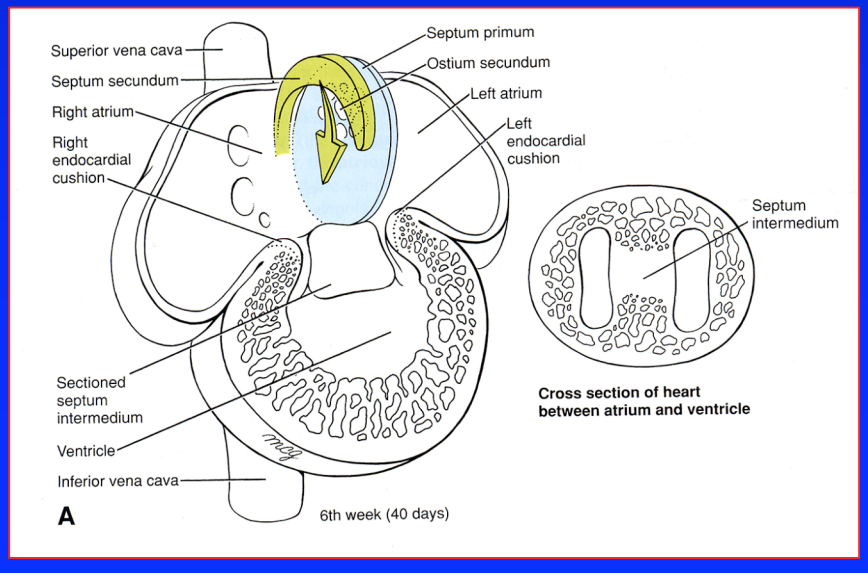
Formation of Primitive Ventricles

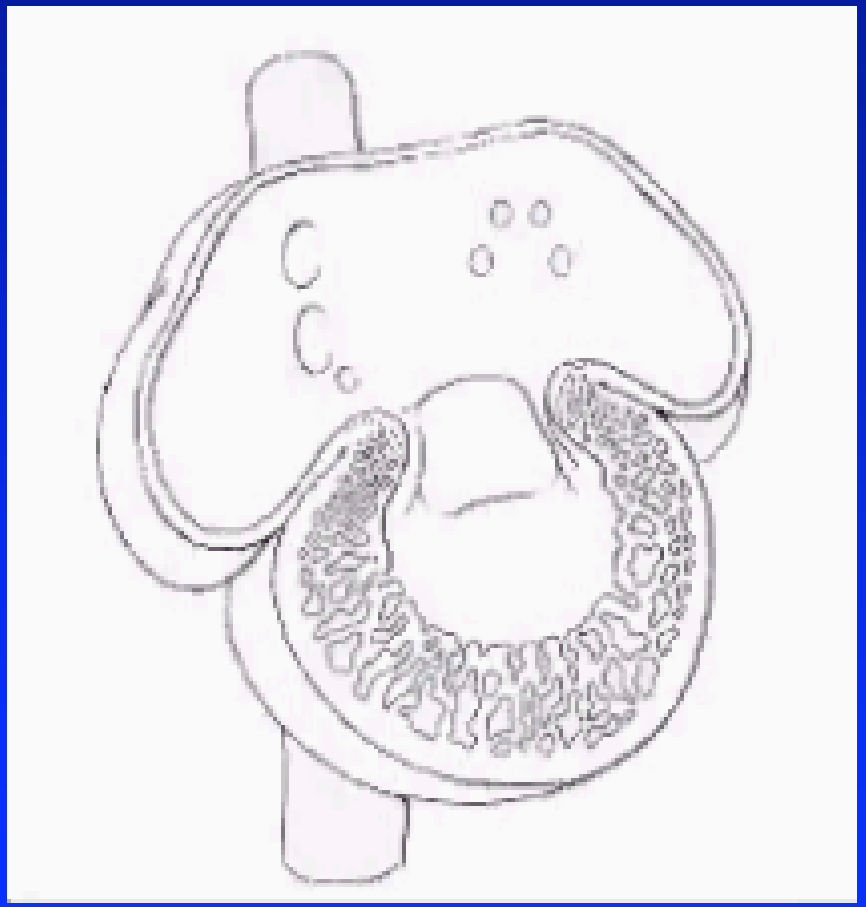




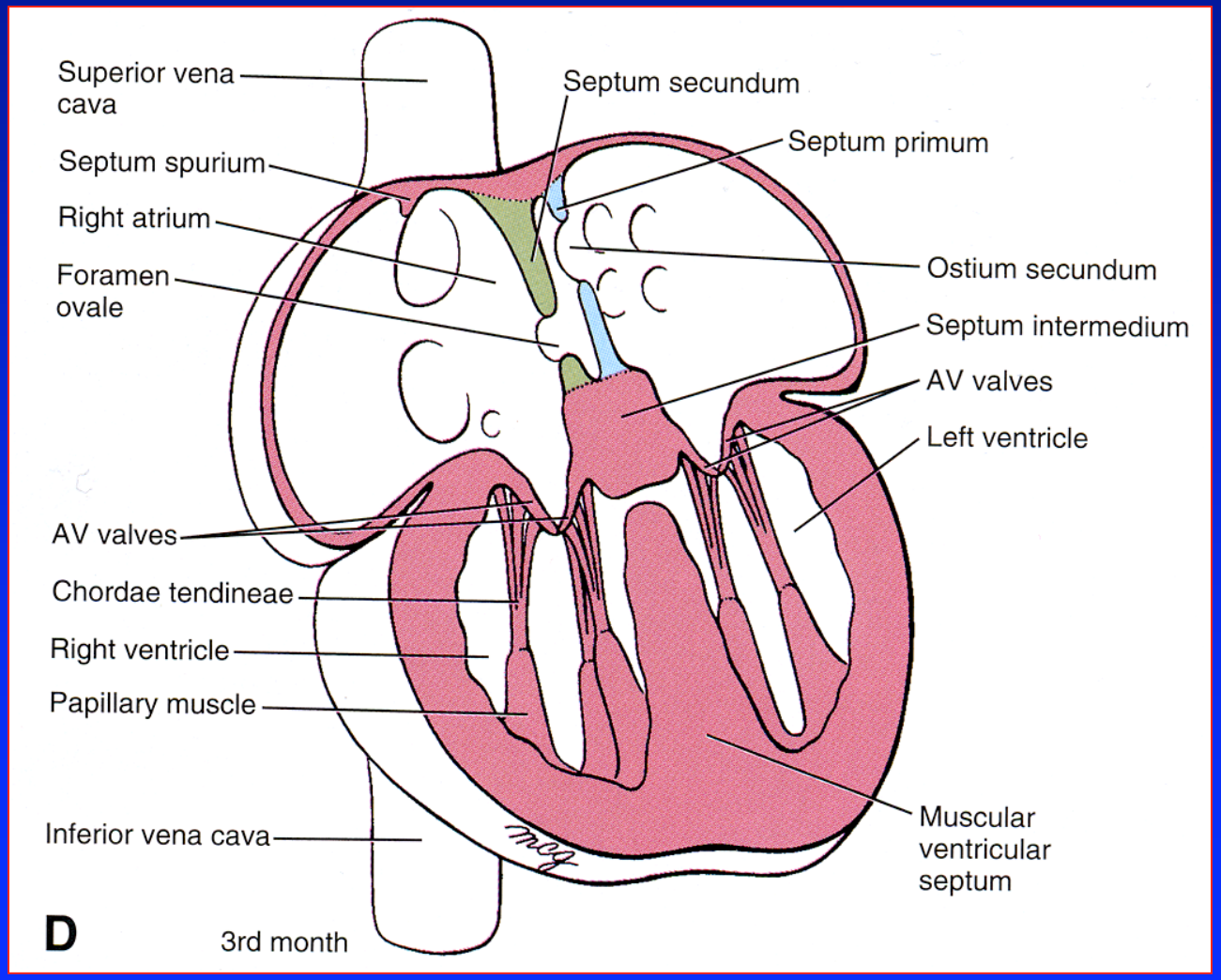
Atrial Septation: 3 Septums
Primum, Secundum, Intermedium







Endocardial Cushion: 80 days



Endocardial Cushions

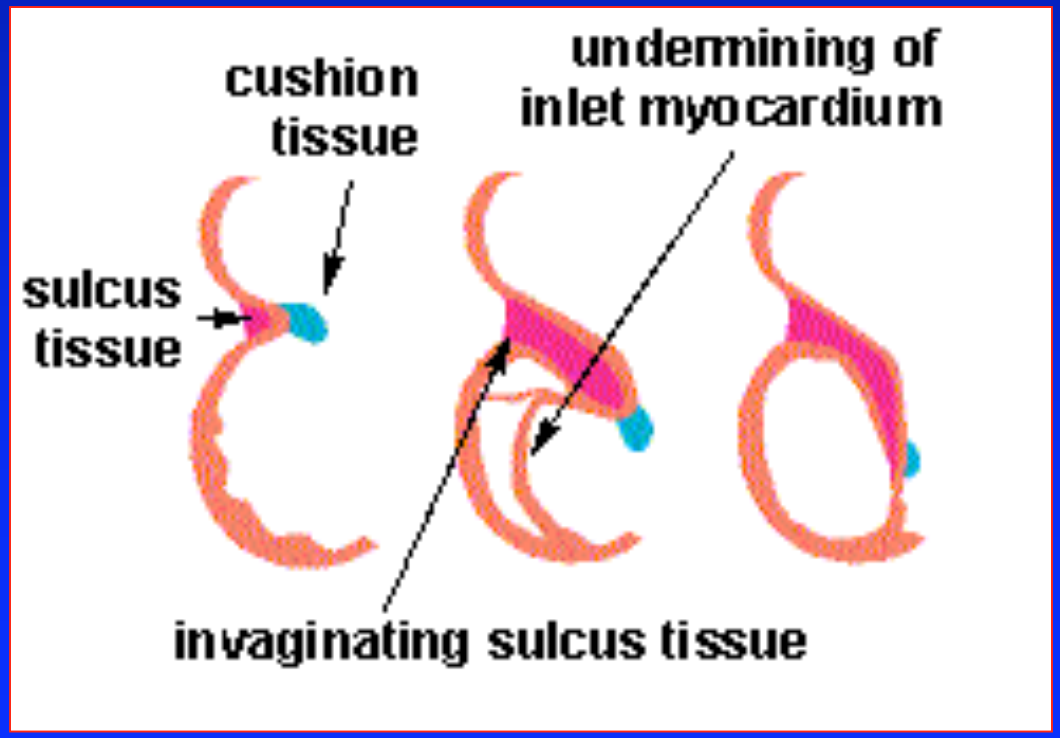
- Atrioventricular Canal: Divide between the atria and ventricles
- Endocardial Cushions: Four tissue expansions found in periphery of AV canal
  - Atrial septation
  - Atrioventricular valve formation: Mitral and Tricuspid Valves
  - Ventricular septation

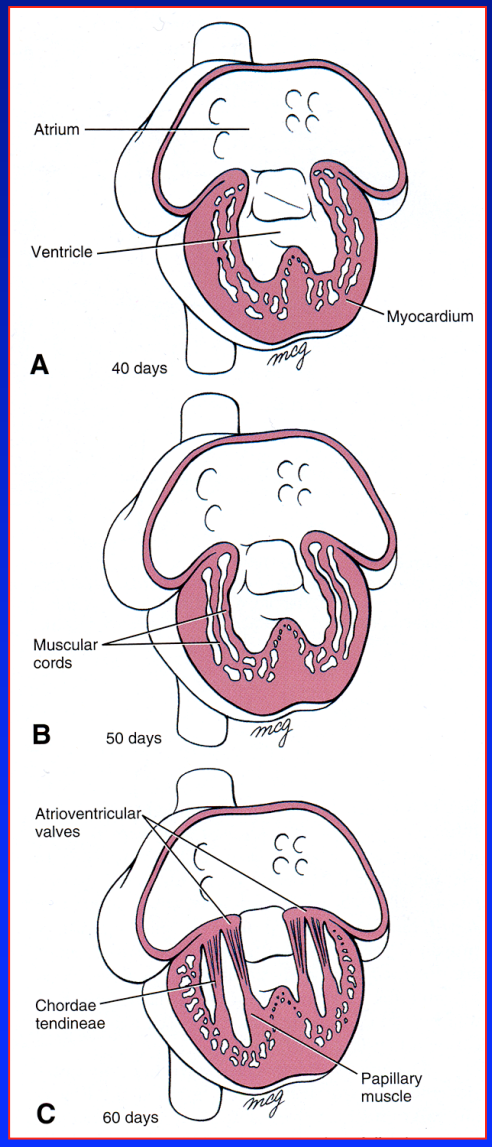
Endocardial Cushions

- **Superior-Inferior cushions**
  - Septum Intermedium
  - Inferior atrial septum
  - Posterior/superior ventricular septum
- **Right and Left Cushions**
  - Ventricular myocardium
  - Mitral valve
  - Tricuspid valve

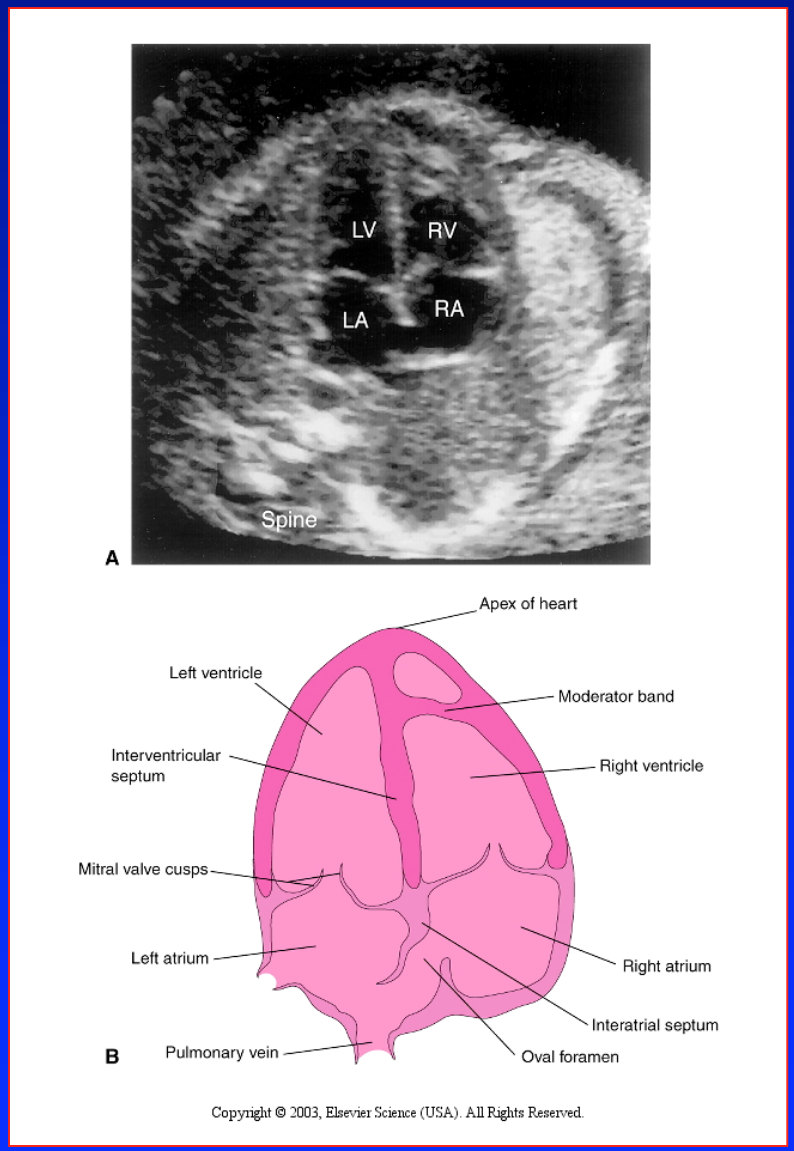
Atrioventricular Valve Formation

- Left and Right Endocardial Cushions

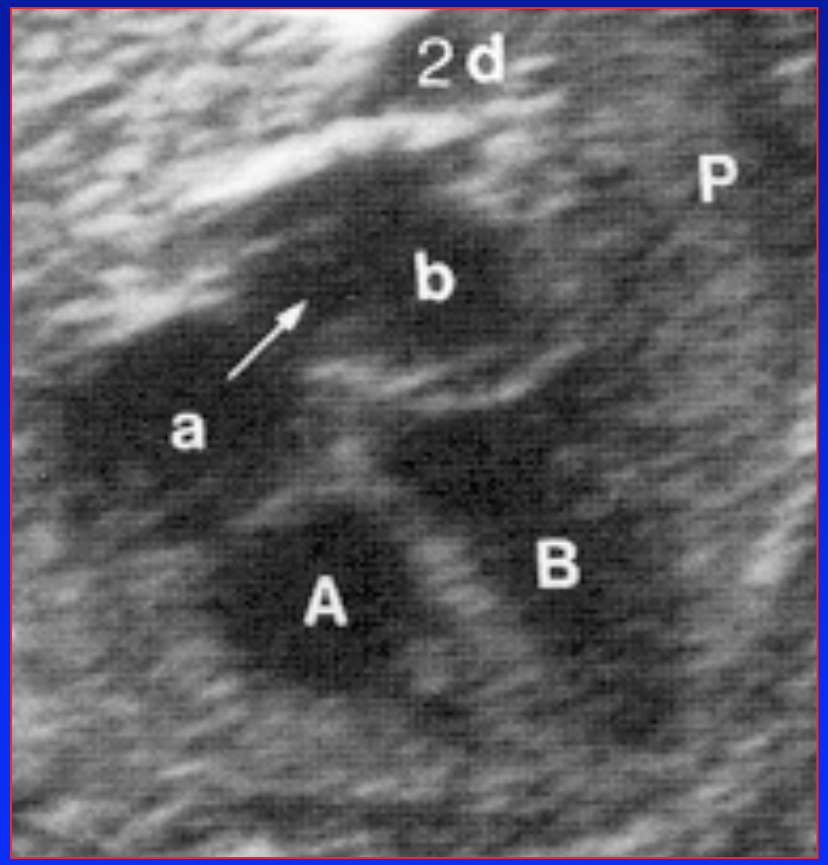




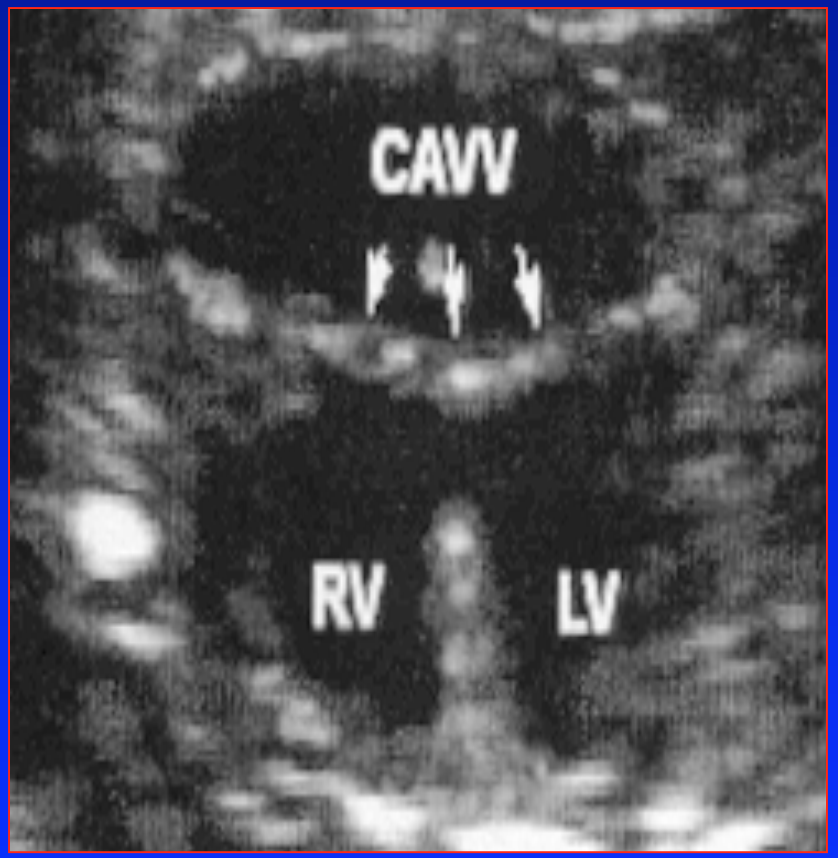
FOUR CHAMBERS- ULTRASOUND VIEW @ 20 wks



Congenital Heart Defect: Endocardial Cushion Defect



Normal

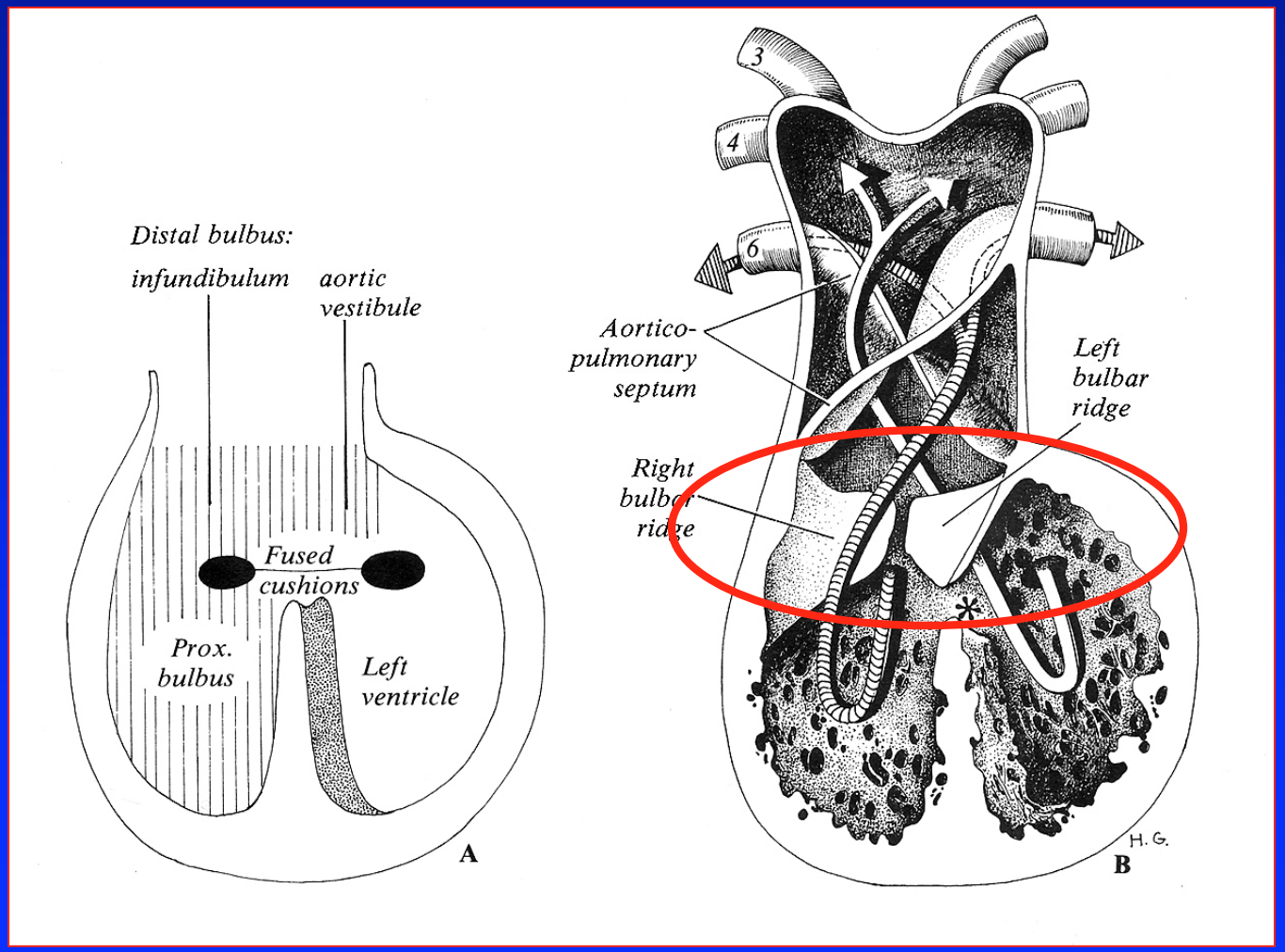


Endocardial Cushion Defect

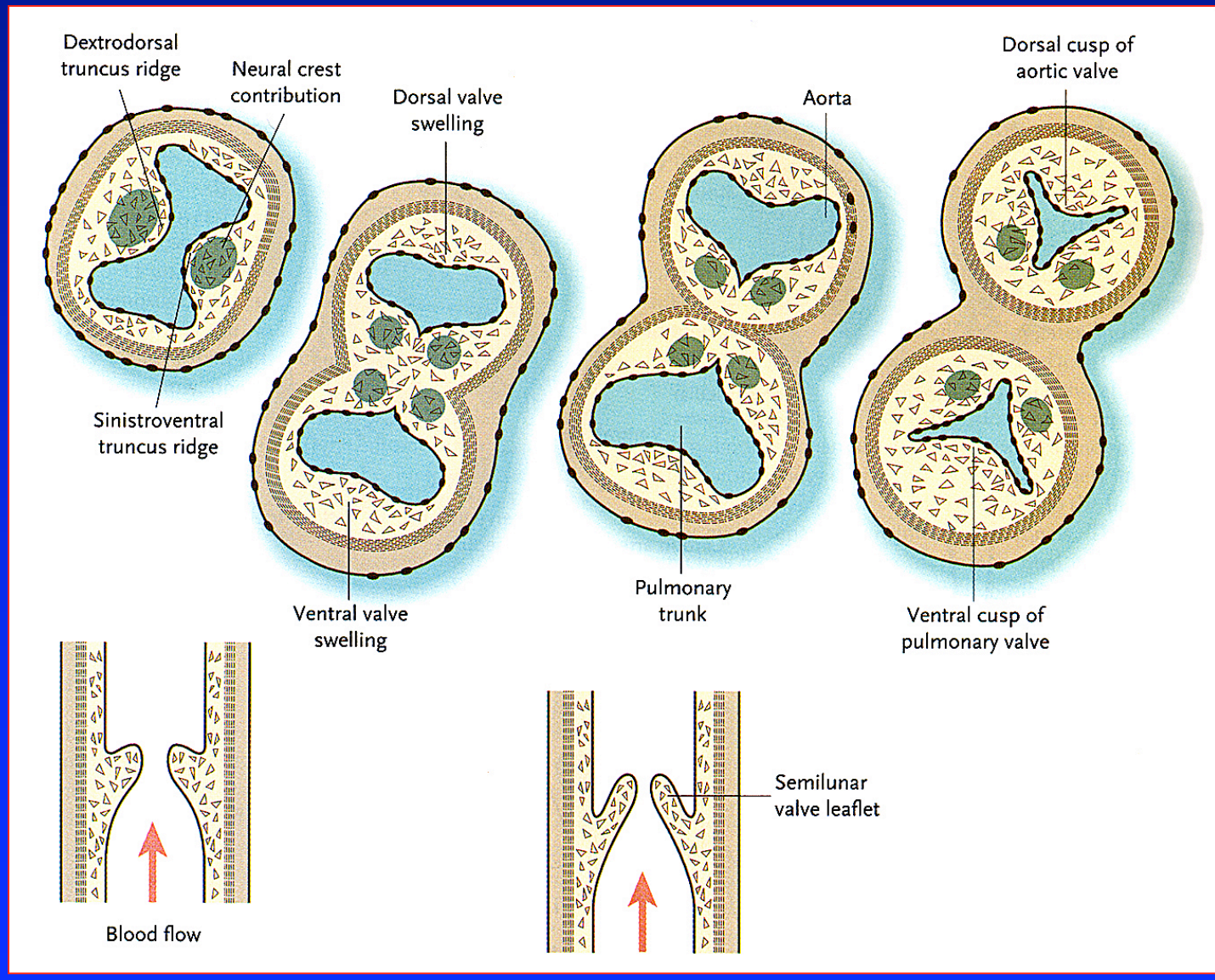
Ventricular Outflow Tracts and Great Arteries

- Truncus Arteriosus: common arterial trunk from the primitive ventricle
- Conus (Bulbus) Cordis: outflow portion of the primitive ventricle
- Bulbar Ridges: Tissue ridges at junction of the conus and truncus
  - Conotruncal septum
  - Semi-lunar valves (aortic and pulmonic)
- Truncal Ridges: Within Truncus
  - Septation of the Aorta and Pulmonary arteries

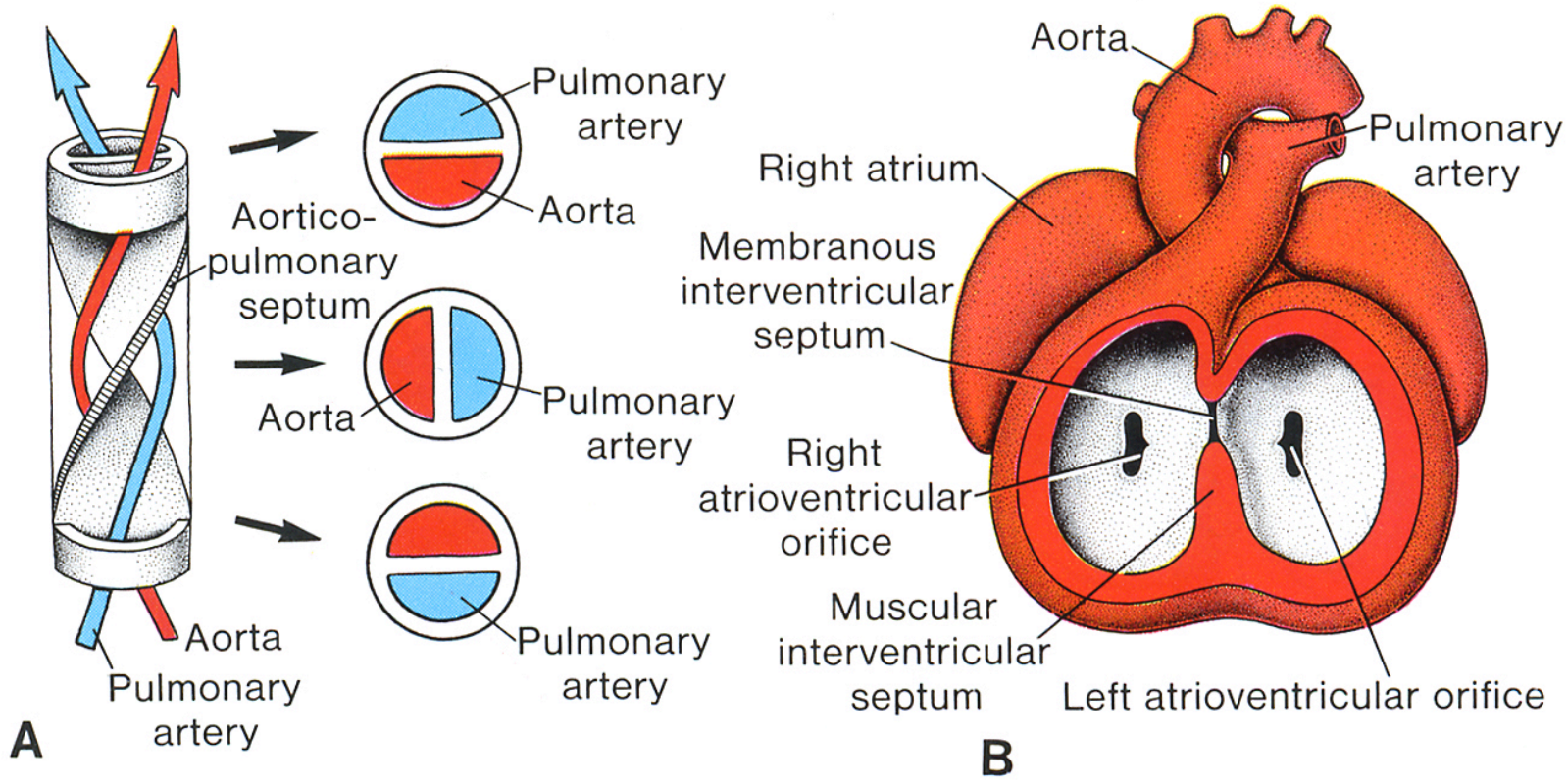
Formation of the Conotruncal Septum



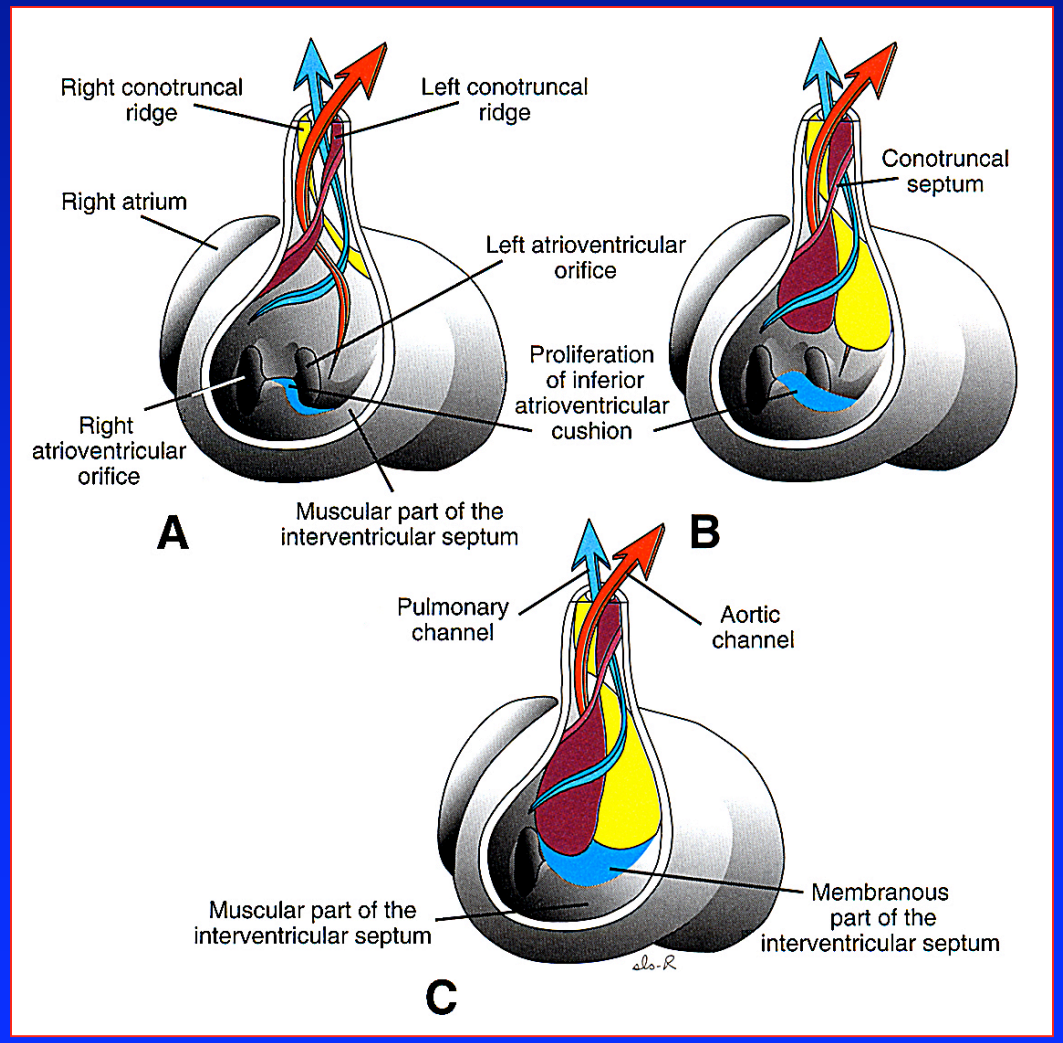
Semilunar Valve Formation

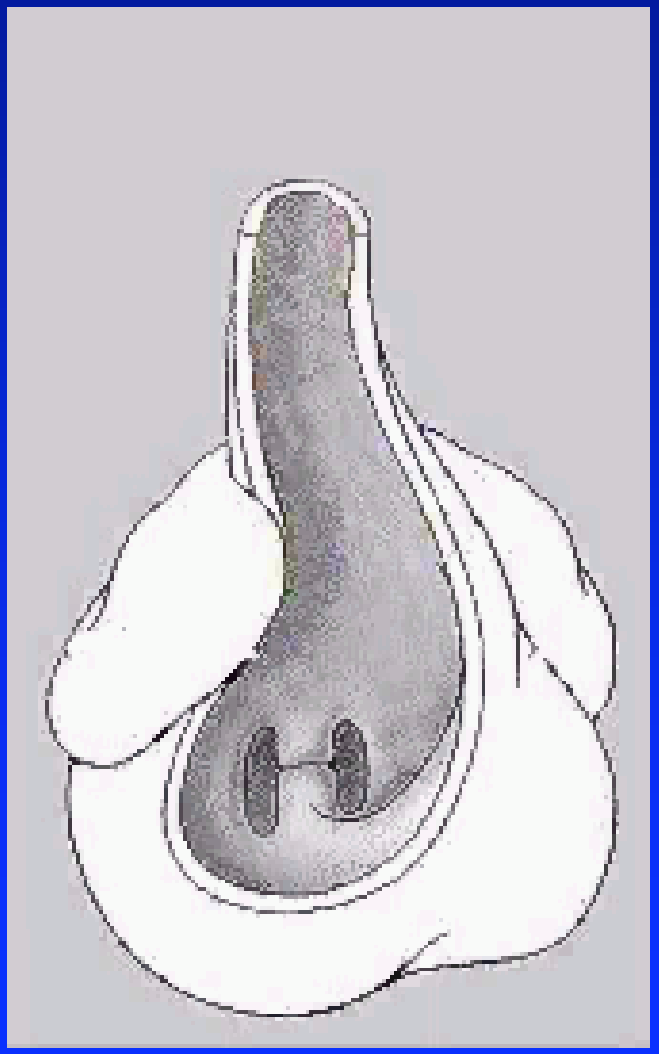


Formation of the Aorta and Pulmonary Artery



Conotruncal Formation

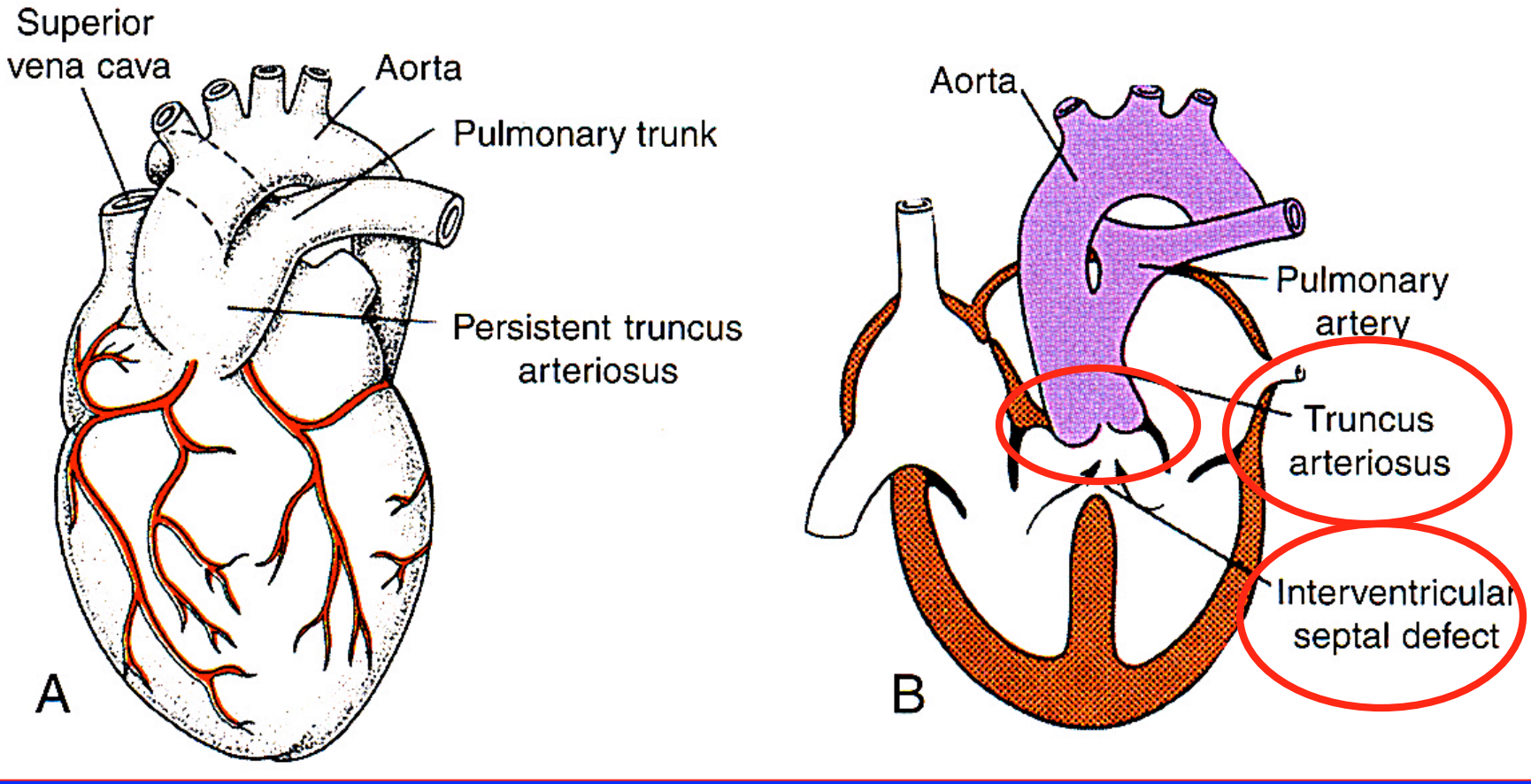




Defects of Conotruncal Septation

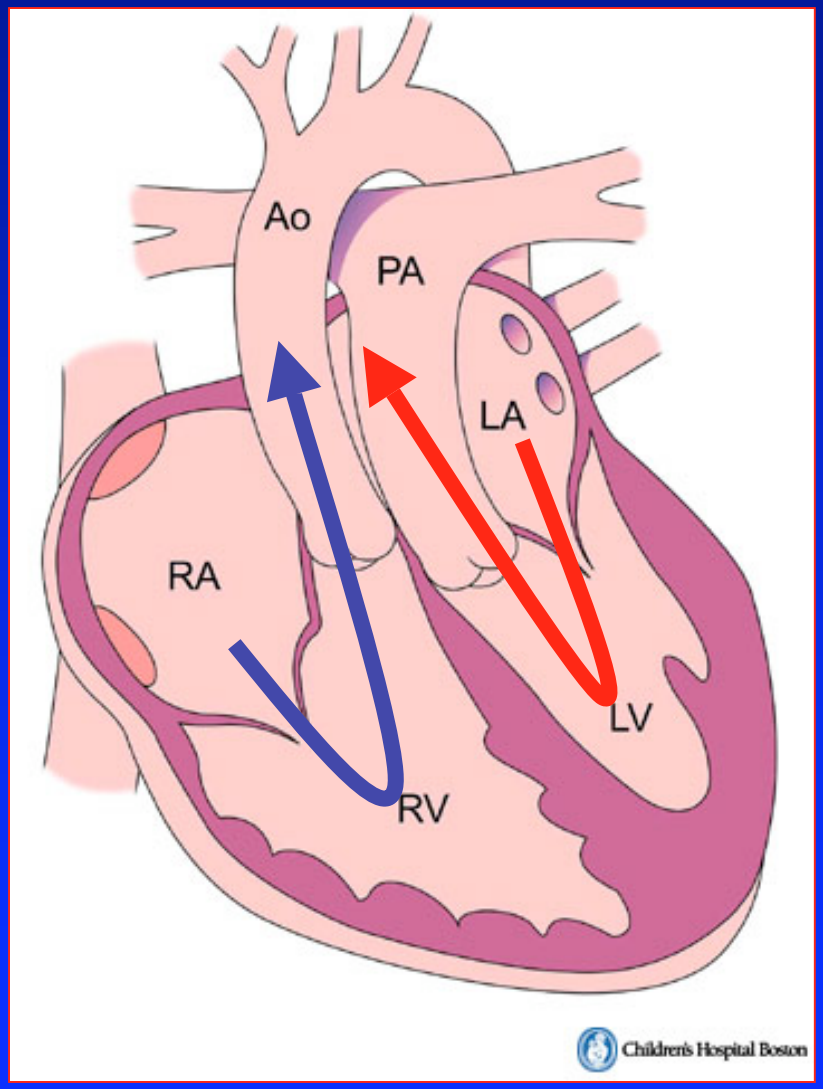
- **Persistent Truncus Arteriosus**
  - Failure of conotruncal septation
- **Transposition of the Great Arteries**
  - Failure of helical twisting during truncal septation
- **Tetralogy of Fallot**
  - Malalignment of conoventricular septum

Persistent Truncus Arteriosus

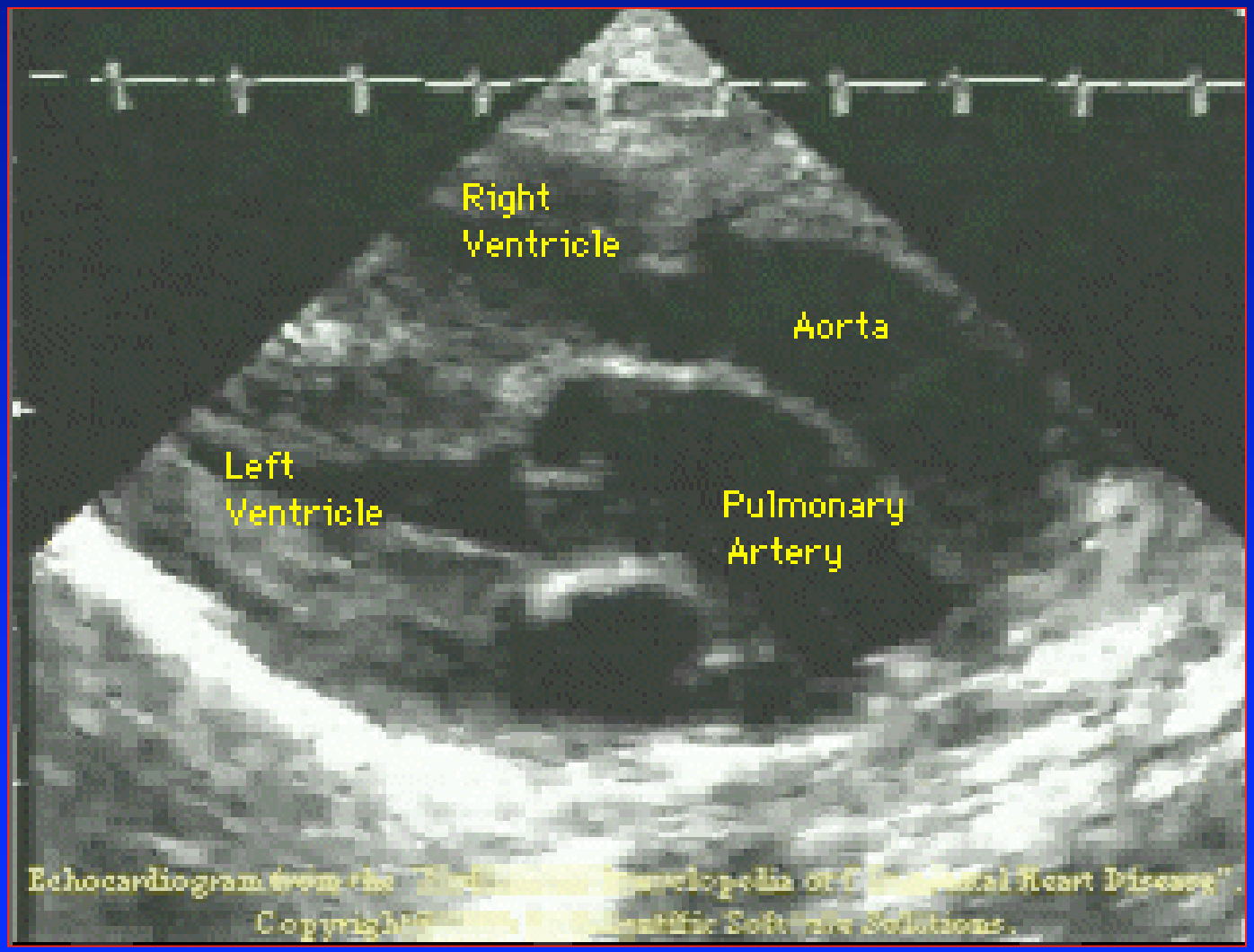


Transposition of the Great Arteries

- Failure of helical twisting during truncal septation
- Aorta arises from RV
- Pulmonary artery arises from LV
- VSD in 20% of cases



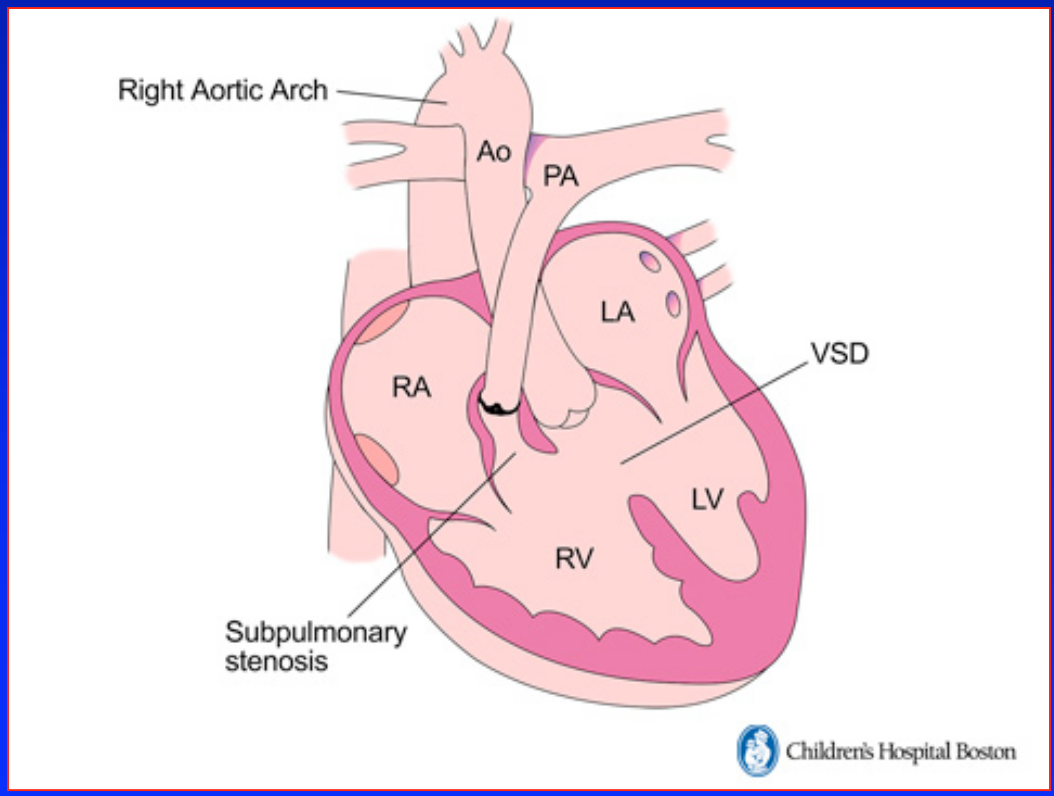
Transposition of the Great Arteries



Tetralogy of Fallot

- **Malalignment of conoventricular septum**

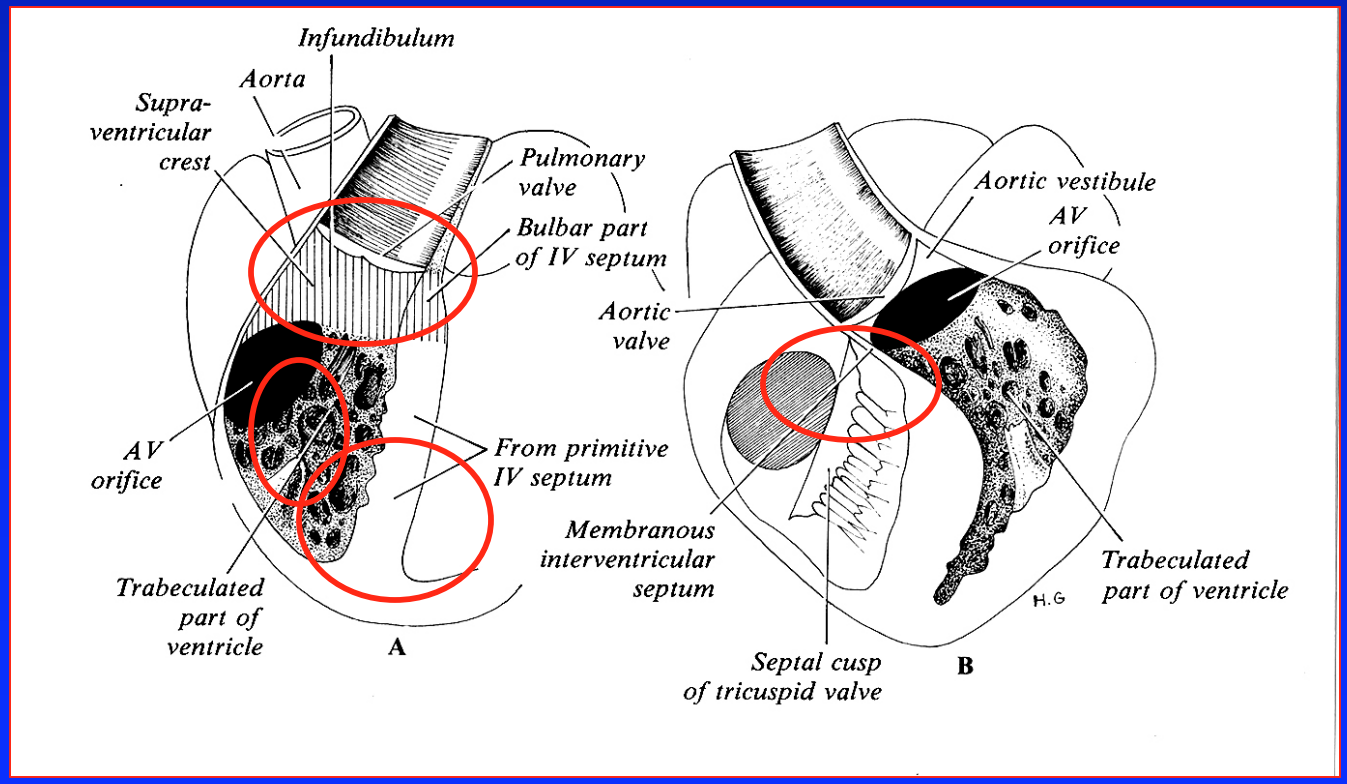
1. **Ventricular septal defect**
2. **Aortic valve override**
3. **Pulmonary stenosis**
4. **Right ventricular hypertrophy**



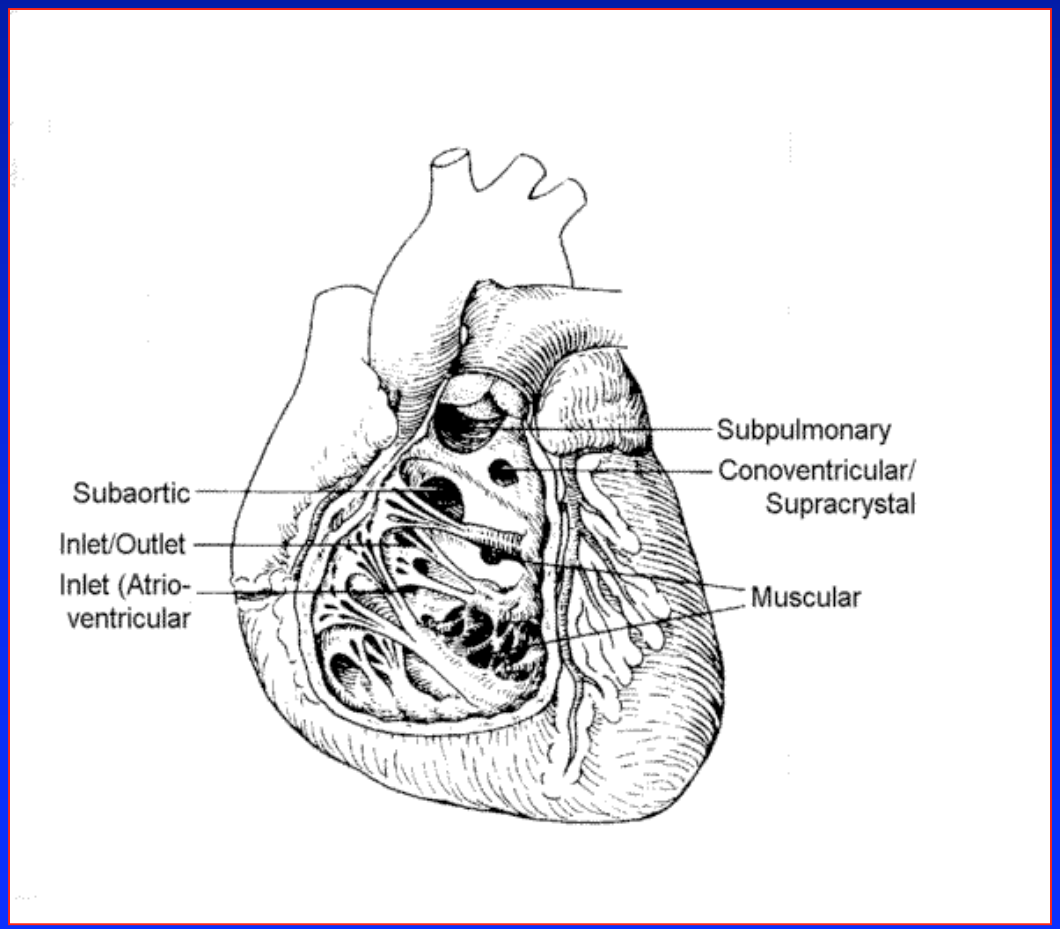
Ventricular Septum

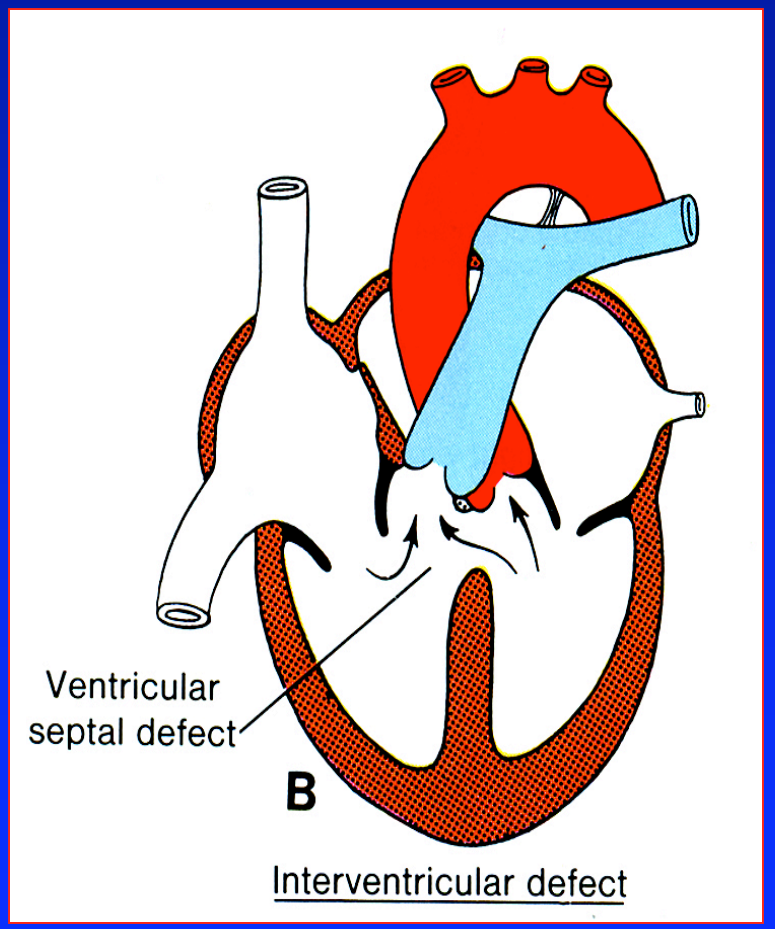
Primitive Septum **Conotruncus**

Endocardial Cushion **Membranous**



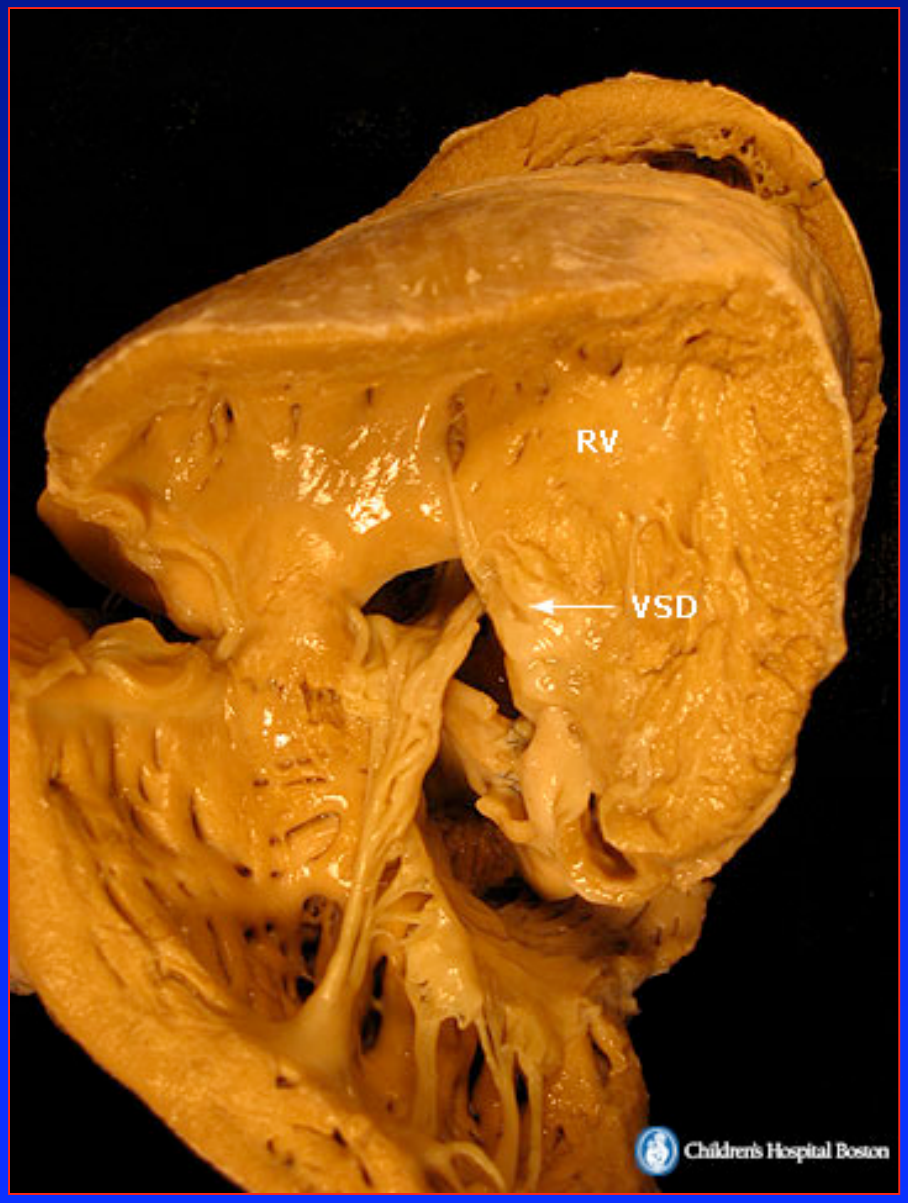
Ventricular Septal Defect (VSD)

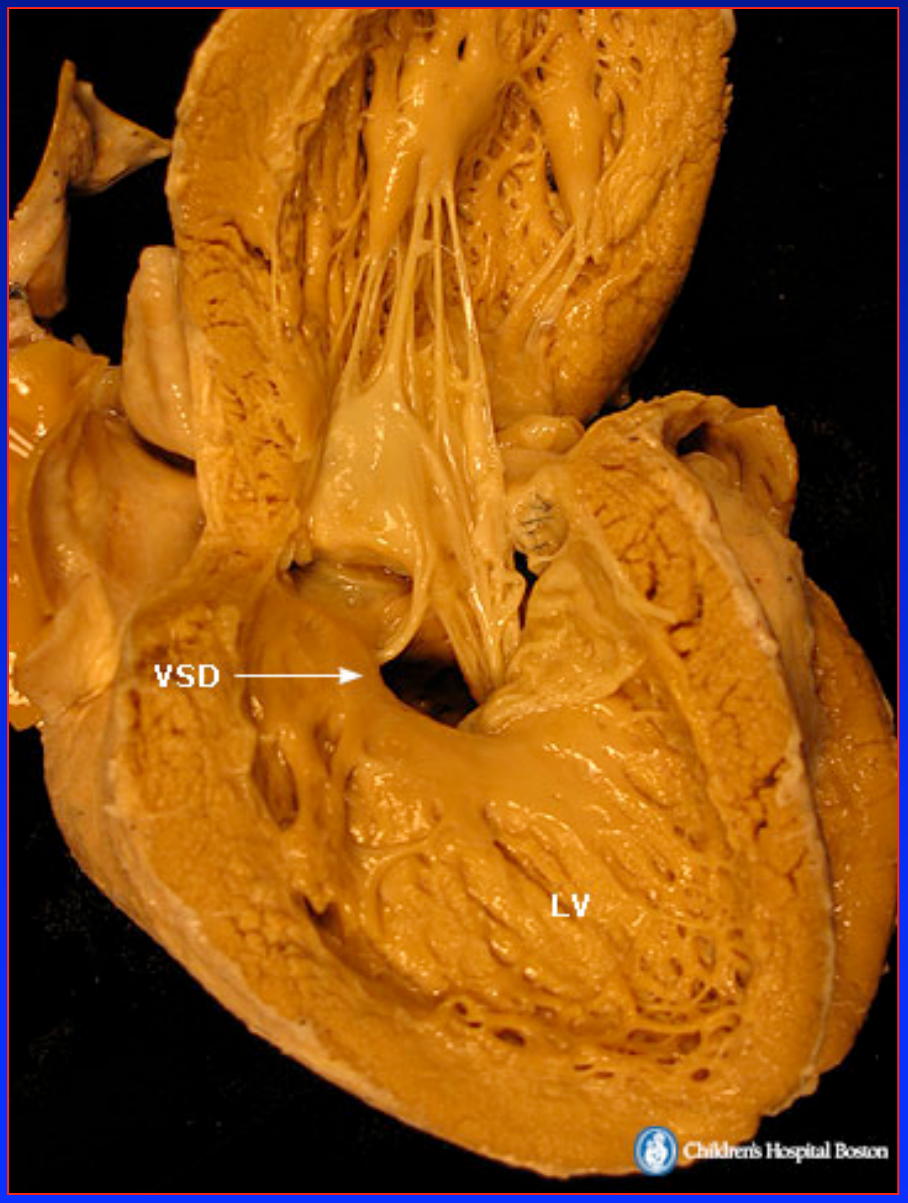




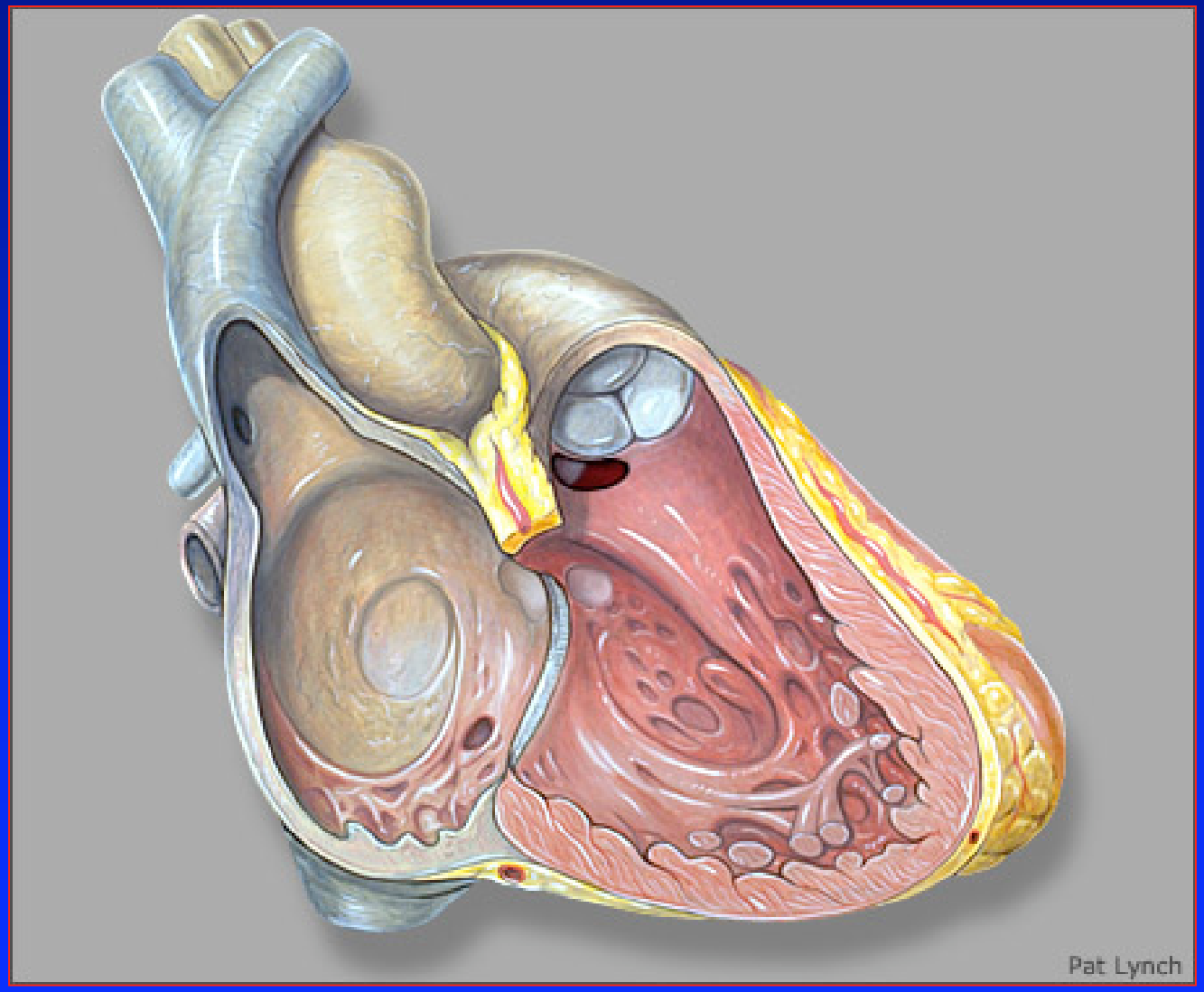
Muscular VSD

Endocardial Cushion (Inlet VSD)

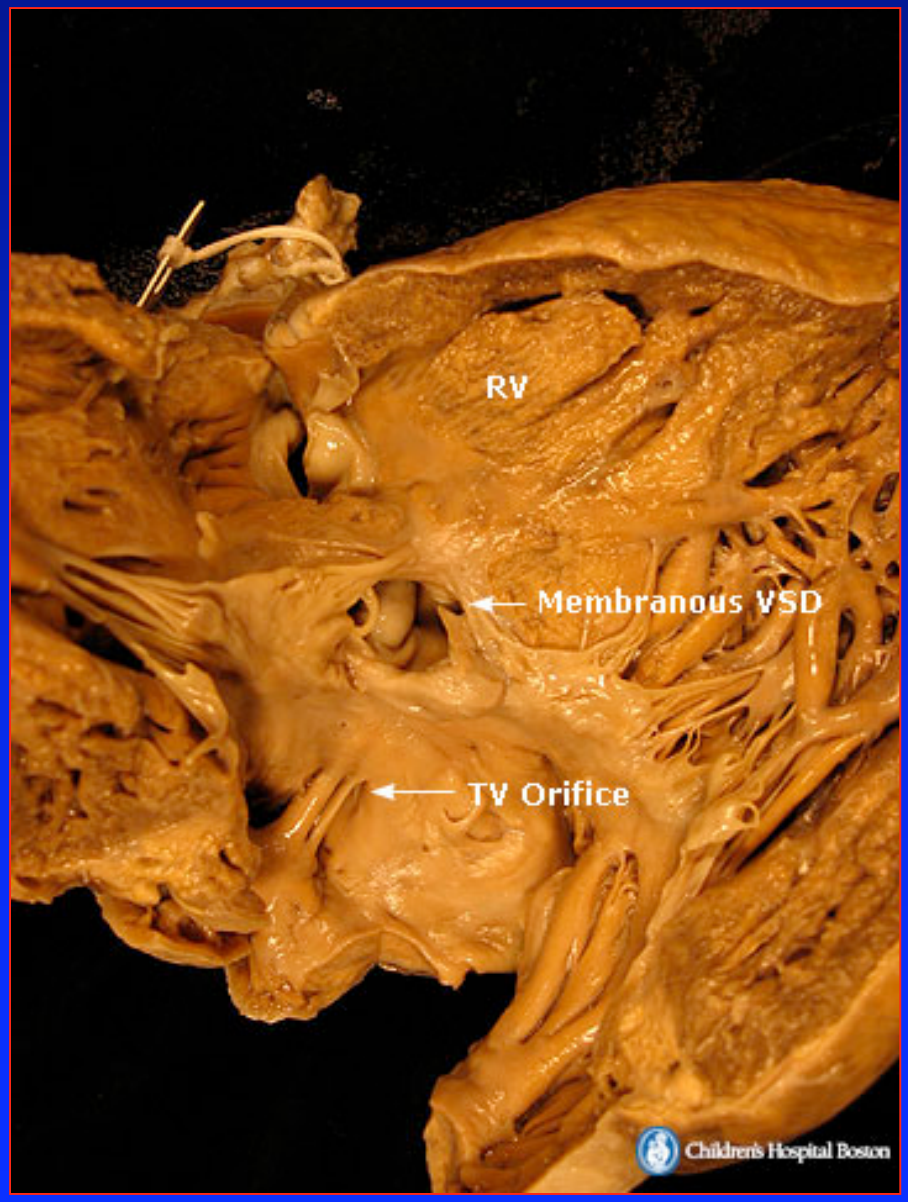




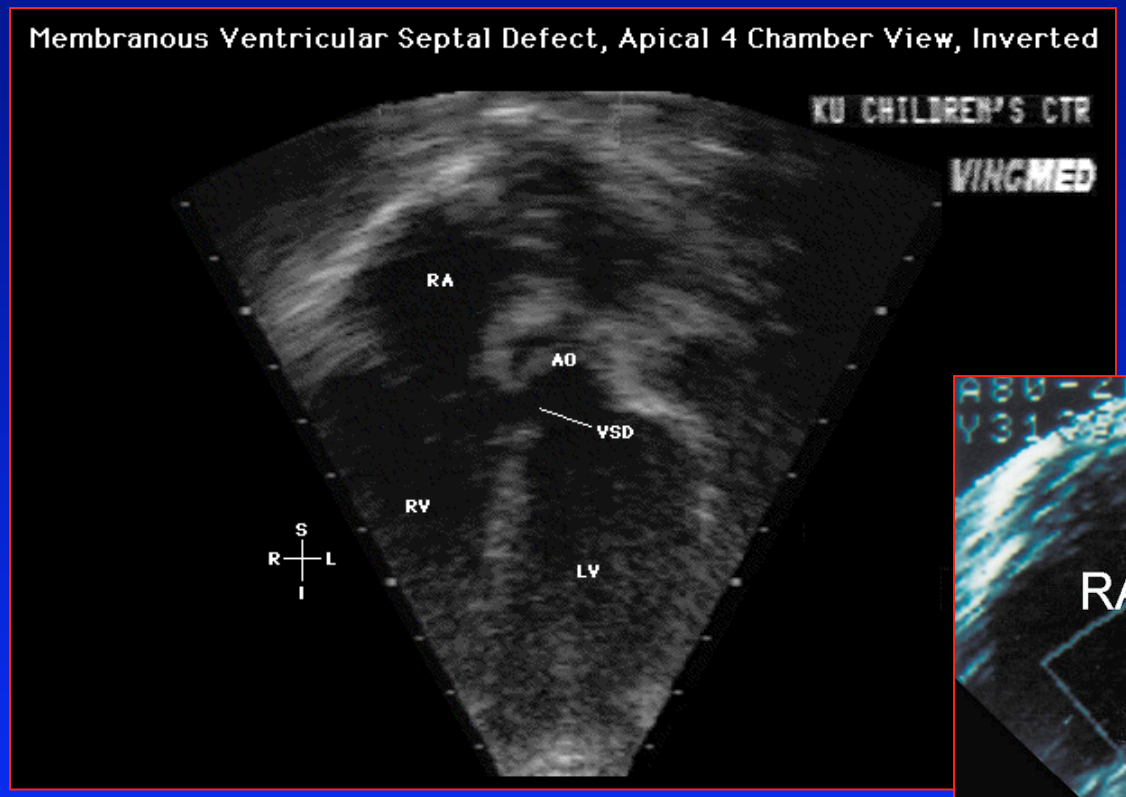
Supracristal VSD

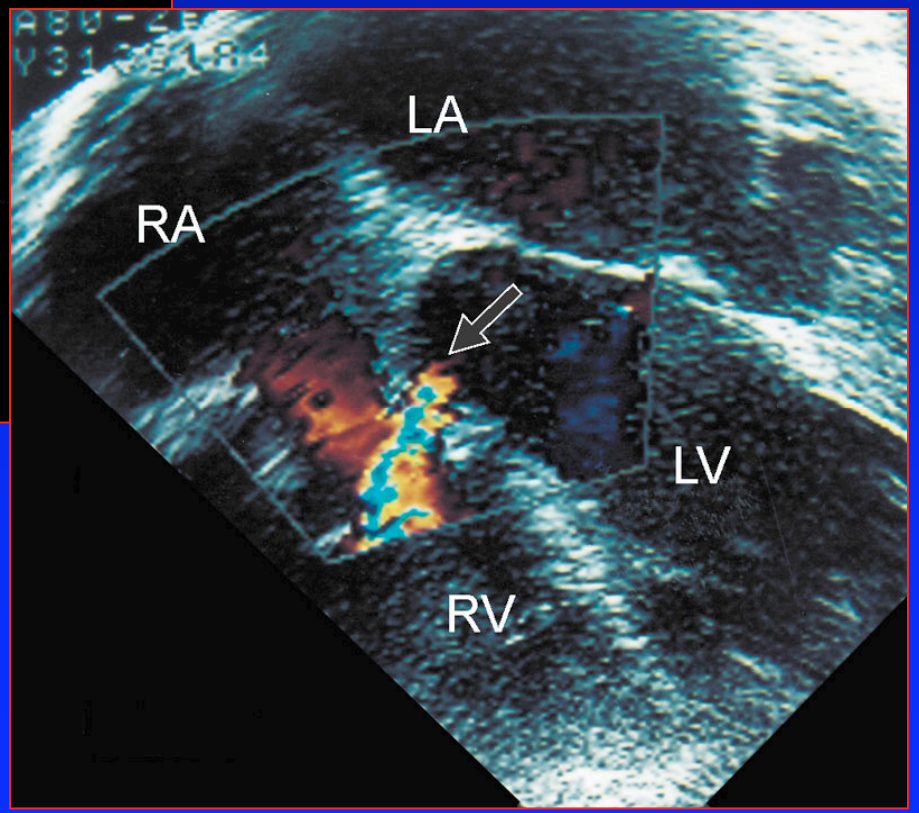


Membranous VSD

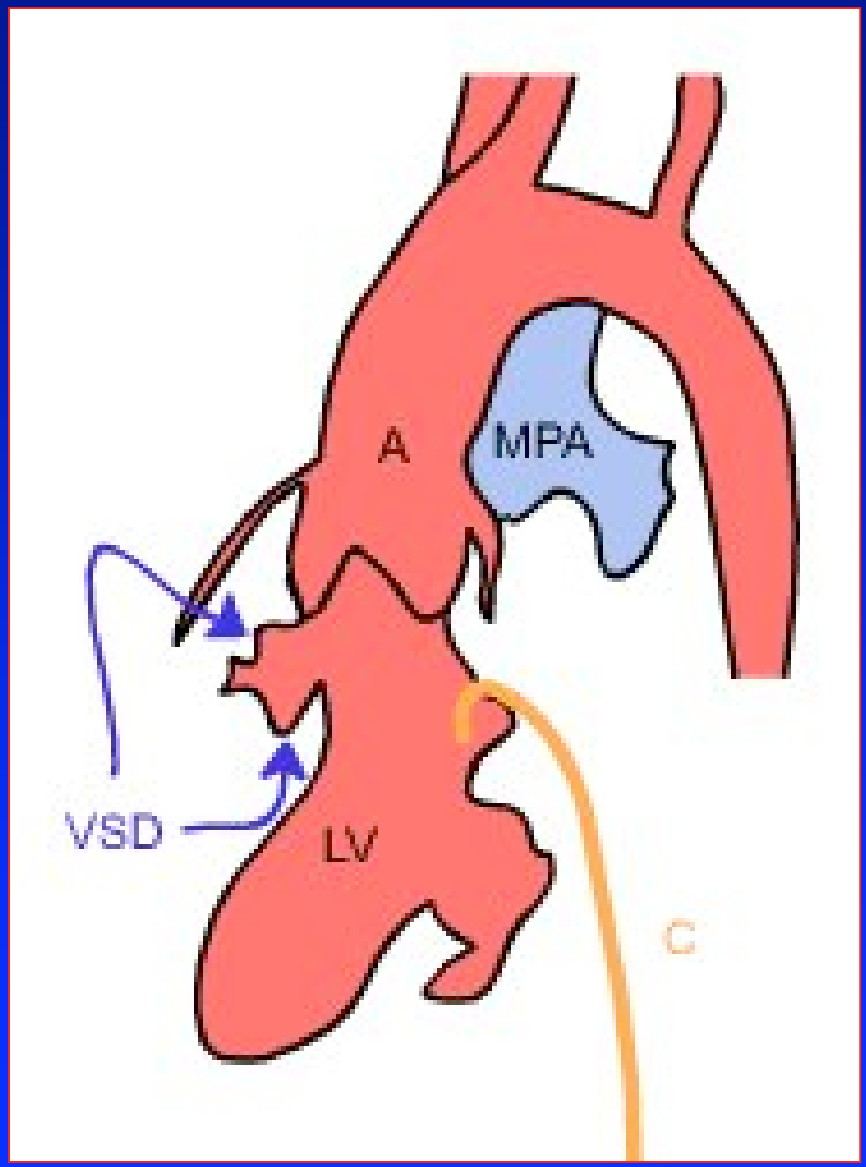


Echocardiogram: Membranous VSD





Angiogram: VSD



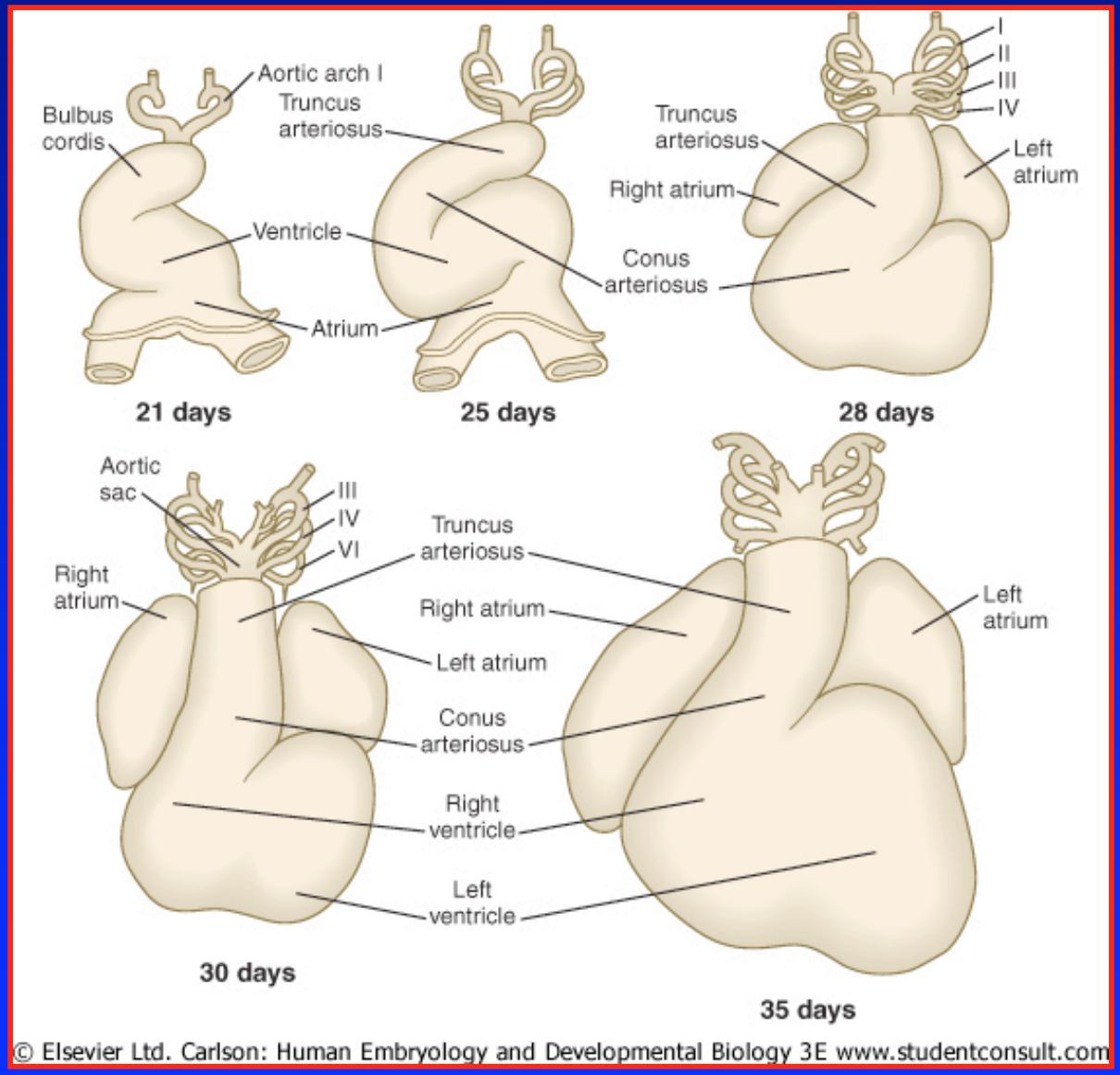
Heart Formation

- Heart structures form completely by week 12
- Week 3-4: Vitelline Stage
  - Heart tube forms
  - “Looping” occurs
  - Venous system starts to form
- Week 5
  - Placenta provides nutrients and liver takes of hematopoeis

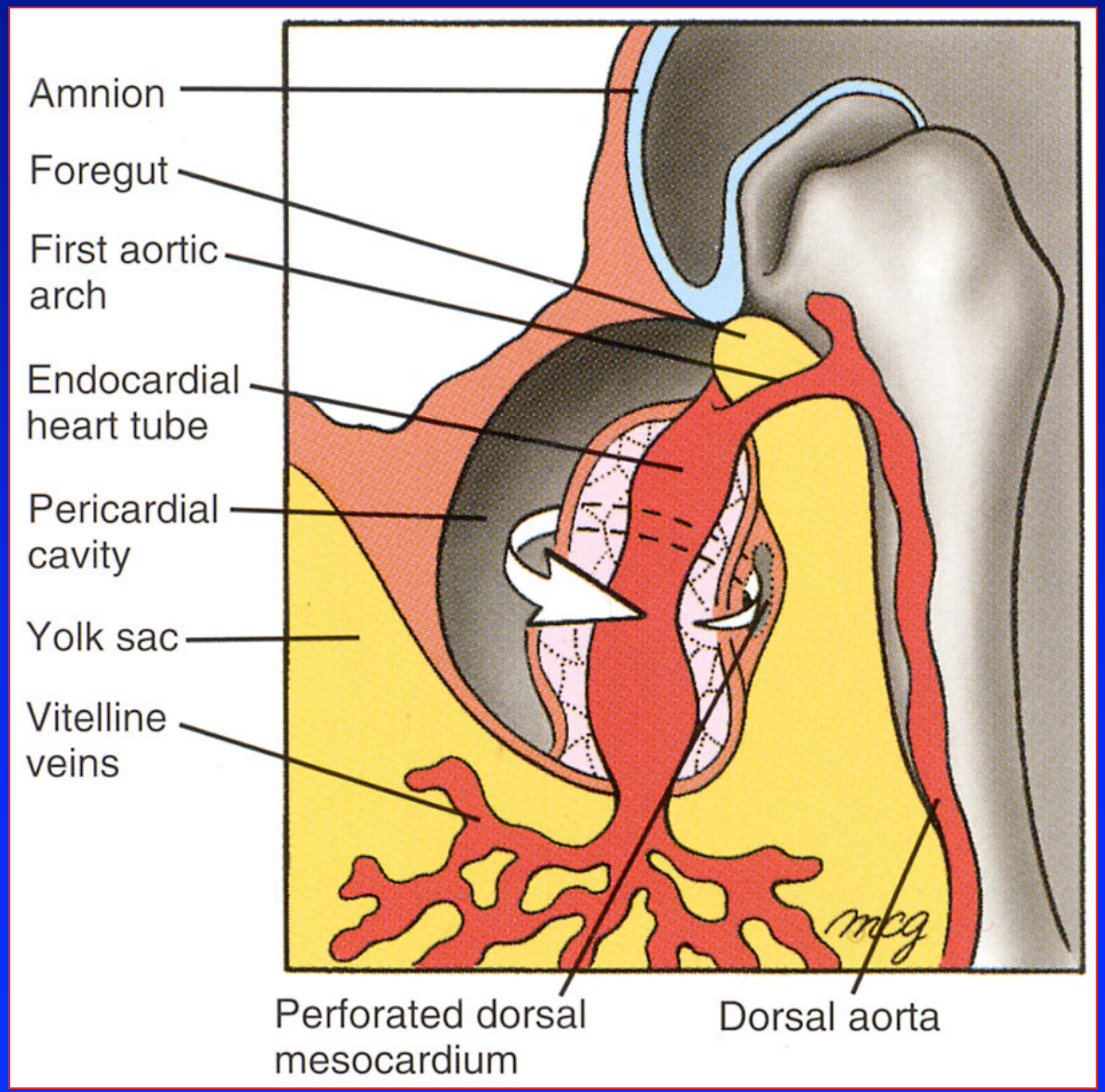
Heart Formation con't

- **Weeks 5-8**
  - SVC, IVC form
  - Right and Left Atrium divide
  - Ventricles start to form
  - Aorta and pulmonary arteries
- **Weeks 8-12**
  - Ventricles and mitral and tricuspid valves
  - Aorta and pulmonary artery, aortic arch
- **Week 12:Fetal Circulation begins**

From Primitive Heart Tube to Four Chambers: External View

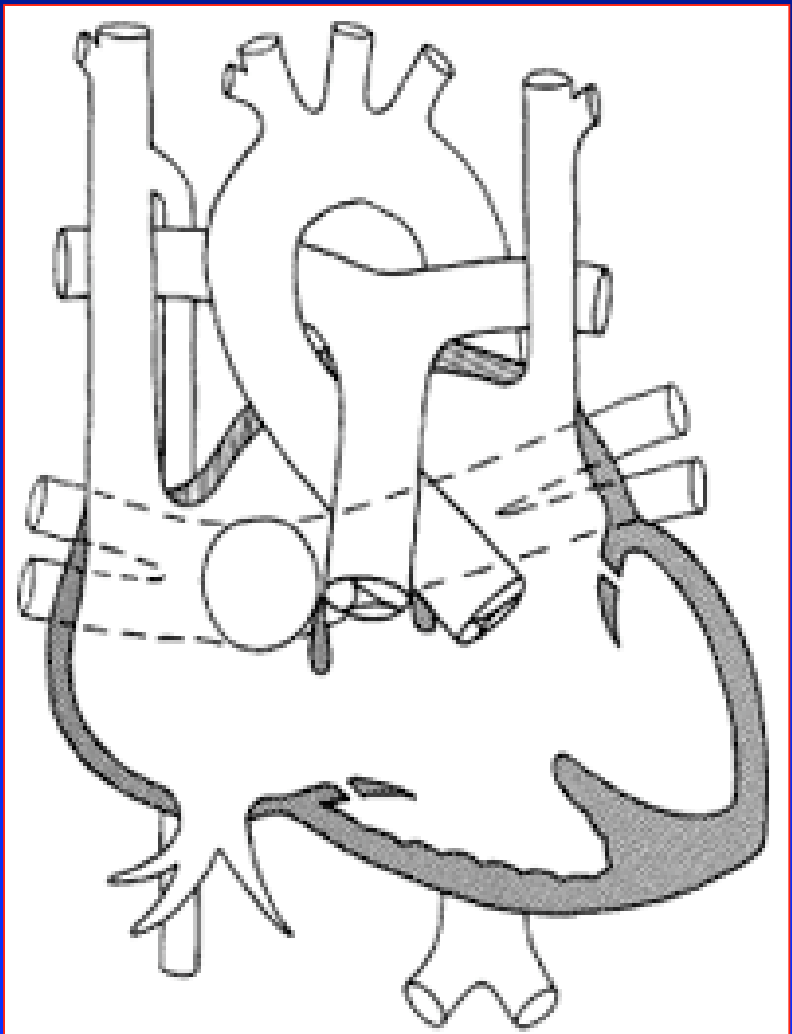


HEART AND ITS NEIGHBORHOOD: WEEK 4



Multiple Defects: Bilateral Left-Sidedness

- **Systemic Veins**
  - Interrupted IVC
  - Bilateral SVC
- **Common Atrium**
- **Common Ventricle**
  - VSD: endocardial cushion
- **Pulmonary veins:**
  - Ipsilateral
- **Pulmonary Stenosis**



Heterotaxy Syndrome
with Bilateral "Left-Sidedness"