

Blue is Not the New Pink: An Introduction to Congenital Heart Defects

Becky Anderson RN, MSN, NRP, etc.
Bon Secours Critical Care Transport Team
VCU Health



Objectives

- Have a better understanding of the anatomy and physiology behind your patient's congenital defect in order to provide better care to that patient.
- Feel more confident caring for patients whose parents are telling you to do things that might not make sense.



Outline

- Review Normal Circulation
- Review Normal Fetal Circulation
- **Left-to-Right Shunts**
 - When Fetal Circulation Sticks Around
 - **PFO, PDA**
 - **Septal Defects**
- Cyanotic Defects/ Right-to-Left Shunts
 - Truncus Arteriosus
 - D-Type Transposition,
 - Tricuspid Atresia,
 - Tetralogy of Fallot
 - Total Anomalous Pulmonary Venous Return,
 - Hypoplastic Left Heart
- Notes on Babies Post-Op
- Broncho-Pulmonary Dysplasia



- 
- Scenario 1: You are dispatched to a 2-month-old with “hemorrhage.” You arrive to find a baby in no apparent distress. Mom reports the baby is on daily aspirin for a heart condition and today has had bloody diapers when he poops. She called the cardiologist, who recommended she go to the ED. However, most concerning to you is the fact that the baby appears blue. Lungs are clear and the baby has no increased work of breathing or tachypnea. Your pulse ox shows a sat of 85% with excellent pleth/waveform. Mom says the sats are always low. What do you do?

- 
- Scenario 2: You are dispatched to a 1-week-old infant with “respiratory distress.” You arrive to find a lethargic, limp baby who looks grey. Oral mucosa is tinged blue. You notice increased work of breathing, tachypnea, and a very fast heartrate. Cap refill is 5-6 seconds, skin is clammy and wet. No recent ill contacts. Baby was a normal delivery, born at a birthing center 6 days ago and seemed normal until this morning. Lungs are clear to your auscultation. Pulse oximetry is not reading.
 - What do you think is going on?
 - How are you going to treat this infant?

- 
- Scenario 3: You are dispatched to a 2-month-old baby with “respiratory distress.” You arrive to find an alert but tired looking baby with tachypnea and some increased work of breathing. Under the baby’s eyes appear swollen, and he looks very skinny. Cap refill is 4-5 seconds, no wet diapers today. PMH from mom is that he has always gotten sweaty and “pants like a puppy” when eating from a bottle, has gotten progressively worse. Heartrate feels very fast, and lungs have crackles bilaterally.
 - What do you think is going on with this patient?
 - How will you treat him?

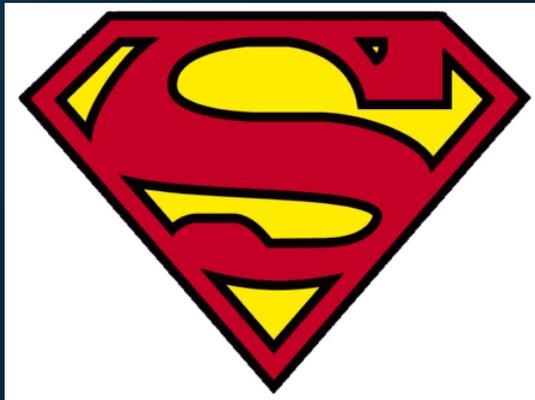
What Is Congenital Heart Disease?

- Congenital = Present at birth
- Wide range of severity of symptoms
- Includes defects of great vessels and/or structures of the heart

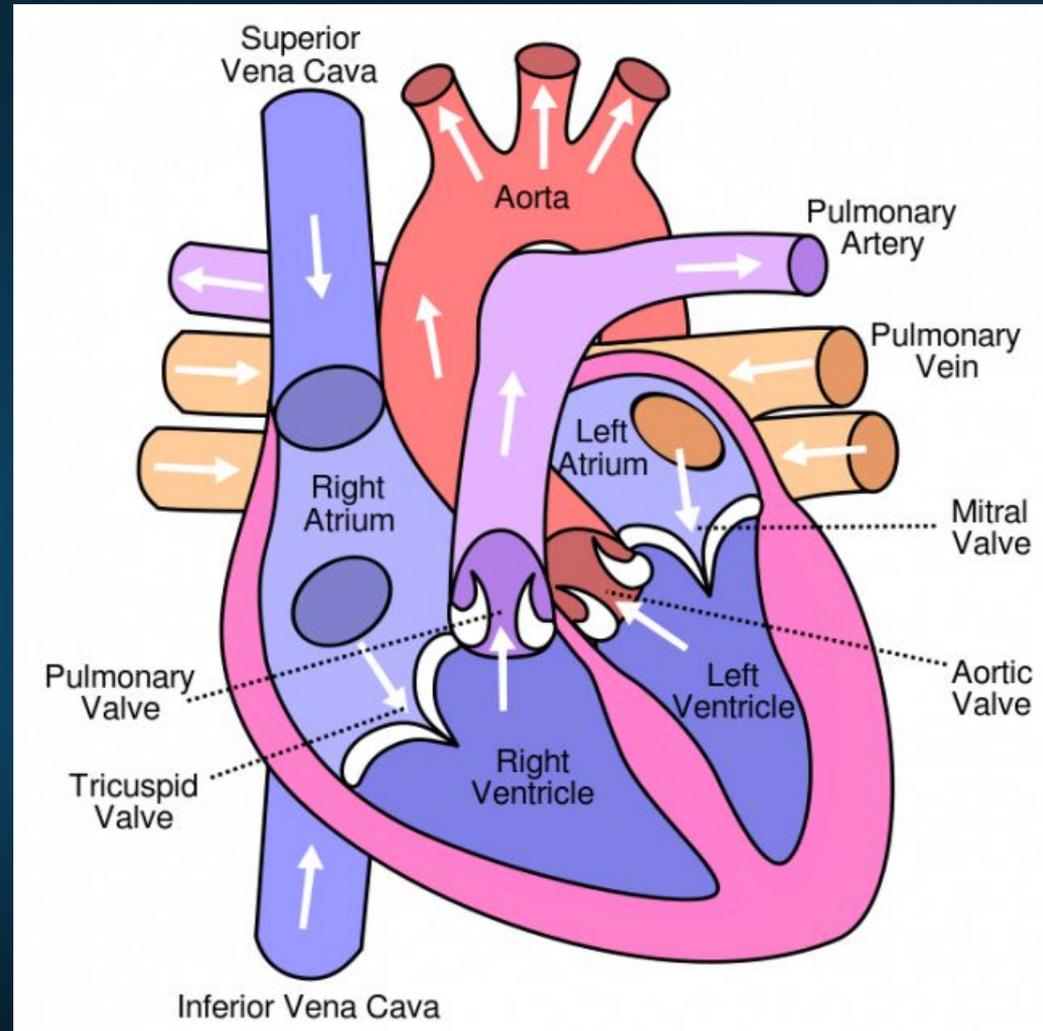


Cyanotic vs Acyanotic Defects

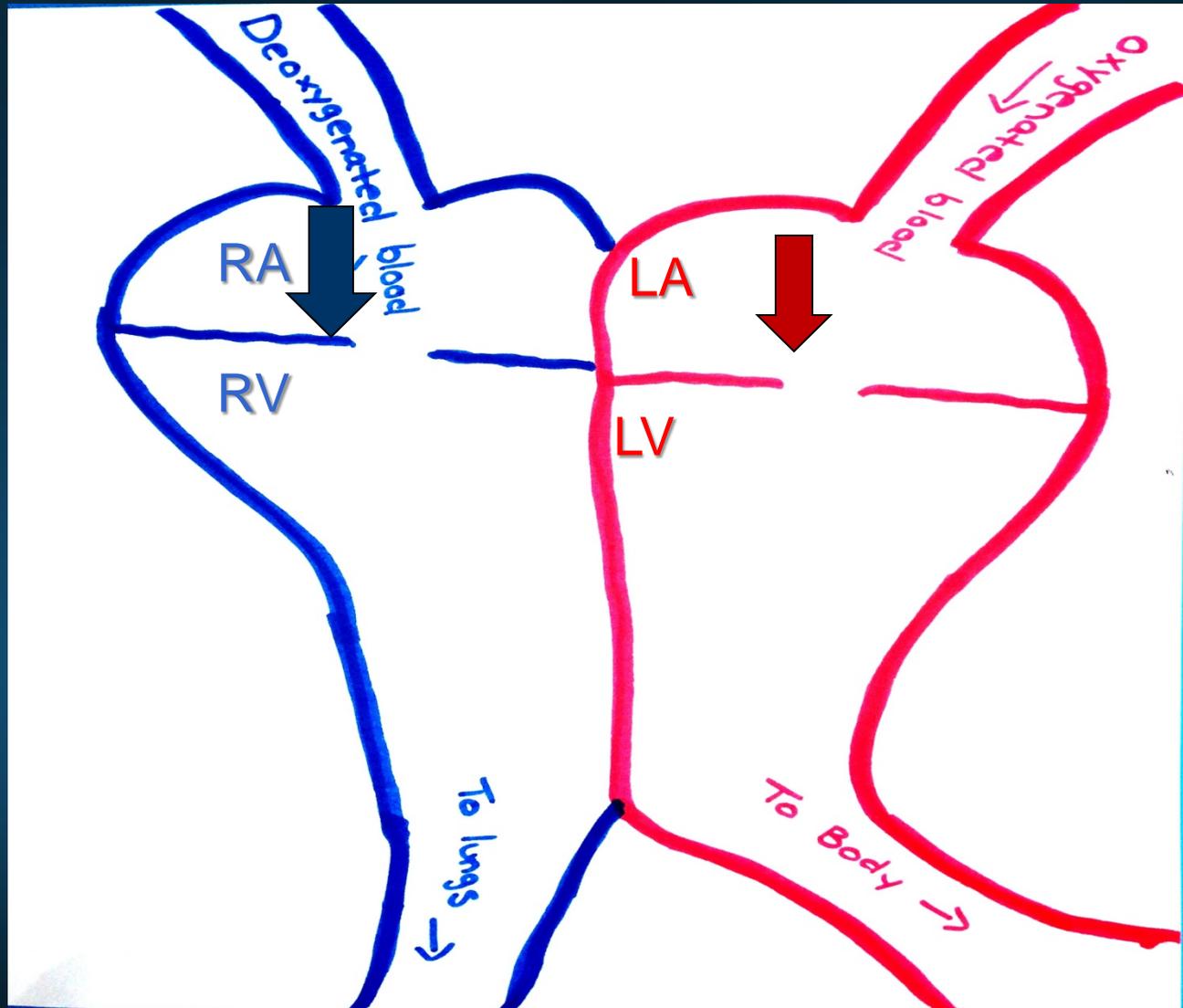
- Cyan = Blue
- Present when blood skips the lungs and goes out to the body
- Oxygenated blood mixes with deoxygenated blood
 - Healthcare Providers can make this mixing better or worse – without ever touching oxygen!!!!



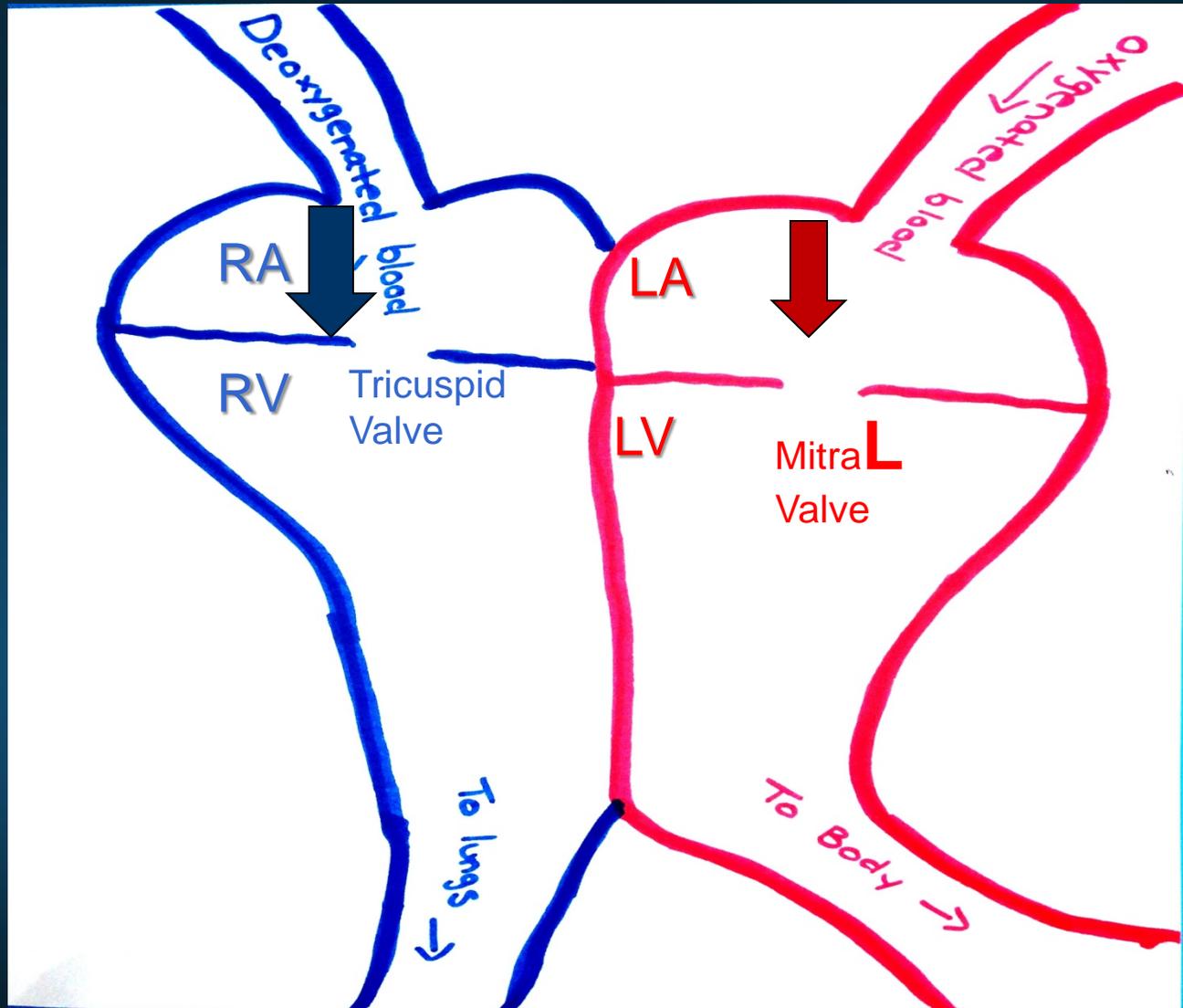
Normal Adult Circulation



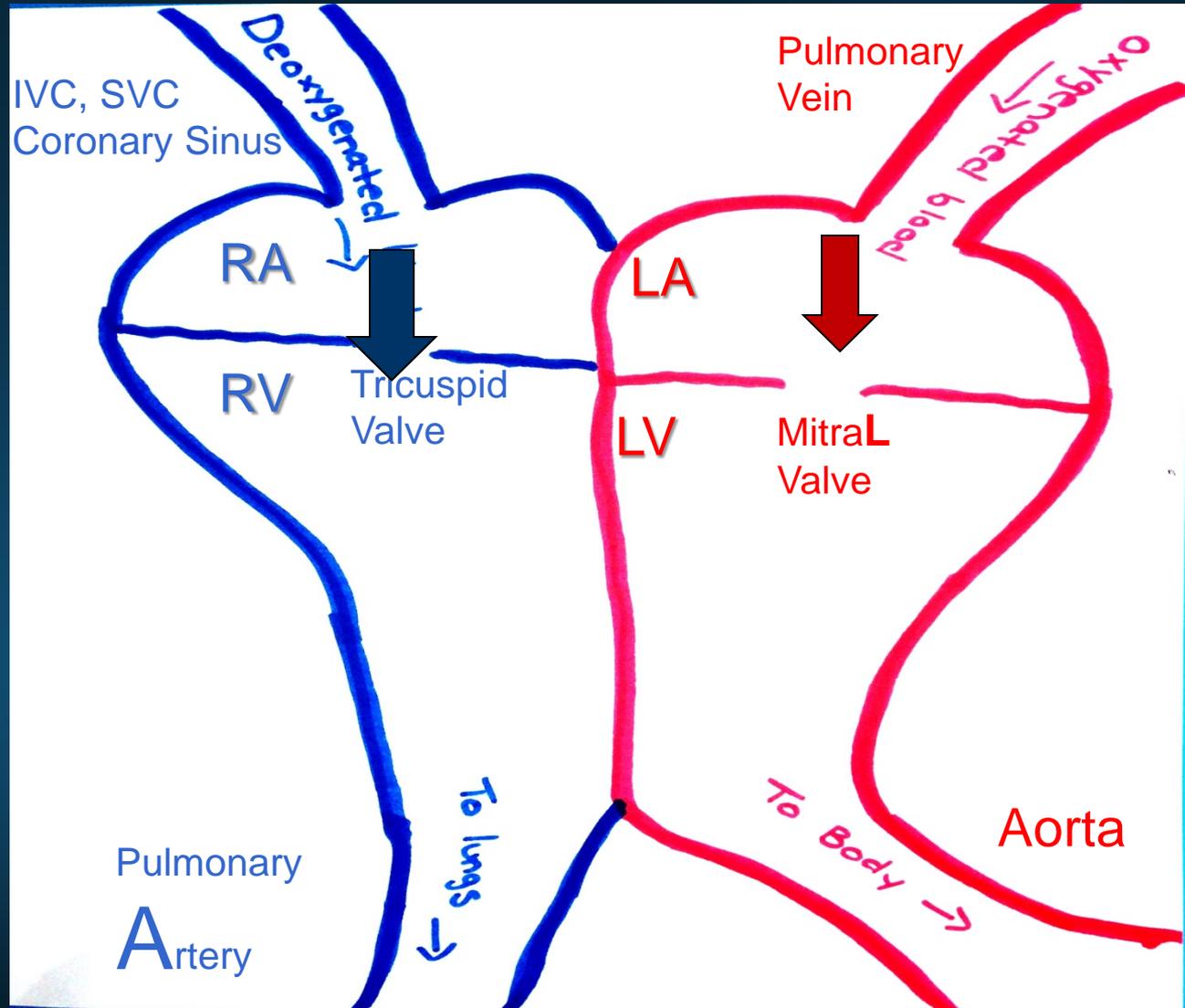
Normal Adult Circulation

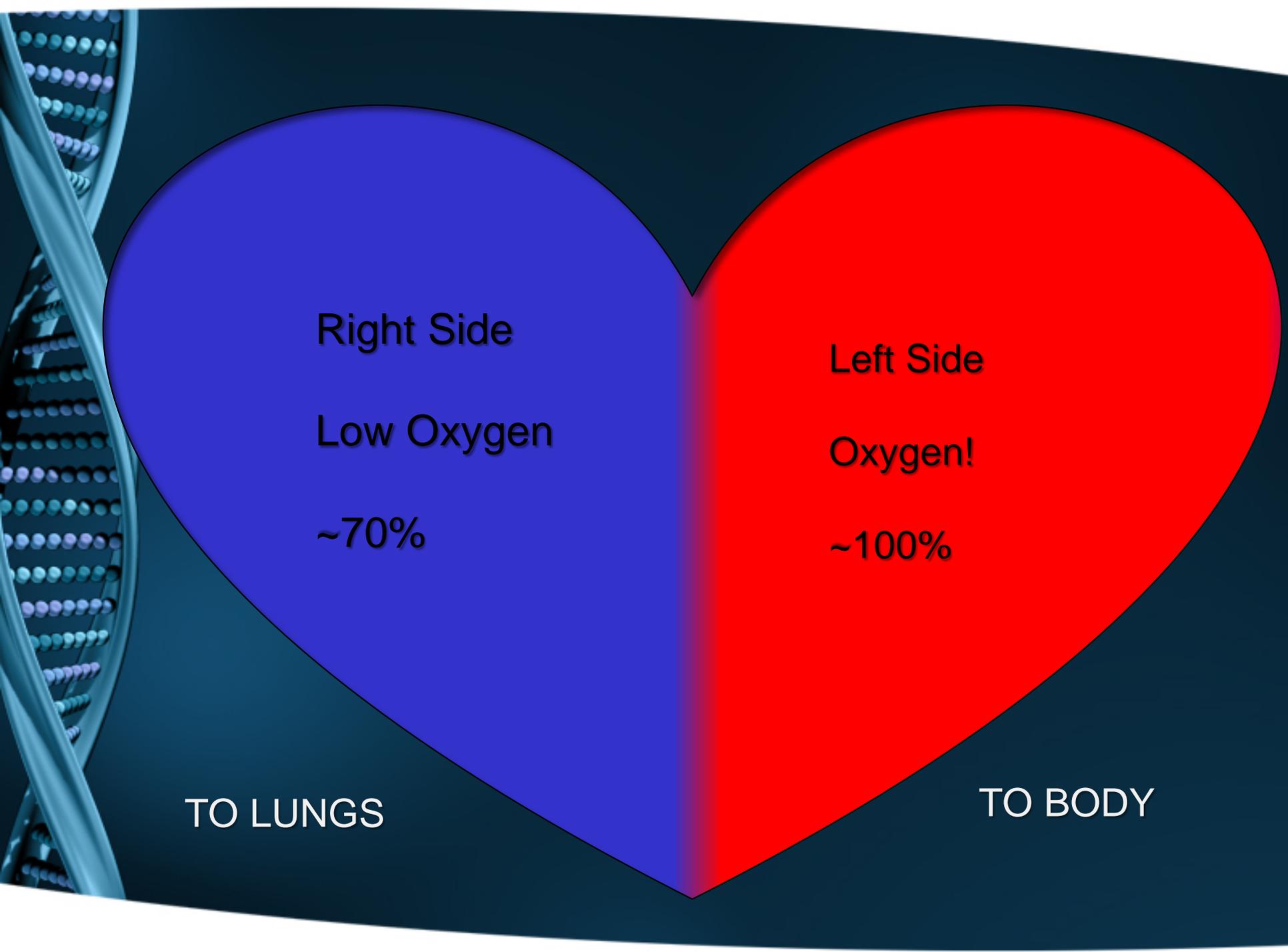


Normal Adult Circulation



Normal Adult Circulation





Right Side

Low Oxygen

~70%

TO LUNGS

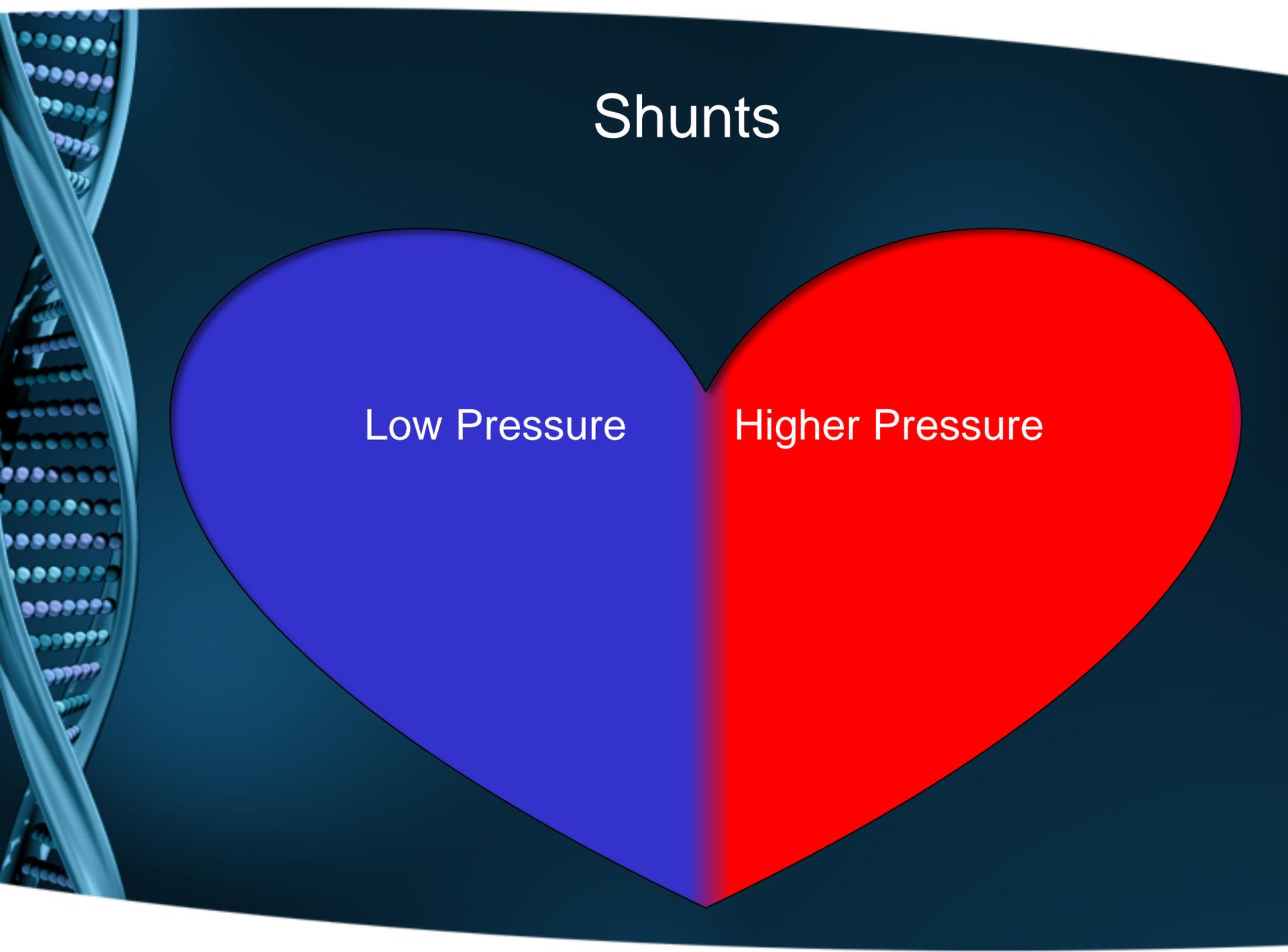
Left Side

Oxygen!

~100%

TO BODY

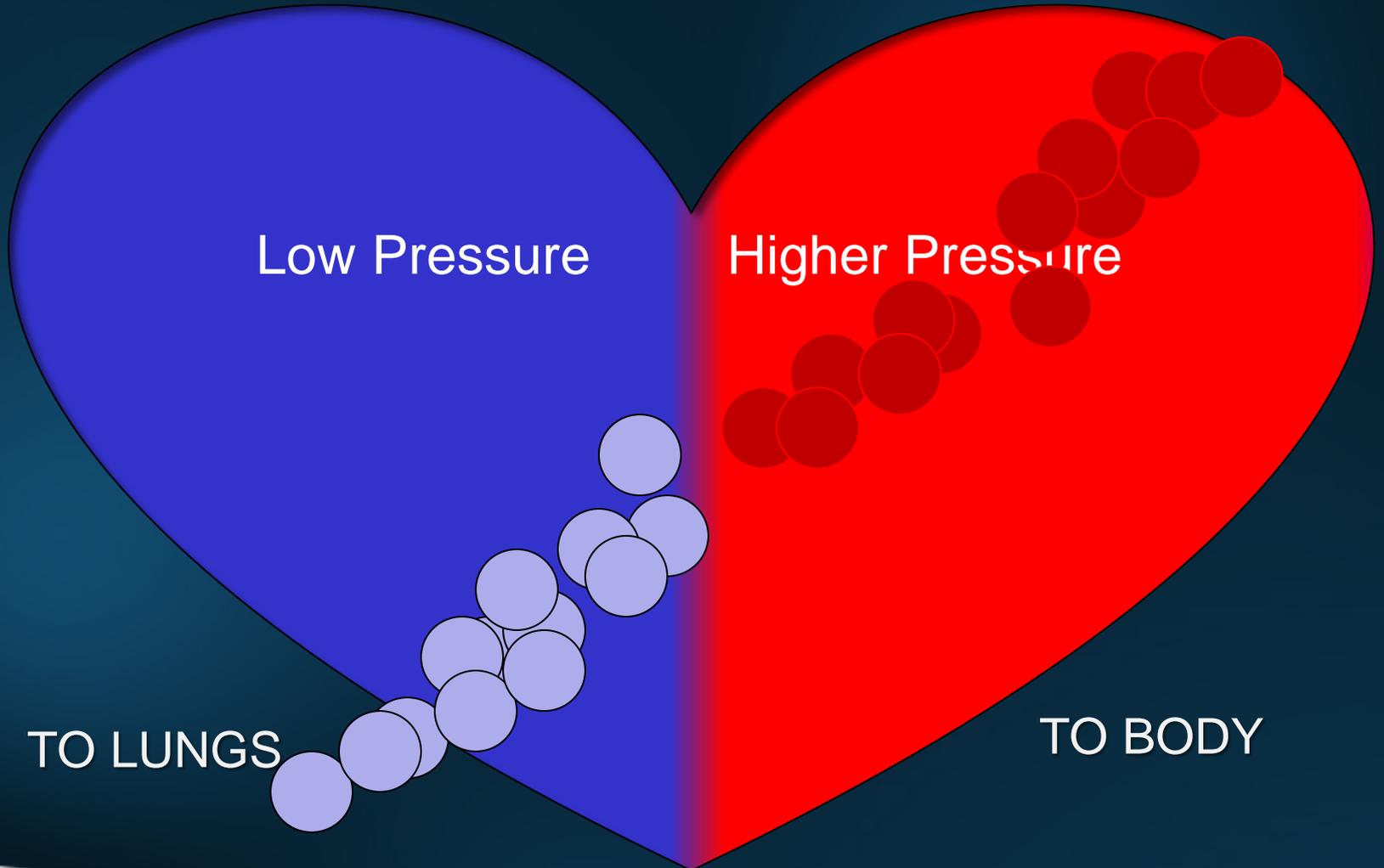
Shunts



Low Pressure

Higher Pressure

Shunts



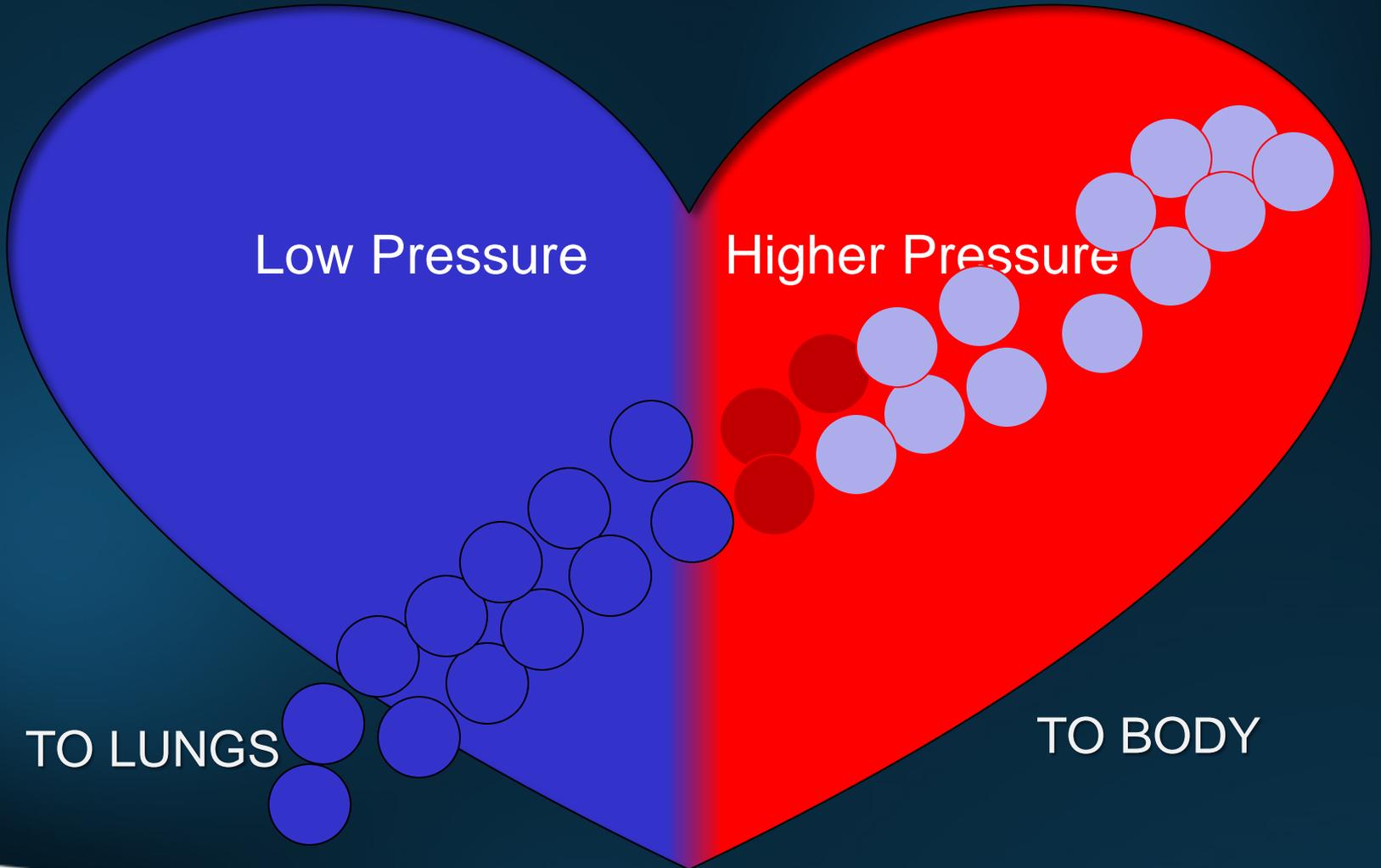
Low Pressure

Higher Pressure

TO LUNGS

TO BODY

Shunts



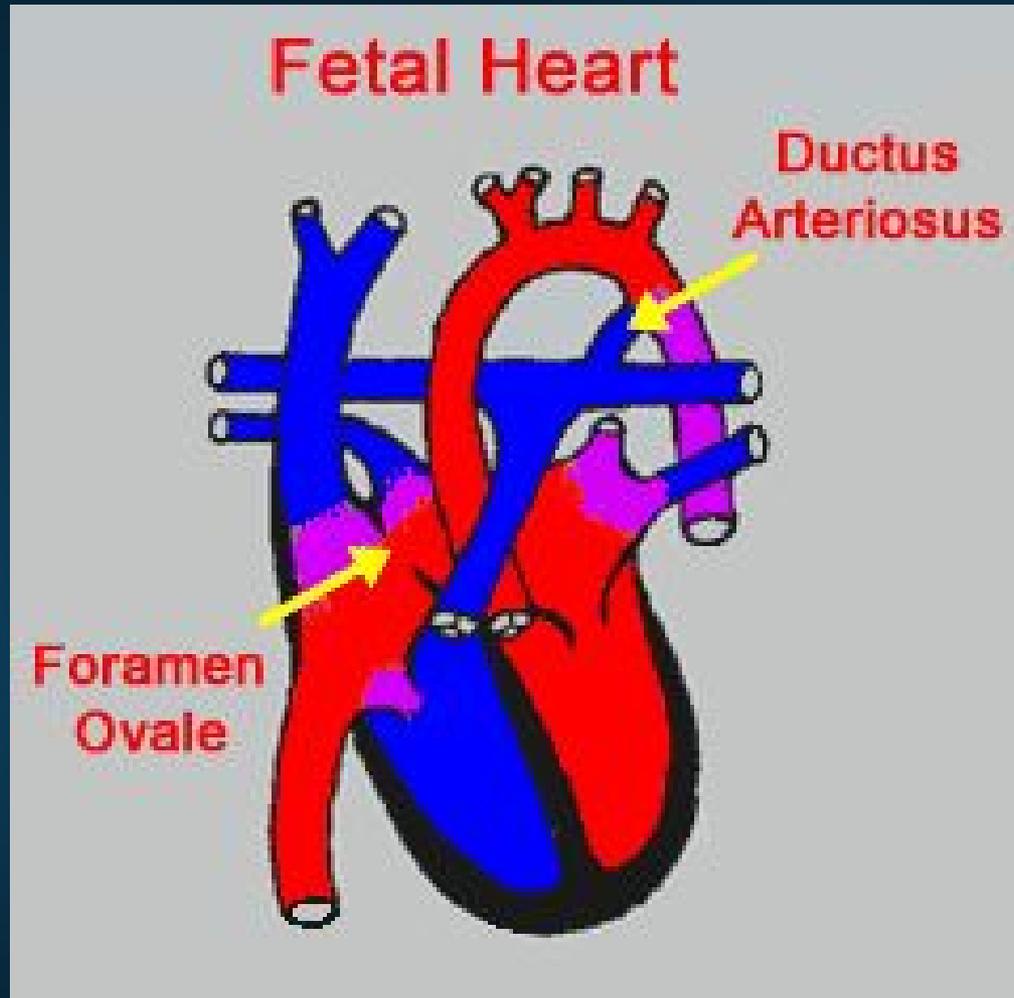
Low Pressure

Higher Pressure

TO LUNGS

TO BODY

Fetal Circulation



Fetal Circulation

Oxygenated
blood from
placenta

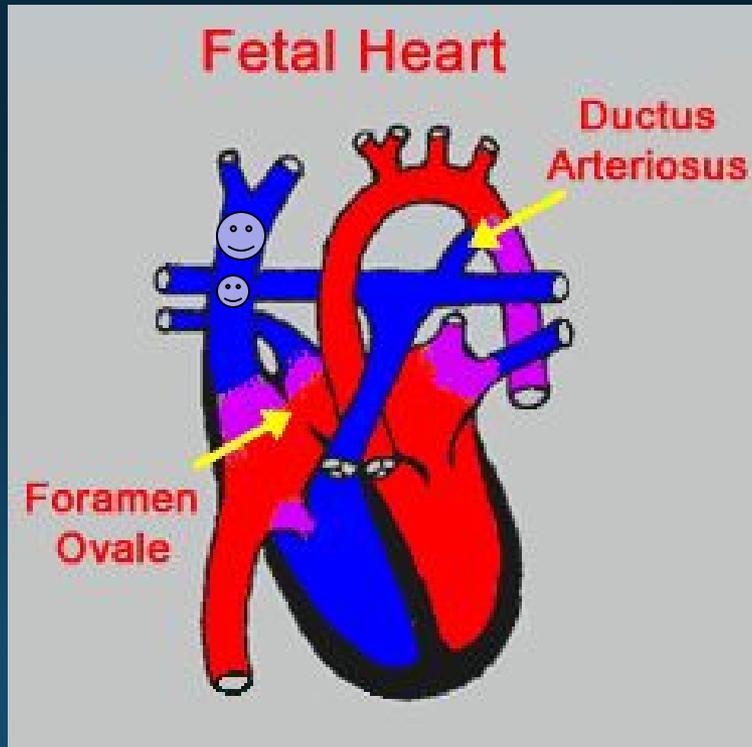
ALL PRESSURES EQUAL!

Oxygen sat ~60-70%

High Pressure
and Fluid
In Lungs



Fetal Circulation

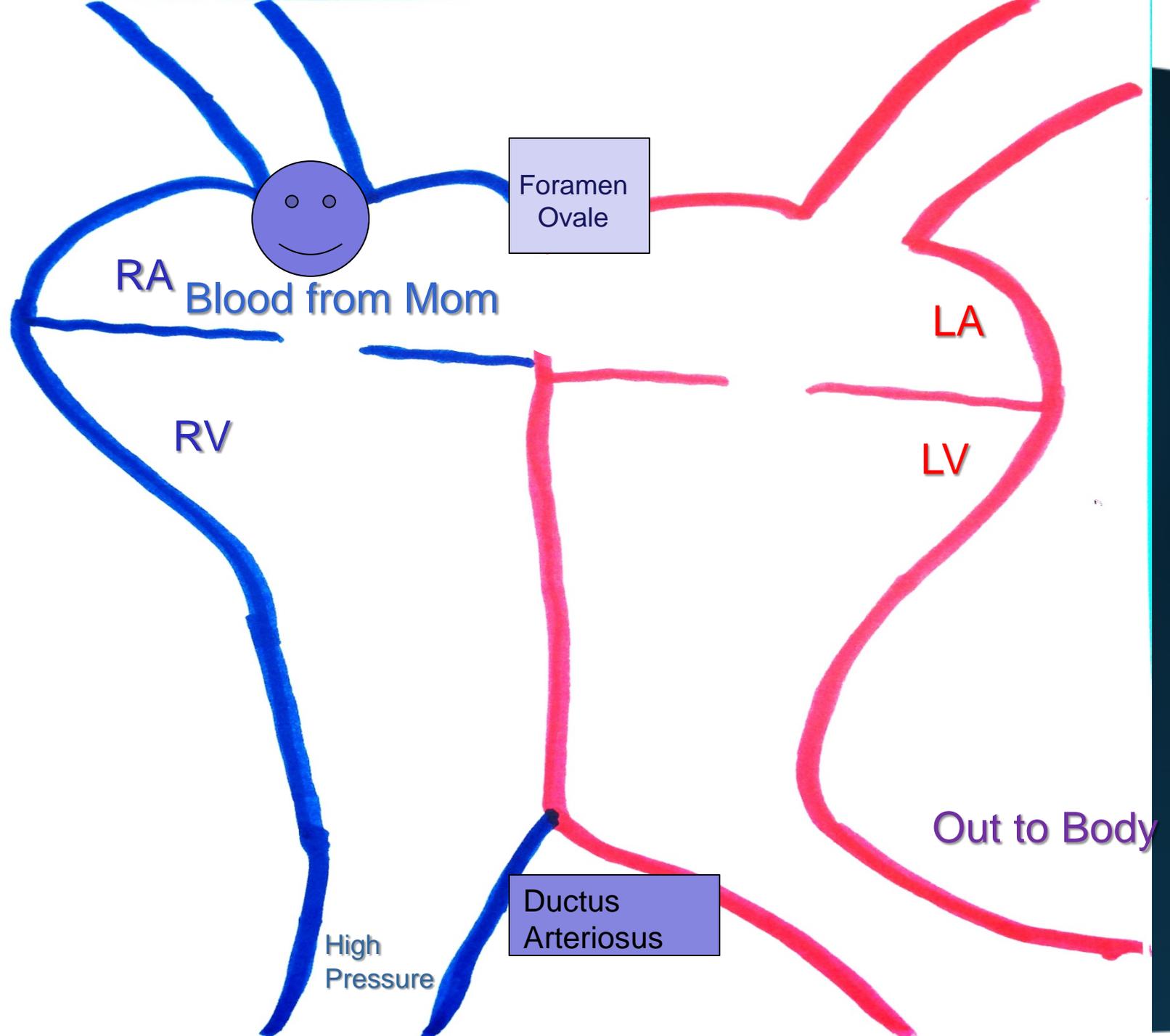


Foramen Ovale: Connects the right and left atria

Ductus Arteriosus: Connects pulmonary artery to aorta

Placenta → Right Atrium → Left Atrium → Left Ventricle → Aorta → Body

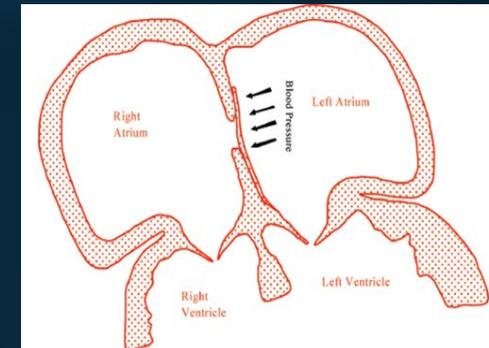
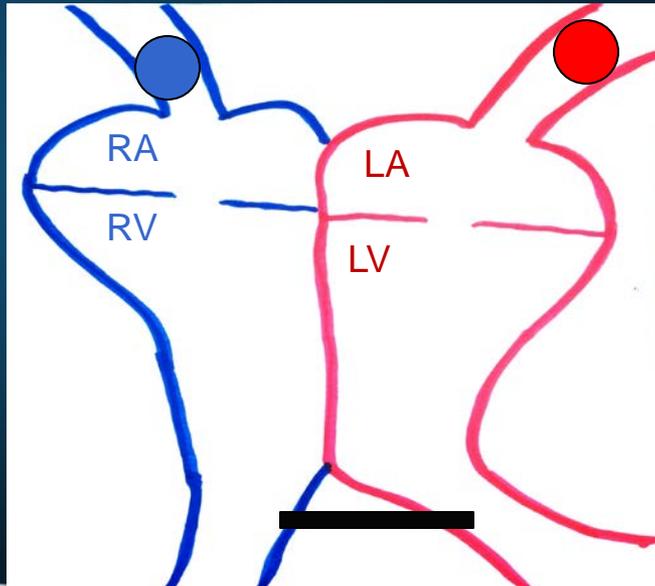
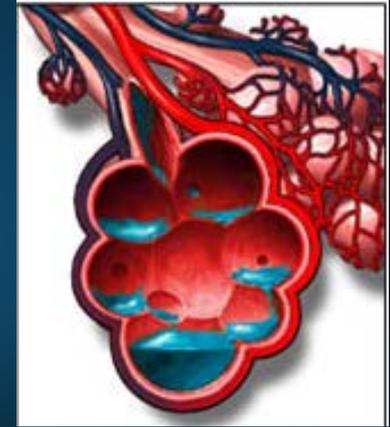
*1/3 of blood still goes to Right Ventricle: Right Ventricle → Pulmonary Artery → Ductus Arteriosus



Birth!

Baby takes first breath, fluid pushed out of lungs.

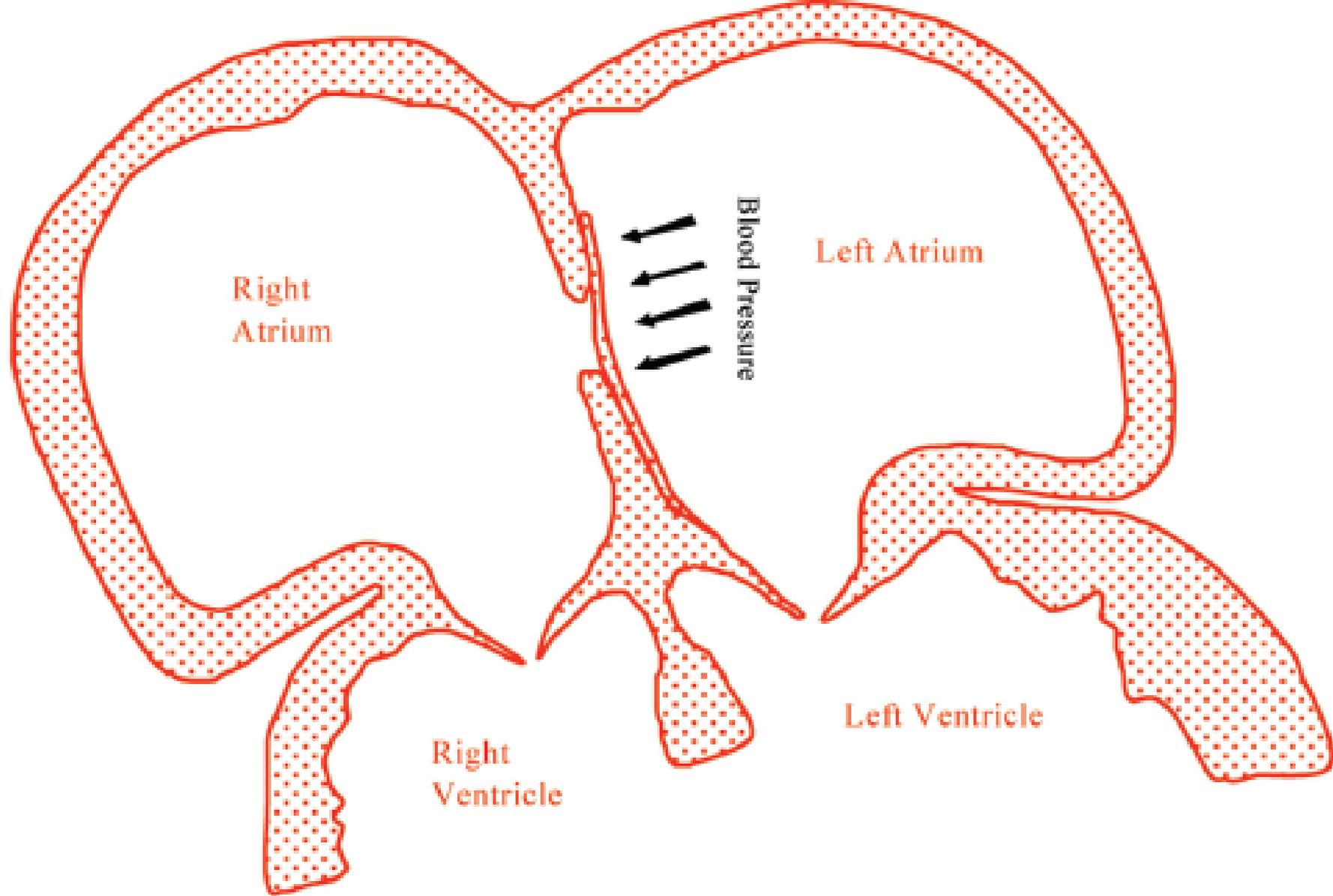
- Pressures change, blood flow changes
- PFO is pushed closed
- Increased oxygen content in blood reaches lining of the ductus arteriosus, which starts constricting and eventually closes





Targeted Pre-ductal SpO₂ After Birth

1 min	60%-65%
2 min	65%-70%
3 min	70%-75%
4 min	75%-80%
5 min	80%-85%
10 min	85%-95%

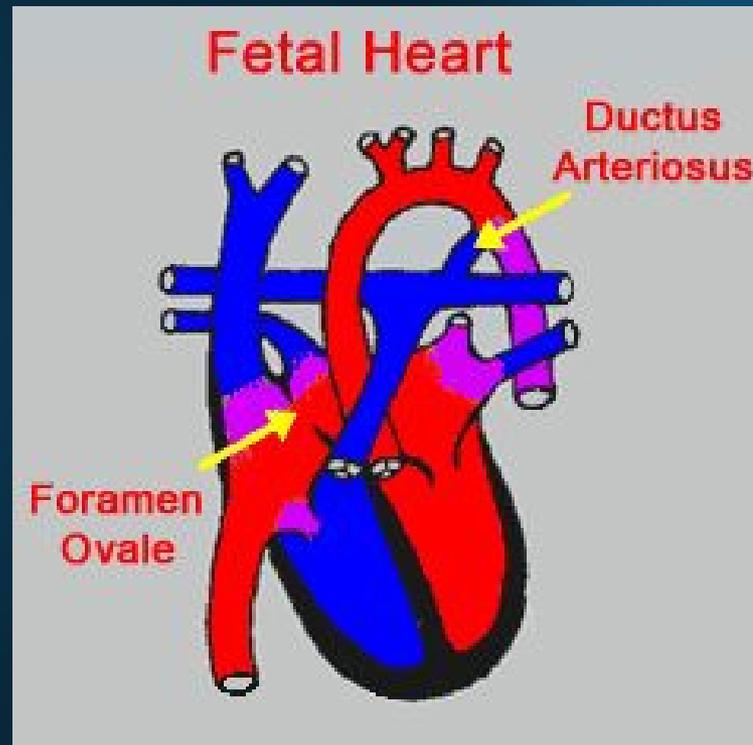


Normal status — higher left atrial pressure keeps the PFO

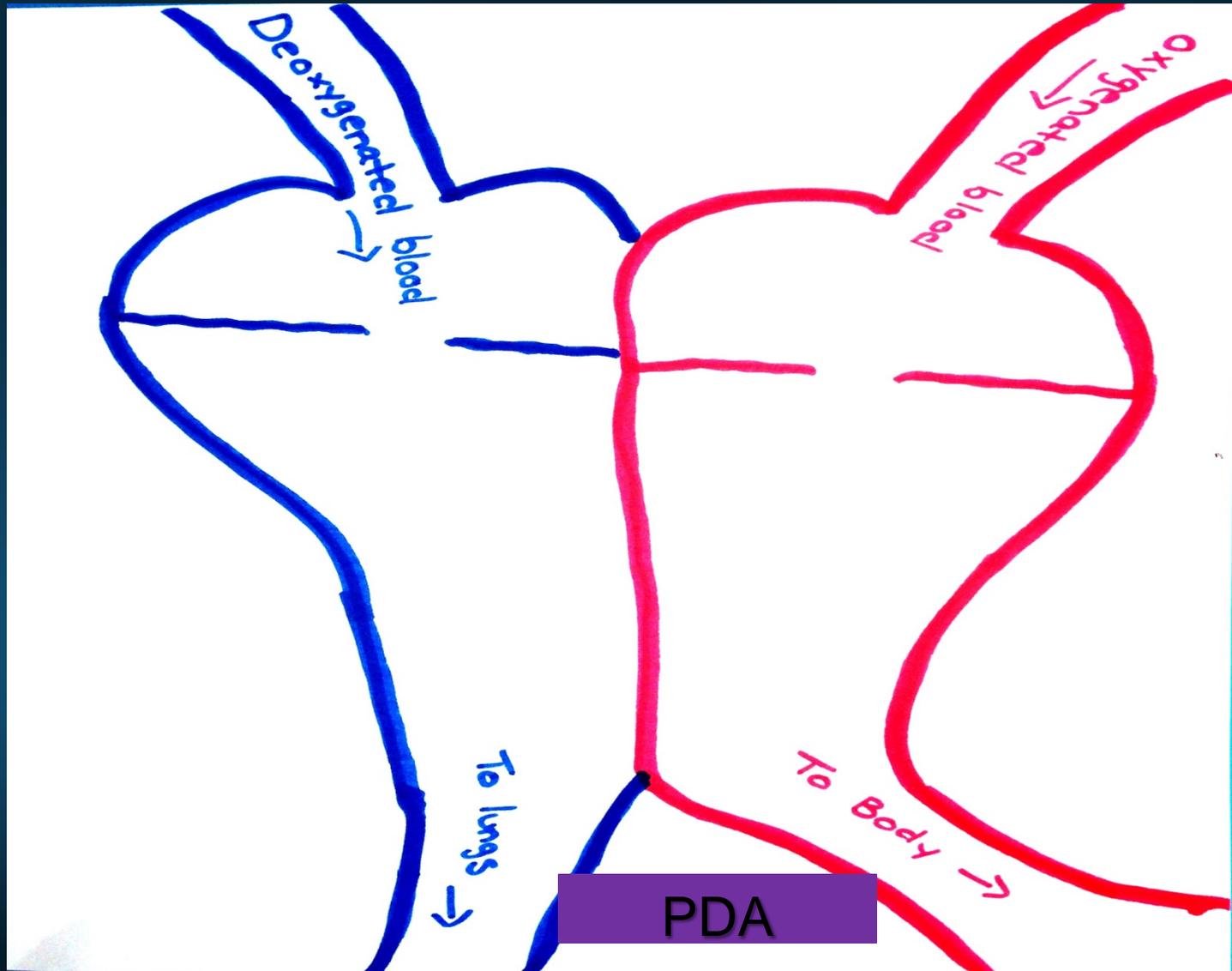
closed.

Sometimes Fetal Circulation Isn't Ready to Say Goodbye

- Patent Ductus Arteriosus
- Patent Foramen Ovale

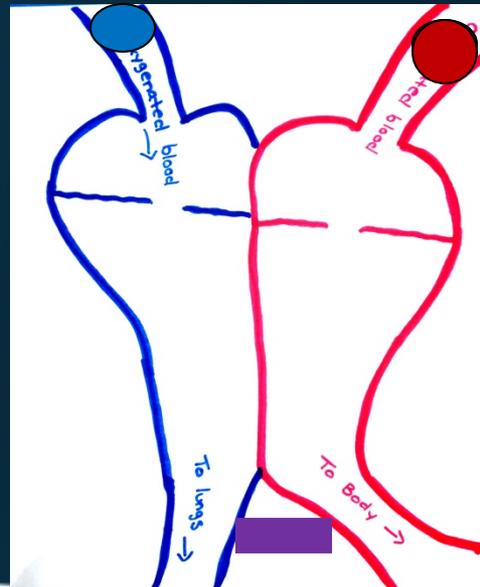
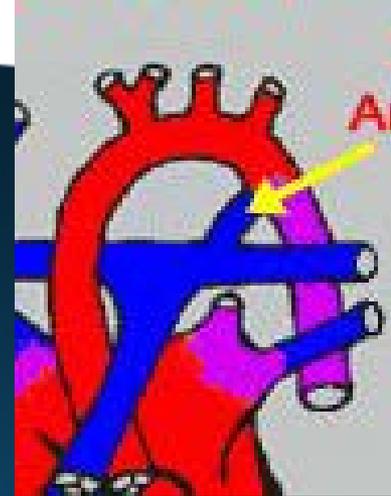


Patent Ductus Arteriosus



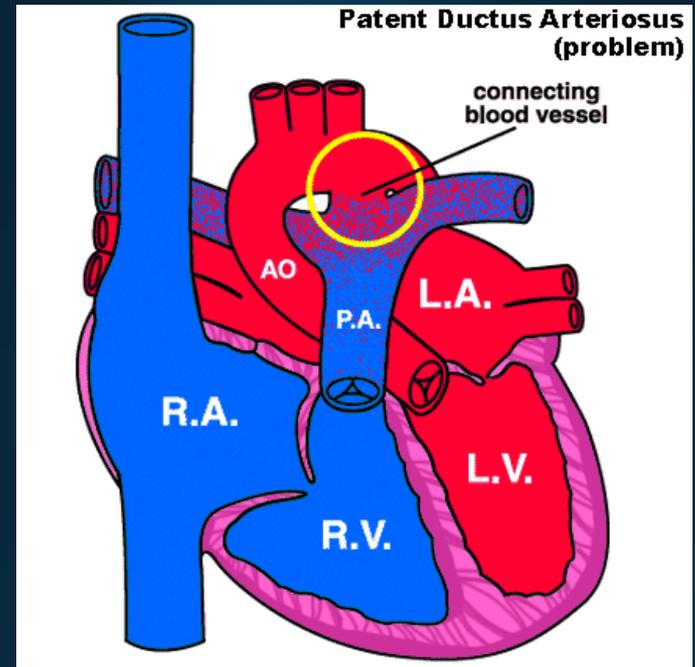
Patent Ductus Arteriosus (PDA)

- Connects pulmonary artery and aorta
 - Which way do you think the blood will flow?
- Blood backs up into the right side of the heart
 - What do we expect to see clinically?



Patent Ductus Arteriosus (PDA)

- Signs in baby:
 - Eventually, baby tires easily, trouble eating, crackles and murmur on auscultation (heart failure)
 - Only if PDA is large.
 - If small, the shunt is insignificant and patient is only at risk for endocarditis.





Congestive Heart Failure in Babies

- Tachycardia
- Tachypnea with crackles in bases
- Irritable at first then lethargic
- Pallor/cool skin
- Decreased cap refill
- Mottled lower extremities
- Sweaty
- Hepatomegaly
- May be underweight
 - Big head, tiny little body
- Dependent edema
 - Not legs if they're not walking or sitting up!
 - Eyes
 - Vulva/Scrotum

Develops
Slowly Over
Time

This Baby is Exercising



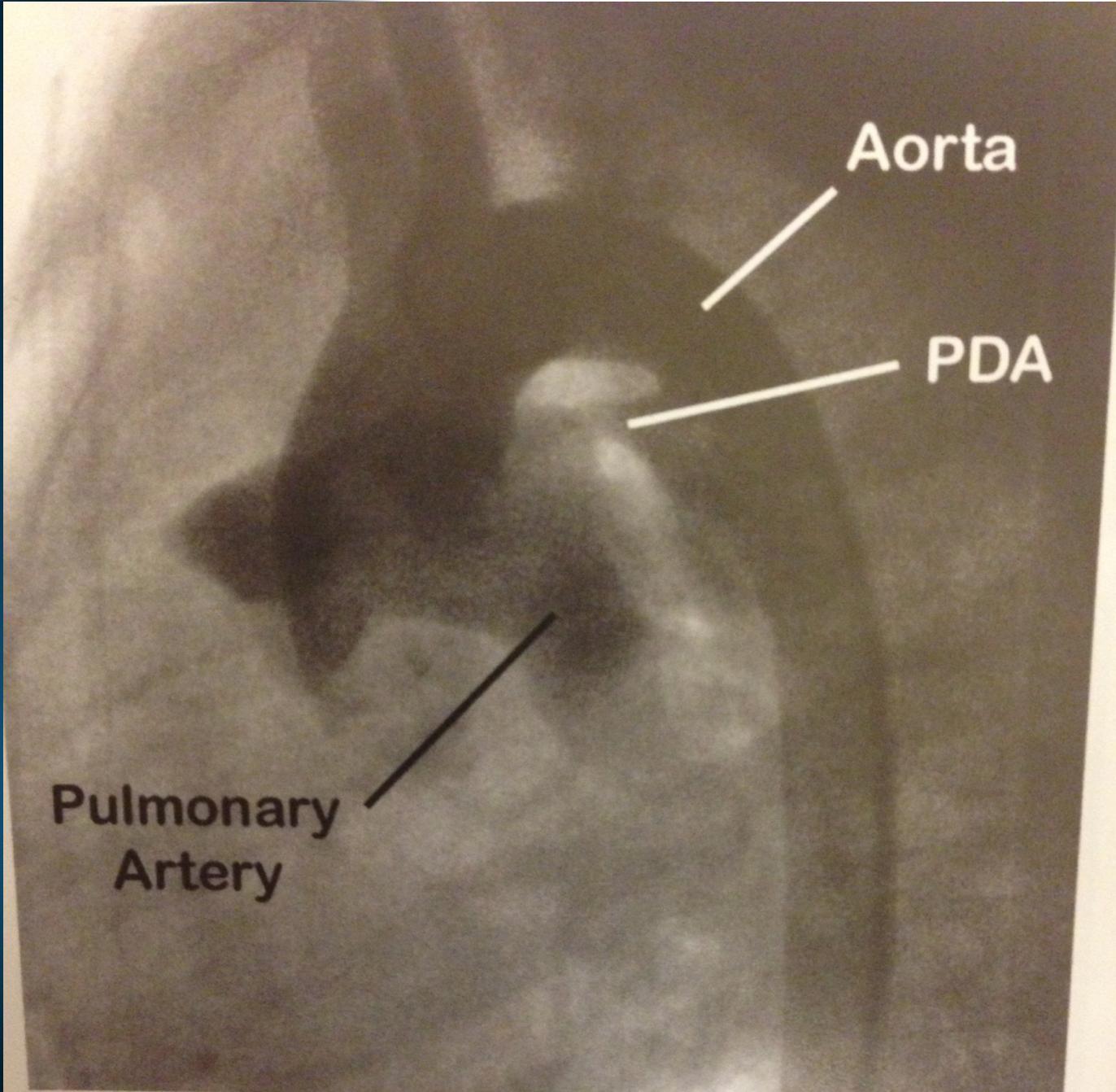
OXYGEN DEMAND

- Two ways to increase oxygenation:
 - Add Oxygen



- Take Away Less Oxygen/
Decrease Oxygen Demand
 - Crying
 - Fever
 - Tachycardia





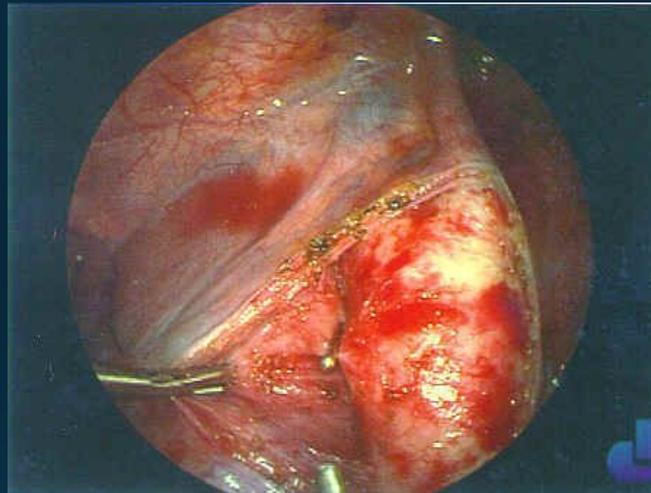
Aorta

PDA

Pulmonary
Artery

PDA Hospital Treatment

- Close with indomethacin or ibuprofen
 - Why we don't give pregnant women these drugs in 3rd trimester
 - If PDA doesn't close with medical therapy, can be closed in the cath lab with coil embolization or other device.
 - Surgical closure is by ligation or metal clip



PDA Hospital Treatment

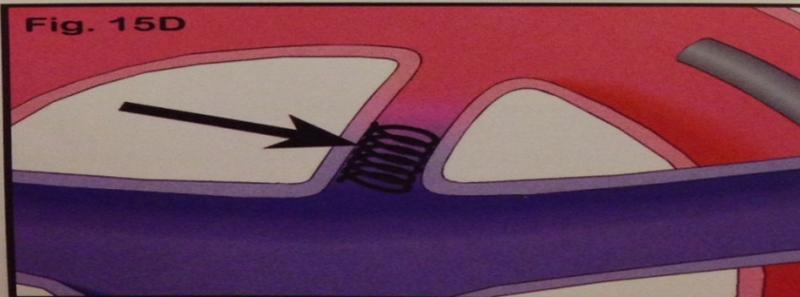
Fig. 15B



Fig. 15C



Fig. 15D

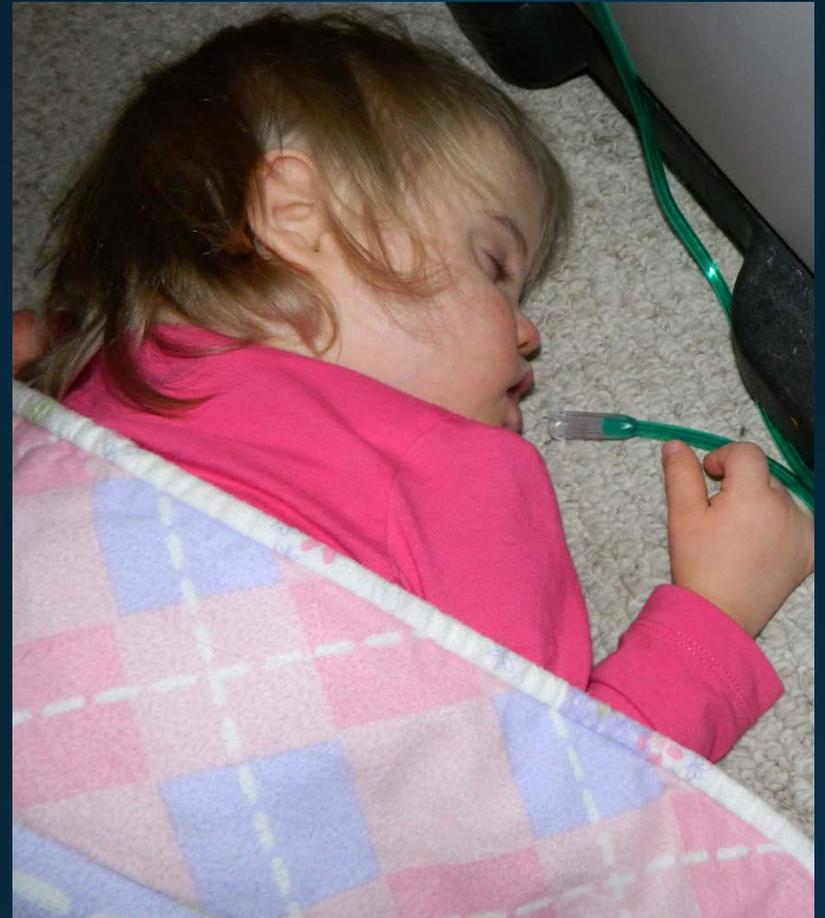


PDA EMS Treatment

- You won't know it's a PDA in your ambulance
 - Baby with gradual trouble breathing and crackles, dependent edema possible.
 - Keep happy
 - Assist breathing if necessary
 - Oxygen?
 - PPV if needed



Blow-By Oxygen



Nasal Cannula in Children



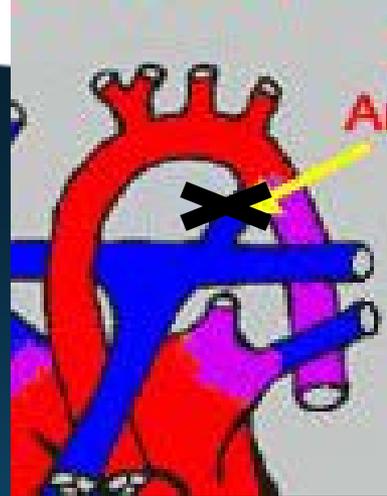
Effects of Oxygen

- Oxygen is a vasodilator
 - Decreases pulmonary pressures
 - Increases blood flow to the lungs, which decreases blood flow from the systemic circulation
- Oxygen is a part of what makes the PDA close at birth



PDA Dependent Lesions

- The PDA usually closes within the first week of life. If the child has an undiagnosed defect that was dependent on this shunt, the baby will experience SEVERE, ACUTE decompensation.
- EMS Presentation:
 - Acute rapid onset cyanosis, tachycardia, tachypnea
 - Signs of shock (decreased cap refill, grey appearance)
- Treatment: Symptom-dependent, get to hospital



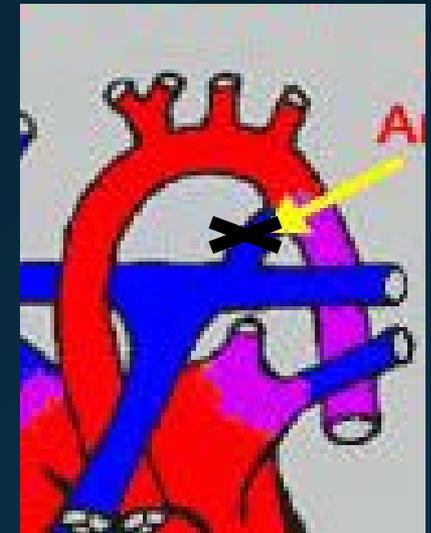
PDA-Dependent Lesions

Hospital Treatment:

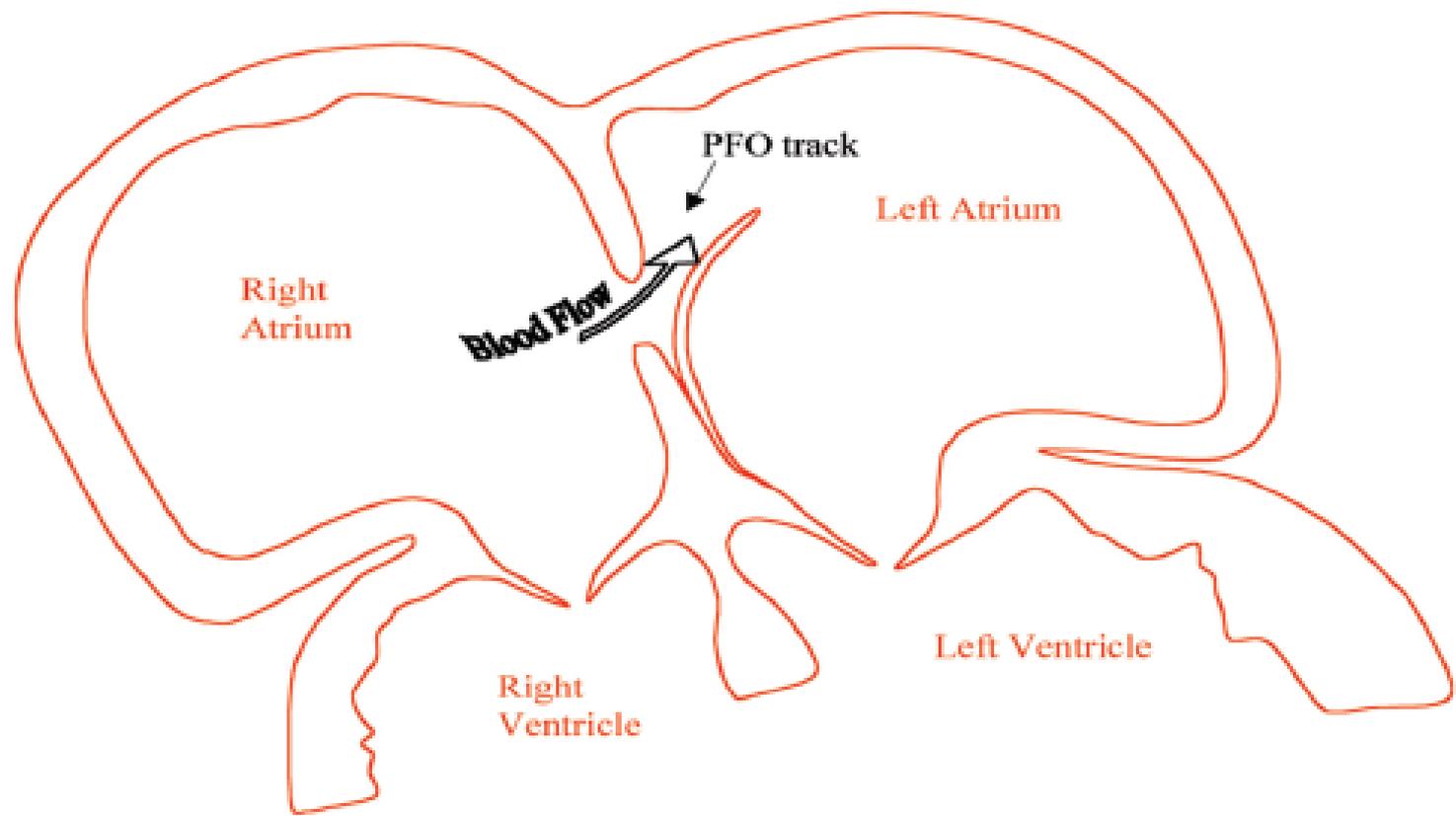
- Keep open with prostaglandin E₁ infusion in hospital (“Keeeeeep open”)
- Begin surgery workup with echo and consults

Prehospital Treatment:

- Get to hospital as quickly as possible. Call ahead as soon as possible with report.



Patent Foramen Ovale (PFO)



. Right atrial pressure exceeds left atrial pressure — PFO opens.

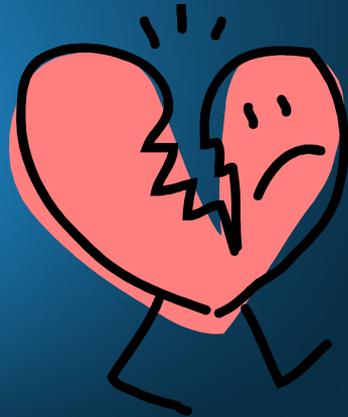


PFO vs Other Atrial Septal Defects (ASDs)

- PFO is technically a type of atrial septal defect
- PFO is a flap, so it's only open when pressures are higher on the right
 - Thus Right to Left shunt
 - Mixes deoxygenated blood with oxygenated blood
 - Air and thrombi can pass directly to left atrium and then to the brain

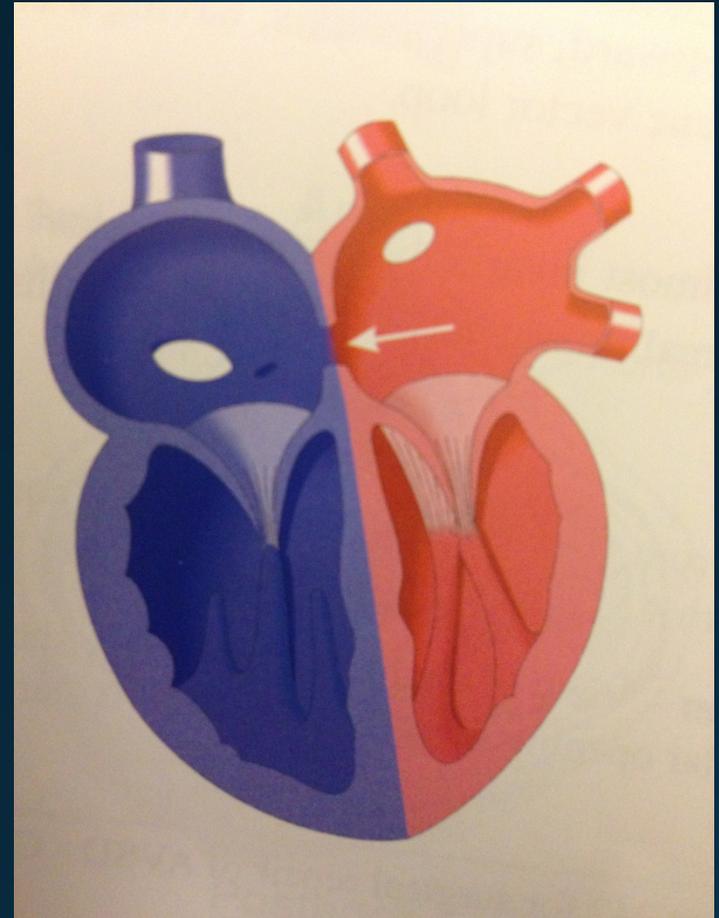
The Septal Defects

Atrial and Ventricular



Atrial Septal Defect

- Creates a left-to-right shunt
- Effect depends on size

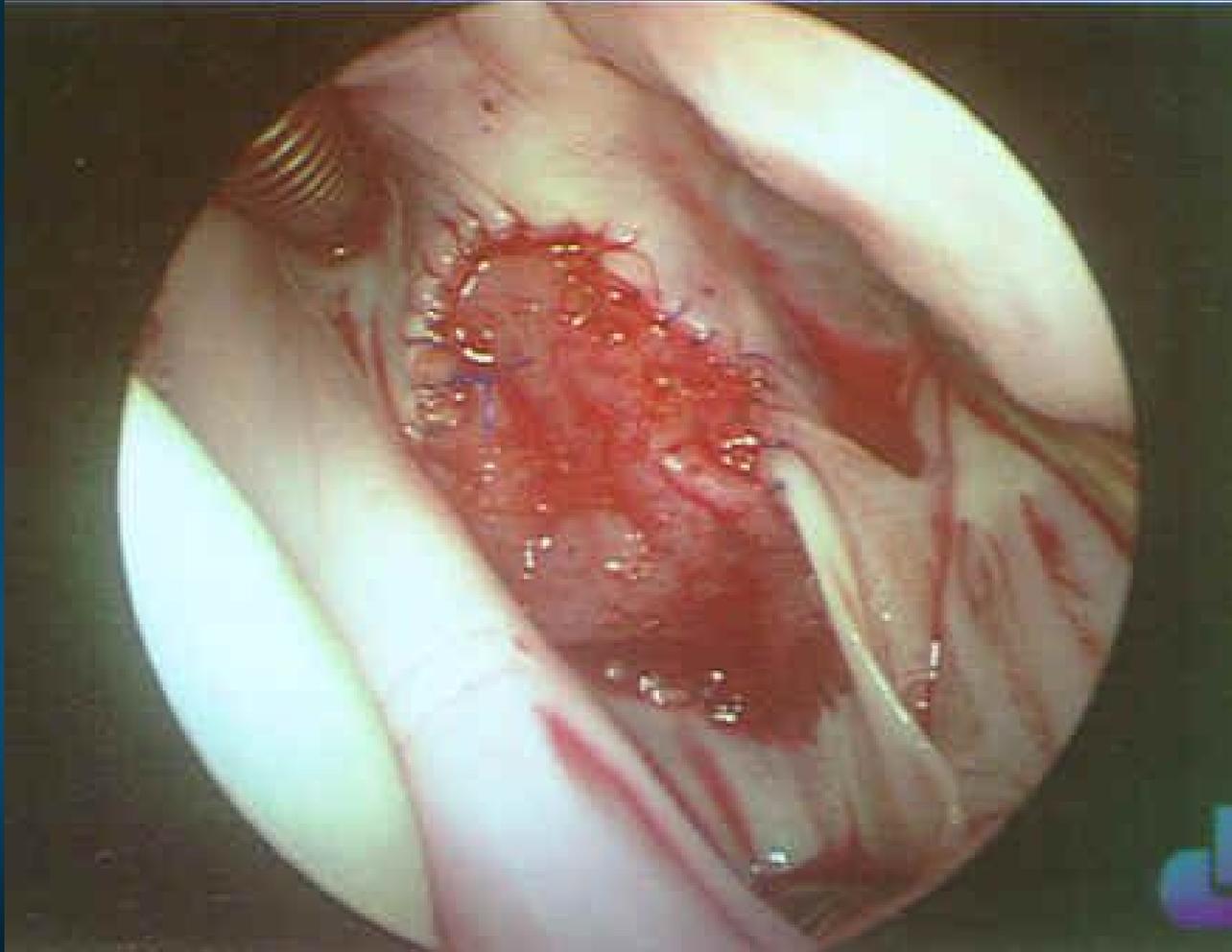




ASD Complications

- Increased blood flow to the right side of the heart and pulmonary circulation
- Over time, right atrial and ventricular dilation
- Pulmonary congestion
- Eventually if untreated, pressure on right side keeps increasing until right-to-left shunt develops
 - Risk for emboli and stroke

Hospital Treatment



Hospital Treatment

Fig. 18C

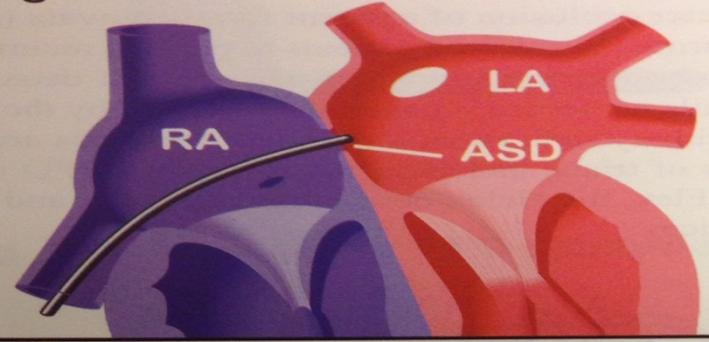


Fig. 18D



Fig. 18E

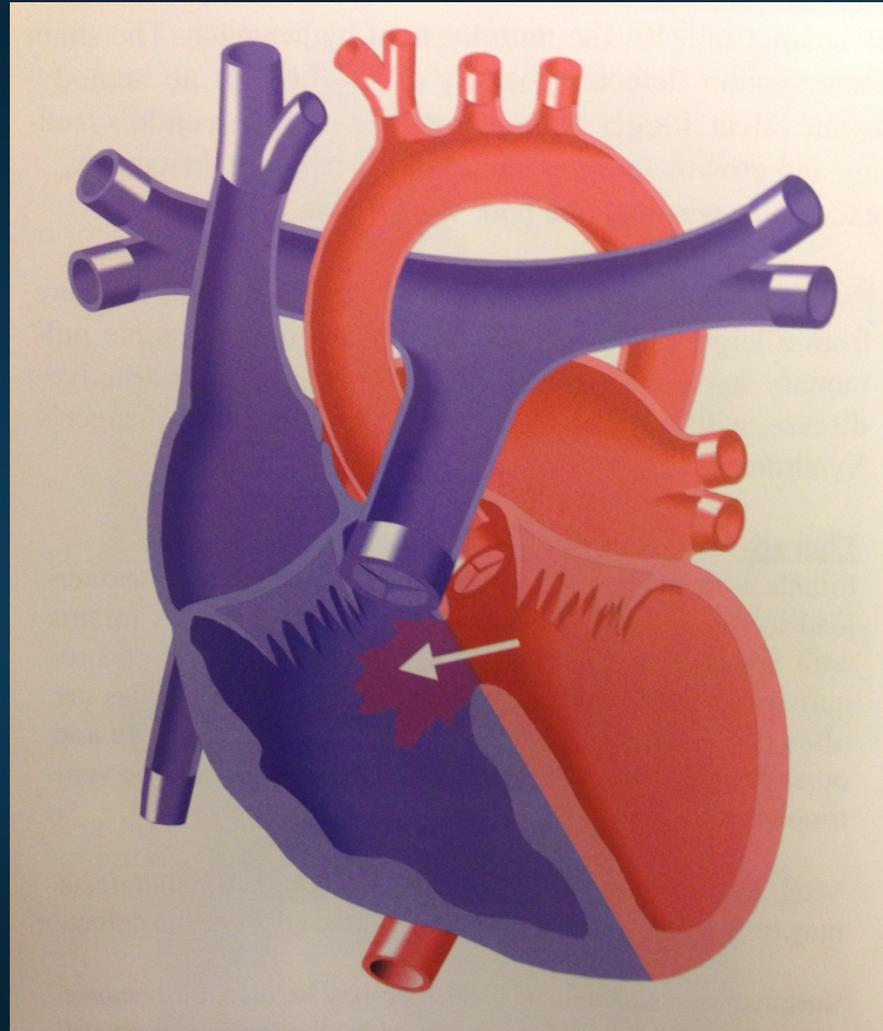


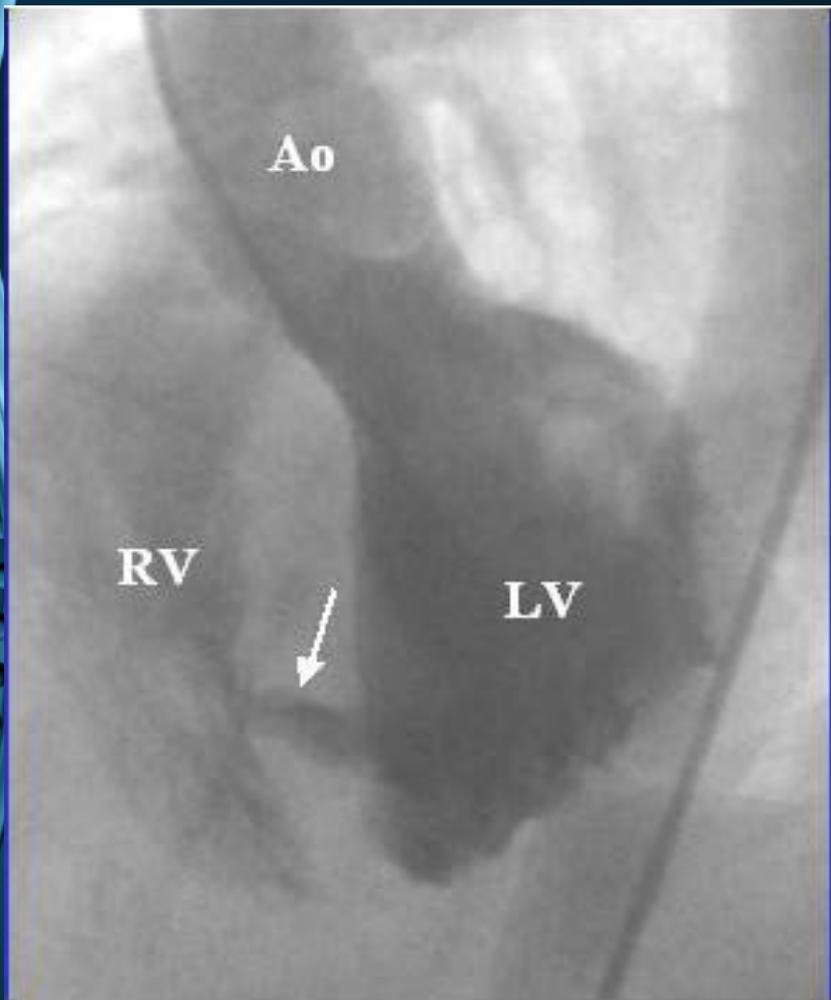
EMS Treatment

- Keep the baby happy!
- Signs of heart failure?
 - Oxygen Options?
 - PPV if needed
 - Support to hospital



Ventricular Septal Defect







VSD Complications

- All the same left-to-right complications we've discussed already
 - Pulmonary congestion, pulmonary HTN, eventual heart failure and eventually right-to-left shunt
- VSDs are larger, so L-R shunt is worse, symptoms are worse
 - More commonly associated with poor feeding and growth, rapid breathing, irritability, excessive sweating

EMS Treatment

- Keep baby happy
- Signs of heart failure?
 - Oxygen ?
 - PPV if needed
 - Support to hospital
- Diuretics?

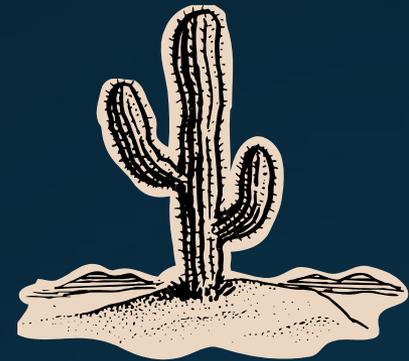


Heart Failure vs PDA Closure

- Slow and progressive vs sudden decompensation
- Crackles vs Dry lung sounds
- Tachypnea vs Increased WOB



VS





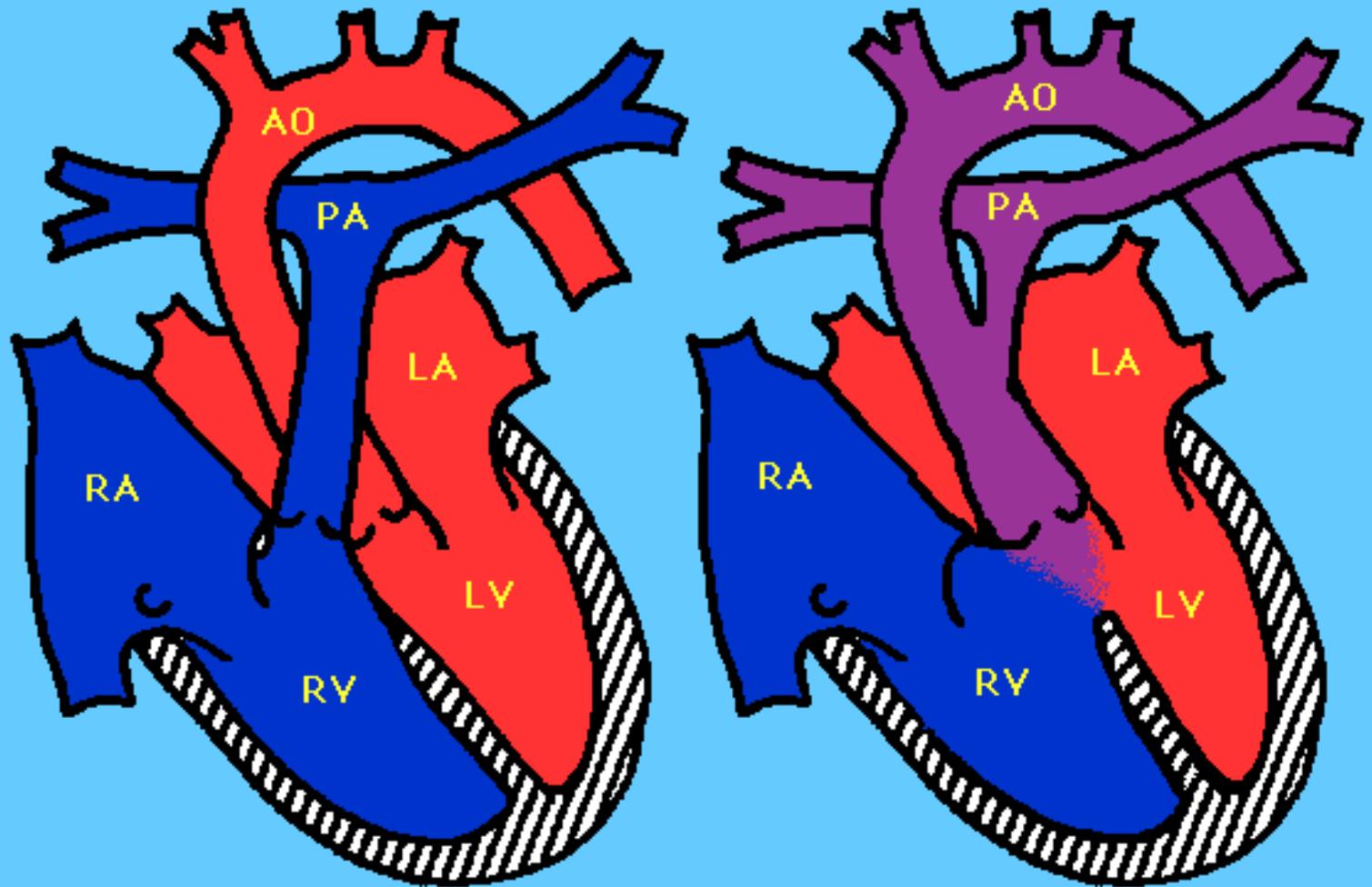
Eisenmenger Syndrome

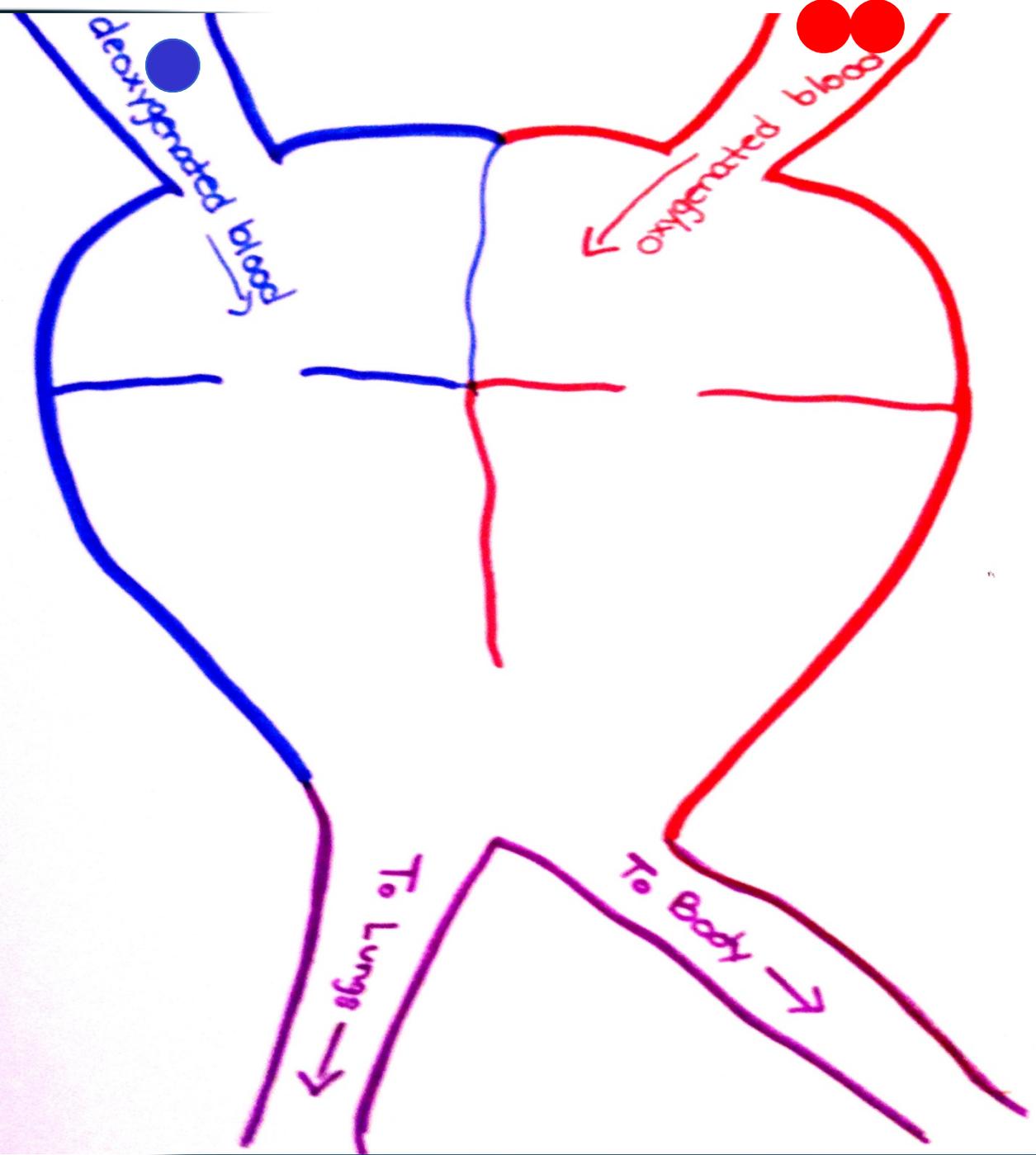
- Eventually the Left to Right shunts cause enough damage that pressure is higher on the right and the shunt switches
- Right to Left Shunt = Cyanotic RIGHT now
- Left to Right shunt = Cyanotic Later

CYANOTIC DEFECTS

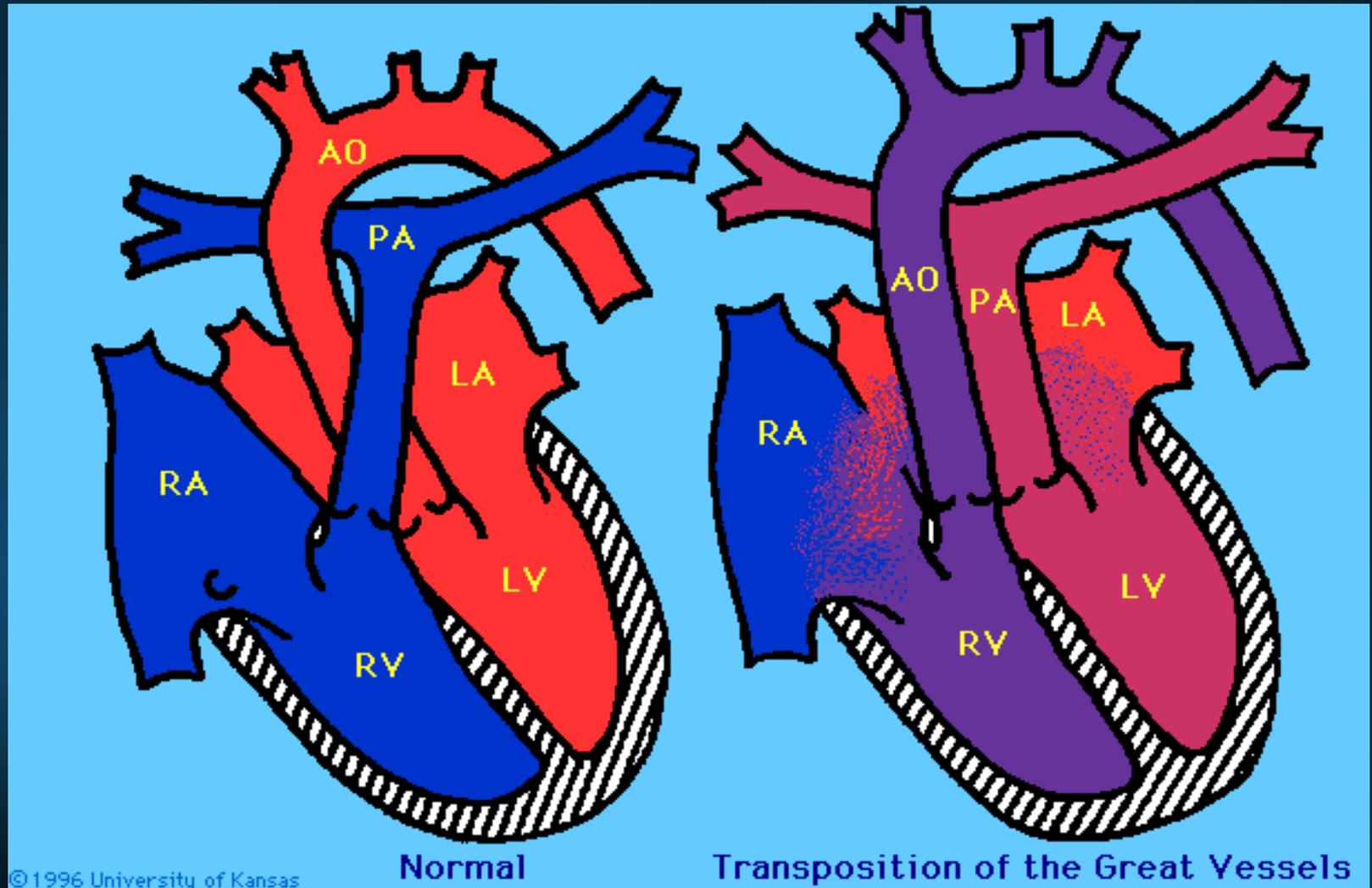


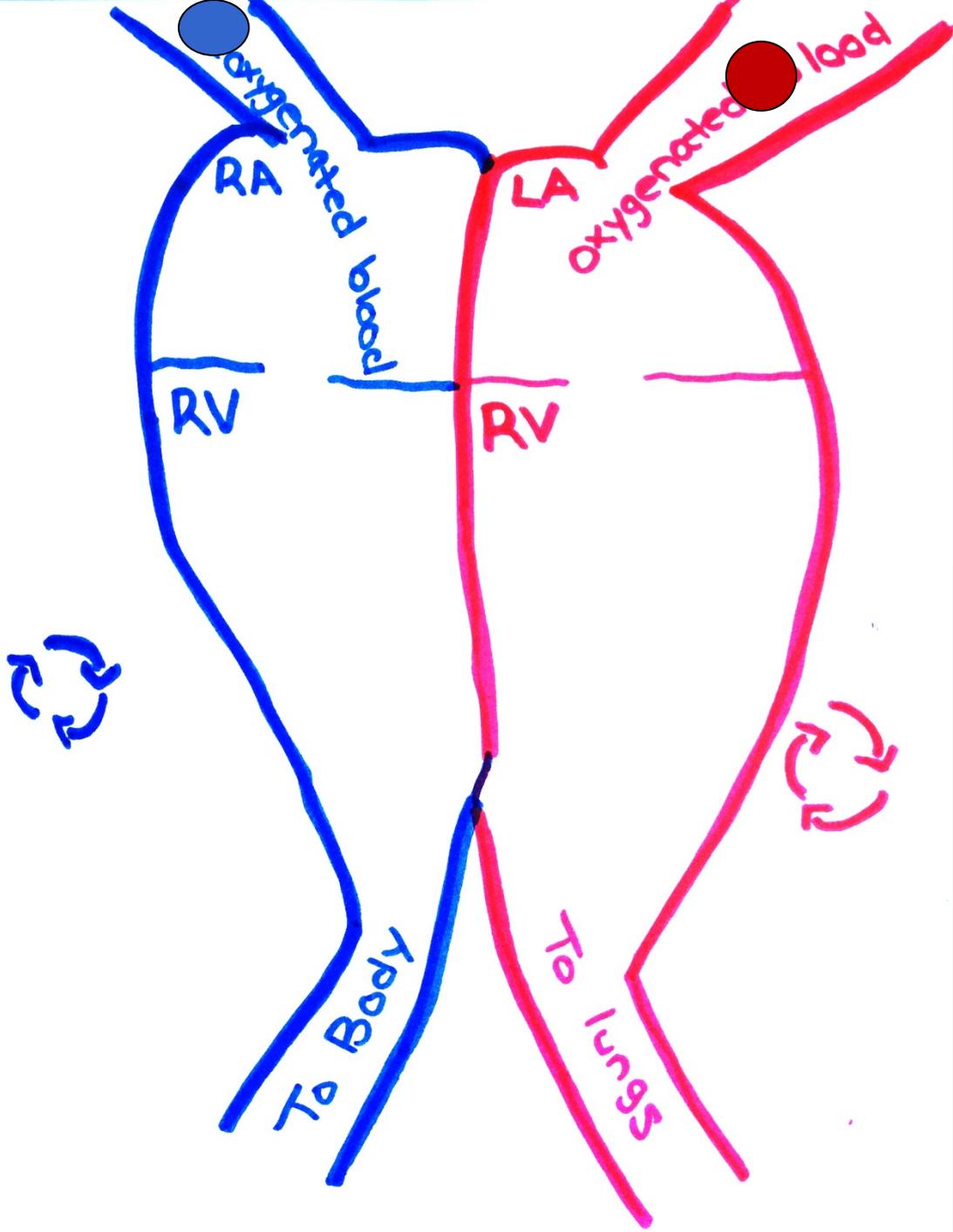
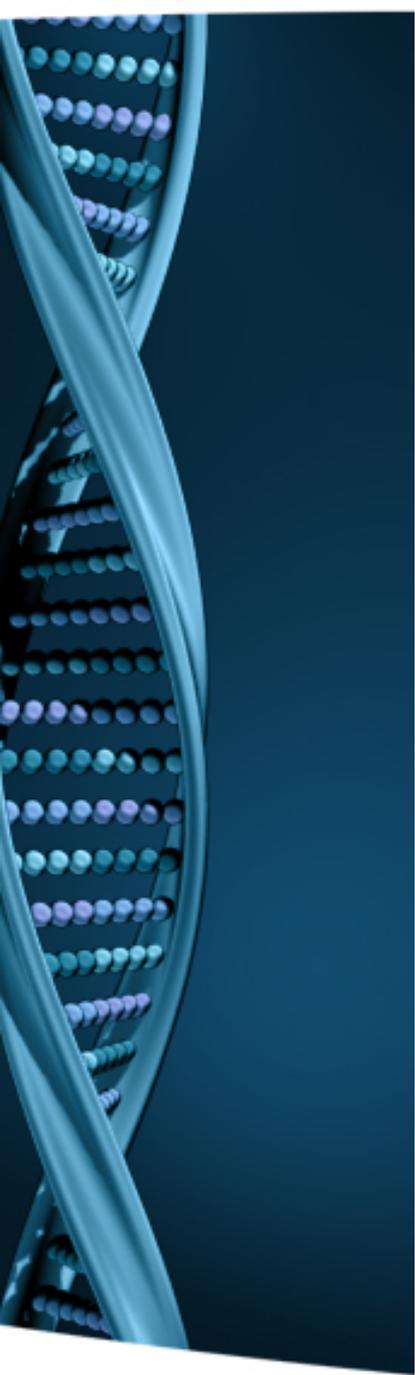
Truncus Arteriosus

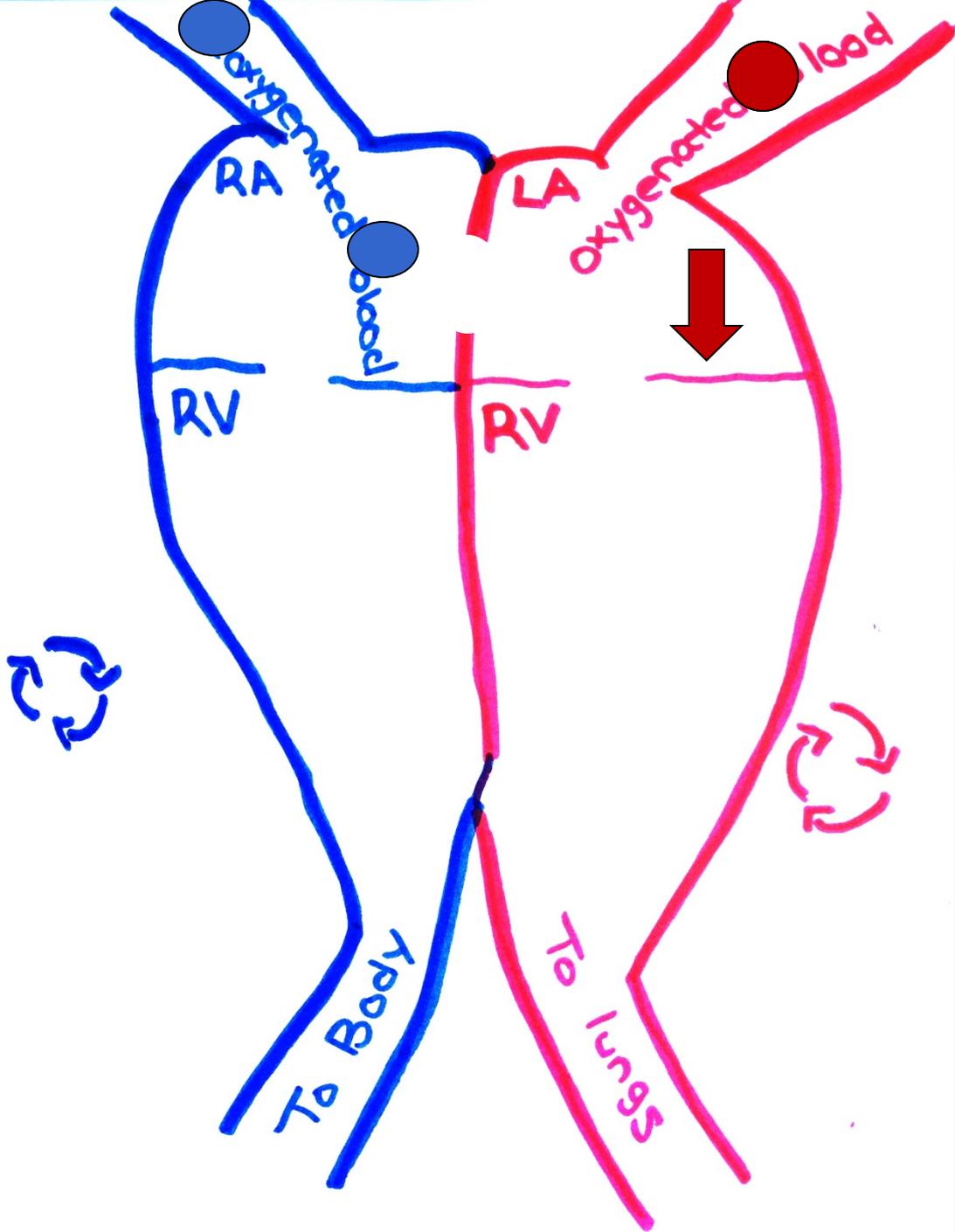
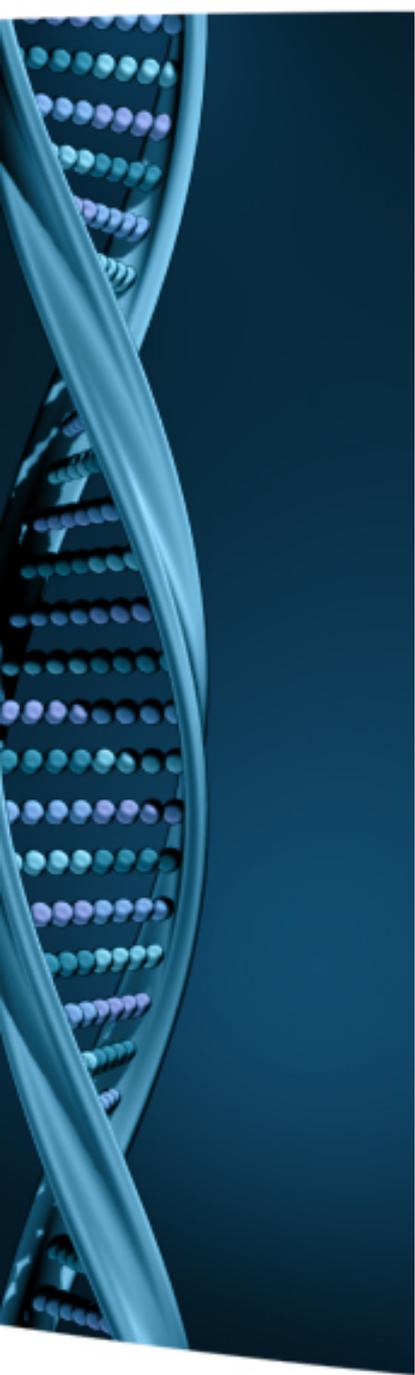


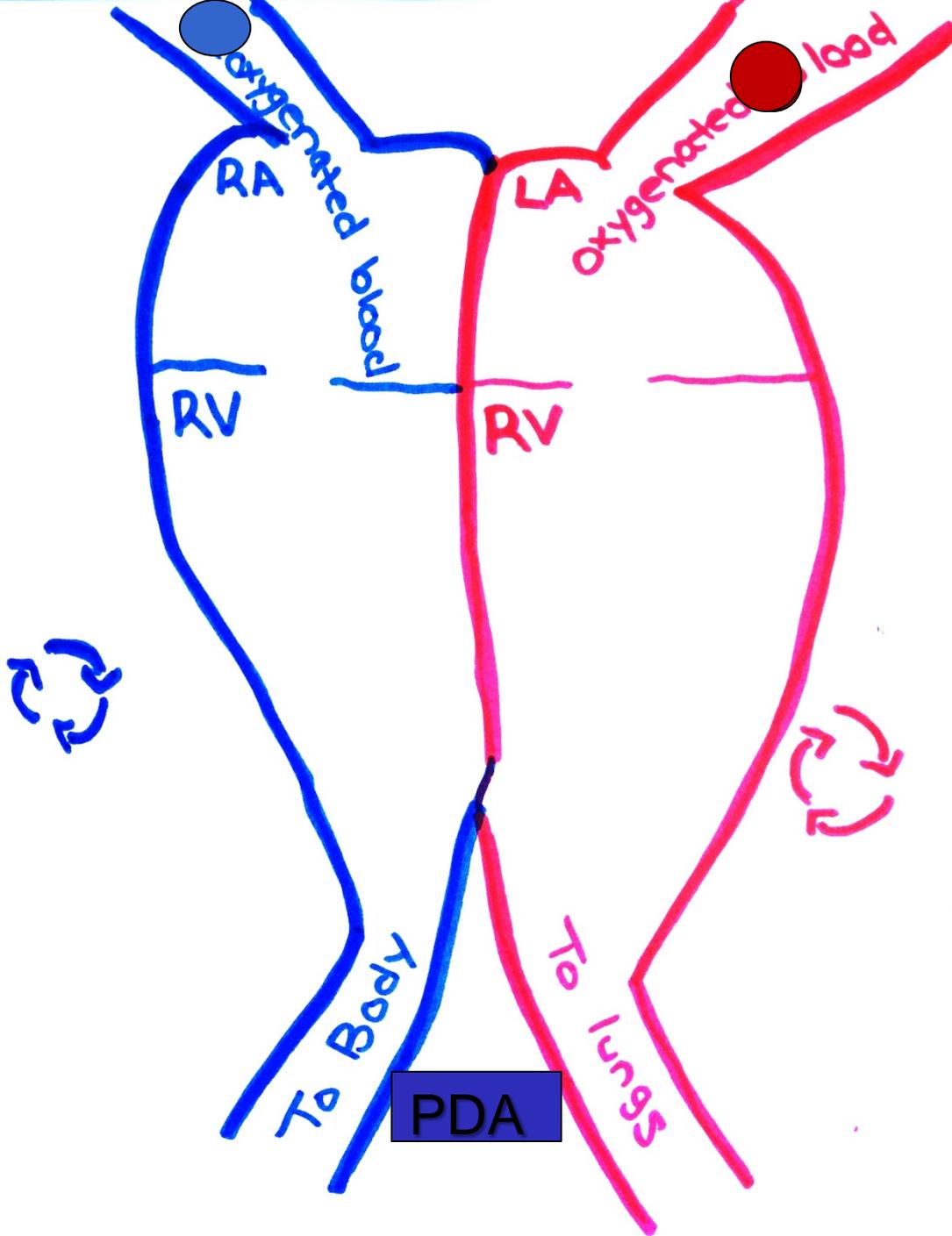
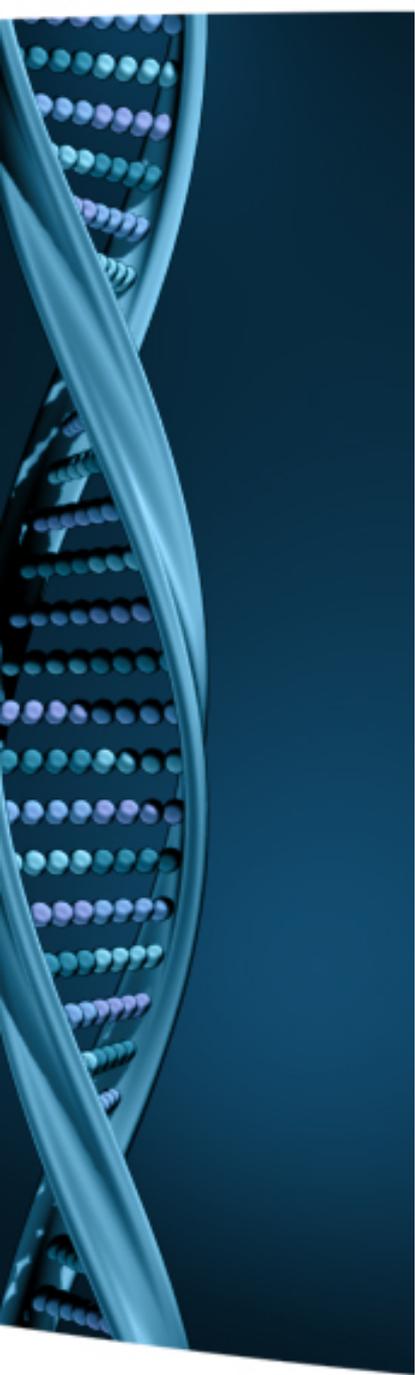


Transposition of the Great Arteries (D-type)



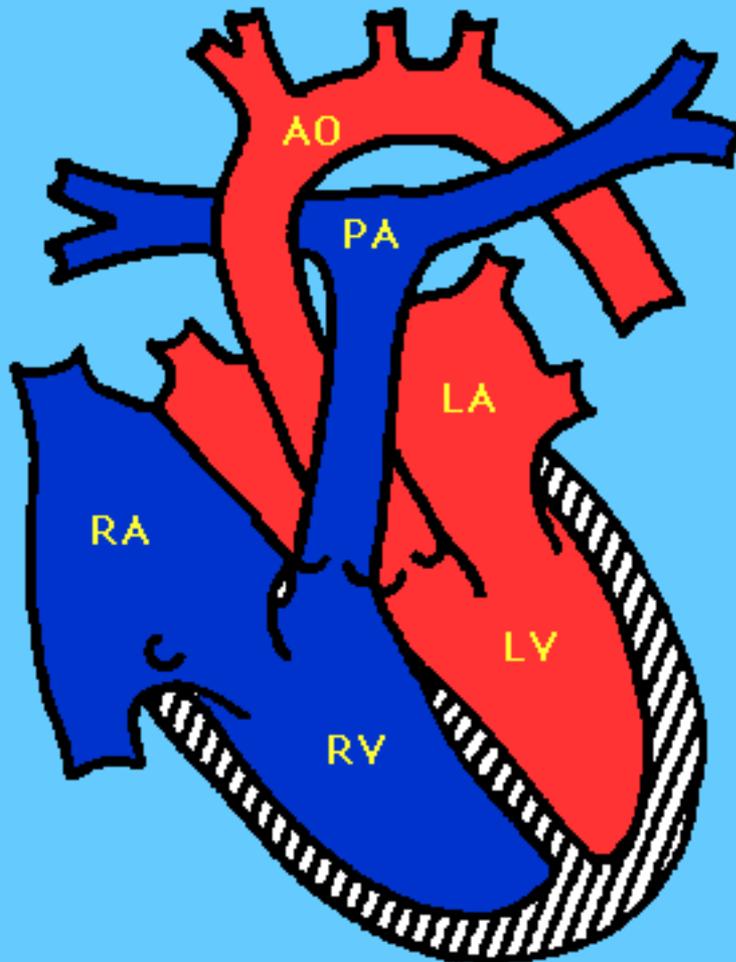




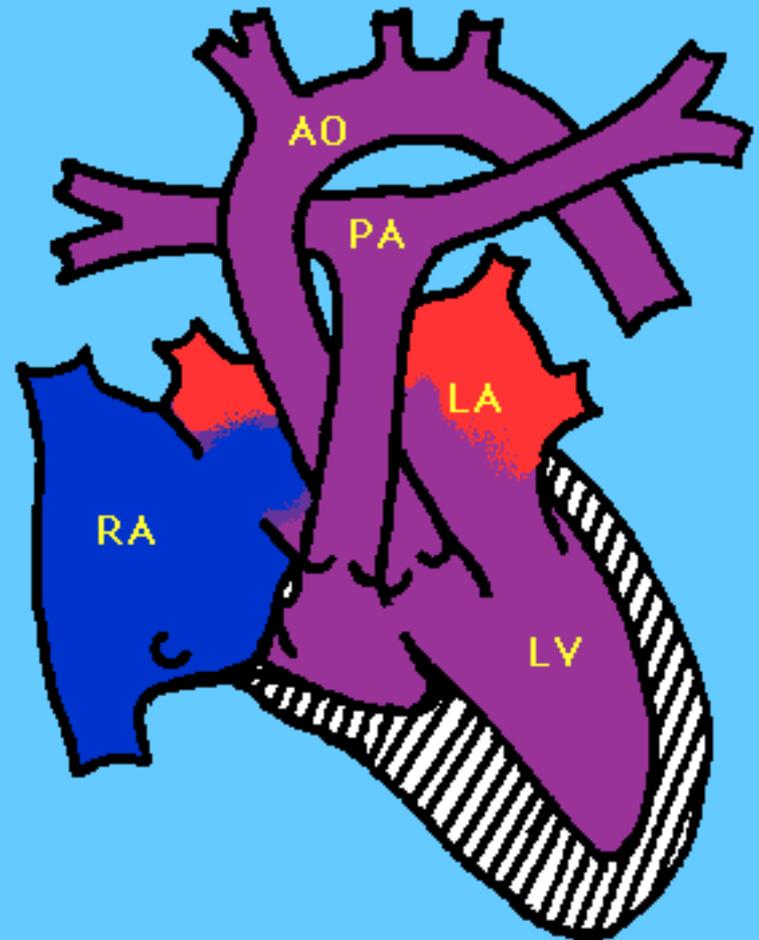


Tricuspid Atresia

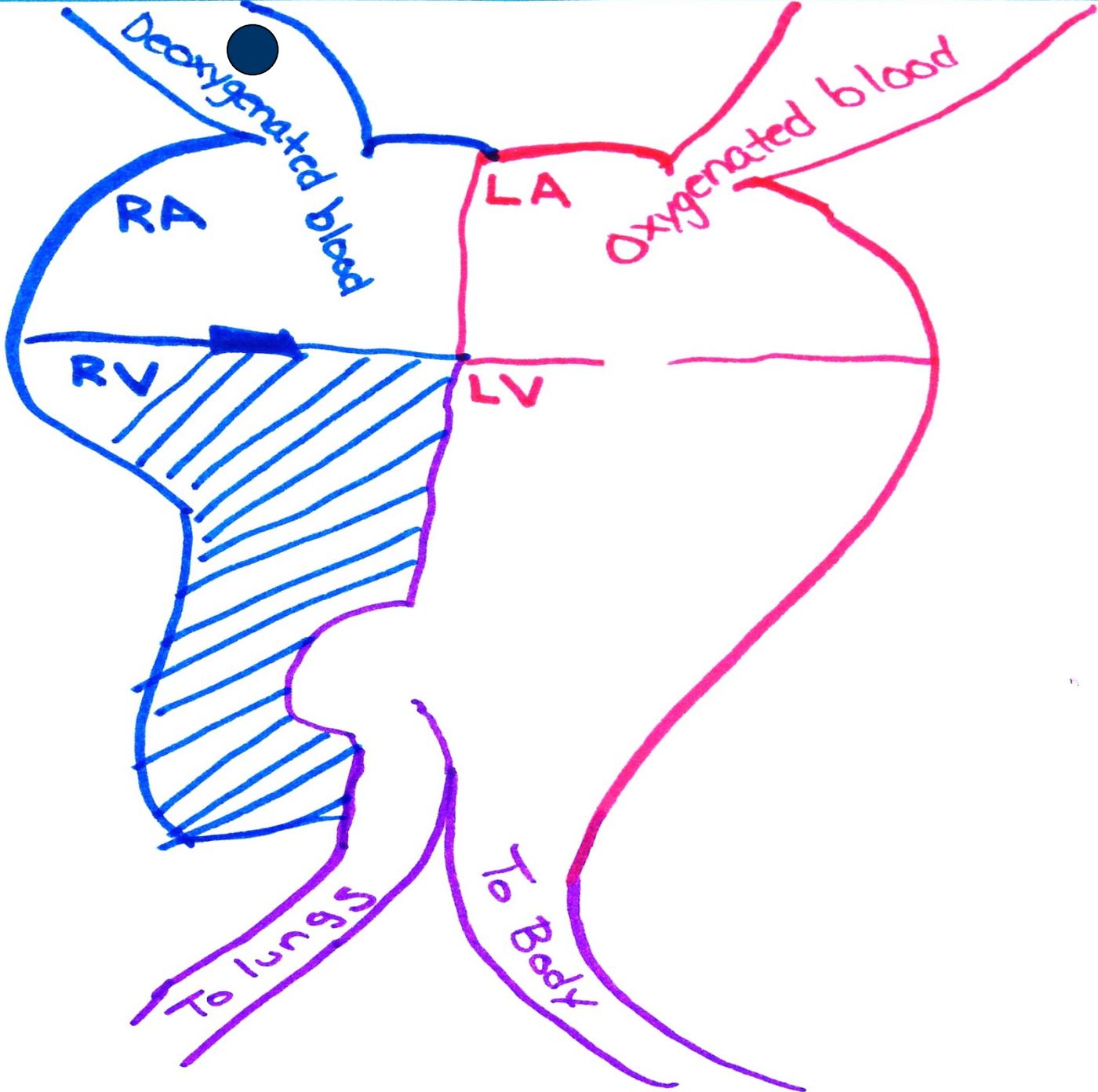
- “Atresia” = Absent/Never formed/Closed

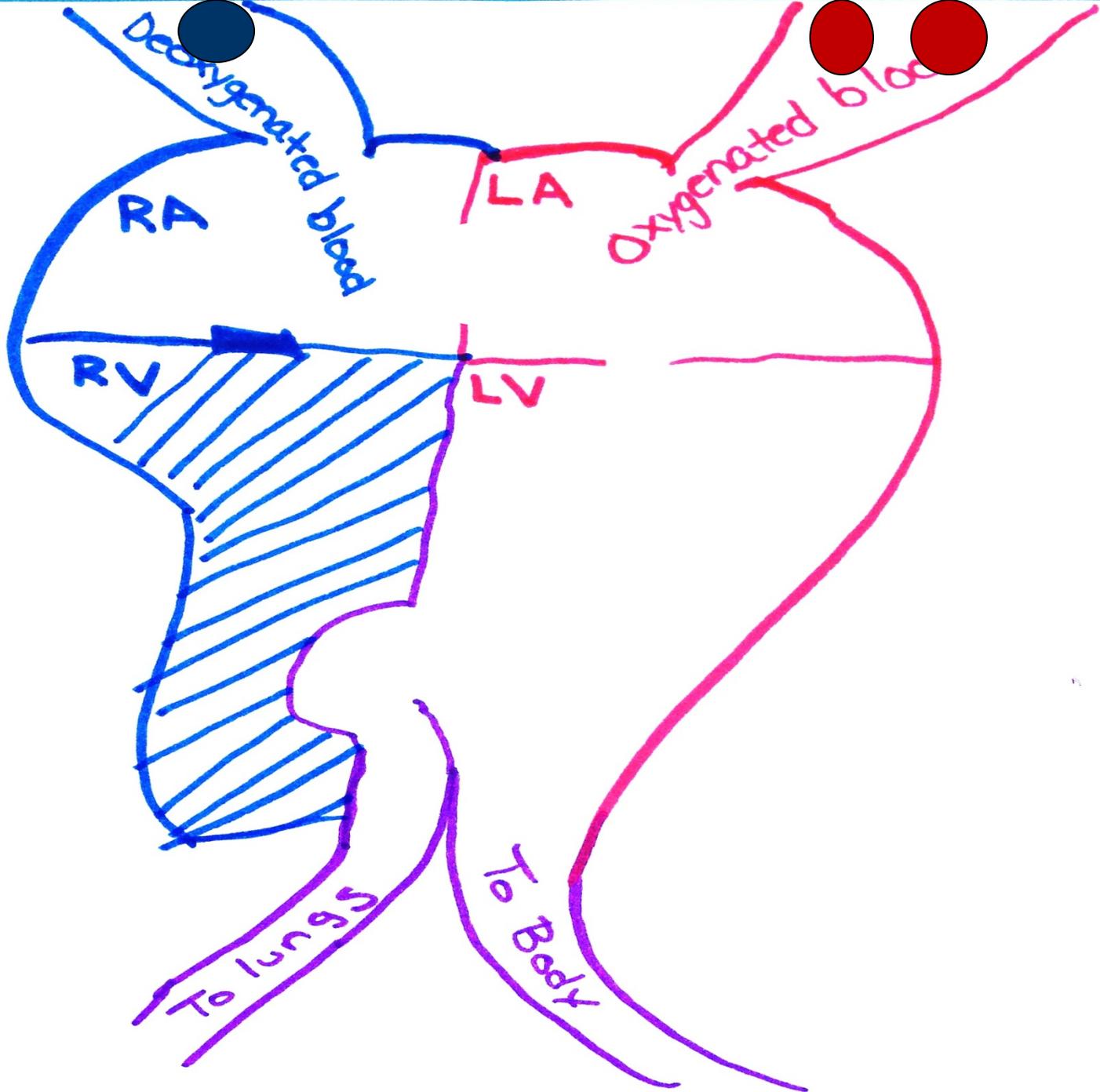


Normal

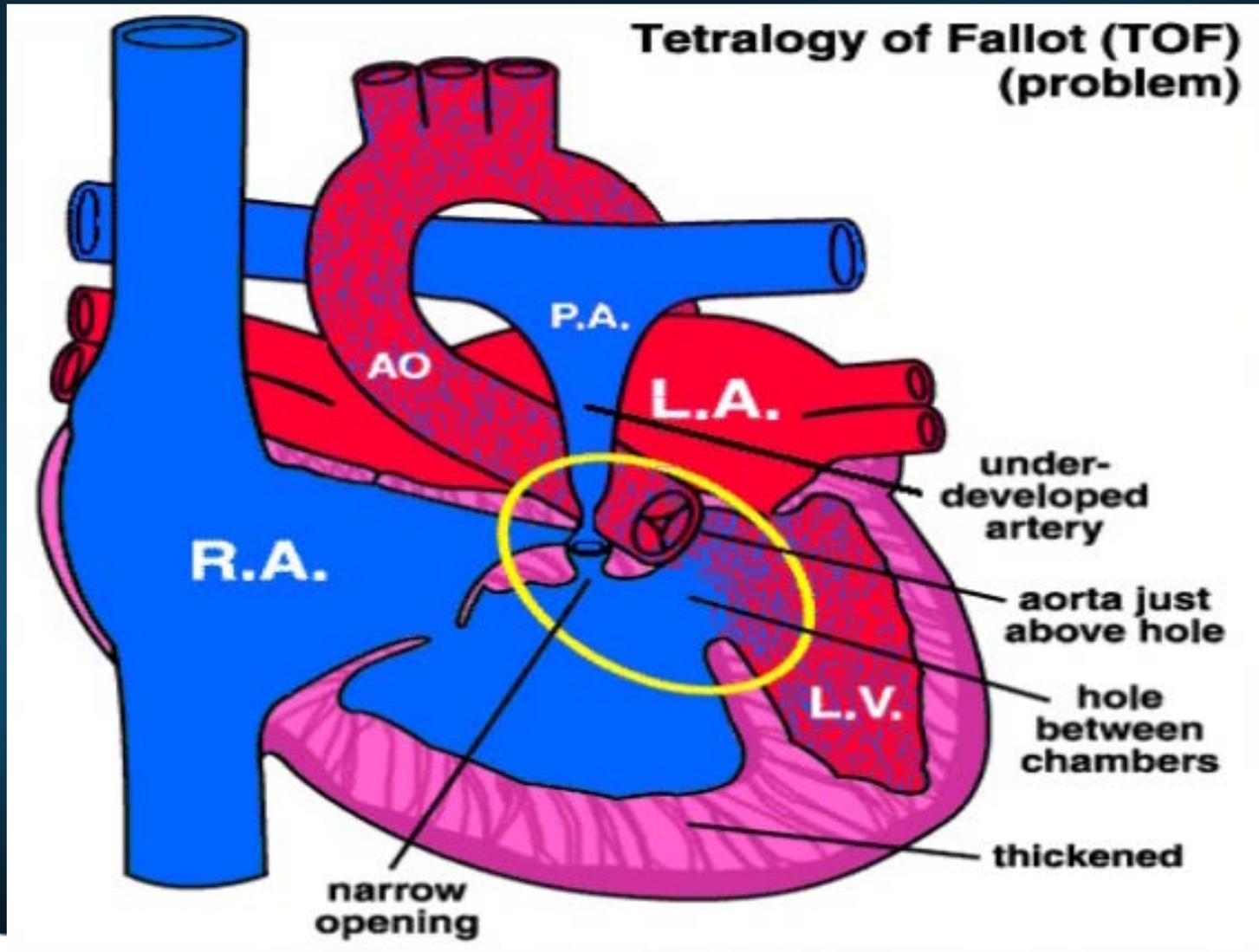


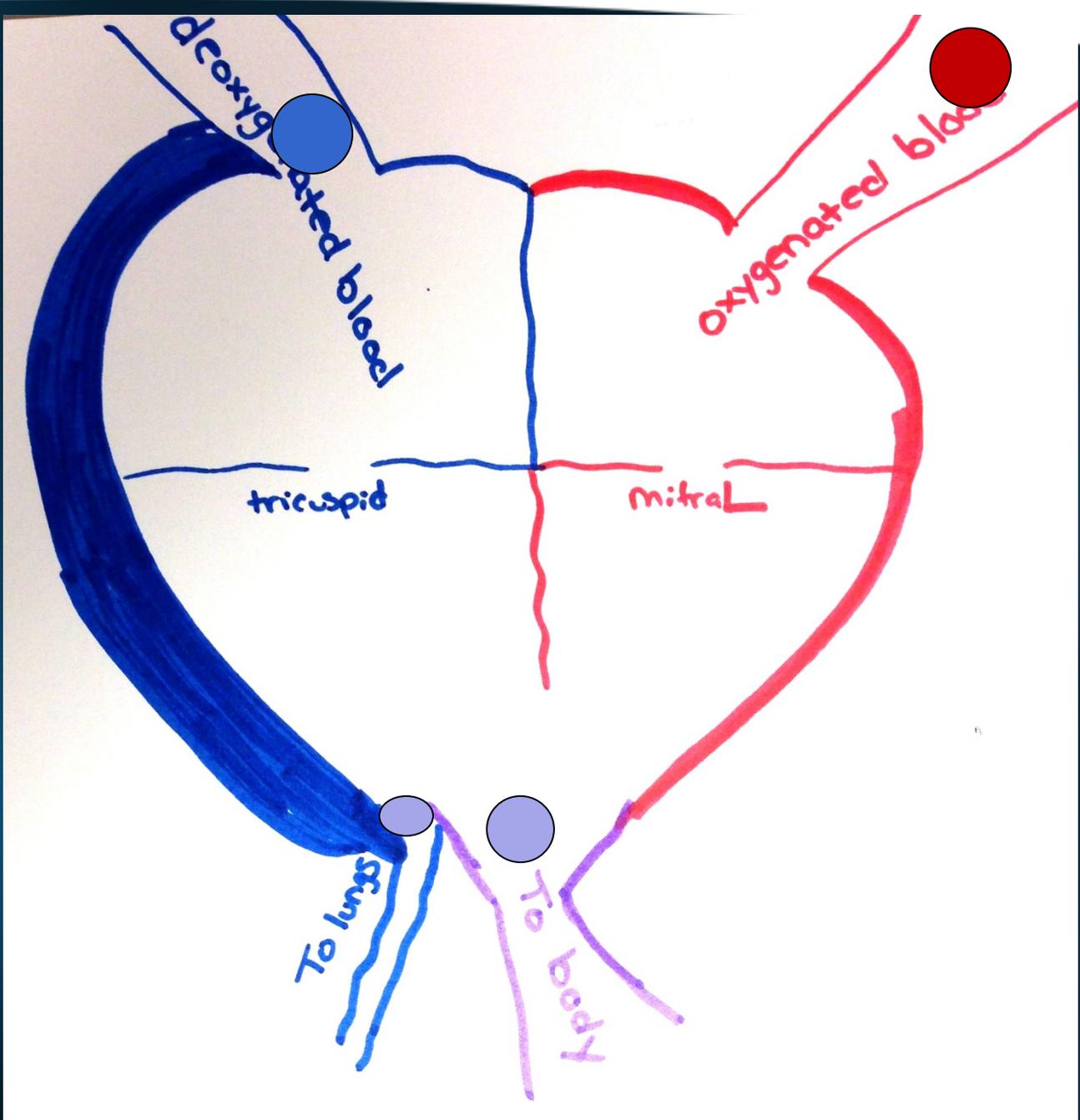
Tricuspid Atresia

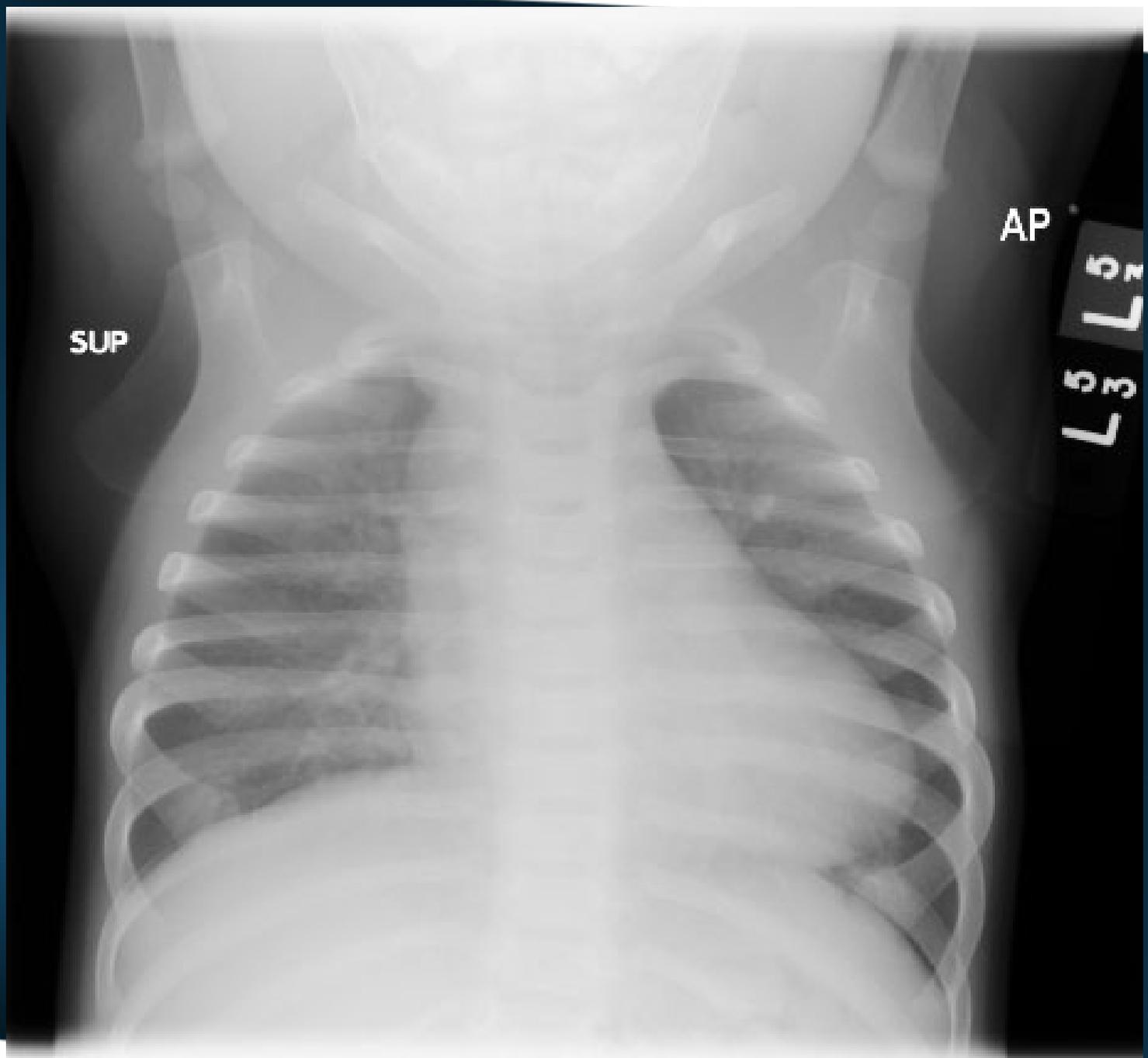




Tetraology of Fallot







Hypercyanotic Spells



Hypercyanotic Spells

Pulmonary stenosis worsens when child is stressed

Crying causes tachycardia

Right to Left shunt worsens with vasodilation:

fever

bowel movement

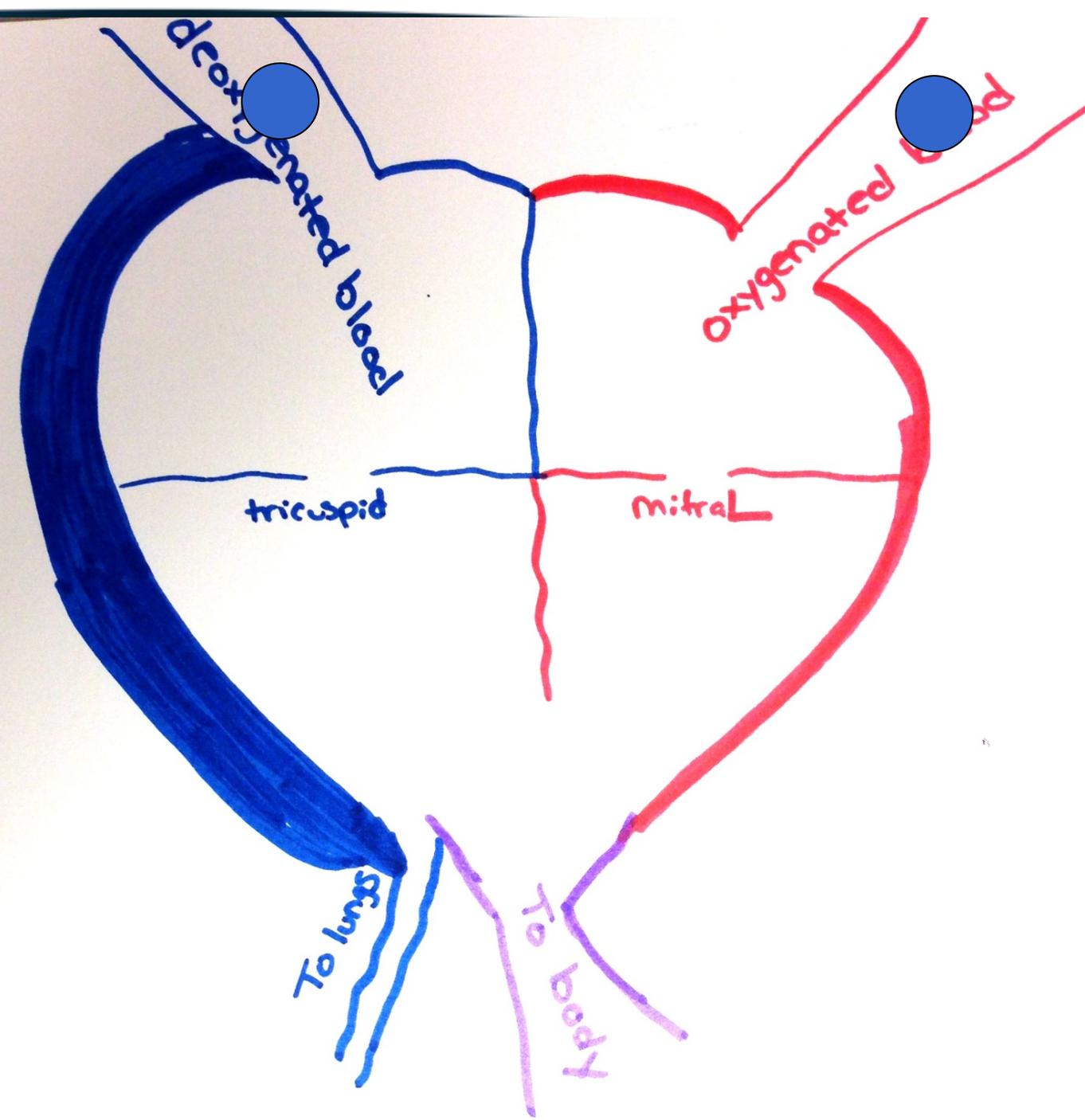
↓ blood flow to lungs, even worse than normal

↓ Systemic Vascular Resistance/ afterload (↓ pressure on L)

↑ Pressure on the Right = ↑ right-to-left shunt/mixed blood

Can be fatal





Treatment of the Hypercyanotic Spell

Knee to Chest Position

Helps calm child and \uparrow SVR/Pressure on L

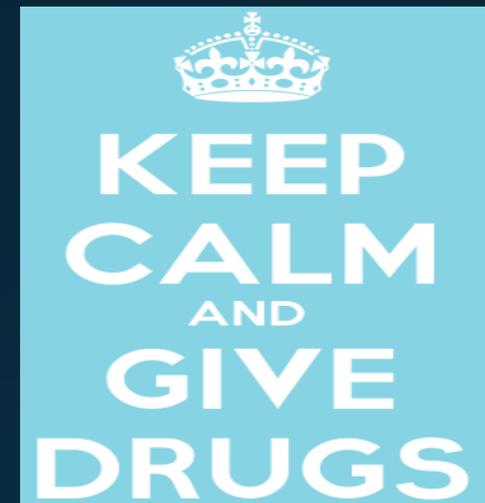
Oxygen??

Can decrease pulmonary pressure, increasing blood flow to lungs. (If there is any blood getting to lungs)

**Can also upset baby, worsening problem!!

Morphine? Other agents?

KEEP THE BABY HAPPY!!!!



Tetraology of Fallot Repair

Preferred to fix all at once around 6 months of age

If severe, may have BT shunt placed as newborn (manmade PDA) before sent home.

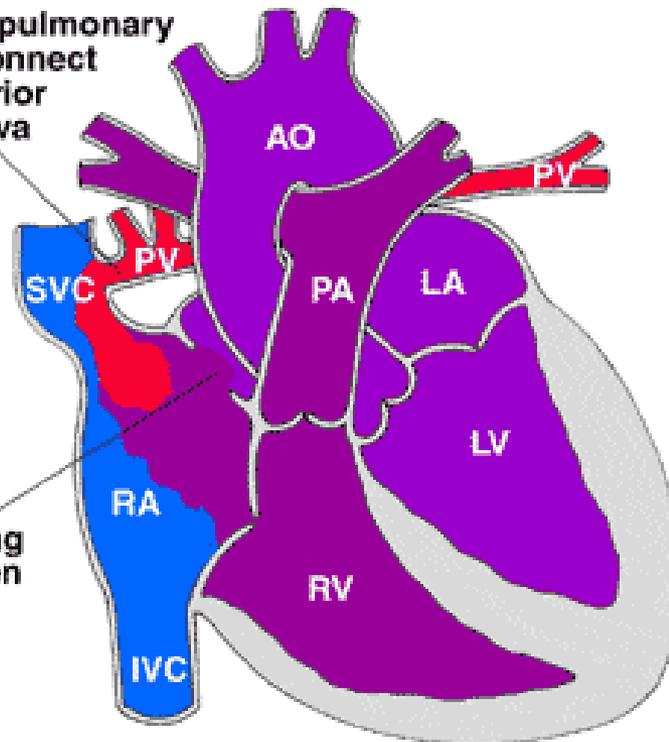


Total Anomalous Pulmonary Venous Return

Total Anomalous Pulmonary Venous Return

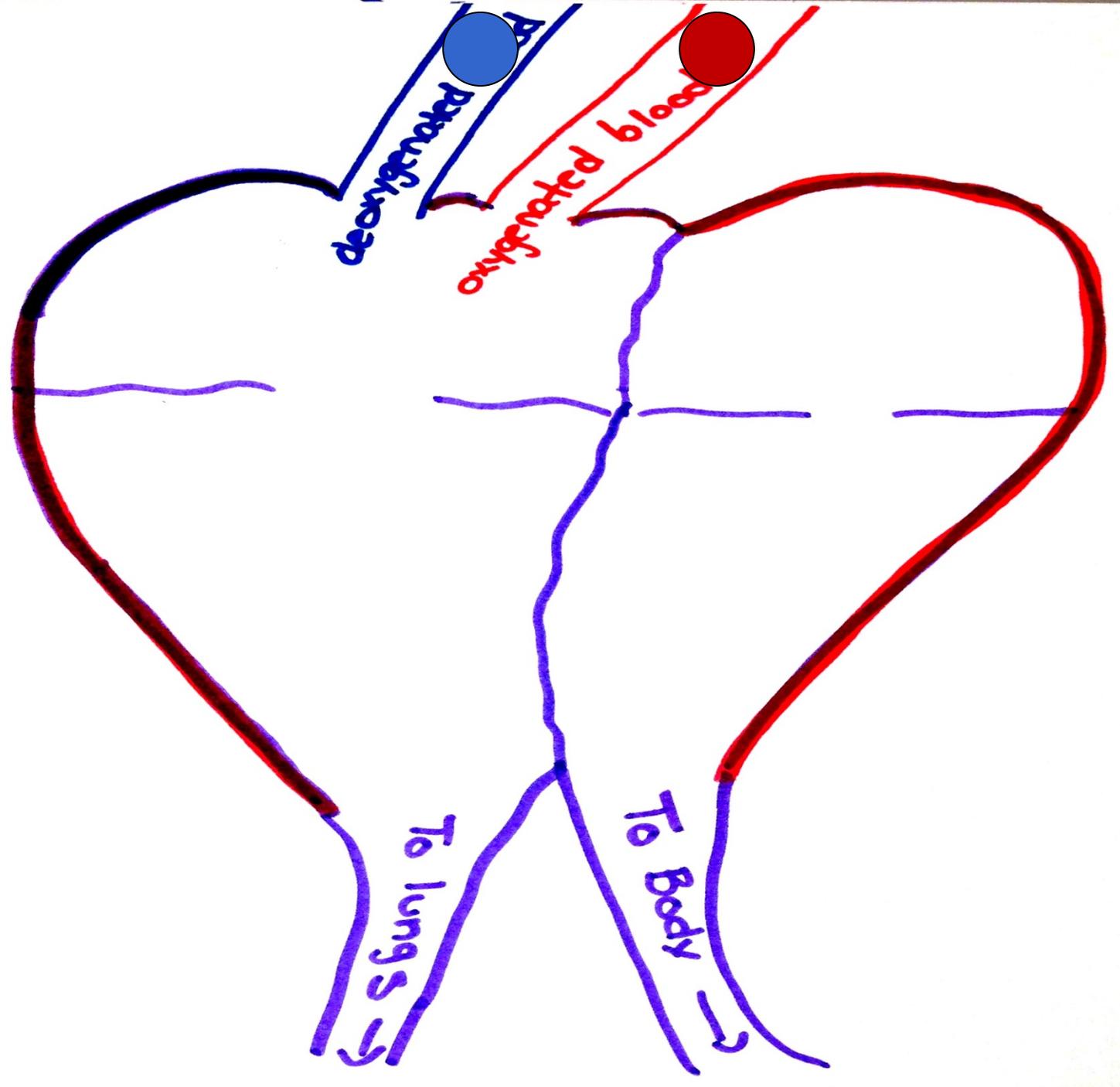
All four pulmonary veins connect to superior vena cava

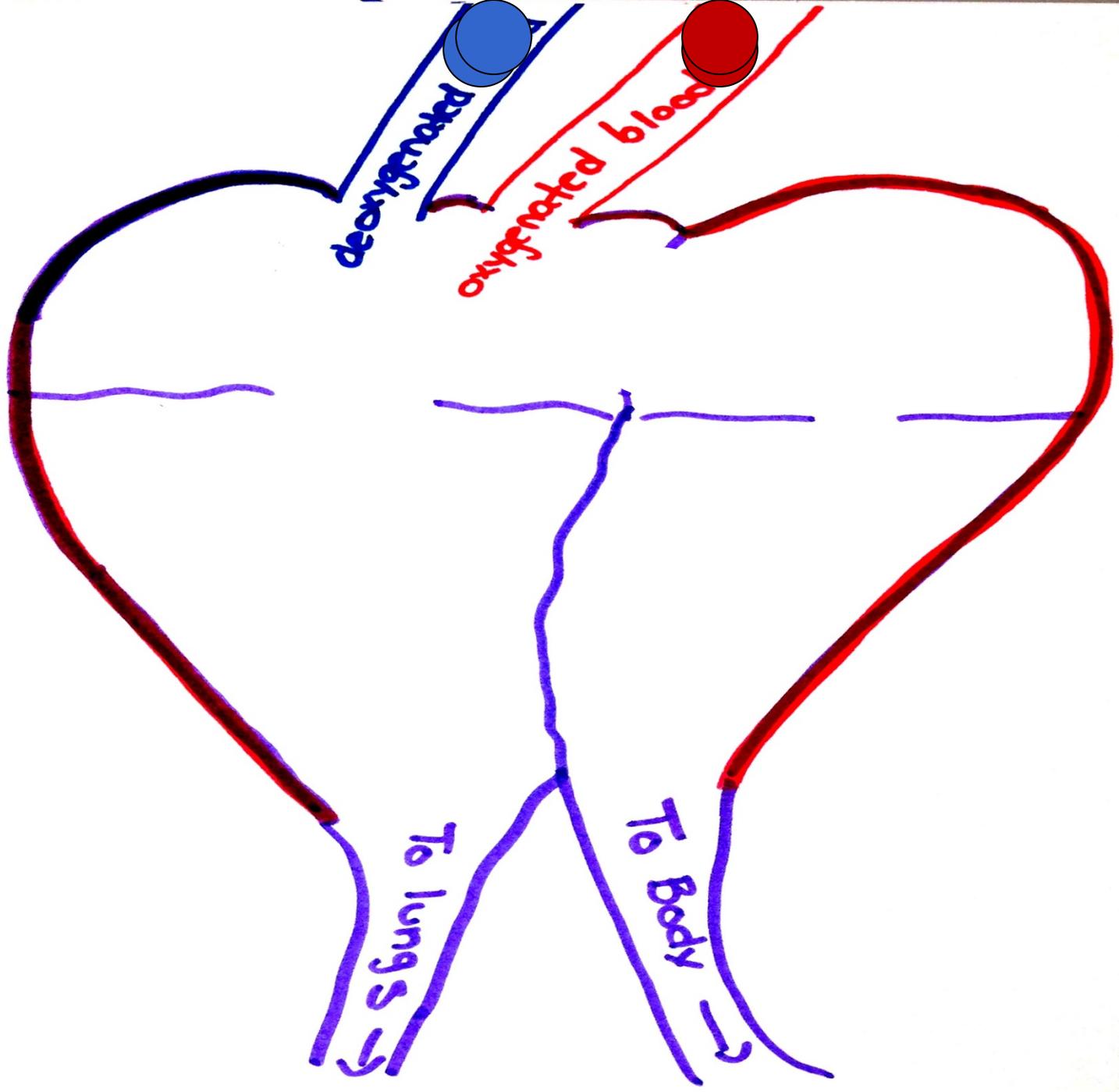
Opening between atria



-  Oxygen-rich blood
-  Oxygen poor blood
-  Mixed blood

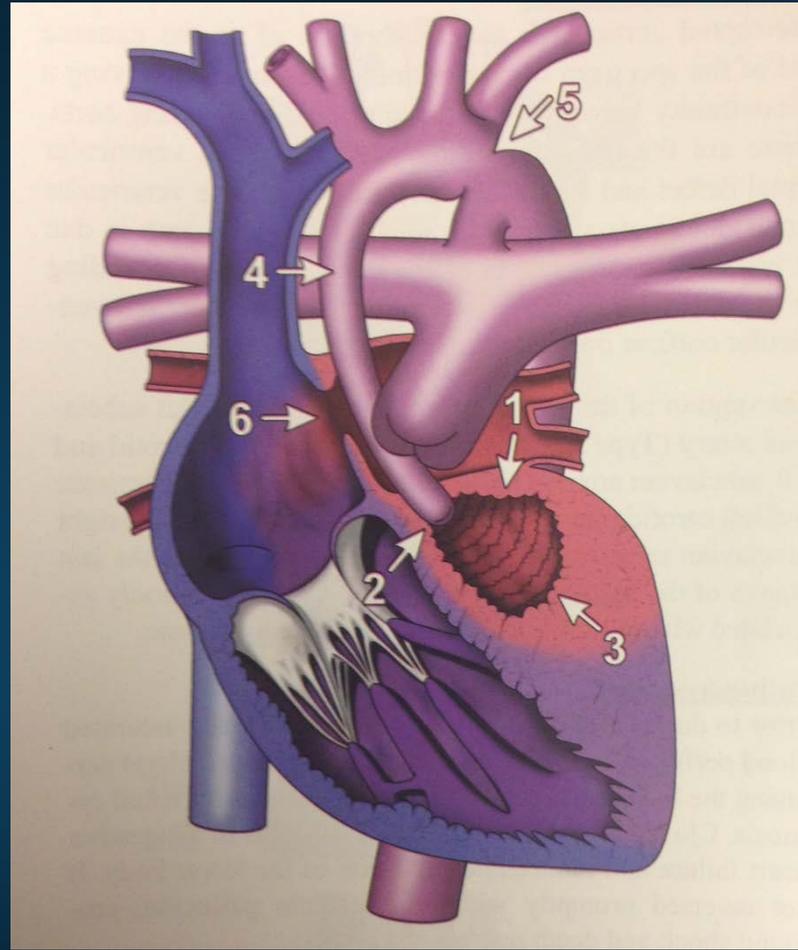
AO = aorta
PA = pulmonary artery
LA = left atrium
RA = right atrium
LV = left ventricle
RV = right ventricle





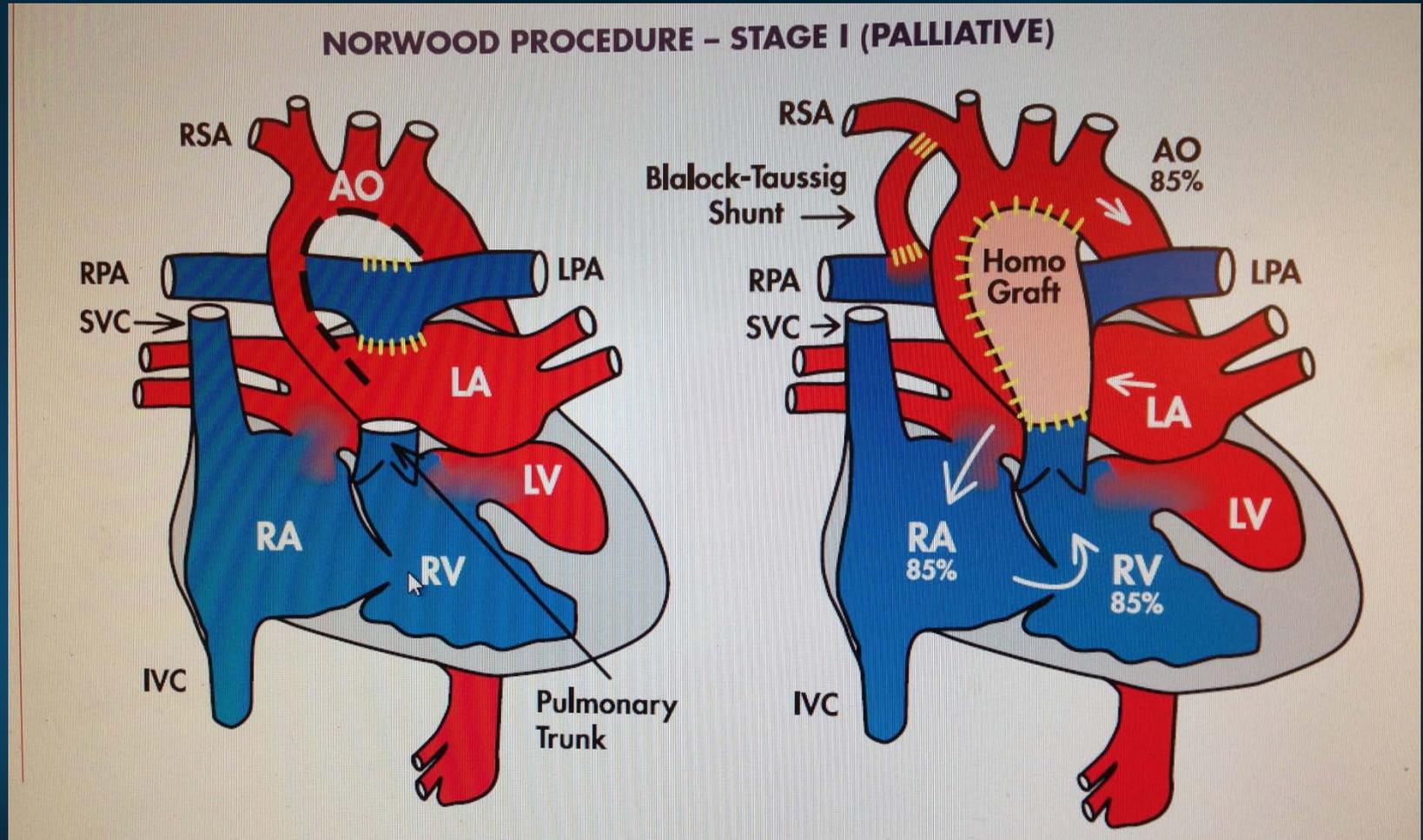


Hypoplastic Left Heart



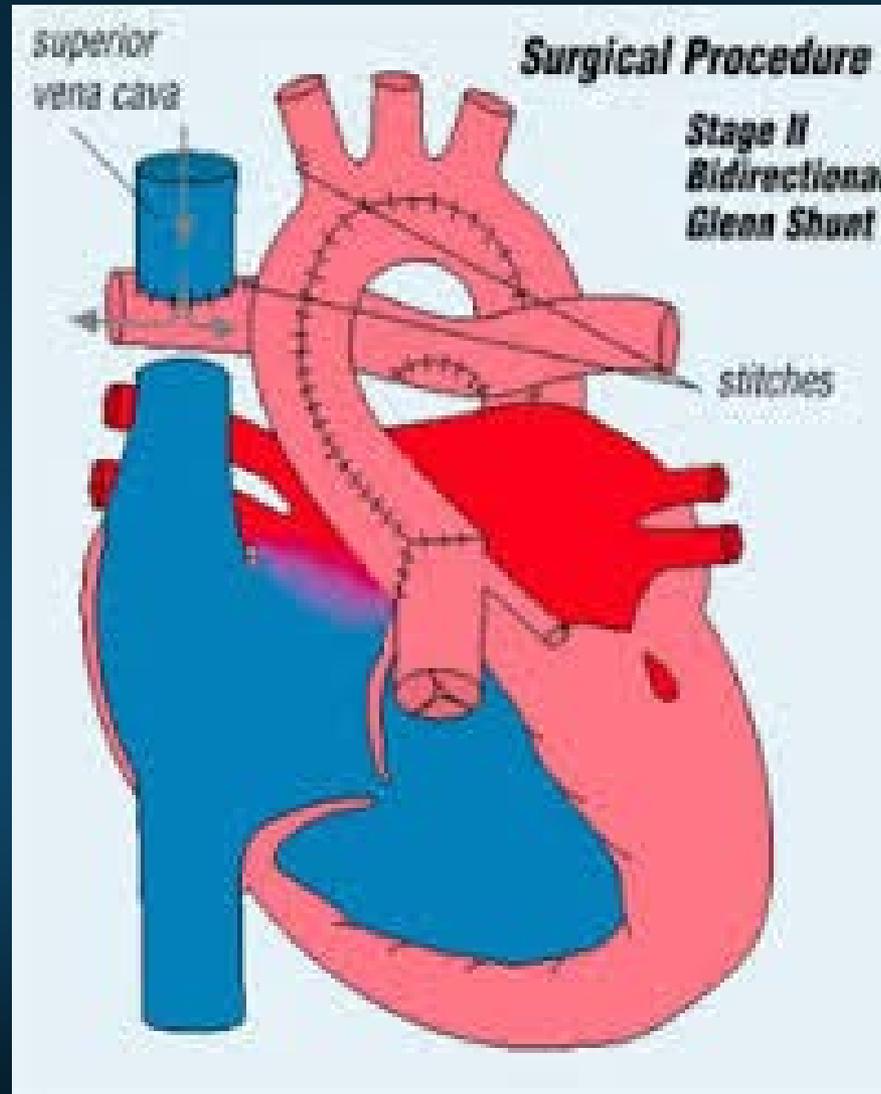
1. Absent or small mitral valve
2. Small ascending aorta
3. Hypoplastic LV
4. Hypoplastic ascending aorta
5. Coarctation of Aorta
6. Atrial Septal Defect

Phase I Repair: Norwood



First few days of life

Phase II Repair: Glenn or Hemi-Fontan

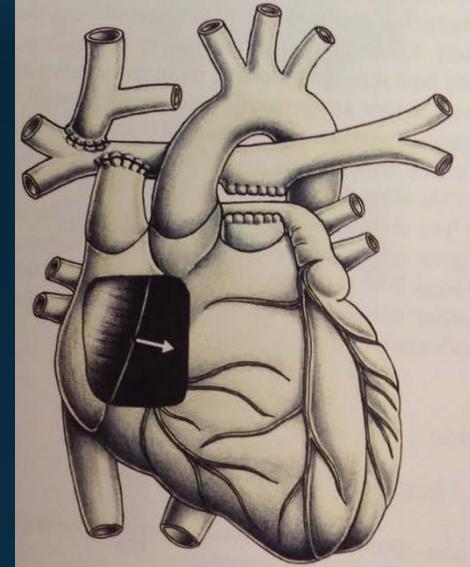
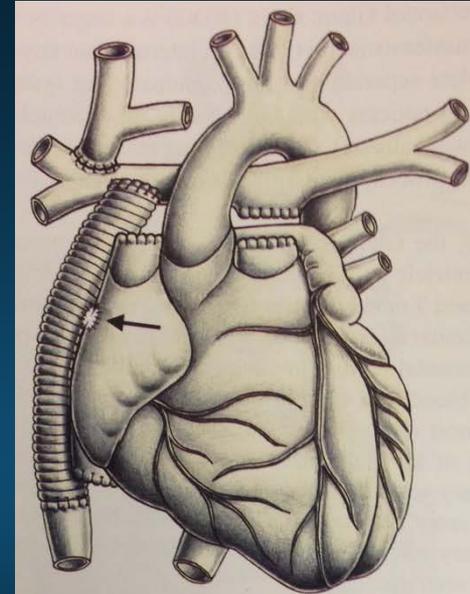
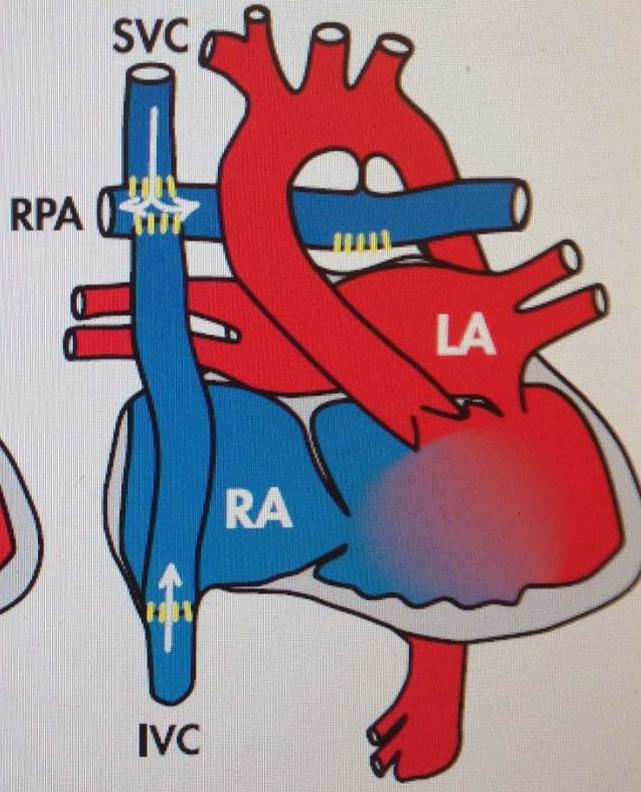
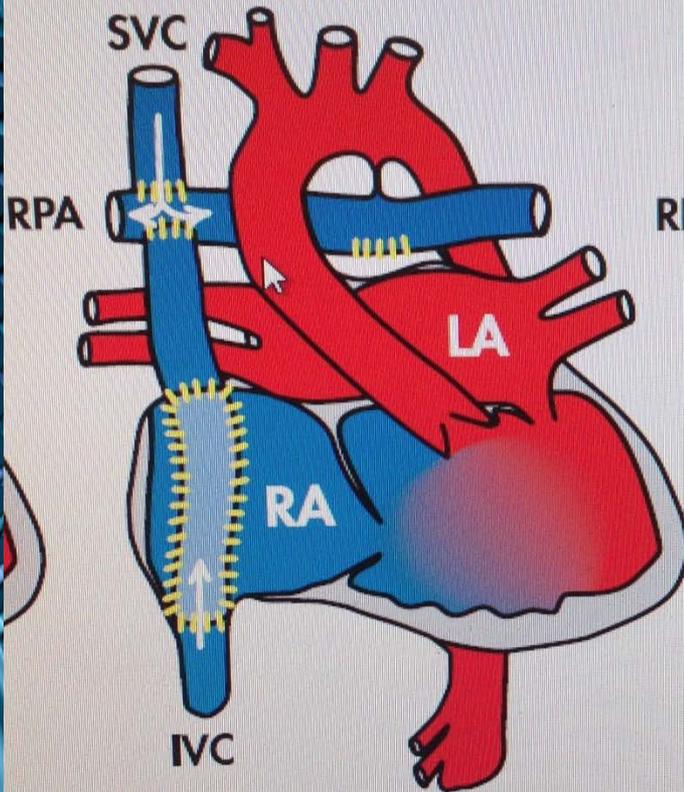


Age 2-9 months

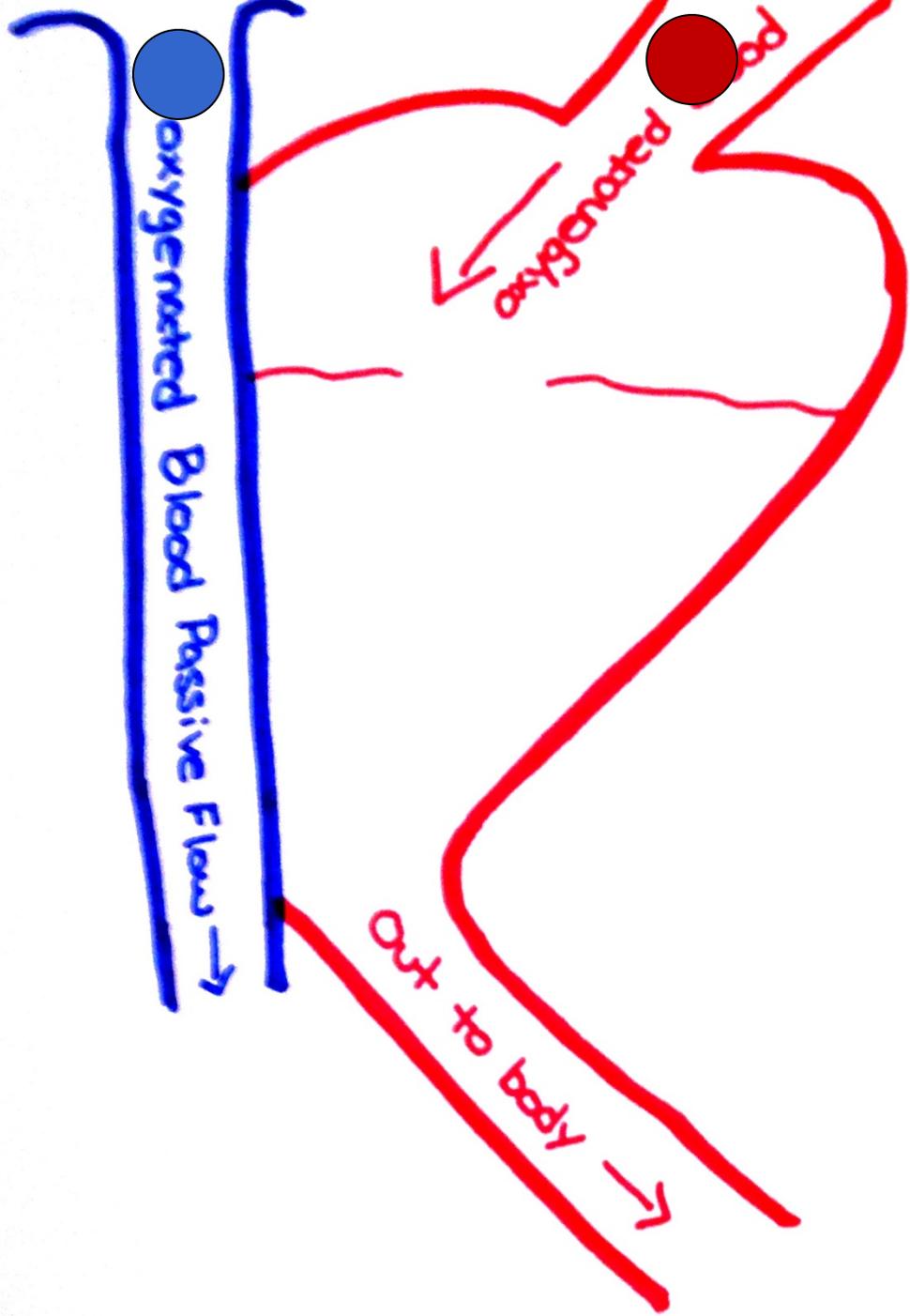
Phase III Fenestrated Fontan

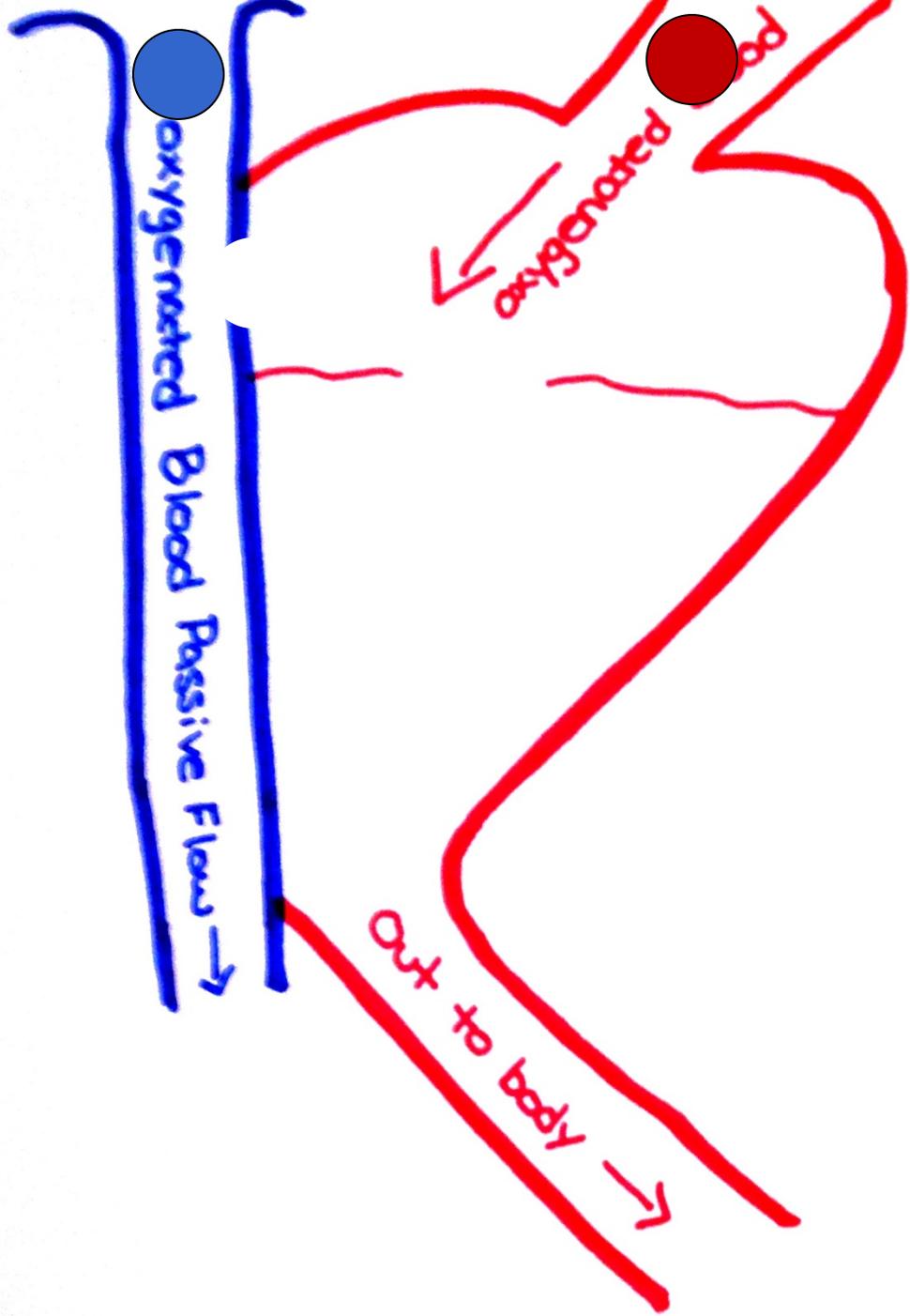
Lateral Tunnel Fontan

Extracardiac Fontan



Age 2-3 years







Babies Post-Surgery

“My baby had a fountain surgery...”

- Prone to arrhythmias, typically tachyarrhythmias
 - Ask about home medications, many on digoxin or amiodarone or a beta blocker
 - Try other ways to get heartrate down including PALS guidelines, fluid bolus 10mL/kg, rule out fever, keep happy
- Passive flow to RV means very pre-load, fluid dependent
 - Respond well to fluid boluses

Other Times It's OK for the Baby to be Blue

- Acrocyanosis



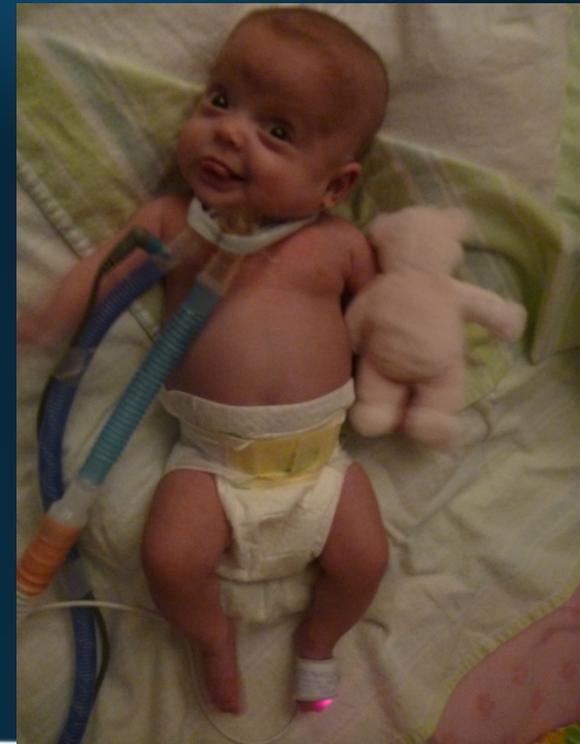
Other Times it's OK to be Blue

- Are they cold?
 - *Cutis marmorata*



BronchoPulmonary Dysplasia (BPD)

- NICU grads
- Characterized by severe desaturations with irritation progressing to bradycardia and potentially cardiac arrest



Summary: Blue Baby Treatment



Review: The 5 T's of Cyanotic Heart Defects





Key Points to Remember

- Decreasing heart rate and oxygen demand can make a big difference!!
 - Calm baby down—with parents and/or medications
 - Sedatives
 - Anti-fever medications
 - Fluid bolus cautiously 10cc/kg to maximize preload/fill the tank
- Oxygen is not always helpful
 - Can irritate baby more (consider blow-by)
 - Takes blood away from systemic circulation/worsens pulmonary congestion
 - Keep baby at baseline saturations, if unknown, around 70 - 85%

If You Have to Intubate



- Prepare for acute decompensation
 - Know your epi dose & have drawn up
 - Fluid bolus ready to go
 - Premedicate with Atropine
 - Keep vent pressures low as possible for effect (PIP and PEEP)
 - If equipped with oxygen blender, try to keep FiO₂ as low as possible to goal saturation of ~70 - 85% or baseline (PALS recommends 80%)
- If patient arrests, PETCO₂ will not be reliable indicator of CPR quality because pulmonary bloodflow depends on other factors



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- Scenario 1: You are dispatched to a 2-month-old with “hemorrhage.” You arrive to find a baby in no apparent distress. Mom reports the baby is on daily aspirin for a heart condition and today has had bloody diapers when he poops. She called the cardiologist, who recommended she go to the ED. However, most concerning to you is the fact that the baby appears blue. Lungs are clear and the baby has no increased work of breathing. Your pulse ox shows a sat of 85% with excellent pleth/waveform. Mom says “that number” is always low. What do you do?

- 
- Scenario 2: You are dispatched to a 1-week-old infant with “respiratory distress.” You arrive to find a lethargic, limp baby who looks grey. Oral mucosa is tinged blue. You notice increased work of breathing, tachypnea, and a very fast heartrate. Cap refill is 5-6 seconds, skin is clammy and wet. No recent ill contacts. Baby was a normal delivery, born at a birthing center 6 days ago and seemed normal until this morning. Lungs are clear to your auscultation. Pulse oximetry is not reading.
 - What do you think is going on?
 - How are you going to treat this infant?

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- Scenario 3: You are dispatched to a 2-month-old baby with “respiratory distress.” You arrive to find an alert but tired looking baby with tachypnea and some increased work of breathing. Under the baby’s eyes appear swollen, and he looks very skinny. Cap refill is 4-5 seconds, no wet diapers today. PMH from mom is that he has always gotten sweaty and “pants like a puppy” when eating from a bottle. Heartrate feels very fast, and lungs have crackles bilaterally.
 - What do you think is going on with this patient?
 - How will you treat him?



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