Pediatric Echocardiography

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What is your career?

A. Adult Echocardiographic Sonographer
B. Pediatric Echocardiography Sonographer
C. Adult and Pediatric
D. Radiology
E. Other

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Objectives

Overview of Embryology

Understand Pediatric Echocardiography

Congenital Heart Disease

- Common lesions
- Complex lesions

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Congenital Heart Defects
7-10/1,000 Live Births

DIAGNOSIS (Balt-Wash)	PERCENT
Ventricular septal defect	26%
Tetralogy of Fallot	9%
Atrioventricular septal defect	9%
Atrial septal defect	8%
Pulmonary valve stenosis	7%
Coarctation of the Aorta	7%
Hypoplastic left heart syndrome	6%
D-Transposition	5%

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CHD in Adults

30,000 babies born with CHD per year

20,000 surgeries for CHD per year

85% survive into adulthood

Over 1.2 million adults with CHD

Increasing at 5% per year

8,500 per year reach adulthood

Less than 10% disabled

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Diagnosis	1950's	1960's	1970's	1980's	1990's	2000's
ASD	Rare Repair	Repair older child	Repair age 4	Repair age 2	Repair age 2-3	Device closure
VSD	Rare Repair	Repair >10 kg or palliate	Repair < 1 year or palliate	Repair 6 months or prn	Repair premature infants	
PDA	Repair	Repair	Repair	Repair	Repair	
TOF	Palliate	Late Repair in adults	Repair after palliation		Repair 2-8 months or prn	
TGA	No survivors	Rare Survivors	Atrial Repair	Transitional Decade	Arterial Repair	
Single Ventricle	Comfort care	Palliate	Rare Fontan	Fenestrated Fontan	Lateral Tunnel	Extra-cardiac Fontan
HLHS	Comfort care	Comfort care	Surgery in Boston	Comfort vs. high risk surgery	Surgery & Fetal Diagnosis	

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Embryology 101

19 Days: Two endocardial tubes have formed – these tubes will fuse to form a common, single primiative heart tube

22 Days: Heart tube begins to beat

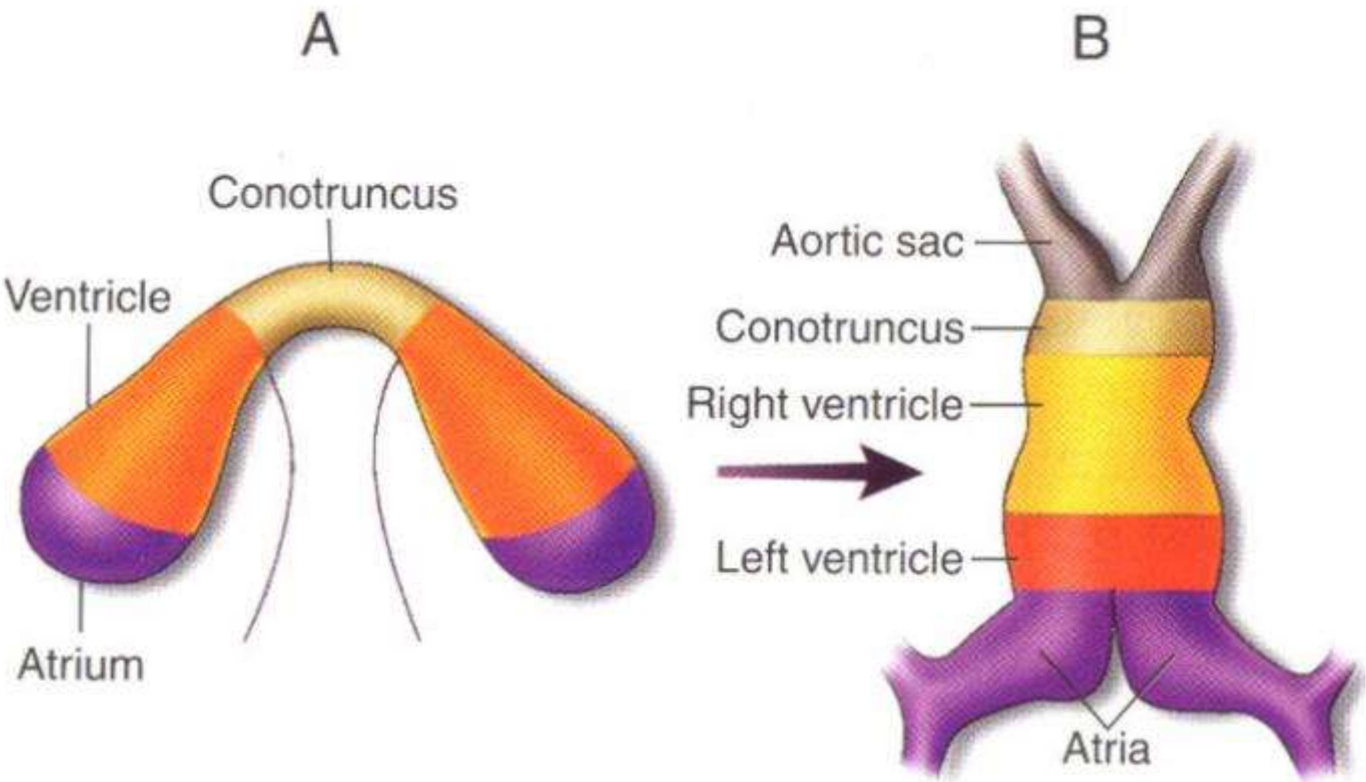
23 Days: Folding commences

30 Days: Primitive circulation

9 weeks (56 Days): All major structures identified

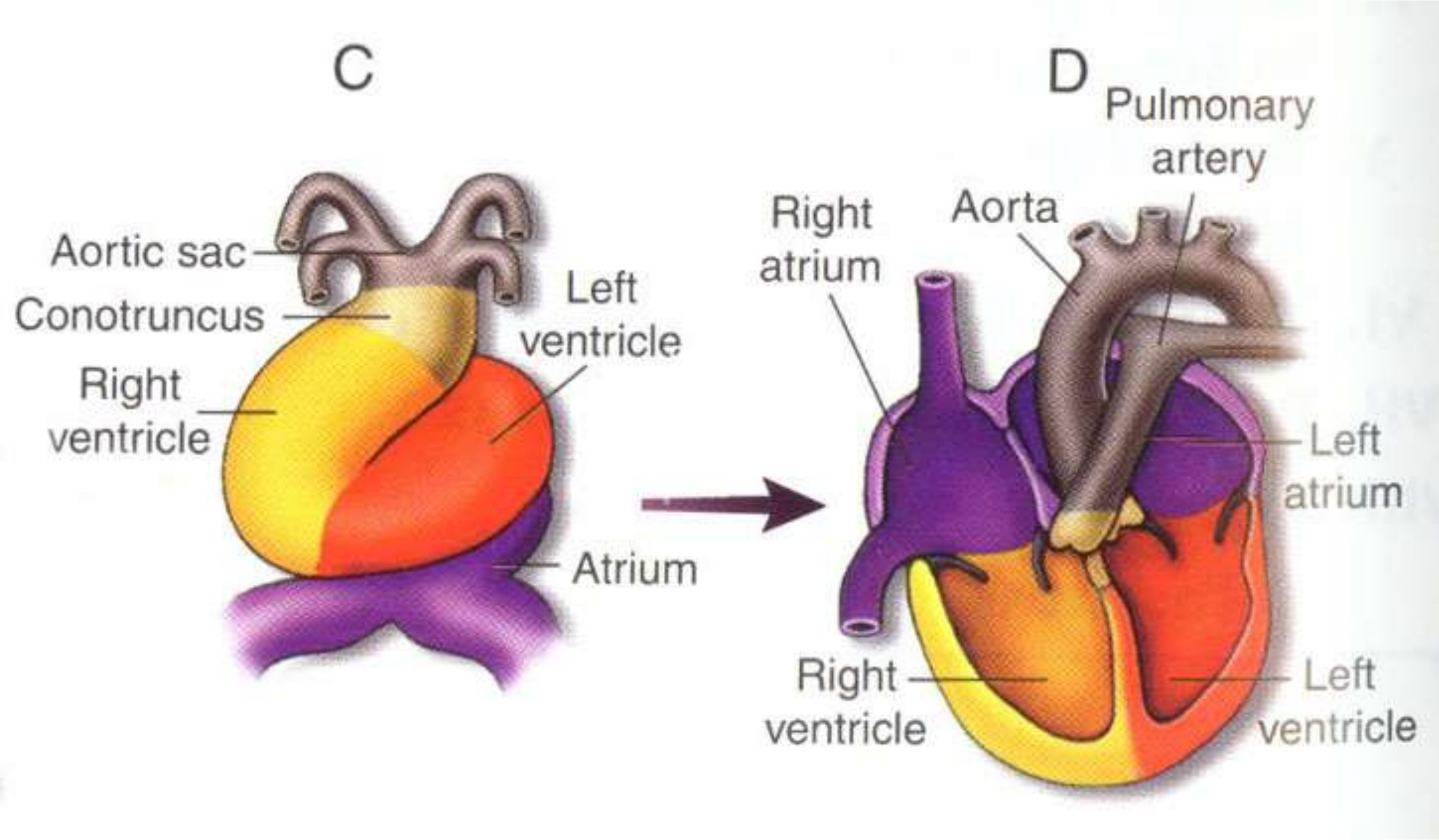
(In humans, several months of gestation remain for emergence of HLHS, PS, etc)

The Cardiac Crescent and the Tube Heart

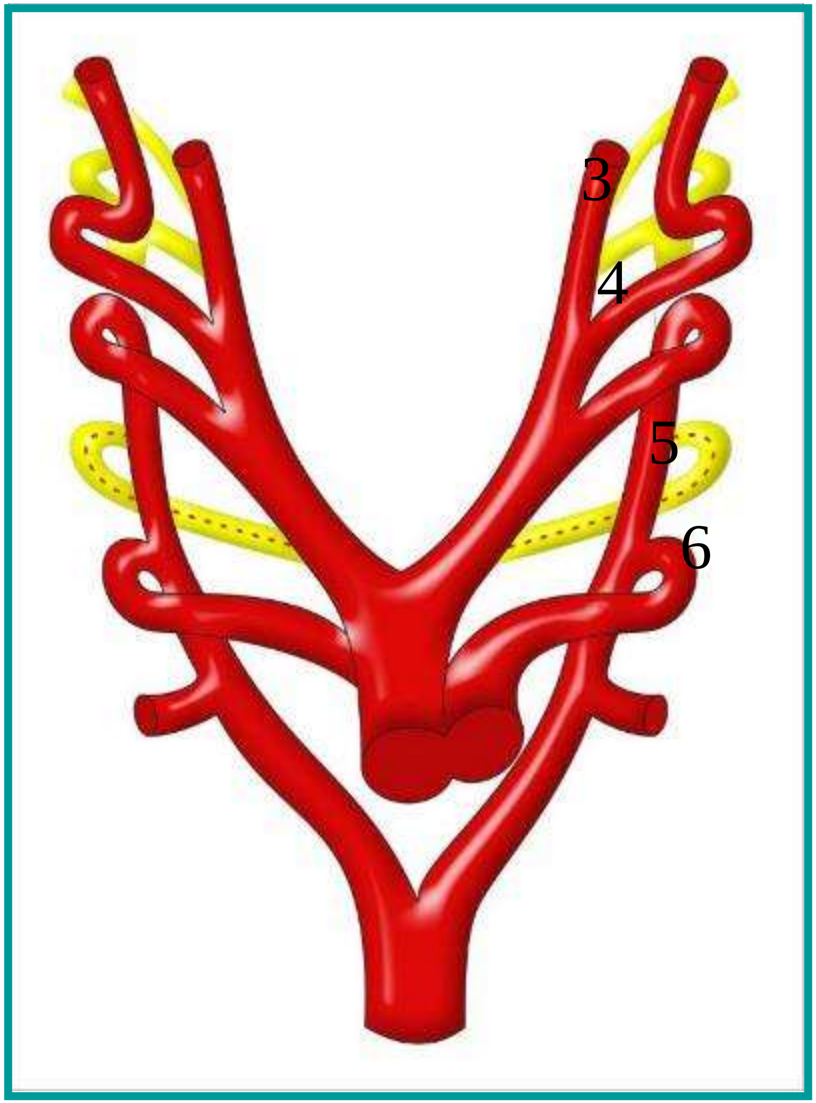


From *Heart Development*, 1999

Looping and Septation



From *Heart Development*, 1999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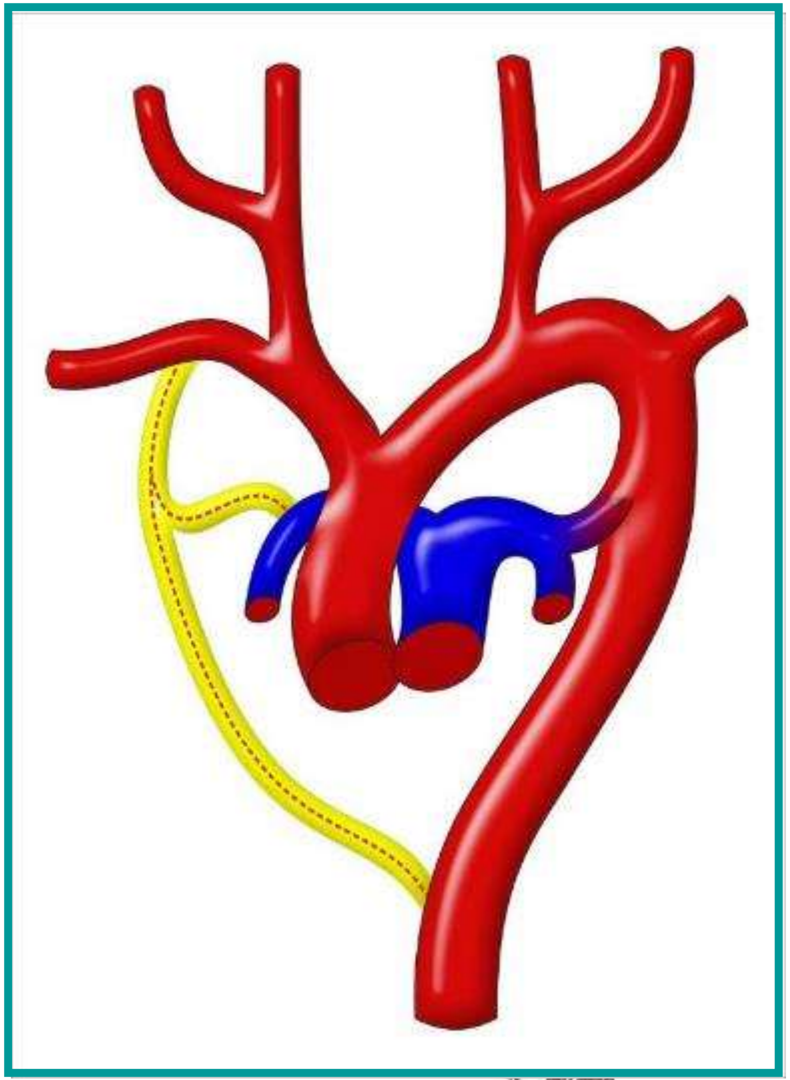

How do Congenital Heart Defects form?

Complex interaction between environmental and genetic etiology

- Multifactorial
- 5-8% chance of recurrence

Environmental exposures may influence micro-uterine environment and either turn on or off needed protein development

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Echo timeline

1793 Italian priest studied bats

1845 Austrian scientist Christian Doppler

WWII Sonar detected submarines

1954 Hertz & Edler

- (A&B mode echocardiogram)

M-mode ultrasound early 1970's

2D echo late 1970's

Doppler Echo 1980's

- Pulsed wave Doppler
- Continuous wave Doppler
- Color Doppler

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Pediatric Echo is Different

Anatomy and physiology over function

Segmental approach for complex patients

Improved resolution

- Heart is closer to chest wall
- Higher frequency transducers
- TEE rarely necessary for diagnosis

Inversion of apical and subcostal images

Echo in CHD

Doppler echo

- Pulsed wave Doppler
  - Quantitation of intracardiac hemodynamics (**Modified Bernoulli Equation $\Delta P = 4 \times v_2^2$**)
    - Valvar regurgitation
    - Intracardiac shunts
    - LVOT/RVOT obstruction
  - Ventricular function
    - Systolic
    - Diastolic (mitral inflow, pulmonary venous inflow)

Echo in CHD

Continuous wave Doppler

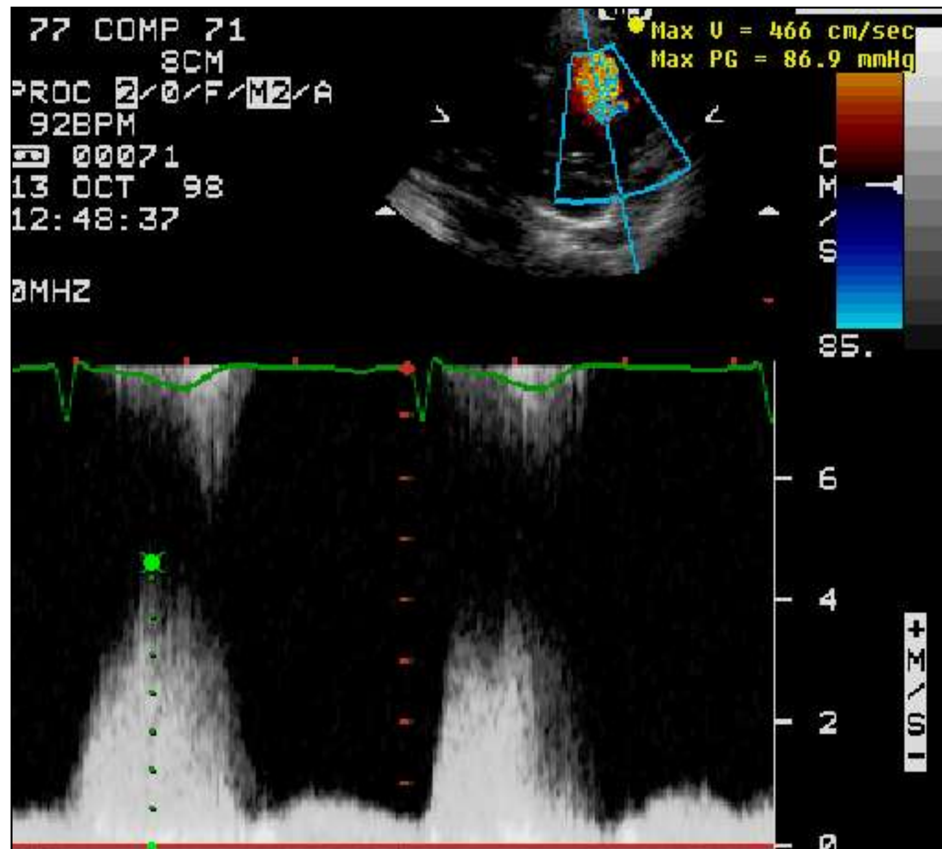
- Non-invasive measurements of mean and peak transvalvar gradients
  - Valvar stenosis
- Prediction of Ventricular Pressure (modified Bernoulli equation)
  - VSD-→ LV: RV pressure gradient
  - TR/PR→ RV, PA pressure

Doppler
Spectral Display

Pulsed Wave (PW)

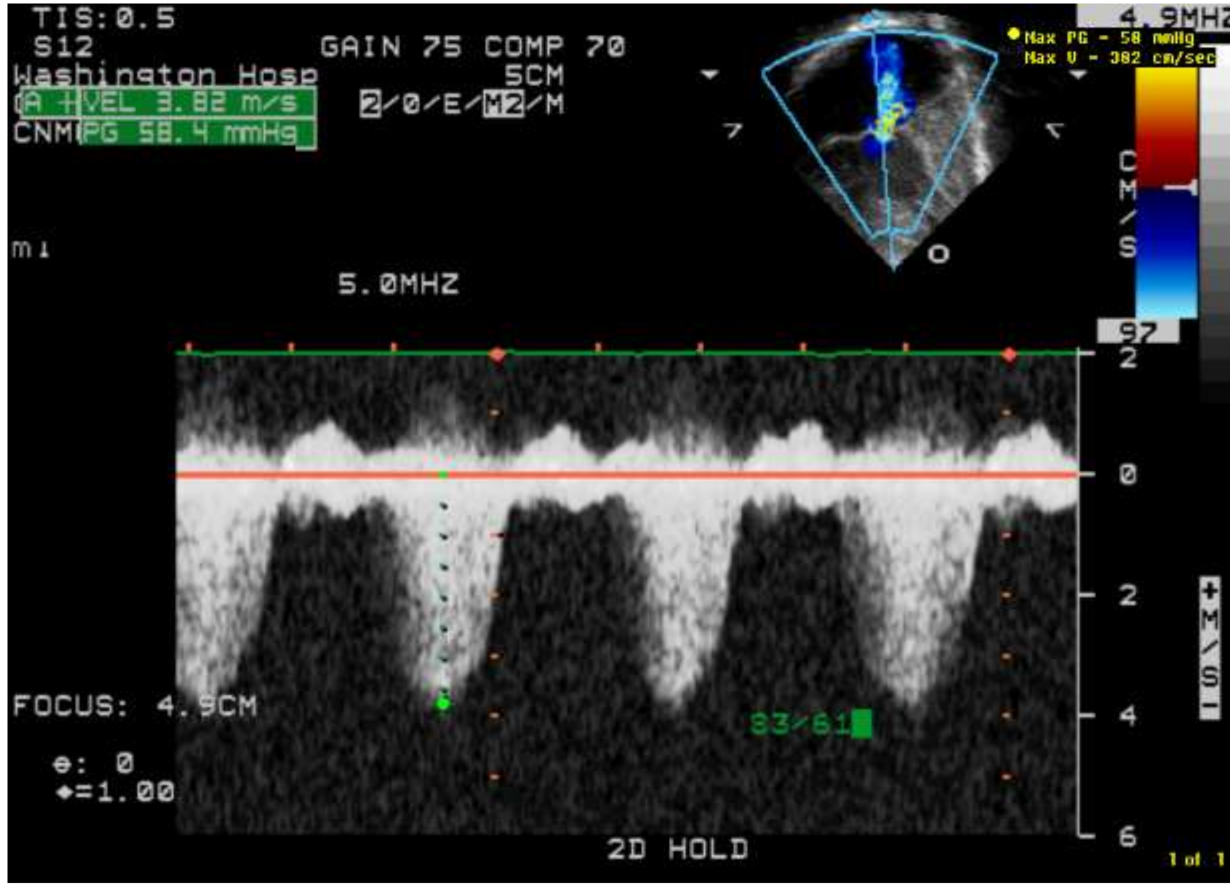
Continuous Wave (CW)

Aortic Valve Velocity



RV pressure by TR estimation

The pressure in the right ventricle is 55+ 10= 65mmHg. The pressure in the LV is 83 mmHg; we know it from the blood pressure which was 83/61. So, it the RV systemic pressure is ¾ of the systemic LV pressure. It means RV systemic pressure is elevated. It should normally be 1/3 systemic.



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Echo in CHD

Color Doppler

- Direction of cardiac flow
  - TAPVR vs. LSVC
- Velocity and Turbulence of cardiac flow
  - Conduit obstruction
  - Identification of intracardiac shunts
    - VSD, PDA, ASD
  - Assessment of Post-op CHD
    - Shunt patency, residual intracardiac shunt

Questions….

1. How much time should you spend trying to obtain Doppler of TR when there is a HUGE ventricular septal defect?
2. What if your patient has a single ventricle, if you measure the TR what does that estimate?
3. Why is it important to Doppler a VSD?
4. If you see funny blood flow, should you invert your color scale?
5. The doctor wants to know if there is pulmonary hypertension in a NICU baby, but there is no TR, is there another way to answer that question?

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Guidelines and Standards for Performance of a Pediatric Echocardiogram: A Report from the Task Force of the Pediatric Council of the American Society of Echocardiography

Wyman W. Lai, MD, MPH, FASE, Tal Geva, MD, FASE, Girish S. Shirali, MD,
Peter C. Frommelt, MD, Richard A. Humes, MD, FASE, Michael M. Brook, MD,
Ricardo H. Pignatelli, MD, and Jack Rychik, MD, Writing Committee, *New York, New York; Boston, Massachusetts; Charleston, South Carolina; Milwaukee, Wisconsin; Detroit, Michigan; San Francisco, California; Houston, Texas; and Philadelphia, Pennsylvania*

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Table 4 Structures viewed from standard examination views

Subxiphoid (subcostal) views

- Inferior vena cava
- Hepatic veins
- Abdominal aorta
- Diaphragm
- Superior vena cava
- Left atrium
- Right atrium
- Atrial septum
- Coronary sinus
- Pulmonary veins
- Mitral valve
- Tricuspid valve
- Left ventricle
- Right ventricle
- Ventricular septum
- Left ventricular papillary muscles
- Aortic valve
- Pulmonary valve
- Ascending aorta
- Coronary arteries
- Main and branch pulmonary arteries
- Pericardium

Apical views

- Inferior vena cava
- Left atrium
- Right atrium
- Atrial septum
- Coronary sinus
- Selected pulmonary veins
- Mitral valve
- Tricuspid valve
- Left ventricle
- Right ventricle
- Ventricular septum
- Left ventricular papillary muscles
- Aortic valve
- Pulmonary valve
- Ascending aorta
- Main and branch pulmonary arteries

Left parasternal views

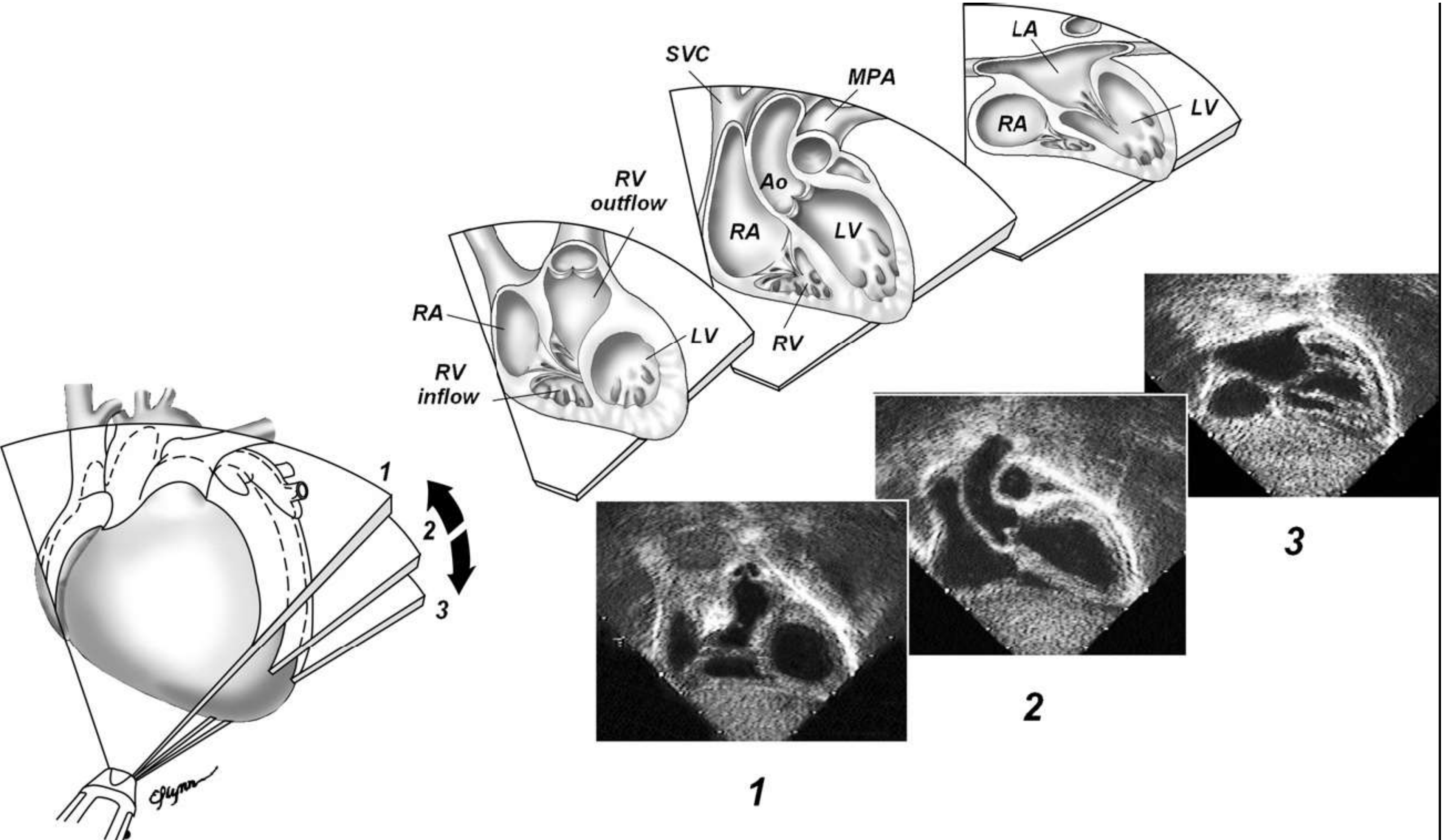
- Inferior vena cava
- Superior vena cava
- Left atrium
- Right atrium
- Atrial septum
- Coronary sinus
- Pulmonary veins
- Mitral valve
- Tricuspid valve
- Left ventricle
- Right ventricle
- Ventricular septum
- Left ventricular papillary muscles
- Aortic valve
- Pulmonary valve
- Ascending aorta
- Coronary arteries
- Main and branch pulmonary arteries
- Pericardium

Suprasternal notch views

- Superior vena cava
- Left atrium
- Pulmonary veins
- Ascending aorta
- Superior thoracic aorta
- Main and branch pulmonary arteries
- Aortic arch
- Proximal brachiocephalic arteries
- Left innominate vein

Right parasternal views

- Inferior vena cava
- Superior vena cava
- Right atrium
- Atrial septum
- Right pulmonary veins
- Ascending aorta
- Right pulmonary artery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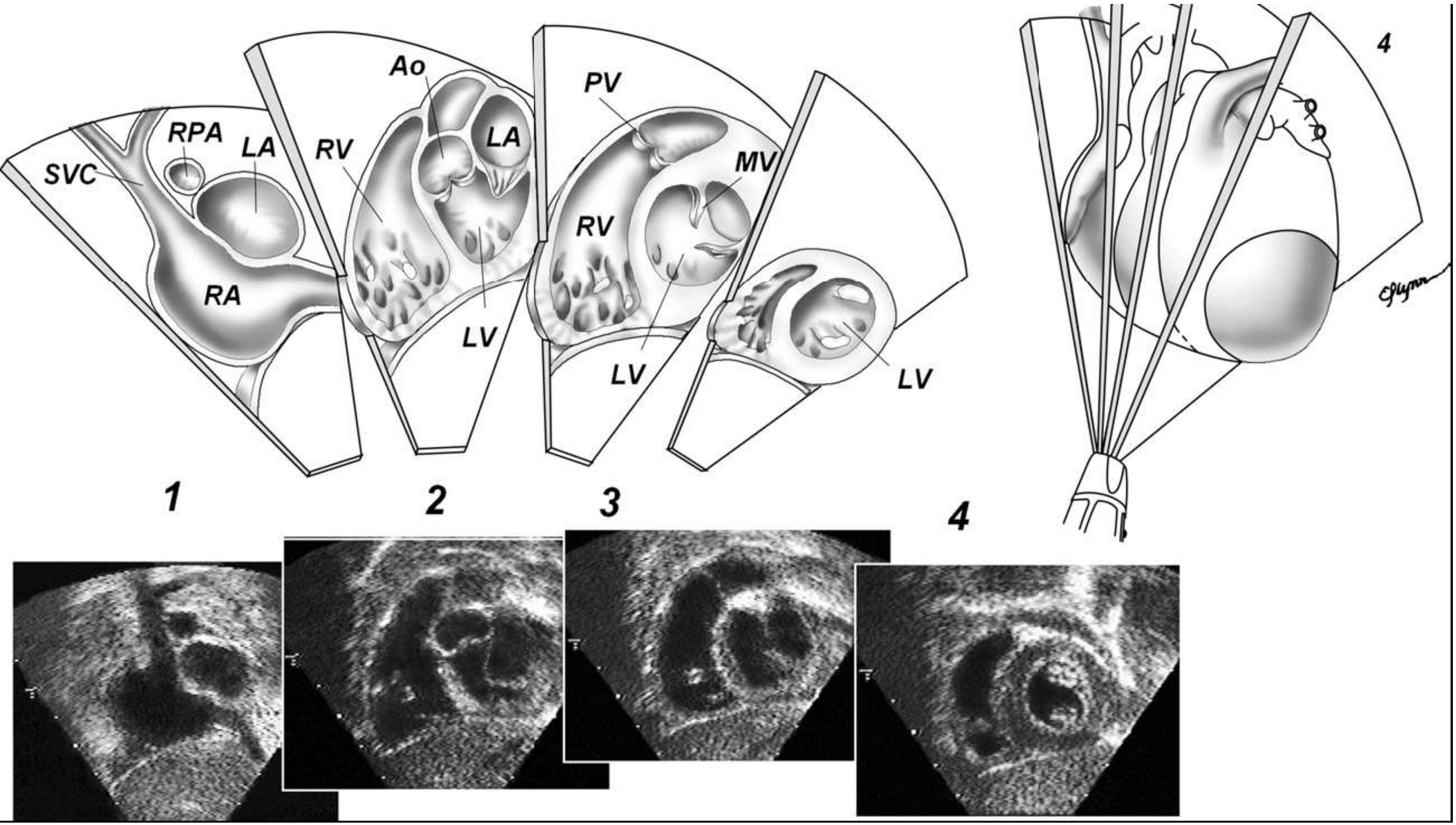
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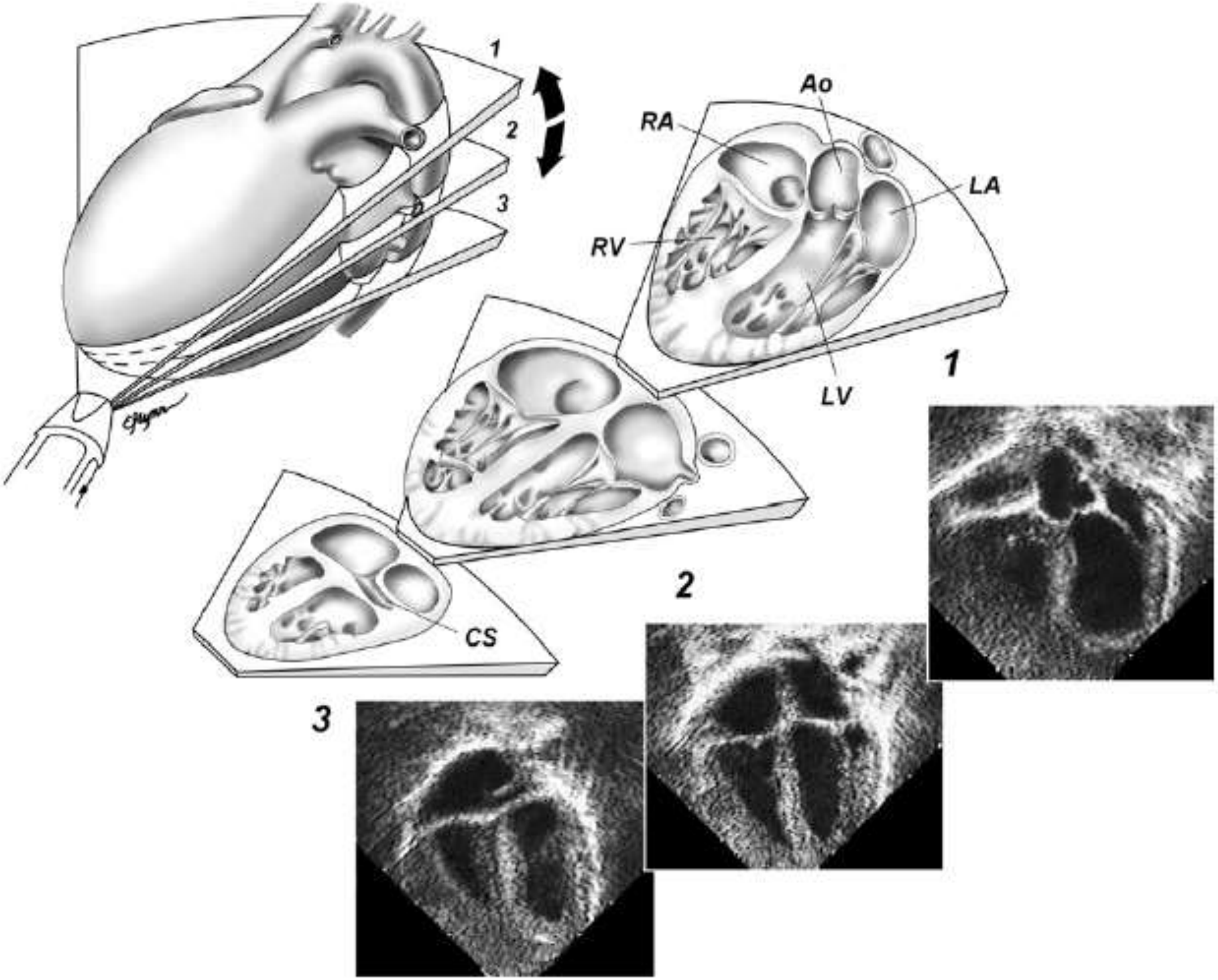
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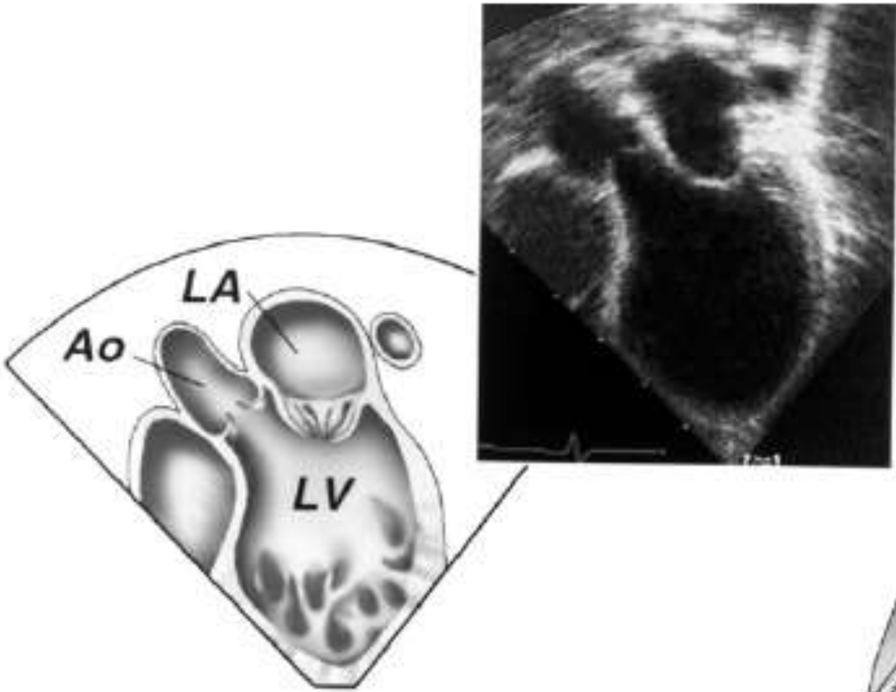
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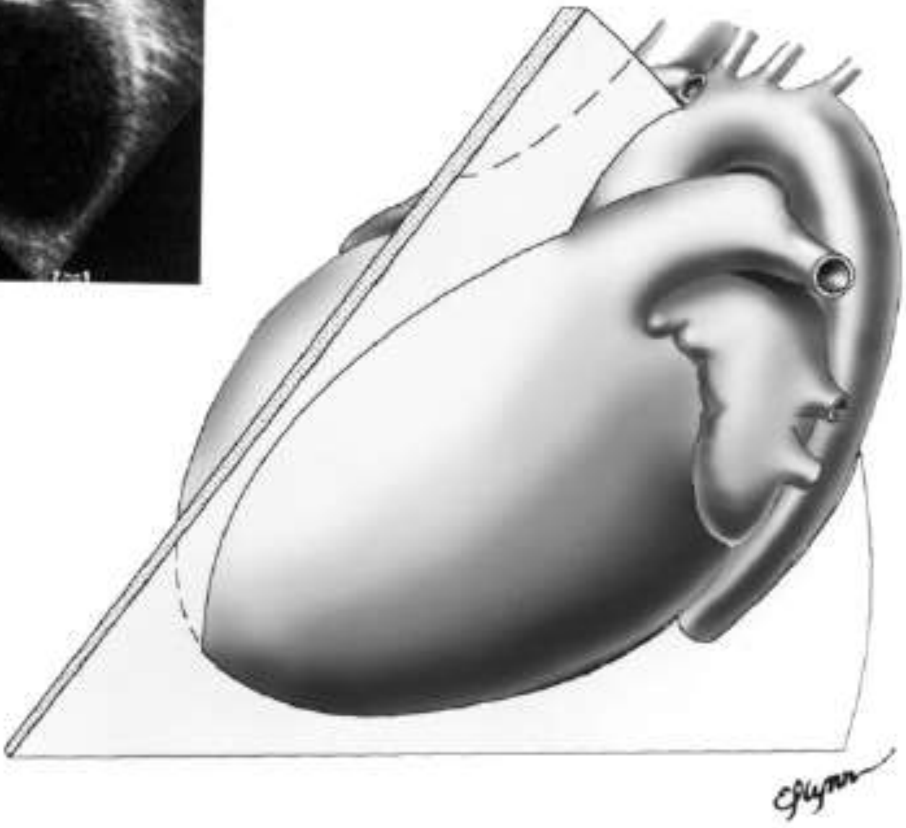

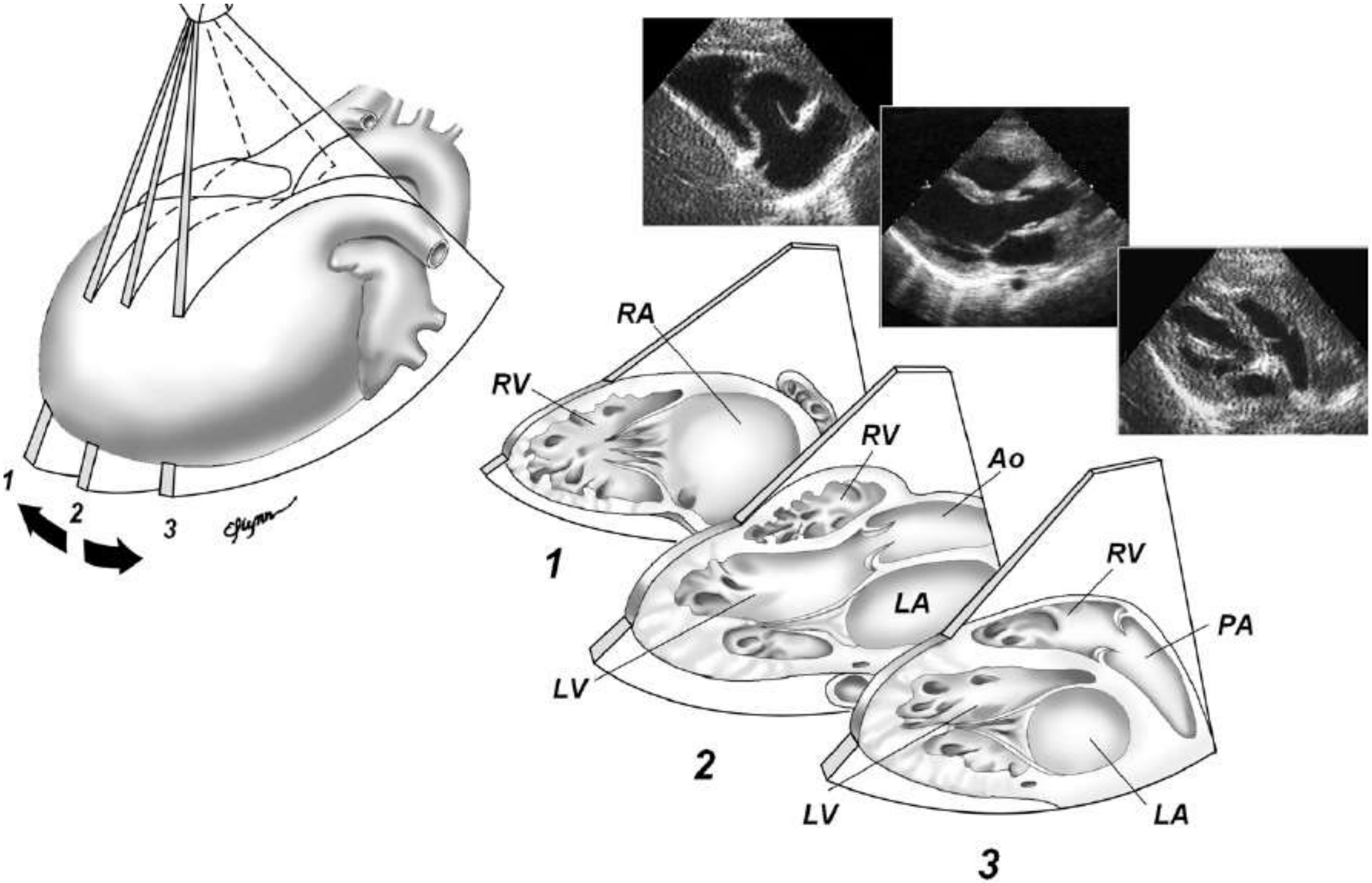
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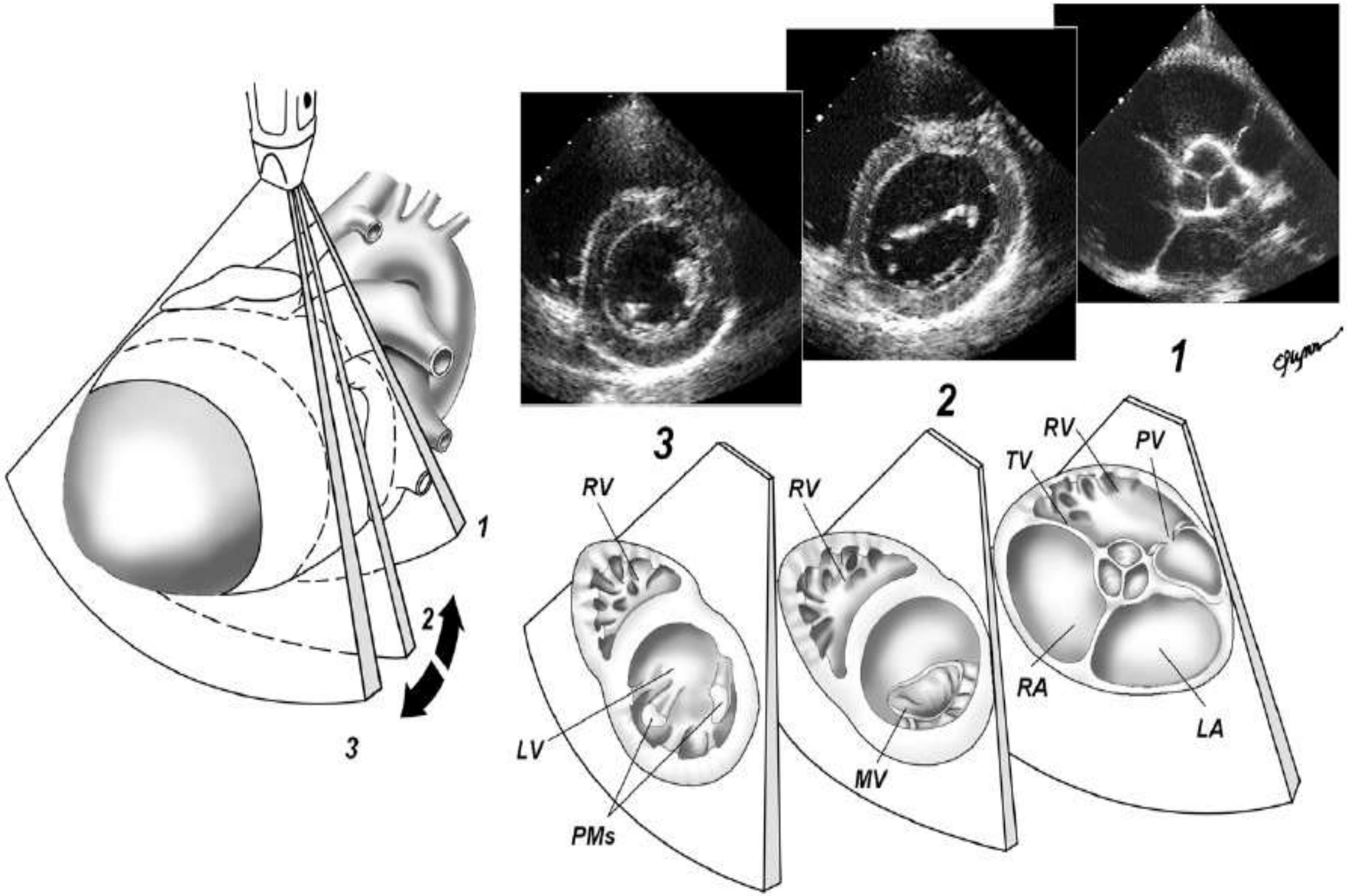
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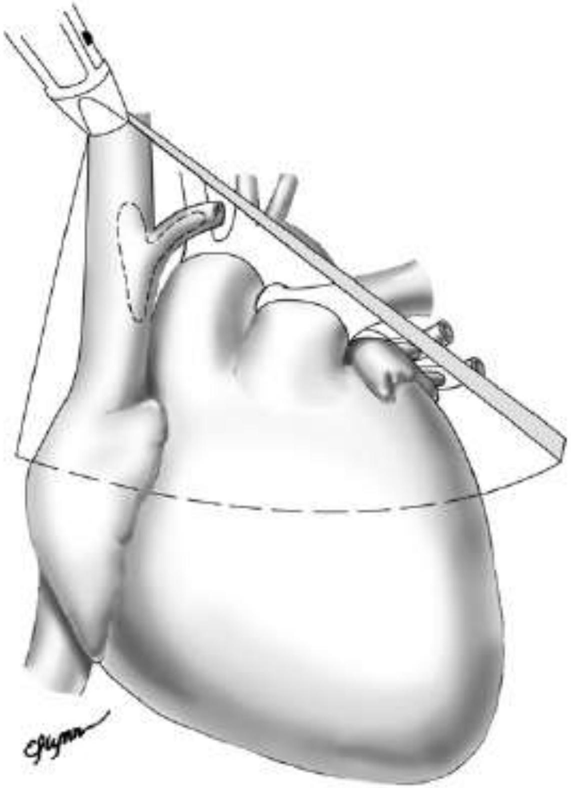

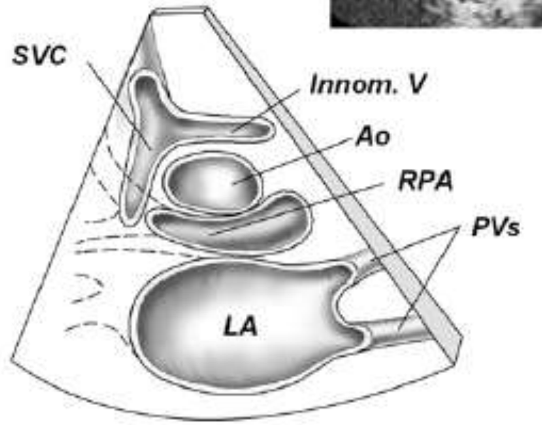
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RPA
PVs
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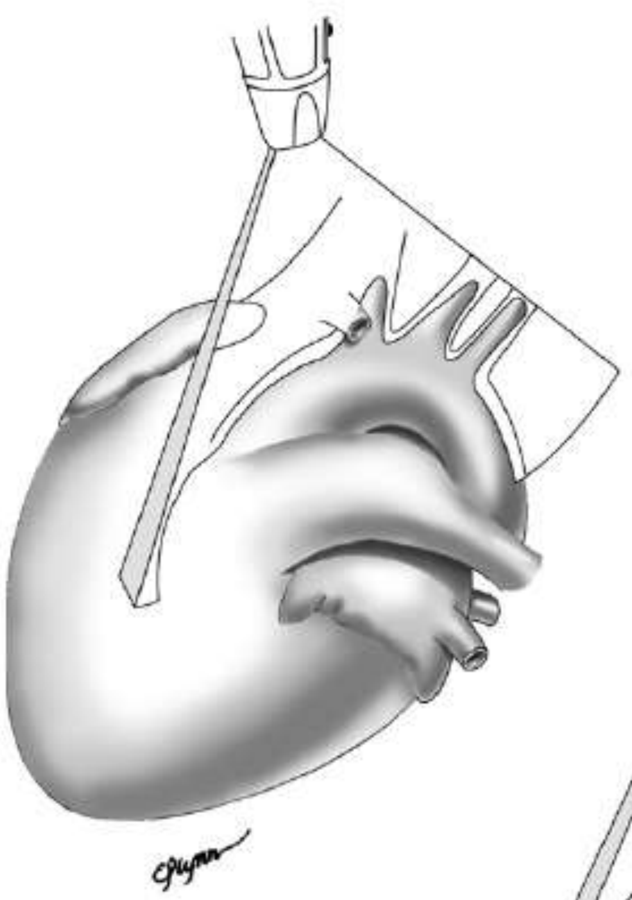

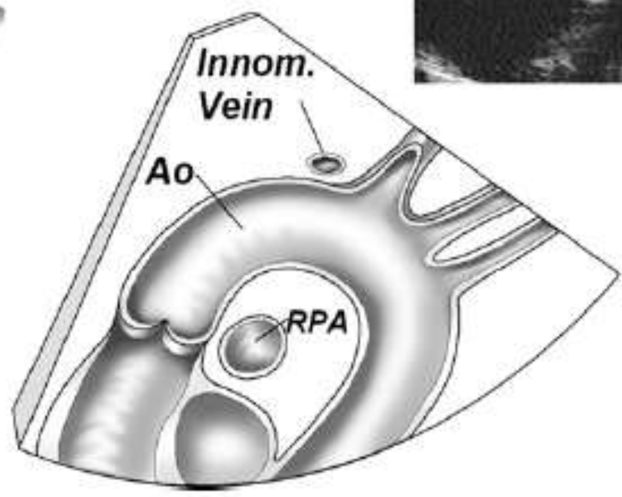
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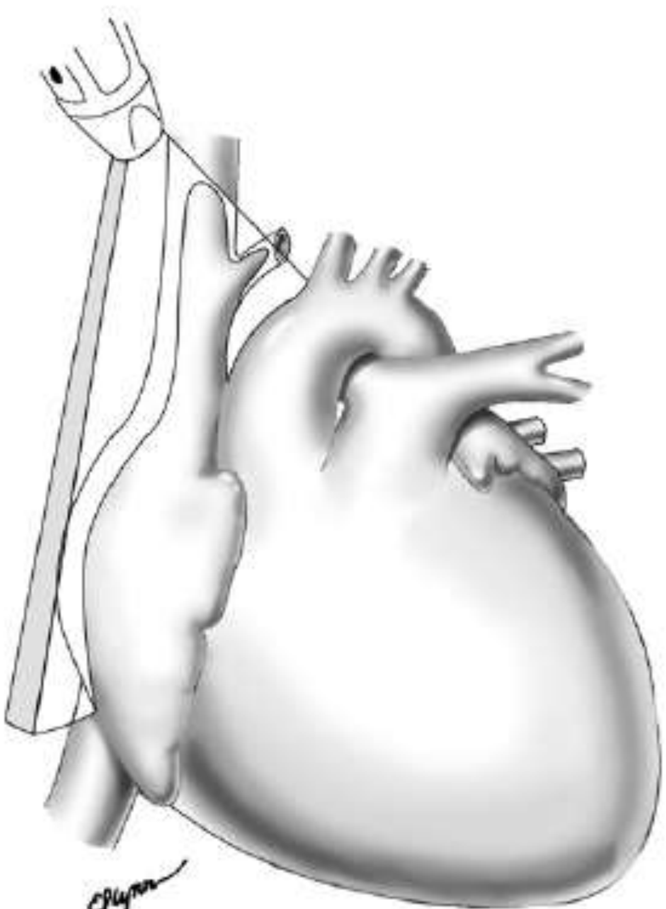

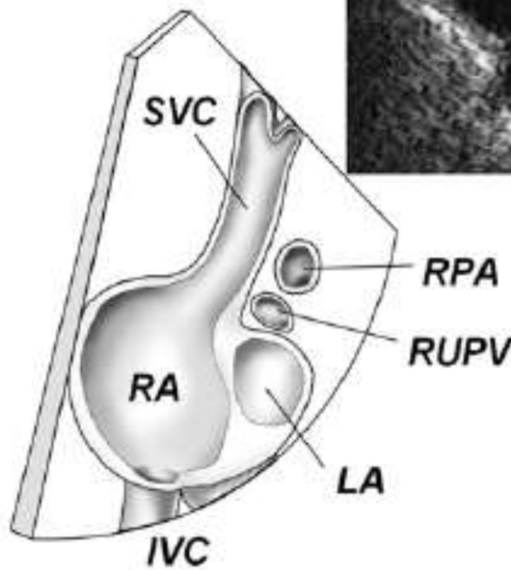
SVC
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RA
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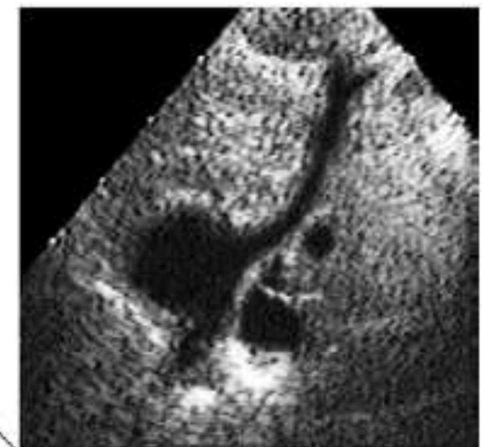
IVC



Classification and Terminology of Cardiovascular Anomalies

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Morphologic/Segmental approach

Define morphologic—not spatial—anatomy

- Which atrium is the Right? Left?
- Which ventricle is the Right? Left?
- Which great artery is which?

Define segmental anatomy

- Segments: Atrium, Ventricles, Great Arteries
- What is the position of each segment relative to each other?
  - Is the RA on the right? Is it connected to the RV? Is it connected to the PA?
  - Is the LA on the left? Is it connected to the LV? Is it connected to the Aorta?

Predict the physiology

- What is the physiology predicted by the segmental connections?
  - Normal? Transposition? Obstructed flow?
- What is the physiology predicted by flow in the ductus? Across the foramen?

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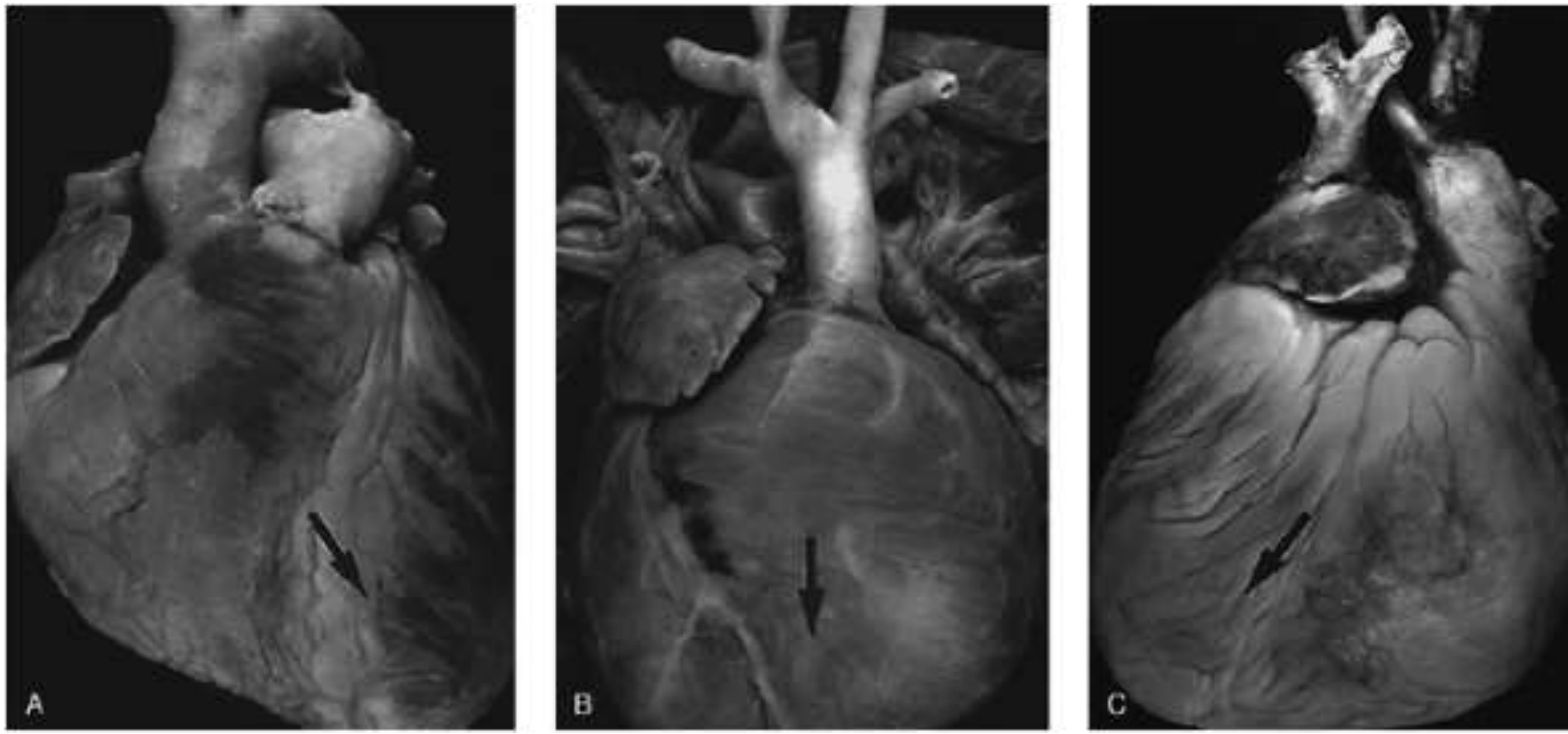
Cardiac base-apex axis and orientation in the chest



Levocardia   Mesocardia   Dextrocardia

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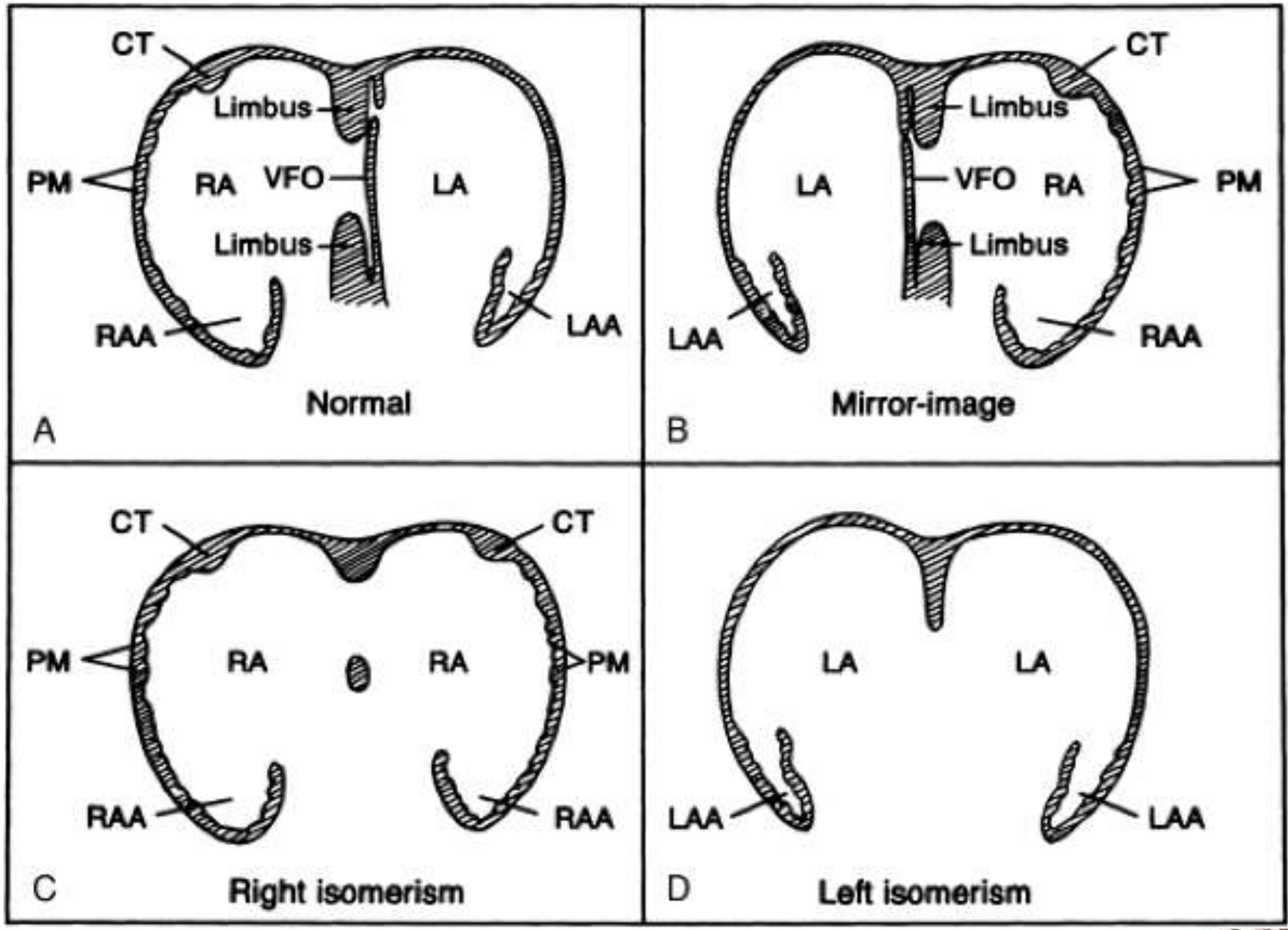
Cardiac situs (sidedness)



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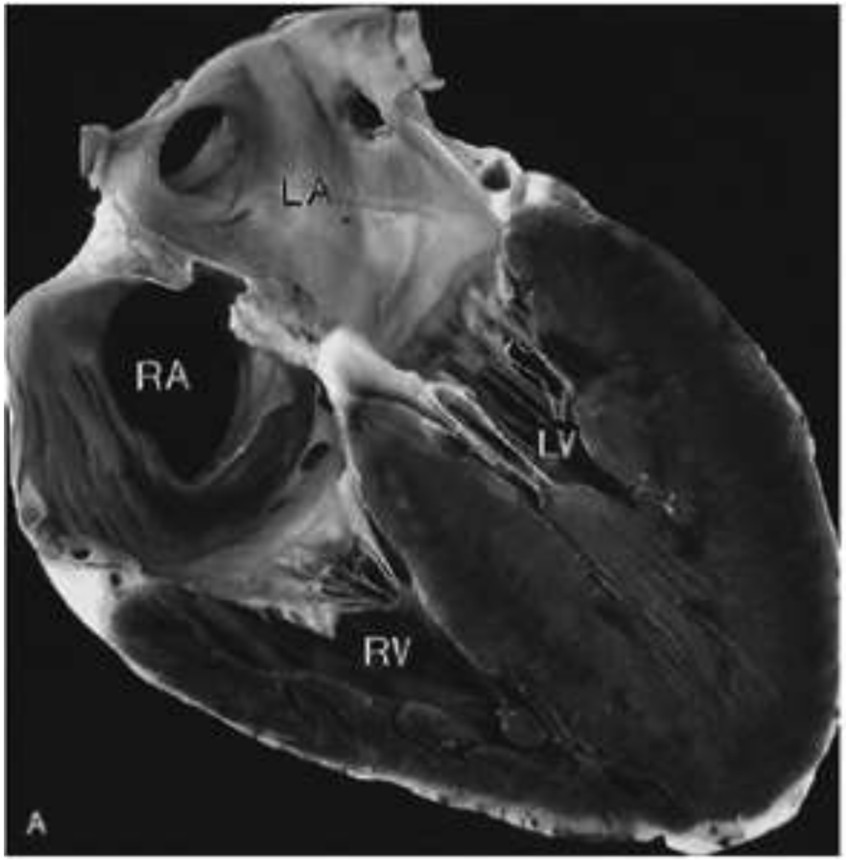
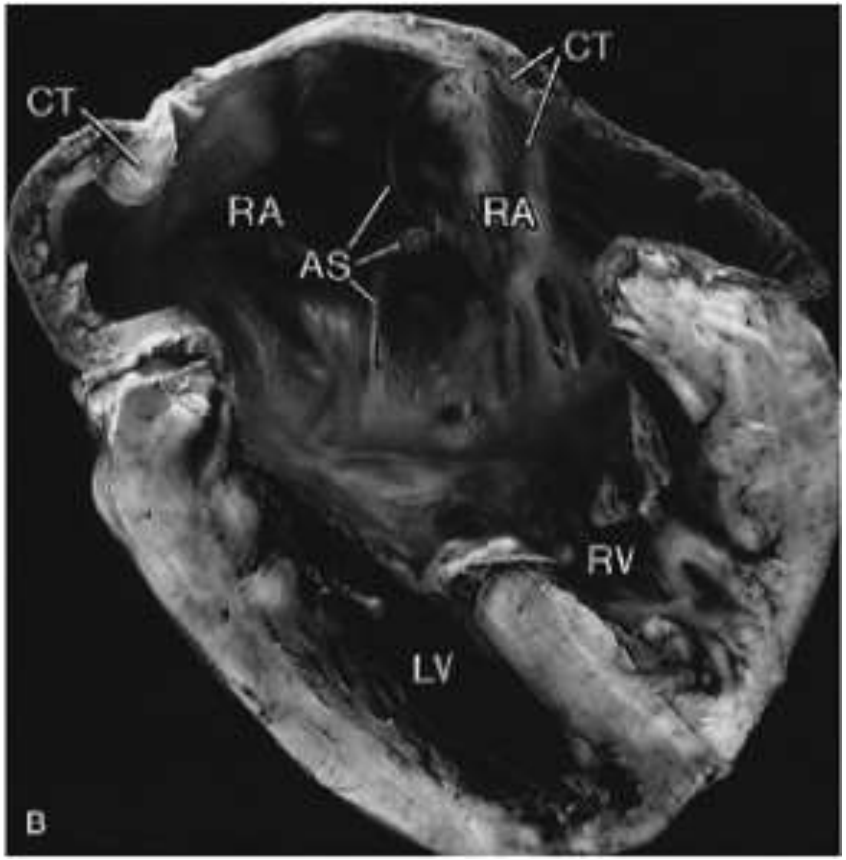
Example: Cardiac sidedness





Situs solitus normal cardiac sidedenss

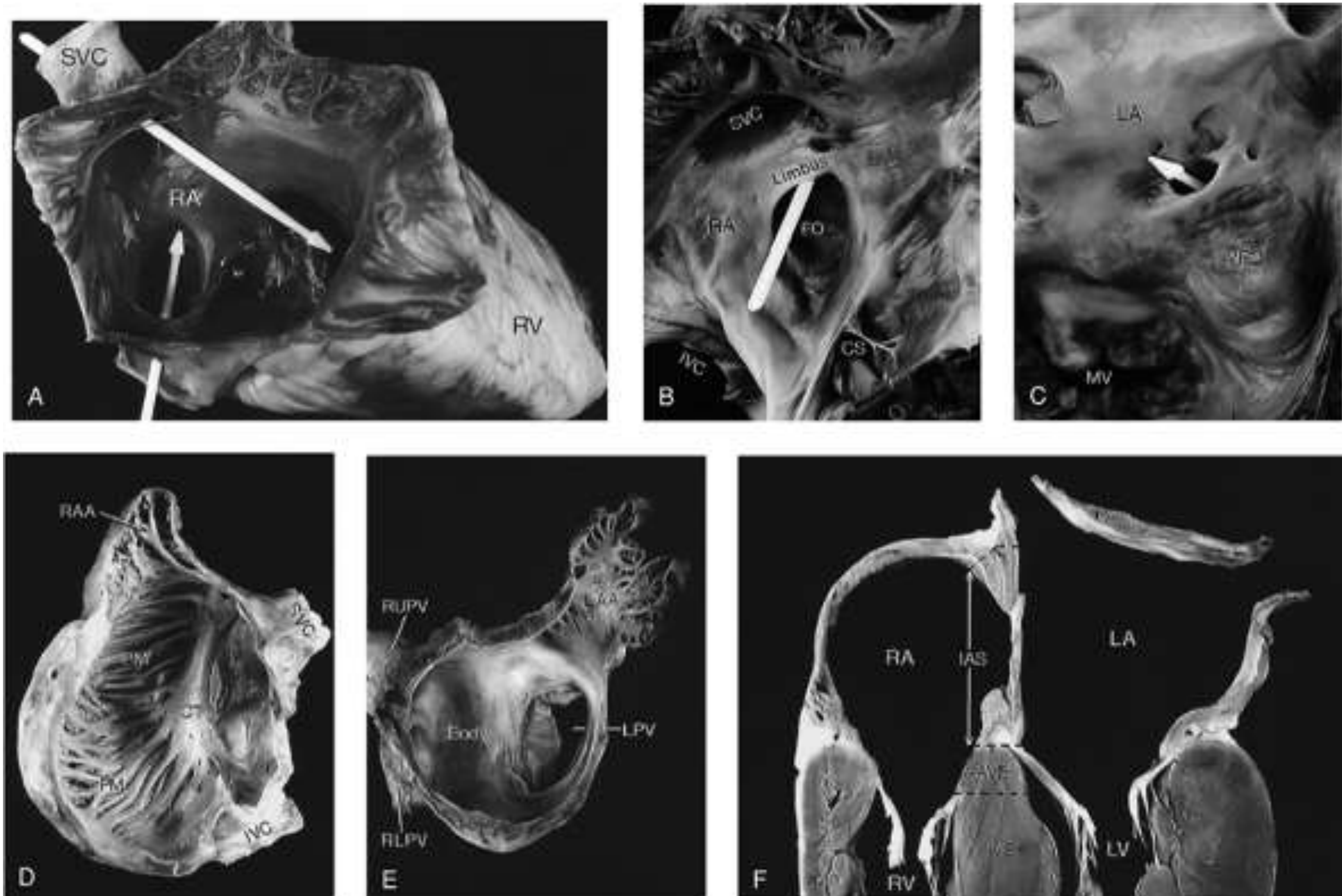
Situs ambiguus, right isomerism

Differentiation between the atria

The morphologic RA has a smooth or sinusal portion, which is found between the interatrial septum and the crista terminalis. It receives the drainage of the superior and inferior venae cavae and the coronary sinus. The trabecular portion is characterized by the presence of pectinate muscles, which are directed from crista terminalis to the base of the right atrial appendage. The RA appendage is wide and its edge is blunt.

RA appendage is broad based and triangularly shaped (like Snoopy's nose), with pectinate muscles that extend into the body of the right atrium.

The anatomic LA is totally smooth and lacks pectinate muscles. It receives the drainage of the pulmonary veins, and LA appendage has a narrow base and fingerlike appearance (like Snoopy's ears) with pectinate muscles confined within the appendage.



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RA and TV valve characteristics

Right atrium:
- Limbus of fossa ovalis (limb of oval fossa)
- Large pyramidal appendage (Snoopy’s nose)
- Crista terminalis (terminal crest)
- Pectinate muscles
- Receives venae cavae and coronary sinus*

Tricuspid valve:
- Low septal annular attachment
- Septal cordal attachments
- Triangular orifice (midleaflet level)
- Three leaflets and commissures
- Three papillary muscles
- Empties into right ventricle

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LA and MV valve characteristics

Left atrium:
- Ostium secundum
- Small fingerlike appendage (Snoopy's ear)
- No crista terminalis
- No pectinate muscles
- Receives pulmonary veins*

Mitral valve:
- High septal annular attachment
- No septal cordal attachments
- Elliptical orifice (midleaflet level)
- Two leaflets and commissures
- Two large papillary muscles
- Empties into left ventricle

Differentiation between the atria

The only structures that are constant and allow differentiation between the right and left atria are the appendages!

The drainage of the systemic and pulmonary veins does not permit the conclusive identification of the atria, as drainage sites are sometimes anomalous. The atrial septum cannot always be used either, because it can have defects or be absent.

Ventricles-characteristics

Right ventricle:

Tricuspid-pulmonary discontinuity
Muscular outflow tract
Septal and parietal bands
Large apical trabeculations
Coarse septal surface
Crescentic in cross sections* (* variable)
Thin free wall (3–5 mm)*
Receives tricuspid valve
Pulmonary valve empties into main pulmonary artery

Ventricles-characteristics

Left ventricle:

- Mitral-aortic continuity
- Muscular-valvular outflow tract
- No septal or parietal band
- Small apical trabeculations
- Smooth upper septal surface
- Circular in cross section* (* variable)
- Thick free wall (12–15 mm)*
- Receives mitral valve
- Aortic valve
- Empties into ascending aorta

Ventricular features (summary)

- Features of the morphologic RV:
- Coarse trabeculae with prominent septal band, parietal band, and moderator band.
- Septophillic attachments of the tricuspid valve (attachments to septum and free wall)
- Well-developed infundibulum (= conus= cone of muscle beneath the semilunar valve) which results in fibrous dyscontinuity between the tricuspid and semilunar valves
- Features of the morphologic LV:
- Smooth septal surface, fine trabeculae
- Septophobic attachments of the mitral valve (attachments only to free wall)
- No infundibulum which results in fibrous continuity of the mitral and semilunar valves

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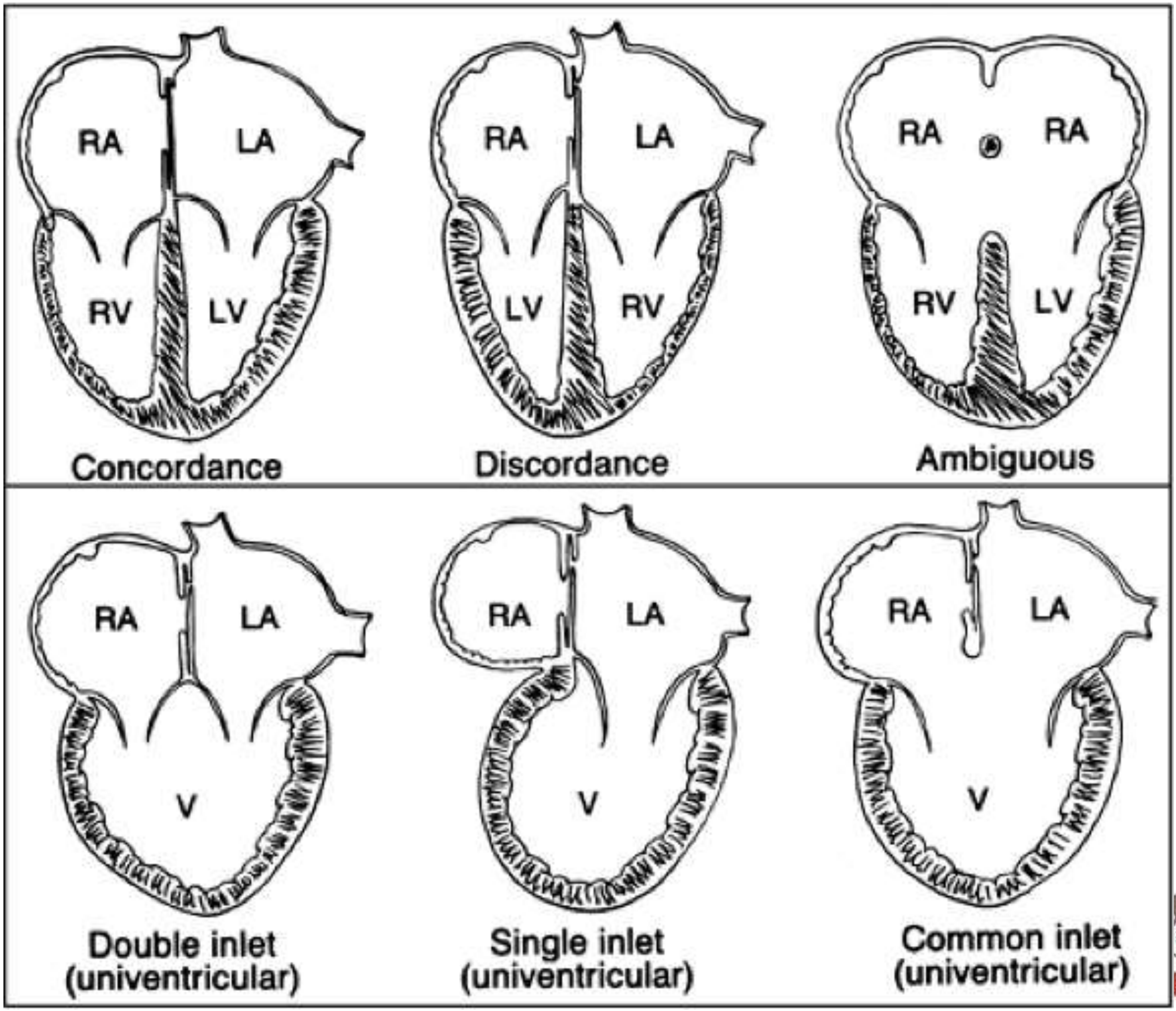
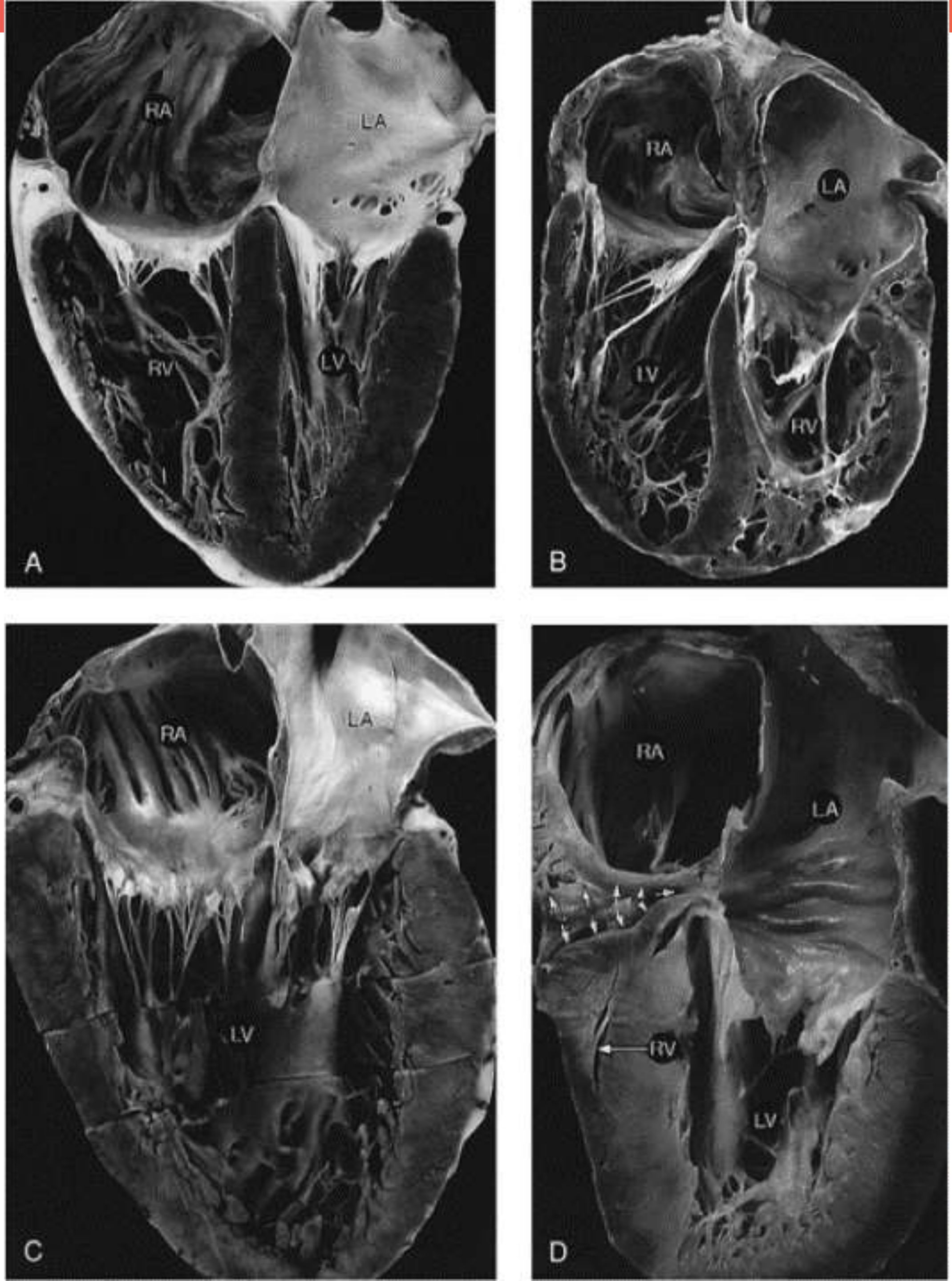
Atrioventricular connections





Examples of atrioventricular connections:

A. Concordance
B. Discordance
C. Double-inlet LV
D. Ticuspid atresia: absent right A-V connection

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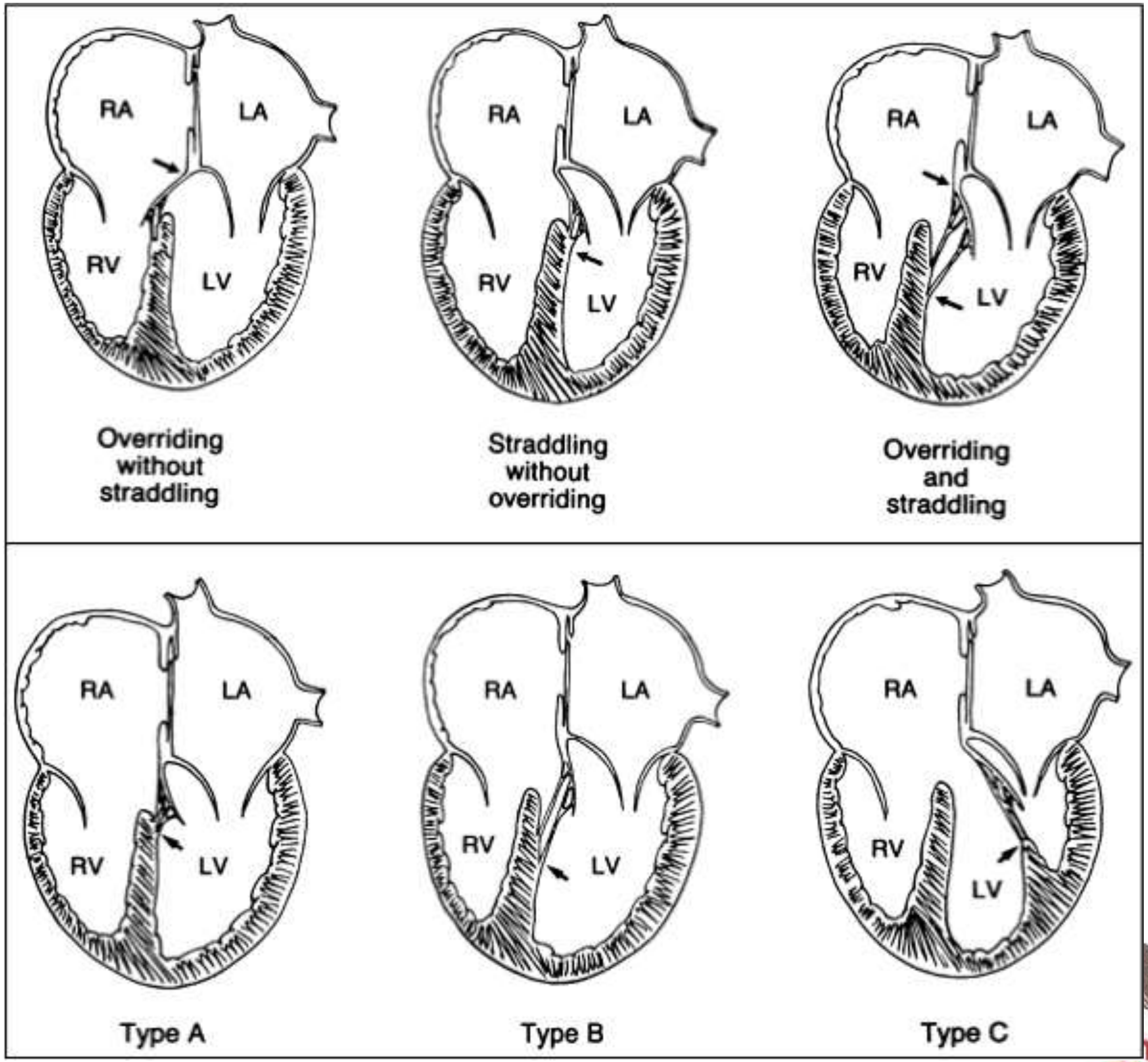
Overriding and straddling



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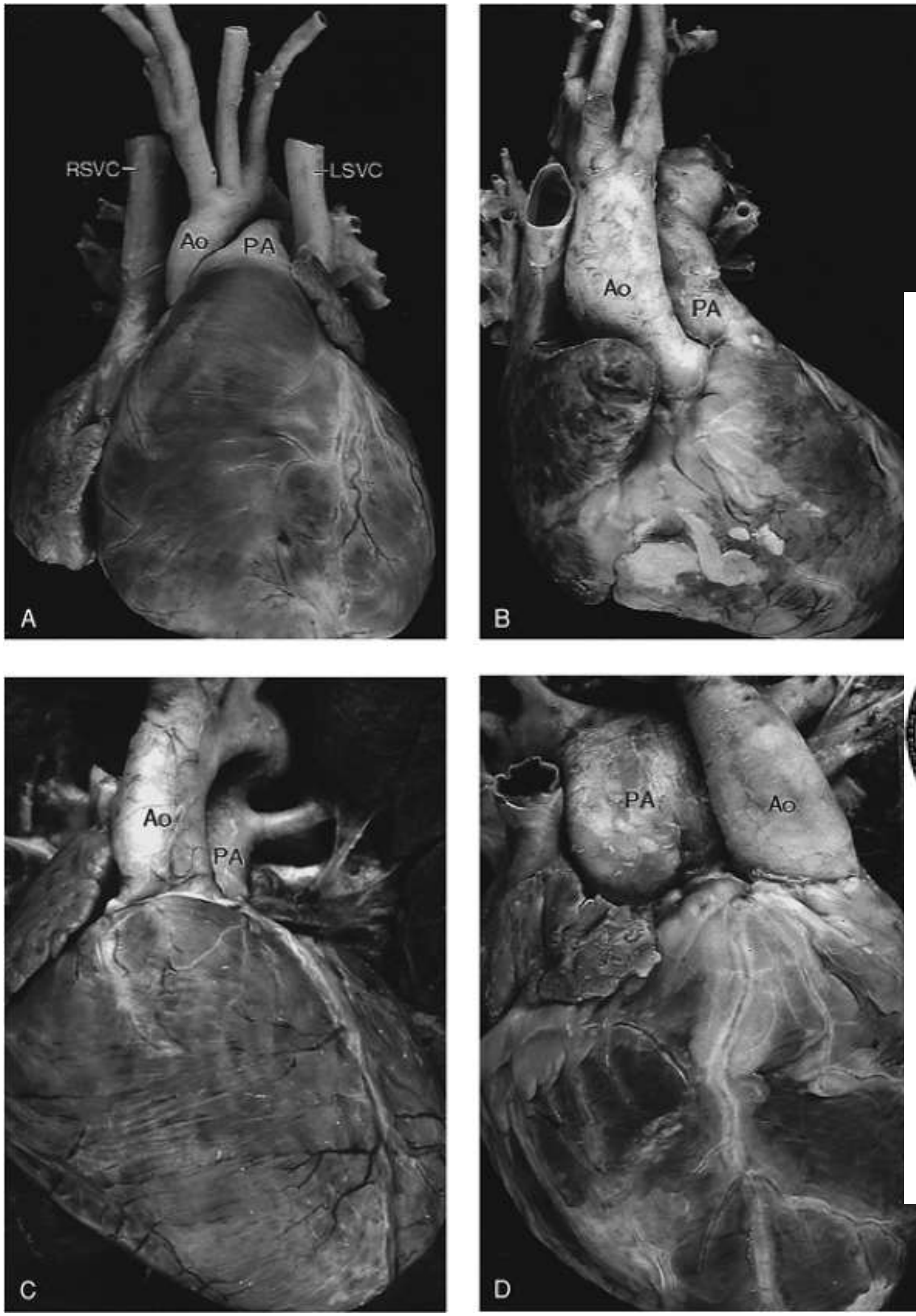
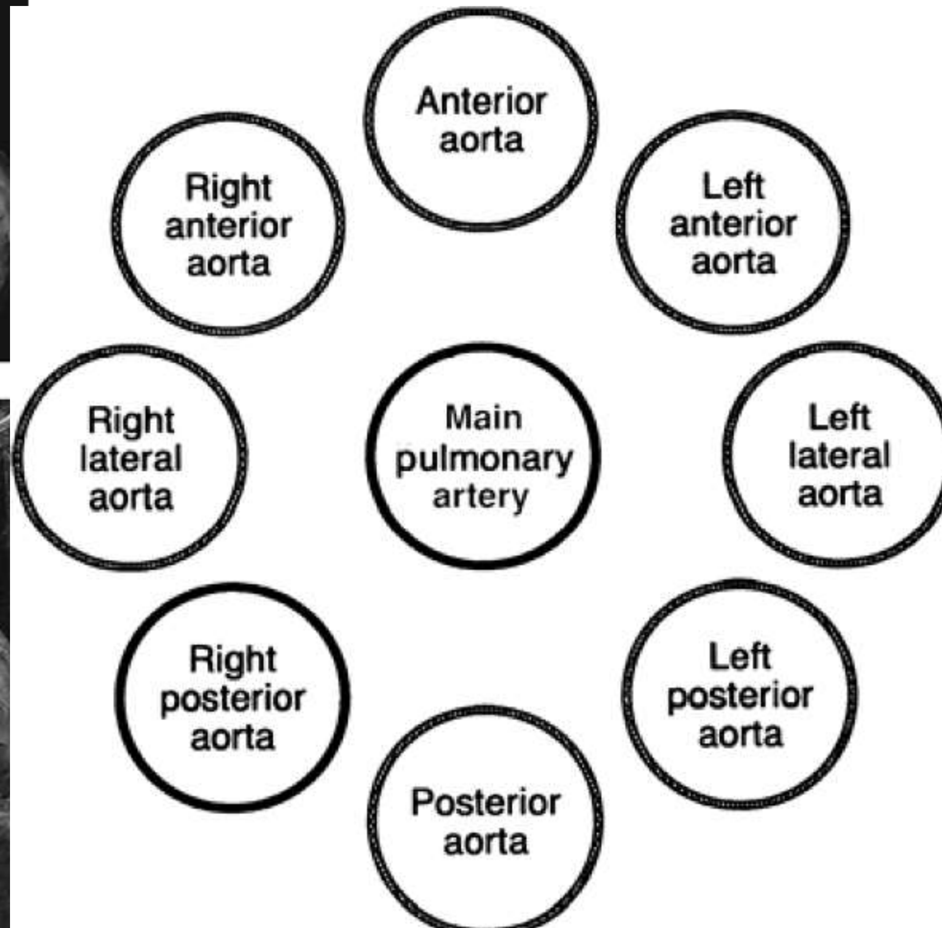


Arterial Segment

A- normal, Pa- anterior, left
Ao-posterior, right



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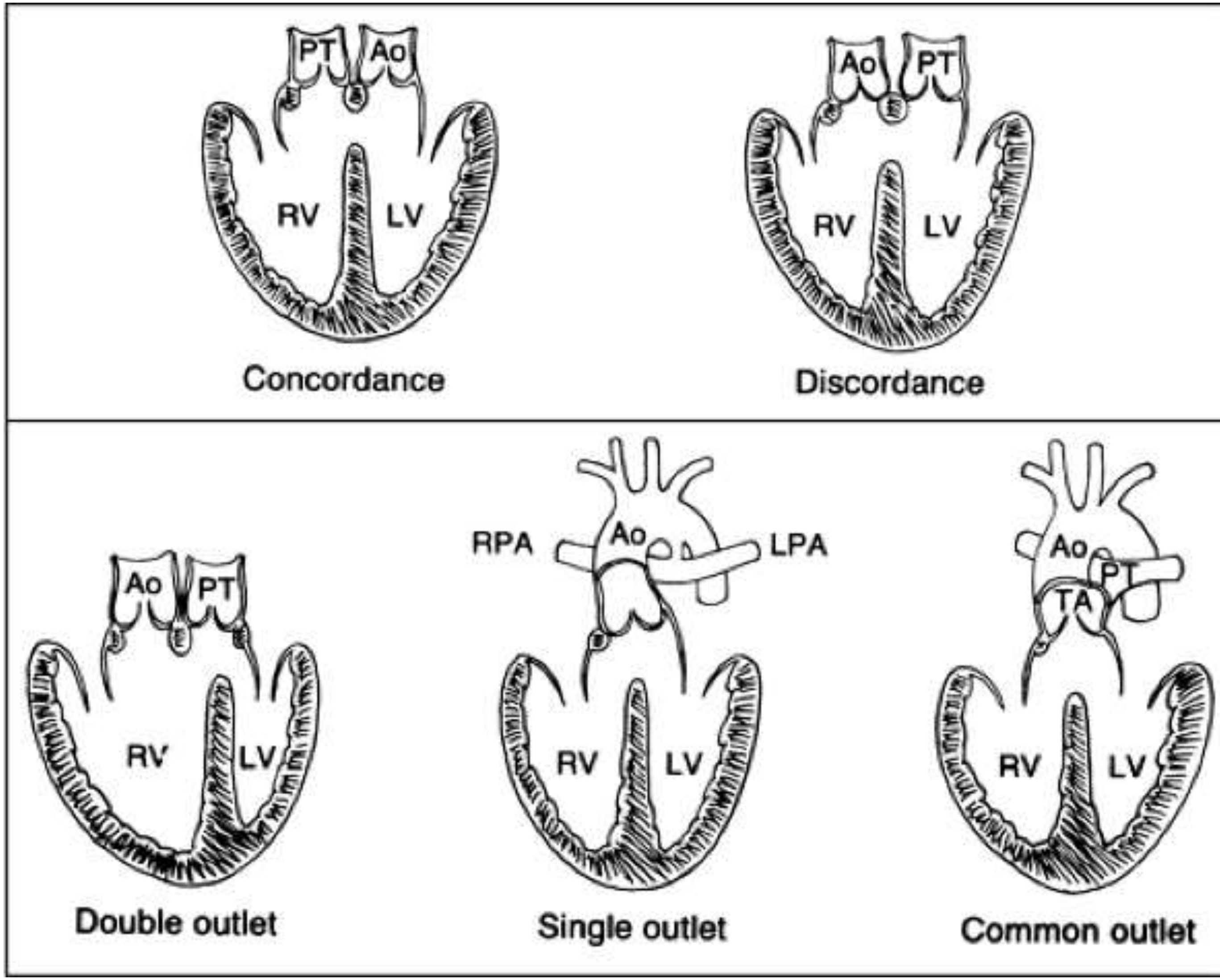
Ventriculoarterial connection- possible



To summarize…….The Cardiac Segments

Viscera and atria

- Abdominal situs
- Systemic and pulmonary venous return
- Atrial anatomy

Atrioventricular canal

- AV valves and atrioventricular septum

Ventricles

- Ventricular anatomy (D- or L-looping)
- Ventricular size and proportion
- Ventricular septum

Conus

- Ventricular outflow tracts

Great arteries

- Semilunar valves
- Great arteries

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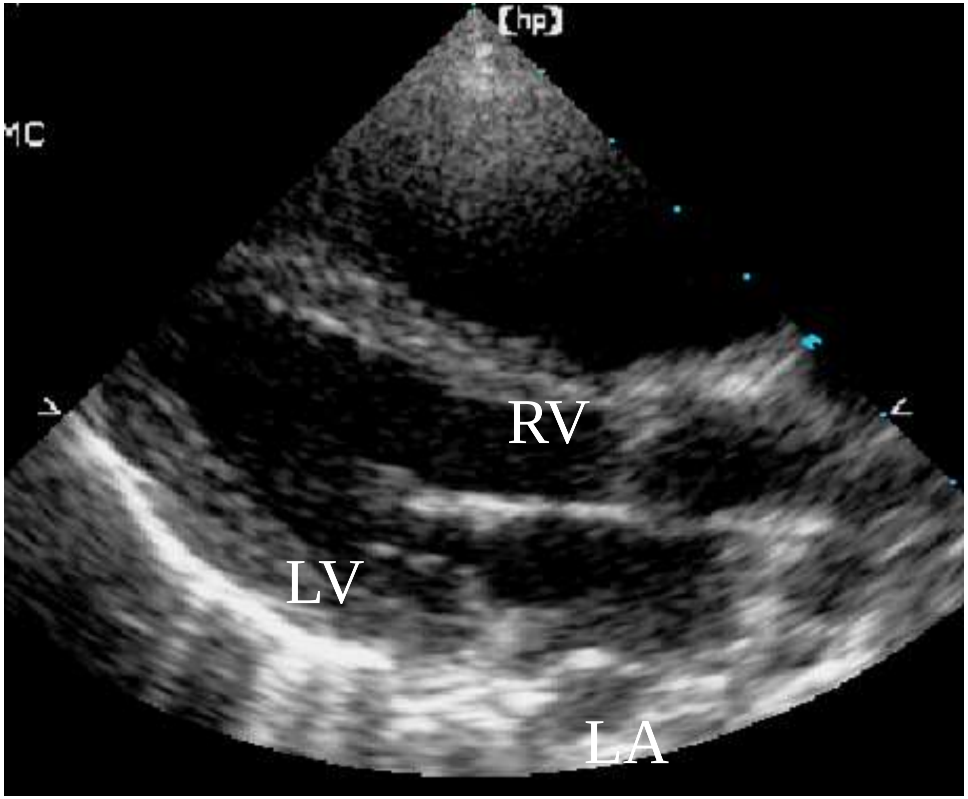
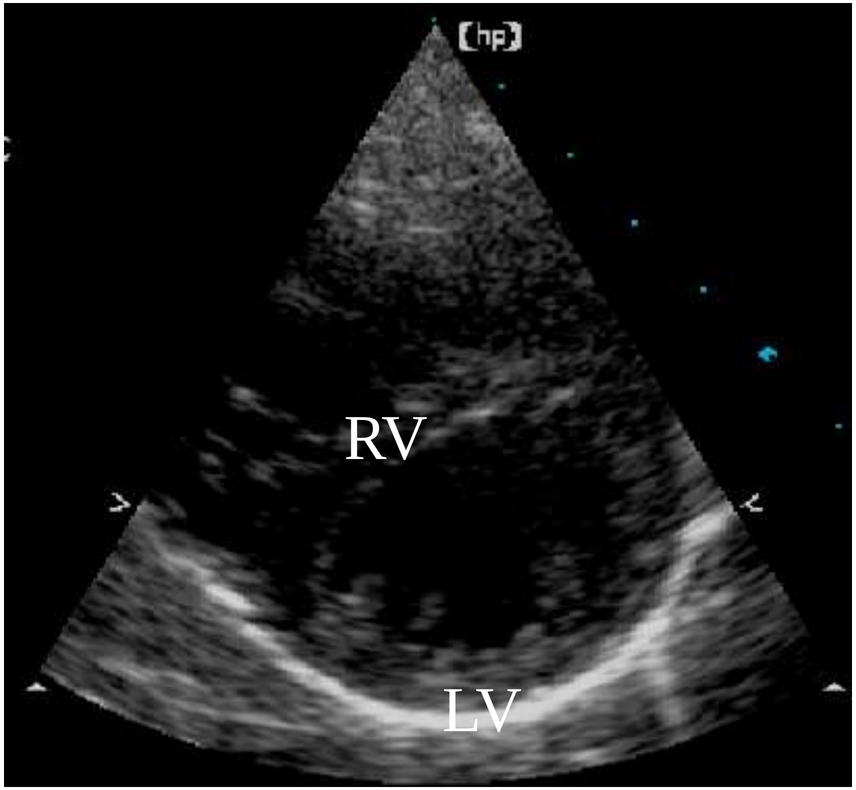
Common Lesions

ASD



RV Dilation



Diastolic Septal Flattening

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Atrial Septal Defects

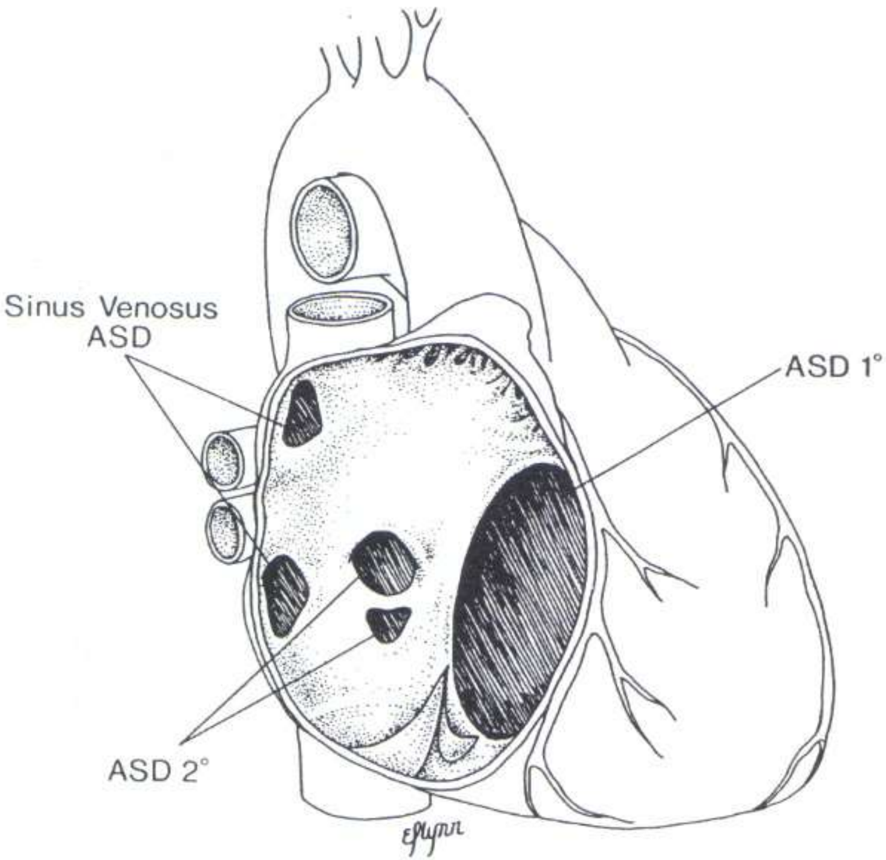
Secundum ASD

Primum ASD

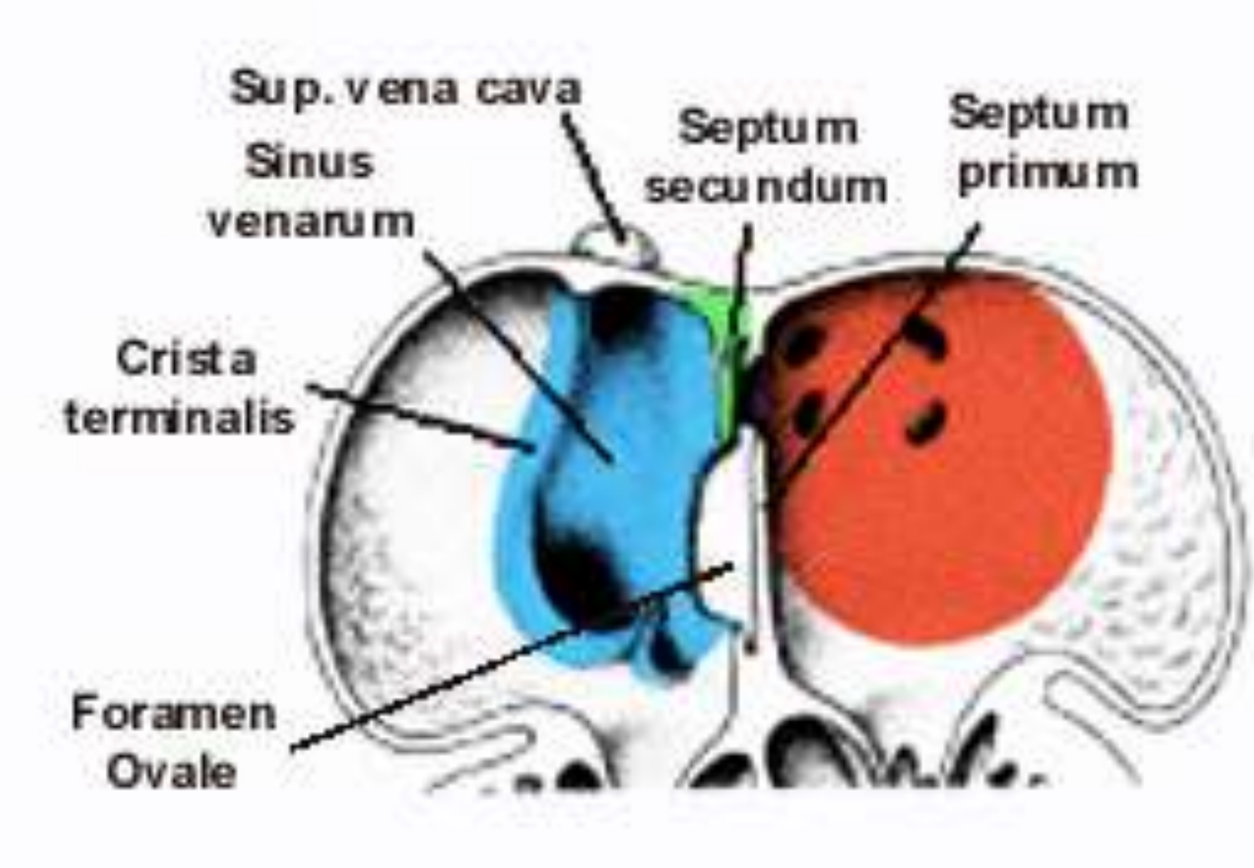
Sinus Venosus defect

- Not truly a deficiency of the atrial septum, but the same physiology as an ASD

Common atrium



Atrial Septal Development



http://www.med.unc.edu/embryo_images/unit-welcome/welcome_htms/contents.htm

Primum ASD

Part of spectrum of AV canal defects

Defect is contiguous with AV valves

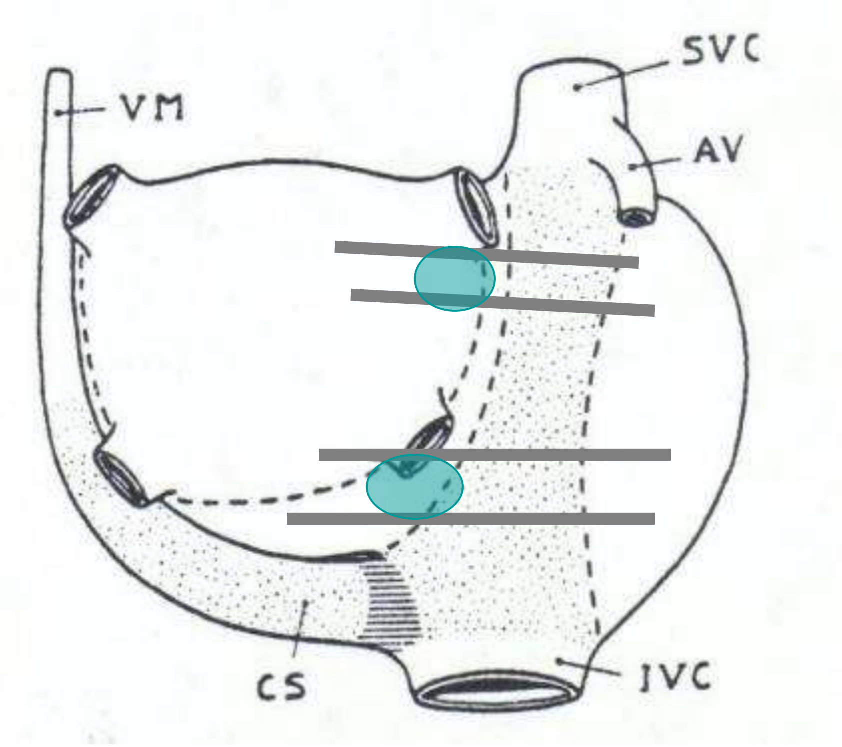
Associated with cleft mitral valve

Sinus Venosus Defects

Deficiency in the wall between the right pulmonary veins and the RA

PAPV-DRAINAGE

- SVC type = RUPV
- Inferior type = RLPV



Sinus Venosus ASD

ASD: Clinical Correlation

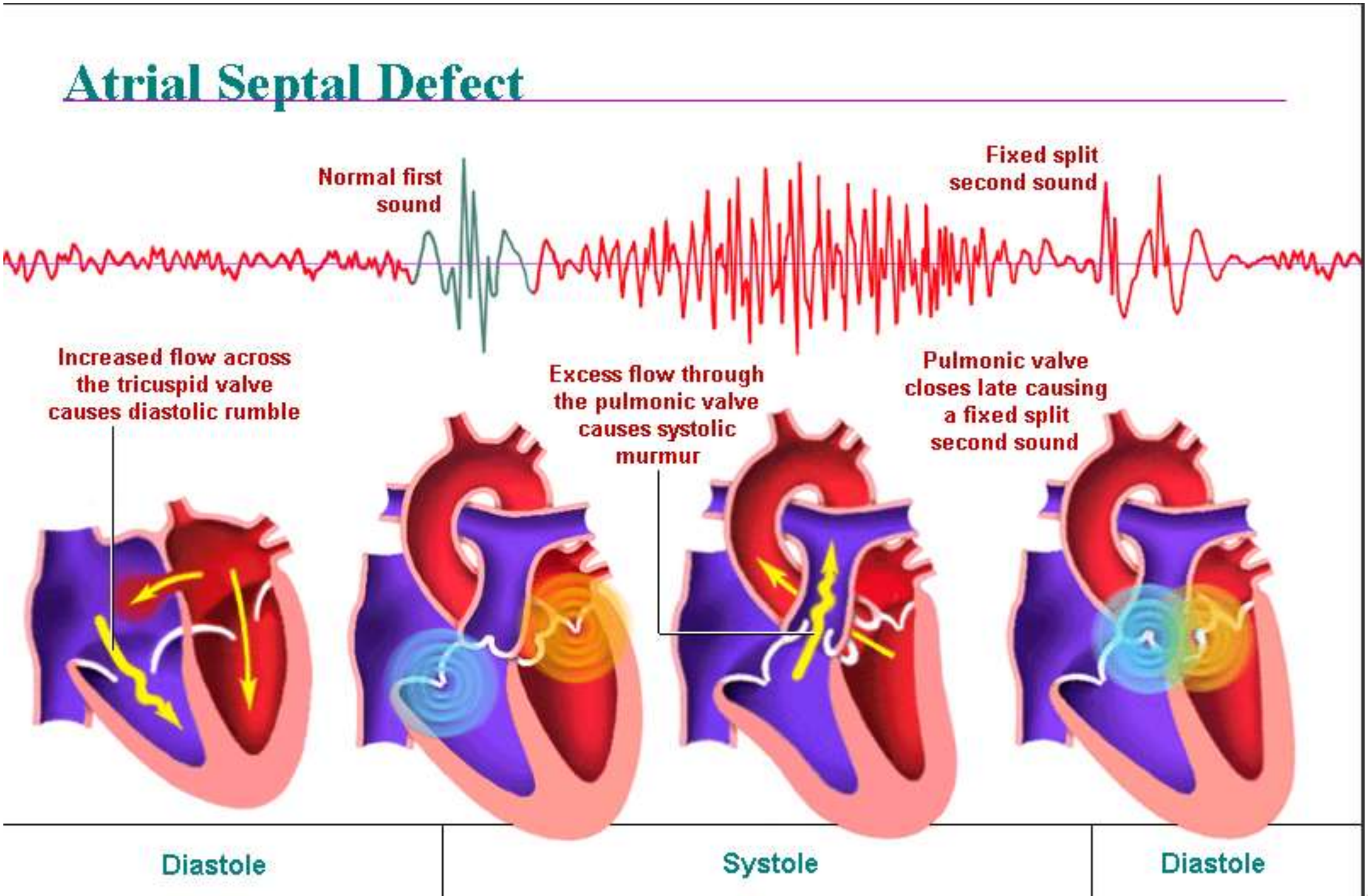
Usually diagnosed in childhood

Asymptomatic

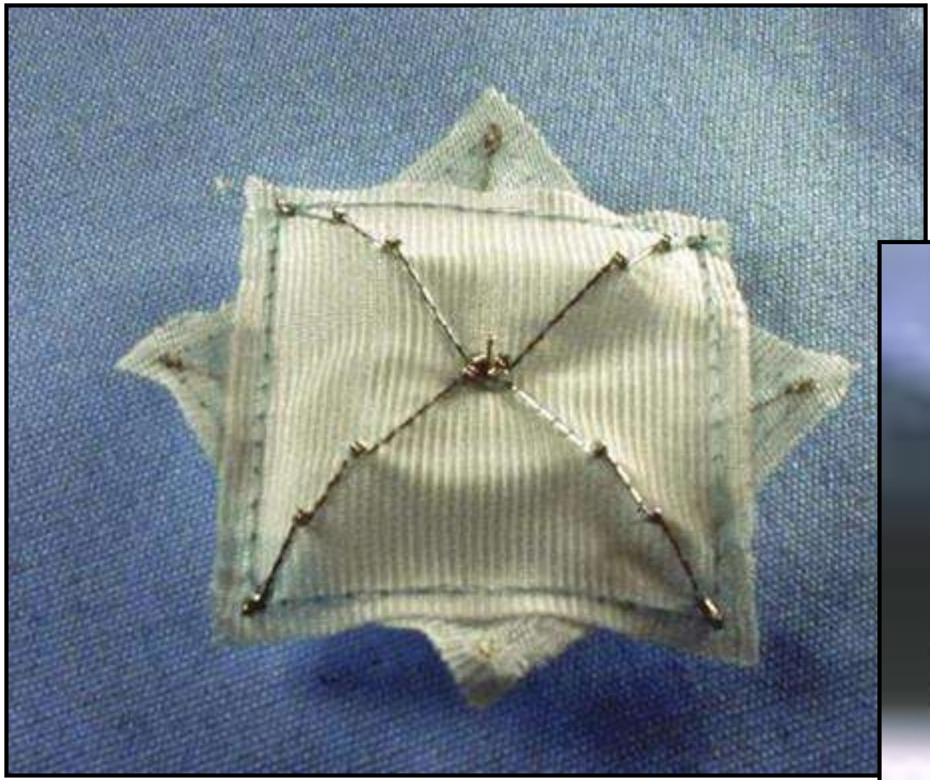
F>M

Systolic ejection murmur and widely split fixed S_2

EKG may show RBBB or RVH


Atrial Septal Defect
Normal first sound
Fixed split second sound
Increased flow across the tricuspid valve causes diastolic rumble
Excess flow through the pulmonic valve causes systolic murmur
Pulmonic valve closes late causing a fixed split second sound
Diastole
Systole
Diastole

Devices for ASD Closure

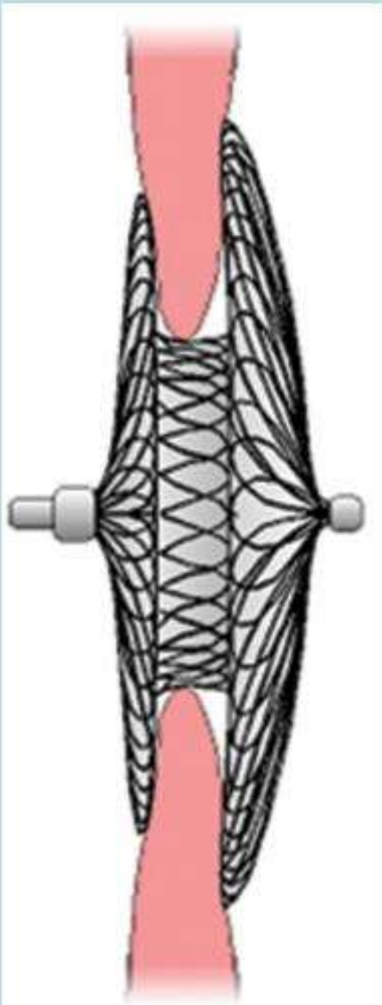


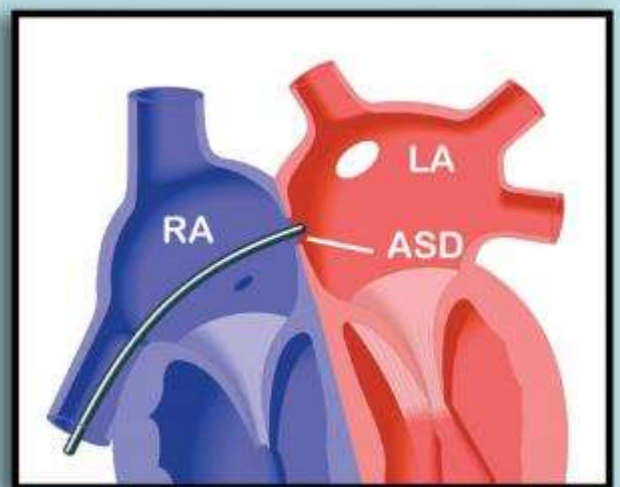
Cardio-SEAL

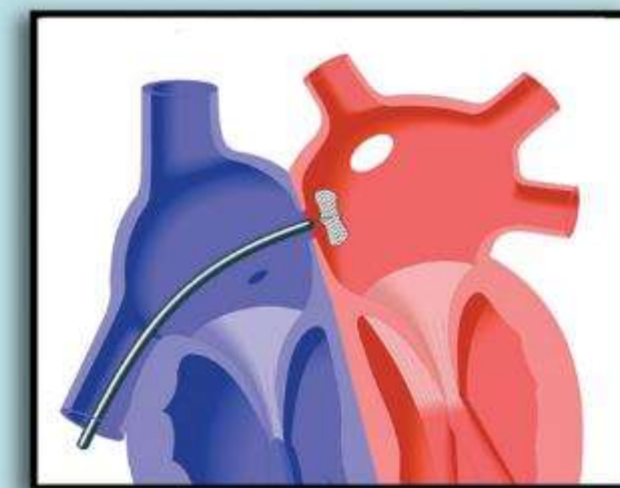
Amplatzer

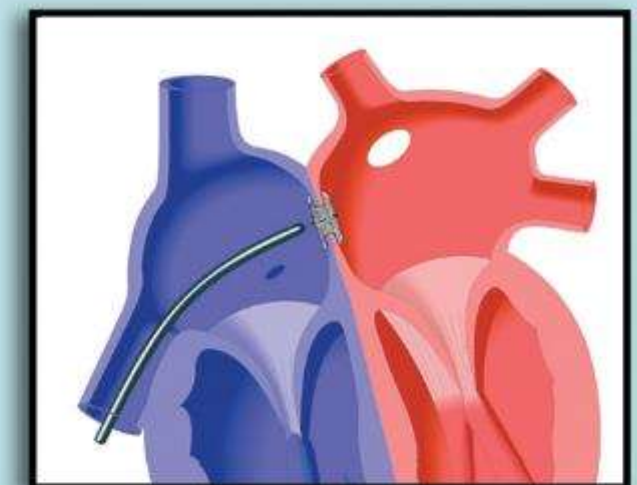
Children's National

Amplatzer Occlusion of Atrial Septal Defect









Clockwise from above: Transcatheter delivery of Amplatzer device, which is positioned across the atrial septal defect

Left: Amplatzer device in place

Newborn infant noted to be breathing heavy in New born nursery

Chest xray demonstrates increased lung markings.

Total Anomalous Pulmonary Venous Return (TAPVR)

I: Supracardiac: common pulmonary vein drains into the right superior vena cava from the left superior vena cava (vertical vein) and the left innominate vein (50%)

II: Cardiac: coronary sinus, right atrium (20%)

III: Infracardiac: subdiaphragmatic (portal vein, inferior vena cava, ductus venosus) (20%)

IV: Mixed: any combination of types I, II, III, the least common

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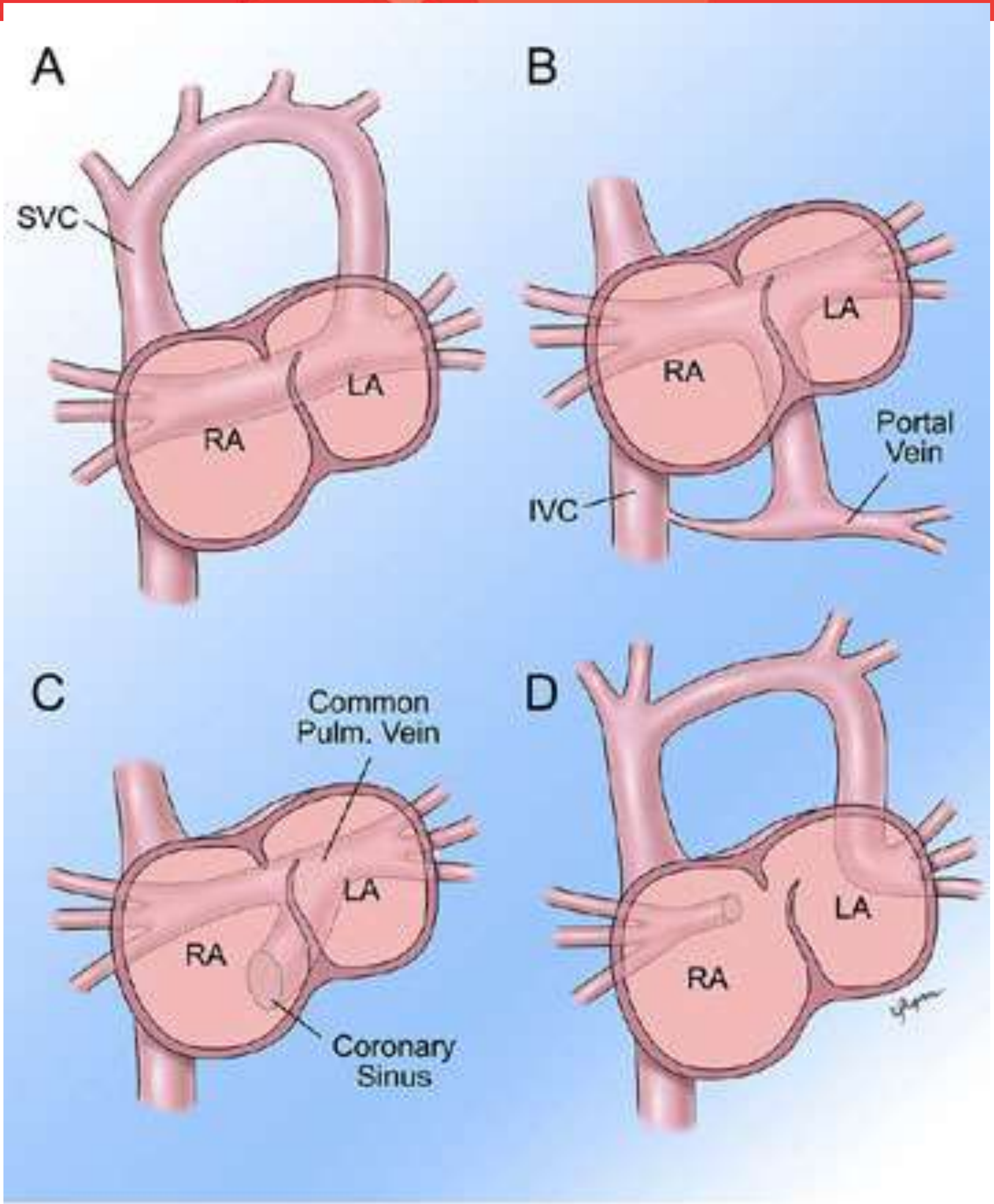
TAPVR



A
SVC
RA
LA
B
RA
LA
Portal Vein
IVC
C
Common Pulm. Vein
RA
LA
Coronary Sinus
D
RA
LA

Partial Anomalous Pulmonary Venous Return (PAPVR)

Right veins (more common):

RA

SVC (RUPV to the RA or base of the SVC-sinus venosus ASD)

IVC

Left veins:

Innominate vein

Coronary sinus

Rarely: SVC, IVC, right atrium, or left subclavian vein

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“Very loud murmur” heard prior to hospital discharge

Baby is well, feeding, growing, pink, passed new pulse ox screening

Children's National

The Ventricular Septum

AV canal septum (1)

Muscular septum including the trabecular portion (2) and the septal band (3)

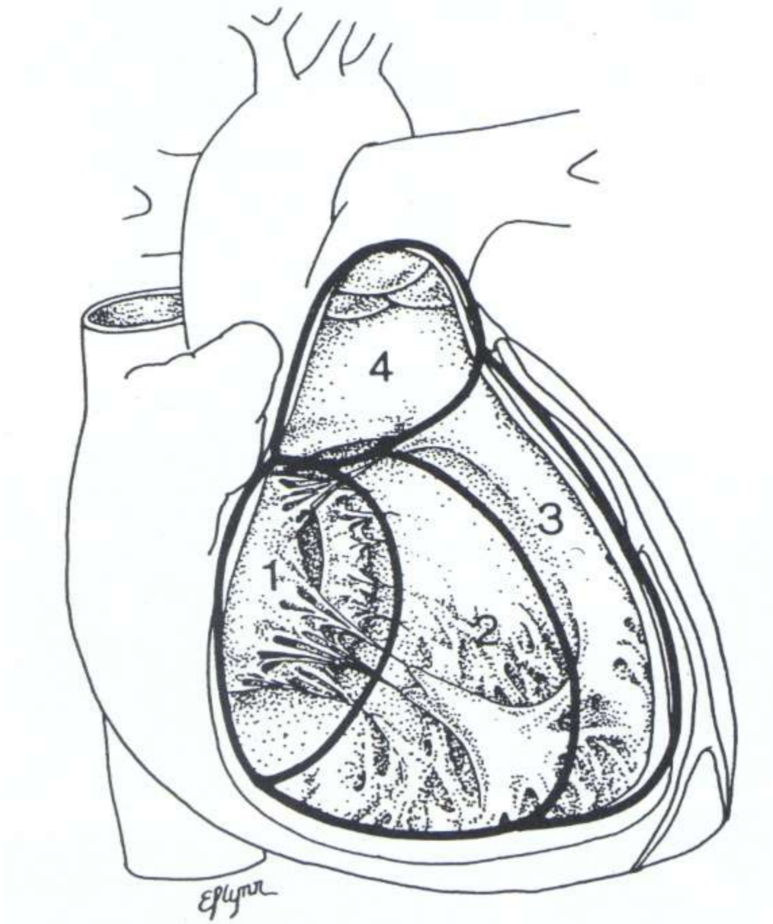
Conal septum (4)

Conoventricular

Membranous

Inlet

Malalignment



The Ventricular Septum

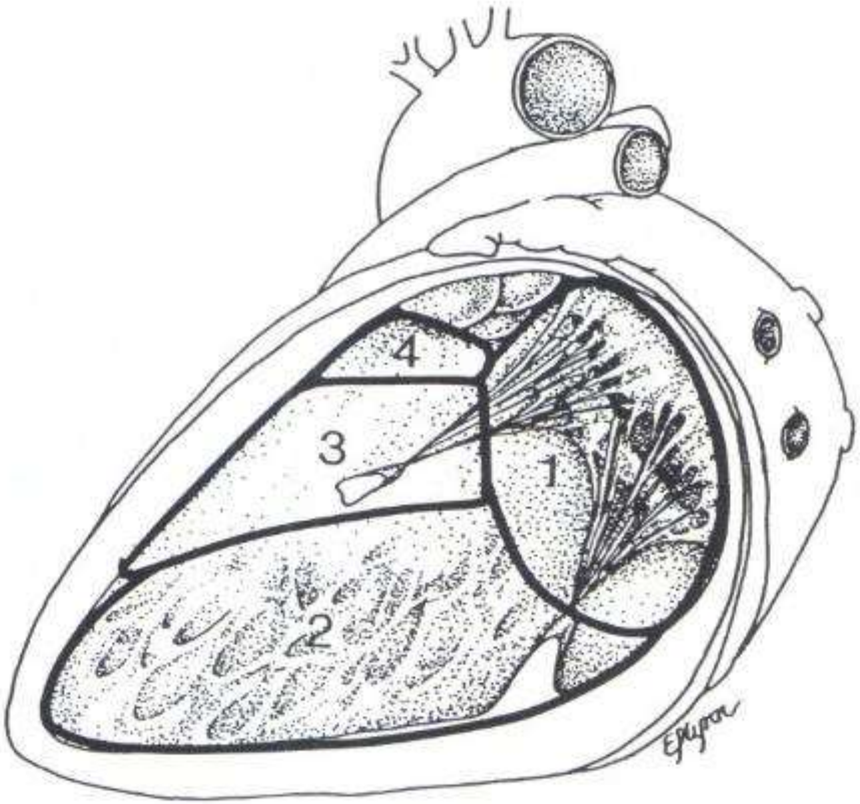
Left ventricular view

AV canal septum (1)

Muscular septum including th

and the septal band (3)

Conal septum (4)



VSD: Clinical Correlation

Size and pulmonary vascular resistance determines clinical presentation

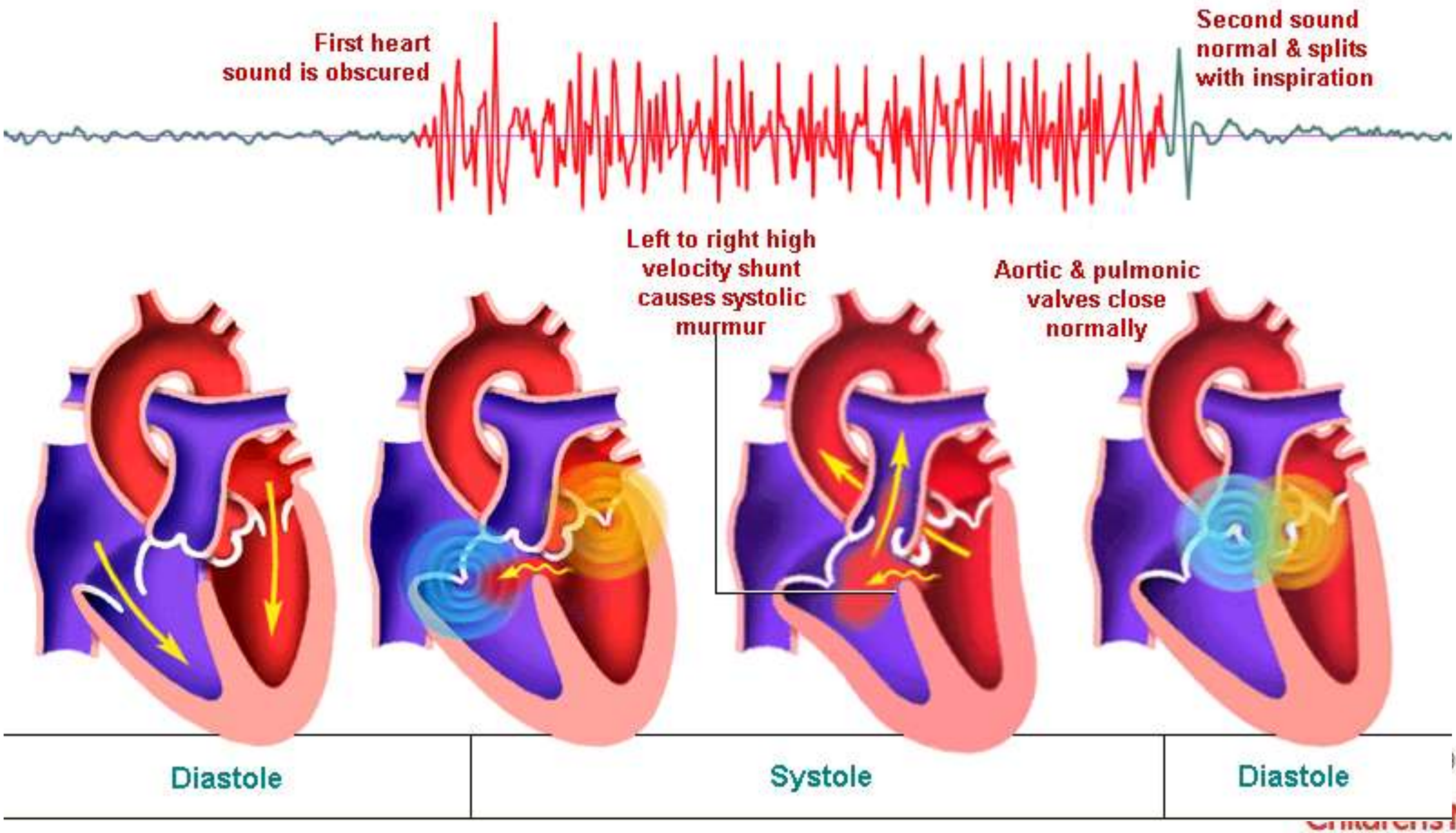
- Fetal transition

Symptoms are determined by the size of the shunt

- Size of defect
- Presence of other anomalies
- Extracardiac abnormalities


(.wav)

Small Ventricular Septal Defects



VSD: Clinical Correlation

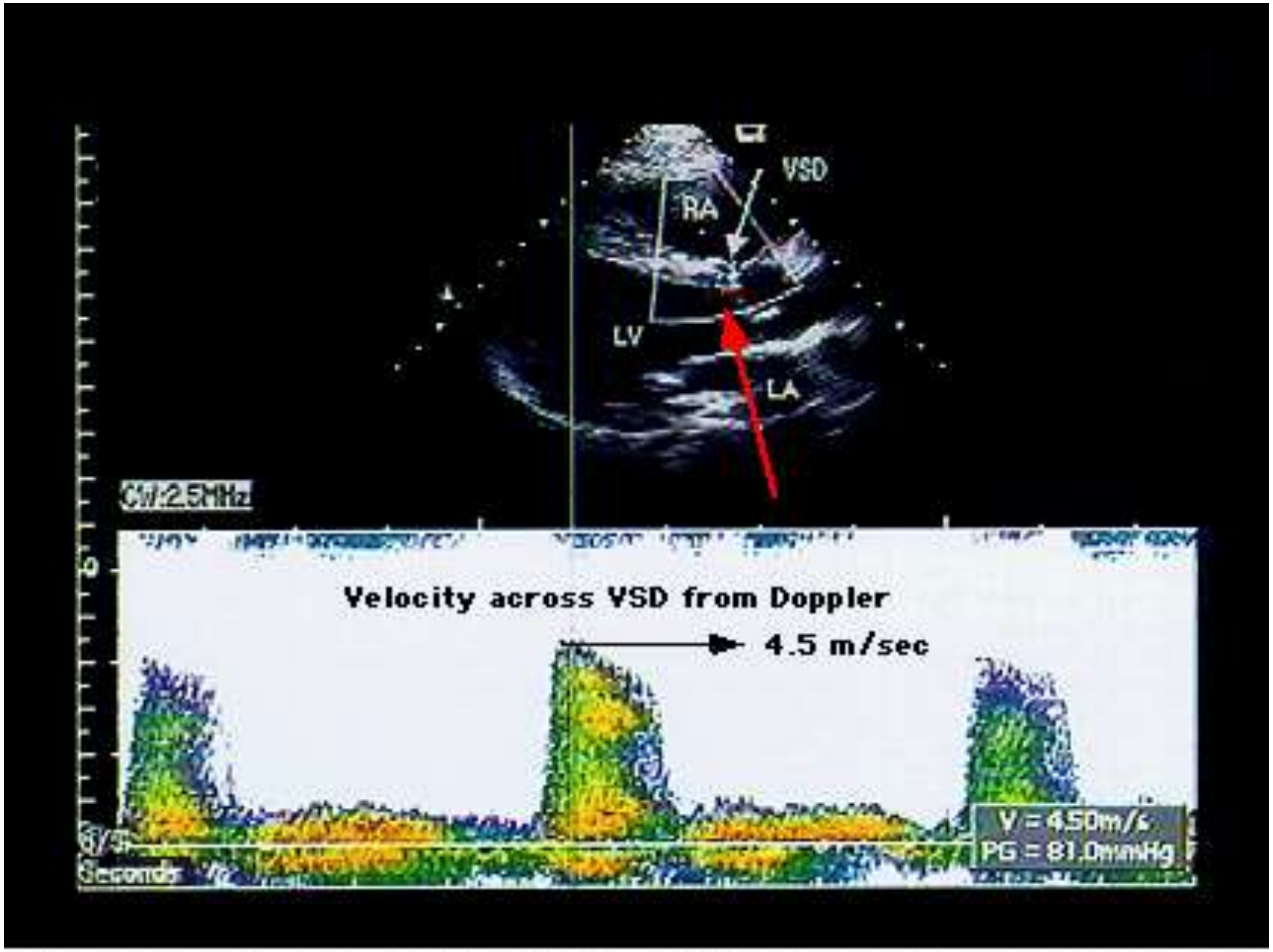
Spontaneous resolution

Or not...

Pulmonary disease

- Eisenmenger's syndrome

Aortic regurgitation



Continuous wave Doppler in ventricular septal defect The echocardiographic frame demonstrated the Doppler determination of pressure gradient across a membranous ventricular septal defect (VSD) (white arrow). The direction of the continuous wave Doppler beam used to obtain the velocity across the ventricular septal defect is illustrated by the red arrow. The velocity (V) is 4.5 m/sec and based upon the modified Bernoulli equation, (pressure = [velocity] 2 x 4) the gradient is 81 mmHg. (Courtesy of Ann Kavanaugh-McHugh, MD.)

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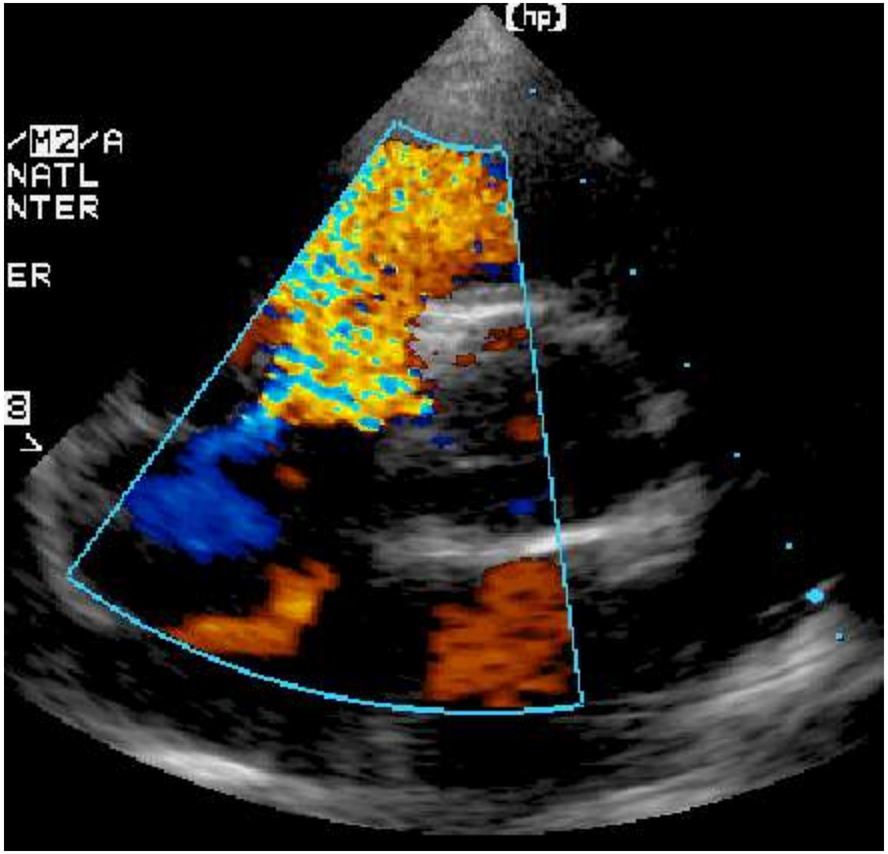
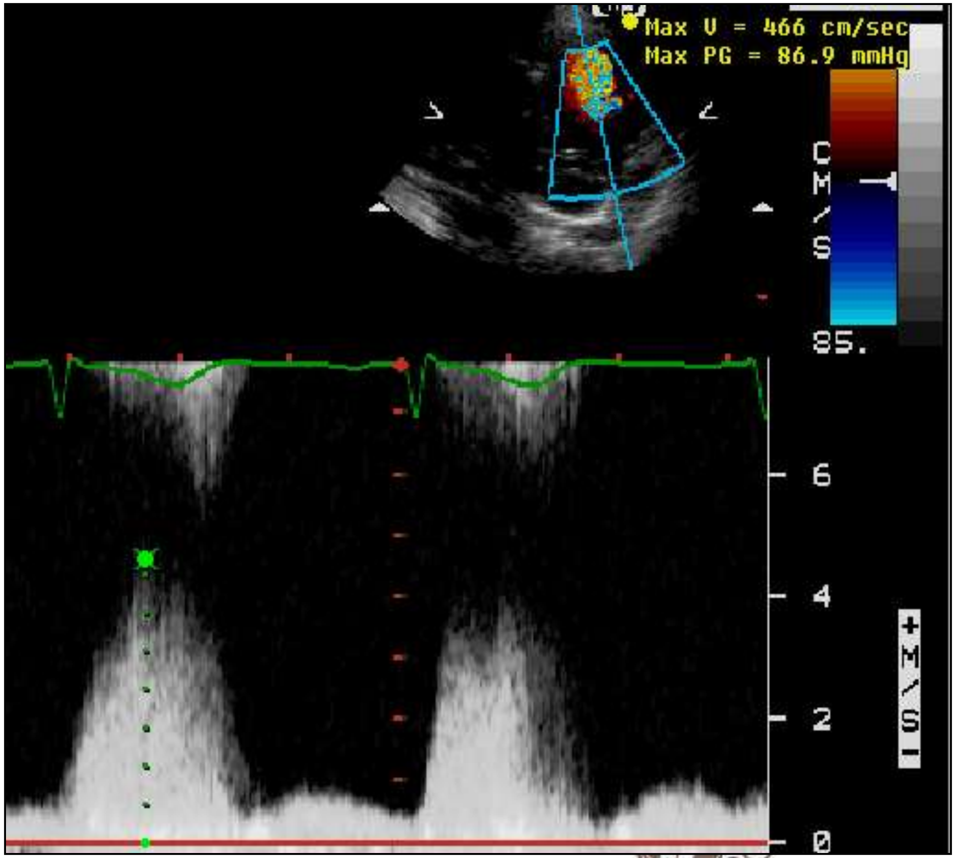
Restrictive Membranous VSD





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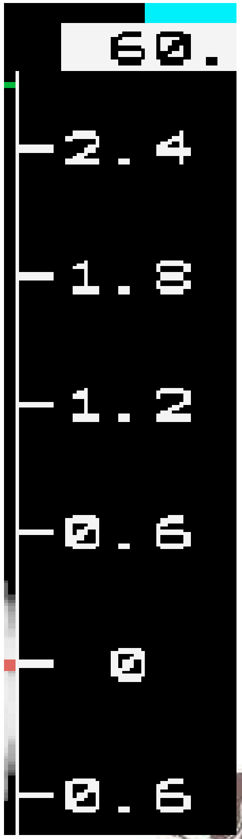
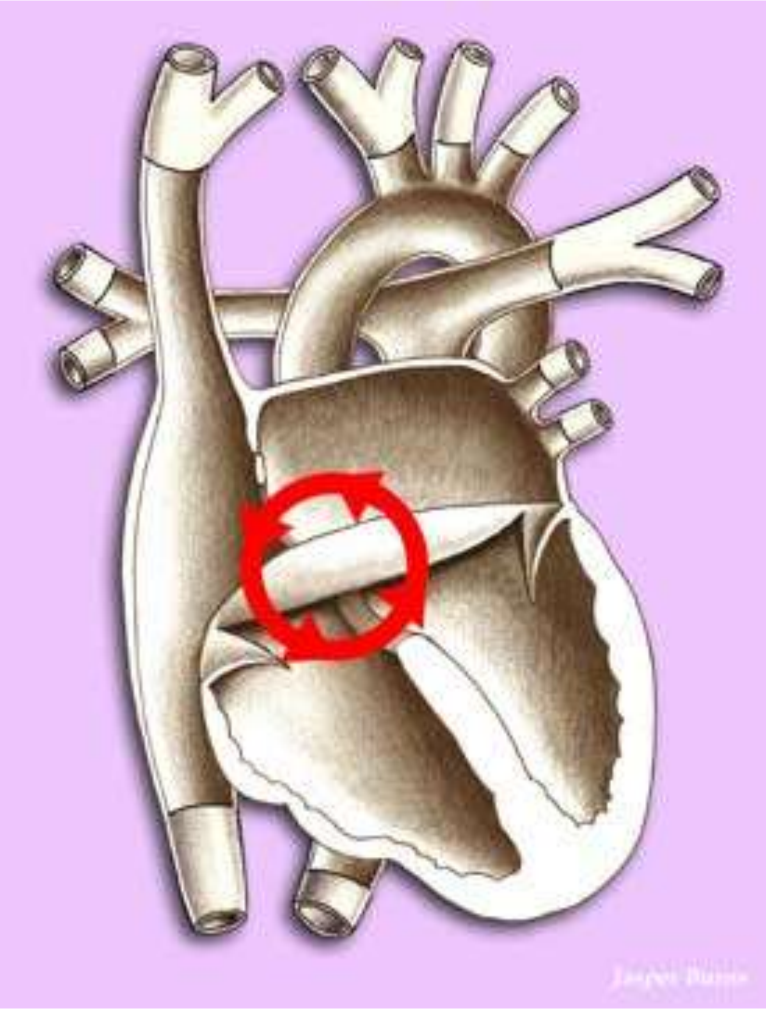
Unrestrictive Membranous VSD



Atrioventricular Canal Defect- Complete



Common AV Canal (CAVC)

Endocardial Cushion Defect (ECD)

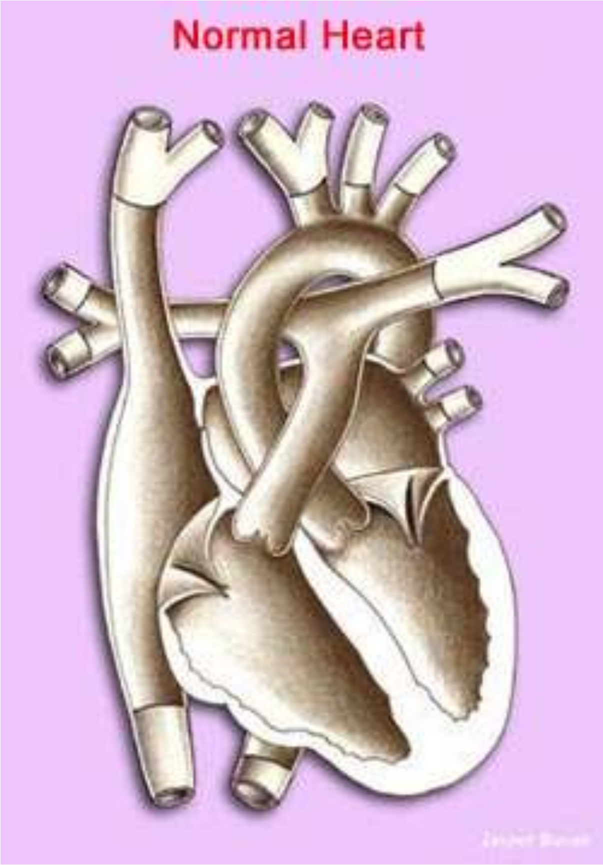
Atrioventricular Septal Defect (AVSD)

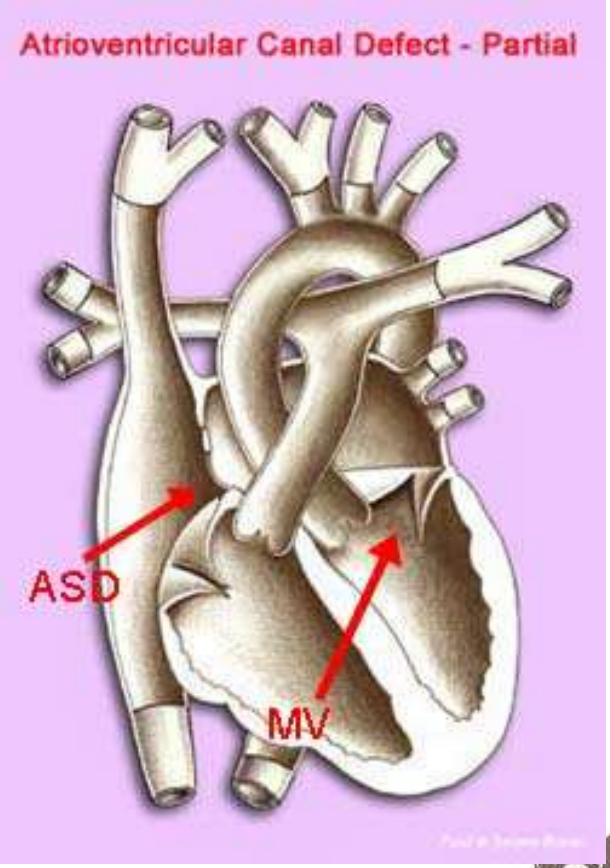
Failure of the AV canal to develop properly and form tricuspid, mitral valves and portions of atrial and ventricular septae

Definitions
Spectrum of defects

- Incomplete CAVC = lack the VSD component or ASD component
- Partial CAVC = synonym for incomplete CAVC *OR* = primum ASD with cleft mitral valve
- Transitional CAVC = small VSD component
- Balanced/Unbalanced

Atrioventricular Canal Defect – Partial





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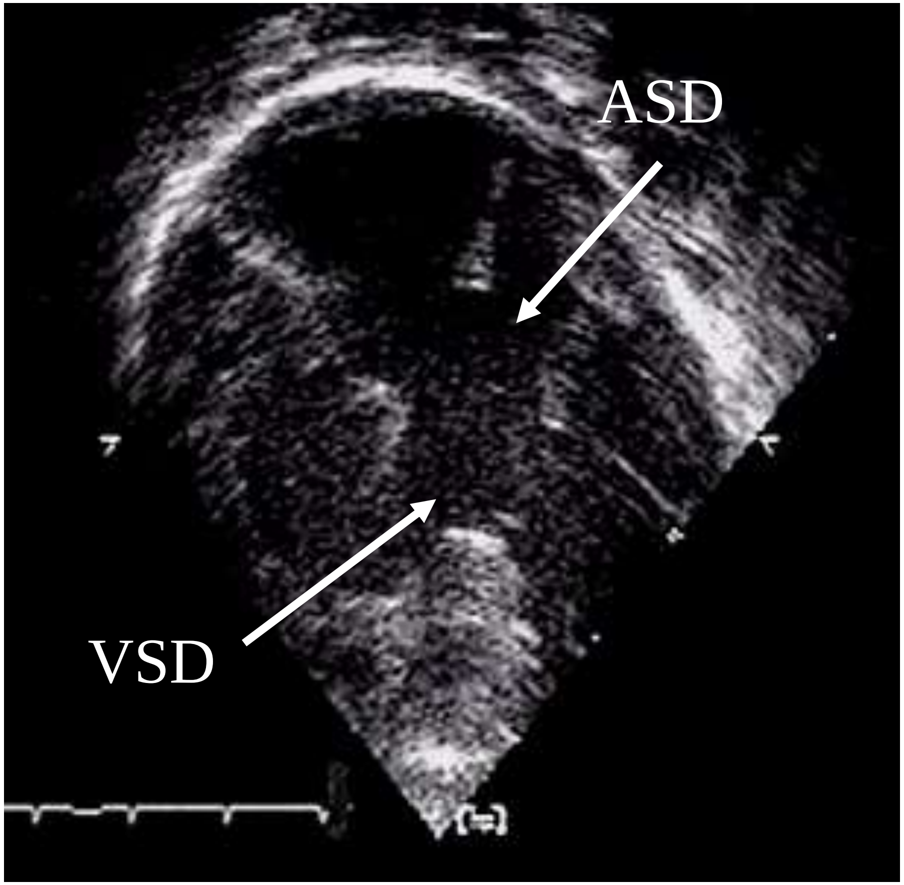
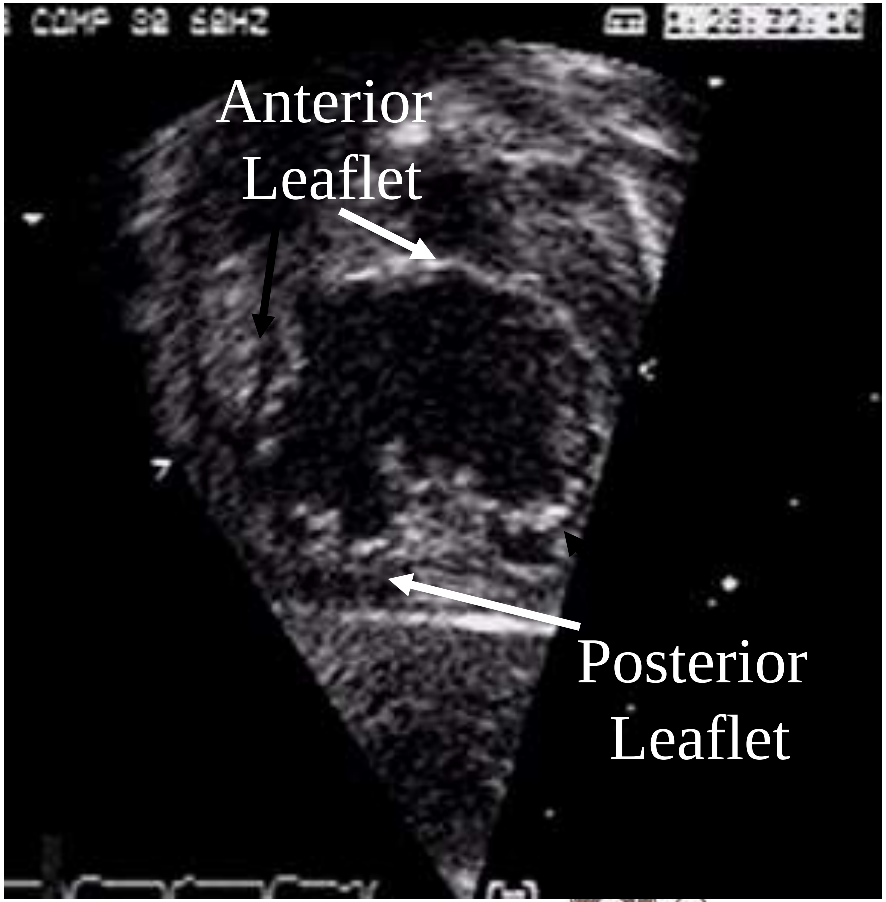
AV Septal Defect

Complete





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Bonus points…

You are doing the echo on a baby and diagnosis her with an Unbalanced AVC.

You are having a hard time imaging the aortic arch.

Are you concerned, or do you think to yourself, the arch is always hard to image, these babies have no necks, they can't stand when I put my transducer there….I am sure its fine, I just can't see it right now…..

You are called to NICU to echo 28 week premature baby, weight is 600 gm, every time you try to image the baby's HR falls and alarms go off…

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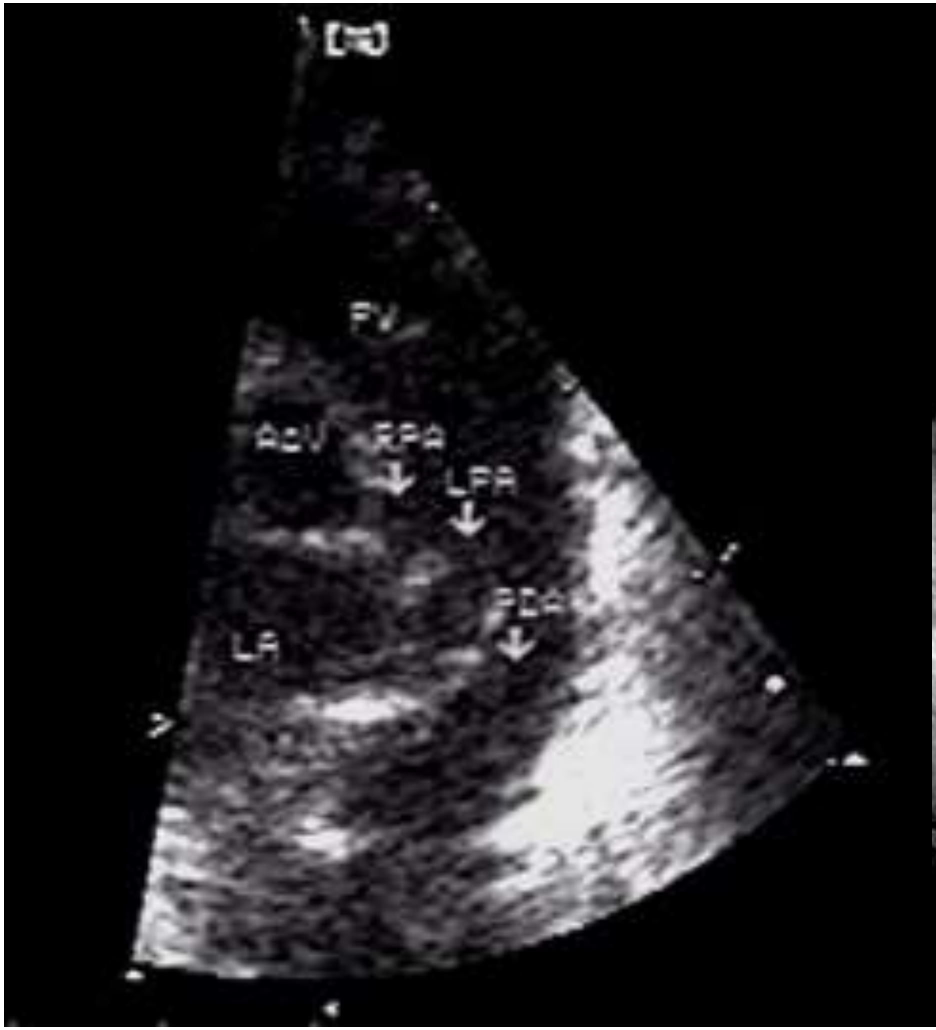
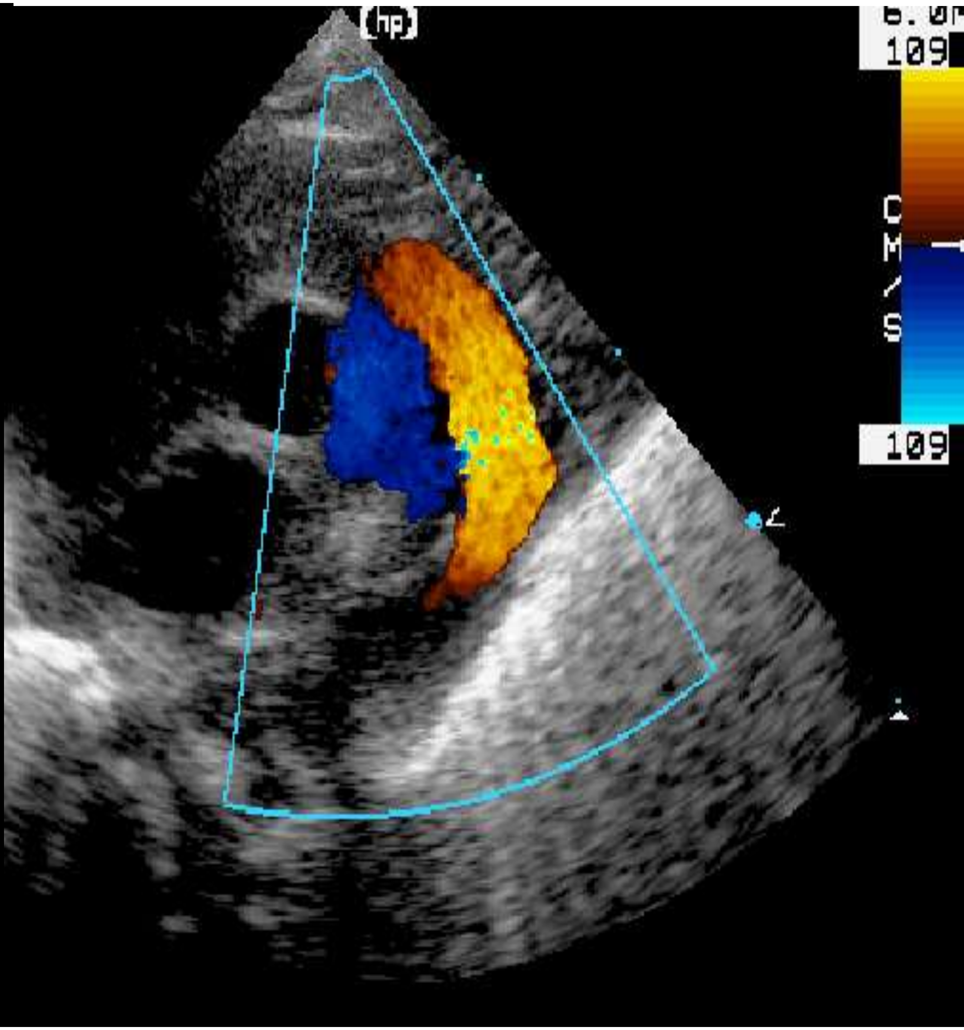
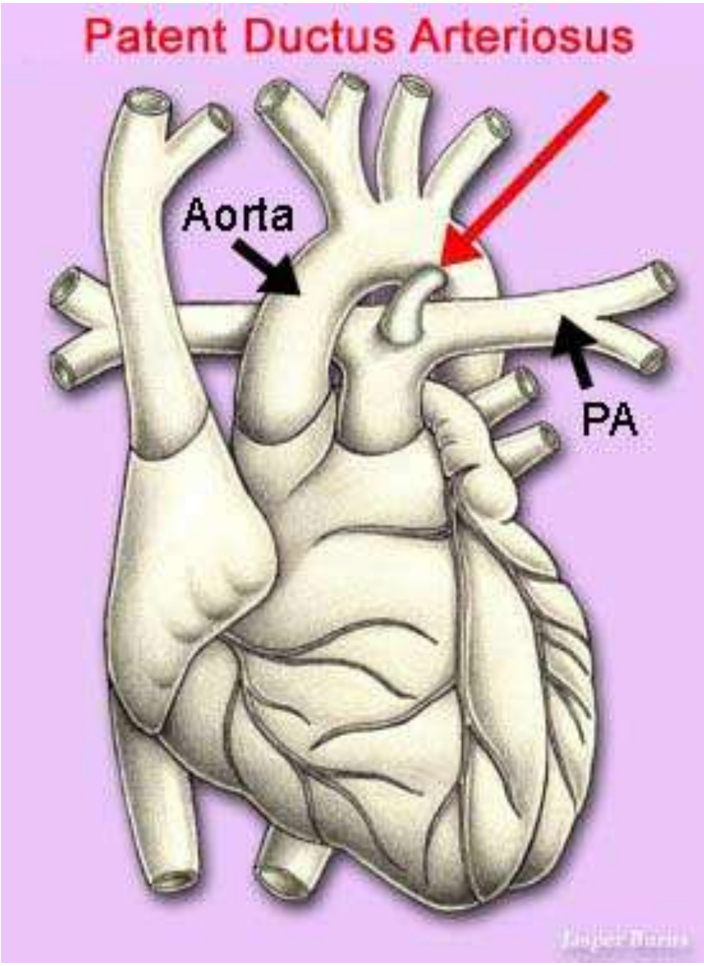
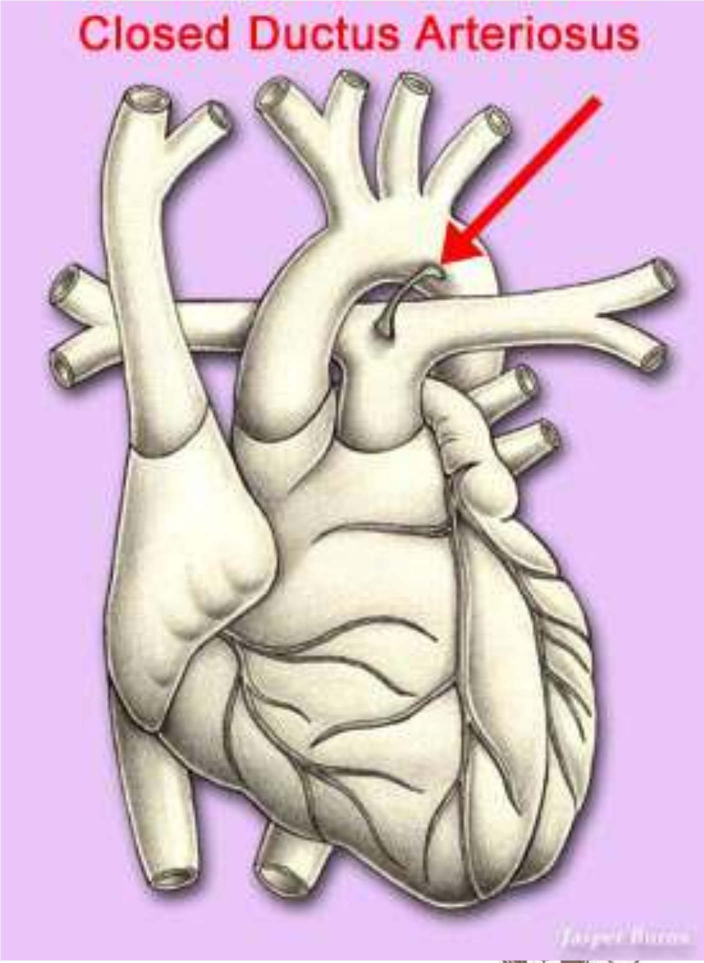
Patent Ductus Arteriosus





Patent Ductus Arteriosus





PDA: Clinical Correlation

Closed in 90% of infants by 48 hours of life

- Prematuring, altitude

Anatomy

- Derived from the left 6^{th} embryonic arch

Closure

- Muscular constriction→endothelium→thrombosis→fibrous strand

Physiology↔ shunting

- Symptoms proportional to shunting

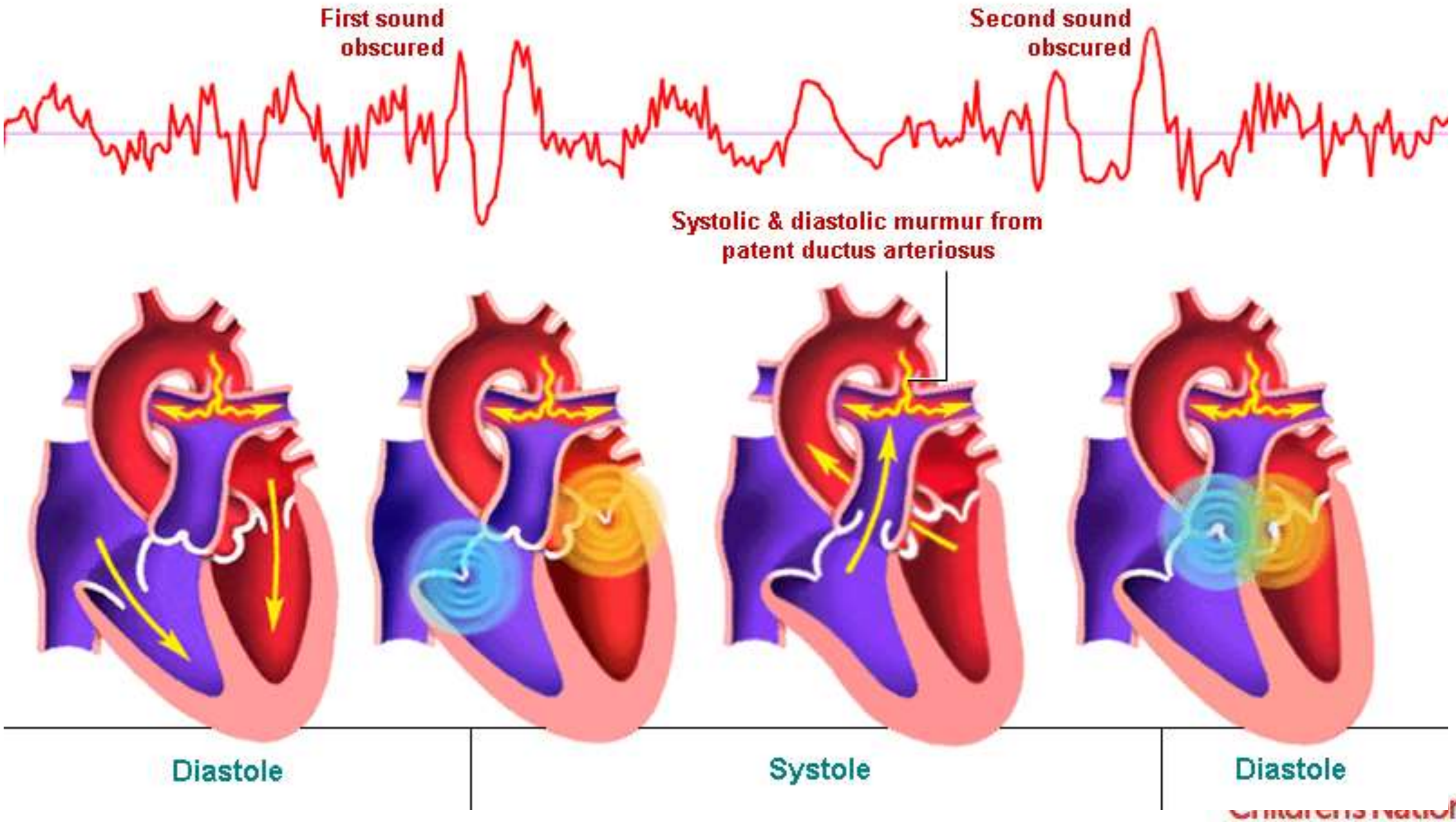
Murmur

EKG

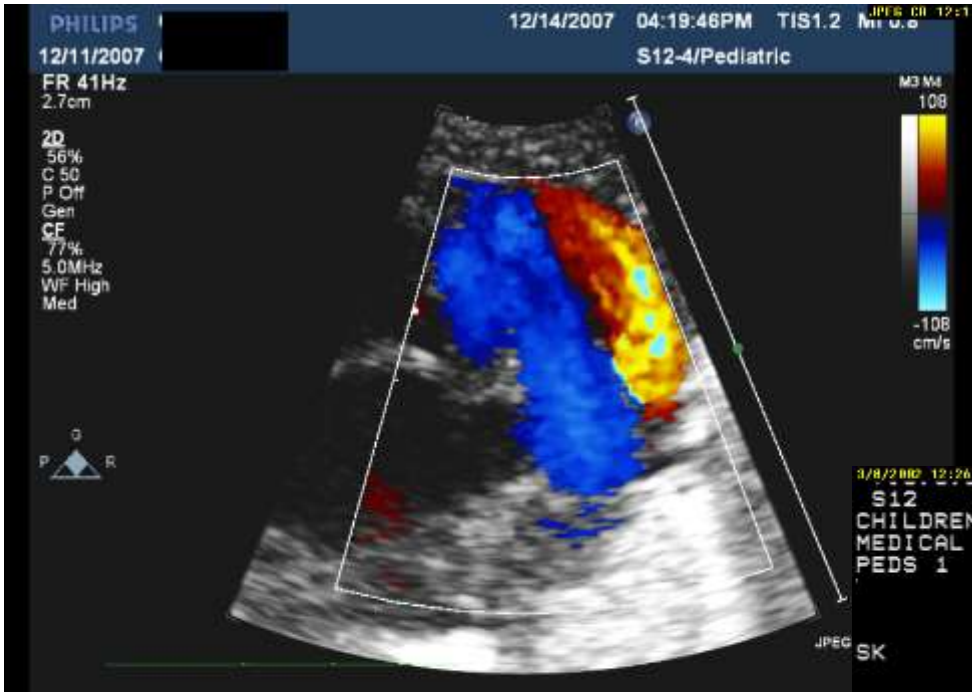
- Ventricular hypertrophy

(.wav)

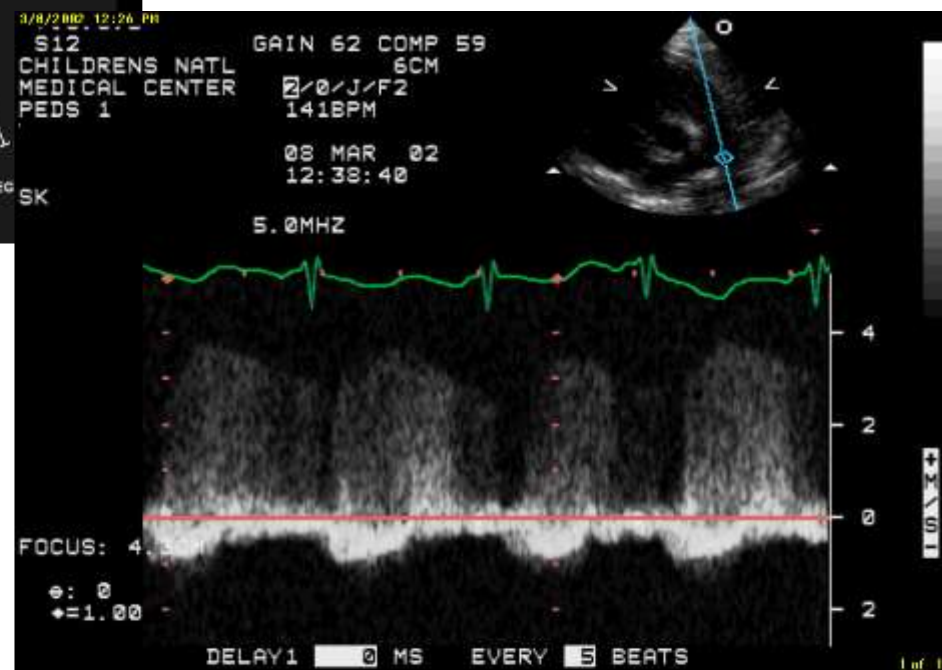
Patent Ductus Arteriosus



Doppler of the PDA (L-R shunt)

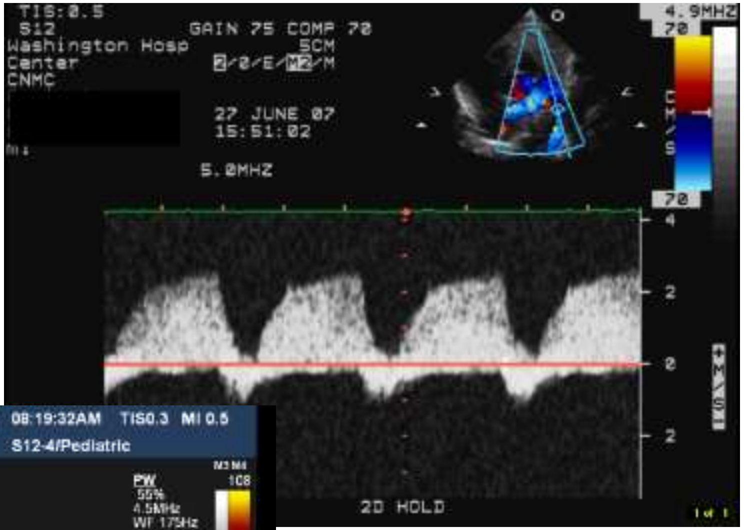


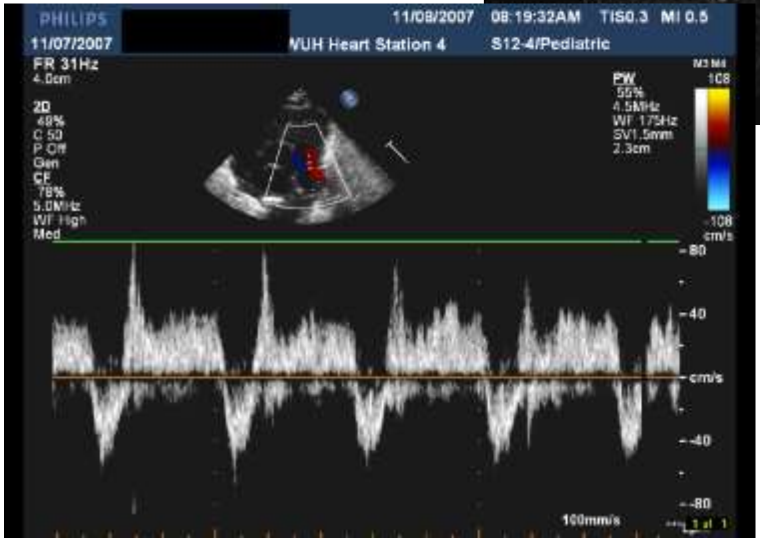
Color flow Doppler (left) showing a L-R shunt from the descending aorta through the PDA to the PA (red: towards the probe)



CW Doppler tracing (right) seen above the baseline indicating flow toward the probe from the descending aorta through the PDA to the PA. The peak velocity is reached in late systole 4 m/s. L-R shunt

Doppler of the PDA (bidirectional shunt)

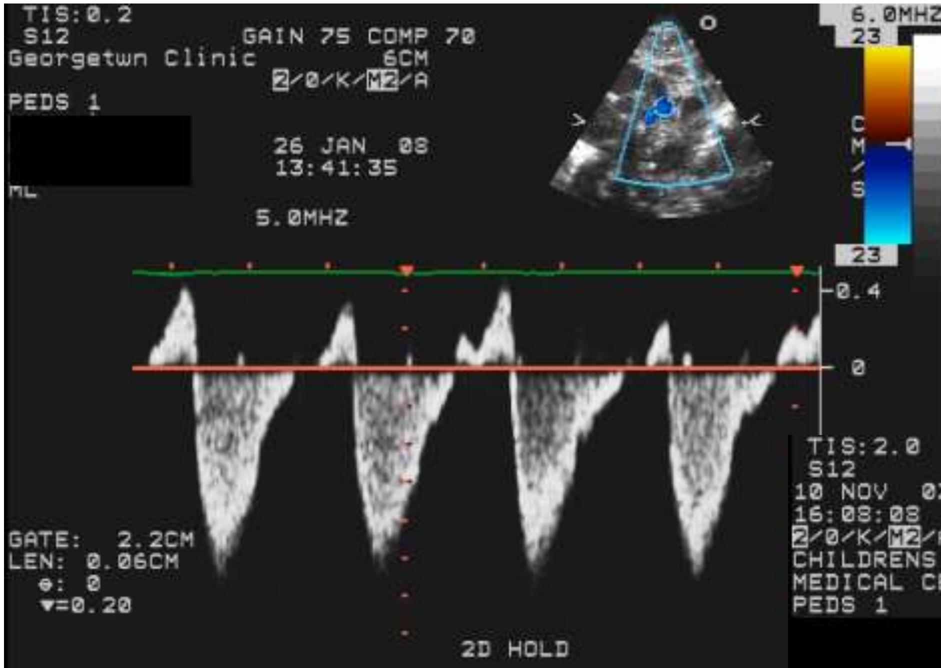


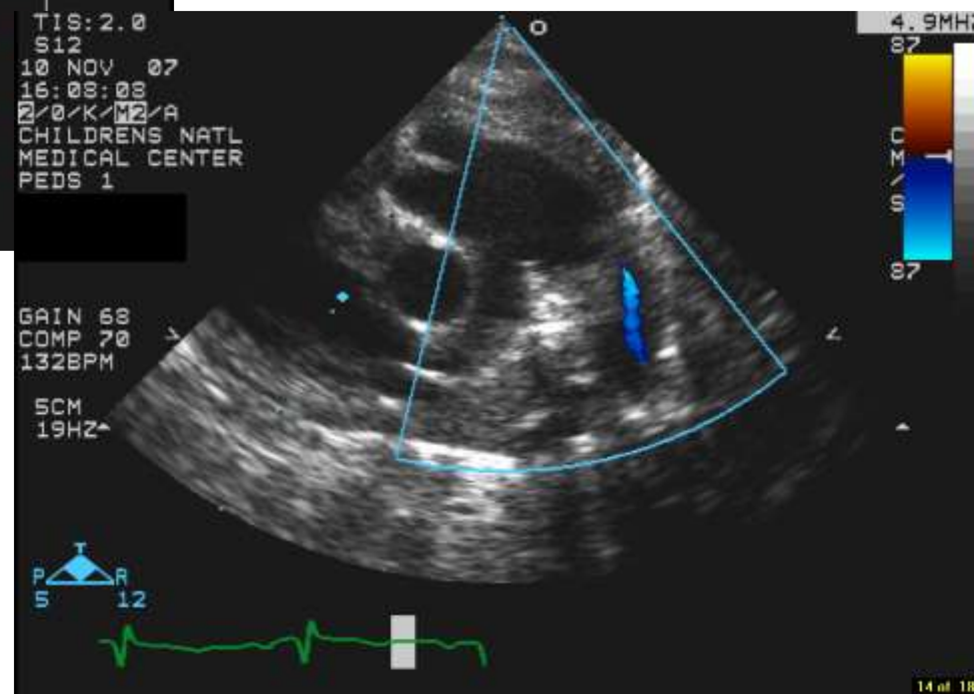


Bidirectional blood flow through the PDA can be a normal finding in newborn infants due to high pulmonary resistance

CW Doppler from an infant with pulmonary artery hypertension and PDA. The negative deflection in systole below the baseline arises from the R-L shunt through the PDA from the PA to the Dao (away from the TDX).
The positive deflection (late systole-into late diastole) arises from L-R shunt through the PDA from the Dao to the PA

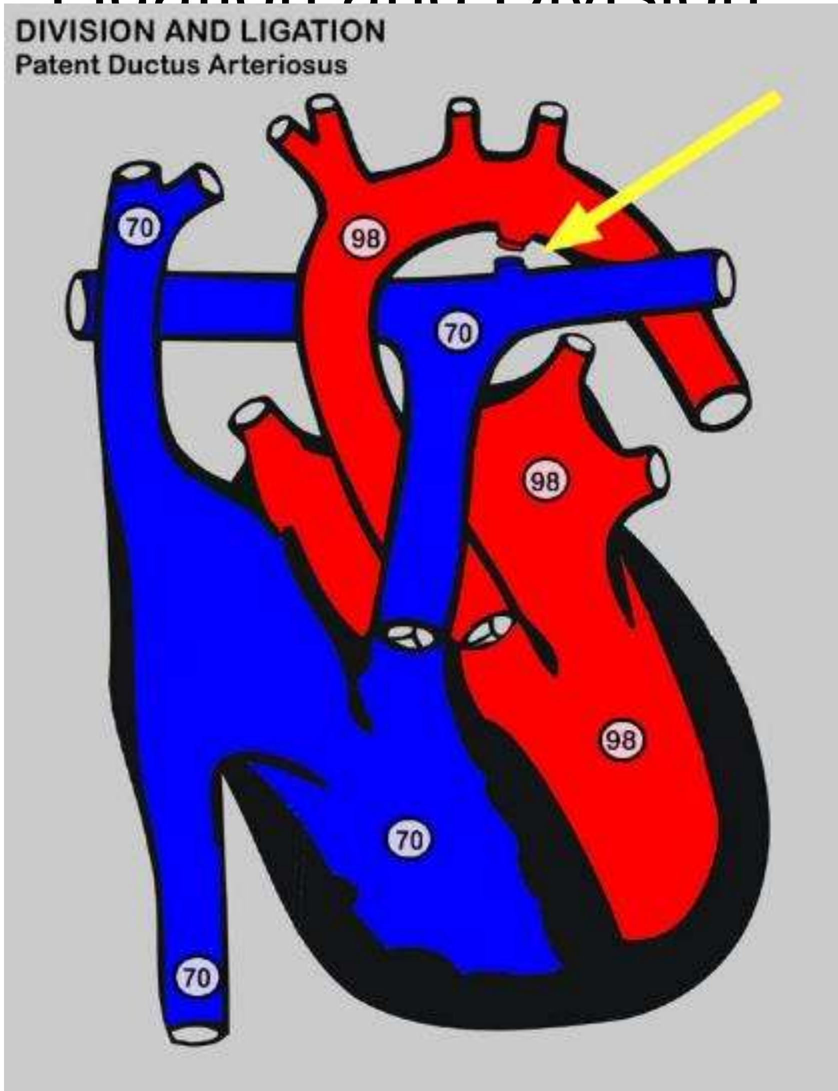
Doppler of the PDA (R-L shunt)

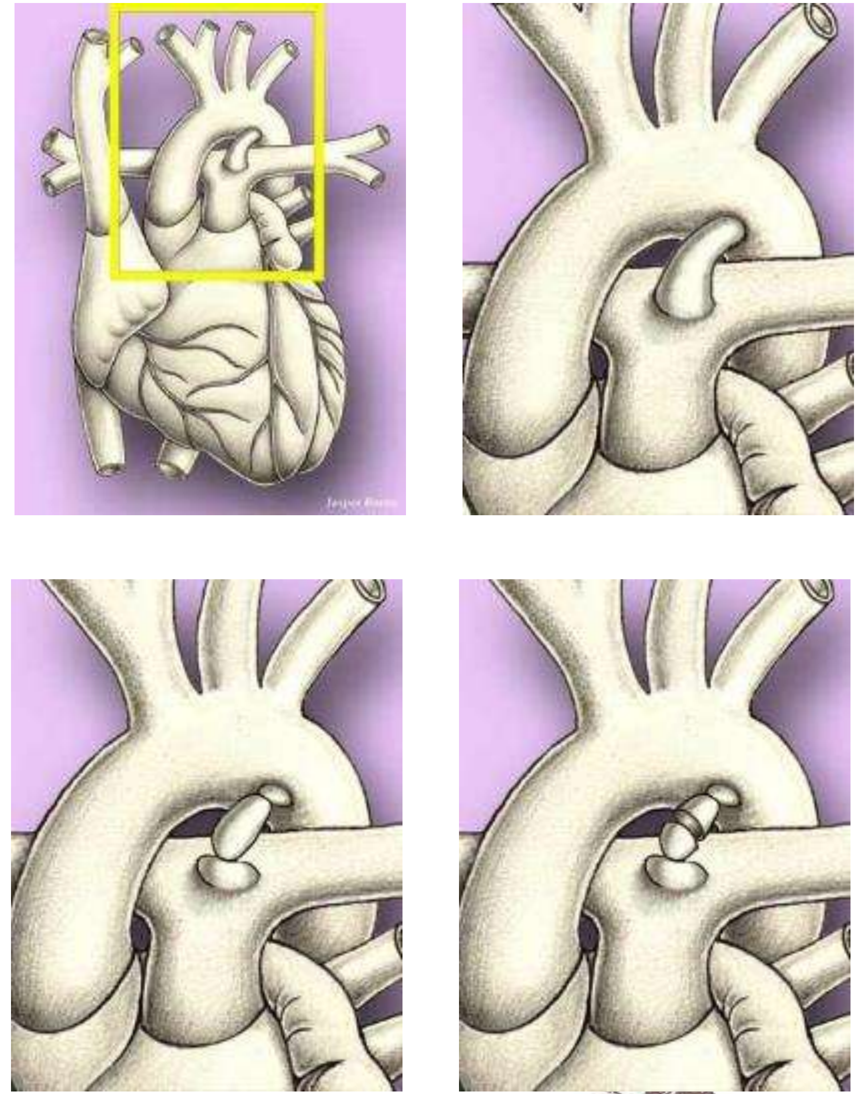




The Doppler spectral tracing shows evidence of severe pulmonary hypertension and no evidence of a L-R shunt through the PDA. The shunt is R-L from the ductus arteriosus to the Dao (blue: away from the TDX)

Patent Ductus Arteriosus – Ligation and Division





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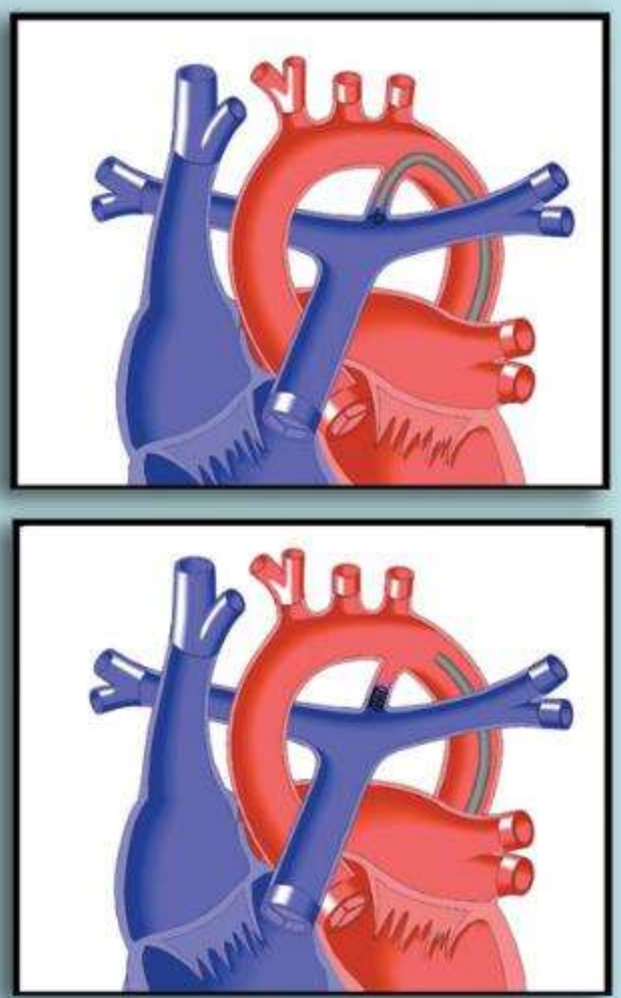
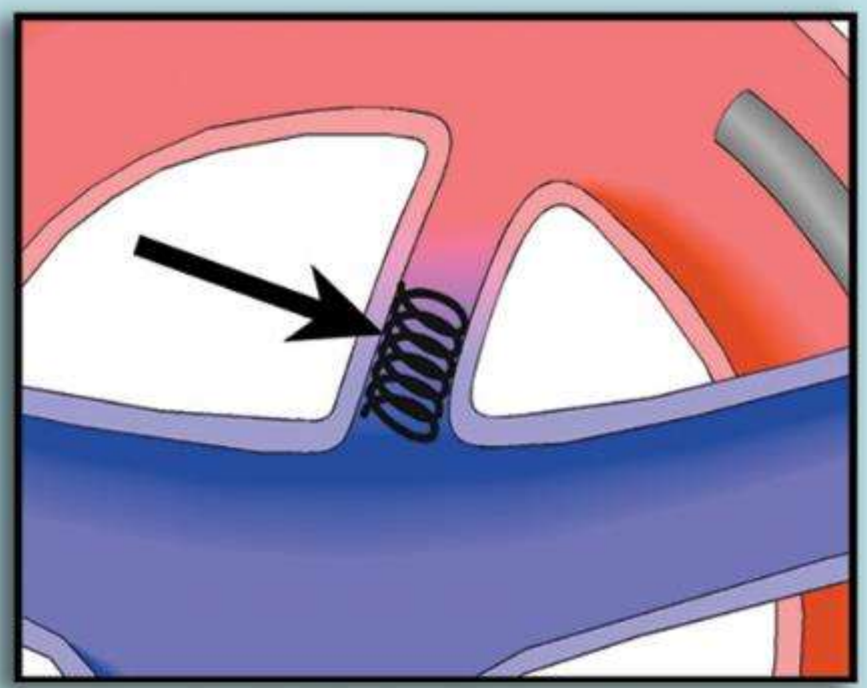
Occlusion of Intracardiac and Vascular Shunts

Coil embolization of PDA





Left, top: Catheter crosses the PDA from the aortic side and delivers a coil.

Left, bottom: Withdrawal of catheter, leaving coil in PDA

Amplatzer Ductal Occluders

Amplatzer ductal occluder

Illustration courtesy AGA Medical Group

Aorta angiogram with device occlusion of PDA, lateral view

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Right Heart Obstructive Lesions

Pulmonary Valve Stenosis

Valve anatomy

- Doming, fused commissures
- Thickened, immobile
- Subvalvar obstruction
- Supravalvar obstruction

Post stenotic dilation

RVH

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PS: Clinical Correlation

Asymptomatic

Murmur at birth

EKG

- RAD, RVH proportional to obstruction

Management

- Balloon dilation

Excellent outcome

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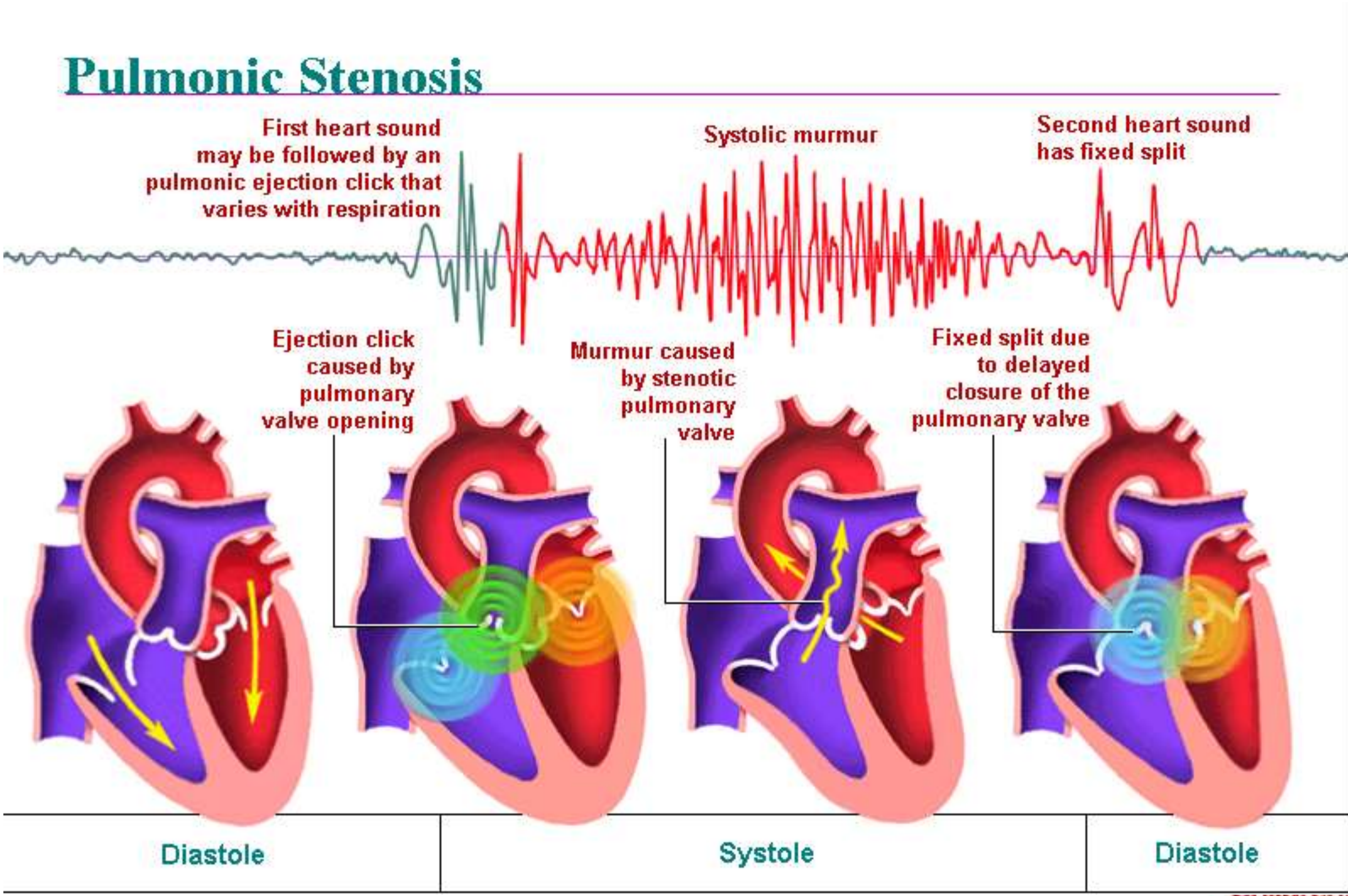

Pulmonic Stenosis
First heart sound may be followed by an pulmonic ejection click that varies with respiration
Systolic murmur
Second heart sound has fixed split
Ejection click caused by pulmonary valve opening
Murmur caused by stenotic pulmonary valve
Fixed split due to delayed closure of the pulmonary valve
Diastole
Systole
Diastole

Pulmonary Artery Branch Stenosis

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This adorable baby was just adopted from Russia. She has a history of a heart condition….

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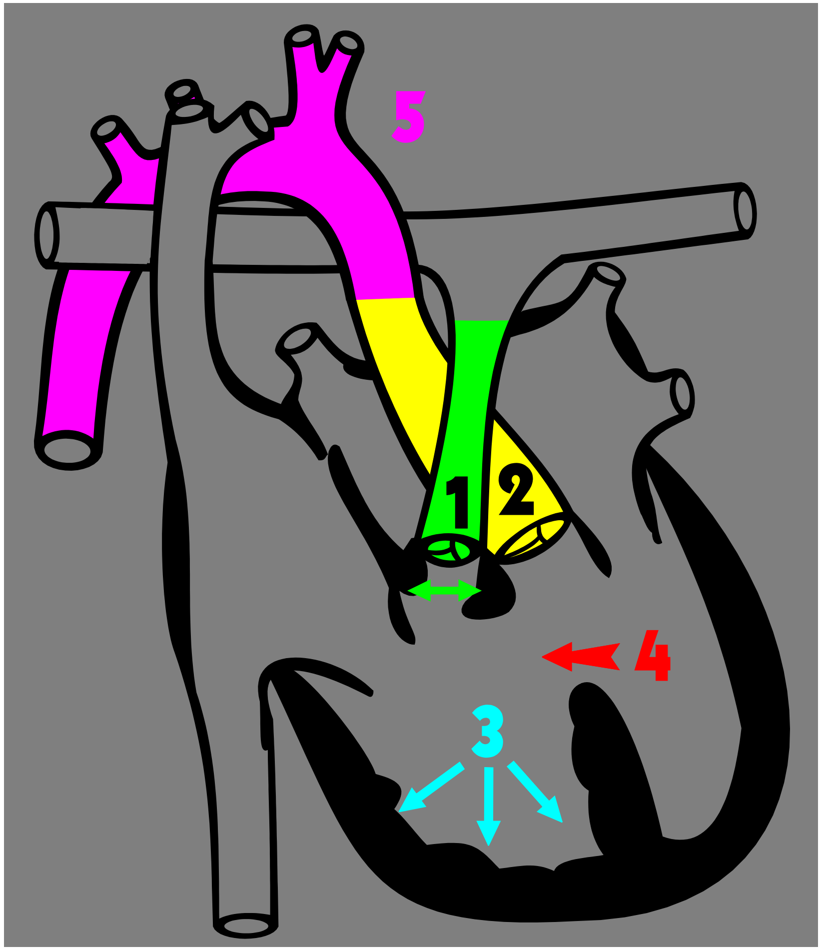
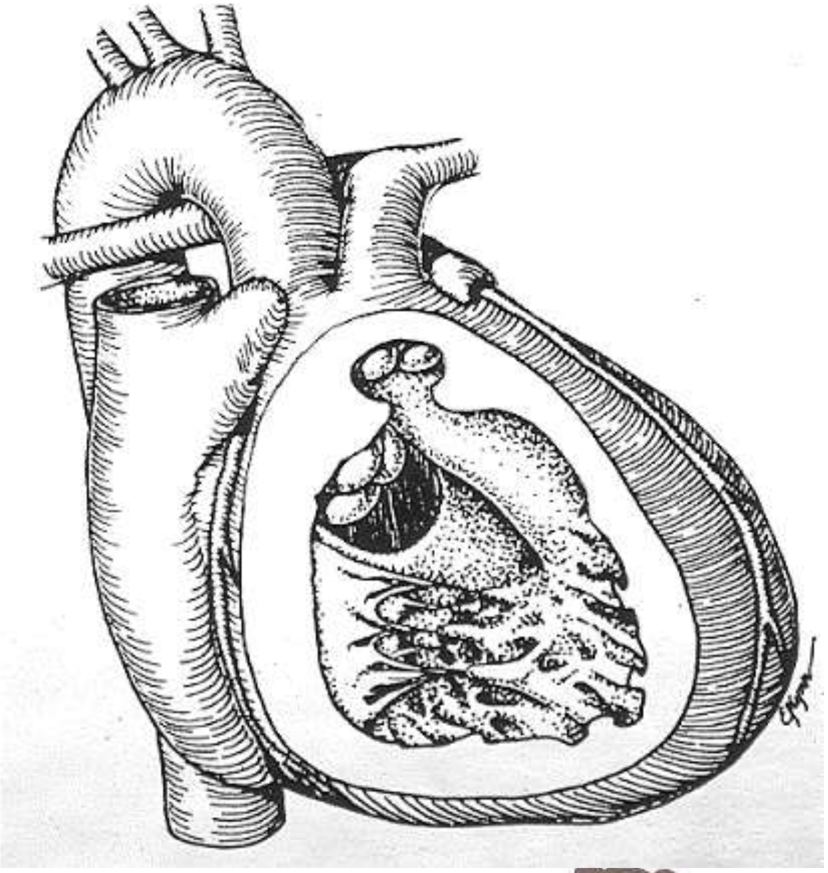
Tetralogy of Fallot : "Maladie Bleu" 1888





TOF: Clinical Correlation

Most common cyanotic defect

Defective neural crest migration resulting in abnormal conotruncal development

Clinical presentation depends on degree of subpulmonary narrowing

- This may change over time

Presentation

- Fetal dx
- Murmur

Variations in TOF

- "Mexican Tet"
  - Hypoplastic or absent conal septum
- Tetralogy with absent pulmonary valve
  - Rudimentary pulmonary valve leaflets result in fetal pulmonary regurgitation, PA dilation
  - Airway and lung development is compromised in severe cases
- Tetralogy with CAVC
- Tetrology with pulmonary atresia

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TOF: Clinical Correlation

- Cyanosis due to right to left shunting at ventricular level
- Degree of cyanosis is proportional to amount of right ventricular outflow tract obstruction
- Dynamic factors may worsen cyanosis
  - Tet Spell→ no murmur→ deeply cyanotic
- EKG
  - RVH, RAD, RAE
- CXR
  - Boot shaped heart

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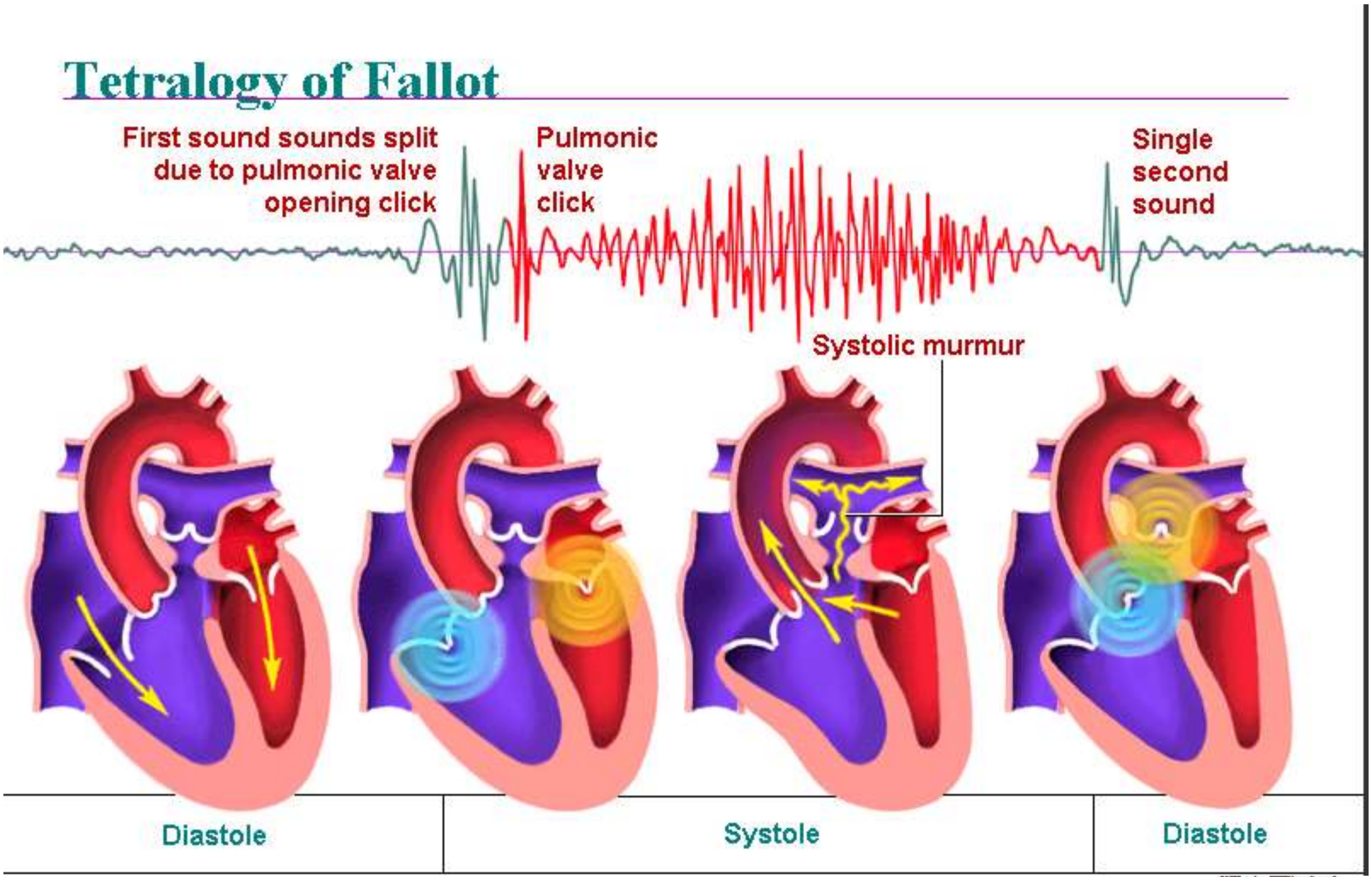
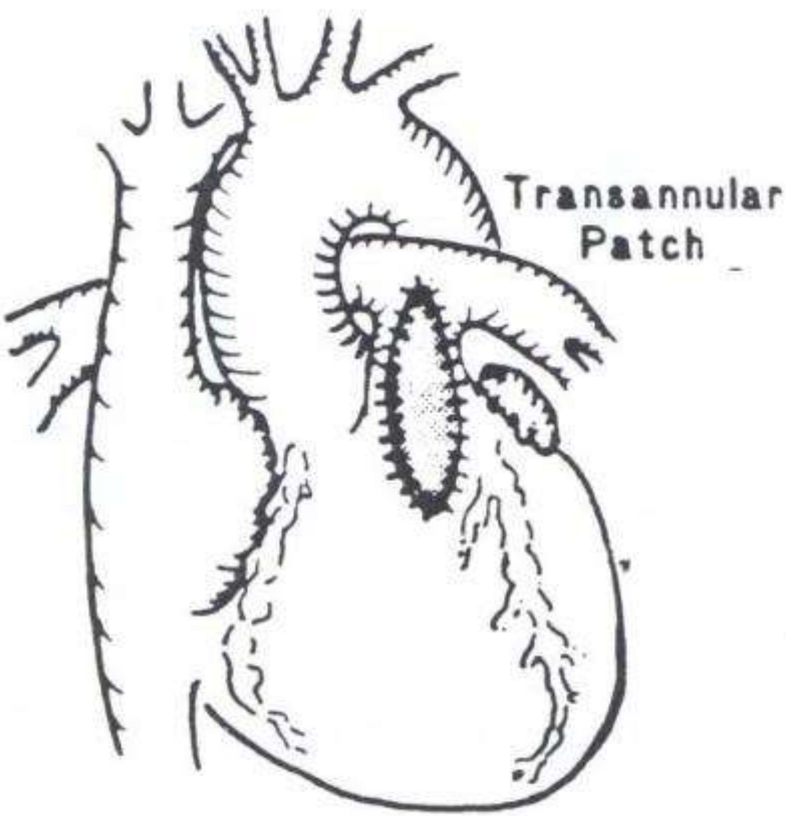
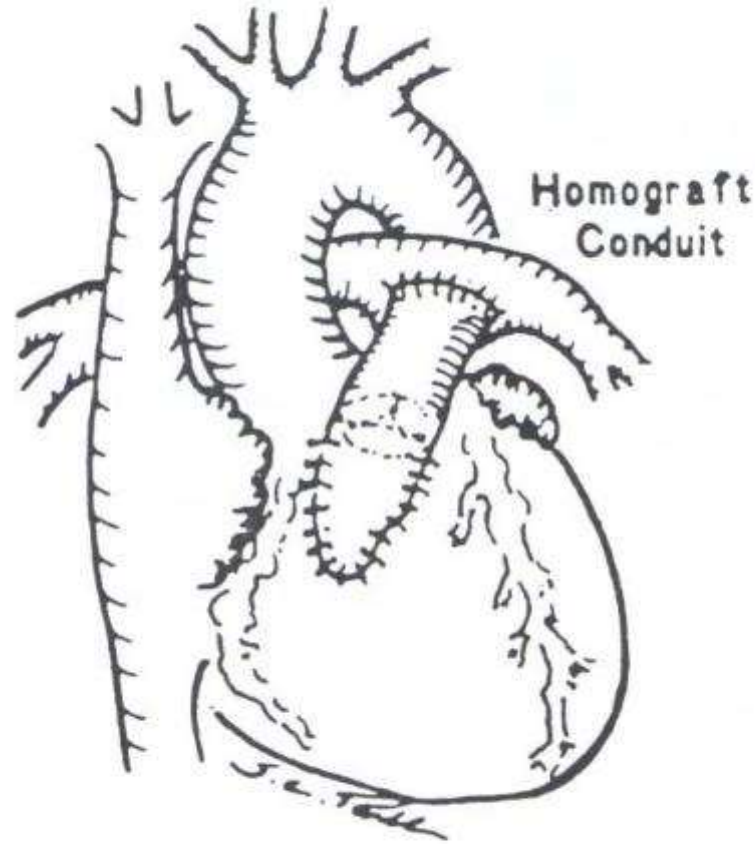

Tetralogy of Fallot
First sound sounds split due to pulmonic valve opening click
Pulmonic valve click
Single second sound
Systolic murmur
Diastole
Systole
Diastole

Tetralogy of Fallot





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Transcatheter Pulmonary Valve- 2010

- Catheter delivered prosthetic pulmonary valve
- Made from bovine jugular vein
- Sewn within a platinum-iridium ballon expandable stent
- For use in patients with a surgically placed conduit from the RV to the PA
- Used to treat significant conduit valve insufficiency and/or stenosis that would otherwise require surgical conduit replacement

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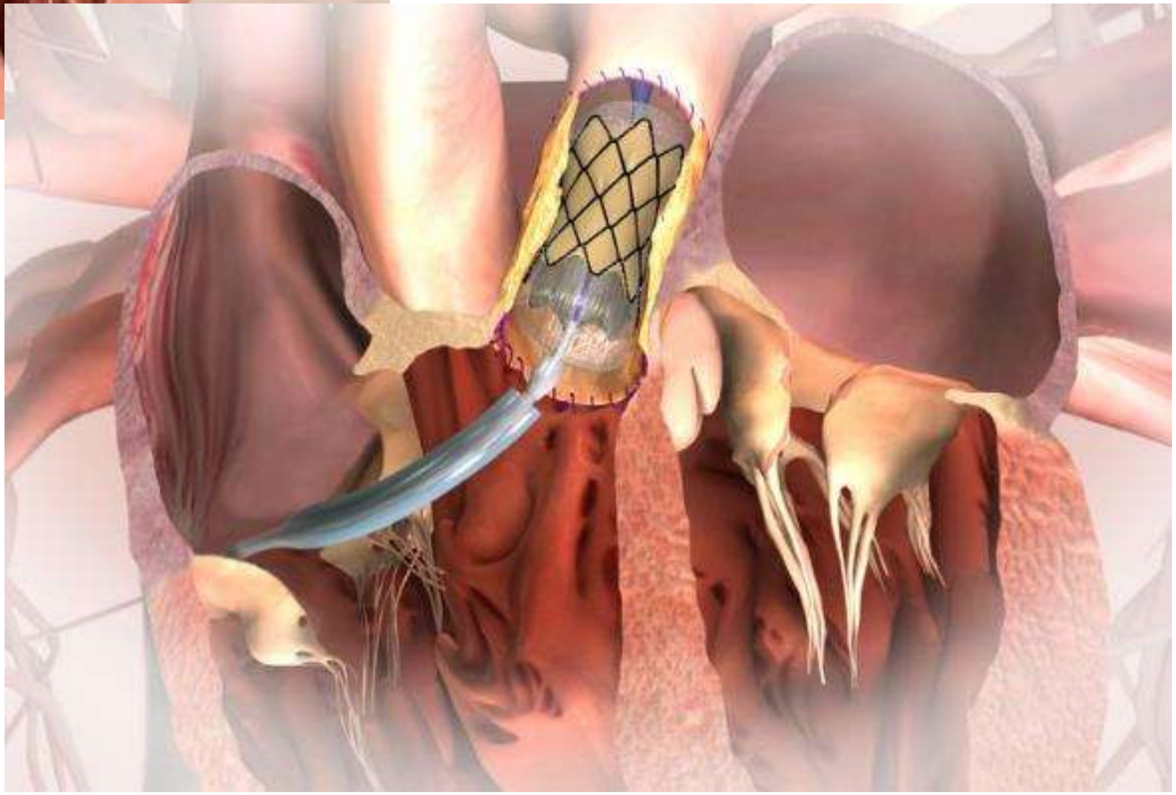

Double Outlet Right Ventricle (DORV)

- Describes a relationship where the PA and Aorta both arise from the anatomic RV
- “DORV” is normal during heart development
- Incidence 1 – 1.5% of patients with CHD
- 1 per 10,000 live births
- Possible association with trisomy 13 and trisomy 18
- Van Praagh – both great arteries arise from the morphologically RV
- NO mitral - aortic fibrous continuity
- Two functional ventricles in which a VSD provides the only outlet for one ventricle
- Anderson - 50% override rule – “if >50% of the aorta is over the RV, its DORV”

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Left Heart Obstruction

Aortic Stenosis

Valve, sub-valvar or supravalvar

Clinical manifestations

- Mild-moderate assymptomatic
- Severe
  - Depends on age of patient
- Management
  - Cath vs. surgery

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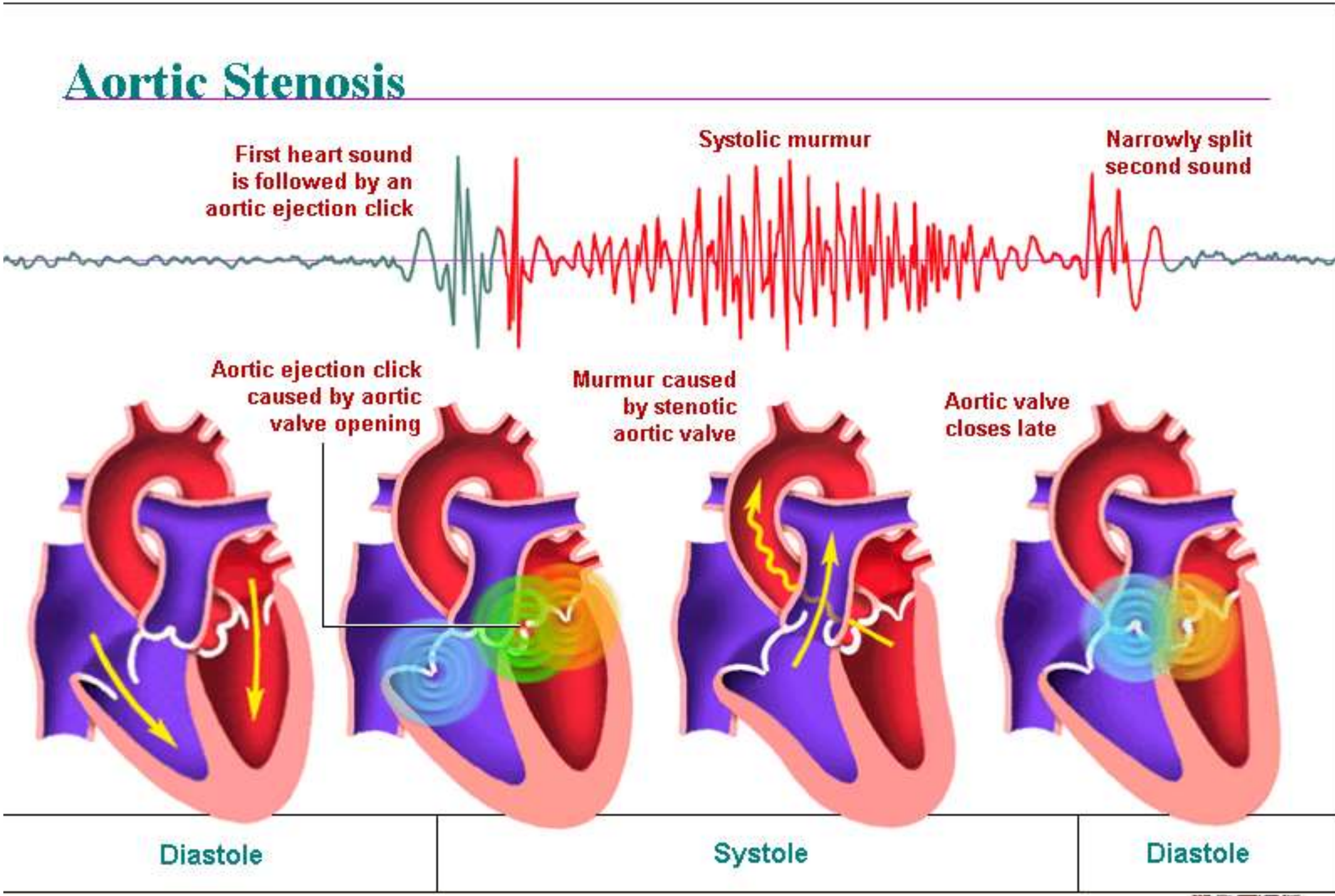

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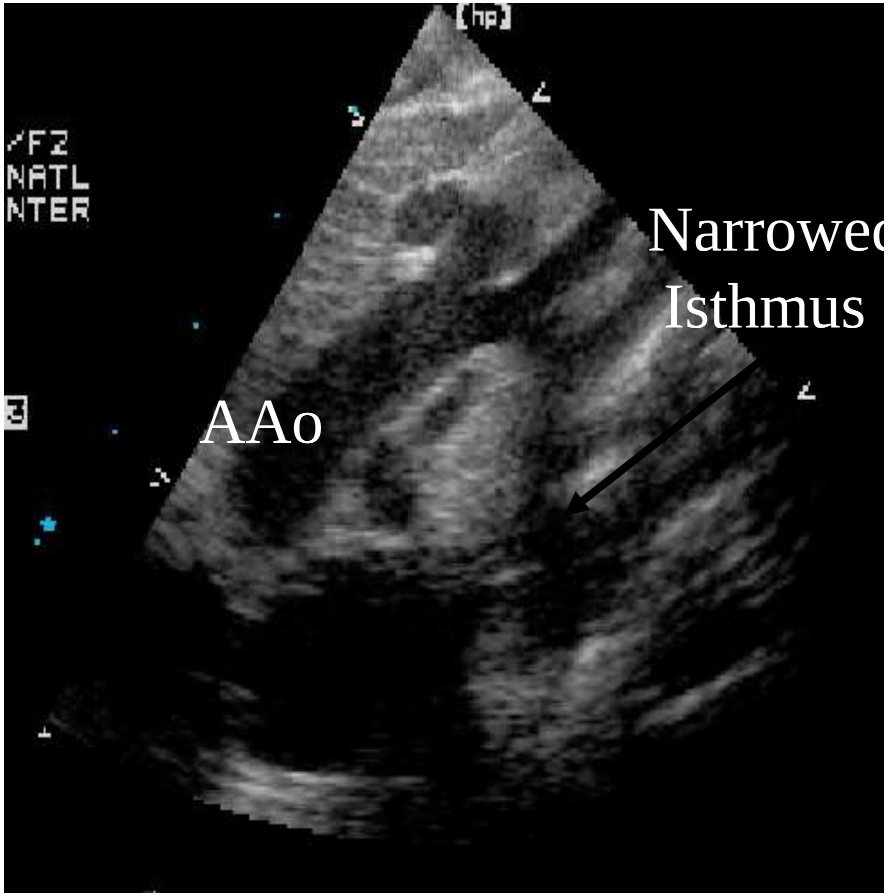
You are called to the emergency room to perform an echo on a baby that is listless and pale.

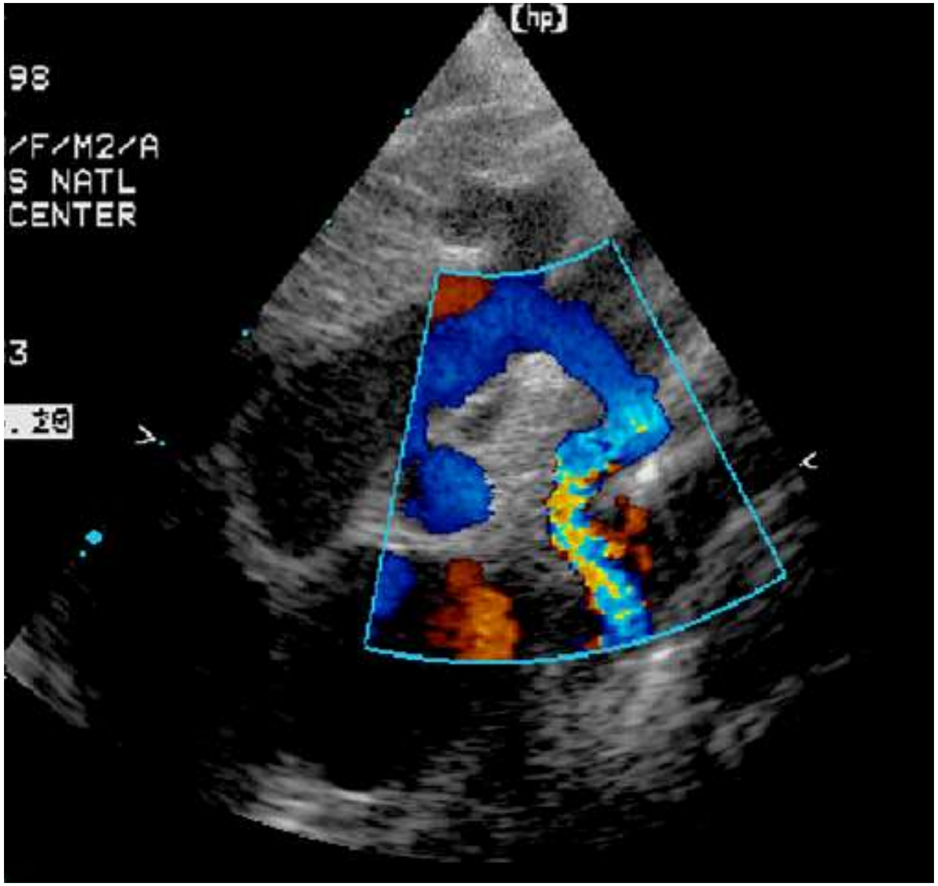
He has not been eating well over the last 24 hours

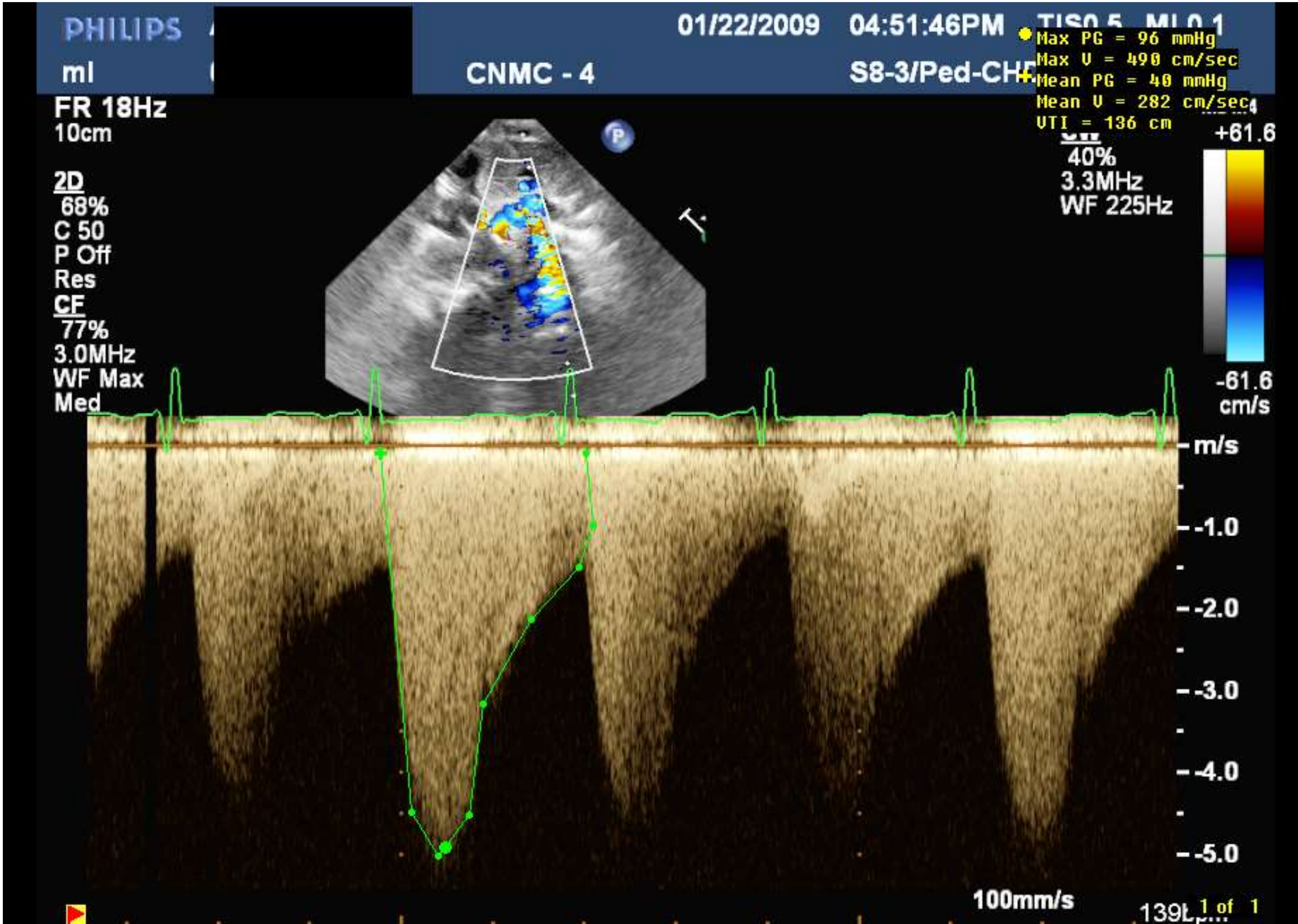
The ER doctor wants to know if they need to call cardiology….

You decide to start with parasternal imaging, you notice the LV function is very very bad....where should you image next?

Coarctation of the Aorta





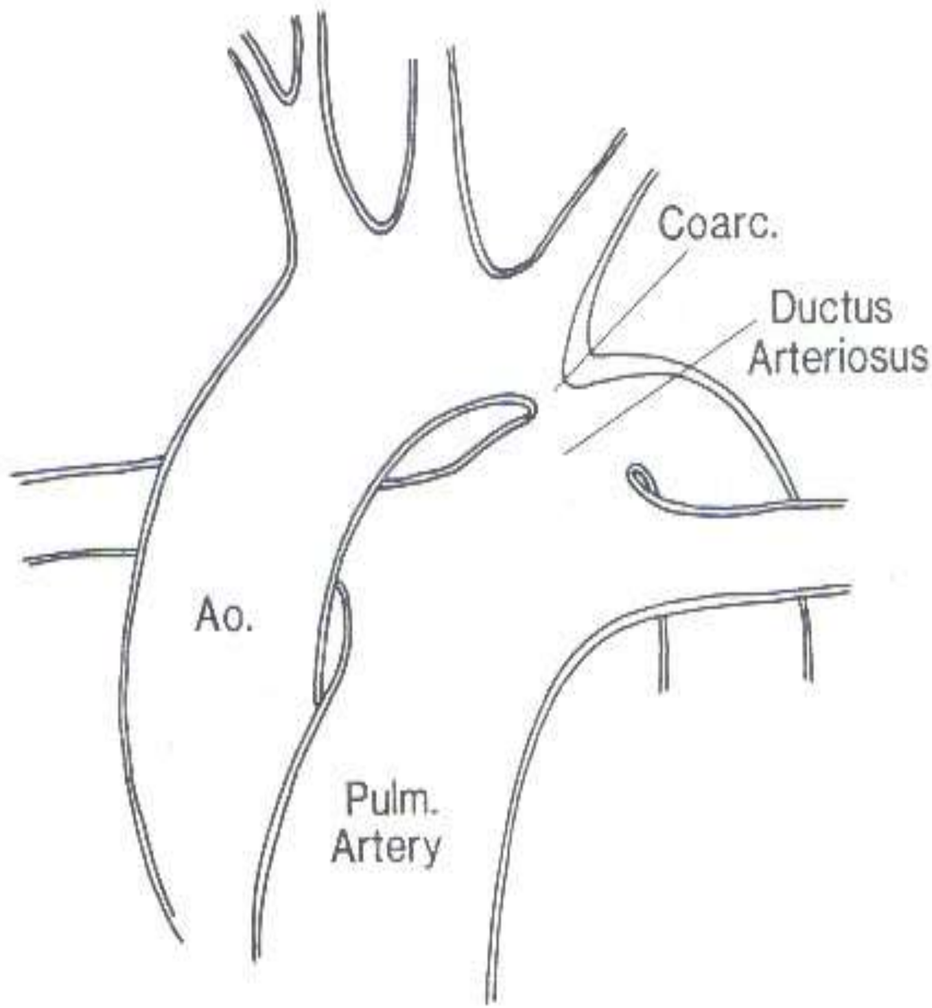

PHILIPS
ml
[redacted]
01/22/2009
04:51:46PM
CNMC - 4
Max PG = 96 mmHg
Max V = 490 cm/sec
Mean PG = 40 mmHg
Mean V = 282 cm/sec
VTI = 136 cm
FR 18Hz
10cm
2D
68%
C 50
P Off
Res
CF
77%
3.0MHz
WF Max
Med
40%
3.3MHz
WF 225Hz
+61.6
-61.6
cm/s
m/s
-1.0
-2.0
-3.0
-4.0
-5.0
100mm/s
1 of 1

Coarctation of the Aorta

Aberrant ductal tissue within the wall of the aorta

All coarcts are “juxtaductal”

Must look for other left heart Disease (aortic & mitral valve)



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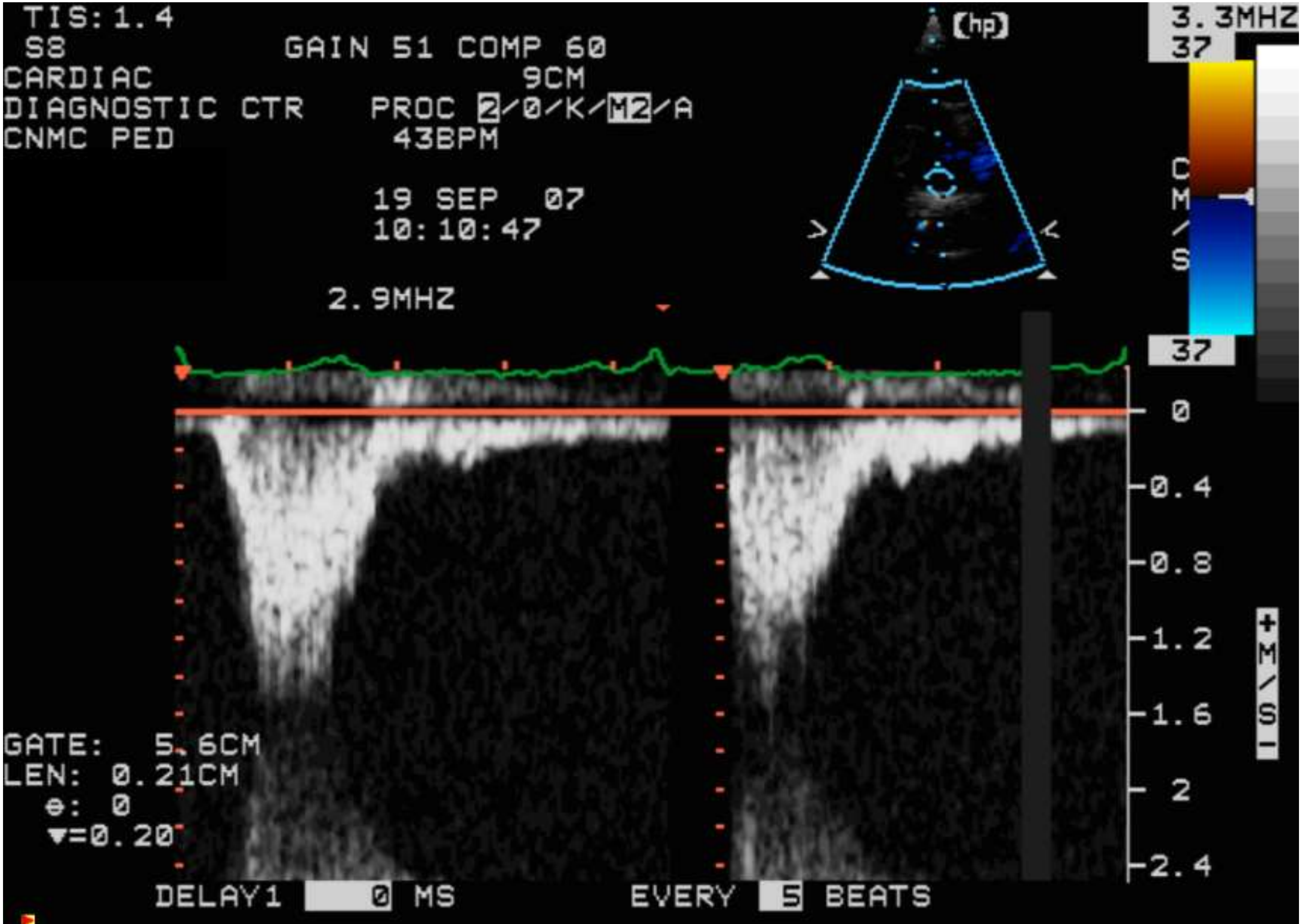
The next day you are staffing a Children's clinic and the nurse tells you the blood pressure in the legs of the next patient are the same as the arms.

The doctor is busy and asks that you perform an echo while she finishes the previous patient…



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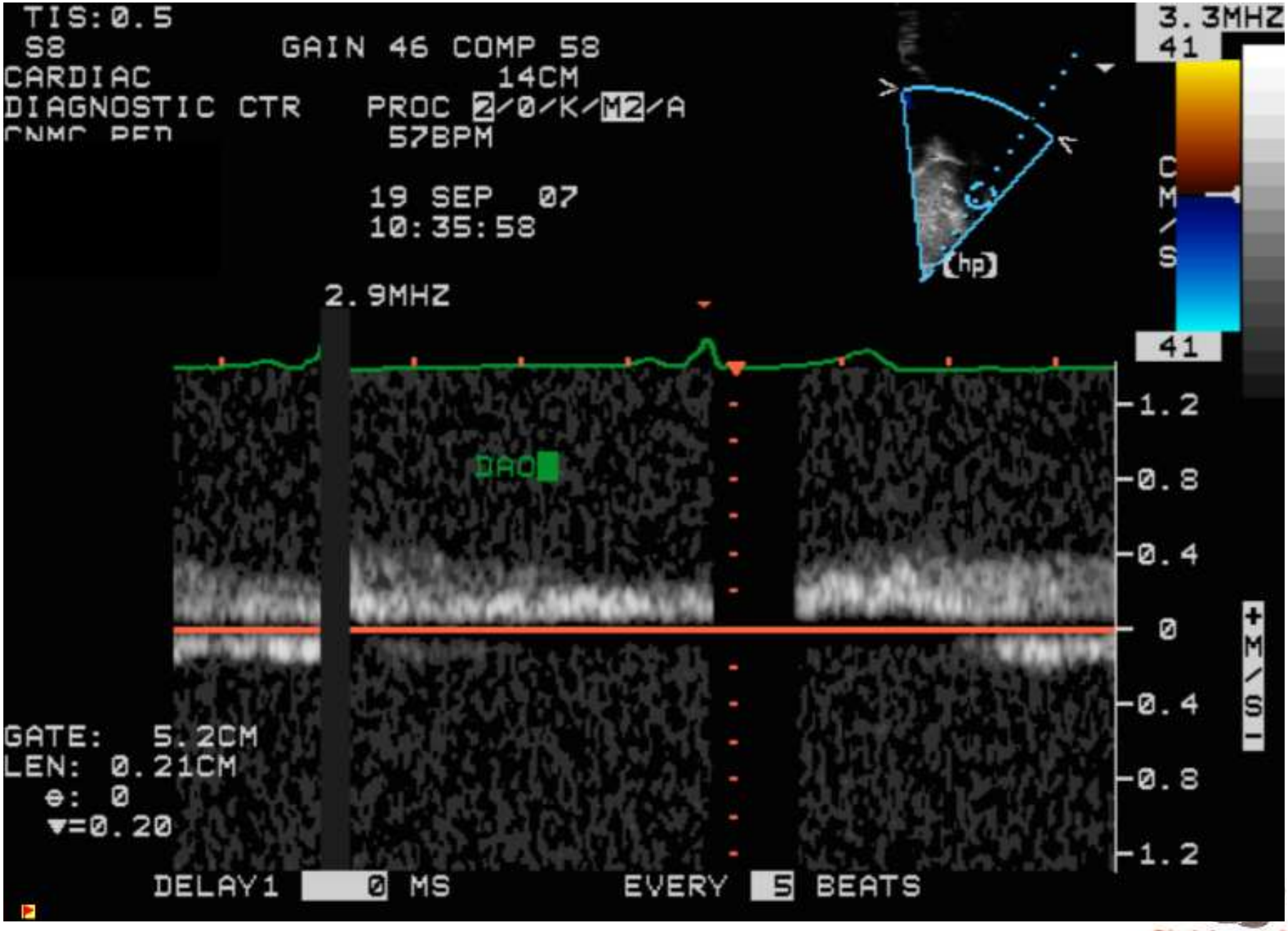
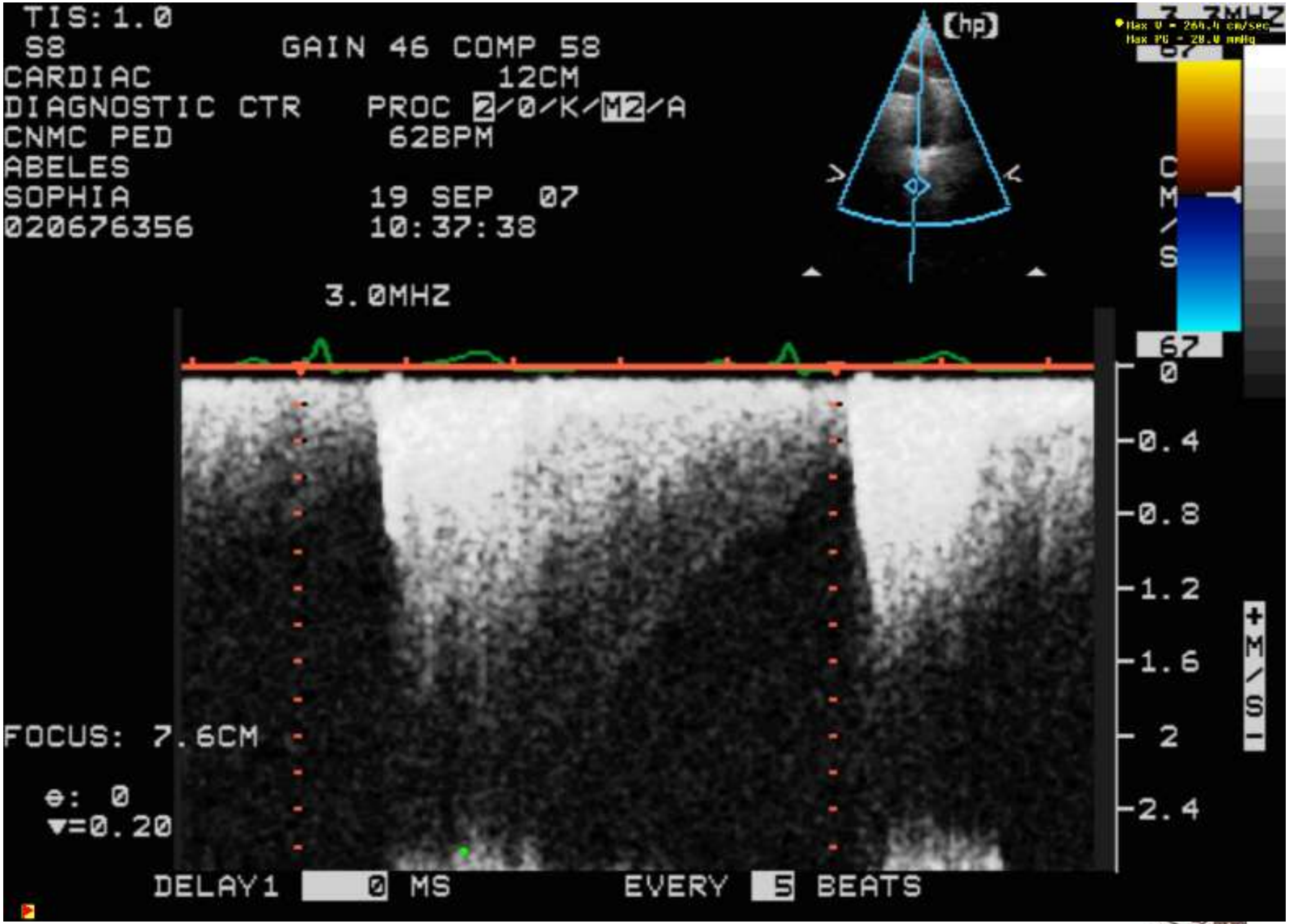
Descending AO Doppler

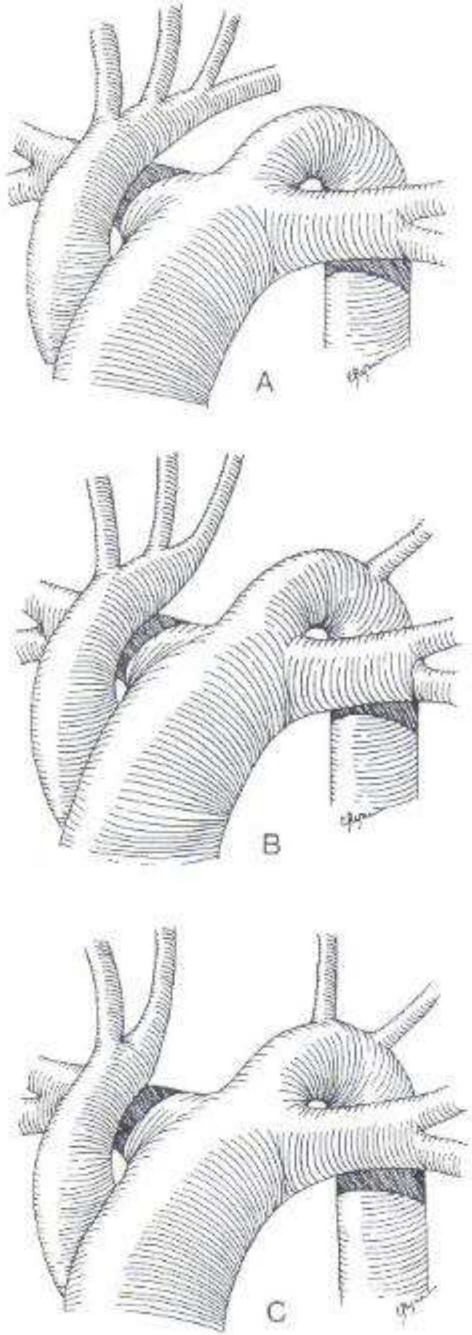


Doppler "drag"



Interrupted Aortic Arch

-Type A = After the subclavian artery,
probably an extreme form of
coarctation with obliteration of the lume
-Type B = Between the LCC and LSCA,
most common, defect of arch
remodeling/neural crest
-Type C = Between the Carotid arteries,
most rare



Complex Lesions

The nurse from the nursery calls you frantic, there is a baby that is blue.

He was born earlier today, he seemed ok, his birth weight was 8 pounds, uncomplicated pregnancy and delivery…

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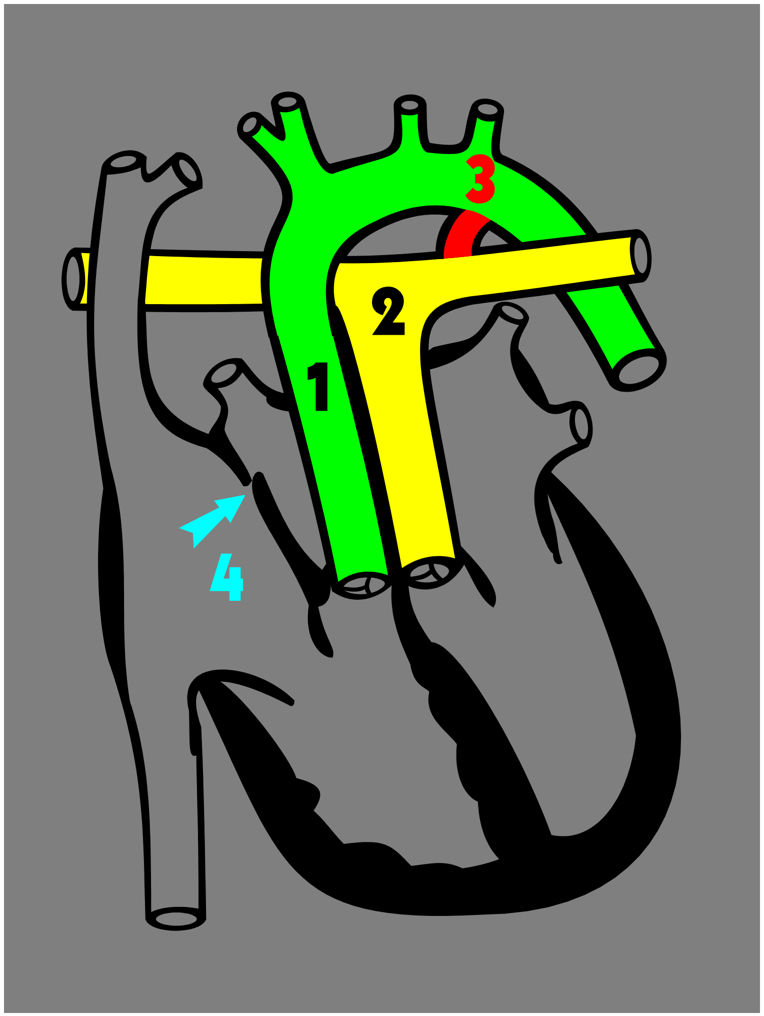
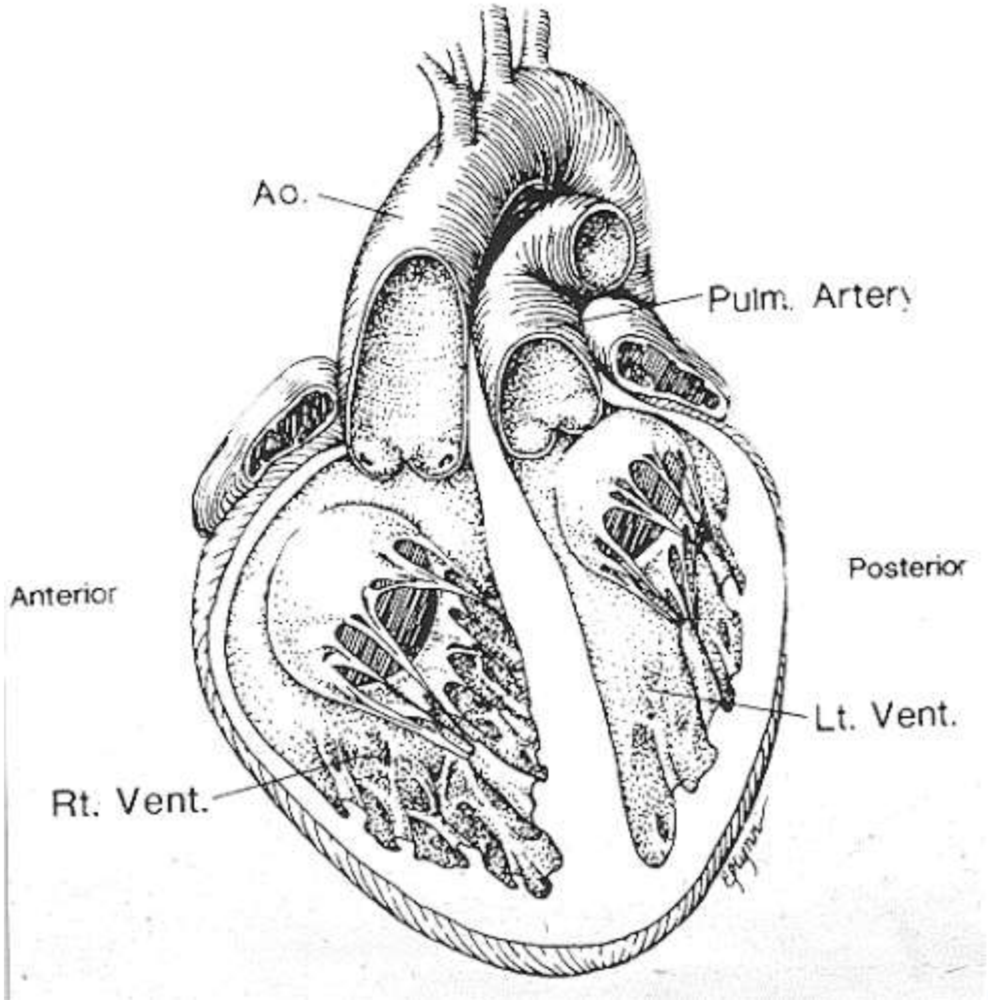
D-Transposition of the Great Arteries



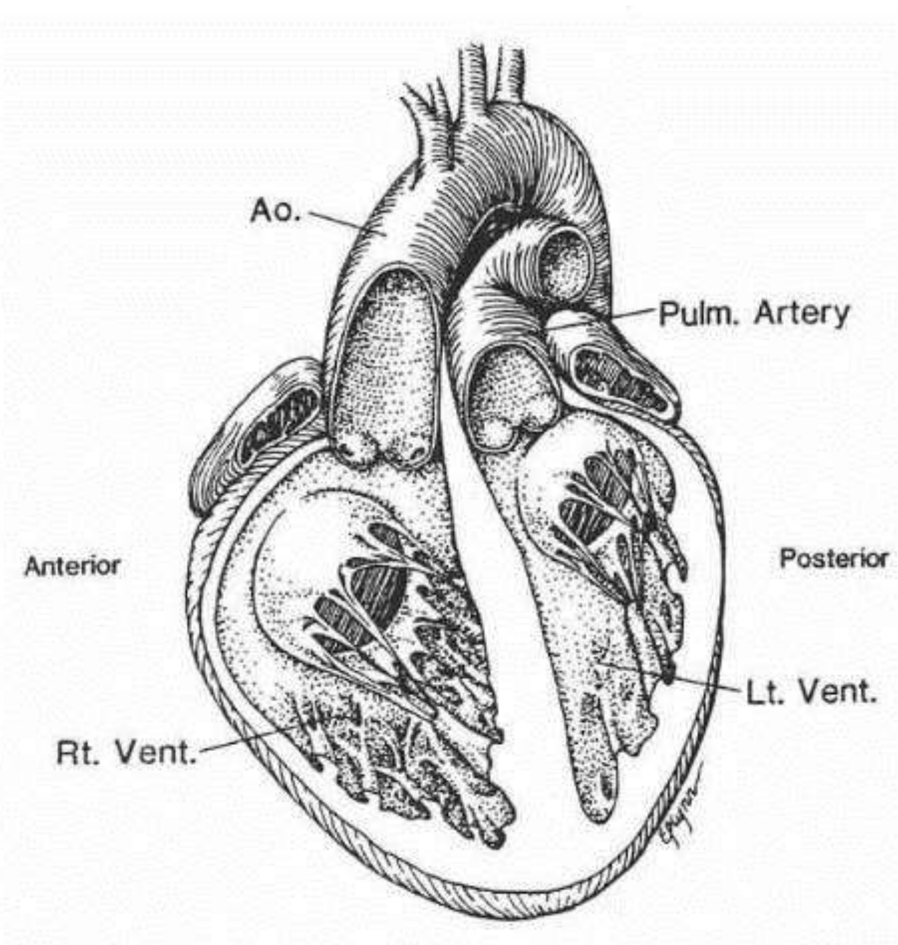


D-TGA

First described by Baillie 1797
Natural history: >90% mortality in infancy
Incidence: ~5% of congenital heart disease
Rare association with syndromes or other anomalies
Male:Female = 2:1
Possible association with infant of diabetic mother

D-TGA

- Ventriculo-arterial discordan
- Circulation in parallel
- RA=>RV=>Ao
- LA=>LV=>PA
- Must have mixing at atrial or survive



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D-Transposition
Balloon Septostomy

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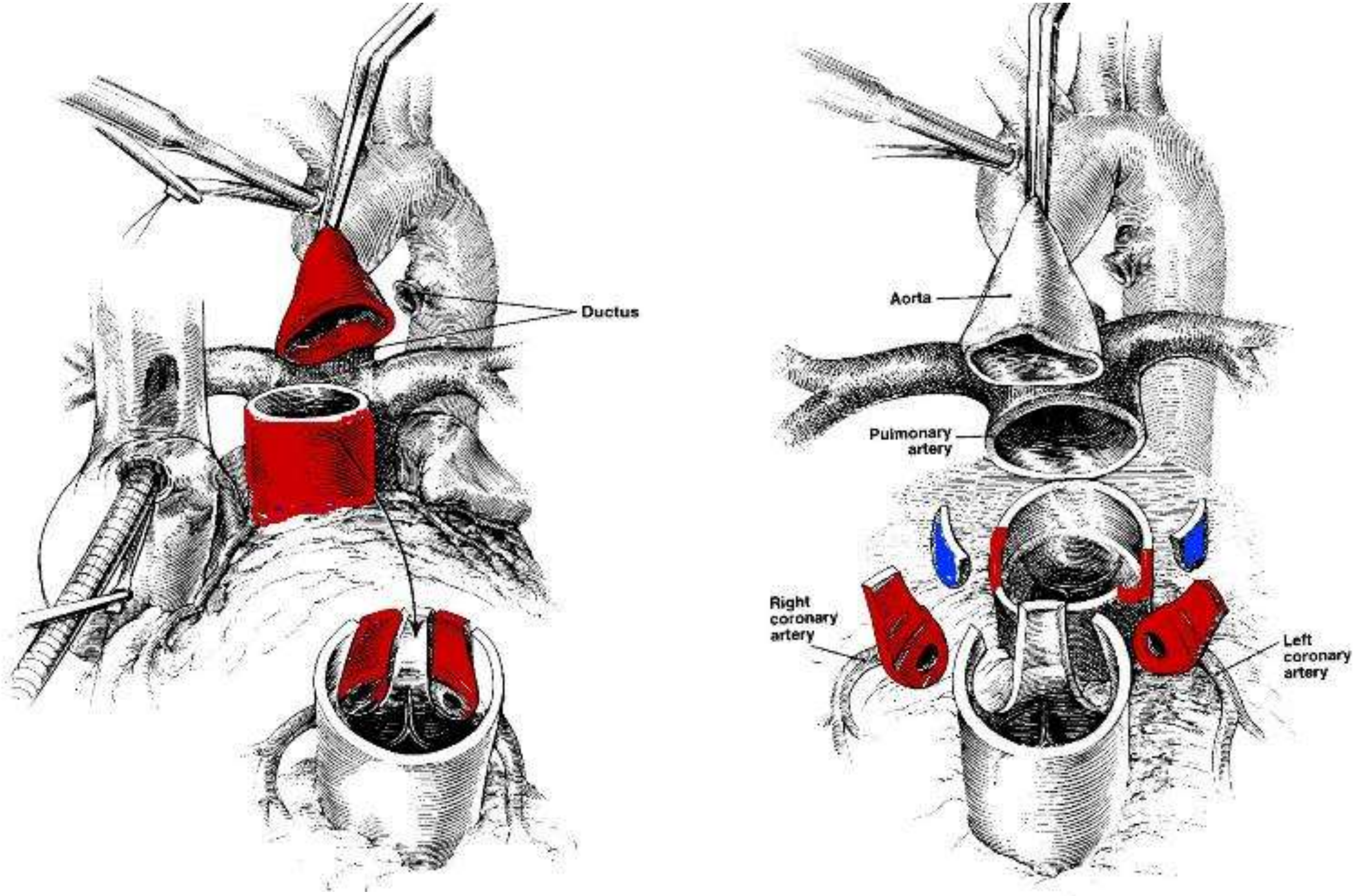
Arterial Switch Procedure



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Long Term Postoperative Concerns Arterial Switch Operation

Neo-pulmonary stenosis

Coronary abnormalities

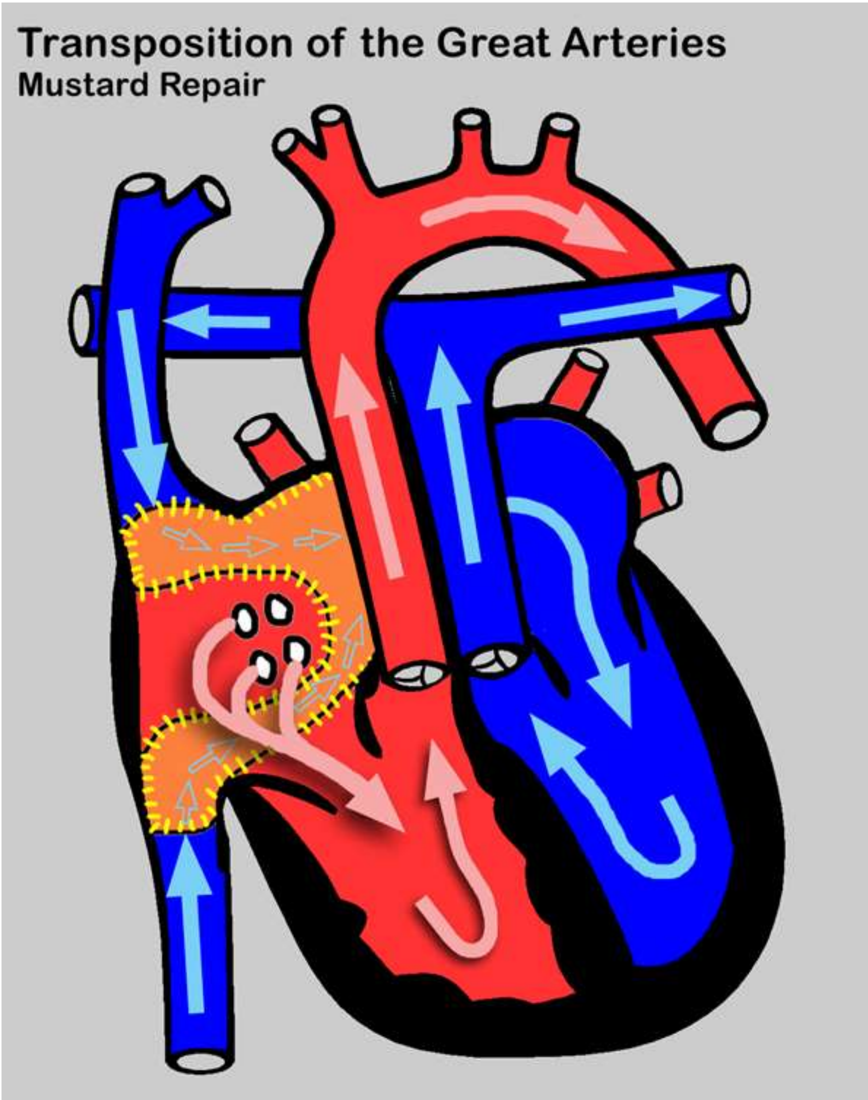
- Obstruction and stenosis
- Decreased flow reserve

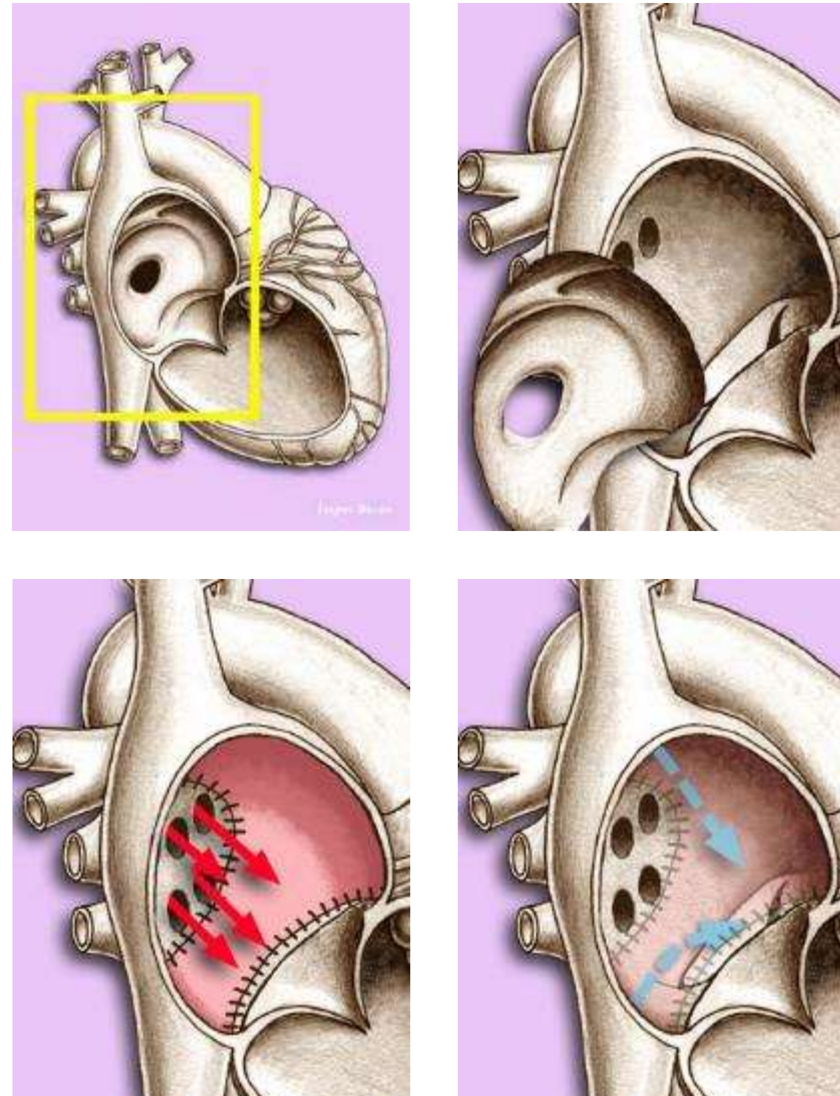
Neo-aortic insufficiency

- Almost always trivial/mild

LV function

Mustard Repair





Atrial Baffle Repair
Long Term Sequelae

On going late mortality risk

- 20% mortality at 20 years

Arrhythmia

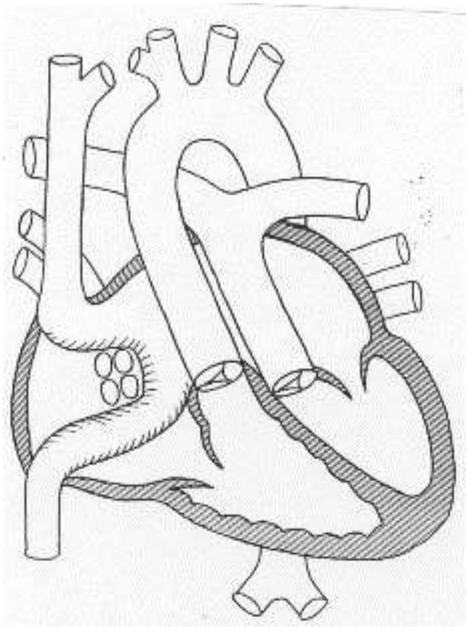
SVC obstruction -- 14-17%

IVC obstruction -- 1%

Baffle Leak -- Significant 1-2%

Systemic AV valve regurgitation -- 30%

Systemic Ventricular Failure -- 15-20%



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Transposition of the Great Arteries – L Type

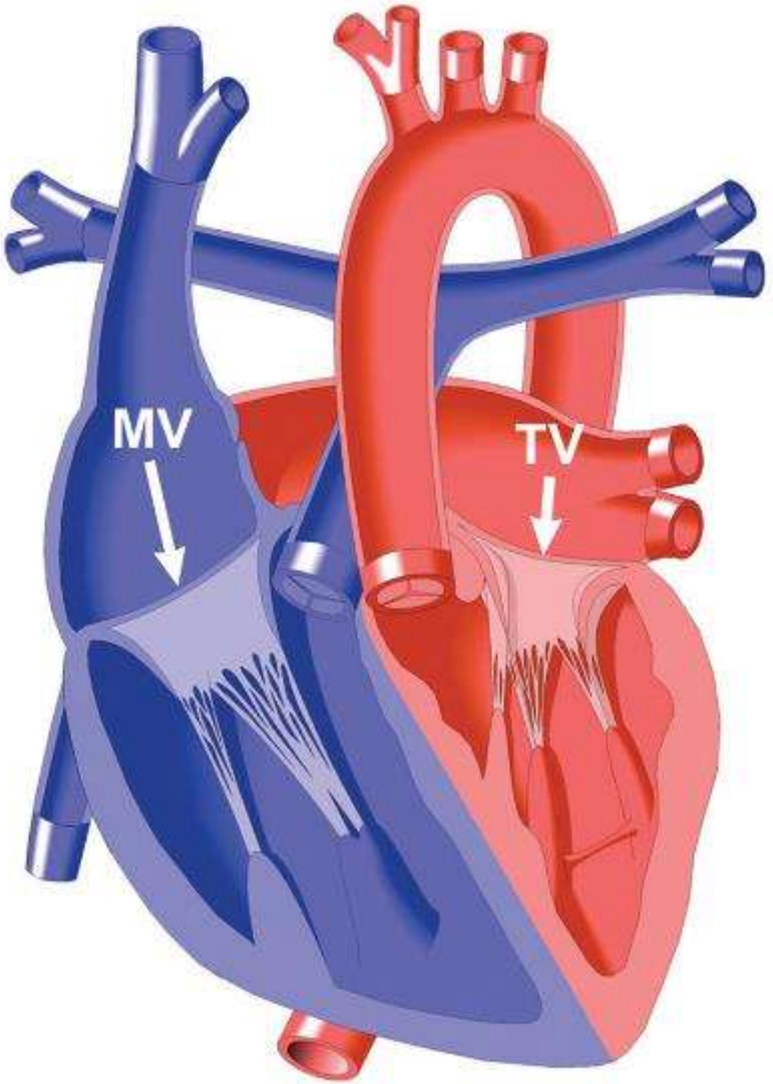
Congenitally Corrected Transposition"

Atrio-ventricular and ventriculo-arterial discordance ("double discordance")

RA ⇒ LV ⇒ PA

LA ⇒ RV ⇒ Ao

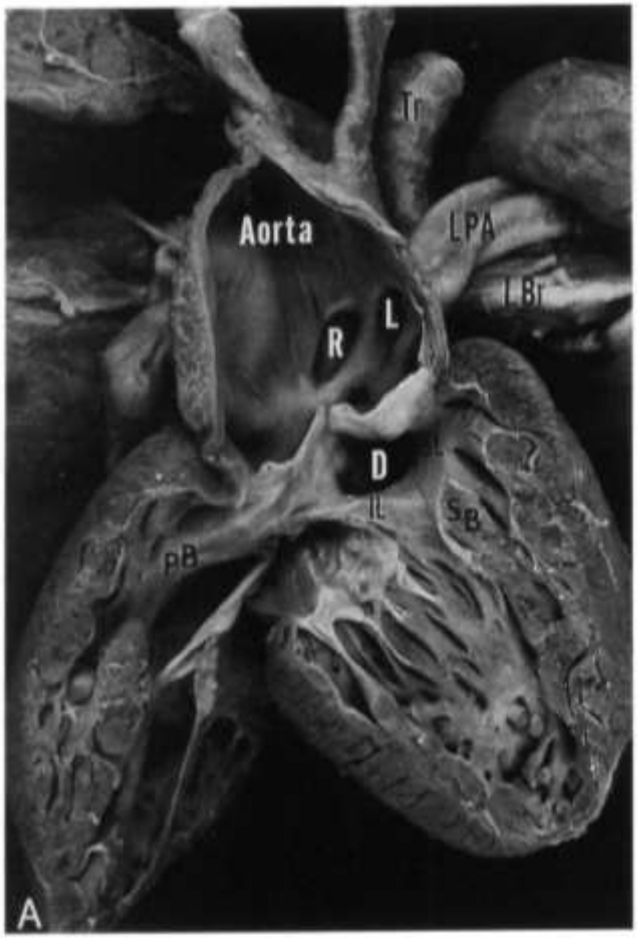
May be an isolated, asymptomatic finding or may be associated with other heart malformations



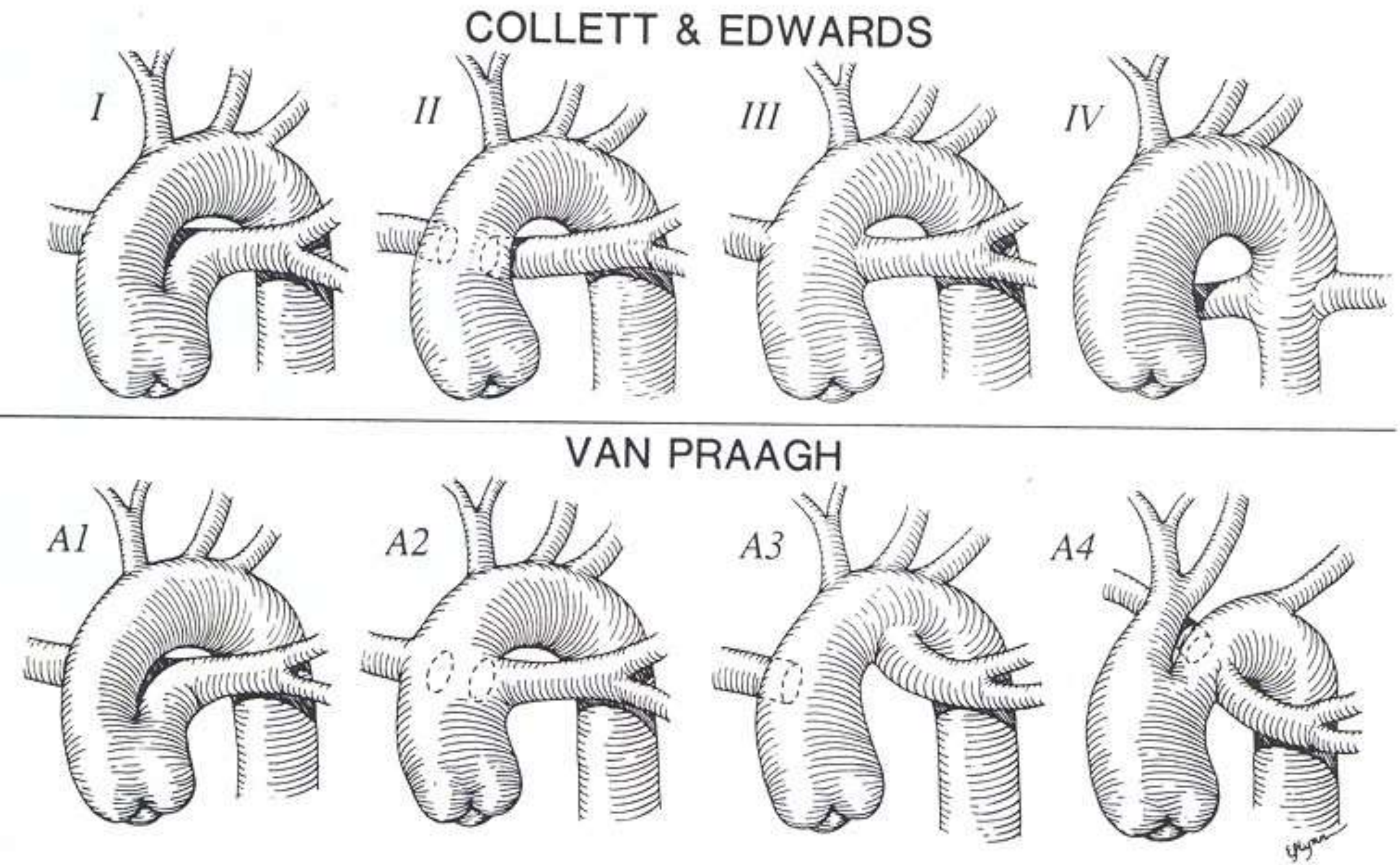
Truncus Arteriosus

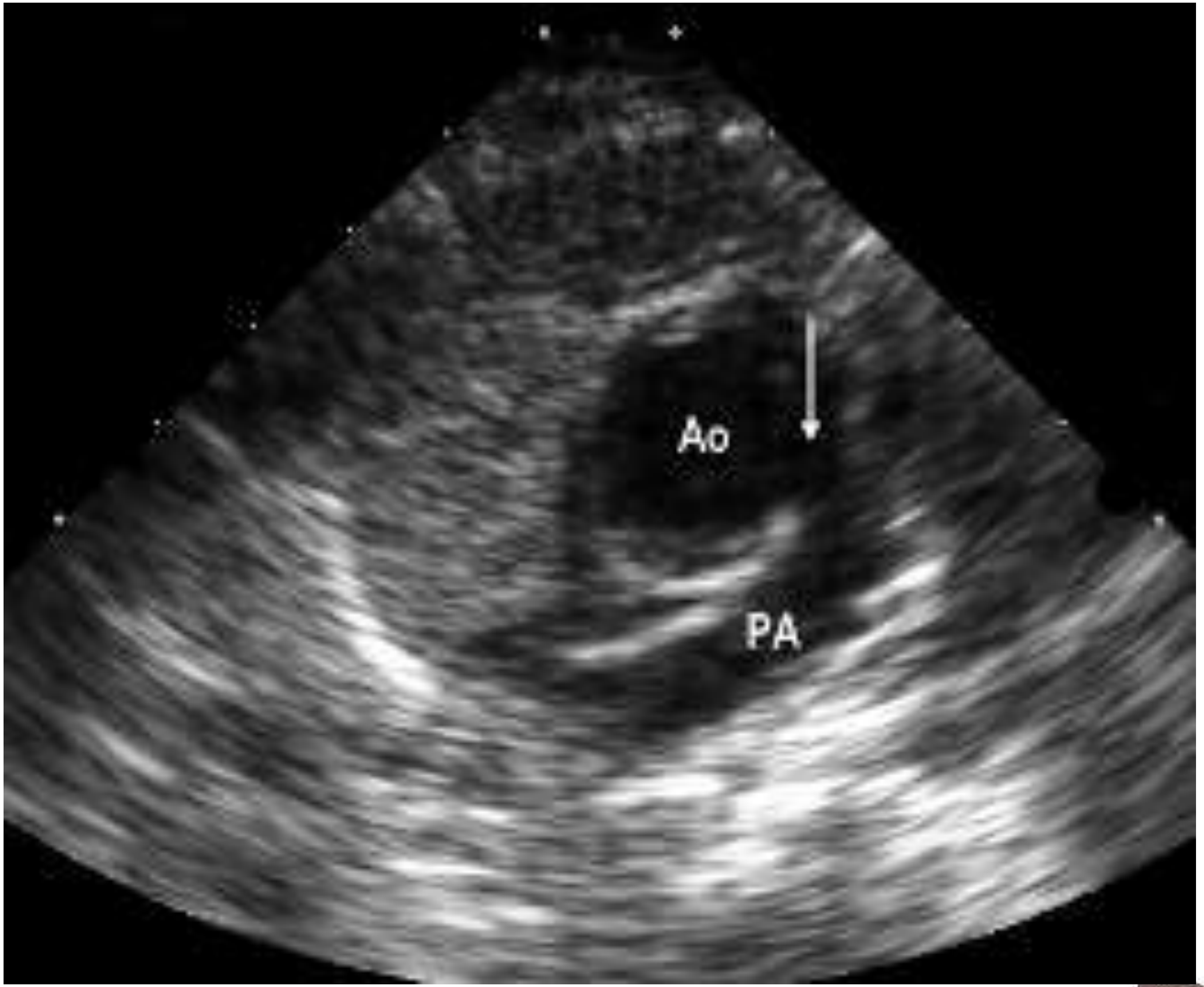
A single vessel arising from the heart and giving rise to the coronary, pulmonary and systemic circulations

The VSD is the same as TOF



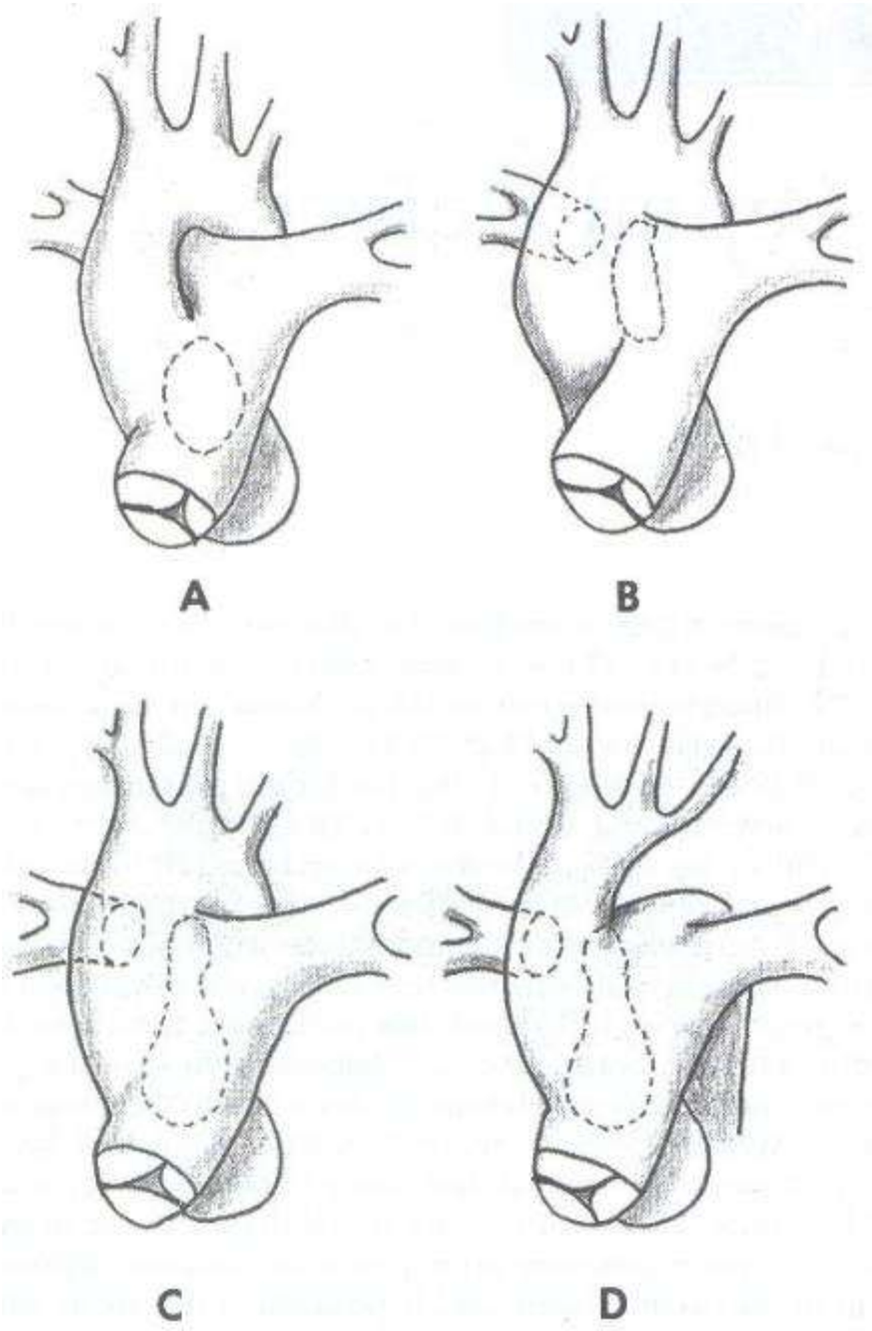
Truncus Arteriosus



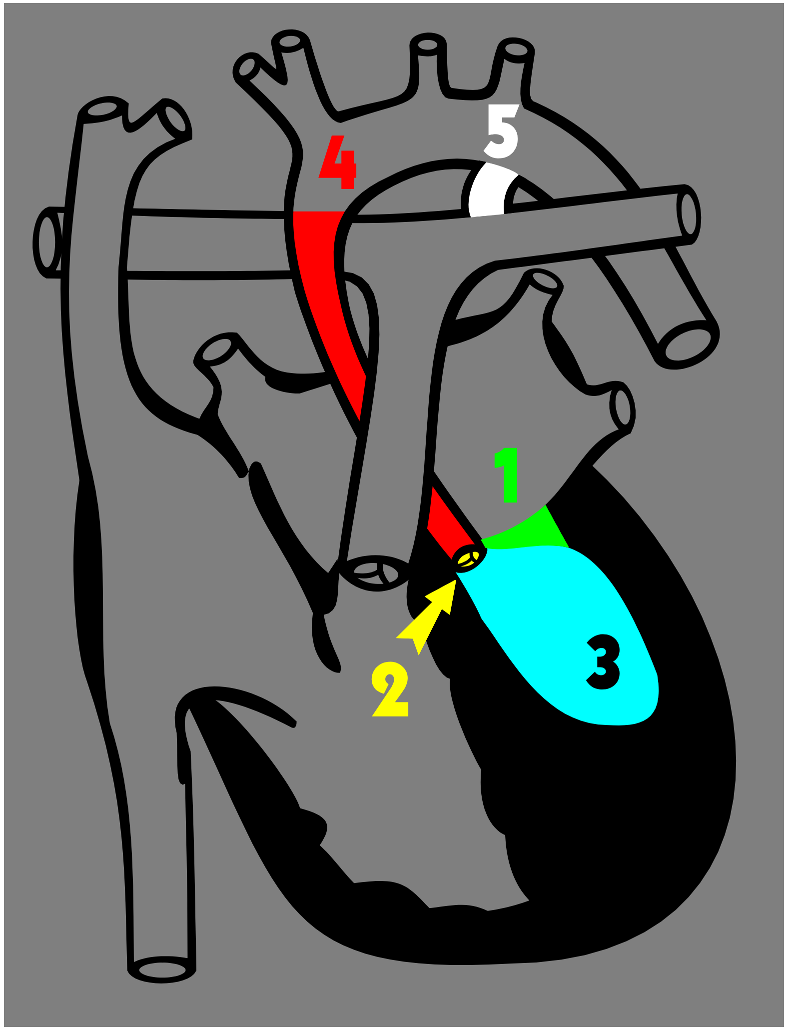

Ao
PA

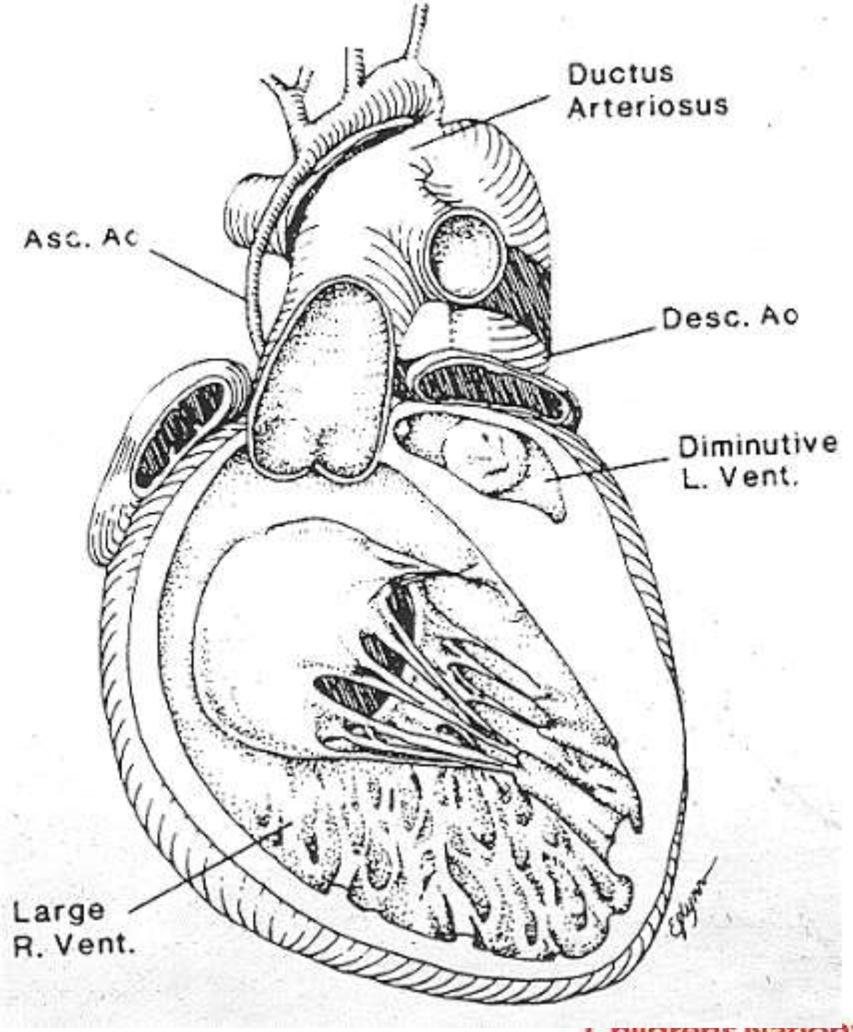
AP Window

Communication between aorta and PA

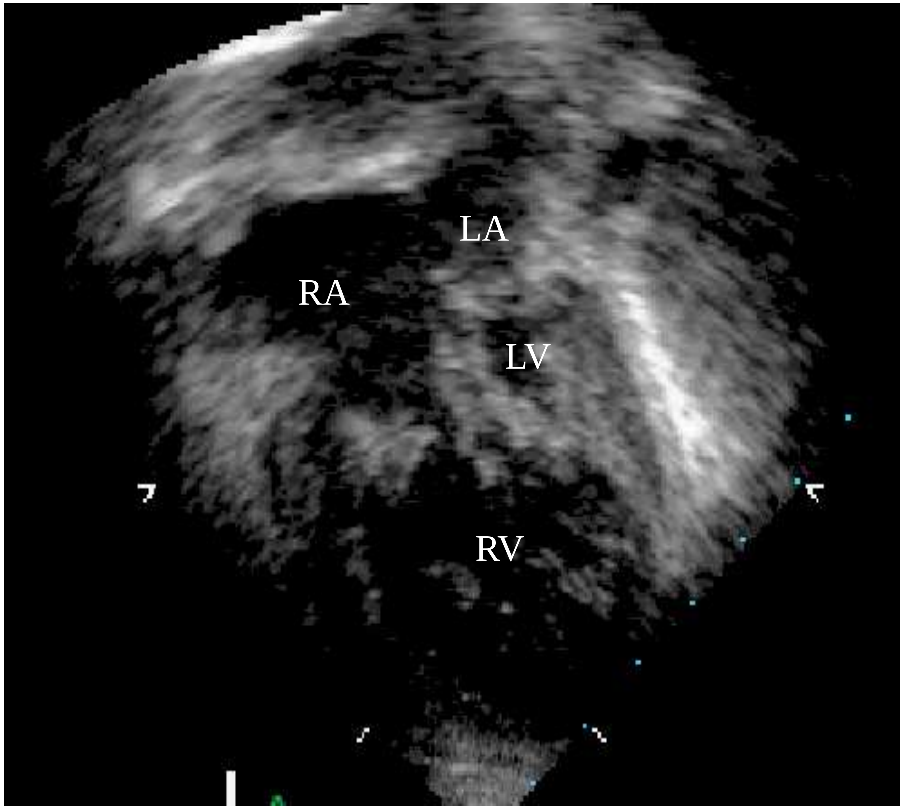


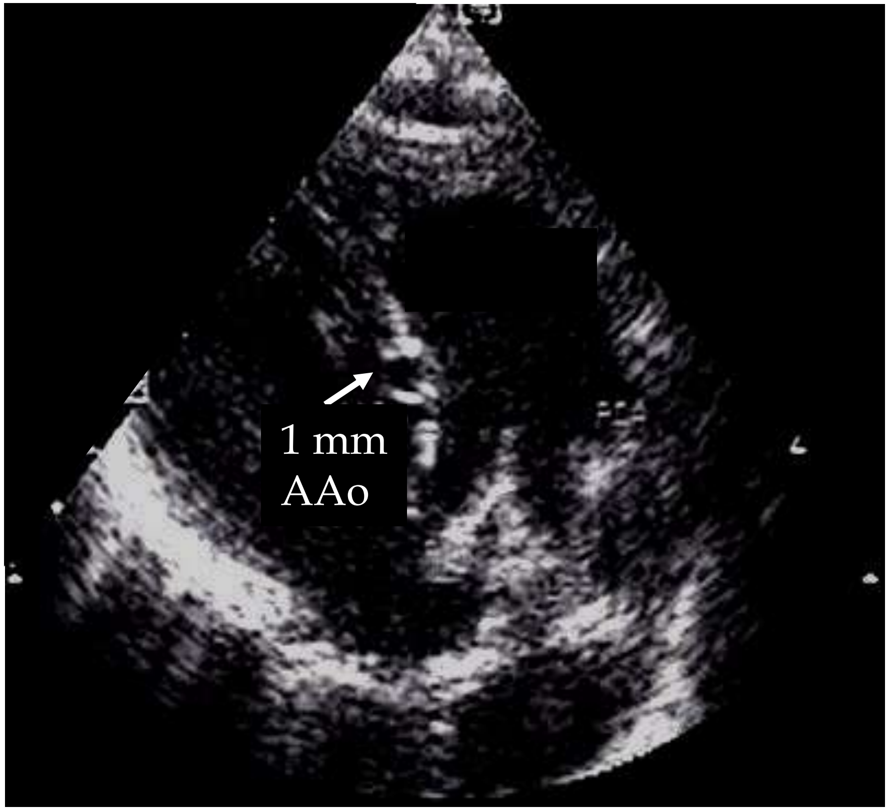
Hypoplastic Left Heart Syndrome





Hypoplastic Left Heart Syndrome





BT Shunt: History

1924: Failing to obtain a surgical residency at Hopkins, Alfred Blalock goes to Vanderbilt and begins research on traumatic shock

1930: Vivien Thomas hired as Alfred Blalock's lab assistant

1938: Rabbit models with subclavian to PA anastomosis fail to produce pulmonary HTN

1941: Blalock and Thomas move to Hopkins

1941: Coarctation relief with subclavian to descending aorta shunt

1943: Helen Taussig, a Hopkins pediatrics residency graduate, approaches Blalock about help for "blue babies"

1944: "Anna," a dog with a surgically created mixing lesion, successfully undergoes end-to-side subclavian-to-PA anastomosis, lives 15 years

November 29, 1944: Eileen Saxon, a 15-month-old 4.5 kg undergoes successful systemic-to-pulmonary shunt by Blalock with Thomas directly over his shoulder

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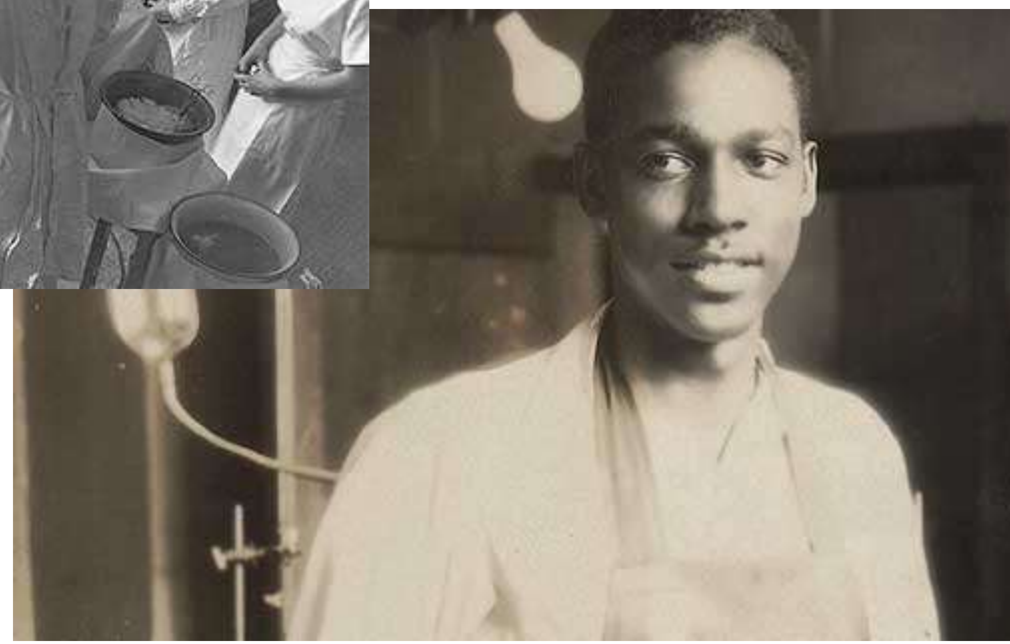

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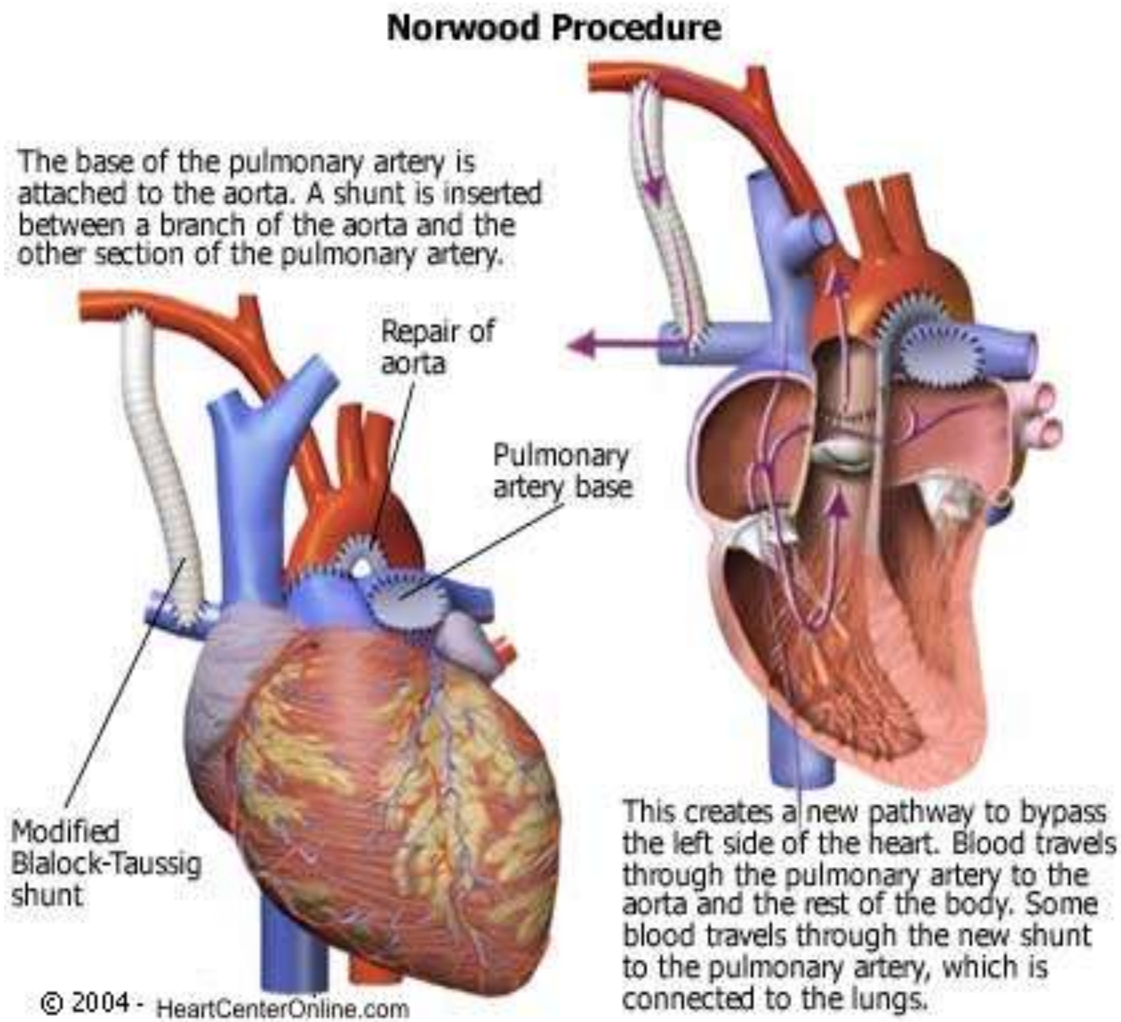
Norwood I: Anatomy

1. Atrial septectomy

2. Ligation of main pulmonary artery and construction of neo-aorta

3. Sano Modification/ Modified BT Shunt



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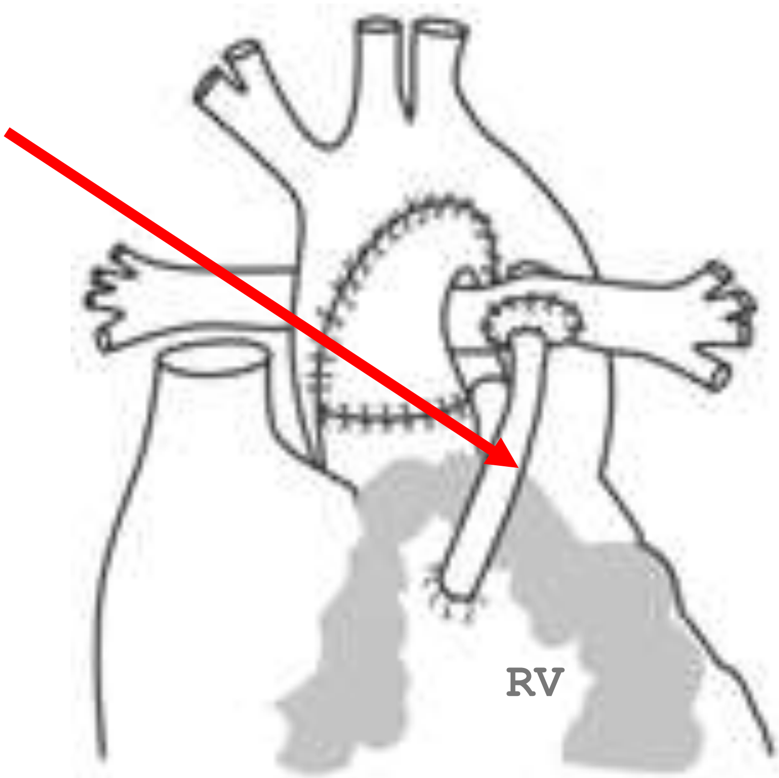
BT Shunt

Norwood I: Sano

Sano modification

- RV-to-PA conduit
- Eliminates competitive flow to PAs in diastole
- Enhances coronary perfusion



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Sano Shunt

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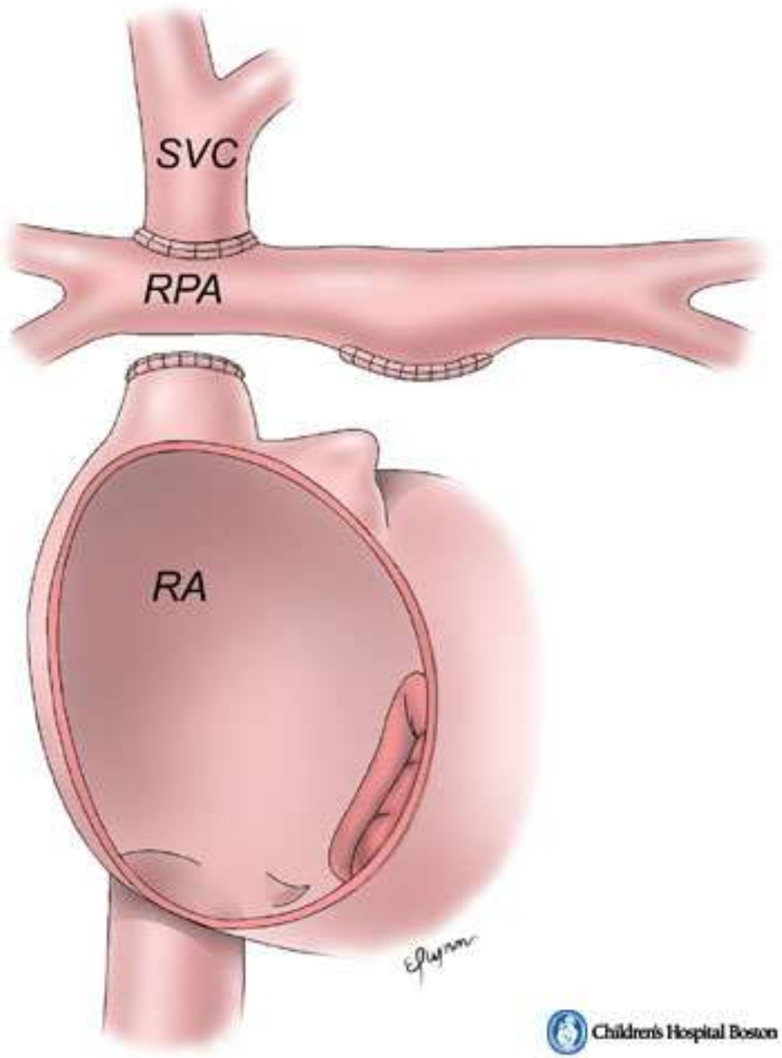
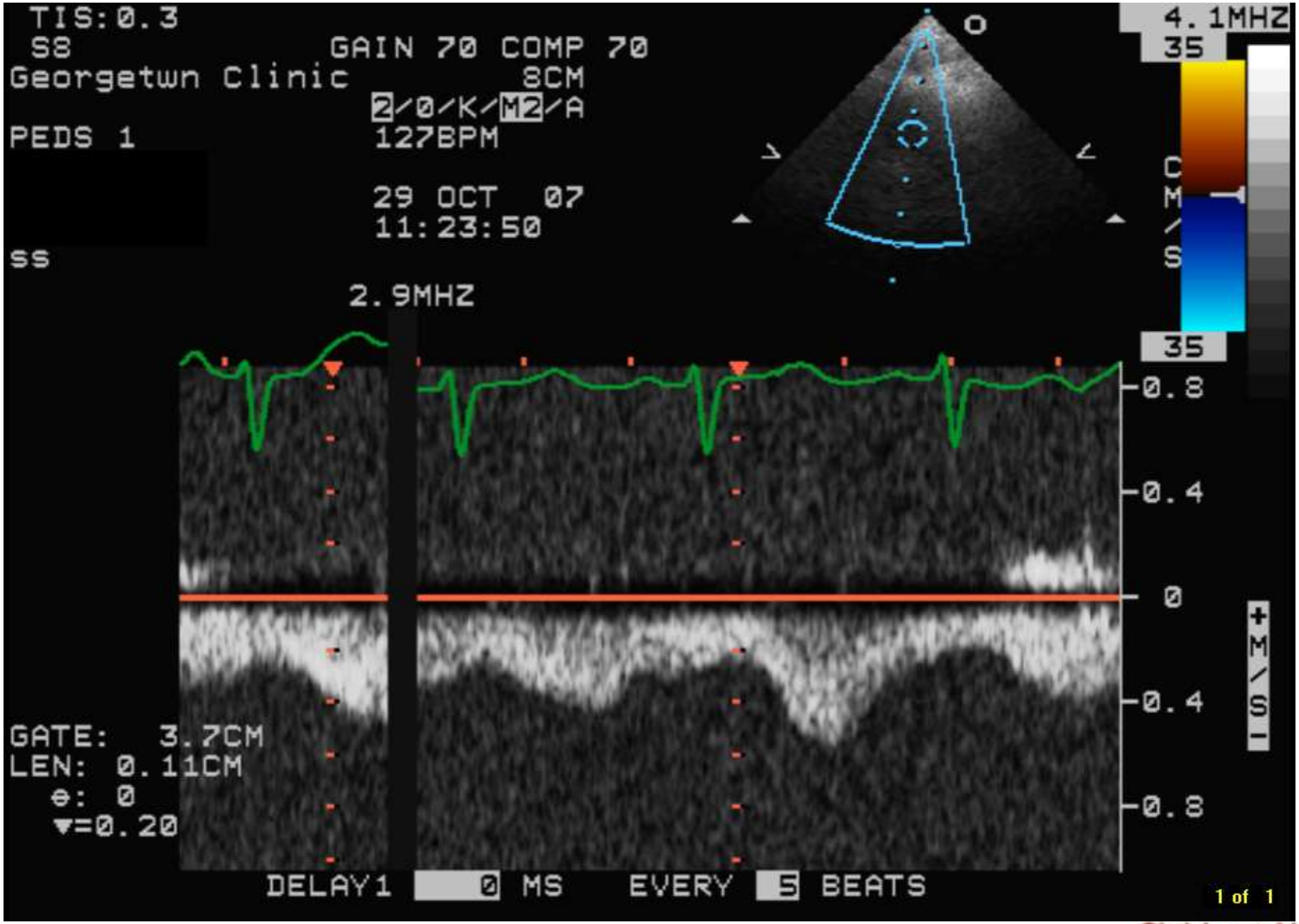
Bidirectional Glenn: Anatomy



- End-to-side anastomosis of SVC to undivided right pulmonary artery
- Includes takedown of BT shunt
- Allows flow to both lungs from SVC via passive flow

Glenn Shunt

Glenn Doppler



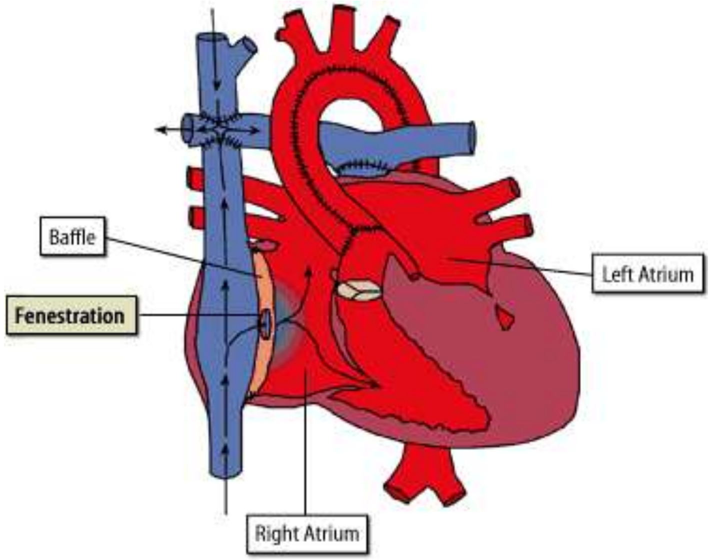
Fontan: Variations

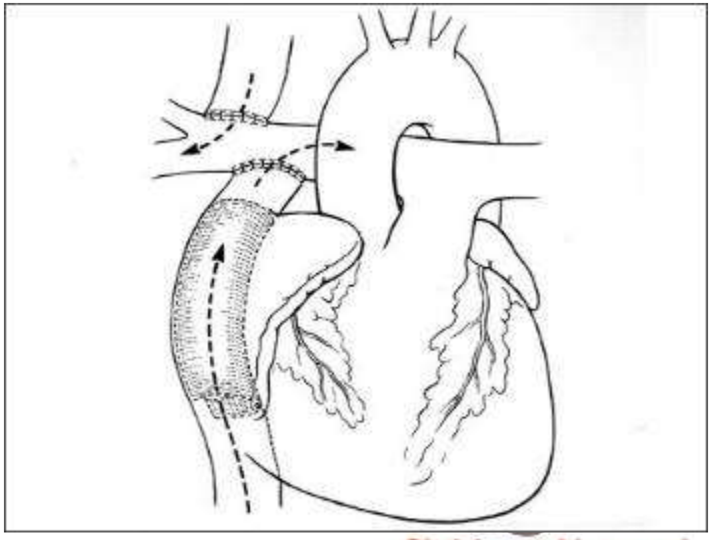
Lateral tunnel runs within RA, using free wall plus conduit as baffle for IVC blood

- Fenestrations: R-to-L shunting through the fenestration → hypoxemia
- Improve cardiac output, minimize systemic venous hypertension, decrease post-op thoracostomy drainage
- Can later be closed by cath

Extracardiac is IVC to MPA

- Generally has lower rate of complications





Fenestrated Fontan

Hypoplastic Left Heart Syndrome Palliative Reconstruction

Stage I -- Norwood Procedure

- Birth

Stage II -- Bi-directional Cavopulmonary Shunt

- 4-6 months

Stage III-- Fontan Procedure

- 18-24 months for lateral tunnel procedure
- > 15 kg for extracardiac procedure

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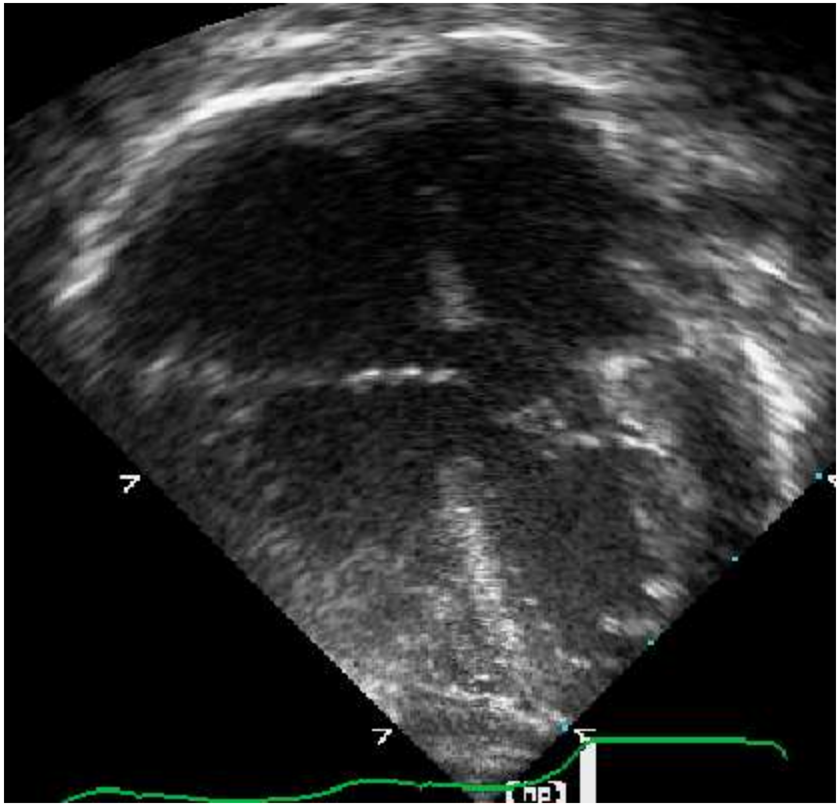
QUESTION 1

A tachypneic 2 month old is not growing well and has a murmur. An echocardiogram is obtained:



SYSTOLE

DIASTOLE

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E

QUESTION 1 (CONT)

All of the following statements are likely to be true except:

A. The patient is at increased risk to have Down Syndrome

B. The patient may not need surgery

C. The patient has an endocardial cushion defect

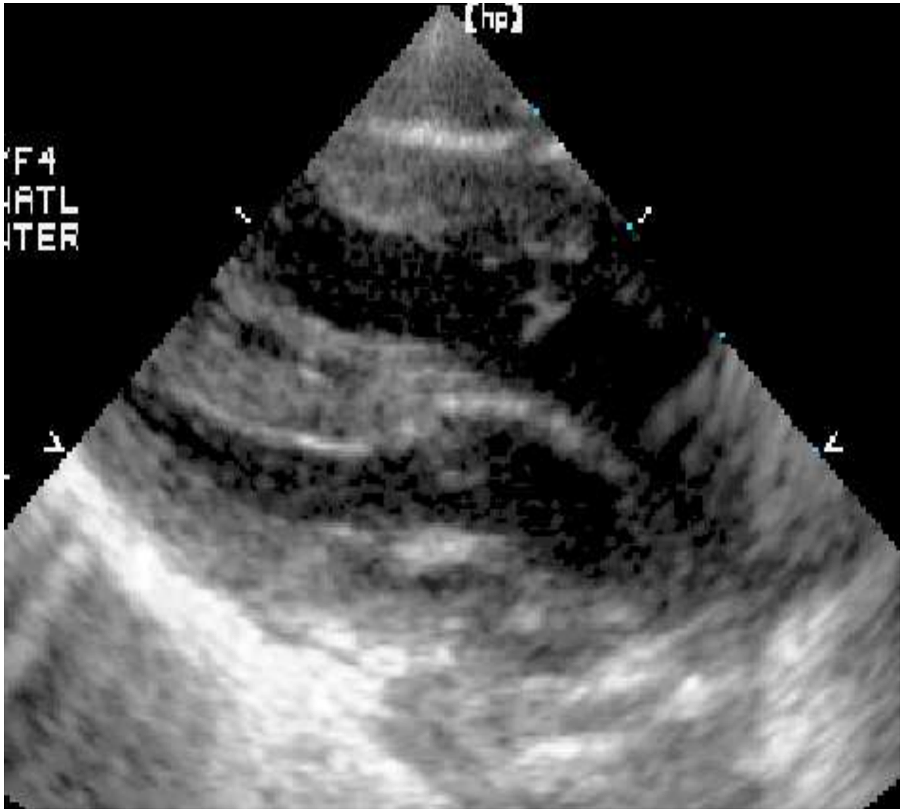
D. The patient has a normal oxygen saturation

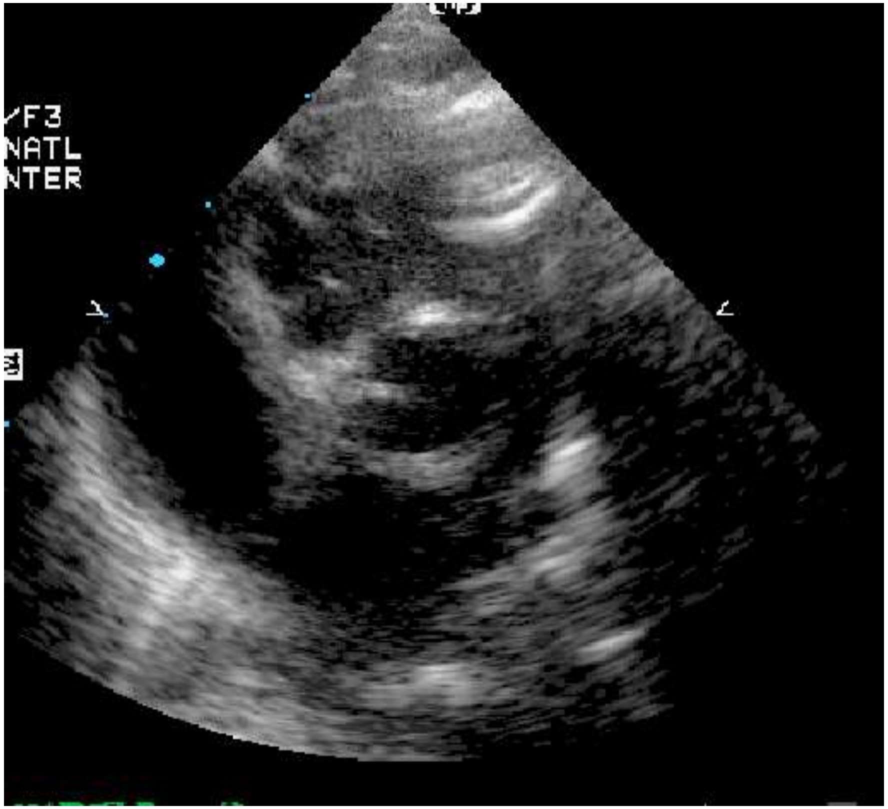
E. The patient may have a small mitral valve cleft after surgical repair

E

QUESTION 2

A cyanotic newborn has the following echocardiogram:





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QUESTION 2 (CONT)

All of the following statements are likely to be true except:

A. The pulmonary artery gives rise to the coronary arteries.

B. The right ventricle pumps blood to the body

C. Oxygenated blood is pumped to the lungs

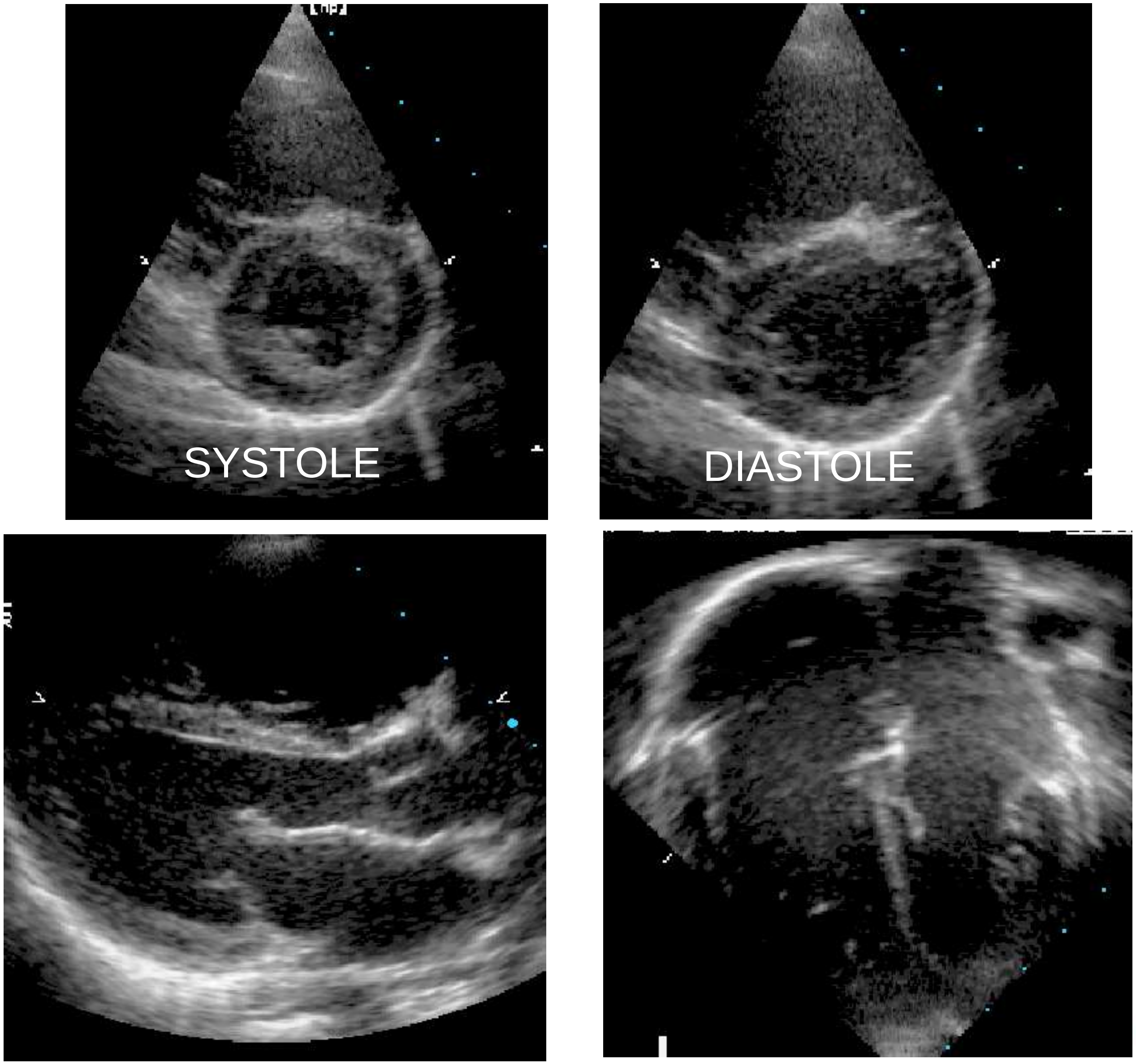
D. The left ventricle pumps blood to the body

E. The right ventricular pressure is greater than or equal to the left ventricular pressure

E

QUESTION 3

A 40 year old with atrial fibrillation has the following echo:



E

QUESTION 3 (CONT)
Subsequent imaging is most likely to reveal the following

A. Tetralogy of Fallot

B. Large membranous ventricular septal defect

C. Large patent ductus arteriosus

D. Large secundum atrial septal defect

E. No structural cardiac defect

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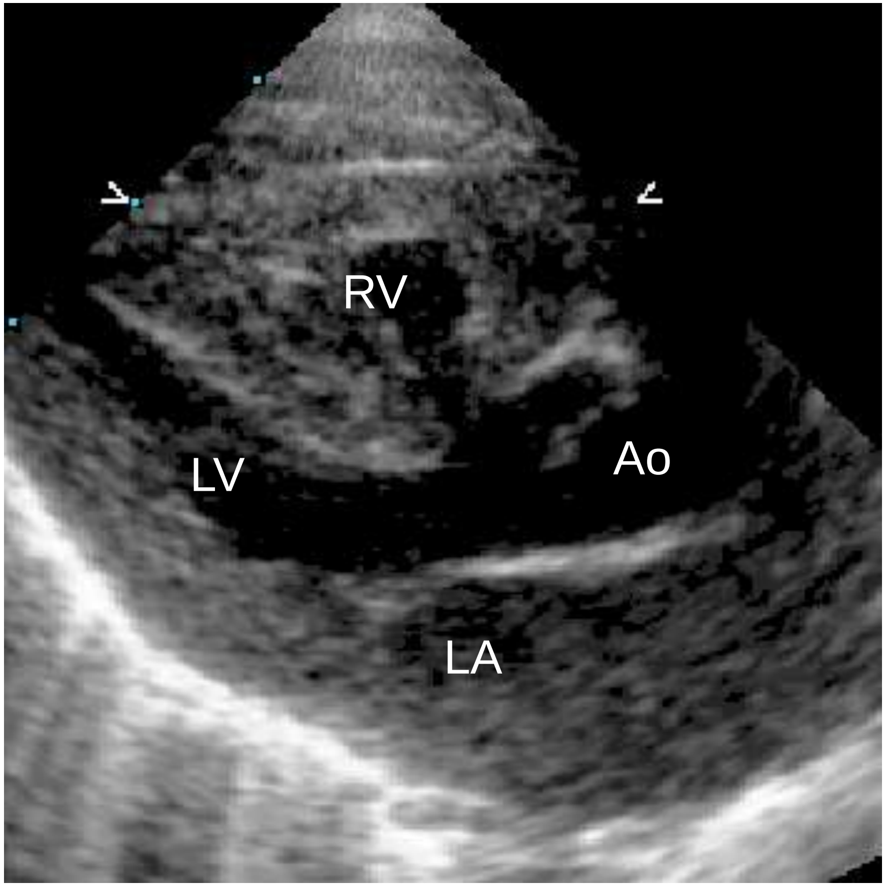
QUESTION 4

A 3 month old with a loud murmur and intermittent perioral cyanosis has the following echo:



E

QUESTION 4 (CONT)

All of the following statements are likely to be true except:

A. The aorta is overriding the left and right ventricle

B. There is a large ventricular septal defect

C. There is pulmonary stenosis

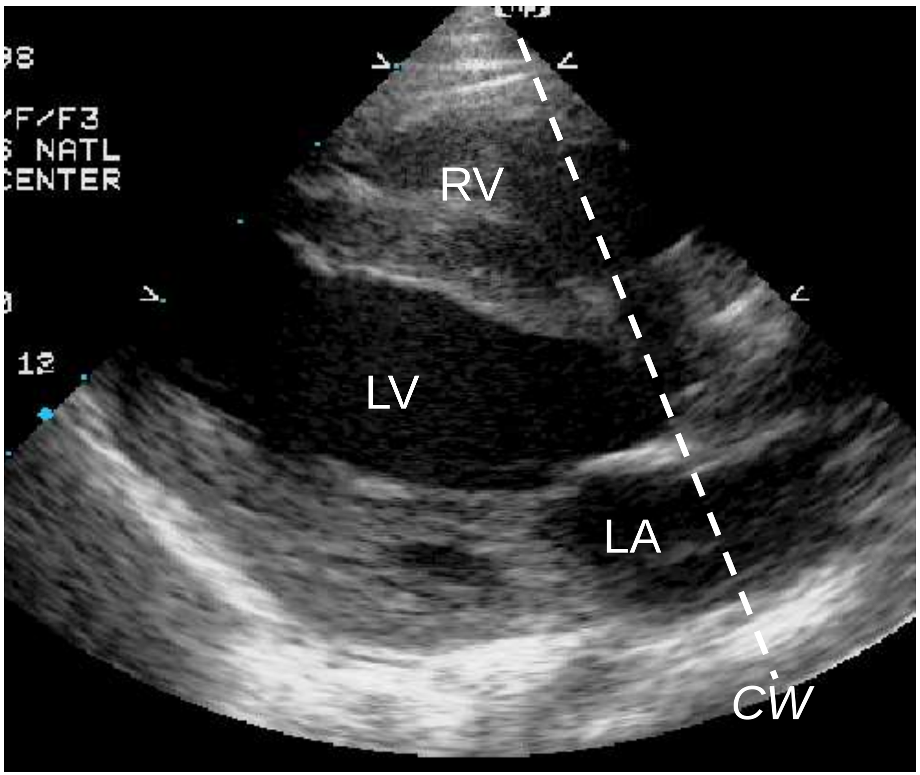
D. The right ventricular pressure is increased

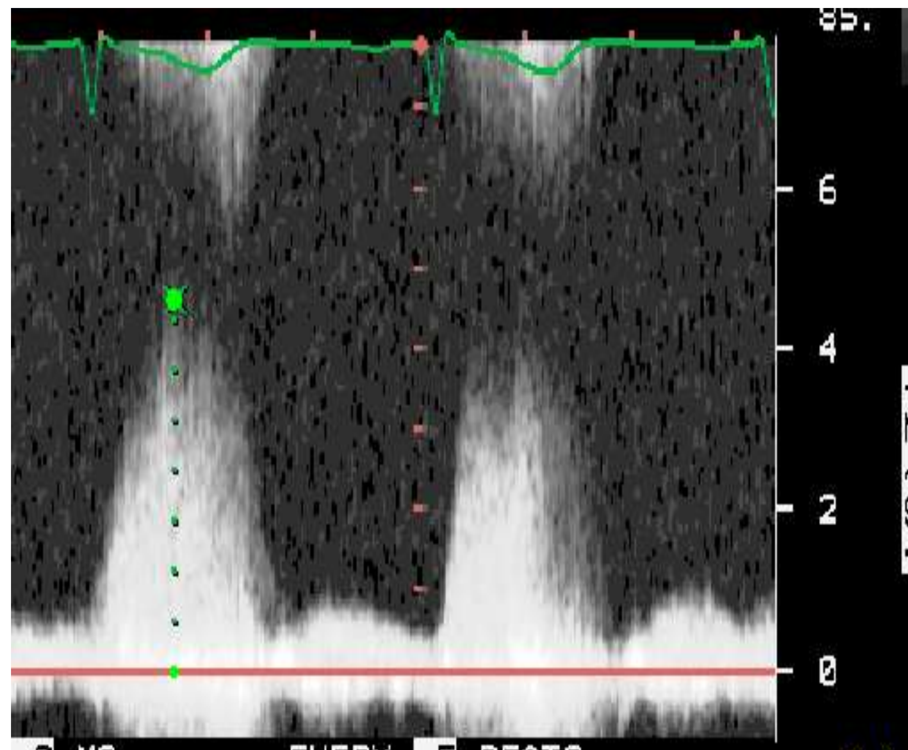
E. The pulmonary artery pressure is increased

E

QUESTION 5

An asymptomatic 9 month old with a loud murmur and a BP of 79/48 and has the following parasternal long axis 2D and CW Doppler findings:





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QUESTION 5 (CONT)
The most likely diagnosis is:

A. Membranous VSD, normal RV pressure

B. Membranous VSD, elevated RV pressure

C. Muscular VSD, normal RV pressure

D. Muscular VSD, elevated RV pressure

E. Tricuspid regurgitation, elevated RV pressure

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Acknowledgements

Unattributed illustrations are from Nadas' Pediatric Cardiology

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- **202-476-4880 Physician Line**
- **202-476-5579 Echo Lab**

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