# Fetal Balloon Aortic Valvuloplasty

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# Disclosures

- None

# Objectives

- Review fetal case examples of critical aortic stenosis
- Define criteria for referral for fetal cardiac intervention
- Understand what fetal balloon aortic valvuloplasty entails
- Review outcomes of fetal balloon aortic valvuloplasty

# Case 1

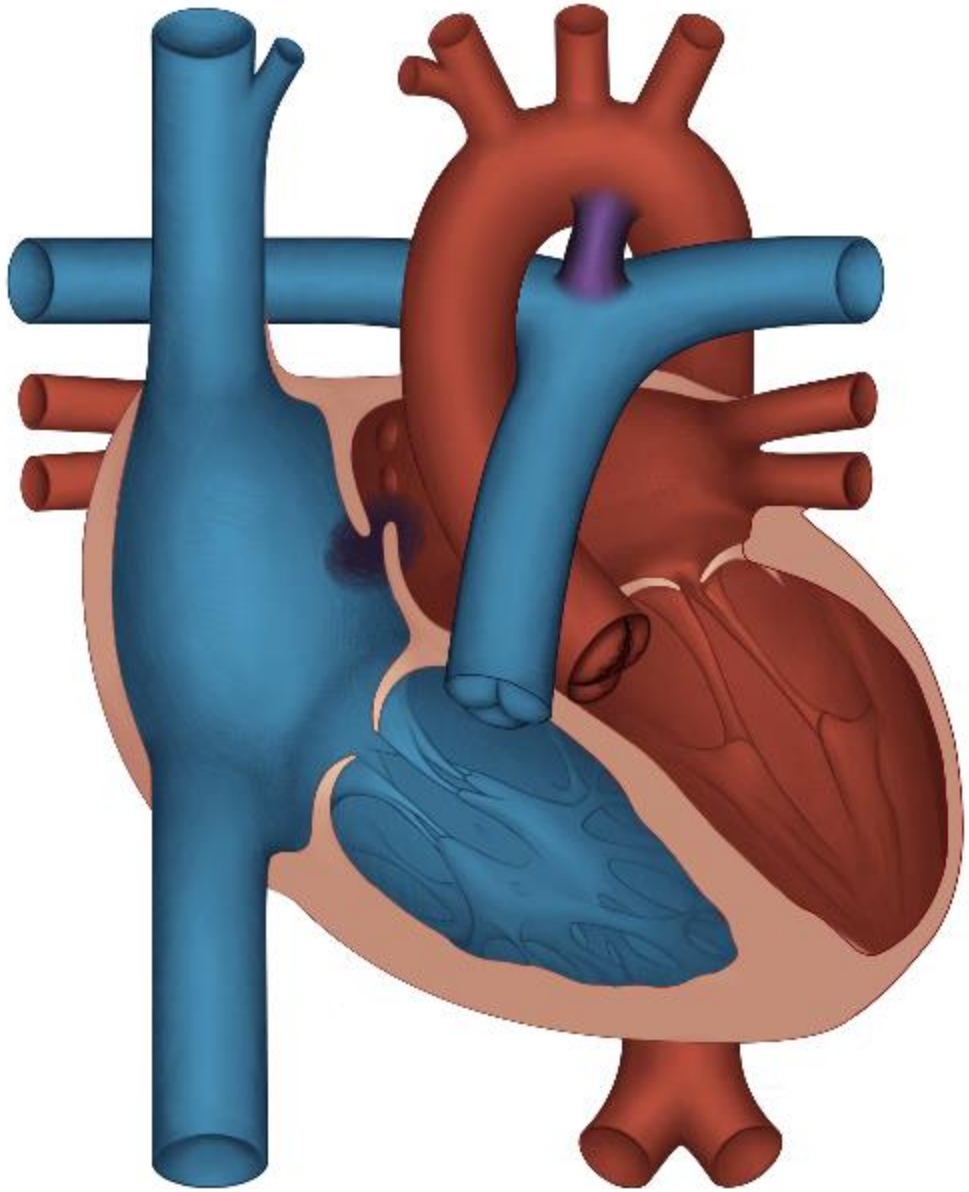
- 24-year-old G2P1 female
- Abnormal 20-week ultrasound with dilated left ventricle
- 23 weeks: referred to local MFM
- 26 weeks: referred to local pediatric cardiologist, concern for critical aortic stenosis
- 26 and 2/7 weeks: Children's Mercy fetal cardiology visit

# Fetal Echo 26 and 2/7 weeks

- Thickened, mildly hypoplastic aortic valve with narrow antegrade flow
- Mildly hypoplastic mitral valve with minimal antegrade flow and mild mitral regurgitation
- Severely dilated left ventricle with septal wall endocardial fibroelastosis (EFE) with severely decreased systolic function
- Mildly hypoplastic aortic arch with retrograde flow
- Small atrial level defect with left to right flow; not restrictive by pulmonary venous Doppler

# Critical Aortic Stenosis

- Severe stenosis of the aortic valve leading to poor LV function
- Left ventricle is initially enlarged, but in some cases, failure of further LV growth due to poor flow through LV
  - Can result in hypoplastic left heart syndrome (HLHS) with subsequent need for single ventricle palliation
- **Fetal balloon aortic valvuloplasty is proposed with the hope of biventricular repair strategy**



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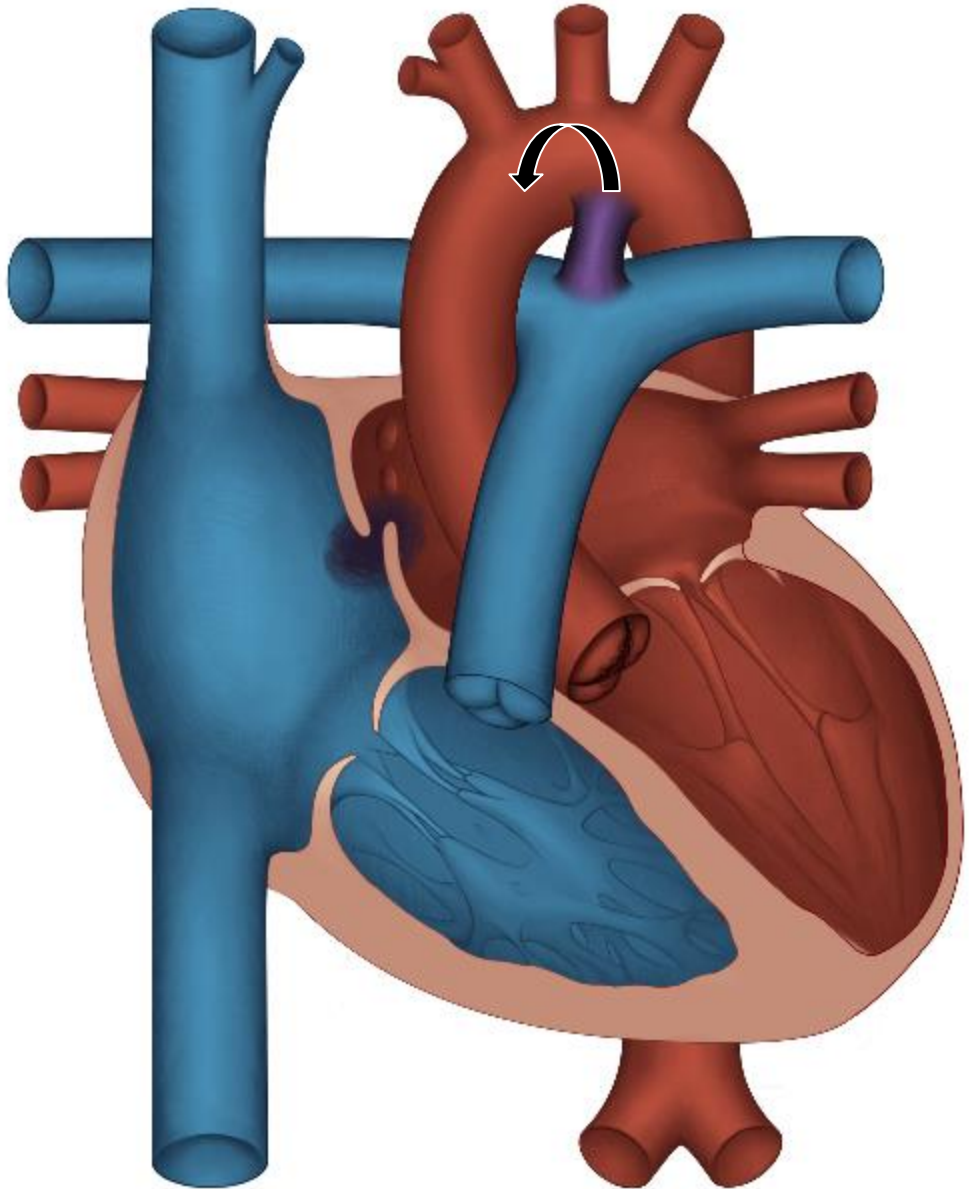
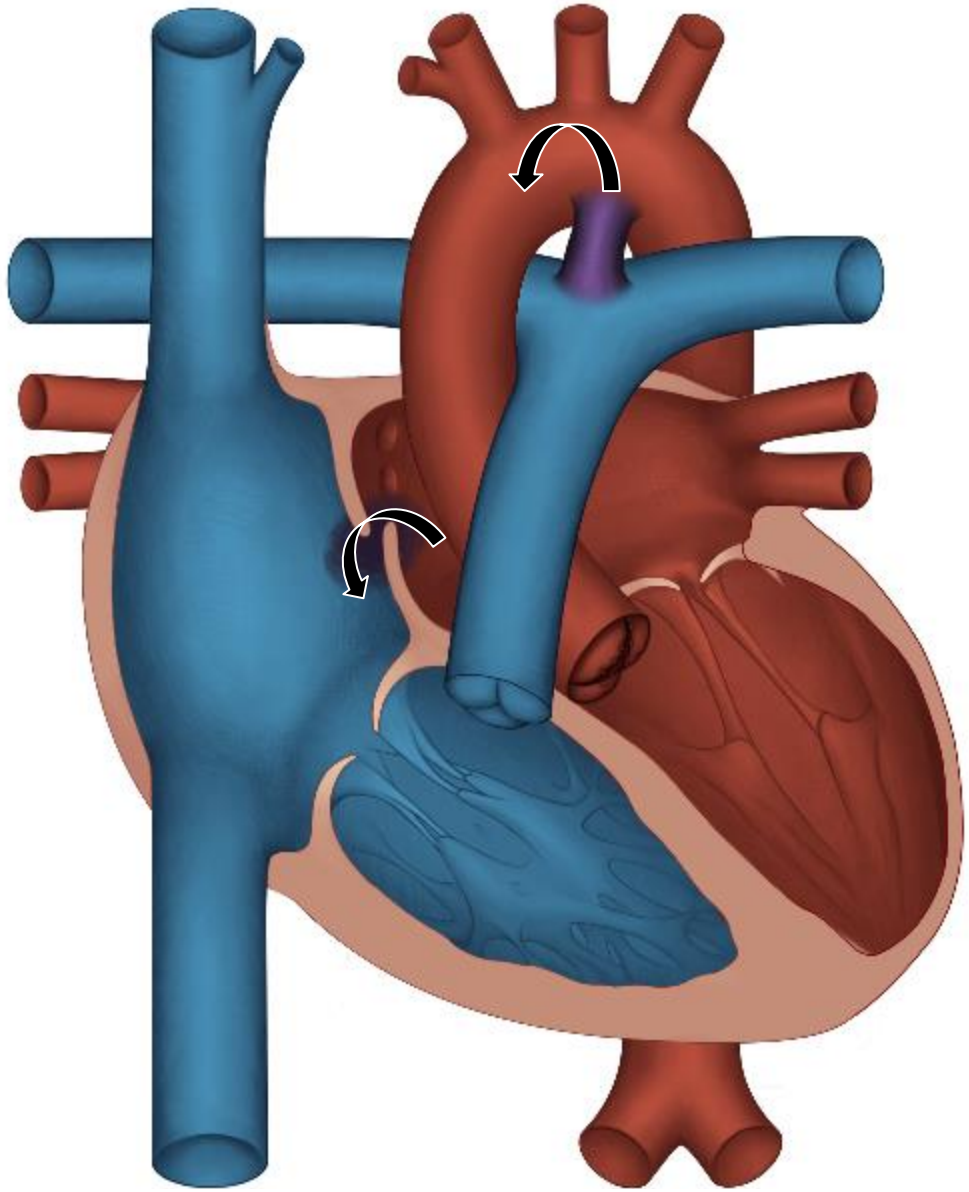
# Critical Aortic Stenosis

- Retrograde or bidirectional flow in the transverse aortic arch



# Critical Aortic Stenosis

- Retrograde or bidirectional flow in the transverse aortic arch
- Left to right flow across the atrial septum



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# Normal Fetal Echo



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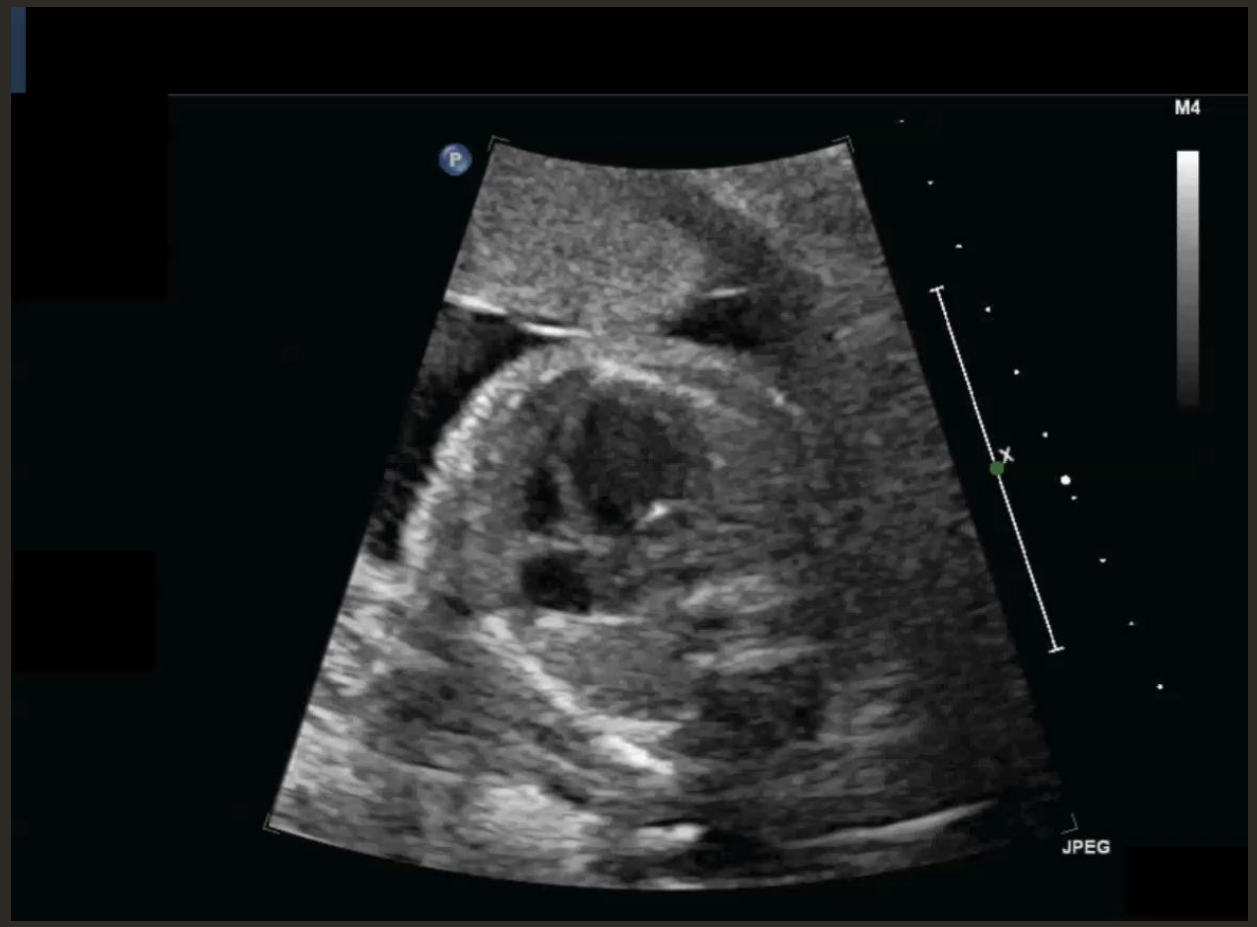
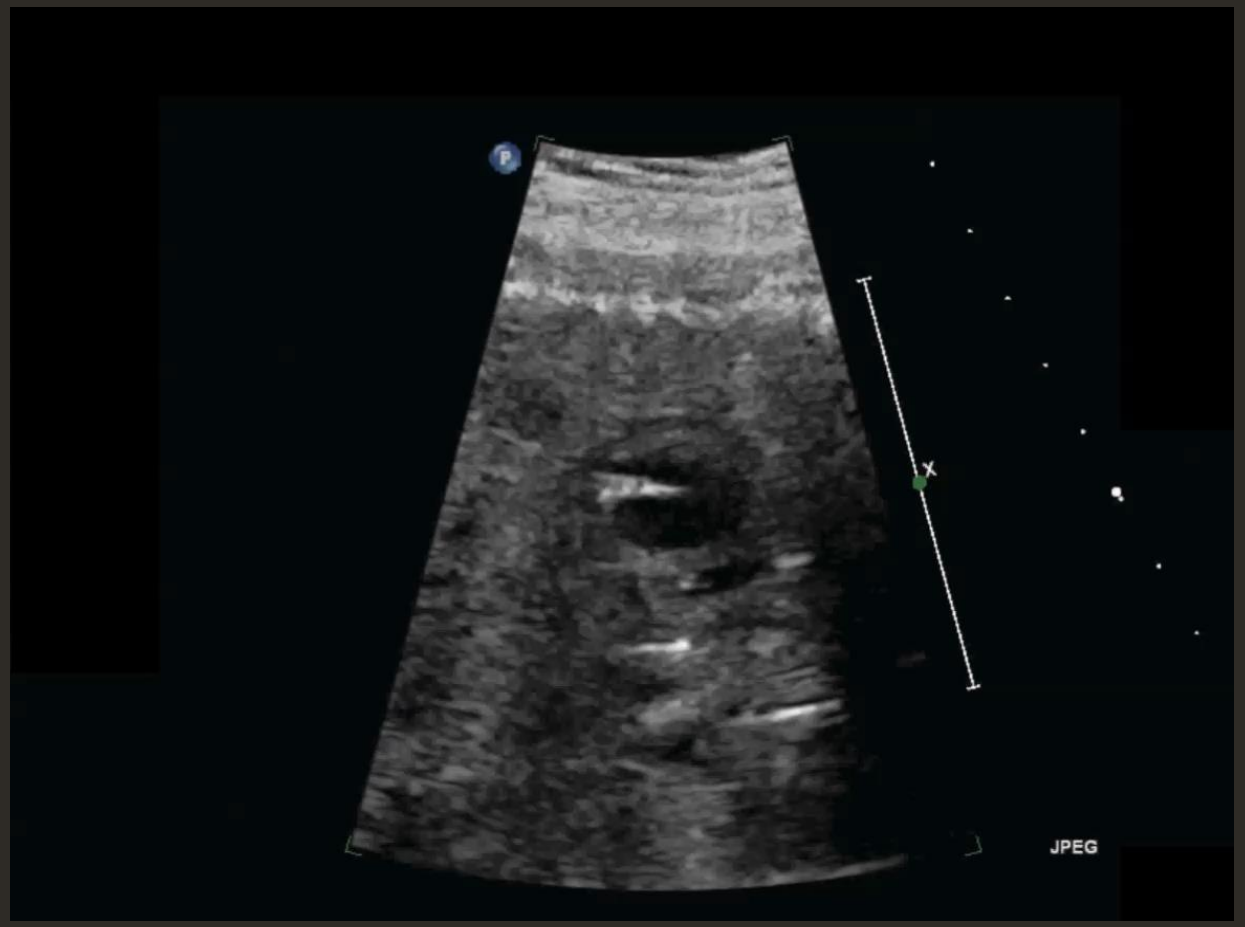
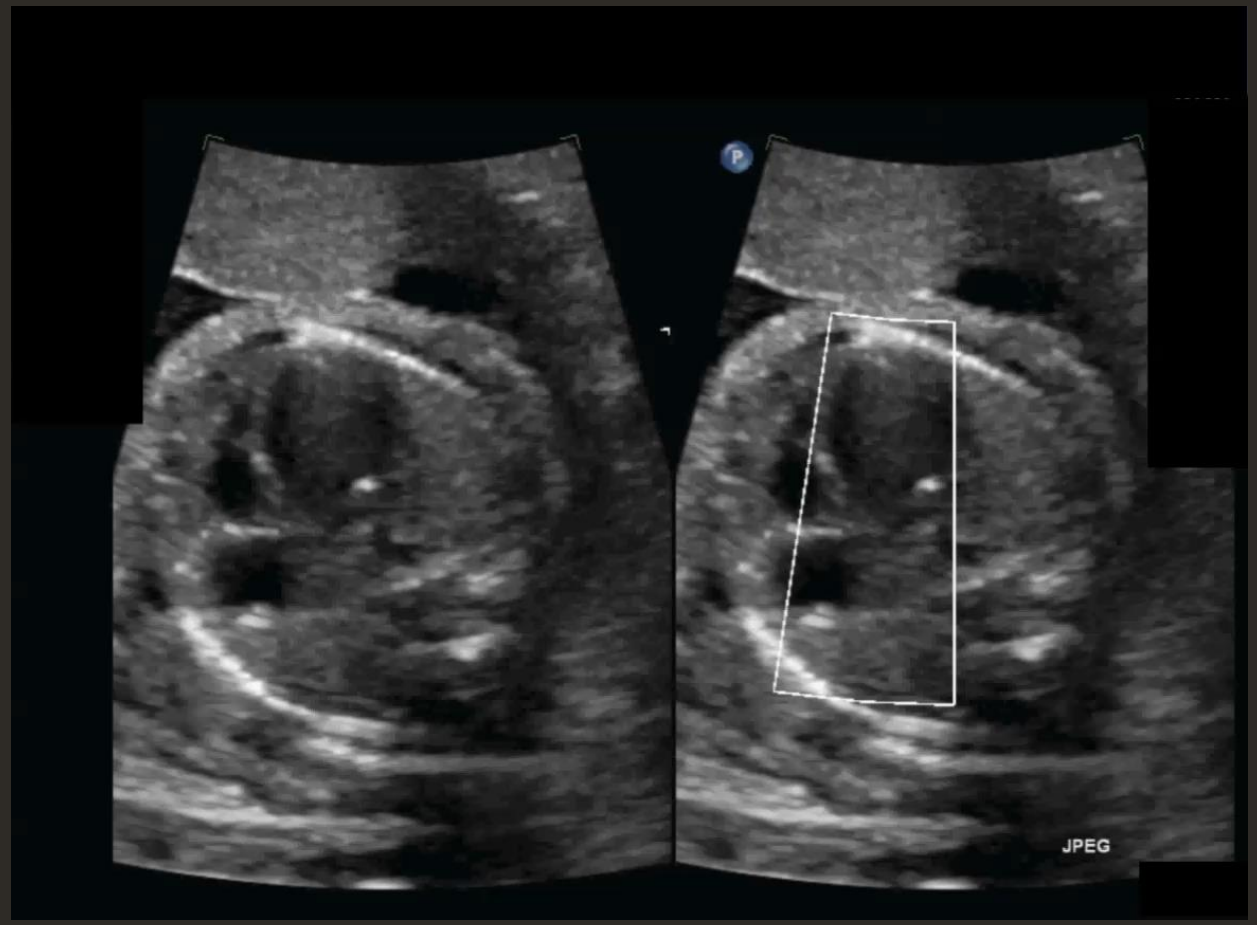
# Critical Aortic Stenosis Fetal Echo

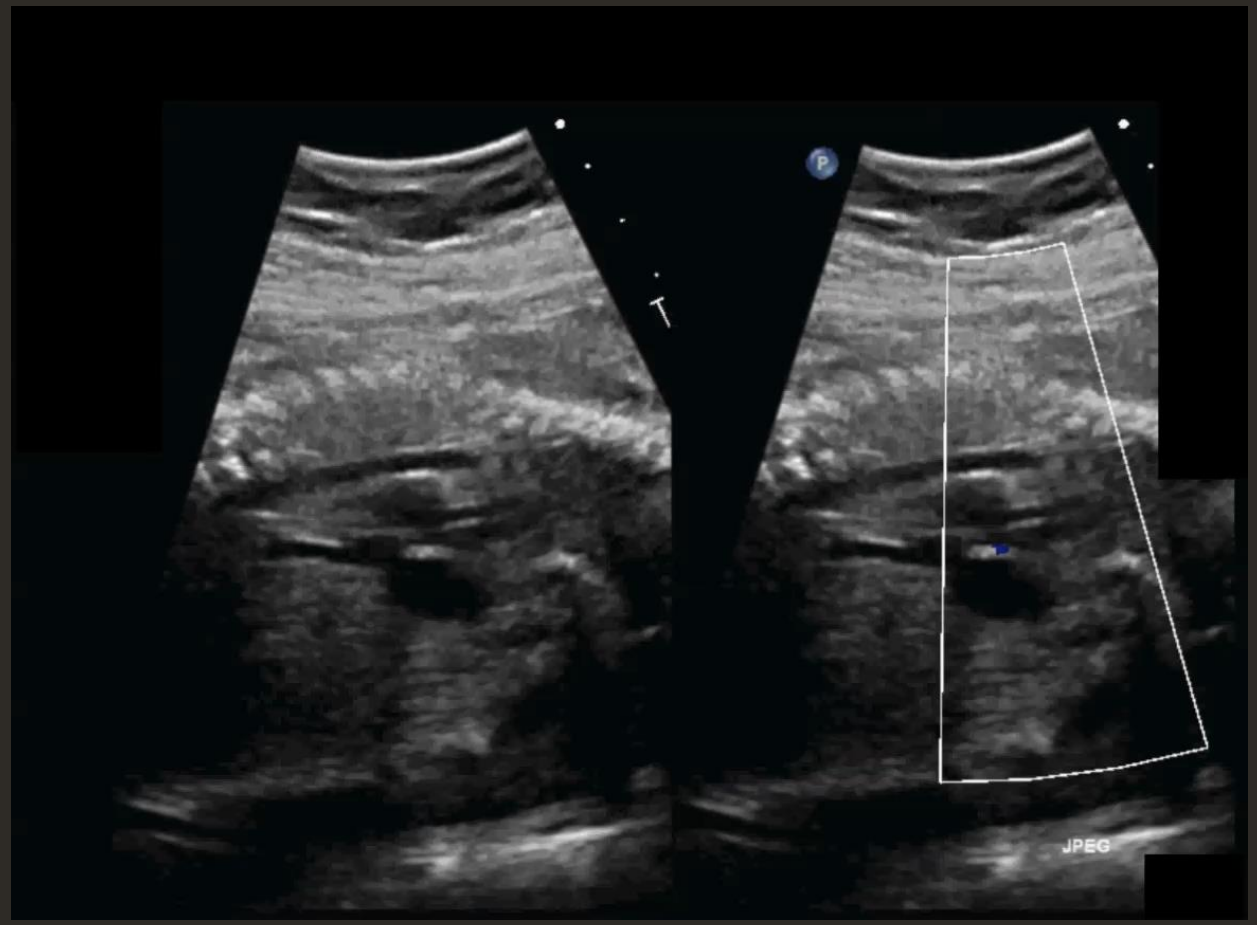


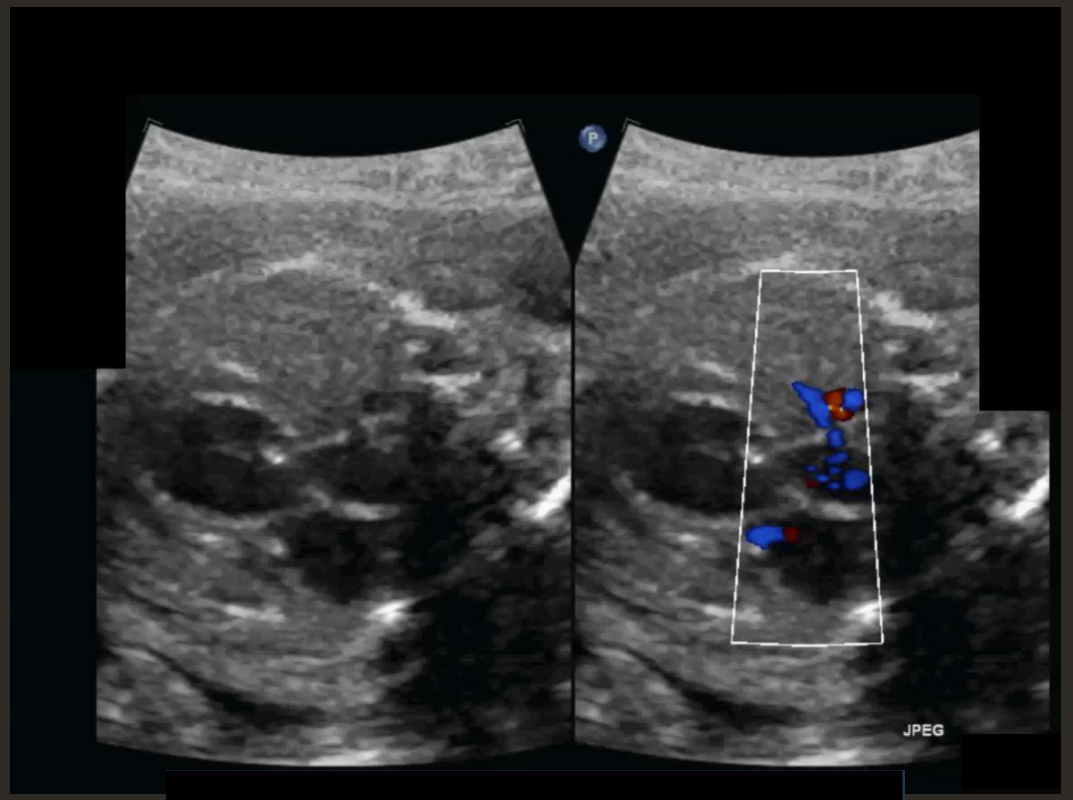



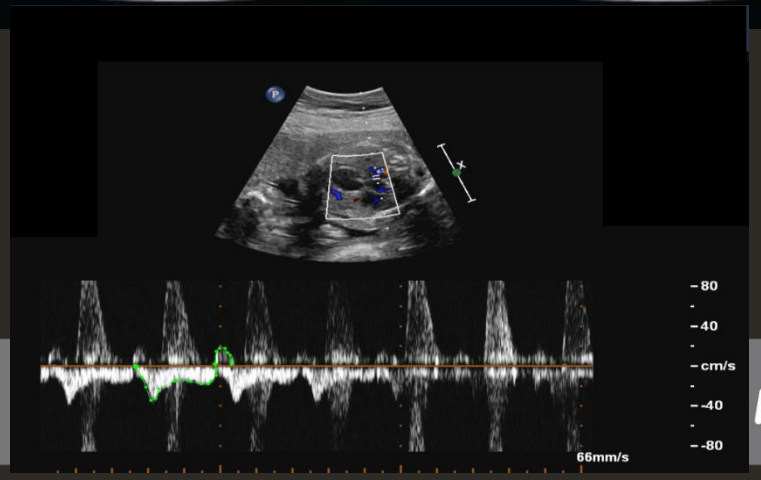
# Critical Aortic Stenosis Fetal Echo




# Critical Aortic Stenosis Fetal Echo

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# Criteria for Referral

| A | B | C | D |
|---|---|---|---|
| **1. Dominant cardiac anatomic anomaly is valvar AS with all of the following:** | **y/n?** | | |
| Decreased mobility of valve leaflets | | 0 | |
| Antegrade Doppler color flow jet across aortic valve smaller than the valve annulus diameter | | 0 | |
| No or minimal subvalvar LV outflow obstruction | | 0 | |
| | | **0** | |
| | | **Does not meet criteria** | |
| **2. Evolving HLHS** | **y/n?** | | LV function qualitatively depressed AND EITHER retrograde or bidirectional flow in the transverse aortic arch OR 2 of the following: Monophasic MV inflow, Left-to-right flow across atrial septum, or bidirectional flow in pulmonary veins |
| LV function qualitatively depressed | | 0 | |
| Retrograde or bidirectional flow in the transverse aortic arch (between the first 2 brachiocephalic vessels) at any time during the cardiac cycle | | 0 | |
| Monophasic MV inflow | | 0 | |
| Left-to-right flow across atrial septum or intact atrial septum (bulging left to right) | | 0 | |
| Bidirectional flow in pulmonary veins | | 0 | |
| | | **0** | |
| | | **Does not meet criteria** | |
| **3. Potential for a technically successful procedure and biventricular outcome postnatally** | **y/n? or Z score** | | |
| Unequivocal AS (vs aortic atresia) | | 0 | |
| LV long-axis *Z* score | | 4 | LV long-axis *Z* score >−2 |
| *Threshold score:* | | | *Threshold score ≥4 (≥4 of the following):* |
| LV long-axis *Z* score | | 0 | LV long-axis *Z* score >0 |
| LV short-axis *Z* score | | 0 | LV short-axis *Z* score >0 |
| Aortic annulus *Z* score | | 1 | Aortic annulus *Z* score >−3.5 |
| MV annulus *Z* score | | 1 | MV annulus *Z* score >−2 |
| MR or AS maximum systolic gradient | | 0 | MR or AS maximum systolic gradient ≥20 mm Hg |
| | | 6 | |
| | | **Does not meet criteria** | |
| **Fetus must meet criteria for all three questions to be a candidate for fetal intervention** | | | |

# Criteria for Referral: Valvar Aortic Stenosis

| A | B | C | D |
|---|---|---|---|
| **1. Dominant cardiac anatomic anomaly is valvar AS with all of the following:** | **y/n?** | | |
| Decreased mobility of valve leaflets | | 0 | |
| Antegrade Doppler color flow jet across aortic valve smaller than the valve annulus diameter | | 0 | |
| No or minimal subvalvar LV outflow obstruction | | 0 | |
| | | **0** | |
| | | **Does not meet criteria** | |

# Criteria for Referral: Evolving HLHS

| 2. Evolving HLHS | y/n? | | LV function qualitatively depressed AND EITHER retrograde or bidirectional flow in the transverse aortic arch OR 2 of the following: Monophasic MV inflow, Left-to-right flow across atrial septum, or bidirectional flow in pulmonary veins |
|---|---|---|---|
| LV function qualitatively depressed | | 0 | |
| Retrograde or bidirectional flow in the transverse aortic arch (between the first 2 brachiocephalic vessels) at any time during the cardiac cycle | | 0 | |
| Monophasic MV inflow | | 0 | |
| Left-to-right flow across atrial septum or intact atrial septum (bulging left to right) | | 0 | |
| Bidirectional flow in pulmonary veins | | 0 | |
| | | **0** | |

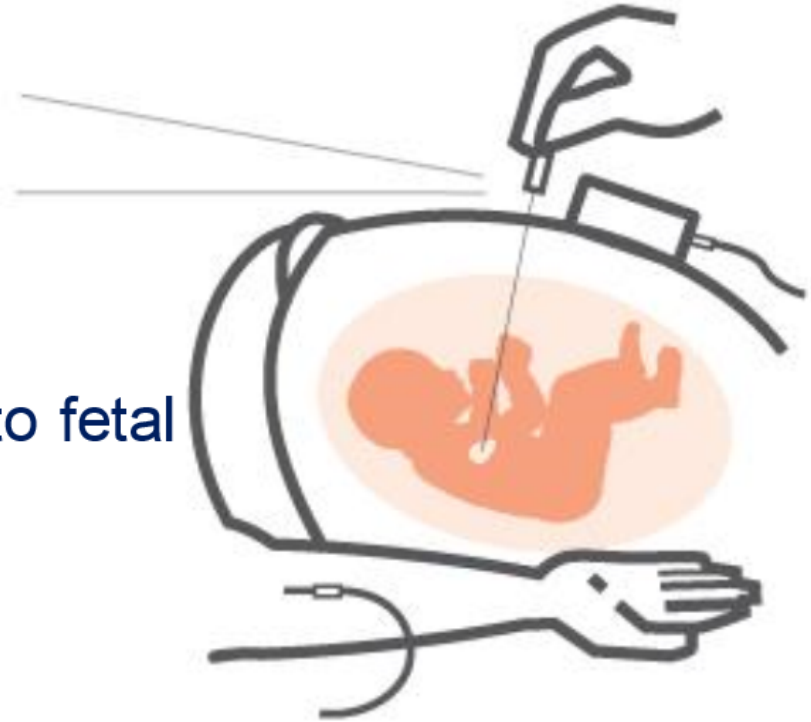
# Criteria for Referral: Potential for technically successful procedure and biventricular outcome postnatally

| 3. Potential for a technically successful procedure and biventricular outcome postnatally | y/n? or Z score | | |
|---|---|---|---|
| Unequivocal AS (vs aortic atresia) | | 0 | |
| LV long-axis Z score | | 4 | LV long-axis Z score >−2 |
| *Threshold score:* | | | *Threshold score ≥4 (≥4 of the following):* |
| LV long-axis Z score | | 0 | LV long-axis Z score >0 |
| LV short-axis Z score | | 0 | LV short-axis Z score >0 |
| Aortic annulus Z score | | 1 | Aortic annulus Z score >−3.5 |
| MV annulus Z score | | 1 | MV annulus Z score >−2 |
| MR or AS maximum systolic gradient | | 0 | MR or AS maximum systolic gradient ≥20 mm Hg |
| | | 6 | |

# Intervention: Fetal Aortic Valvuloplasty

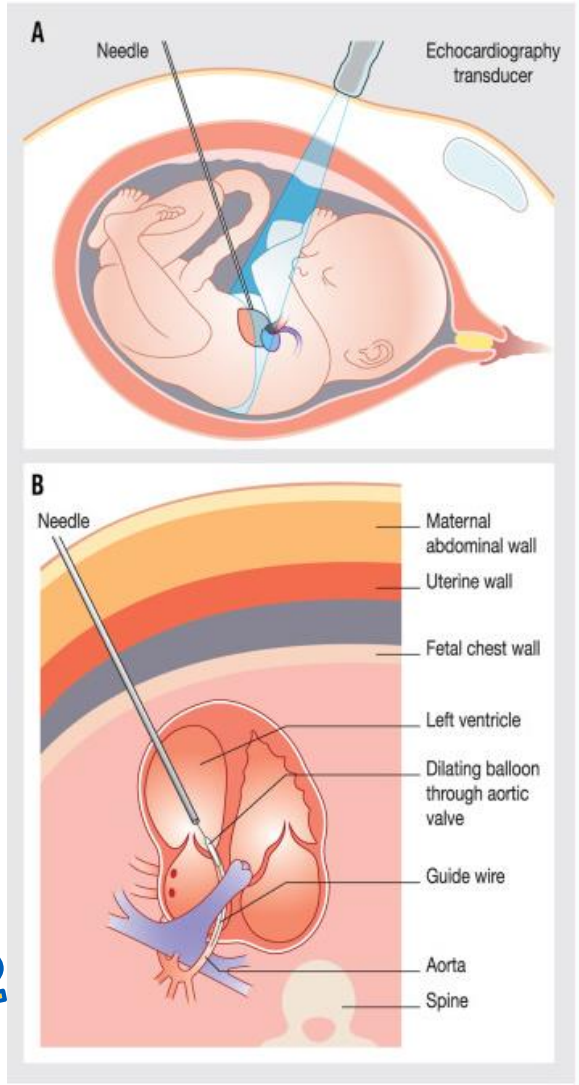
## Fetal Percutaneous Cardiac Intervention

- Percutaneous
- Ultrasound guided
- Epidural maternal anesthesia
- Fetal anesthesia via direct injection into fetal muscle
- Fetal position is critical




Gelehrter S. *Fetal Cardiac Intervention*. National Advisory Board 2018. University of Michigan Congenital Heart Center.

# Intervention: Fetal Balloon Aortic Valvuloplasty




REVIEW

## Fetal cardiac interventions: Where do we stand?

*La cardiologie fœtale interventionnelle : état des lieux*

**Kevin G. Friedman*, Wayne Tworetzky**

*Department of Cardiology, Boston Children's Hospital, 300 Longwood Avenue, Boston, MA 02115, USA*

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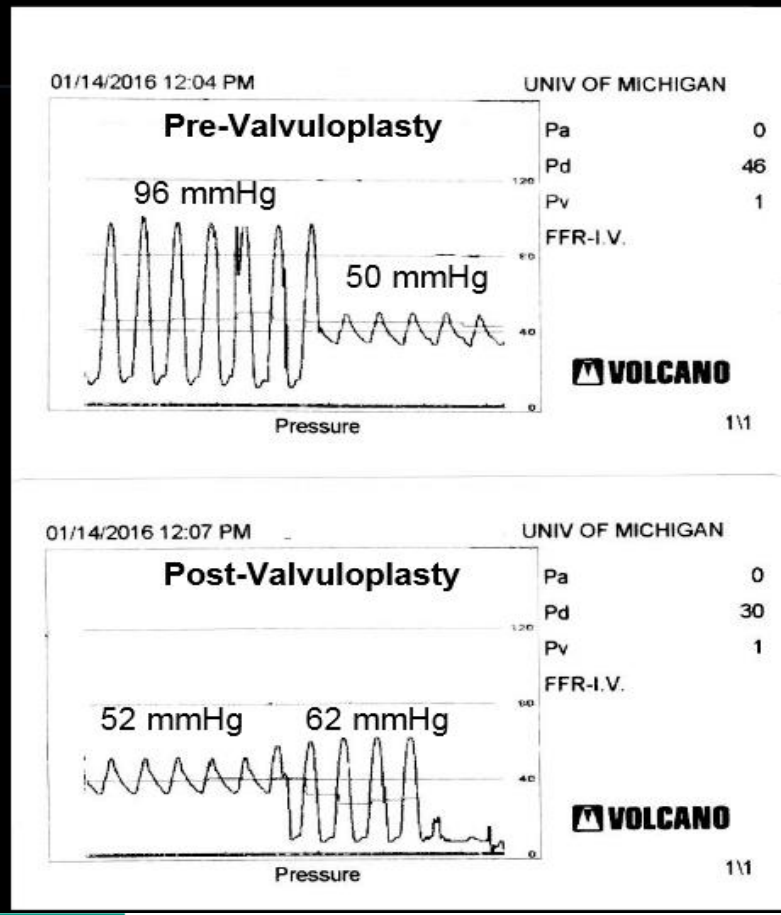
# Fetal Cardiac Intervention: Aortic Valvuloplasty

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Gelehrter S. *Fetal Cardiac Intervention*. National Advisory Board 2018. University of Michigan Congenital Heart Center.

# Case 1: Critical Aortic Stenosis

- Family traveled to Michigan at 29 and 1/7 weeks for a fetal intervention, but could not be attempted due to fetal position
  - Spine up for 6 hours 2 days in a row
- Postnatally
  - Hybrid → Heart Transplant

# Case 2: Critical Aortic Stenosis

- Referral for fetal cardiology consultation at 21 weeks: mild-moderate aortic stenosis which progressed at subsequent visits
- Fetal aortic valvuloplasty performed at Boston at 25 and 3/7 weeks
- Fetus monitored closely with post-procedural moderate-severe aortic insufficiency
- Postnatally, baby underwent neonatal Ross/Konno procedure with coarctation of the aorta repair → biventricular circulation

# Critical Aortic Stenosis Fetal Echo 21 weeks


- Peak aortic valve velocity 2.6 m/sec, gradient 25-29 mmHg
- Mildly hypoplastic aortic valve annulus (Z-score -2.2)
- Normal LV systolic function
- Seen again at 23 weeks with progression: peak gradient 67 mmHg

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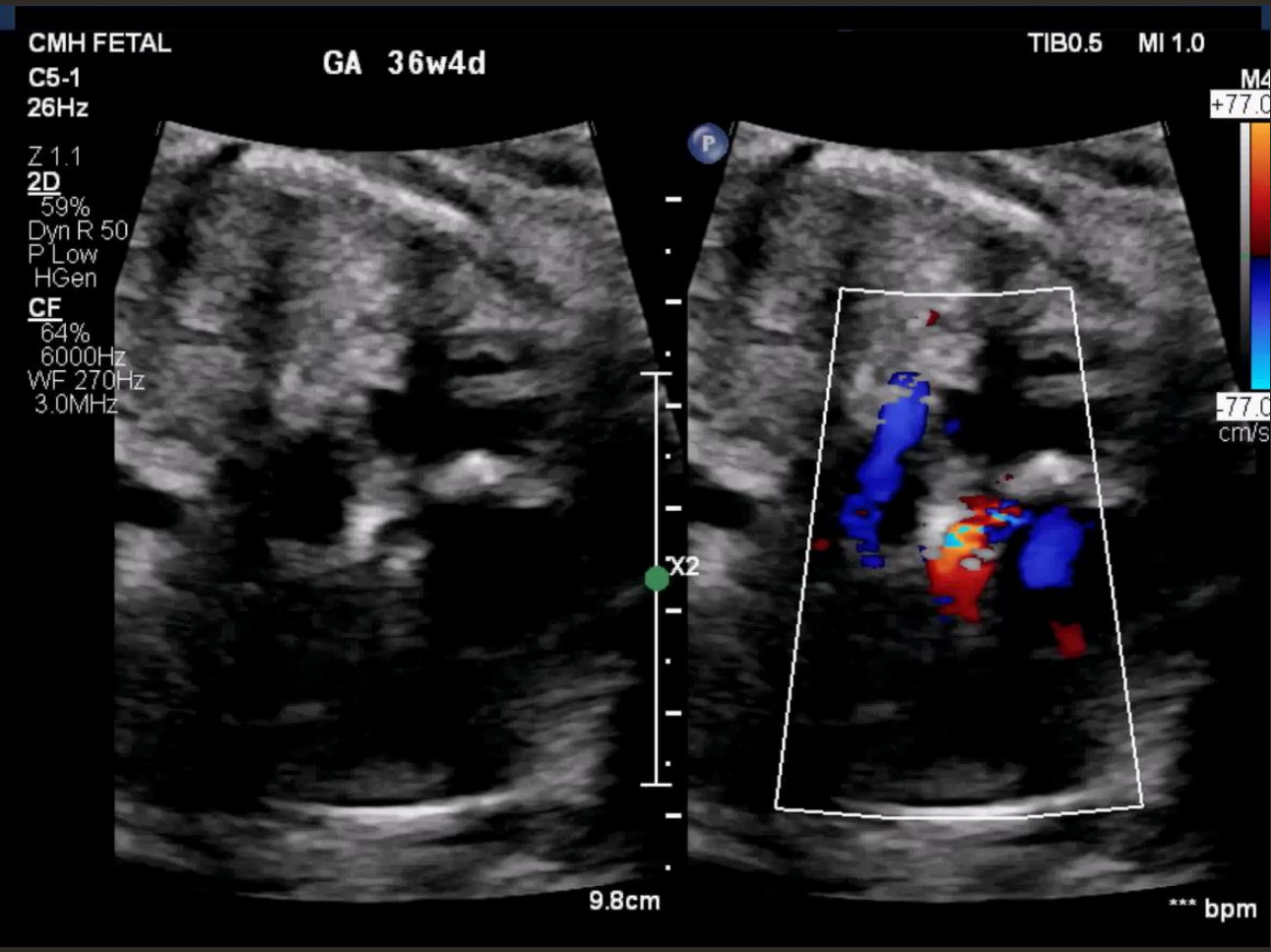
# Critical Aortic Stenosis Fetal Echo 24 weeks



- Peak velocity 3.6 m/sec, peak gradient 52 mmHg
- Worsened LV systolic function (moderately to severely reduced)
- PFO bidirectional flow
- Bidirectional flow in the transverse aortic arch

# Critical Aortic Stenosis Fetal Echo Post Balloon Aortic Valvuloplasty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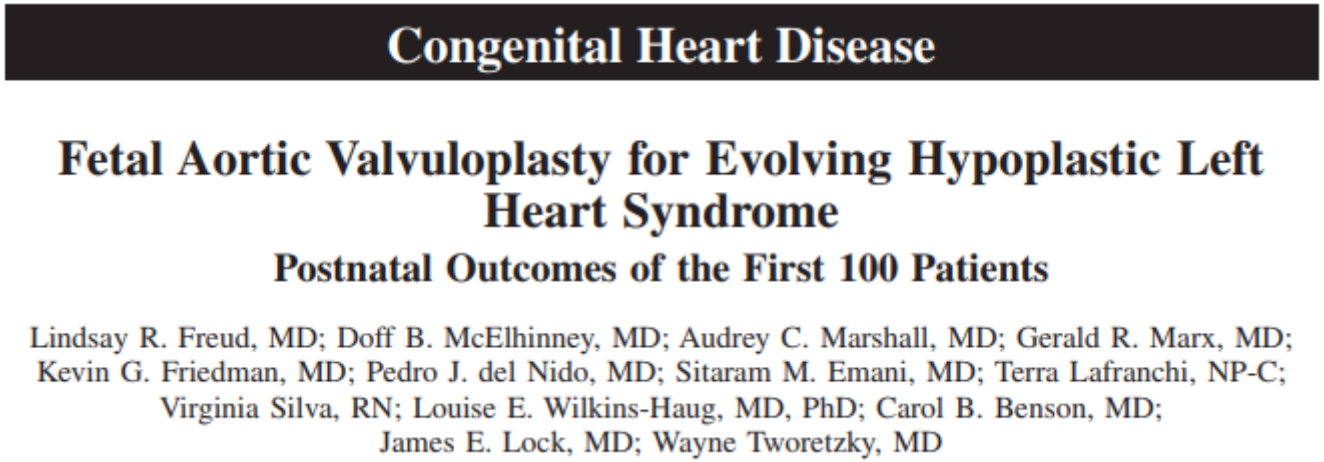
# Fetal Cardiac Interventions: Centers

- Boston Children's Hospital
- Children's Hospital of Philadelphia
- Texas Children's Hospital
- University of Michigan
- Benioff Children's Hospital, UCSF
- Wexner Medical Center/Nationwide Children's Hospital OH

# Outcomes: Critical Aortic Stenosis



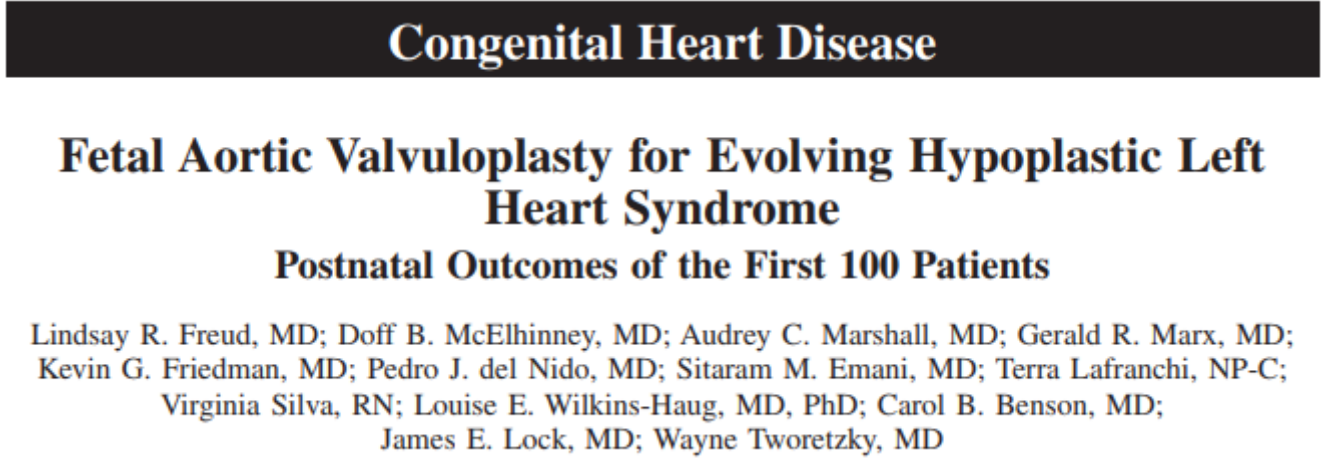
Congenital Heart Disease

**Fetal Aortic Valvuloplasty for Evolving Hypoplastic Left Heart Syndrome**

**Postnatal Outcomes of the First 100 Patients**

Lindsay R. Freud, MD; Doff B. McElhinney, MD; Audrey C. Marshall, MD; Gerald R. Marx, MD; Kevin G. Friedman, MD; Pedro J. del Nido, MD; Sitaram M. Emani, MD; Terra Lafranchi, NP-C; Virginia Silva, RN; Louise E. Wilkins-Haug, MD, PhD; Carol B. Benson, MD; James E. Lock, MD; Wayne Tworetzky, MD

Boston Experience from 2014



Congenital Heart Disease

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- 100 patients underwent fetal BAV at BCH 2000-2013
- 88 live-born fetuses
  - 11 fetal deaths (11%)
  - 38 (43%) → biventricular circulation
  - Left-sided structures significantly larger in BiV group
  - Postnatal survival, freedom from cardiac death worse for HLHS
- Clinical success: 43-45% with fetal cardiac intervention versus 17-19% without FCI in other studies

# Fetal Aortic Valvuloplasty for Evolving Hypoplastic Left Heart Syndrome

## Postnatal Outcomes of the First 100 Patients

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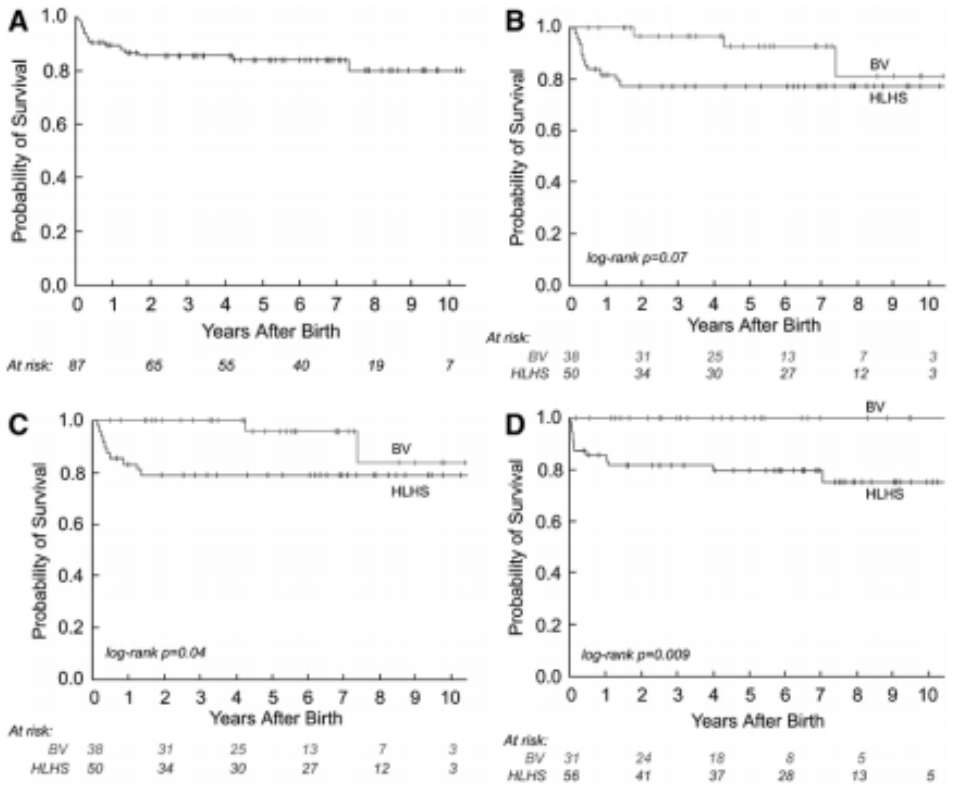


**Figure 4.** Kaplan–Meier curves depicting survival from the time of birth. **A**, All-cause mortality in the live-born cohort. **B**, All-cause mortality between hypoplastic left heart syndrome (HLHS) and biventricular (BV) outcome groups. **C**, Cardiac mortality between HLHS and BV outcome groups (excluding the 2 noncardiac deaths: 1 in the HLHS and 1 in the BV outcome group). **D**, Cardiac mortality between HLHS and BV outcome groups based on the initial postnatal management strategy.

- After median f/u of 5.4 yrs, freedom from cardiac death among BiV patients was 96+/-4% at 5 years and 84+/- 12% at 10 years
- No cardiac mortality in patients with BiV circulation from birth
- All but 1 of BiV patients required postnatal intervention
- 42% underwent aortic or mitral valve replacement

# Left Ventricular Diastolic Function and Characteristics in Fetal Aortic Stenosis

Kevin G. Friedman, MD[a,b,*], David Schidlow, MD[a,b], Lindsay Freud, MD[a,b], Maria Escobar-Diaz, MD[a,b], and Wayne Tworetzky, MD[a,b]

- Worse diastolic dysfunction associated with poor BiV outcome
- **Lower LV predicted pressure by AS or MR jet**
- MV inflow time shortened and fused E/A or monophasic inflow
- MV inflow/TV inflow time 60-80%
- **Greater EFE burden**

*Ultrasound Obstet Gynecol* 2018; **52**: 212–220
Published online 4 July 2018 in Wiley Online Library (wileyonlinelibrary.com). **DOI:** 10.1002/uog.17530

# Improved technical success, postnatal outcome and refined predictors of outcome for fetal aortic valvuloplasty

K. G. FRIEDMAN[1,2], L. A. SLEEPER[1,2], L. R. FREUD[1,2], A. C. MARSHALL[1,2], M. E. GODFREY[1,2], M. DROGOSZ[1], T. LAFRANCHI[1], C. B. BENSON[3,4], L. E. WILKINS-HAUG[3,4] and W. TWORETZKY[1,2]

[1]*Department of Cardiology, Boston Children's Hospital, Boston, MA, USA;* [2]*Department of Pediatrics, Harvard Medical School, Boston, MA, USA;* [3]*Department of Obstetrics and Gynecology, Brigham and Women's Hospital, Boston, MA, USA;* [4]*Department of Obstetrics and Gynecology, Harvard Medical School, Boston, MA, USA*

- BCH 2000-2015
- 123 fetal aortic valvuloplasties
- Independent predictors of favorable BiV outcome:
  - *Predicted LV systolic pressure >47 mmHg*
  - *Larger ascending aorta size (z-score ≥0.57)*
  - *Better LV diastolic function*
  - *Higher LV long axis z-score*
- → Strict criteria for referral

Circulation: Cardiovascular Quality and Outcomes

ORIGINAL ARTICLE

# Fetal Aortic Valvuloplasty for Evolving Hypoplastic Left Heart Syndrome

## A Decision Analysis

See Editorial by O'Byrne and Peyvandi

Sarah S. Pickard, MD, MPH

- 2020 paper from Boston group
- 143 fetuses who underwent fetal aortic valvuloplasty (FAV), 2000-2017 + secondary analysis from the PHN SVR trial
- Estimated probability of transplant-free survival from fetal diagnosis to age 6 years

Circulation: Cardiovascular Quality and Outcomes

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## A Decision Analysis

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Sarah S. Pickard, MD, MPH

- Technically successful FAV in 84%; Fetal demise 8%
- BiV circulation in 50% w/ successful FAV; 16% w/ unsuccessful FAV
- Transplant-free survival to age 6 years 75% vs 72% w/ expectant fetal management
- Analysis limited to improved FAV experience since 2009, increased probability of survival to 82% (modest medium-term survival benefit)

# Conclusions

- **Early diagnosis and referral of fetal aortic stenosis matters**
- Referral criteria for fetal aortic valvuloplasty are key to capture all potential candidates
- Fetal interventional procedures are complex and require multifaceted family counseling
- Outcomes are encouraging but multiple postnatal surgeries are still likely

# Our Fetal Cardiology Team

Maria Kiaffas, MD

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Geetha Haligheri, MD

Nitin Madan, MD

Amanda McIntosh, MD

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Jenna Schermerhorn, MD

Laura Schoeneberg, MD

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# Our Fetal Cardiology Team

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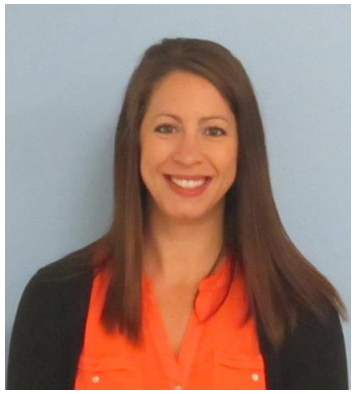
Ashley Warta, RDCS

Andrea Fargo, RDCS

Laura Kuzava, RDCS

Carol Moser-Dungan, RDCS



Mindy Parry, RDCS

Christen Schulz, RDCS

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# Thank you!

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