CVS

PBL

Modified NO: 1

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تدقيق: Done

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Clinical Pediatric Cardiology
CVS- 3rd year

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Pediatric Cardiology

Color code

- Slides
- Doctor
- Additional info
- Important

Introduction

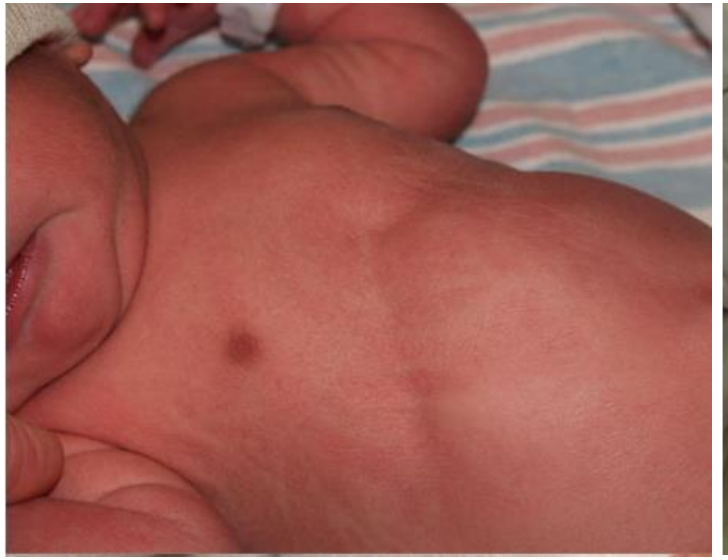
- Pediatric cardiac disease are largely congenital defects
- Some acquired heart disease may happen in Pediatrics, but they are the minority of diseases.
- Will present 4 common cases of children with heart disease

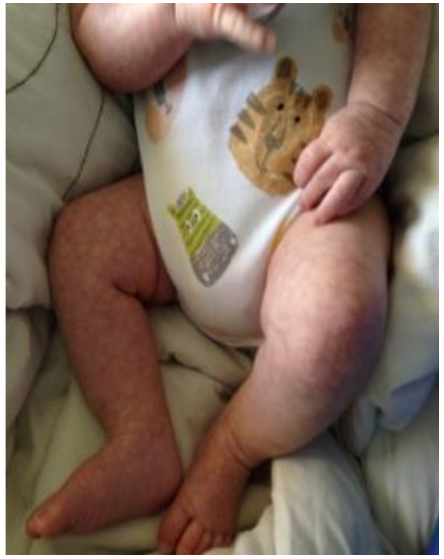
- In each case we will focus on:
  - Embryologic consideration leading to the anatomic defect
  - Physiology and hemodynamics leading to the clinical picture

Congenital means it developed before birth and it manifested after birth, many of them are genetics based, and others are developmental which means it happened during the development of the fetus but it manifested later.

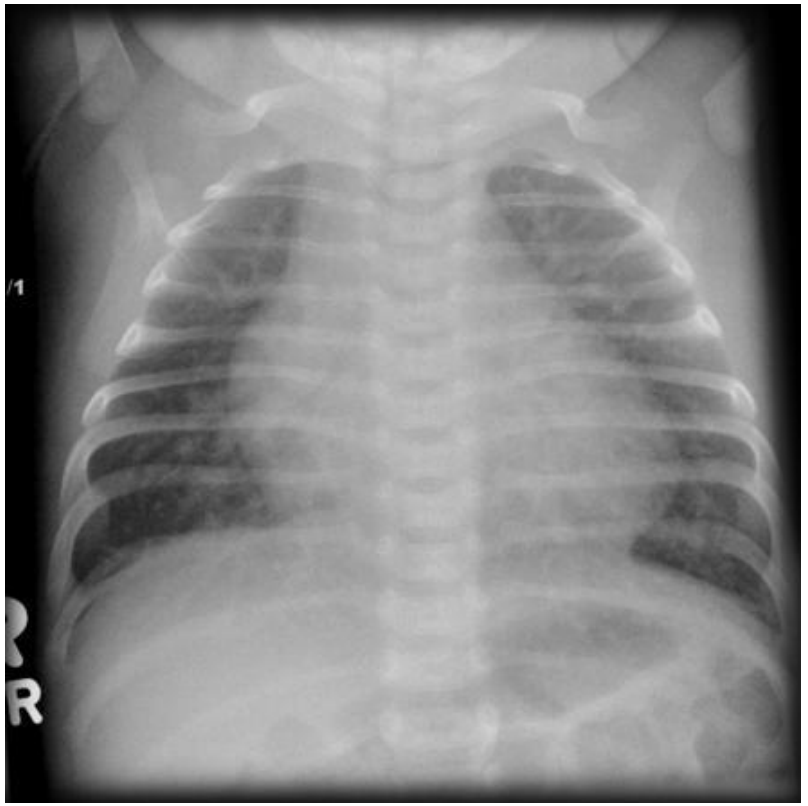
Case 1

- Two month old infant who has:
  - Rapid breathing (tachypnea)
  - Difficulty of feeding
  - Not gaining weight appropriately
- Examination:
  - Signs of respiratory distress (difficulty when taking breath)
  - Rapid heart rate (tachycardia)
  - Weak pulses, skin hypo-perfusion (the skin looks cold, pale → the perfusion to the skin is not normal)
  - Systolic heart murmur
- Chest X ray showed:
  - Enlarged heart (cardiomegaly)
  - Congested lungs (the lungs are receiving more fluid (blood) than normal)





abnormal sound when we listen to the heart, which indicates that there is a high velocity blood going somewhere in his heart.



In the pictures above we see the following:

1) Intercostal Muscle Retraction: Because of the difficulty of breathing, we will see that the intercostal muscles target in when he takes a breath, and that because the pressure in the chest is negative and less than normal.

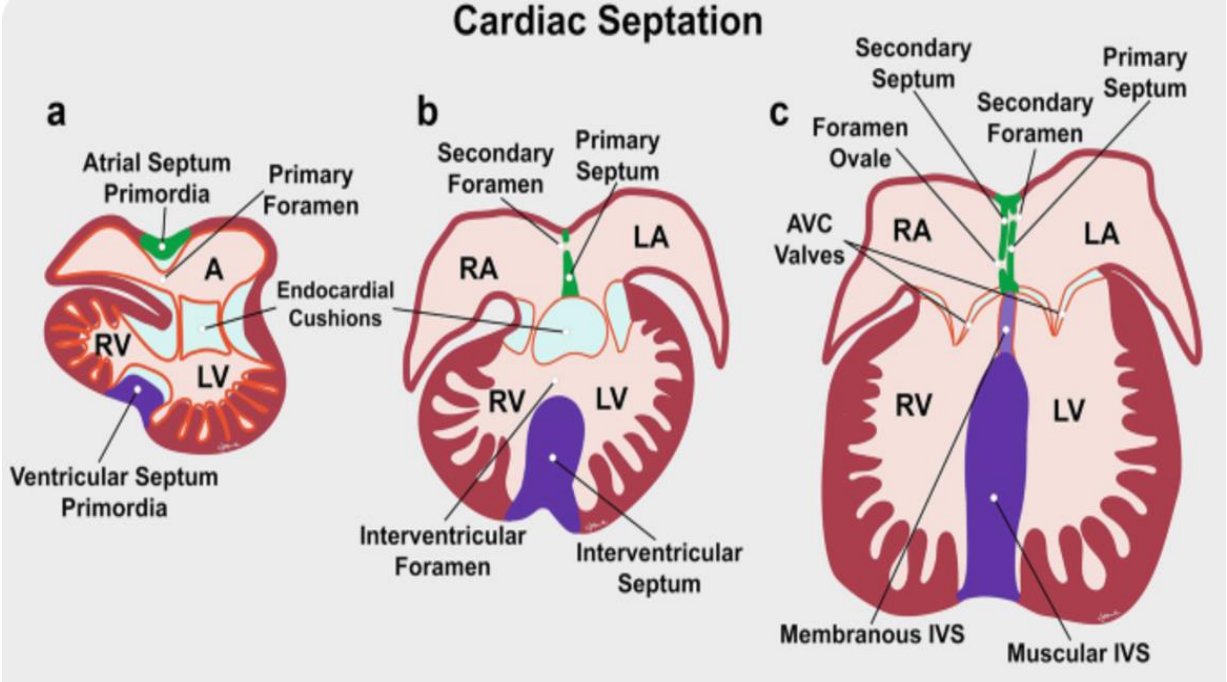
2) The skin might be either pale or mottled, mottling of the skin means that there is a **net**-like pattern on the skin that might indicate decreased perfusion of the skin.

3) In the chest X-Ray image, it shows that the heart shadow is large (cardiomegaly), and the lungs are congested (there is more fluid in the lungs), normally the lungs should appear black because they are full of air (the air is black on the x-ray), but there is a lot of white in the lung which means that the lung are congested.

Additional info regarding the negative pressure: When breathing becomes harder, the muscles that help us breathe, like the diaphragm and intercostal muscles work harder. As the diaphragm moves down more forcefully, it creates a vacuum (negative pressure) in the chest. This causes the intercostal muscles to pull inward (retract) with each breath, trying to help the body get more air.

Diagnosis: Ventricular septal defect

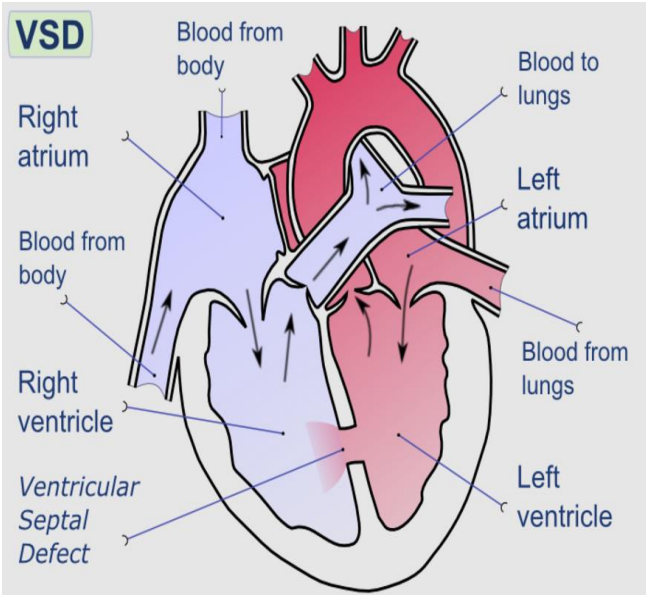
The above symptoms may indicate another diagnosis, but VSD is the most common.



- Embryologic defect: failure of ventricular septation
  - Septation of the primitive ventricle starts after cardiac looping. to form the two ventricles
  - Interventricular foramen (primary and secondary)
  - Ventricular septum is composed of muscular part, membranous part, and endocardial cushings
- Anatomical result:
  - Defect between the ventricles
  - Types (muscular, membranous, inlet/canal types..etc)

Revision:
First, the muscular septum begins to grow. Along with the Cushing's septum and the membranous septum, they will all meet and form the interventricular septum.

Deficiency of this process



The upcoming slides is a bit long, but they are easy!

Note that the location of the intraventricular septum defect is not important in physiology, but we need it more in anatomy.

❑ But now, what is the effect of this defect in the heart and how does it cause all this symptoms? To know that we need to understand the hemodynamic first.

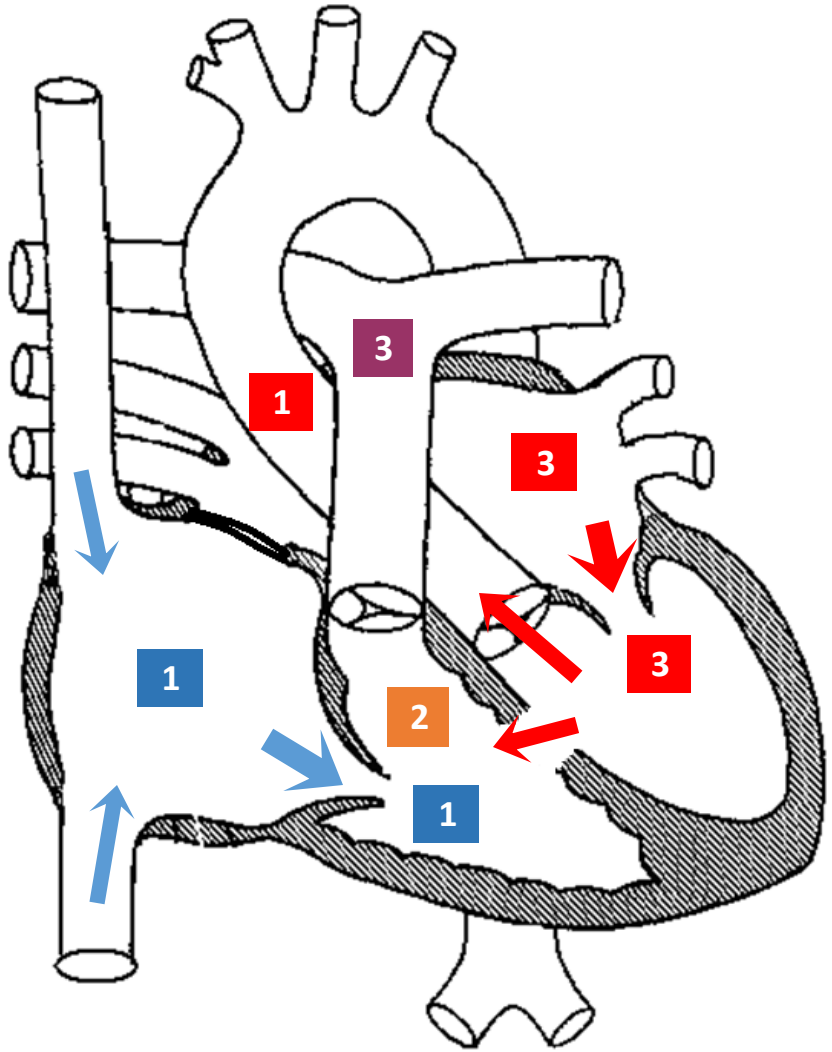
We have a systemic and pulmonary circulation that are connected at the heart, but normally they don't mix with each other.

But now we have a defect that can allow the blood to move from (right to left) ventricle or (left to right), what determine the flow direction is the **pressure** (flows from higher to lower pressure) and the **resistance**.

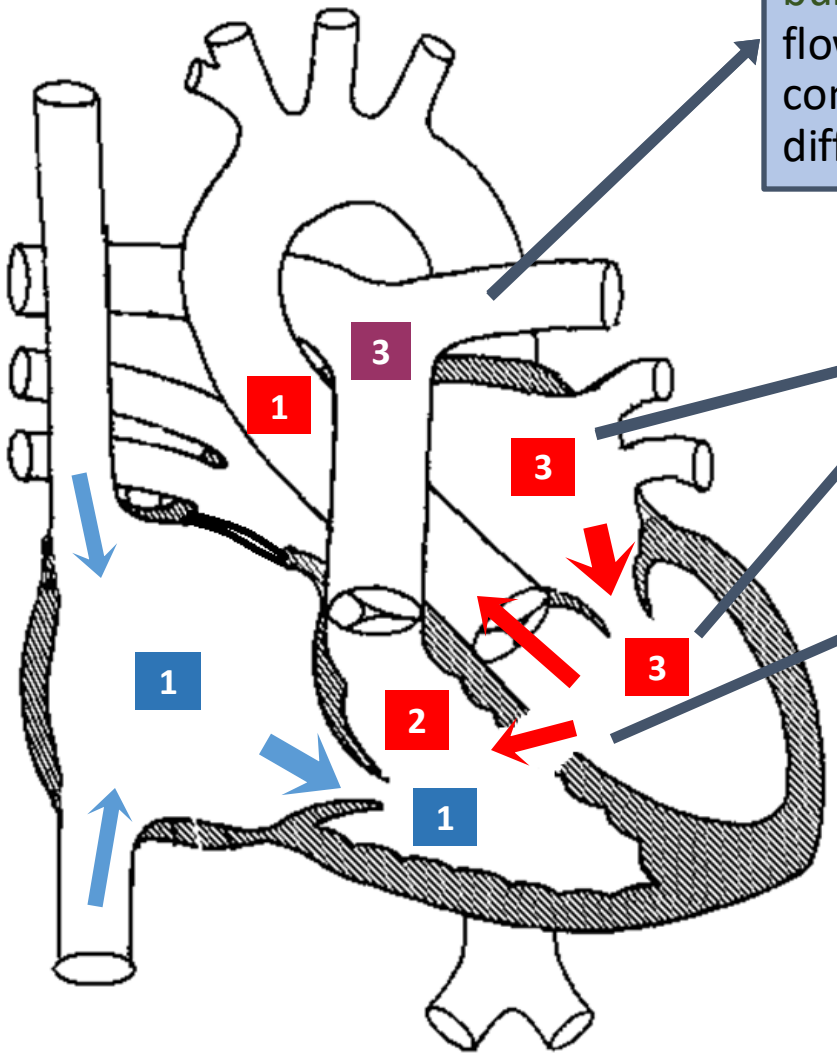
Here we know that the left ventricular pressure is higher → the blood will flow from **left to right**, also the vascular resistance in the pulmonary artery is much less compared to the systemic arteries, this will allow the blood flow to the lungs much easier than the aorta.

So, it's easier for blood in the left ventricle to cross the VSD to the right ventricle.

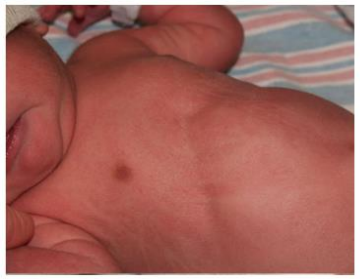
Let's assume that this child has 1 cardiac output that meats his demand and go into the systemic circulation, it will come back through the veins into the right atrium, then to the right ventricle, so there will be 1 cardiac output in the right ventricle now.
Now let's assume that the defect is large, and it can allow a lot of blood to flow to the right ventricle, let's say it's about 2 cardiac output crossed from left to right during systole.
So now you have 3 cardiac output in the right ventricle that will go to the lung (1 cardiac output + 2 cardiac output), and all of this will come back to the left atrium.
After that, the blood will go to the left ventricle during diastole, and now the left ventricle have 3 cardiac output.
Now in the next systole, you will get 1 cardiac output (the normal cardiac output) + 2 cardiac output (cross VSD … to the lungs)
So, you will have a lot of saturated blood that is going back to the lungs, so it's only causing problems to the lungs and it's not benefiting the baby.



Physiology of VSD ➔ clinical picture

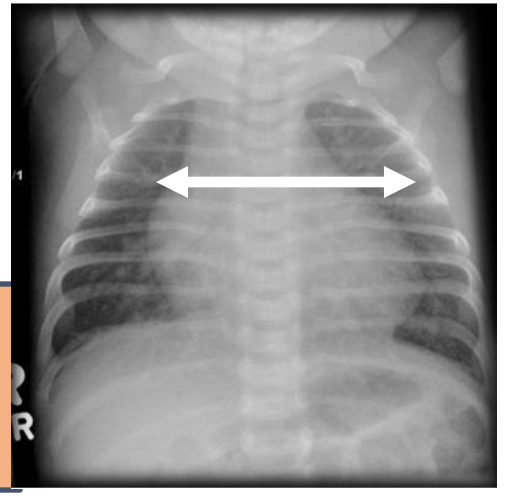


A lot of blood going to the lungs in high pressure (because the left ventricle is bumping)➔ Increased pulmonary blood flow and pulmonary pressure➔ congested lungs + stiff lungs ➔ breathing difficulty and rapid breathing (tachypnea)



Increased left atrial and left ventricular filling ➔ dilatation (cardiomegaly)

You will see the heart shifted to the left in physical examination

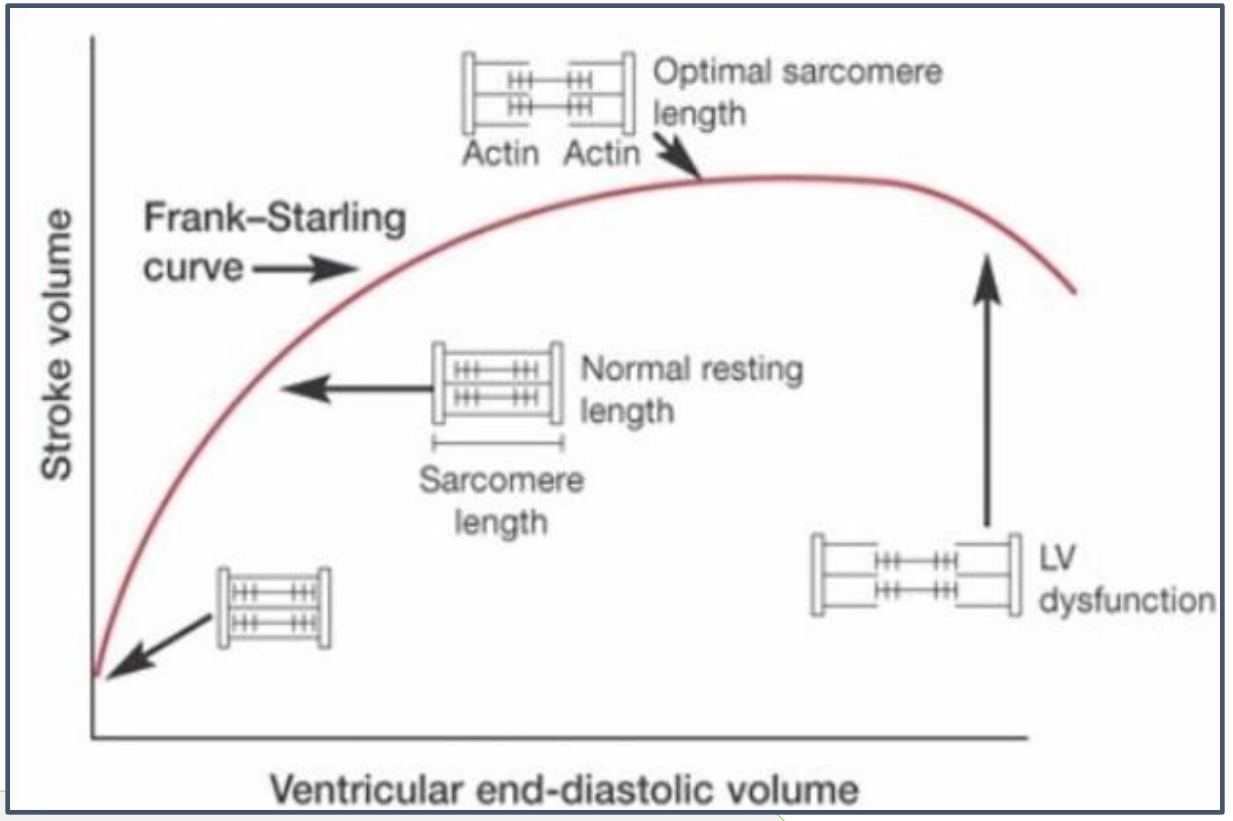


Flow between high pressure (left) and low pressure chamber (right) ➔ systolic murmur (turbulence)

Abnormal sound you can hear by the stethoscope.

Excessive dilatation eventually leads to systolic dysfunction ➔ low tissue perfusion

Excessive dilatation eventually leads to systolic dysfunction ➔ low tissue perfusion



Frank-starling law:
the more you stretch the muscle, initially you will get more contractility, but if you stretch the muscle too much, you will **lose that contractility.**

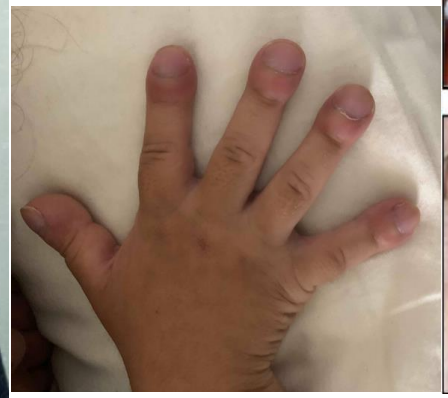
If you keep dilating and dilating, you will lose the interaction between the actin and myosin and you start losing the contractility.

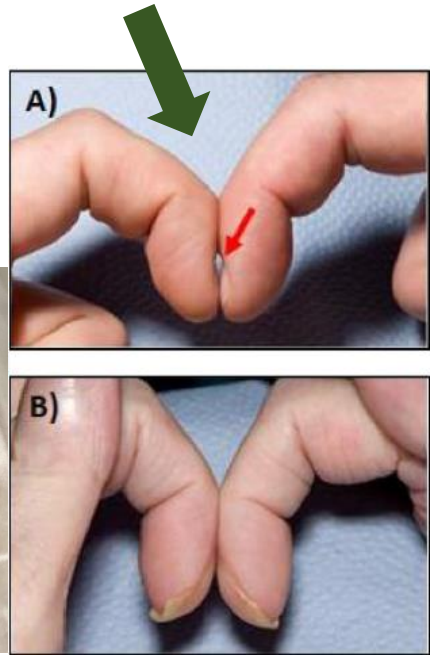
if we don't do anything to this condition, this will lead to **compensated heart failure**.

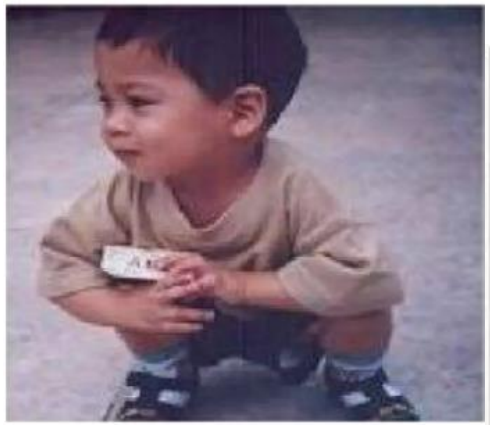
Case 2

- Two year old child who has:
  - Cyanosis that increases when he plays and cries (blue, ↓O_2 in blood).
  - Occasional squatting when cyanosed.
  - Generally growing well with good weight no symptoms of the other child (difficulty of feeding, breathing, heart failure …..)
- Examination:
  - Oxygen saturation of 80% (the normal is above 95%)
  - Plethoric face (opposite of pale) (congested with blood, purple in color)
  - Good pulse and perfusion (Normal cardiac output)
  - Systolic heart murmur
  - Finger clubbing (sign seen in patients with chronic cyanosis) (losing the angle between the nail and the nail bed)
- Chest X ray showed:
  - Heart not enlarged, but boot shaped, with dark lungs
- Hemoglobin level was 17 gm/dL (polycythemia) (higher than normal)

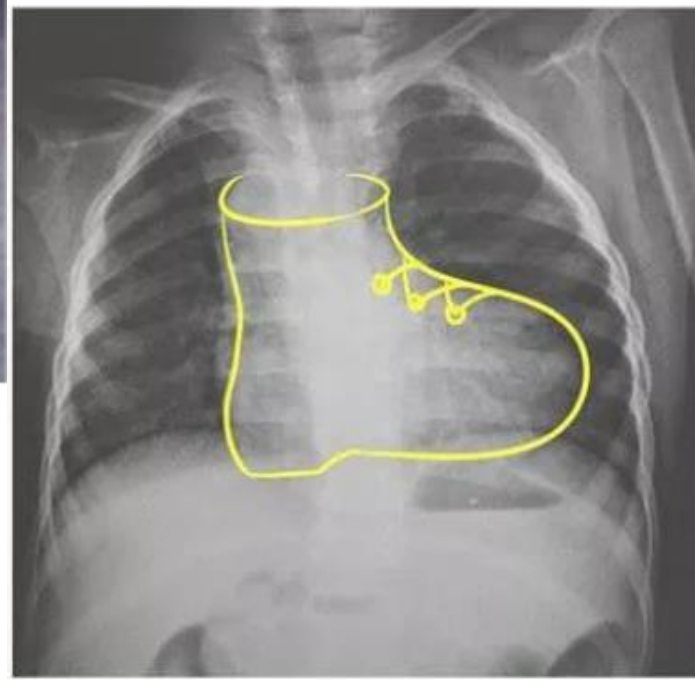
finger clubbing: no space between fingers.





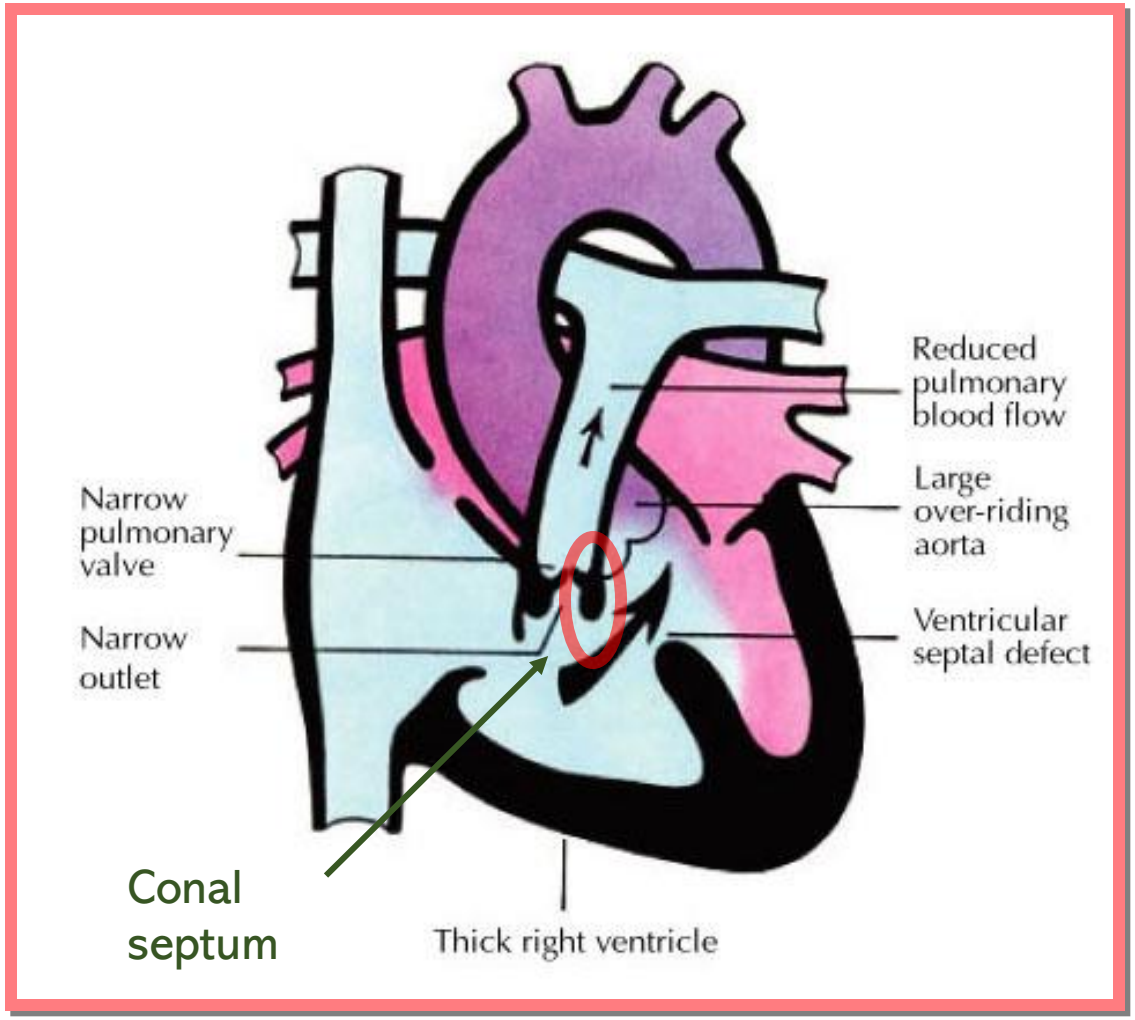


Notice the lung is more black than normal.



Diagnosis: Tetralogy of Fallot

- Embryologic defect: anterior and rightward deviation of the "conal" septum
- Anatomical result:
  - Failure of the muscular ventricular septum to fuse with conal septum (VSD)
  - Sub-pulmonary and pulmonary stenosis (crowdedness under the pulmonary valve)
  - Aorta overrides the septum (it's pulled to the right side, so it appears between the two ventricles)
  - Secondary right ventricular hypertrophy (now both ventricles are pumping with the same pressure)



How does that result in the symptoms we mentioned? Let's talk about the hemodynamics.

Let's assume that this child has 1 cardiac output that meats his demand and go into the systemic circulation, it will come back through the veins into the right atrium, then to the right ventricle, so there will be 1 cardiac output in the right ventricle now.

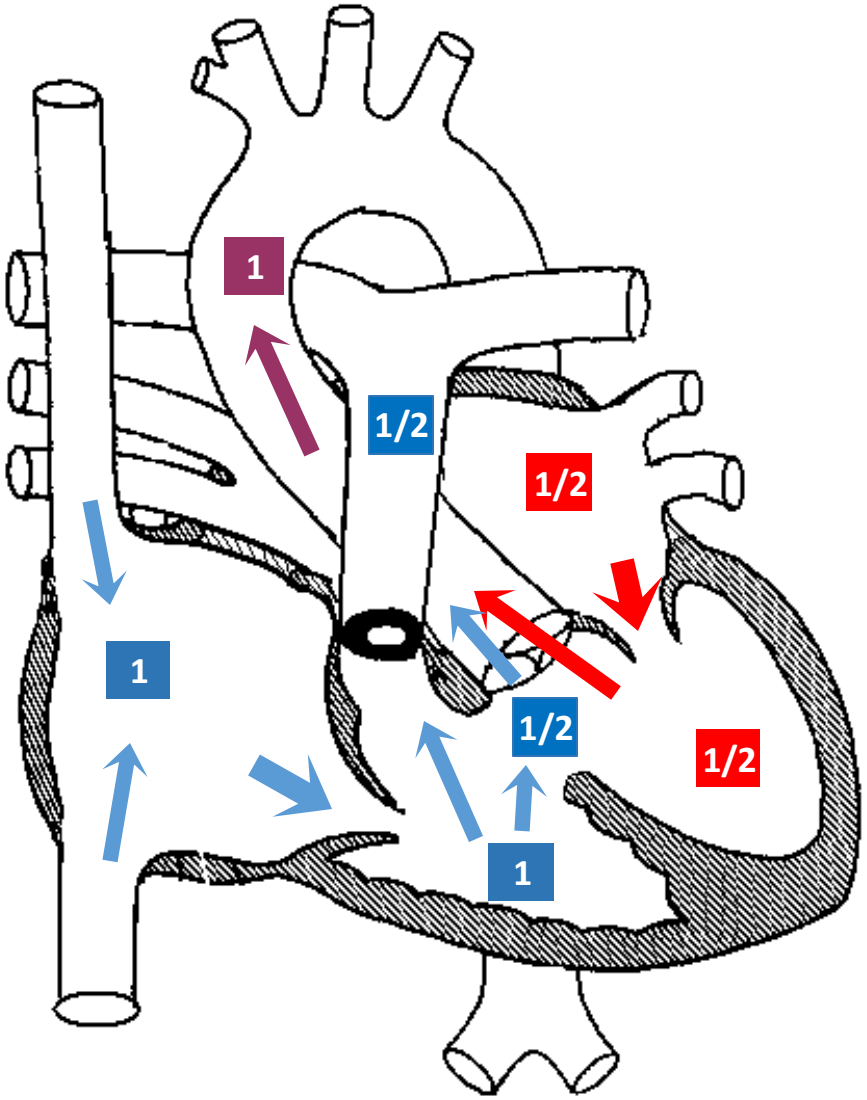
Here we also have VSD, but why this patient doesn't have the same symptoms as the other one?

Here there is a difference which is the **pulmonary stenosis**, so the right ventricle will not be able to pump all the blood because there is an obstruction (fixed or dynamic) in the pulmonary tract. So the shunt will be from **Right to left** because it's easier for the blood to flow this way.

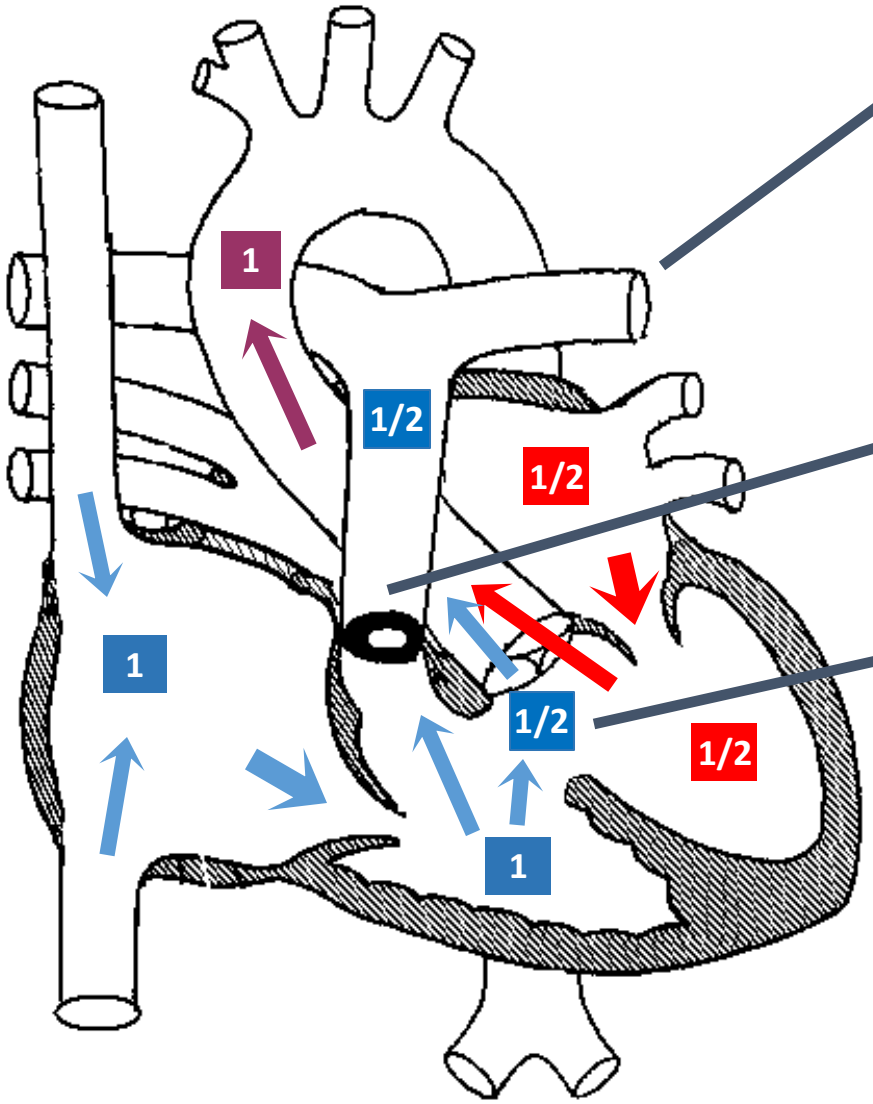
So, let's assume only ½ cardiac output that makes it to the lungs, the other half has to cross the VSD to the aorta.

As for the half that goes into the lung, it will reach the left atrium fully saturated (red) → left ventricle.

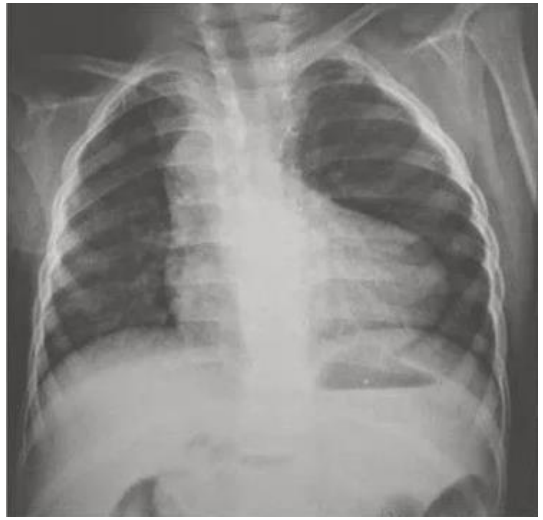
So now, the blood that go into the aorta is a mix between the half from the right ventricle + half from the left ventricle. So you will get 1 cardiac output to the aorta, but it's **mixed** between red and blue.



Physiology of TOF ➔ clinical picture

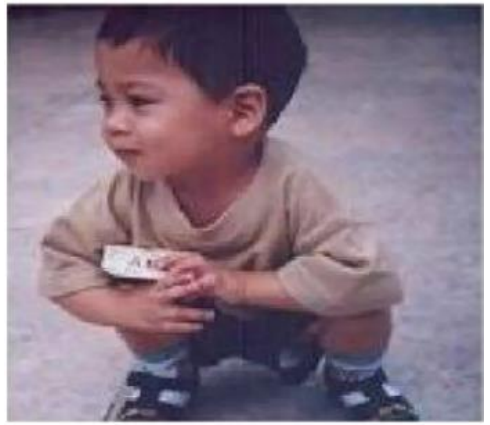


Decreased pulmonary blood flow ➔ No respiratory distress (dyspnea) or rapid breathing (tachypnea), and darker lungs on the X ray (no congestion), small pulmonary artery



Obstruction leads to turbulence of blood flow ➔ systolic murmur (here it's because the pulmonary stenosis, VSD murmur was because of the flow across the VSD itself)

Shunting of deoxygenated blood to the aorta ➔ cyanosis (blood saturation is 80 % not 100 %) ➔ polycythemia, clubbing



Squatting helps the child to decrease the right to left shunting ➔ alleviates the cyanotic episodes

❑ But why does the patient squat when he plays?

Let's say that this child has 80% saturated blood, which is a mix between 100% saturated blood and 60% saturated blood, let's assume this is the baseline.

If he tries to exercise or run around, he's consuming more oxygen, so the 80% that goes by the aorta to the systemic circulation will not come back as 60% saturated blood in the right atrium because he is consuming more oxygen right now, so it will come back with a lower saturation, let's say 40% saturated.

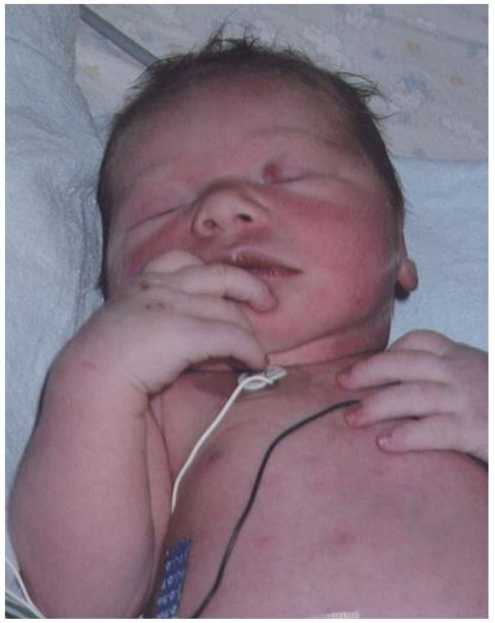
So now you are mixing 40% saturated blood with 100 saturated blood, you will not get 80% saturated blood, you will get less.
So, every time this child works harder, run or have an infection …. (the metabolic demand is higher), his oxygen saturation will drop, and when that happens, he feels he is not right and he is getting bluer, so he squats.

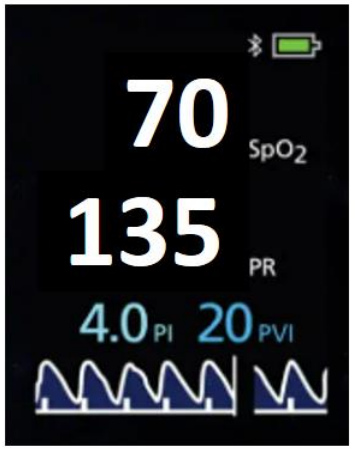
When he squats:

- ❑ He is **stopping** the exercise that he is doing.
- ❑ He is **compressing** large arteries in the lower limb (femoral and external iliac), this will lead to increase in the resistance of the aorta, which prevent some of the blood shunt from right to left (increase the resistance in the systemic circulation, so it works against the shunt), which force the blood to go to the lungs more to get oxygen, so his oxygen saturation will improve.

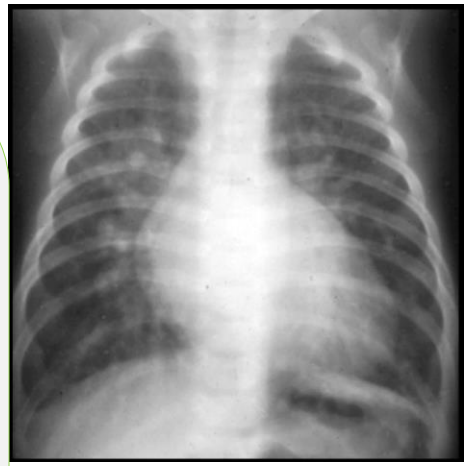
Case 3

- One day old newborn who has:
  - Severe cyanosis
  - No respiratory distress
- Examination:
  - Oxygen saturation of 70%
  - Good pulse and perfusion
  - No murmur
- Chest X ray showed:
  - Egg shaped heart,
  - narrow mediastinum
- Cyanosis is getting worse with time





Worse than the previous case

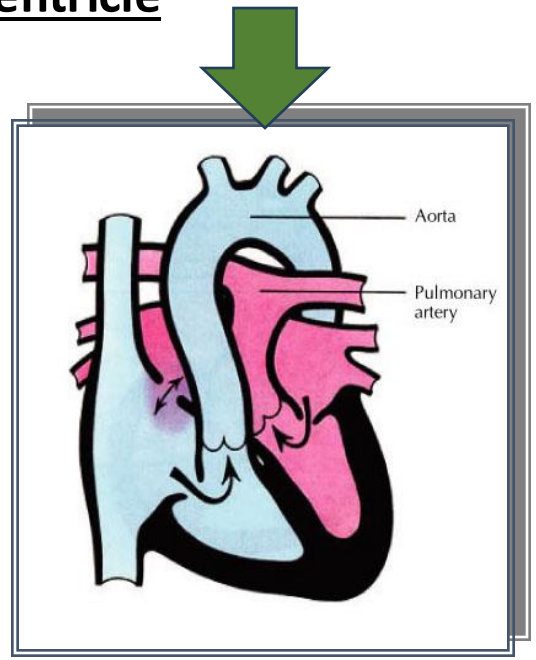


In a typical configuration, the great arteries (aorta and pulmonary artery) are positioned side-by-side within the mediastinum, which contributes to its normal width.

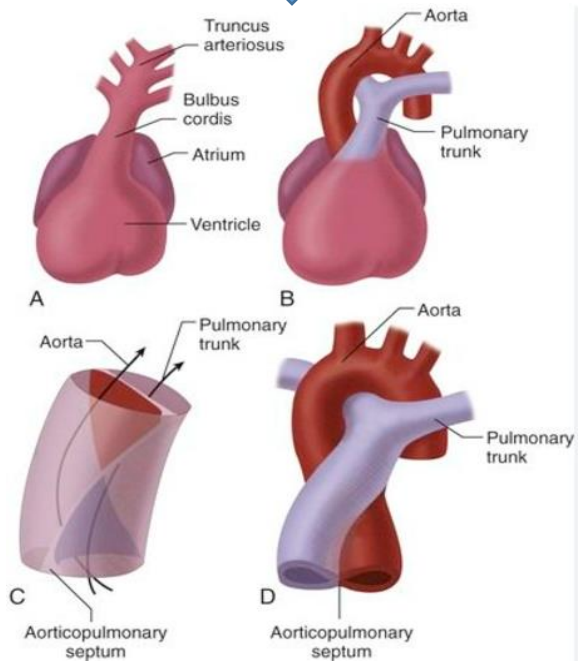
If the great arteries are not side-by-side but instead positioned anterior-posterior, the mediastinum appears narrower on imaging

Diagnosis: Transposition of the Great Arteries

- Embryologic defect:
  - Abnormal septation of the truncus arteriosus (failure of spiral septation)
- Anatomical result:
  - **Aorta** –anterior-- arises from the **right ventricle**
  - **Pulmonary artery** –posterior- arises from the **left ventricle**

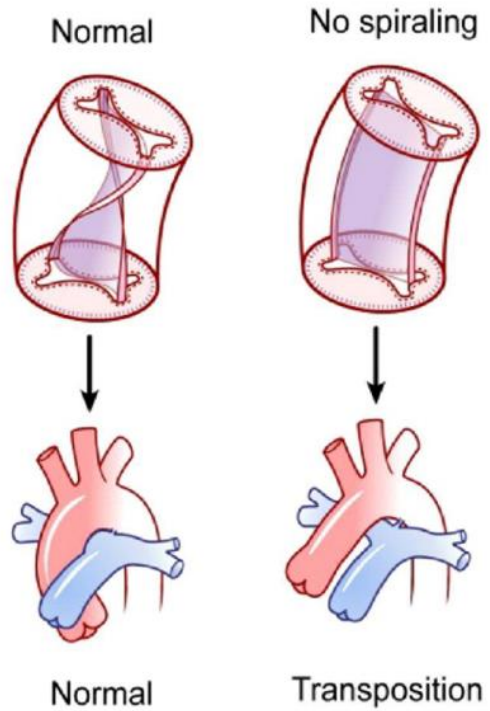


Normal spiraling of AP septum



The pulmonary is pushed anterior and aorta to posterior

Normal vs abnormal spiraling

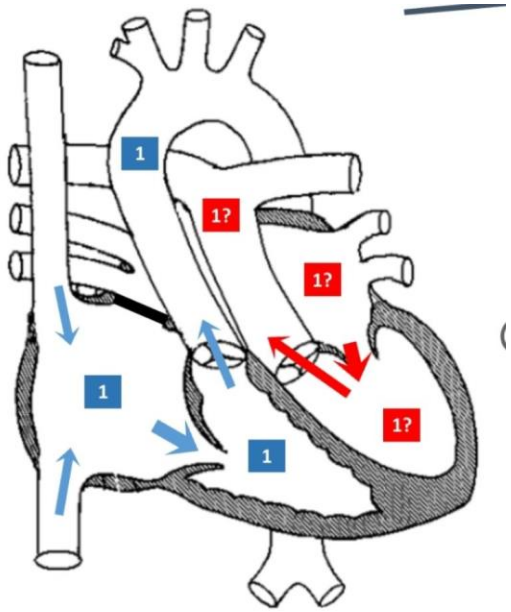


Parallel

Physiology of TGA ➔ clinical picture

Systemic Circulation (blue part):
Blood is pumped by the left ventricle into the aorta, **returning deoxygenated blood** to the right atrium via systemic veins.
This deoxygenated blood then enters the right ventricle and is pumped back into the aorta without passing through the lungs.
With each cycle, oxygen saturation decreases progressively (e.g., from 80% to 60%, then to 40%), leading to critical hypoxia.

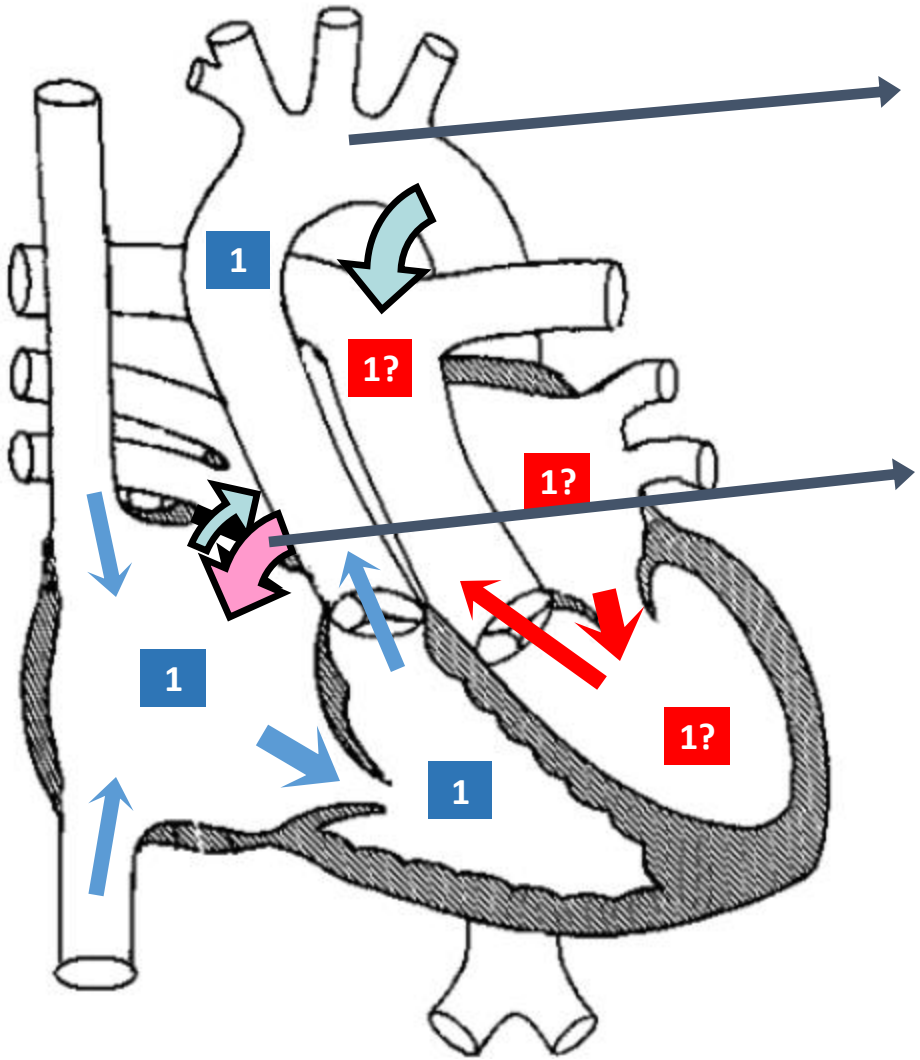
Pulmonary Circulation (red part):
Similarly, **oxygenated blood is recirculated** within the lungs. Blood from the pulmonary veins enters the left atrium, moves to the left ventricle, and is pumped into the pulmonary artery.



The lack of mixing results in two separate, non-communicating circulatory systems.**Unlike** conditions such as Tetralogy of Fallot, where cyanosis develops over time and is manageable for months or years, **neonates with parallel circulation have severe cyanosis birth leads to death after birth, requiring urgent intervention.**

To survive even temporarily, neonates rely on **fetal shunts** that allow mixing of oxygenated and deoxygenated blood

Physiology of TGA ➔ clinical picture



The two circulations are parallel, systemic and pulmonary blood re-circulate, resulting is severe cyanosis

Clinical Interventions 👇 👇
This approach ensures better survival outcomes while the baby awaits definitive surgical repair (Arterial Switch Operation)

For survival, atrial level mixing is mandatory (bidirectional flow)

Allows for some oxygenated blood to body, and deoxygenated blood to lung

Emergency balloon septostomy can be life-saving

Additional shunting via patent ductus arteriosus can also help the baby survive until surgical repair is done

Prostaglandin infusion can be life-saving

Case 4

• 12 year old child complains of headache, and occasional chest pain with exercise.
• He reports cramps of his leg when he walks for long distances.
• Examined by school nurse, and was found to have blood pressure of 160/100 high pressure to a child
• Other findings on examination:
• Good radial / brachial pulse, but poor femoral pulse
• BP in the leg measured 110/70 there is a gradient between the upper and lower limbs
• Cardiac palpation showed strong apical impulse (apical heave)
• ECG showed left ventricular hypertrophy Force of left ventricule on ECG is high
• Chest X ray showed:
• Notched ribs

Blood pressure measurements:
Arms: 160/70 mmHg
Legs: 110/70 mmHg
This demonstrates a significant pressure gradient between the upper and lower extremities.

It was difficult to palpate femoral pulses, a condition known as radiofemoral delay.
Blood pressure measurements:

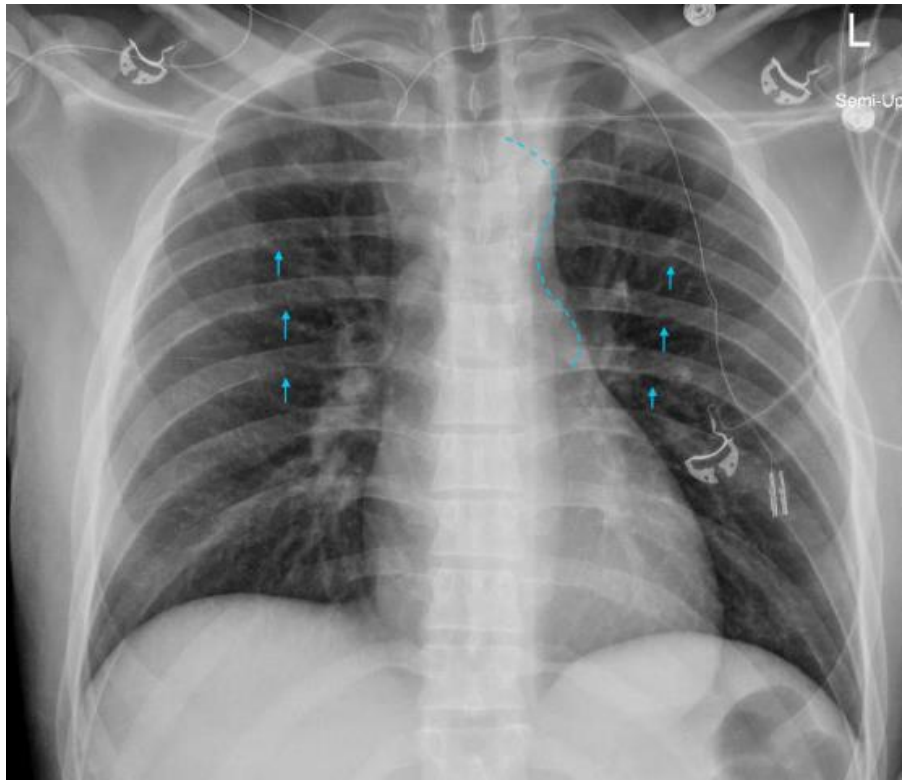
indicating that the left ventricle is hypertrophic and pumping with greater force due to high pressure.

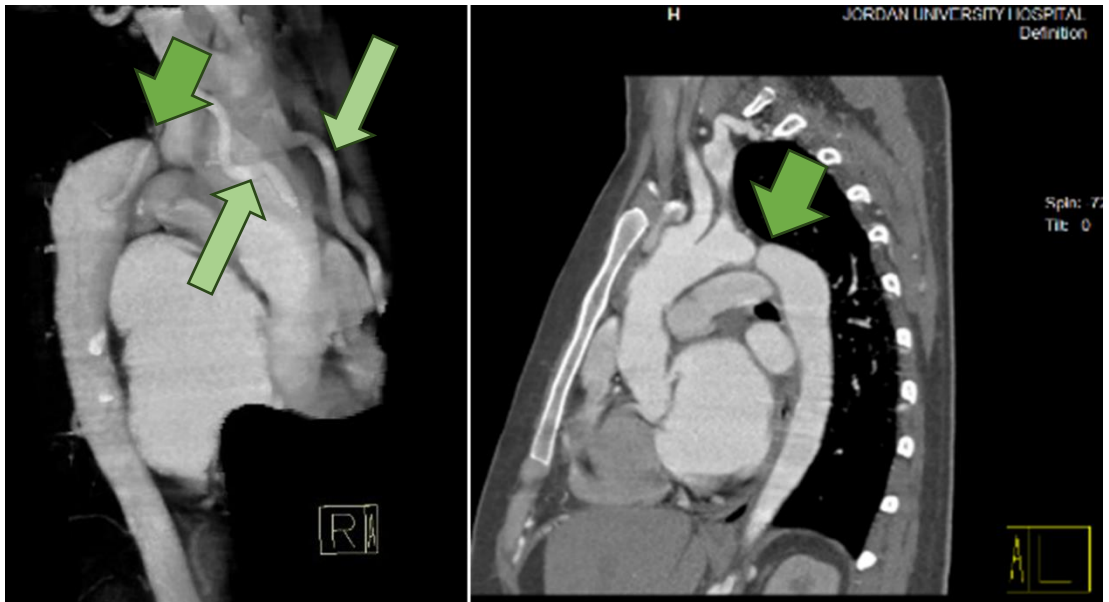
Case 4

- Chest X ray showed:
  - Notched ribs

The ribs showed notching along their undersurface, appearing irregular, as if eroded The heart appeared slightly enlarged, particularly on the left side. This rib notching is a subtle sign that requires vigilance to detect.
A radiologist must carefully assess the x-ray when a significant blood pressure gradient is reported.

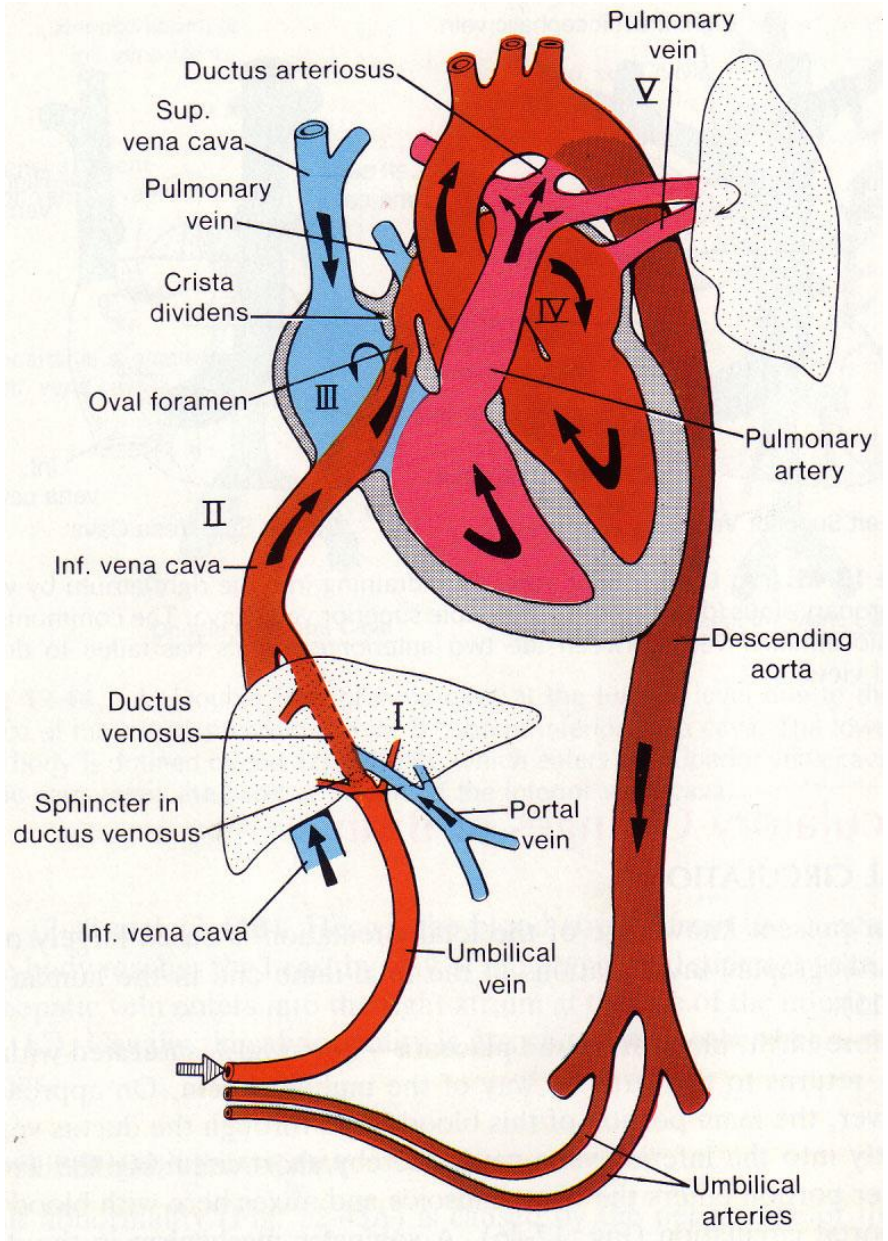
:The CT scan confirmed severe obstruction of the aortic arch. The reconstruction showed the narrowed segment and enlarged collateral blood vessels, such as the internal mammary and intercostal arteries, arising from the subclavian arteries

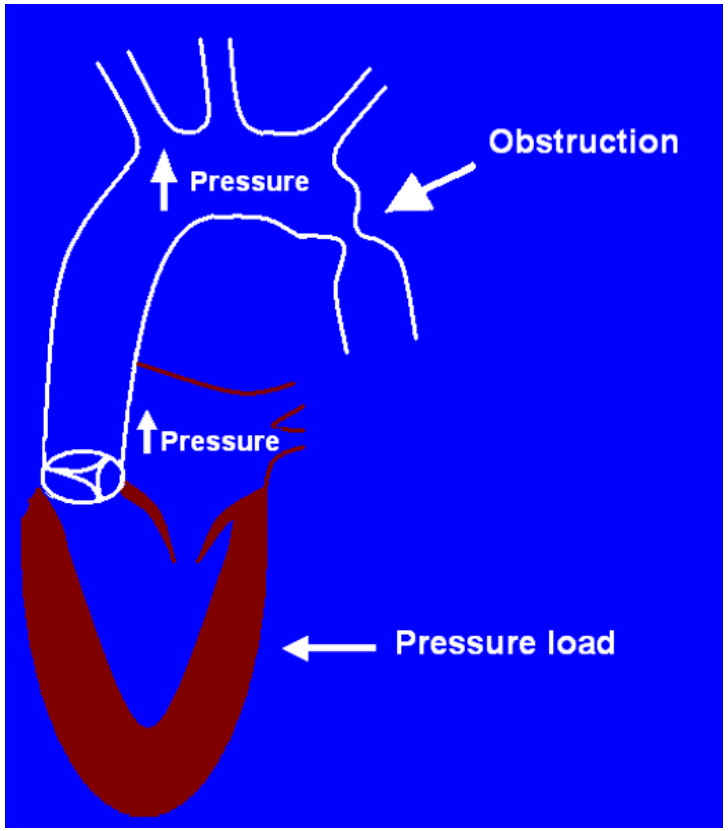




Diagnosis: Coarctation of Aorta

- Embryologic defect: decreased flow across the distal aortic arch during fetal life (most accepted theory)
- Anatomical result:
  - Narrowing of the distal arch (Isthmus), usually distal to the left subclavian artery





- The heart, like most organs, undergoes significant development during the first trimester of gestation,
- However, some **developmental defects** can **occur later**, such as coarctation of the aorta, which is **influenced by fetal blood flow dynamics**.

Normal Fetal Circulation

↓

Oxygenated Blood Enters Right Atrium

↓

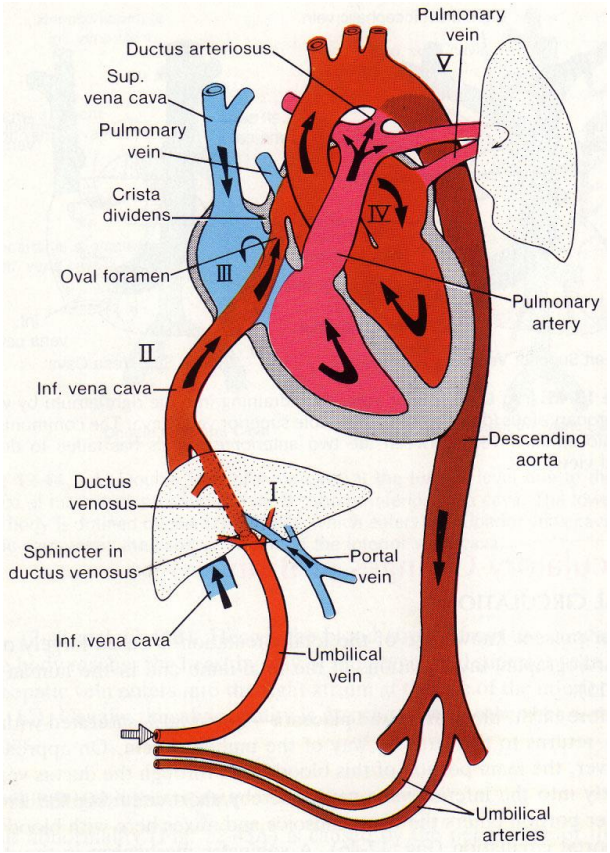
Most Blood Flows Through Foramen Ovale to Left Atrium → **Left Ventricle → aorta →Upper Body & Brain**

↓

Remaining Blood Flows to **Right Ventricle → Pulmonary Artery**

↓

Bypasses Lungs Through Ductus Arteriosus → **Descending Aorta** → **Lower Body** & **Placenta** (via Umbilical Arteries)



Pathophysiology of Fetal Coarctation of the Aorta

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compromised filling of the left atrium and left ventricle

↓

Reduced Blood Flow to Upper Body and Aortic Isthmus

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Insufficient Blood Crossing the Aortic Isthmus

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Developmental Weakness in Distal Aortic Arch

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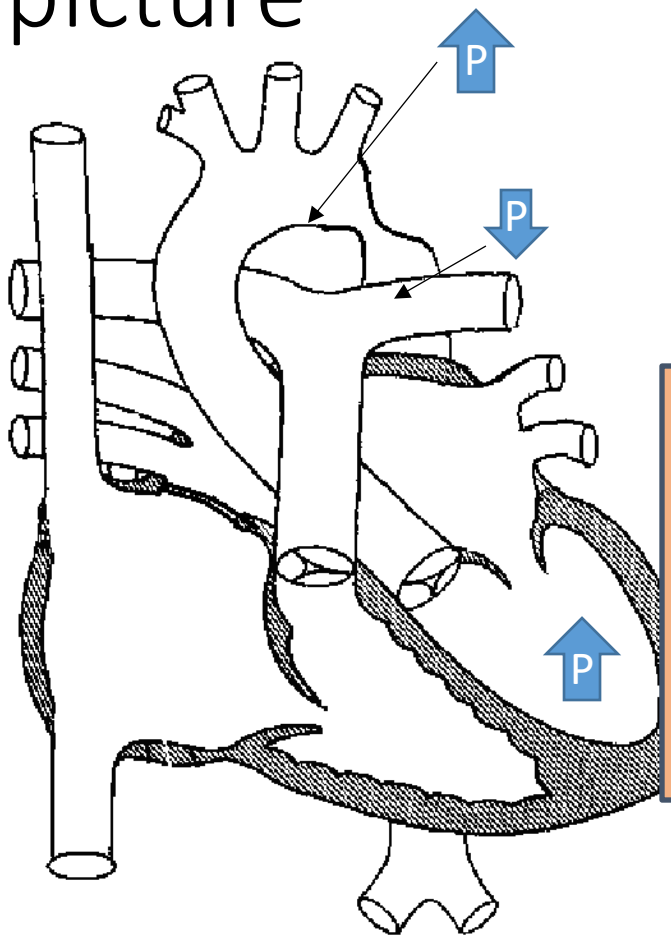
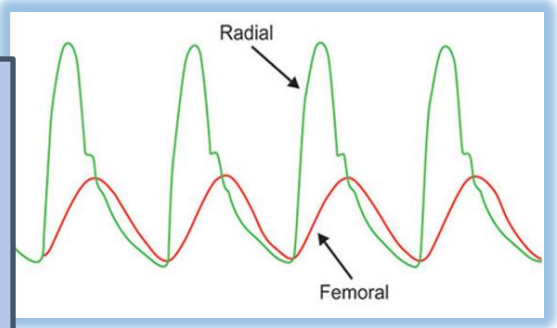
Narrowing of Distal Aortic Arch (Coarctation of the Aorta)

Physiology of Coarctation of Aorta ➔ clinical picture



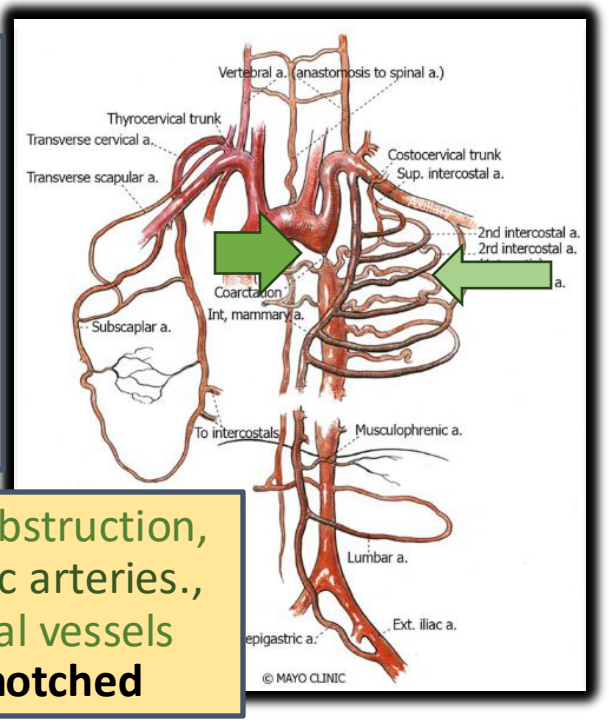


Hypertension in upper limbs, (Proximal to the narrowing) with normal or low pressure in lower limb, (distal to the narrowing)

Coarctation is a recognized secondary cause of **hypertension. Blood pressure measurements often reveal a significant gradient between the radial (high) and femoral (low) pressures.**

Left ventricle becomes hypertrophic to be able to handle the high pressure➔ increased myocardial oxygen demand specifically when exercise ➔ susceptible for exertional ischemia and exertional **angina**

The left ventricle works harder to overcome the resistance caused by the narrowed aortic segment, leading to hypertrophy.



Collateral circulation develops slowly to bypass the obstruction, include the internal mammary arteries, and epigastric arteries., involving the **intercostal arteries** ➔Enlarged collateral vessels erode the undersurface of the ribs, causing **ribs are notched**

Lower limb blood supply may get compromised with exercise leading to muscle pain and cramping (claudications)

Chat GPT

What is the primary hemodynamic abnormality in a Ventricular Septal Defect (VSD)? A. Increased left-to-right shunting of blood B. Increased right-to-left shunting of blood C. Decreased pulmonary blood flow D. Decreased systemic circulation	What is essential for survival in a newborn with Transposition of the Great Arteries (TGA)? A. Development of collateral circulation B. Mixing of oxygenated and deoxygenated blood at the atrial or ductal level C. Closure of the ductus arteriosus D. Complete surgical correction at birth
Why do children with Tetralogy of Fallot often squat during cyanotic episodes? A. To decrease systemic vascular resistance B. To increase systemic vascular resistance and reduce right-to-left shunting C. To increase cardiac output D. To improve oxygen saturation by increasing heart rate	Which of the following diagnostic findings is characteristic of Tetralogy of Fallot on chest X-ray? A. Cardiomegaly with congested lungs B. Egg-shaped heart and narrow mediastinum C. Boot-shaped heart with dark lungs D. Rib notching with left ventricular hypertrophy
Which of the following is a hallmark clinical finding in Coarctation of the Aorta? A. Cyanosis of the upper limbs B. Absence of a murmur C. Rib notching and weak femoral pulses D. Increased blood pressure in the lower limbs	A 12-year-old child presents with hypertension in the upper limbs, weak femoral pulses, and rib notching on X-ray. What is the most likely diagnosis? A. Ventricular Septal Defect B. Coarctation of the Aorta C. Tetralogy of Fallot D. Transposition of the Great Arteries

A,B,C

B,C,B

الحمد لله و لا إلا إله إلا الله و الله أكبر

Additional sources

VERSIONS	SLIDE #	BEFORE CORRECTION	AFTER CORRECTION
V1→ V2			
V2→V3			

امسح الرمز و شاركنا بأفكارك لتحسين أدائنا!!