# Hypoplastic left heart syndrome


Nitin Madan, MD

Associate Director, Echo Lab

Ward Family Heart Center, Children's Mercy Kansas City

Assistant Professor of Pediatrics, UMKC School of Medicine


August 10th 2021

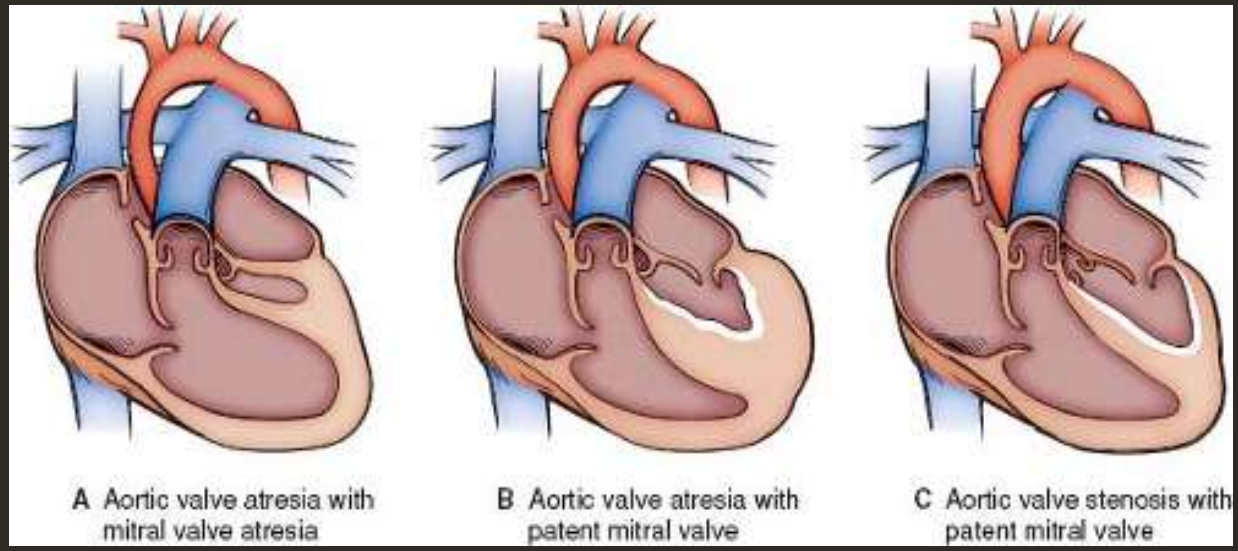
# Objectives

- Hypoplastic left heart syndrome (HLHS) definition
- Classification of HLHS
- Incidence
- Prenatal physiology and management
- Postnatal management
- Outcomes
- Case discussion for HLHS and rare interesting variants

# HLHS definition

Left side of the heart is inadequate to sustain systemic perfusion

- MA/AA
- MS/AA
- MS/AS
- Additional variant of MVD/AS



# Frequency

- 0.26 per 1000 live births
- Most commonly diagnosed prenatal CHD
- Multiple theories to HLHS occurrence
  - Abnormal atrial septal anatomy
  - Abnormal LV compliance
  - Aortic stenosis as primary anomaly
  - Compression from large cystic hygroma
  - Perhaps multiple pathways contribute to a nonviable left side

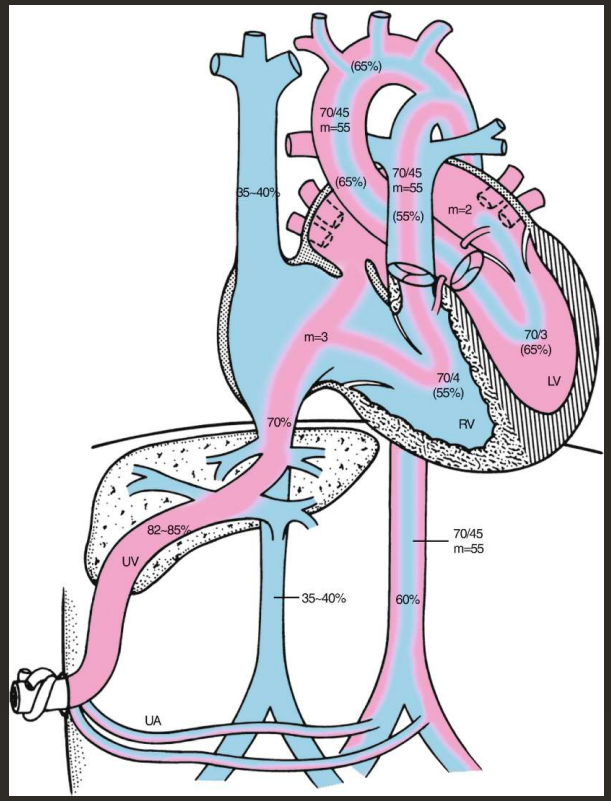
# Genetics

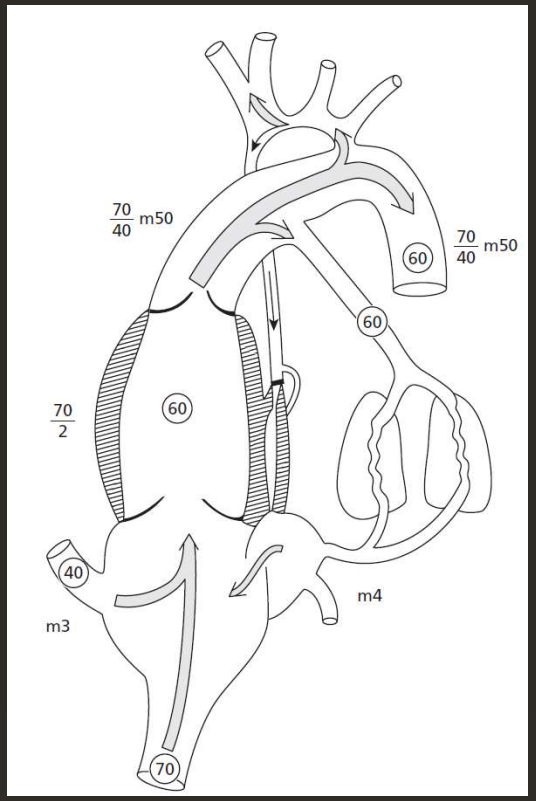
- Chromosomal abnormalities in 5-10% (Trisomy 13, 18 and Turner's syndrome)
- Smith-Lemli-Opitz syndrome (microcephaly, mental retardation, CHD, extra fingers and toes, fused second and third toes, cleft palate, underdeveloped external genitals)
- Holt-Oram (Skeletal anomalies of hands/arms and CHD)
- VACTERL (Vertebral anomalies, anal atresia, cardiac abnormalities, TEF/esophageal atresia, renal agenesis/dysplasia and limb defects)
- Rarely chromosome 22q11 deletion

# Recurrence risk

- Normal CHD incidence 1%
- Recurrence risk with other CHD 2-3%
- One child with HLHS, risk of CHD in another sibling 13-14%

# Prenatal physiology normal vs. HLHS



# Fetal imaging goals

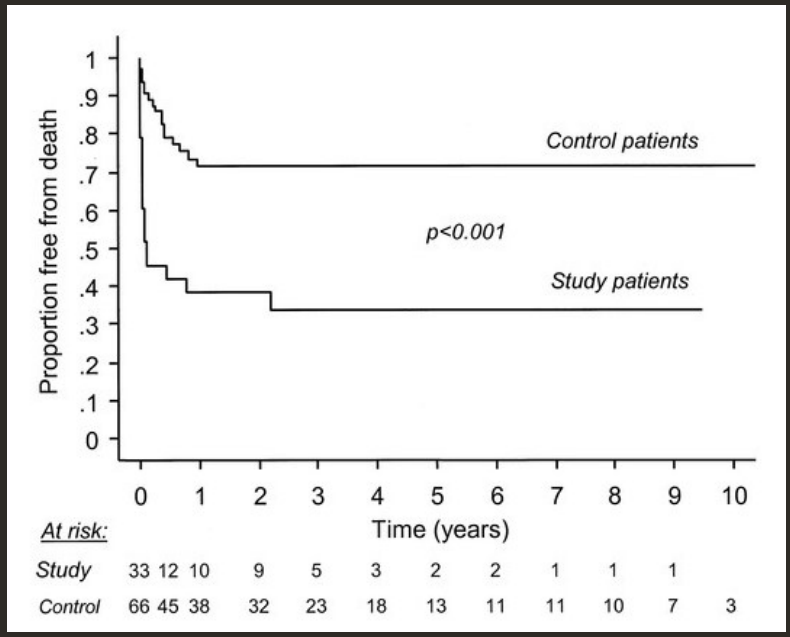
- Size and function of LV
- Atrial septal anatomy and direction of shunting at the atrial level
- Mitral valve size and function
- Aortic valve size and function
- Direction of flow in aortic arch
- Pulmonary vein dopplers
- Tricuspid valve and right ventricular function
- Assessment for coronary sinusoids

# Predictors of LV inadequacy

- Left to right shunt at the atrial level
- Mitral valve Z score < -3
- Aortic valve Z score < -3
- Severe LVOT obstruction
- Endocardial fibroelastosis
- Retrograde blood flow in the ascending aorta

# The atrial septum and pulmonary vasculature

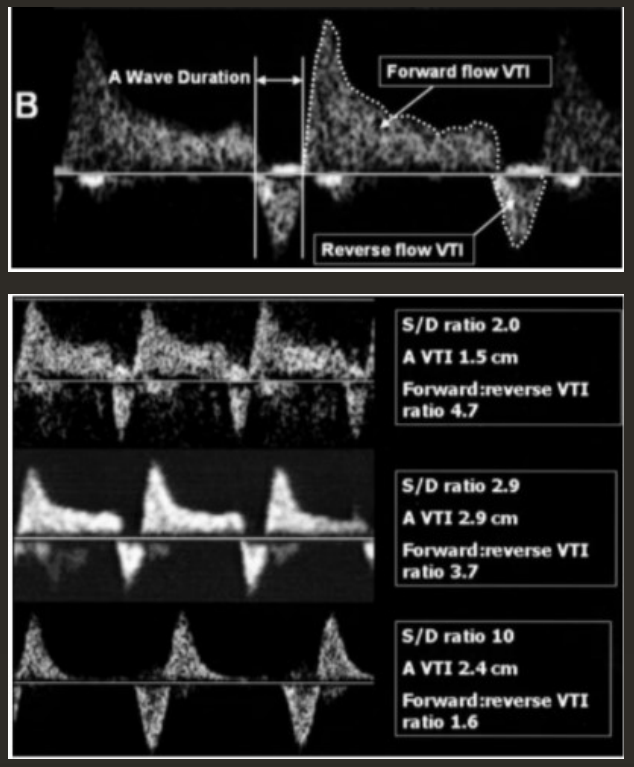
- Atrial septum patency is critical in HLHS
- A severely restrictive or intact atrial septum ~6% HLHS
- Elevated left atrial pressure
- Dilated pulmonary veins with arterialized walls
- Fetus asymptomatic but extreme cyanosis after birth
- Even early postnatal intervention associated with poor outcomes




Children's Mercy

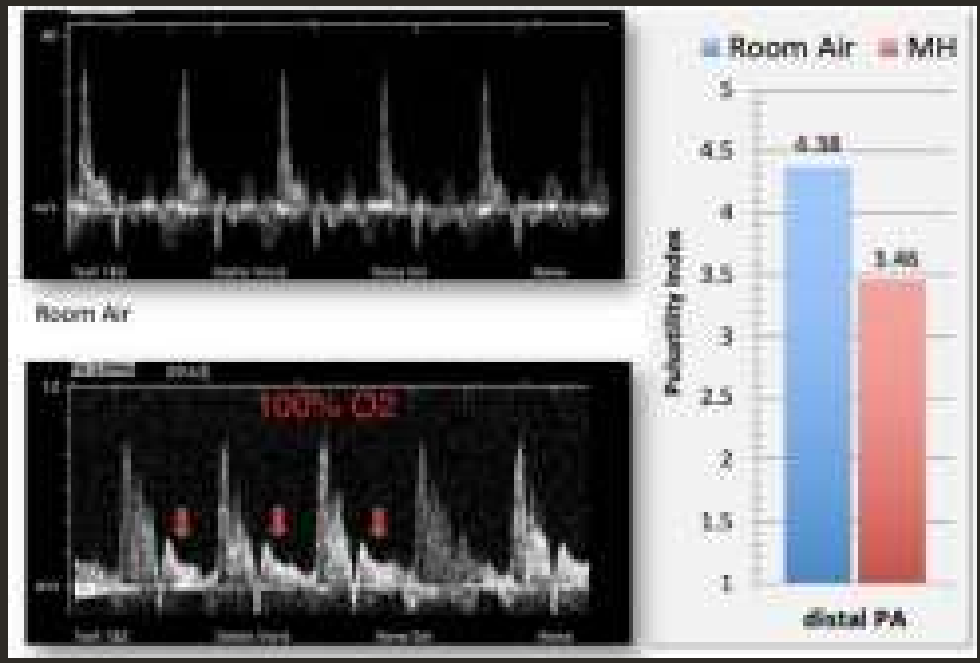
# Fetal pulmonary vasculature

- Pulmonary vein doppler assessment
- Forward to reverse velocity time integral (VTI)
- Forward/Reverse VTI <5 predictor of atrial septal restriction and need or urgent BAS



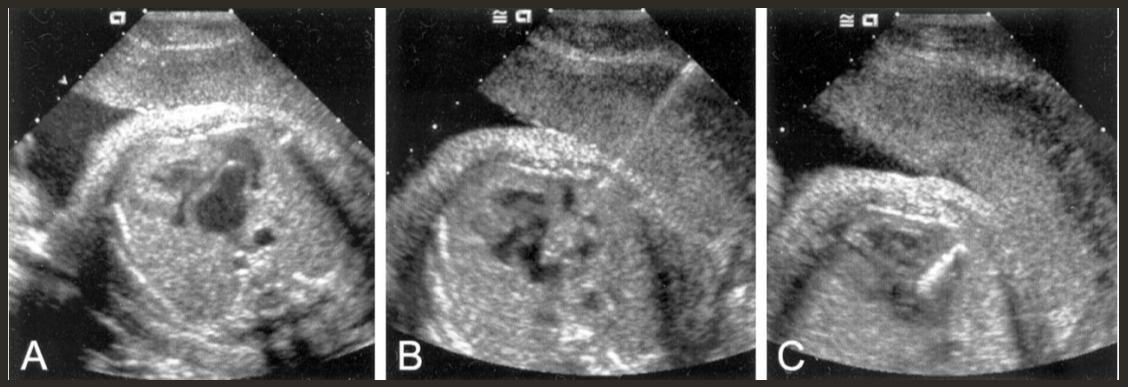
# Maternal hyperoxygenation (MH)

- To assess pulmonary vasoreactivity
- Hyperoxygenation results in pulmonary vasodilation
- Typically evaluation of PA dopplers on RA followed by 15 minutes of MH
- Response is blunted with atrial septal restriction <10% decrease in PI



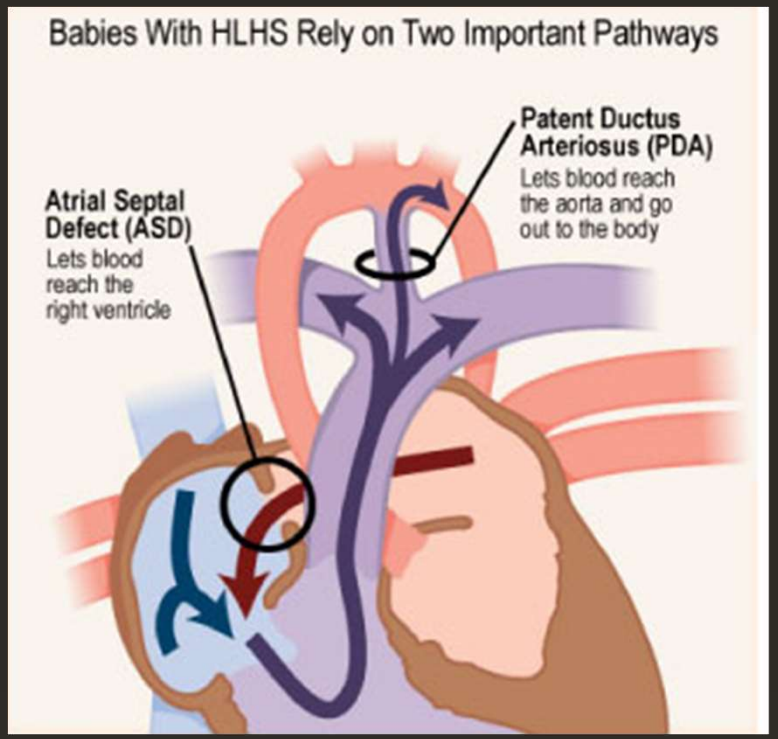
# Prenatal atrial septal intervention

- Balloon atrial septoplasty or stenting
- 26-34 weeks of gestation
- Few centers
- 2017 International Fetal Cardiac Intervention Registry: Survival to discharge did not differ between FCI or no FCI



# Postnatal management newborn

Prostaglandin initiation
Non-restrictive atrial septum
Surgical strategy - 3 stage palliation

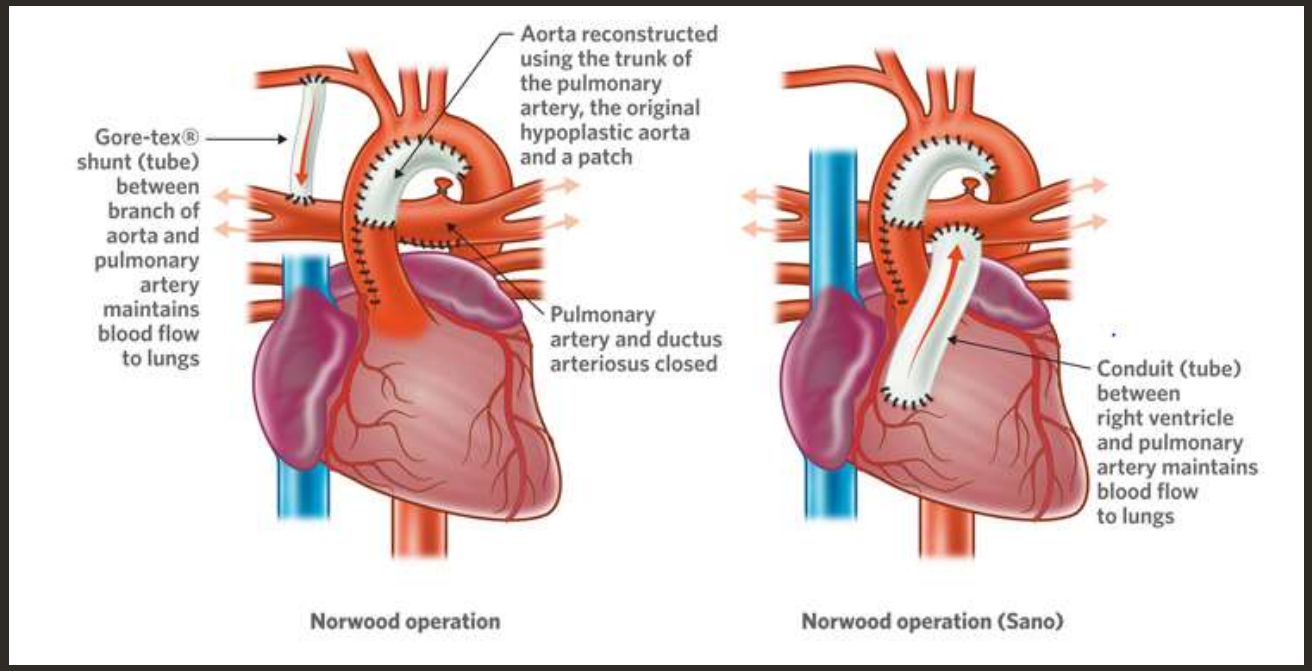


# Stage I Norwood

Norwood Procedure

(first week of life)

- PA to aorta anastomosis
- Arch reconstruction
- BT shunt or Sano
- PDA ligation
- Atrial septectomy



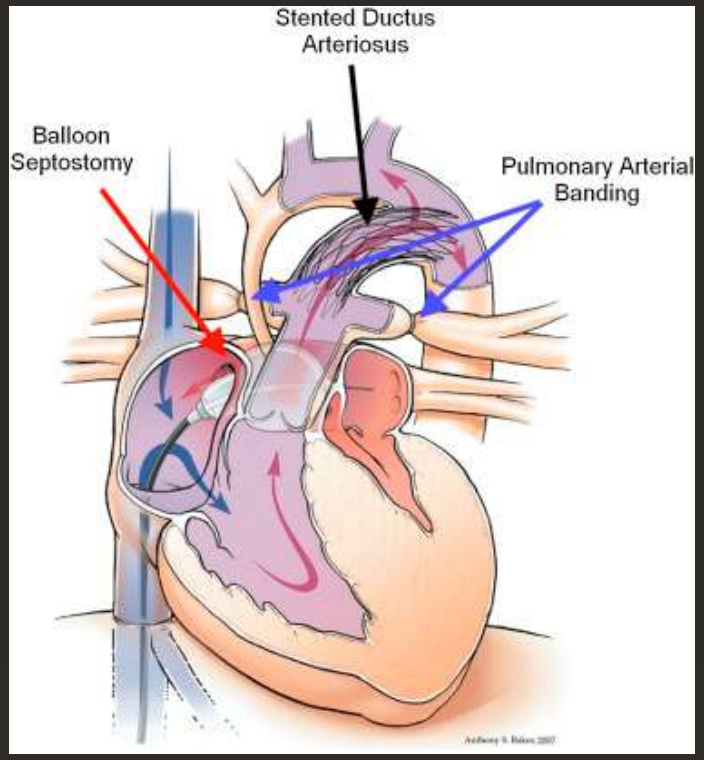
# Hybrid

Hybrid procedure

- PDA stenting
- Balloon atrial septostomy/stent
- Bilateral PA bands

Commoner with risk factors

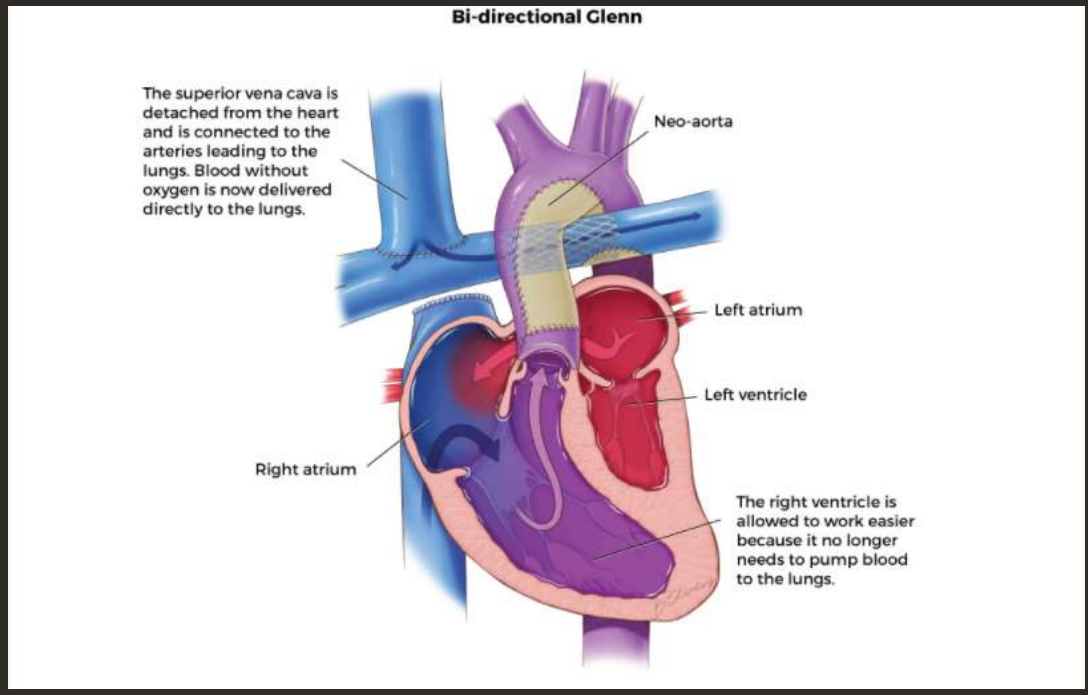
- Prematurity
- Low birth weight
- Chromosomal abnormalities
- Organ dysfunction (shock, NEC etc)



# Stage II Glenn

Glenn (4-6 months of age)

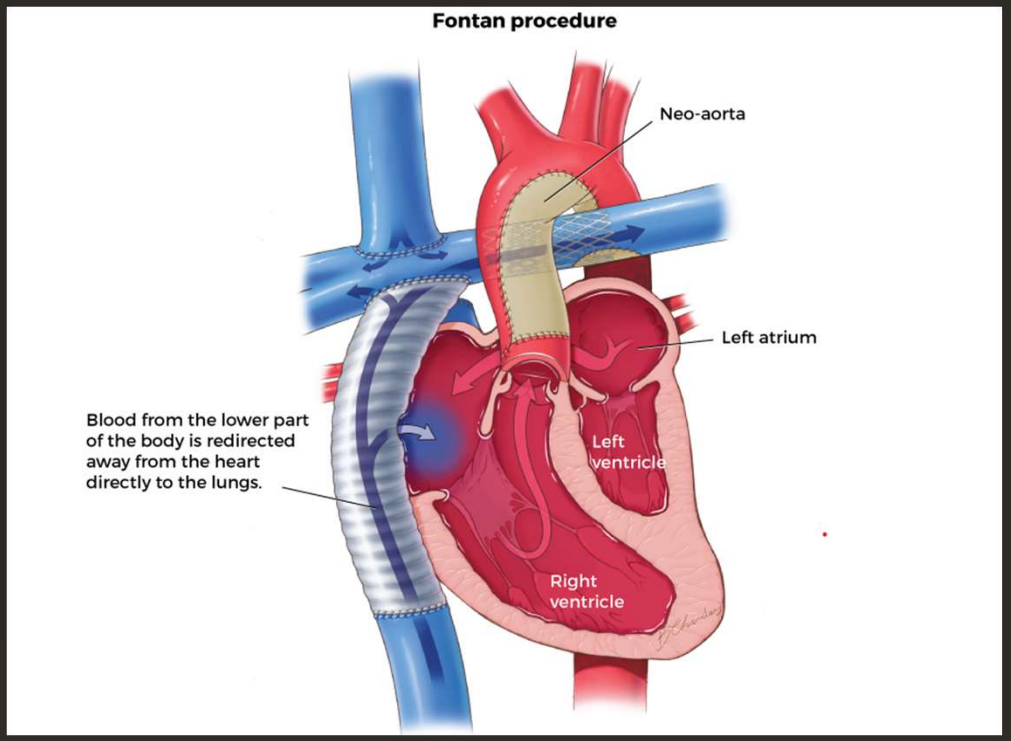
- SVC anastomosis to RPA
- Shunt/Sano is disconnected
- Eliminates high pressure pulmonary blood flow
- Early reduction of the volume work on the single ventricle



# Stage III Fontan

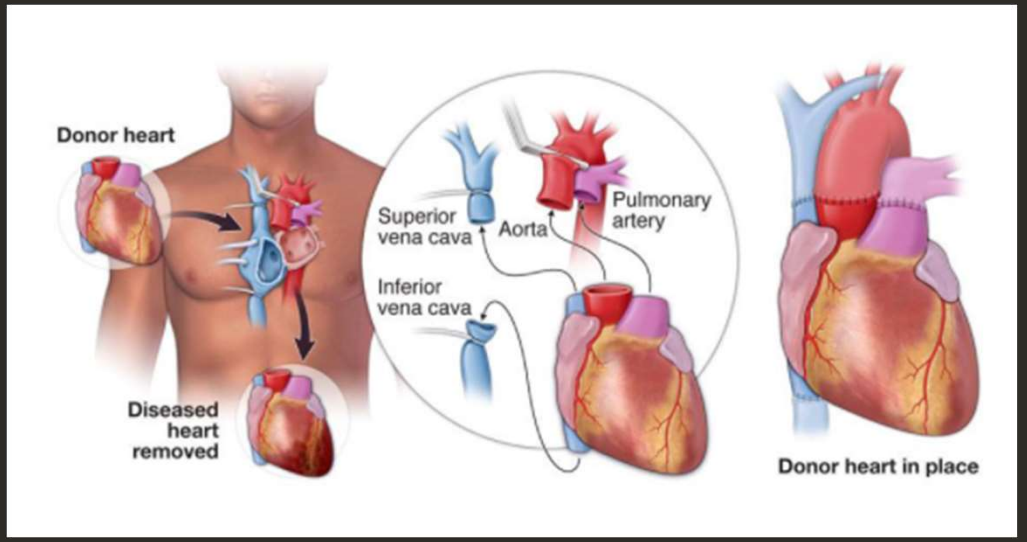
Fontan (3-5 years of age)

- IVC anastomosis to PA
- Remaining desaturated blood directly to PAs



# Primary Heart Transplant

- HLHS with reduced ventricular function and severe atrioventricular valve insufficiency
- Arch reconstruction necessary

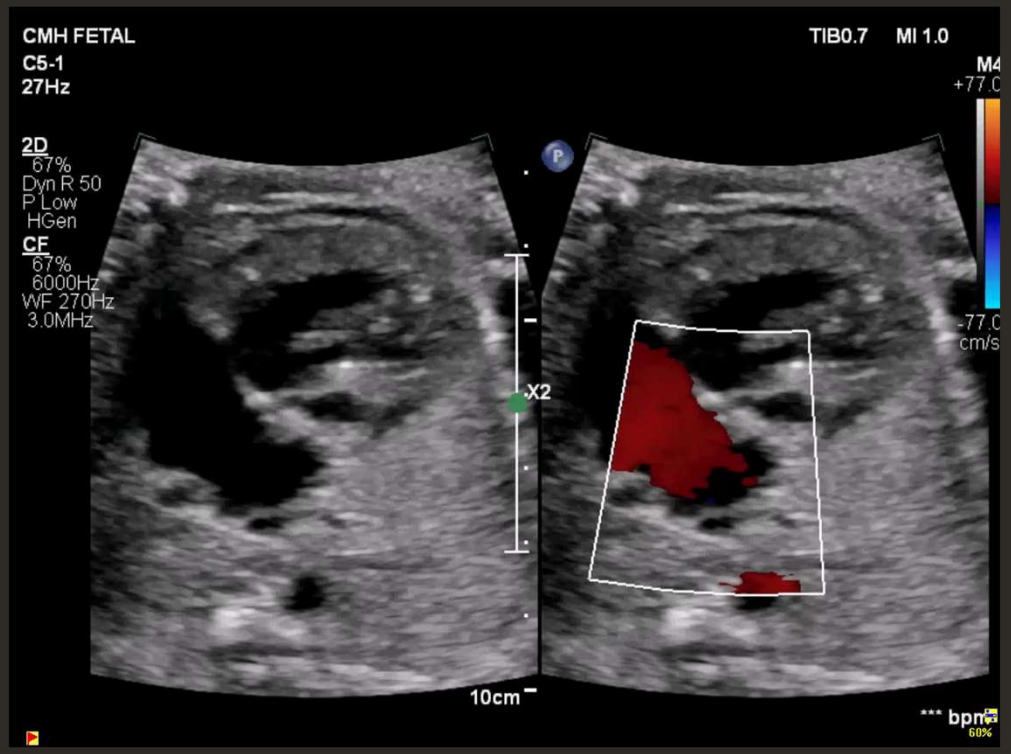


# Outcomes

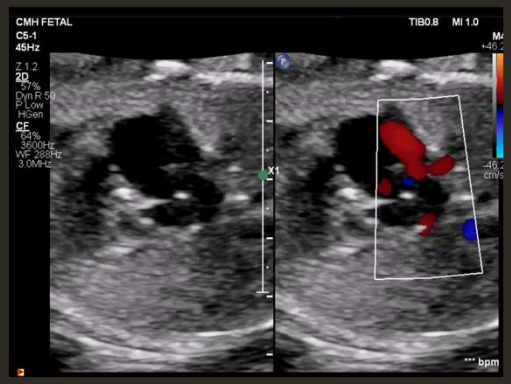
- Three-to six-year survival rates of ~70% for infants who undergo stage I repair
- For children who survive to age of 12 months, long term survival up to 18 years ~90%
- Stage I: Surgical mortality is sufficiently high. Multicenter single ventricle reconstruction trial, 1 year mortality ~30%
- High risk factors for Stage I: Prematurity, weight <2.5Kg, non cardiac diagnoses, restrictive atrial septum, MS/AA variant, very small ascending aorta, significant TR and RV dysfunction
- Stage II-III: Mortality rates are low ~ 1-3%

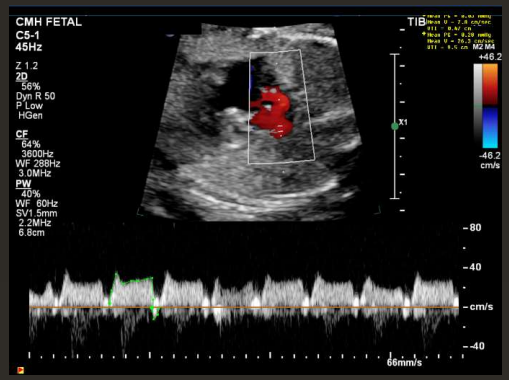
# Case 1

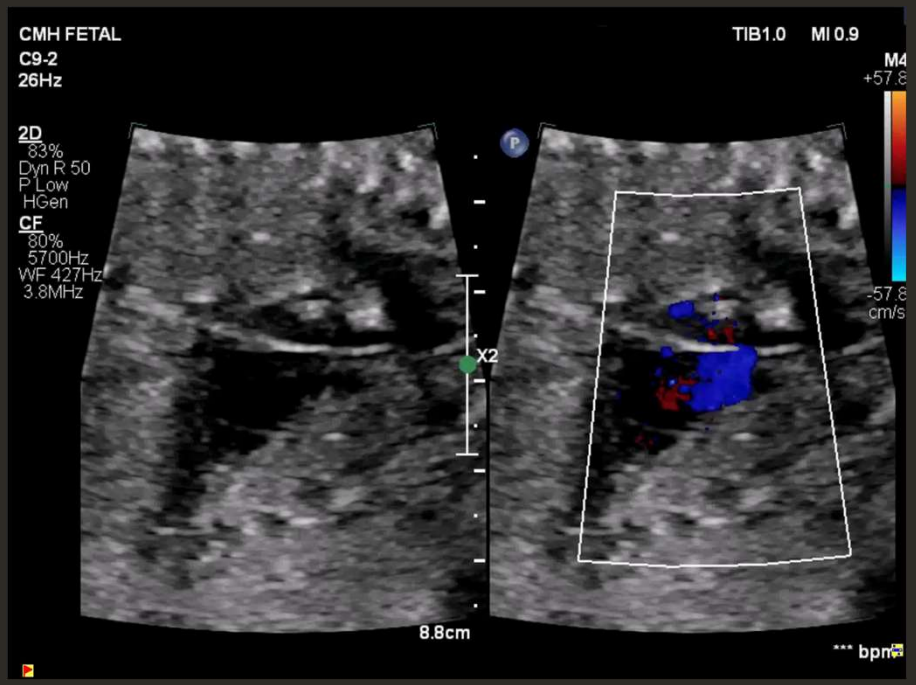
- G2 P1 referred at 24 4/7 weeks of gestation for suspected HLHS
- No family history of CHD



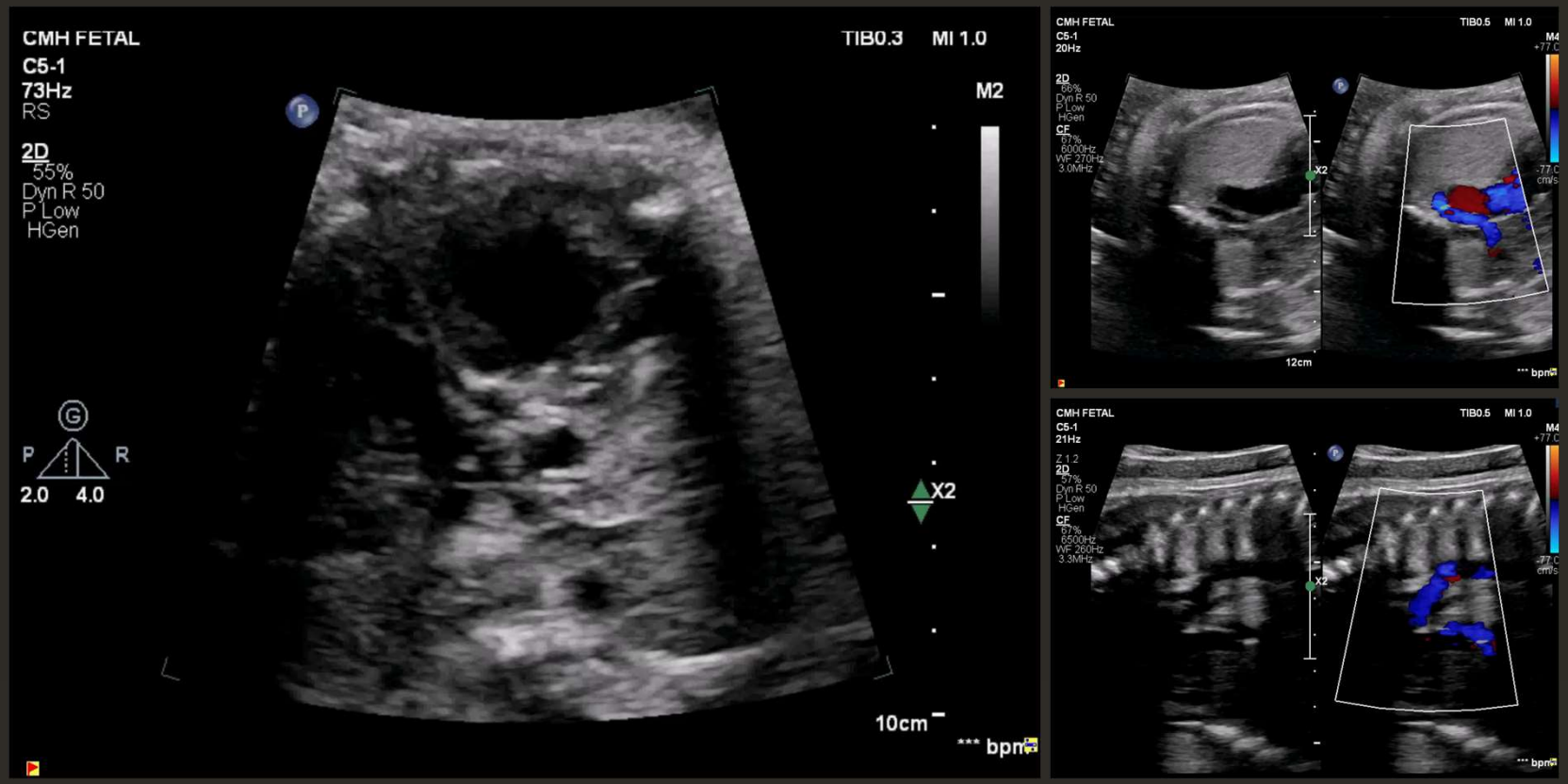
# Case 1 Atrial Septum



# Case 1 Aortic Root/Arch



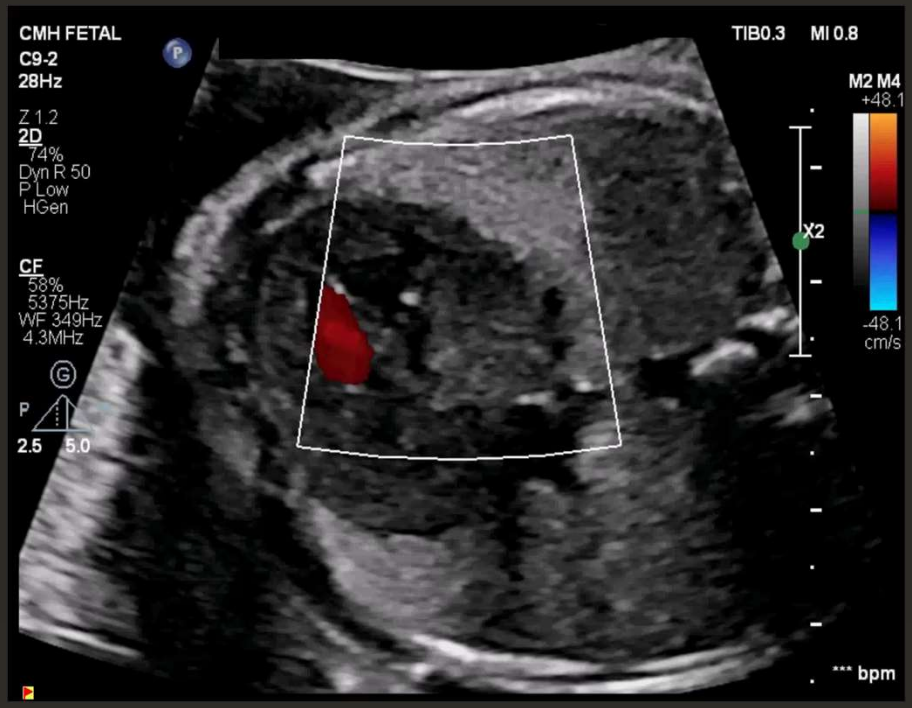

Children's Mercy

# Case 1 Postnatal outcome

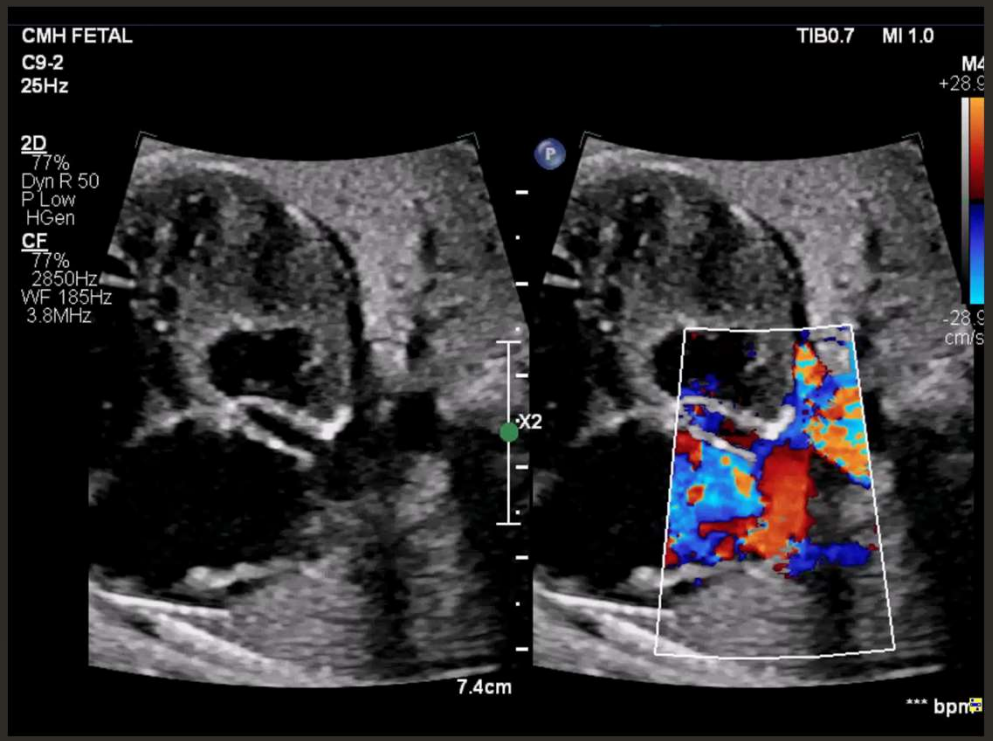
- Infant delivered at 39 weeks
- Birth weight 3.4 Kg
- HLHS MA/AA
- Norwood Sano at DOL 6
- Recovering postoperatively

# Case 2

- G5 P3 referred at 28 weeks of gestation for suspected HLHS
- No family history of CHD



# Case 2 Veins/Aorta

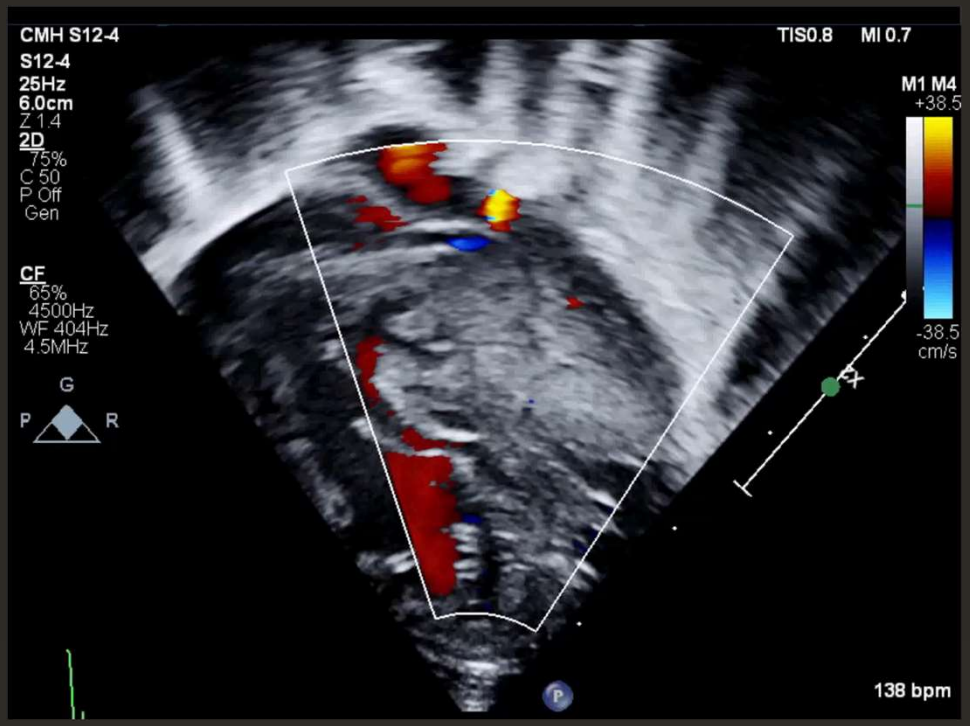




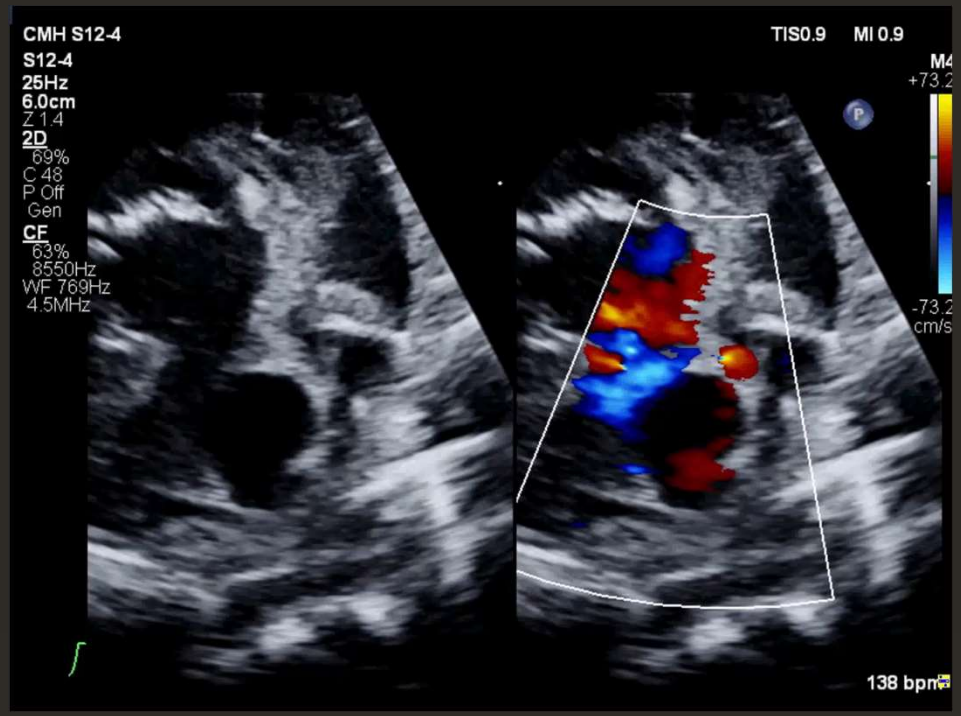
Children's Mercy

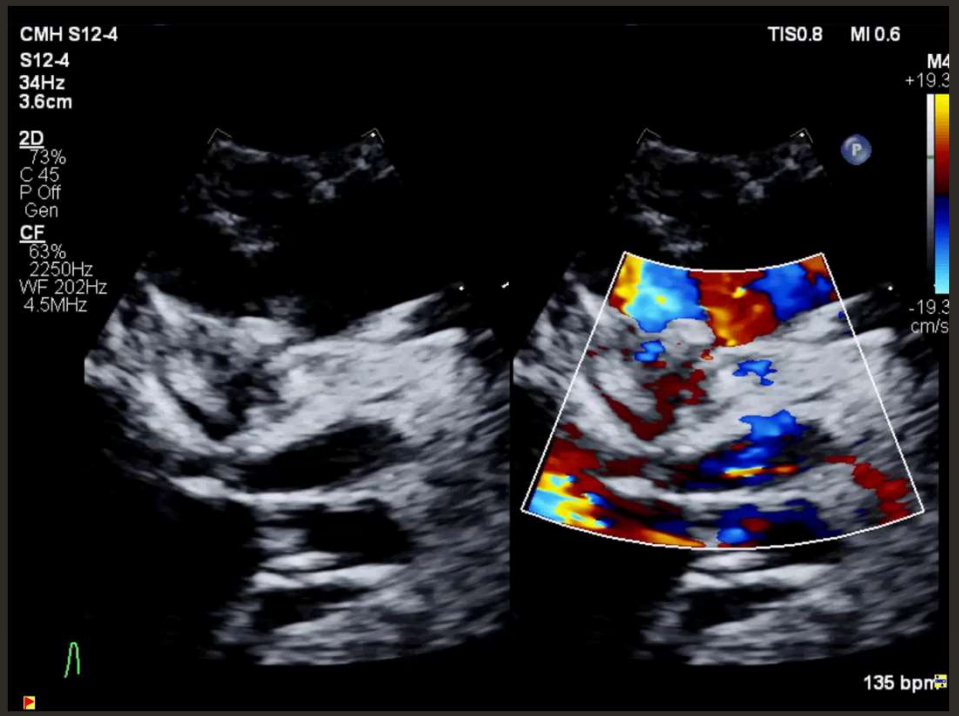
# Case 2 Postnatal

- Term infant delivered
- IUGR
- Birth weight 2.2 Kg
- HLHS MS/AA
- Concern for sinusoids



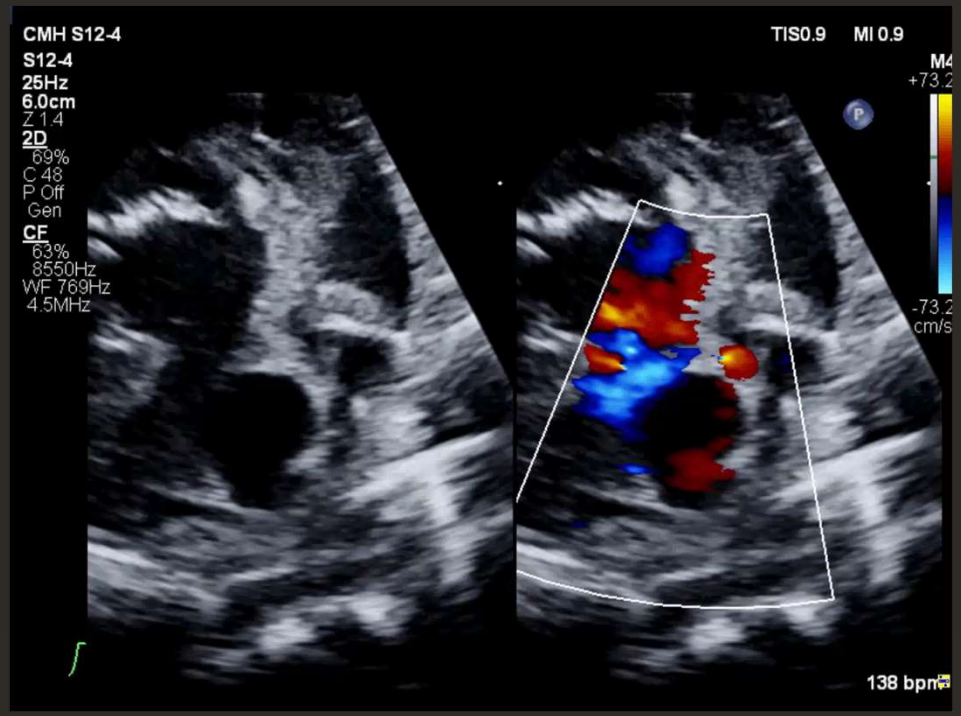
# Case 2 Postnatal Atrial Septum/ Coronaries

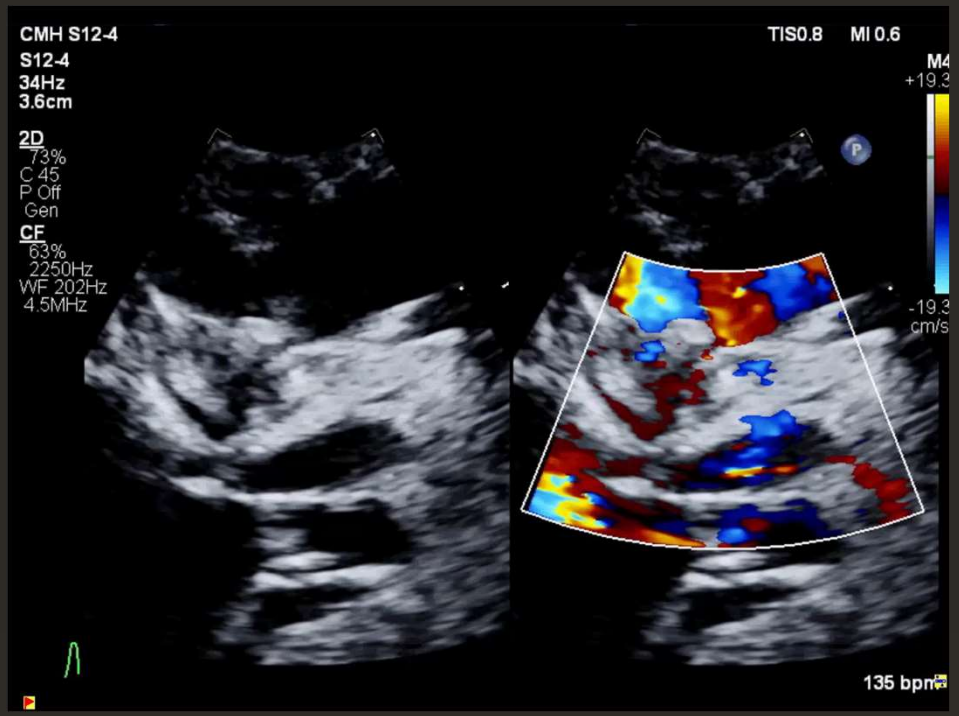



Children's Mercy

# Case 2 Postnatal Atrial Septum/ Coronaries




Children's Mercy

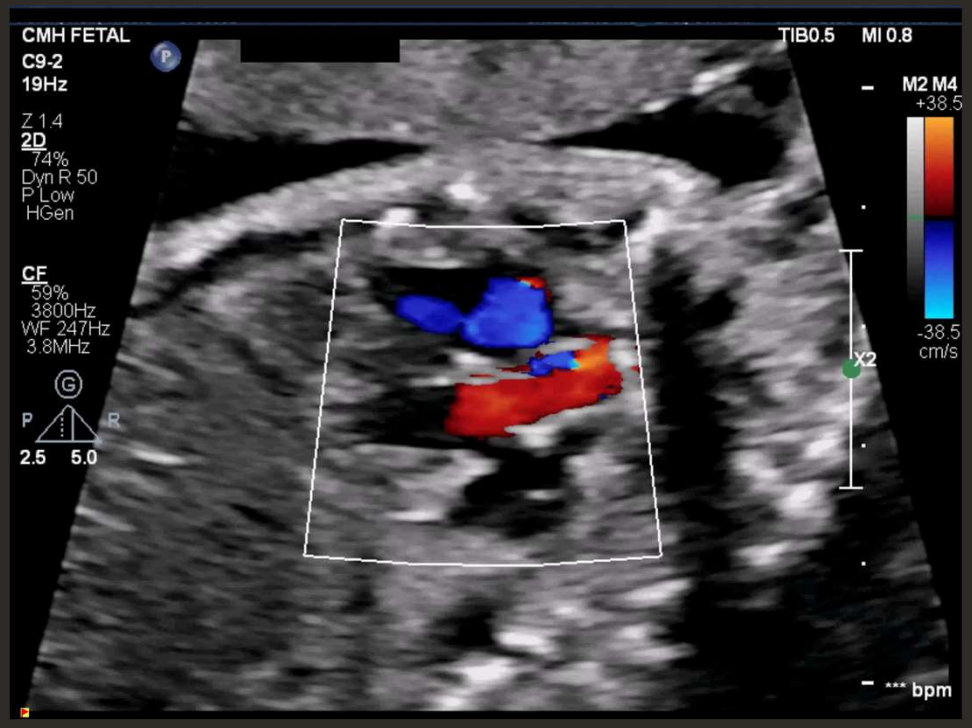
# Case 2 Postnatal outcome

- Bilateral PA bands at 1 week of life
- Atrial septostomy 3 weeks of life
- Heart block
- ECMO
- Decannulation from ECMO
- Continued critical state
- Redirection of care

# Case 3

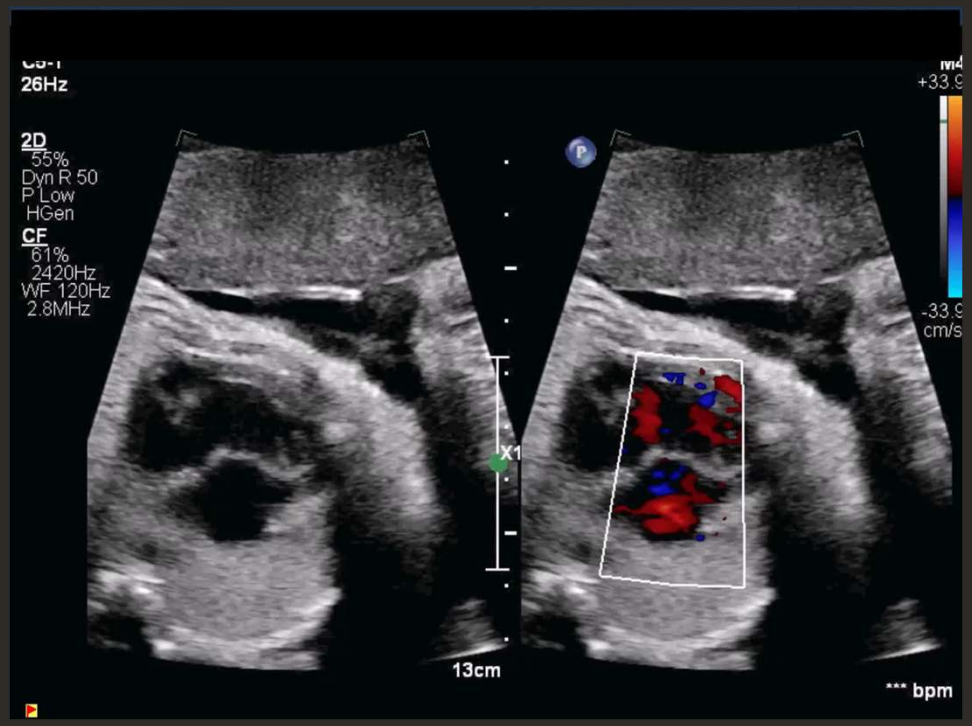
- G1P0 referral at 28 5/7 weeks
- Suspicion for HLHS
- No family history of CHD


# Case 3 Mitral Valve/Aortic valve

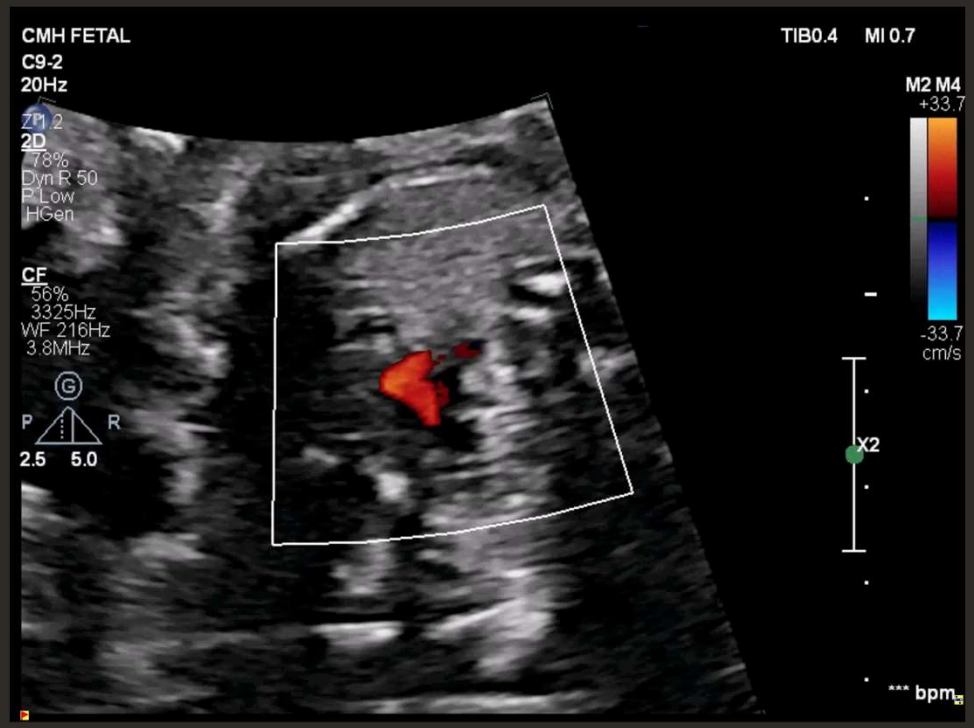

Children's Mercy

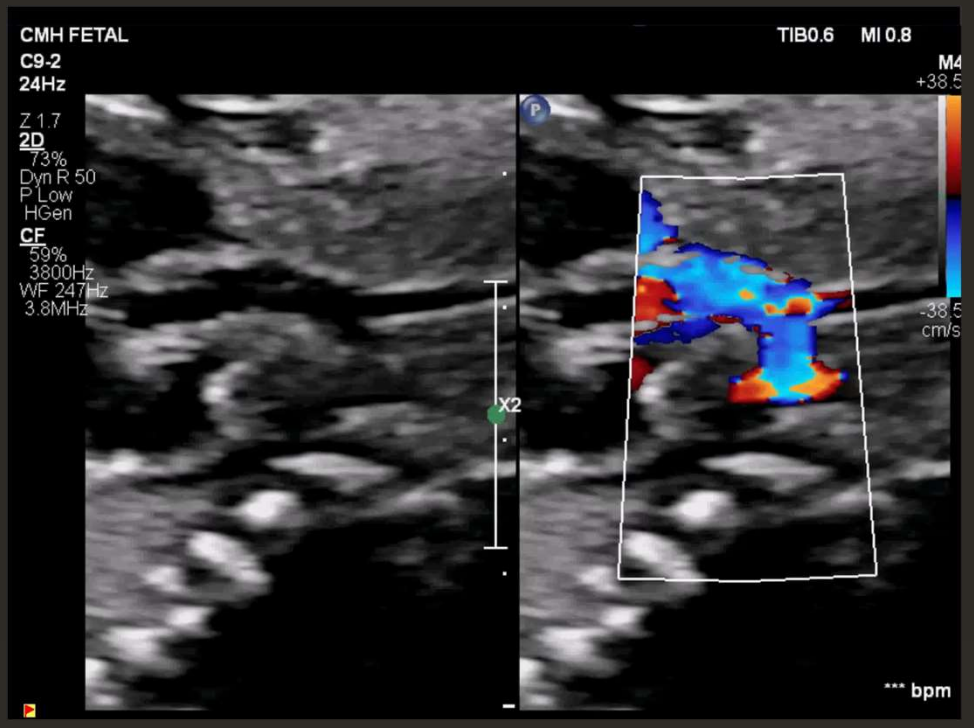
# Case 3 Atrial Septum/Pulm Vein


Children's Mercy

# Case 3 Arch




Children's Mercy

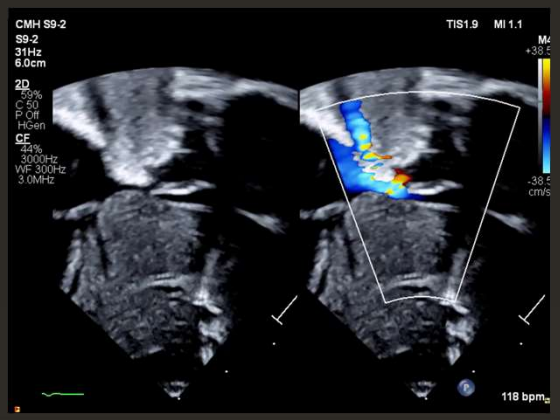
# Case 3 Branch PA/ Head



Children's Mercy

# Case 3 Postnatal

- Born at 37 weeks gestation
- Birth weight 3.1 Kg
- Echo confirmed diagnosis Hypoplastic left heart structures, severe MV hypoplasia (-5Z), mildly hypoplastic left ventricle, severe coarctation, large VSD, absent RPA, scimitar vein to IVC


Children's Mercy

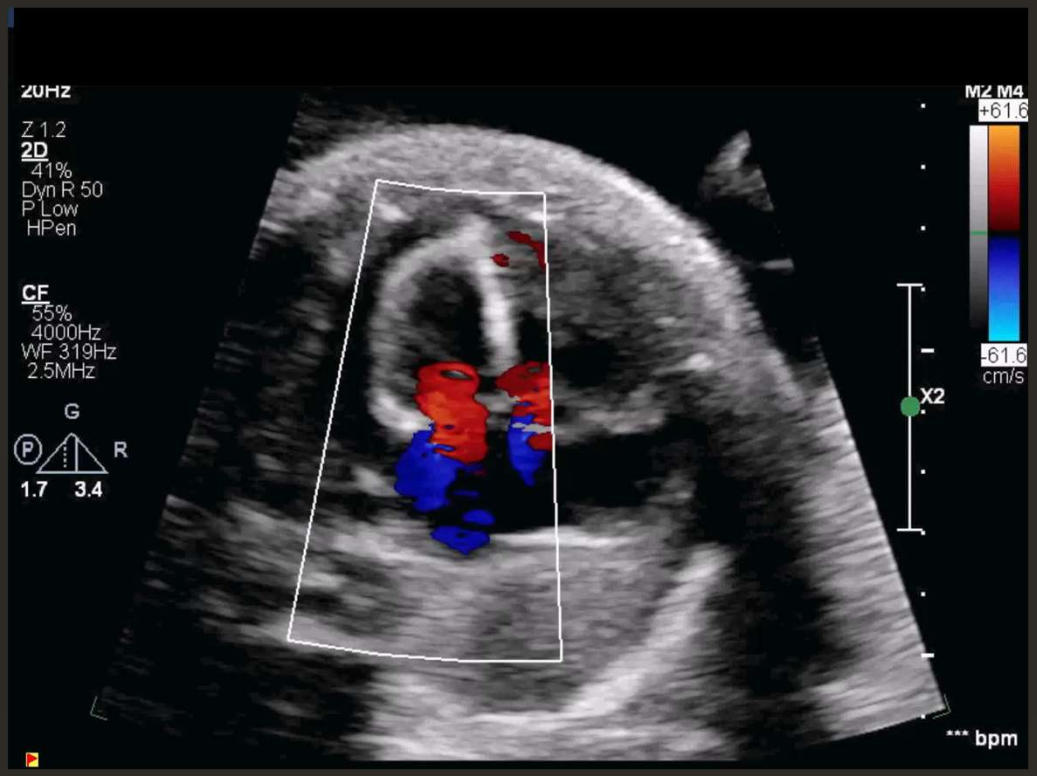
# Case 3 Postnatal outcome

- Multidisciplinary discussions
- Not a two-ventricle candidate
- Very poor single ventricle candidate
- Neurological concerns, need for VP shunt
- Redirection of care

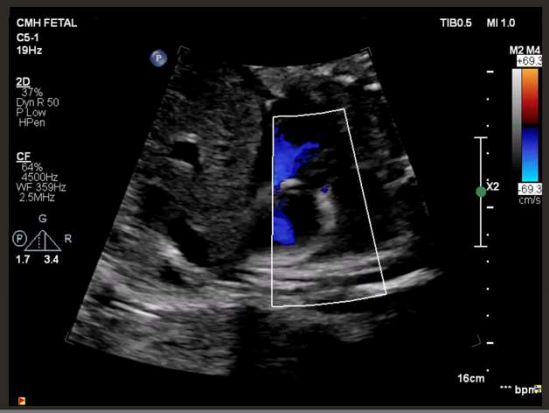
# Case 4

- G3 P2 referred at 33 6/7 weeks of gestation for ventricular fibroelastosis
- No family history of CHD

# Case 4 AV valves, aortic valve

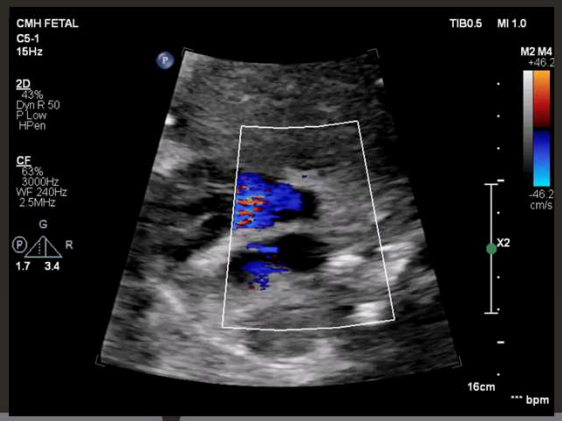


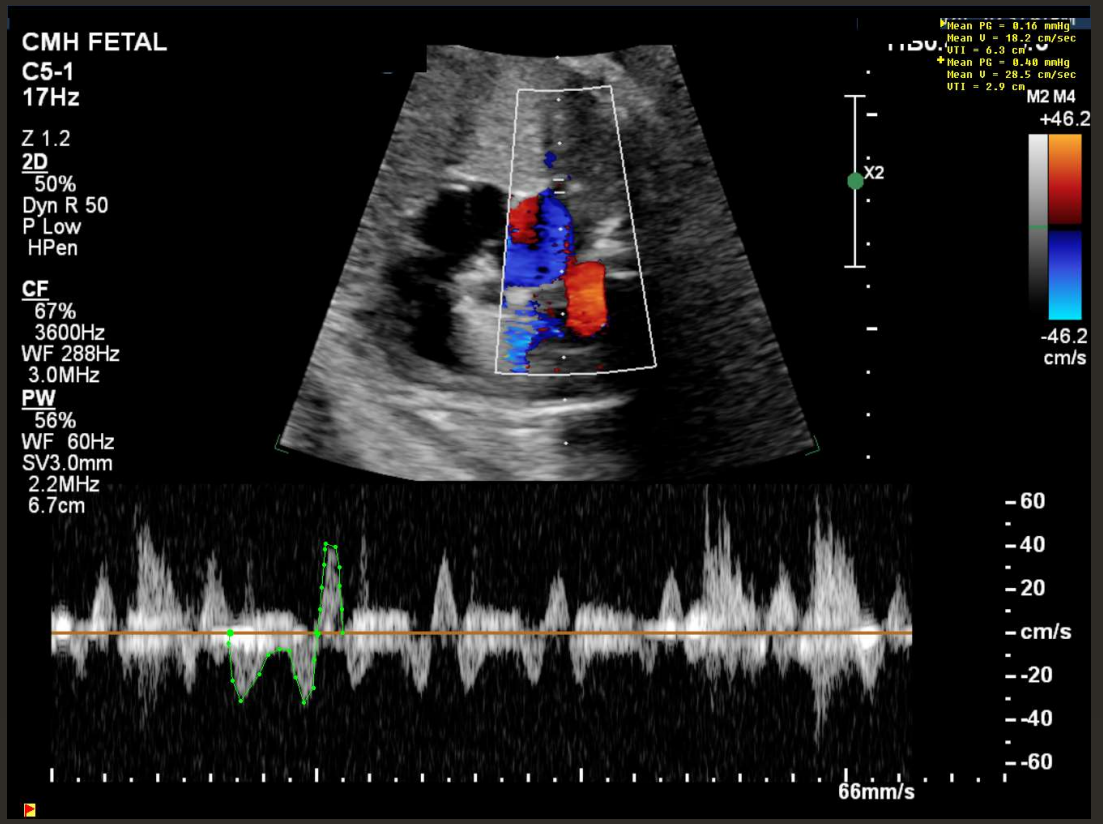





Children's Mercy

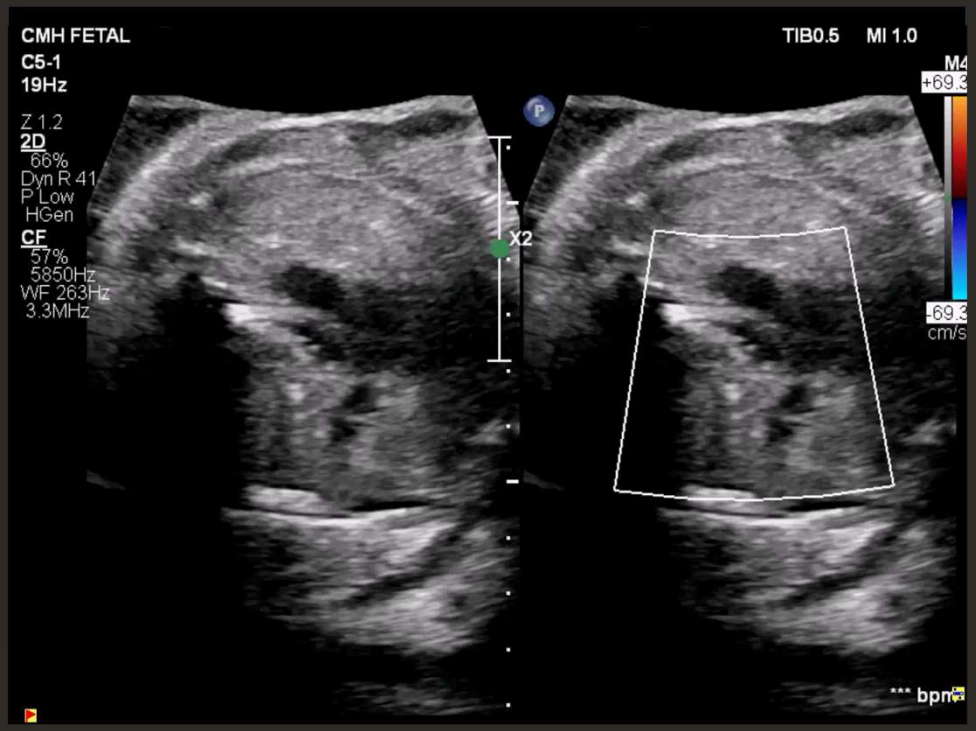
# Case 4 Atrial septum, pulmonary veins

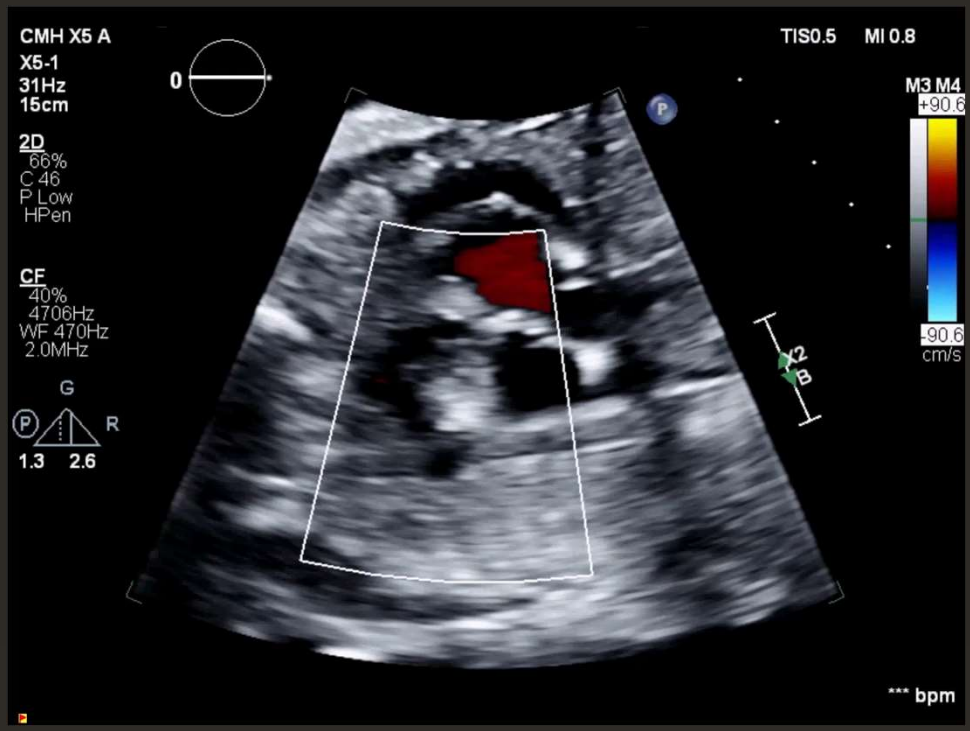




Children's Mercy

# Case 4 Arch 3VT/Sagittal

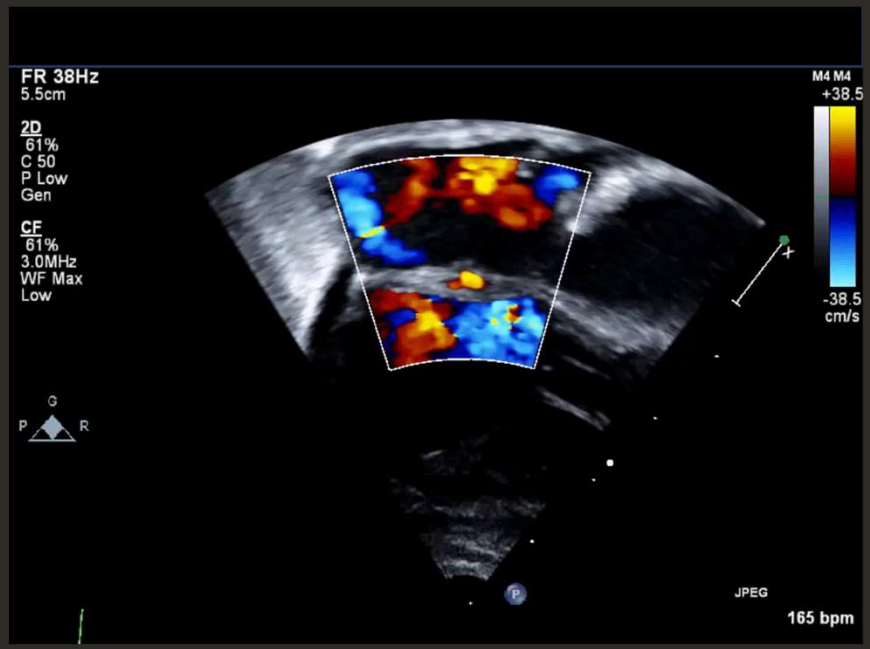



Children's Mercy

# Case 4 Postnatal

- Hydropic fetus (pericardial effusion, pleural effusion, ascites, scalp edema) along with macrosomia and polyhydramnios
- PROM at 36 4/7 weeks of gestation
- Delivery via C. Section in CVOR. Birth weight 4Kg
- Initial O2 saturations 50's. Intubated.
- Cath lab for urgent BAS



# Case 4 Postnatal outcome

- Mitral valve dysplasia, hypoplastic left ventricle with severe dysfunction with EFE, mildly hypoplastic aortic valve, moderate TR and RV dysfunction
- Bilateral PA bands and atrial septal stenting at 2 weeks
- Listed for primary heart transplant
- Heart transplant at 4 months of age

# Conclusions

- HLHS is defined by left side of the heart being inadequate to sustain systemic perfusion
- Characteristic abnormal 4 chamber view makes prenatal diagnosis quite feasible
- Important to assess details of HLHS types and variants
- Prenatal assessment of adequacy of atrial septum is critical
- Palliative surgical pathways have reasonable outcomes in standard risk patients
- Primary heart transplant in special circumstances

# Thank you!

Acknowledgements:

Melanie Kathol
Rita France
Alison Samrany
Ashley Warta
Bryan Beaven
Kelsey Pinnick
Maria Kiaffas