Critical aortic stenosis

Disclosures

No disclosures

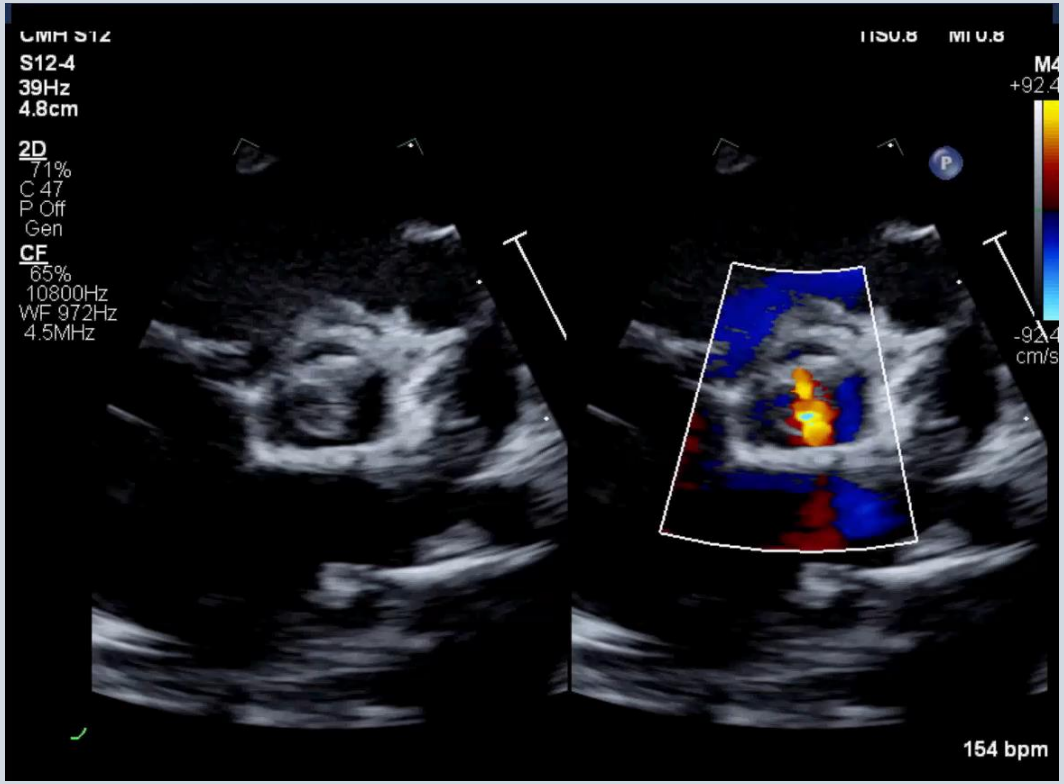
Objectives

Gain a better understanding of Critical AS in fetal life

Explain the role of fetal cardiology and echo in diagnosing critical AS in a fetus

Learn the different pathways a critical AS patient may take in the management of this cardiac defect

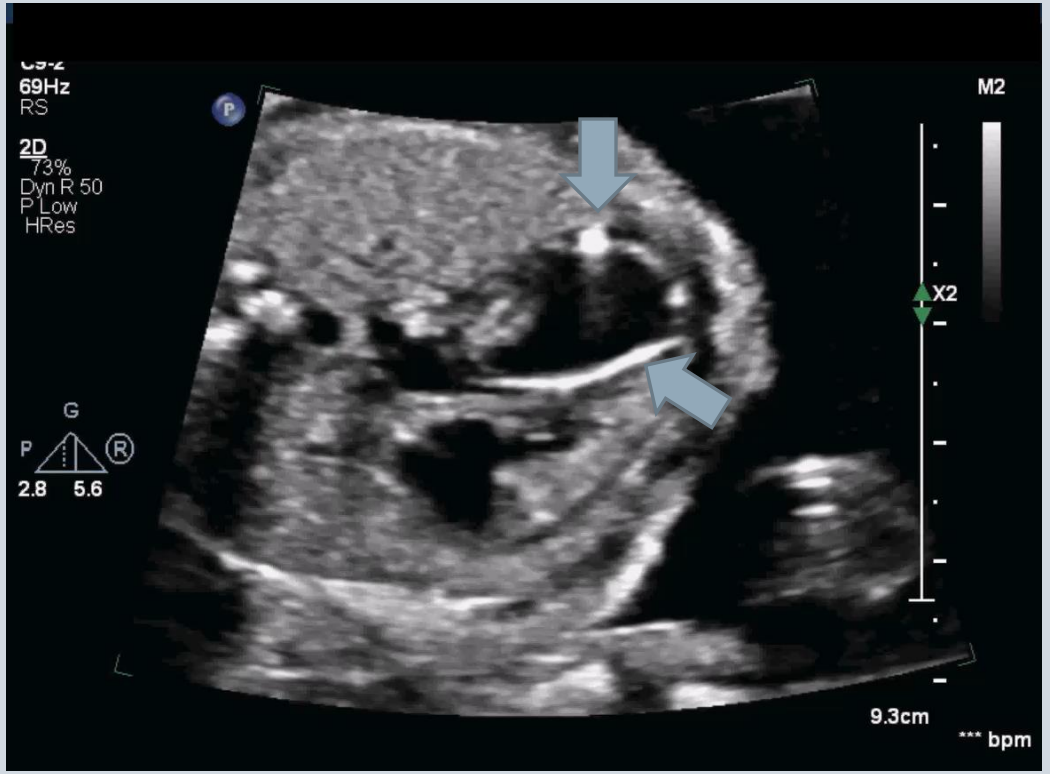
Aortic stenosis



- Aortic valve stenosis in children is usually due to a congenitally abnormal aortic valve rather than degeneration of a normal valve
- Morphologically, this may be due to
  - Commissural underdevelopment
  - Thickening of the valve leaflets
  - Annular hypoplasia
  - Combination of these elements

Pathophysiology

- Obstruction to the left ventricular outflow tract results in an increased afterload on the left ventricle
  - In fetal life, mild stenosis of the aortic valve is usually well tolerated
  - In fetuses with critical aortic valve stenosis the left ventricle dilates, contractile function decreases and endocardial fibroelastosis(EFE) often develops



“No flow no grow”

- Decreased blood flow in fetal life can lead to hypoplasia of cardiac structures
- In severe or critical AS the aorta, aortic arch, left ventricle and mitral valve can become hypoplastic by term
  - Can turn an aortic stenosis case into a single ventricle pathway
    - “Hypoplastic left heart variants”
  - This progression of aortic valve stenosis to a situation like hypoplastic left heart syndrome has led to prenatal intervention being considered for affected fetuses

Prenatal detection Critical cases

- Left ventricle is typically dilated early in gestation with reduced function
- Endocardial fibroelastosis is commonly seen
  - Appears as a bright echogenic lining in the LV
  - Severity may vary
- Patent foreman ovale (PFO) which is right to left in fetal circulation, may be left to right due to severe LV obstruction
  - Can become severely restricted or even intact
  - Pulmonary veins can help determine degree of atrial restriction
- Aortic arch often has retrograde filling
  - Studies have indicated this is a precursor to evolving HLHS

Why prenatal detection is important

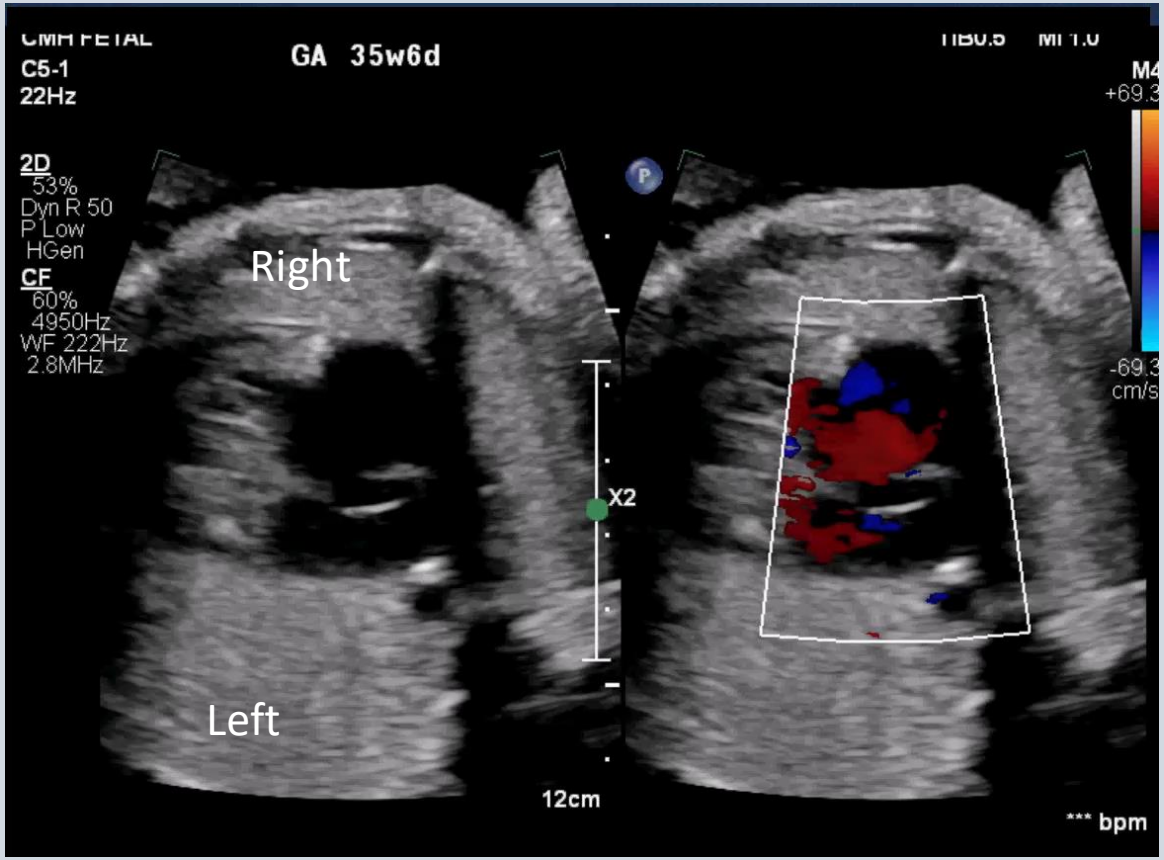
- In infants with severe aortic stenosis with severe left ventricular dysfunction and/or underdevelopment of left sided structures; post natal circulation may not be supported
  - Imperative to distinguish if aortic stenosis is considered “critical AS” and needs prostaglandins after birth to keep duct open
  - In some cases, the fetus may benefit from fetal intervention
  - Family and fetus benefit having a plan in place

The fetal echo

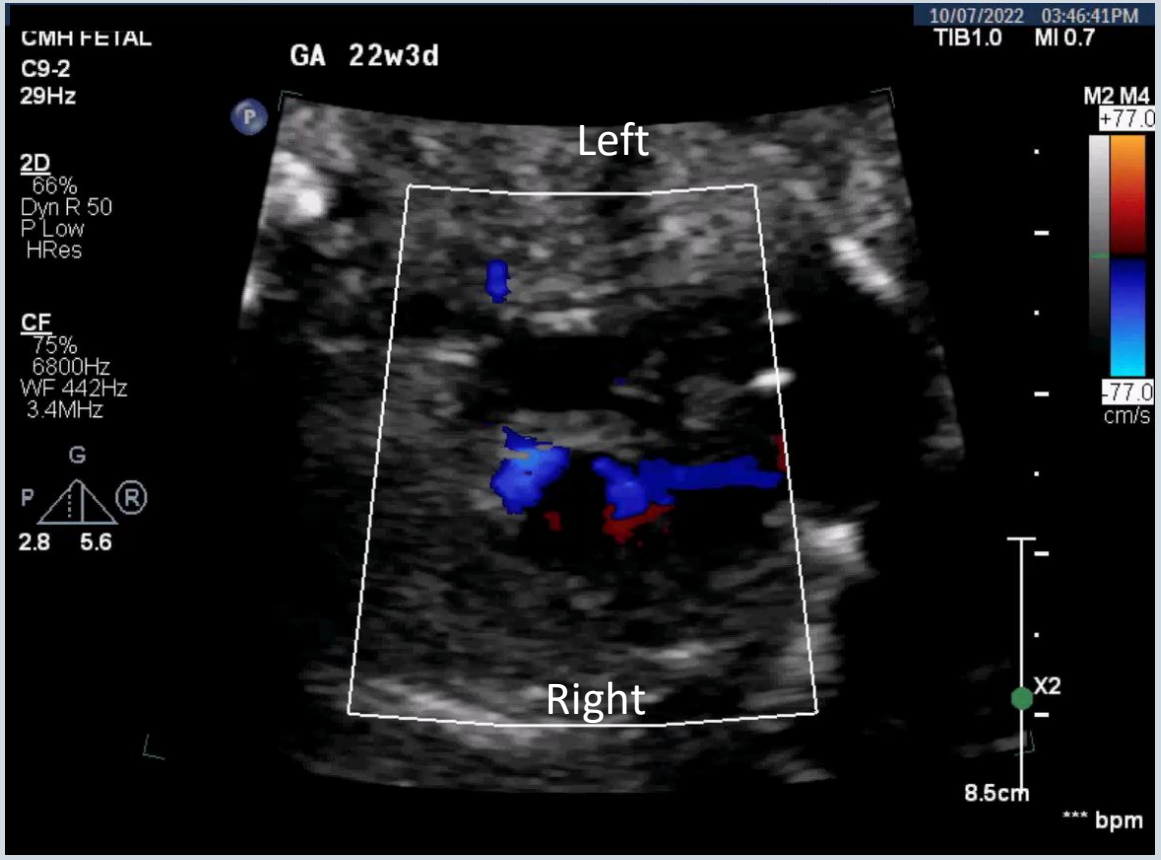
- Assessing aortic stenosis
- Diameters of left and right ventricles
- Lengths of right and left ventricles
- Diameters of the mitral and tricuspid annuli
- Severity of mitral valve regurgitation
- Diameters of aortic valve annulus and aortic arch
- Direction of systolic flow in the distal aortic arch (antegrade versus retrograde)
- Pulmonary vein dopplers for VTI
- Patent foramen ovale

Atrial septum

NORMAL (RIGHT TO LEFT)



ABNORMAL (LEFT TO RIGHT)

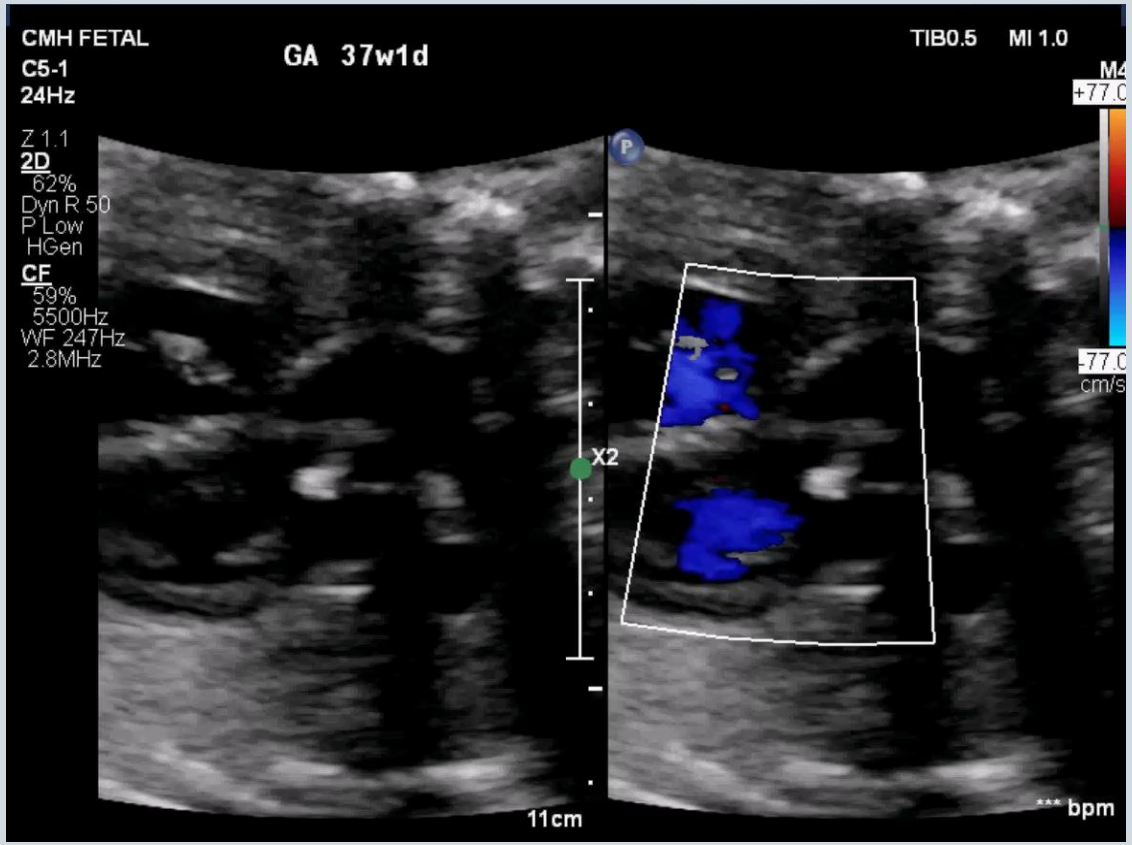


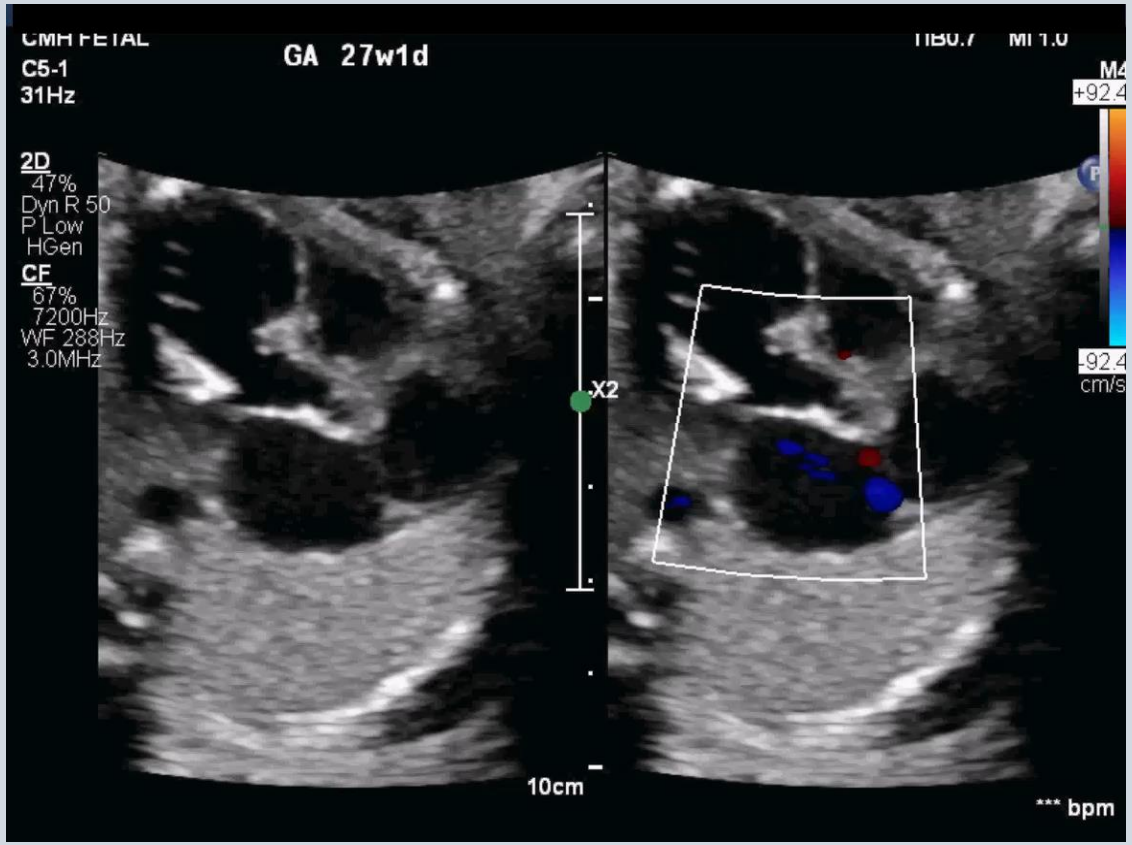
Function


GA 26w2d
C9-2
89Hz
RS
2D
65%
Dyn R 50
P Low
HGen
M2
X2
P
R
2.5
5.0
6.7cm
*** bpm


GA 27w1d
C5-1
81Hz
RS
2D
44%
Dyn R 44
P Low
HGen
M2
X2
P
R
2.0
4.0
8.6cm
*** bpm

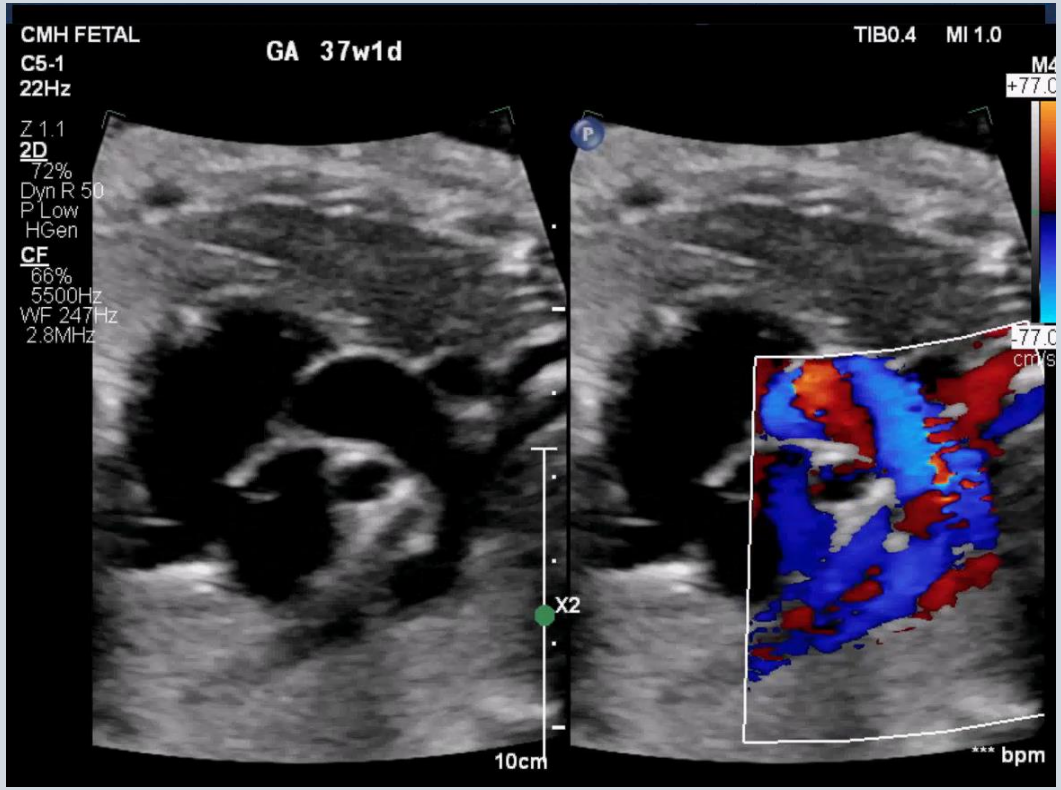
Aortic stenosis



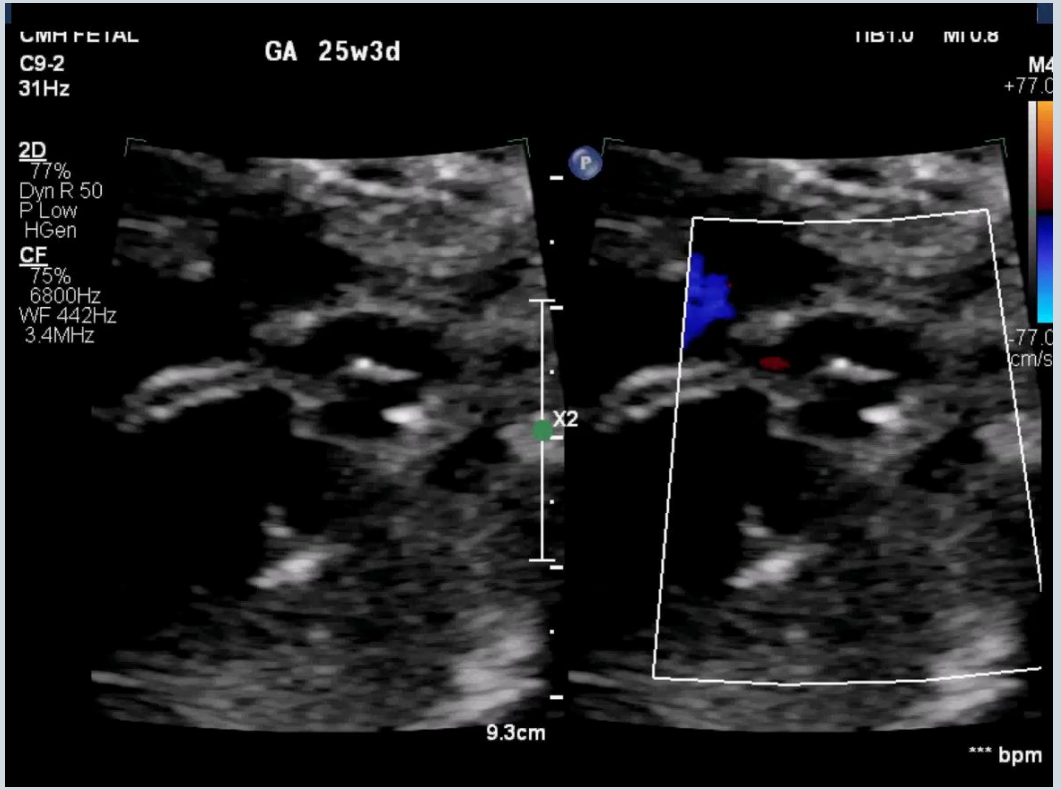


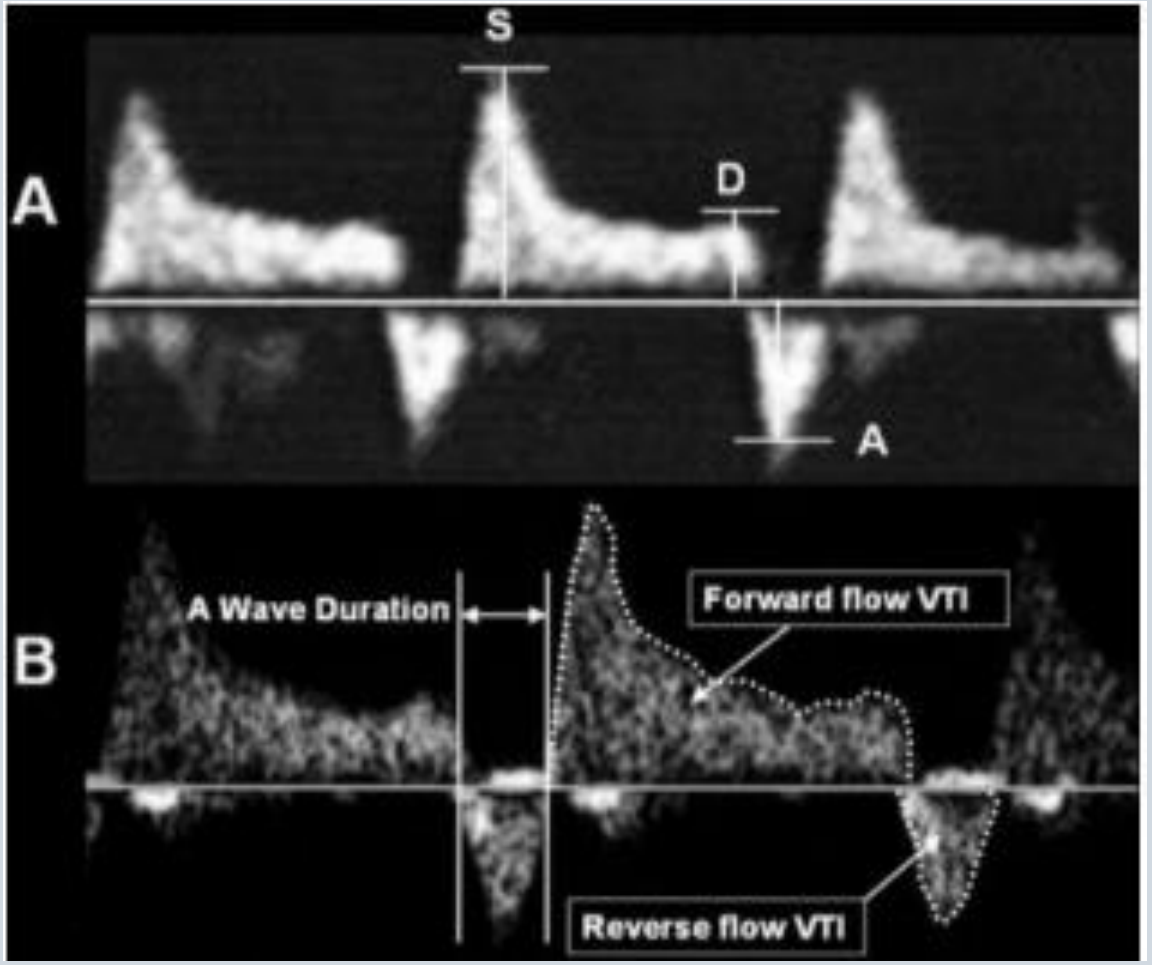
Aortic arch

ANTEGRADE FILLING



RETROGRADE FILLING





Pulmonary vein VTI

- Velocity-time integral
- In the fetus with HLHS, a pulmonary vein VTI <5 is the strongest predictor of the need for emergent atrial septostomy in the newborn period
  - Dividing forward flow VTI by reverse flow VTI to get ratio
- Predictor if fetal intervention is needed (if additional criteria met)

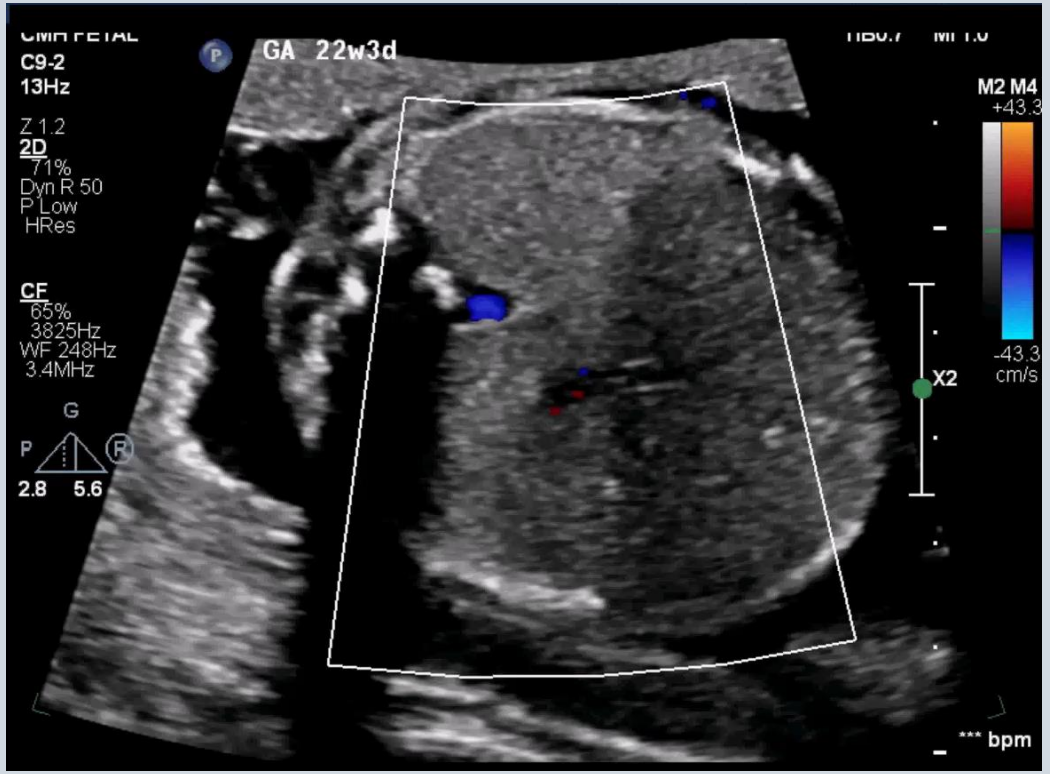
Critical aortic stenosis-options

- Fetal atrial stent ⬅
- Comfort care ⬅
- Single ventricle pathway ⬅
  - Hybrid→Norwood
- Prenatal aortic valve balloon
- “IMPACT” procedure
  - Immediate postpartum access to cardiac therapy

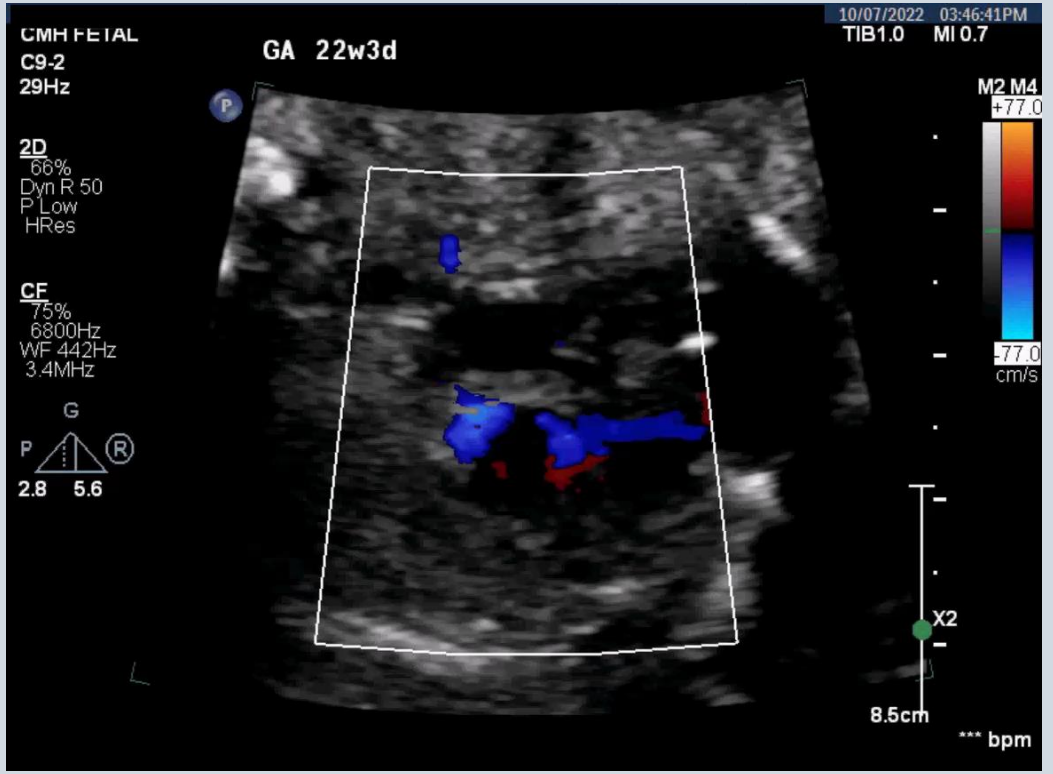
Case #1- Atrial Stent

- G1 P0
- Singleton gestation at 22 weeks 3 days
- Diagnosis:
  - Critical aortic stenosis with evolving hypoplastic left heart syndrome
  - Restrictive atrial septum

Initial sweeps

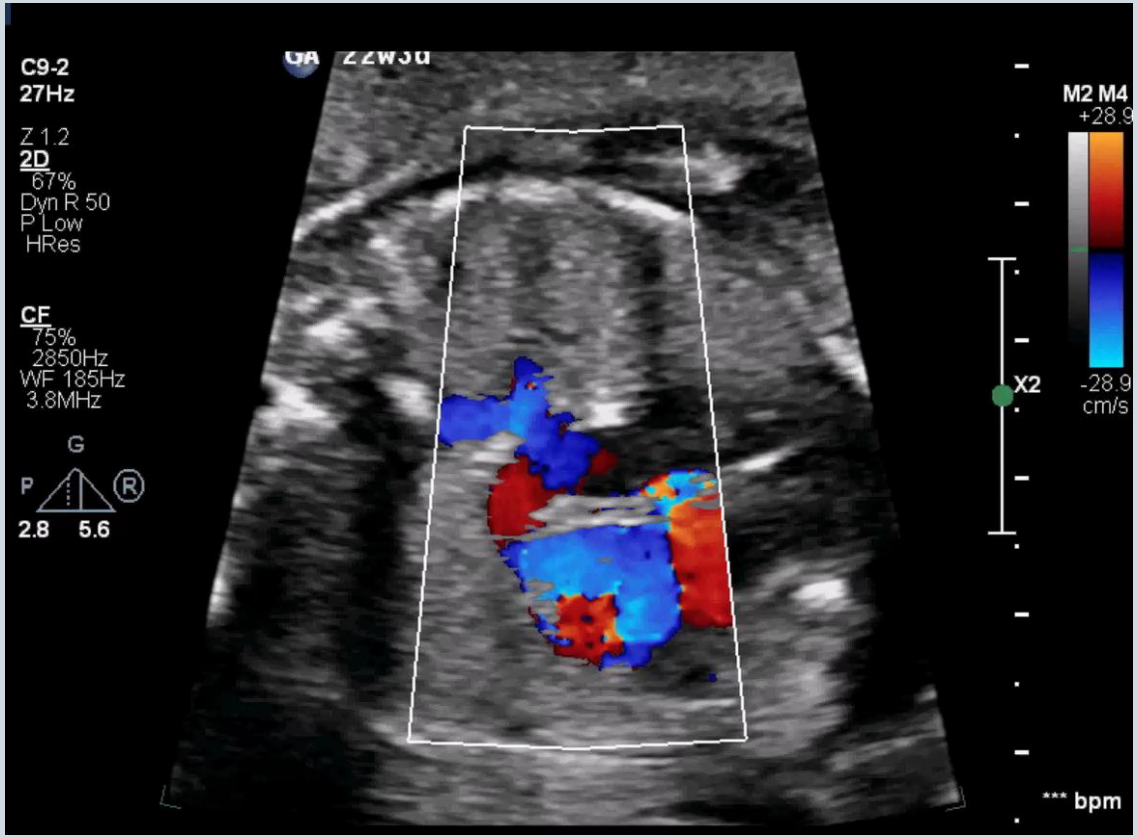


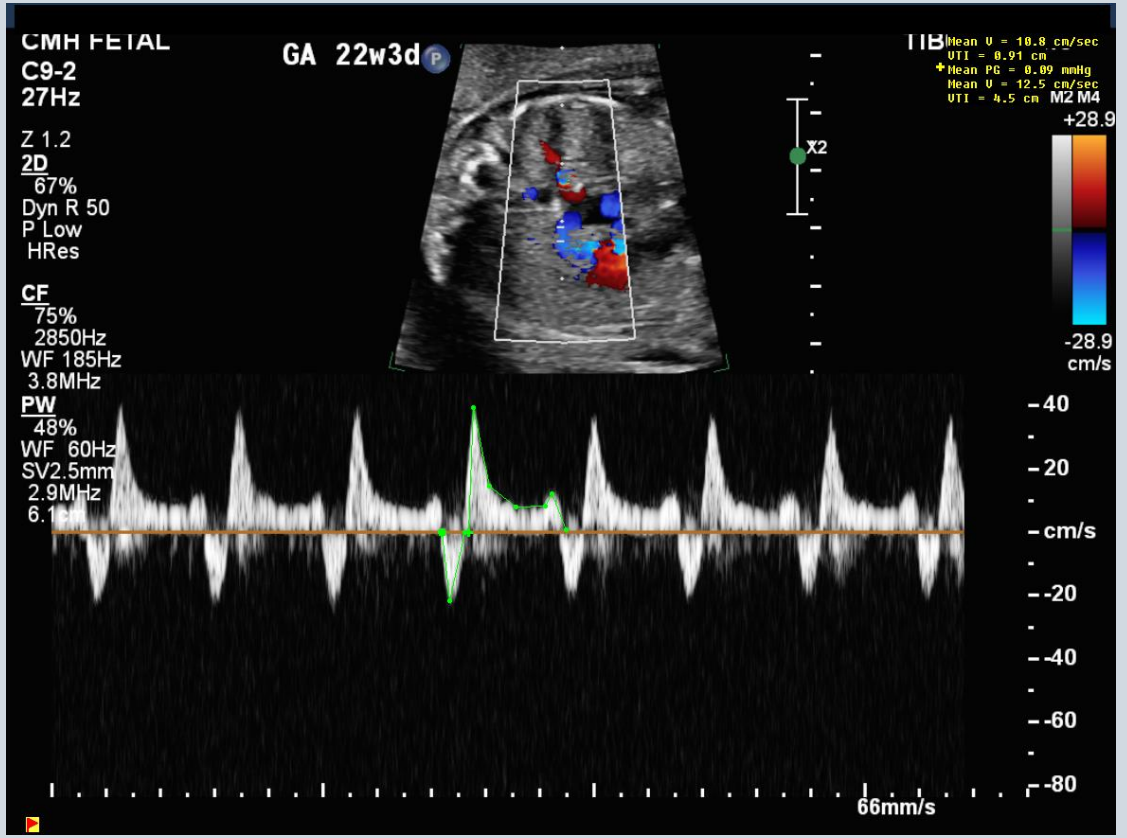
Atrial septum

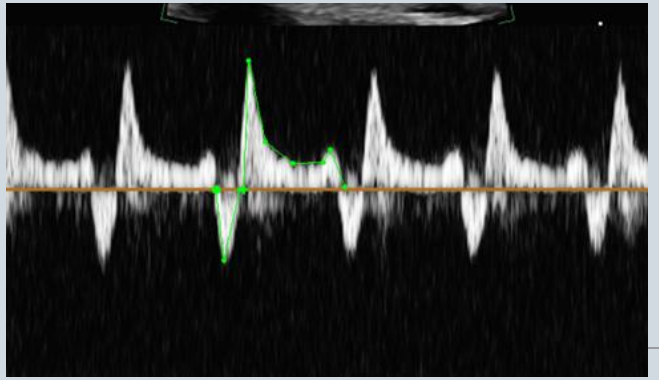


Pulmonary veins

VTI 4

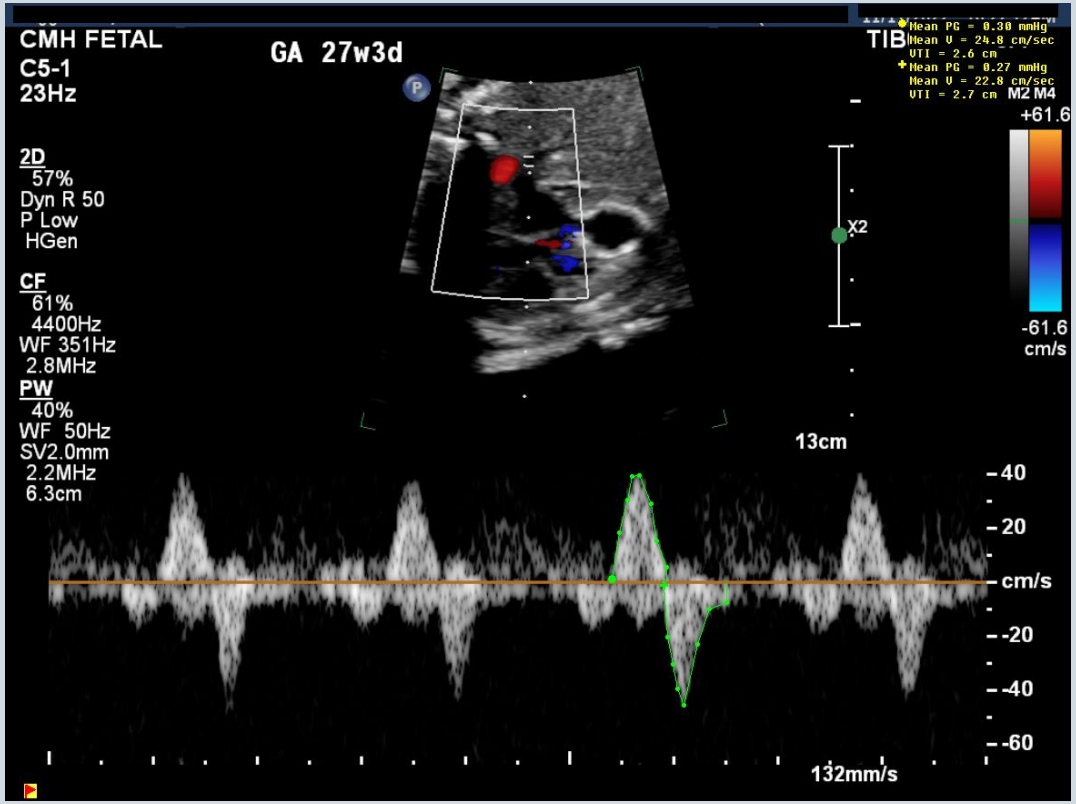


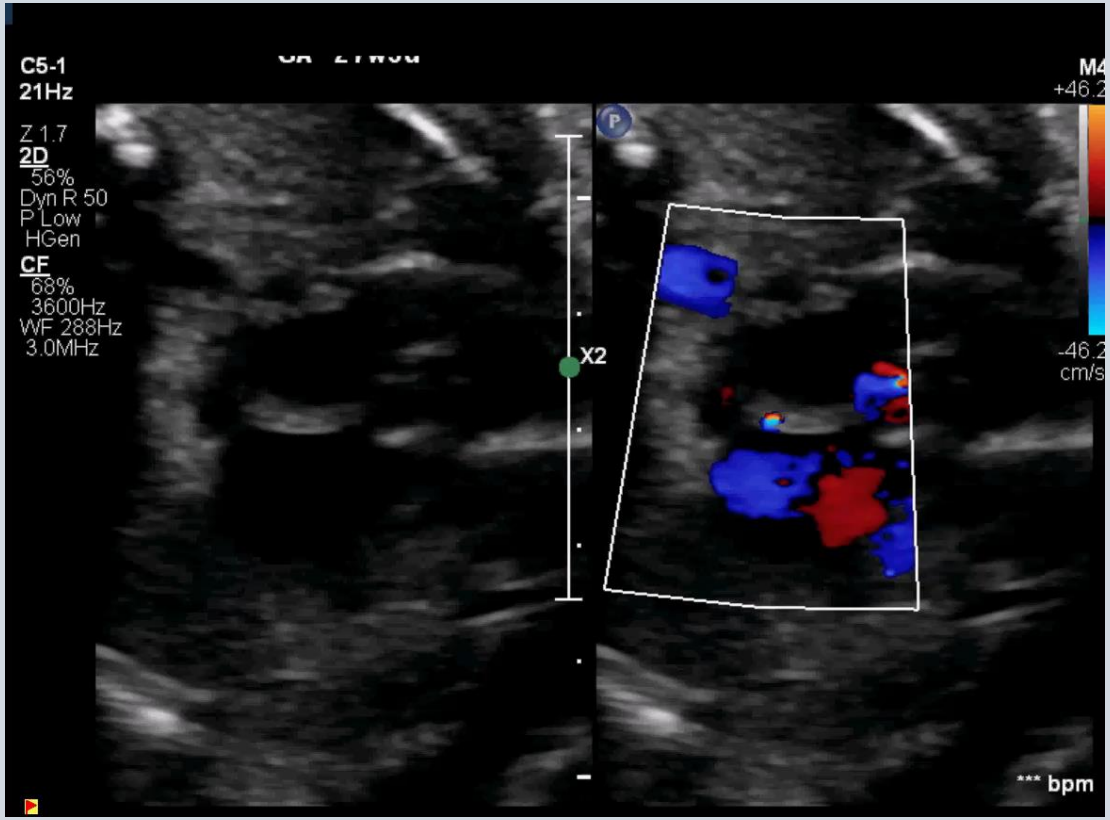




27 weeks 3 days

VTI 2.1





Criteria for atrial stent

INCLUSION CRITERIA

- Single gestation
- Agreement to social work assessment
- Documented understanding of the voluntary nature of the intervention
- Severely restrictive or intact atrial septum with VTI ratio < 3 and/or pulmonary lymphangiectasia on fetal MRI
- Adequate left atrial size for intervention
- No aneuploidy (specifically T13/T18, Turner syndrome, or T21 by amniocentesis)

EXCLUSION CRITERIA

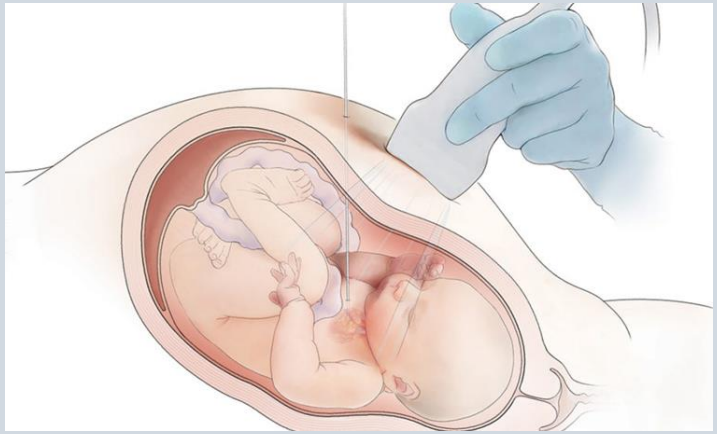
- Other major fetal anomaly
- Major uterine anomaly
- Cerclage
- Maternal coagulopathy
- Maternal HIV+, Hep-B +, Hep-C+
- Hypertension not well controlled
- Contraindication to surgery/anesthesia
- Did not clear psychosocial evaluation
- Significant maternal comorbidities

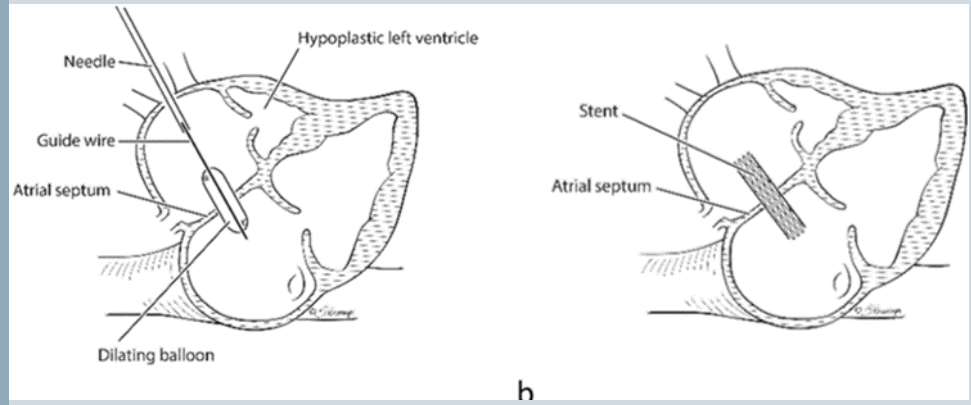
Underwent initial atrial septal stent placement

- The initial stent was successful; however, the following day was noted to have no flow thru stent with a pulmonary vein VTI ratio of about 2.

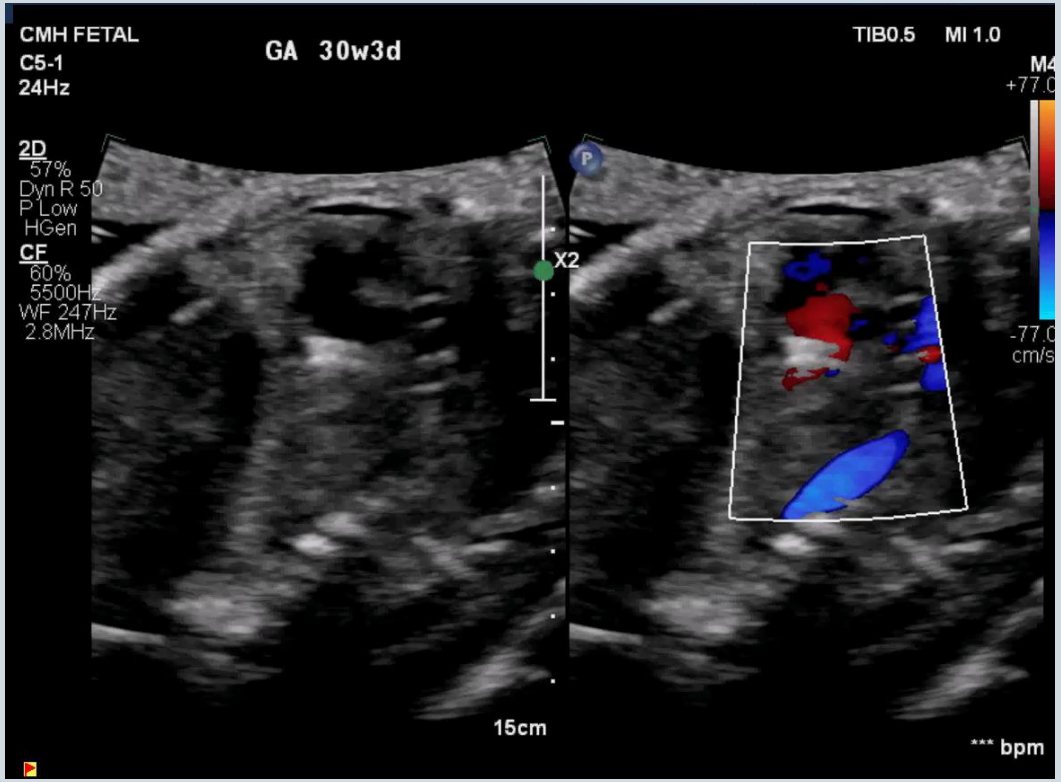
An additional atrial septal stent was placed 7 days later

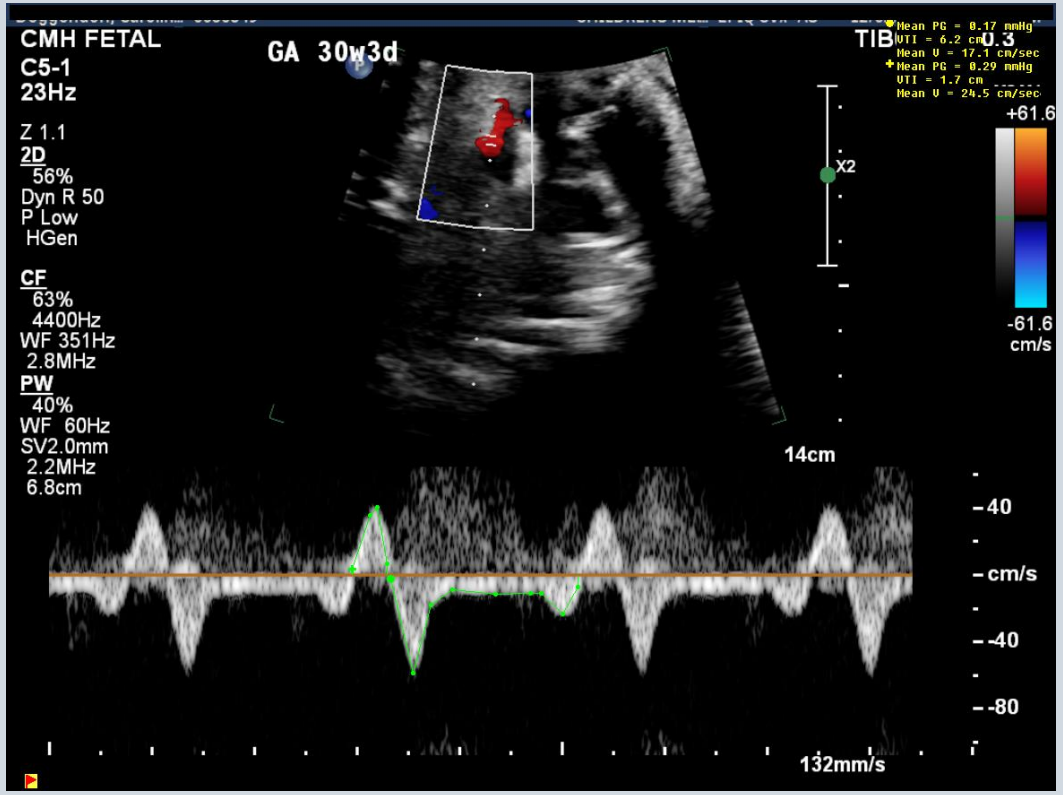
- During second procedure, the fetus developed a large pericardial effusion and bradycardia requiring resuscitation. A right sided pleural effusion also developed with no intervention under the expectation this would most likely self-resolve.





S/p Stent


CMH FETAL
C5-1
24Hz
GA 30w3d
TIB0.5 MI 1.0
2D
57%
Dyn R 50
P Low
HGen
CF
60%
5500Hz
WF 247Hz
2.8MHz
X2
15cm
*** bpm


CMH FETAL
C5-1
23Hz
GA 30w3d
Mean PG = 0.17 mmHg
VTI = 6.2 cm
Mean V = 17.1 cm/sec
Mean PG = 0.29 mmHg
VTI = 1.7 cm
Mean V = 24.5 cm/sec
Z 1.1
2D
56%
Dyn R 50
P Low
HGen
CF
63%
4400Hz
WF 351Hz
2.8MHz
PW
40%
WF 60Hz
SV2.0mm
2.2MHz
6.8cm
+61.6
-61.6
cm/s
X2
14cm
-40
cm/s
-40
-80
132mm/s

- VTI improved
- Will likely have postnatal single ventricle physiology
- Single ventricle pathway

Case #2- Comfort care

G4 P2

Singleton gestation at 20 weeks 4 days

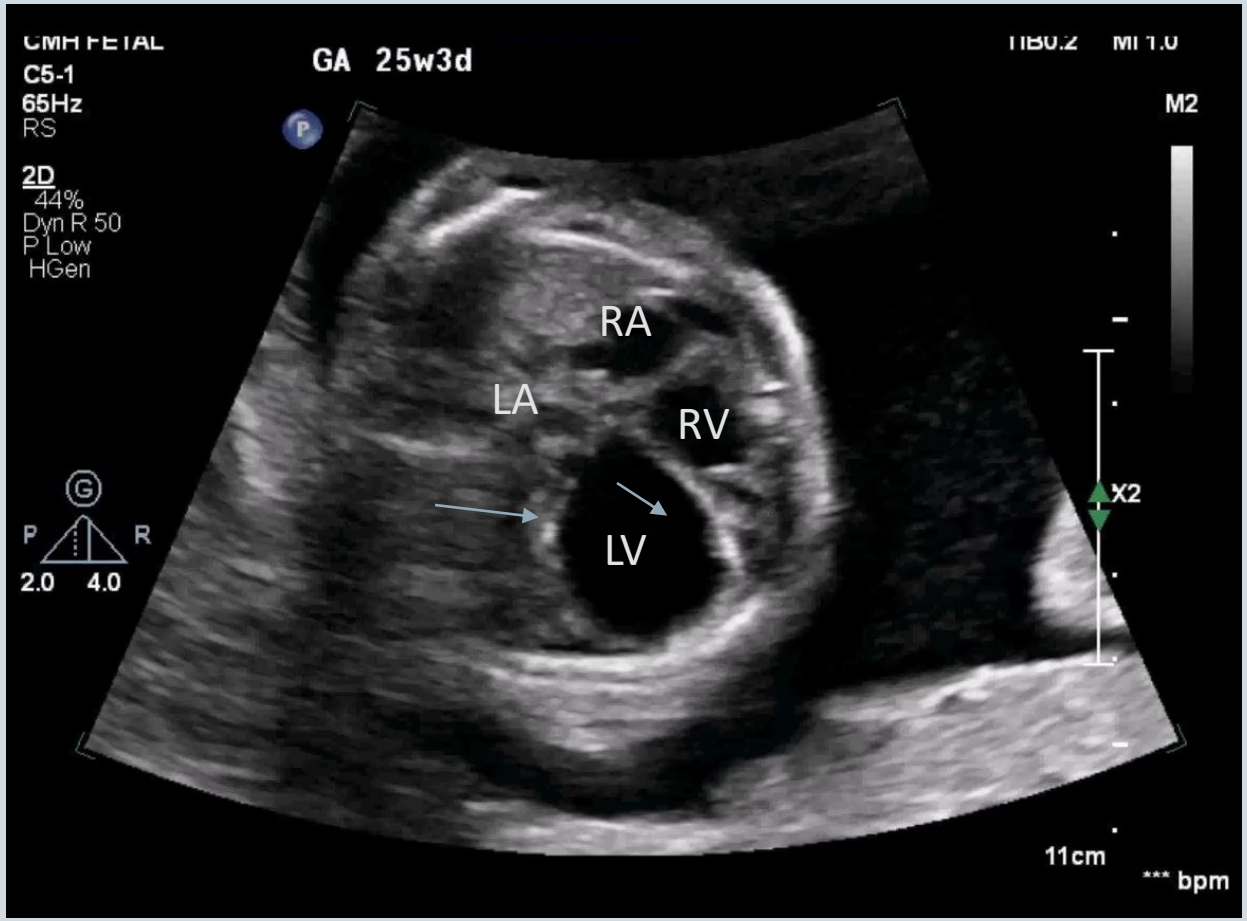
Reason for study:

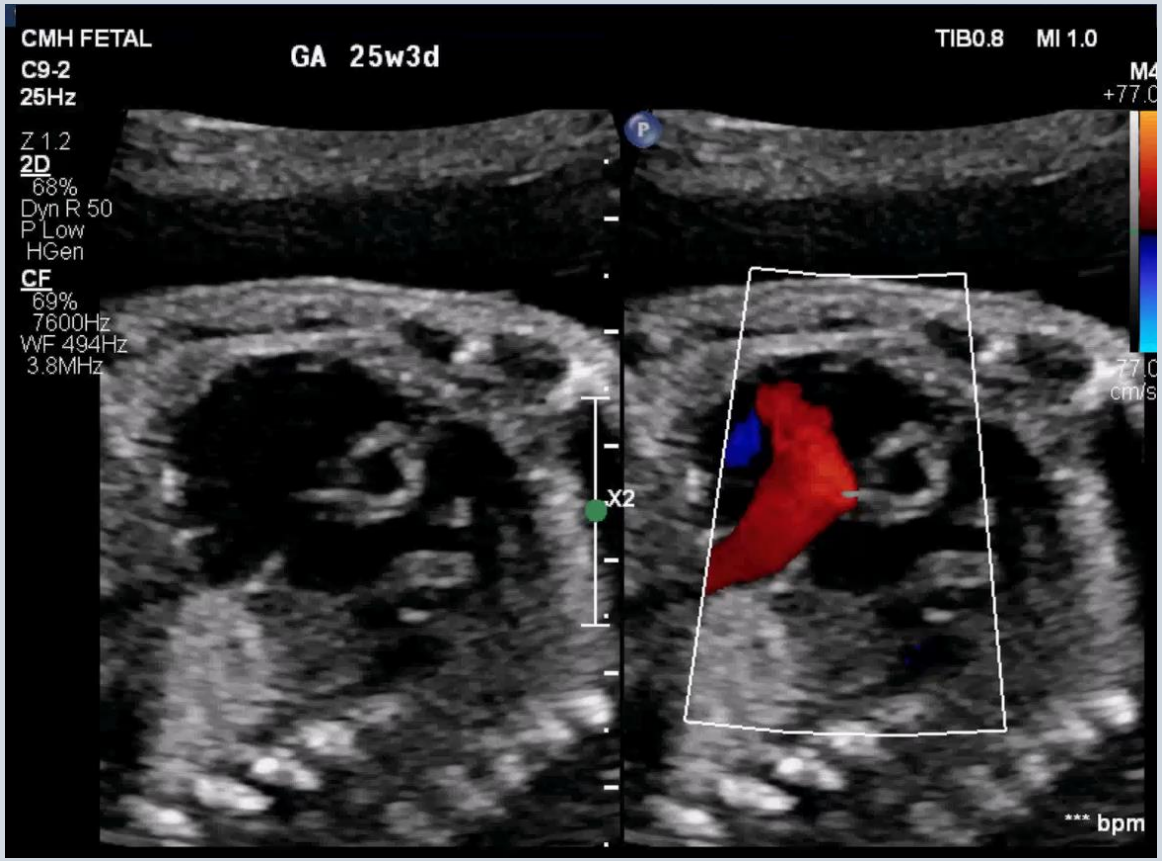
- Aortic stenosis

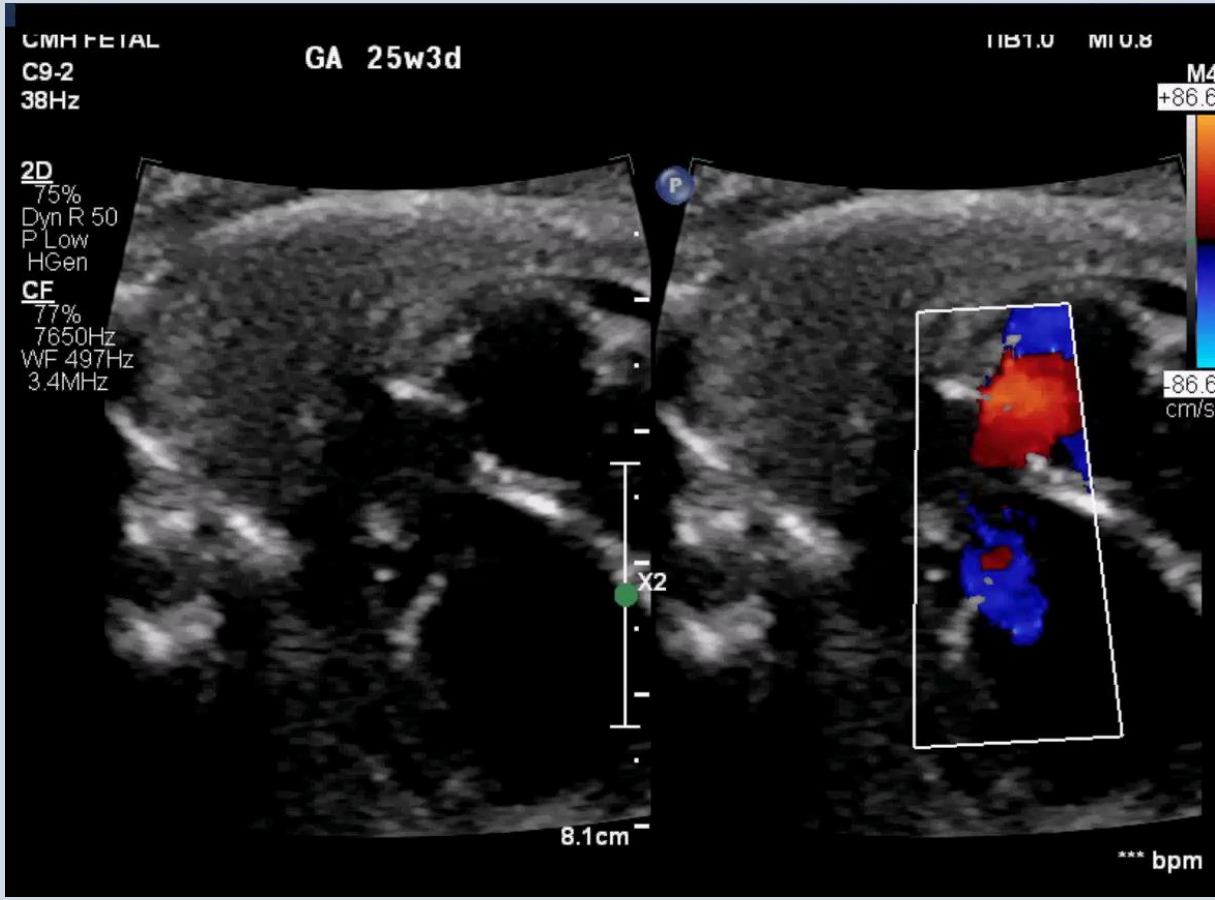
Diagnosis:

- Hypoplastic left heart syndrome variant (MS/AS)
- Normal mitral valve annulus size but with echo bright papillary muscles and chords suggestive of EFE
- Serial echoes to monitor progression

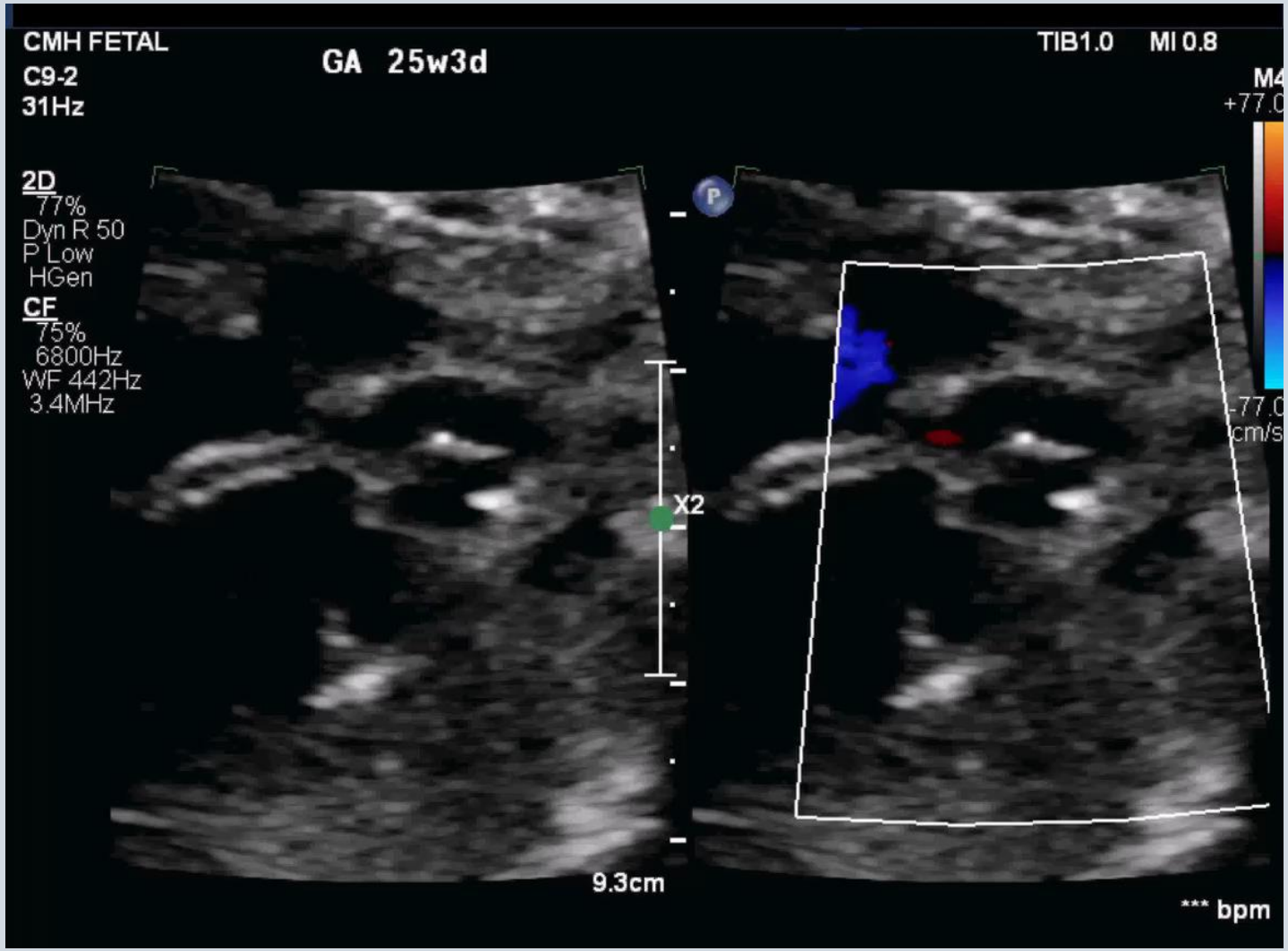
25 weeks 3 days

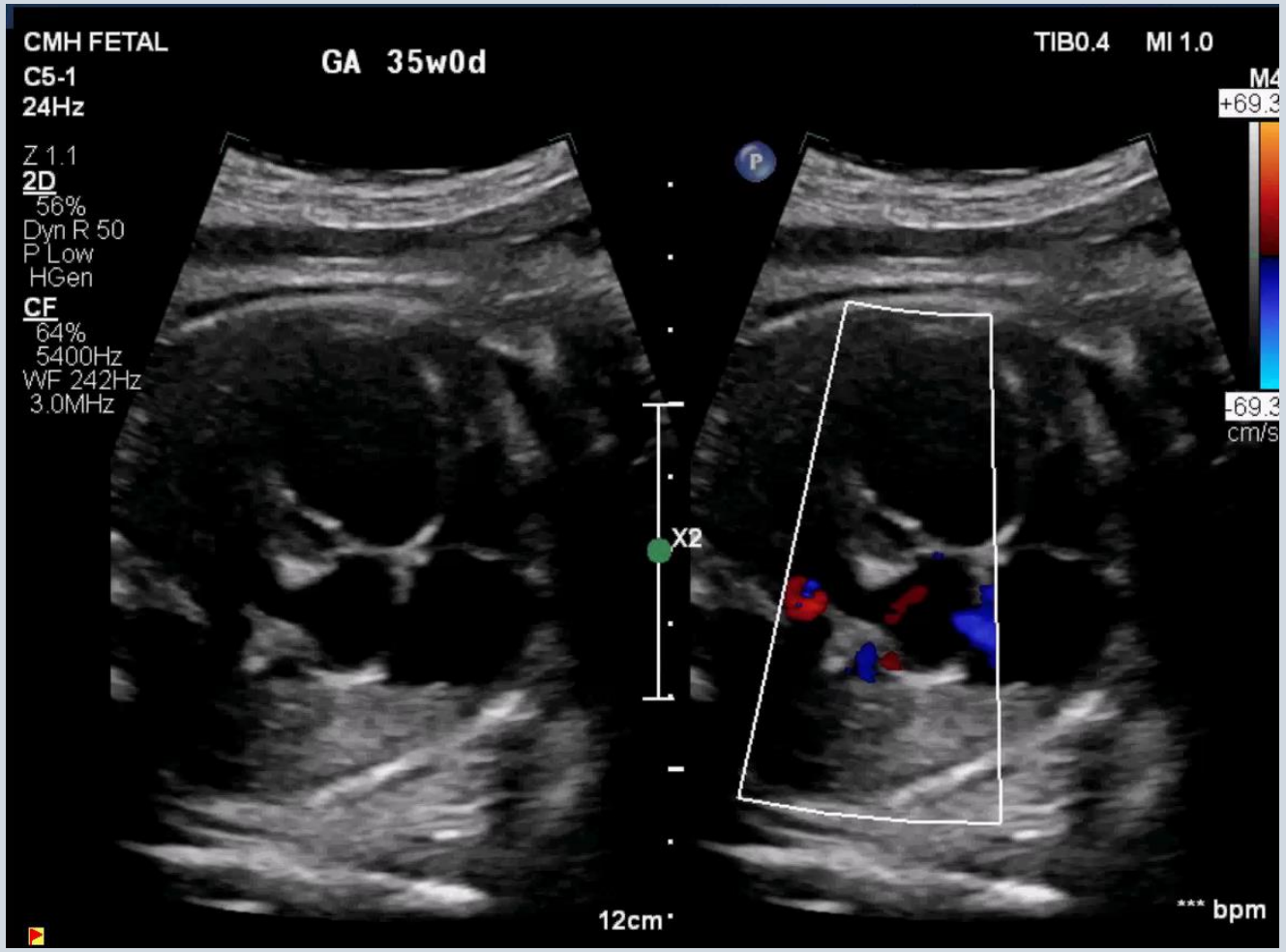



CMH FETAL
C9-2
25Hz
GA 25w3d
TIB0.8 MI 1.0
Z 1.2
2D
68%
Dyn R 50
P Low
HGen
CF
69%
7600Hz
WF 494Hz
3.8MHz
X2
*** bpm


GA 25w3d
C9-2
38Hz
2D
75%
Dyn R 50
P Low
HGen
CF
77%
7650Hz
WF 497Hz
3.4MHz
X2
8.1cm
*** bpm

- Hypoplastic transverse arch with retrograde filling
- No antegrade flow thru the aortic arch determined fetus was not a candidate for fetal intervention

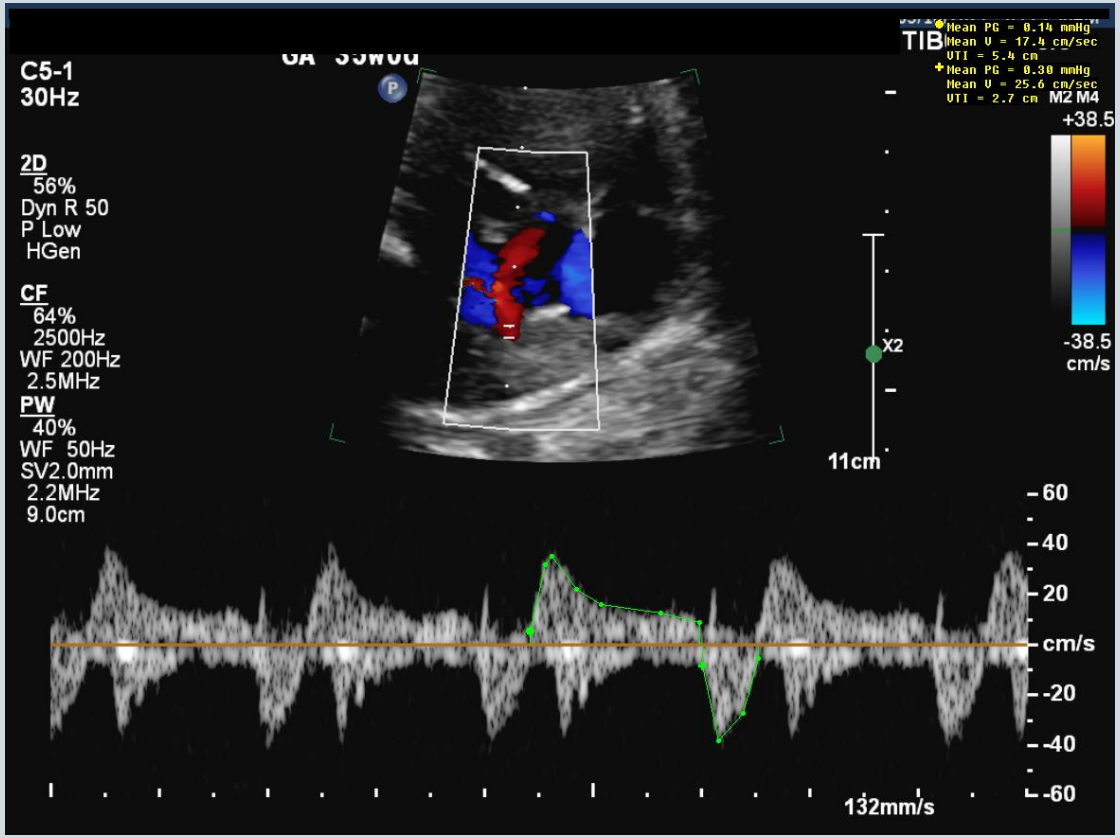


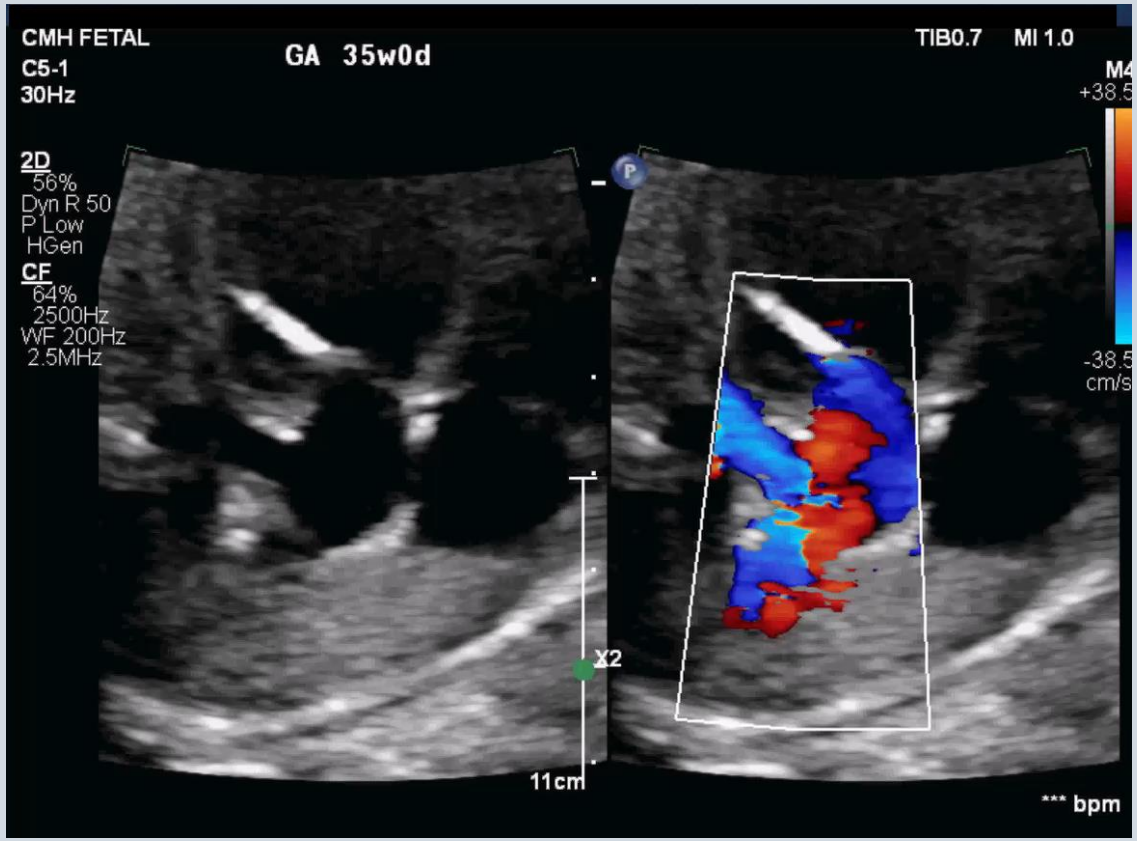

CMH FETAL
C5-1
24Hz
Z 1.1
2D
56%
Dyn R 50
P Low
HGen
CF
64%
5400Hz
WF 242Hz
3.0MHz
GA 35w0d
TIB0.4 MI 1.0
M4
+69.3
-69.3
cm/s
X2
12cm
*** bpm

35 weeks

Pulmonary veins

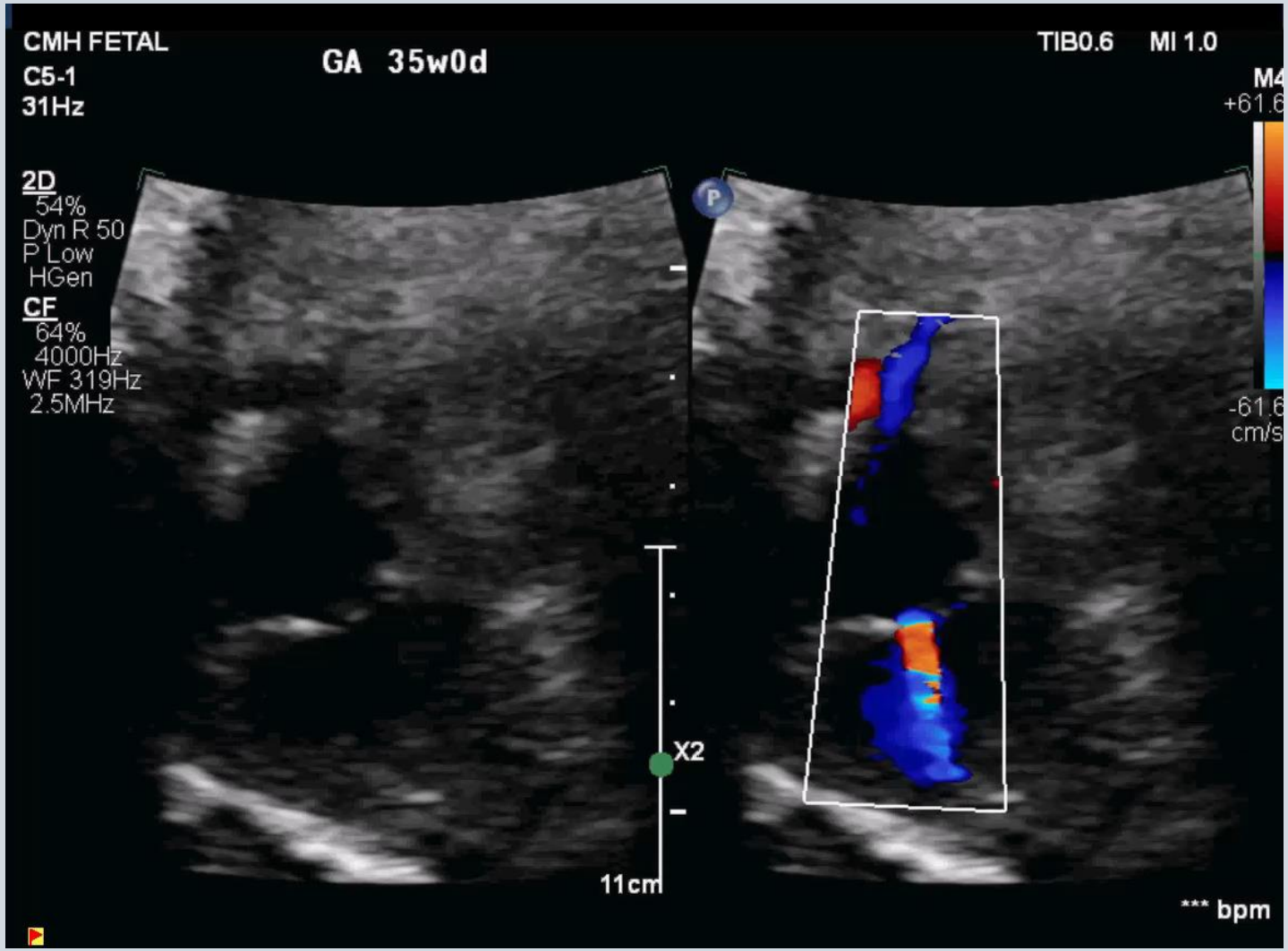
VTI 3-4





Atrial septum

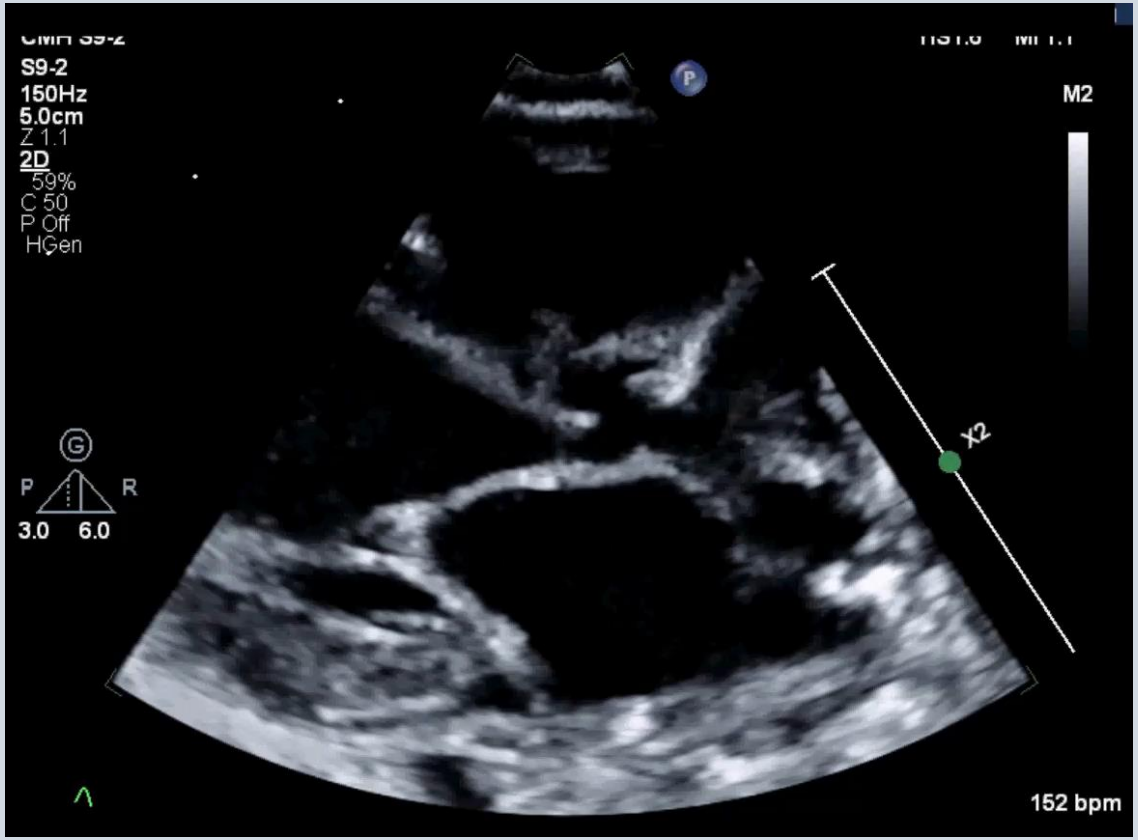
- Left to right flow
- Increased gradient (6-7 mmHg)



Counseling

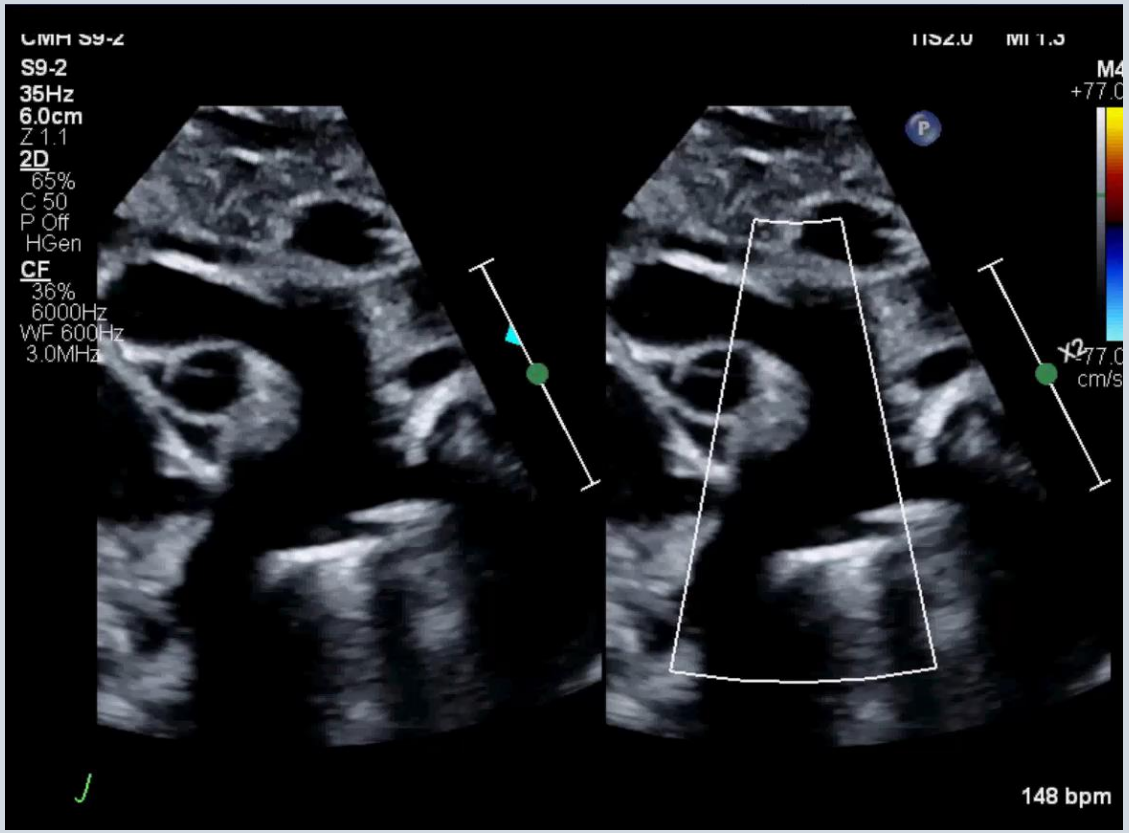
- Family discussion of hypoplastic left heart syndrome with restrictive atrial septum and likelihood of immediate intervention needed after birth with multiple surgeries to follow
- Given post natal options family chose comfort care with the goals of holding and allowing siblings to meet baby soon after birth
- Plan for immediate post natal echo confirming diagnosis

Post natal echo

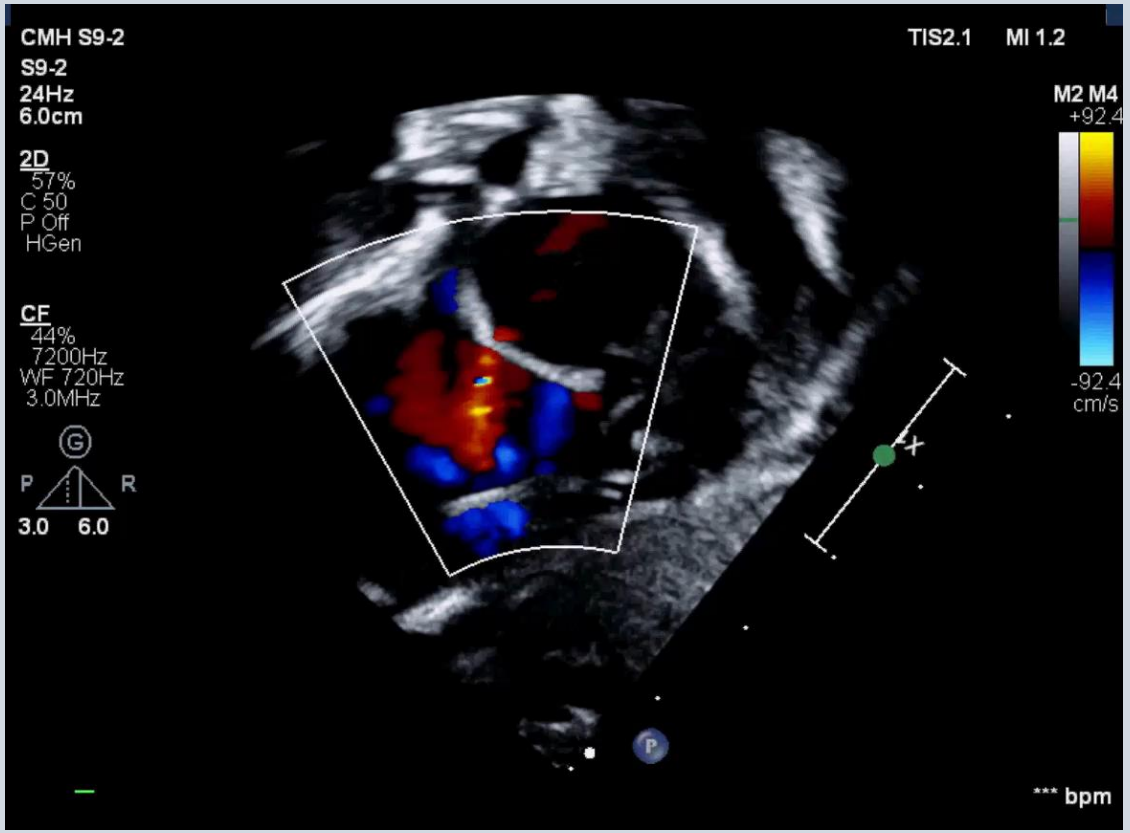


Post natal echo

RETROGRADE ARCH FILLING



RESTRICTIVE IAS

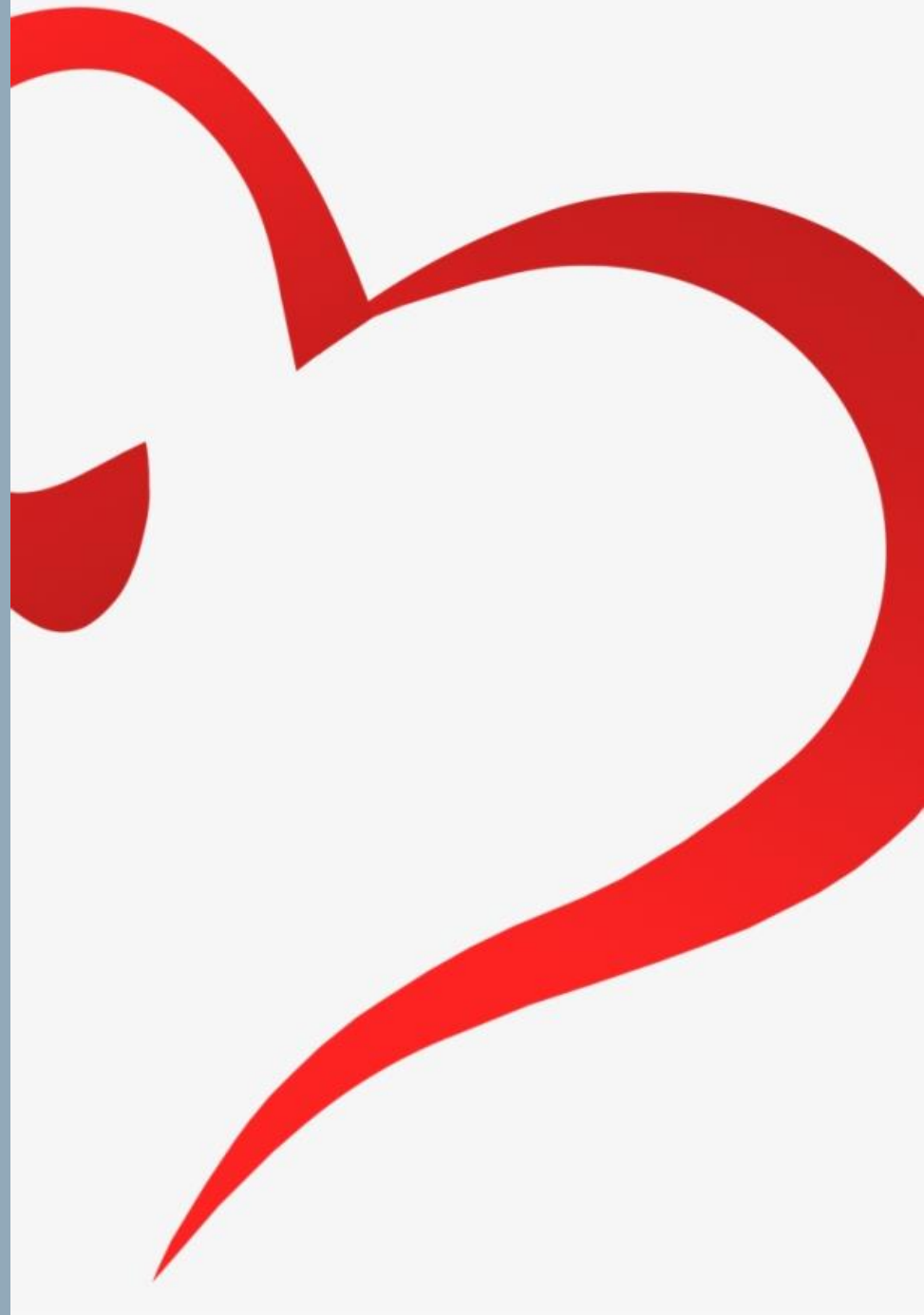


What is comfort care?

- Once a diagnosis has been determined; parents are counseled on possible outcomes, including:
  - Surgical option
  - Nonsurgical option; palliative care/hospice
- Since prenatal diagnosis is obtained parents have time to discuss and process all information and determine what they feel is best for their baby
- Comfort care creates a pathway for families to focus on infant comfort, contact with family members and creating positive memories with their newborn in the time they have together

How we help families navigate

- Create goals
  - Do they want to hold immediately?
  - Sibling involvement
- Meet with palliative team
- Counseling
- Memory makers-handprints, photography, heartbeat bear



Case #3- Hybrid

- G4 P2
- Singleton gestation at 20 and 5 weeks
- Reason for study:
  - Possible HLHS
- Diagnosis:
  - Moderate aortic valve stenosis
  - Mild mitral and tricuspid insufficiency
  - Borderline biventricular systolic function

20 weeks 5 days

The plan…

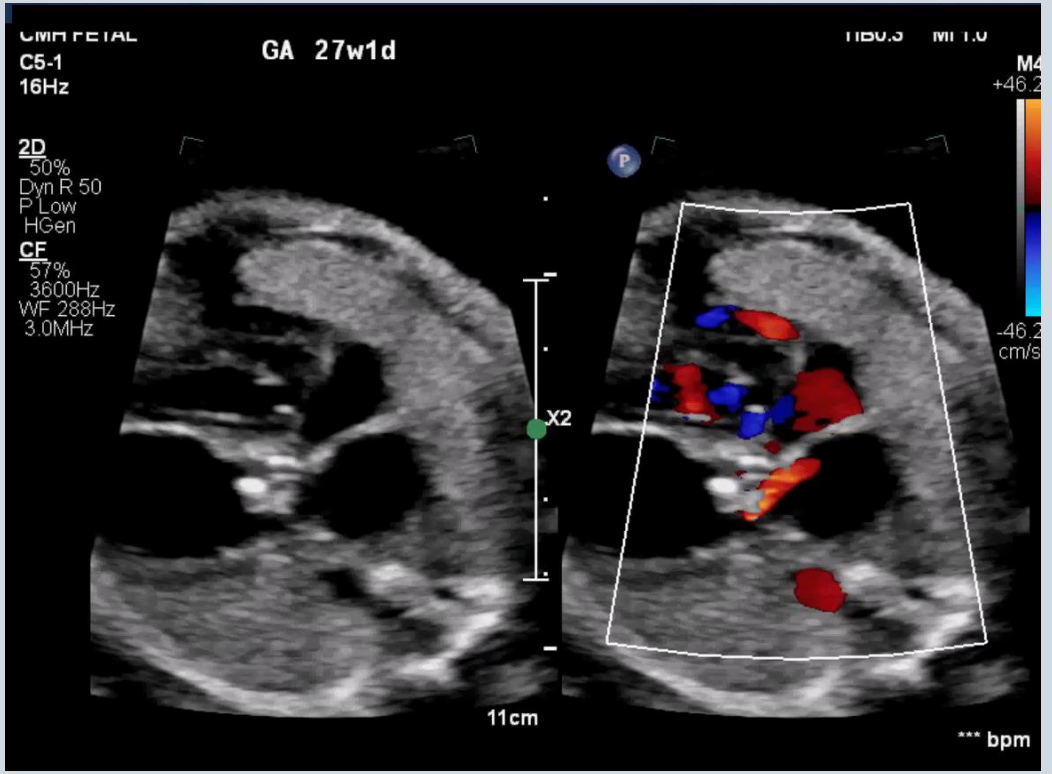
- Family counseled on multiple scenarios
- A wait and see situation
- Echo performed 2 weeks after initial study with little to no changes
- Serial echoes planned

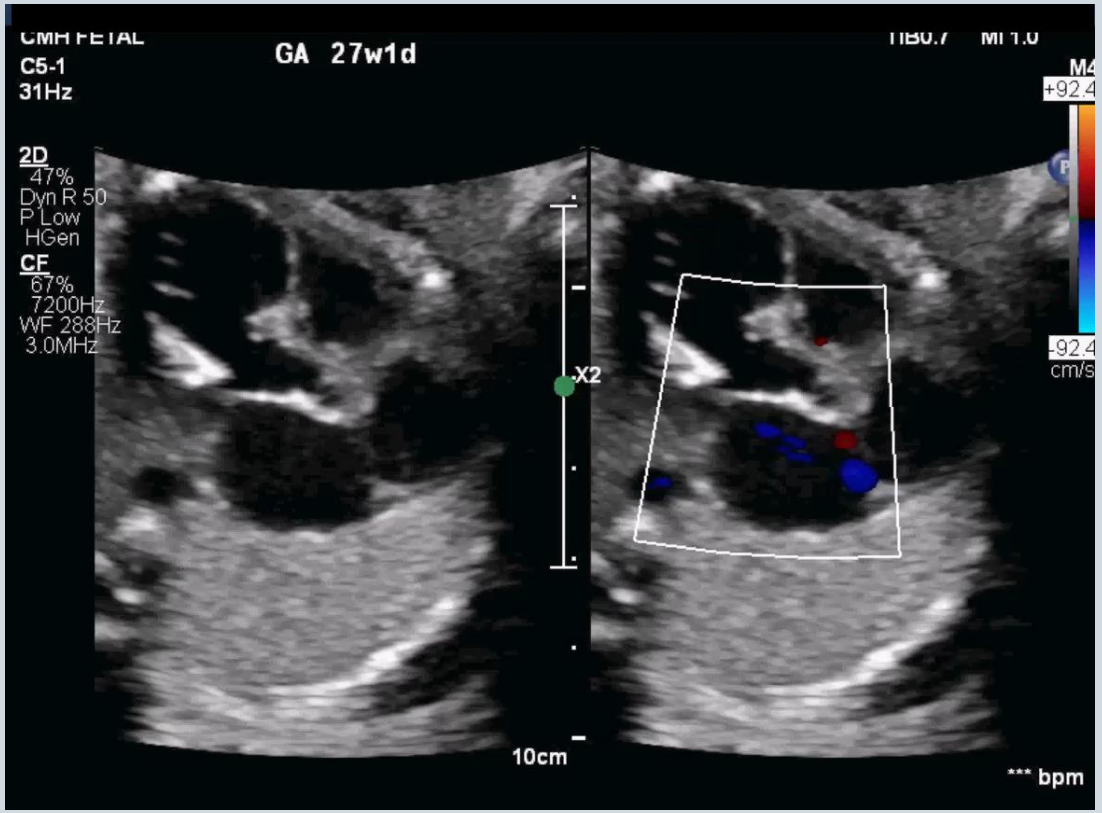
Serial echoes

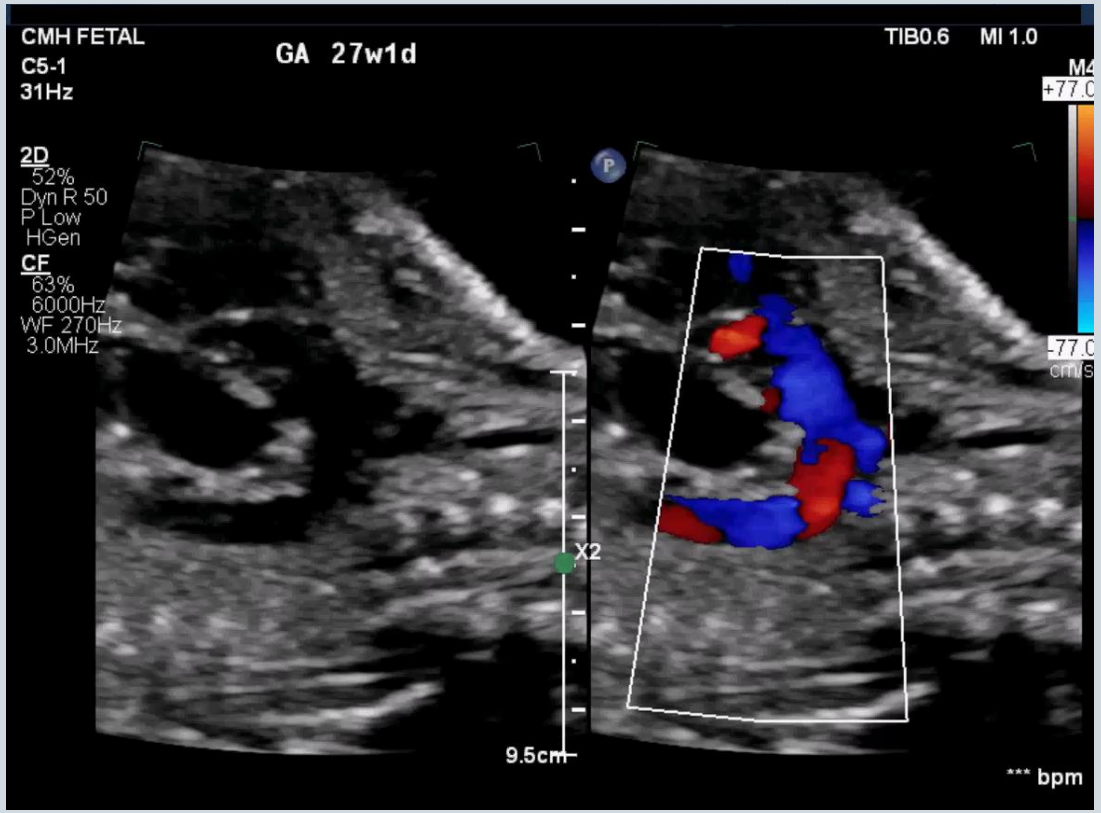
23 WEEKS (2 WEEKS AFTER INITIAL STUDY)

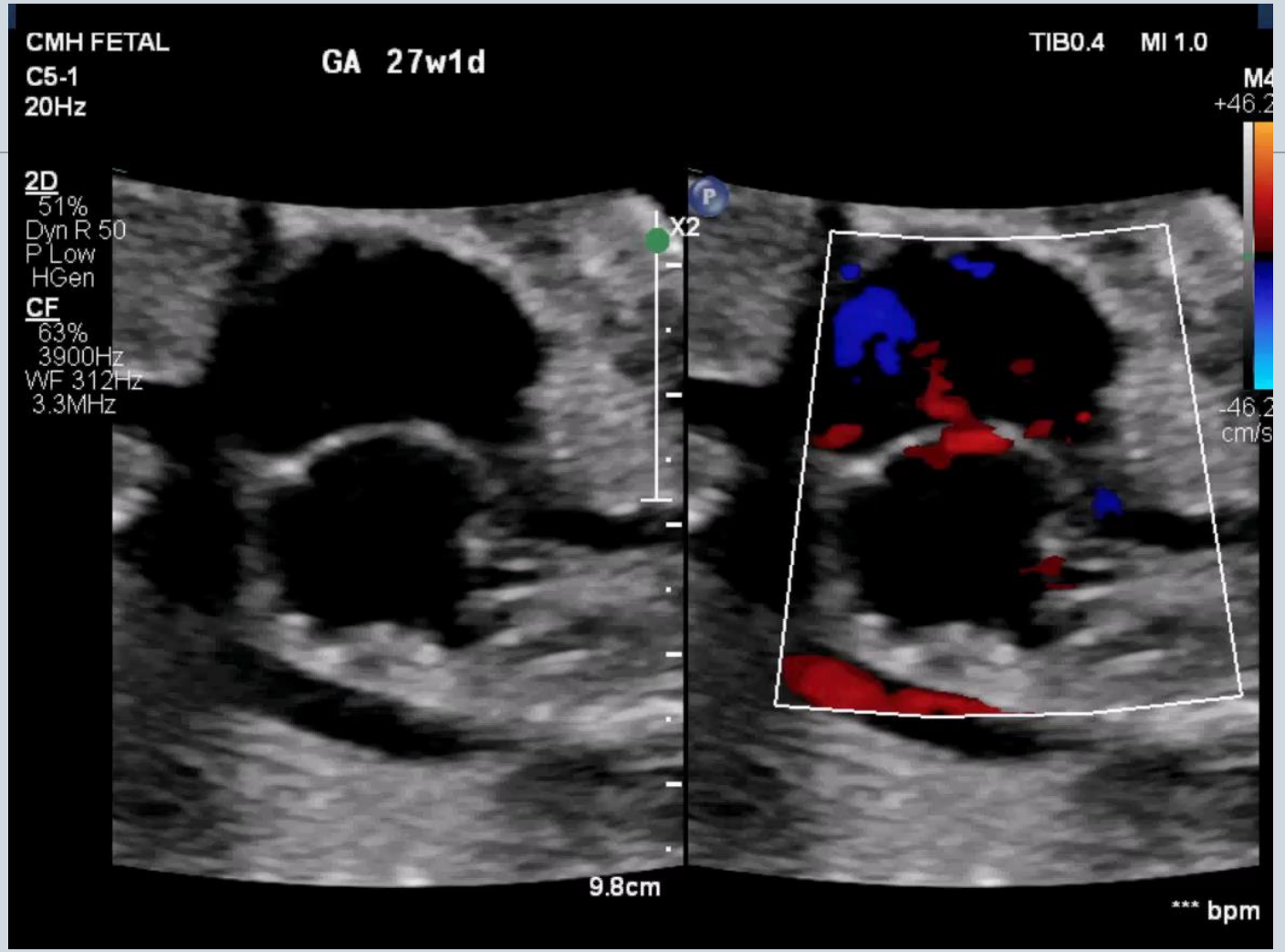
27 WEEKS 1DAY


GA 27w1d
C5-1
81Hz
RS
2D
44%
Dyn R 44
P Low
HGen
M2
X2
8.6cm
*** bpm
2.0 4.0
P R


GA 27w1d
C5-1
16Hz
2D
50%
Dyn R 50
P Low
HGen
CF
57%
3600Hz
WF 288Hz
3.0MHz
X2
11cm
*** bpm
cm/s

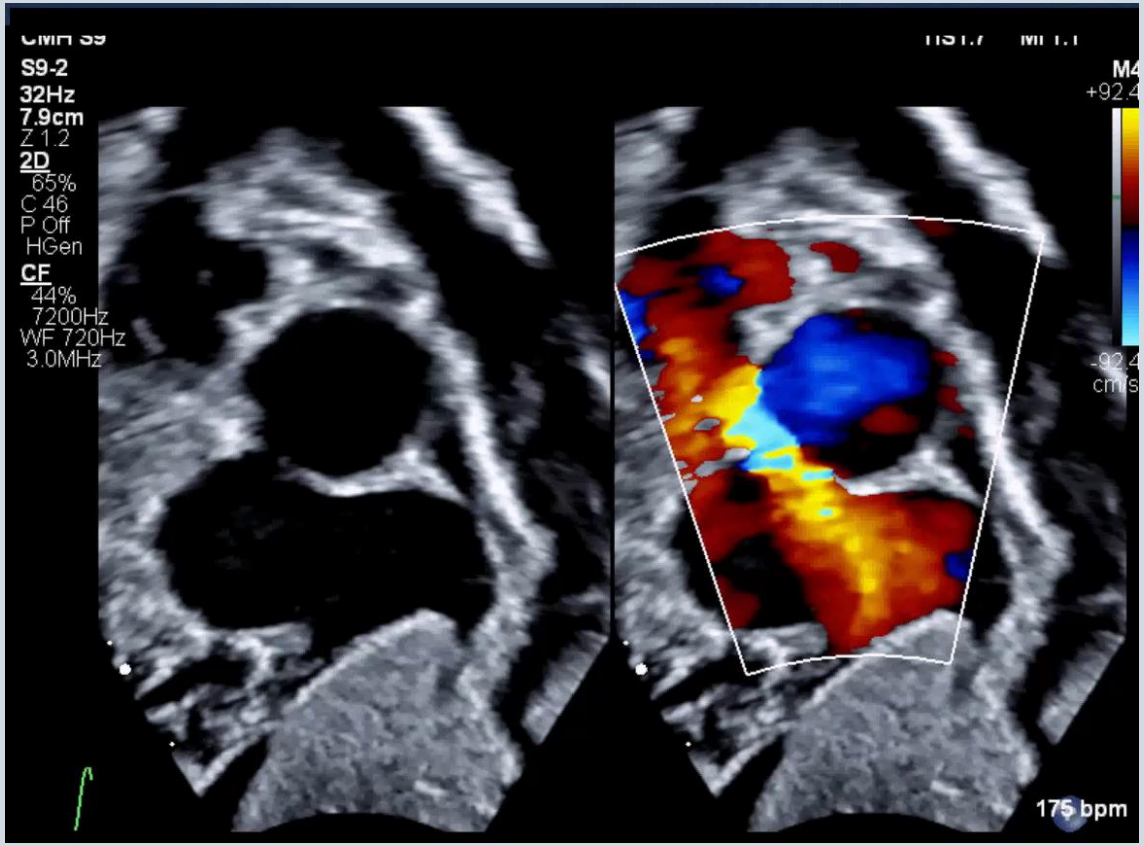

CMH FETAL
C5-1
31Hz
GA 27w1d
2D
47%
Dyn R 50
P Low
HGen
CF
67%
7200Hz
WF 288Hz
3.0MHz
X2
10cm
*** bpm
cm/s


CMH FETAL
C5-1
31Hz
GA 27w1d
TIB0.6
MI 1.0
2D
52%
Dyn R 50
P Low
HGen
CF
63%
6000Hz
WF 270Hz
3.0MHz
X2
9.5cm
*** bpm
cm/s

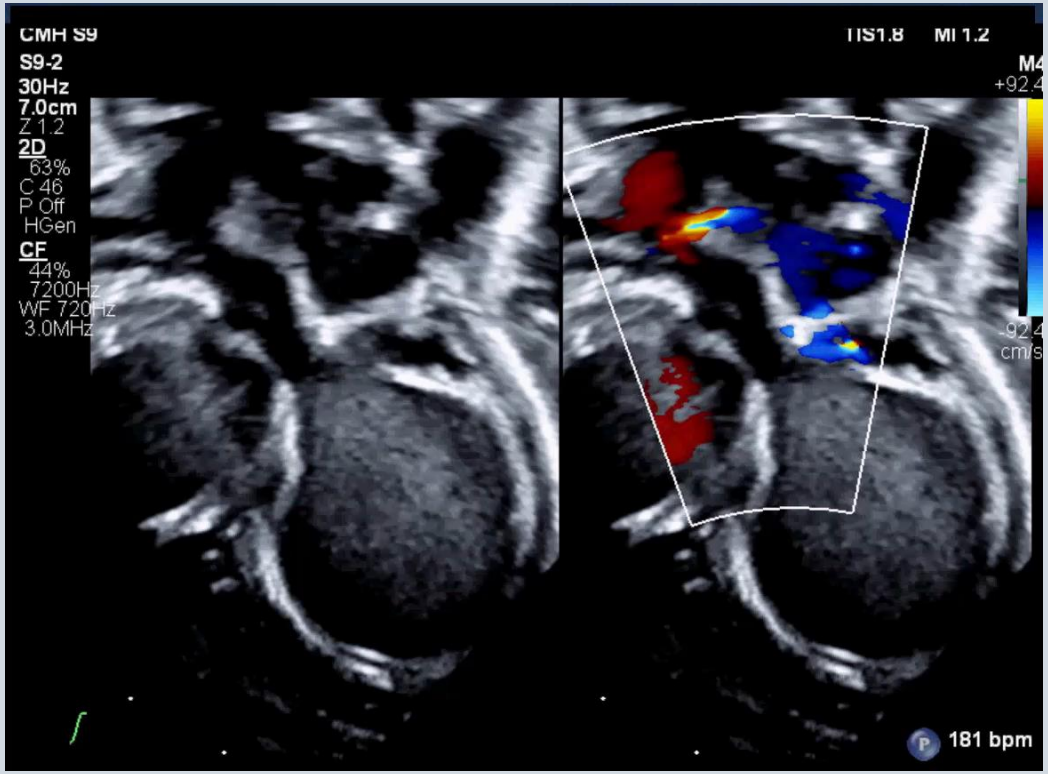

CMH FETAL
C5-1
20Hz
GA 27w1d
TIB0.4 MI 1.0
2D
51%
Dyn R 50
P Low
HGen
CF
63%
3900Hz
WF 312Hz
3.3MHz
X2
9.8cm
*** bpm

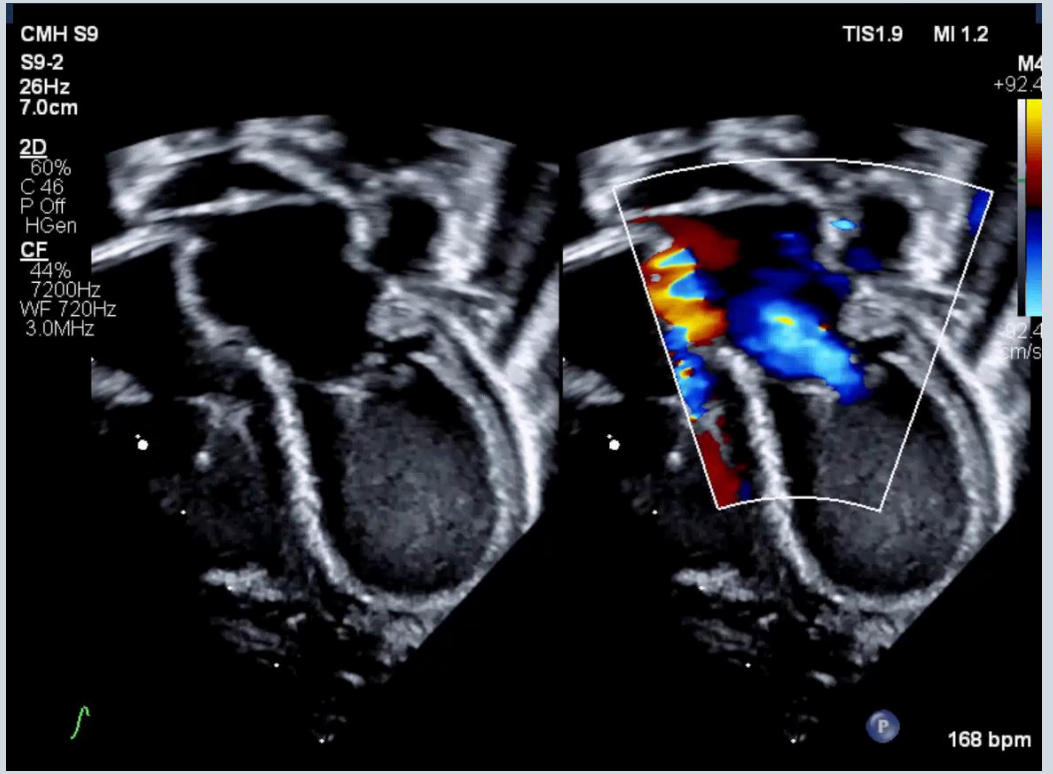
- Fetus determined not a candidate for fetal intervention (balloon valvuloplasty) due to:
  - Left ventricular dilation and function
  - EFE-echo brightness of myocardium and mitral valve papillary muscles- recovery of LV function thought to be unlikely
- Evolving hypoplastic left heart syndrome
  - Single ventricle palliation

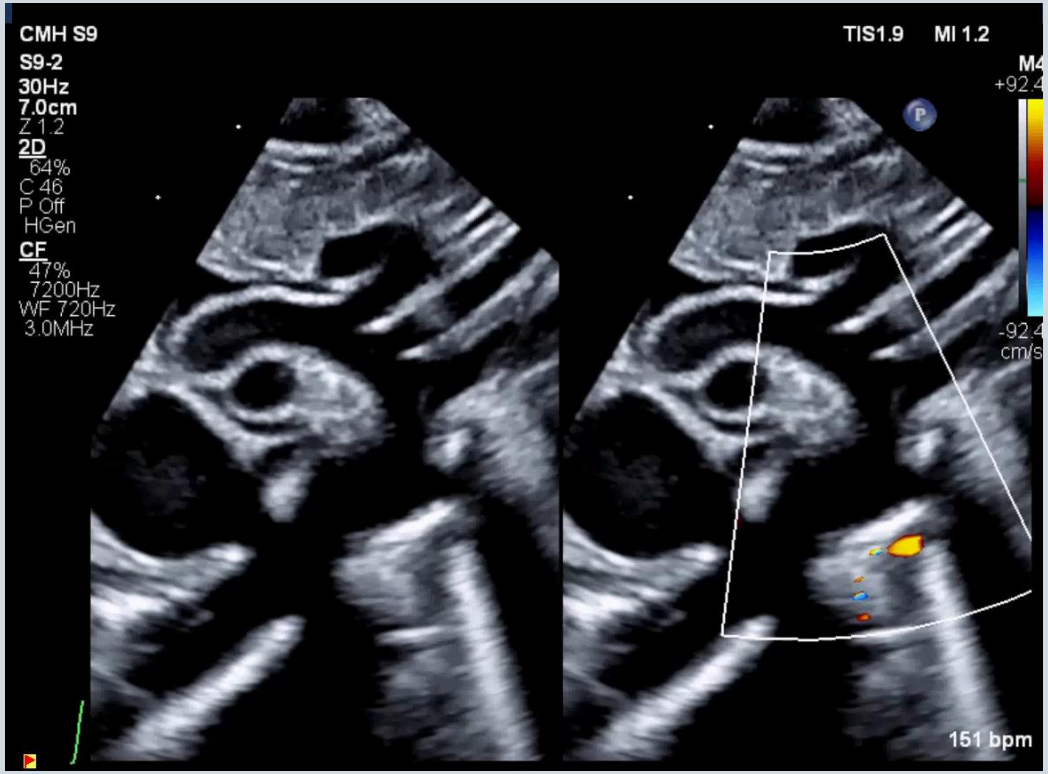
Postnatal echo

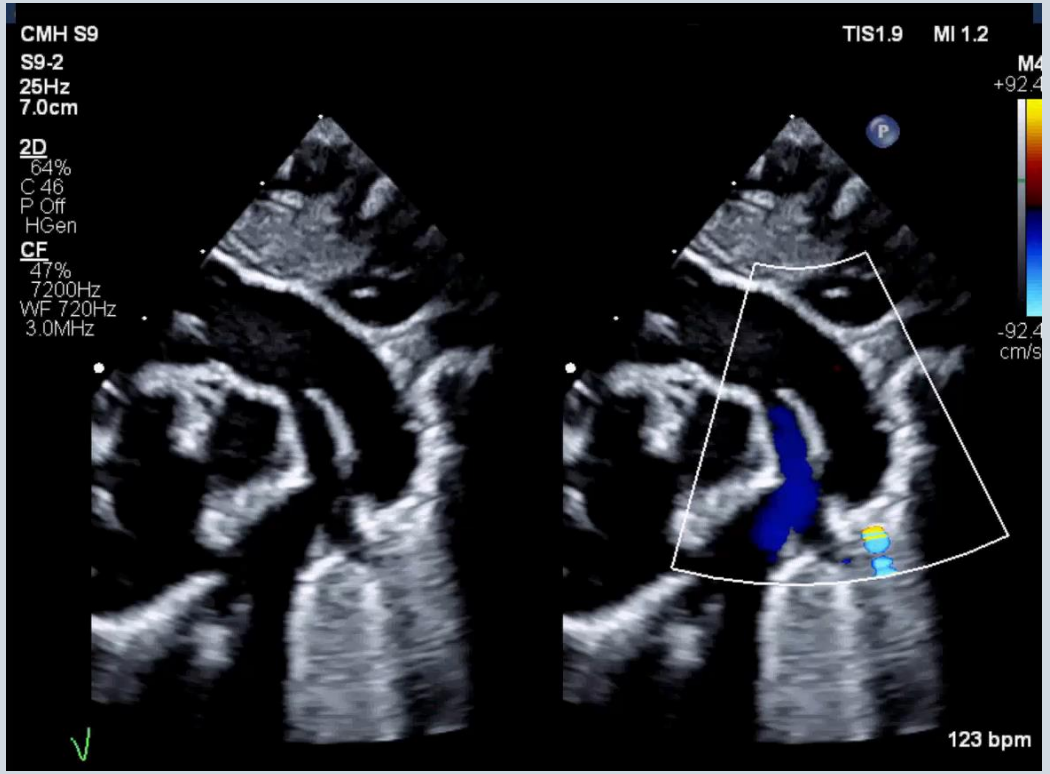


- Baby required prostaglandins immediately after birth to keep patent ductus arteriosus open
- Clinically was shown to have reasonable cardiac output so immediate intervention not needed
- Close monitoring of RV function will determine next steps to take:
  - Hybrid
  - Norwood/shunt

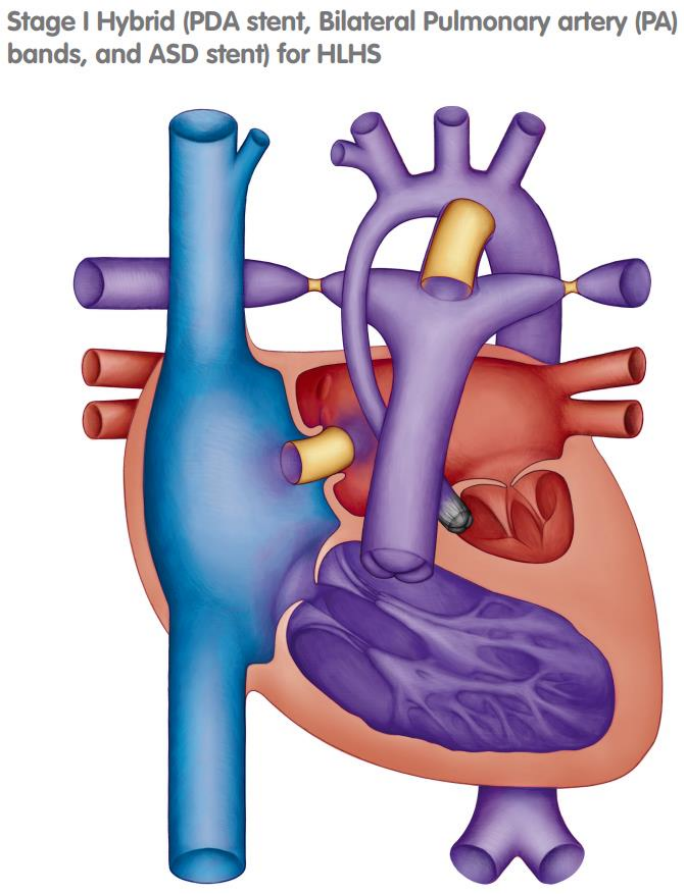

CMH S9
TIS1.8 MI 1.2
S9-2
30Hz
7.0cm
Z 1.2
2D
63%
C 46
P Off
HGen
CF
44%
7200Hz
WF 720Hz
3.0MHz
181 bpm


CMH S9
TIS1.9 MI 1.2
S9-2
26Hz
7.0cm
2D
60%
C 46
P Off
HGen
CF
44%
7200Hz
WF 720Hz
3.0MHz
168 bpm

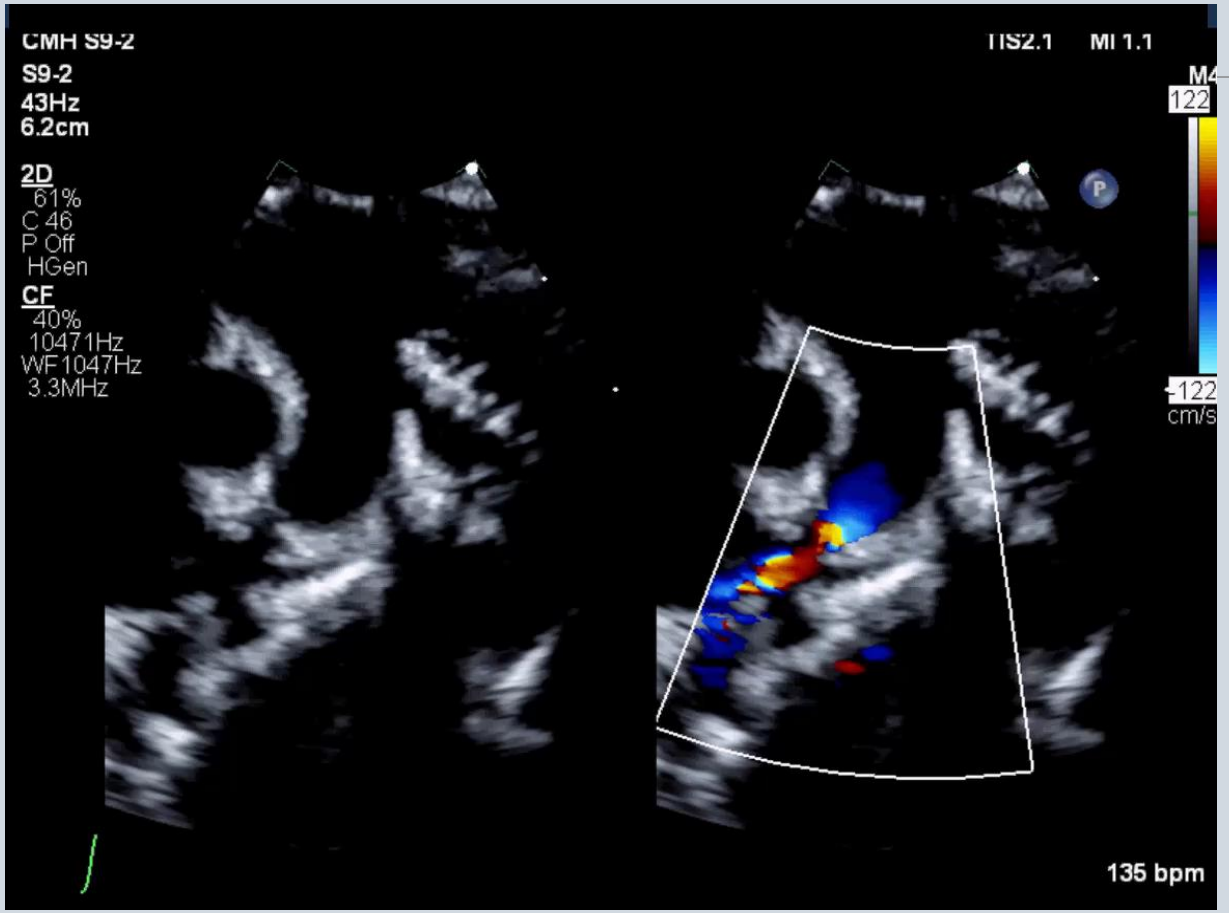

CMH S9
S9-2
30Hz
7.0cm
Z 1.2
2D
64%
C 46
P Off
HGen
CF
47%
7200Hz
WF 720Hz
3.0MHz
TIS1.9 MI 1.2
M4
+92.4
-92.4
cm/s
151 bpm

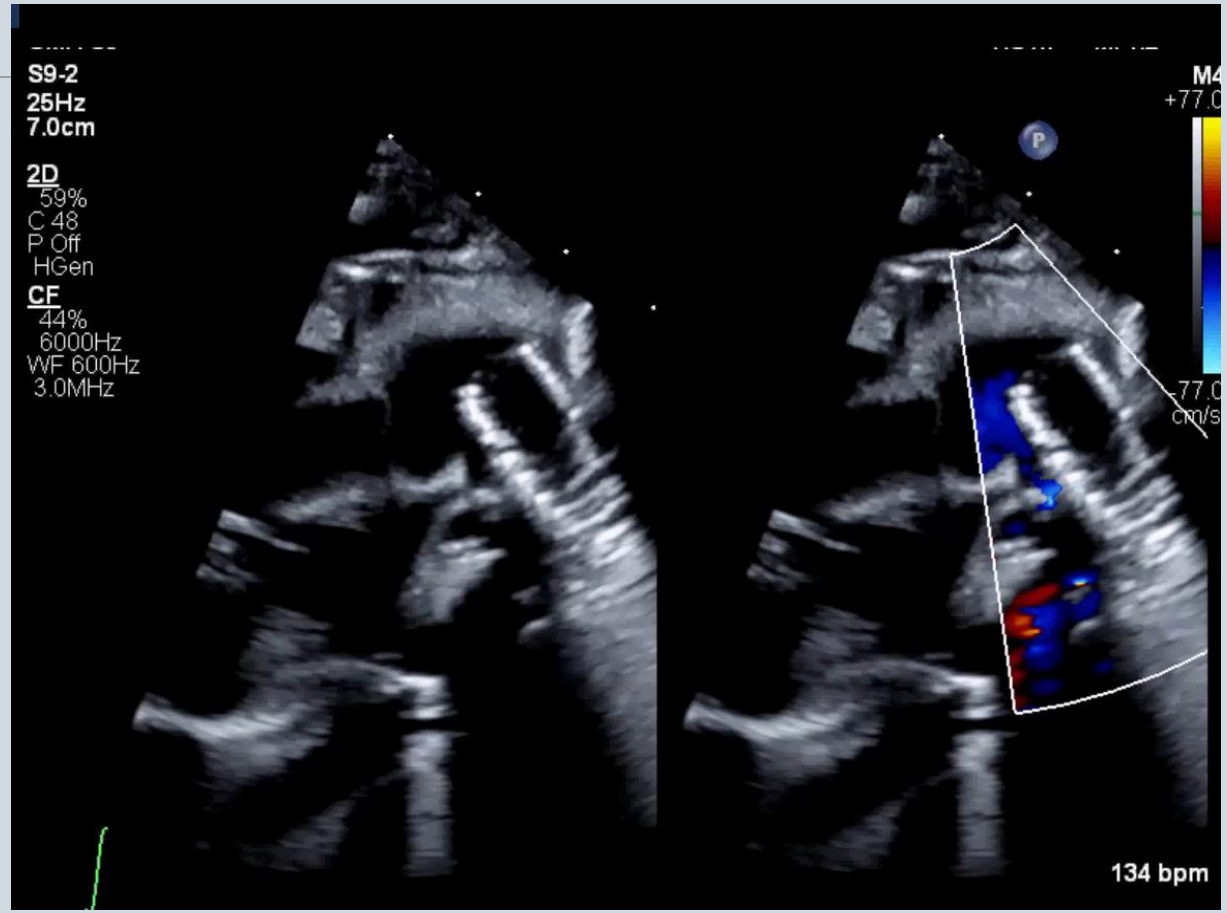

CMH S9
S9-2
25Hz
7.0cm
2D
64%
C 46
P Off
HGen
CF
47%
7200Hz
WF 720Hz
3.0MHz
TIS1.9 MI 1.2
M4
+92.4
-92.4
cm/s
123 bpm

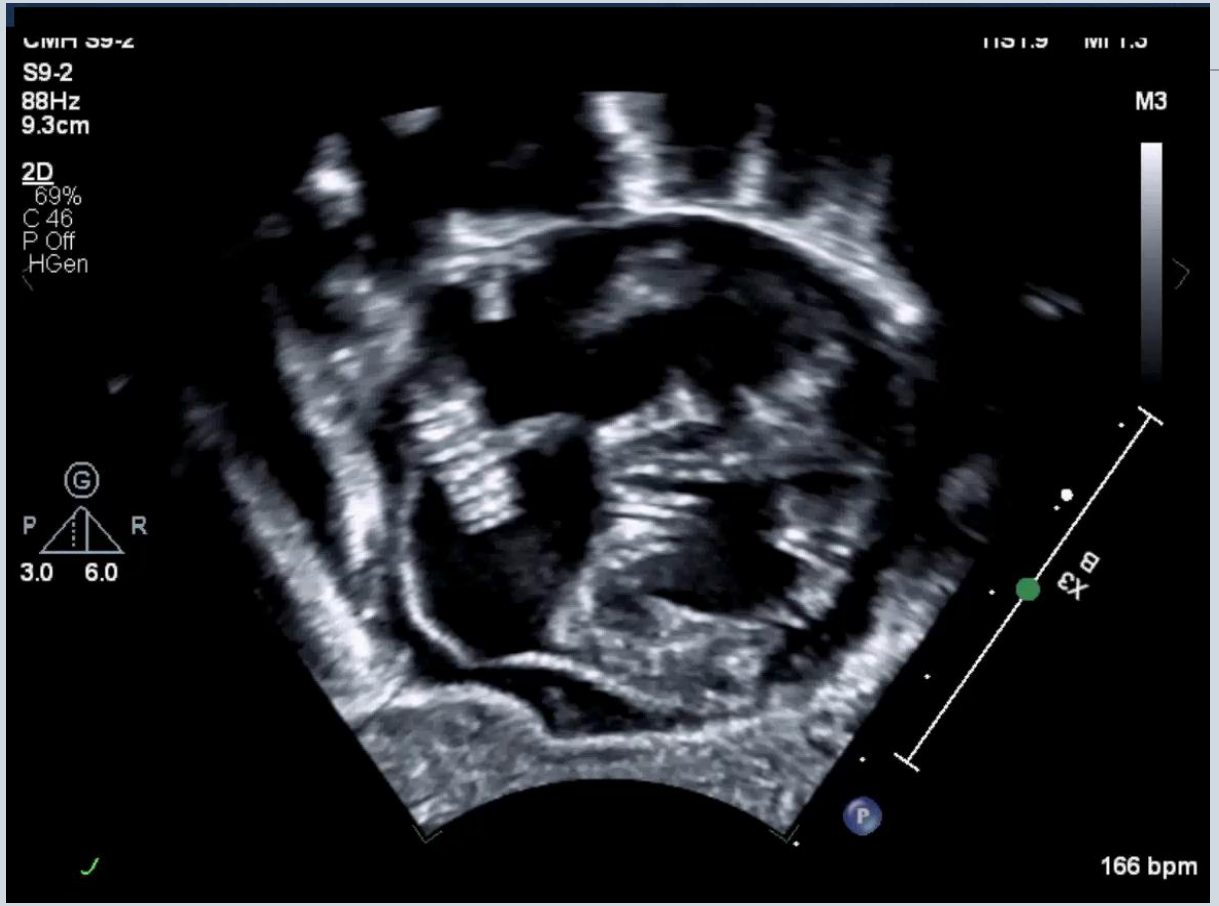
Hybrid procedure

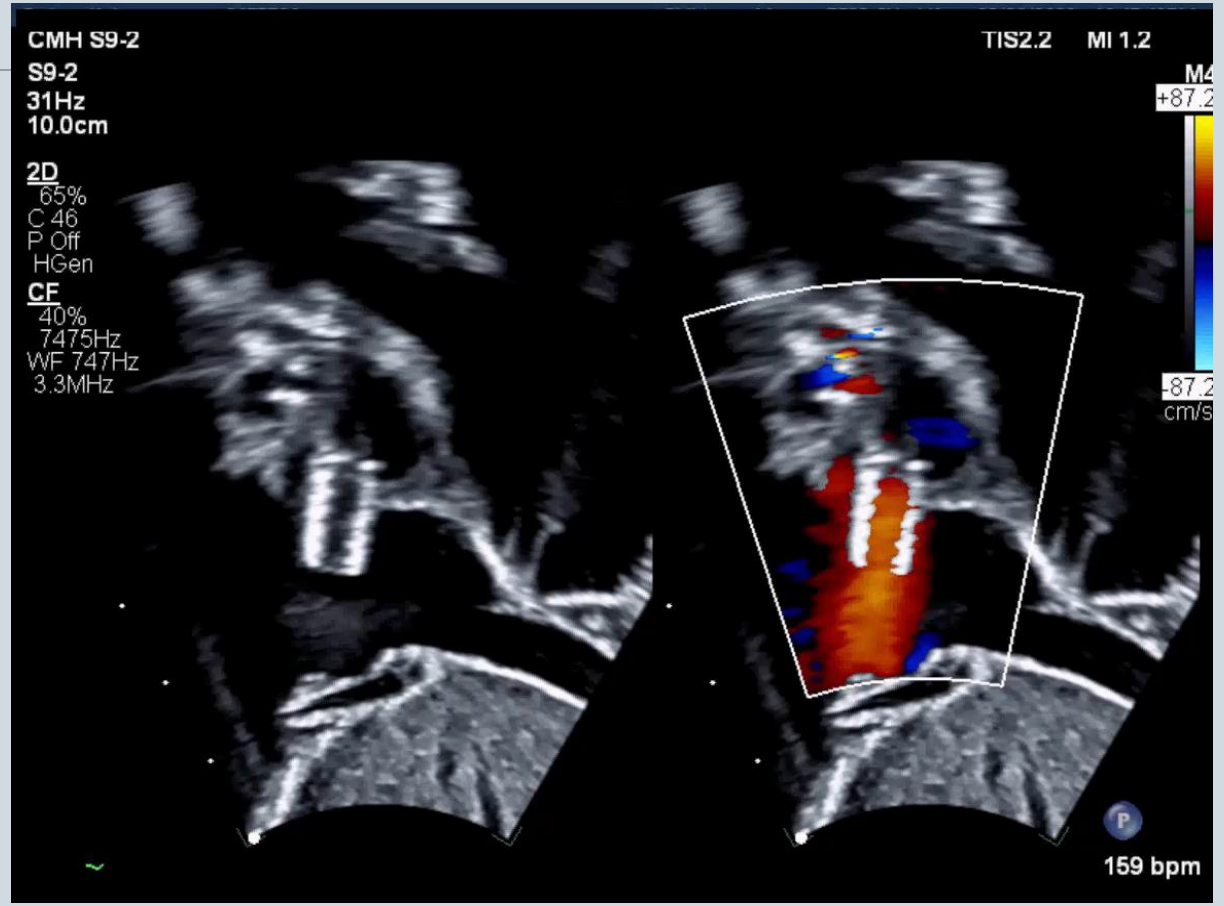


- Placement of PA bands
  - Balances circulation to the lungs and body
- PDA stent
  - Keeps the PDA open and maintains the connection to the aorta and the body's circulation
- Atrial septostomy or stenting of PFO
  - To provide open blood flow and mixing of the oxygenated and deoxygenated blood between the atria


S9-2
43Hz
6.2cm
2D
61%
C 46
P Off
HGen
CF
40%
10471Hz
WF1047Hz
3.3MHz
122
-122
cm/s
135 bpm

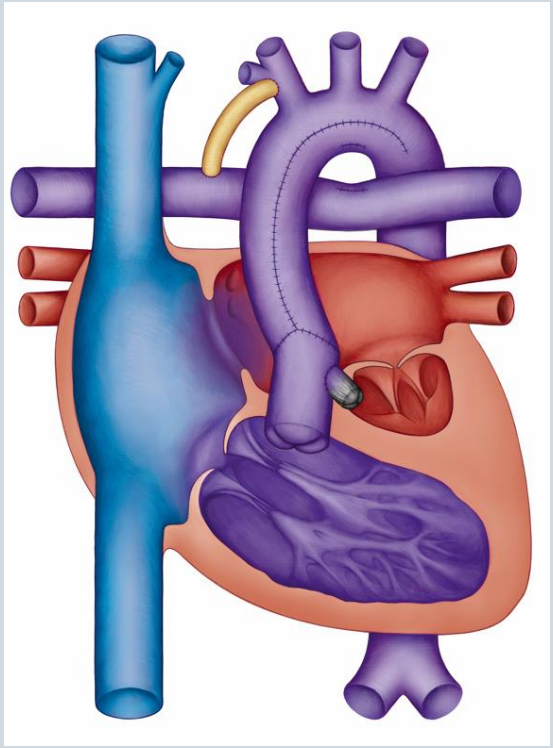

S9-2
25Hz
7.0cm
2D
59%
C 48
P Off
HGen
CF
44%
6000Hz
WF 600Hz
3.0MHz
cm/s
134 bpm


S9-2
88Hz
9.3cm
2D
69%
C 46
P Off
HGen
M3
G
P
R
3.0
6.0
166 bpm

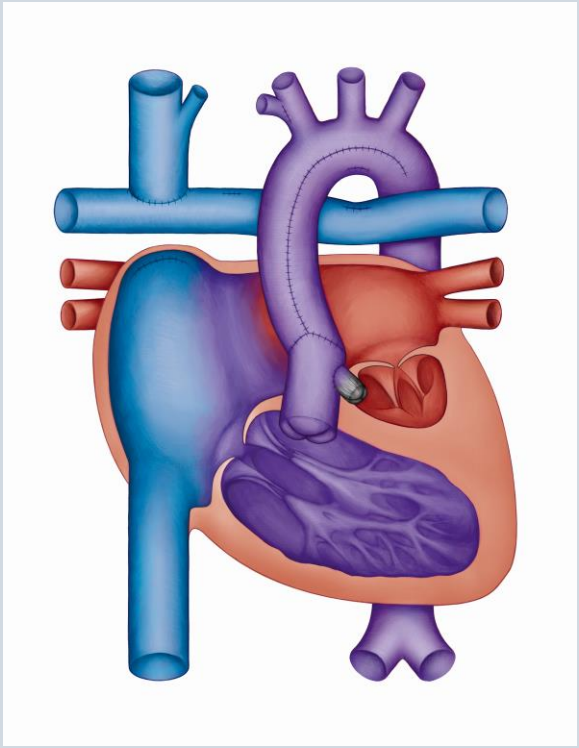

CMH S9-2
TIS2.2
MI 1.2
S9-2
31Hz
10.0cm
2D
65%
C 46
P Off
HGen
CF
40%
7475Hz
WF 747Hz
3.3MHz
+87.2
-87.2
cm/s
159 bpm

The single ventricle

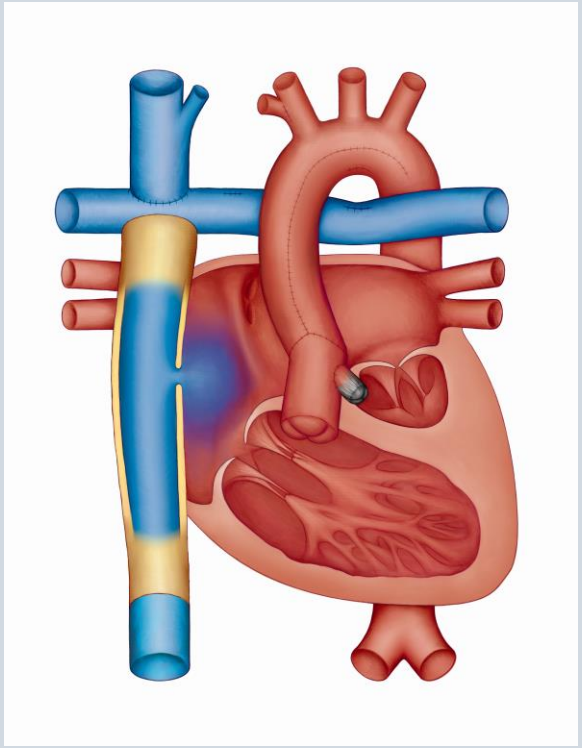
Norwood/shunt



Bidirectional Glenn 4-6 months



Fontan palliation 2-3 years

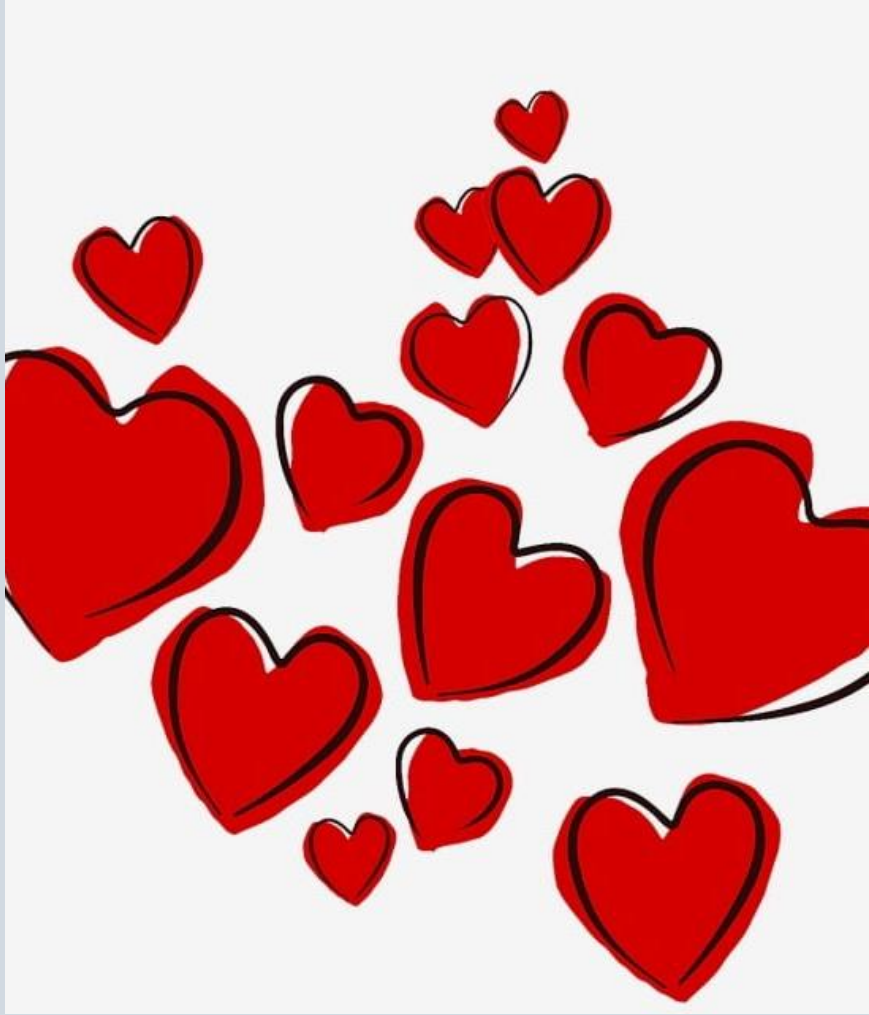


Helpful resources

- Dr. Nitin Madan's prior talk on hypoplastic left heart syndrome
  - August 2021 education series talk
- Anticipation, Accompaniment, and a Good Death in Perinatal Care
  - Bryanna S. Moore , Brian S. Carter, Bryan Beaven, Katie House, and Joel House

Summary

- Prenatal detection is extremely important in the diagnosis of aortic stenosis
  - Aortic stenosis has multiple avenues it can evolve into, and time is of the essence for these fetuses
- Prenatal care and consults give patients time to help determine the route and pathway patient will go down whether they choose fetal intervention, post natal surgery or comfort care
  - Mentally prepares patient for whatever decision they make
- Ultrasound imaging is very important piece in patient care, early detection and monitoring can make a huge difference in decisions



Thank YOU!! And Happy Valentines Day!

CONTACT: ALISON SAMRANY AMSAMRANY@CMH.EDU

Echocardiography in pediatric and congenital heart disease: from fetus to adult

Lai, Wyman W. 2009

Predictive Value of Fetal Pulmonary Venous Flow Patterns in Identifying the Need for Atrial Septoplasty in the Newborn With Hypoplastic Left Ventricle

- Erik Michelfelder, MD; Carlen Gomez, MD; William Border, MBChB; William Gottliebson, MD; Cheri Franklin, CNP

Fetal Aortic Valve Stenosis and the Evolution of Hypoplastic Left Heart Syndrome

Patient Selection for Fetal Intervention

Kaarin Mäkikallio, Doff B. McElhinney, Jami C. Levine, Gerald R. Marx, Steven D. Colan, Audrey C. Marshall, James E. Lock, Edward N. Marcus and Wayne Tworetzky