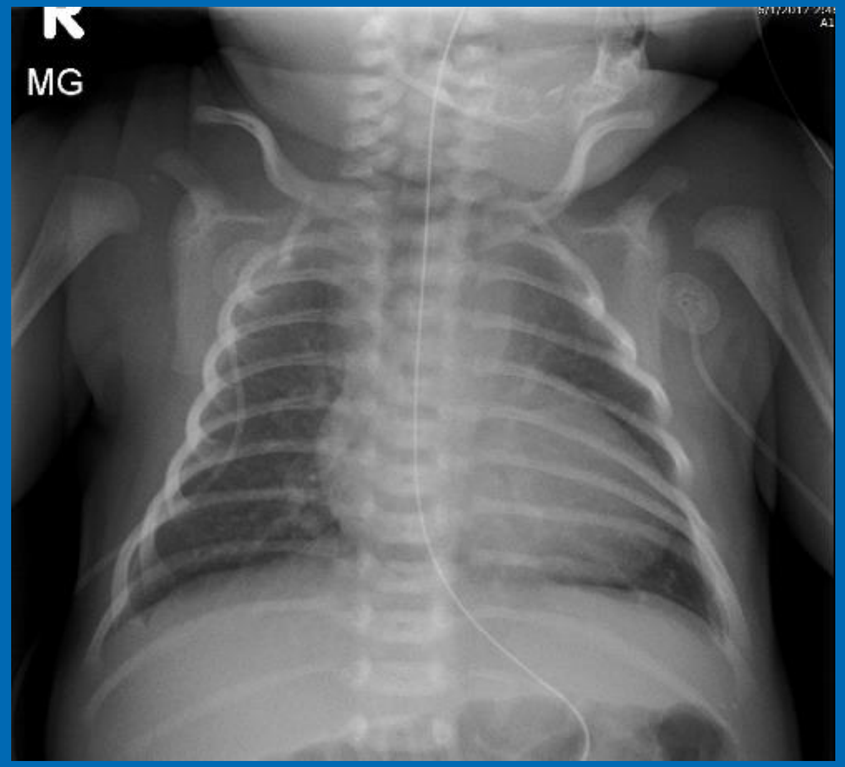
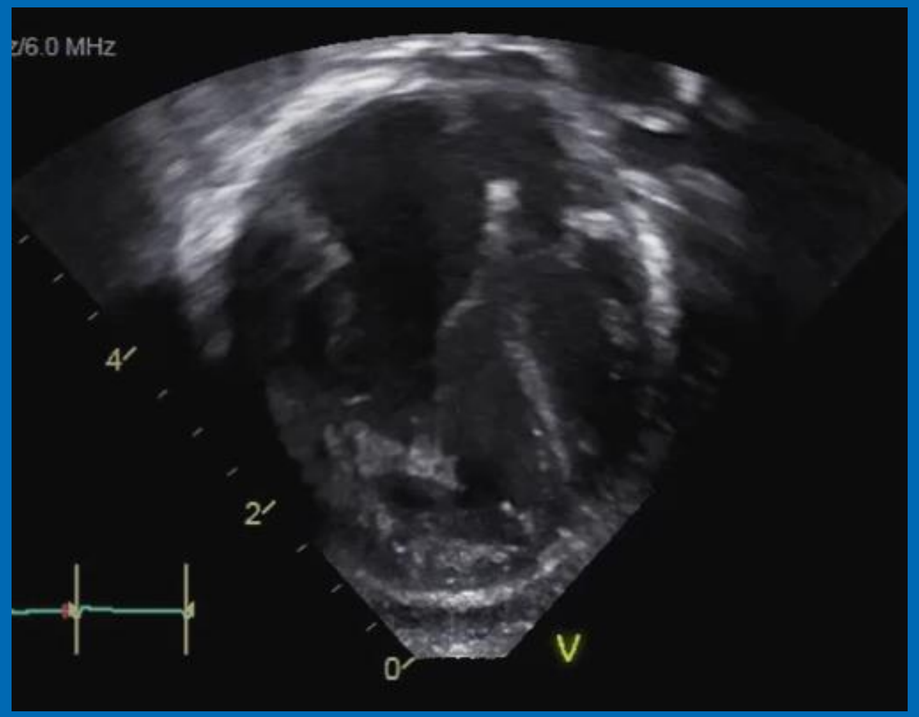
CYANOTIC HEART DISEASE





Michael V. Di Maria, MD
michael.dimaria@childrenscolorado.org
Office: (720) 777-2940

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Conflicts of Interest: None

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LEARNING OBJECTIVES

- **Review definition and differential diagnosis of neonatal cyanosis**
- **Discuss Initial evaluation of cyanotic heart disease**
- **Describe when & how to start Prostaglandin**

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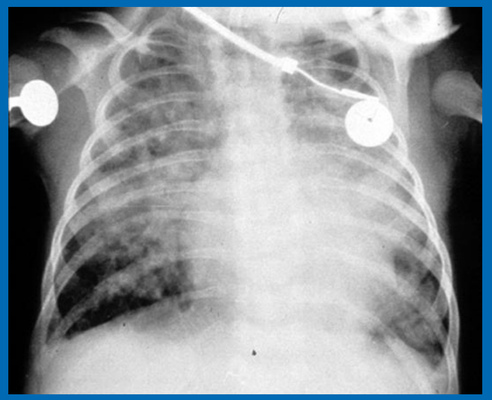
5 CAUSES OF HYPOXIA

(If you're a pulmonologist)

Low FiO2

Hypoventilation

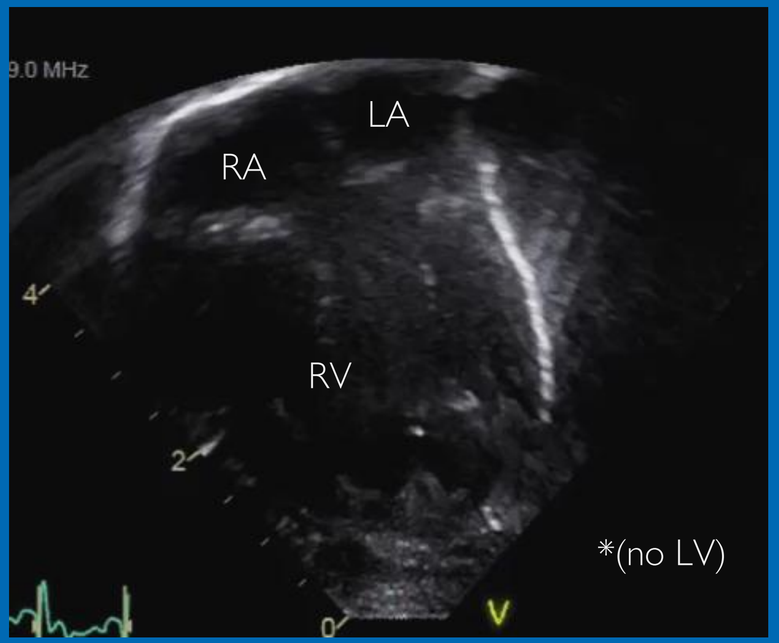
A-a gradient



Intrapulmonary shunt

Pulmonary Capillary

Intracardiac Shunt



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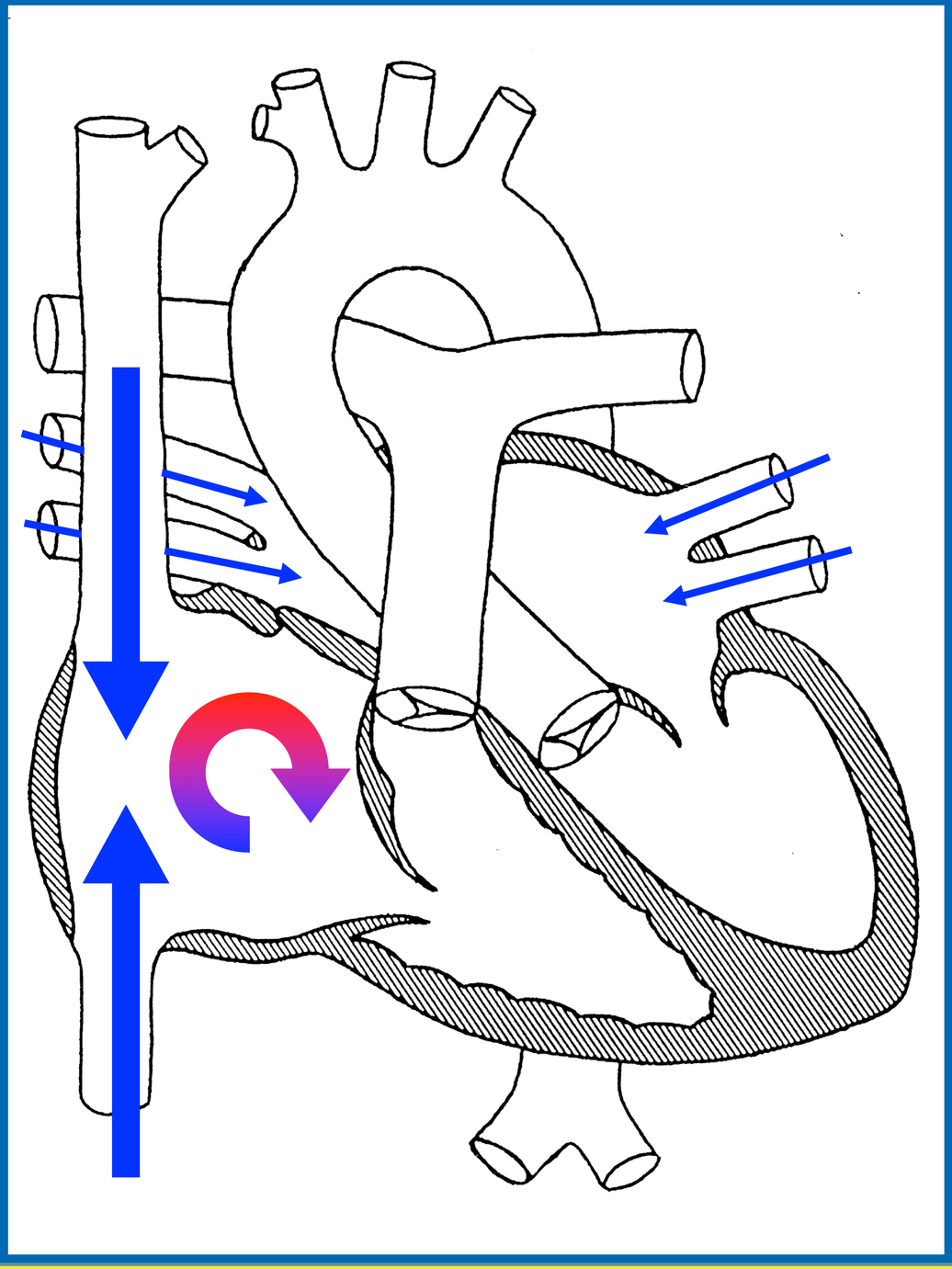


3 CAUSES OF HYPOXIA

(If you're a cardiologist)

1. Pulmonary vein blood too blue

- A-a gradient
- Hypoventilation
- Intrapulmonary shunt

2. Systemic vein blood too blue

- Sepsis
- Low cardiac output
- Left-sided obstructive lesions

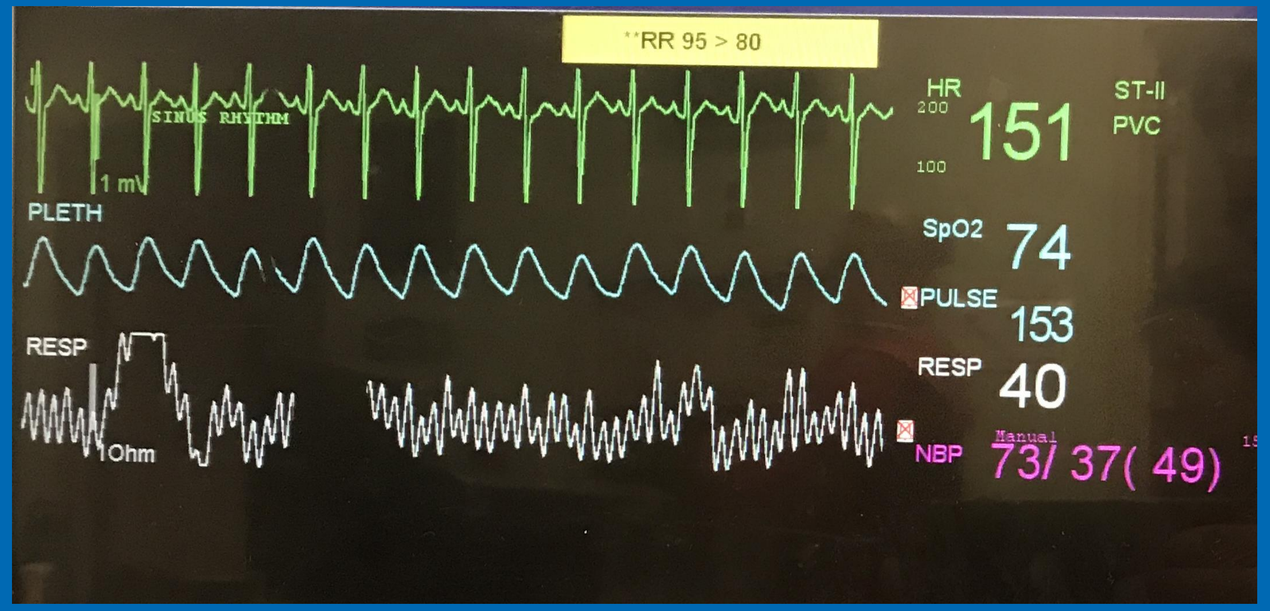
3. Pulmonary and Systemic Venous Mixing

- "Cyanotic congenital heart disease"

Mullins, C., & Mayer, D. (1991). *Congenital Heart Disease: A Diagrammatic Atlas*. Wiley-Liss.

CASE: NEWBORN WITH CYANOSIS

- Term male, uncomplicated pregnancy and delivery.
- Baby appeared vigorous but “dusky”, no improvement in saturations with CPAP.
- Initial saturations 74% on RA in the RUE
- Birth weight 3.2kg
- Normal S1, loud S2, no murmur



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THE BLUES

Foundational concepts:

1. What is cyanosis?
2. Acrocyanosis vs. “central” cyanosis?

You need 3-5g/dL deoxyhemoglobin to look blue!

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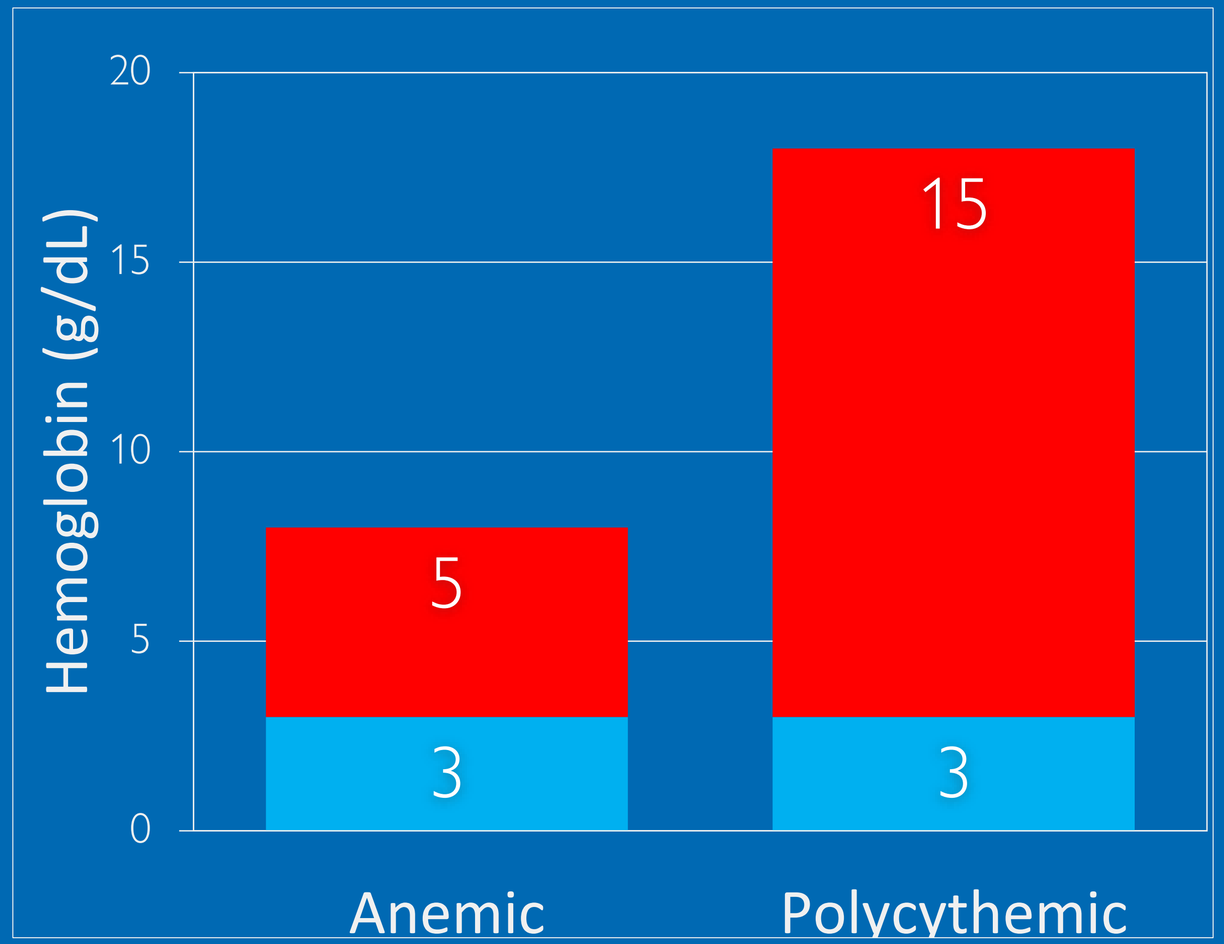
TOTAL HEMOGLOBIN MATTERS



Key Concept

- If your total hemoglobin is 8: 3g/dL is almost half the hemoglobin
- If your total hemoglobin is 18: 3g/dL is a small fraction (<20%).

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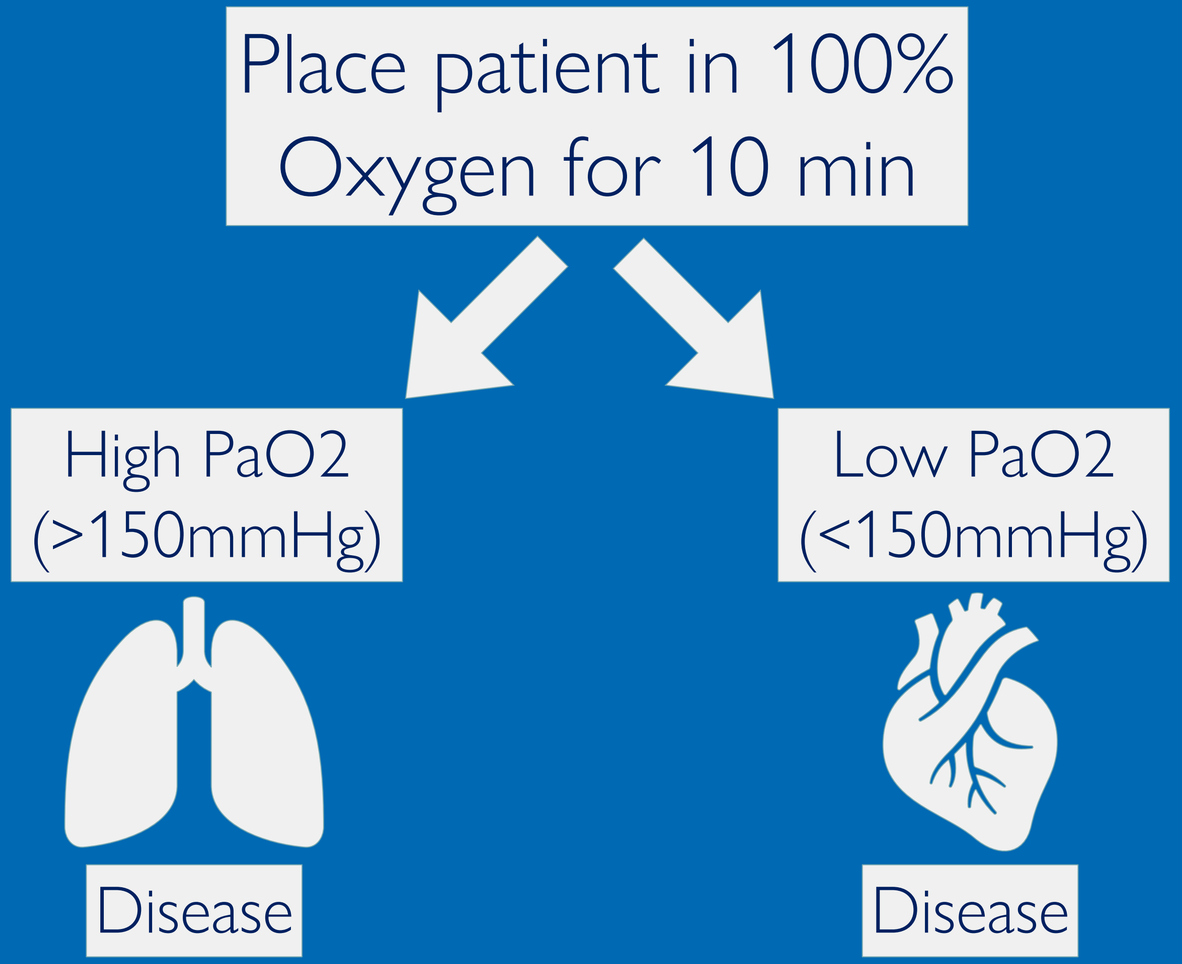
IS IT THE HEART OR THE LUNGS?

The Hyperoxia Test

- By placing the patient in 100% oxygen, we can differentiate pulmonary vs. cardiac causes of cyanosis.
- Key Concept: oxygen can usually "fix" pulmonary causes of hypoxemia, but not intracardiac mixing.



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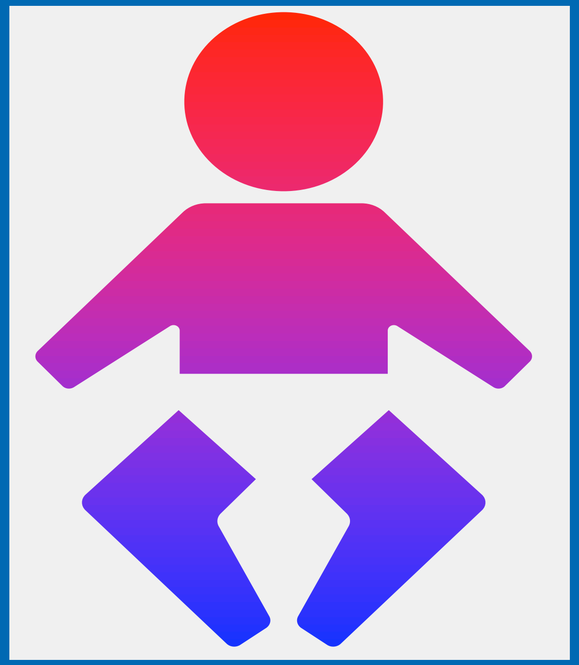
DIFFERENTIAL CYANOSIS

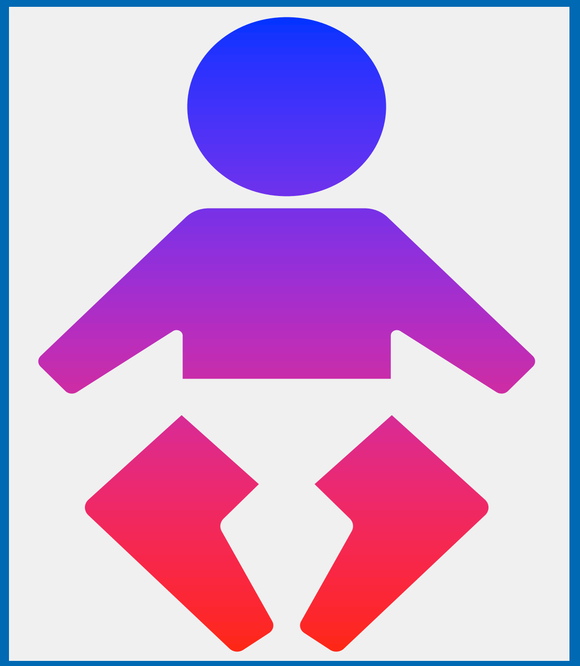
Head and arms: High



Abdomen and legs: Low

REVERSE DIFFERENTIAL

Head and arms: Low



Abdomen and legs: High

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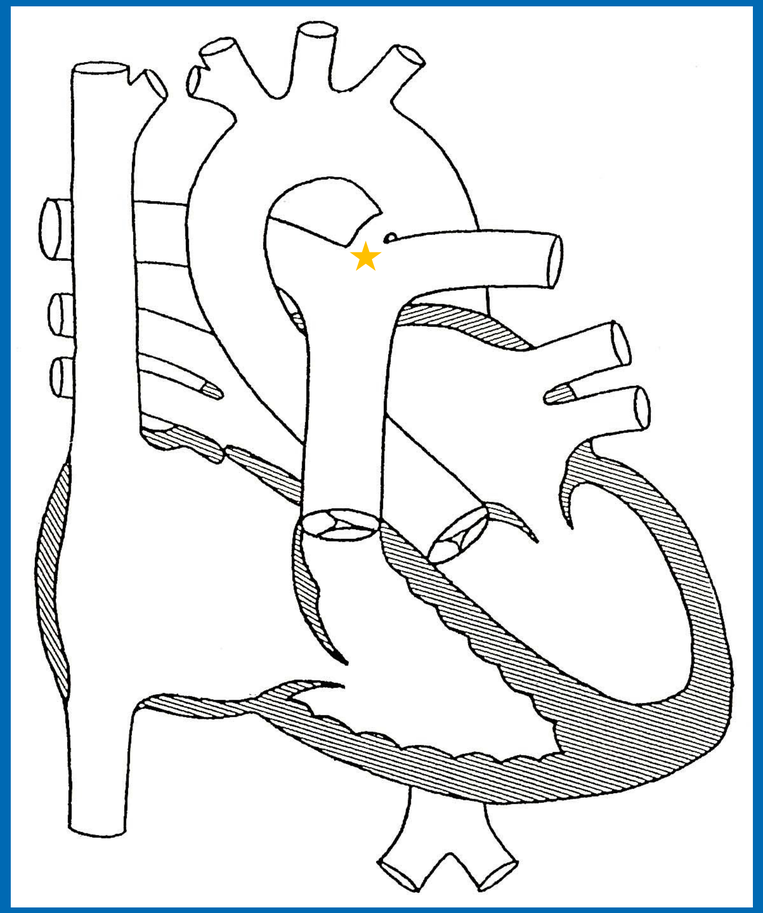
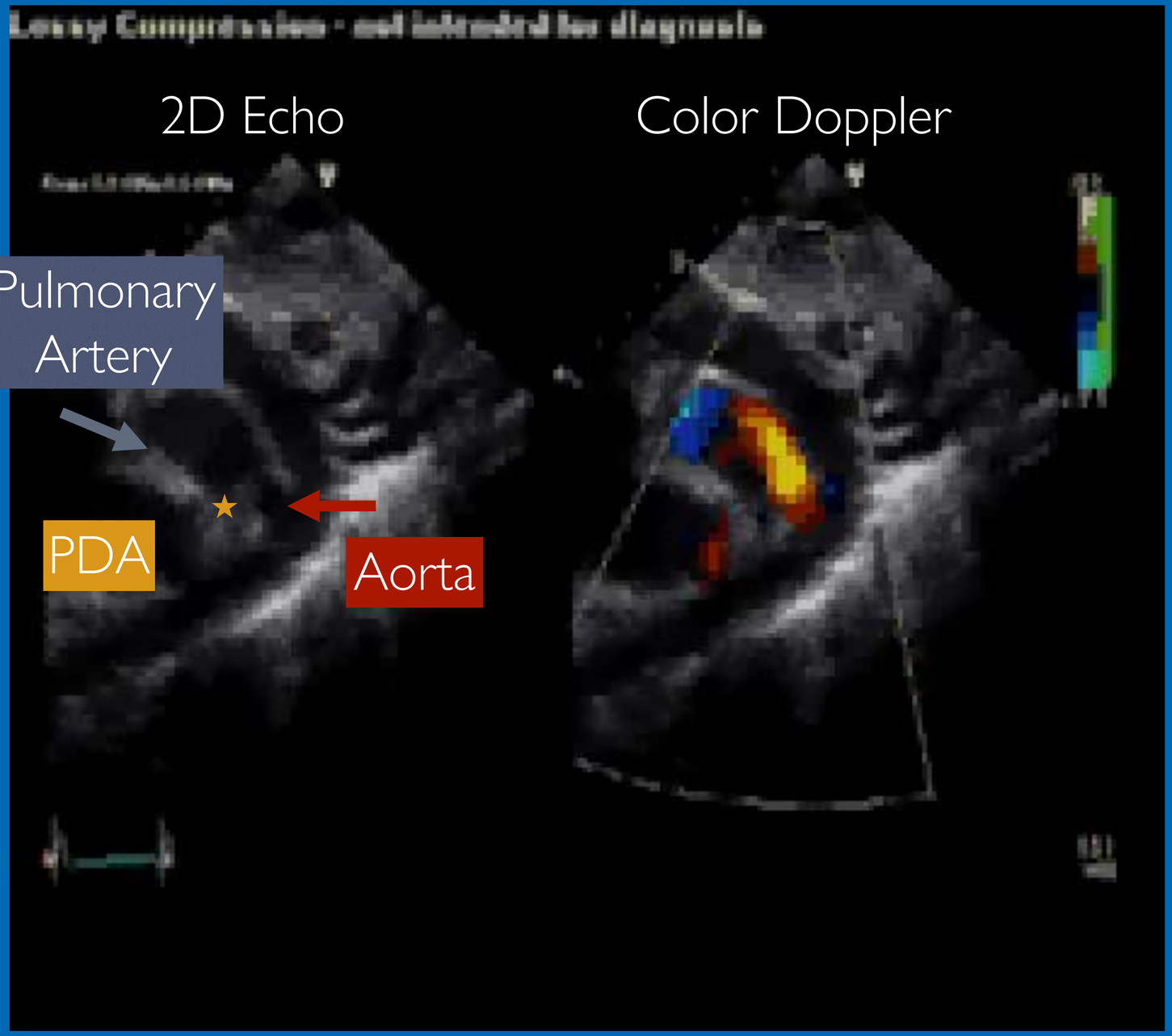
PATENT DUCTUS





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DIFFERENTIAL CYANOSIS

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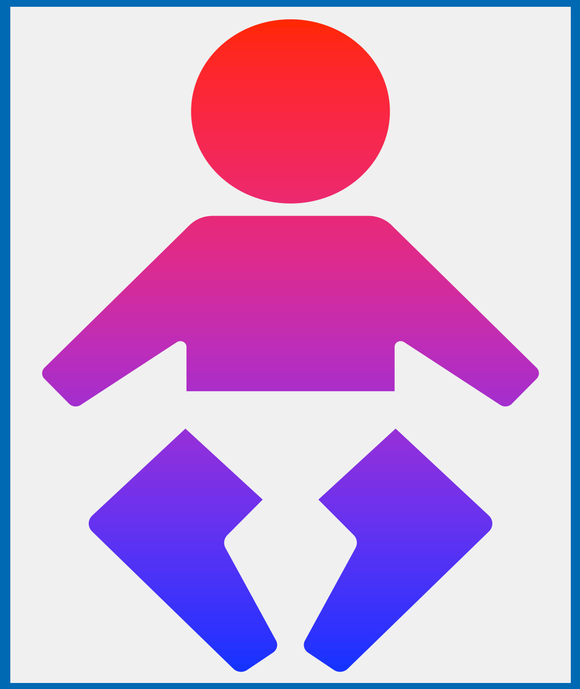
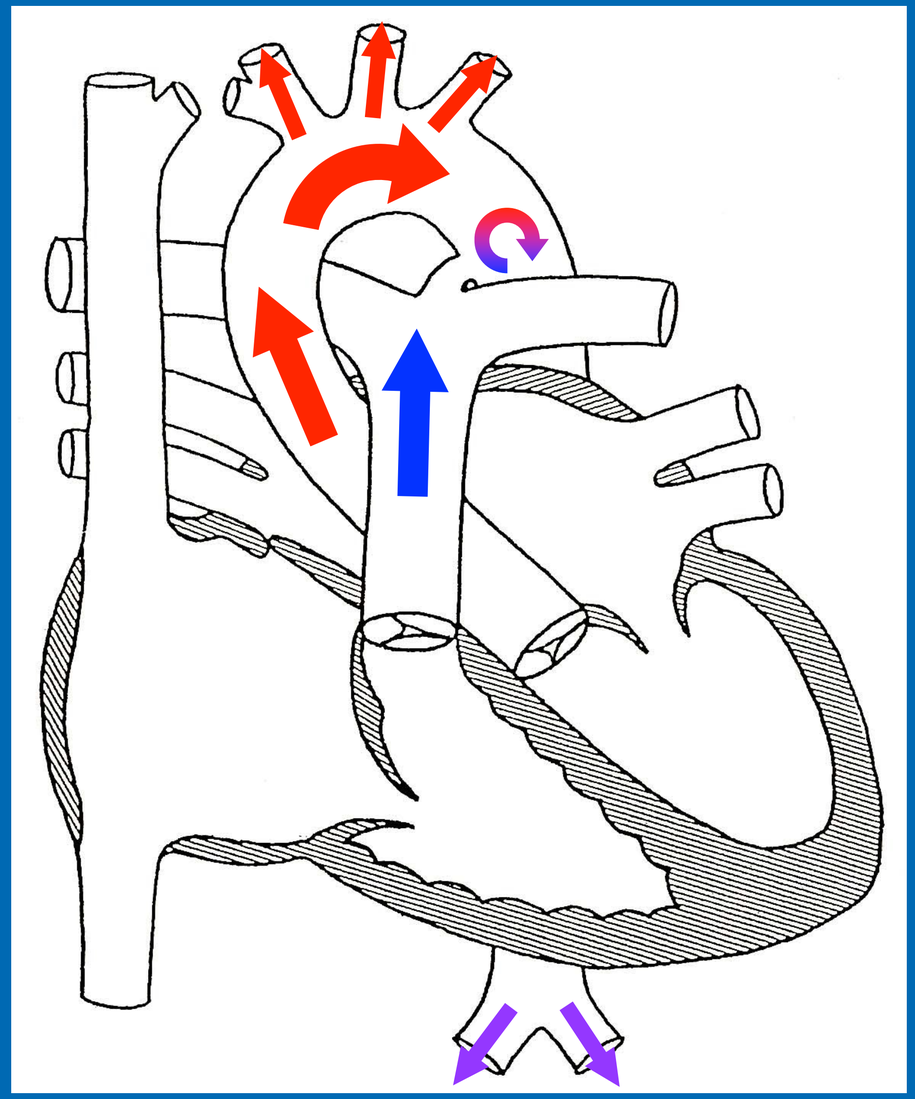


Abdomen and legs: Low



Key Concept:
BLUE blood from the PDA enters the descending aorta, making it desaturated

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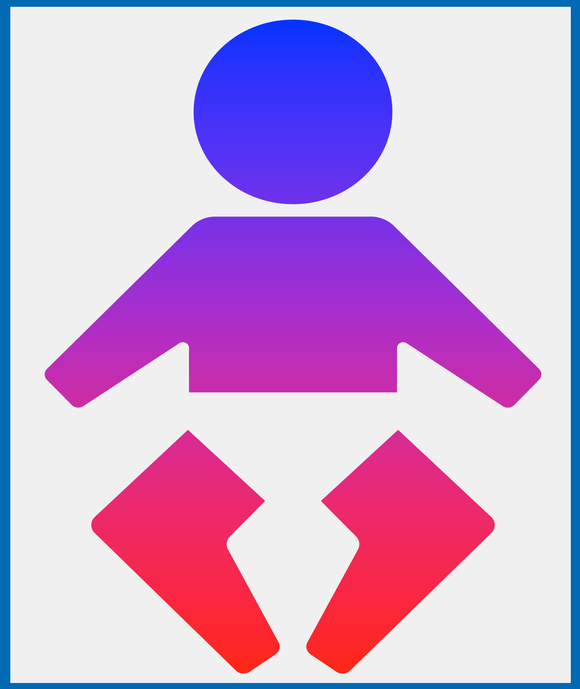
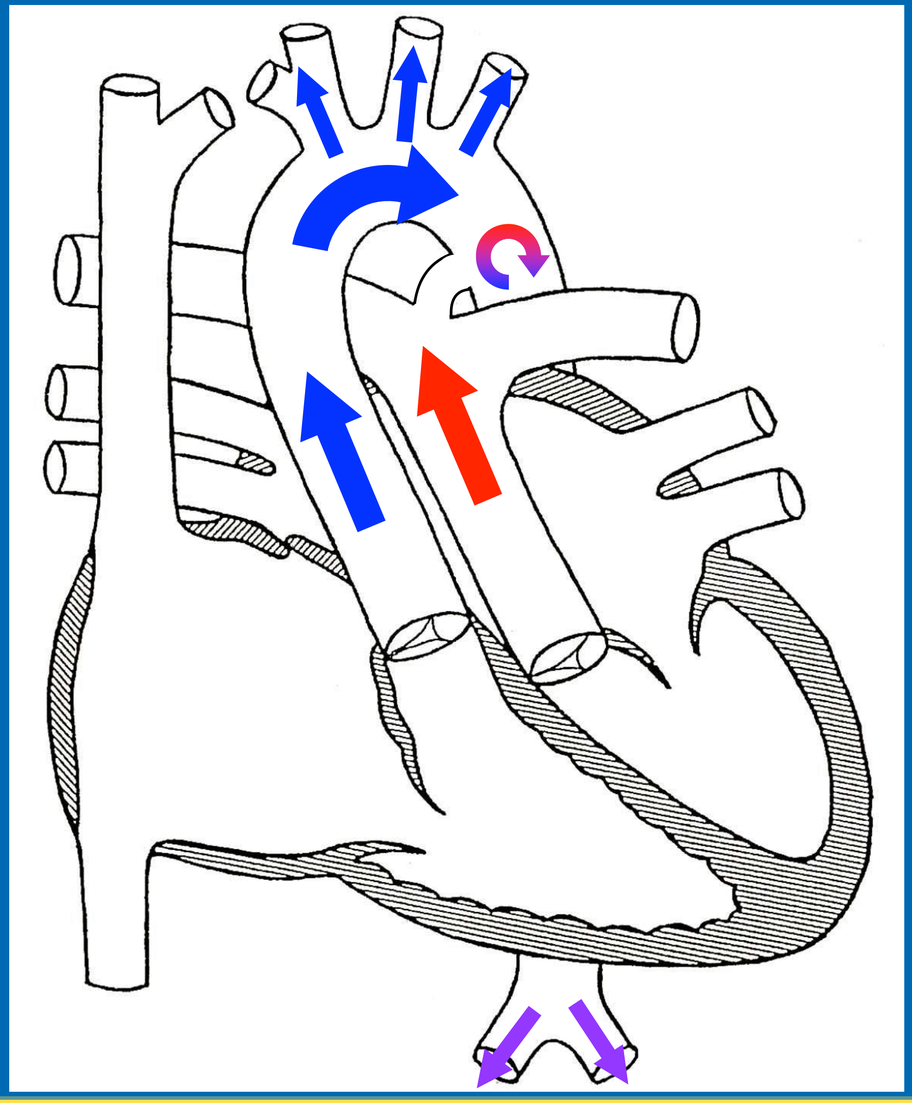
REVERSE DIFFERENTIAL CYANOSIS

Head and arms: Low



Abdomen and legs: High



Key Concept:
RED blood from the PDA enters the descending aorta, making it LESS desaturated

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Mullins, C., & Mayer, D. (1991). *Congenital Heart Disease: Diagrammatic Atlas*. Wiley-Liss.

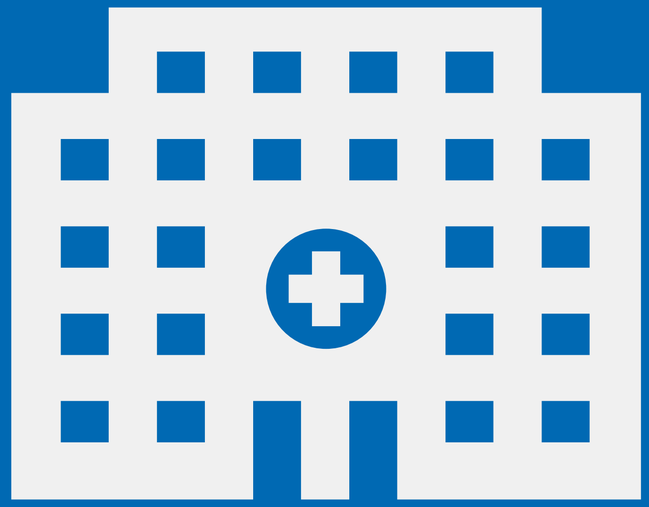
INITIAL EVALUATION

Physical Exam

Chest x-ray

ECG

Echo



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MURMURS

Intensity	Pitch	Quality	Timing	Radiation
1. Barely audible 2. Soft 3. Prominent 4. (+)Thrill 5. * 6. No stethoscope	Low (bell) vs High (diaphragm)	“Vibratory” “Harsh” “Blowing” “Machine-like”	Systolic Diastolic Continuous	Where is it loudest? Audible elsewhere?

“Grade 2/6, high-pitched, harsh systolic ejection murmur at the left-upper sternal border”

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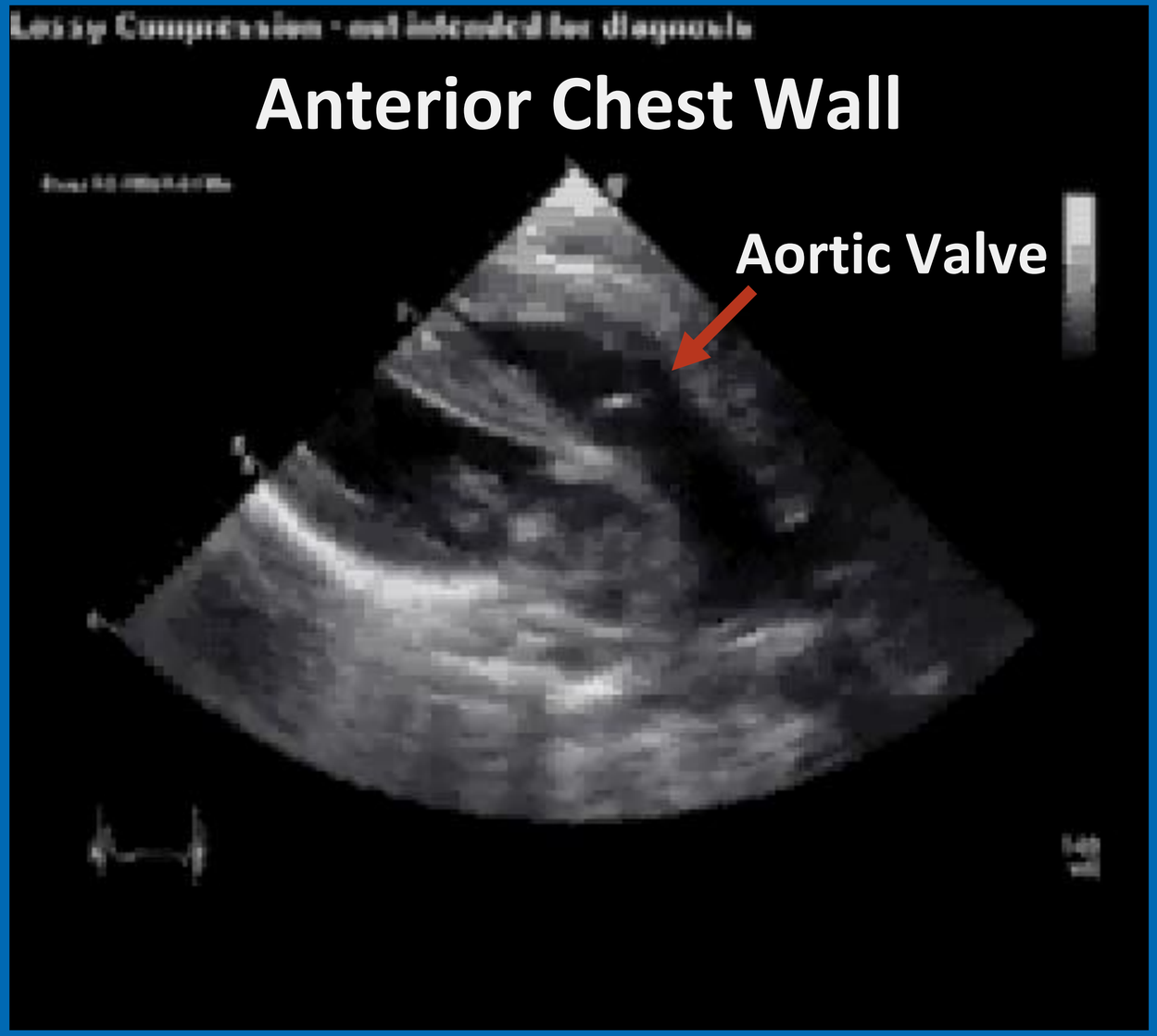
CYANOTIC & NO MURMUR

- Transposition of the Great Arteries*
- Total anomalous pulmonary veins
- Hypoplastic left heart syndrome*

* Loud second heart sound



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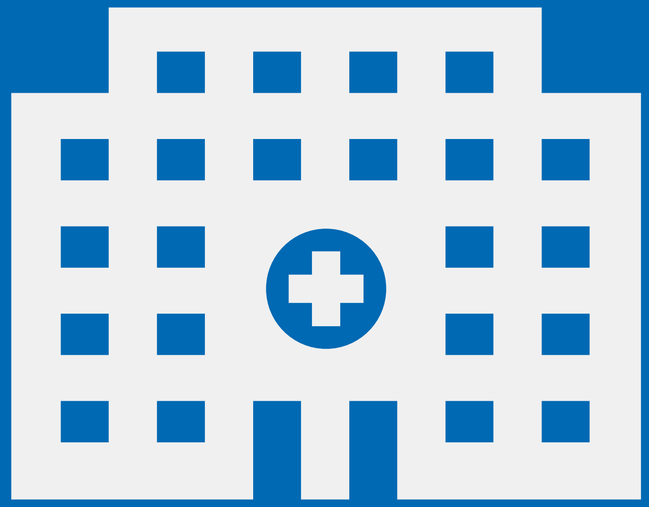
INITIAL EVALUATION

Physical Exam

Chest x-ray

ECG

Echo



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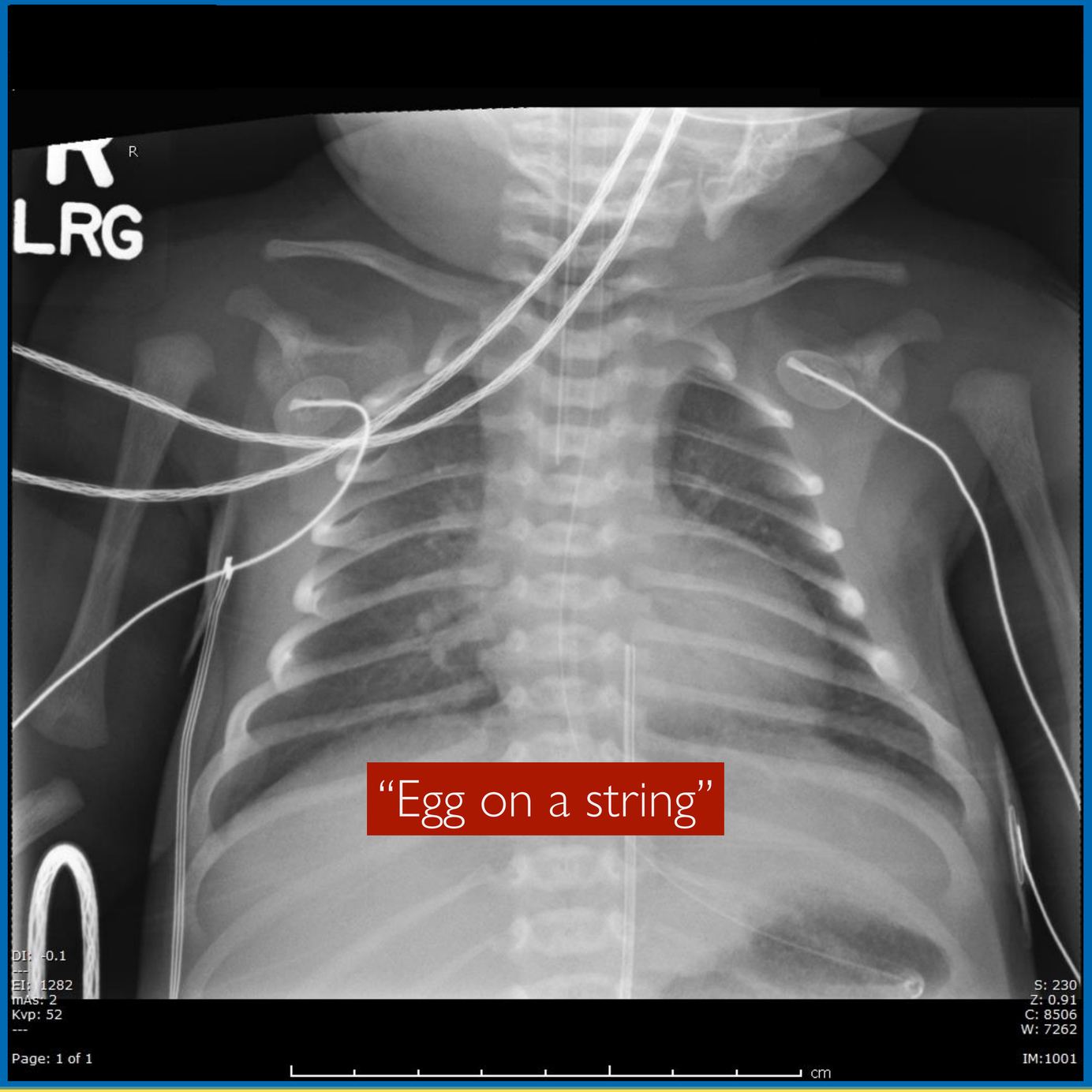
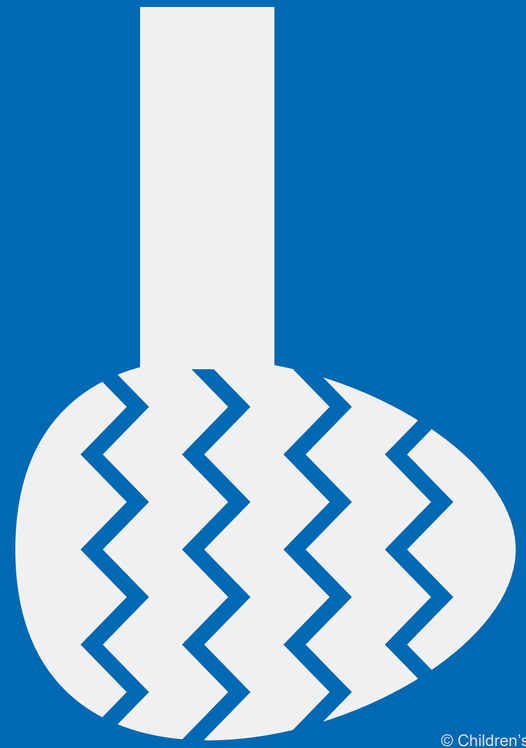
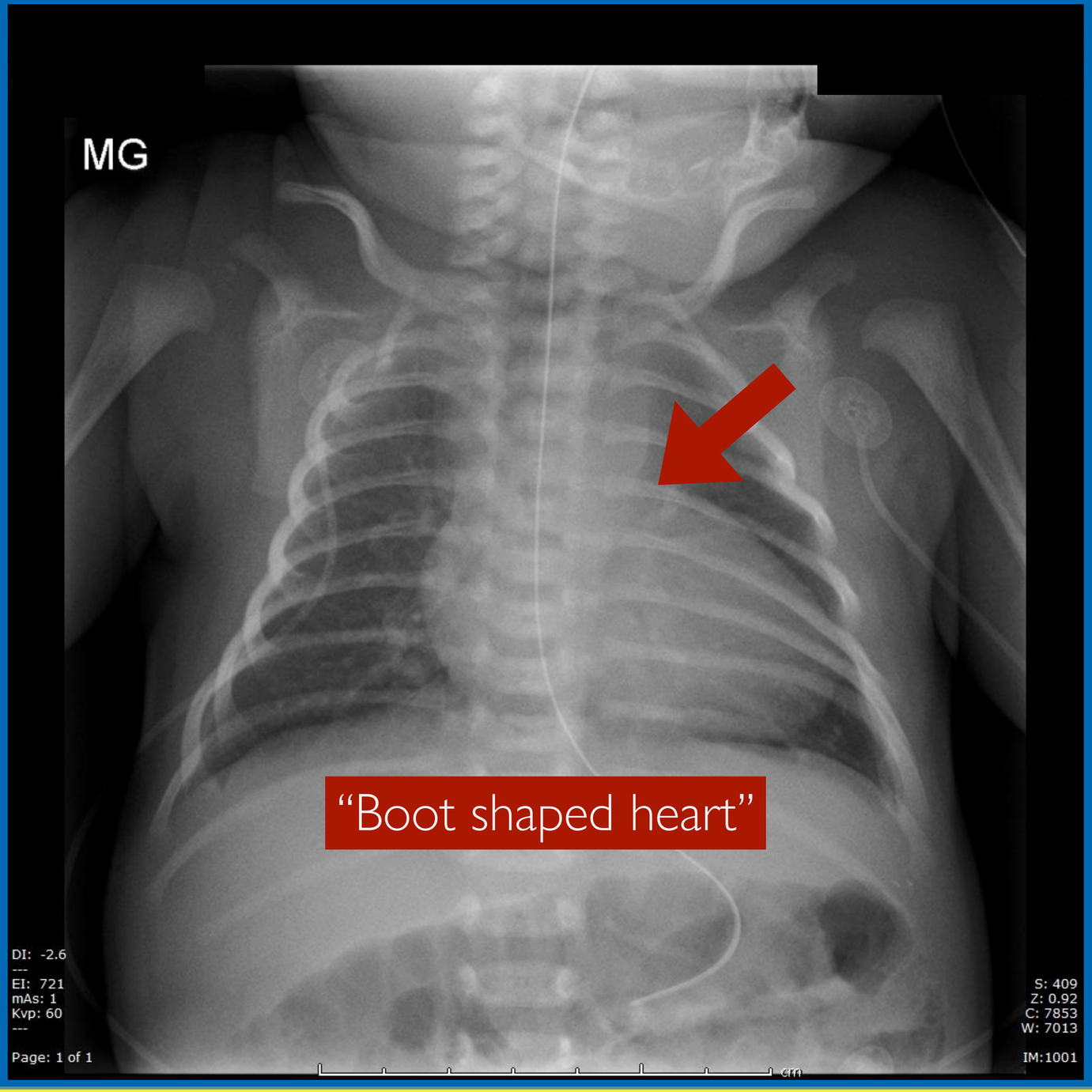


CHEST X-RAY

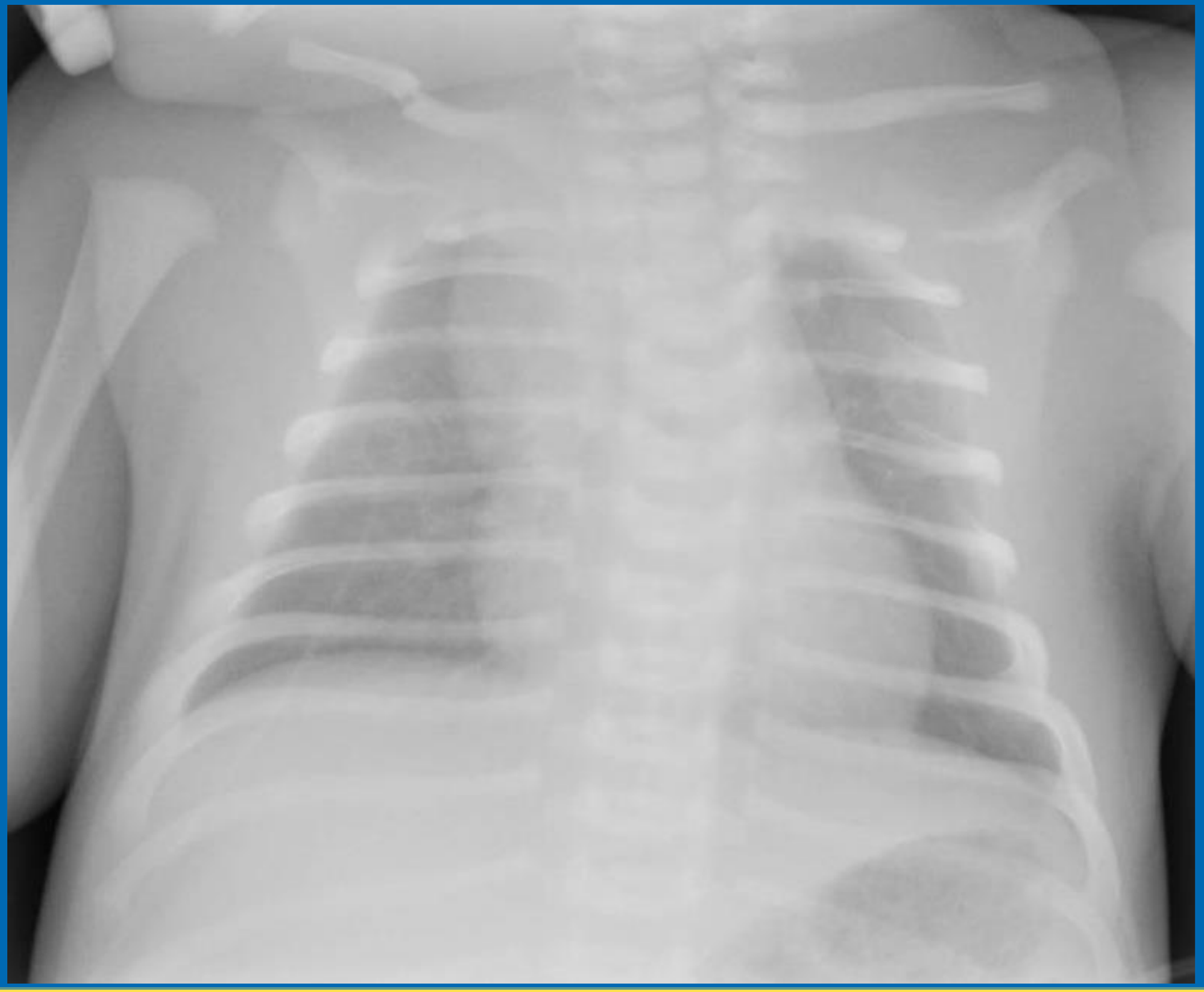
Transposition of the Great Arteries





CHEST X-RAY

Tetralogy of Fallot



CASE: CHEST X-RAY

- Pulmonary edema
- Normal heart size
- Prominent thymus
- Clavicle fracture

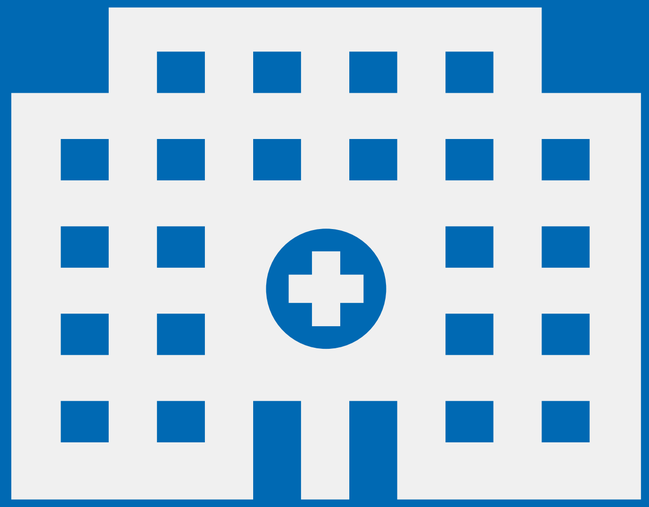
INITIAL EVALUATION

Physical Exam

Chest x-ray

EKG

Echo



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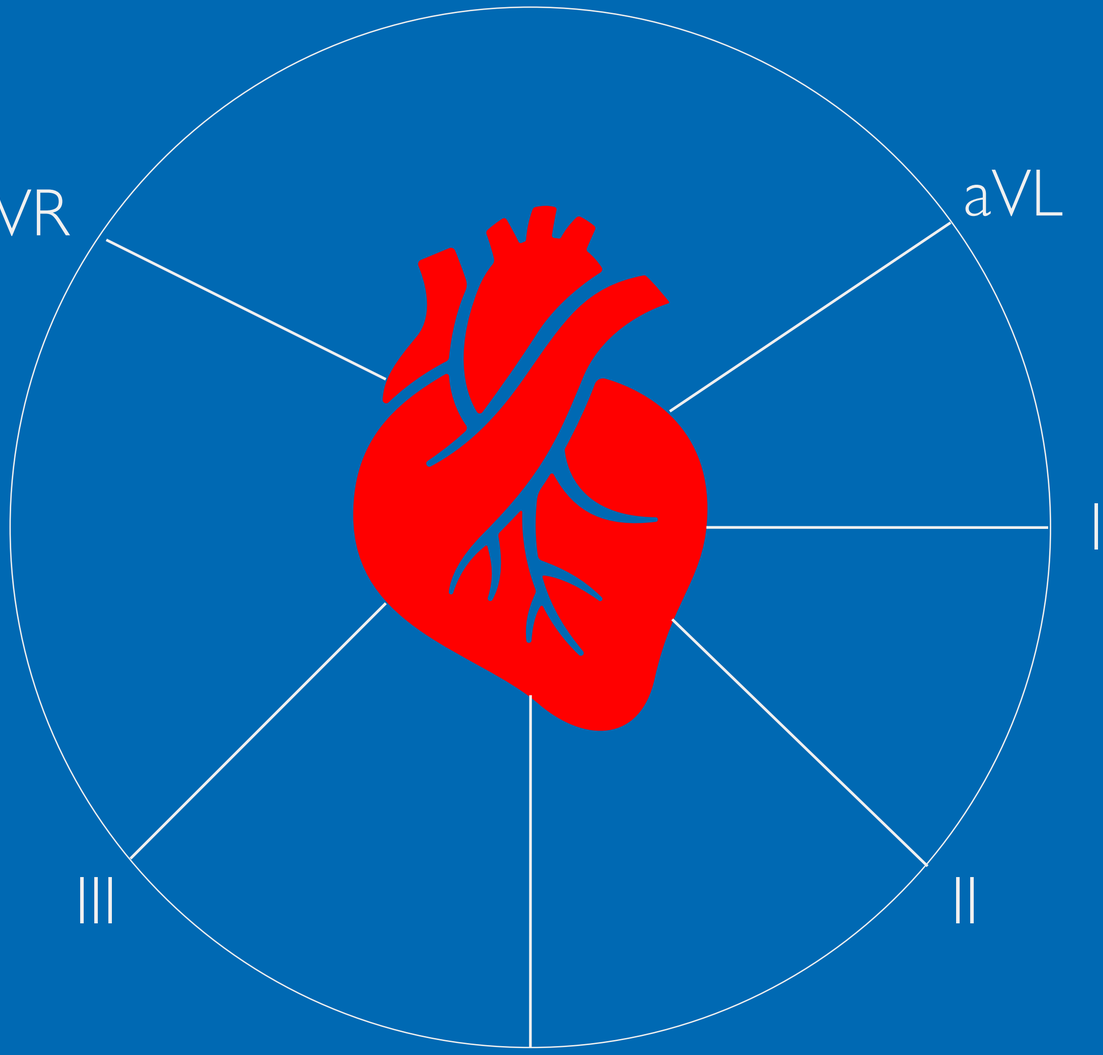
ELECTROCARDIOGRAM: AXIS



Left Axis Deviation

Right Axis Deviation

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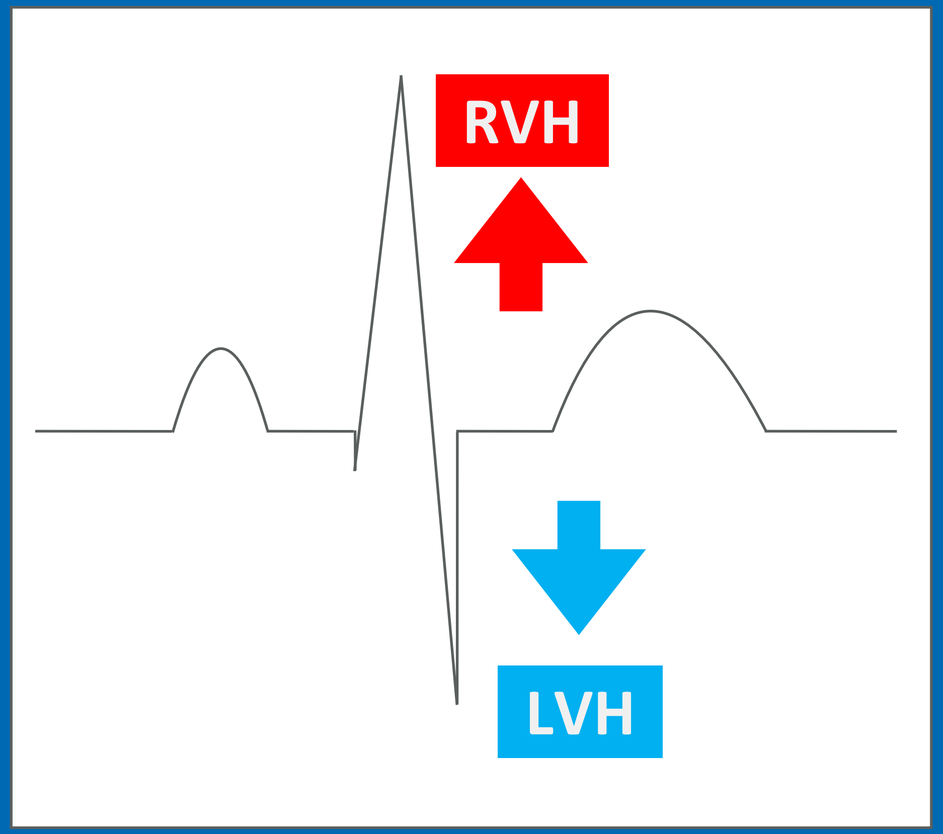
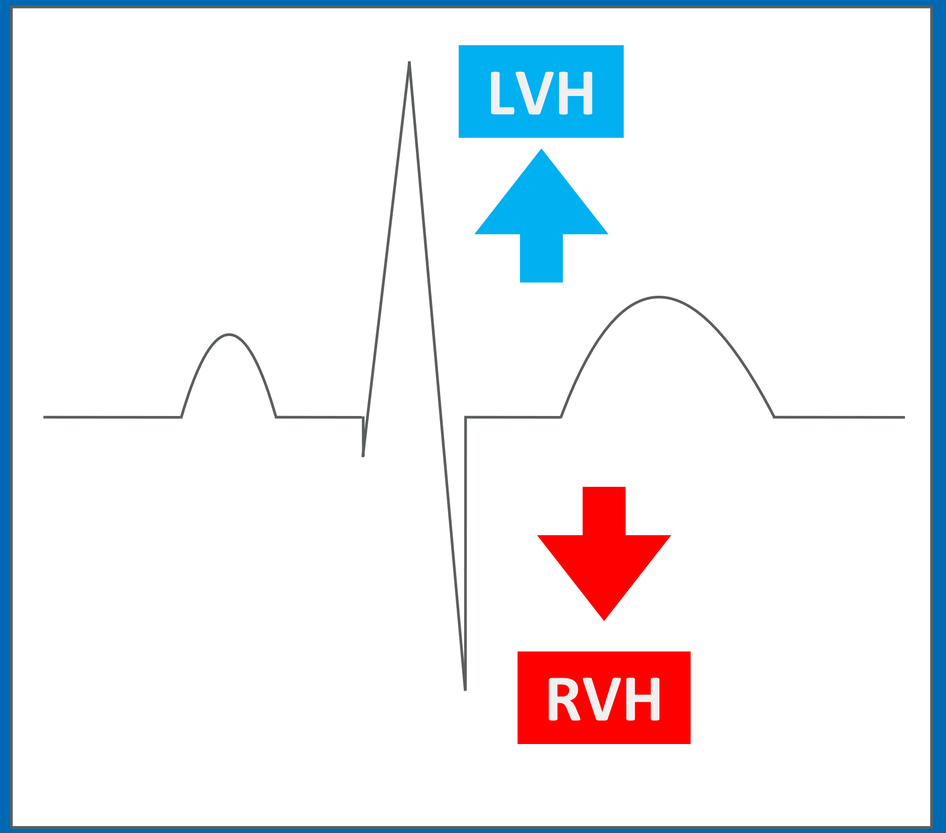
ELECTROCARDIOGRAM: HYPERTROPHY

“20,20,20,5” Rule

V1



V6



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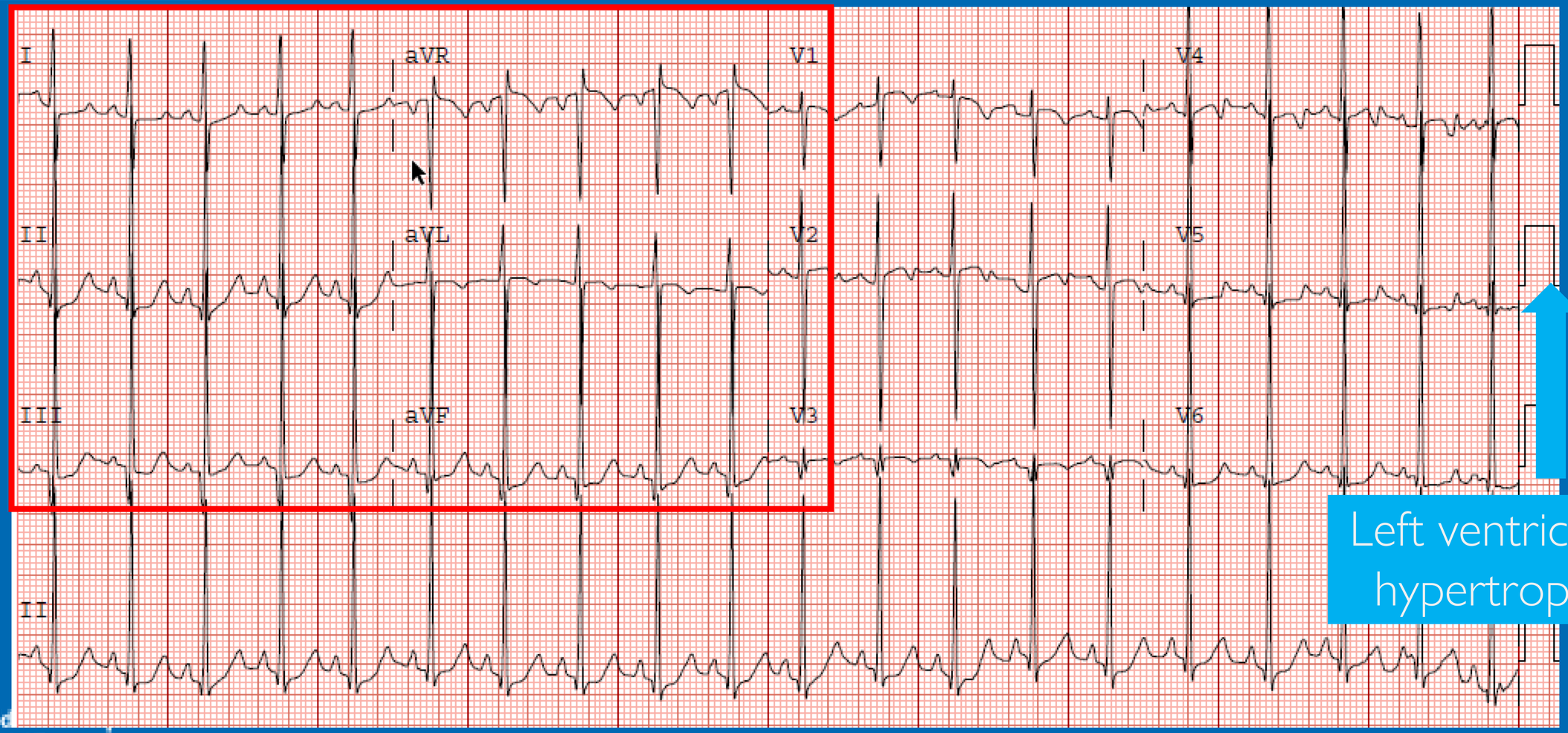
ECG

Newborn with severe aortic stenosis

Left axis deviation

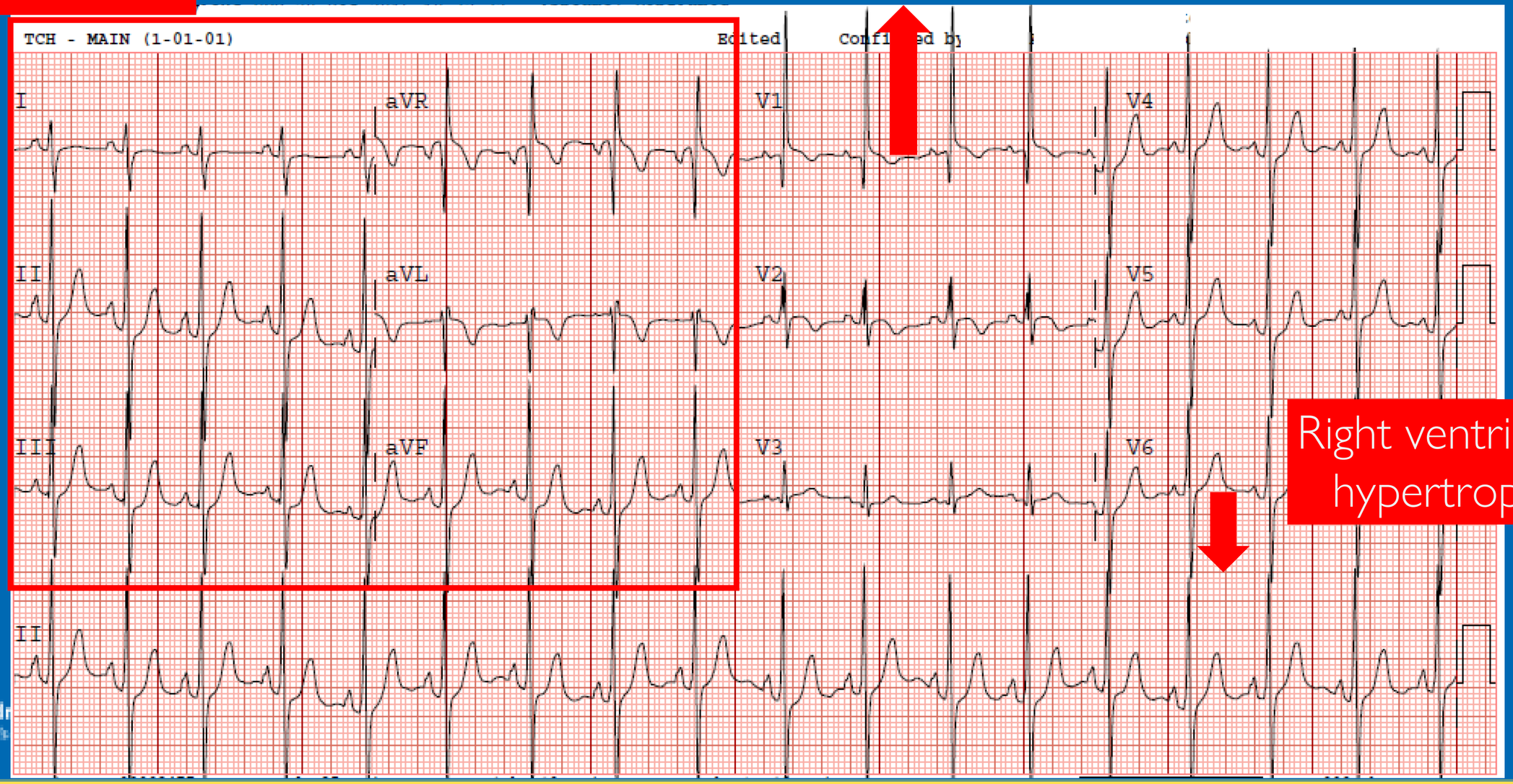


Left ventricular hypertrophy

ECG

Newborn with hypoplastic left heart syndrome

Right axis deviation

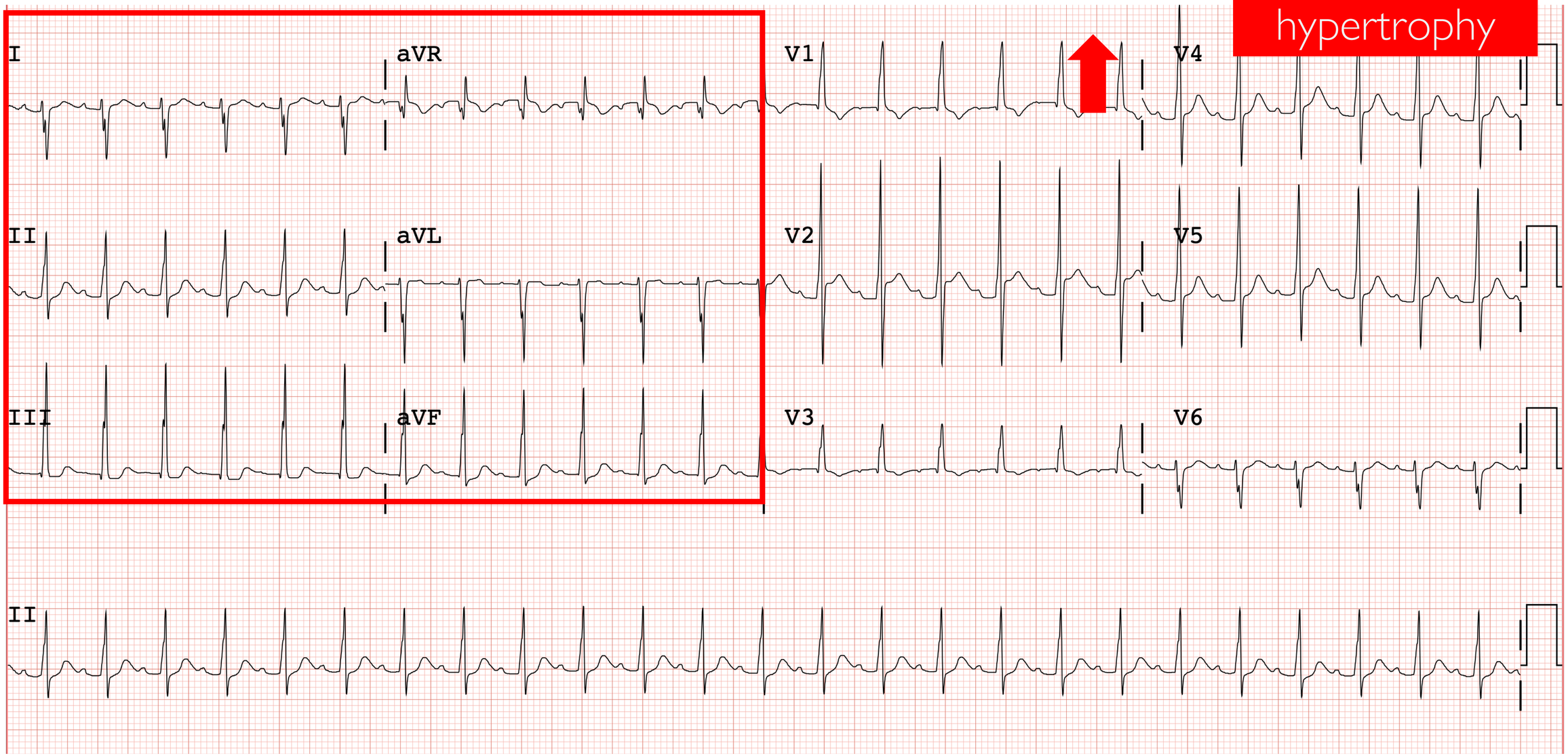


Right ventricular hypertrophy

CASE: EKG

Right axis deviation

Right ventricular hypertrophy



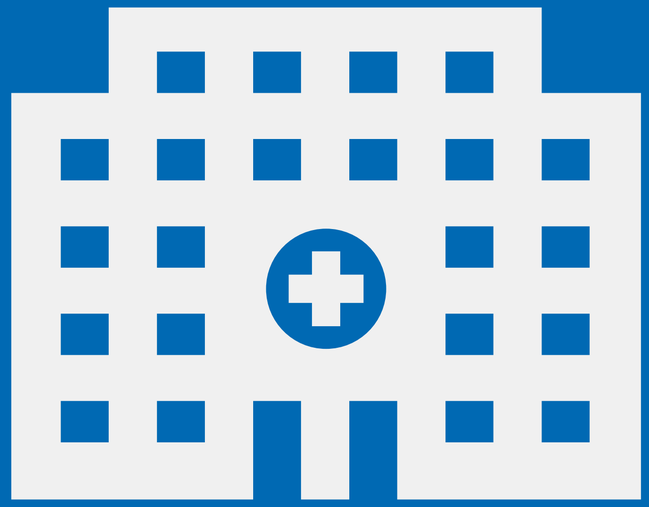
INITIAL EVALUATION

Physical Exam

Chest x-ray

EKG

Echo



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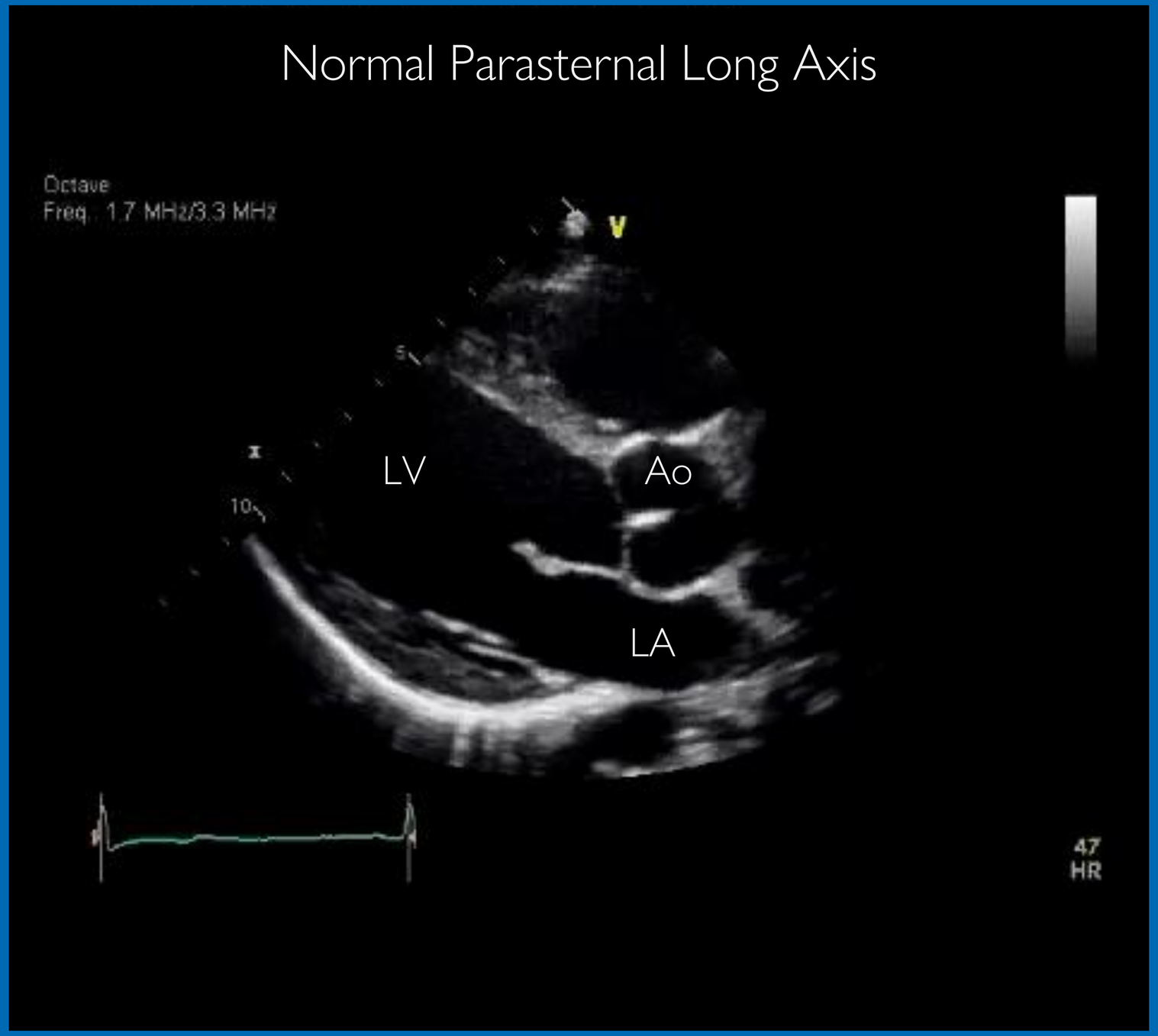
ECHO

"2D" Echo is the primary modality for assessing heart structure and function



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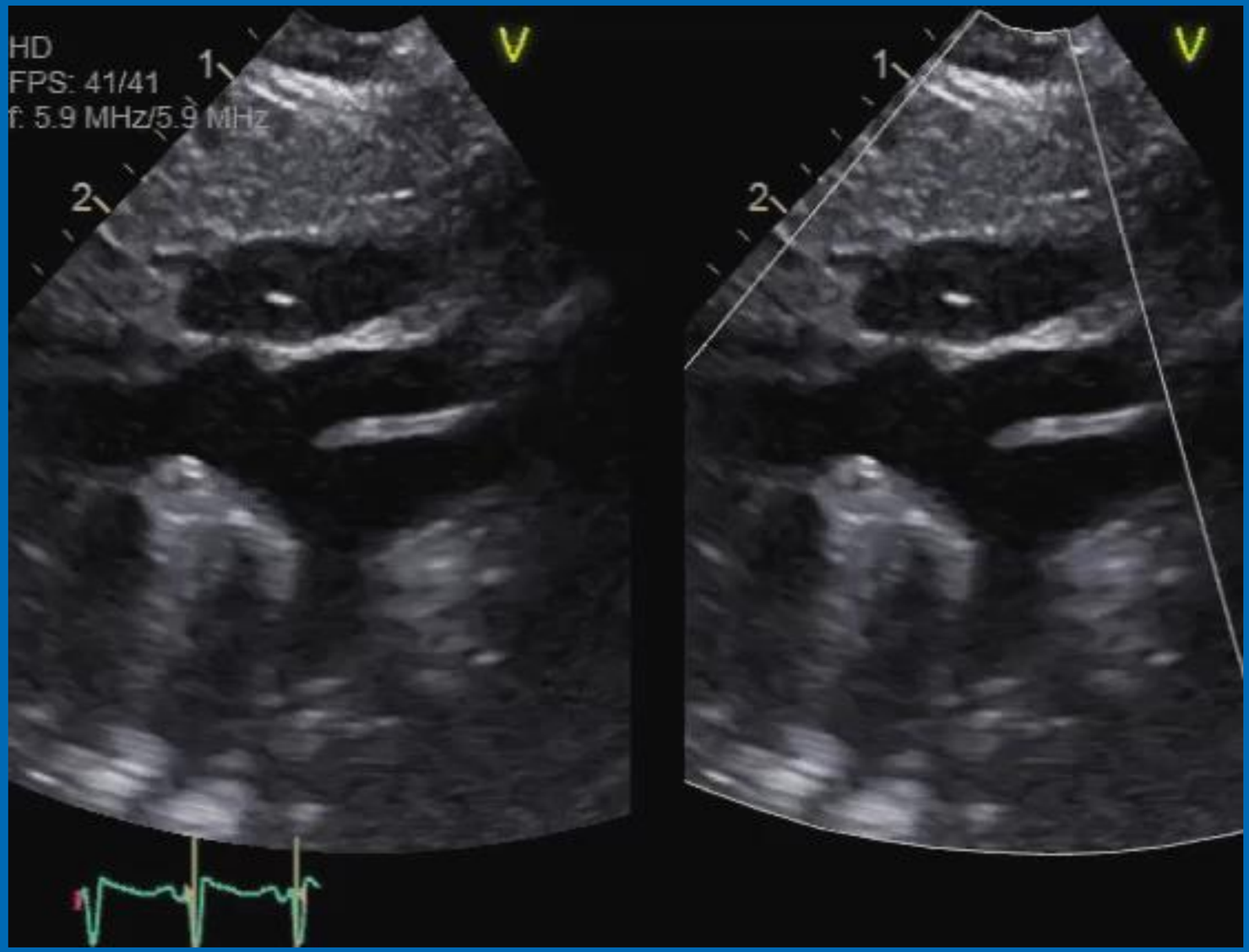
COLOR DOPPLER

Color Doppler tells you about direction of flow

BLUE = Away
RED = Toward



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"ALIASING"

"the misidentification of a signal frequency"

Translation: the blood moves too fast to accurately show direction of flow

Nyquist Limit = the maximum velocity that can be accurately evaluated

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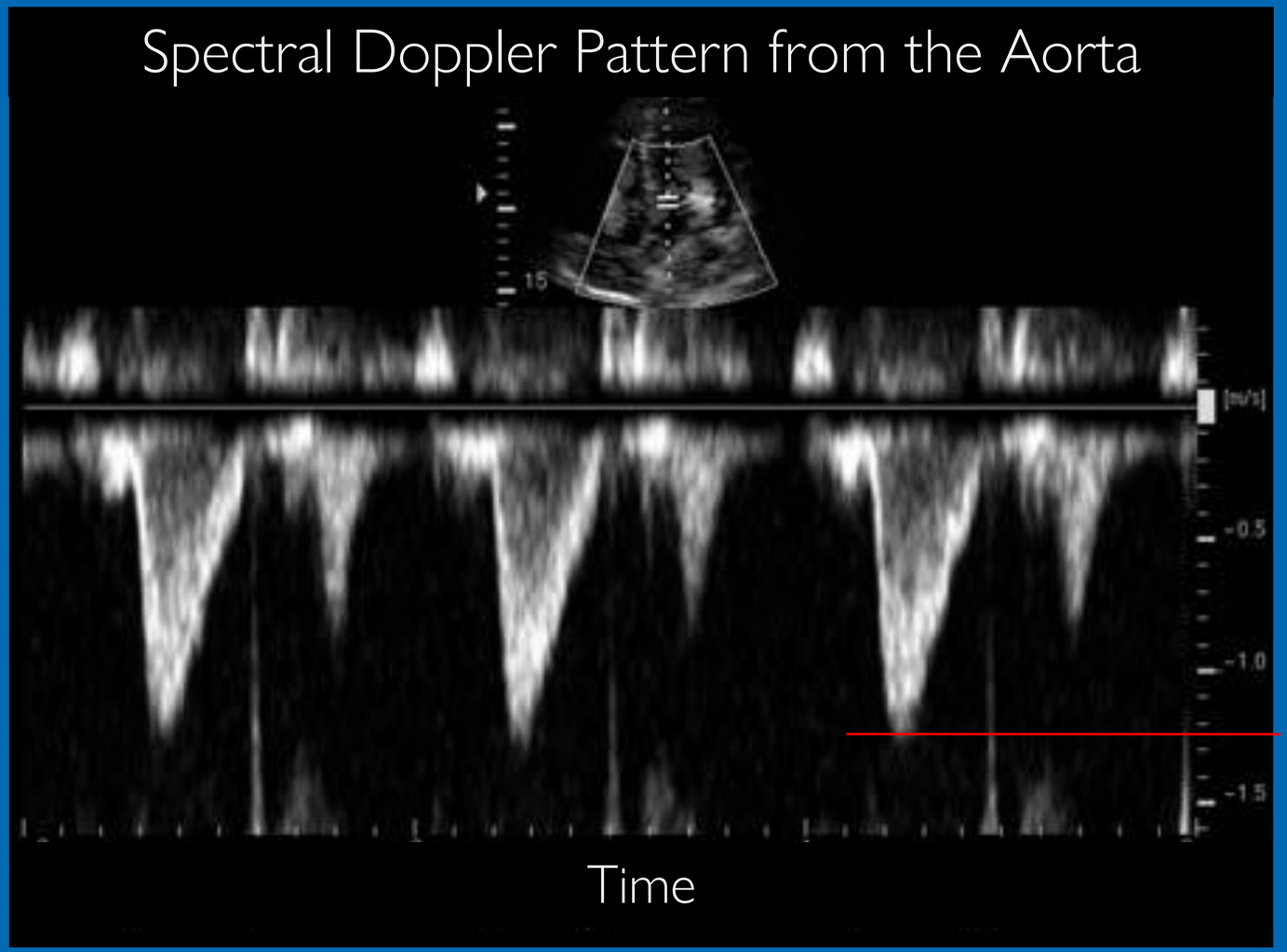
SPECTRAL DOPPLER

A graph of time versus velocity



Peak Velocity 1.25m/s

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CASE: ECHO

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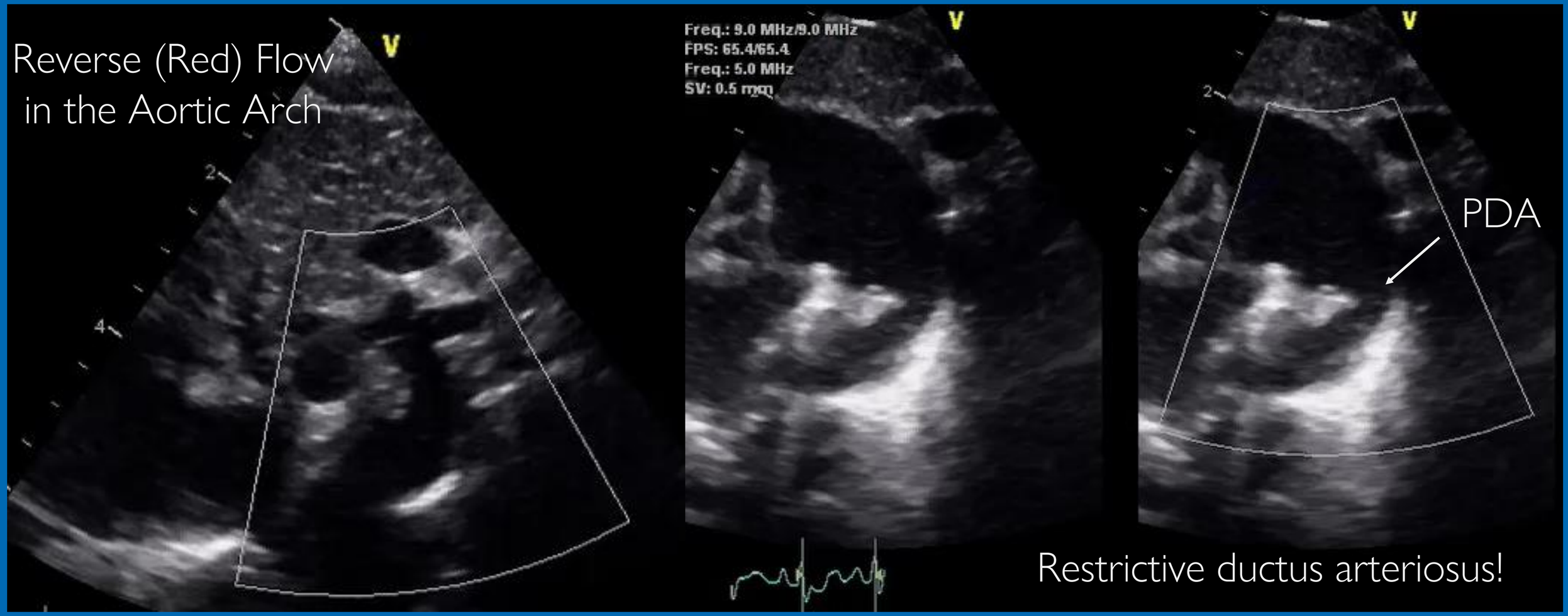
CASE: ECHO



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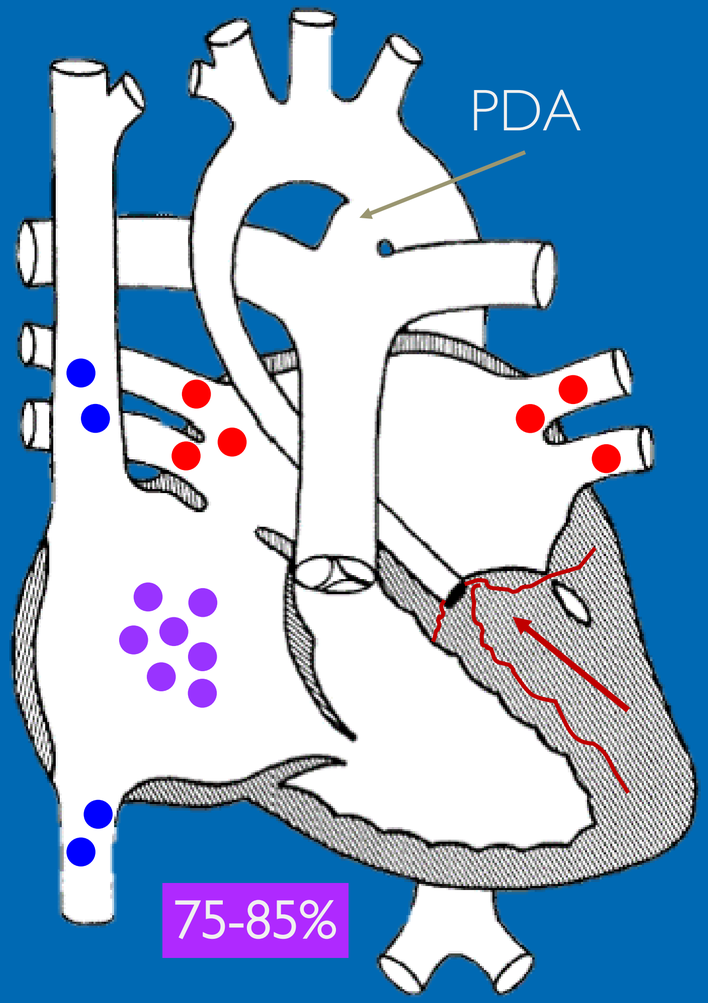
PHYSIOLOGY - HLHS CIRCULATION



"Single Ventricle Physiology"

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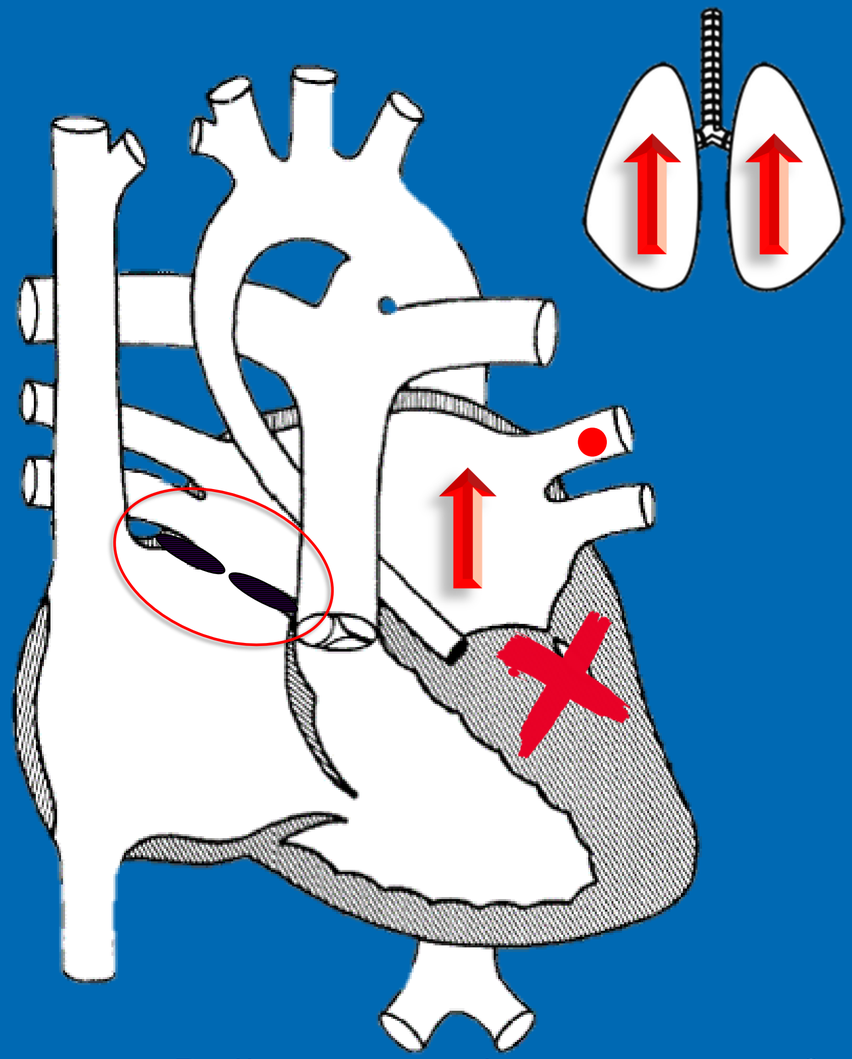
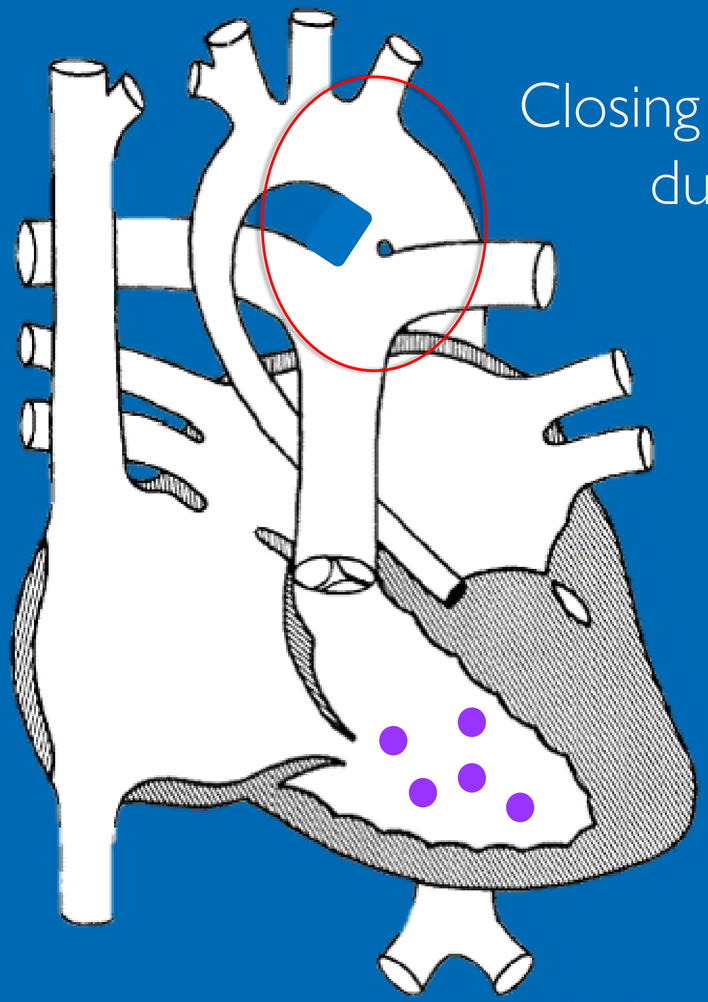
PROBLEM AREAS

Restrictive atrial septum



Balloon atrial septostomy?

Closing arterial duct



Prostaglandin (PGE)?

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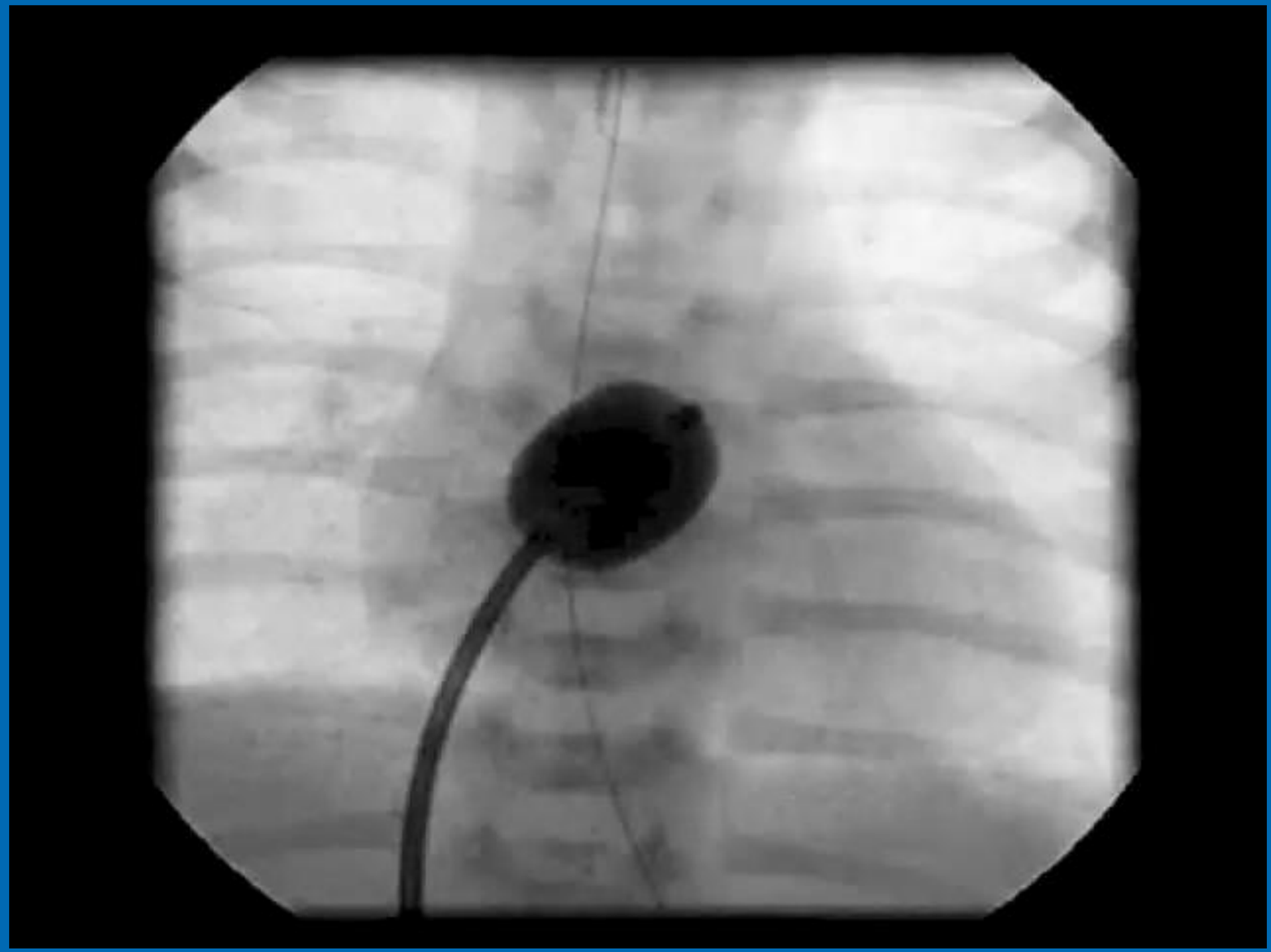
ATRIAL SEPTOSTOMY

A restrictive atrial septum may cause left atrial hypertension, pulmonary edema & clinical decompensation!



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PROSTAGLANDIN

Key Concept: When & How start PGE

- **Suspected "ductal-dependent" heart disease**
- **Starting dose: 0.05 mcg/kg/min**
- **Beware apnea & hypotension!**
- **If the baby worsens, may need emergent interventional cardiology procedure!**

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LEARNING OBJECTIVES

- Definition and differential diagnosis of neonatal cyanosis
  - **Use the hyperoxia test to differentiate cyanotic heart disease**
- Initial evaluation of cyanotic heart disease
  - **Pre/Post ductal sats, physical exam, CXR, EKG, Echo**
- When & how to start Prostaglandin
  - **Suspected ductal dependent heart disease**

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THANK YOU!

My contact information:
michael.dimaria@childrenscolorado.org
Office: (720) 777-2943

Special thanks: Scott Kirby, RDCS

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