

Critical Heart Disease in the Newborn

What you need to know

DISCLOSURES

- Nothing to report

OBJECTIVES

- DESCRIBE NEONATAL CARDIOVASCULAR PHYSIOLOGY
- RECOGNIZE NEONATAL CARDIAC EMERGENCIES
- FORMULATE TREATMENT PLANS

DEFINITION

- Critical congenital heart disease
 - all cardiovascular lesions that would result in neonatal demise unless immediate intervention to palliate or correct the anatomic defect is undertaken
- Signs and symptoms of severe heart disease in the newborn
 - cyanosis
 - discrepant pulses and blood pressures
 - congestive heart failure
 - cardiogenic shock

GENERAL PRINCIPLES OF TREATMENT

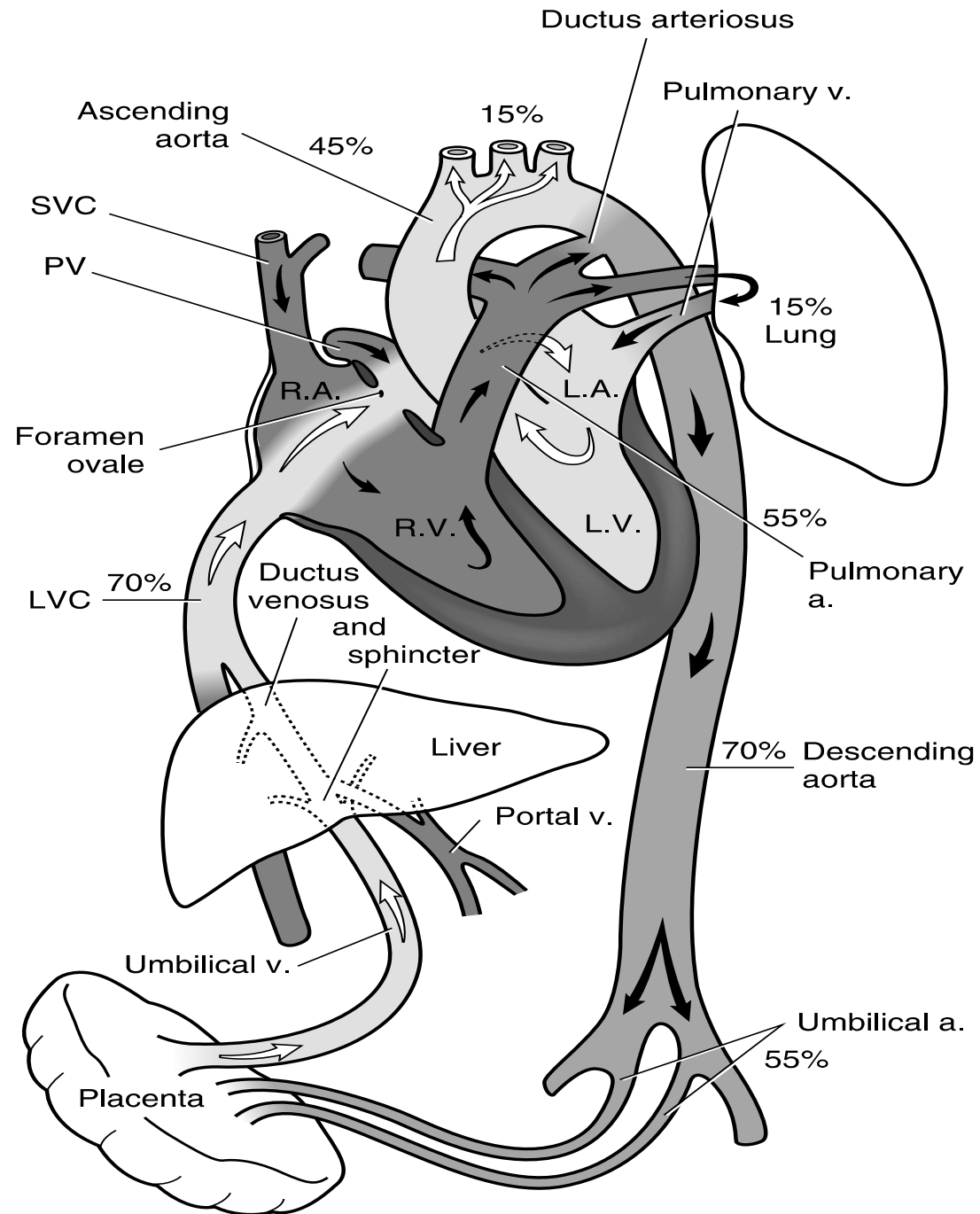
- Early diagnosis is the key
- High index of suspicion is very important
- ABCs trump other considerations, take care of the airway and treat shock
- Start prostaglandins early, don't wait for cardiologist or echocardiogram
- Volume resuscitate
- Correct acidosis
- Inotropic support
- Keep sats in the 75-85 range, be gentle with oxygen

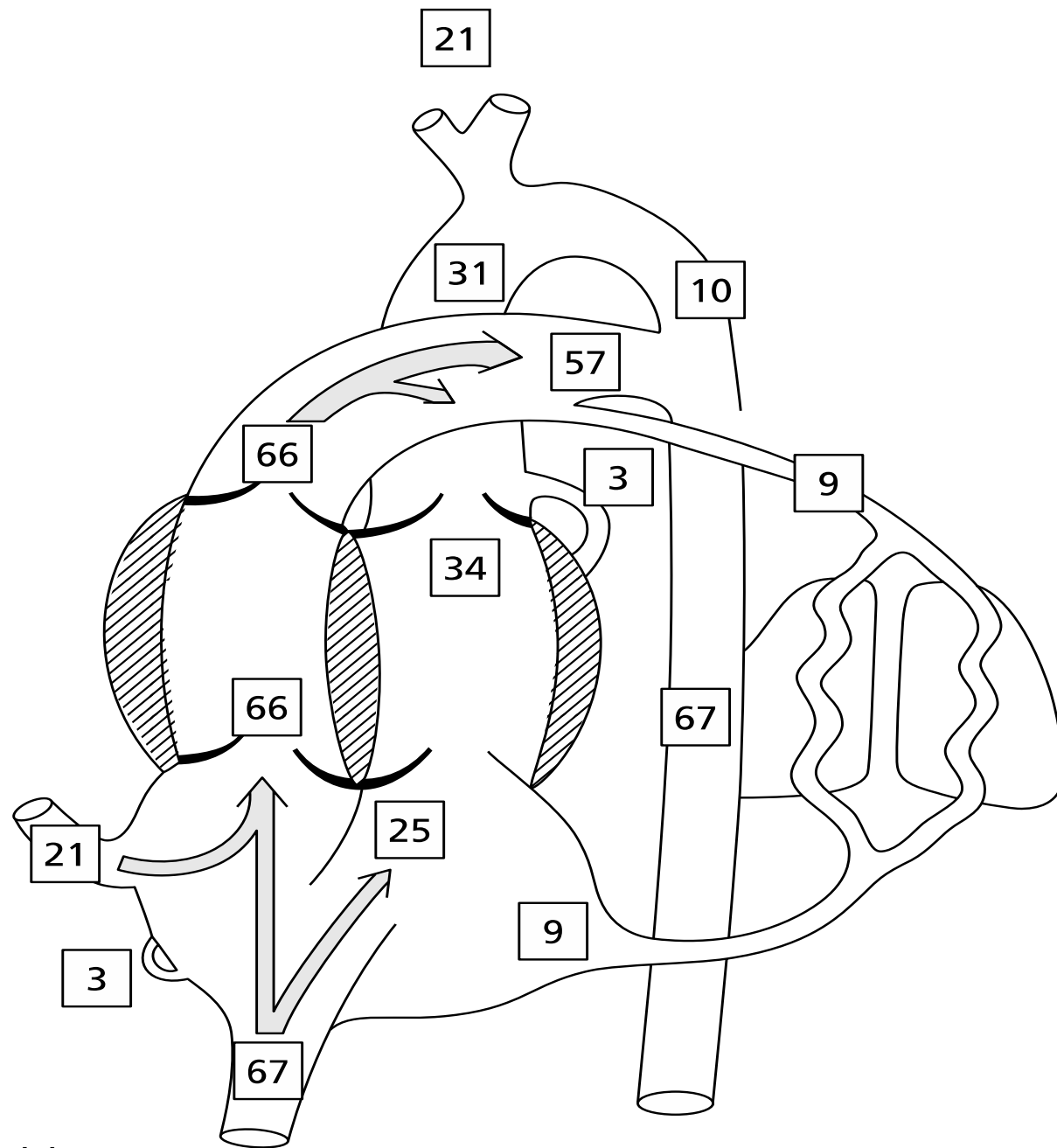
EPIDEMIOLOGY

- In the United States
 - congenital heart disease (CHD) occurs in 8/1000 live births
 - the subpopulation with critical CHD is roughly 3.5/1000 live births
 - approximately 32,000 children each year are born with CHD, and
 - 14,000 of them are born with critical heart lesions

FETAL CIRCULATION

- critical heart disease is well tolerated in utero but is uniformly fatal postnatally without intervention
- the ventricles work together to deliver blood to systemic tissues
- placenta serves as the organ of oxygen delivery
- the umbilical vein carries oxygenated blood from the placenta to the inferior vena cava through the ductus venosus
- a series of central shunts and preferential streaming patterns, directs oxygenated blood to vital organs
- deoxygenated blood is diverted to organs with lower oxygen consumption and to the placenta





TRANSITIONAL CIRCULATION

- at birth, with separation of the umbilical cord, the responsibility of oxygenation shifts from the placenta to the lungs
- in order for this to occur, PVR must fall rapidly
- physical expansion of the lungs and the replacement of fluid-filled alveoli with gas promotes the dilation and distention of the pulmonary arteries decreasing PVR
- mechanical distention of the lungs promote local production of prostacyclin, a pulmonary artery vasodilator, further decreasing PVR
- increased oxygen tension in the pulmonary artery acts as a vasodilator both directly and through its ability to stimulate nitric oxide production

TRANSITIONAL CIRCULATION

- when the placenta is removed from the circulation, blood return to the heart through the inferior vena cava is significantly diminished, causing right atrial pressures to fall
- the increase in Q_p brings about an increase in pulmonary venous return and subsequent elevation in left atrial pressures, the flap over the foramen ovale closes
- there is a dramatic reduction in the production of prostaglandin E2 (by the placenta) and an increase in its metabolism (by the lungs) in combination with increased oxygen content in the blood, provides the stimulus for the PDA to constrict
- the neonatal circulation transitions to a series configuration, thereby establishing separate systemic and pulmonary circulations

NEONATAL CIRCULATION

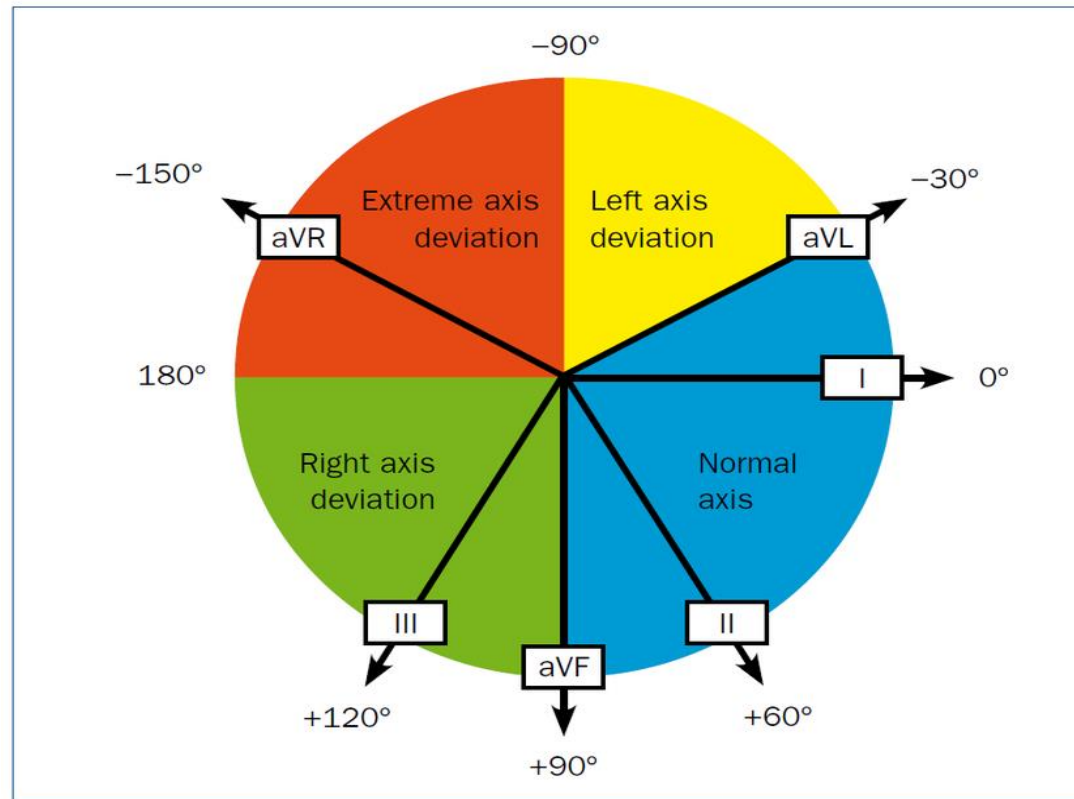
- cardiac output (CO) is directly proportional to the heart rate (HR) and stroke volume (SV): $CO = SV \times HR$
- stroke volume is dependent on three determinants
 - (1) preload, or the distention of the ventricle prior to systole
 - (2) afterload, or the resistance to ejection from the ventricle, and
 - (3) myocardial contractility
- neonatal myocardium is relatively stiff and has fewer contractile myofibrils compared to adults
- the newborn cannot increase cardiac output by increasing stroke volume, and relies mainly on increases in heart rate
- neonatal myocytes are deficient in sarcoplasmic reticulum calcium stores, newborn cardiac output is exquisitely sensitive to calcium
- calcium is an inotrope for the newborn

Basic Evaluation of the Newborn for Congenital Heart Disease

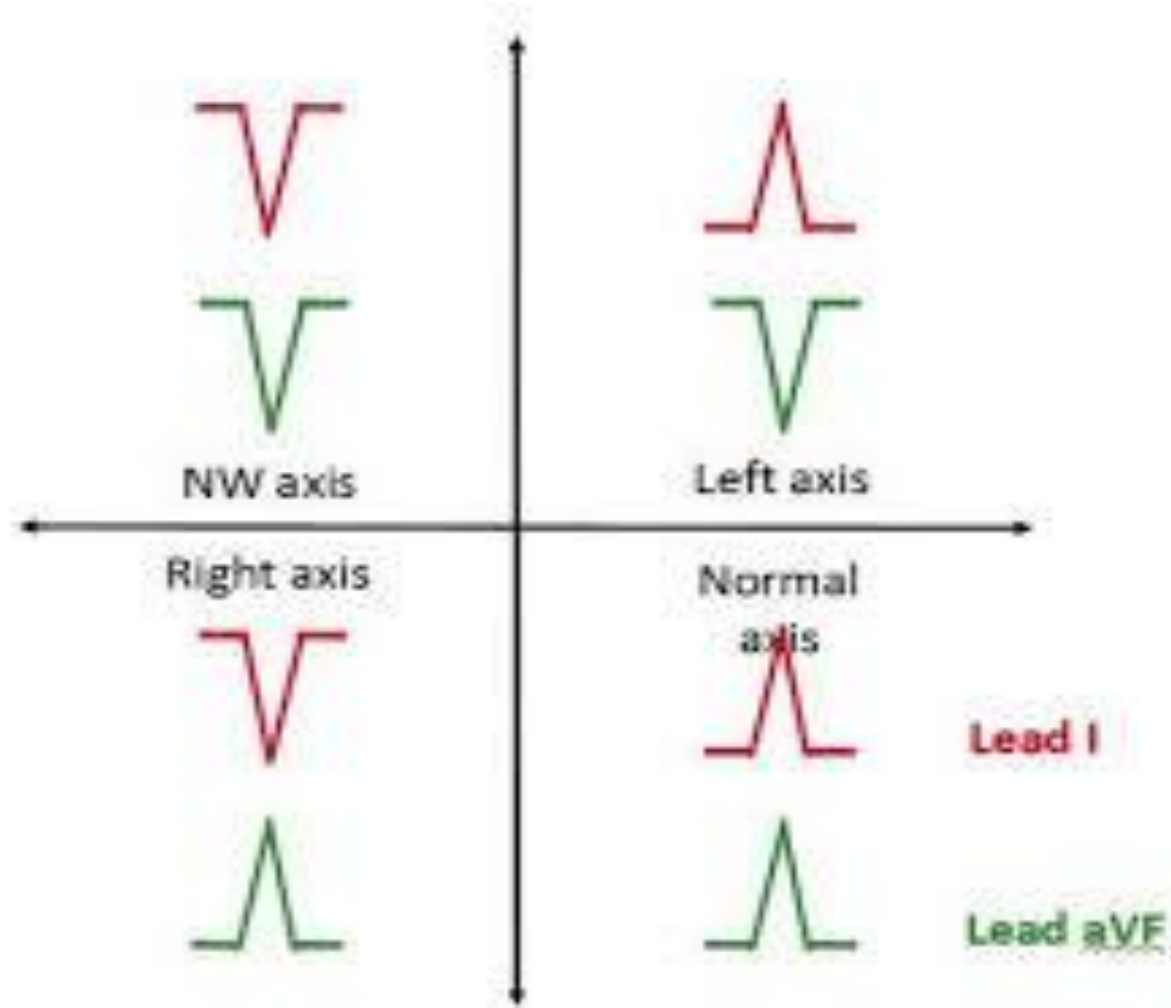
- **EXAM:**
 - Systolic murmur:
 - valvar stenosis (pulmonic or aortic) (Ejection Systolic)
 - tricuspid or mitral regurgitation (Holosystolic)
 - Diminished pulses and BP:
 - Diminished lower extremity pulse and BP: Coarctation
 - Diminished four-extremity pulse and BP: Left-sided obstructive lesions
 - Tachypnea:
 - High Qp:Qs
 - Diminished left ventricular function

Basic Evaluation of the Newborn for Congenital Heart Disease

- ECG:
 - Check for sinus rhythm
 - Superior axis: consider atrioventricular septal defect or tricuspid atresia



EKG AXIS DETERMINATION



Basic Evaluation of the Newborn for Congenital Heart Disease

- CXR

- Severe cardiomegaly: consider neonatal Ebstein's or cardiomyopathy
- Look for right aortic arch
- Look for normal abdominal situs (stomach on left)
- Hypoxemia with normal lung fields: Consider congenital heart disease
- Progressive interstitial pattern: Consider obstruction to pulmonary venous return

Basic Evaluation of the Newborn for Congenital Heart Disease

- **OXYGEN SATURATIONS:**

- Differential cyanosis: Consider pulmonary hypertension (PPHN), coarctation, or interrupted arch
- Reverse differential cyanosis: Consider the above with transposition of the great arteries

- **ARTERIAL BLOOD GAS:**

- Hyperoxia test: PaO₂ in right radial artery on 100% FiO₂ with less than 150 mmHg: Consider intracardiac mixing
- Hypoxemia that improves markedly with oxygen: Consider lung disease

PROBLEM 1

- A newborn infant with prenatal diagnosis of hypoplastic left heart syndrome is on PGE1 infusion, D3 of life, not intubated, waiting for Norwood procedure. He is on 2L/min nasal canula oxygen, sats are 90%. He is not being fed, is on maintenance D10 iv fluids. You notice that his BP has been progressively decreasing, his extremities are cold, cap refill is prolonged. What is going on? What should you do now?
- 1. start TPN
- 2. intubate
- 3. start Dopamine
- 4. discontinue nasal canula O2
- 5. take him to the OR immediately

Qp:Qs – Ratio of Pulmonary to Systemic Perfusion – Single Ventricle Physiology

- newborns with critical CHD whose PDA is kept open with PGE1, balancing Qs and Qp is essential
- particularly important for patients with single-ventricle physiology and complete intracardiac mixing
- systemic venous return (desaturated blood) and pulmonary venous return (saturated blood) usually completely mix within the heart
- there is competitive Qp and Qs blood flow and the relative resistances to flow govern the ratio of distribution of flow between the two circuits

Qp:Qs and Oxygen

- oxygen must be used with caution in the neonate with congenital heart disease, particularly those with single-ventricle physiology
- oxygen is a potent pulmonary vasodilator and will increase pulm circulation (Qp) at the expense of the systemic circulation (Qs)
- oxygen should be minimized in ductal-dependent congenital heart disease
- should be given when there is concurrent underlying lung disease
- may be required in newborns with cyanotic right-sided obstructive lesions and cyanotic transposition of the great arteries
- In general, **arterial oxygen saturations should be maintained between 80% and 85%,** which in a neonate with single-ventricle physiology and good cardiac output **translates to a balanced circulation with a Qp:Qs ratio of 1:1.**

Qp:Qs – Ratio of Pulmonary to Systemic Perfusion – Single Ventricle Physiology

- management goal of patients with single ventricle physiology
 - provide adequate pulmonary blood flow
 - without compromising systemic oxygen delivery and tissue perfusion
- $Qp:Qs = \text{Aortic Sats} - \text{Mixed venous Sat} / \text{Pulm v sat} - \text{Pulm a sat}$
- $Qp:Qs = 25 / 95 - \text{Ao sat}$
- $\text{Ao sat} = 90$
- $Qp:Qs = 25 / 95 - 90 = 25 / 5 = 5:1$

Qp:Qs – Ratio of Pulmonary to Systemic Perfusion – Single Ventricle Physiology

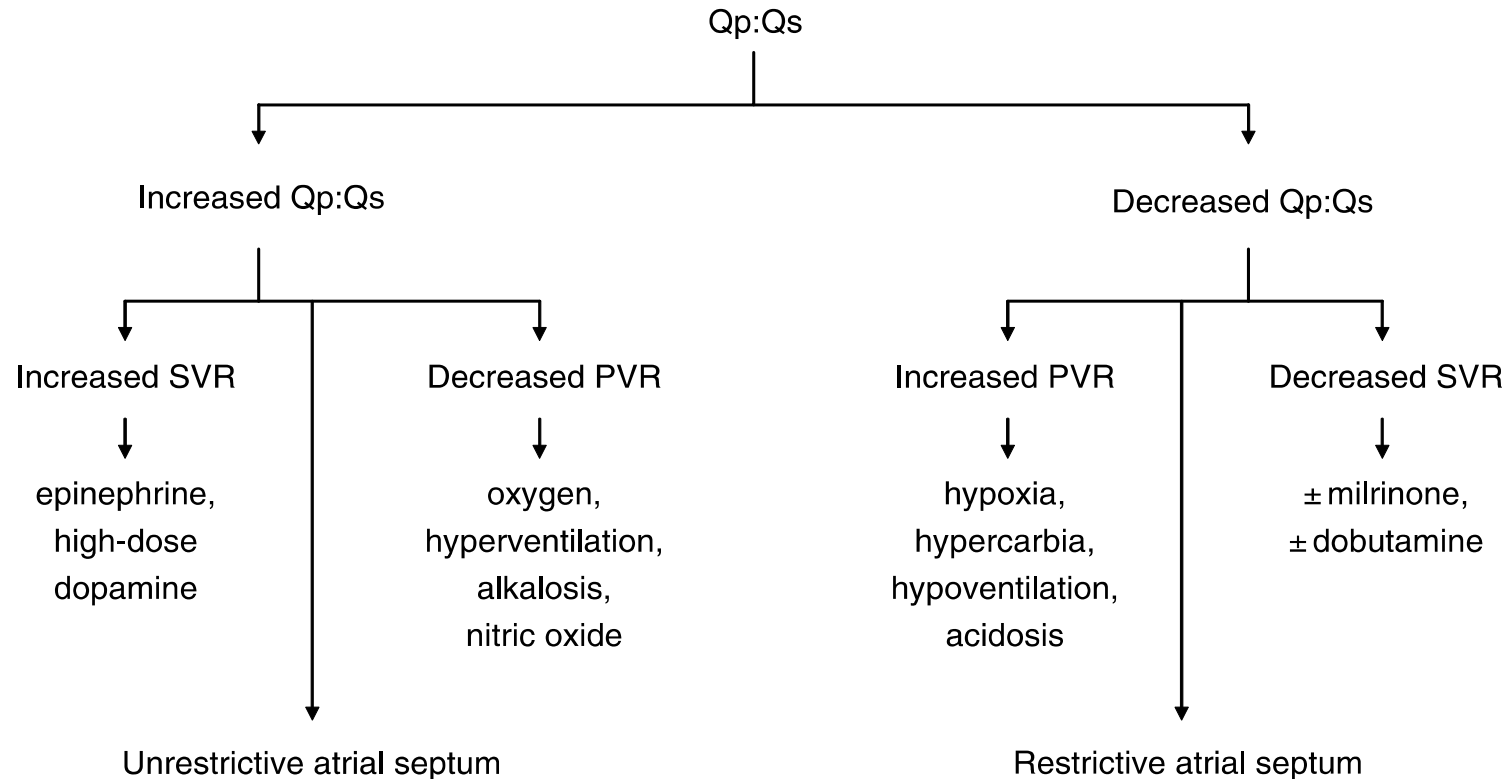


Figure 2-3 The balance between Qp and Qs blood flow is dependent on the relative resistances of the systemic (SVR) and pulmonary vasculature (PVR). Maneuvers that lower PVR (oxygen, hyperventilation, and alkalosis) will increase Qp:Qs at the expense of systemic perfusion. Maneuvers that increase PVR (hypoxia, hypercarbia, acidosis) will decrease Qp:Qs and improve systemic perfusion. Other factors being equal, an unrestrictive atrial septum will encourage pulmonary blood flow, whereas restriction at the atrial septum will help to limit pulmonary blood flow.

The Cost of a High Systemic Saturation in Single Ventricle Physiology

Systemic Arterial Saturation	Systemic Venous Saturation	Pulmonary Venous Saturation	Qp:Qs	Work Imposed on the Single Ventricle (Qp + Qs)
65%	40%	98%	0.75:1	1.75 cardiac outputs
73%	48%	98%	1:1	2 cardiac outputs
86%	61%	98%	2:1	3 cardiac outputs
90%	65%	98%	3:1	4 cardiac outputs
92%	67%	98%	4:1	5 cardiac outputs

Qp, Pulmonary blood flow; *Qs*, systemic blood flow; *Qp:Qs*, the ratio of pulmonary to systemic blood flow.

*The systemic venous saturation is usually approximately 25 percentage points lower than the systemic arterial saturation if the cardiac output and hemoglobin are normal.

PROBLEM 2

- Term newborn in delivery room, called to see for low sats. Difficult resuscitation, sats in the 50s and 60s at 10 mins, even after bagging with 100% O₂, poor peripheral pulses, hypotensive, prolonged capillary refill. Intubated and transferred to the NICU. Lines placed, cultures sent, antibiotics given. ABG pH 7.1 PaCO₂ 29 PaO₂ 27 HCO₃ 12, BD -16. Lactate 8. CXR shows bilateral pulm edema. Cardiac lesion suspected, awaiting Echocardiogram. Prostaglandin infusion started, but no improvement. What is going on?

Timing of Presentation of Critical CHD

- Shock in delivery room
- Symptoms on first day of life
- Symptoms in the first week of life

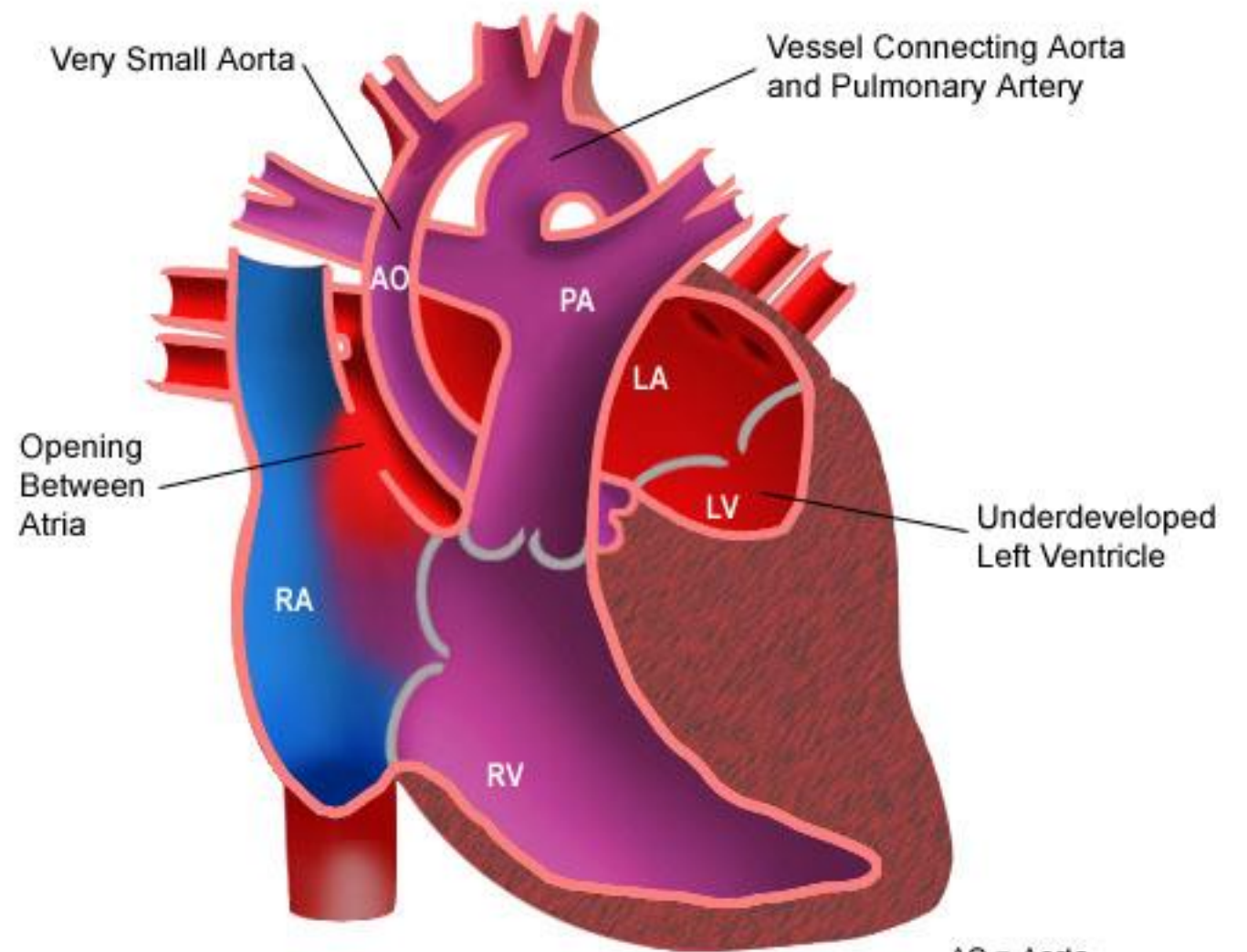
Shock in delivery room

- Cardiac lesions that are unstable in the delivery room
 - represent severe abnormalities of oxygen delivery
 - that are often not stabilized by PGE1 alone and
 - require immediate intervention in order to sustain life
- Hypoplastic Left Heart Syndrome with **Intact Atrial Septum**
- Transposition of the Great Arteries with **Restrictive or Intact Atrial Septum**

Hypoplastic Left Heart Syndrome

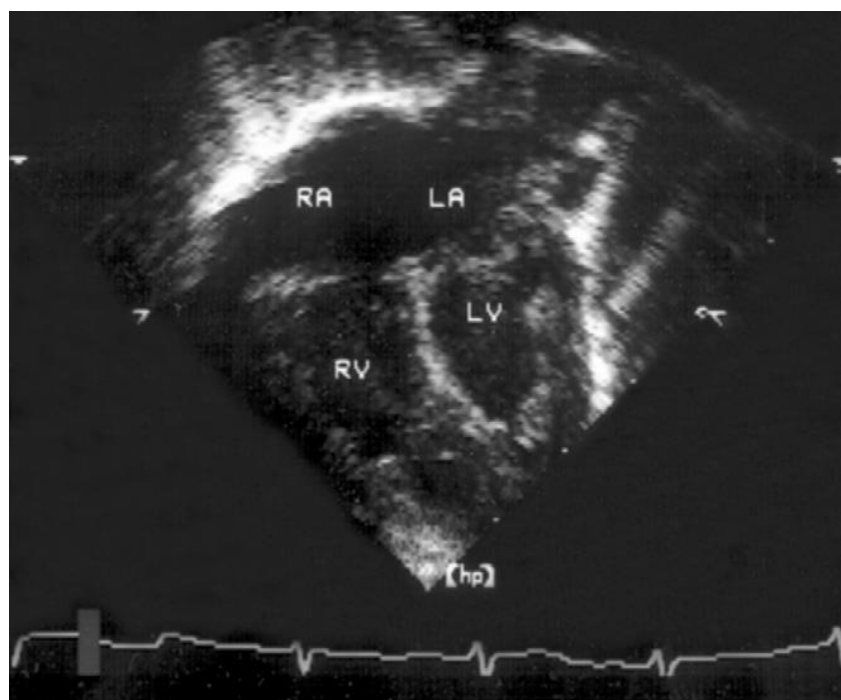
- underdevelopment of the mitral valve, LV, left ventricular outflow tract, aortic valve, and aorta
- the right ventricle is responsible for maintaining both pulmonary and systemic circulation, systemic perfusion is dependent on the PDA – **single ventricle physiology**
- majority present within the first week, when the PDA closes, with signs and symptoms of shock
- **If an intact or restrictive atrial septum (IAS) is present, effective egress from the left atrium is not possible – presents at birth**
- pulmonary venous obstruction develops and causes pulmonary hypertension

Hypoplastic Left Heart Syndrome

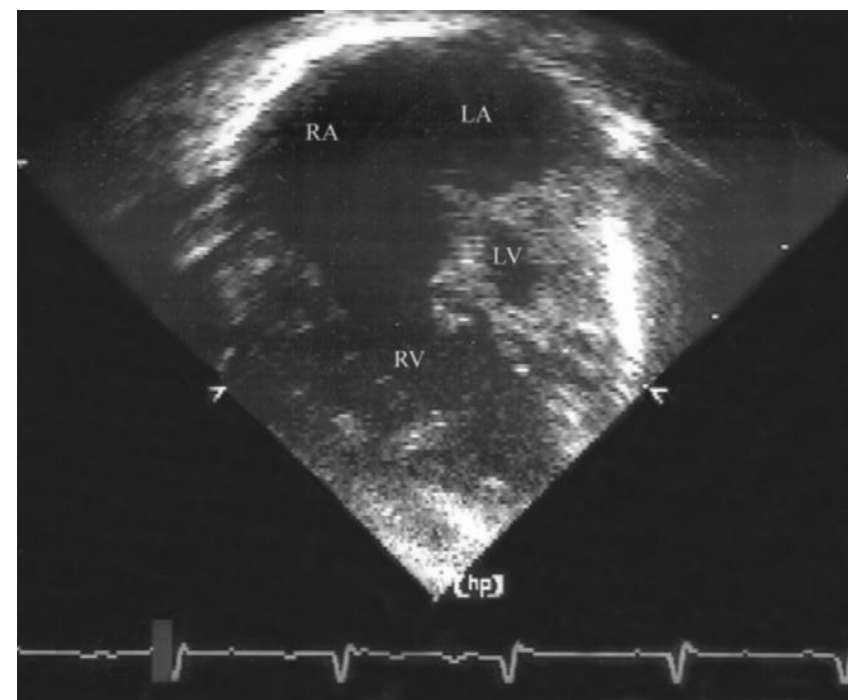


■ Oxygen-rich Blood
■ Oxygen-poor Blood
■ Mixed Blood

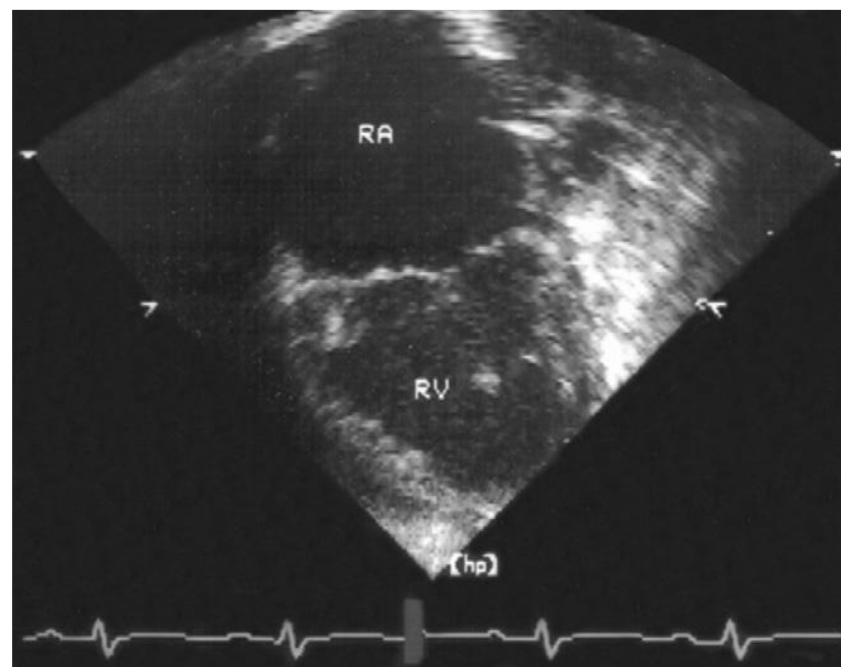
AO = Aorta
PA = Pulmonary Artery
LA = Left Atrium
RA = Right Atrium
LV = Left Ventricle
RV = Right Ventricle



A



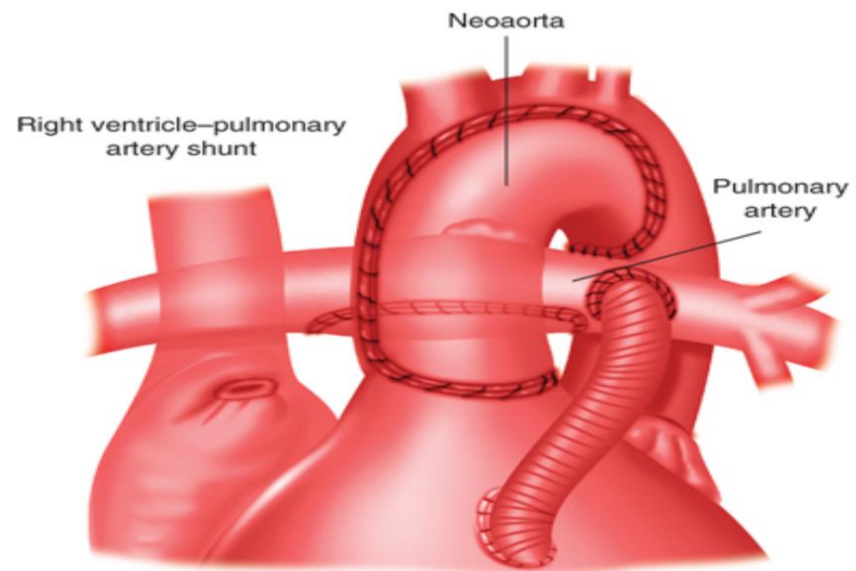
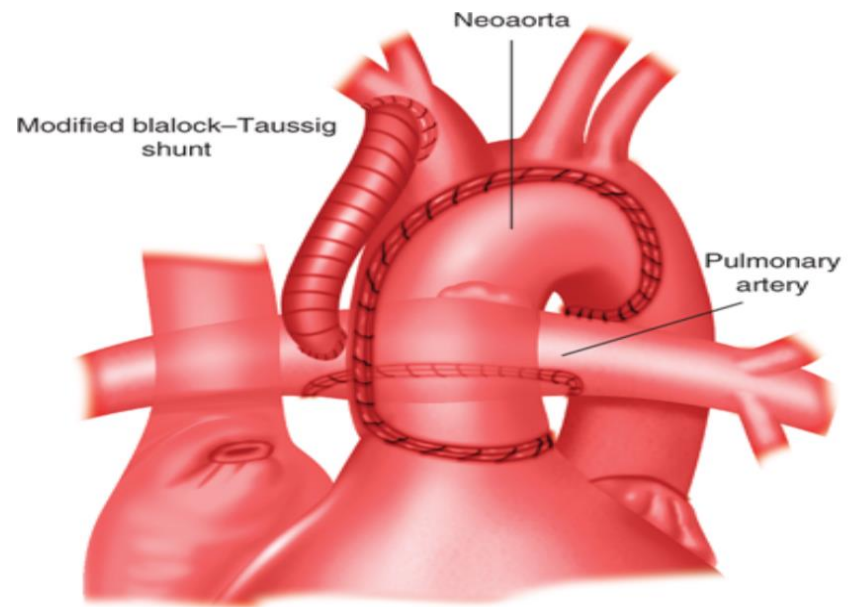
B



C

Hypoplastic Left Heart Syndrome with **Intact Atrial Septum**

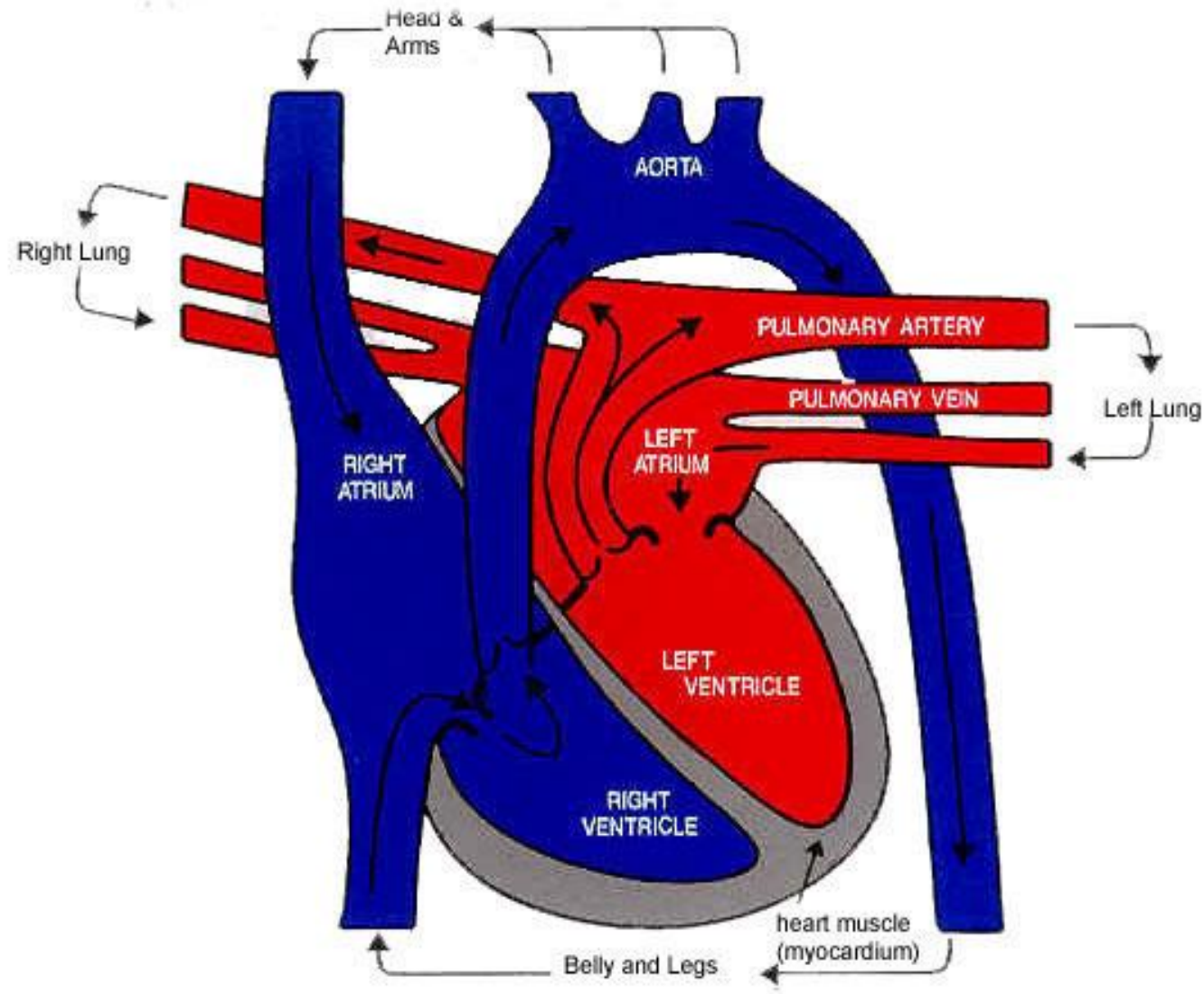
- at delivery, infants with HLHS/IAS present with profound cyanosis, metabolic acidosis, respiratory distress, and cardiovascular collapse
- patients are critically ill, markedly tachypneic, and often have a PaO₂ of less than 20 mmHg
- prostaglandin should be administered immediately to ensure ductal patency and systemic perfusion
- **emergent transcatheter (balloon and blade atrial septostomy) intervention must be performed**
 - decompress the left atrium
 - allow for oxygenated blood to reach the circulation



Transposition of the Great Arteries

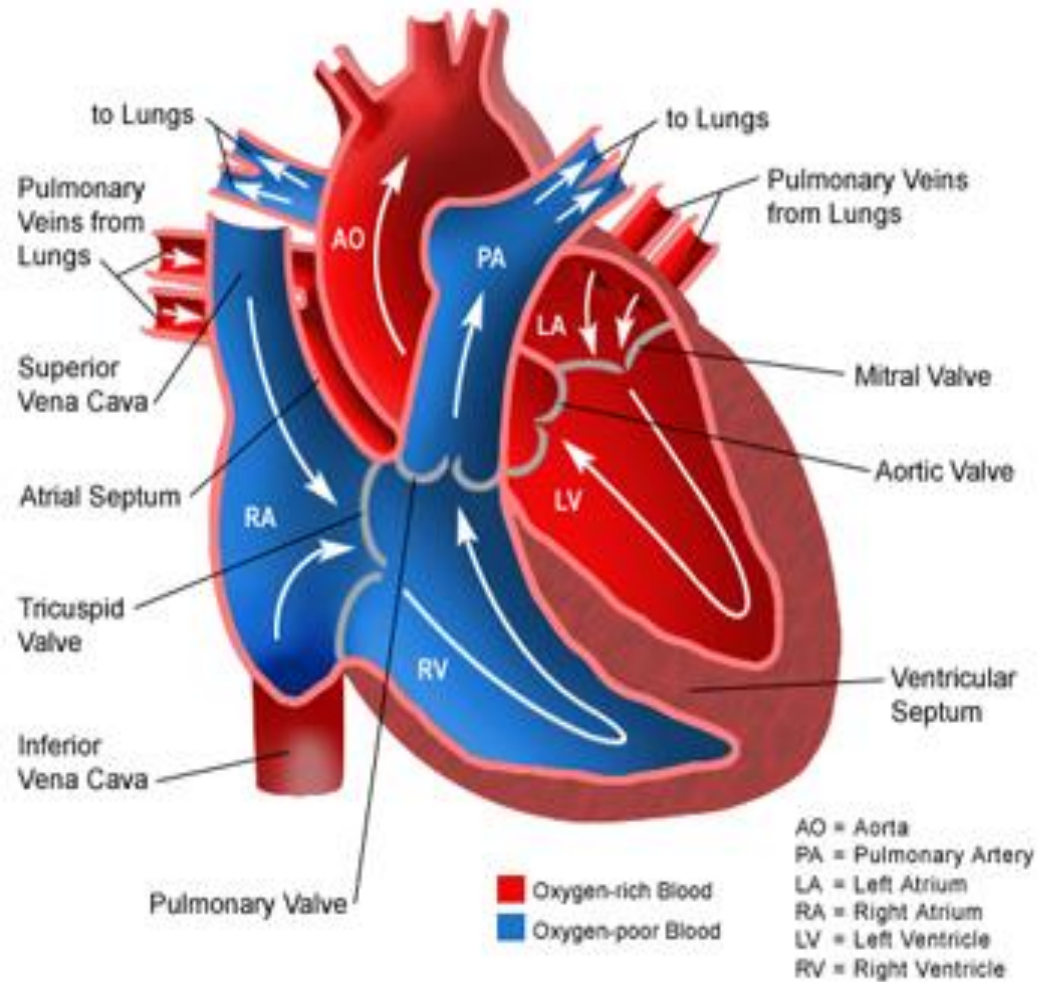
- aorta arises from the RV and pulm artery arises from the LV
- systemic and pulmonary circulations are arranged in parallel rather than in series
- causes recirculated Q_p and a deficiency of oxygen supply to the tissues
- survival with the systemic and pulmonary circulations arranged in parallel is not possible
- requires some blood to exit the pulmonary circuit in order to enter the systemic circuit to provide oxygen (Q_{ES}), and for blood to similarly exit the systemic circuit in order to enter the pulmonary circuit to pick up oxygen (Q_{EP})
- In a completely normal heart, $Q_p = Q_{EP}$, $Q_s = Q_{ES}$, and $Q_p = Q_s$

TRANSPOSITION OF THE GREAT VESSELS

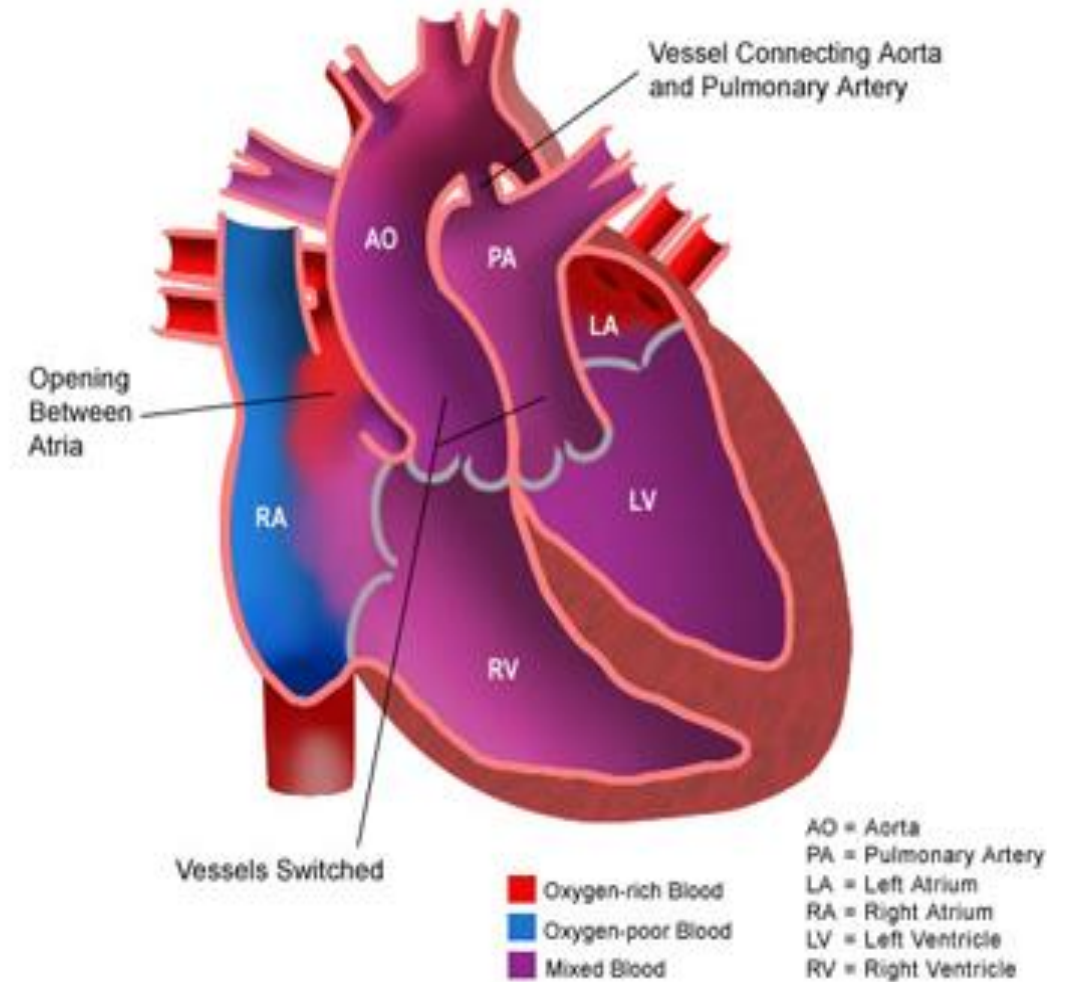


TRANSPOSITION OF THE GREAT VESSELS

Normal Heart



Transposition of Great Arteries



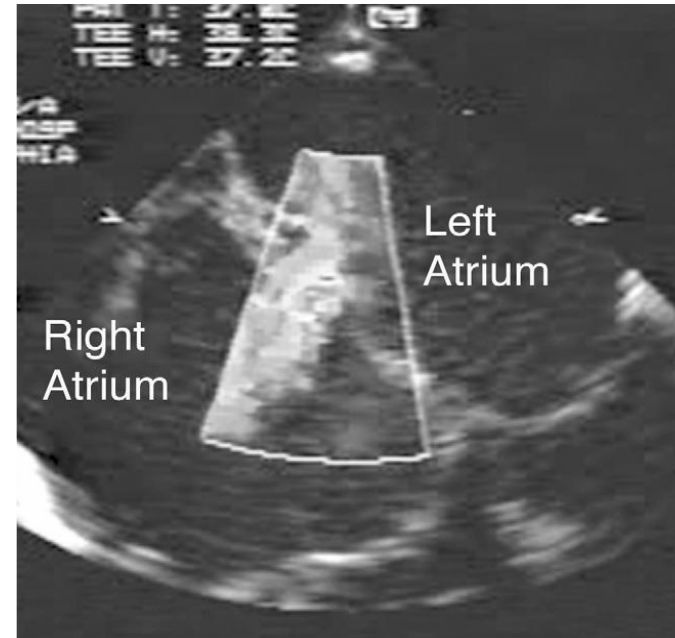


Source: David K. Stevenson, Ronald S. Cohen, Philip Sunshine: Neonatology: Clinical Practice and Procedures
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Transposition of the Great Arteries with Restrictive or Intact Atrial Septum

- a newborn baby with transposition of the great arteries, both the PFO and the PDA are open in the first few minutes or hours of life.
- the two parallel circuits consisting of the pulmonary and systemic circulations are expected to have two possible areas of communication between them
- when the atrial septum is intact or very restrictive , mixing between the two circulations only can occur at the level of the PDA
- significantly impaired O₂ delivery – PaO₂ in the 20s
- presents at delivery with profound cyanosis and metabolic acidosis
- pre- and postductal saturations may demonstrate reverse differential cyanosis
- PGE₁ should be initiated immediately to maintain ductal patency
- emergent balloon atrial septostomy must be performed to allow for adequate mixing at the atrial level

Balloon Atrial Septostomy



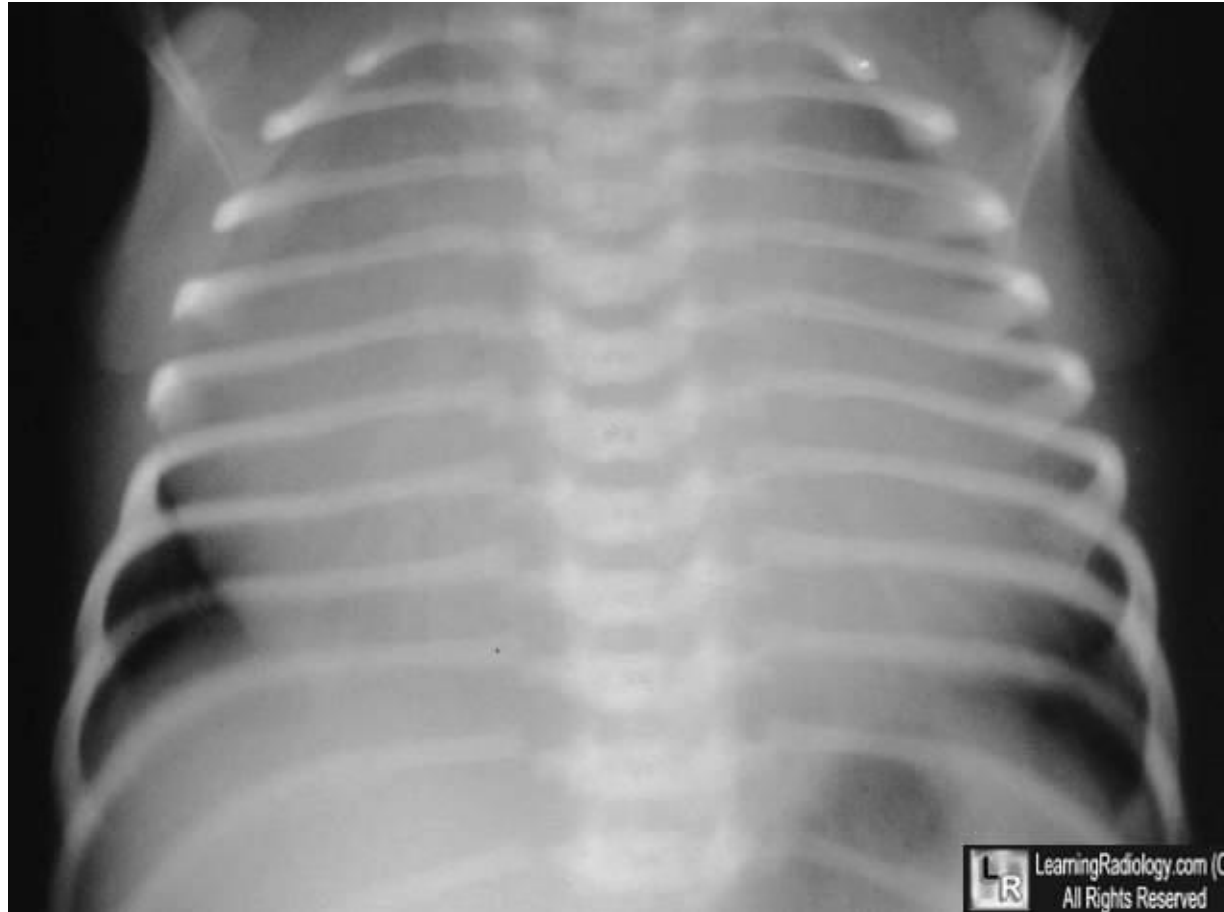
SYMPTOMS ON FIRST DAY OF LIFE

- Airway compromise
 - Severe Ebstein's anomaly of the tricuspid valve
 - Tetralogy of Fallot with absent pulmonary valve
- Obstruction to pulmonary venous return
 - Total anomalous pulmonary venous return (TAPVR) with obstruction – infracardiac type

Severe Ebstein's Anomaly of the Tricuspid Valve

- the septal and posterior leaflets are deformed and displaced inferiorly into the RV
- the tricuspid valve is severely incompetent (TR), resulting in profound right atrial enlargement – Holosystolic murmur audible
- newborns often present with respiratory failure due to airway compression from the profound cardiomegaly and cyanosis
- there may be functional pulmonary atresia, because of severe TR
- causing right-to-left atrial-level shunt, cyanosis and ductal-dependent pulmonary blood flow

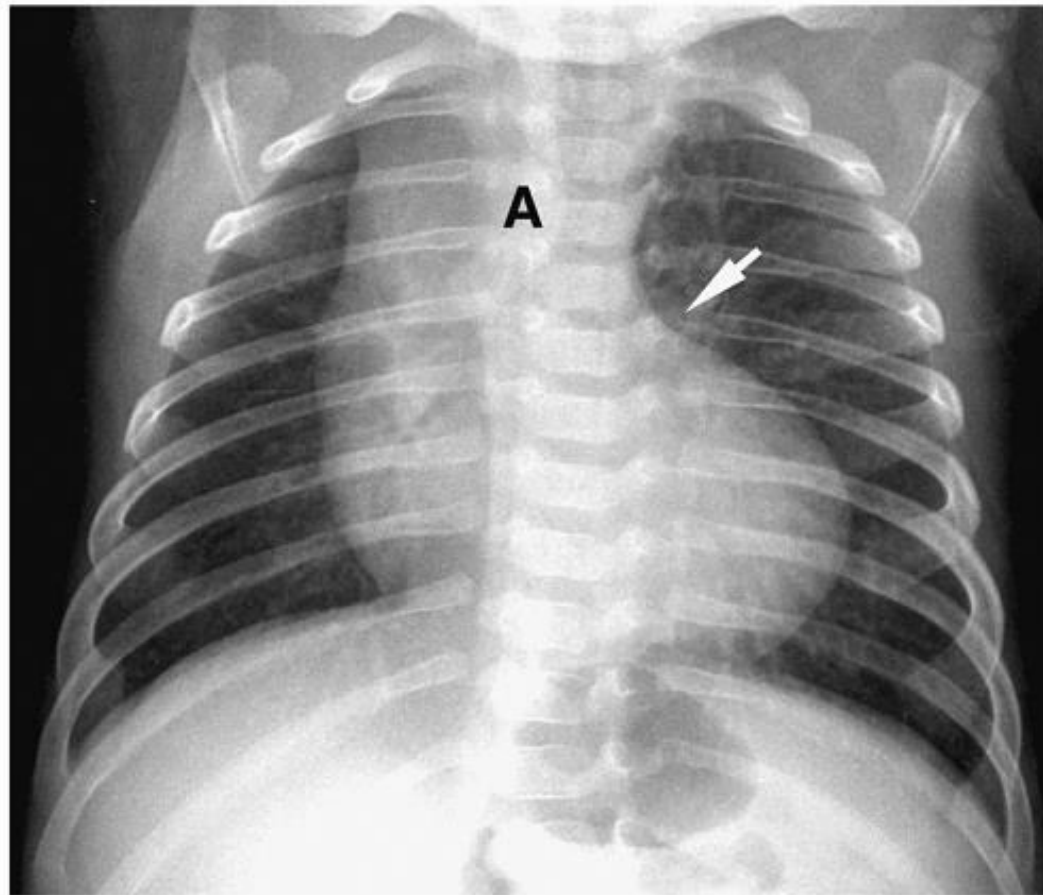
EBSTEIN'S



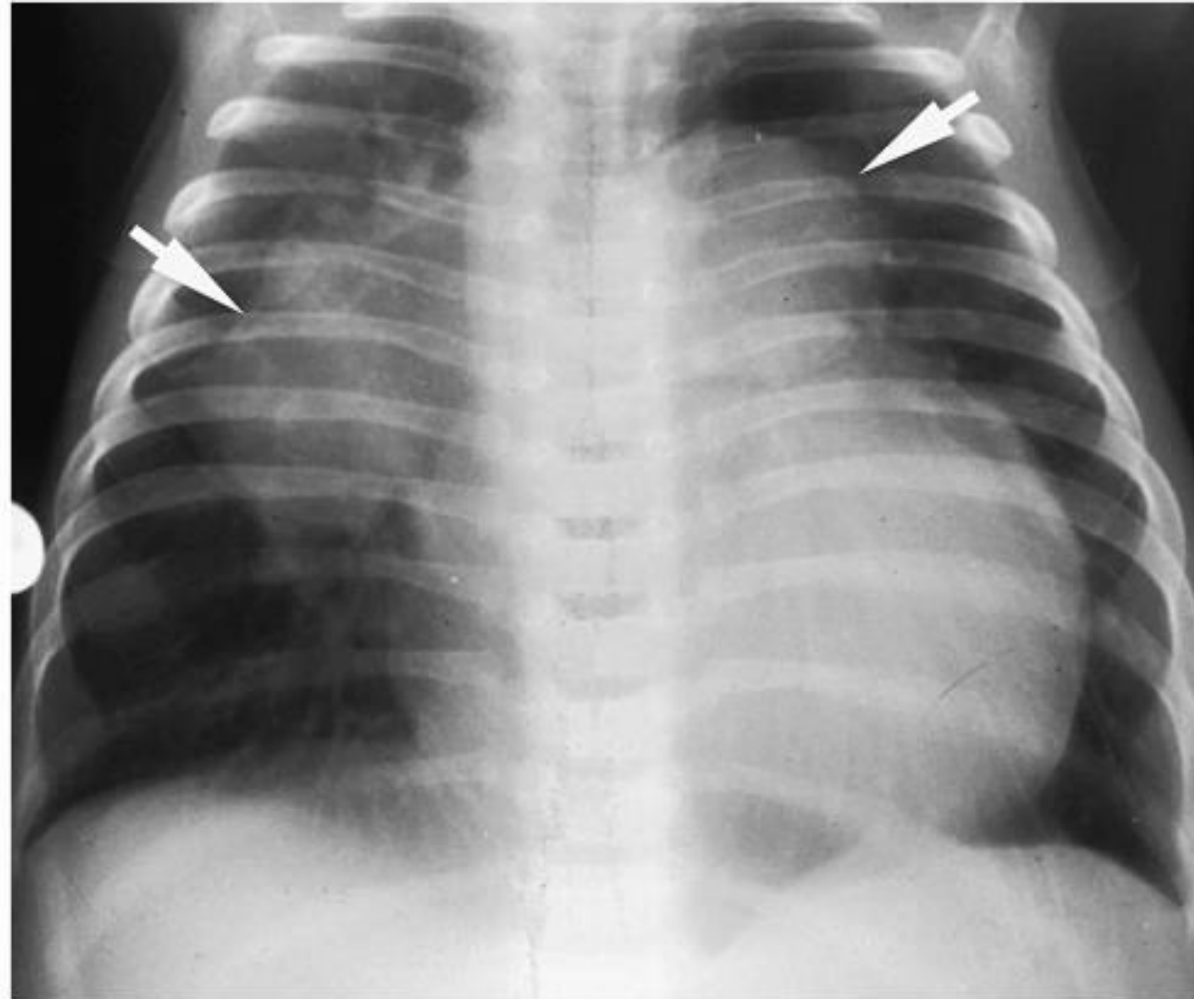
Tetralogy of Fallot with absent pulmonary valve

- the four components tetralogy of Fallot (TOF) are a VSD, an overriding aorta, pulmonary stenosis, and right ventricular hypertrophy
- a variant, TOF with absent pulmonary valve, is associated with markedly dilated central(proximal) pulmonary arteries in utero
- **characteristic “to-and-fro” murmur** at the left upper sternal border, consistent with pulmonary outflow obstruction and regurgitation
- severe bronchomalacia can result from compression of the bronchi by the large central pulmonary arteries
- **require immediate intubation and ventilation**
- prognosis of these patients may be limited by their ventilatory difficulties

TETRALOGY OF FALLOT



TETRALOGY OF FALLOT WITH ABSENT PULMONARY VALVE



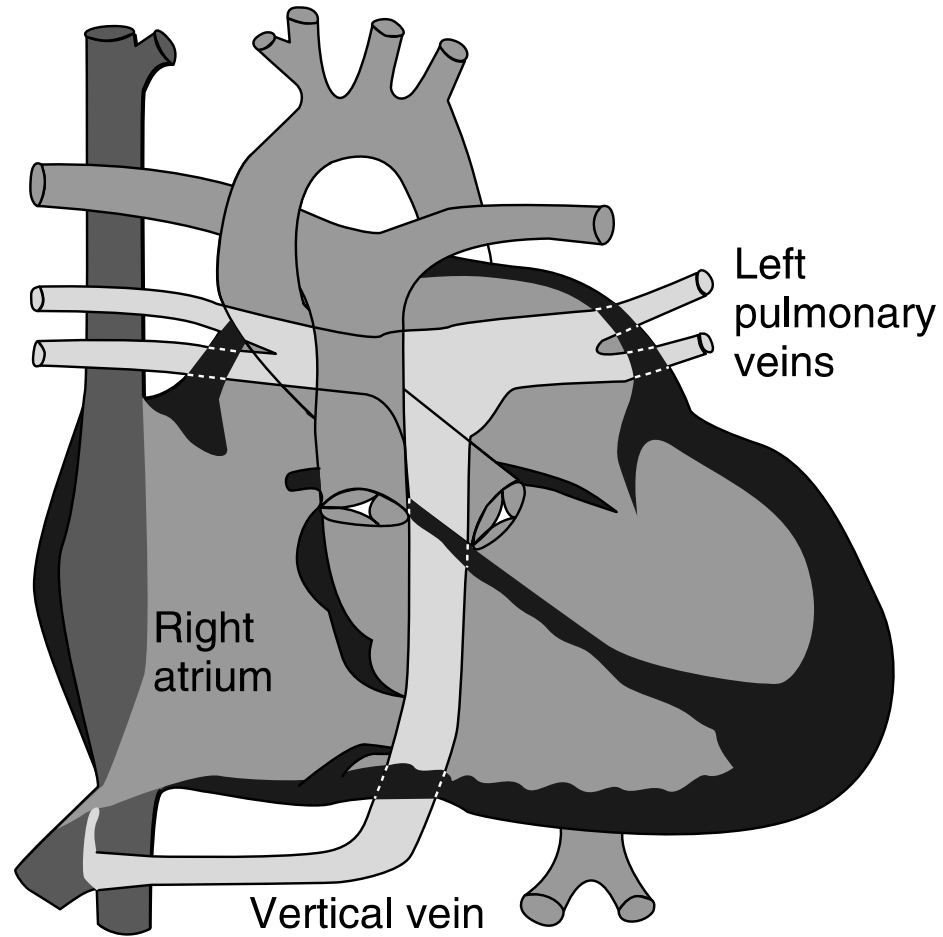
TOTAL ANOMALOUS PULMONARY VENOUS RETURN (TAPVR)

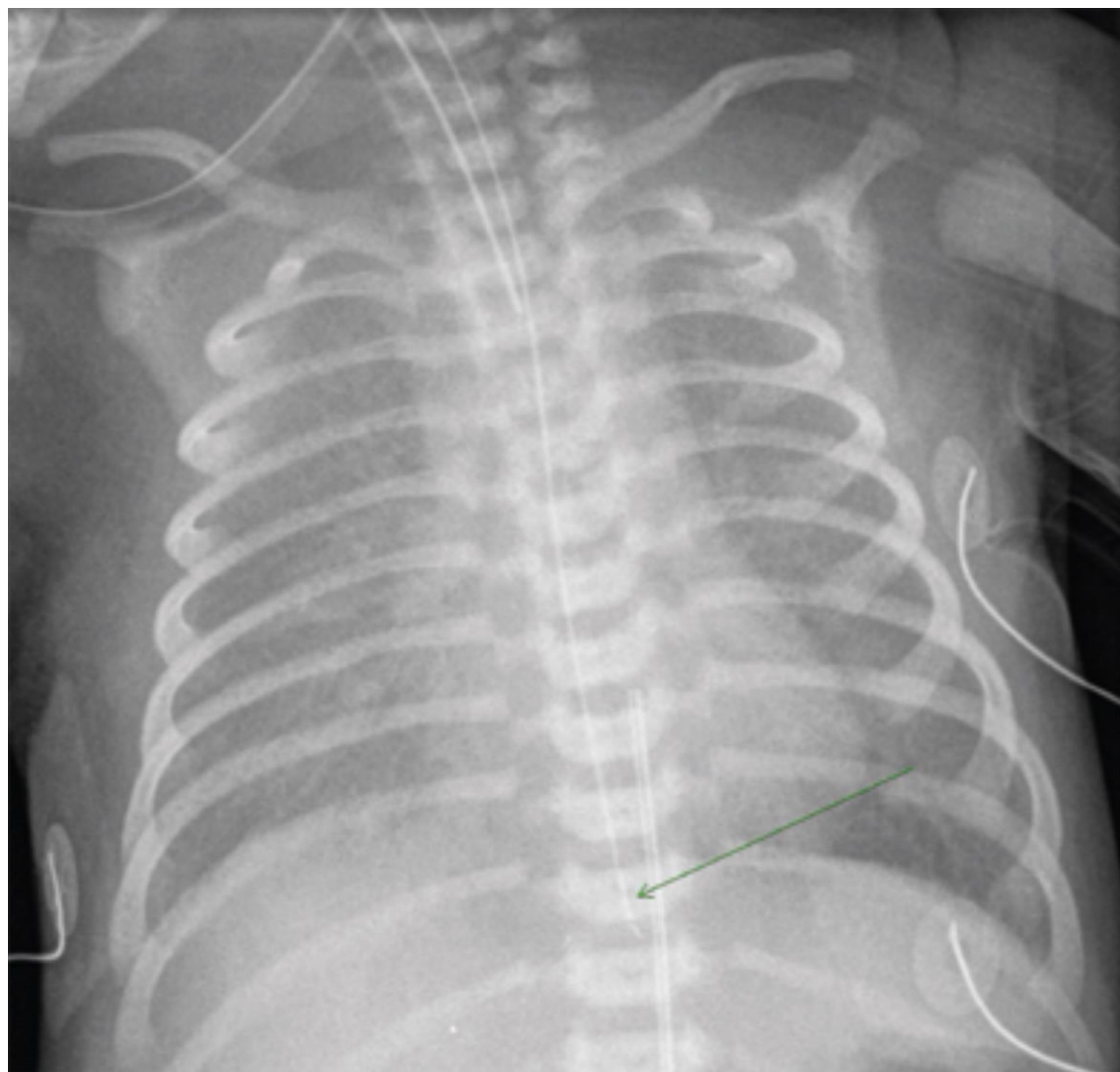
- in TAPVR there is no connection between the pulmonary veins and the left atrium
- pulmonary veins form a confluence behind the left atrium
 - which decompresses through a vertical vein
 - inferiorly below the diaphragm
 - empties either into the portal system or into the ductus venosus before ultimately returning to the right atrium
- complete mixing of systemic and pulmonary venous return occur within the right atrium

Obstruction to Pulmonary Venous Return

- neonates with TAPVR with obstruction usually present within the first hours to days of life – **true surgical emergency**
- cyanosis and respiratory distress secondary to pulmonary venous congestion with pulm edema and small heart size on CXR
- surgical repair should be performed as soon as possible
- if critically ill and immediate surgery is not an option, may be stabilized with veno arterial extracorporeal membrane oxygenation (VA ECMO)
- prostaglandin does not help, may worsen the hemodynamic state by further increasing pulmonary blood flow

Obstruction to Pulmonary Venous Return





Source: David K. Stevenson, Ronald S. Cohen, Philip Sunshine: Neonatology: Clinical Practice and Procedures
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PROBLEM 3

- A 6 day old, male term infant presents to the ER with 2 day history of not eating well, increasing lethargy, and tachypnea. HR 190, Sats are 88%, pulses are difficult to feel, extremities are cool, cap refill is 5 seconds, the ER physician is not sure whether he is feeling his own pulses. Glucose is 40, ph is 7.1, BUN is 20 and Creatinine 1.9. Mother's GBS status is unknown. Had a previous child who died early. He has been given 20 ml/kg fluid bolus, and antibiotics are ordered. Your institution is one of the few who will admit a neonate up to 7 days of age into the NICU. Lucky you! What is going on?
- 1. Sepsis
- 2. Metabolic disorder
- 3. Non accidental trauma
- 4. Duct dependent circulation
- 5. acute renal failure from dehydration

SYMPTOMS IN THE FIRST WEEK OF LIFE

- Lesions with Ductal-Dependent Systemic Blood Flow
 - Hypoplastic Left Heart Syndrome
 - Critical Aortic Stenosis
 - Critical Coarctation of the Aorta
- Lesions with Ductal-Dependent Pulmonary Blood Flow
 - Critical Pulmonary Valve Stenosis
 - Pulmonary Atresia, Including RV-Dependent Coronary Circulation
 - Severe Tetralogy of Fallot
- Lesions with Large Left-to-Right Shunts
 - Truncus Arteriosus
 - VSD with Arch Obstruction

Critical Coarctation of the Aorta

- newborns with critical coarctation usually present in the first 6 weeks (typically in the first 7–10 days) of life
- with tachypnea, tachycardia, and severe hypotension
- if the PDA is partially open
 - preductal saturations will be higher than postductal saturations
 - evidence of upper extremity hypertension and diminished palpable femoral pulses
- if the PDA has closed
 - no discrepancy between saturations will be noted and
 - femoral pulses often will be absent.

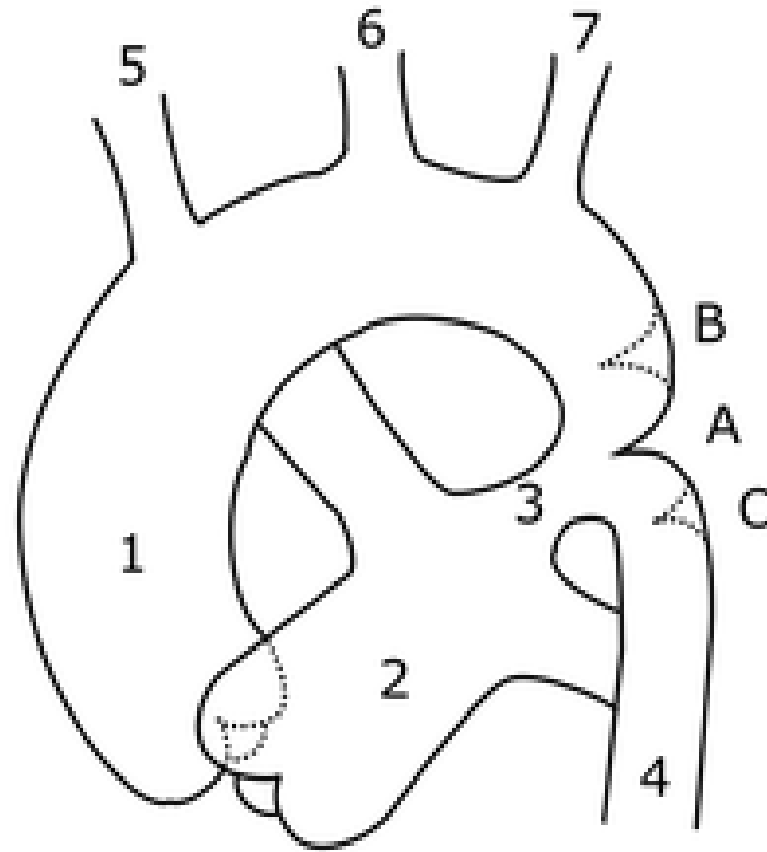
Critical Coarctation of the Aorta

- occurs at the insertion site of the PDA into the descending aorta
- **can be difficult to appreciate** in the face of a widely patent PDA
- with closure of PDA, the entire cardiac output must cross the area of coarctation(narrowing) to enter the descending aorta
- in the case of critical coarctation, severe obstruction is present and the LV cannot supply adequate flow to the descending aorta
- congestive heart failure and cardiogenic shock occurs

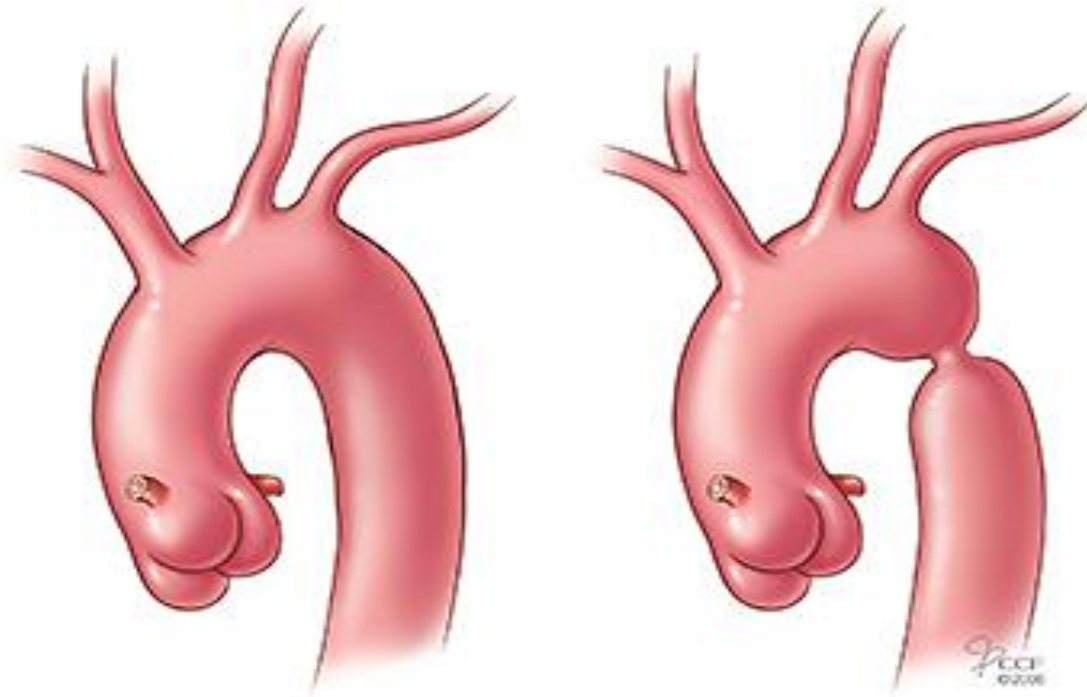
COARCTATION AND BLOOD PRESSURE

- BP should be measured in all four extremities. A difference of greater than 10 mmHg in upper compared to lower-extremity blood pressure suggests the **presence of aortic coarctation, aortic arch hypoplasia, or interrupted aortic arch**
- there are 2 caveats
- in the event of low cardiac output and systemic hypotension, blood pressure differences are diminished. Coarctation may be present in the absence of significant blood pressure gradient. Hypotension should be corrected, and cardiac output should be maximized prior to interpretation of blood pressure differences
- if the PDA is widely patent, a blood pressure difference between upper and lower extremities may not be noted, despite underlying coarctation. A complete assessment of the newborn with cyanosis includes preductal and postductal measurements of oxygen saturation

Coarctation of the Aorta



Coarctation of the Aorta



SUMMARY

- As the newborn separates from the placenta, oxygenation and ventilation are dependent on the infant; PVR falls, and the PFO and PDA begin to close
- Cardiac output is proportional to heart rate and stroke volume. The infant's stroke volume is fairly fixed, and cardiac output is predominantly heart rate-dependent
- Newborns are deficient in calcium stores in their sarcoplasmic reticulum. They are very responsive to calcium administration
- Potential side effects of PGE1 are hypotension, apnea and fever
- Reverse differential cyanosis is only seen with the physiology of transposition of the great arteries
- A failed hyperoxia test ($\text{PaO}_2 < 150$ mmHg in the right radial artery) is consistent with an intracardiac mixing lesion. Congenital heart lesions with normal hyperoxia tests include coarctation, aortic stenosis, and isolated interrupted aortic arch

SUMMARY

- Many factors impact Qp:Qs. Oxygen, hyperventilation, alkalosis, and inspired nitric oxide increase Qp:Qs. Hypoventilation and inspired carbon dioxide decrease Qp:Qs
- Cardiac lesions that are unstable in the delivery room represent abnormalities of oxygen delivery that are often not stabilized by PGE1 alone and require immediate intervention (HLHS with intact atrial septum, TGA with intact atrial septum, or TAPVR with severe obstruction)
- Neonatal Ebstein's anomaly and TOF with absent pulmonary valve syndrome can result in immediate respiratory compromise caused by compression of the airways by the large right atrium or large central pulmonary arteries, respectively

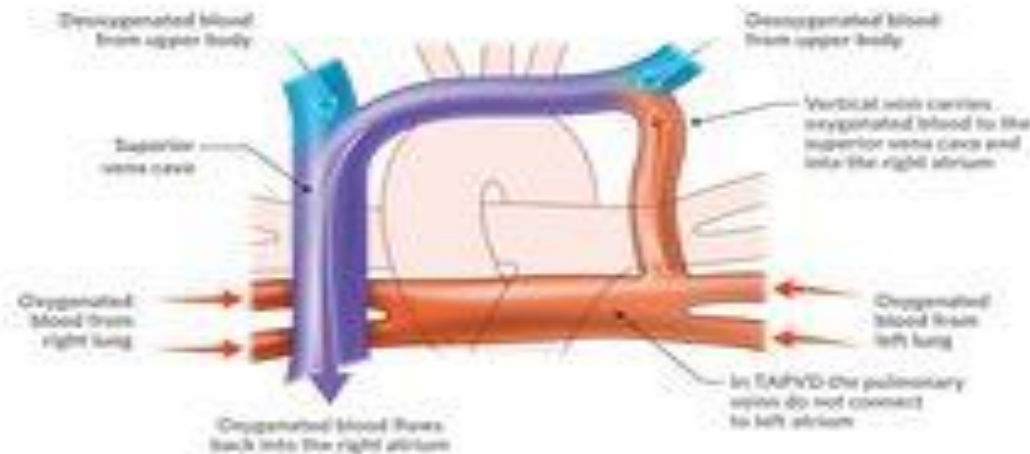
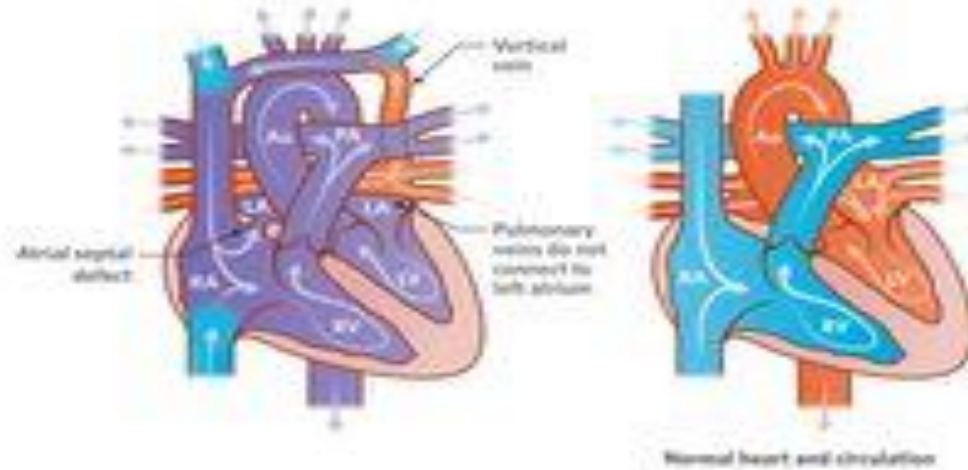
SUMMARY

- Patients with left-sided obstructive lesions (e.g., critical AS, HLHS, or critical coarctation of the aorta) can present in shock in the first few weeks of life with diminished pulses and a profound metabolic acidosis. Coarctation can present in the first several months.
- Patients with ductal-dependent Qp on PGE 1 are also at risk for pulmonary overcirculation and systemic hypoperfusion.
- Patients with truncus arteriosus often have low systemic diastolic pressures and are at risk for pulmonary overcirculation. They have an increased preoperative incidence of ventricular fibrillation.

- **THANK YOU FOR LISTENING**

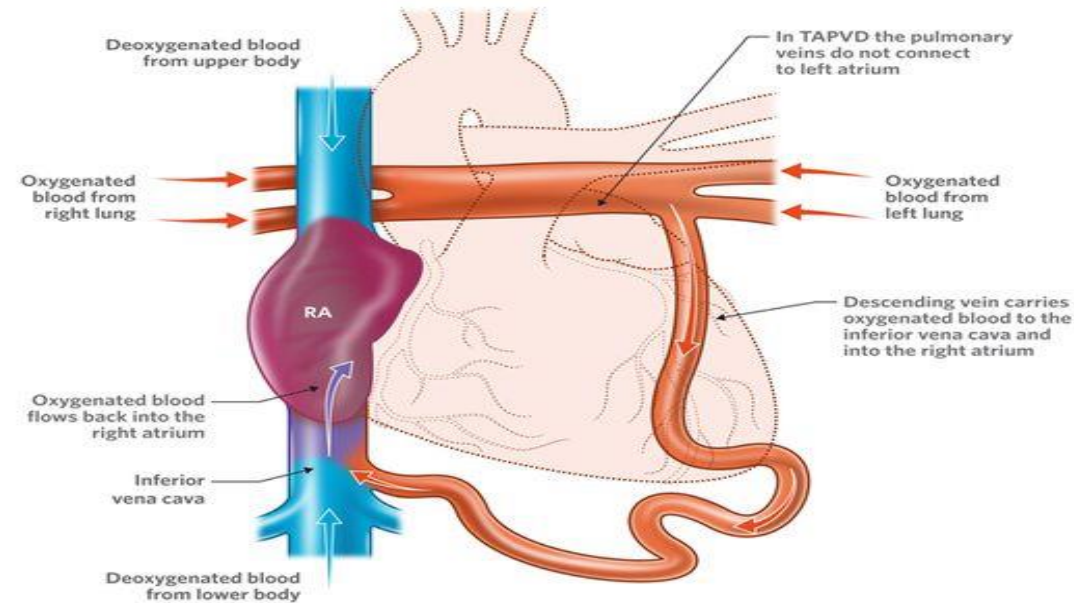
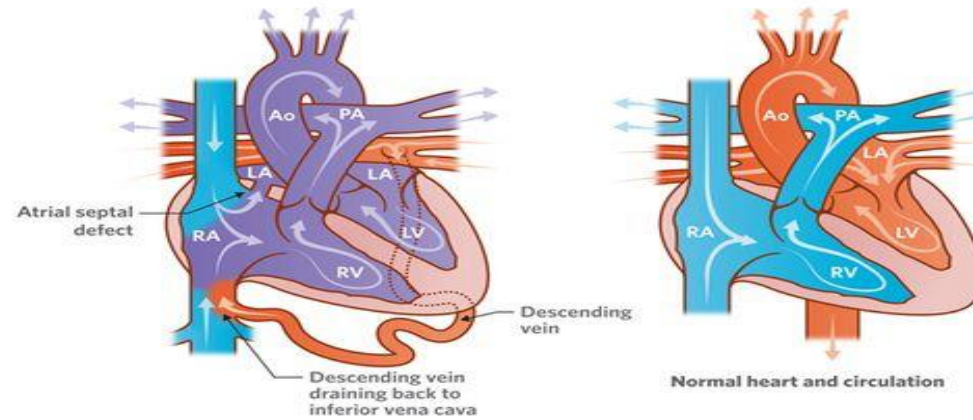
TOTAL ANOMALOUS PULMONARY VENOUS RETURN - SUPRACARDIAC

Total anomalous pulmonary venous drainage (TAPVD)

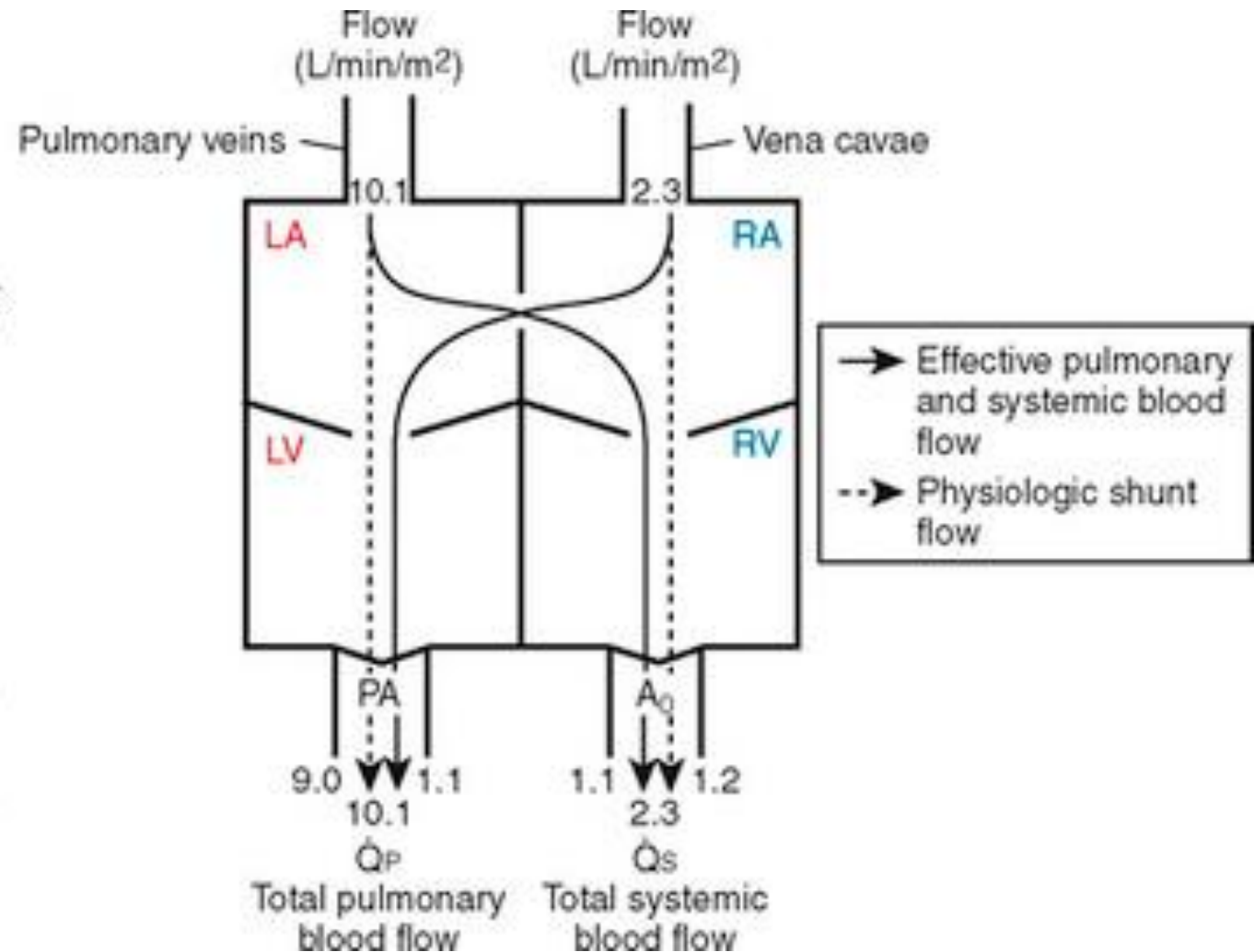
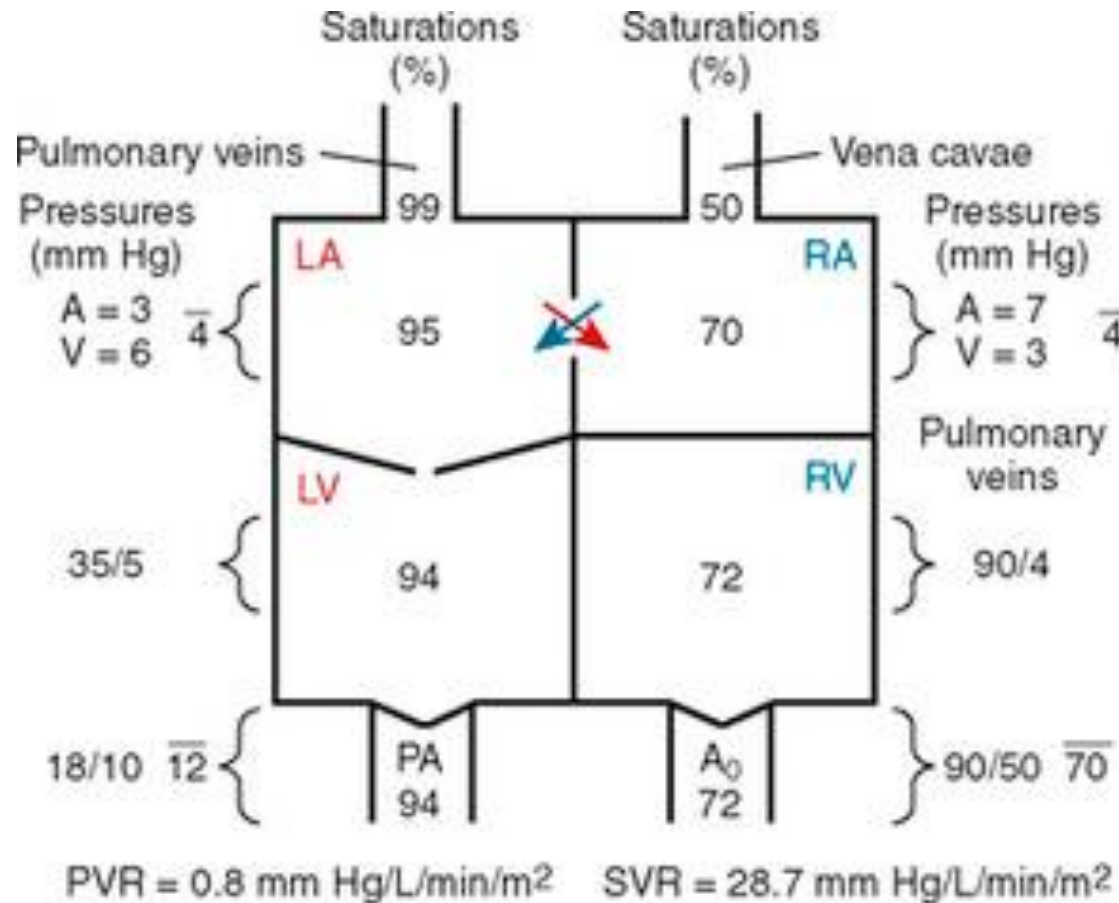


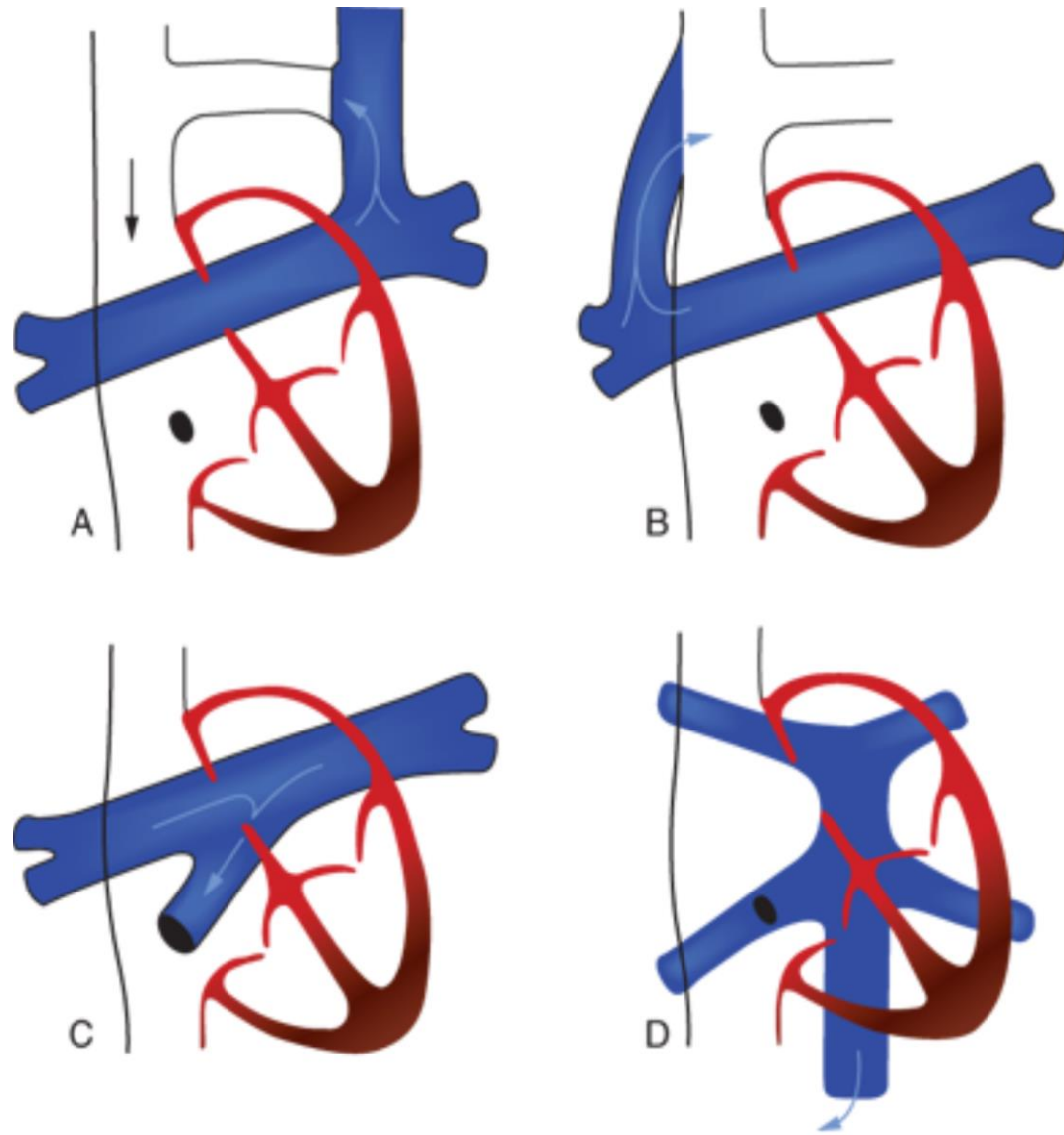
TOTAL ANOMALOUS PULMONARY VENOUS RETURN - INFRACARDIAC

Infracardiac TAPVD



TRANSPOSITION OF THE GREAT VESSELS





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