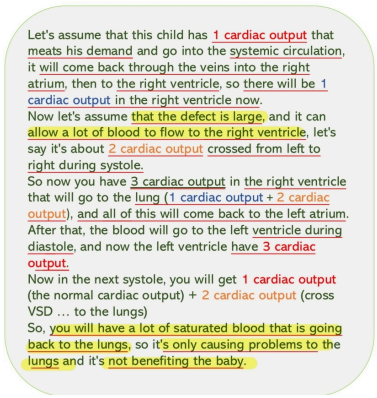
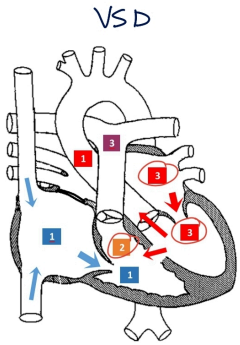
L1 (Clinical Pediatric Cardiology)

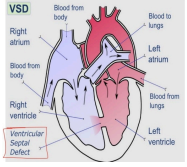
NOTE: Most of Pediatric cardiac diseases are congenital defects.

- Case 1: A two-month-old infant presents rapid breathing (tachypnea), difficulty feeding, and poor weight gain. On examination, the baby shows signs of respiratory distress, including intercostal muscle retraction, a rapid heart rate (tachycardia), weak pulses, and skin hypoperfusion, making the skin cold and pale. A systolic murmur is heard on auscultation. A chest X-ray reveals an enlarged heart (cardiomegaly) and congested lungs, indicating an abnormal increase in pulmonary blood flow.

😊 Firstly: understand the Physiology of VSD:



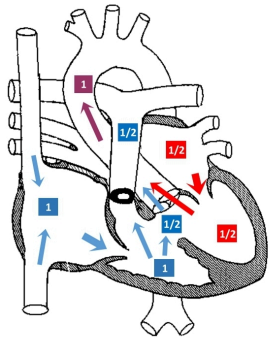


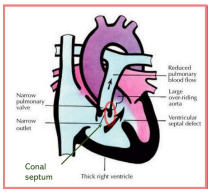
Symptoms and signs	Diagnosis	Embryologic Defect	Anatomical Result	Physiology and Clinical Picture
Tachypnea, difficulty feeding, poor weight gain, respiratory distress, tachycardia, weak pulses, intercostal muscle retraction [Because of the difficulty of breathing, we will see that the intercostal muscles target in when he takes a breath], the skin might be either pale or mottled, mottling of the skin means that there is a net-like pattern that might indicate decreased perfusion of the skin , systolic murmur, enlarged heart (cardiomegaly), congested lungs [there is more fluid in the lungs].	Ventricular Septal Defect (VSD). - 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. 	Failure of ventricular septation during embryogenesis	Defect between ventricles (muscular/membranous types)	1. Left-to-right shunting of blood occurs due to higher pressure in the left ventricle compared to the right ➔congested lungs + stiff lungs ➔ breathing difficulty and rapid breathing (tachypnea). 2. The left atrium and ventricle are overloaded due to excess blood return, causing dilation (cardiomegaly). 3. The systolic murmur results from high-velocity turbulent blood flow through the septal defect. 4. Excessive dilation eventually leads to systolic dysfunction ➔ low tissue perfusion. [Remember 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].

- Case 2: A two-year-old child presents with cyanosis that worsens during physical activity or episodes of crying. The child often squats during cyanotic episodes to alleviate symptoms. Despite these issues, the child is growing well, with no significant difficulty feeding, breathing, or gaining weight. Examination reveals oxygen saturation of 80% (normal >95%), a plethoric (congested, purplish) facial appearance, and finger clubbing, a sign of chronic cyanosis. A systolic murmur is noted on auscultation. A chest X-ray shows a boot-shaped heart and dark lungs, indicating decreased pulmonary blood flow. Hemoglobin levels are elevated at 17 g/dL (polycythemia), a response to chronic hypoxia. No murmur.

☺ Firstly: understand the Physiology of TOF:

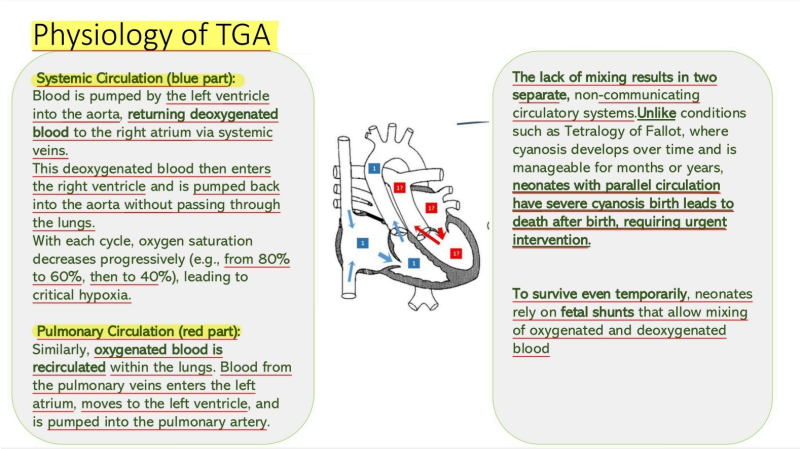
How does that result in the symptoms we mentioned? Let's talk about the hemodynamics.
Let's assume that this child has 1 cardiac output that meats his demand and go into the systemic circulation, it will come back through the veins into the right atrium, then to the right ventricle, so there will be 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.



Symptoms and signs	Diagnosis	Embryologic Defect	Anatomical Result	Physiology and Clinical Picture
Cyanosis, plethoric face, finger clubbing [sign of chronic cyanosis] (it means losing the angle between the nail and the nail bed), oxygen saturation 80%, elevated hemoglobin level boot-shaped heart and dark lungs.	Tetralogy of Fallot (TOF).	Anterior and rightward deviation of the conal septum. 	VSD, sub-pulmonary and pulmonary stenosis [crowdedness under the pulmonary valve], overriding aorta [it's pulled to the right side, so it appears between the two ventricles], and right ventricular hypertrophy [both ventricles are pumping with the same pressure].	1. Right-to-left shunting occurs due to pulmonary stenosis, preventing adequate blood flow to the lungs. 2. Deoxygenated blood enters the aorta, causing cyanosis. 3. Chronic hypoxia leads to compensatory polycythemia and finger clubbing. 4. Reduced pulmonary flow results in dark lungs on X-ray and no respiratory distress. 5. The systolic murmur arises from turbulent blood flow due to pulmonary stenosis. 6. Squatting increases systemic vascular resistance, reducing shunting and improving oxygenation.

- Case 3: A one-day-old newborn is brought to medical attention with severe cyanosis since birth. The baby shows no signs of respiratory distress but has an oxygen saturation of only 70%. A chest X-ray reveals an egg-shaped heart with a narrow mediastinum. Cyanosis worsens progressively due to the absence of adequate mixing between the systemic and pulmonary circulation.

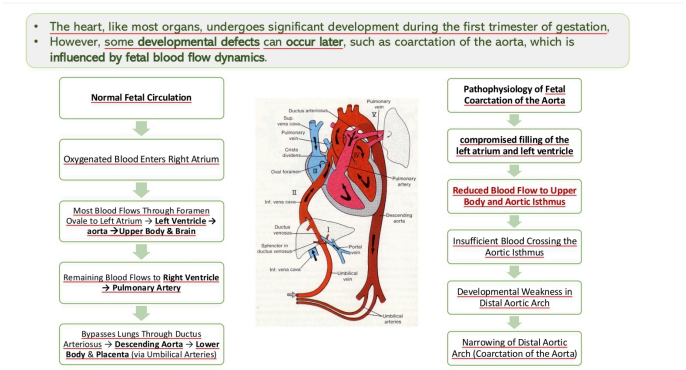
😊 Firstly: understand the Physiology of TGA:



Symptoms and signs	Diagnosis	Embryologic Defect	Anatomical Result	Physiology and Clinical Picture
Severe cyanosis since birth, oxygen saturation of 70%, no respiratory distress, egg-shaped heart and narrow mediastinum. 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	Transposition of the Great Arteries (TGA)	Failure of spiral septation of the truncus arteriosus.	**Aorta** arises from the **right ventricle; pulmonary** artery arises from the **left ventricle**.	1. The pulmonary and systemic circulations are parallel and non-communicating resulting in severe cyanosis. 2. Deoxygenated blood recirculates in the systemic circuit, progressively worsening hypoxia and Oxygenated blood recirculates in the pulmonary circuit, without systemic benefit. 3. Survival depends on fetal shunts (e.g., patent ductus arteriosus or atrial septal defect) to allow blood mixing until surgical correction (arterial switch operation). 4. Emergency balloon septostomy and prostaglandin infusion can be lifesaving.

- Case 4: A 12-year-old child presents with hypertension (160/100 mmHg in the upper limbs) and complaints of headaches, chest pain during exercise, and leg cramps after walking long distances. On examination, radial and brachial pulses are normal, but femoral pulses are weak (radio femoral delay). Blood pressure measurements reveal a significant gradient, with 160/70 mmHg in the arms and 110/70 mmHg in the legs. Cardiac examination shows a strong apical impulse (apical heave). A chest X-ray demonstrates rib notching caused by enlarged collateral vessels and mild left ventricular hypertrophy.

😊 Firstly: understand the Physiology of Coarctation of the Aorta:



Symptoms and signs	Diagnosis	Embryologic Defect	Anatomical Result	Physiology and Clinical Picture
Hypertension in the upper limbs, weak femoral pulses (radio femoral delay), rib notching, mild left ventricular hypertrophy, leg cramps during exercise, chest pain, and headaches.	Coarctation of the Aorta	Reduced flow across the distal aortic arch during fetal development.	Narrowing of the distal aortic arch (usually distal to the left subclavian artery).	1. Hypertension in upper limbs, (Proximal to the narrowing) with normal or low pressure in lower limb, (distal to the narrowing). 2. Left ventricle becomes hypertrophic to be able to handle the high pressure ➔ increased myocardial oxygen demand ➔ susceptible to exertional ischemia and exertional angina. Lower limb ischemia results in claudication and leg cramps during activity 3. Collateral circulation forms over time, bypassing the obstruction include the internal mammary arteries, and epigastric arteries involving the intercostal arteries but eroding the ribs (rib notching).

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