WHERE IS THE FOURTH VALVE? TRICUSPID ATRESIA

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DISCLOSURES

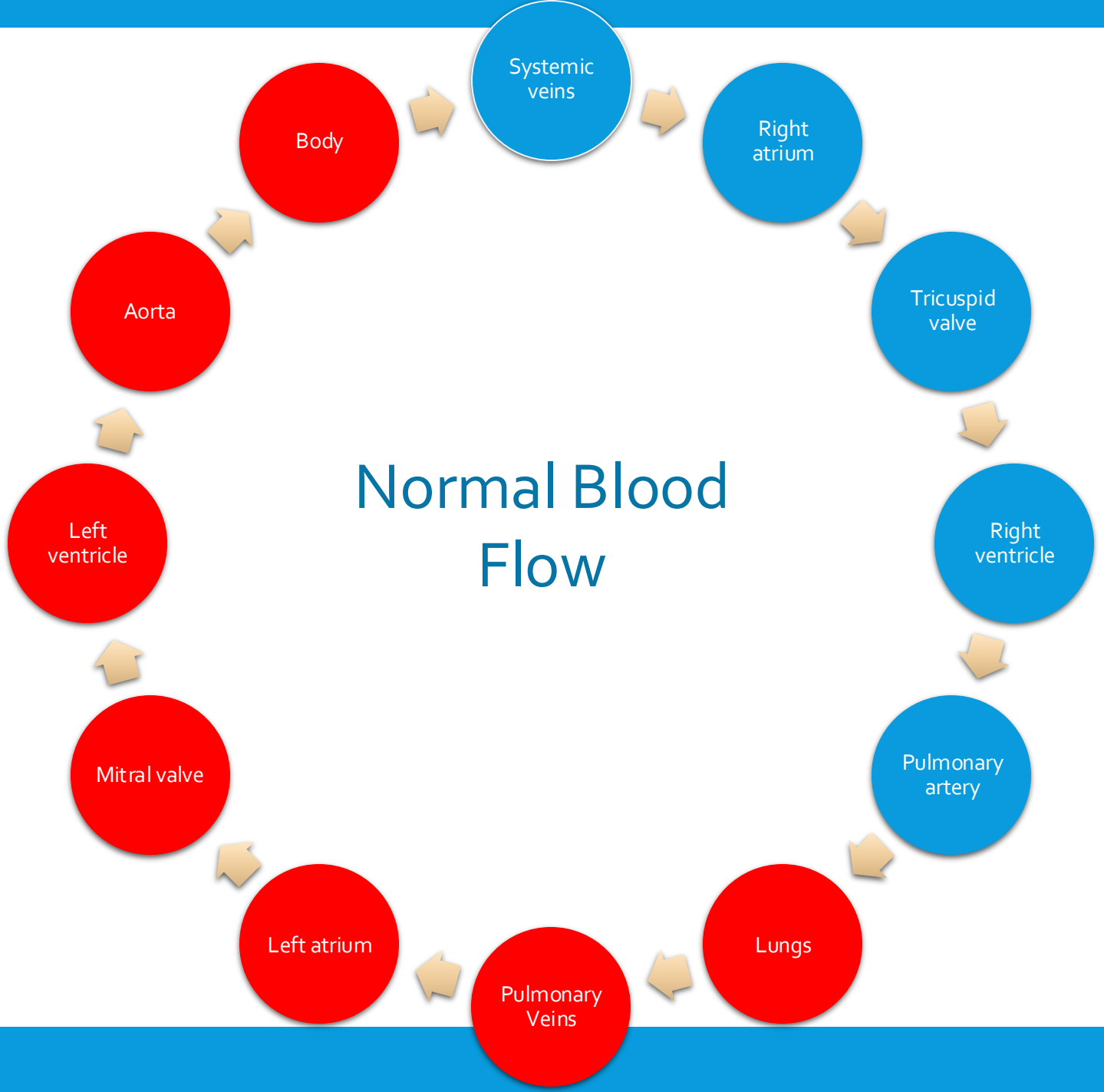
- No disclosures

OBJECTIVES

- To comprehend the severity of tricuspid atresia.
- To understand the different types of tricuspid atresia.
- How to perform a high quality ultrasound to prove tricuspid atresia

NORMAL FETAL HEART

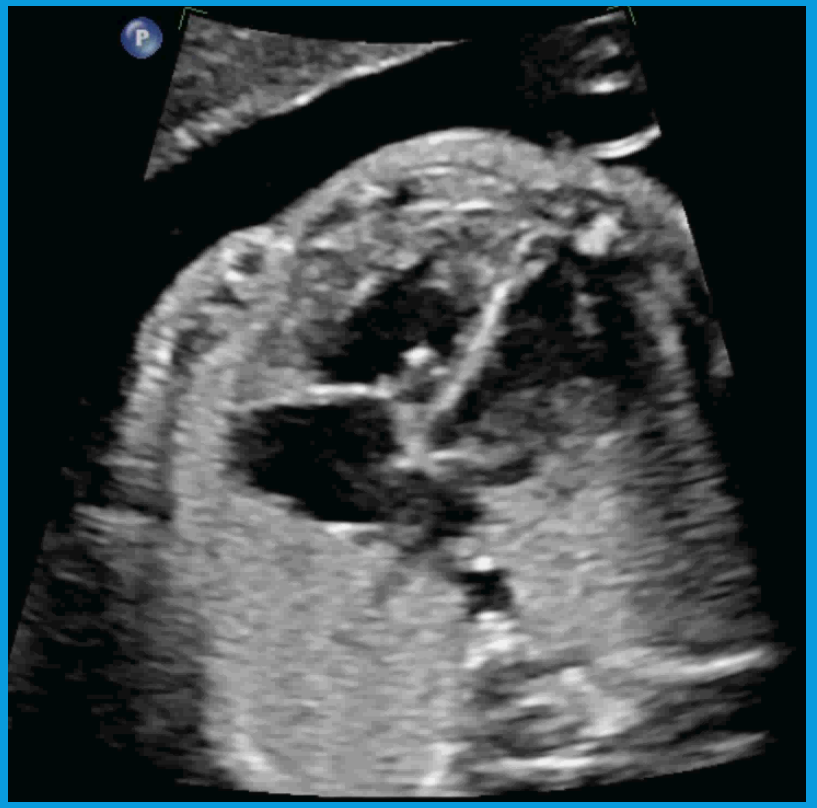
Normal Blood Flow



DIFFERENT TYPES OF TRICUSPID VALVE DISEASE

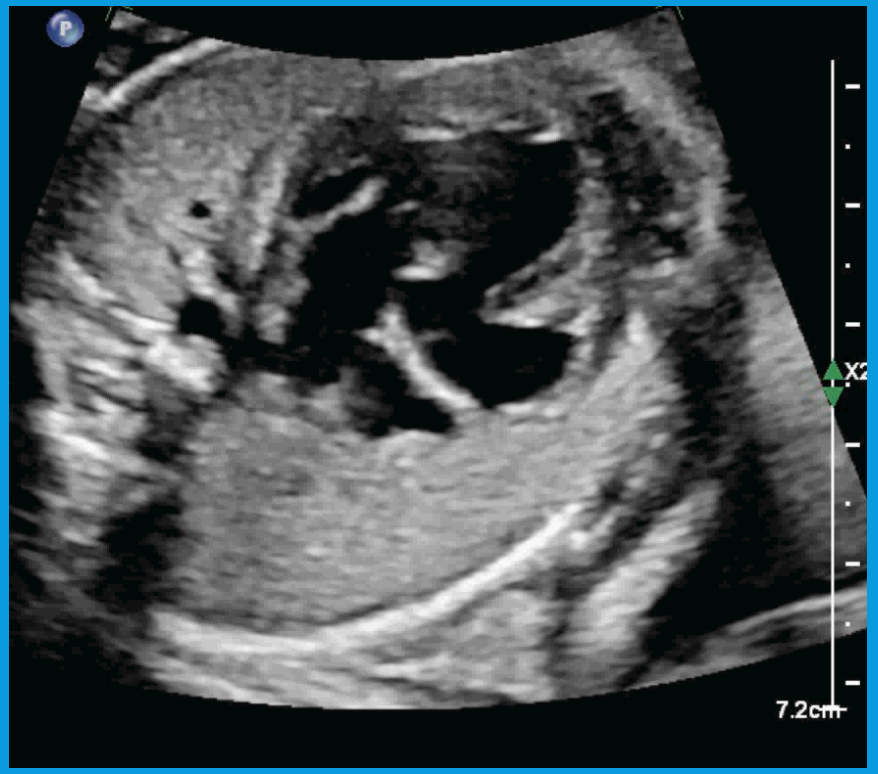
Normal leaflet excursions

Dysplastic TV
Thickened, non coapting leaflets.



Hypoplastic TV Annulus
Limited flow across TV, monophasic Doppler
Almost atretic

DIFFERENT TYPES TRICUSPID VALVE DISEASE



Tricuspid Atresia
No inflow, valve is replaced with plate like tissue

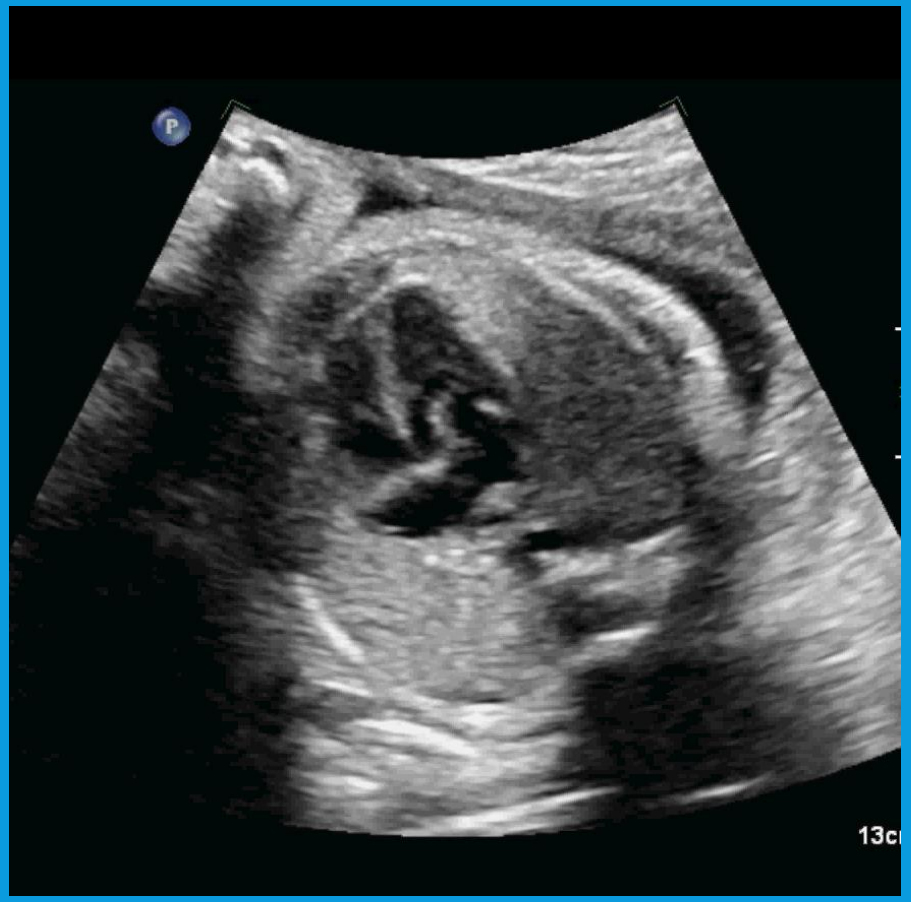
Tricuspid atresia, hypoplastic RV, with pulmonary atresia

NORMAL VS. ABNORMAL 4 CHAMBER HEART

NORMAL VS. ABNORMAL

27 weeks Normal

31 weeks TA



TRICUSPID ATRESIA

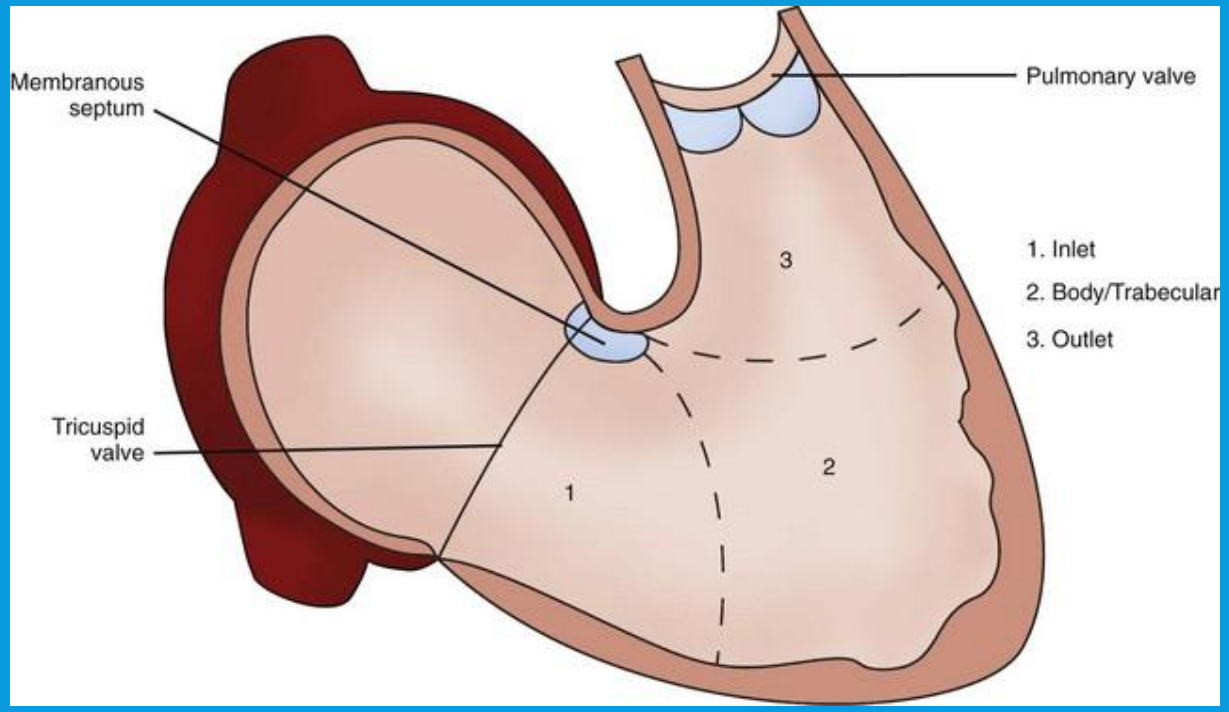
- Tricuspid atresia is a congenital heart disease occurring in 2 out 10,000 live births
- Tricuspid atresia is a CHD that consists a sealed off tricuspid valve. No prograde flow from the right atrium to the right ventricle.
- In tricuspid atresia, the oxygen rich blood is mixed with the deoxygenated blood flow.
- An ASD and VSD is needed to provide the blood mixing.
- Symptoms of tricuspid atresia postnatal:
  - Cyanosis
  - Sweating
  - Rapid breathing and heartbeat
  - Poor weight gain

TRICUSPID ATRESIA COMPONENTS

- Tricuspid atresia consists of:
  - Plate like tricuspid valve – instead of a valve there is a solid plate of tissue, obstructing the blood flow from the right atrium to the right ventricle.
  - Underdeveloped right ventricle – no flow no grow. Without a tricuspid valve inflow, the RV will appear smaller.
  - ASD – without the tricuspid valve, the blood flow will cross the atrial septum to the left side.
  - Ventricular septal defect – Bulboventricular foramen, allows blood to cross into the RV and supply blood flow to the pulmonary valve (in normally related great arteries)
  - Normal or malposed great arteries – determines the type of tricuspid atresia in the fetal heart. The size and position will also determine if the baby requires prostaglandins or not.

RIGHT VENTRICLE

- The right ventricle has 3 components making it a tripartite
  - Inlet – Tricuspid valve inflow
  - Apical/trabecular portion
  - Outflow
- Tricuspid atresia is missing the inlet portion making the RV a bipartite.



Thoracictexbox

TRICUSPID ATRESIA - VSD

- Tricuspid atresia typically is associated with a VSD or bulboventricular foramen.
- The severity of RV hypoplasia and hypoplastic pulmonary artery is determined by the size of the VSD.
- If the VSD is small, the RV will have a smaller cavity and the pulmonary artery (or aorta if great arteries are malposed) will have a certain degree of stenosis/hypoplasia
- Most patients with tricuspid atresia should have VSD.

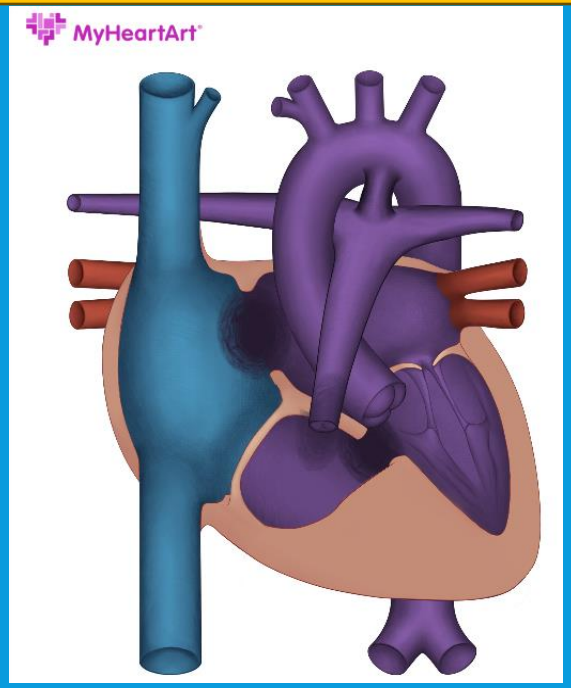
ASSOCIATED LESIONS

- Pulmonary stenosis – pulmonary artery coming from the small right ventricle, may be hypoplastic with pulmonary stenosis
- Transposition of the great arteries
- Double outlet RV
- VSD

Types of Tricuspid Atresia

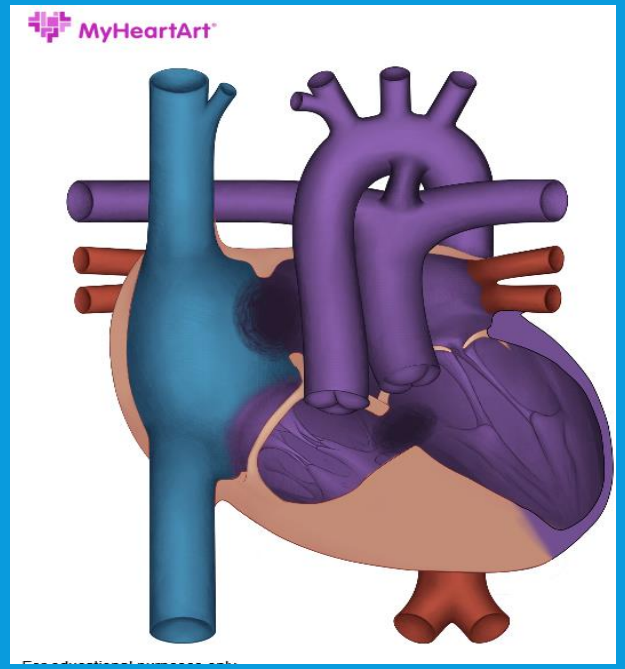
Type 1

Tricuspid atresia with normally related great arteries (70-80%)



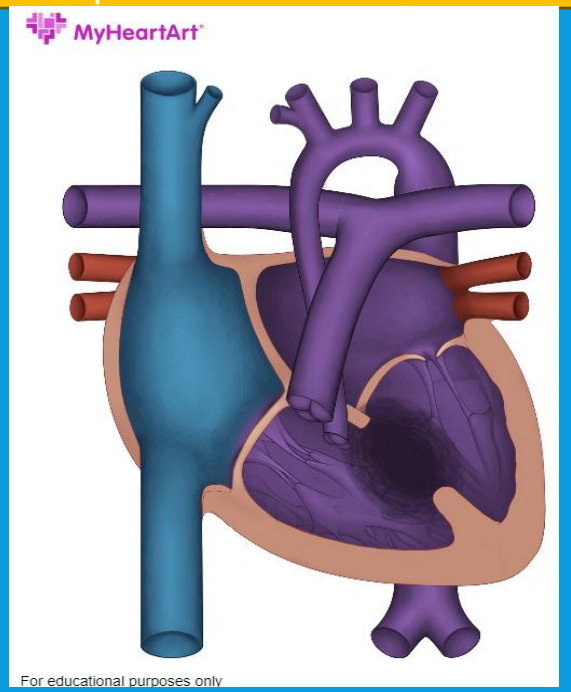
Type 2

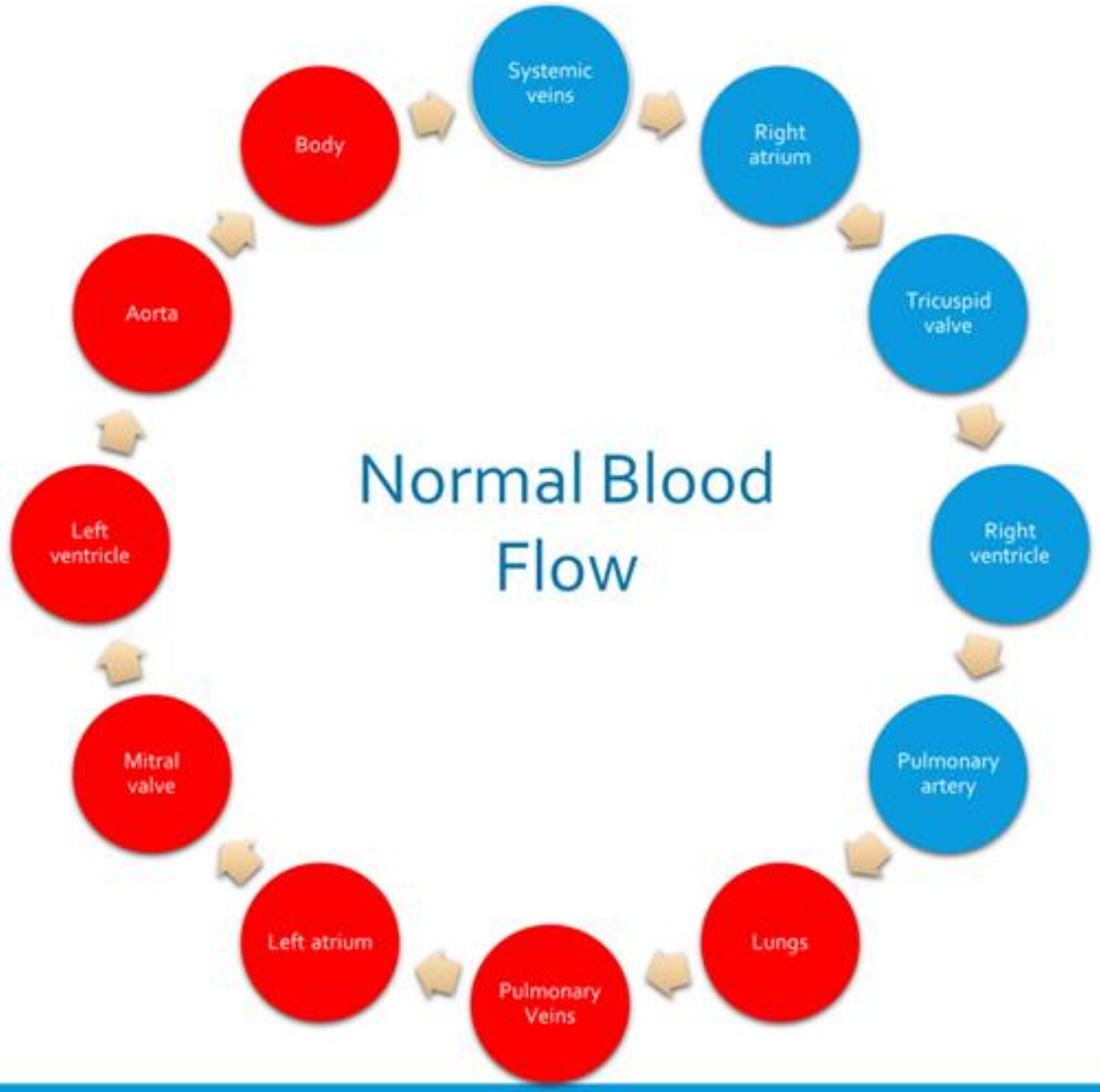
Tricuspid atresia with transposed great arteries(12-25%)

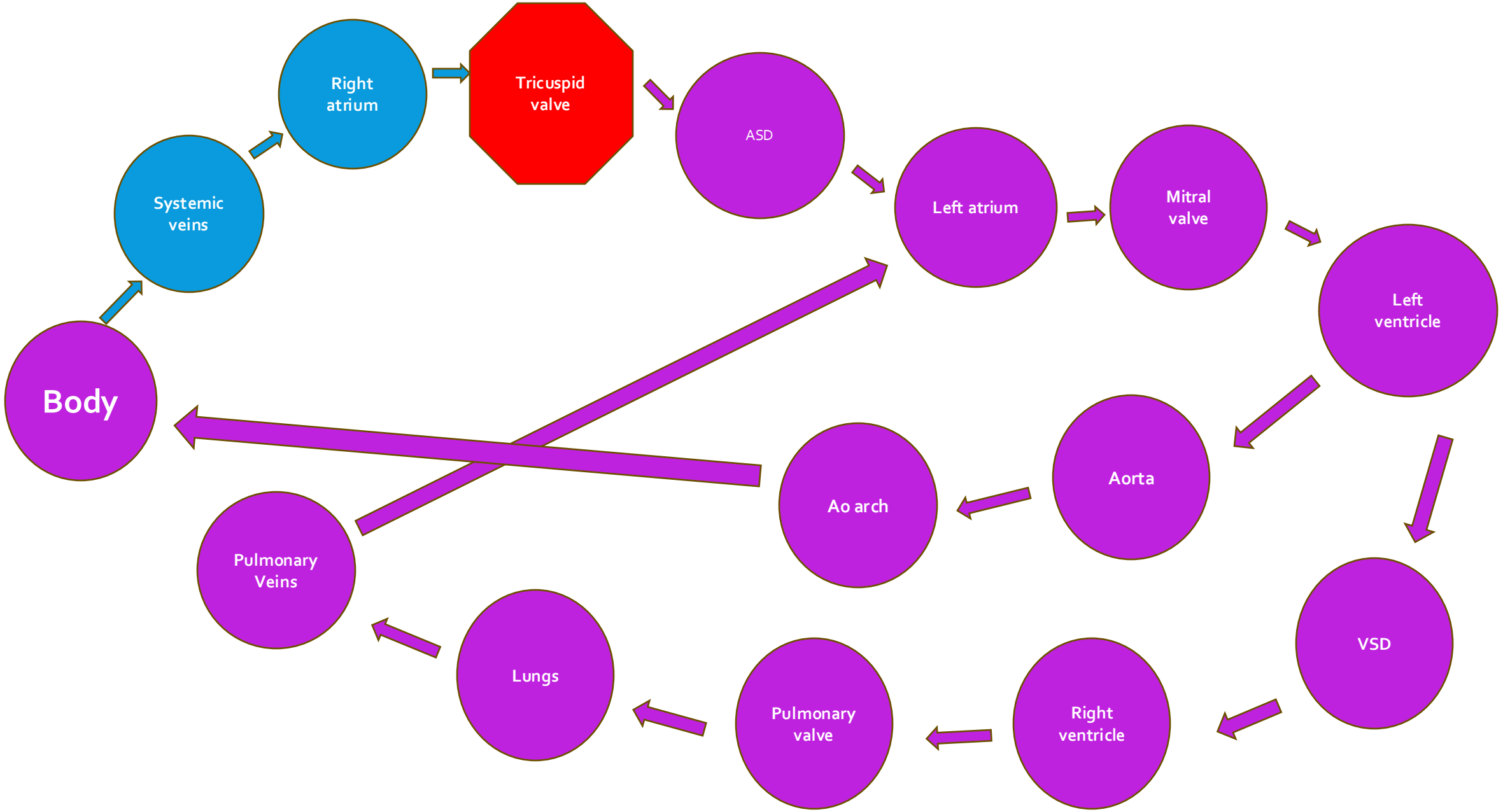


Type 3

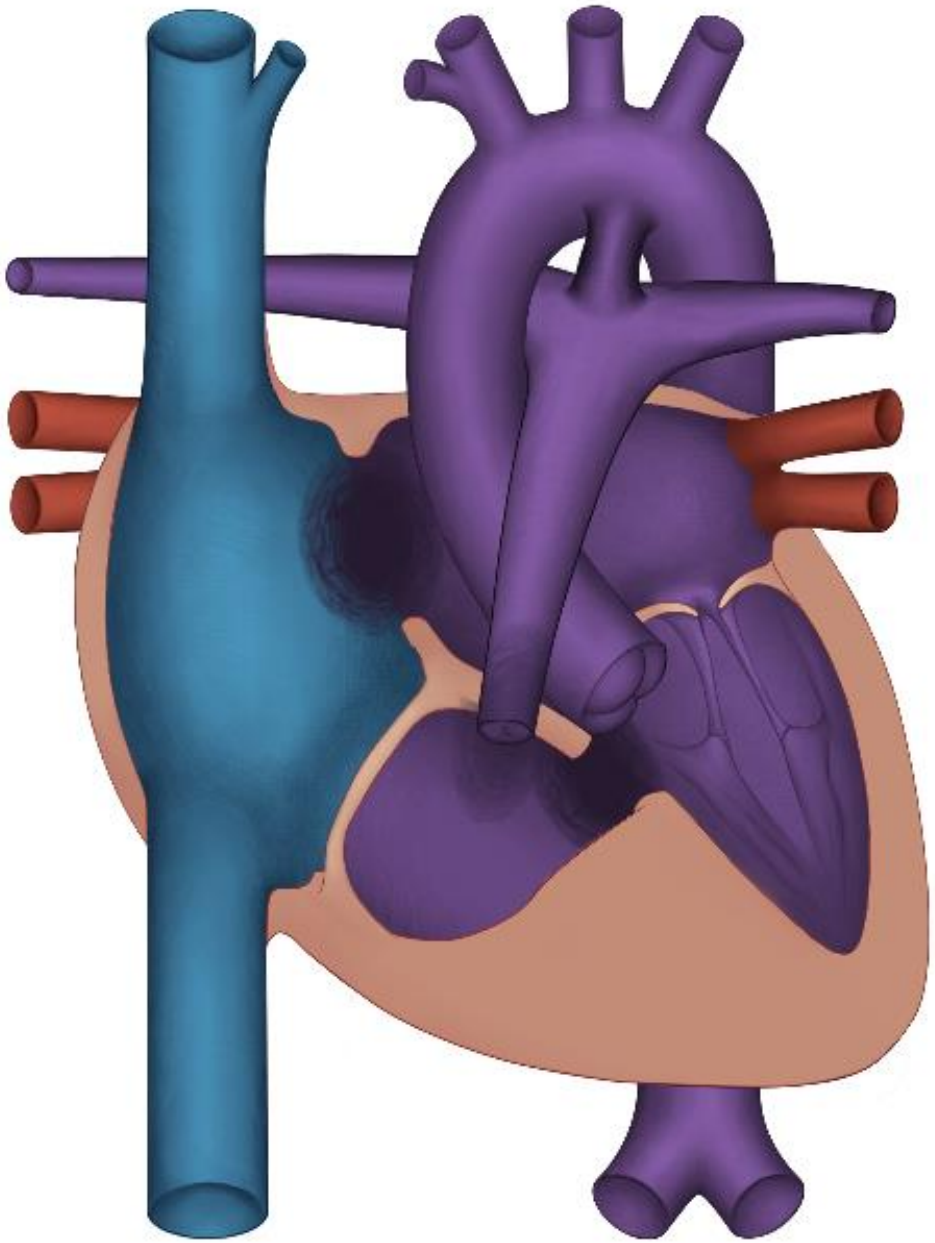
Tricuspid atresia with more complex heart disease such as L-looped ventricles (very rare, 3-6% patients with TA)




Normal Blood Flow
Systemic veins
Right atrium
Tricuspid valve
Right ventricle
Pulmonary artery
Lungs
Pulmonary Veins
Left atrium
Mitral valve
Left ventricle
Aorta
Body


Right atrium
Tricuspid valve
ASD
Left atrium
Mitral valve
Left ventricle
Systemic veins
Body
Aorta
Ao arch
Pulmonary Veins
VSD
Lungs
Pulmonary valve
Right ventricle

MyHeartArt®



For educational purposes only

TRICUSPID ATRESIA TYPE I NORMALLY RELATED GREAT ARTERIES

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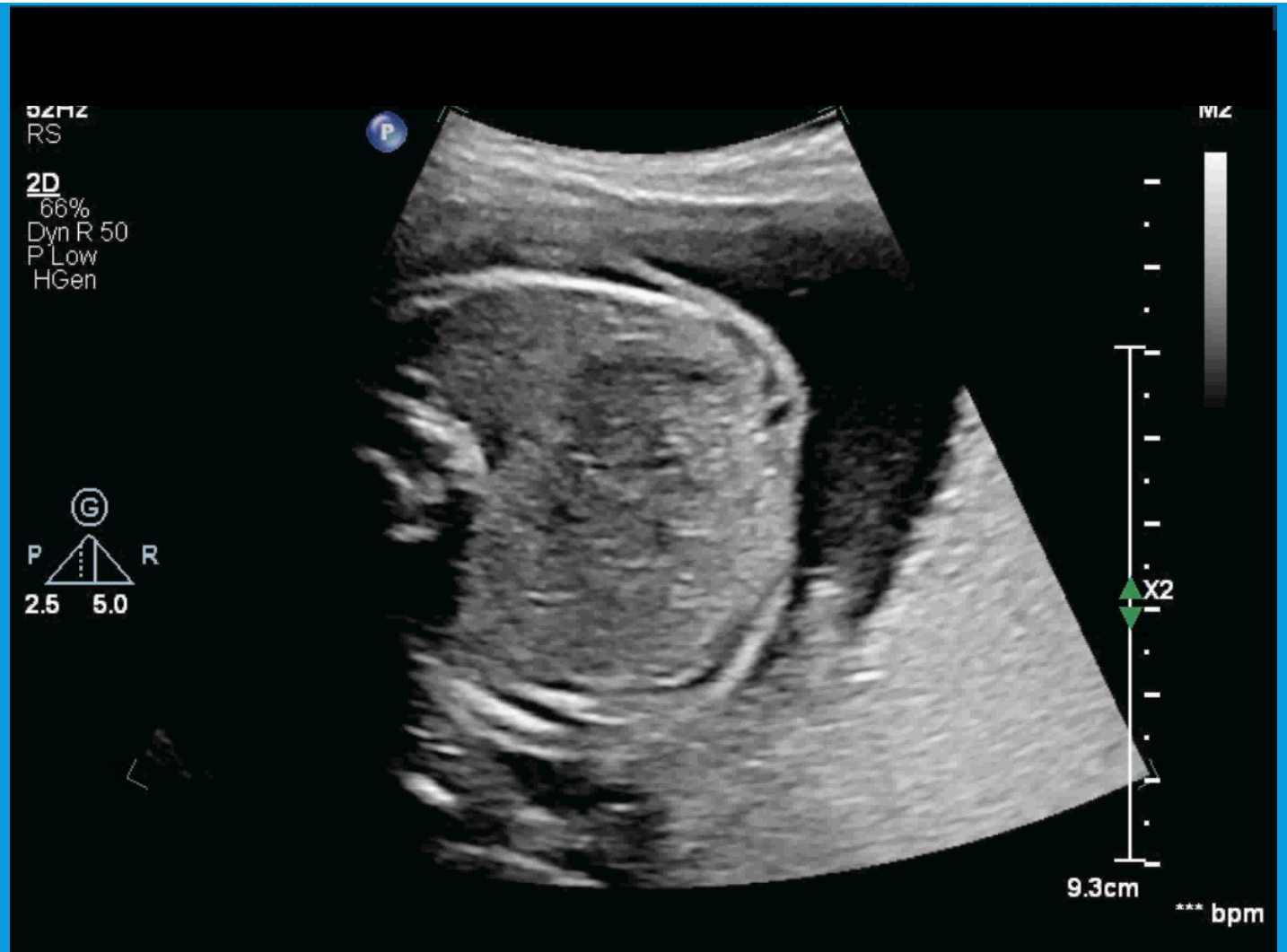

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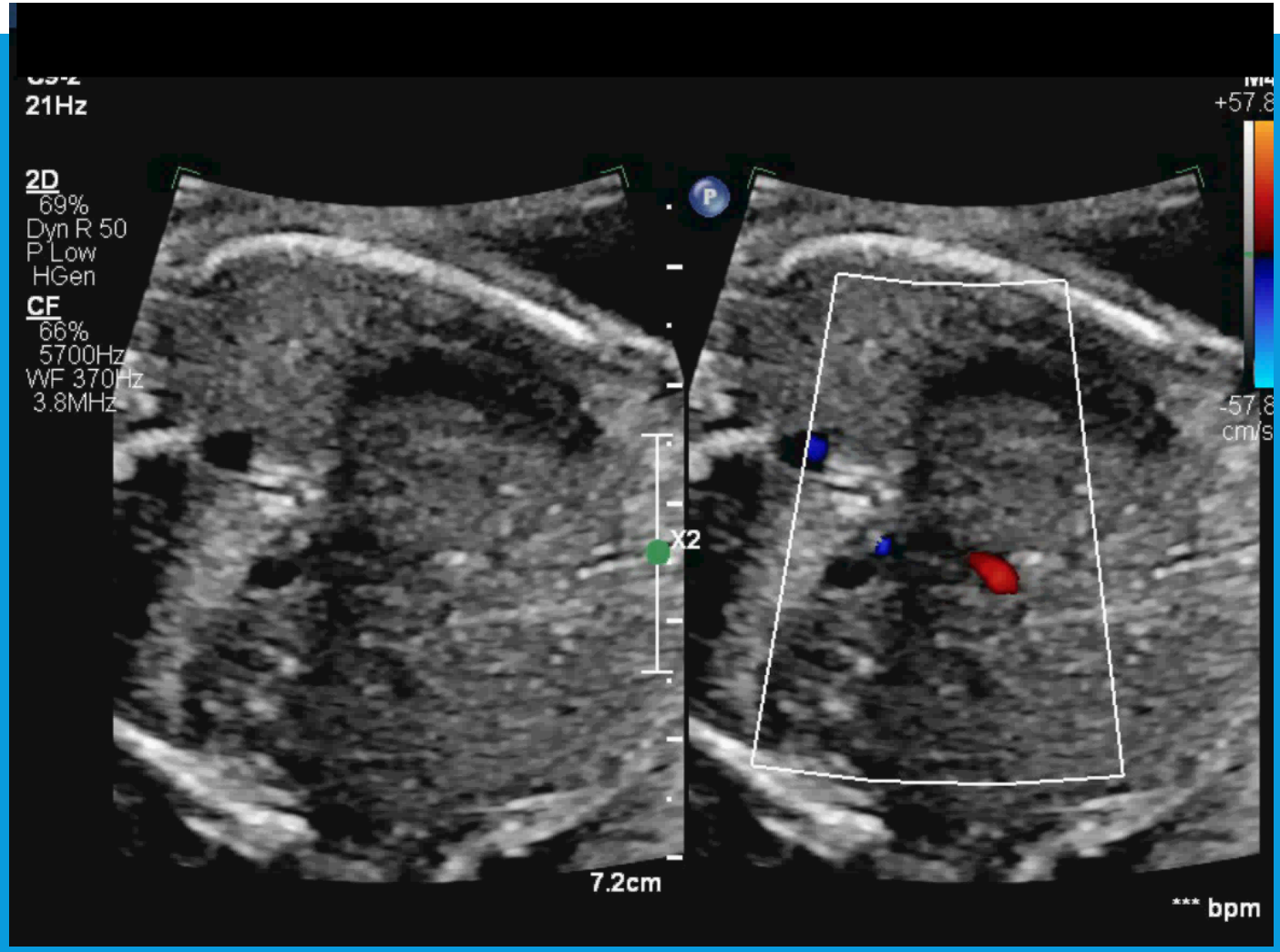

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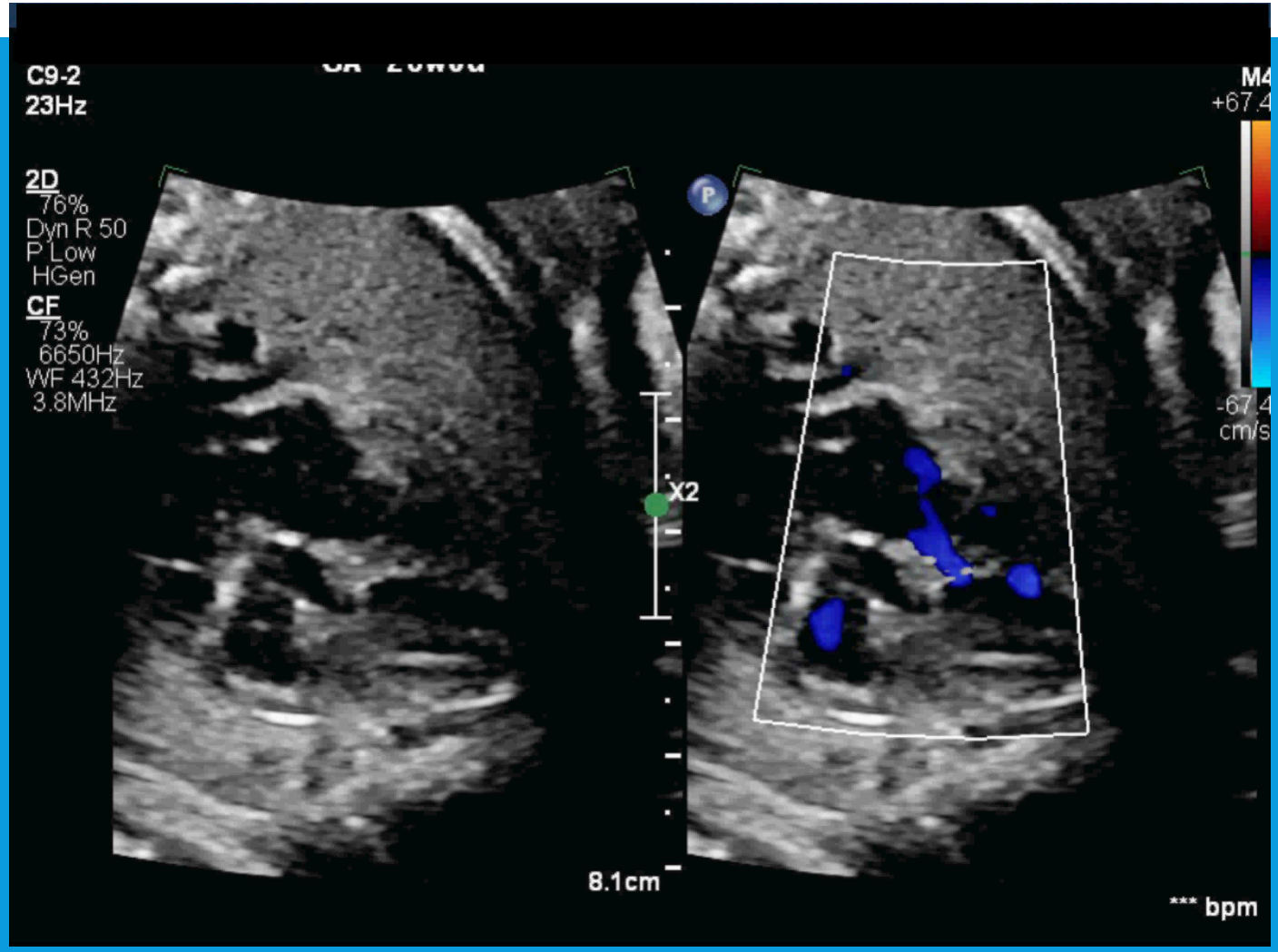

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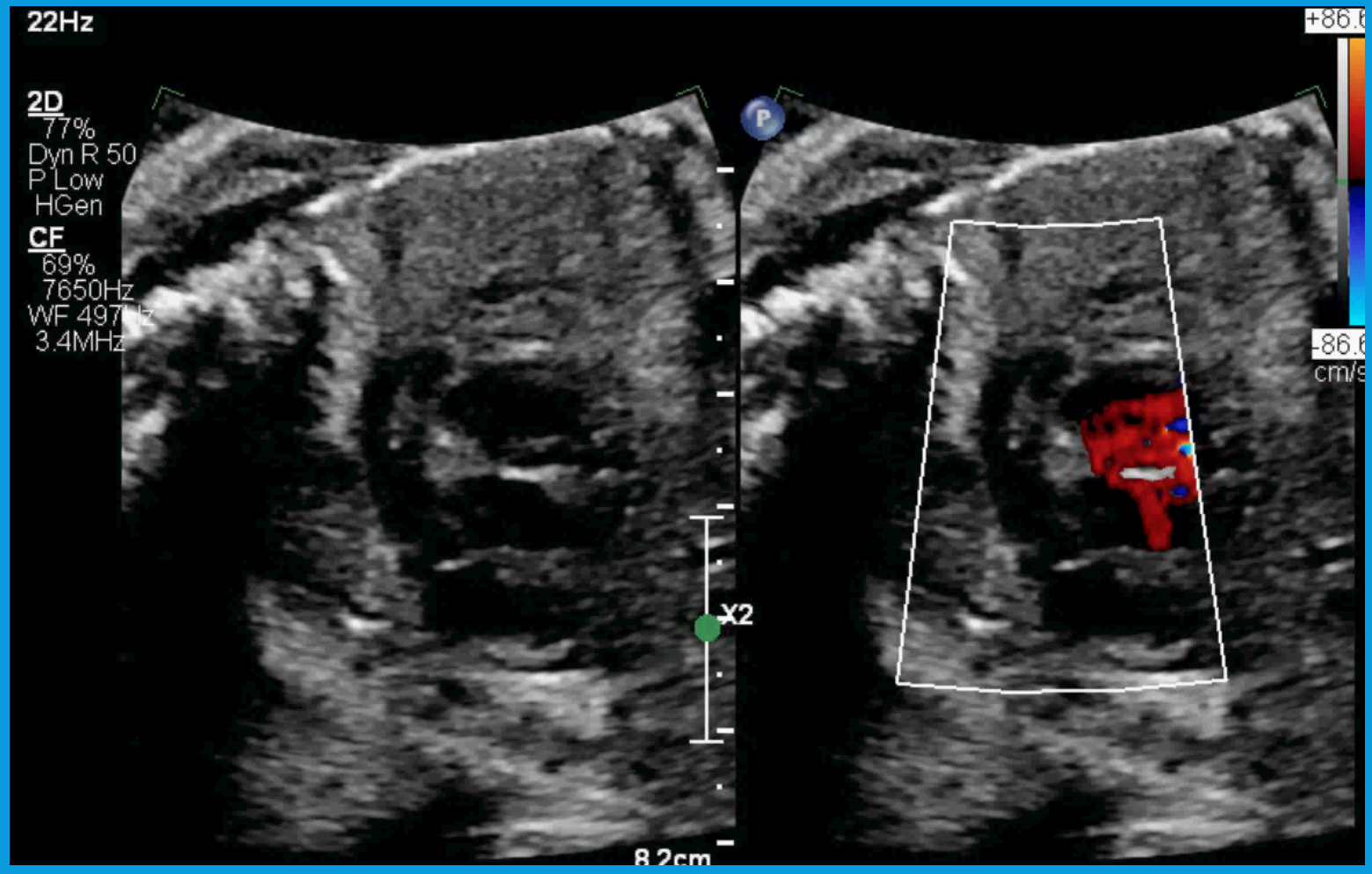
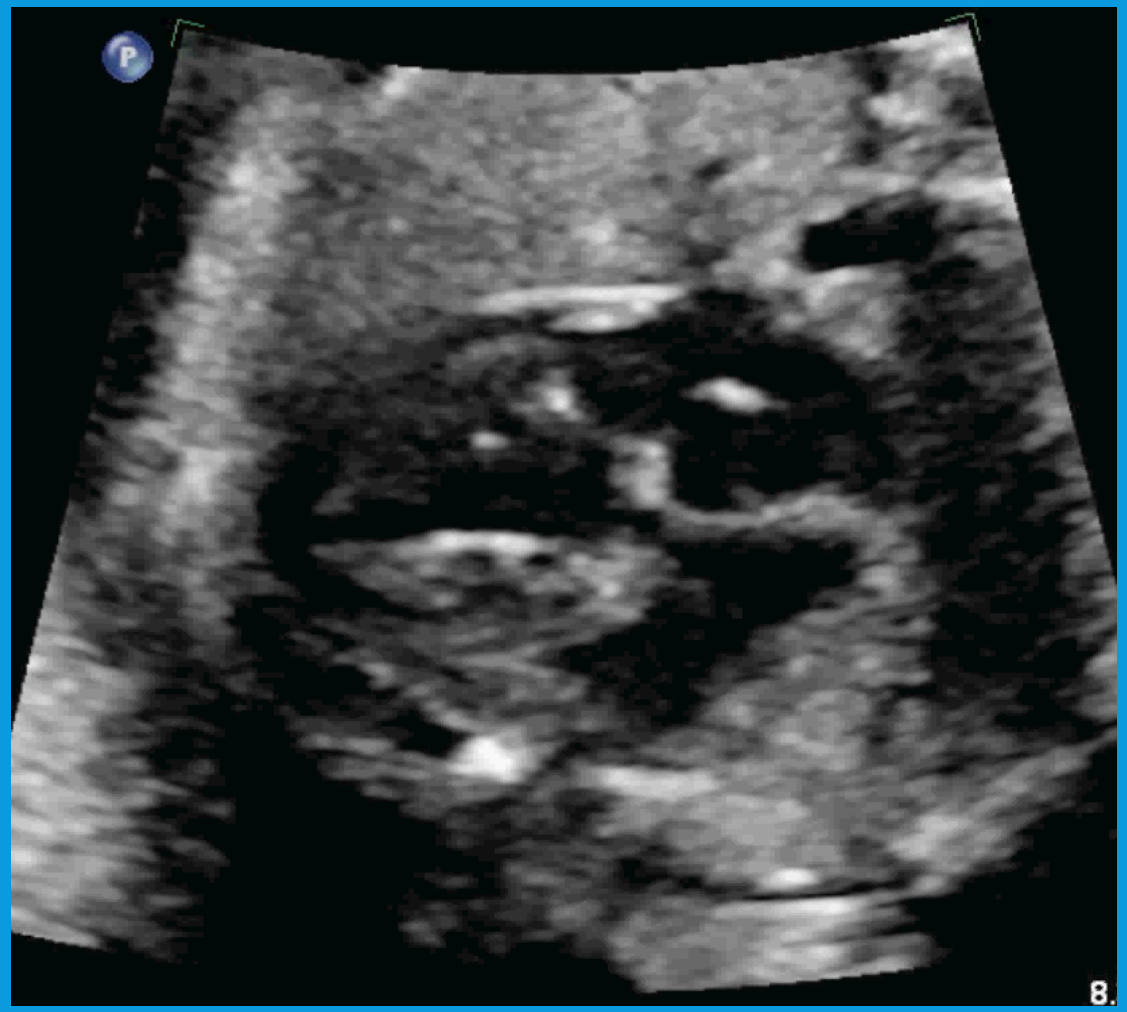


TRICUSPID ATRESIA NORMALLY RELATED GREAT ARTERIES WITH HYPOPLASTIC PV



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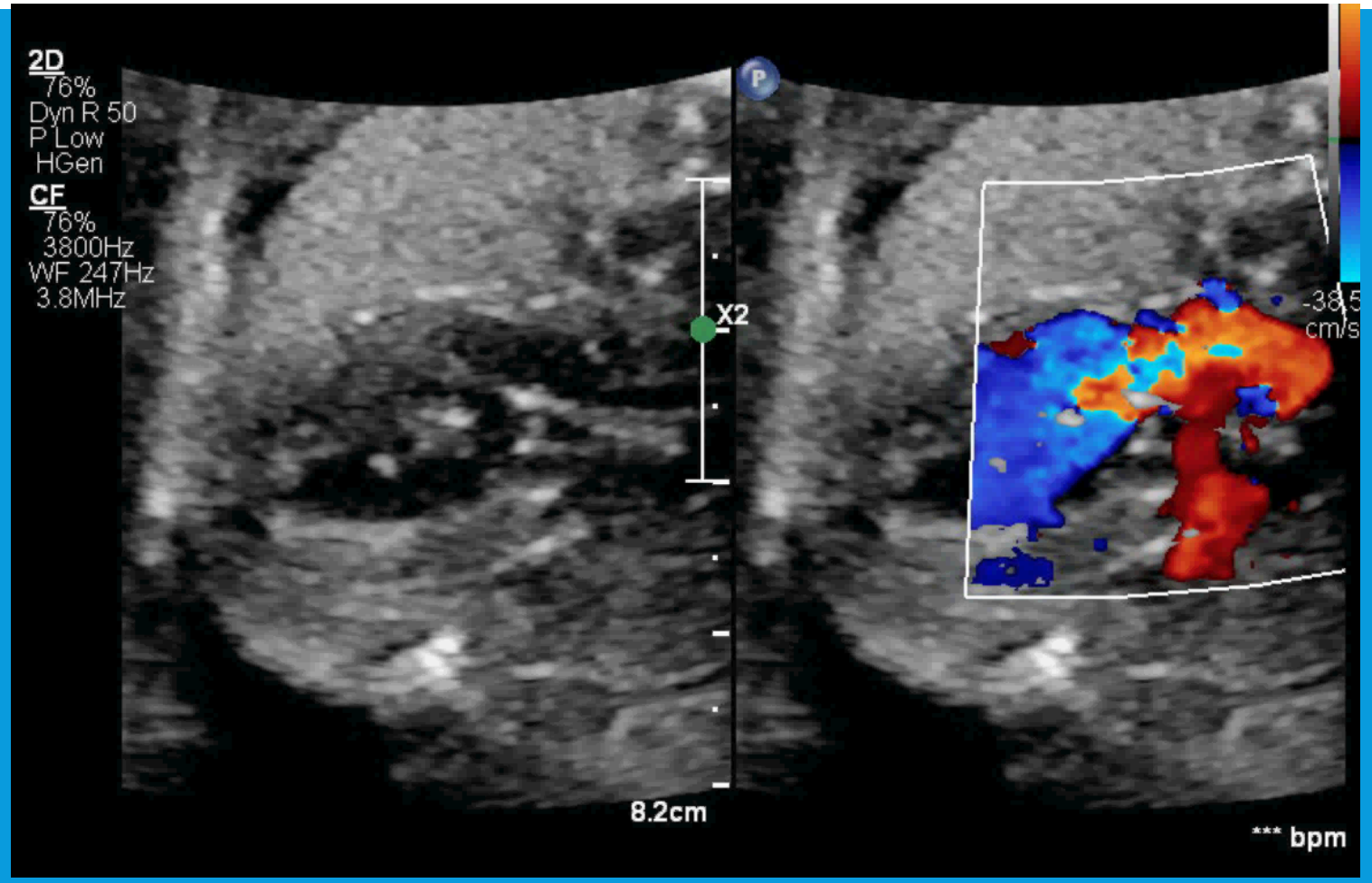

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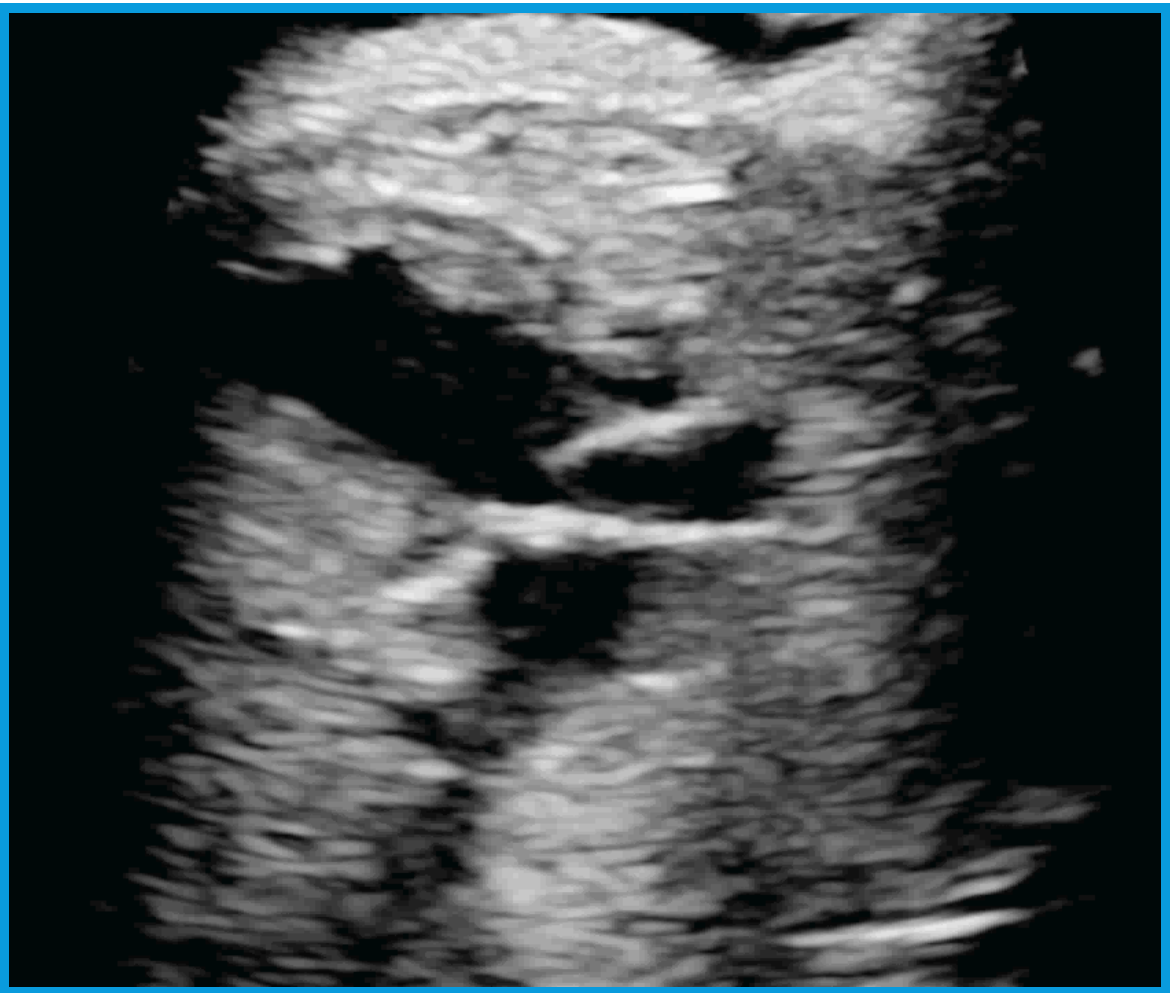

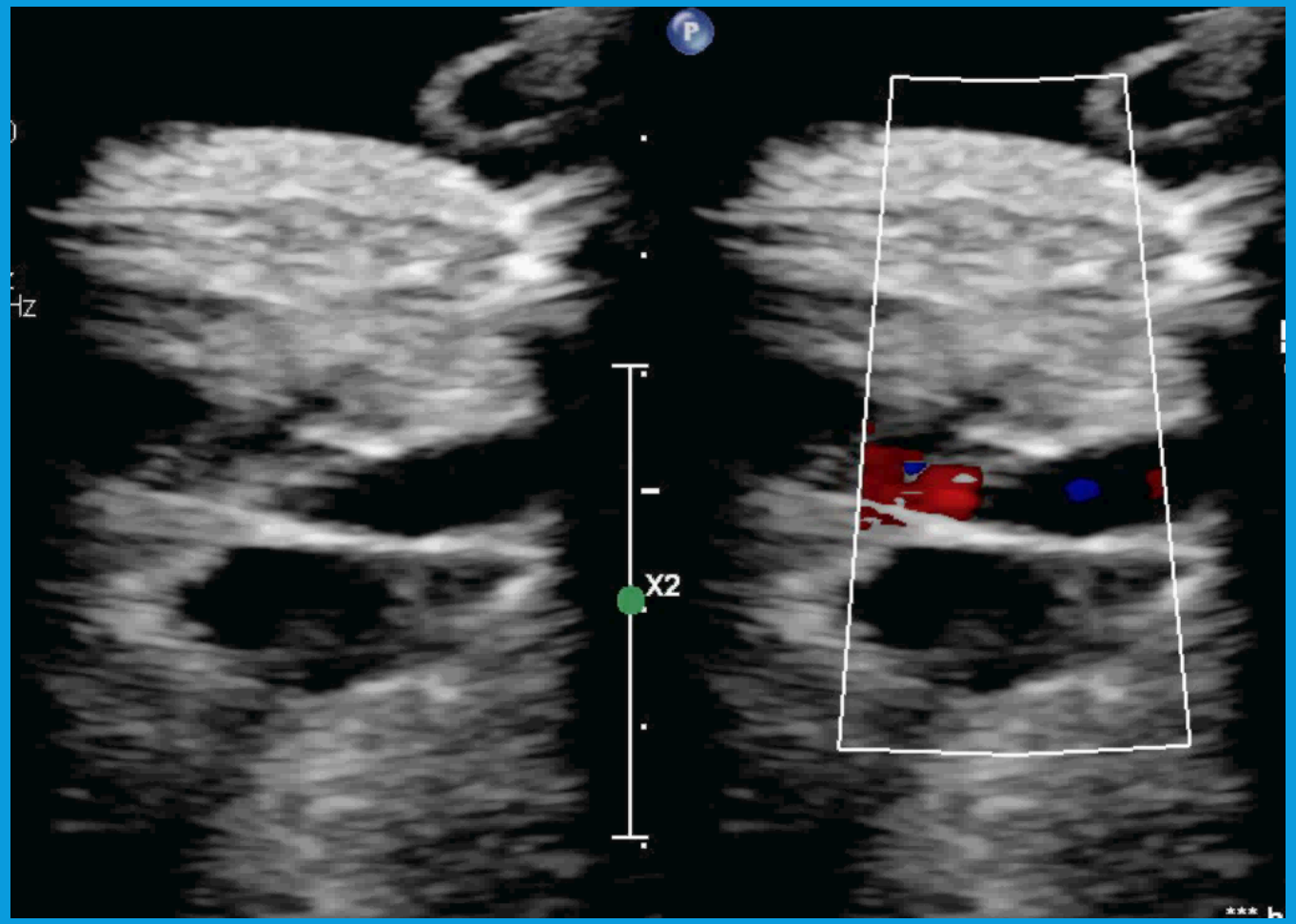

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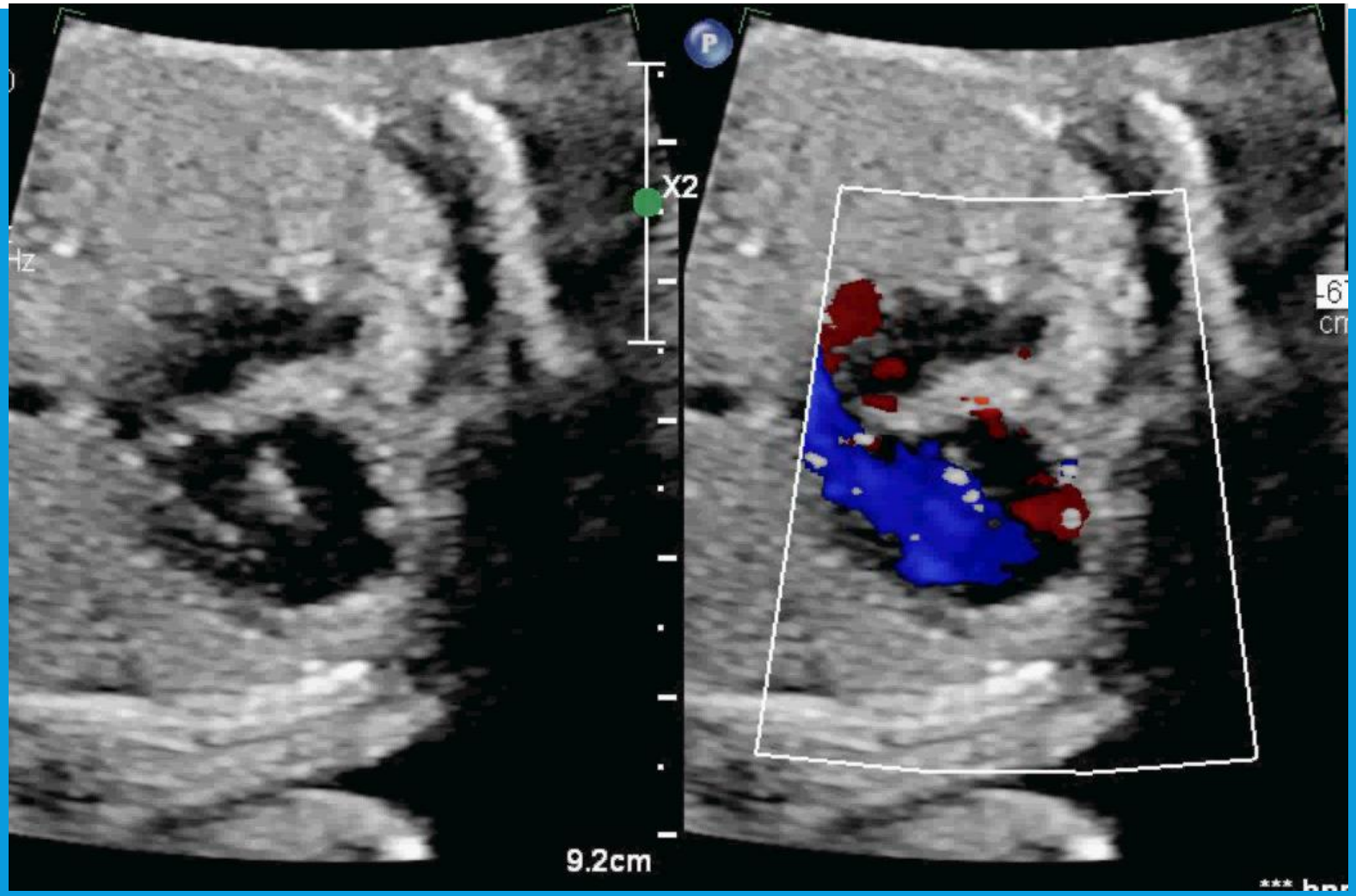

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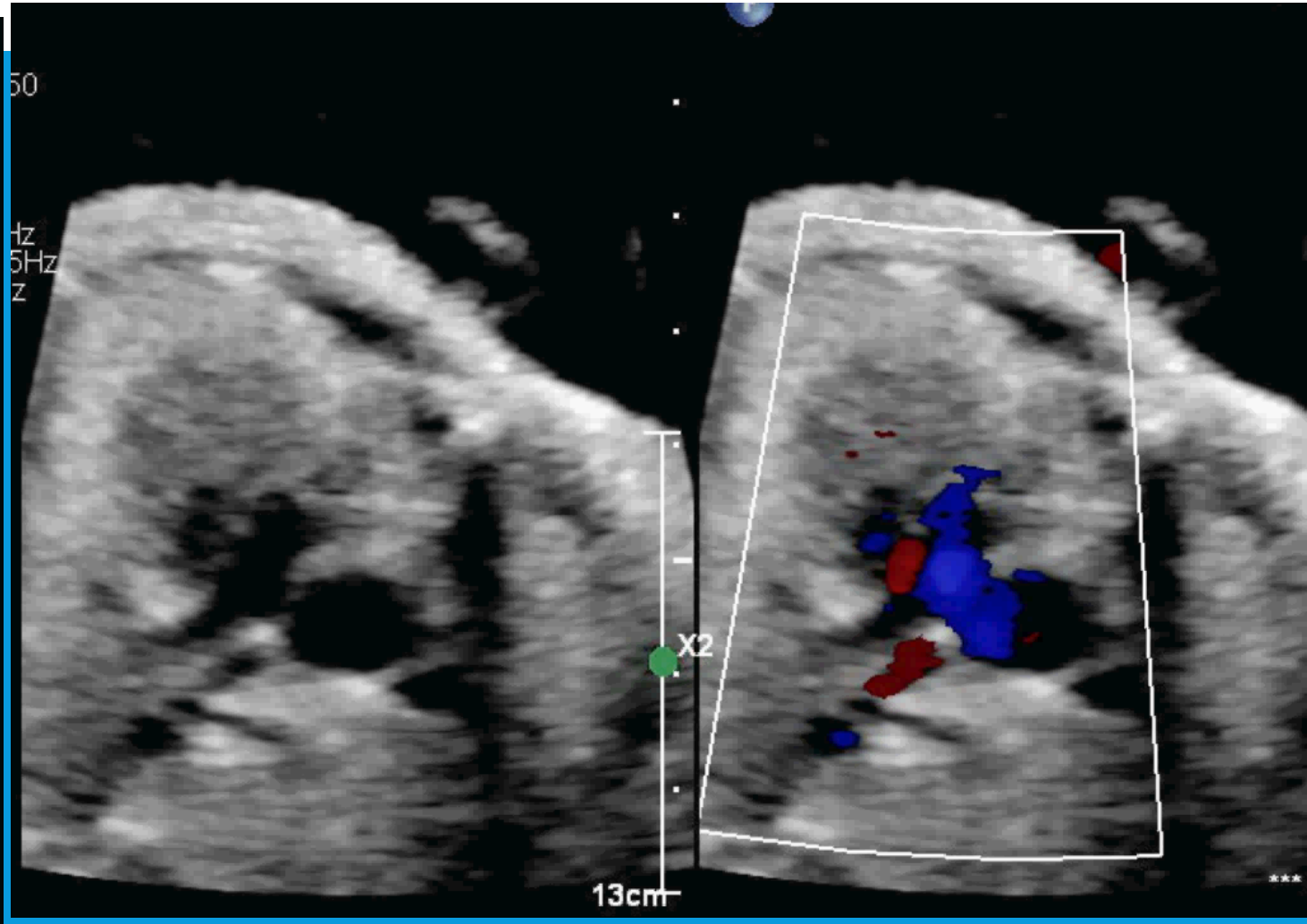
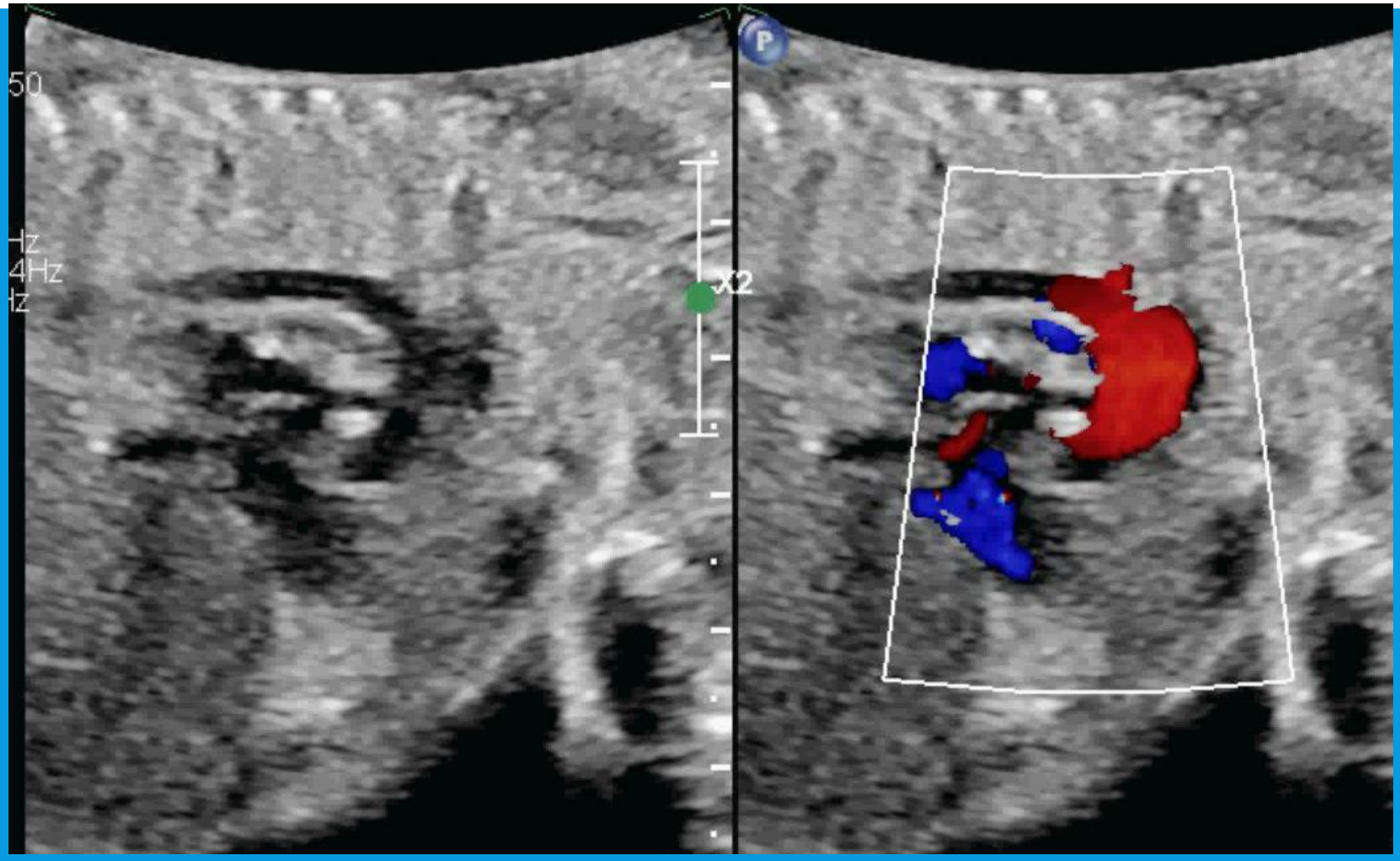
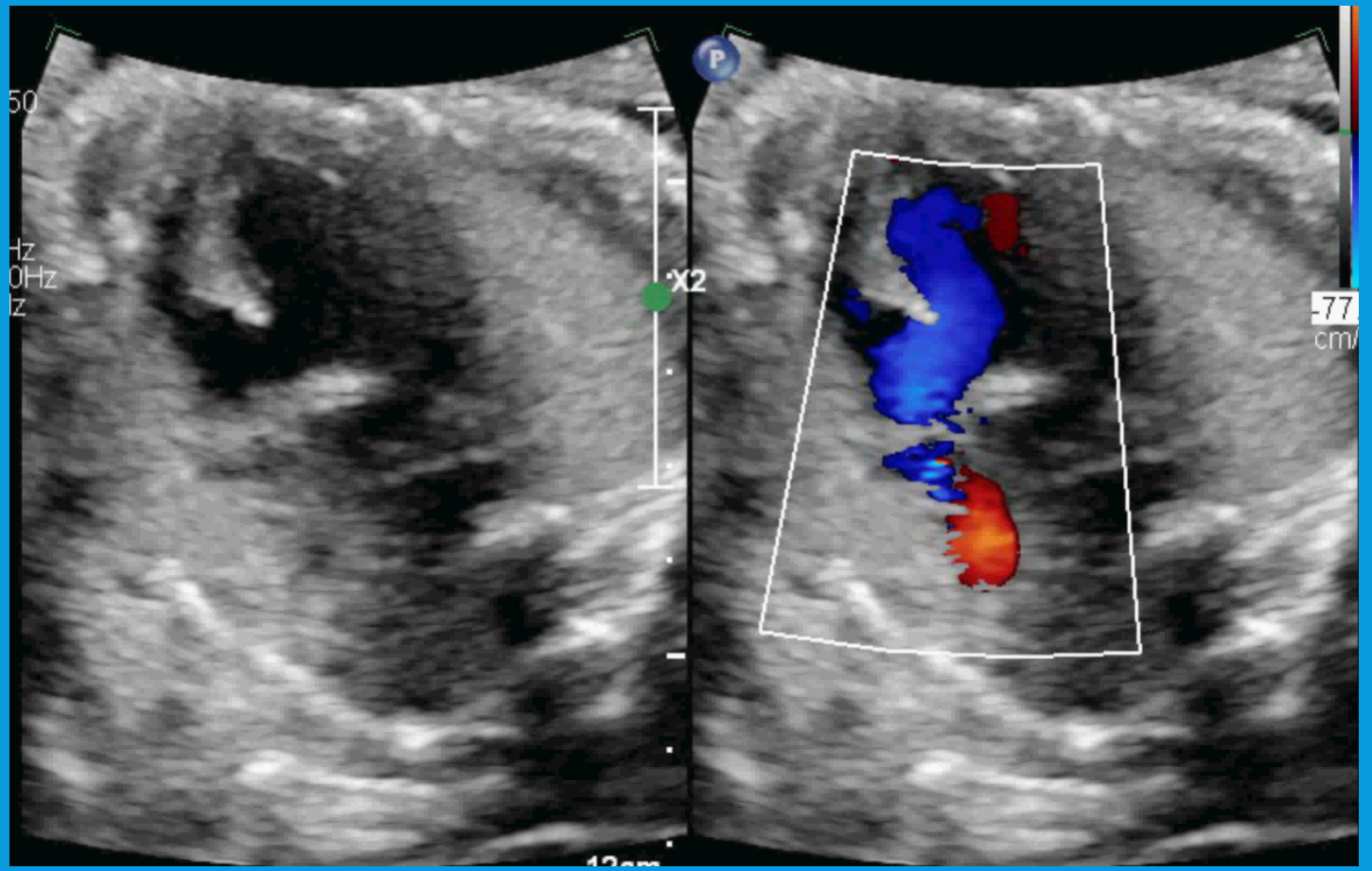


TRICUSPID ATRESIA NORMALLY RELATED GREAT ARTERIES WITH HYPOPLASTIC PV

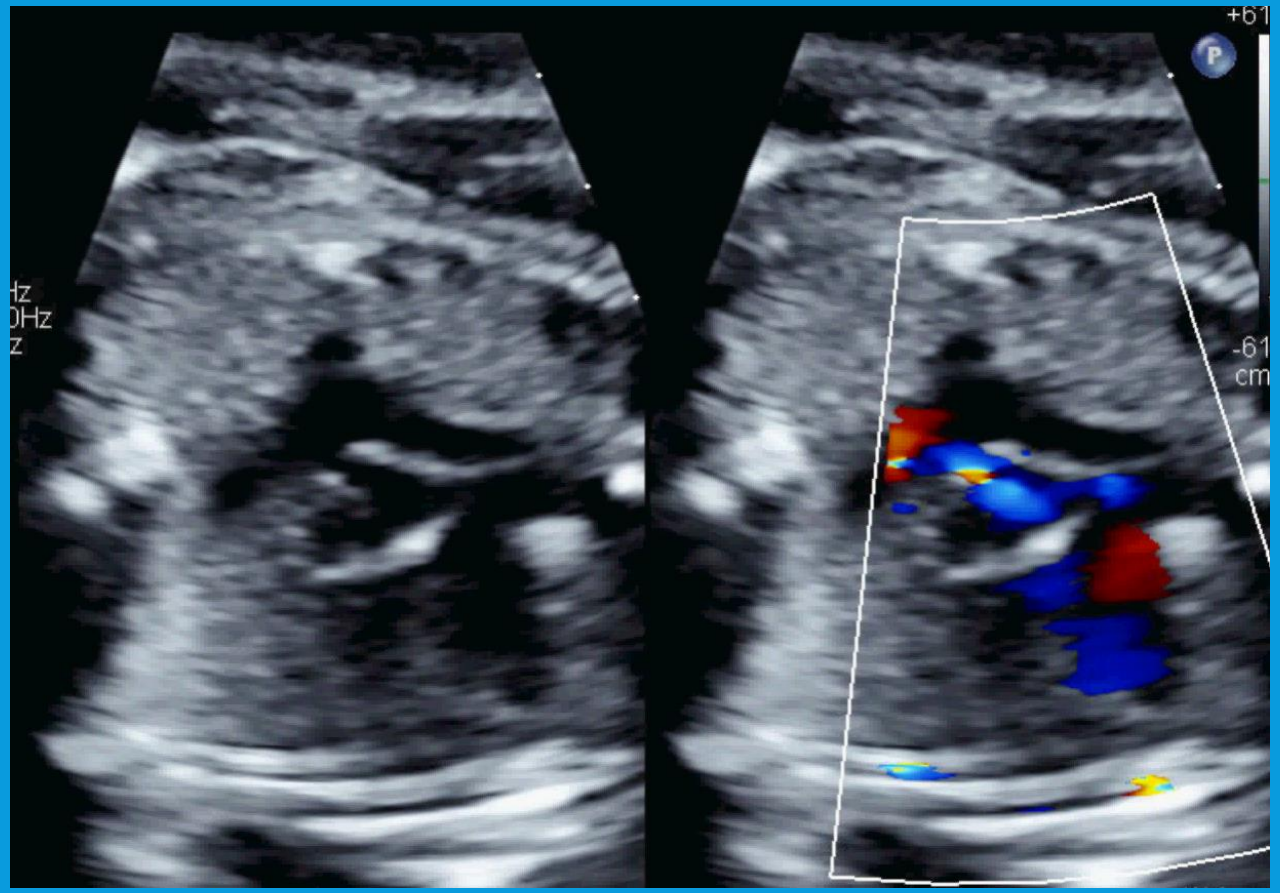


TRICUSPID ATRESIA TYPE 2 WITH D-MALPOSED GREAT ARTERIES

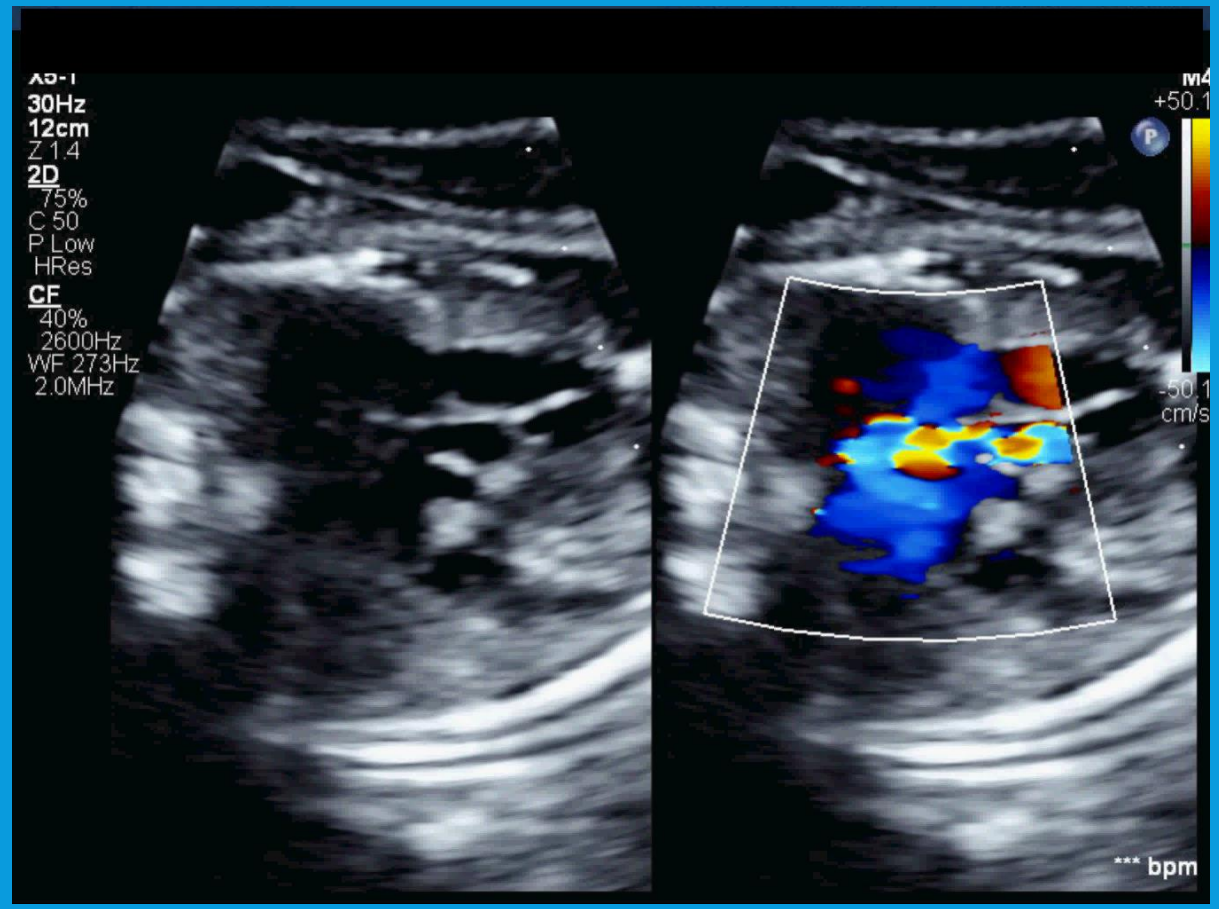
TRICUSPID ATRESIA D-MALPOSED GREAT ARTERIES TYPE 2



TRICUSPID ATRESIA D-MALPOSED GREAT ARTERIES

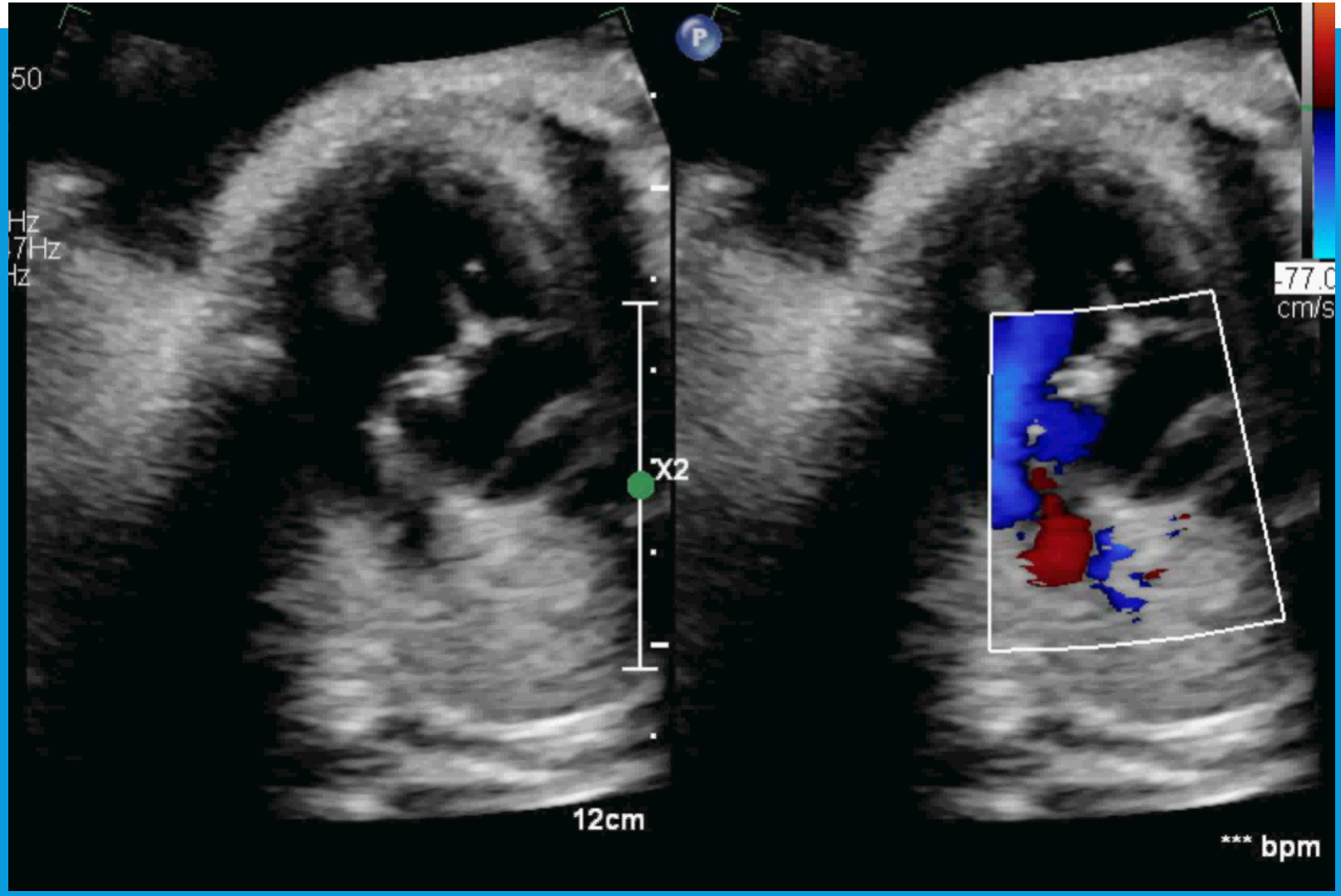


Aortic outflow

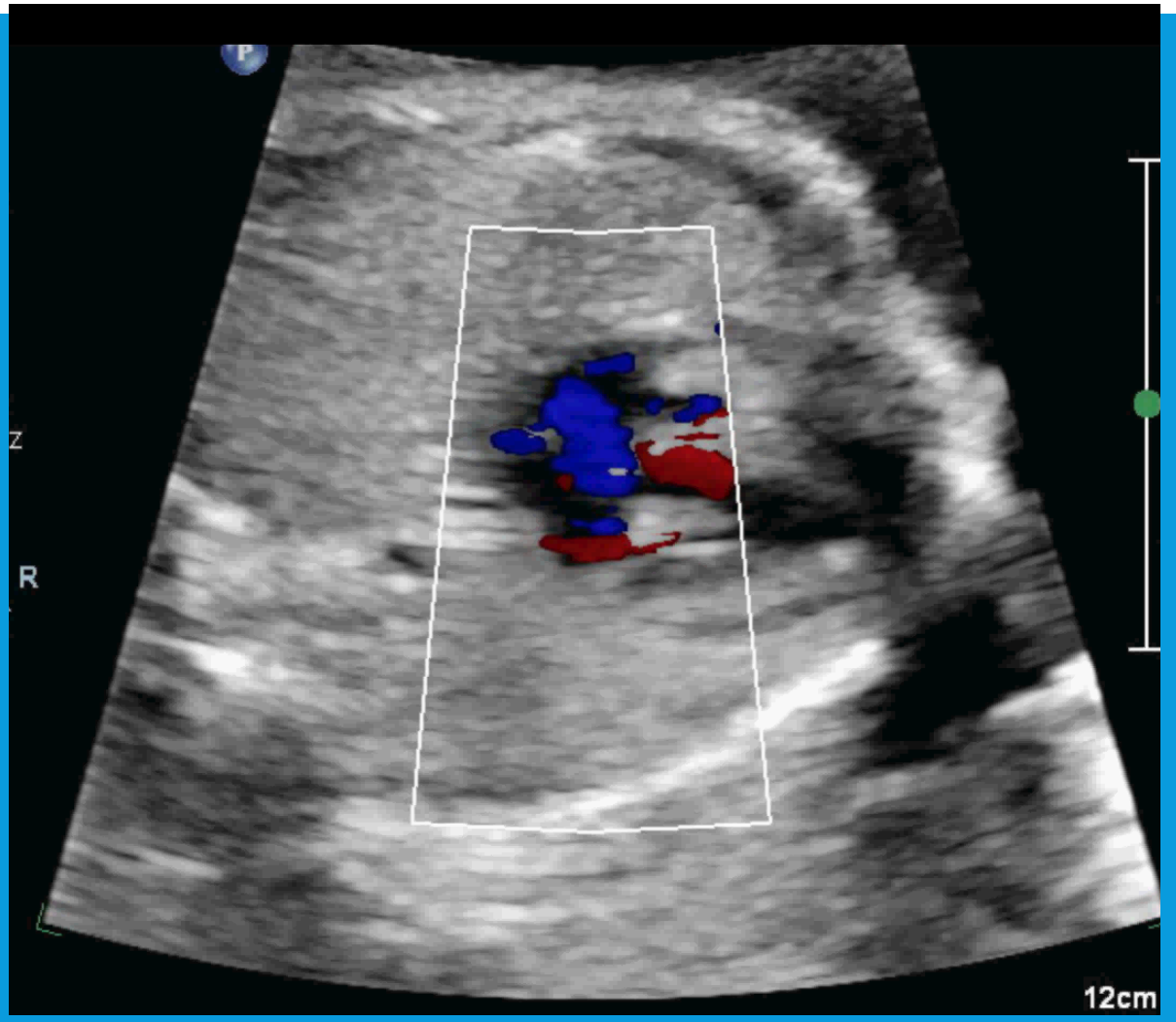


Pulmonary Outflow

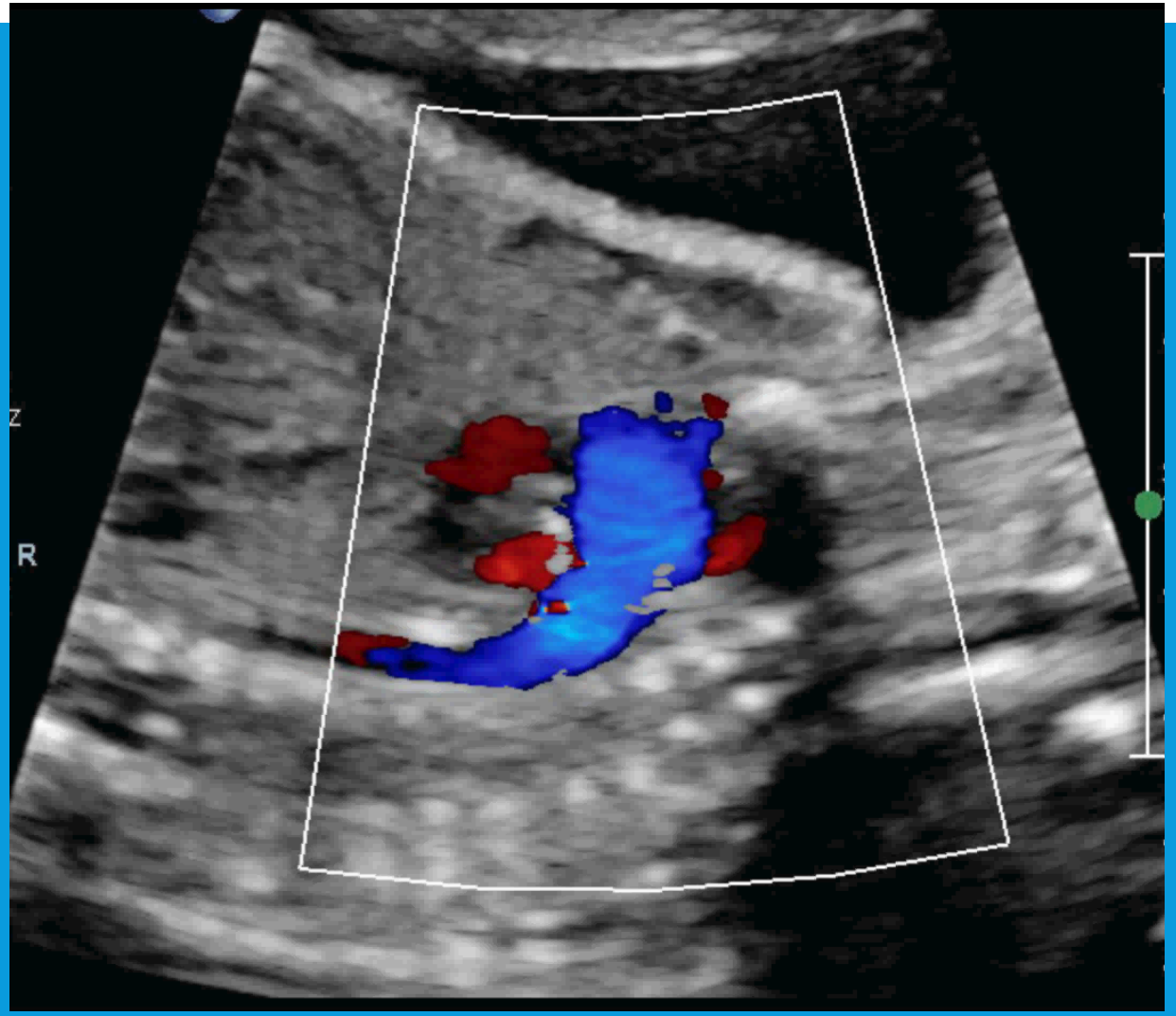
TRICUSPID ATRESIA D-MALPOSED GREAT ARTERIES



TRICUSPID ATRESIA D-MALPOSED GREAT ARTERIES



TRICUSPID ATRESIA D-MALPOSED GREAT ARTERIES



IMAGING TIPS

- Power of the Sweep – a sweep through the heart can rule out multiple heart defects. With tricuspid atresia, we need to sweep through the heart to determine which type of tricuspid atresia we have. This will help determine the route the baby will go for delivery.
- Is there flow across the TV? If not, how is the blood getting out of the heart. Check the atrial septum and VSD for blood flow direction.
- Always look at your outflow tracts, the size and direction of flow determine if the baby needs to be on prostaglandins after birth.

TESTING AND DIAGNOSIS

- Tricuspid atresia can be detected by ultrasound then referred to cardiology for further imaging
- If not picked up prenatally, a baby will show symptoms within the first few hour or days of life.
  - Cyanosis, rapid breathing, poor feeding
- If the patient is diagnosed in a timely manner the baby will have good long-term outcomes.

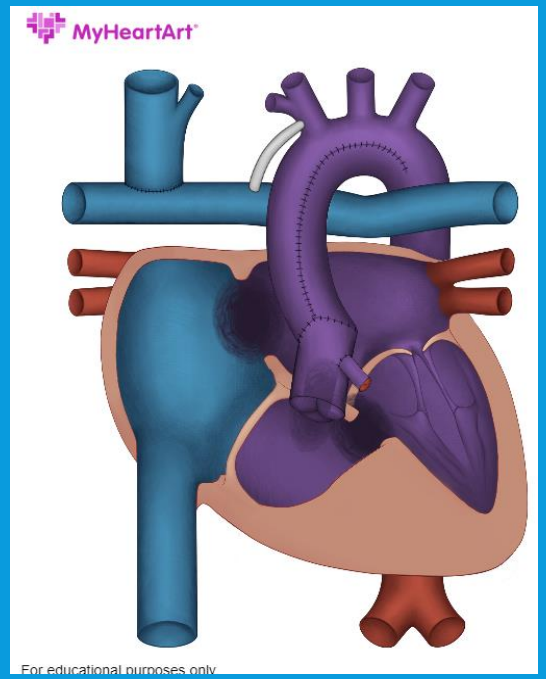
DELIVERY

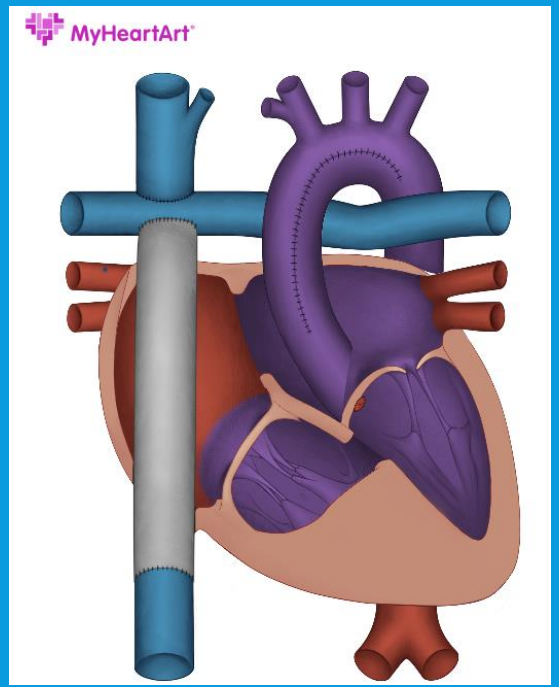
- Before delivery, the mom will have multiple fetal echoes to evaluate the baby's heart and a fetal cardiologist will discuss the findings.
- An integrated consult will be held to discuss the patient and prepare the family for delivery.
- Baby will be delivered at a fetal health center.
- IV lines will be placed immediately after birth.
- Prostaglandins – yes and no. Depends on the great arteries (normal size, prograde flow, transposition of the great arteries)
- Post natal echo will be performed within a few hours of delivery

Surgery

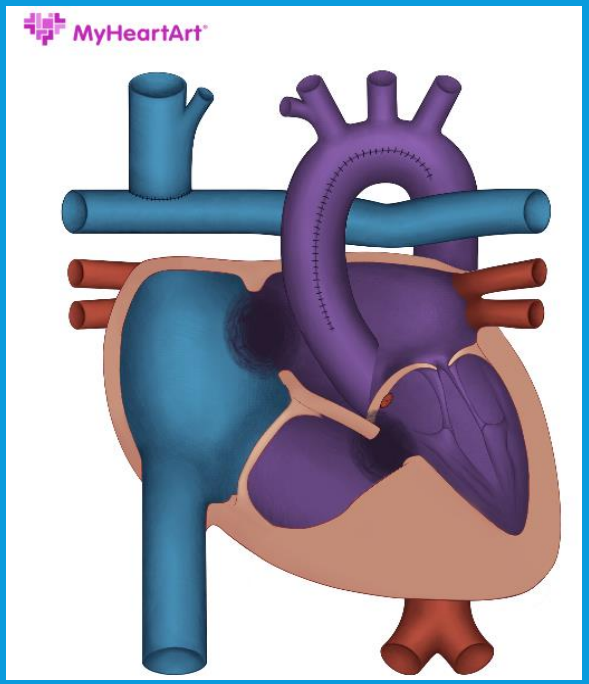
- Without surgical intervention, the baby will not make it past the first year of life.
- Babies with tricuspid atresia will need 2-3 open heart surgeries
  - Blalock Tausig Shunt - first couple weeks of life
  - Glenn – 4-6months of life
  - Fontan – 2 to 4 years of life
  - Fun fact – the first fontan procedure was done on a tricuspid atresia patient

BTT Shunt





Glenn



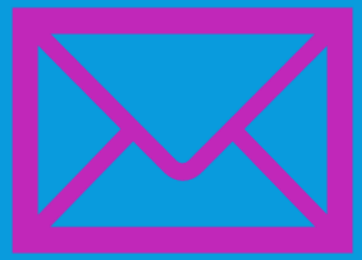
Fontan

SUMMARY

- There are different types of tricuspid atresia that need to be determined prenataly for the baby's best outcome.
- Tricuspid atresia is a single ventricle heart disease that will need surgical attention within the first few days of life or first few months.
- Sweeping through the heart will help determine the type of tricuspid atresia and whether or not if the baby is prostaglandin dependent or not.

QUESTIONS

Contact us if you have questions



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LET'S GO ROYALS and TAYLOR SWIFT'S BOYFRIEND!!

RESOURCES

- Rychik, J. (n.d.). In *Tricuspid Atresia* (pp. 298–302). essay.
- Philadelphia, T. C. H. of. (n.d.). *Tricuspid Atresia*. Children's Hospital of Philadelphia. https://www.chop.edu/conditions-diseases/tricuspid-atresia
- myheartart.org