Congenital Heart Disease

Mohammed Alghamdi, MD, FRCPC, FAAP, FACC
Associate Professor and Consultant
Pediatric Cardiology, Cardiac Science
King Fahad Cardiac Centre
King Saud University

The dr didn't give his slides and mentioned illustrated as the reference

INTRODUCTION

The most common malformation in children

- CHD ~ 0.8% of live births.
- Major CHD: CHD is typically 1 in a 100.
  - Ventricular Septal Defect: 35% The most common
  - Atrial Septal Defect: 7 %
  - Patent Ductus Ateriosus: 7 %
  - Coarctation of Aorta: 6 %
  - Tetralogy of Fallot: 6 % Most common cyanotic heart disease
  - Pulmonary valve stenosis: 6 %
  - Aortic valve stenosis: 5 %
  - D-Transposition of great arteries: 4 %

Is CHD something inherited from your parents?
Nobody can answer that, it is multifactorial, and your percentage of having it with normal parents is 1% but if one of your parents have CHD then that percentage increases to 2% other diseases like (left sided obstructive lesions: coarctation, bicyspid aortic valve) the percentage will rise to 4-5%

Congenital Heart Disease

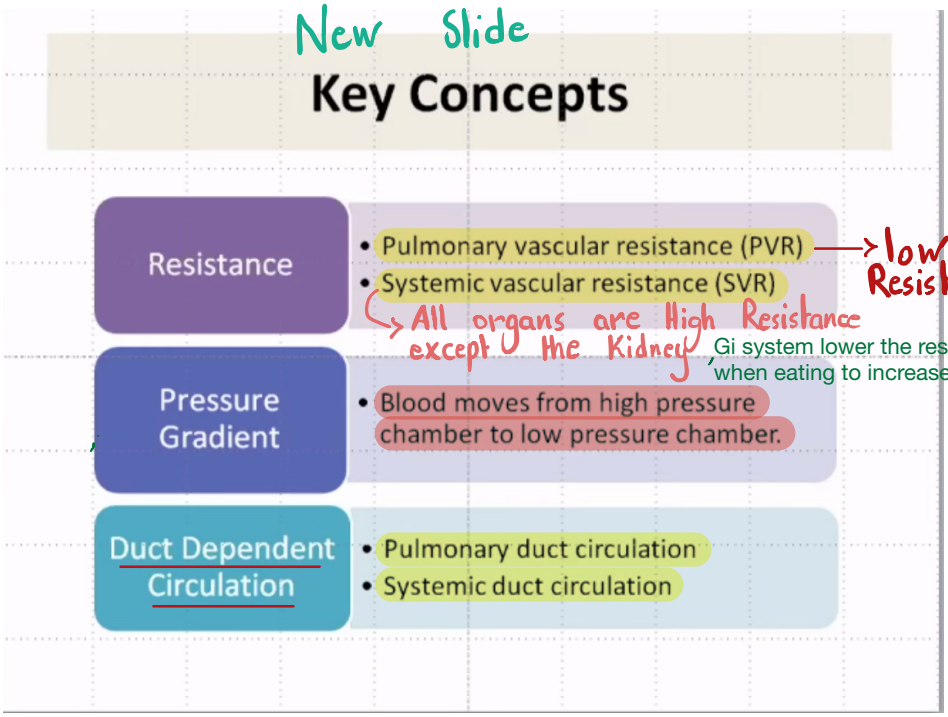
imp.

- Etiology: Mostly unknown
- Chromosomal abnormalities can cause
  - Trisomy 21: AVSD — 50% of the cases of downsyndrome have CHD, most commonly AVSD
  - Trisomy 18: VSD
  - Trismoy 13: PDA, VSD, ASD

The last 4 are above your level

  - DiGeorge Syndrome: Arch, Conotruncal abnormalities → Arch obstruction
  - Turner syndrome: Coarctation of Aorta + Bicuspid Aortic Valve
  - Williams Syndrome: Supra-aortic stenosis, PA stenosis → Pulmonary Artery Branch Stenosis
  - Noonan Syndrome: Dysplastic pulmonary valve → Also called male Turner

New Slide



1. Resistance :

- We all Know that fluids like to move from High resistance to low resistance. What are the low resistance organs in our bodies? The lungs and Kidneys, So if there is any hole in the Heart, the Blood will choose to go to the lungs. This is a Key to understand all the diseases.

3. Duct Dependant Circulation:

- Some Congenital Heart disease are called "Duct Dependant Circulation Diseases", means that if the duct Closes, the Baby will die !!, We can divide them into :

- Pulmonary duct dependant : The Forward flow of the Blood from the Heart to the lungs is very limited, therefor there should be another Pathway, which is the PDA
- Systemic duct dependant : The Forward flow from the left Ventricle to the Body, if it gets significantly Reduced by a "Critical Aortic Valve Stenosis" or "Critical Coartication", and source or way for the Blood to reach the Body has to be there.

Classification of CHD

- Divided into 2 major groups: Presence or absence of cyansosis
  - Cyanotic heart diseases.
  - Acyanotic heart diseases.
- Subdivided further according to:
  - Physical Finding
  - Chest X-ray finding
  - ECG finding
- Diagnosis is confirmed by:
  - Echo, Cardiac CT/MRI or Cardiac Catheterization.

- Best modality to diagnose CHD is **Echo** (very good for detecting intra-cardiac abnormalities because it has motion)
- **Cardiac CT/ MRI** is used to investigate something the echo can't show (detects extra-cardiac abnormalities)

(**MRI** is good for 1. Volume 2. Function 3. Extra-cardiac structure like veins and arteries)
The **CT** is the best when it comes to extra-cardiac structures but it has radiation. So we usually go for MRI

- **Cardiac catheter** is good for hemodynamic assessment measuring the pressure gradient or for interventions

KEY CONCEPTS:

- End result of L-R shunt is pulmonary **overcirculation**
- It is the mixing of red blood with blue blood that increases the volume and that increased volume goes to the pulmonary circulation

- R-L shunt end result is pulmonary **hypoperfusion**
- The blue blood mixes with the red blood so the blue blood in the right ventricle decreases leading to pulmonary hypoperfusion reducing the pulmonary flow.

- The aorta should receive pure red blood, when you have R-L shunt you have mixed blood so you have a cause of cyanosis

- in L-R shunt the aorta is receiving PURE red blood but LESS amount so it does not cause cyanosis, the right ventricle is getting more blood which will eventually go to the lung leading to **heart failure**

What defines a L-R shunt vs. R-L Shunt (blood flow)

1. **Pressure difference** (blood goes from high pressure to low pressure)
2. **Resistance difference** (the blood goes to where there is less resistance)

- you have 2 circulations the pulmonary vs the systemic resistance. The systemic circulation has the **Higher** resistance unlike the pulmonary which has a **Lower** resistance this is because of the pressure difference (systemic 120/80 vs pulmonary 20/8) so the blood will choose to go to the lungs

3. **Location of the defect**
4. **Size of the defect**
5. **Presence or abscence of valvular stenosis**

- When you have an obstructive lesion (aortic valve stenosis, pulmonary valve stenosis, coarct etc) if its in the mild to moderate or begining of the severe stage it is **Acyanotic** Heart Disease but once it reaches the critical i.e. very severe then it becomes **Cyanotic** Heart Disease

What is the difference between fetal circulation and post-natal circulation?

- the **FETAL** circulation has

1. The placenta: is a low resistance organ (it sucks blood)
2. The lungs: they are not used during fetal life, only 7% of the blood goes to the lung and has a **HIGH resistance** → 10 %

- The Right Atrium (RA) in the fetal circulation receives blood from **four** sources (SVC, IVC, coronary sinus, Placenta) whereas the adult receives it from three only (SVC, IVC, Coronary sinus) therefore the pressure in the RA is higher in the fetus. The Left atrium pressure is lower because as we mentioned the fetus only uses 7% of the blood to his lungs so the left atrium receives less blood. And since the blood moves from high pressure (RA) to low pressure (LA) the **foramen ovale in the fetus** opens from RIGHT TO LEFT. Once the fetus is born (post natal circulation) the lungs resistance is low and the lung receives 100% of the blood so now the systemic circulation has a higher resistance and the foramen ovale opens from LEFT TO RIGHT.

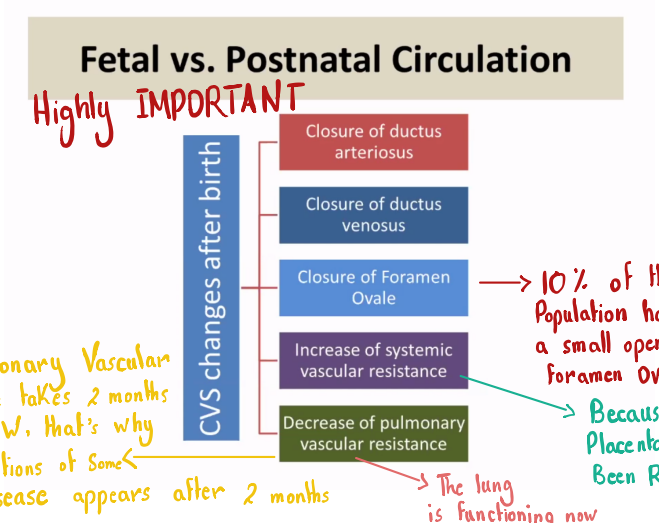
→ Three , the doctor didn't mention the Coronary Sinus

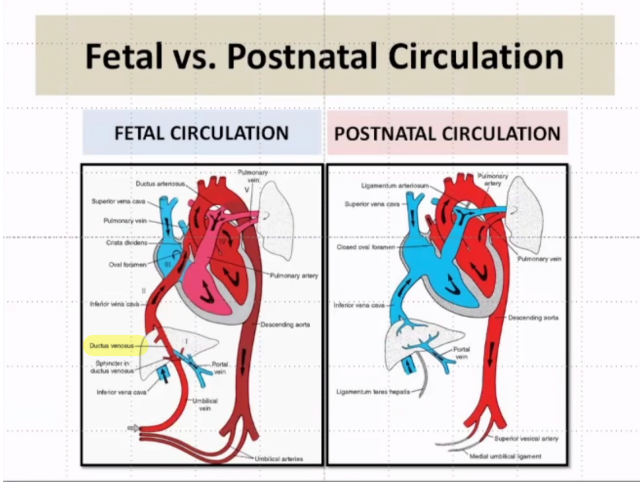
- We previously mentioned that the fetal lung only recieves 7% of the blood, where does the rest of the blood go? It goes to the ductus arteriosus. After birth the baby cries and there will be a release of prostaglandins and prostacyclins that will eventually physiologically and anatomically closes the PDA initially in the first few to 24h and the. Completely in the next 48h to 7 days.
This goes back to two things 1. The lung is now using 100% of the blood and 2. The systemic resistance is now higher (the placenta is out which was the reason the systemic resistance was initially low in the fetal life)

At birth baby cry and breath>lung expands%>less resistance

IMP QUESTION, you might get in the exam:
What is the difference between fetal and postnatal circulation?
CVS changes after birth:

1. **Closure of ductus arteriosus**
2. **Closure of ductus venosus**
3. **Closure of foramen ovale** → Because LA Pressure is Higer than RA
4. **Systemic resistance increases in the post natal circulation because the placenta is out**
5. **Pulmonary circulation resistance decreases because the lungs opened up**





Classification of CHD

Cyanotic Heart Disease

- Decreased pulmonary flow:
  - Tetralogy of Fallot
  - Tricuspid atresia Very complicated subject with multiple types
  - Other univentricular heart with pulmonary stenosis.
- Increased pulmonary flow:
  - Transposition of great arteries
  - Total anomalous pulmonary venous return.

  - The degree of cyanosis depends on the degree of pulmonary flow

Acyanotic Heart Disease

- Left – Right shunt lesions:
  - Ventricular septal defect
  - Atrial Septal Defect
  - Atrio-ventricular Septal Defect
  - Patent Ductus Arteriosus
- Obstructive lesions:
  - Aortic stenosis
  - Pulmonary valve stenosis
  - Coarctation of Aorta

If it critical it will present with cyanosis

*Left to Right Shunt:

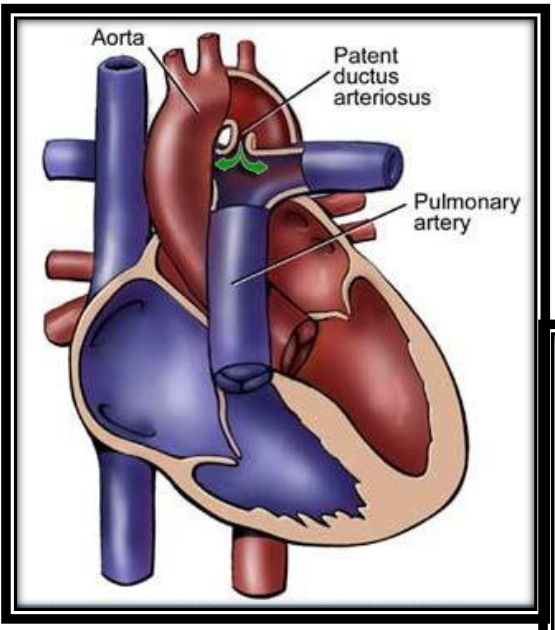
- means that a part of the Blood in left chambers will mix with blood in the right chambers, so the lung will receive mixed blood (Oxygenated and non Oxygenated).
- The Body will receive pure red blood, so no cyanosis.

Acyanotic Heart Disease
Left – to- Right Shunt lesions

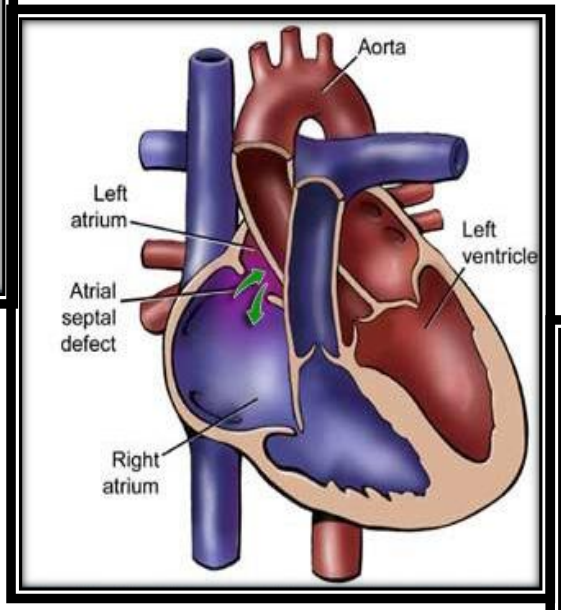
When we classify shunts we have 3 segments:

1. At the level of the hearts atria (ASD)
2. At the level of the hearts ventricles (VSD)
3. At the level of the great arteries (PDA)

Left – to- Right Shunt lesions

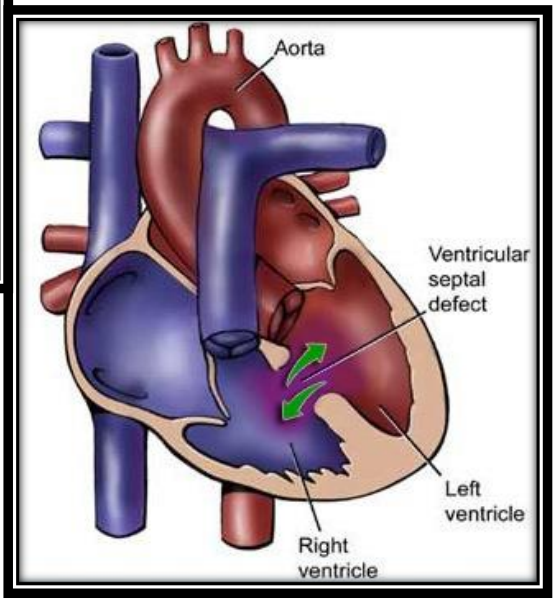


PDA



ASD

VSD



VSD

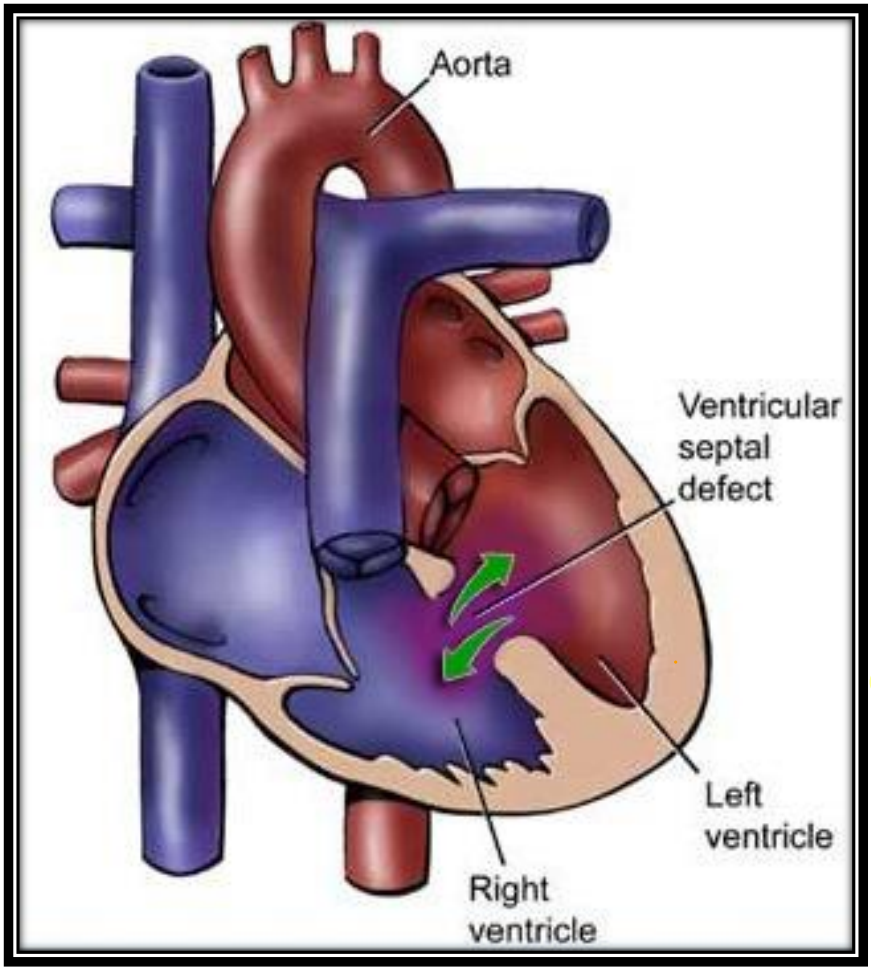
The main Problem in VSD is: lung Congestion

- The LV pressure is 120/8 No elastic in diastolic recoil like aorta

The RV pressure is 20-25/8
So the pressure difference is only in all of systole but no difference in diastole.
Thats why VSD causes a **pansystolic** murmur

When it comes to resistanc: the blood moving from the LV has 2 options either it goes to the aorta (**high resistance**) or through the VSD to the RV to the pulmonary which is (**low resistance**) the blood will choose the lower resistance path thats why it will end up being a left to right shunt eventually leading to pulmonary over circulation leading to conjestive heart failure.

- if we had the same dx but the patient had **severe pulmonary stenosis** then the resistance changes and becomes higher than the aorta and then it will become a cyanotic disease not an acyanotic one cuz the blood follows the less resistant path



Do you expect a patient with VSD to have heart failure from day one? Why?

No, it happens after 2-3 months.

The size,site, pressure didn't change, **But** the resistance did.. when you are born you have high resistance pulmonary circulation and for it to decrease you need 4 to 6 to 8 weeks to become low resistance.

Difference between VSD HF and ischemic HF

Assuming the pt had Mitral valve regurgitation the blood will go back to the LV to the pulmonary veins to the lungs causing venous pulmonary congestion so they have pump failure because of the back flow unlike VSD which is a forward pressure.

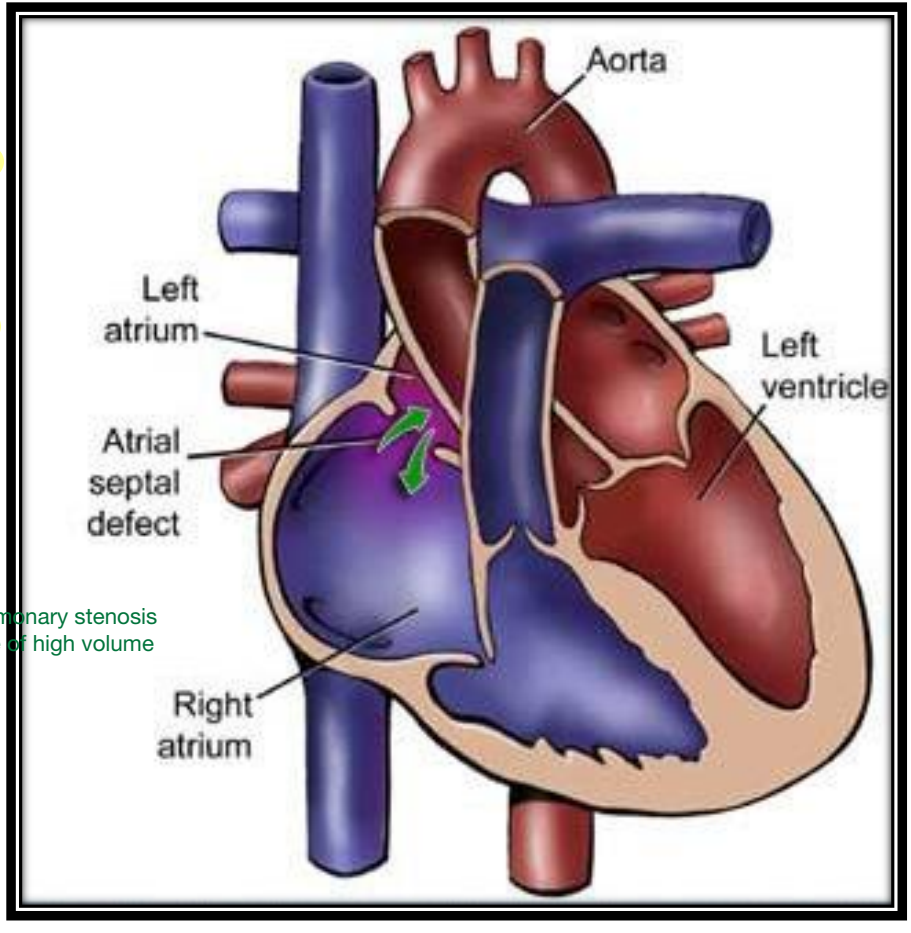
The normal ratio is 1:1,but here the ratio will be 2:1
So for example 4 litres of blood will go to the lung and 2 litres to the system

ASD

The mean LA pressure is 6 to 8 while the RA pressure is 4 to 6 so the LA has a higher pressure by little difference.

Therefore the LA will drain blood to the RA which will go the the RV eventually draining into the lungs and causing pulmonary overcirculation **BUT without** getting exposed to high pressure such as the VSD thats is why ASD doesn't cause eisenmenger. Syndrome as fast.

Here the murmurs resemble pulmonary stenosis
The valve is normal but because of high volume



ASD doesnt cause symptoms of CHF it is usually subtle, silent no body knows about it and usually shows up during child bearing period, onset of pregnancy shoiwng signs of CHF, SOB

PDA

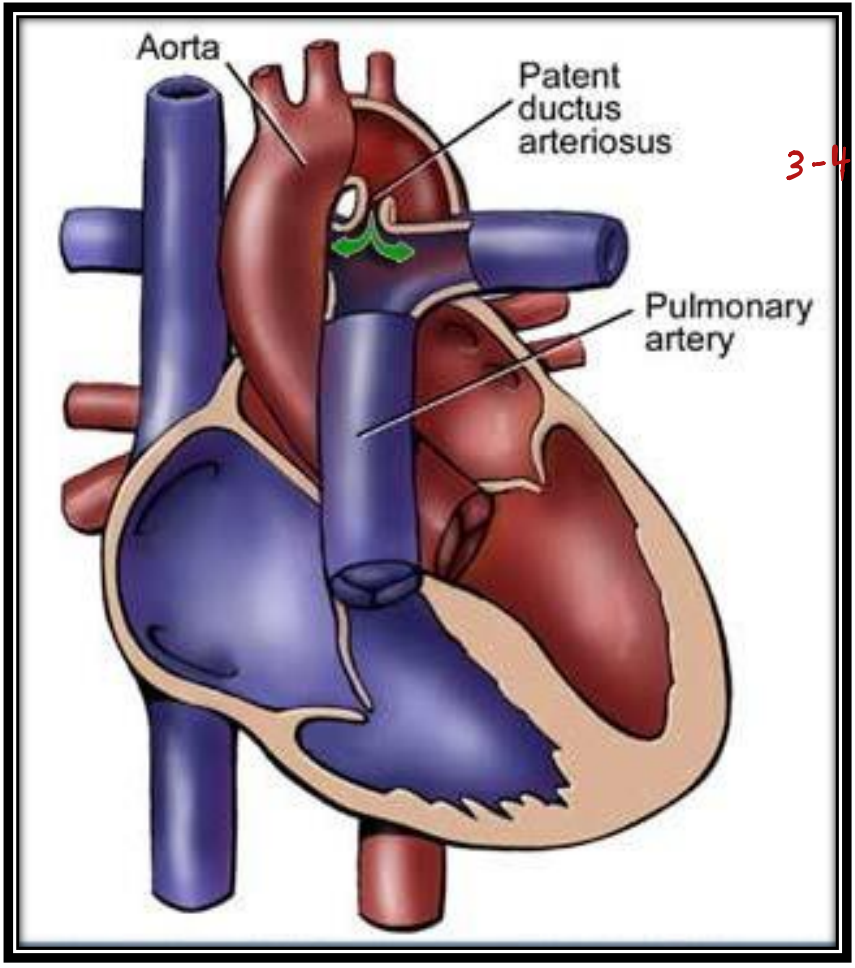
* PDA : Shunts during Both Systole and Diastole
* VSD : Shunts only in Systole

- Aorta is high resistance and the pulmonary is low resistance. So the blood will go from the aorta to the pulmonary
- The aorta's pressure is 120/80 and vs the pulmonary artery pressure 20/5 so the pressure difference is in both diastole and systole that is ehy it gives you **machinery murmur** (Continuous)

- the aorta pressure is 120/80 and the LV is 120/8 why?

Because of the elastic recoil in the aorta, and the aorta is directly related to high resistance organs while the LV is protected by a valve if the the LVks pressure was 180/20 then the LA wouldn't be able to drain into the LV

120/80



3-4

A 3-4mm PDA is considered large whereas a 4mm VSD is considered ok because of the pressure difference

A large ASD 12-18mm might even reach 30

A large VSD 6-8 mm

The reason for this is the location

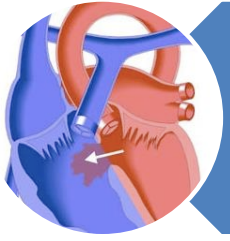
PATHOPHYSIOLOGY: L-R SHUNT

A normal Qp:Qs ratio is 1:1

- L-R shunt toward pulmonary circulation.
  - Increased Qp:Qs ratio
    - Increased cardiac output to the pulmonary circulation (Qp)
    - Reduced of cardiac output to the systemic circulation (Qs)

PATHOPHYSIOLOGY: L-R SHUNT

VSD



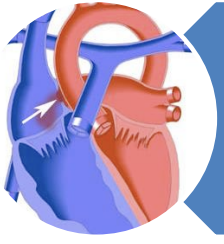
L-R shunt at ventricular level:

Dilated LA and LV

Enlarged pulmonary arteries

* The Blood doesn't stay in the RV for a long Period.

ASD

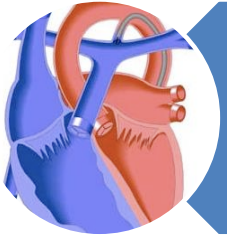


L-R shunt at atrial level:

Dilated RA and RV

Enlarged pulmonary arteries

PDA

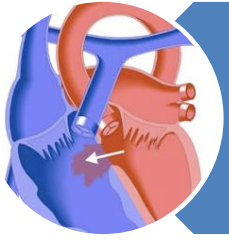


L-R shunt at artery level:

Dilated LA and LV

SYMPTOM: L-R SHUNT

VSD

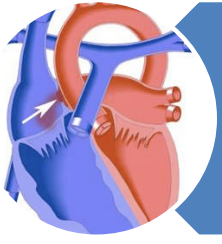


Small VSD: Asymptomatic

There is no change in the pressure difference or the resistance the thing that changes was the size if you have a small VSD you have a normal life.

Moderate to large VSD: CHF (Congested lung)

ASD



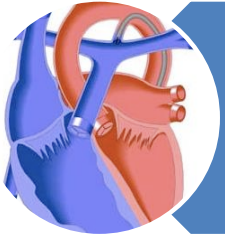
Usually asymptomatic

Samall medium or even large are asymptomatic because the pressure increased.

Older children: Activity related SOB & Fatigability

Rare: CHF , FTT

PDA



Small PDA: Asymptomatic

Moderate to large PDA: CHF

CONGESTIVE HEART FAILURE

SYMPTOMS IMP

- Diaphoresis
- Poor feeding
- Failure to thrive
- Shortness of breath
- Recurrent chest infection
- Exercise intolerance

6

Good media for the pathogens to grow

SIGNS IMP

- Tachypnia
- Tachycardia
- Cardiomegally
- Hepatomegally
- Active precordium

Tetrad of heart failure

IMPORTANT

The liver is the JVP of pediatrics

One of the best signs that the VSD is significant

you can feel the heart pumping كأن واحد مرتاع و خايف

يجري

VSD

- No symptom during neonatal period
  - due to high pulmonary vascular resistance

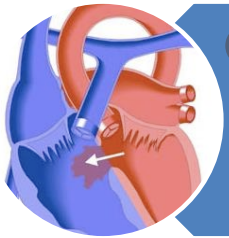
Even no murmurs

Lungs need 4-8 weeks after birth to decrease the resistance

- Symptoms of CHF started ~ 2/12 of age
  - diaphoresis, poor feeding, and failure to thrive.
  - shortness of breath, recurrent chest infection.
  - exercise intolerance.

EXAMINATION: L-R SHUNT

VSD

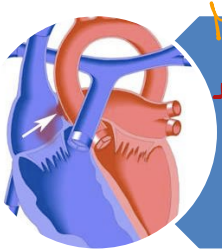


Pan

- Holosystolic murmur
- Large: mid-diastolic murmur
- Small muscular: ejection systolic murmur

صب مهمة !!

ASD

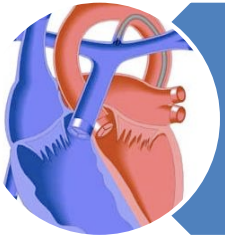


MCQ IMPORTANT

- Fixed Widely splitted second heart sound
- \+ Ejection systolic murmur
- Large: mid-diastolic murmur

Because the RV is pumping more blood to the pulmonary artery so it resembles a pulmonary valve stenosis

PDA



- Small PDA: Silent
- Large PDA: Continuous "machinery" murmur

Heard under the Left Clavicle

- Large PDA: Widened pulse pressure

Means high pressure difference between systole and diastole.e.g:110/40

→ Can be assessed by Clincal Examination, you will find the Pulse to be "Pounding" or "Strong"

INVESTIGATION: L-R SHUNT

- Diagnosis:
  - Chest X-ray: To look at the pulmonary congestion
    - Increased pulmonary vascular marking Congestion
    - +/- cardiomegally
  - ECG: You don't have to know how to read ECG or Echo you will have scenario only
    - Small lesion: Normal
    - Mod to large: chambers enlargement
  - ECHO: Most imp investigation
    - Confirm Diagnosis You can see the cardiomegaly
  - Cardiac Cath: not required for Dx

4

- We rarely use it expect of we want to assess hemodynamics especially if we suspect a pt has eisenmenger syndrome and we want to check if they are operable
- Most PDA are now closed with a cath except for premature babies.
- Most ASDs are closed with caths nowadays
- VSDs most common mode of closure is surgical

MEDICAL Rx: L-R SHUNT

Medical Mgx

- Anti-congestive therapy:
  - Diuretics · Lasix +/x spironolactone
  - ~~Digoxin~~. We use digoxin in pump failure only it's an ionotropic agent.here it's not useful
  - After load reducing agents
    - captopril (Mainly to Balance the Potassium disturbance caused by the diuretics)
- Nutritional support
- ASD: usually no medication needed

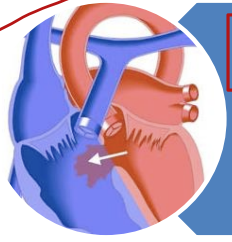
- One of the most common mistakes when someone comes with ASD or VSD is to ask them to reduce fluids if anything you should ask them to give their baby more milk because it is their only source of calories and you dont want to make their failure to thrive worse. You will treat the excess fluids with diuretics but never ask them to give their baby less milk

↑ Their milk
+
↑ Diuretics

INTERVENSION: L-R SHUNT

By default Surgical , In Rare cases "Cath"

VSD

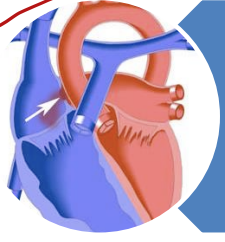


Surgical closure 4-8 months

Some types in older children can be closed by device via CATH.

The opposite

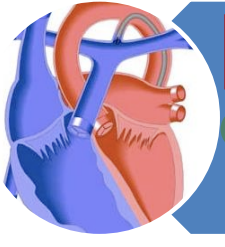
ASD



Most closed by device via CATH around 3-6 years

Some types need surgical closure.

PDA



Most closed by device via CATH

Surgery needed in premature baby and symptomtic neonate less than 6 kg.

PROGNOSIS: L-R SHUNT

When the pt is 6-9 months has HF to pulmonary circulation there is too much blood in the lungs, this will expose the lungs to high pressure for a very long time: so they will have evolving hypertension which starts as reversible then they will have fibrosis and thickening which eventually becomes irreversible (eisenmenger's syndrome)

- Untreated VSD and PDA beyond infancy: It is a long standing significant L-R shunt ultimately leading from reversible to irreversible changes to the pulmonary vascular bed which turns it from reversible to irreversible hypertension
  - Eisenmenger's syndrome = Death certificate

Their only chance Would be Heart-lung Transplant

- Signs for you to start worrying
  1. Sign and symptom of CHF will disappear
  2. Patient will become cyanotic
     - R-L shunt reversed shunt.
  3. Clubbing
  4. Heart murmur disappears & a LOUD second heart sound appears

Because of the fibroses the lung will no longer accept the blood coming from the RV, so instead of heart failure it will become cyanosis, the RV pressure will elevate until it equalizes with the LV, so the RV will shift to the LV and the patient will become cyanotic so the symptoms of HF will disappear. The heart murmur wil also disappear becuse the heart pressure equalized.

- If the mother comes to you telling you my child is now ok and he's no longer showing signs of HF then there are two options either there was closure of VSD and you become happy it became eisenmenger and the baby starts to become cyanotic with **clubbing** in which case you start worrying.
- You also listen to a loud second heart sound which is a sign of pulmonary hypertension.

PROGNOSIS: L-R SHUNT

- Untreated ASD:
  - complication happened during adult Life:
    - Eisenmenger's syndrome
    - Atrial arrhythmias
    - Paradoxical embolism (rare)

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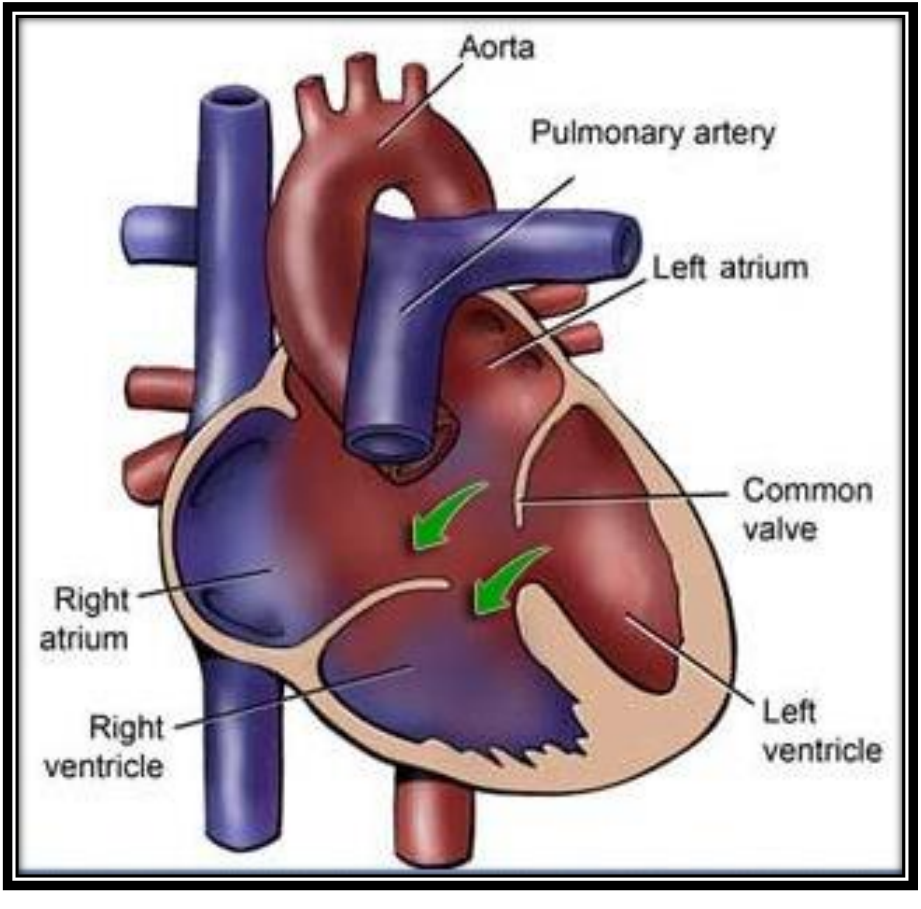
AVSD

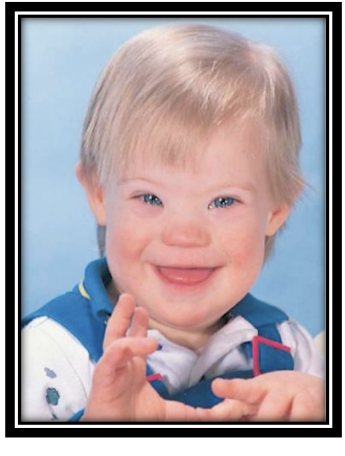
Atrio-Ventricular Septal Defect



AVSD

The most important thing here that I want to know that it's associated with Down syndrome,ECG will show left axis deviation and if it's complete there is no rule of cath in treatment only Surgery

- Incidence: 4 % of all CHD
  - Associated with Down Syndrome (50%)
- Divided into:
  - Complete AVSD
    - ASD primum/ inlet VSD / common AV valve
    - Balanced vs. Unbalanced AVSD
  - Partial AVSD
    - ASD primum
    - No VSD



AVSD

* It will cause CHF similar to VSD + little Cyanosis

- Pathophysiology:
  - Similar to VSD and ASD
    - left to right shunt across the atrial level
    - Left to right shunt at and ventricular level
    - In addition: AV valve regurgitation
  - Significant L-R shunting:
    - Pulmonary over-circulation
    - Increase Qp:Qs ratio.

AVSD

- Clinical Features:
  - Usually asymptomatic at neonatal period
    - Due to high pulmonary vascular resistance
    - Baby may have slightly lower oxygen saturation
  - Symptoms of CHF started at few months of age
    - Diaphoresis
    - Poor feeding
    - Failure to thrive.
    - Shortness of breath
    - Recurrent chest infection.
    - Exercise intolerance.

Exactly the same as ASD and VSD

AVSD

- Clinical Features:
  - Physical Examination:
    - Feature of Down Syndrome
    - Tachypnia
    - Tachycardia
    - Active precordium
    - Murmur:
    - Pan-systolic (holosystolic) murmur
    - Hepatomegaly

AVSD

- Diagnosis:
  - Chest X-ray:
    - Increased pulmonary vascular marking
    - Cardiomegaly
  - ECG: Imp
    - Left Axis deviation with RVH is very suggestive of AVSD
  - ECHO:
    - Confirm Diagnosis
  - Cardiac Cath: not required for Dx

AVSD

- Treatment:
  - Medical Rx:
    - Anti-congestive therapy:
    - Nutritional suppor
  - Surgical closure for complete VSD:
    - Usually done before 6 months of age to ovoid development of Eisenmenger's syndrome.
      - Balanced AVSD: Biventricular repair
      - Unbalanced AVSD: may need single ventricular repair

Only Surgical and there is No role for Cath

They develop Esemngier Faster !!

Acyanotic Heart Disease
obstructive lesion

Coarctation of Aorta (CoA)

- There is a narrowing in the aortic arch, whenever the narrowing gets more severe there is no blood flow to the lower body, there will be ischemia and acidosis.

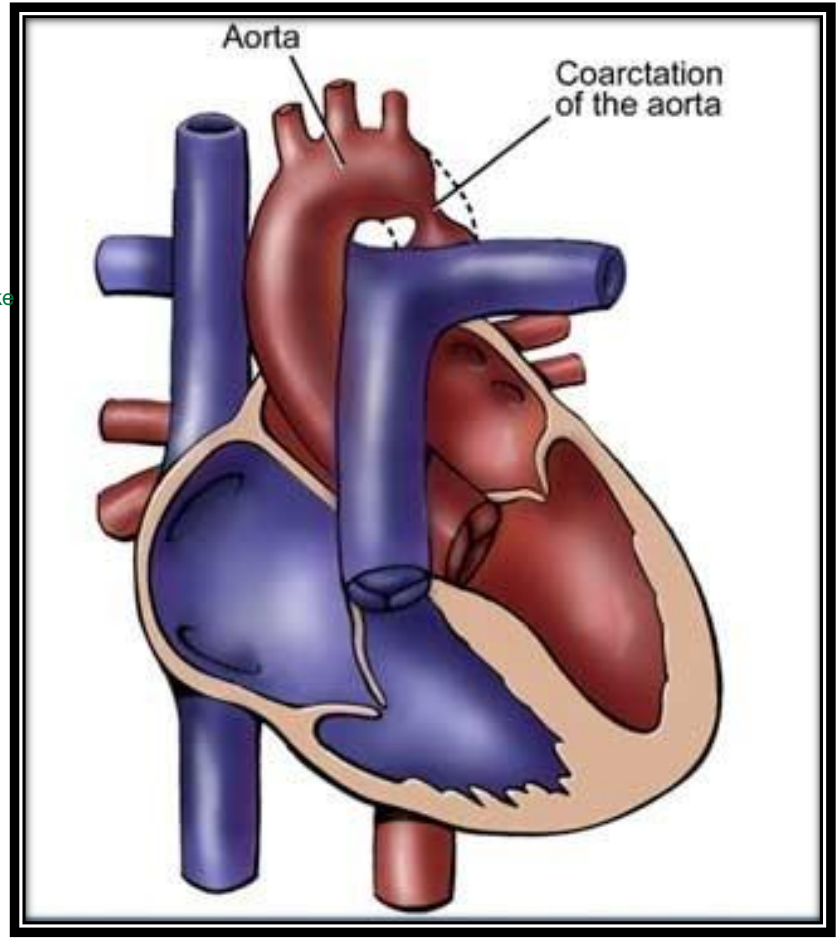
- the LV has no way to come out which leads all the blood to be directed to the brain leading to brain hemorrhage Hemorrhagic stroke

- so the degree if stenosis could be so severe tha we need to open a source of blood for the lower body

so we give prostaglandins to have PDA. Critical coartication

Because this is considered a **systemic duct dependent lesion**

Unlike TOF wich is considered a **pulmonary duct dependent lesion**



Patient will have secondary HTN.
Causes of secondary HTN:
1-Kidney disease(most common).
2-heart.like coartication.
3-endocrine disease.

Coarctation of Aorta (CoA)

- Incidence: 5-7 % of all CHD
  - Associated with Turner syndrome in female
  - Arch interruption: seen in DiGeorge syndrom
- Can be: Discrete or Diffuse

If small

- Can be mild to severe

PATHOPHYSIOLOGY: CoA

CRTICAL CoA

- Spontaneous PDA closure
  - Obstruction of blood flow to distal arch
  - Hypotension and Shock
  - Acute increase of LV after-load
  - LV dysfunction
- "DUCT DEPENDENT CHD"

If you have critical congenital heart disease you have to open the duct, in this situation you need it to be a right to left shunt. By prostaglandins

MILD CoA

- Collateral vessels develop overtime
- Flow maintained between proximal and distal aorta
- Present later on life :

In young Adults and adolscent

Appears in adult life as an Unexplained Hypertension

"Their Blood Pressure in the Upper body is Higher than the lower Body"

"Radio-femoral Delay"

CLINICAL PRESENTATION: CoA

CRTICAL CoA

- Presented 2-3 wks of life
  - Sign of CHF
  - Circulatory collapse
  - Shock Cardiogenic
  - Death

- **"DUCT DEPENDENT CHD"**

They usually Present at 1-2 weeks with Cardiogenic Shock and Acidosis.
Why ? Because the duct has Closed !!
What would you do ? Open the duct Prostaglandin then Surgery

MILD CoA

- Present later on life
  - Murmur
  - Chronic hypertension
  - Headache
  - Fatigue
  - Stroke
    - » Rupture cerebral aneurysm

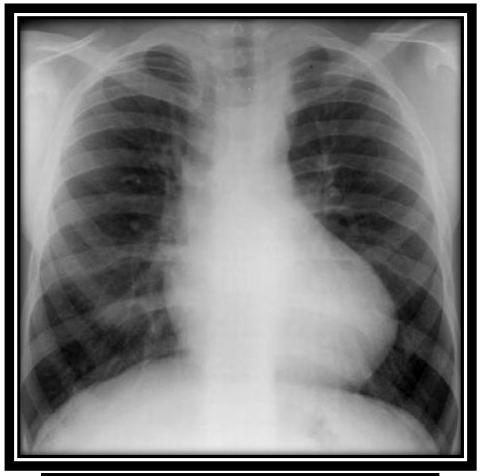
On examination they will have weak femoral pulse,low blood pressure in the lower side of the body.normally lower side of the body has higher pressure because of gravity and the vessels is more muscular.

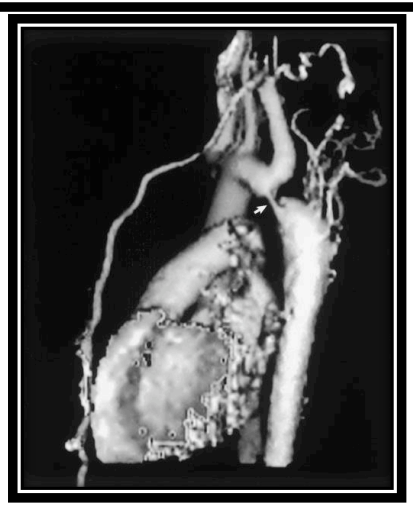
Clinical Features: CoA

- Physical Examination:
  - Differential cyanosis (severe CoA in newborn)
  - Signs of cardiac shock
  - Reduced or absent femoral pulses
  - Brachio- femoral delay → In MILD Only
  - BP in lower limb lower than upper Limb BP → Normally its the Opposite
  - Murmur:
    - Ejection systolic murmur at the back
    - Continues murmur due to collateral at the back

DIAGNOSIS: CoA

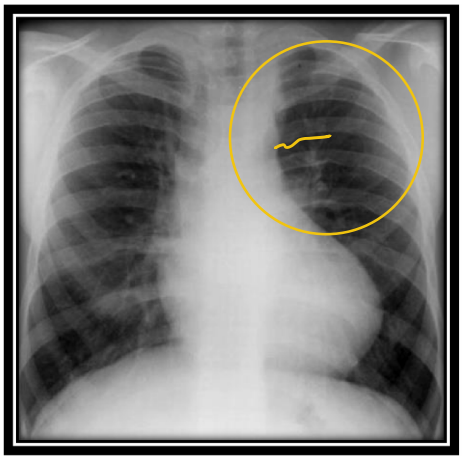
- Chest X-ray:
  - Cardiomegaly
  - Prominent aortic knob
  - Rib notching
    - » Due to d of collateral vessels
    - » Rarely seen before age of 10 years
- ECG:
  - Neonate: RV hypertrophy
  - Older children: LV hypertrophy
- ECHO:
  - Usually will establish the diagnosis
- May need Cardiac CT /MRI



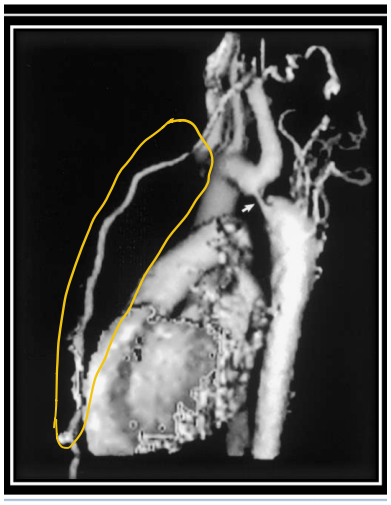


IMPORTANT Question that might come in your exam :

- A 9 Years old Pateint came to your Clinic with Hypertension, We did X-Ray and these are the Results :



* There is Rib notching caused by engourgment of the intercostal arteries because they are a part of the Collateralls.
* This is very rare .



* If you do CT scan for them, you will see the Big Collaterals.
* BTW, CT scan gives you the best delination of the Vascular Anatomy (Outside the Heart).
* But for (Intracardiac Anatomy), Echo is the Best.

TREATMENT: CoA

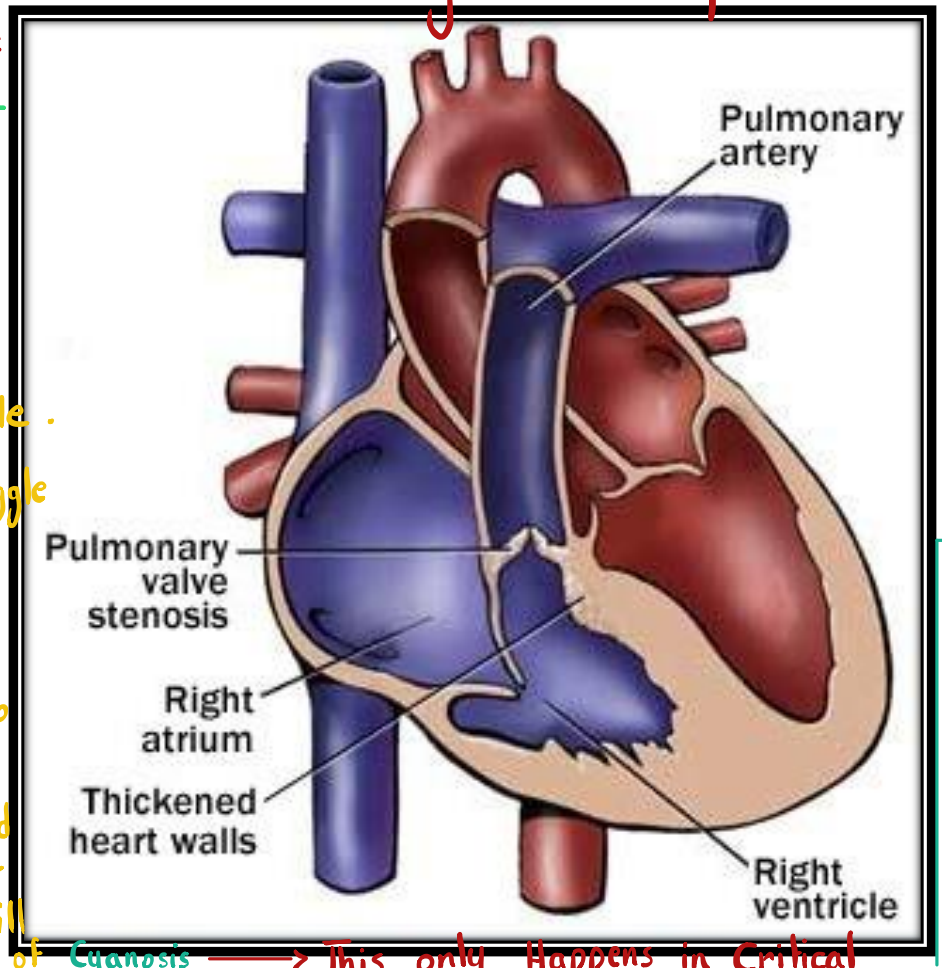
- Critical CoA
  - "Duct Dependent CHD"
    - Prostaglandin E2 to keep PDA open
- Surgery is the primary intervension
  - Depends on the site:
    - Discrete → Thoracotomy
    - Extensive → Median Sternotomy
- Trans-catheter balloon angioplasty +/- stent:
  - Recurrent CoA
  - Primary intervention: Discrete CoA in older children
    - For Late Presentation 3 - 6 years

Pulmonary Valve Stenosis

Pulmonary Duct Dependant

*Divided into:
Mild - Moderate - Severe - Critical

*Critical:
- The valve will be like a Pinhole.
- The RV will struggle to push the Blood through th PV, So the RA will Push the Blood to the LA through Foramen Ovale, and there will mixing of Blood, so they will have some degree of Cyanosis ⟶ This only Happens in Critical



*We can give them Prostaglandins to open the PDA as an alternative Pathway and a Temporarliy Tx.

*Then we can do Ballon opening for the PV.

*Mild and Moderate PV stenosis will live Normal lives without ang intervention.

*What determenes the degree of stenosis? The amount of Blood Going to the lungs.

Didn't talk about it

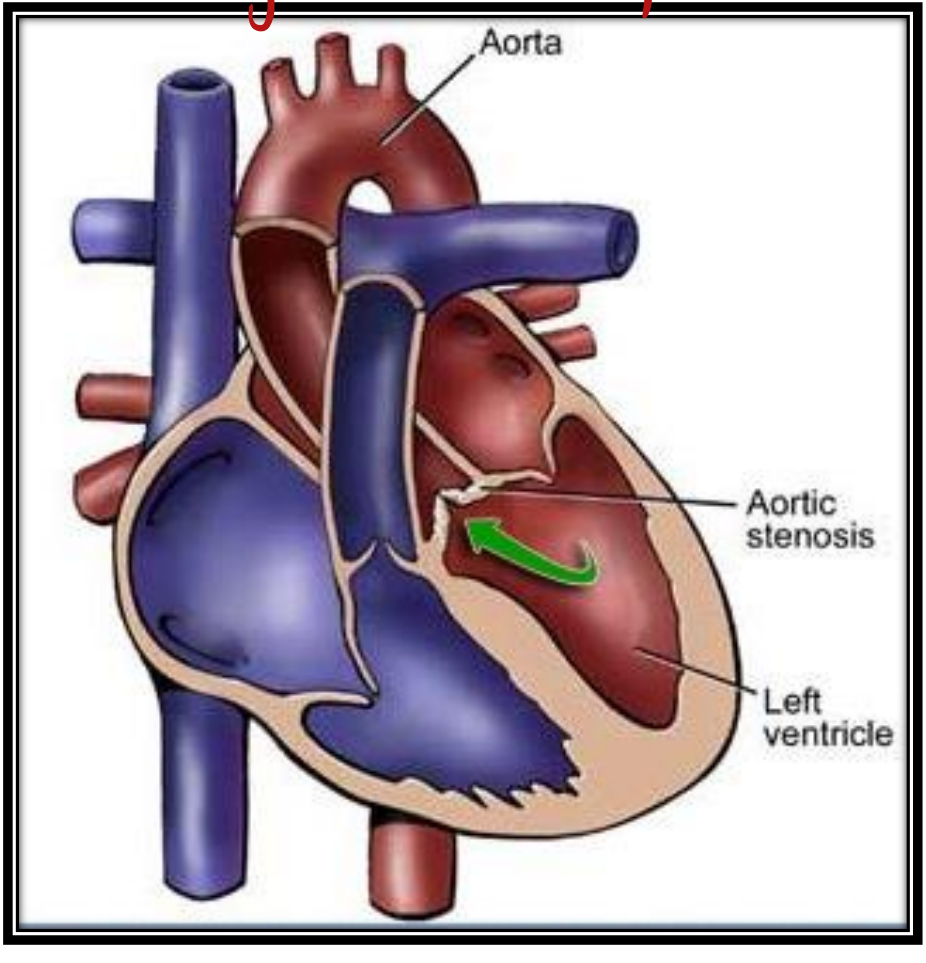
Aortic Valve Stenosis

Systemic Duct Dependant

*Divided into:
Mild - Moderate - Severe - Critical

*One of the Worst conditions that a child can Have, Because there is no Solution (Only mechanical Valve can work for them)

So they will need Anticoagulation for life !!

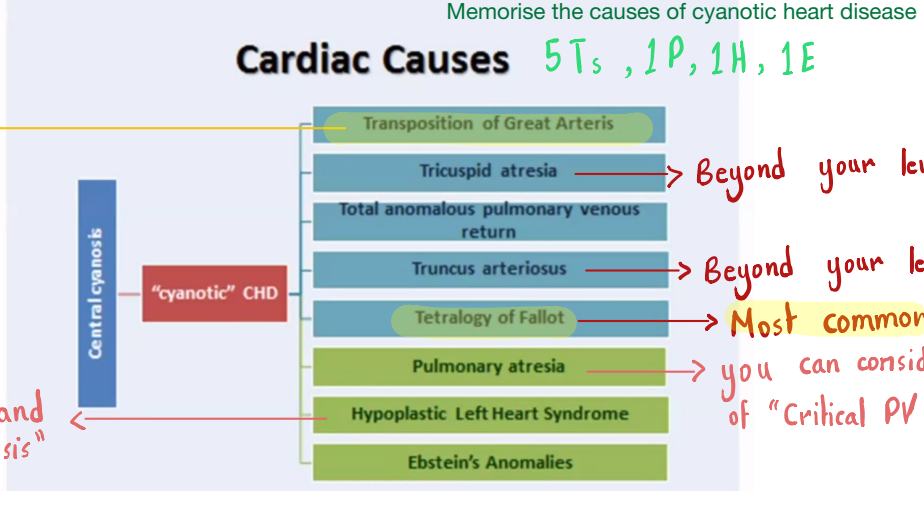


Cyanotic Heart Disease

"MCQs"

What is the most Common Cyanotic Heart disease that presents with "Severe Cyanosis" after Birth?

TGA (Transposition of Great Arteries).



Beyond your level

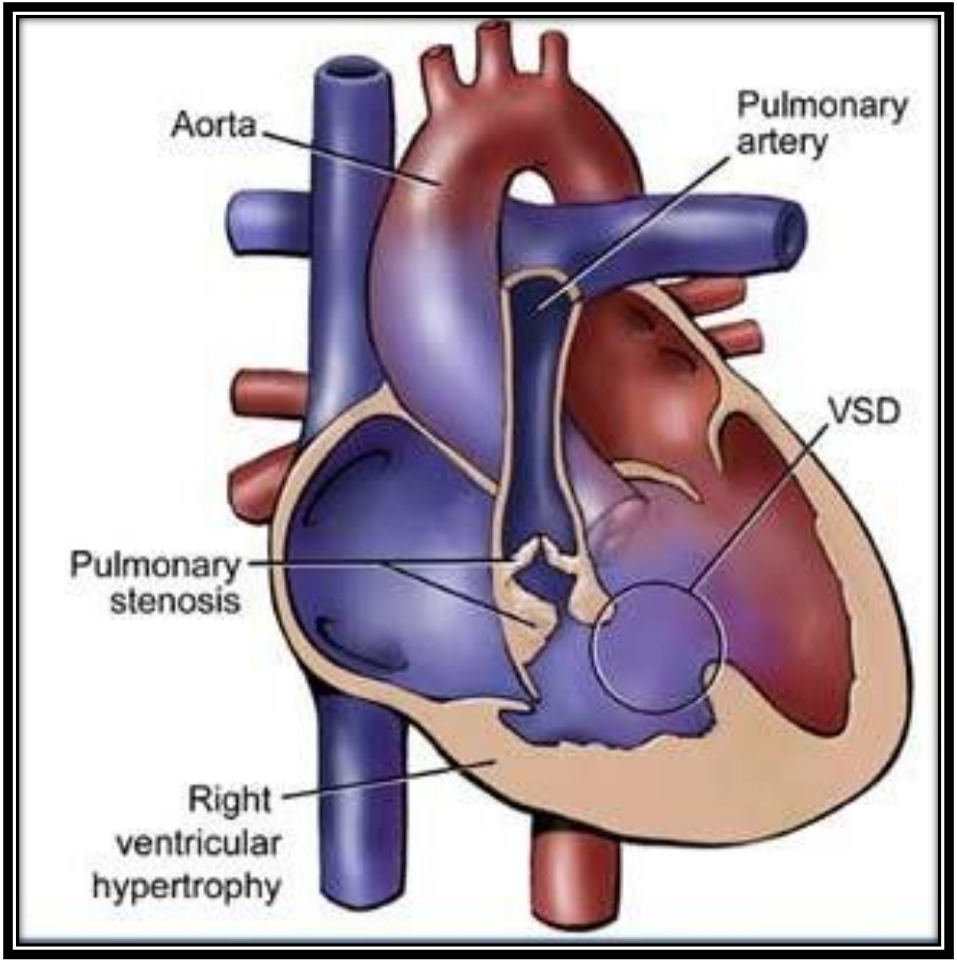
Beyond your level

Most common "MCQs"

you can consider it a sister of "Critical PV stenosis"

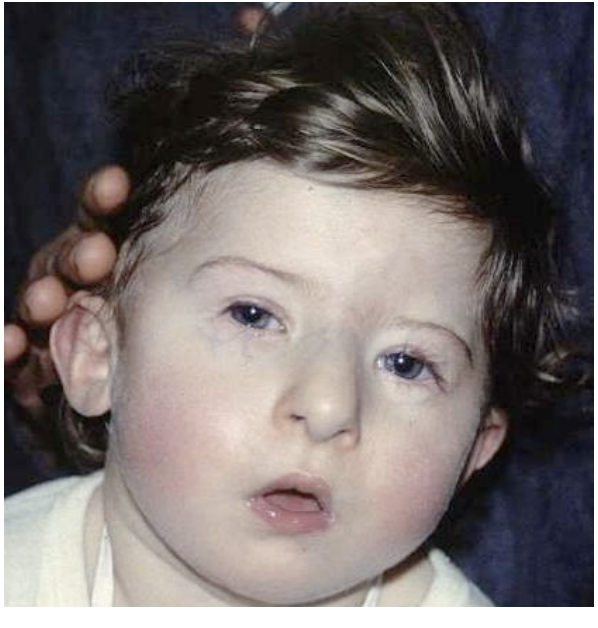
Similar to "CoA" and "Critical AV stenosis"

Tetralogy of Fallot



"رُباعيتي فالوب" Tetralogy of Fallot

- Most common cyanotic CHD
  - Incidence: 6 % of all CHD
- Can be associated with
  - DiGeorge Syndrome

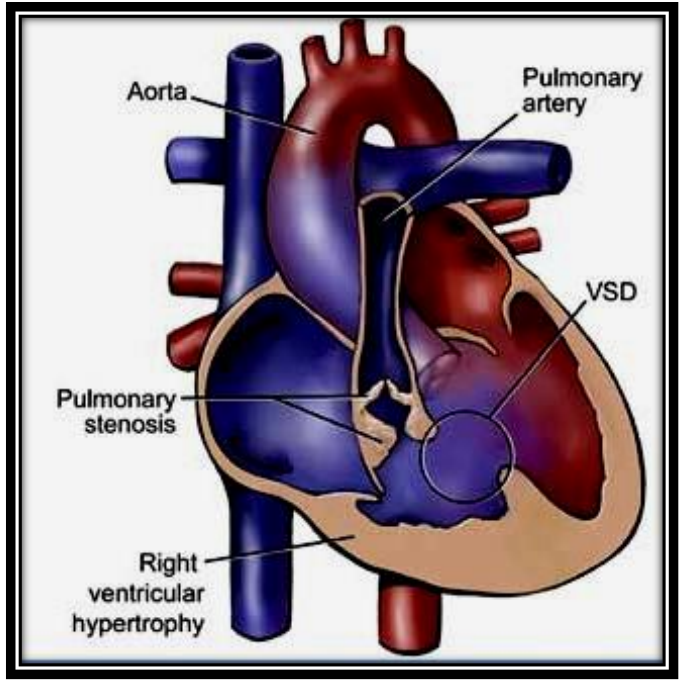


Tetralogy of Fallot

- Which of these components determines the severity of TOF?
  pulmonary stenosis

- What is the thing that makes cyanosis progressive?
  The pulmonary valve stenosis mainly the subvalvular.

- **Four basic components**
  1 – Large VSD
  2 – Pulmonary stenosis (PS)
  3 – Overriding aorta
  4 – RV hypertrophy



- As the VSD gets larger the pressure equalizes between the 2 ventricles if there were no pulmonary stenosis, then the blood would have gone through the pulmonary artery into the lung and caused an Acyanotic heart disease **BUT this is NOT the case**.
- the pulmonary stenosis doesn't allow the blood to pass through it so the blood goes to the aorta and now you have **mixed blood in the aorta** leading to cyanotic heart disease.
- So what determines the severity of the cyanosis is the severity if the pulmonary stenosis. The more stenotic the higher the cyanosis **the earlier you need to intervene**

CLINICAL FEATURES: TOF

- Depend on the severity of PS
  - Most newborn:
    - Asymptomatic
    - Ejection systolic murmur on routine discharge exam
    - Initially have mild cyanosis which progress with time:
      - Might present with hypercyanotic spells "tet spell" if delayed intervention

- Cyanosis in TOF is progressive, stenosis is both at the valve and under the valve, and whats under the valve is a muscle that with time hypertrophies and grows more, so the obstruction here becomes progressive and when that happens it turns into a disease called a hypercyanotic spell (severe cyanosis that could lead to death) ——> Not seen Anymore

CLINICAL FEATURES: TET SPELL

- Usually occur around 9-12 months of age
  - Episodes of acute and severe cyanosis
  - RX:
    - Medical emergency
      - Reduced anxiety "Keep child in his mother lab"
      - Knee-to-chest position
      - Give oxygen
      - Sedation with morphine ↓PVR
      - IV fluid ↑SVR
      - Beta Blocker
      - Phenylephrine
    - Might require emergency surgical intervention

*We have Two options:
- ↑ Systemic circulation:
  1- Knee to chest Position "Bend the Femoral artery to ↑ the resistance and Force the Blood to go to the Pulmonary circulation.
  2- Adrenaline "Phenylephrine"

The other option:
- ↓ Pulmonary Circulation
  1- Calm the Child if He is Crying
  2- The most Potent Pulmonary vasodialator is "Oxygen" by Face Mask.
  3- Beta Blocker

CLINICAL FEATURES: TOF

- **Newborn with severe PS or pulmonary atresia**
  - **Severe cyanosis when PDA close**
  - **"Duct dependent CHD"**
  - **Need IV prostaglandin E2**

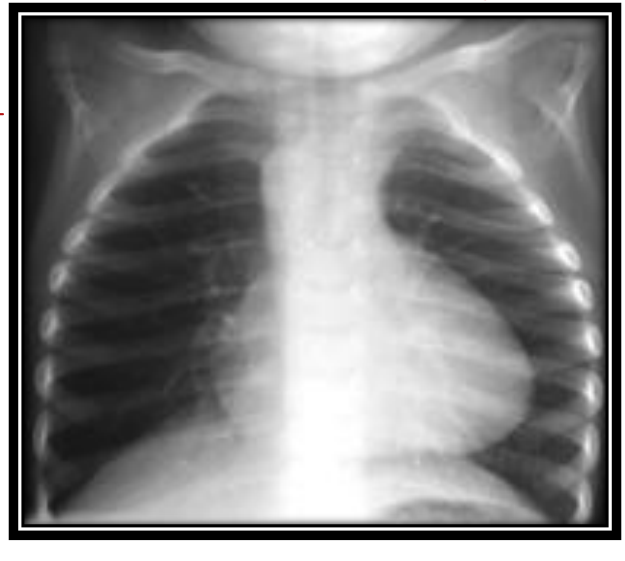
- The worst thing that could happen is for the baby to have and atretic pulmonary valve. So if they have TOF with pulmonary atresia, there is no blood coming to the lung so the baby wil die immediately.
- This is called **critical congenital heart disease.**
- We need a source of blood to go to the lung that is why we give them **prostaglandins** so that we have a PDA. **This is because it is a "duct dependent lesion"**
- In this situation you need to supply the pulmonary stenosis so the shunt that you will need is left to right shunt.

INVESTIGATION: TOF

*Boot Shaped Heart:
1- Narrow mediastinum
2- Heart Apex is going up
3- The lung is Oligemic (No Blood is going there)

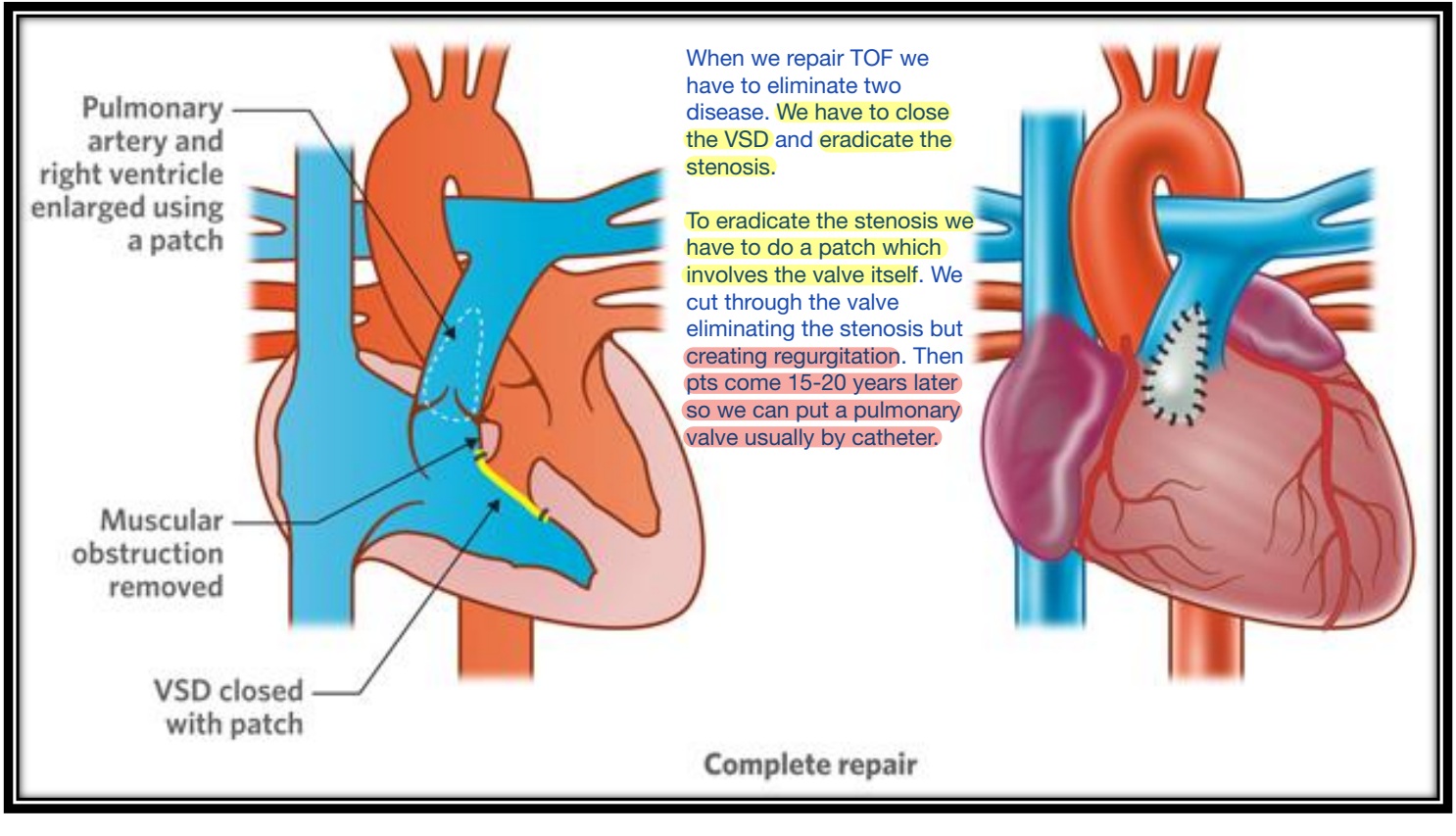
We might ask you about it !!

- CHEST X-RAY:
  - "boot-shaped heart"
- ECG: RVH
- ECHO: confirm diagnosis
- CT/MRI rarely needed



Lung is black and the heart is boot shaped which is characteristic of TOF

TREATMENT: TOF



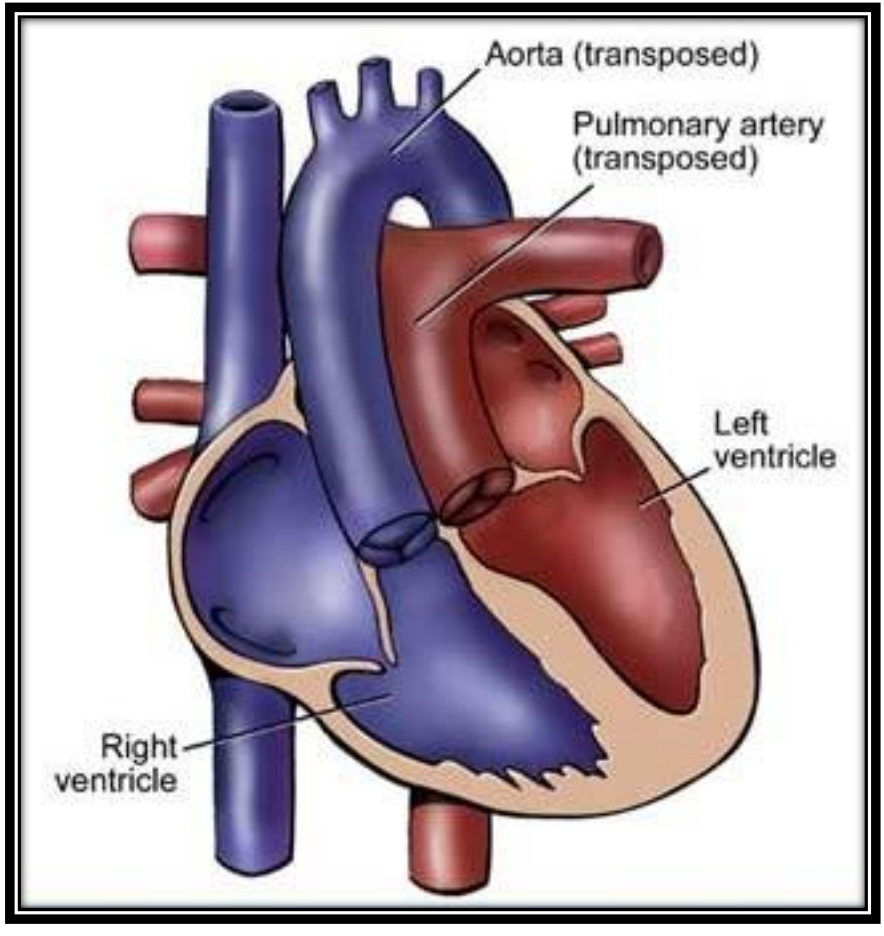
When we repair TOF we have to eliminate two disease. We have to close the VSD and eradicate the stenosis.

To eradicate the stenosis we have to do a patch which involves the valve itself. We cut through the valve eliminating the stenosis but creating regurgitation. Then pts come 15-20 years later so we can put a pulmonary valve usually by catheter.

Transposition of the Great Arteries

The pulmorary Artery now became the Aorta and Vice Versa.

They are parallel to each other>systemic circulation not communicating with pulmonary circulation



D-TGA

- Incidence: 4 % of all CHD
- Most common CHD presented with cyanosis at birth.
  - TOF is the most common cyanotic heart disease 5%
  - TGA is the most common cause of severe cyanosis in newborn baby
- More common in male
- Higher incidence in infant of diabetic mother

  + large for Gestational Age

PATHOPHYSIOLOGY: D-TGA

- In Normal heart:
  - Pulmonary and systemic circulations are in series
- In D-TGA:
  - Pulmonary and systemic circulations are in parallel

- Follow the blood in TGA:
- Blue blood comes from SVC/IVC –> RA –> RV –> Aorta to systemic circulation so the blue is becoming more and more blue.
- Red blood comes from the lung –> LA –> LV –> pulmonary system circulation so they red is becoming more red
- The two bloods are seperate therefore its a parallel circulation (the blood should meet in the lungs but its not)

PATHOPHYSIOLOGY: D-TGA

- Mixing of oxygenated and deoxygenated blood can occur at three levels:
  - Atrial level via ASD/PFO (most important)
  - Great arteries level via PDA
  - Ventricular level via VSD (if present)

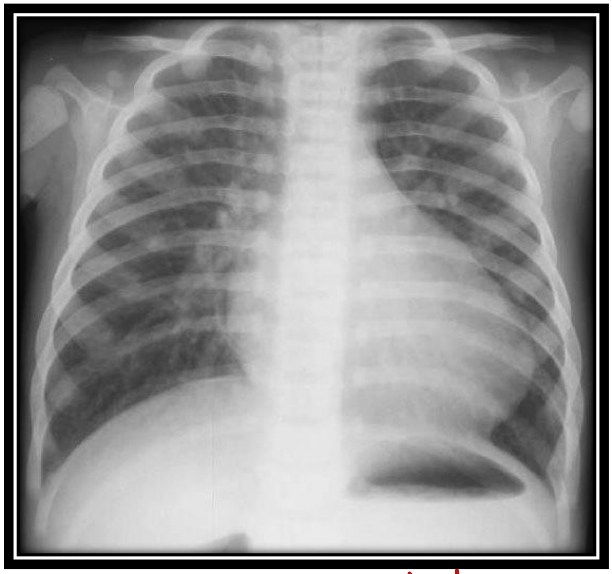
- You basically want a place for the blood to mix so you need to create room for the blood to mix and the best place for blood mixing is a place where the pressure is equal in both sides i.e the atrial shunt → The Best thing to do initially is : Atrial Septostomy
- We dont open a VSD if the VSD was not originally there.
- You can initially manage the pt by opening PDA by giving them prostaglandins but it's not the best way to shunt (it is just temporary)

PRESENTATION: D-TGA

- Severely Cyanosis after birth
- "**Duct Dependent CHD**"
- "**Reverse differential cyanosis**" if Pulm HTN
- No signs of respiratory distress
- Single second heart sound → Bec. the Aorta is moved Anteriorly, so it will mask the Pulmonary.
- Typically: no murmur
- Hyperoxic test: **FAIL**

INVESTIGATION: D-TGA

- Chest X-ray:
  - "egg on a string" appearance (MCQ)
- ECG:
  - Typically normal
- ECHO: confirm diagnosis
- Cardiac Cath:
  - For septestomy
  - +/- coronary arteries anatomy



1- Narrowed mediastinum
2- Egg shaped Heart

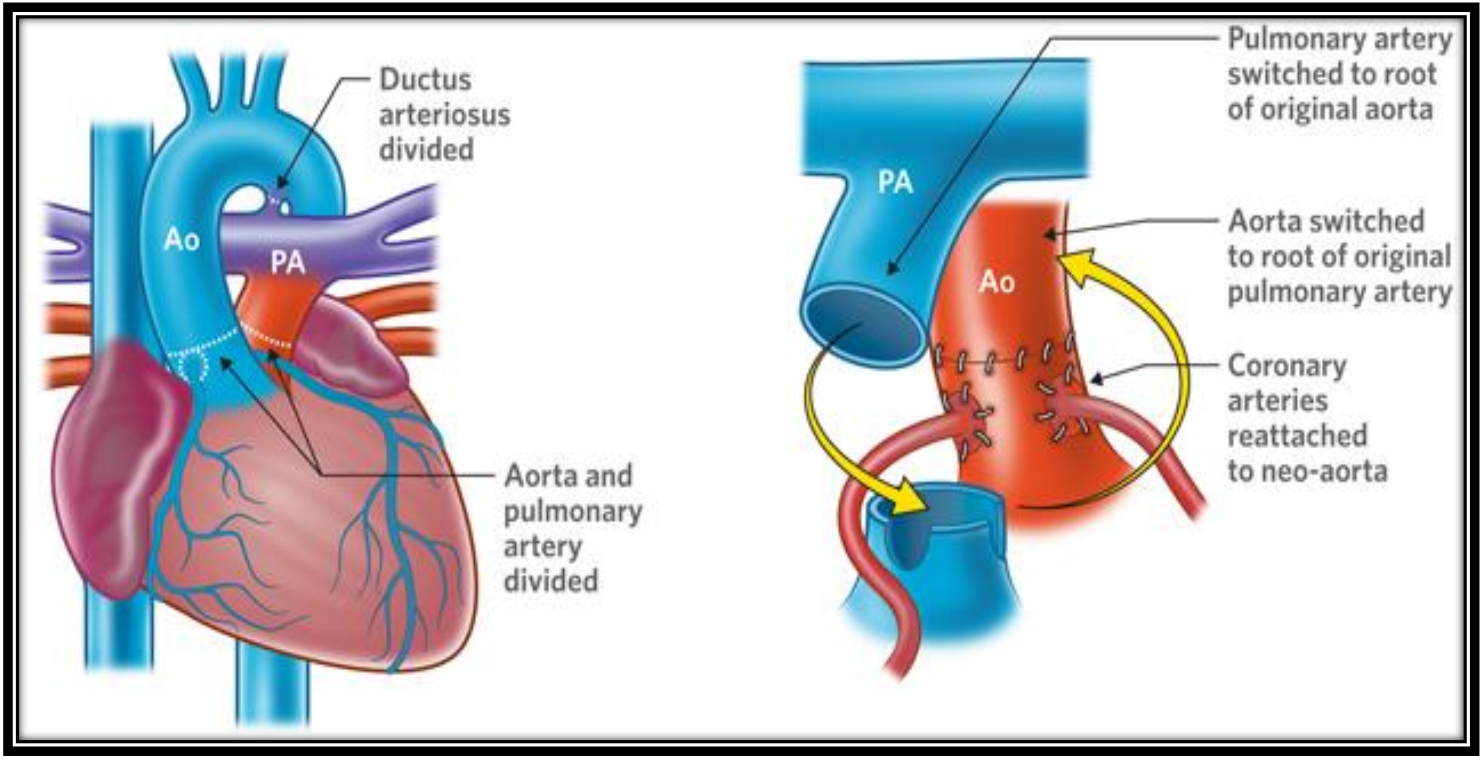
The Best intervention for them, to move them from severe cyanosis and death to "Acceptable Cyanosis"
↳ After that we have to do surgery.

MANAGEMENT: D-TGA

- Supportive:
  - Prostaglandin E2 Will mix the two vessels,from Aorta to pulmonary the lung will have more blood>increase pressure in left atrium>ballon>right atrium
  - Balloon atrial septostomy (for better mixing) At the level of atrium
    - This is a temporary measure to allow the pt to be more stable, less cyanotic and acidotic

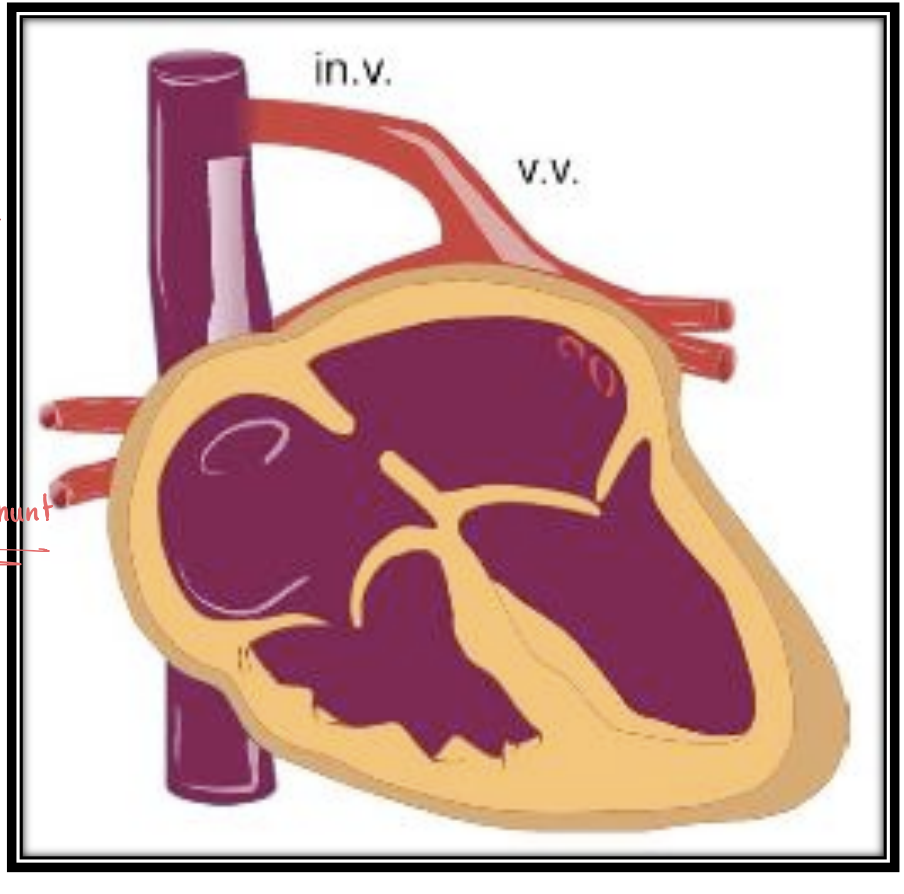
MANAGEMENT: D-TGA

- Arterial switch operation (definitive treatment)
- The timing of the operation should be in the first two weeks of life



Total Anomalous Pulmonary Venous Return : TAPVD

* The LA will only receive Blood From "Foramen Ovale" Which will Not Close.
Why ?
The RA Pressure Postnataly remains High, Which is also called: "Obligatory" R-L shunt

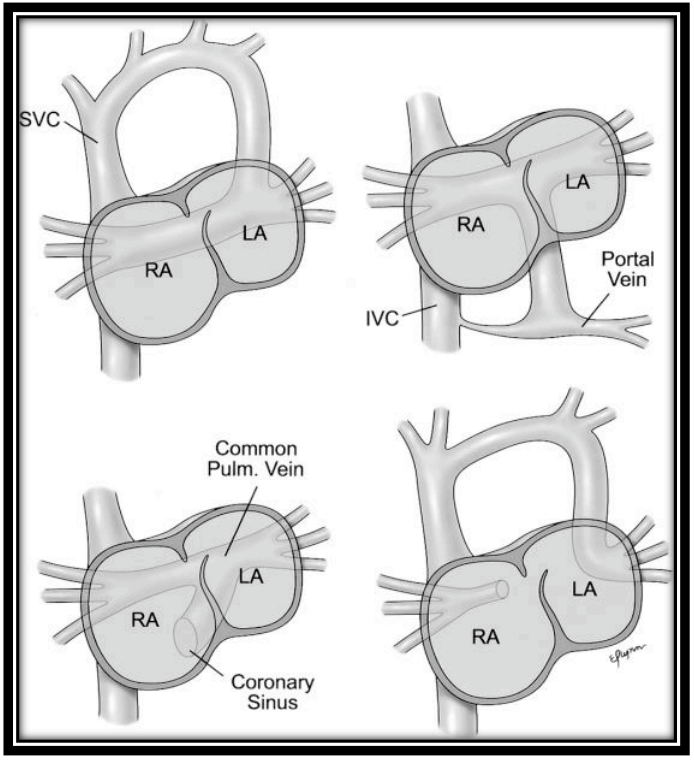


TAPVD

- All 4 pulmonary veins returns to the right atrium
- Can be:
  - Supracardiac (50%) → SVC
  - Cardiac (25%) → Coronary Sinus to the RA
  - Infracardiac (20%) → IVC
  - Mixed (5%)

4

- Can be:
  - Obstructed TAPVR
  - Non-obstructed TAPVR



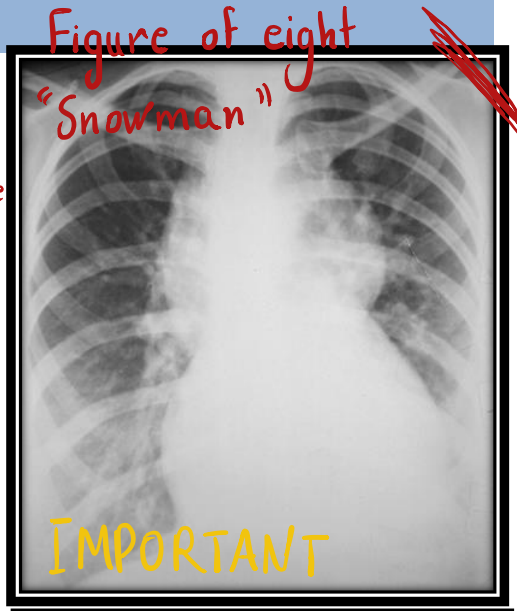
PRESENTATION: TAPVD

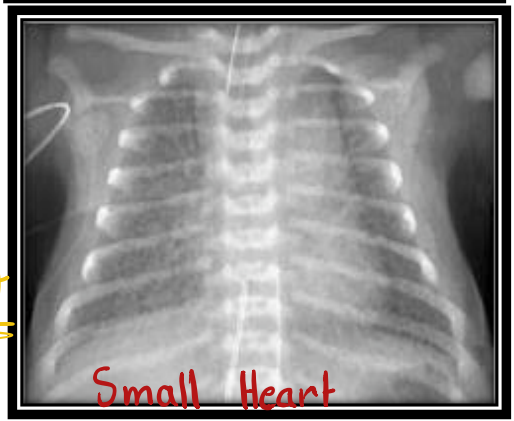
- Clinical Feature:
  - Cyanosis at birth
- Diagnosis:
  - Chest X-ray:
    - Increased pulmonary vascualr markings
    - "**Figure of eight**" in obstructed supracardiac TAPVR
  - ECG: RVH
  - ECHO: Confirm Dx
  - Cardiac CT/MI: may be need

Snowman

* They usually have No murmurs.
* Normal or Borderline Oxygen Saturation

Treatment : Surgery and the Outcome is Excellent

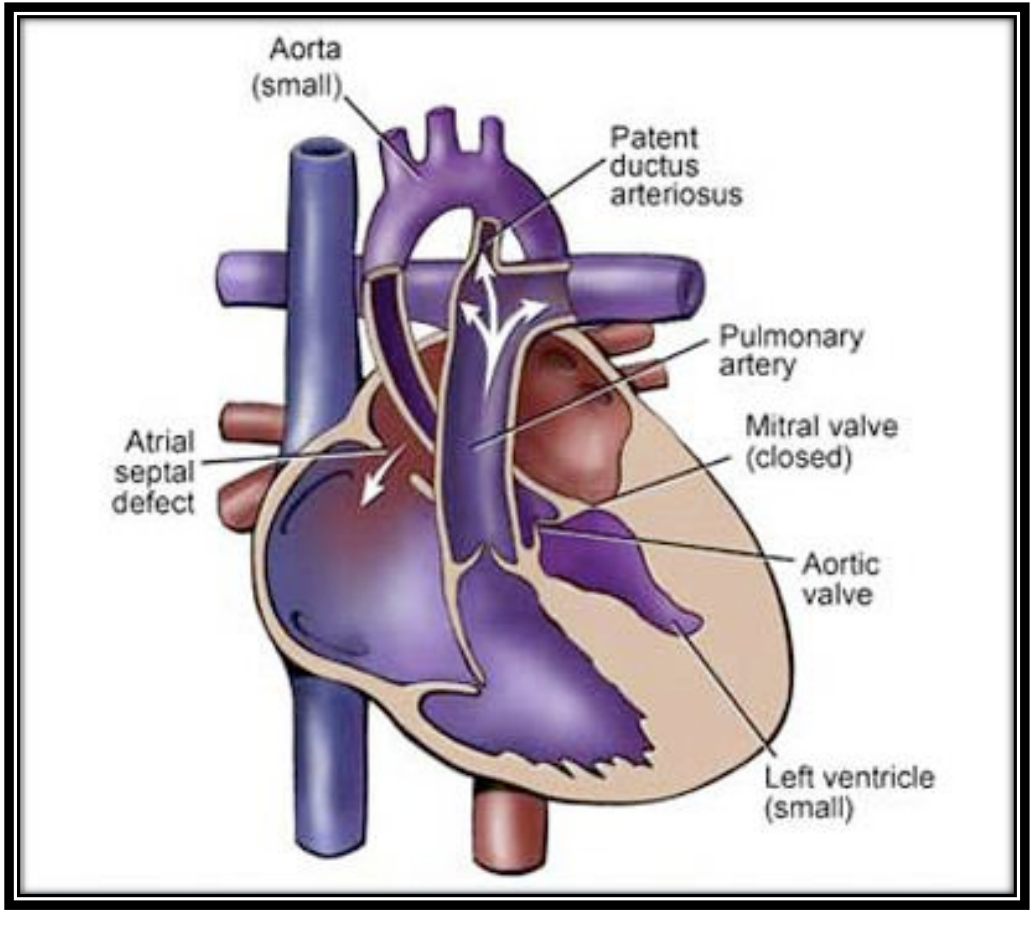




Didn't talk about it

RDS Picture

Hypoplastic Left Heart Syndrome

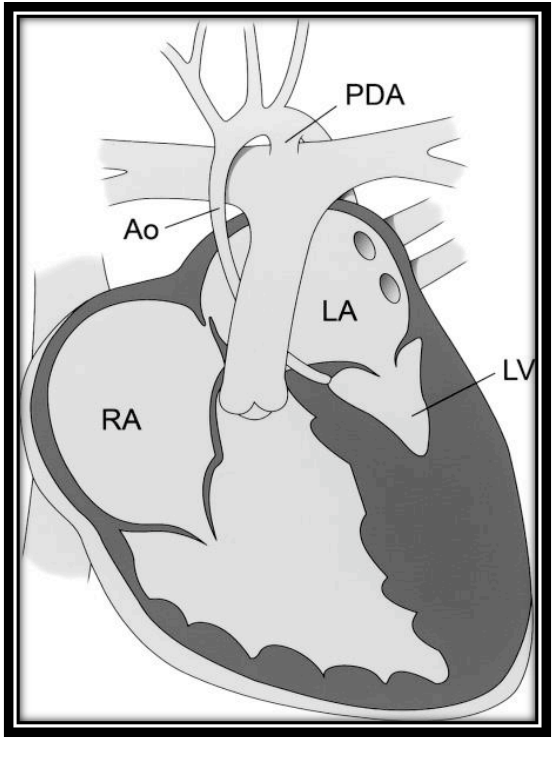


HLHS

- HLHS: one of the most severe form of CHD
  - High morbidity and mortality
- Incidence: 1-2 % of all CHD
- multiple level of obstruction at left heart structures.
  - Mitral stenosis to mitral atresia
  - Variable degree of LV hypoplasia
  - Aortic stenosis to aortic atresia
  - Variable degree of ascending aorta hypoplasia

PATHOPHYSIOLOGY: HLHS

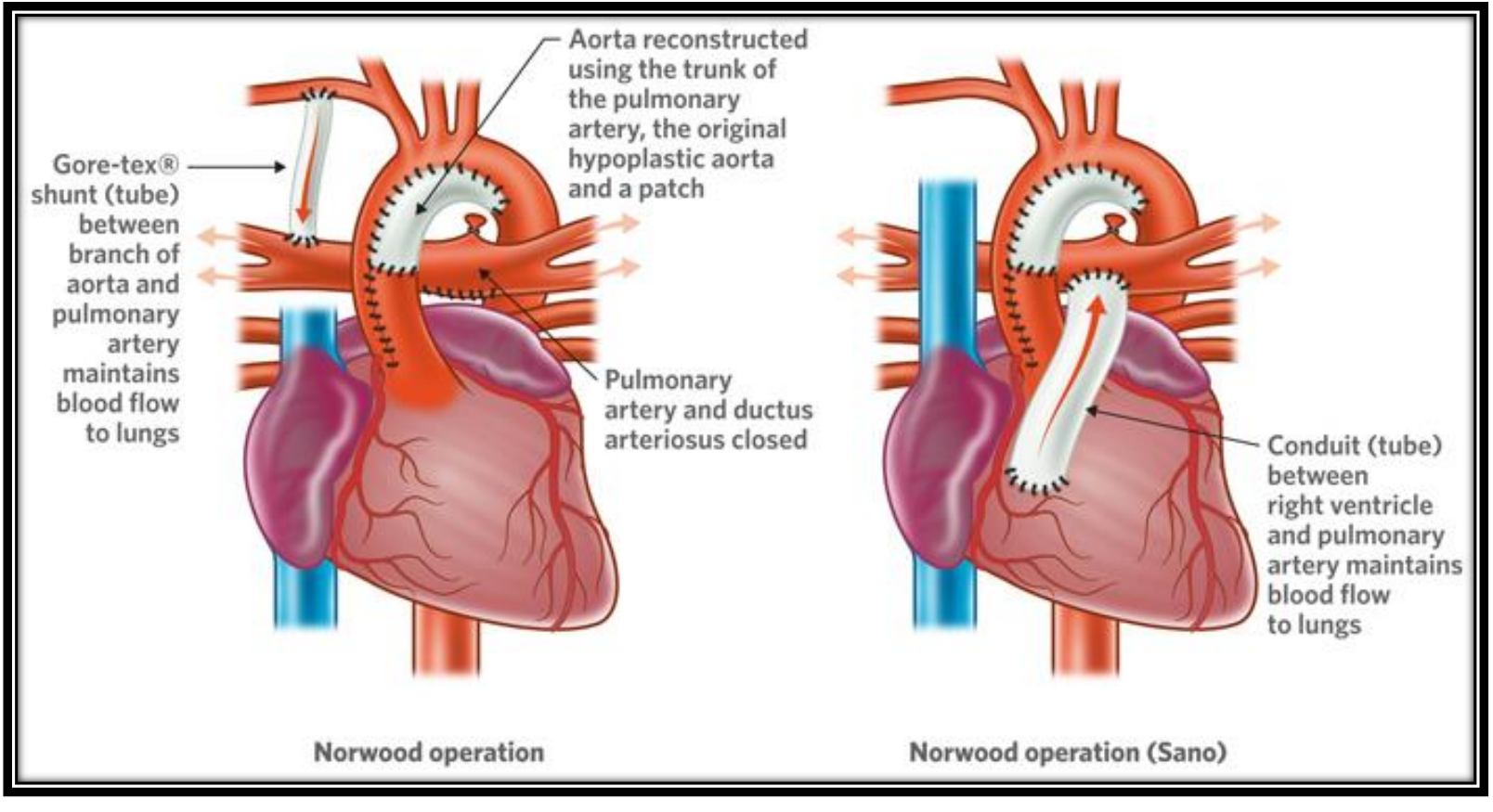
- No adequate flow across aortic valve to ascending aorta
- Relies on retrograde PDA flow to:
  - Brain
  - Coronary arteries
- Need ASD/PFO to shunt blood from LA to RA.



PRESENTATION: HLHS

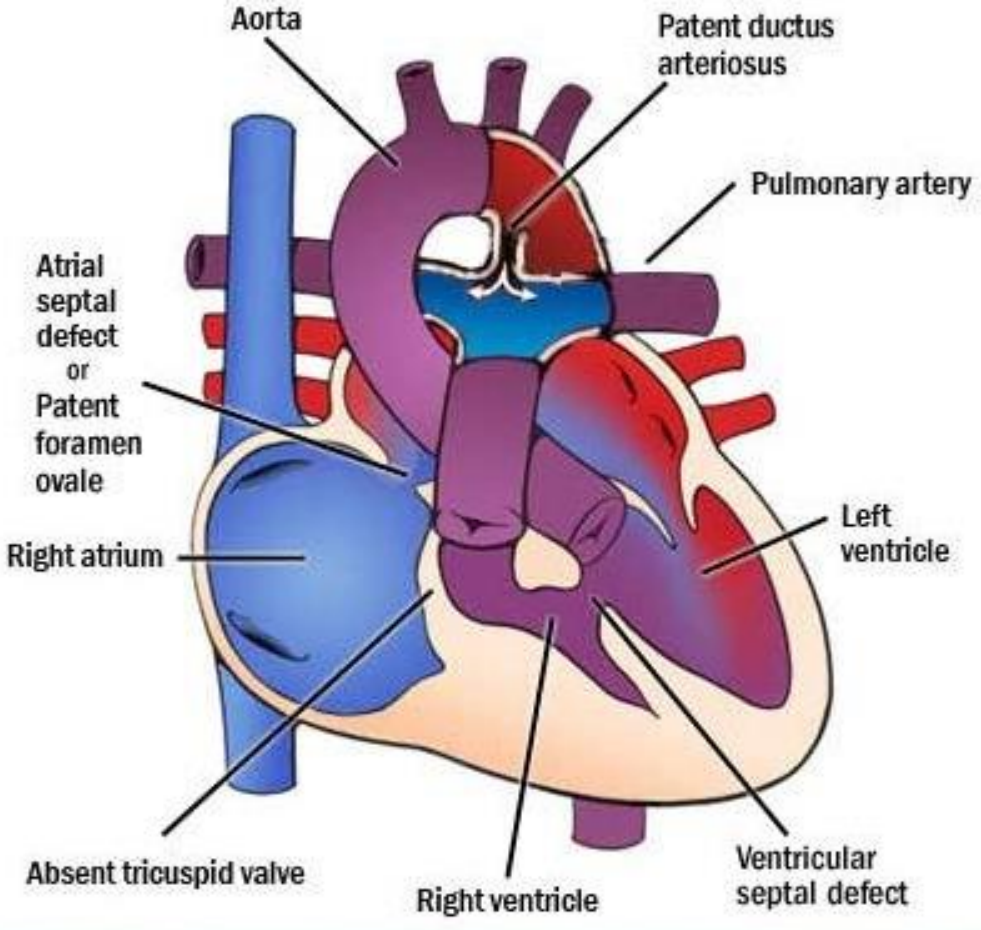
- At birth: Cyanosis
- At 2-4 week of life:
  - Respiratory distress
  - Poor pulses/perfusion
  - Signs of cardiac shock

TREATMENT: HLHS



Tricuspid Atresia

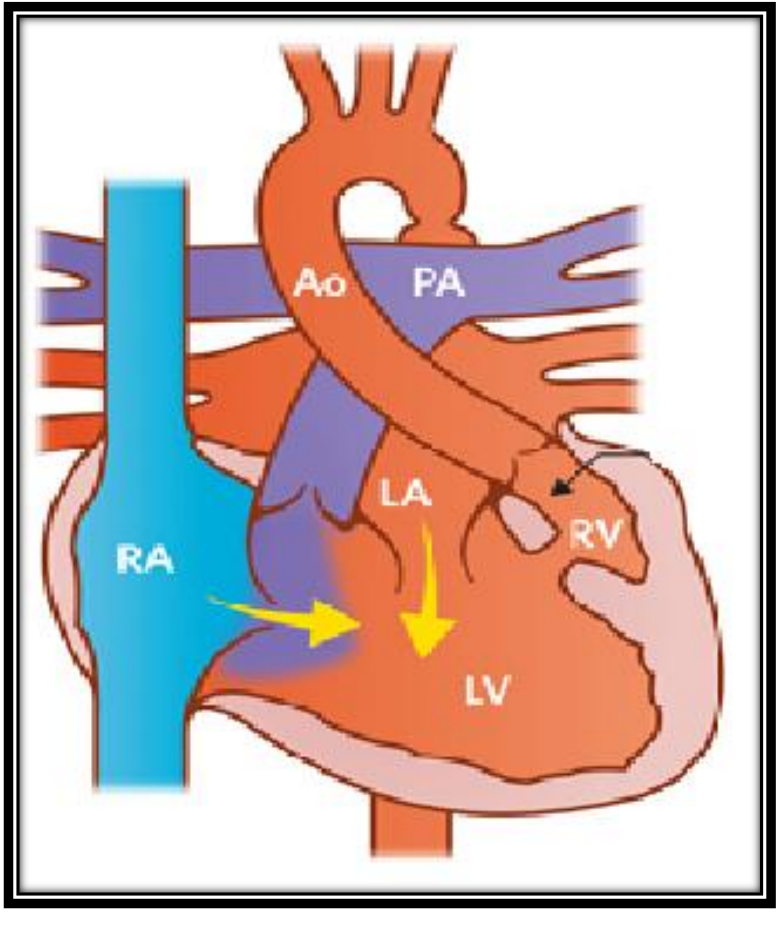
Didn't talk about it



Single Ventricle

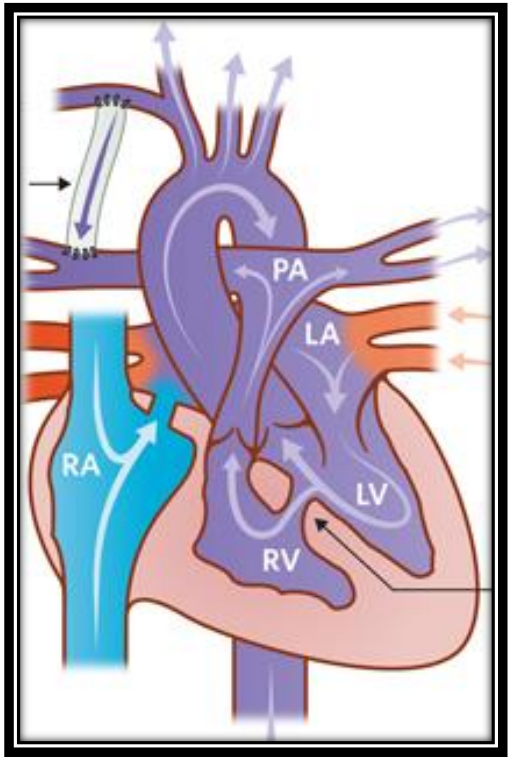
Not v.imp for your knowledge

*Here the Heart Can't be divided into 4 rooms



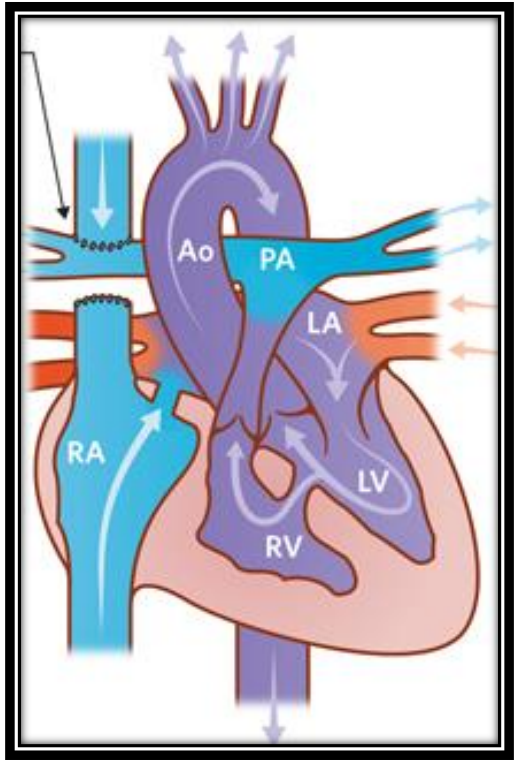
- Multiple stages to fix the single ventricle
- They can have a decent living and they will not differ from normal ppl except for their need to take regular medication

STAGED SURGERY UNIVENTRICULAR HEART

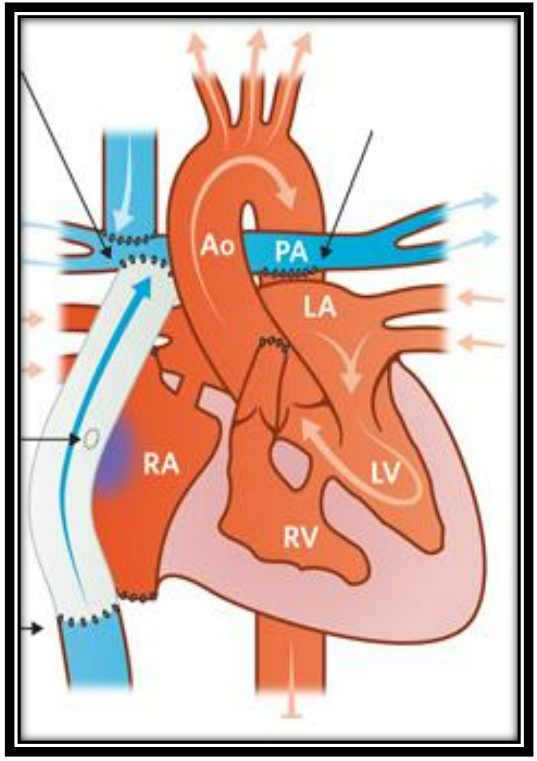


BT SHUNT

The First Stage



GLENN

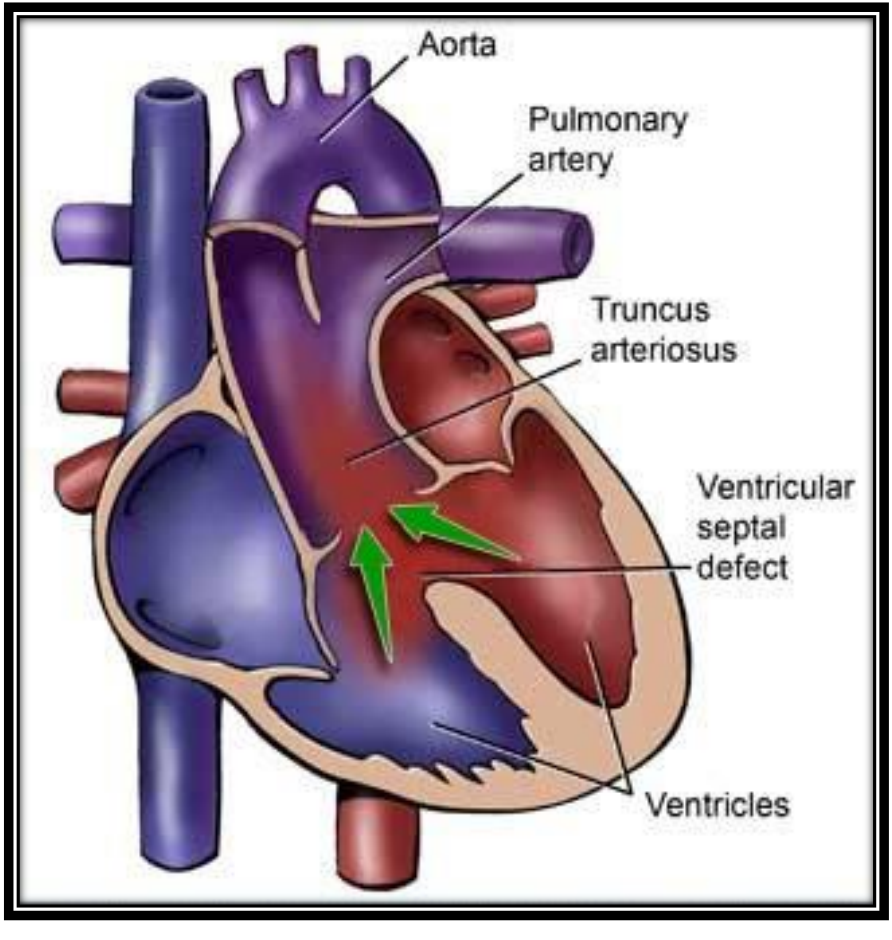


FONTAN

The Ultimate Surgery

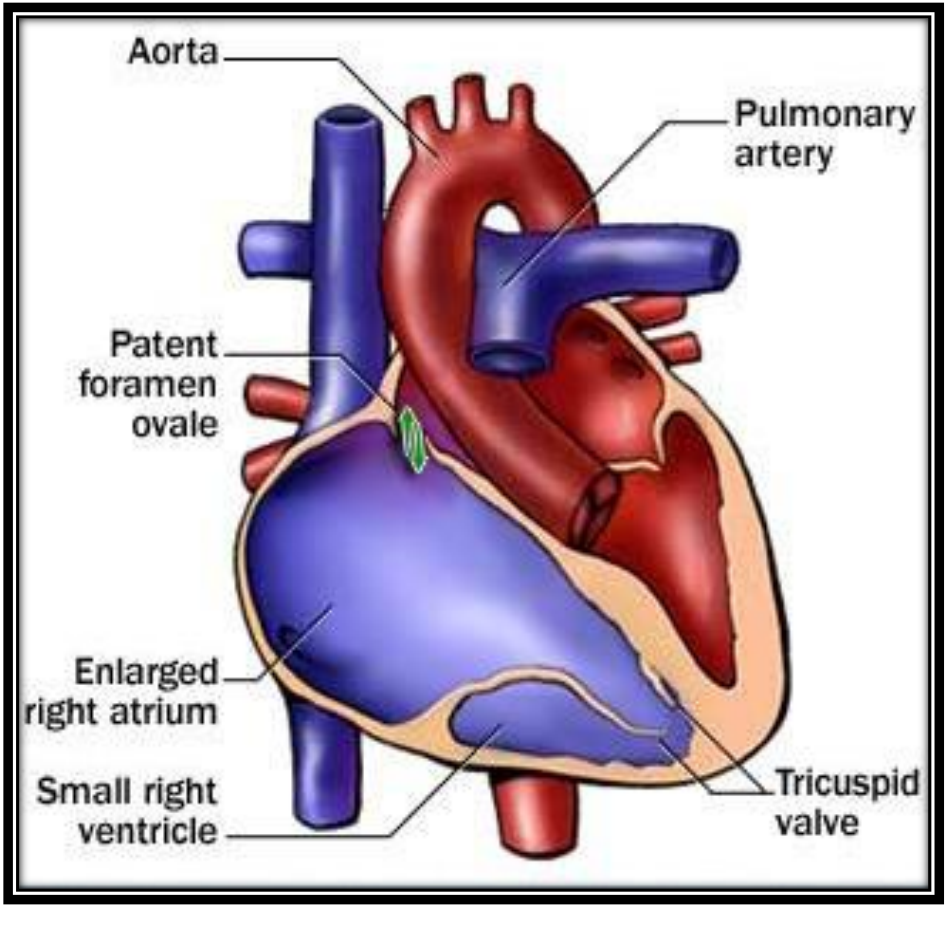
Didn't talk about it

Truncus Arteriosus



Didn't talk about it

Ebstein's Anomaly



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