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North East and North Cumbria Congenital Heart Disease Network

Introduction to Congenital Heart Disease (CHD)

PAEDIATRIC CHD Webinar (NENC-CHDN)

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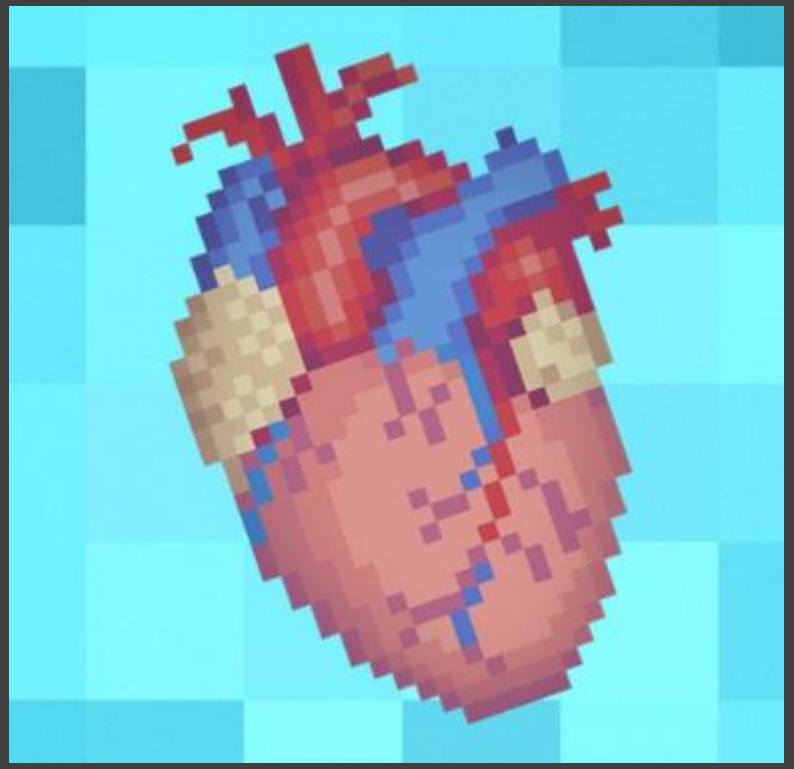
19.05.2020



Outline

- CHD - Overview
- Early Presentation - CHD
- Duct-dependent lesions
- Common CHD (acyanotic lesions)
- Quiz Time

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CHD – Definition

The definition of congenital heart disease is the persistence of any structural abnormality of the heart or great vessels that is present at birth.

These defects occur during fetal development and majority of the lesions can be detected antenatally by fetal echocardiography.

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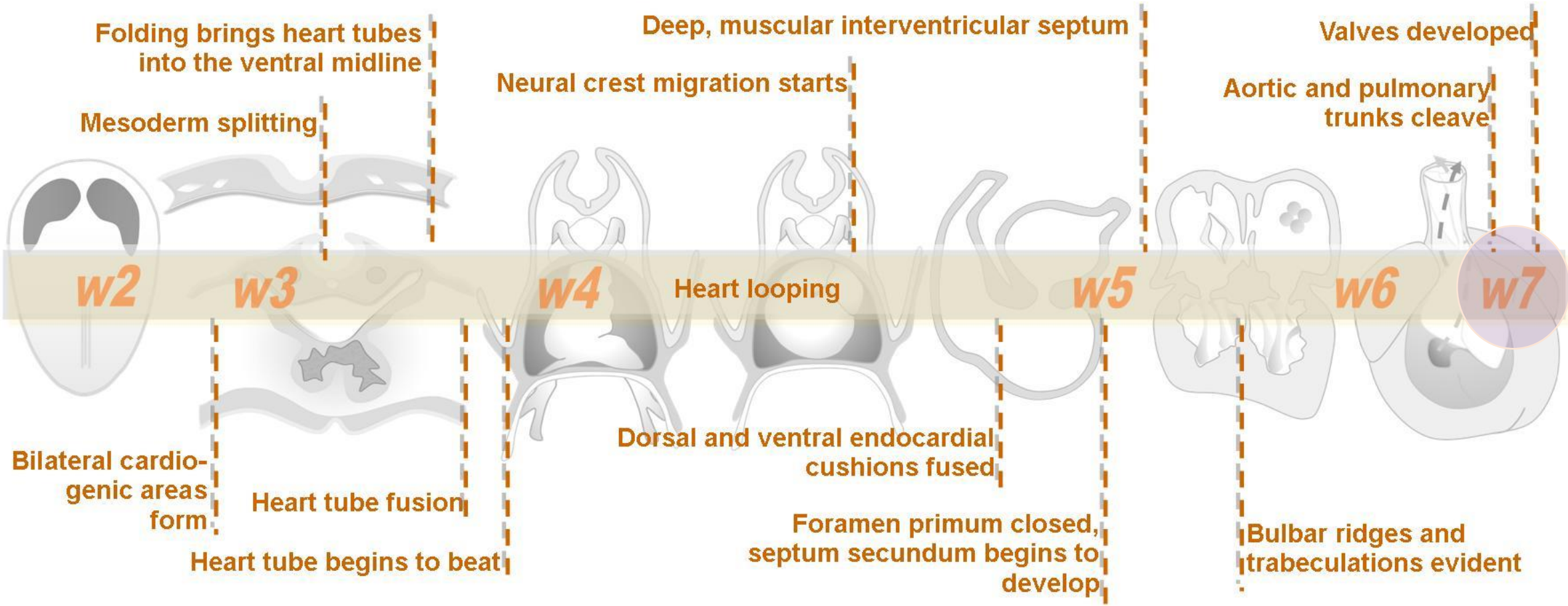
Cardiac Embryology



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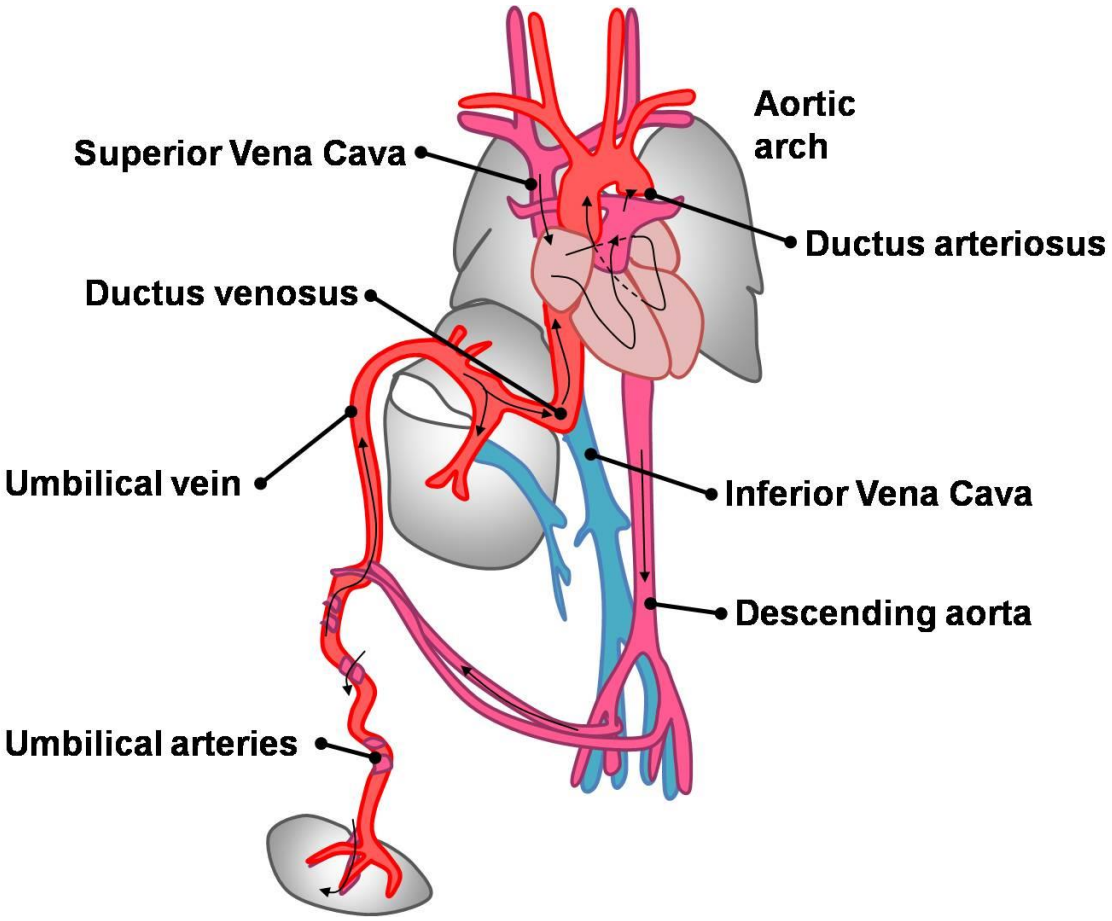
Fetal Circulation



3 major vascular shunts:

- Ductus venous (UV & IVC)
- Foramen ovale (RA & LA)
- Ductus arteriosus (PA & DA)

Redirect oxygenated blood away from the lungs, liver and kidney (placenta)

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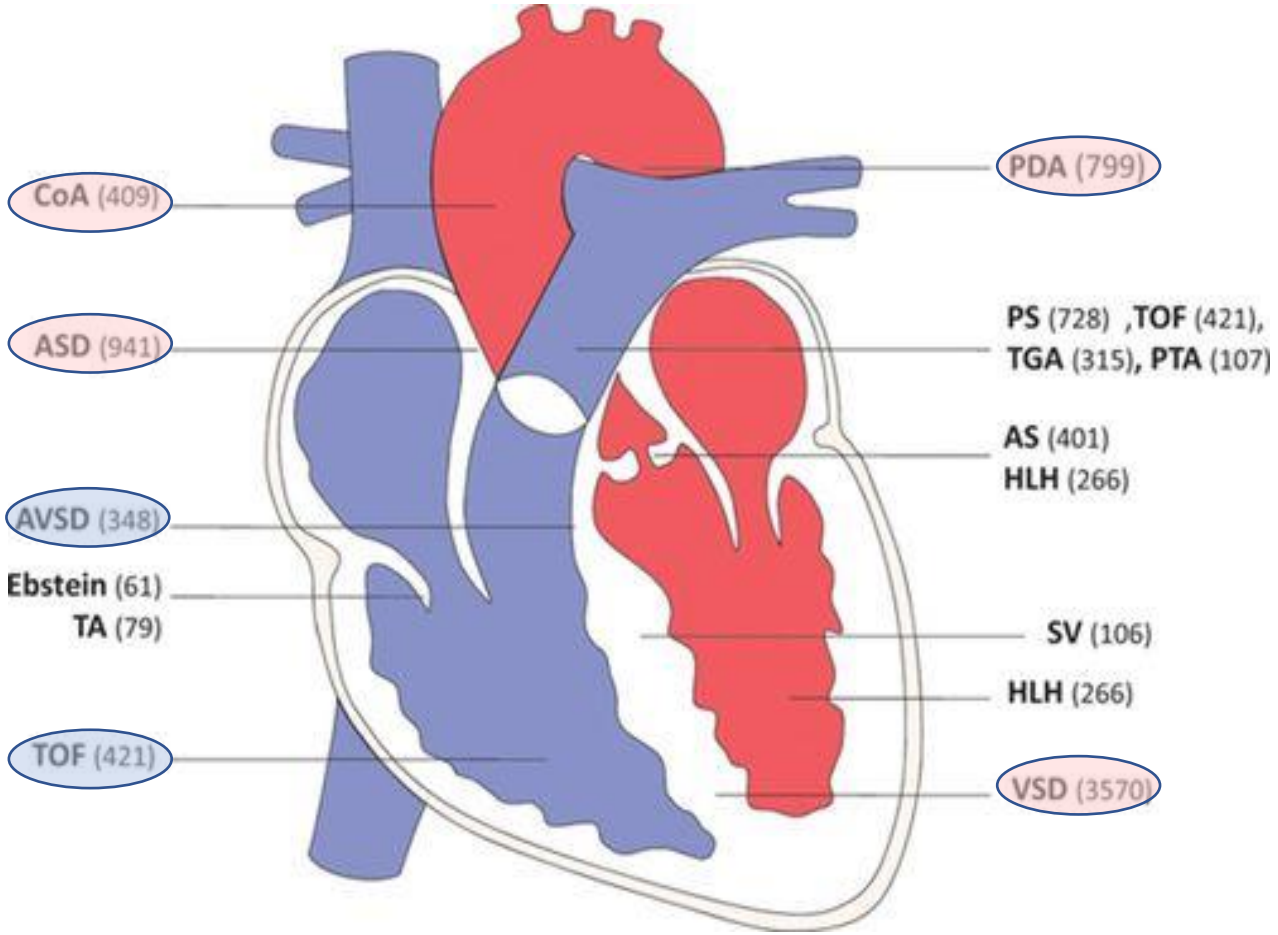
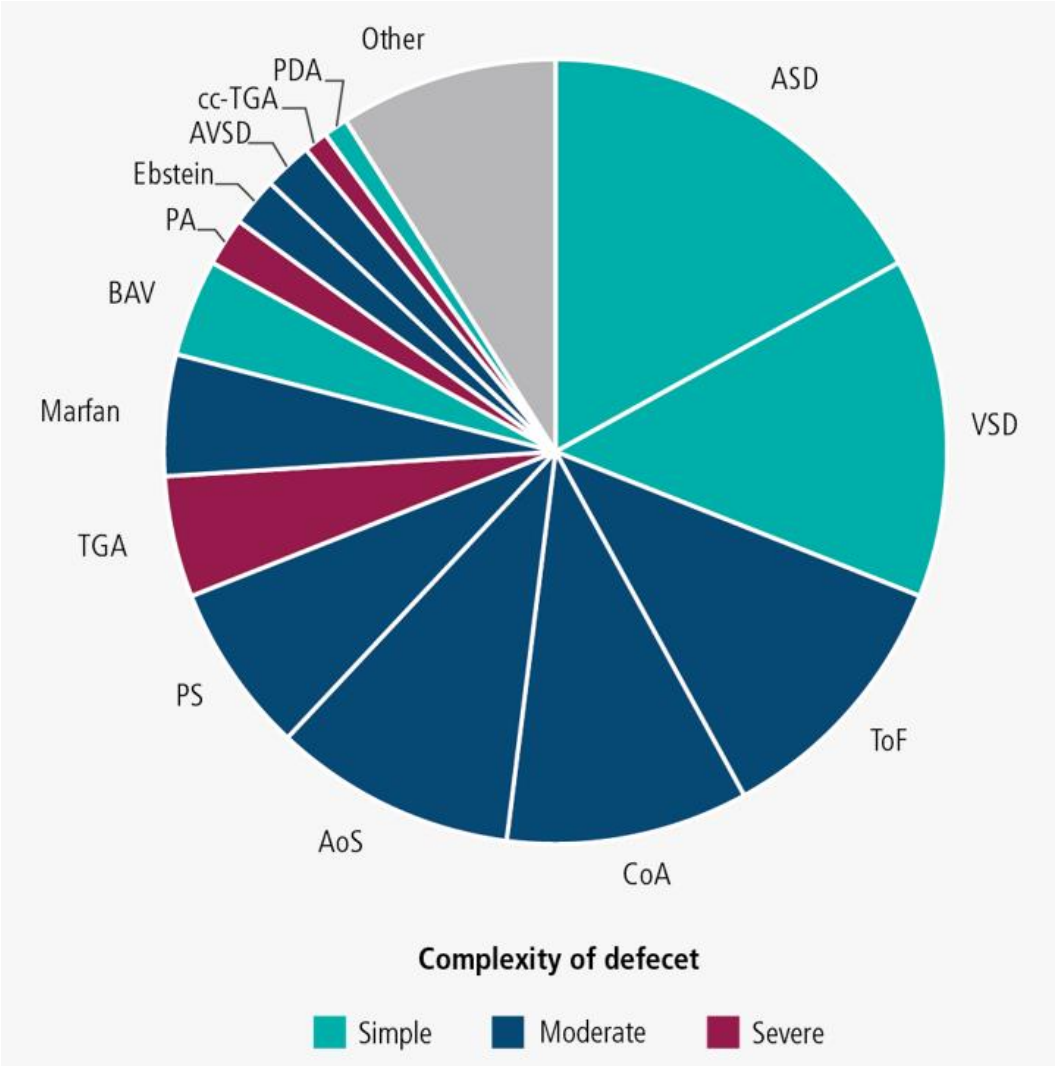
Common CHD



Locations of heart malformations that are usually identified in infancy, and estimated prevalence based on the CONCOR database.
Numbers indicate the birth prevalence per million live births

Fahed et al; Genetics of Congenital Heart Disease The Glass Half Empty; Circulation Research. 2013;112:707–720



Verheugt, Carianne L., et al. European Heart Journal 2010

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Facts - CHD

#1 CHD is the most common birth defect in the world

6-8/1000 live births

25% critical CHD requiring surgery

Surgical survival rates nearly 100% for some procedures

13 babies per day

1-2 % of UK population affected

On average more than 50% CHD* are spotted during pregnancy

> 80% survive to adulthood

Over 250000 adults with CHD (UK)

Most common is VSD

Cost of CHD service to NHS England

£ 175 million pa

Source: NHS, Englnad

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Common Syndromes associated with CHD

Genetic Syndrome	Genetic Etiology	CHD (%)	Cardiac Lesions
Down's	Chr 21 (trisomy)	40-50	AVC VSD, ASD, TOF
22q11.2 Deletion	Chr 22q11; TBX1 (deletion)	> 80	IAA, TA TOF, VSD, aortic arch anomalies
Turner's	Chr X (monosomy)	25-50	COA, BAV AS, HLHS, PAPVD, aortic root dilatation
Williams	Chr 7q11.23; ELN (deletion)	> 80	SVAS, PPS AV & MV abnormalities
Noonan's	RAS-MAPK pathway (single gene mutation)	75-80	PS, HCM VSD, ASD
Kabuki	12q13.12; KMT2D, KDM6A (single gene mutation)	31-55	COA, ASD, TOF, VSD, HLHS PDA, TGA, AS, MS
Alagille	20p12; JAG1, NOTCH2 (single gene mutation)	90	PPS, PS, TOF

Fahed et al; Genetics of Congenital Heart Disease The Glass Half Empty; Circulation Research. 2013;112:707–720

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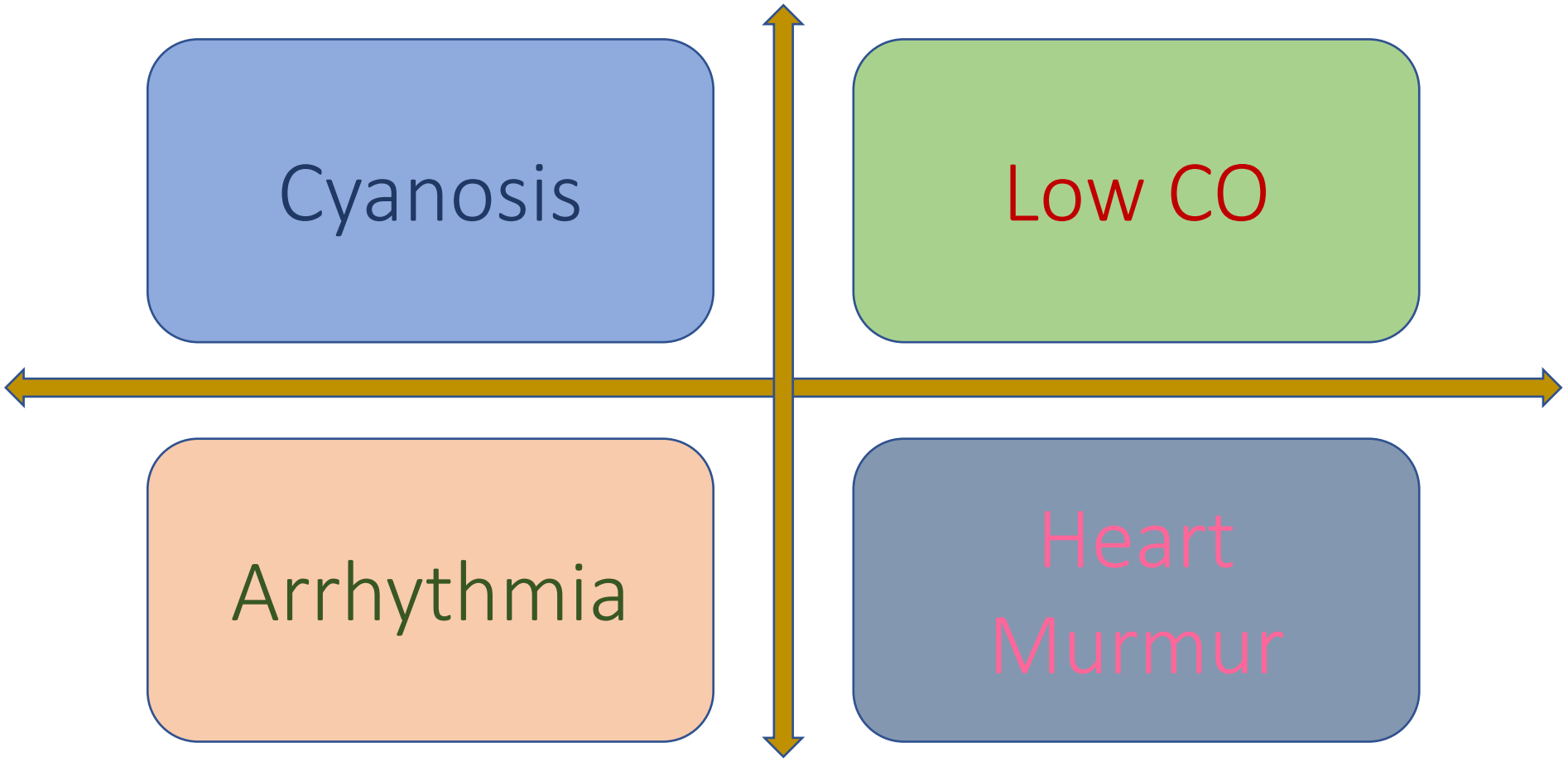
Early Presentation in CHD



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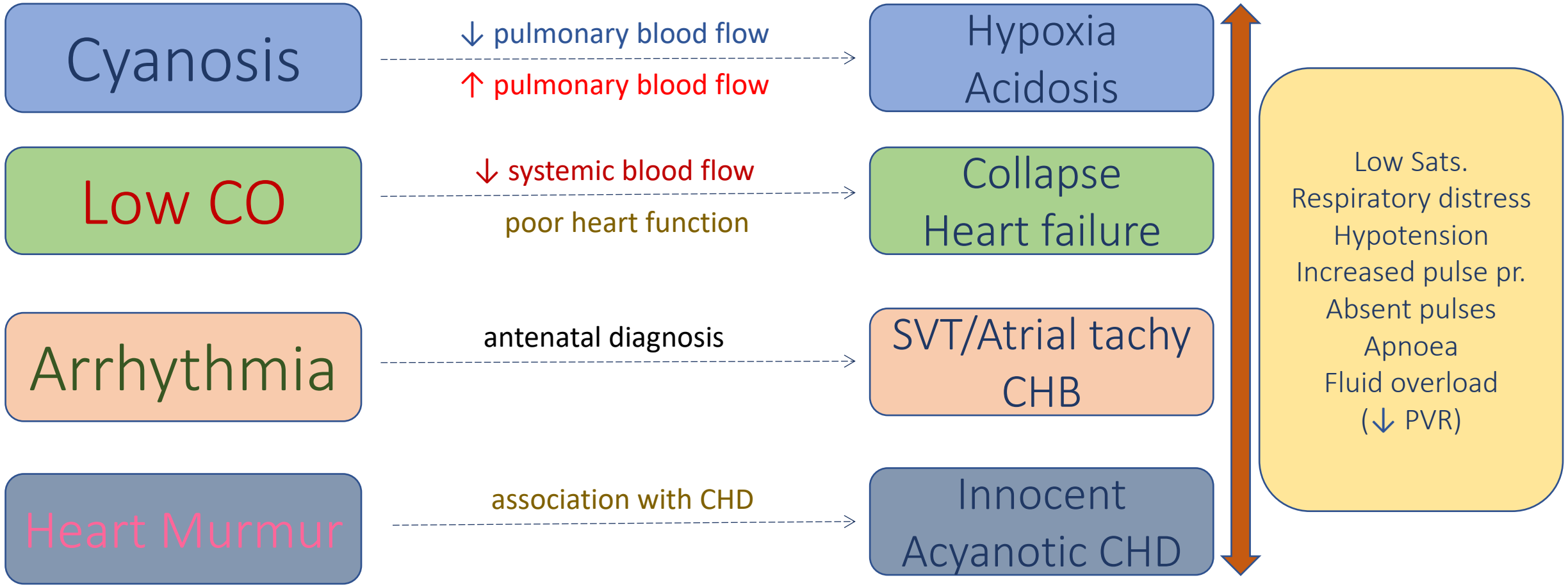
Early Presentation in CHD



Duct-dependent (DD) Lesions

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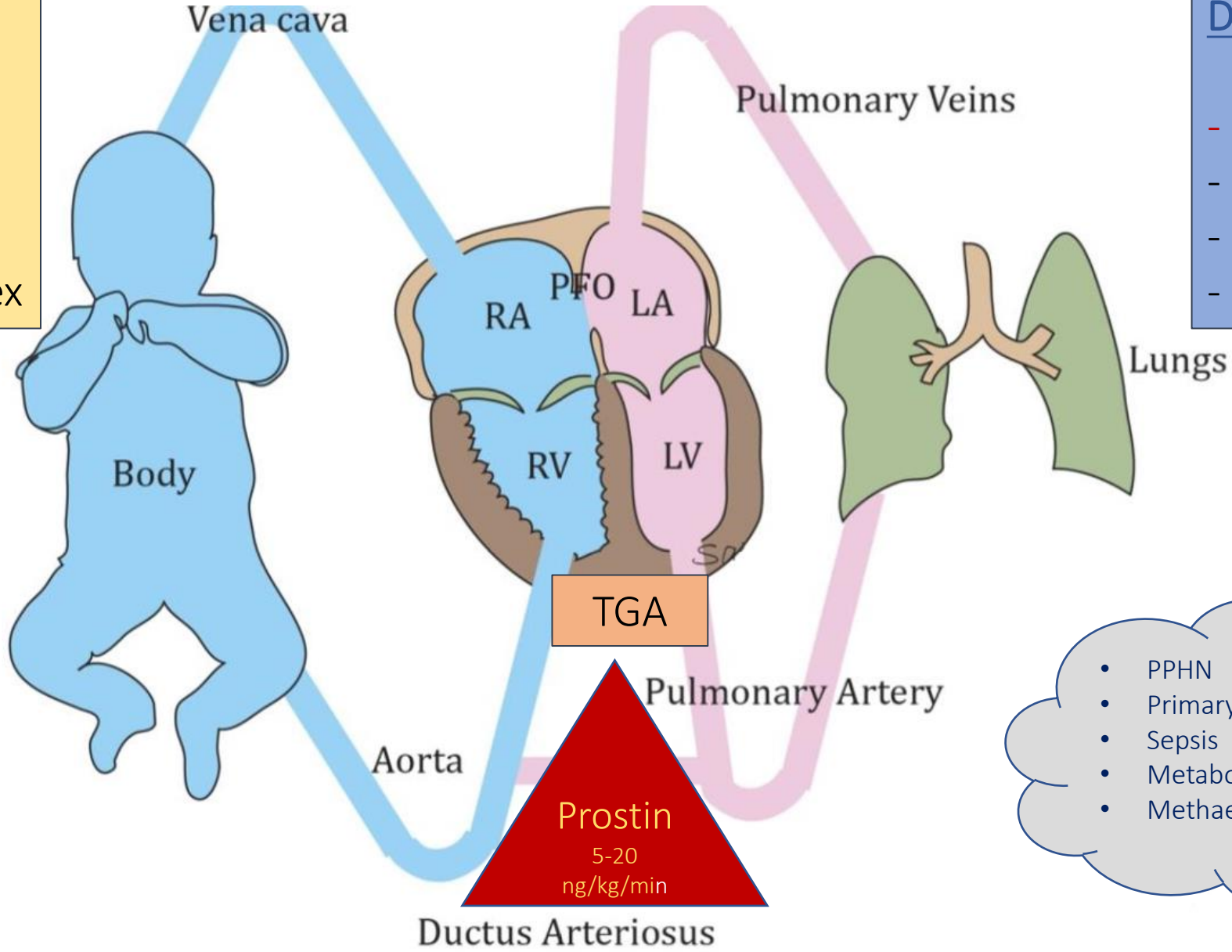
DD Systemic Circ.

- COA/IAA
- Critical AS
- HLHS
- 'Shone' complex



DD Pulmonary Circ.

- Tetralogy of Fallot
- Critical PS
- PA/IVS
- Tricuspid Atresia

- PPHN
- Primary lung ds.
- Sepsis
- Metabolic ds.
- Methaemoglobinaemia

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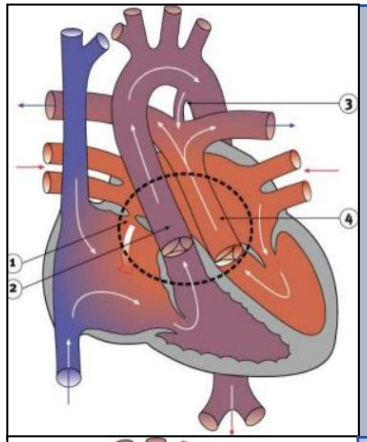
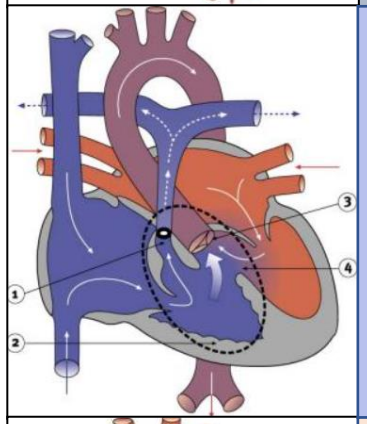
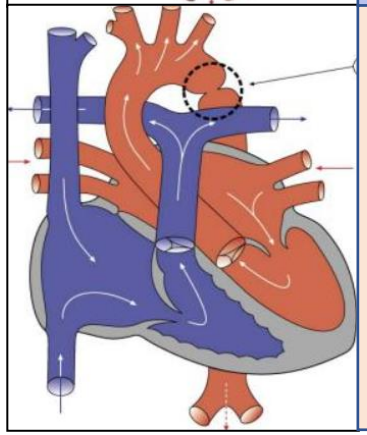
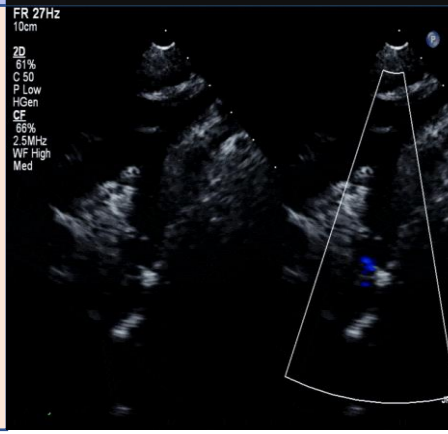
Duct-dependent (DD) Lesions

	1. ASD 2. Aorta 3. PDA 4. PA	Transposition of the Great Arteries (10-11% CHD)	DD Systemic & Pulmonary Circulation	Cyanosis Hypoxemia Metabolic Acidosis	1. Prostin 2. Balloon atrial Septostomy 3. Arterial Switch Op.	
	1. PS 2. RVH 3. Aortic override 4. VSD	Tetralogy of Fallot (9-14% CHD)	DD Pulmonary Circulation	<u>Spells</u>: 1. Calm 2. Oxygen 3. Knee-chest position 4. Sedation 5. Hydration	1. Prostin (+/-) 2. Ductal stent 3. RVOT stent 4. BT shunt 5. Complete repair	
	Narrowing juxta-ductal region	Coarctation of Aorta (5-7% CHD)	DD Systemic Circulation	Collapse Shock Tachypnoea Hypotension Weak pulses	1. Prostin (+/-) 2. Surgical repair 3. Stent insertion (late presentation)	

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Investigations for CHD

- Assessment/Auscultation/Pulse Oximetry
- Chest X-ray
- ECG
- Echocardiography
- Cardiac CT/MR
- Cardiac Catheterisation/Angiography
- Electrophysiological studies

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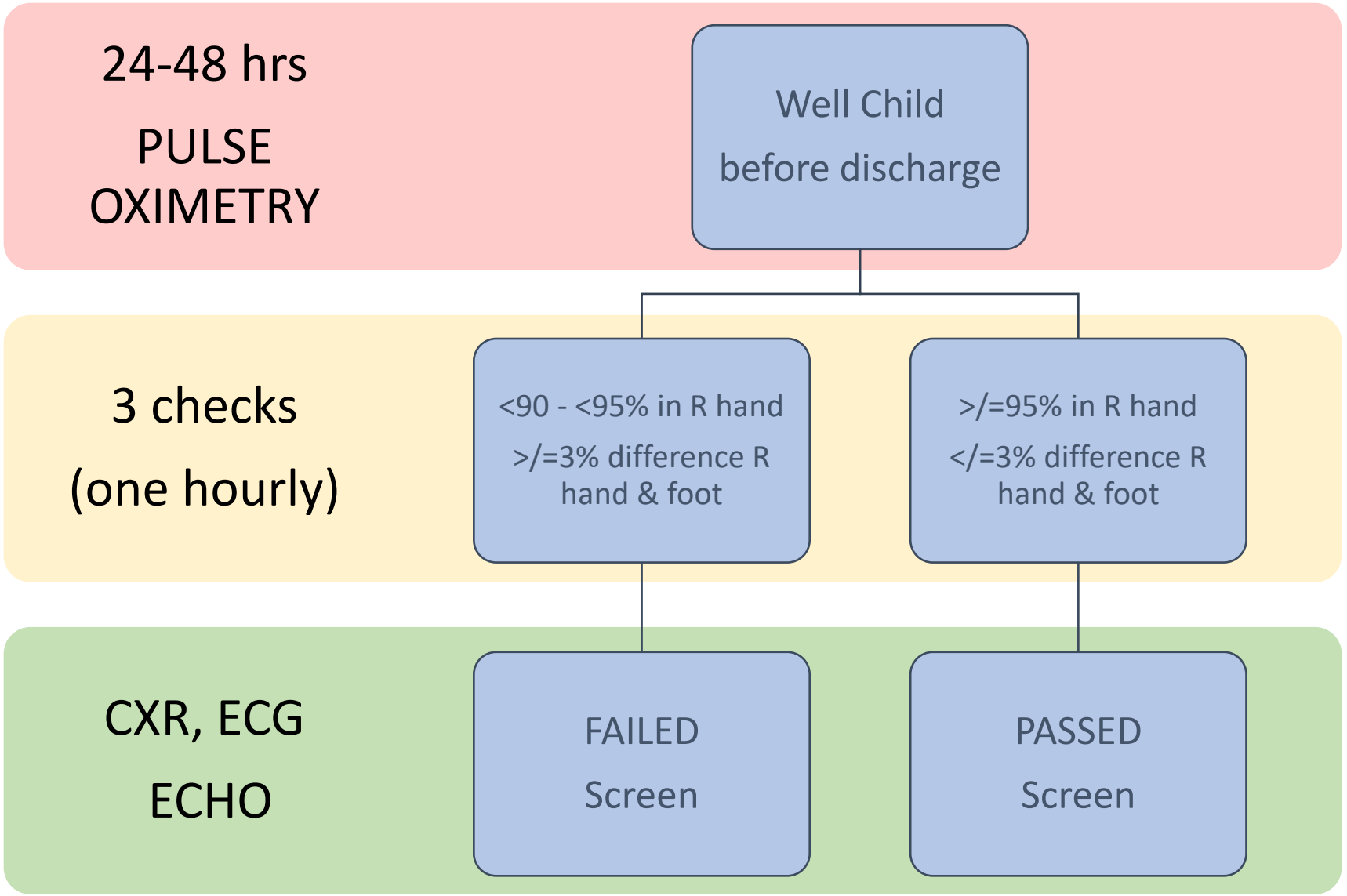
CHD Screening Algorithm



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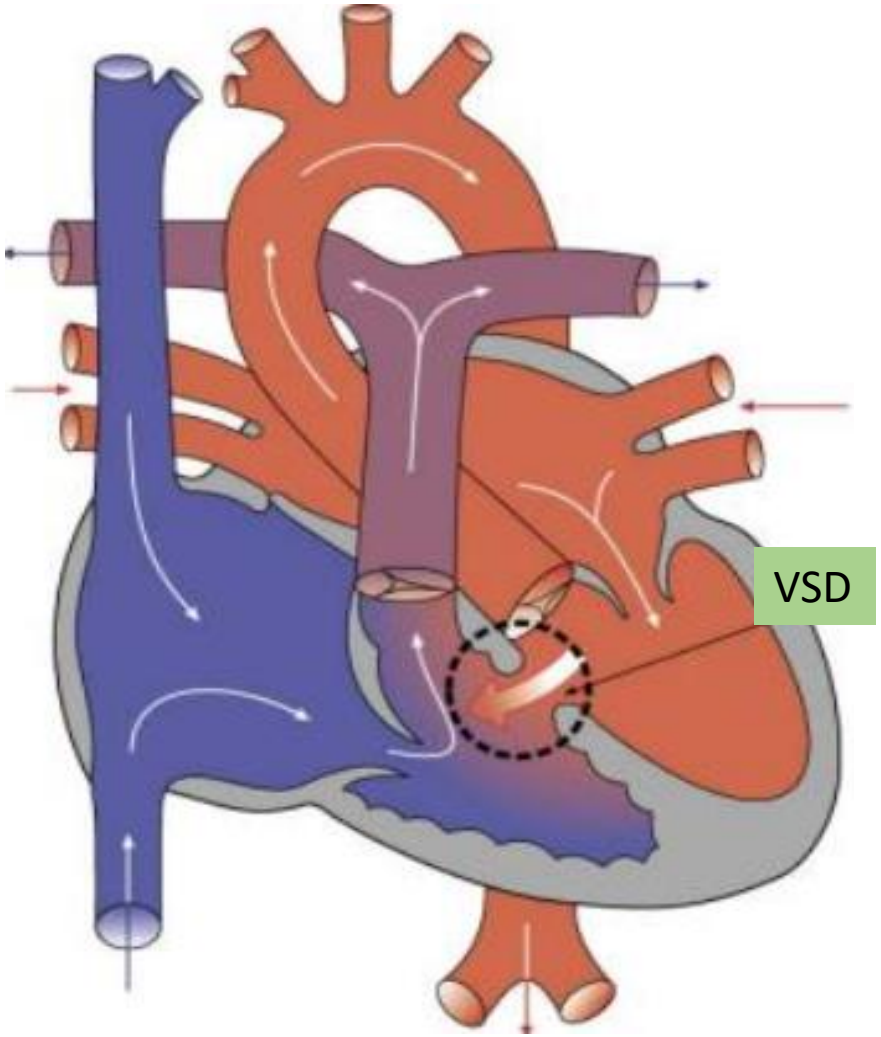
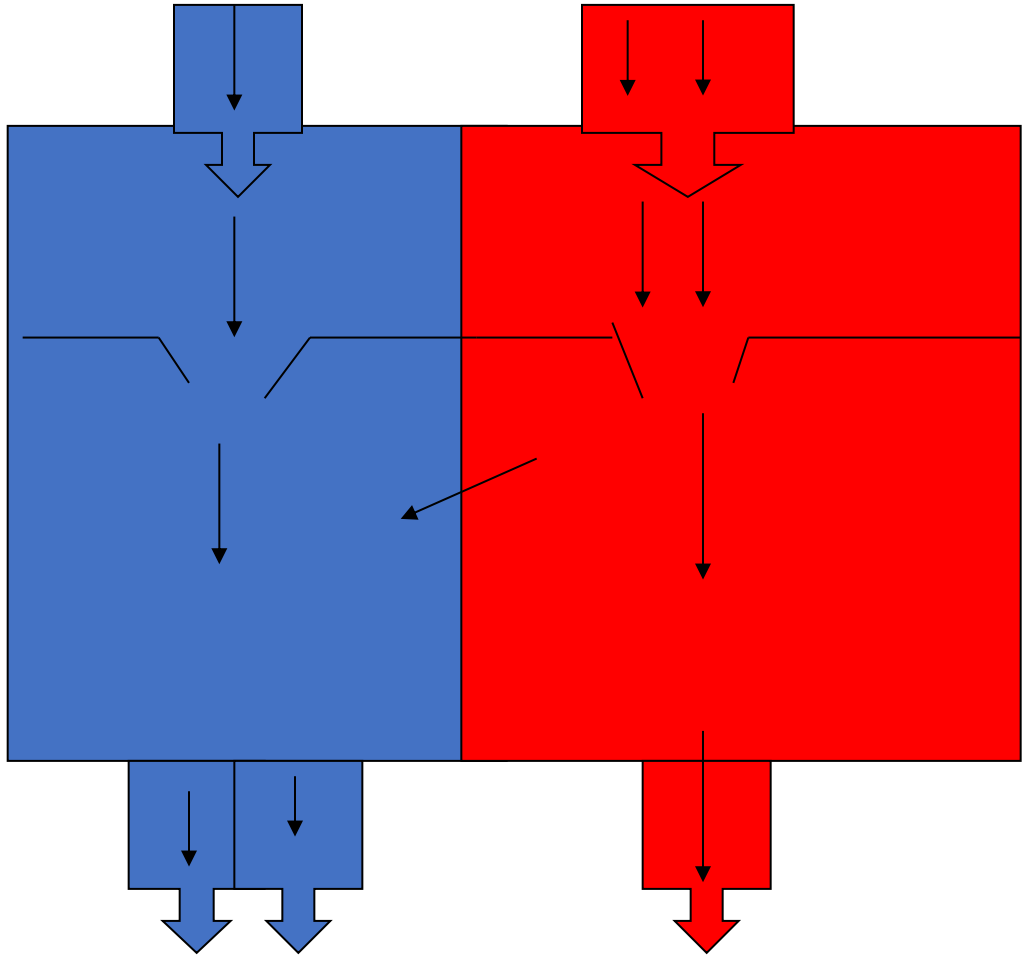
Ventricular Septal Defect (VSD, 30-35%)





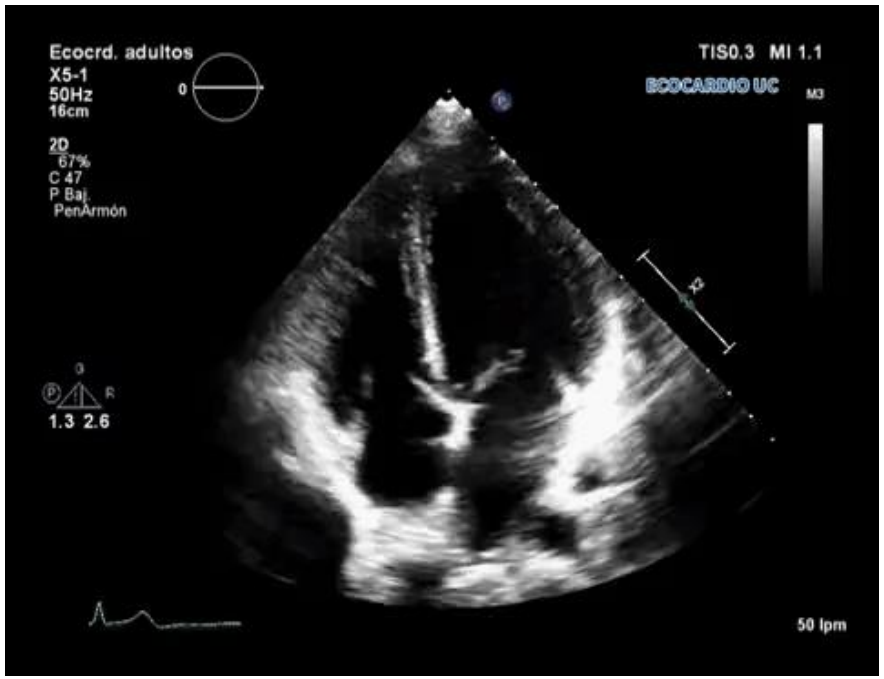
Ventricular Septal Defect (VSD, 30-35%)

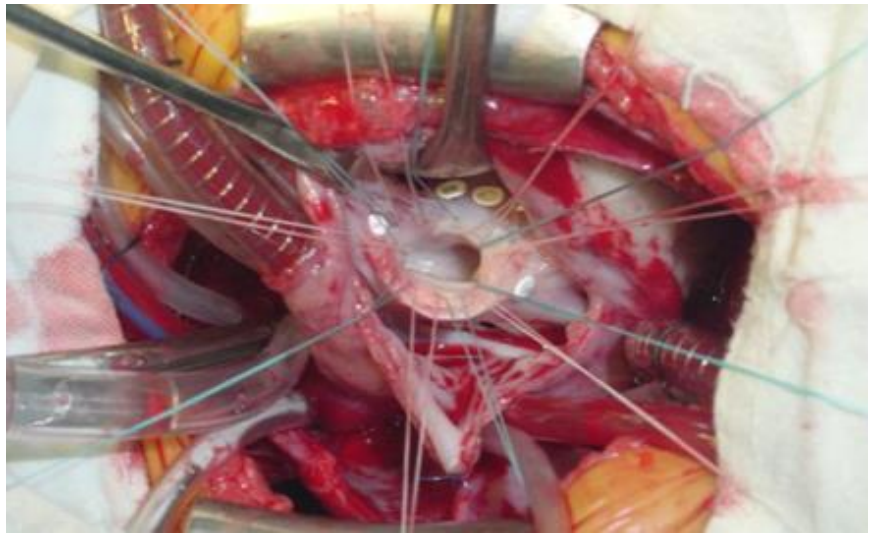
- Left to right shunt (acyanotic)
  - ? Eisenmenger's
  - Type of VSD
- PSM, failure to thrive
- High Qp, Left heart overload (LV dilatation), Heart failure
- Mitral regurgitation, Aortic regurgitation
- Associated lesions: COA, TOF Multiple VSDs

- **Conservative treatment**
  - no medication/ monitor weight
  - high calorie diet/NG
  - Diuretics (Frusemide, Spironolactone), ACEi (Captopril)
- **Surgical treatment**
  - Complete VSD repair
  - PA band f/b complete VSD repair

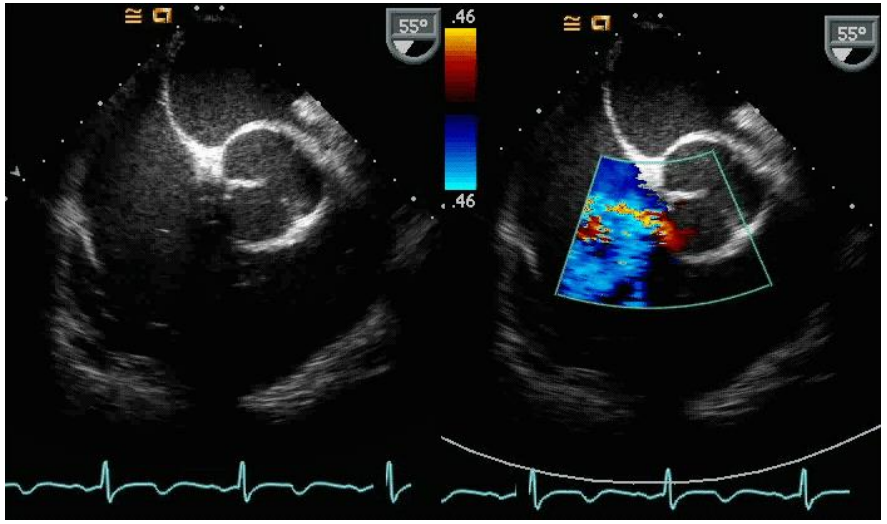
- **Follow up**
  - pericardial effusion
  - VSD patch dehiscence
  - residual VSD, aortic regurgitation
  - no medication

Ventricular Septal Defect (VSD, 30-35%)



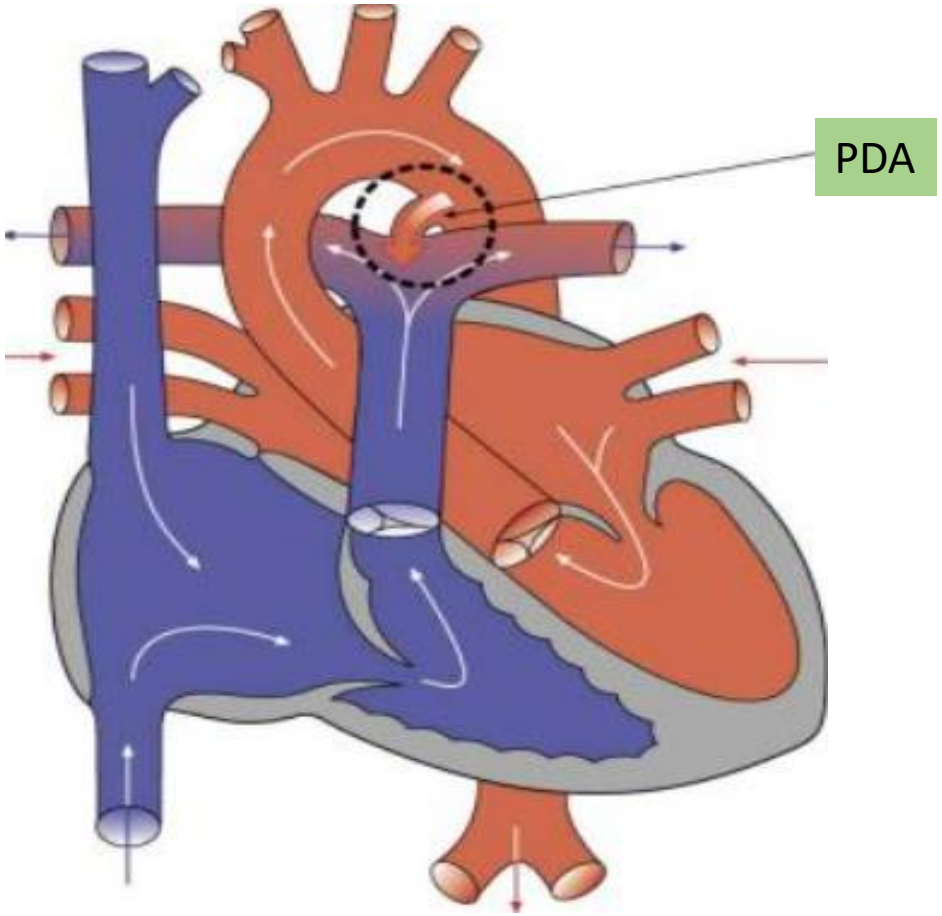


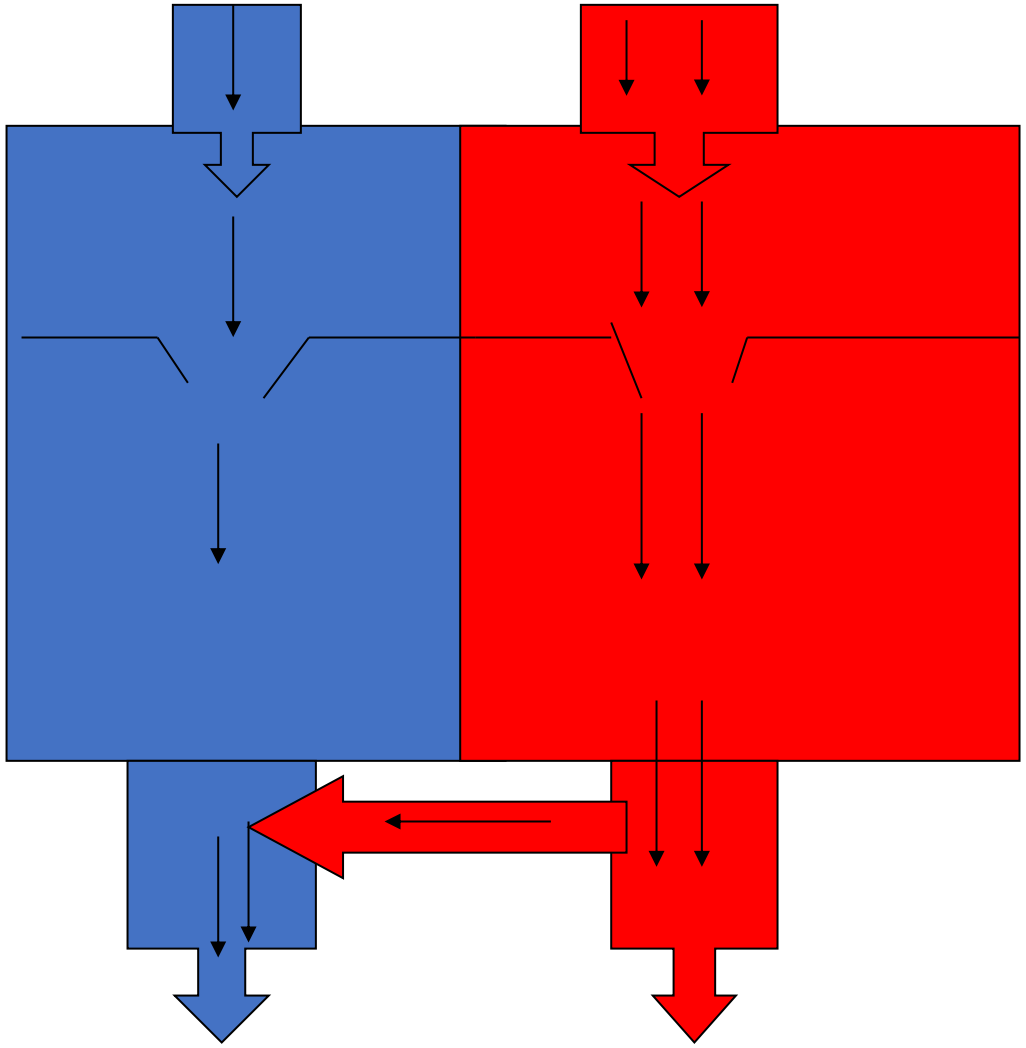
High Qp:Qs
Large heart shadow
Increased lung marking
Pulmonary congestion



Surgical Patch repair
VSD

Patent Ductus Arteriosus (PDA, 10-15%)





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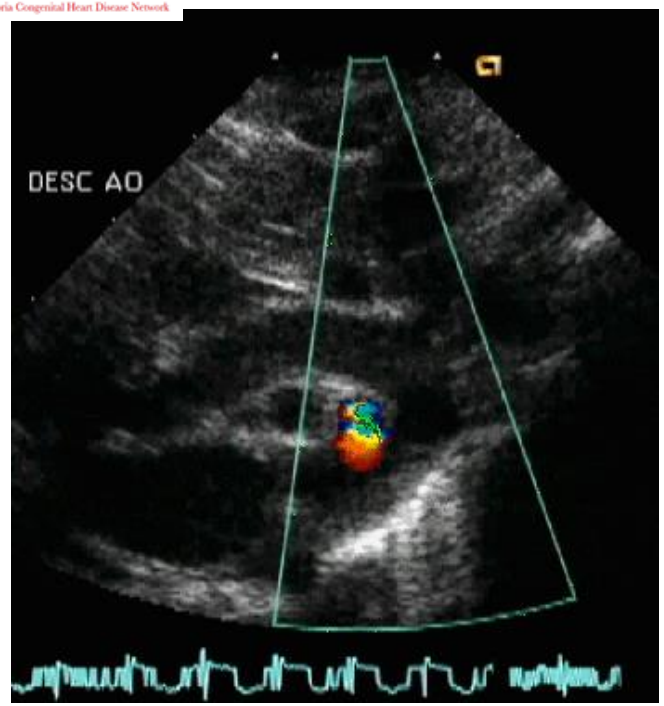
Patent Ductus Arteriosus (PDA, 10-15%)

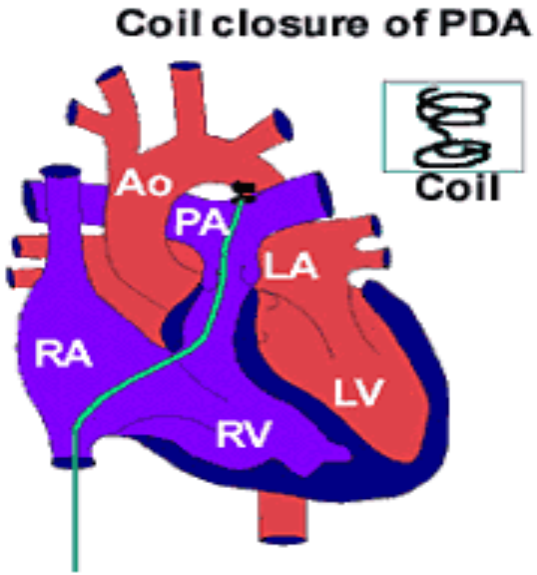
- Left to right shunt (acyanotic)
- Pre-term (80% low BW)
- Size – Small/Moderate/Large
- High Qp, Left heart overload (LA/LV dilatation), Continuous murmur, Wide pulse pressure
- Heart failure, MR
- Pulmonary hypertension (PH)

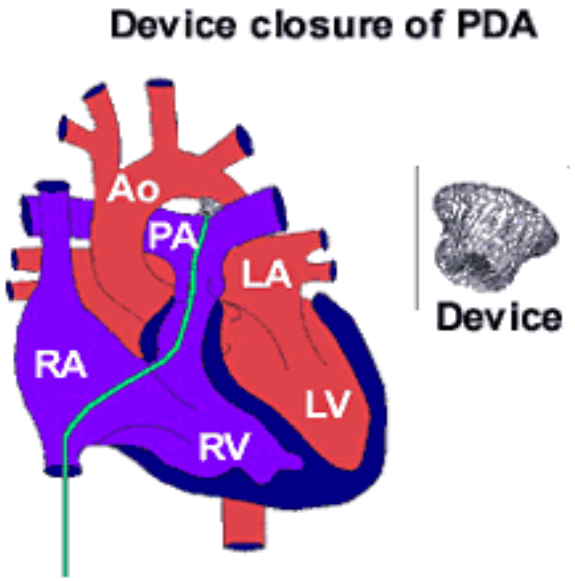
- **Conservative treatment**
  - spontaneous closure
  - Drug therapy (PGEi – Ibuprofen, Aspirin, Indomethacin)
- **Intervention/Surgical**
  - Device/Coil
  - Surgical clip/ligation

- **Follow up**
  - pericardial effusion
  - residual duct
  - obstruction -LPA, aortic arch
  - no medication

Patent Ductus Arteriosus (PDA, 10-15%)







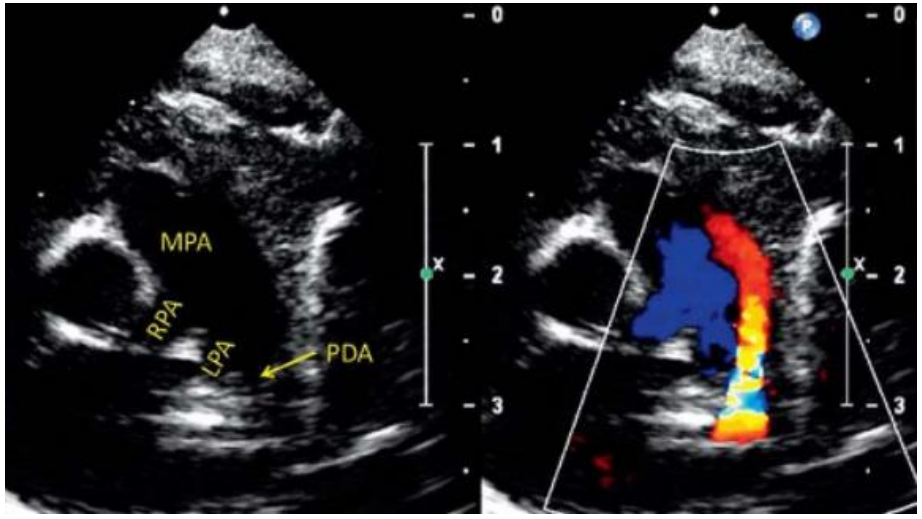
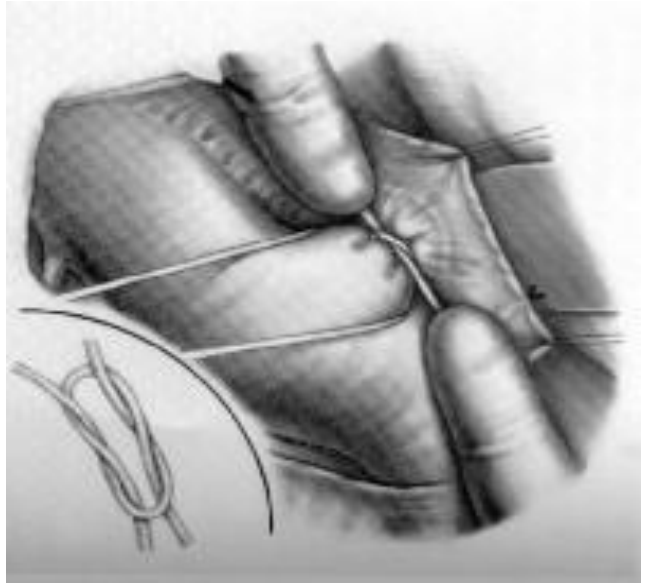


Fig. 4 – Exposure of the double-clipped patent ductus arteriosus. The lung withdrawn anteriorly



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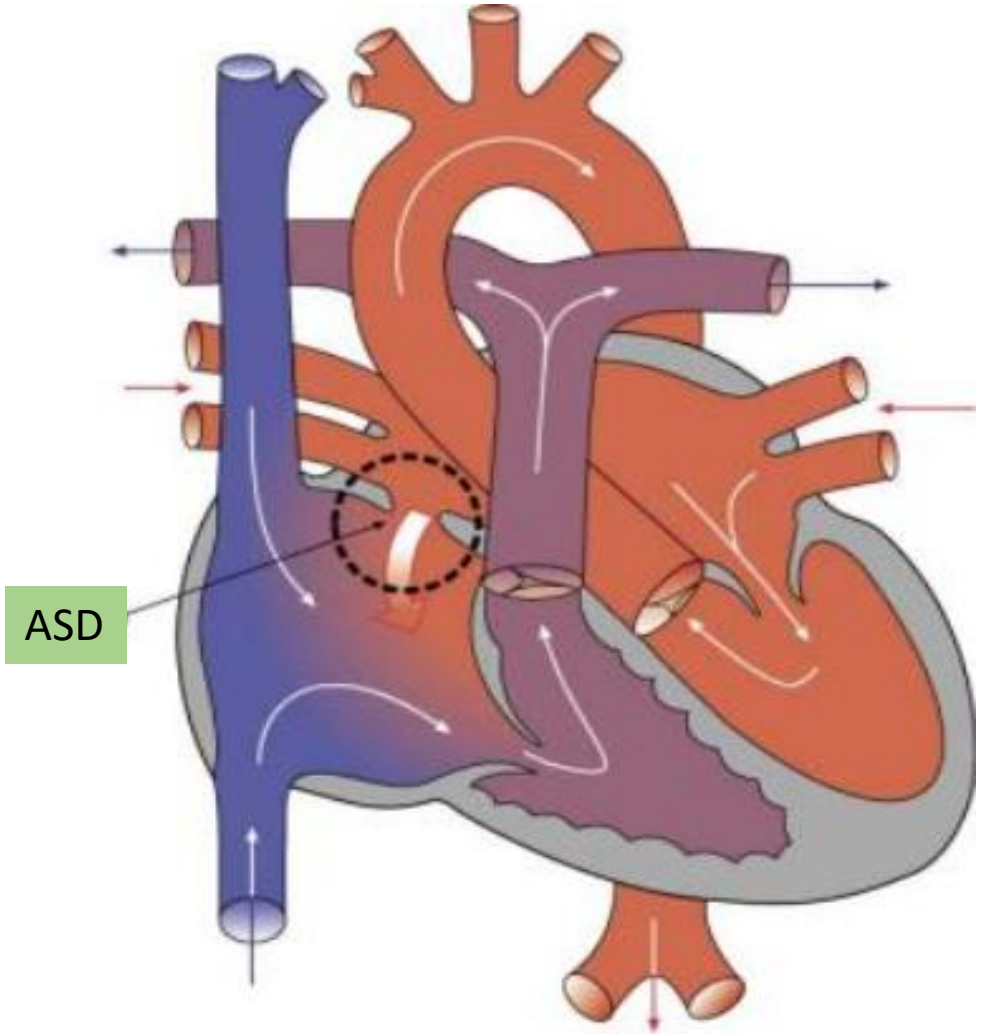
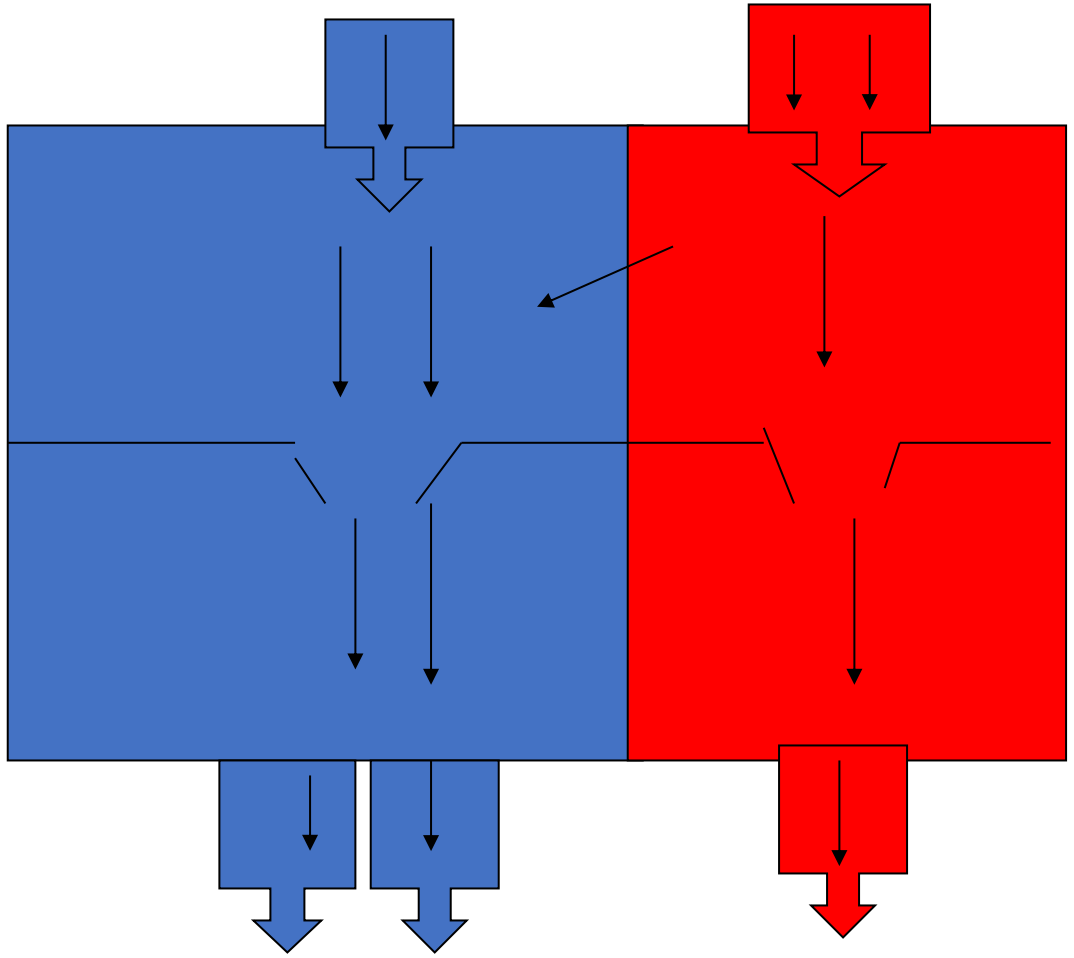
Atrial Septal Defect (ASD, 20-25%)





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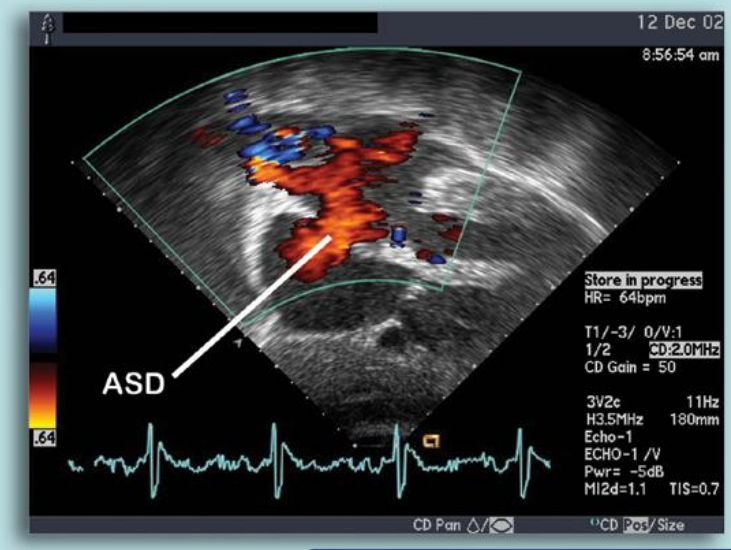
Atrial Septal Defect (ASD, 20-25%)

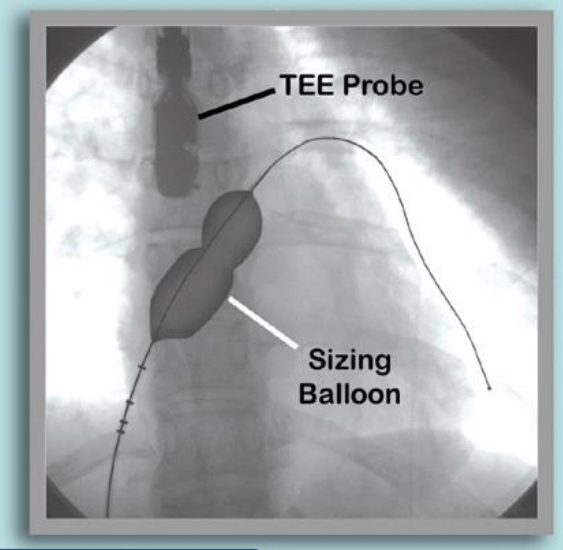
- Left to right shunt (acyanotic)
- Types: primum, secundum, sinus venosus
- SM with split S2, rarely failure to thrive
- Right heart overload (RA/RV)
- Late presentation
- Arrhythmias/Stroke/PH

- **Conservative treatment**
  - spontaneous closure/no treatment
  - rarely, diuretics
- **Intervention/Surgical**
  - Device closure
  - Surgical patch closure

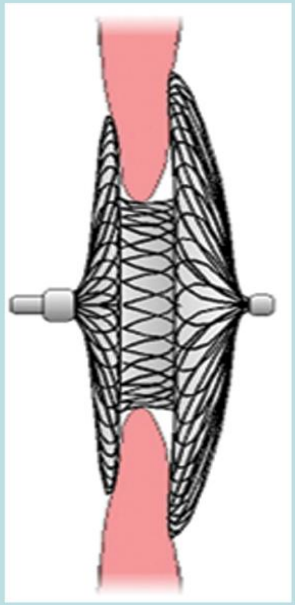
- **Follow up**
  - pericardial effusion
  - residual shunt
  - discharge
  - no medication

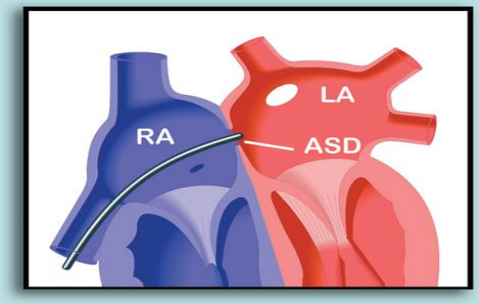
Atrial Septal Defect (ASD, 20-25%)

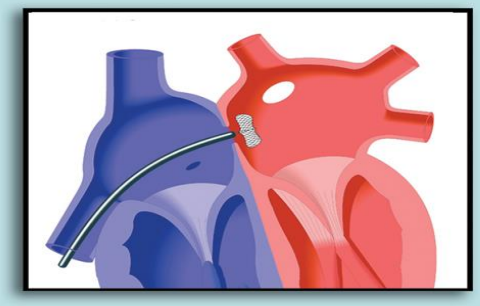


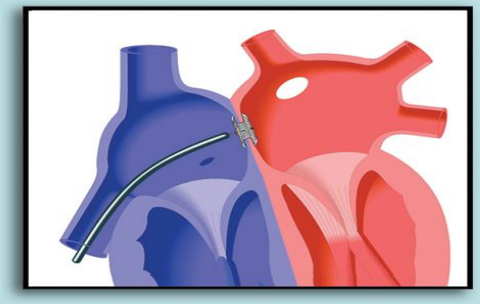


Age 2- 4 years; 20 kg



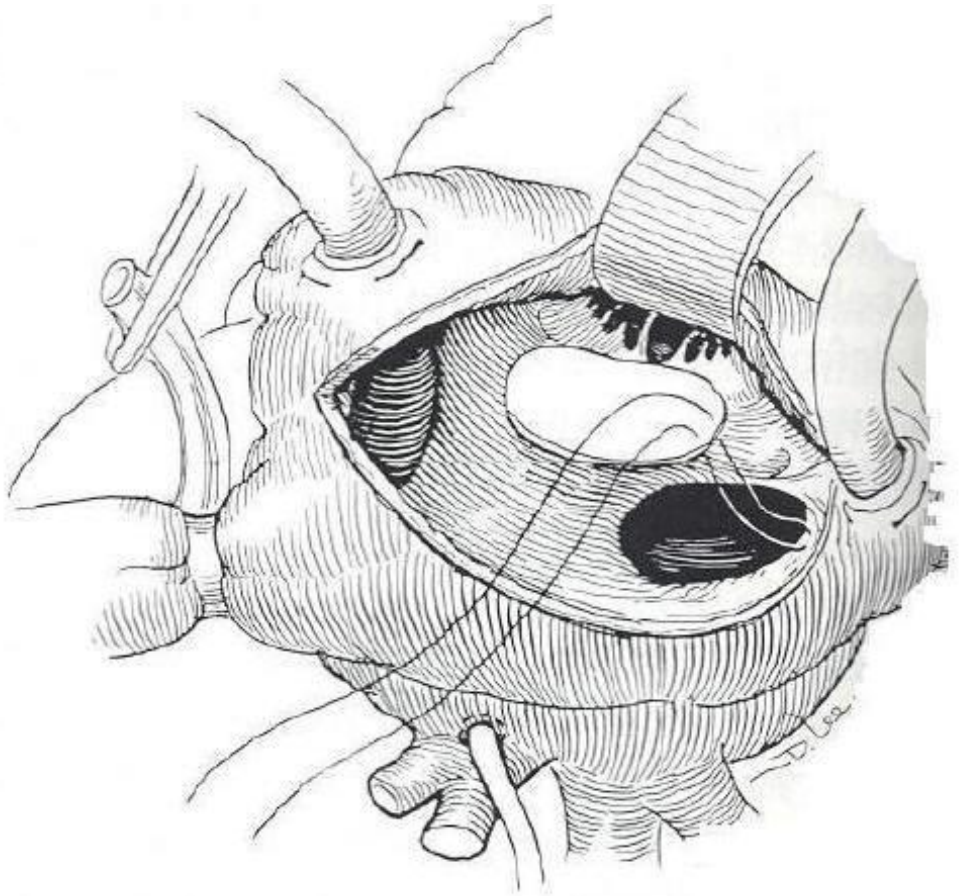






Left: Amplatzer Device

Right: Clockwise
- Catheter
- Device in position
- Device secured & released



SURGICAL ASD PATCH CLOSURE

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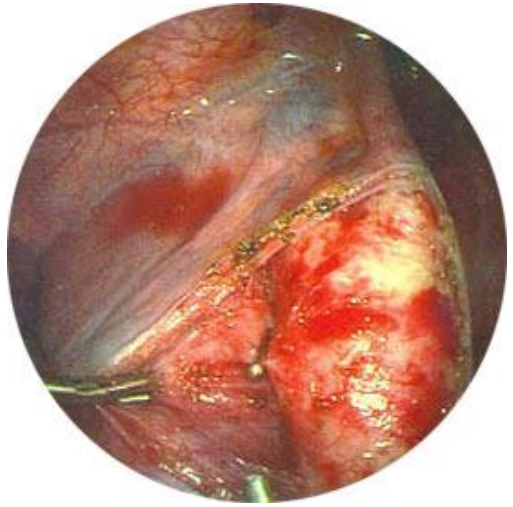
Surgical Management

Beating Heart



versus

"Palliative Surgery"

versus

"Corrective Surgery"

Cardio-pulmonary Bypass (Heart Lung Machine)

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Growing up with CHD, Transition, ACHD

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Question Time

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1. An infant is being prepared for surgical repair of Ventricular Septal Defect (VSD). Which of the following problems will be prevented by closing the defect?

a. Ventricular Arrhythmias

b. Heart Block

c. Failure to thrive √

d. Respiratory Alkalosis

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2. You are preparing to administer Ibuprofen to an infant with a persistent patent ductus arteriosus (PDA). The mother of the baby asks why the medication is being given to her baby. What is the best response?

a. Your baby needs help clearing the extra fluid from the lungs

b. Your baby needs this drug because it interferes with substances that keep the PDA open √

c. This drug is a non-steroidal anti-inflammatory drug, so it will help control your baby's pain

d. This drug will the baby's heart contract with stronger force

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3. The paediatric team is caring for a child with congenital heart disease. When planning care, monitoring for which of the following complications will be included in the plan of care?

a. Bradycardia and hepatomegaly

b. Pulmonary hypotension and cyanosis

c. Increased pulmonary compliance and cyanosis

d. Congestive heart failure and Hypoxemia

√

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4. The paediatric team is planning the discharge for a child with Tetralogy of Fallot. Which of the following is a priority to include in the discharge instructions?

a. Provide instructions for a monthly immunoglobulin injection during RSV season	√
b. Advice family to provide low calorie feeding regime	
c. Monitor the baby's blood pressure at home	
d. Advice that heart condition is likely to spontaneously resolve	

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5. An infant has an atrial septal defect (ASD) and a systolic ejection murmur heard at the upper left sternal border. Which of the statement below is characteristic of this type of murmur?

a. Increased blood is flowing through the pulmonary valve	√
b. The murmur can be heard throughout systole	
c. Increased blood flow through the atrioventricular valves	
d. There is a thrill present	

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6. You are caring for a patient with a congenital heart defect and is reviewing fetal circulation. Which of the following statement most accurately explains the major difference between fetal circulation and the circulation established after birth?

a. Deoxygenated blood flows from fetus to placenta through umbilical vein

b. After umbilical cord is cut, foramen ovale opens to allow more blood flow to the lungs

c. Systemic vascular resistance is lower than pulmonary vascular resistance in the fetus √

d. In fetus, the ductus arteriosus diverts most blood towards the lungs

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7. During an examination of an infant with a patent ductus arteriosus (PDA), the healthcare provider should expect to observe:

a. Clubbing of fingers and toes	
b. Widening pulse pressure	√
c. Diastolic murmur	
d. Profound cyanosis	

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8. What heart condition is Turner's syndrome commonly associated with?

a. Tricuspid Atresia

b. Truncus Arteriosus

c. Coarctation of Aorta √

d. Aortic root dilatation

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9. What is the common cause of Mitral Stenosis from below diagnosis?

a. Infective endocarditis

b. Dilated Cardiomyopathy

c. Acute rheumatic fever

d. Chronic rheumatic valve disease √

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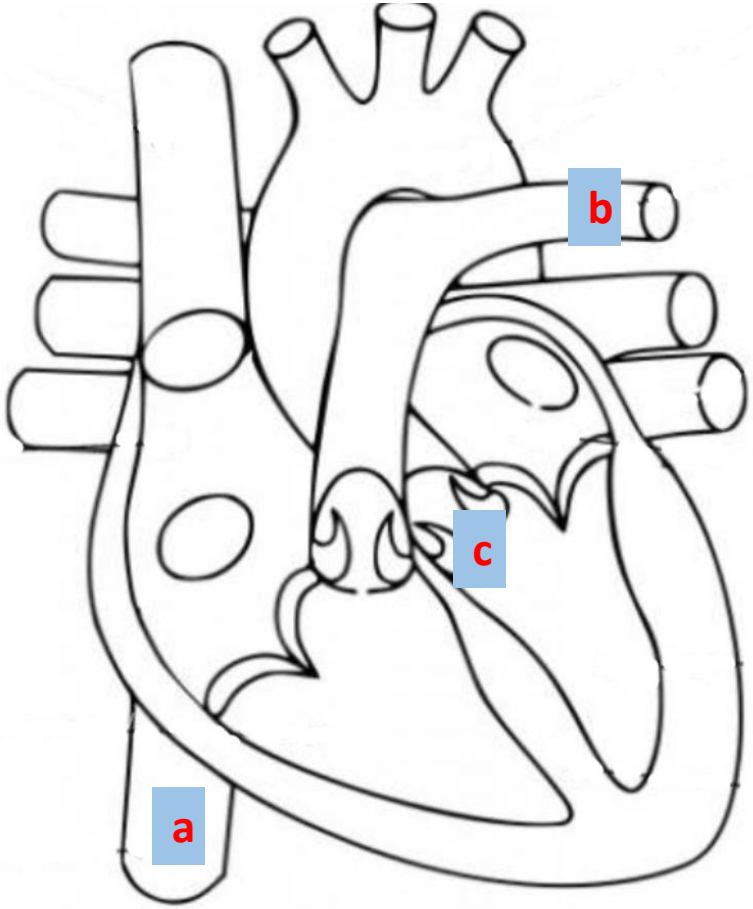
10. Select the correct answer in order of the labelled structures of the heart:



a. Superior vena cava b. Innominate artery c. Mitral valve

a. Inferior vena cava b. Left pulmonary artery c. Aortic valve √

a. Inferior vena cava b. Aorta c. Right pulmonary artery

a. Azygos vein b. Main pulmonary artery c. Pulmonary valve

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Thank You