

# *Developmental anatomy of the heart and the embryological basis for cardiac defects*

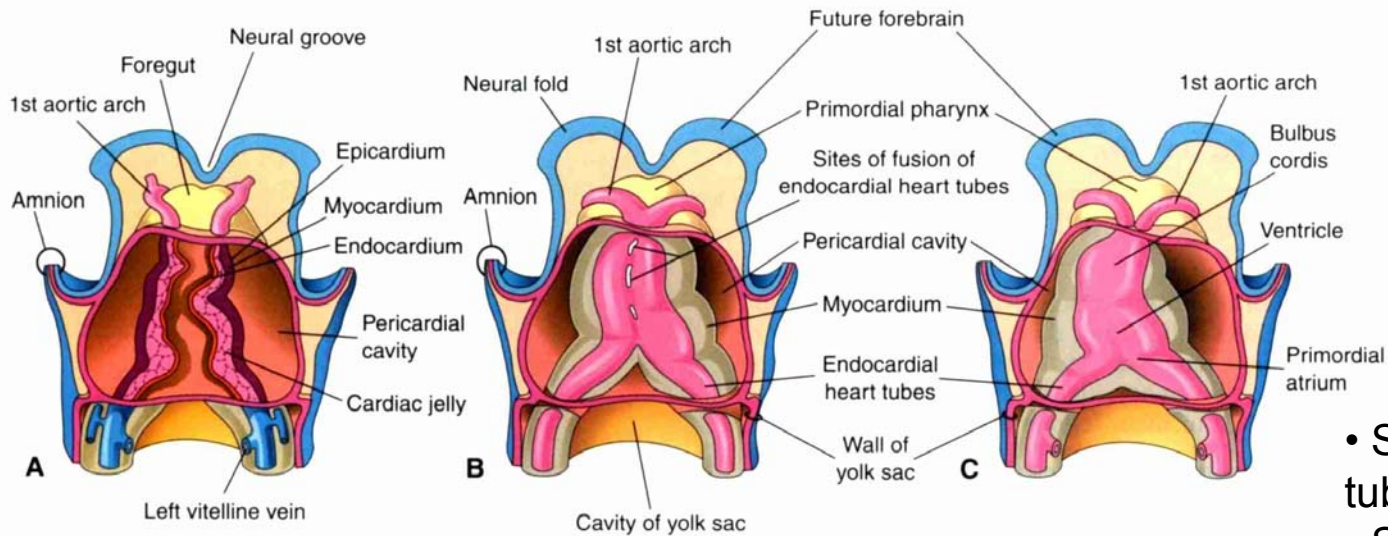
21 August 2002

<http://www.childrenheartinstitute.org/educate/defects/tetra1.htm>

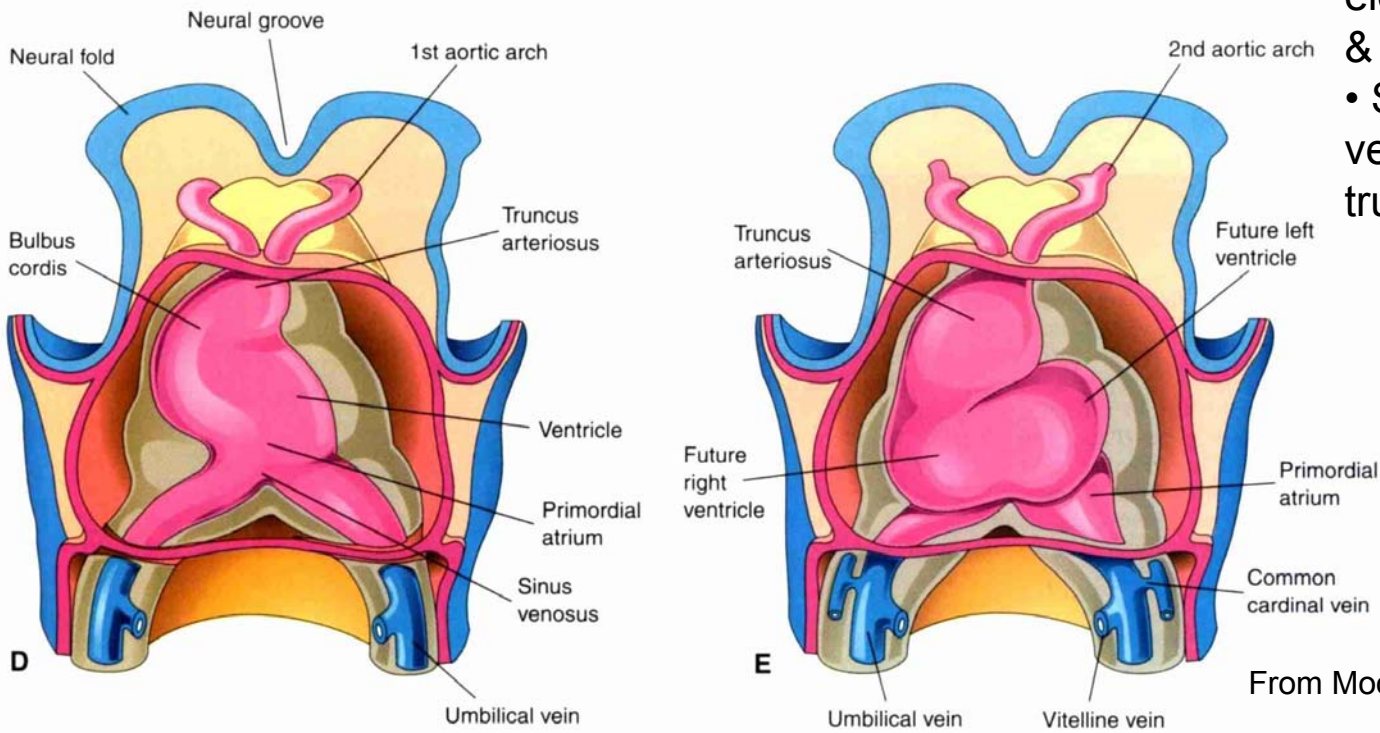
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# Formation of Endocardial Tube

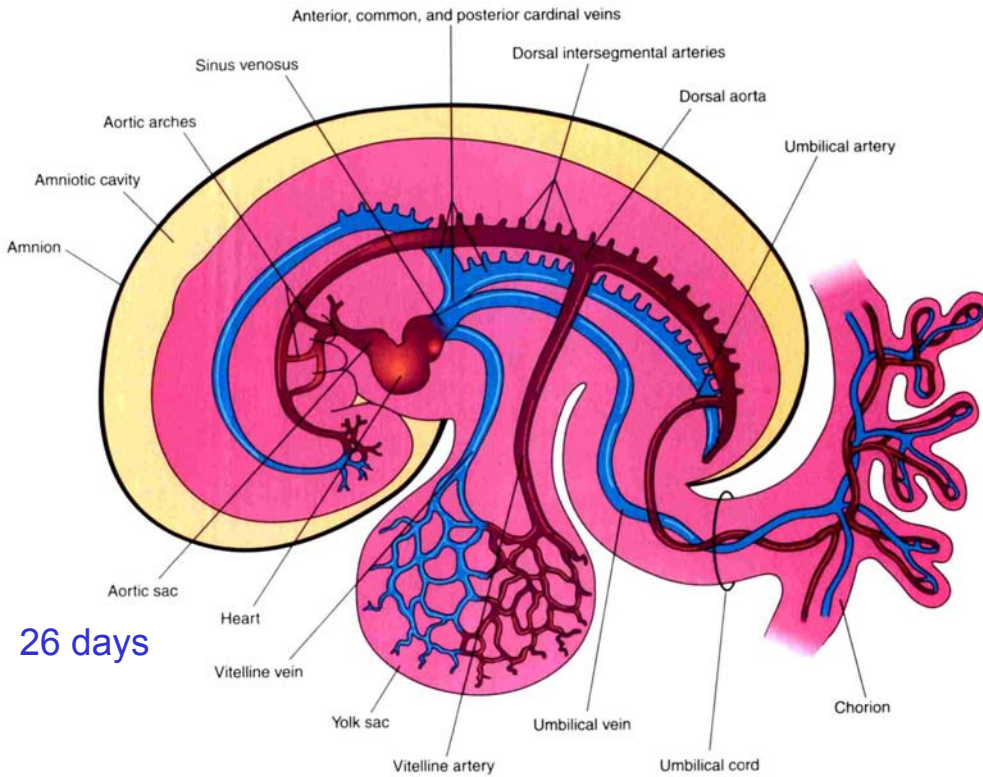


- Separate endocardial tubes that fuse
- Single endocardial tube elongates, forms dilations & constrictions
- Sinus venosus, atrium, ventricle, bulbus cordis, truncus arteriosus



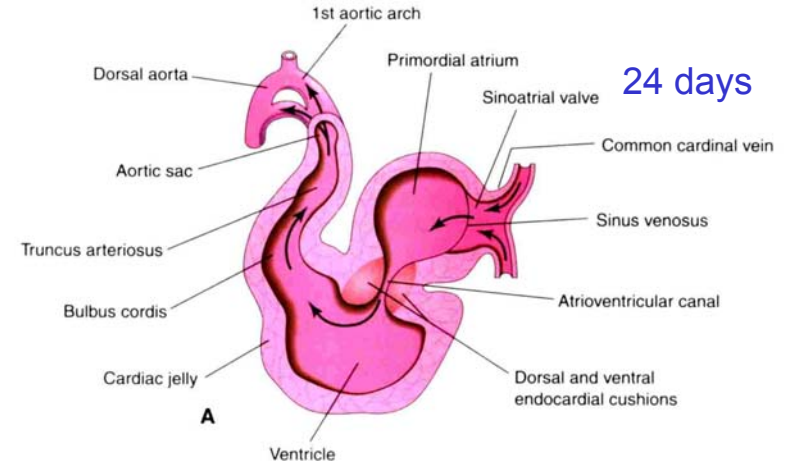
From Moore & Persaud 1998

# General Organization

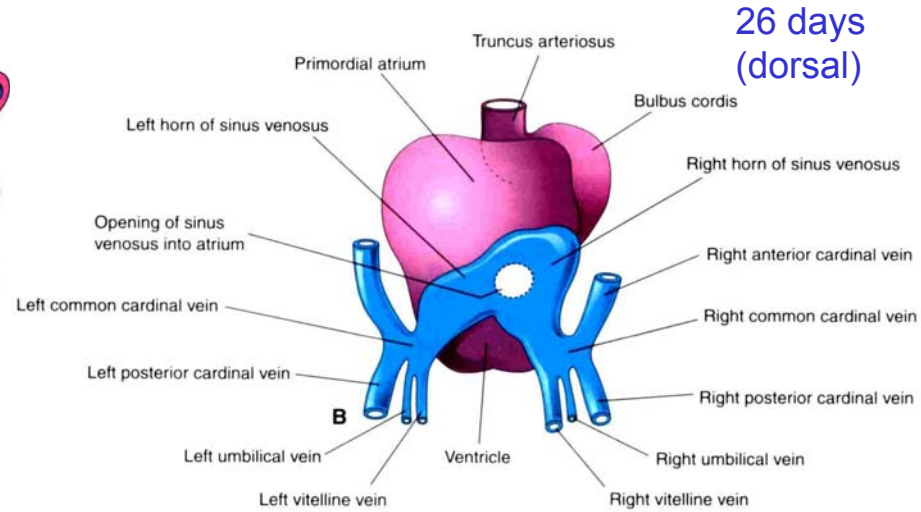


26 days

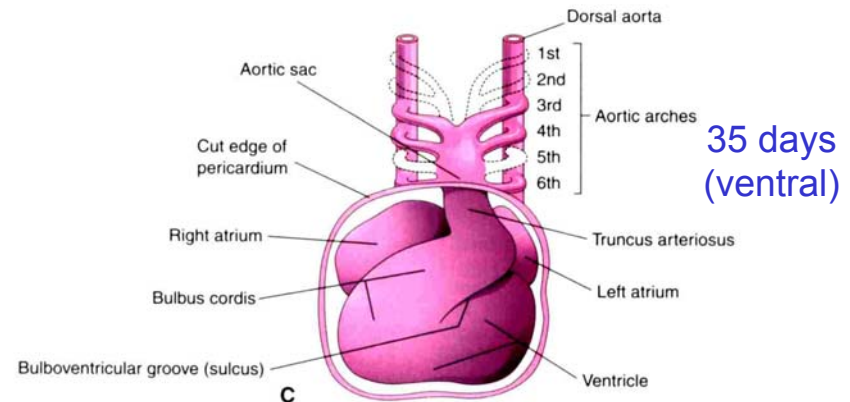
From Moore & Persaud 1998



24 days



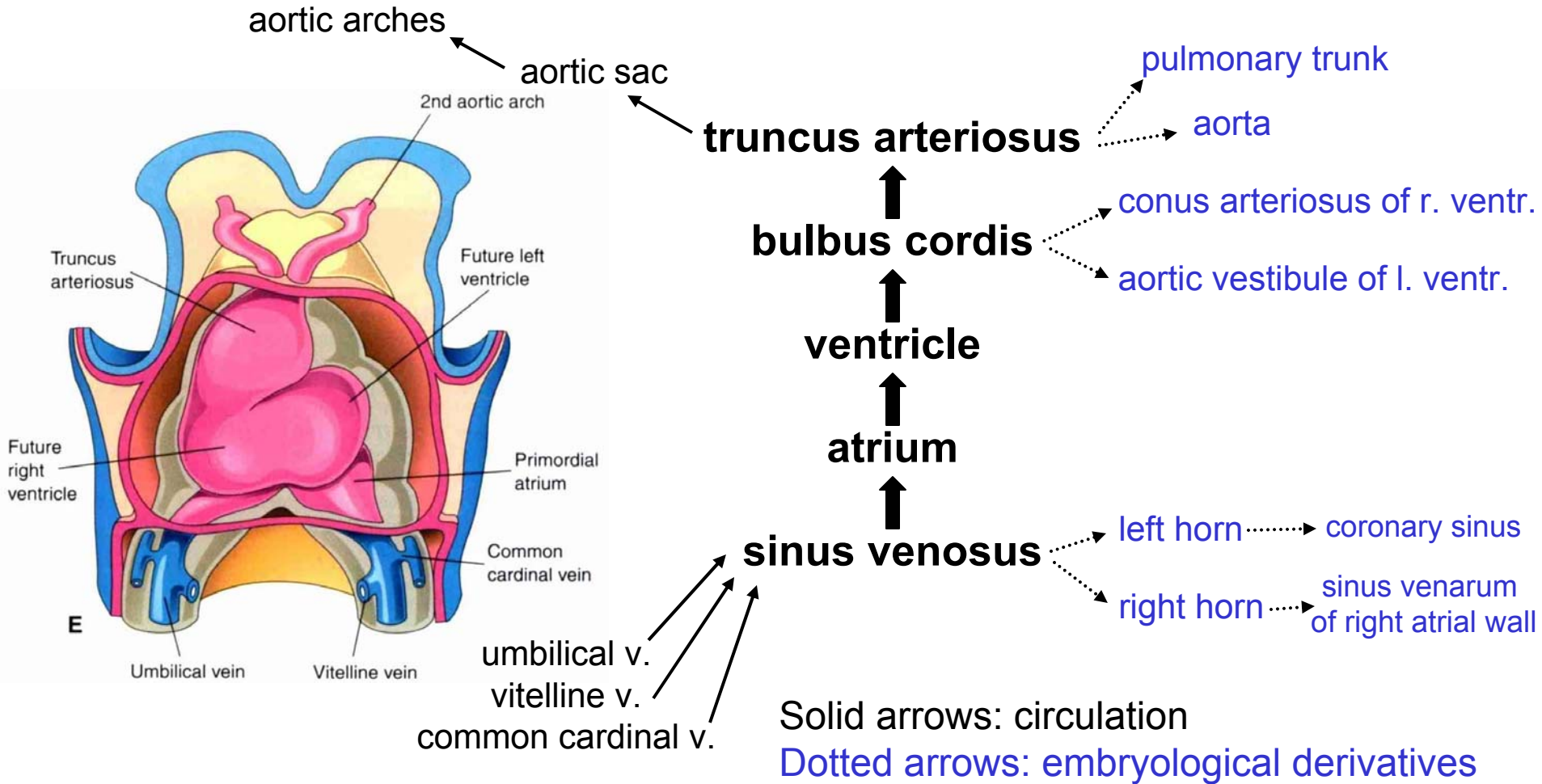
26 days  
(dorsal)



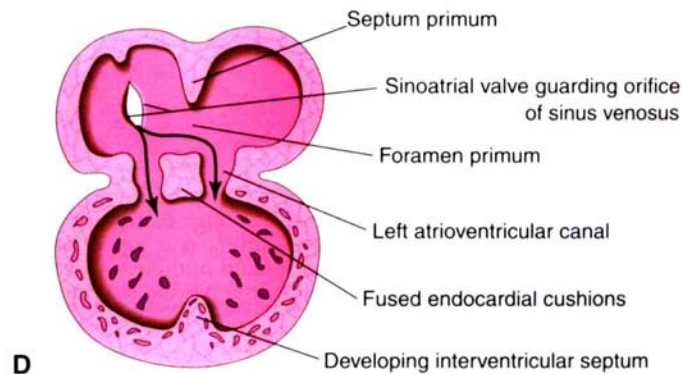
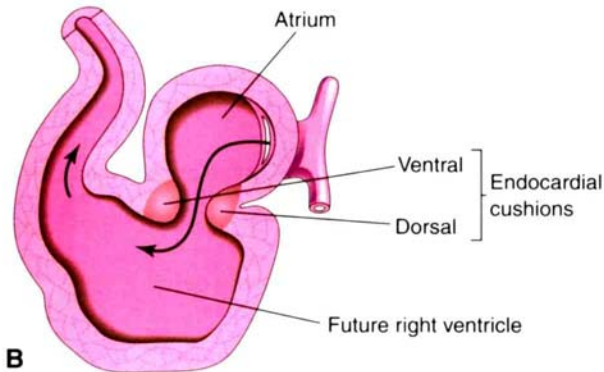
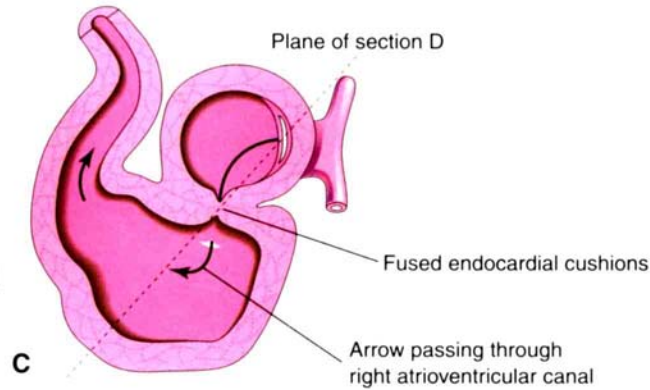
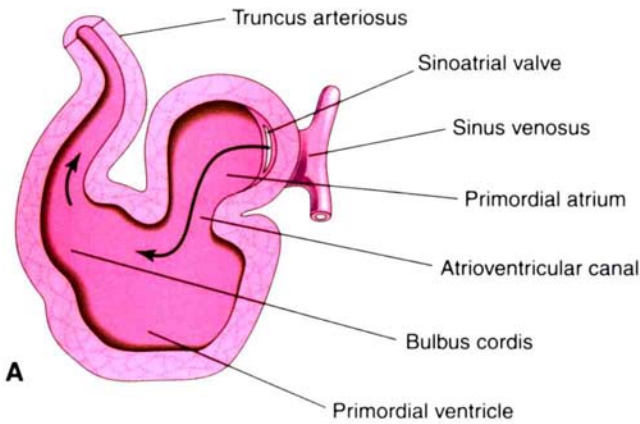
35 days  
(ventral)



# Blood Flow and Embryological Fates



# Partitioning

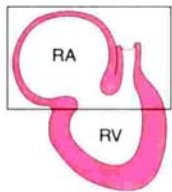


## Endocardial cushions

- dorsal & ventral swellings
- fuse, dividing the single AV canal into paired canals
- involved in formation of interatrial & interventricular septa
- derived from neural crest
- involved in many CHDs

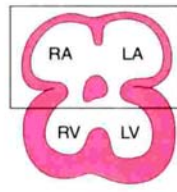
Weeks 4-5

From Moore & Persaud 1998



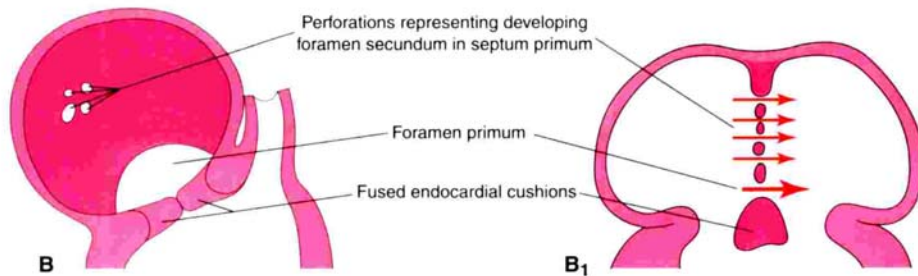
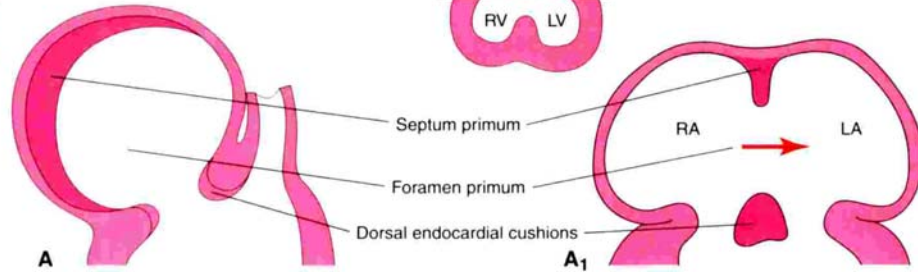
RA, right atrium  
RV, right ventricle

**viewed  
from right**

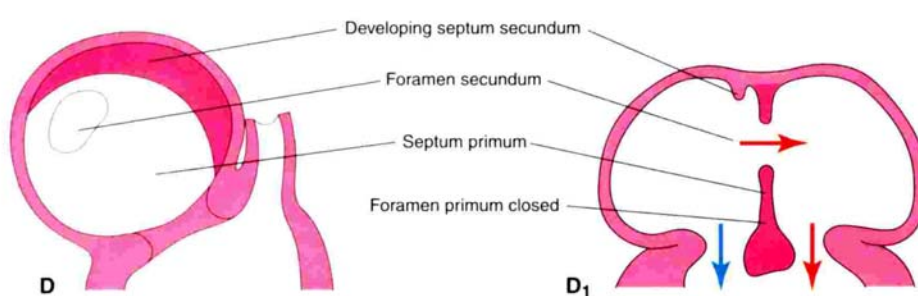
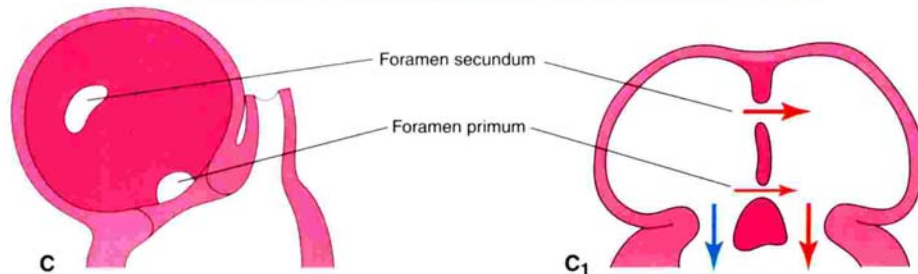


LA, left atrium  
LV, left ventricle

**coronal  
sections**



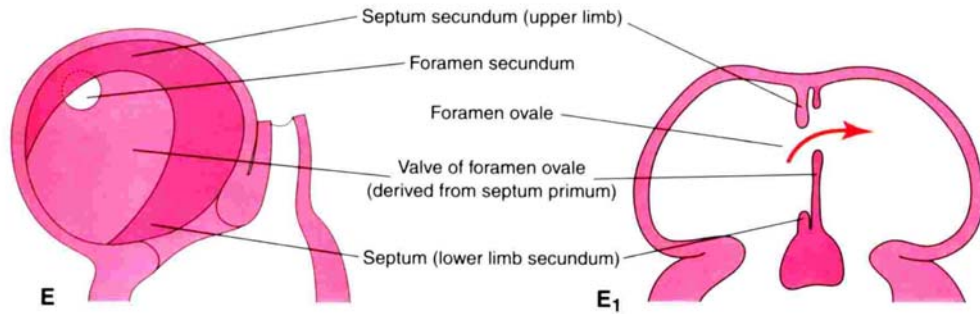
Red arrows—well oxygenated blood    Blue arrows—poorly oxygenated blood



## Atrial Partitioning I

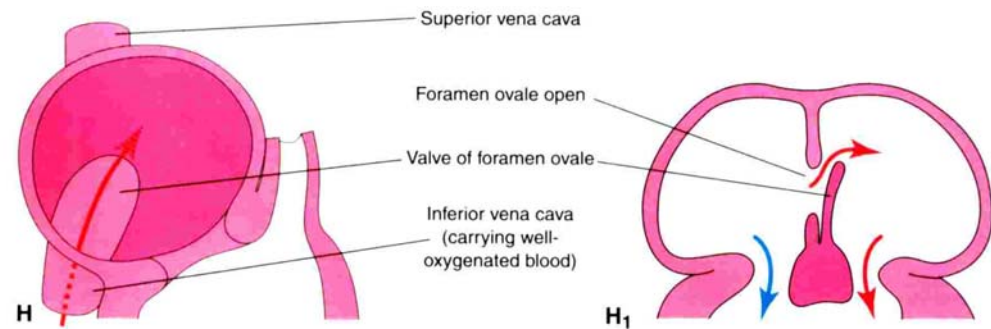
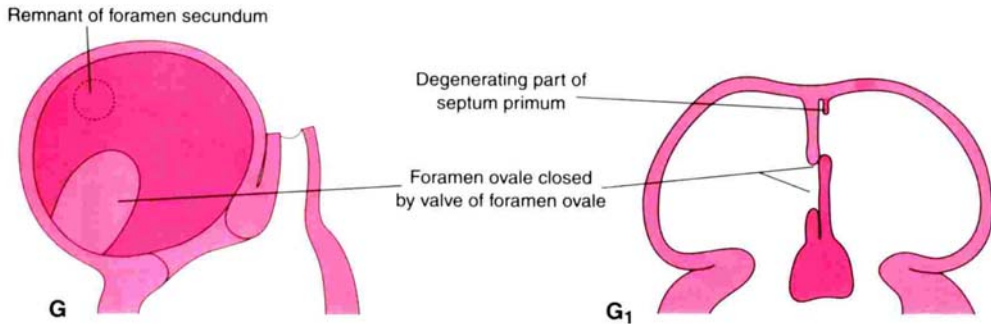
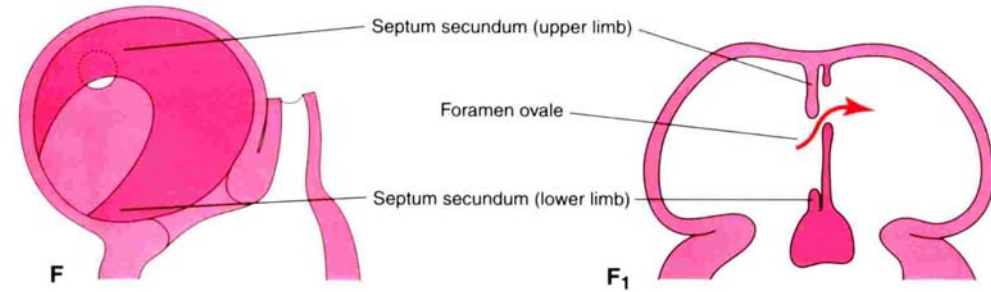
- Septum primum grows from atrial roof toward endocardial cushions
- Foramen primum: shunt that closes
- Foramen secundum: perforates septum primum, allowing shunt
- Septum secundum grows down, overlapping foramen secundum

From Moore & Persaud 1998



## Atrial Partitioning II

- Septum secundum grows down, overlapping foramen secundum
- Foramen ovale: between septum primum & septum secundum
- Remaining portion of septum primum forms valve of foramen ovale



From Moore & Persaud 1998

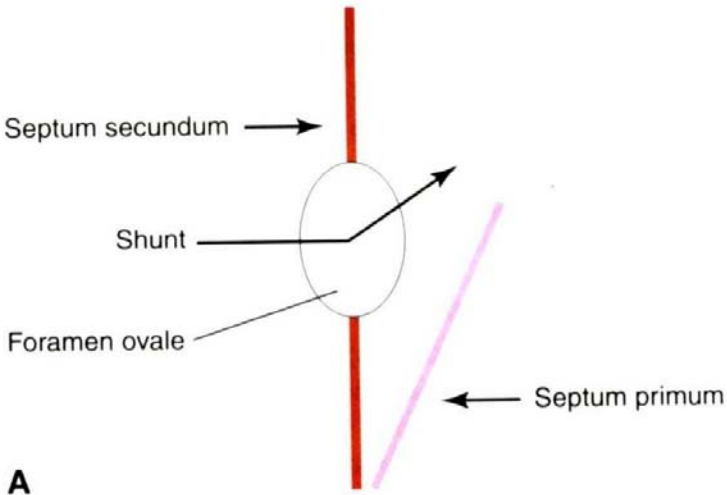


# Atrial Partitioning III

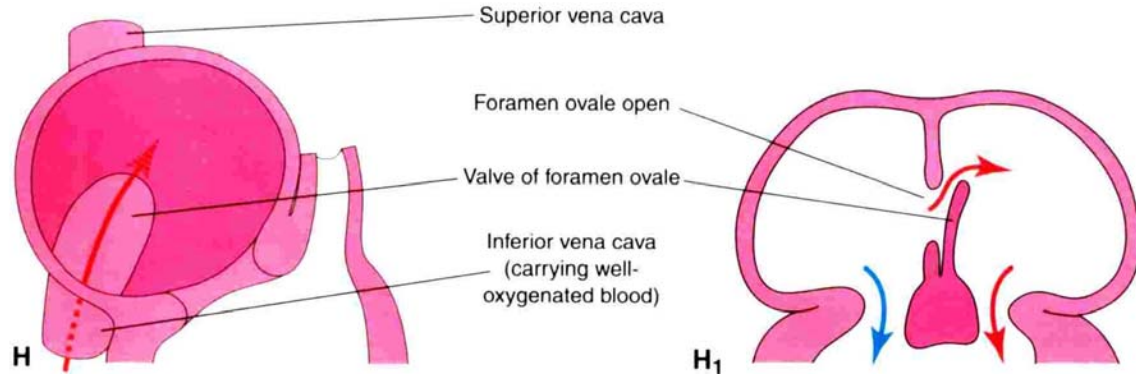
BEFORE BIRTH

**RIGHT ATRIUM**  
HIGHER PRESSURE

**LEFT ATRIUM**  
LOWER PRESSURE



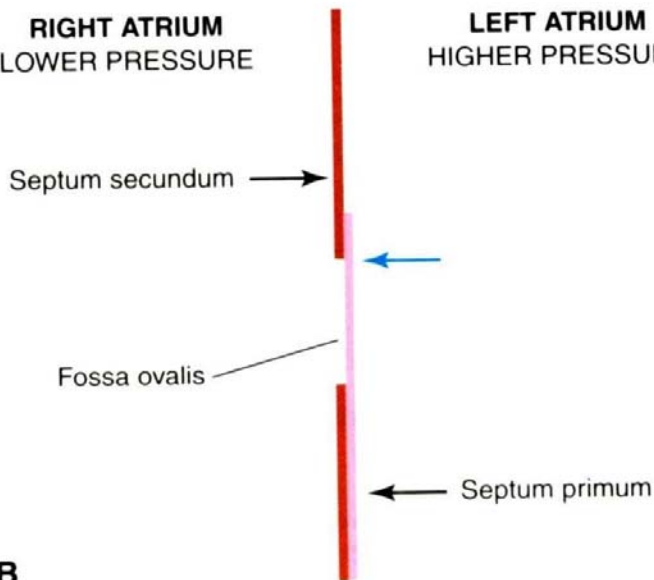
**A**



AFTER BIRTH

**RIGHT ATRIUM**  
LOWER PRESSURE

**LEFT ATRIUM**  
HIGHER PRESSURE



**B**

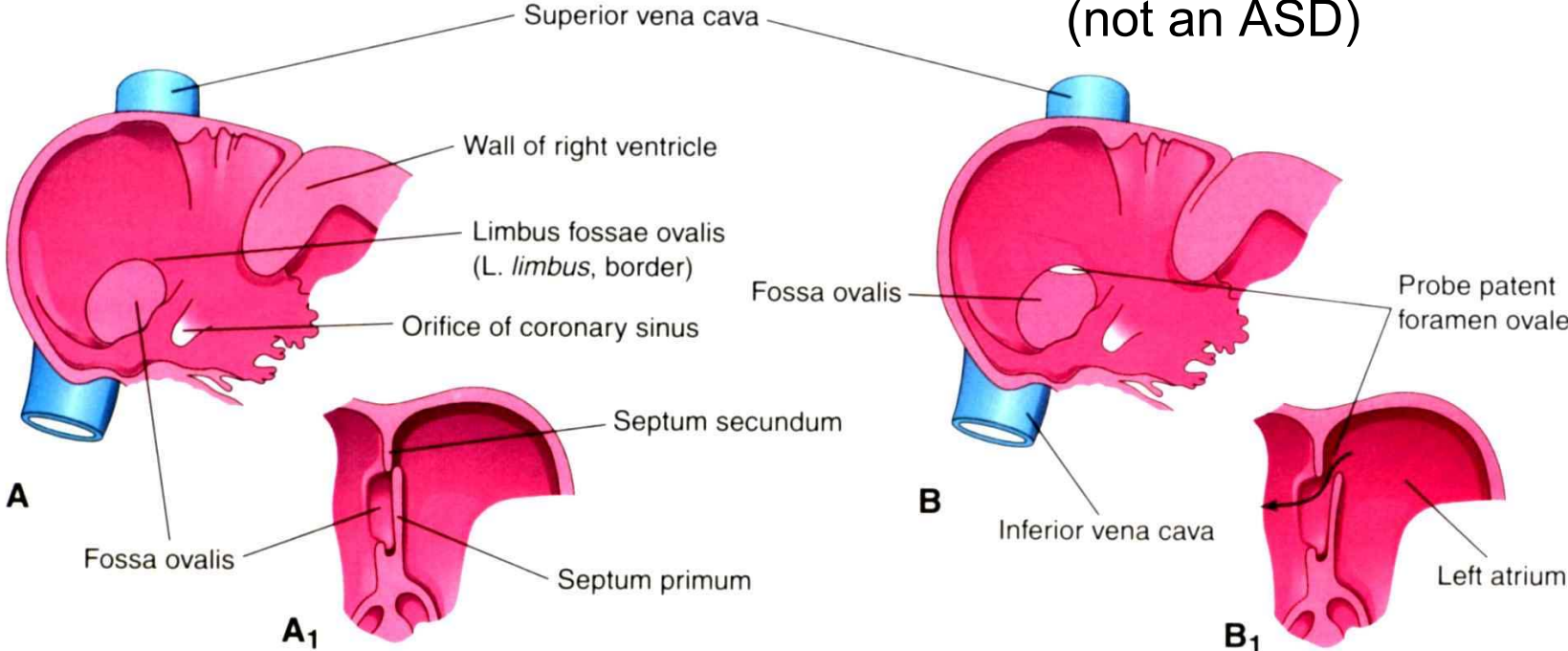
- Fetus
  - right side high pressure (high pulmonary resistance, etc.)
  - well oxygenated blood streams through foramen ovale
  - valve of foramen ovale closes with left atrial contraction
- After birth
  - right side low pressure (low pulmonary resistance)
  - valve remains closed (physiological closure)
  - valve eventually fuses (anatomical closure): fossa ovalis



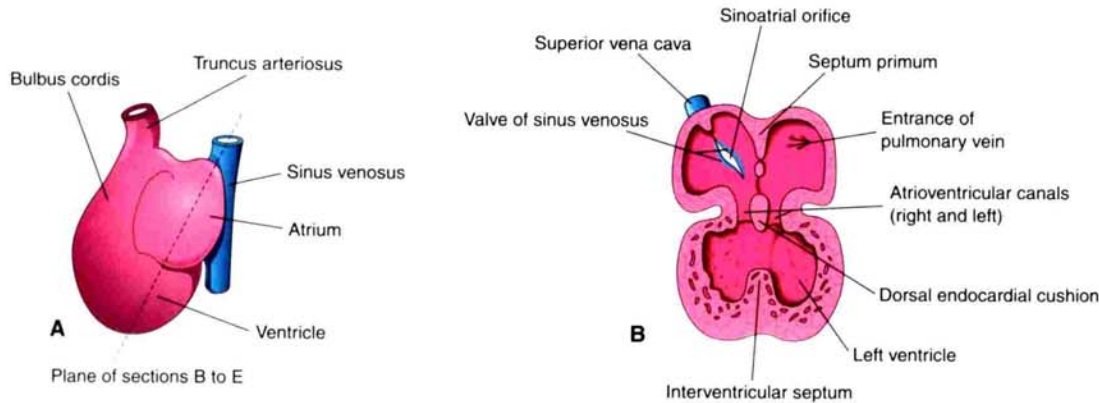
# Atrial Partitioning IV

postnatal

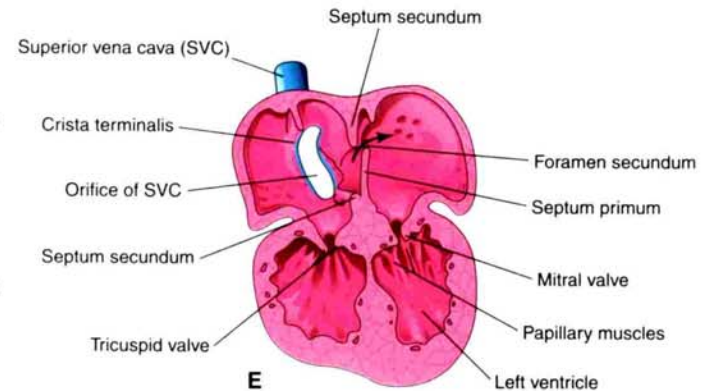
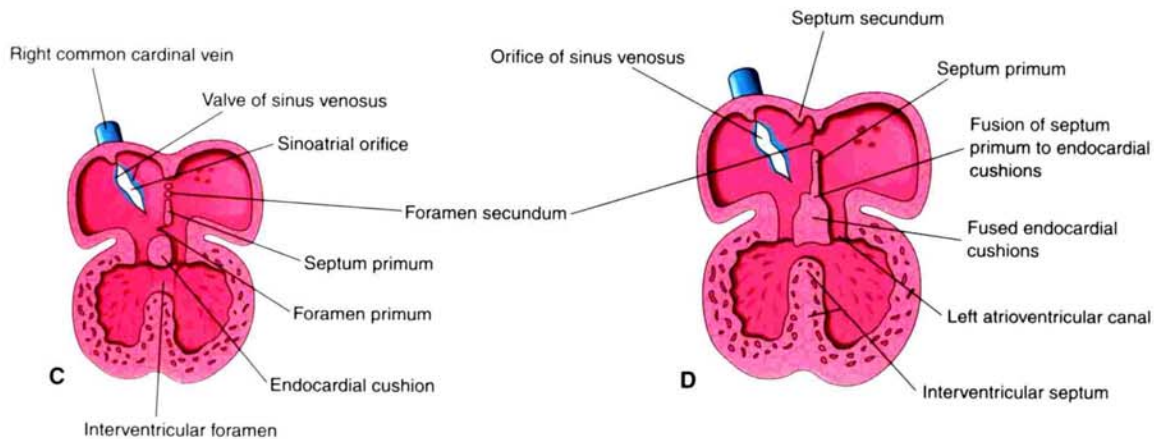
probe patent  
foramen ovale  
(not an ASD)



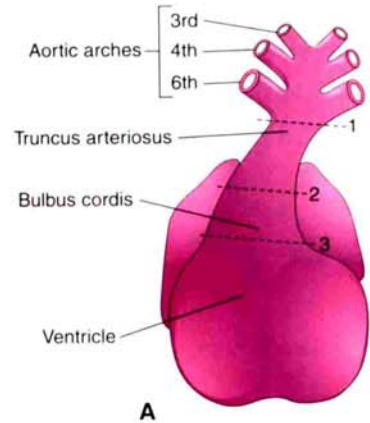
# Ventricular Partitioning



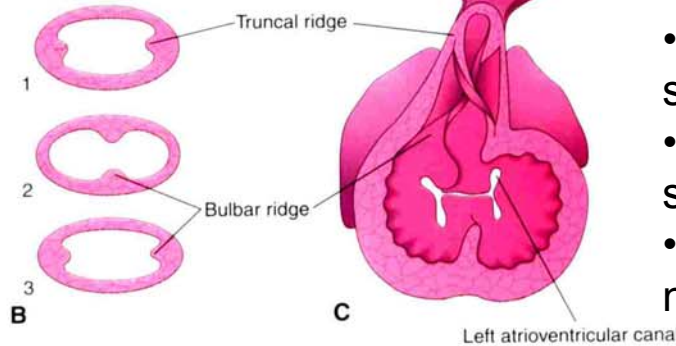
- Closes in week 7: not part of fetal circulation
- Muscular IV septum grows from floor
- Membranous IV septum forms from endocardial cushions and bulbar ridges
- Closure of membranous IV is associated with partitioning of truncus arteriosus



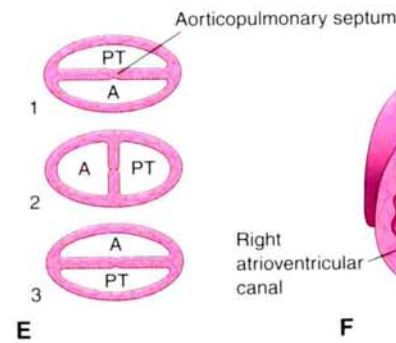
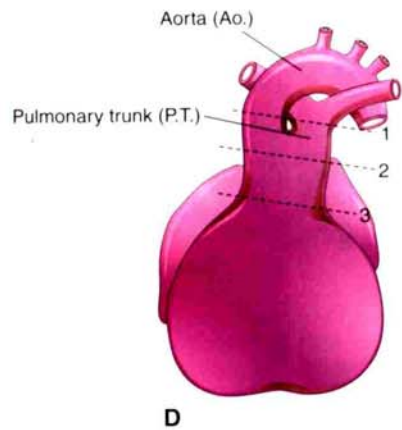
# Partitioning of Truncus Arteriosus



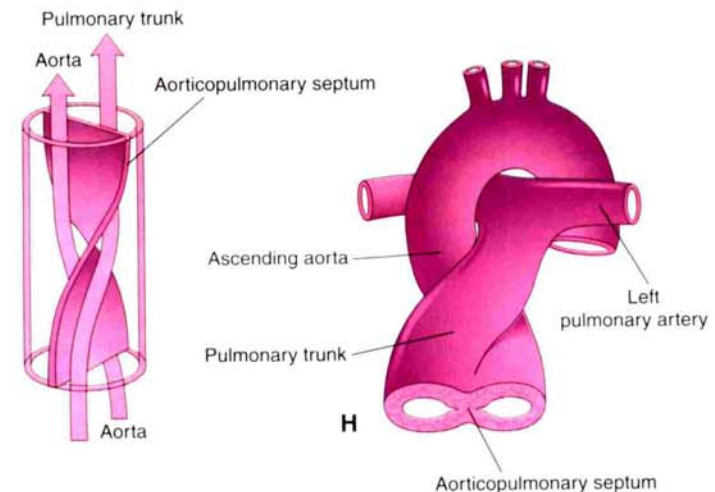
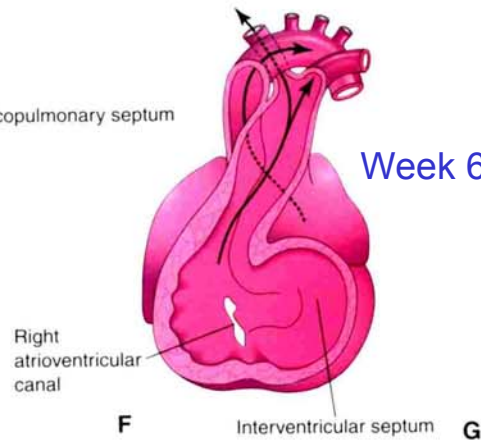
Week 5



- continuous set of ridges in bulbus cordis (bulbar ridges) and truncus arteriosus (truncal ridges)
- grow toward each other, spiraling 180°
- fuse to form spiraling aorticopulmonary septum, dividing aorta & pulmonary trunk
- bulbar ridges involved in formation of IV septum
- bulbar & truncal ridges derived from neural crest cells—clinical implications

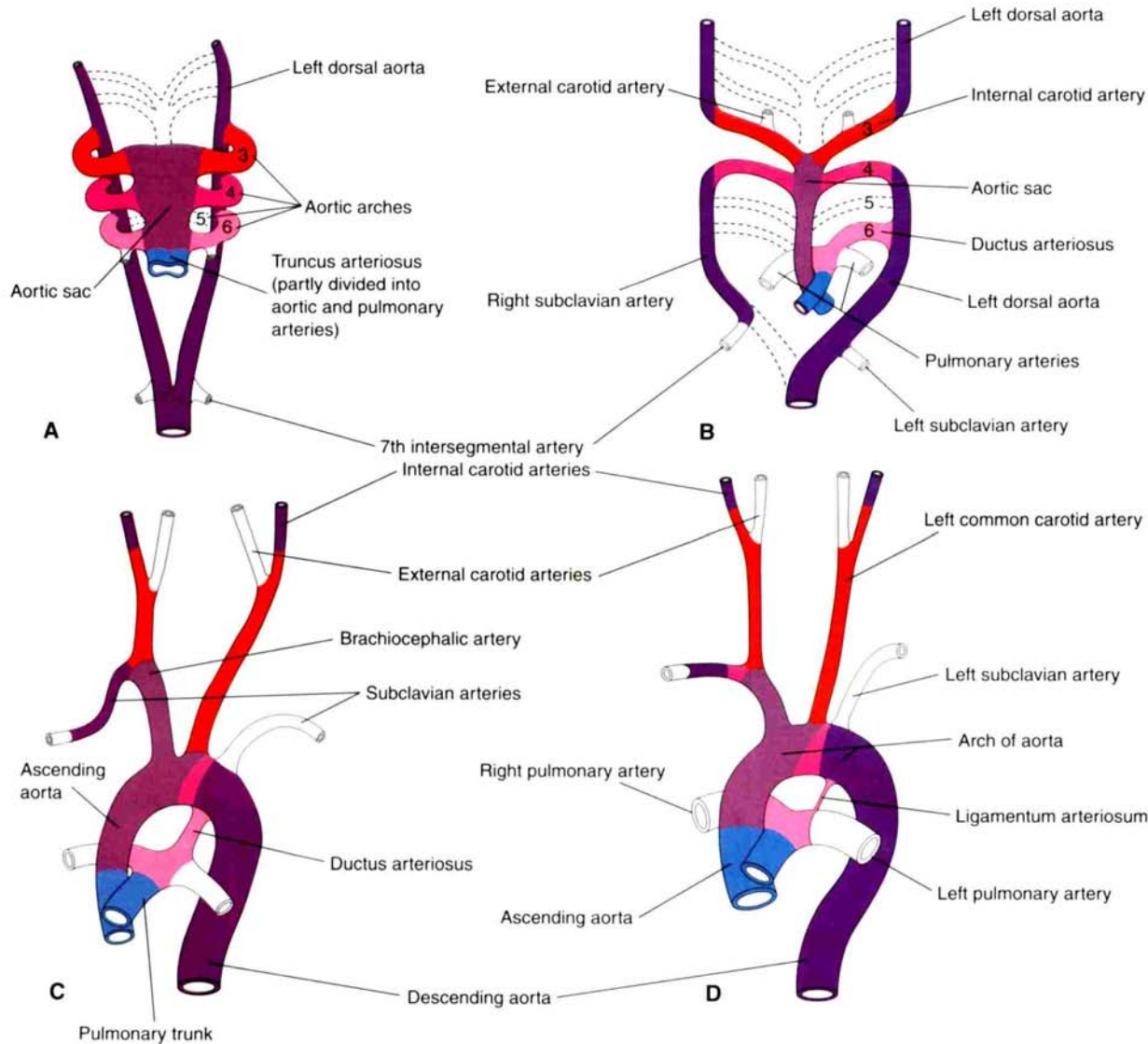


Week 6





# Aortic arches: Ductus arteriosus

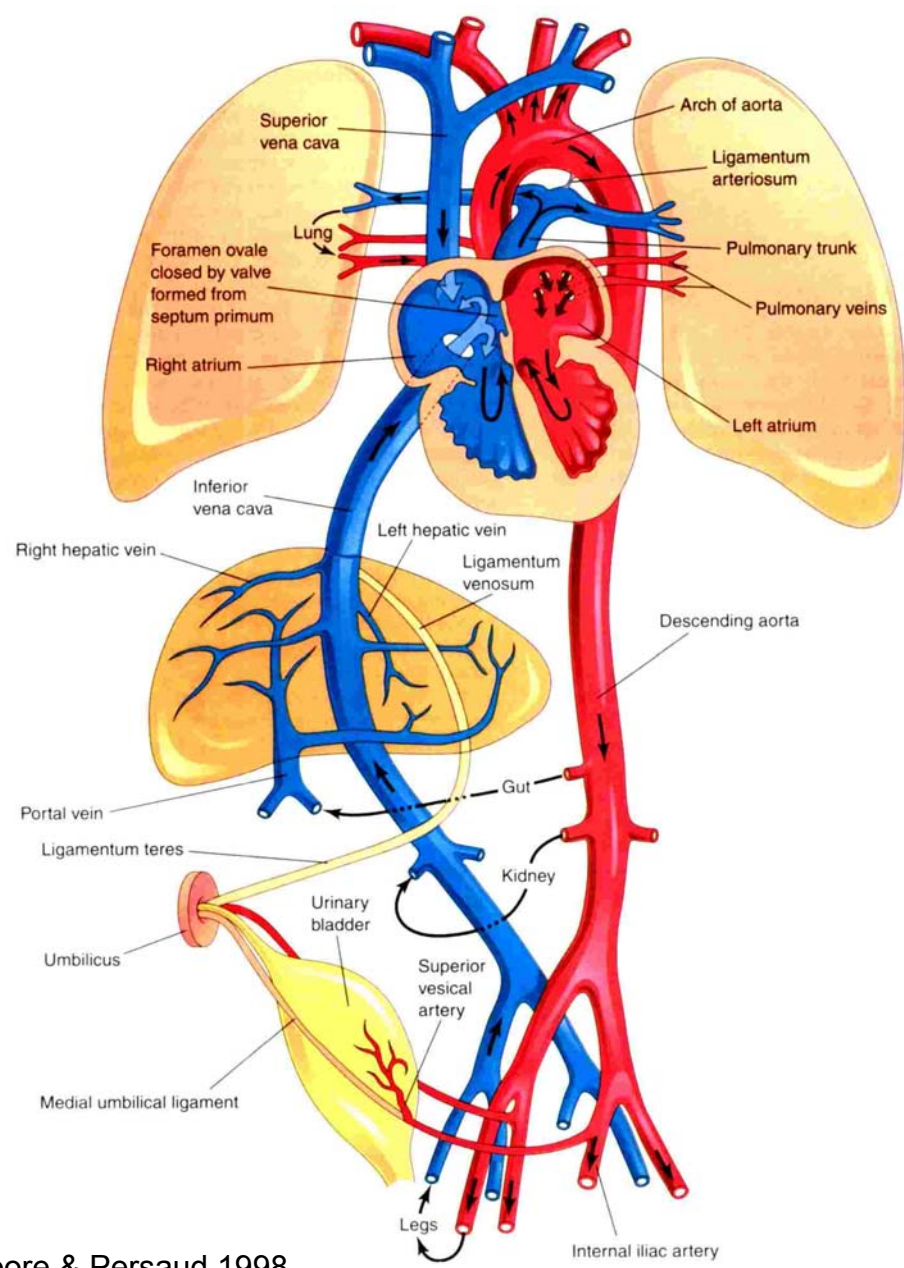
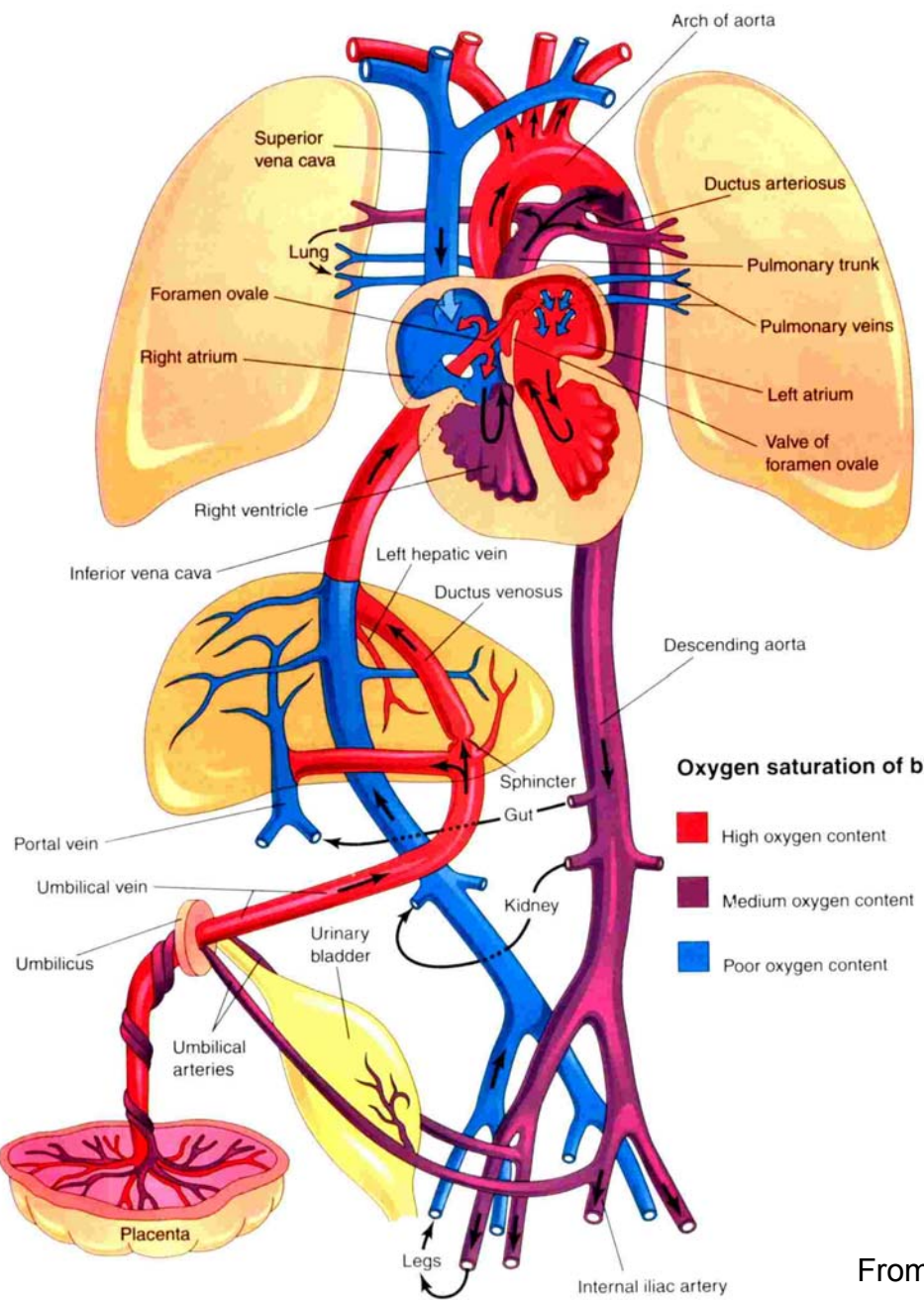


- postnatal vessels cobbled together from aortic arches, aortic sac, TA, & dorsal aortae
- Ductus arteriosus: persistent distal portion of left 6th arch
- DA connects pulmonary trunk to aorta
- DA closes postnatally



From Moore & Persaud 1998

# Perinatal Circulation



From Moore & Persaud 1998

# Congenital Heart Defects

## Acyanotic

volume load

pressure load

left-to-right shunts

obstr. ventric. outflow

- atrial septal defect
- ventricular septal defect
- AV canal
- patent ductus arteriosus

- pulmonary valve stenosis
- aortic valve stenosis
- coarctation of aorta

## Cyanotic

↑ pulmonary flow

↓ pulmonary flow

- transpos. of gr. vessels
- single ventricle
- truncus arteriosus
- total anomalous pulm. return w/o obstruction

- tetralogy of Fallot
- pulmonary atresia
- tricuspid atresia
- total anomalous pulm. return w/ obstruction

modified from Bernstein (1996) and other sources



# Ventricular Septal Defects (VSD)

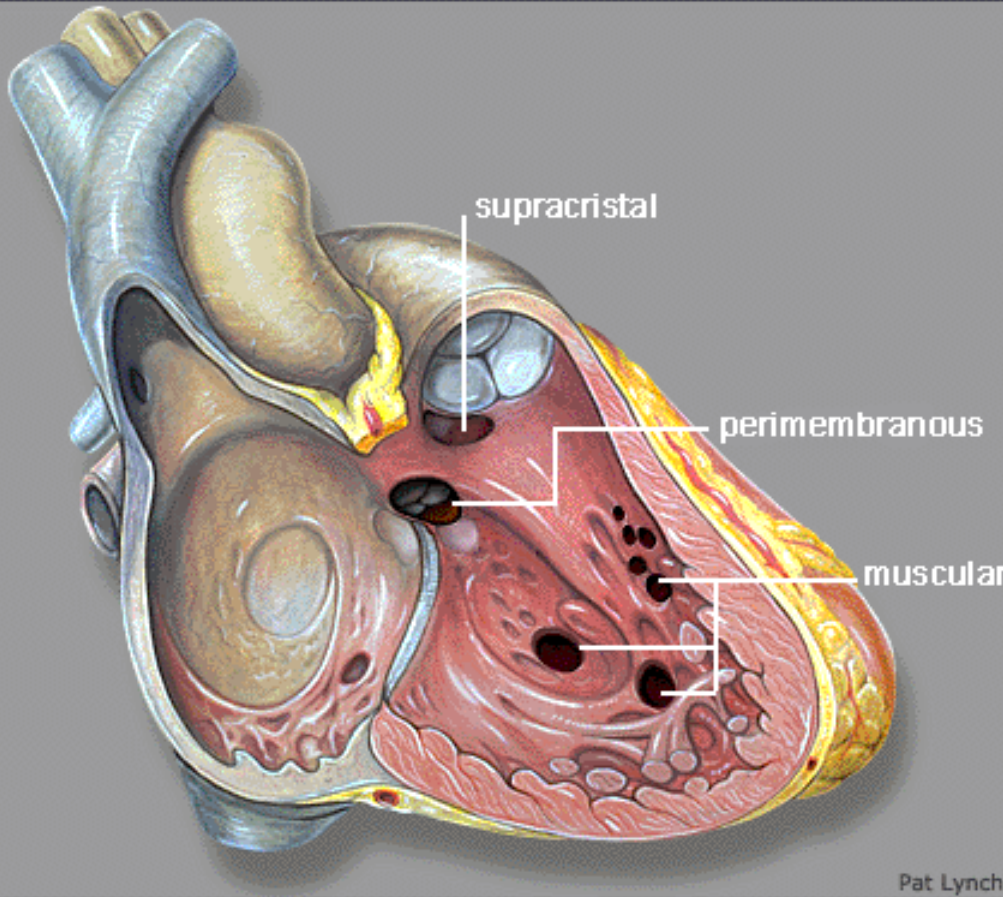
Acyanotic

volume load

left-to-right shunts

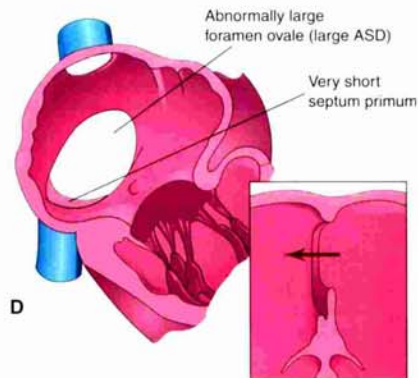
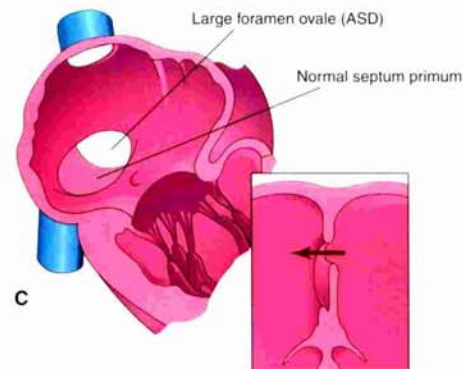
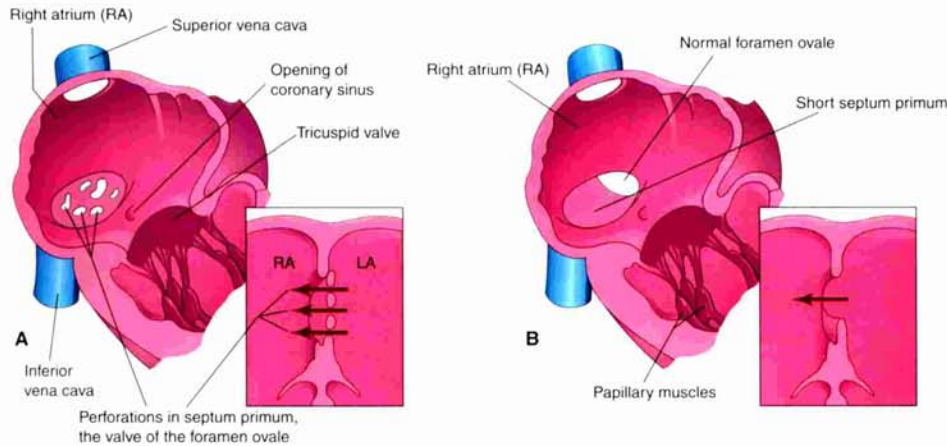
- atrial septal defect
- ventricular septal defect
- AV canal
- patent ductus arteriosus

- Membranous (= perimembranous, conoventricular) VSD
  - Most common CHD (males>females)
  - Endocardial cushions & bulbar ridges fail to fuse with musc. septum
- Muscular VSD
  - In muscular IV septum
  - “Swiss cheese” VSD
- Supracristal VSD
  - Least common



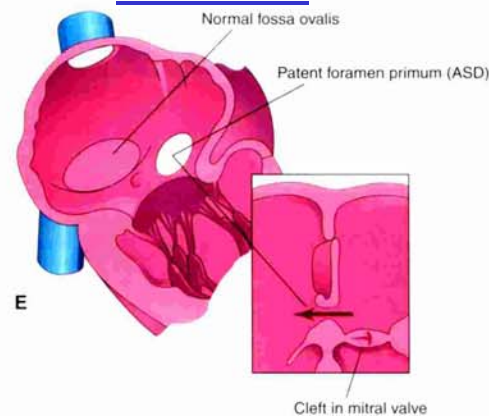
# Atrial Septal Defects (ASD)

## Secundum ASDs

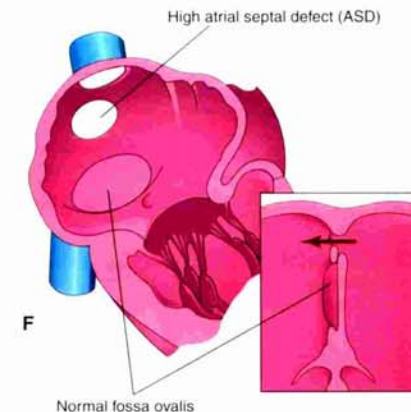


- (Ostium) Secundum ASDs
  - Most common ASD (females > males)
  - Usually due to problems with septum primum (perforated or too short), but sometimes septum secundum or both septa
- AV septal defect (AV canal)
  - Endocardial cushion problems so that septum primum never fuses with cushion tissue
  - Patent foramen (ostium) primum
  - Valve defects
  - Sometimes no fusion of endocardial cushions: AV septal defect
  - 20% of Downs patients
- Sinus venosus ASDs: very rare

## Primum ASDs & AV canal



## Sinus venosus ASD



# Increased pressure load defects: Valve stenosis

- Pulmonary or aortic stenosis
- Unequal partitioning of the truncus arteriosus
- Deviation of the aortopulmonary septum
- One side expanded, other side stenosed

Acyanotic



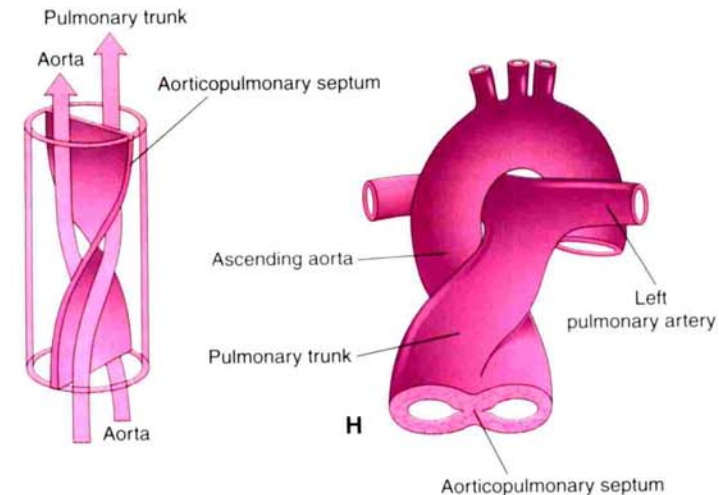
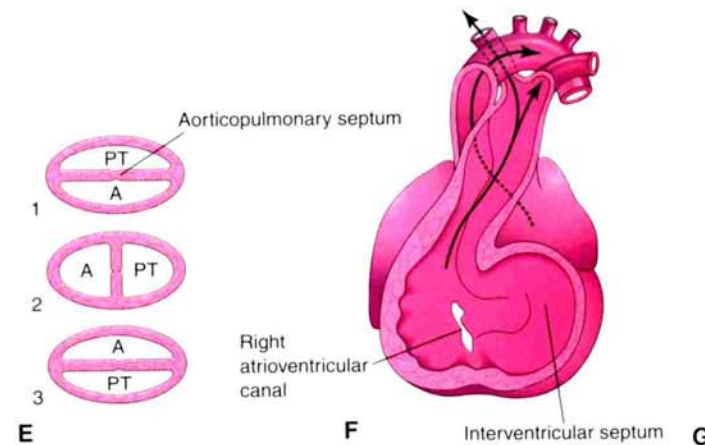
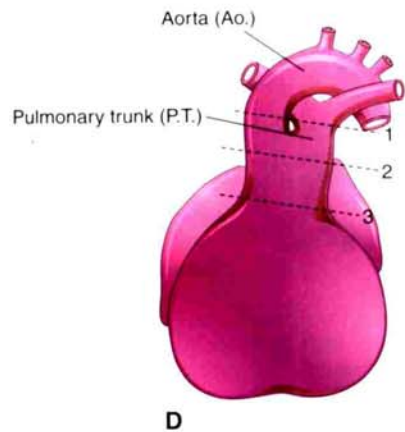
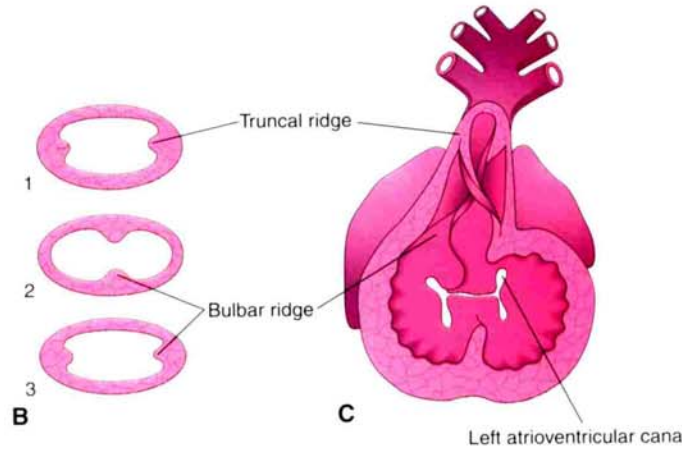
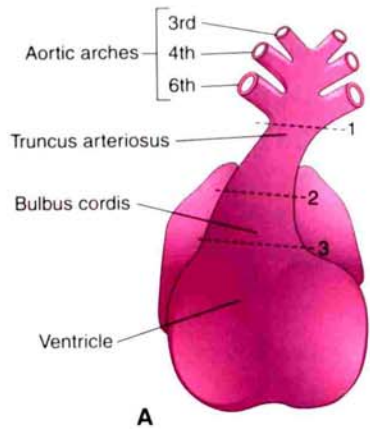
pressure load



obstr. ventric. outflow

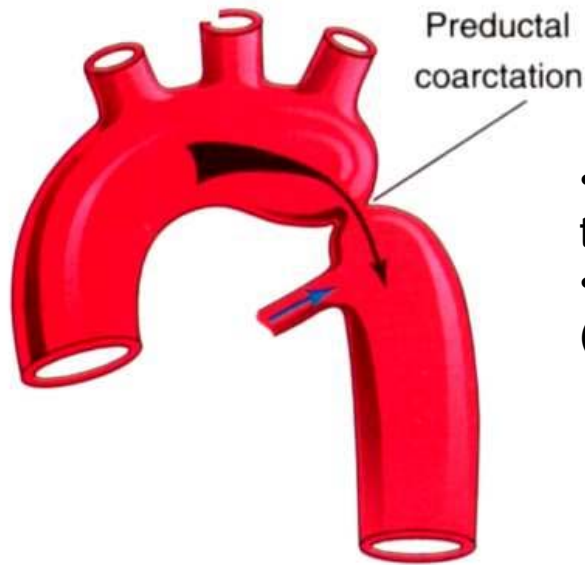


- pulmonary valve stenosis
- aortic valve stenosis
- coarctation of aorta

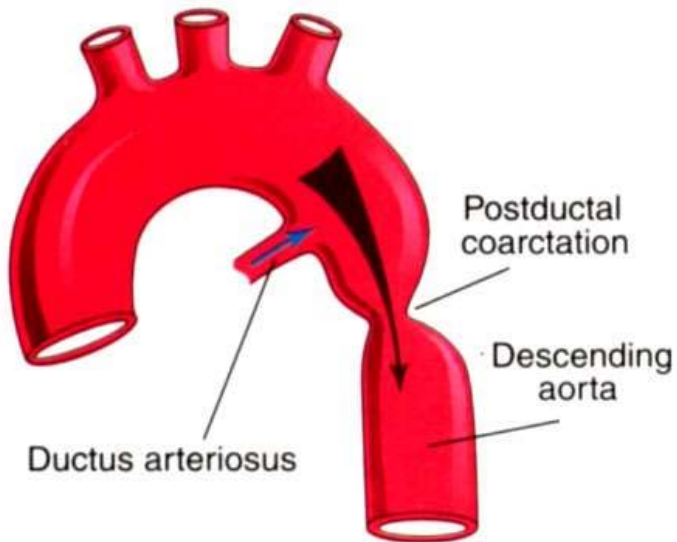




# Increased pressure load defects: Aortic coarctation

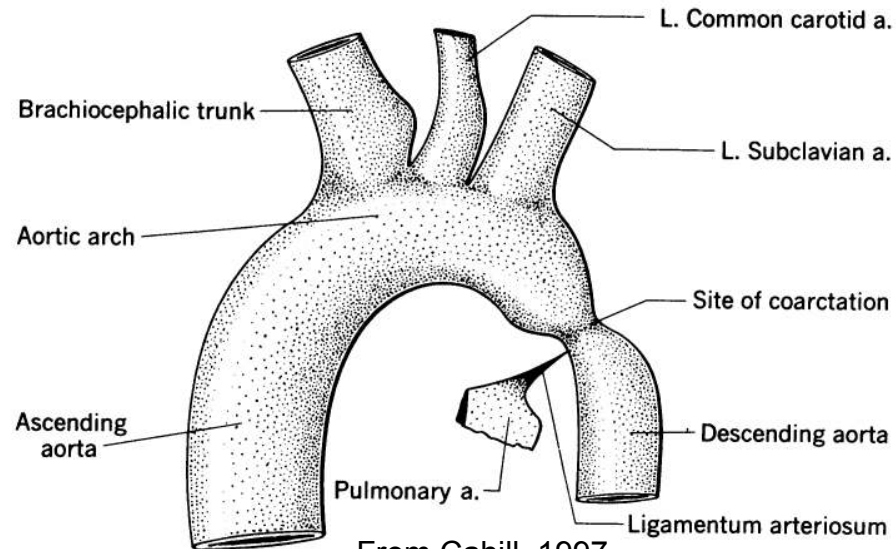
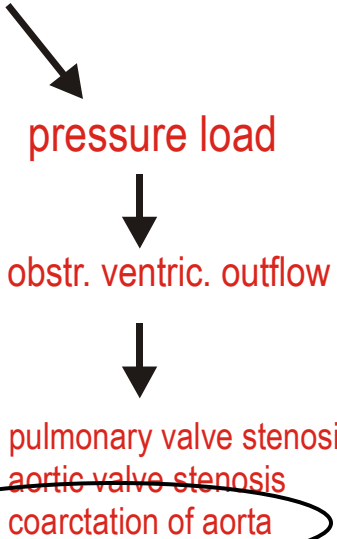


- Constriction of the aorta distal to the left subclavian artery
- Typically near ductus arteriosus (lig. arteriosum)
  - Preductal (= infantile)
  - Postductal (= "adult")
  - Juxtaductal



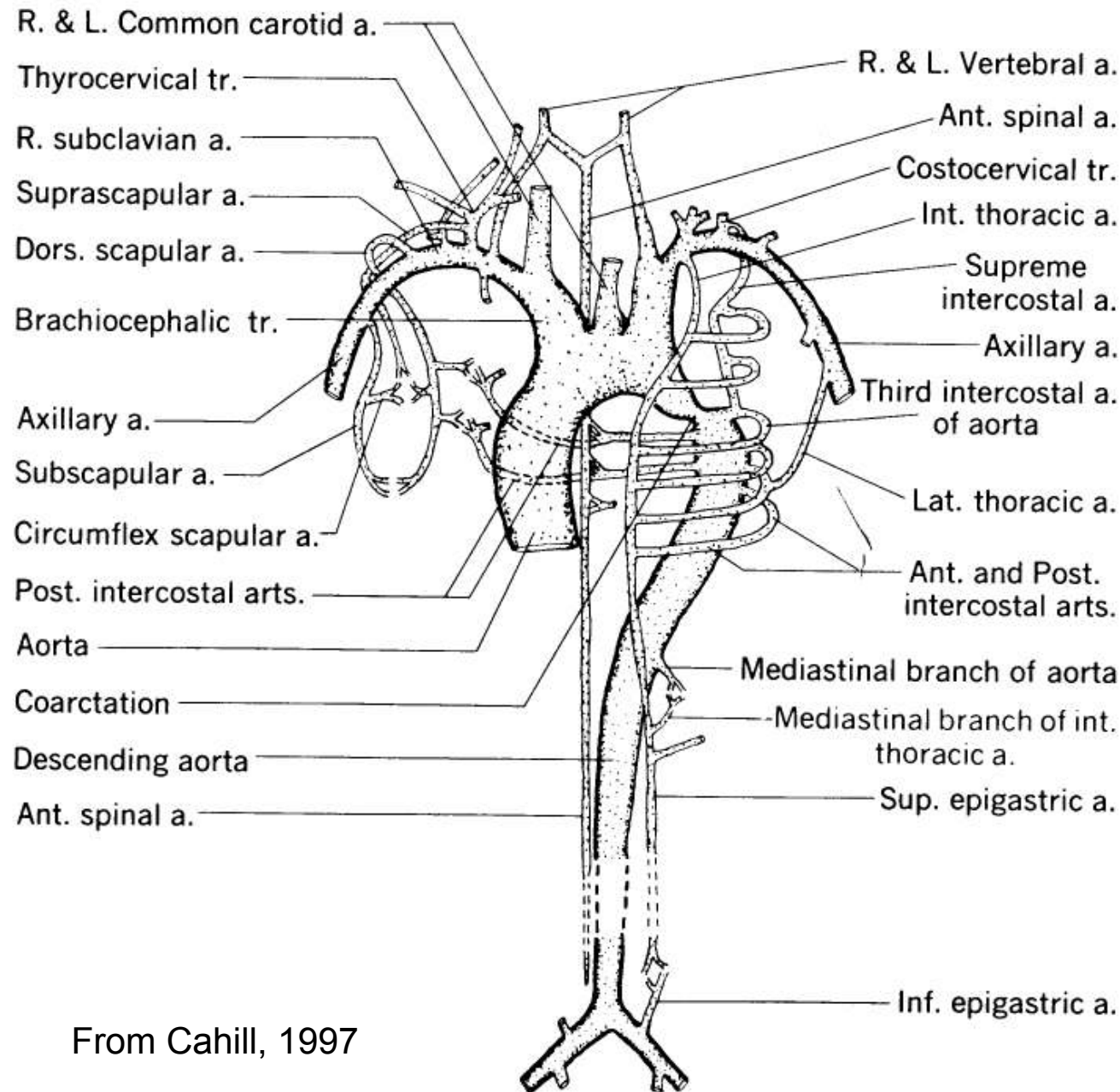
From Moore & Persaud 1998

Acyanotic



From Cahill, 1997

# Increased pressure load defects: Aortic coarctation



## Collateral Circulation

- Subclavian → IMA → intercostals → aorta
- Subclavian → IMA → sup. epigastr. → inf. epigastr. → iliac → aorta
- Subclavian → cervical & scap. branches → intercostals → aorta
- Subclavian → vertebral → ant. spinal → intercostals & lumbar → aorta

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pressure load

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- tetralogy of Fallot
- pulmonary atresia
- tricuspid atresia
- total anomalous pulm. return w/ obstruction

modified from Bernstein (1996) and other sources



# Increased pulmonary load defects: TGA

Cyanotic

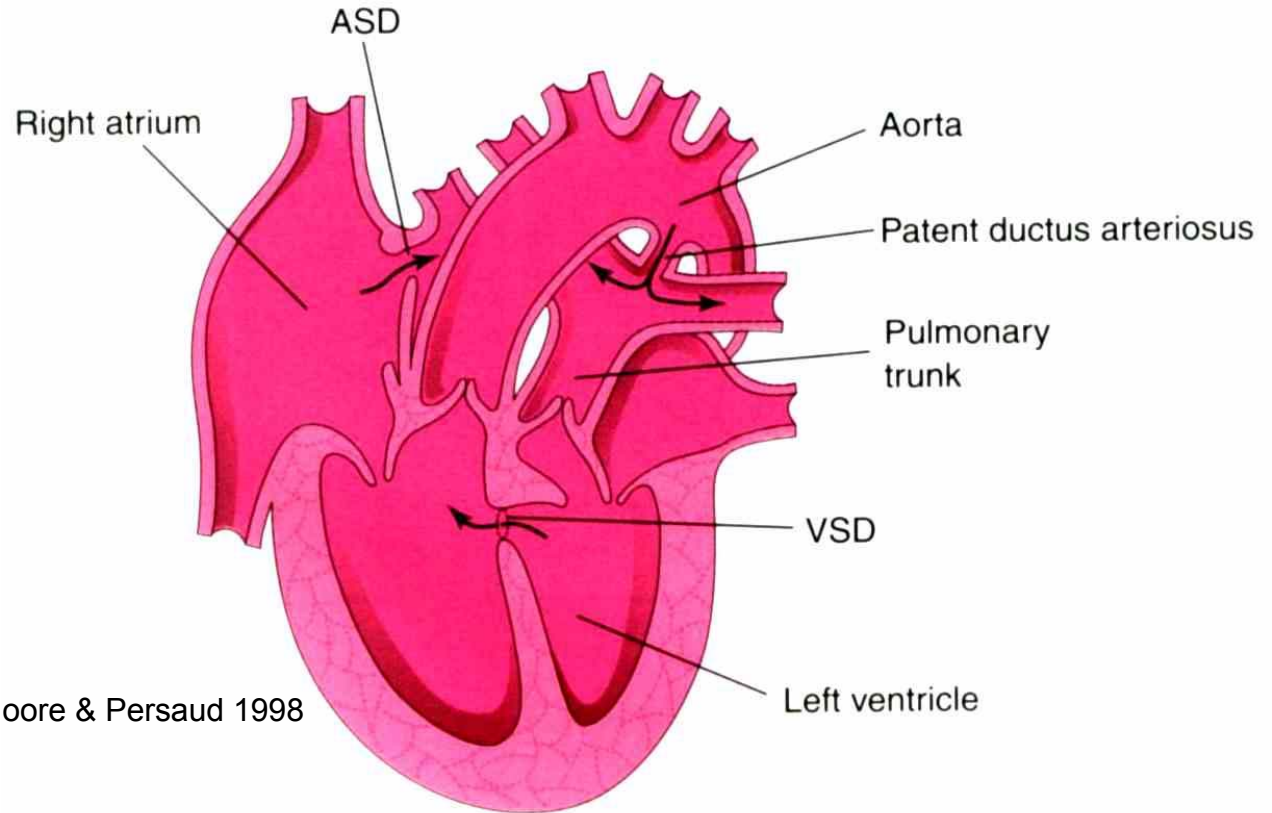


↑ pulmonary flow



- transpos. of gr. vessels
- single ventricle
- truncus arteriosus
- total anomalous pulm. return w/o obstruction

## Transposition of the Great Arteries (d-TGA)



From Moore & Persaud 1998

- Most common cyanotic neonatal heart defect
- Failure of aorticopulmonary septum to take a spiraling course
- Fatal without PDA, ASD, & VSD

# Increased pulmonary load defects: Truncus arteriosus

Cyanotic

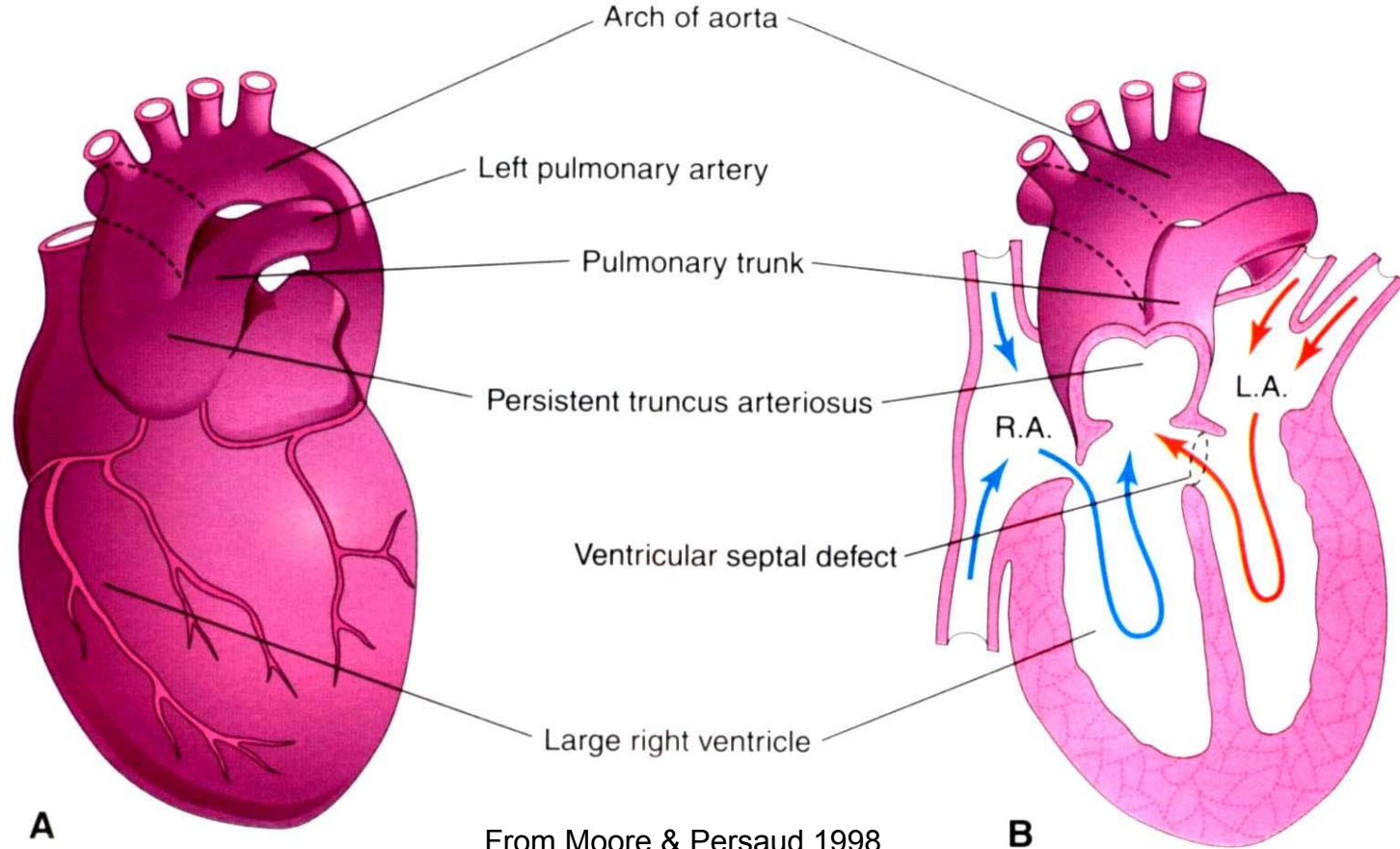


↑ pulmonary flow



- Single outflow tract from the heart
- Improper formation of truncal ridges & aorticopulmonary septum such that aorta & pulmonary trunk are not fully divided
- 1-2% of all CHDs

- transpos. of gr. vessels
- single ventricle
- truncus arteriosus
- total anomalous pulm. return w/o obstruction



From Moore & Persaud 1998

# Decreased pulmonary load defects: Tetralogy of Fallot

- 5-7% of all CHDs
- Four co-occurring heart defects
  - Pulmonary stenosis
  - Ventricular septal defect
  - Overriding aorta (dextroposition)
  - Right ventricular hypertrophy
- Asymmetrical fusion of bulbar & truncal ridges

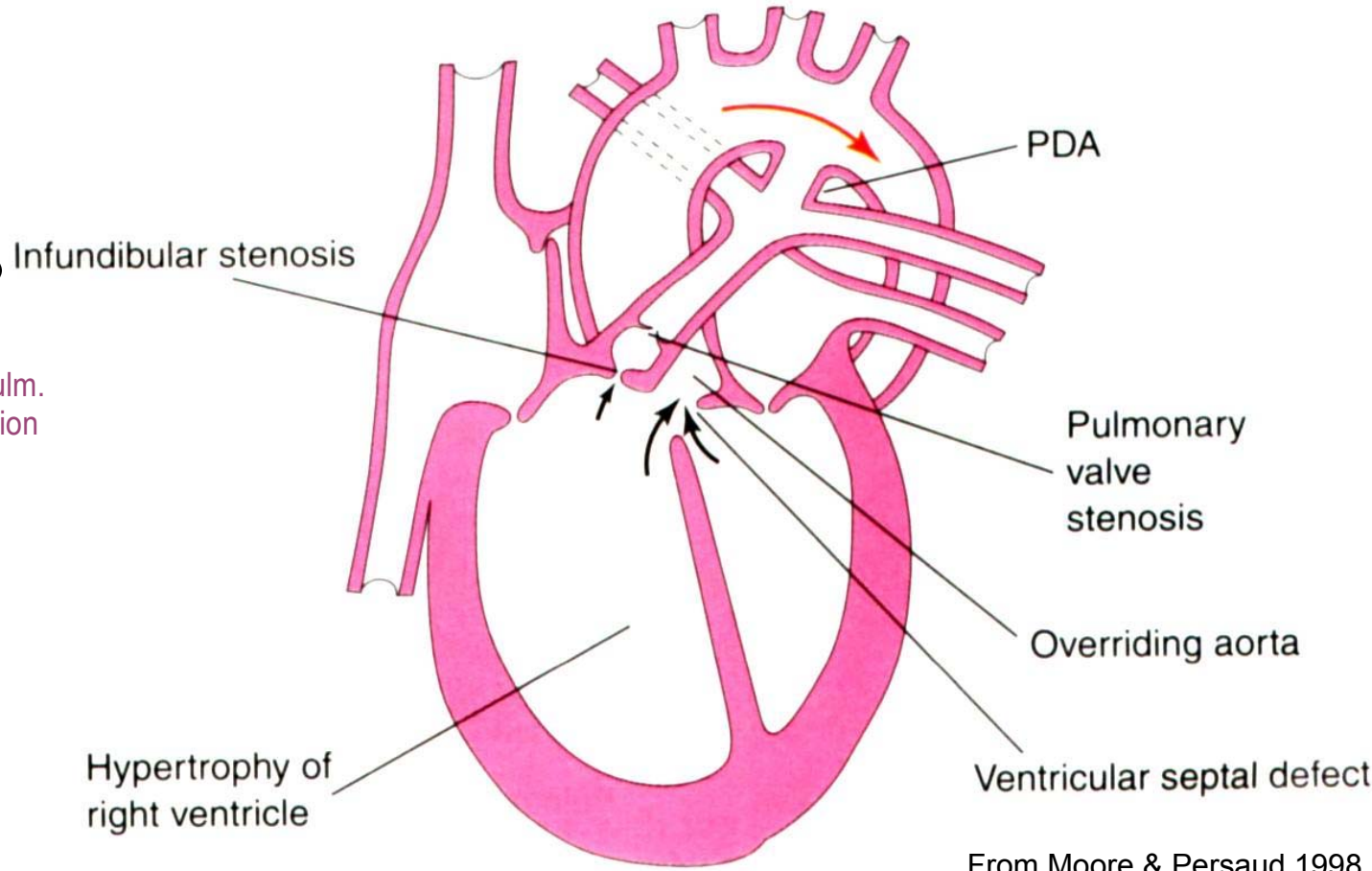
Cyanotic



↓ pulmonary flow



- tetralogy of Fallot
- pulmonary atresia
- tricuspid atresia
- total anomalous pulm. return w/ obstruction





## References: print sources

- Bernstein, D. 1996. The cardiovascular system; in *Nelson's Textbook of Pediatrics*. Saunders, Philadelphia.
- Cahill, D. R. 1997. *Lachman's Case Studies in Anatomy*. Oxford Univ. Press, New York.
- Moore, K. L. and T. V. N. Persaud. 1998. *The Developing Human: Clinically Oriented Embryology, 6<sup>th</sup> Ed.*, Saunders, Philadelphia.

## References: internet

- <http://www.med.yale.edu/intmed/cardio/chd>
- <http://www.pediheart.org/parents/defects/index.html>
- <http://www.childrenheartinstitute.org/educate/eduhome.htm>
- <http://www.tmc.edu/thi/congenit.html>
- <http://www.kumc.edu/kumcpeds/cardiology/cardiology.html>
- <http://www.congenitalheartdefects.com/typesofCHD.html>