Panel discussion of complex fetal cases

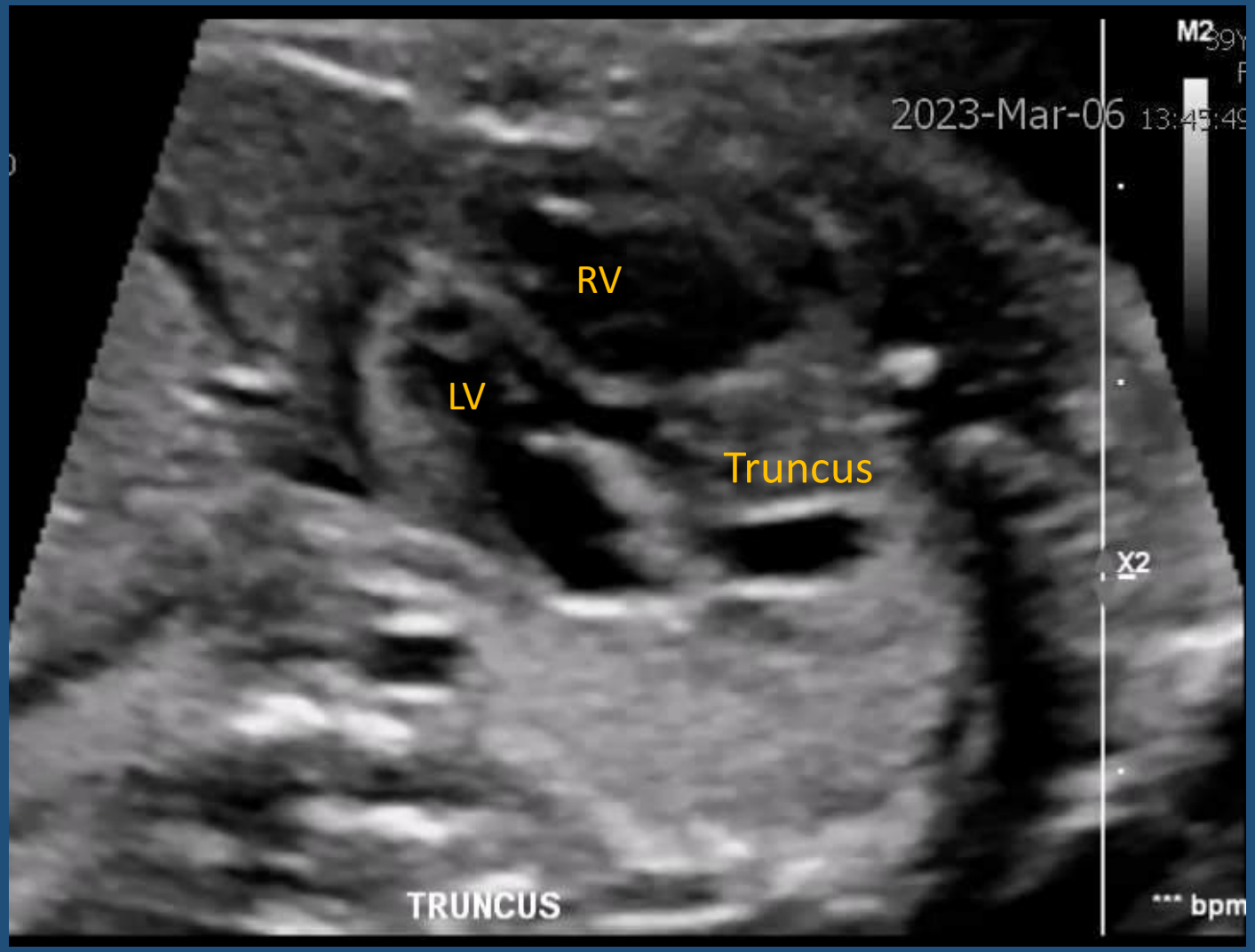
February 18, 2025

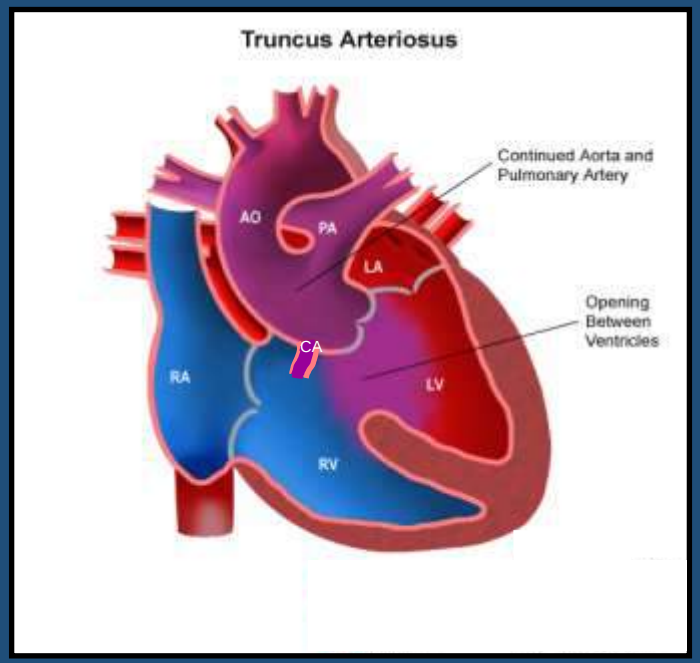
Richard D. Wood Jr. Center for Fetal Diagnosis and Treatment

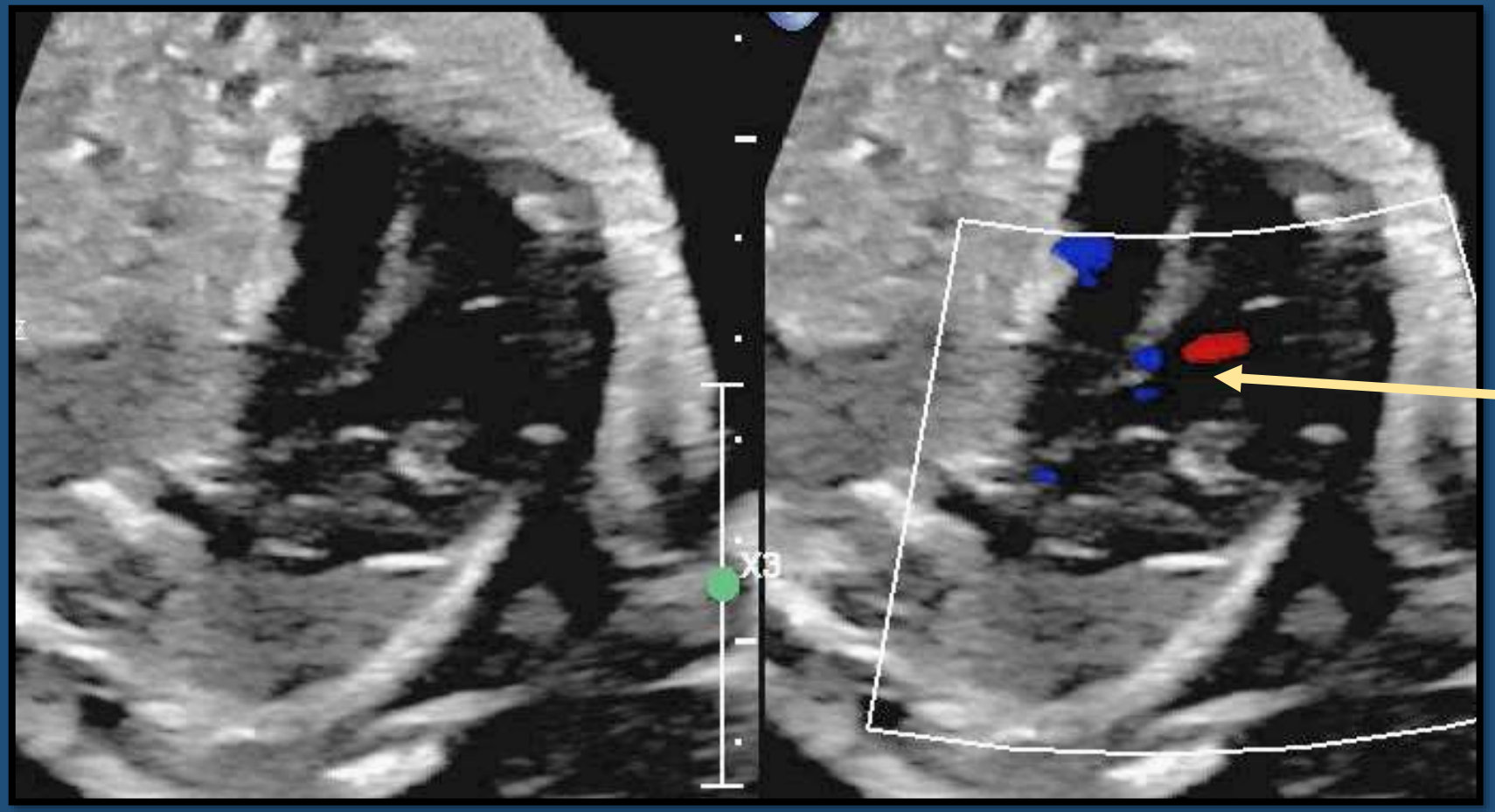
Children's Hospital of Philadelphia
Richard D. Wood Jr.
Center for Fetal Diagnosis & Treatment

Case 1

- 39 yo P3023 at 23 weeks 2 days referred for CHD and CDH
- POBHx:
  - Uncomplicated full-term NSVD x 3; 2nd child has Simpson-Golabi-Behmel syndrome
  - Two chemical pregnancies
- PMH: SVT as a teen
- PSH: None
- Meds: PNV and baby aspirin
- SH: Denies toxic habits; works as a teacher/librarian

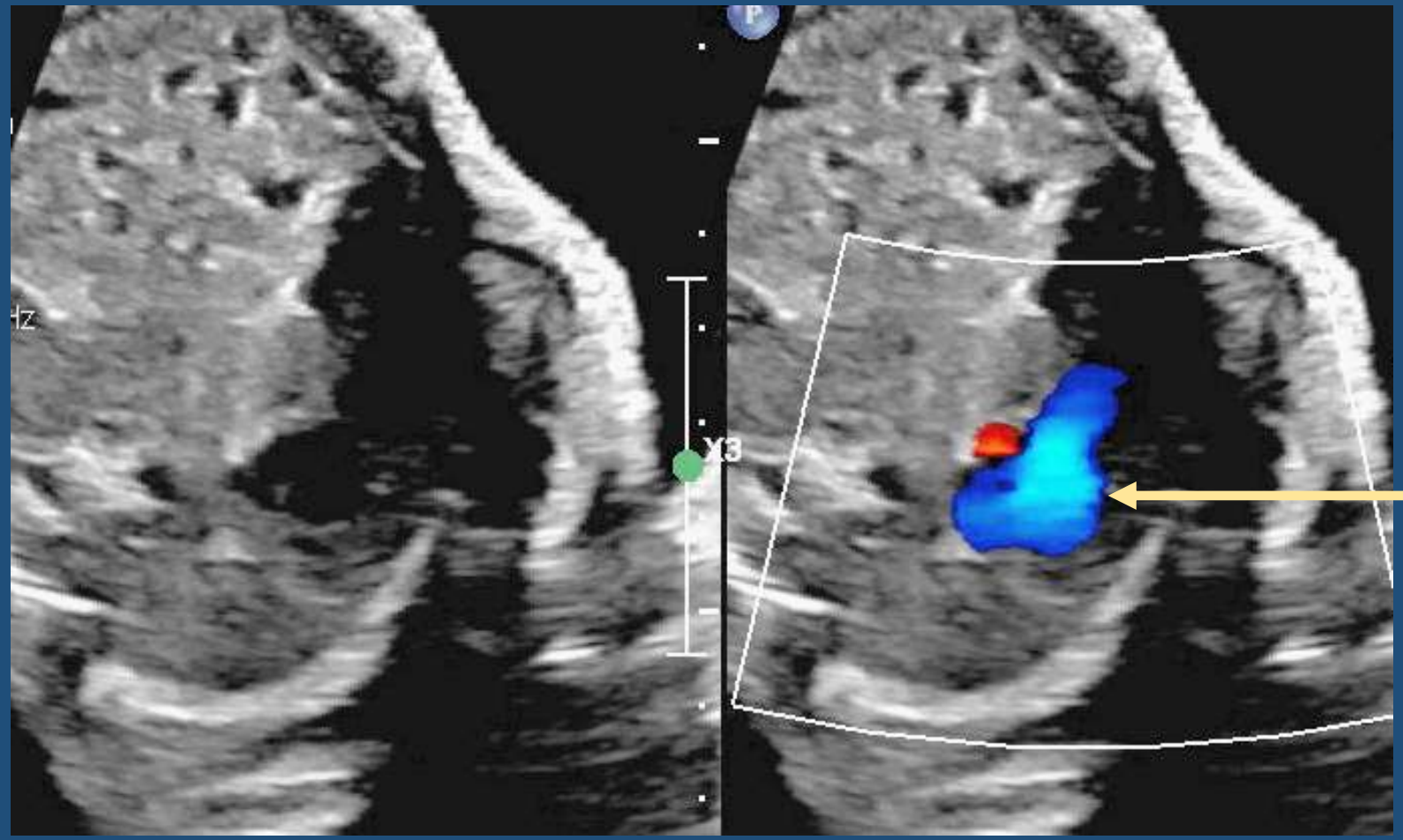

M2
2023-Mar-06
RV
LV
Truncus
X2
TRUNCUS
*** bpm


Truncus Arteriosus
Continued Aorta and Pulmonary Artery
AO
PA
LA
Opening Between Ventricles
CA
RA
LV
RV



Truncal valve

- No stenosis
- No regurgitation

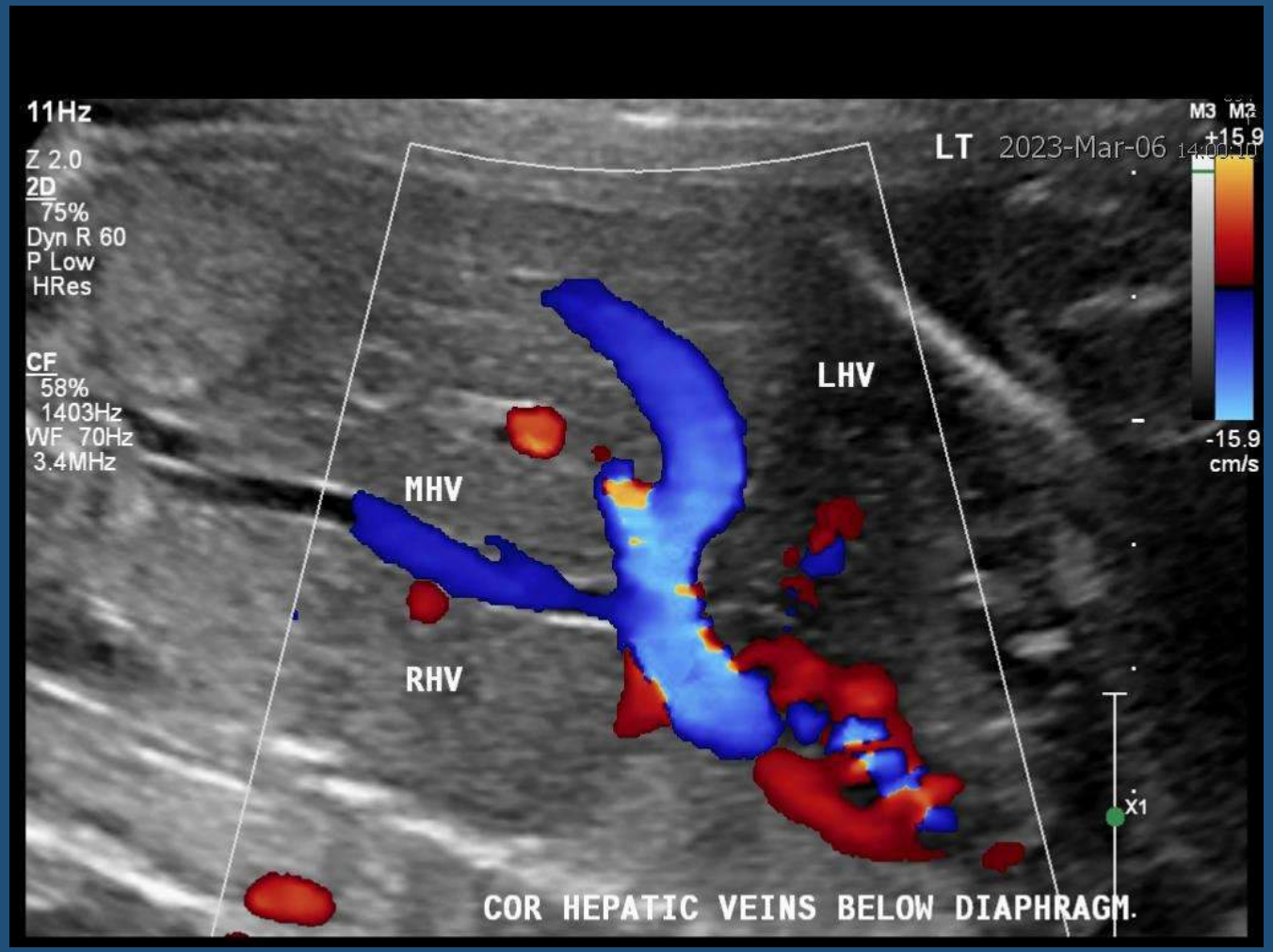


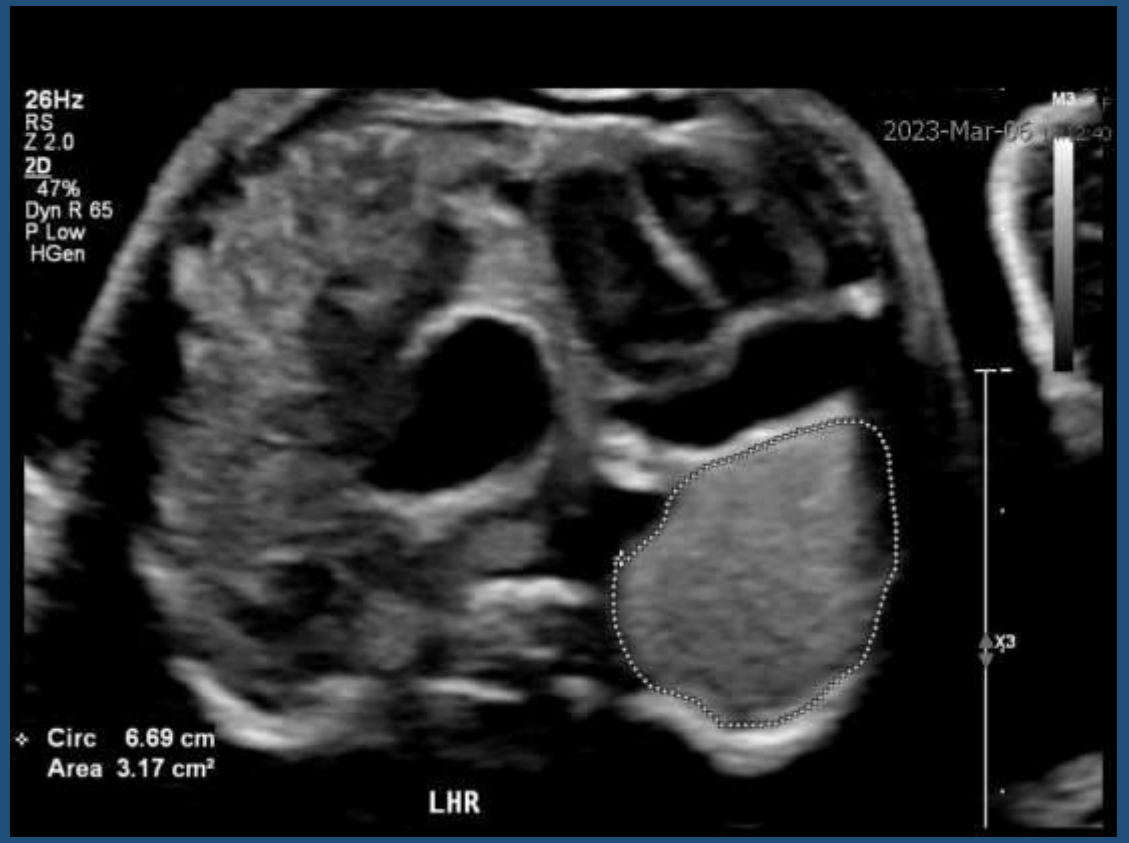
Truncus Arteriosus

- “type A1”
- short Main PA
- confluent PAs


35Hz
R1
Z 2.0
2D
59%
Dyn R 60
P Low
HRes
M3
39Y
F
LT
2023-Mar-06 13:56:40
RT
COR MID DIAPHRAGM
*** bpm


31Hz
R1
Z 2.0
2D
59%
Dyn R 60
P Low
HRes
M3
2023-Mar-06 13:29:21
LHR

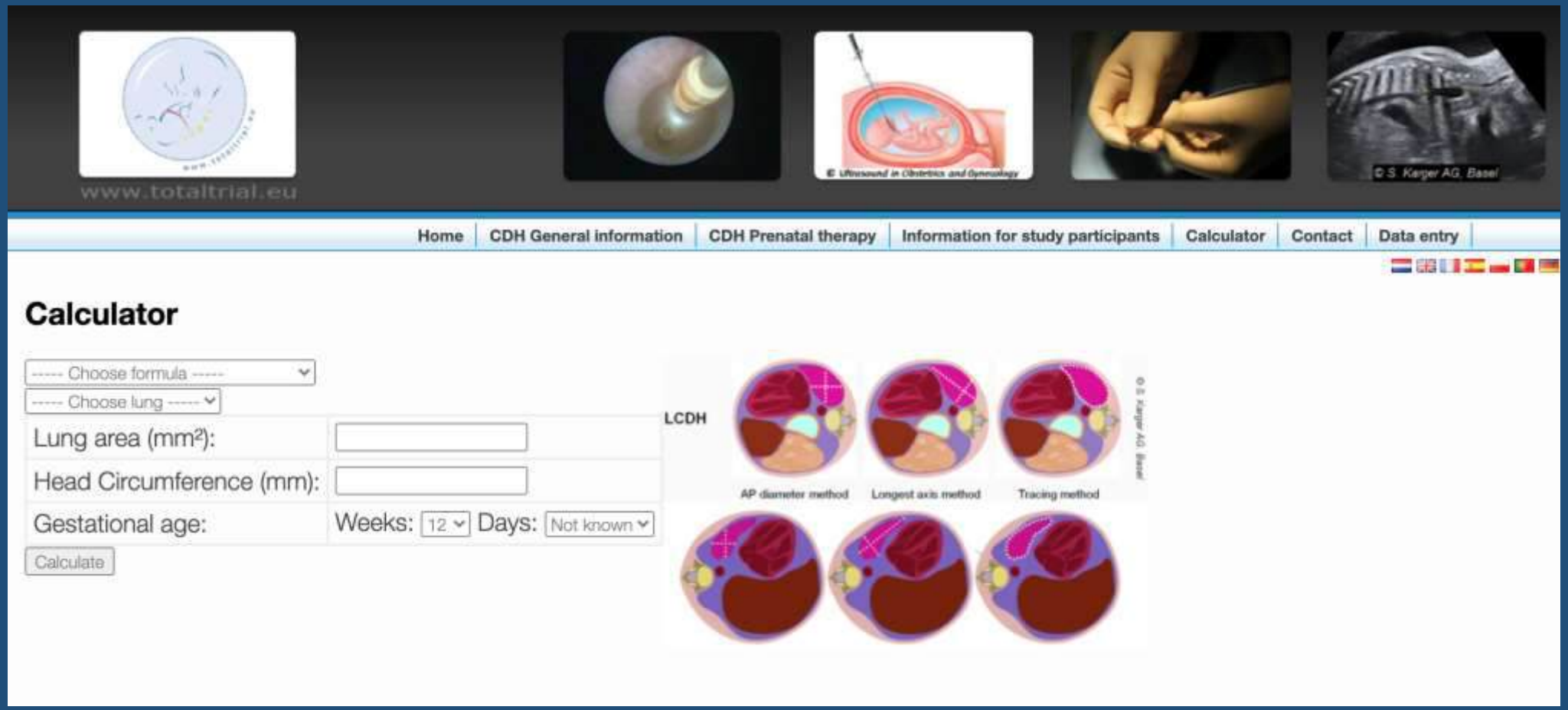

11Hz
Z 2.0
2D
75%
Dyn R 60
P Low
HRes
CF
58%
1403Hz
WF 70Hz
3.4MHz
LT 2023-Mar-06
M3 M2
+15.9
-15.9
cm/s
LHV
MHV
RHV
X1
COR HEPATIC VEINS BELOW DIAPHRAGM.



Lung Head Ratio (LHR)

LHR=Area of contralateral lung/Head circumference = 317/214.3=1.48

Observed/Expected LHR = 62.9%



www.totaltrial.eu/?id=6

Biometry:
Biparietal diameter: 6.1 cm; 24 weeks and 6 days which is within the very upper normal limits at 92 %.

Head circumference: 21.4 cm; 23 weeks and 3 days which is within normal limits at 42 %.

Abdomen circumference: 22.1 cm; 26 weeks and 4 days which is increased at > 98%.

Femur length: 3.9 cm; 22 weeks and 3 days which is within the lower normal limits at
90 15 %.

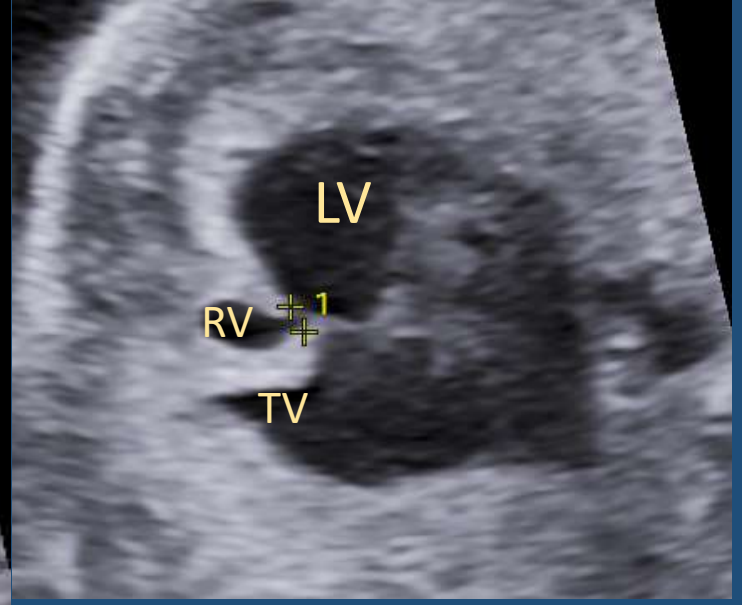
The average ultrasound age is 24 weeks and 3 days, approximately 1 week and 1 day ahead of expected.
HC/AC: 0.97
Estimated fetal weight: 725 g; which is within the very upper normal limits at 96 %.

How would you counsel the expectant parents?

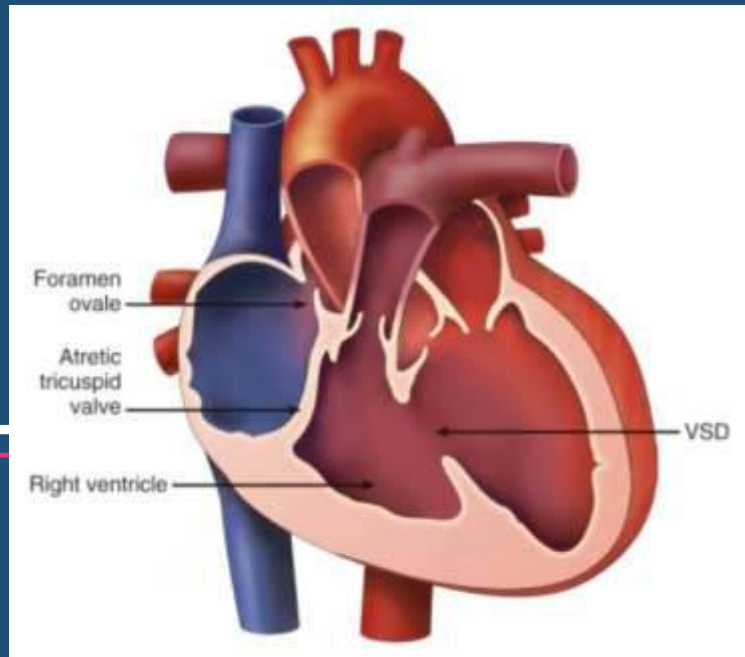
Case 2

- 46 yo P0020 at 25 weeks 0 days referred for CHD
- IVF pregnancy with normal PGT-A
- NT normal in first trimester
- US-indicated cerclage placed at 23 weeks 6 days
- POBHx:
  - Missed abortion
  - Chemical pregnancy
- PMH: Type II DM
- PSH: D&C, egg retrieval, hysteroscopic polypectomy, oral surgery
- Meds: PNV, baby aspirin, and metformin
- SH: Denies toxic habits


TRV
HEART



Tricuspid Atresia, Normally Related Great Arteries



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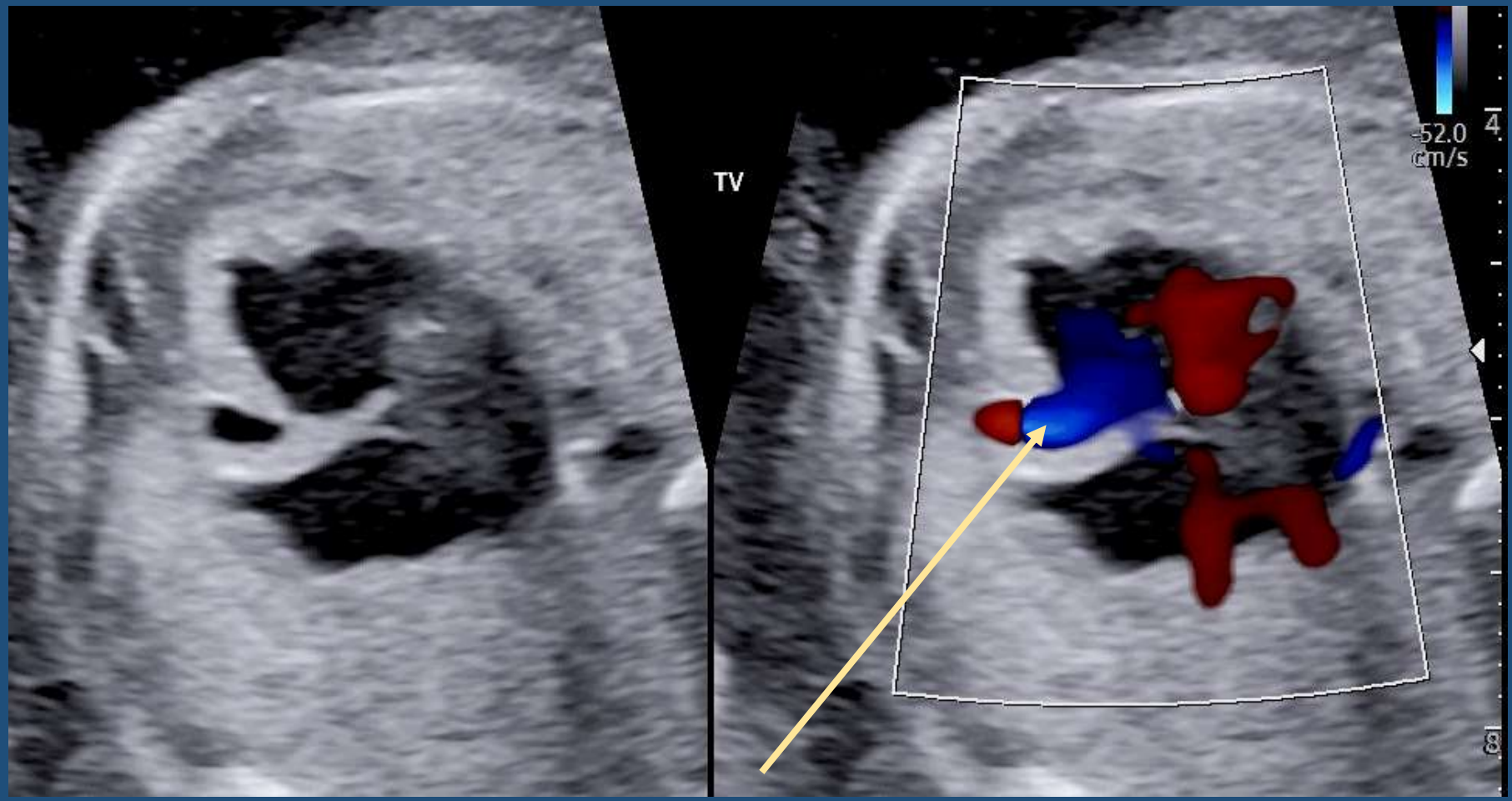
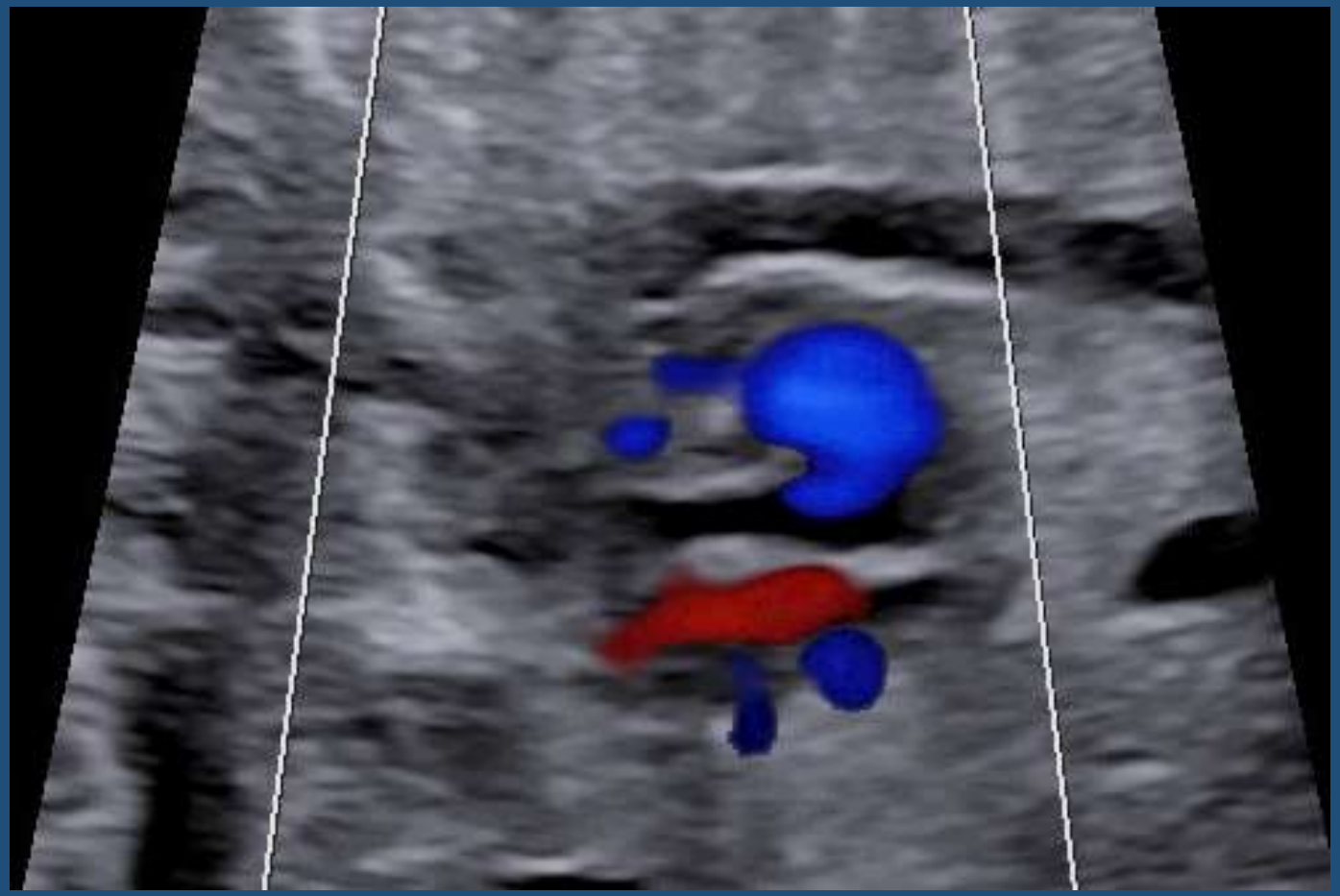
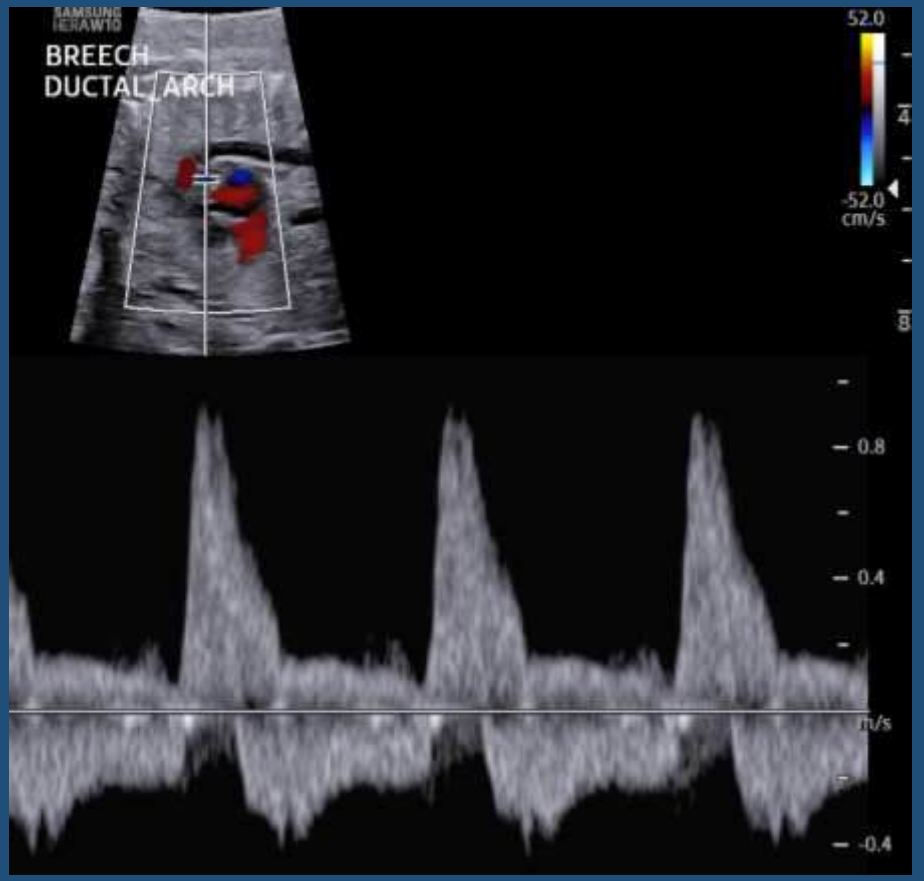


Small VSD

Normally Related Great Arteries
- antegrade flow into pulmonary artery





Flow reversal in Ductus Arteriosus

- suggests pulmonary blood flow will be ductal dependent postnatally

Tricuspid Atresia, Normally Related Great Arteries

- small VSD
- antegrade flow across the pulmonary valve
- flow reversal in the ductus arteriosus

→ ductal dependent pulmonary blood flow

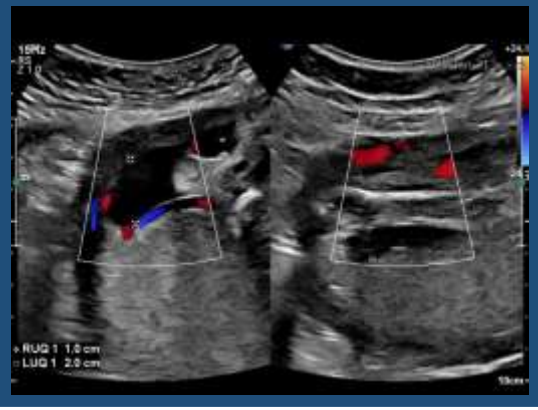

27Hz
RS
2D
48%
Dyn R 56
P Off
HGen
M3
2025-Jan-14
X3
Cerv Length 0.73 cm
4.0cm
SAG CERVIX TV KNOWN CERCLAGE

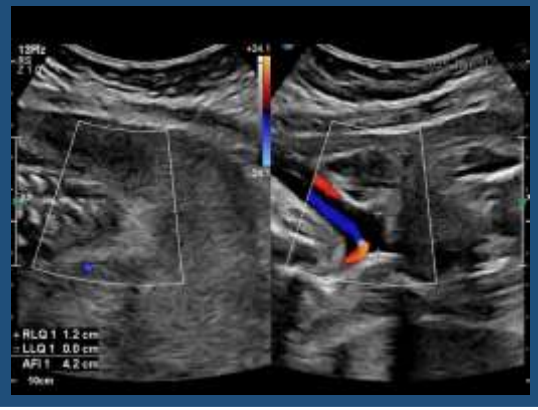

27Hz
RS
2D
48%
Dyn R 56
P Off
HGen
M3
2025-Jan-14
X3
4.0cm
SAG CERVIX TV KNOWN CERCLAGE

PPROM at 25 weeks 5 days at outside hospital

How would you counsel patient now?

- When to transfer?
- When to deliver?
- What are the obstetrical issues to consider?
- What are the postnatal cardiac needs?
- How is cardiac counseling affected by prematurity in this scenario?





Conjoined Twins

- Thoracopagus, Omphalopagus
  75%
- Pygopagus, ischiopagus
  23%
- Craniopagus
  2%

Case #1 NH

- 23yo G3P1, spontaneous conception, 22 weeks GA evaluation
  - Unremarkable medical history, past OB hx
  - Declined genetic testing
- Omphalopagus twins:
  - Shared liver
  - Large shared omphalocele containing liver, small bowel, and the stomach of one of the twins (twin A)
  - Possible shared bowel cannot be excluded.
  - No shared cardiac structures.


A
B
X
JPEG

Prenatal counseling

- Delivery Planning
- Separation suitability
- Timing of separation
- Mortality/Morbidity Risks

Thoraco-Omphalopagus Conjoined Twins

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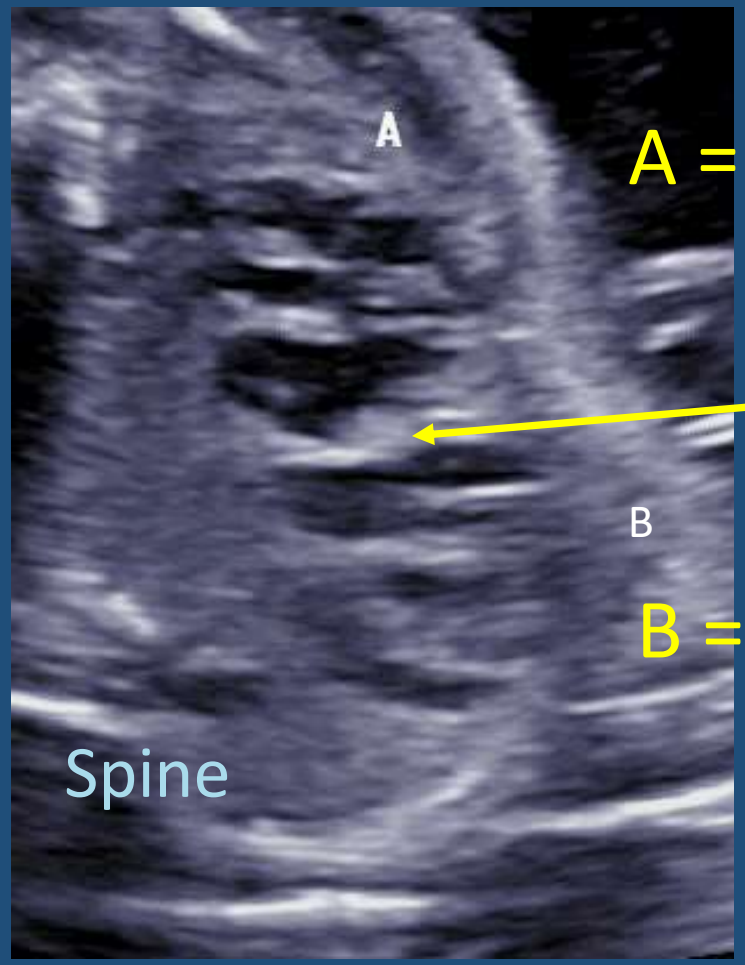
Case #2 JH

- 27yo G2P1, spontaneous conception, 22 weeks GA
  - Noncontributory medical/ OB history
  - Declined genetic testingz
- Dithoracic parapagus conjoined twins:
  - Spines that are in extremely close proximity with abutting spinal canals
  - Shared liver and bowel with separate gallbladders in close proximity
  - Renal tissue in both paraspinal regions with color Doppler demonstrating main renal vessels bilaterally without four distinct kidneys
  - Tribrachias with a total of 3 upper extremities, 2 within the range of normal and a third upper extremity with a radial ray malformation including a foreshortened humerus, a single bone in the forearm with a single digit with persistent sharp flexion at the wrist and elbow
  - Low conus medullaris in both fetuses
  - Two lower extremities with bilateral talipes

Case #2 JH

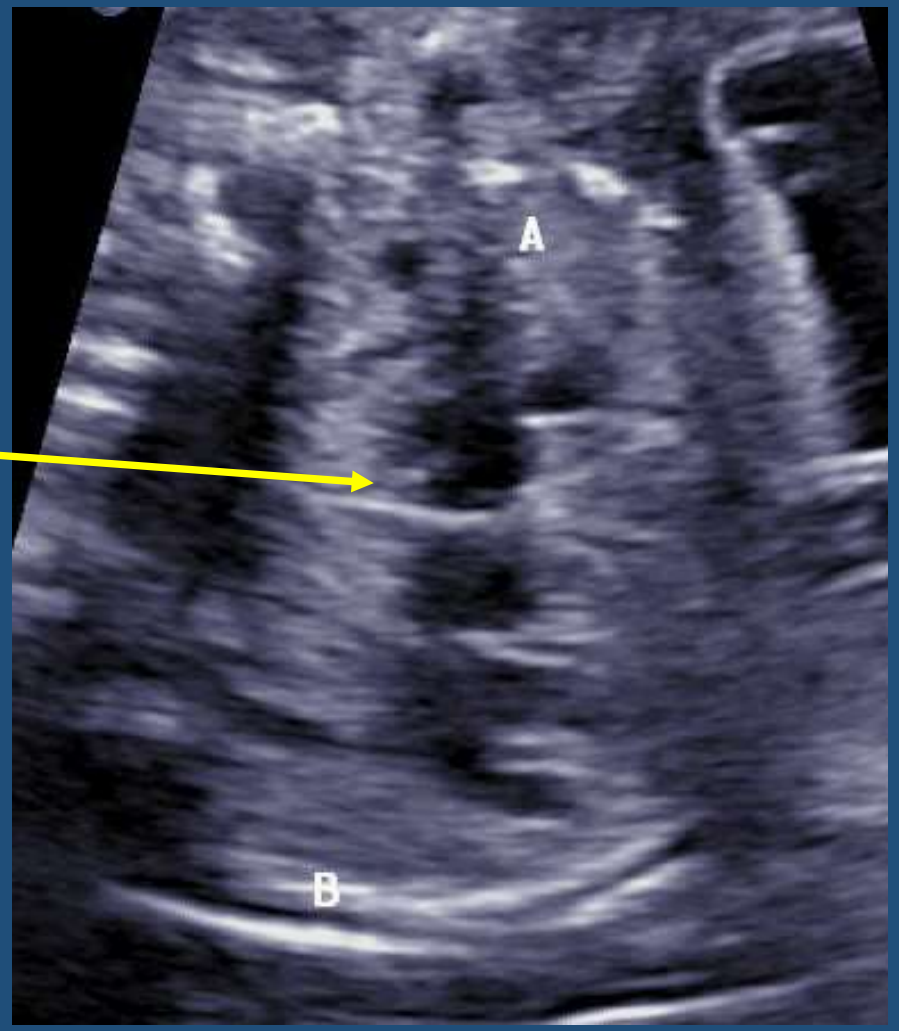
- **Twin A:**
  - Complete AV canal, balanced
  - Double outlet right ventricle, Pulmonary Stenosis (moderate)
    - Bidirectional shunting in PDA
  - TAPVR to SVC
  - Normal aortic arch
  - No IVC detected
  - Possible shared atrial wall with Twin B
- **Twin B:**
  - IVC to RA identified, no SVC clearly identified
  - Otherwise normal intracardiac anatomy, function, rhythm

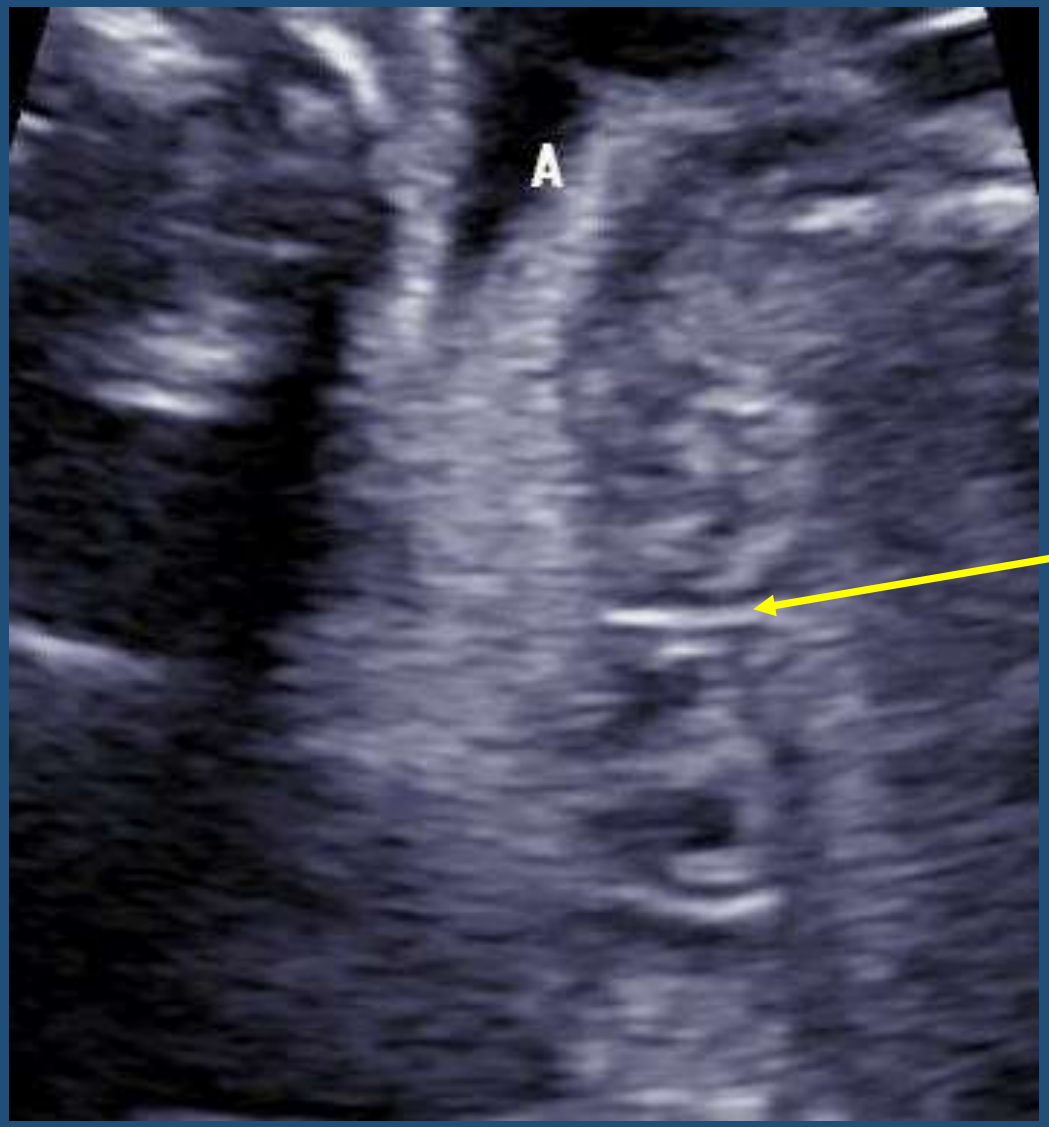


A = CAVC, DORV

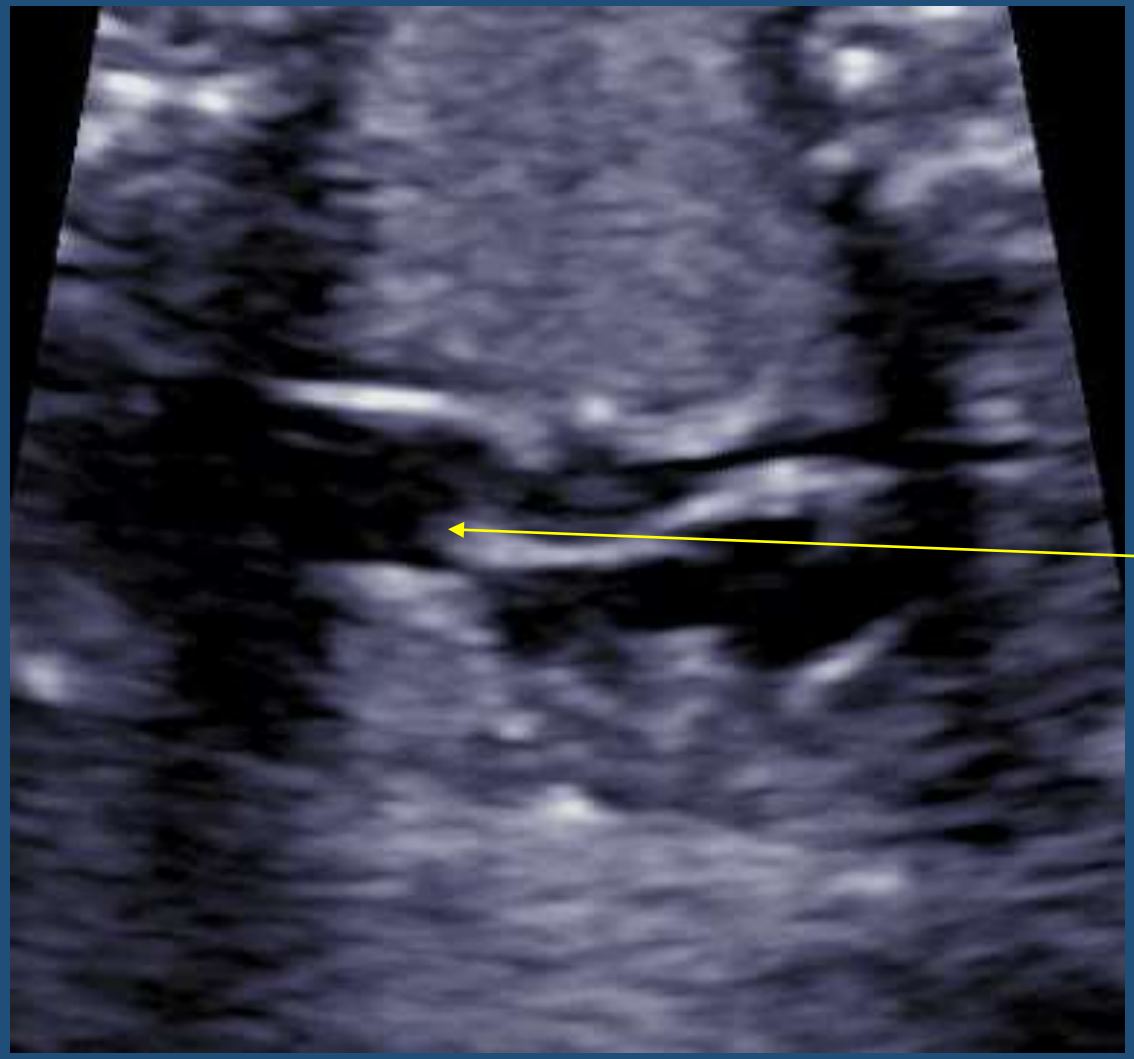
Shared atrial wall

B = Normal heart





LV walls adjacent, but not interconnected



Twin A = CAVC

dilated SVC

Prenatal counseling?

Case #3 VA

- Prenatal diagnosis of thoraco-omphalopagus twins
  - shared pericardium (?myocardium) and liver
  - Twin A with CHD and cleft palate

- **Twin A**: Tetralogy of Fallot w/ moderate PS and RVOT obstruction, large secundum ASD, bilateral SVCs without bridging vein, L-SVC and L-IVC to coronary sinus, left aortic arch w/ aberrant right subclavian
- **Twin B**: Superior/inferior ventricles, otherwise no congenital heart disease

- Prenatal Genetic testing → mosaic Turner's syndrome

Case #3 VA

- Prenatal diagnosis of thoraco-omphalopagus twins
  - shared pericardium (?myocardium) and liver
  - Twin A with CHD and cleft palate

- **Twin A**: Tetralogy of Fallot w/ moderate PS and RVOT obstruction, large secundum ASD, bilateral SVCs without bridging vein, L-SVC and L-IVC to coronary sinus, left aortic arch w/ aberrant right subclavian
- **Twin B**: Superior/inferior ventricles, otherwise no congenital heart disease

- Prenatal Genetic testing → mosaic Turner's syndrome

Prenatal counseling

- What are the anatomic considerations for separation?
  - *Stabilization*
  - *Timing*
- When can the cleft palate be repaired
- How can the heart defect in in Twin A be managed?
  - *What are the implications of a shared pericardium? Shared myocardium?*
- Implications of not separating?
- Mortality risk?

Case #4 SC

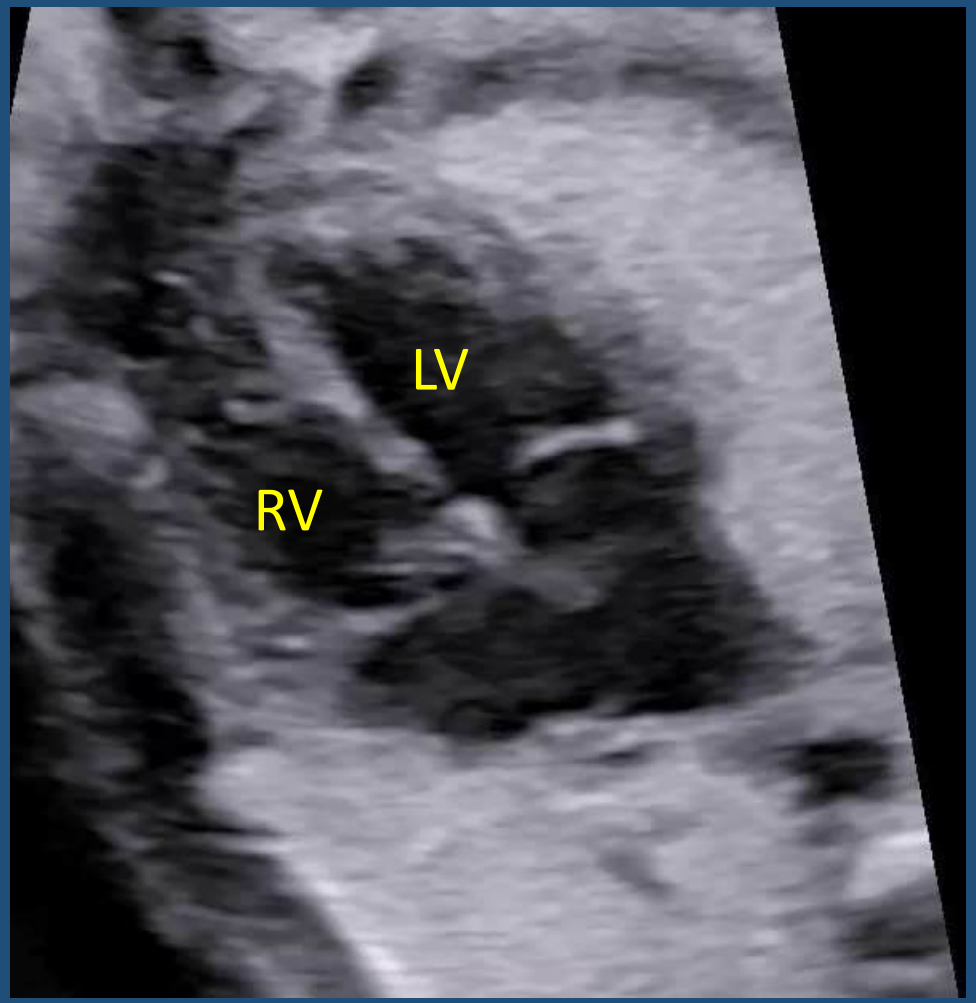
- 35 yo G5P3, spontaneous conception, 25 4/7 weeks
  - Noncontributory past medical / OB history
  - NIPT low risk, declined amnio
- **Thoraco-omphalopagus twins:**
  - diffuse lower sternum/xiphoid
  - Fused liver,shared bowel loops and anterior abdominal wall.
  - Possible fusion of the anterior pleura in the anterior pericardium, however each twin has normal appearing lungs and heart.
  - Each twin has a normal gallbladder.

Case #4 SC

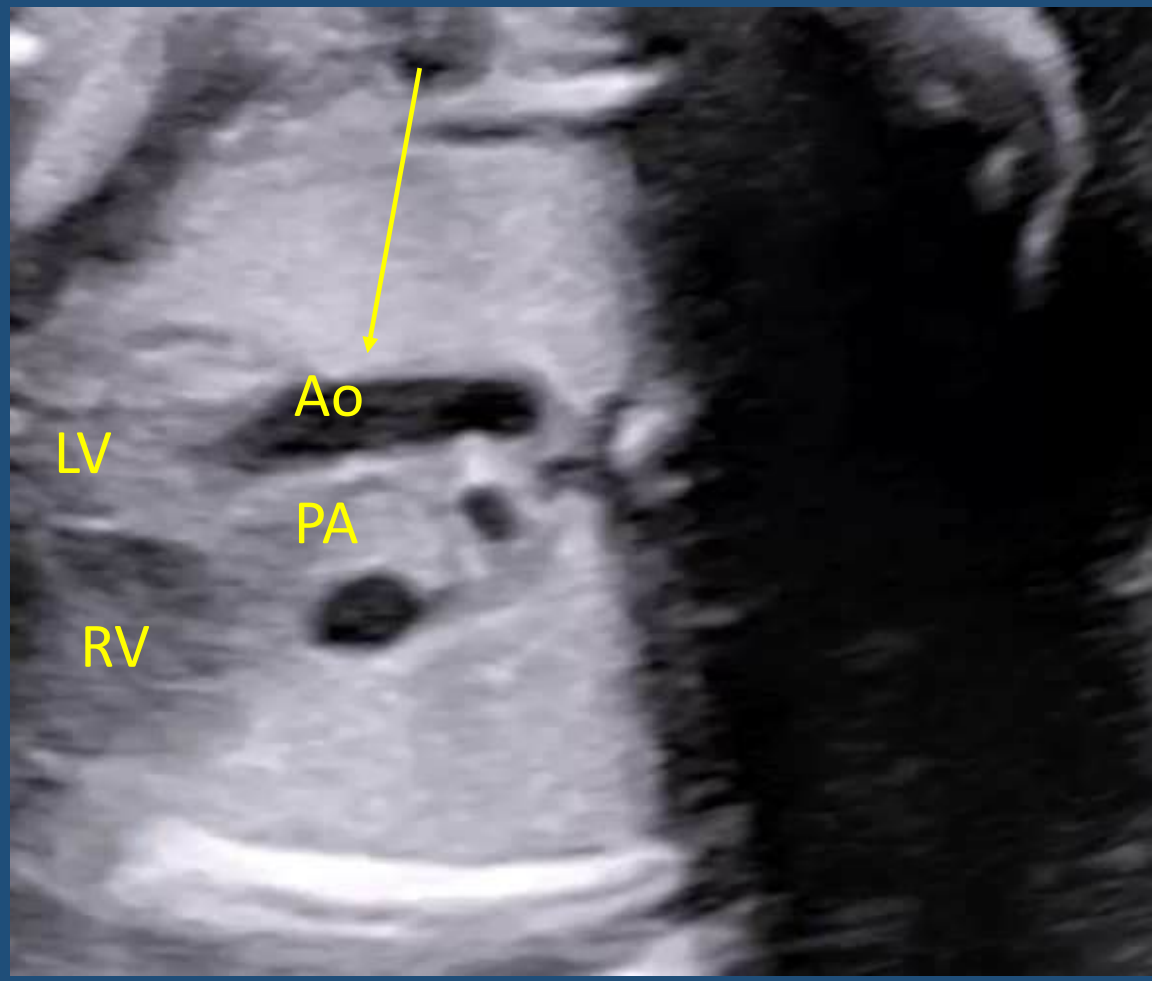
- **Twin A**
  - Structurally normal heart
- **Twin B**
  - {S,D,L} Anatomically corrected malposition (ACM) of great arteries
  - Unobstructed outflow tracts.
  - V-A alignment = RV-PA and LV-Aorta,
    - *but with abnormal subaortic conus (lack of mitral-aortic valve fibrous continuity)*
  - Aortic valve is located anterior and leftward relative to the pulmonary valve, Aortic valve measures slightly smaller than the pulmonary valve
  - Moderate mid-muscular VSD


A MAT RT
BREECH
A
B

Twin B



Aorta leftward, anterior to PA



Prenatal counseling?

Thank you!