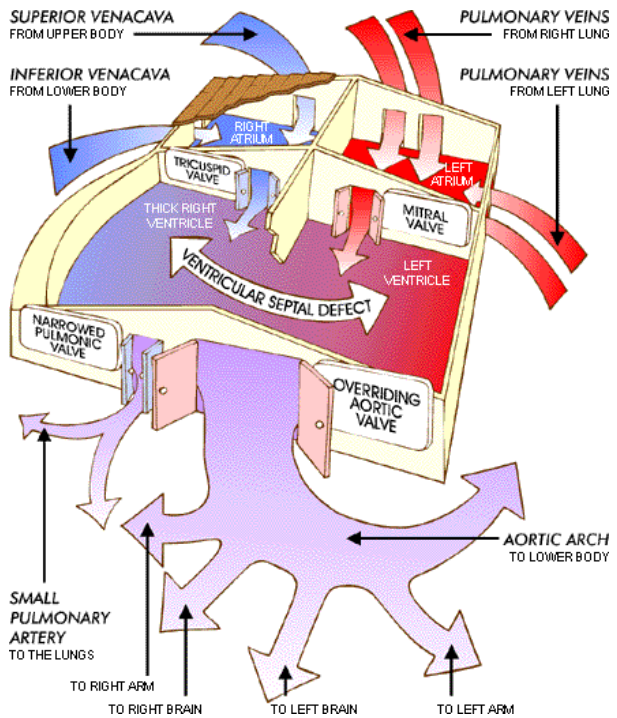




Developmental anatomy of the heart and the embryological basis for cardiac defects

21 August 2002

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

Lawrence M. Witmer, PhD

Department of Biomedical Sciences

College of Osteopathic Medicine

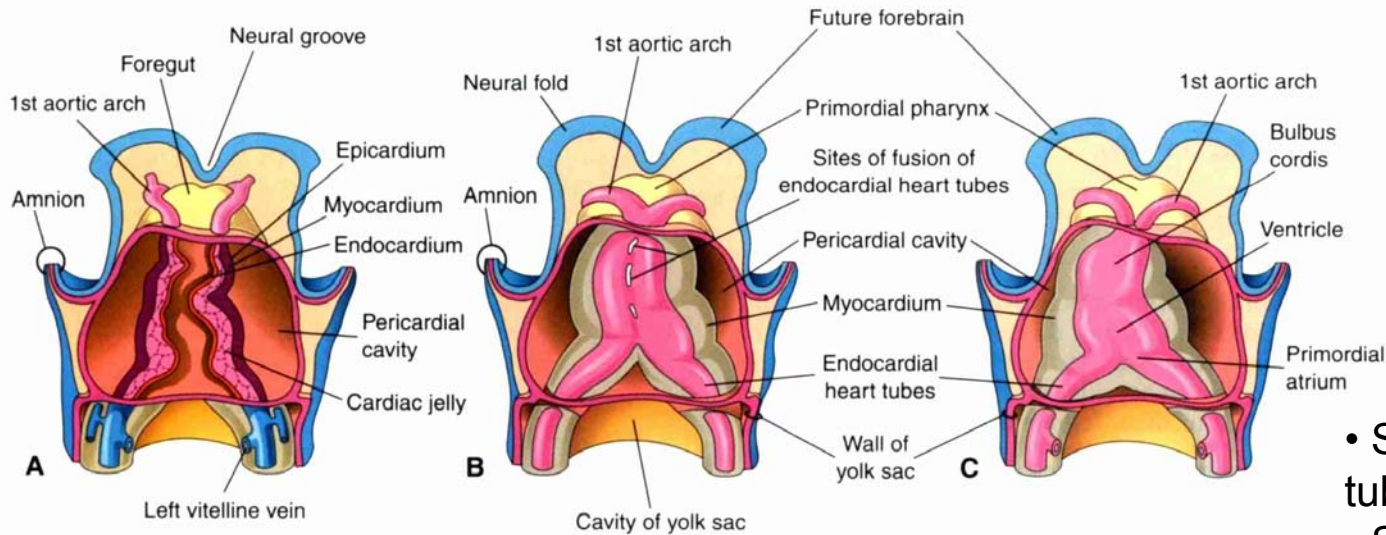
Ohio University

Athens, Ohio 45701

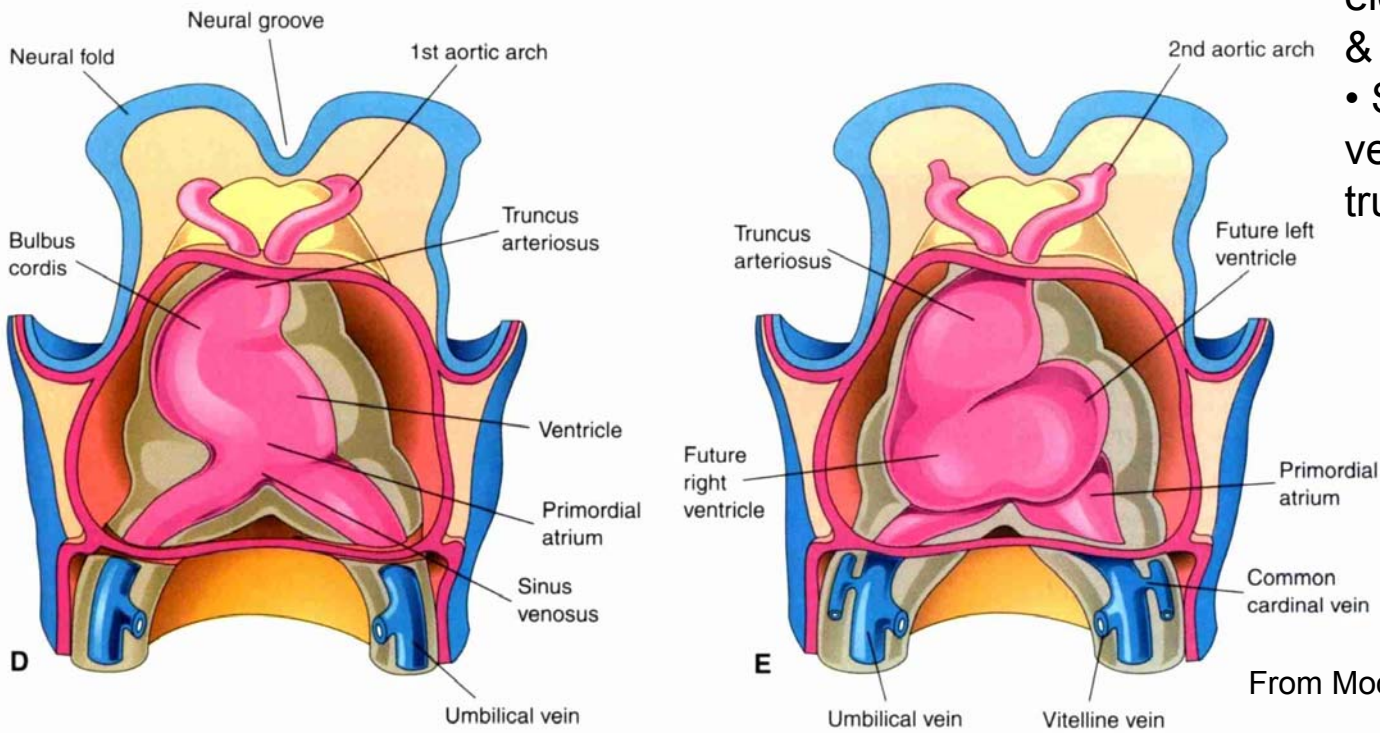
witmer@exchange.oucom.ohiou.edu



Formation of Endocardial Tube

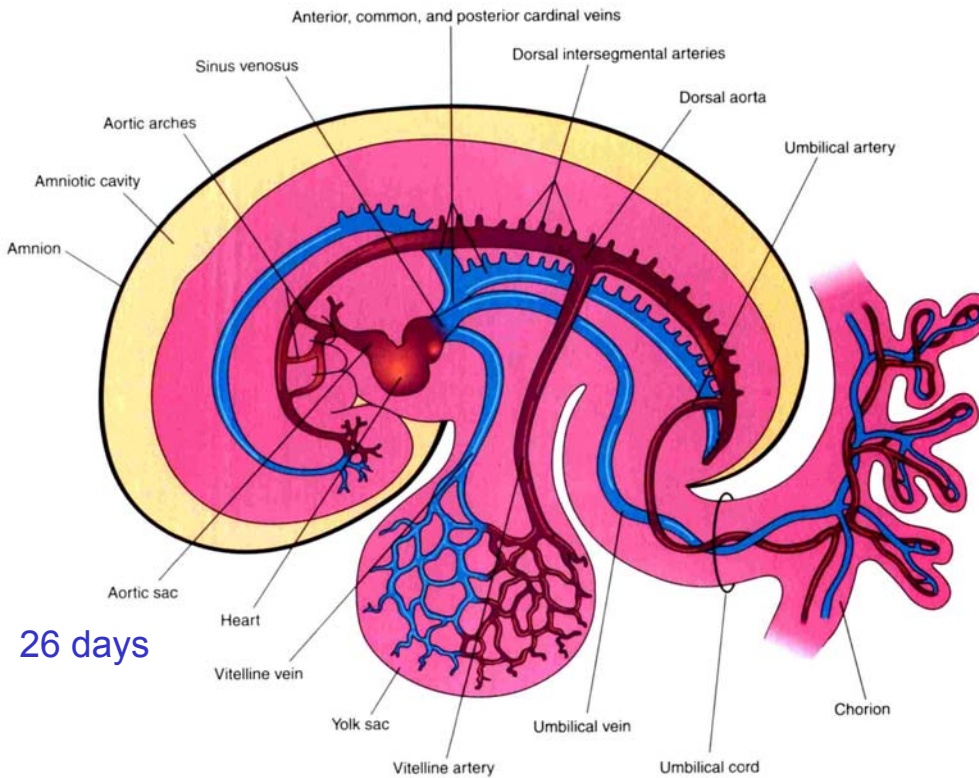


- Separate endocardial tubes that fuse
- Single endocardial tube elongates, forms dilations & constrictions
- Sinus venosus, atrium, ventricle, bulbus cordis, truncus arteriosus

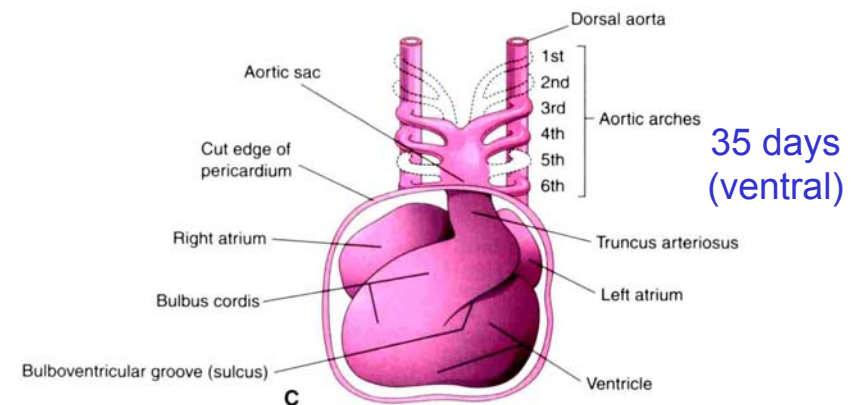
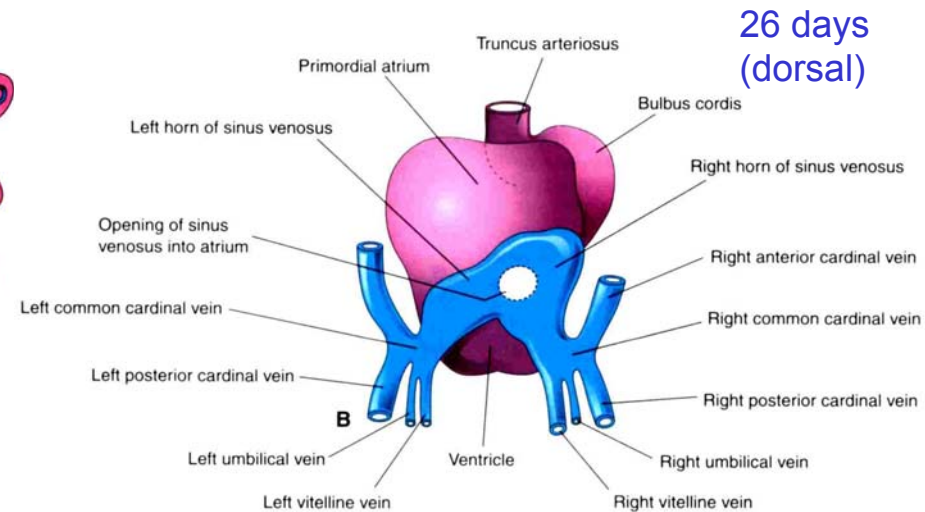
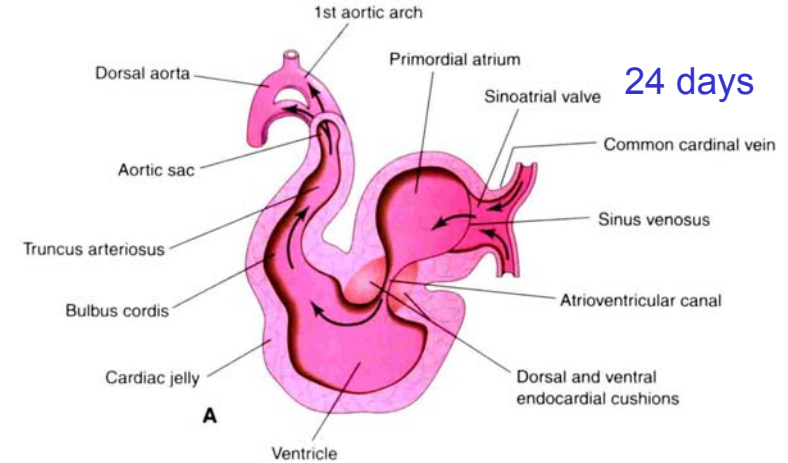


From Moore & Persaud 1998

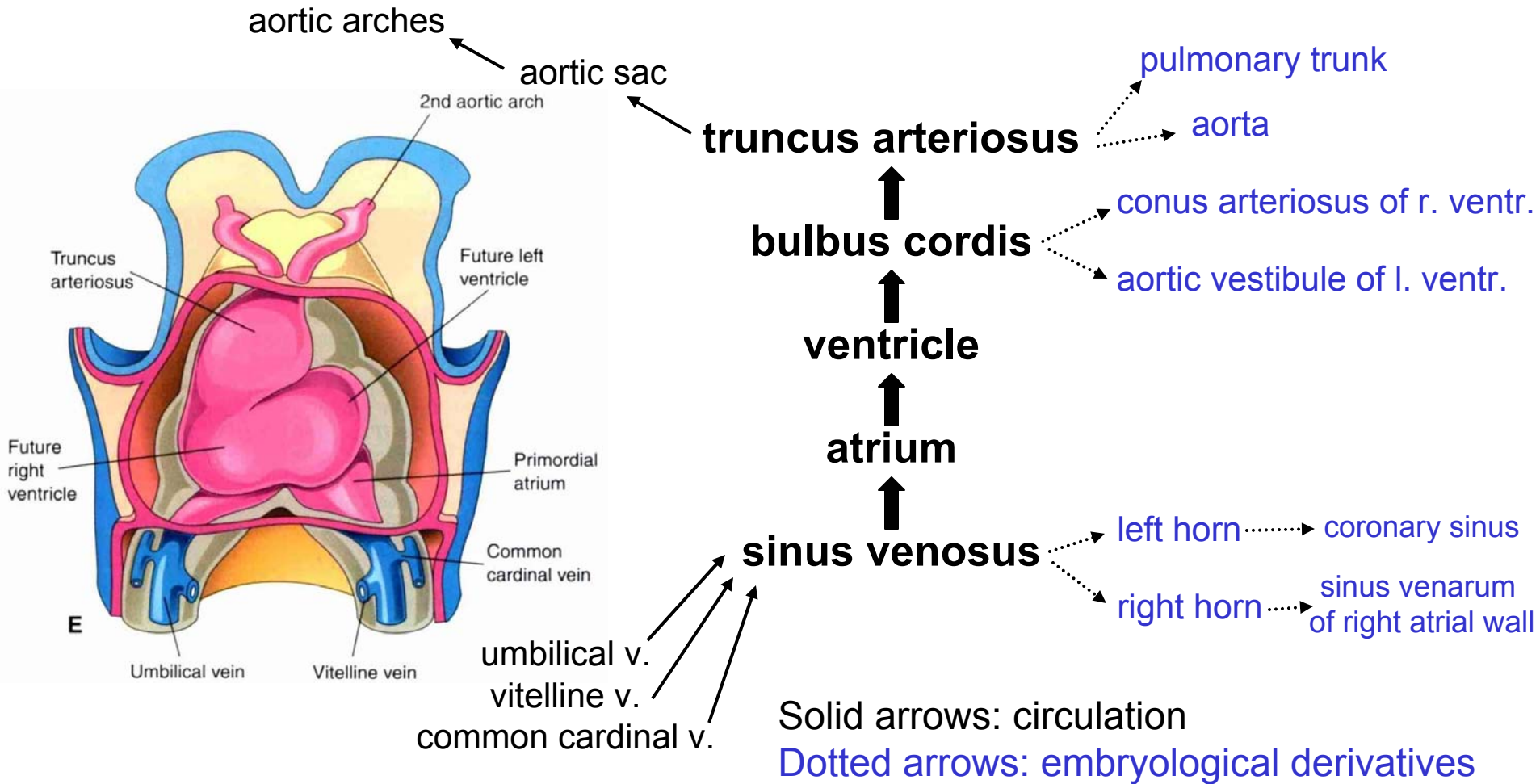
General Organization



From Moore & Persaud 1998



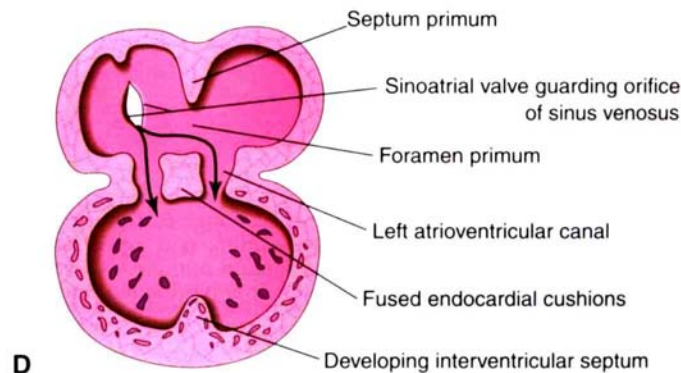
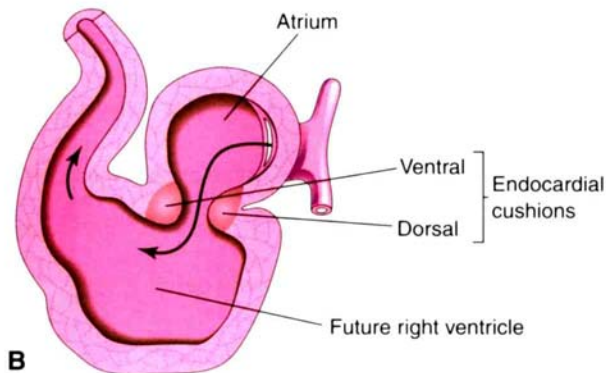
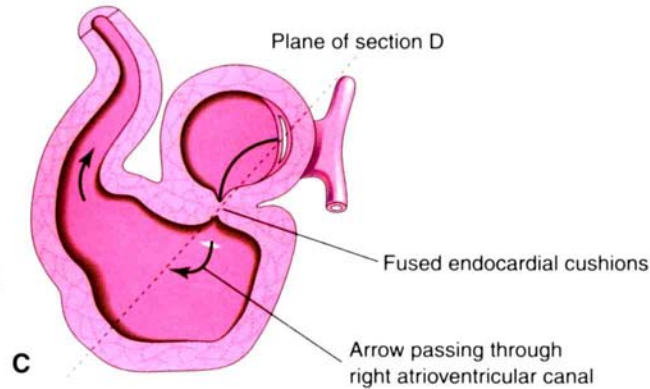
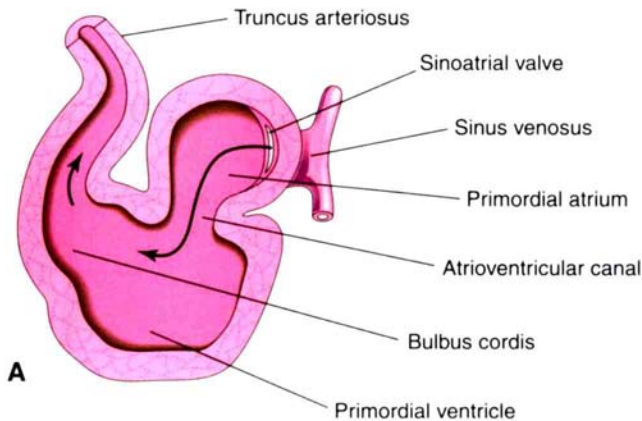
Blood Flow and Embryological Fates



Partitioning

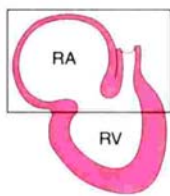
Endocardial cushions

- 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



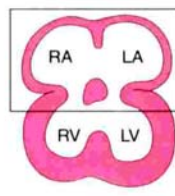
Weeks 4-5

From Moore & Persaud 1998



RA, right atrium
RV, right ventricle

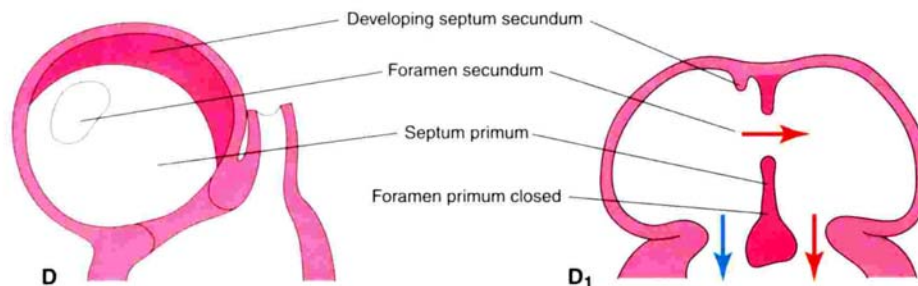
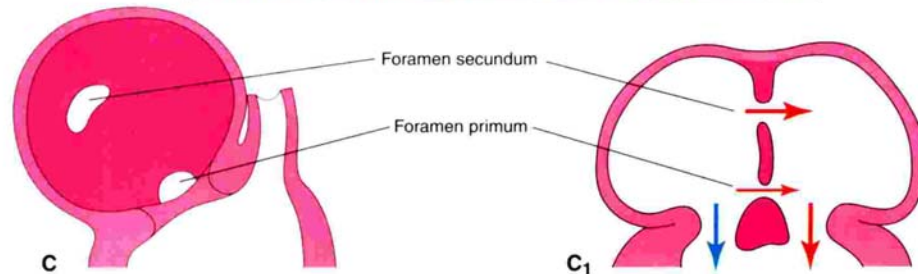
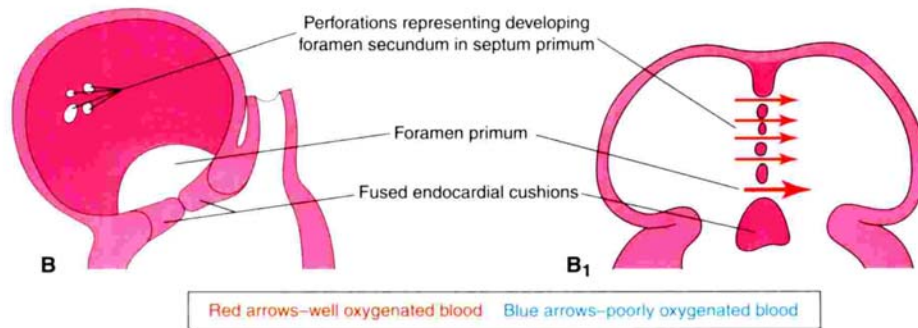
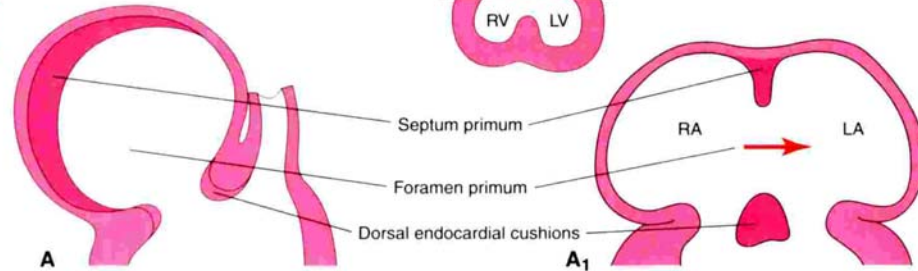
**viewed
from right**



LA, left atrium
LV, left ventricle

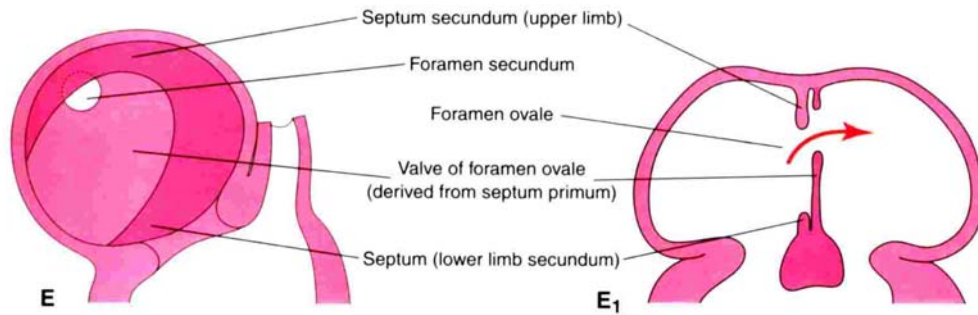
**coronal
sections**

Atrial Partitioning I



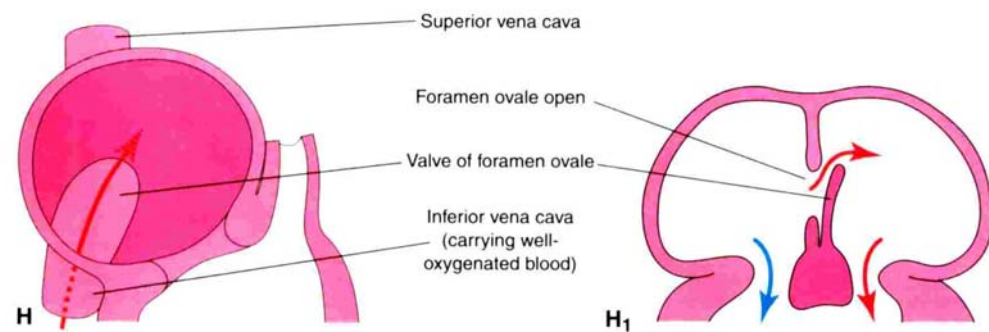
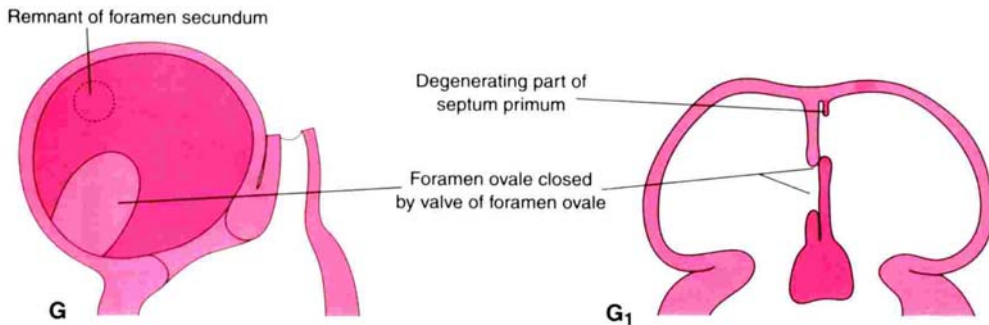
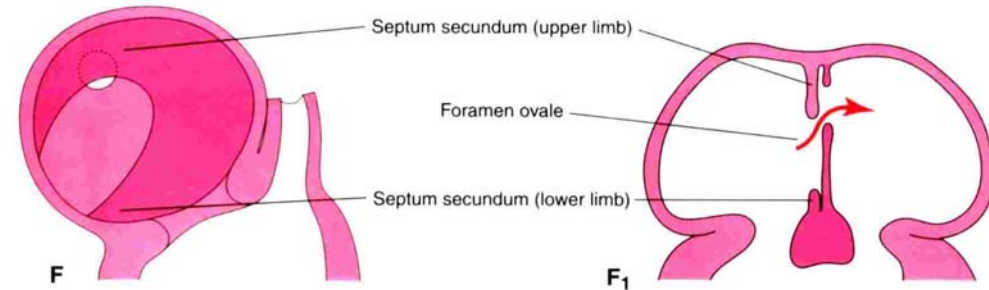
- Septum primum grows from atrial roof toward endocardial cushions
- Foramen primum: shunt that closes
- Foramen secundum: perforates septum primum, allowing shunt
- Septum secundum grows down, overlapping foramen secundum

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

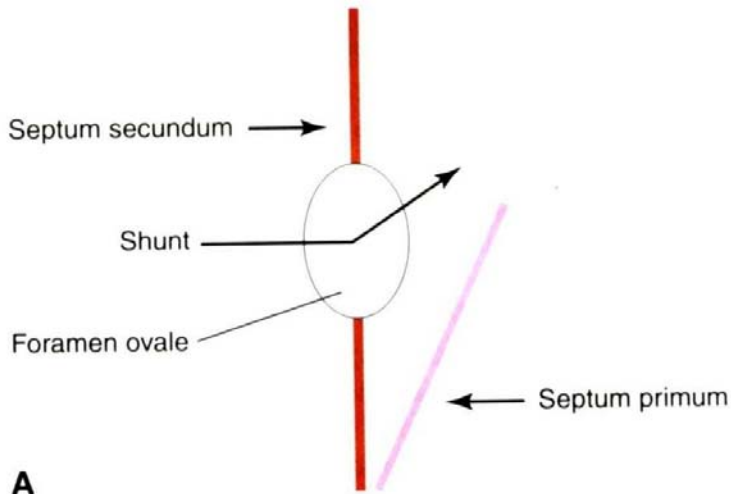


From Moore & Persaud 1998

BEFORE BIRTH

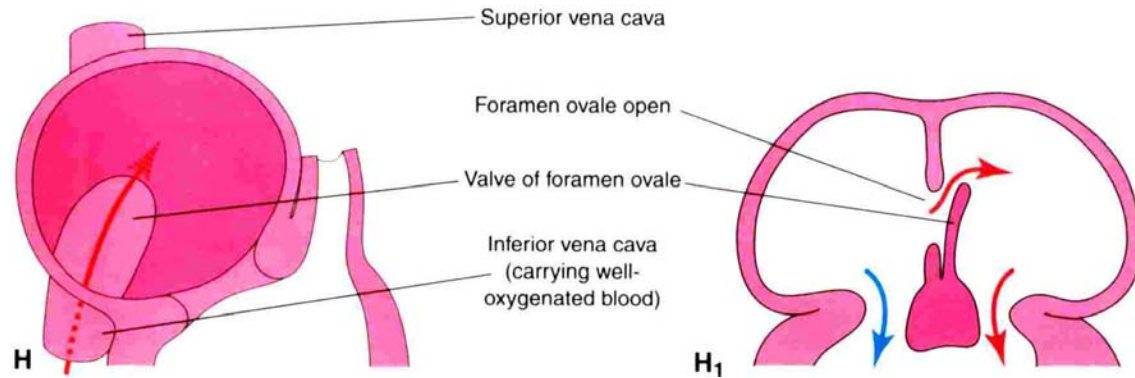
RIGHT ATRIUM
HIGHER PRESSURE

LEFT ATRIUM
LOWER PRESSURE



A

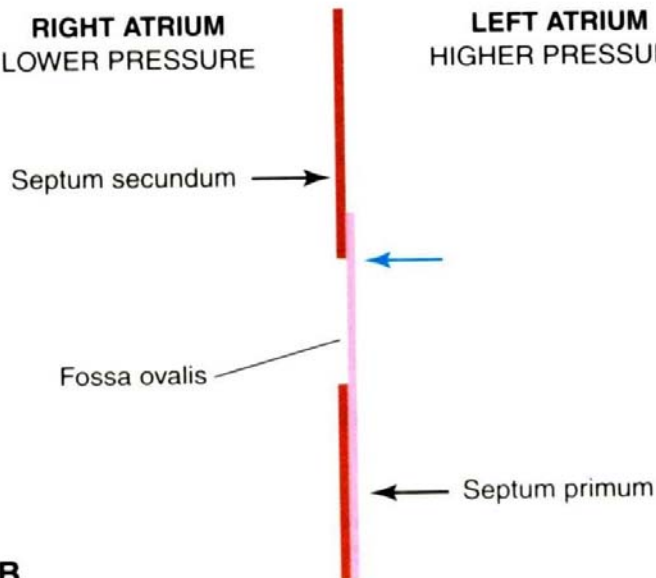
Atrial Partitioning III



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

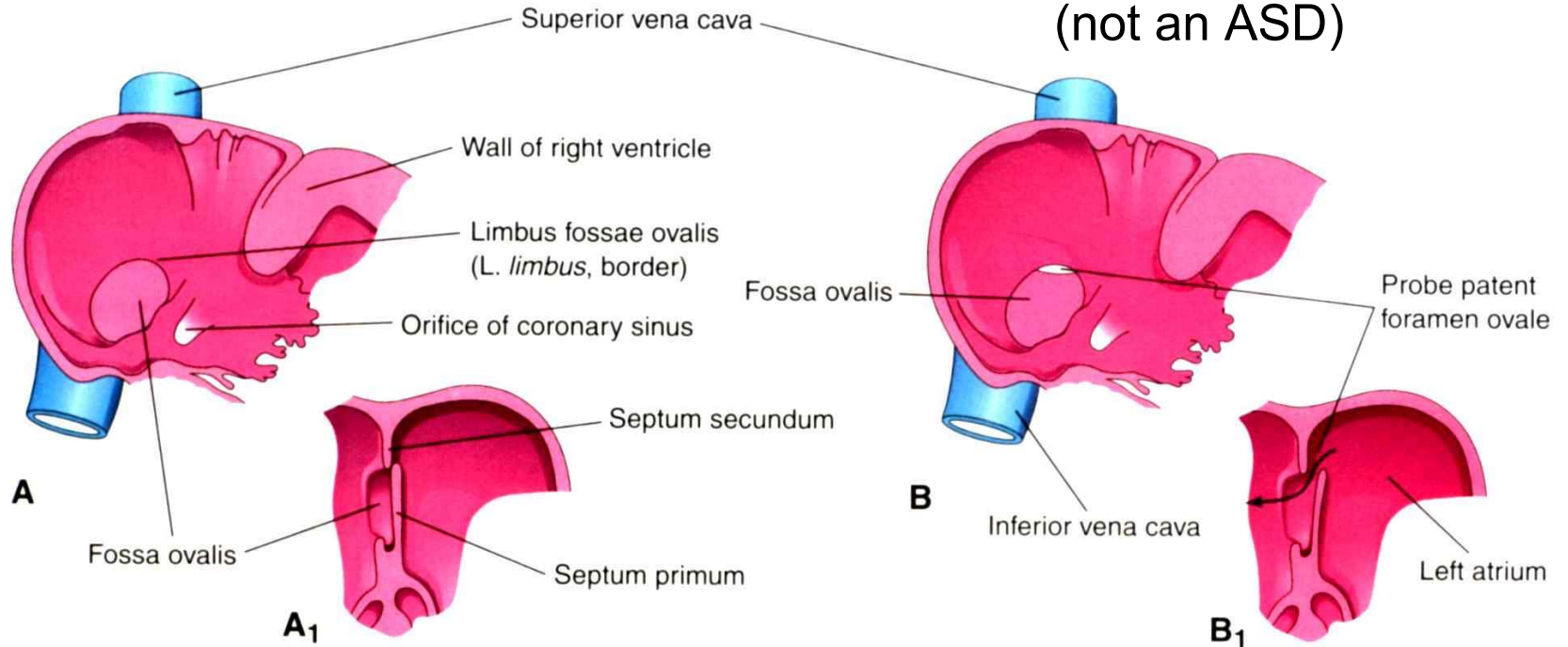
• After birth

- right side low pressure (low pulmonary resistance)
- valve remains closed (physiological closure)
- valve eventually fuses (anatomical closure): fossa ovalis

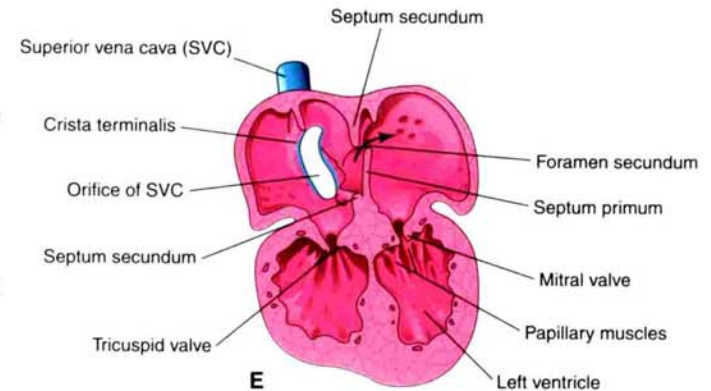
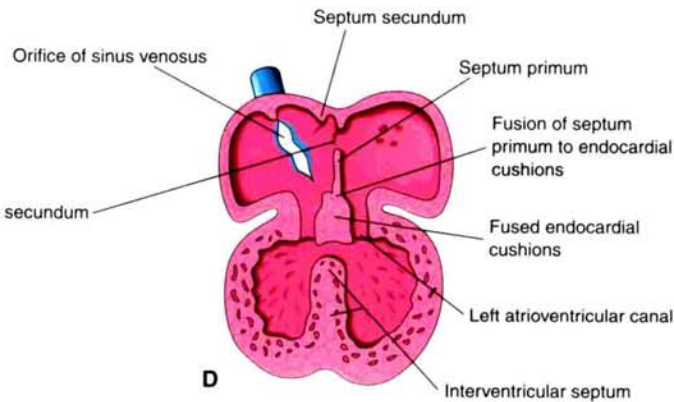
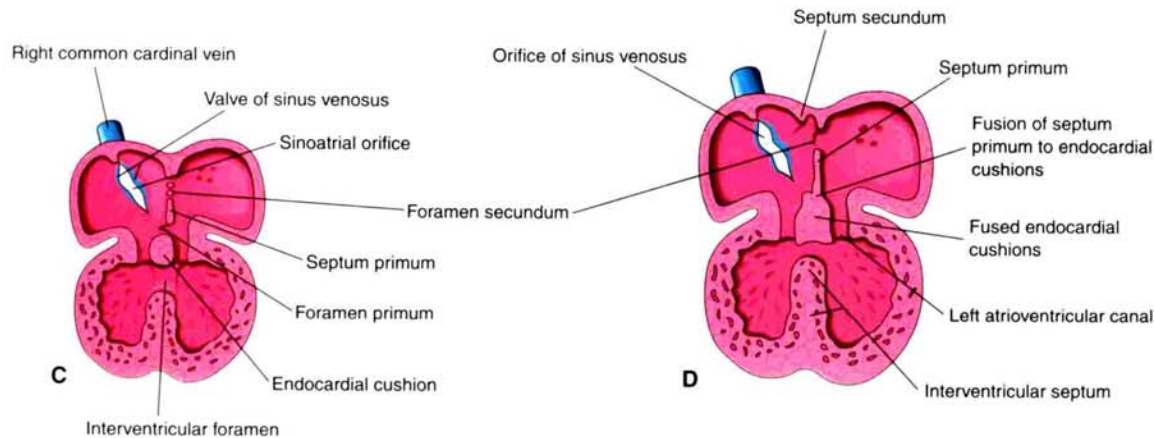
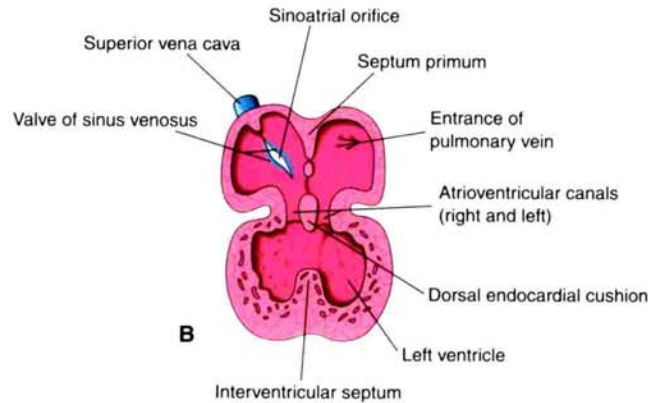
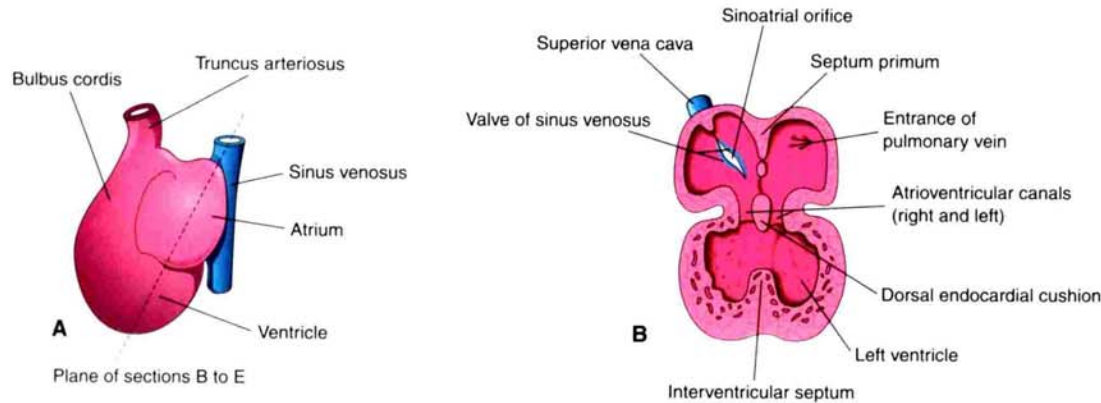
Atrial Partitioning IV

postnatal

probe patent
foramen ovale
(not an ASD)



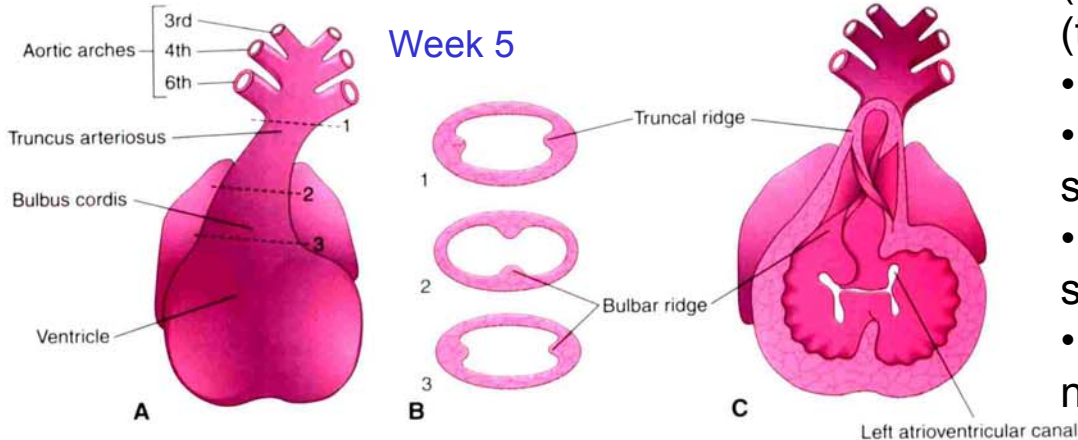
Ventricular Partitioning



- 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

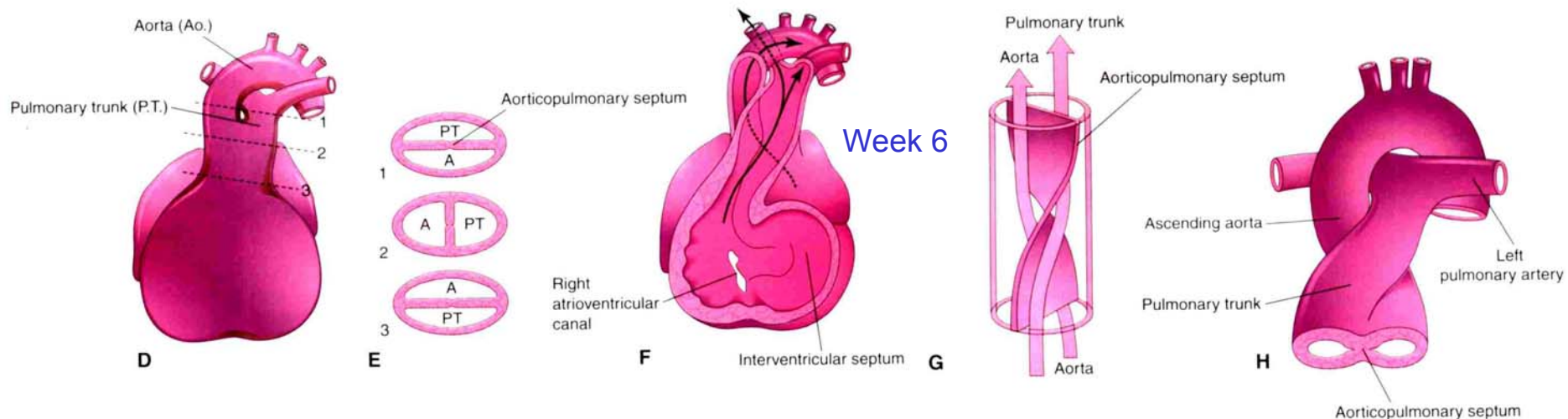
Partitioning of Truncus Arteriosus

Week 5

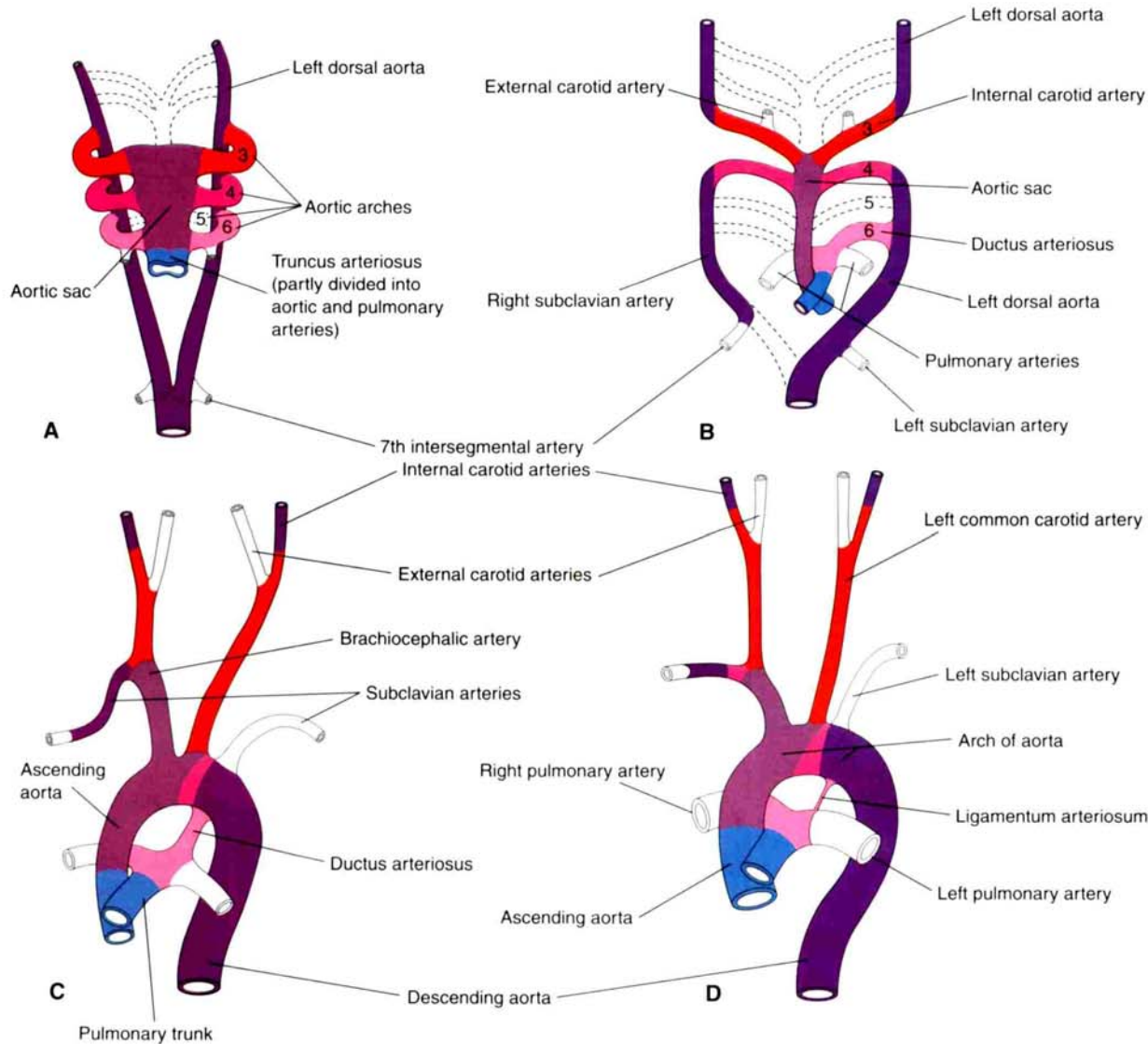


- continuous set of ridges in bulbus cordis (bulbar ridges) and truncus arteriosus (truncal ridges)
- grow toward each other, spiraling 180°
- fuse to form spiraling aortopulmonary septum, dividing aorta & pulmonary trunk
- bulbar ridges involved in formation of IV septum
- bulbar & truncal ridges derived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

Week 6



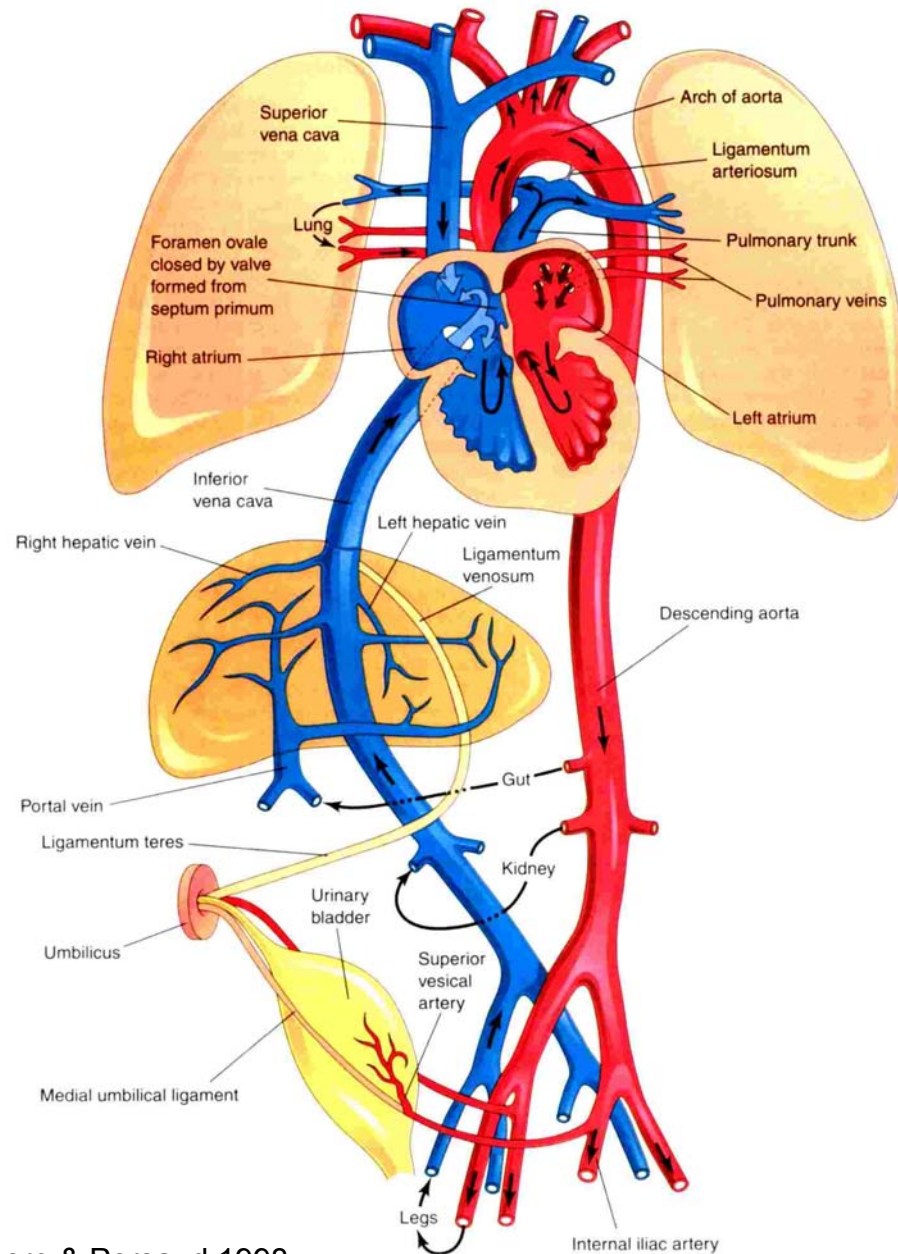
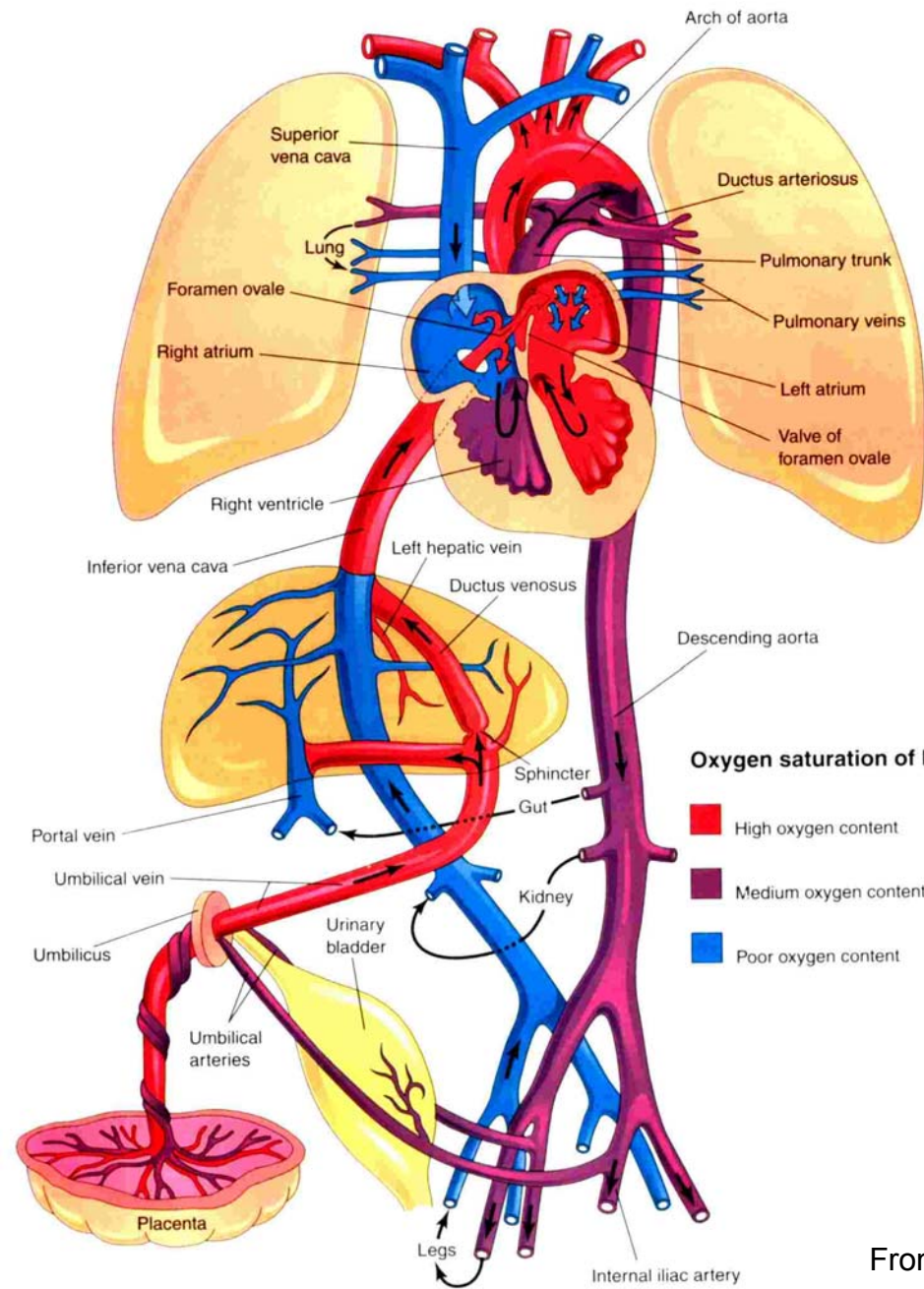
Aortic arches: Ductus arteriosus



- 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

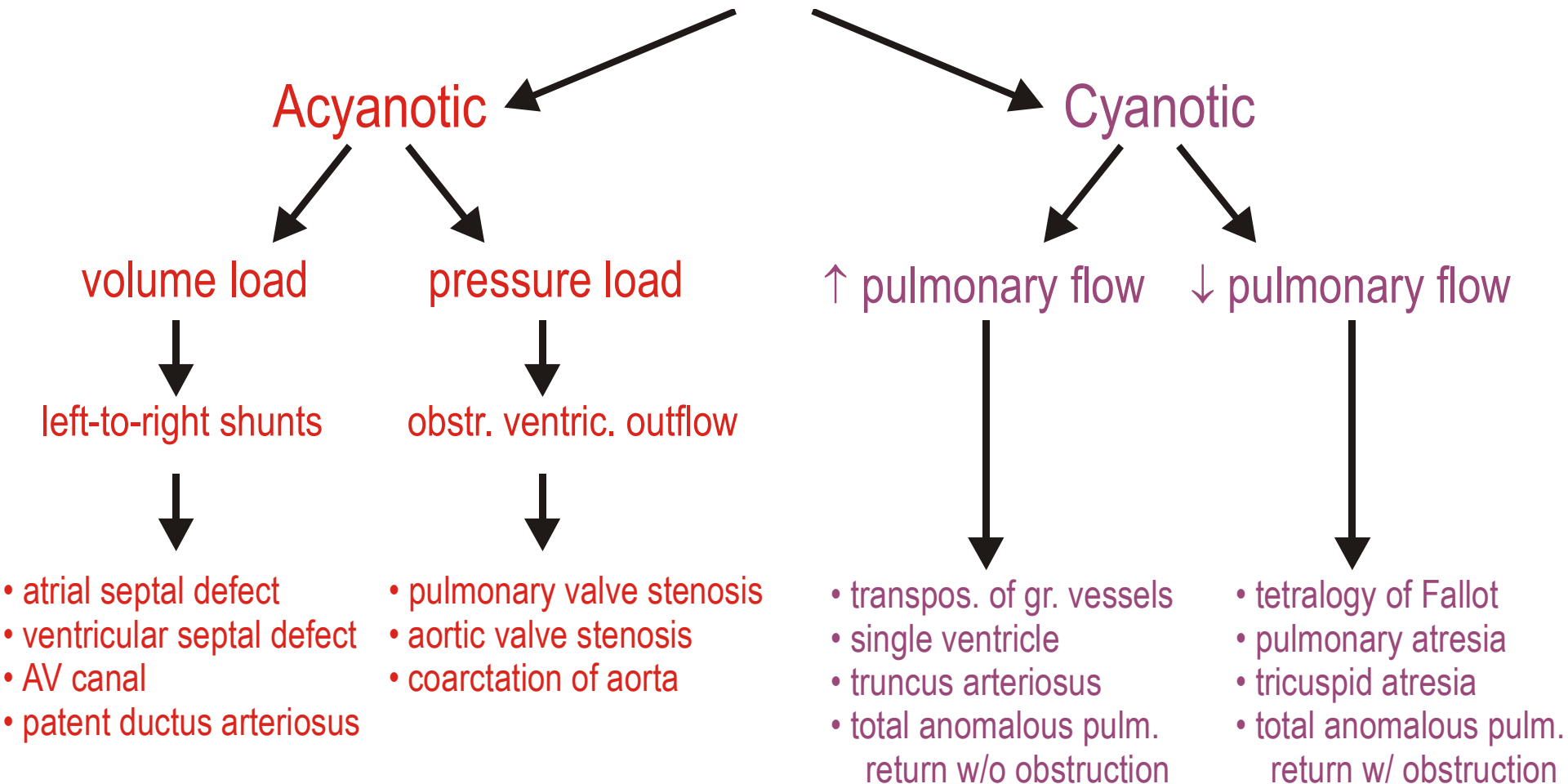
From Moore & Persaud 1998

Perinatal Circulation



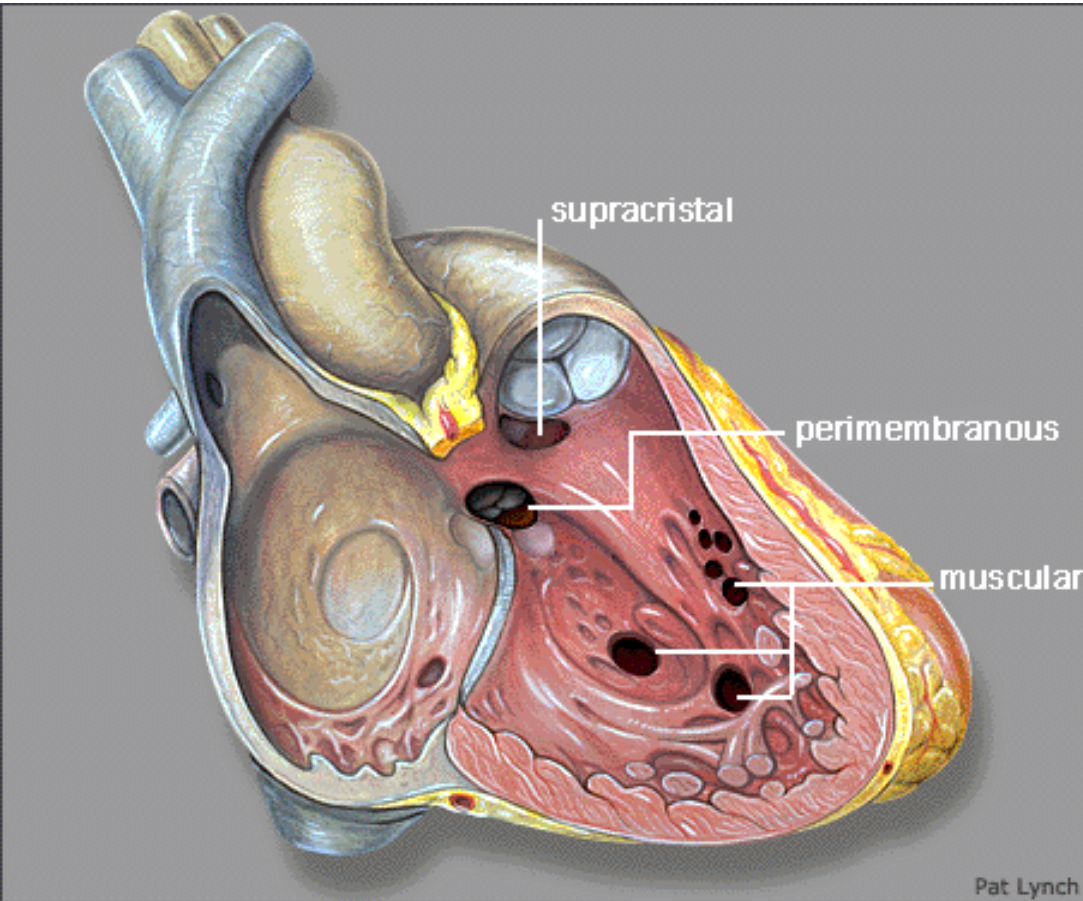
From Moore & Persaud 1998

Congenital Heart Defects



modified from Bernstein (1996) and other sources

Ventricular Septal Defects (VSD)



Acyanotic

volume load

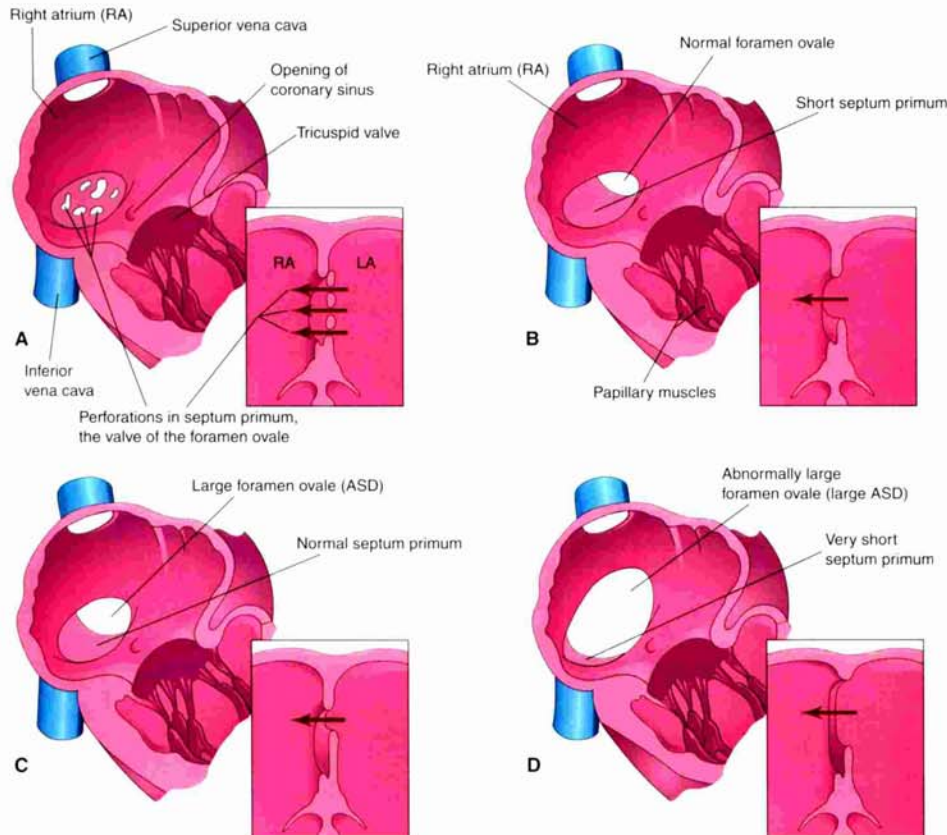
left-to-right shunts

- 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)

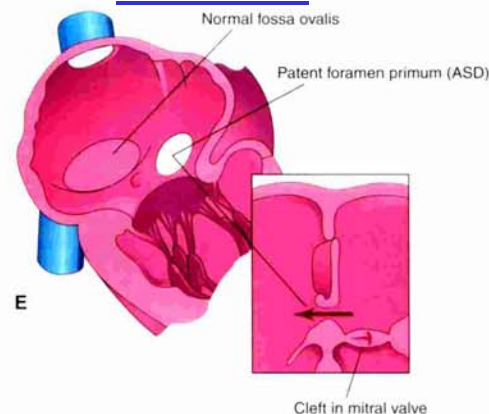
Secundum ASDs



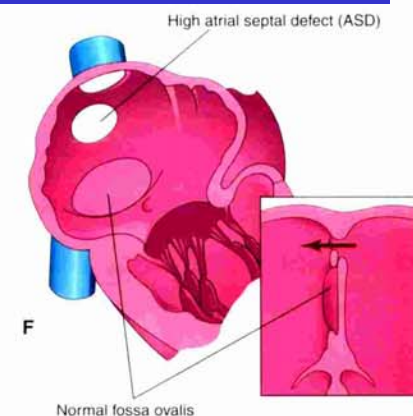
From Moore & Persaud 1998

- (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



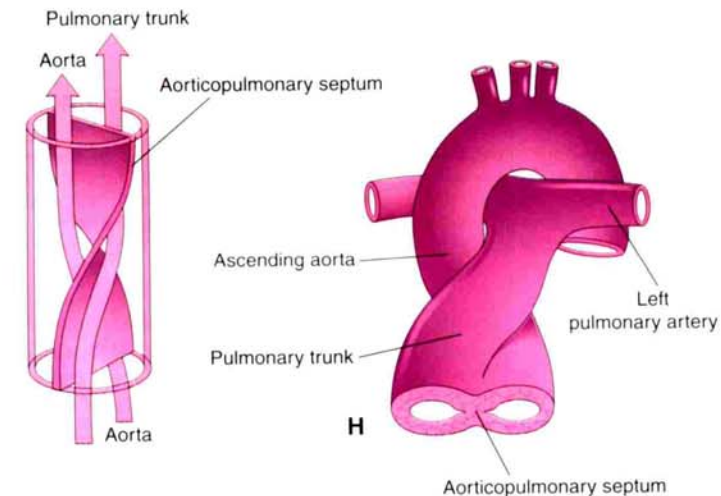
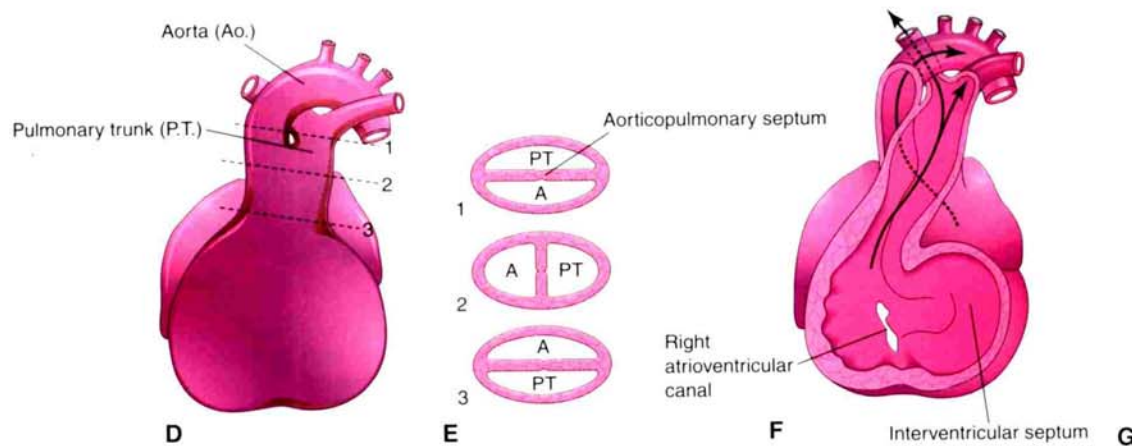
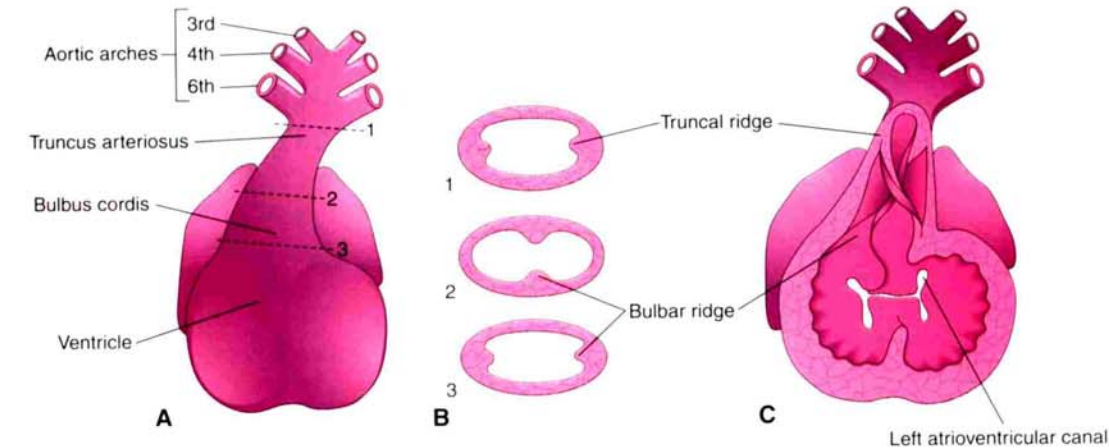
pressure load



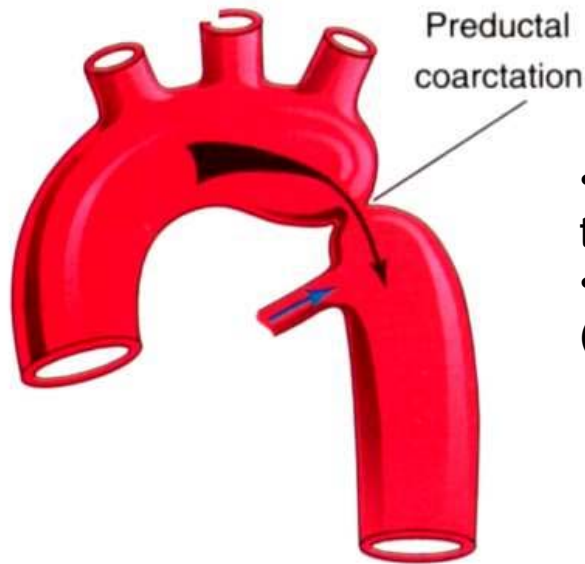
obstr. ventric. outflow



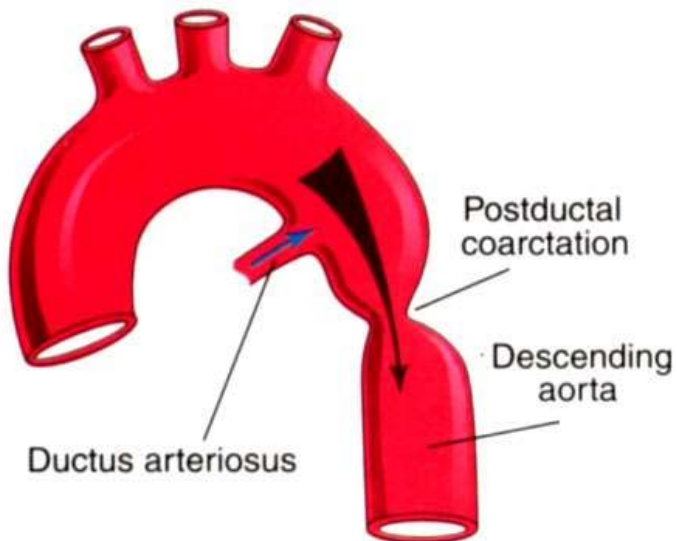
- pulmonary valve stenosis
- aortic valve stenosis
- coarctation of aorta



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



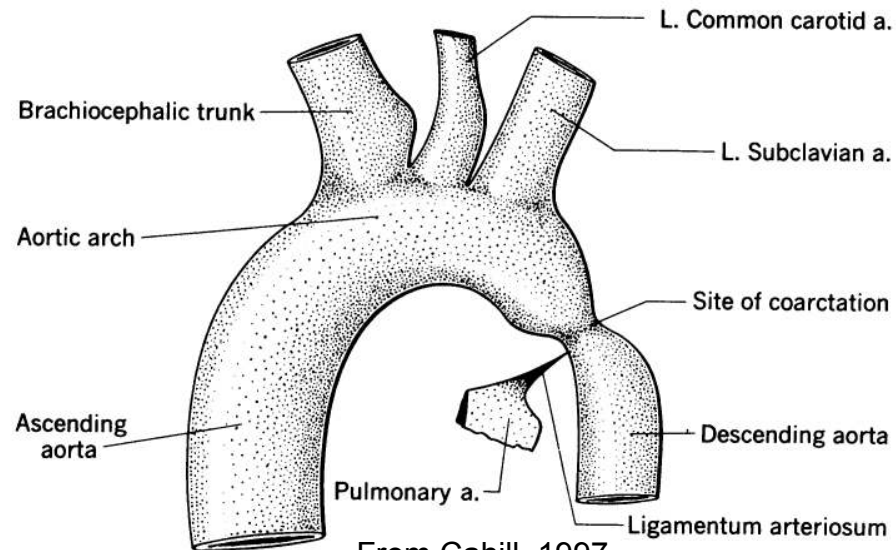
From Moore & Persaud 1998

Acyanotic

pressure load

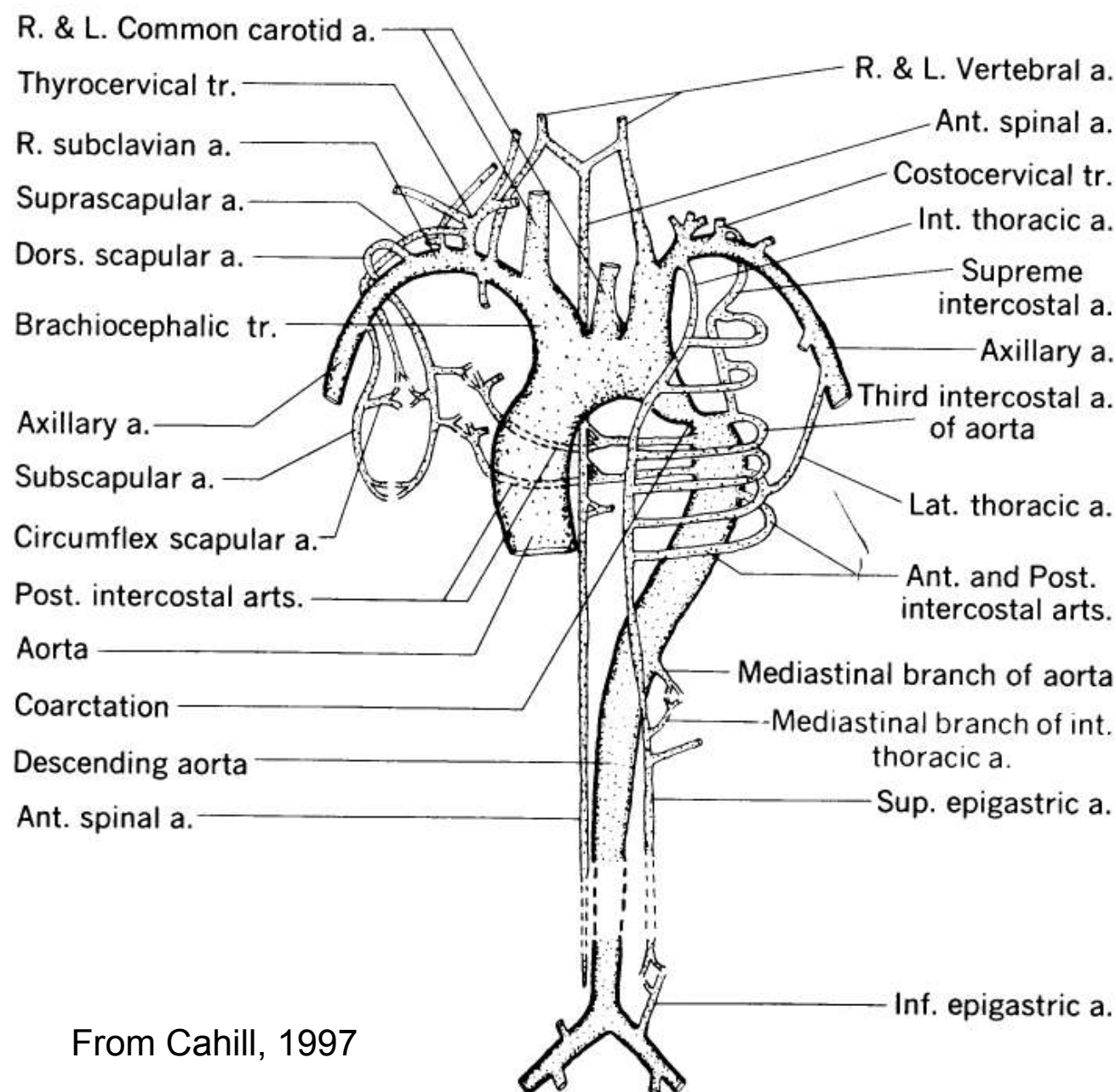
obstr. ventric. outflow

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



From Cahill, 1997

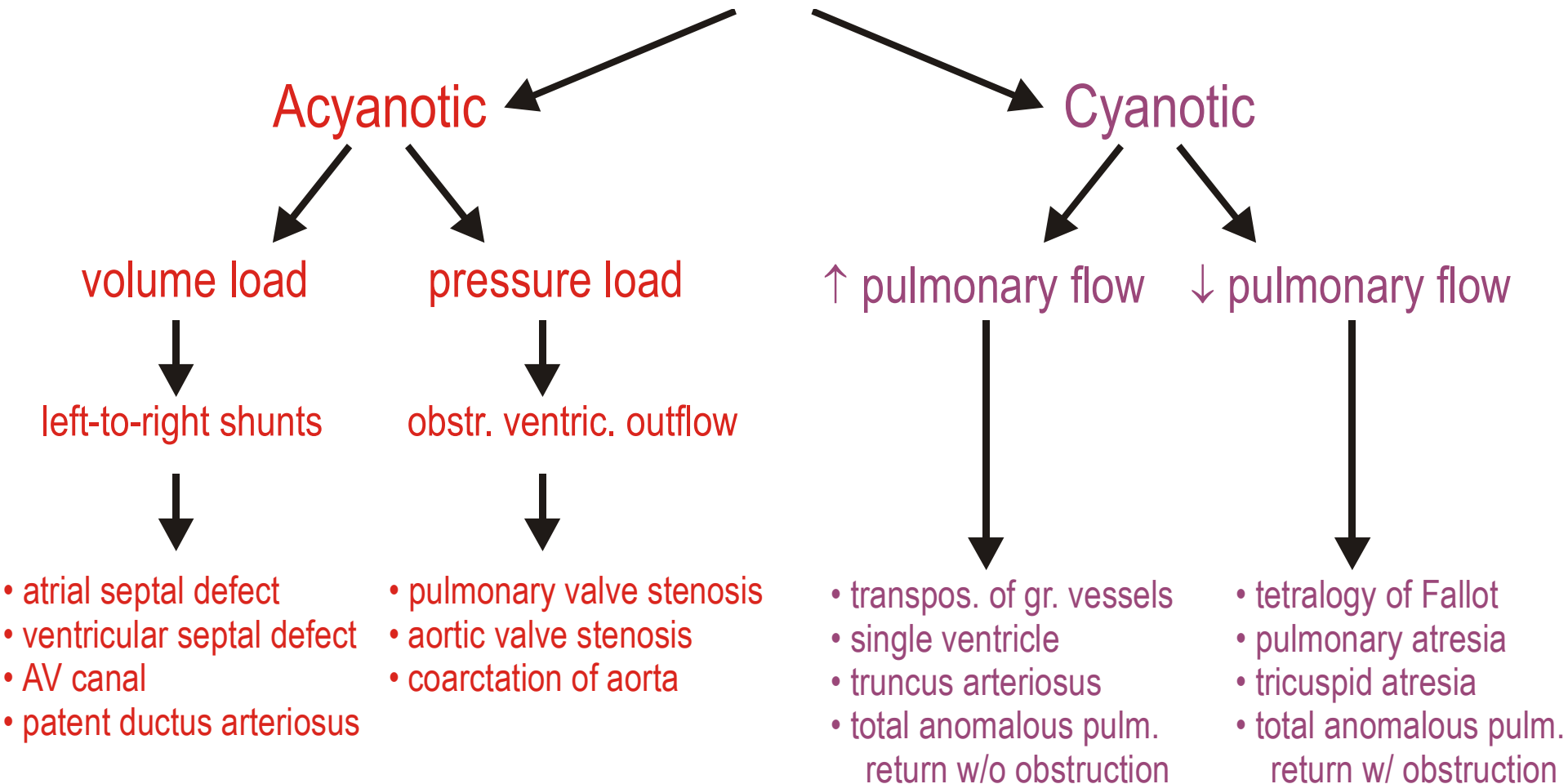
Increased pressure load defects: Aortic coarctation



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

Congenital Heart Defects



modified from Bernstein (1996) and other sources

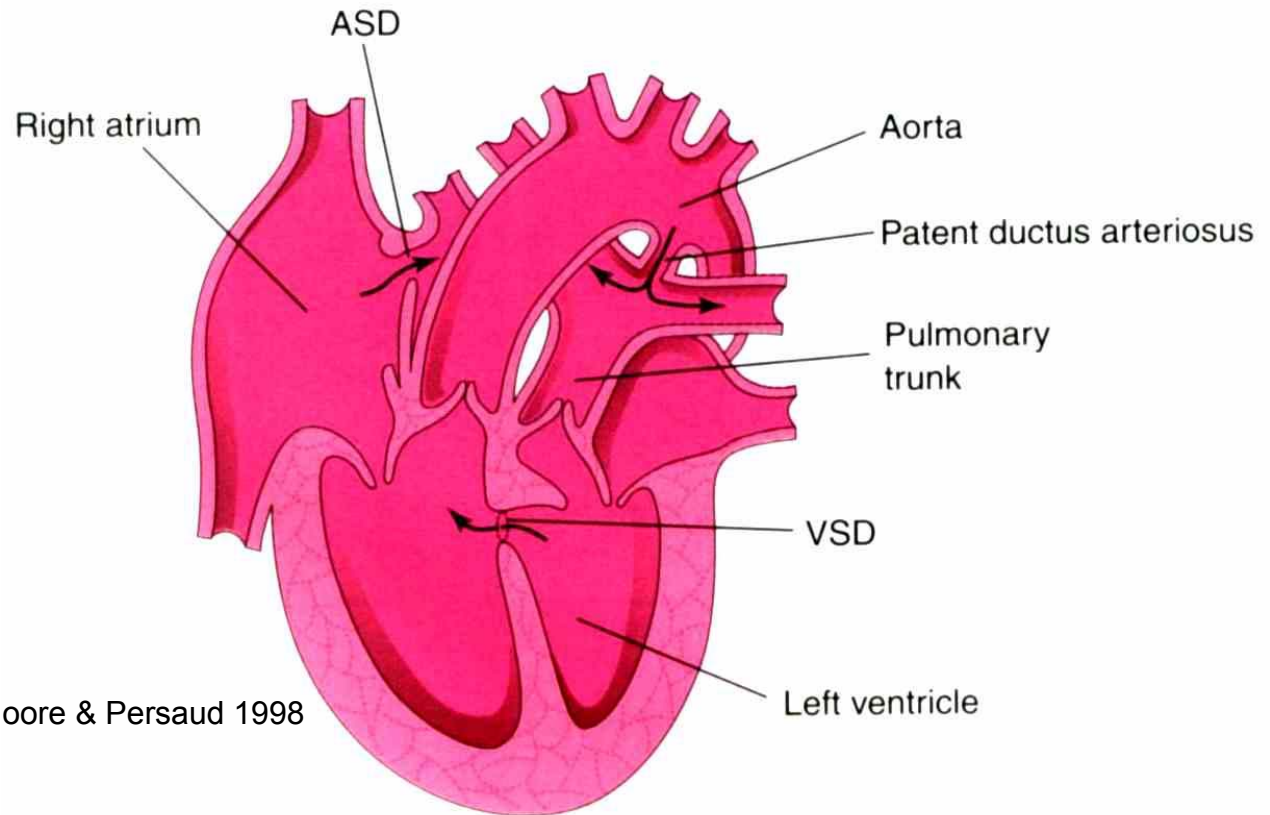
Increased pulmonary load defects: TGA

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

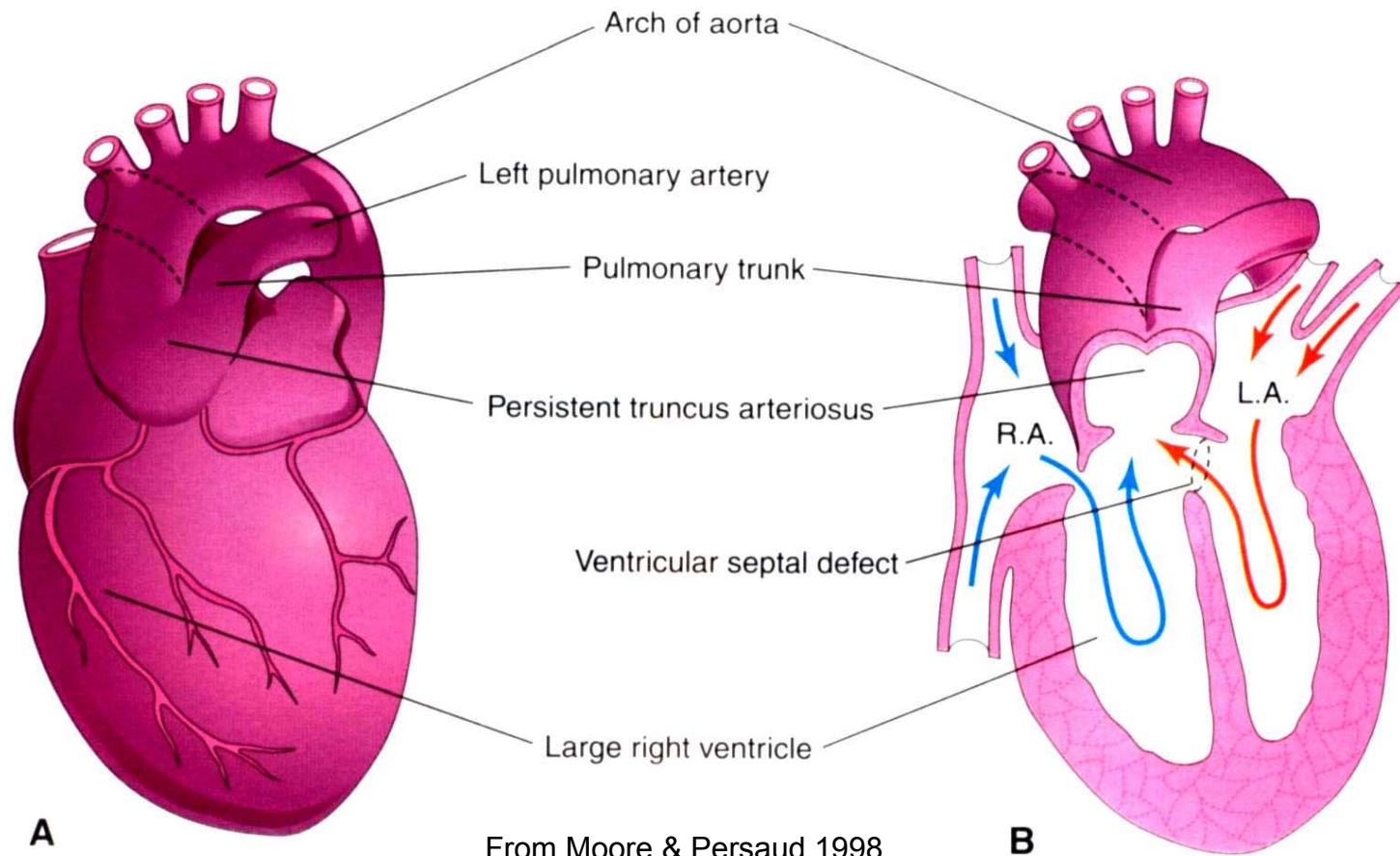


↑ pulmonary flow



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

- 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



From Moore & Persaud 1998

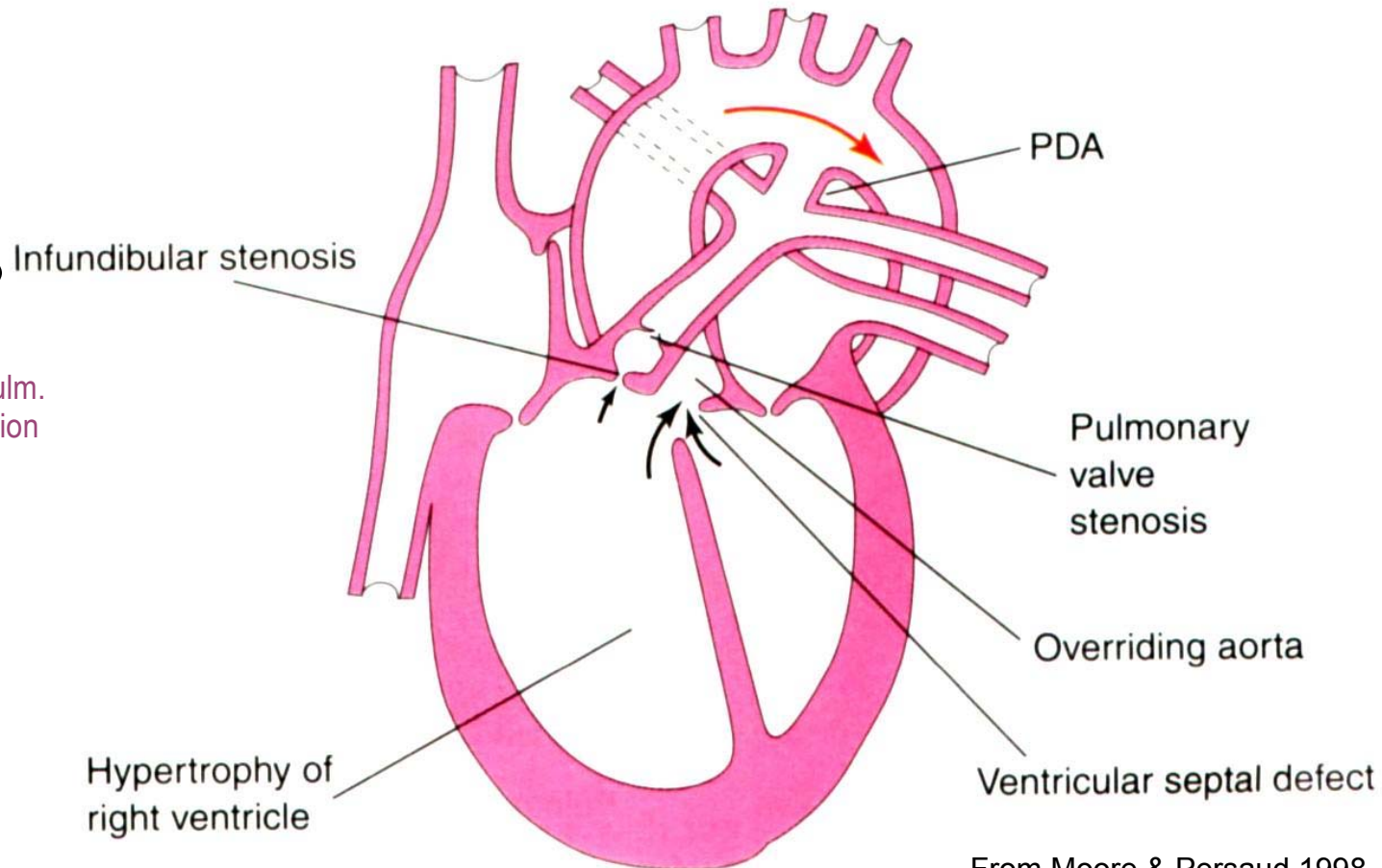
Decreased pulmonary load defects: Tetralogy of Fallot

- 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, 6th 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>