Fetal Cardiac Interventions: Where Are We Today and What Can We Expect in the Future?

Cardiology 2025
Children's Hospital of Philadelphia Annual Meeting
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Introduction

- Disclosures – None
- The following has been teamwork at BCH and BWH
- History of FCI at BCH
- Current FCI
- New Innovations

Introduction

- All you ever wanted to know about Fetal Cardiac Intervention
- …but were too afraid to ask

- Aortic stenosis with evolving HLHS

- HLHS with intact/restrictive atrial septum

- Pulmonary atresia with intact ventricular septum (HRHS)

- To improve postnatal, long-term outcomes

- To improve survival

Timeline Fetal Cardiac Intervention
Boston Children's and Brigham and Women's Hospitals

- First attempted (unsuccessful) FCI in March 2000
  - Patient has HLHS, s/p Fontan, graduated HS, has Fontan related chronic cardiac issues.

- First successful FCI Sept 13th 2001
  - Baby Jack has a 2V circulation and has graduated college

- We have performed n~ 260 FCI
- Peak 24/year (prior to other institutions performing FCI)
- Currently n~ 10-12/year
- Fetal AS procedures n~ 202

Introduction

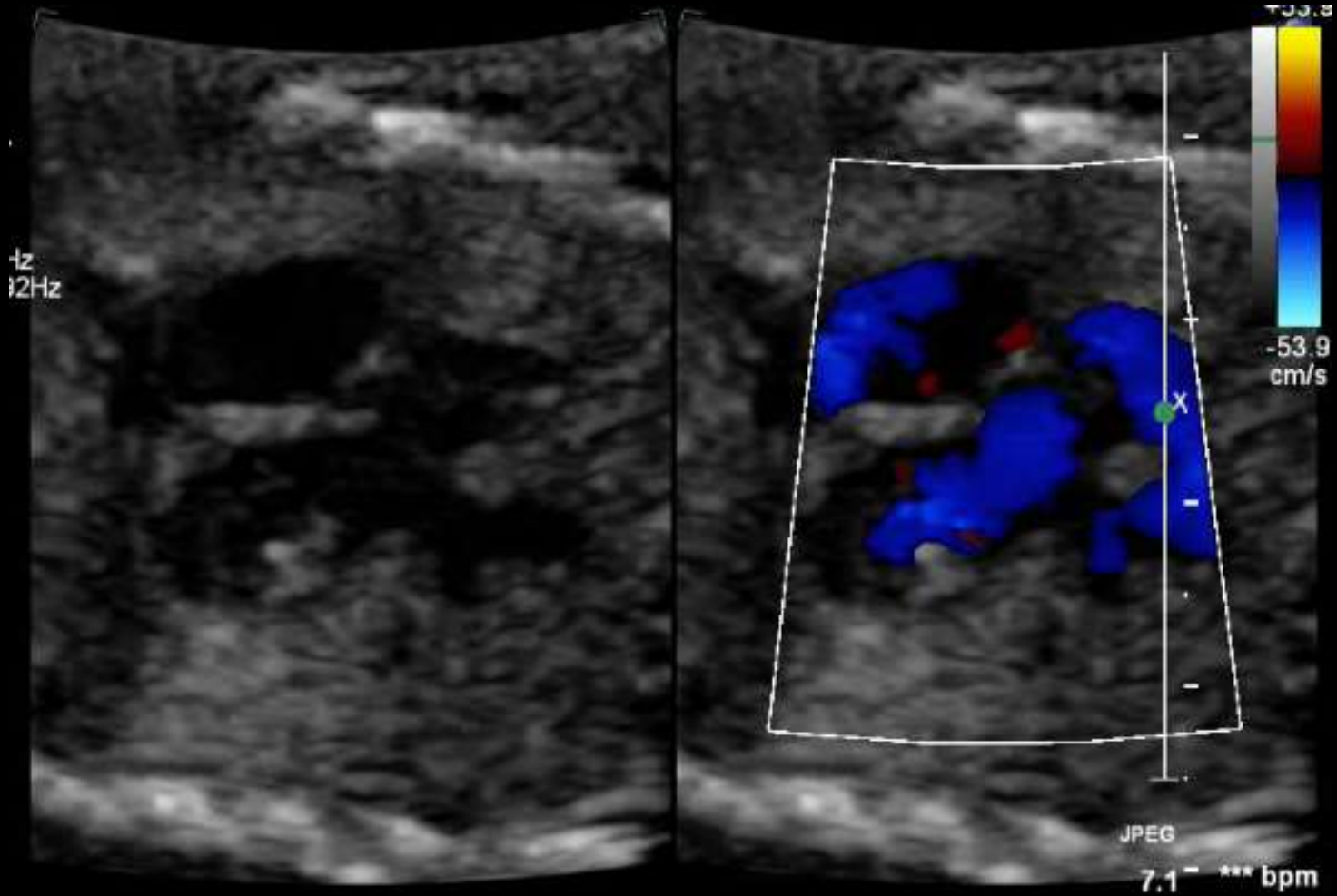
- Aortic stenosis with evolving HLHS
- Why?
- Prevent progression to HLHS
- Salvage LV myocardium. Improve long-term systolic and diastolic function
- Gaps in our knowledge
  - Perfect patient selection (too early vs too late)
  - Optimal postanal management (center variation in experience and management)
  - Long term outcomes (fate of aortic and mitral valves and myocardium)

Fetal Aortic Stenosis progresses to HLHS

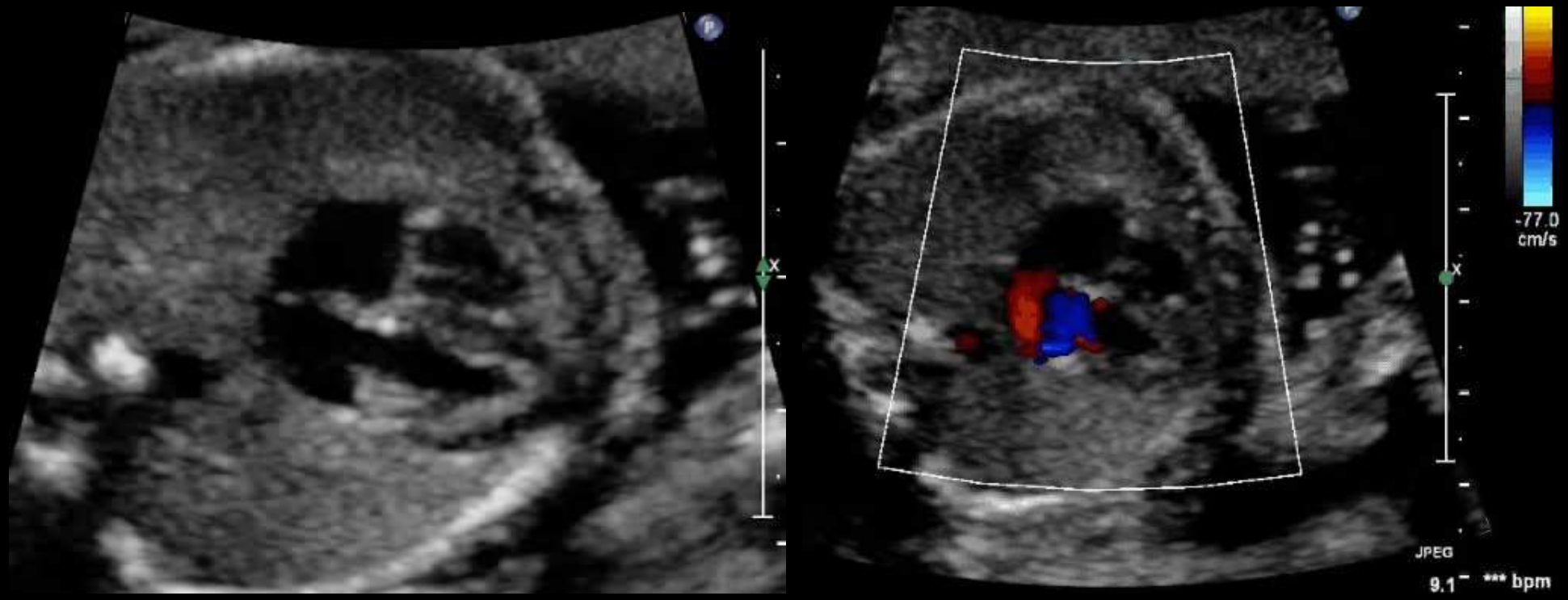
23w 36w

Poor candidate for FCI

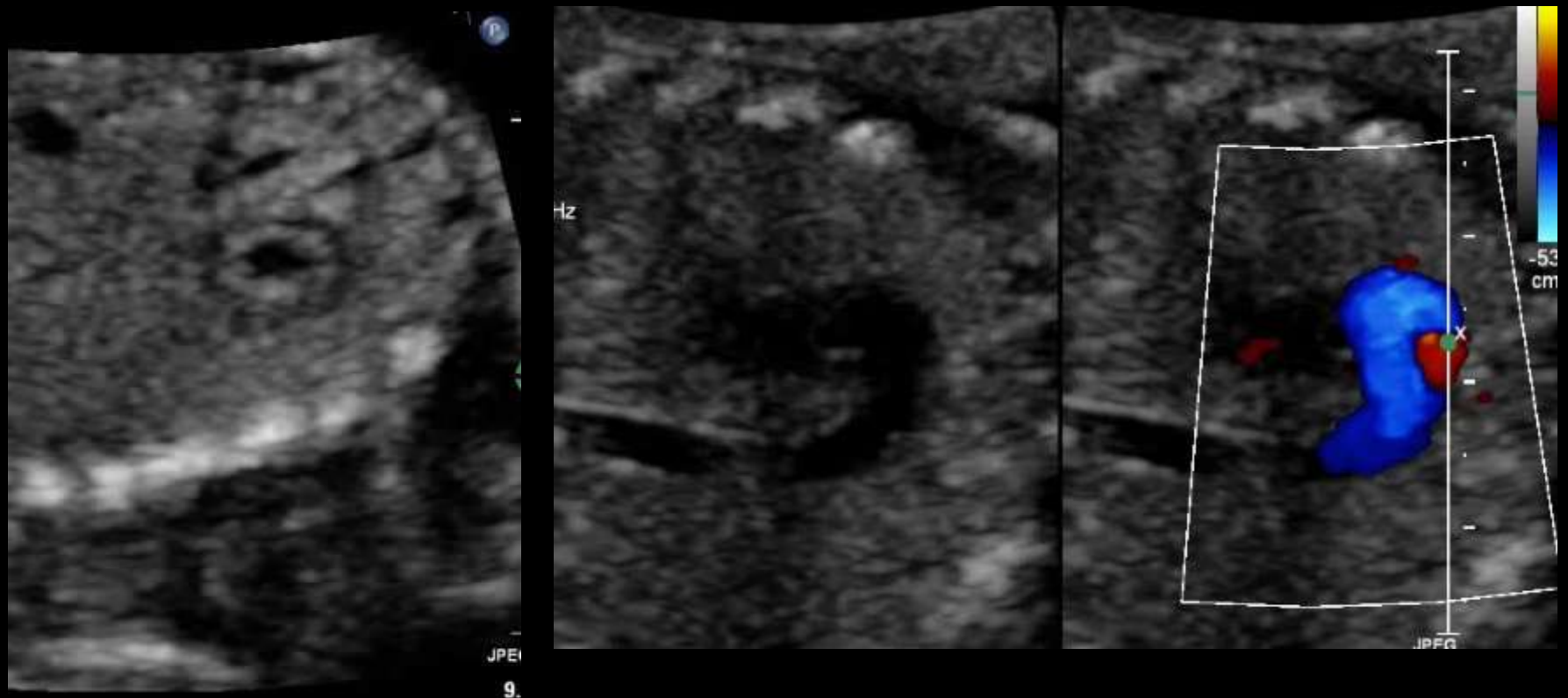
Good candidate for FCI
Fetal echo at 22 weeks GA
Starts with “Mild” aortic stenosis



Fetal echo at 22 weeks GA
”Mild aortic stenosis”
Good LV function - mild MR



Fetal echo at 22 weeks GA
Good LV function – anterograde arch flow



Follow-up Fetal Echo at 25 weeks GA

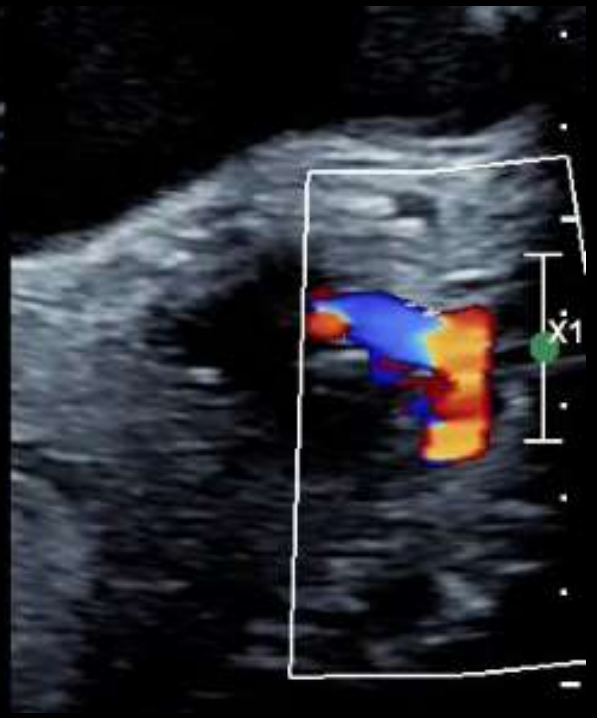
Severe LV dilation and dysfunction

25 weeks

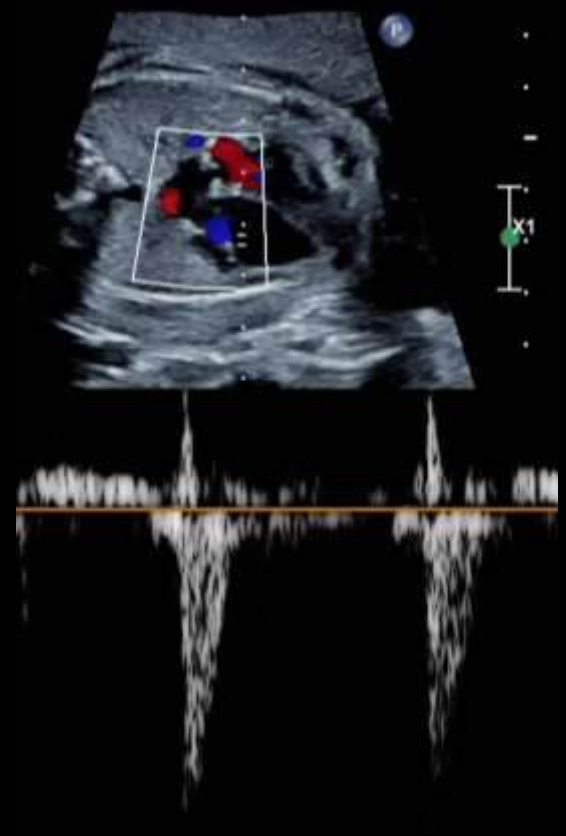
22 weeks

Follow-up Fetal Echo at 25 weeks - Aortic Stenosis

Aortic arch retrograde flow



MV monophasic inflow



Percutaneous Fetal Intervention for Aortic Stenosis
Cannula (19g) access to left ventricle

Fetal Intervention for Aortic Stenosis
Wire across aortic valve and balloon inflation

Fetal Echo 26 weeks GA
4 ch view Post FCI for AS

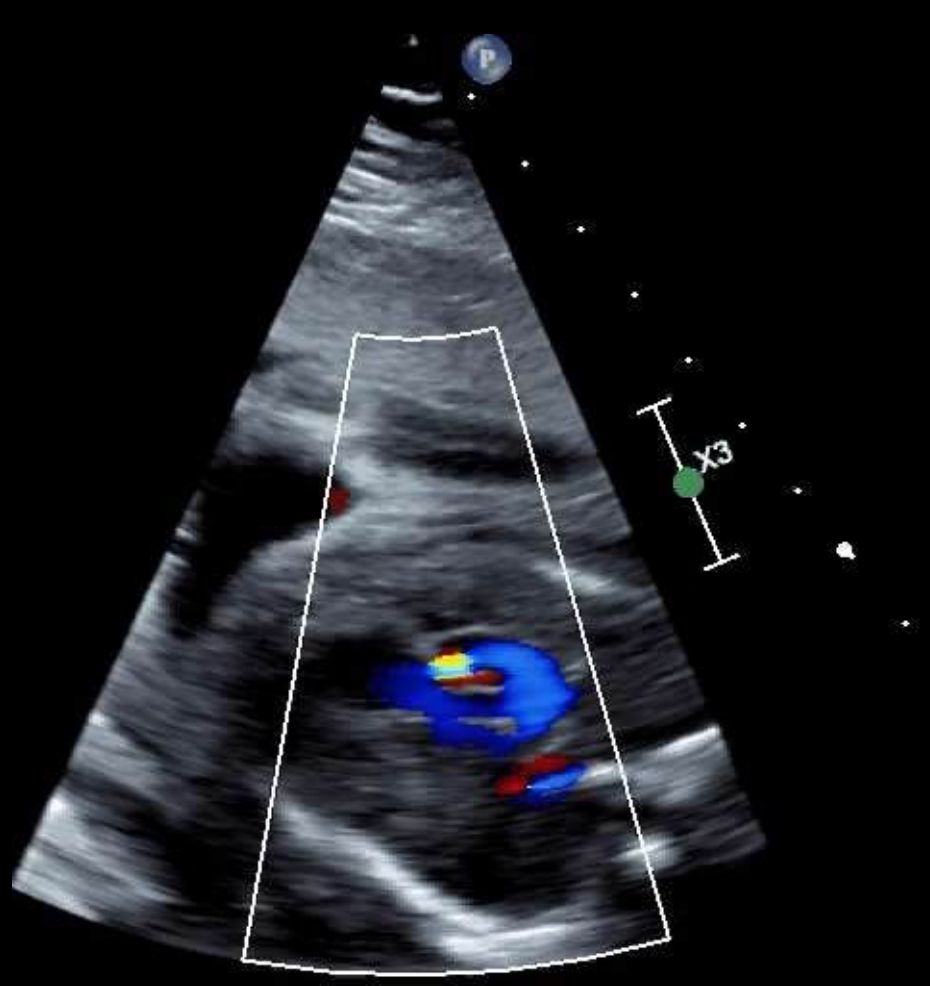
Post FCI

Pre FCI

Fetal echo – one week post FCI at 25 weeks

Improved AS Jet

Antegrade arch flow



JACC: ADVANCES VOL. 3, NO. 3, 2024

ORIGINAL RESEARCH

CONGENITAL HEART DISEASE

Technical Success and Serious Adverse Events for Fetal Aortic Valvuloplasty in a Large 20-Year Cohort

Ryan Callahan, MD,[a] Kevin G. Friedman, MD,[b] Wayne Tworetzky, MD,[b] Jesse J. Esch, MD,[b] Lynn A. Sleeper, ScD,[b] Minmin Lu, MS,[b] Arielle Mizrahi-Arnaud, MD,[c] Roland Brusseau, MD,[c] Terra Lafranchi, NP-C,[b] Louise E. Wilkins-Haug, MD,[d] Stephanie H. Guseh, MD,[d] Carol B. Benson, MD,[e] Mary C. Frates, MD,[e] Mirjam Keochakian, MS,[b] Diego Porras, MD[b]

Callahan et al

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Circulation. 2014 August 19; 130(8): 638–645. doi:10.1161/CIRCULATIONAHA.114.009032.

Fetal Aortic Valvuloplasty for Evolving Hypoplastic Left Heart Syndrome: Postnatal Outcomes of the First 100 Patients

Lindsay R. Freud, MD[1], **Doff B. McElhinney, MD**[1], **Audrey C. Marshall, MD**[1], **Gerald R. Marx, MD**[1], **Kevin G. Friedman, MD**[1], **Pedro J. del Nido, MD**[2], **Sitaram M. Emani, MD**[2], **Terra Lafranchi, NP-C**[1], **Virginia Silva, RN**[3], **Louise E. Wilkins-Haug, MD, PhD**[3], **Carol B. Benson, MD**[4], **James E. Lock, MD**[1], and **Wayne Tworetzky, MD**[1]

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Short term survival advantage BiV vs. HLHS

Freud LR et al

ULTRASOUND
in Obstetrics & Gynecology

isuog

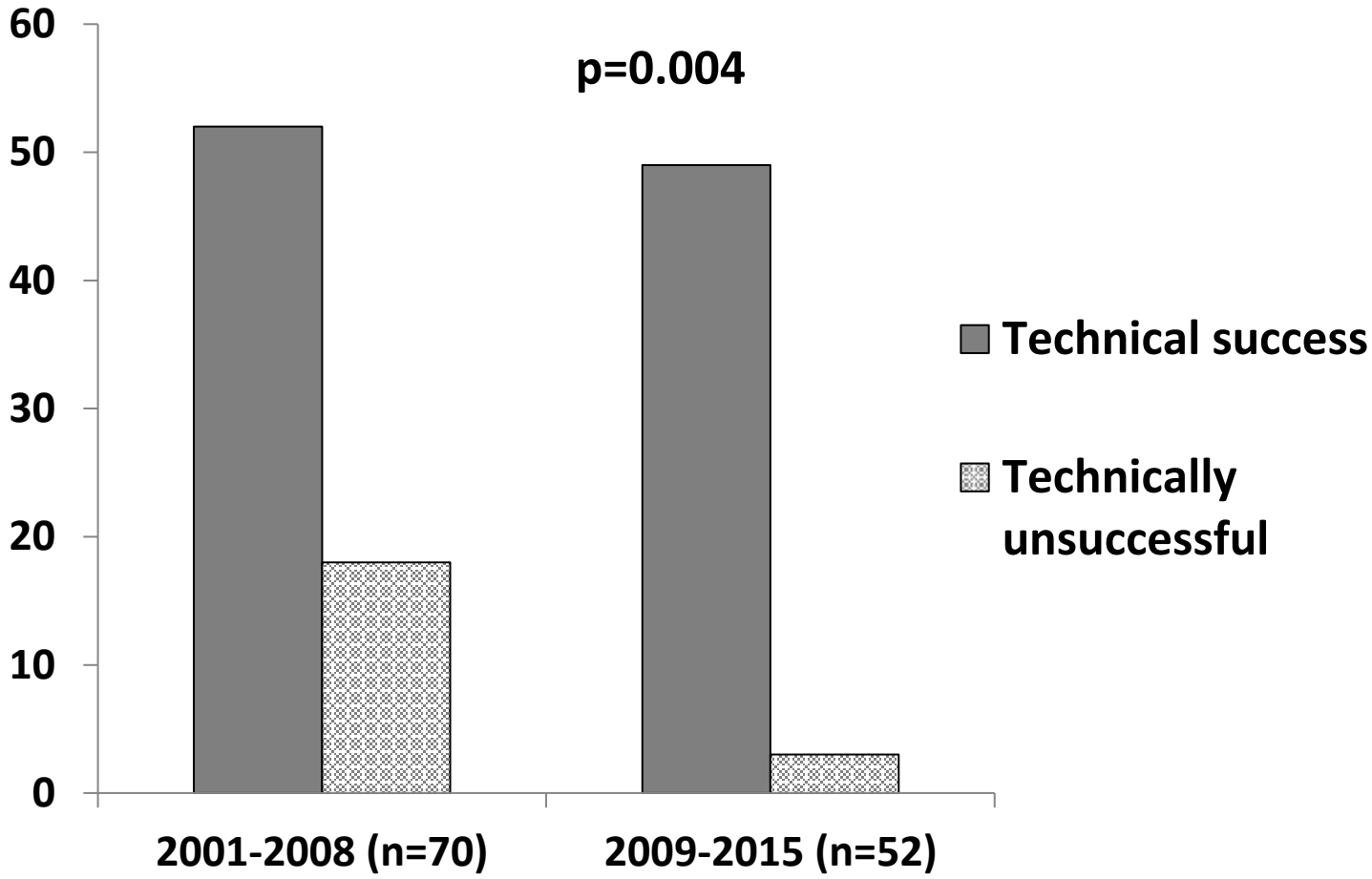
Original Paper | Free Access

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

K. G. Friedman, L. A. Sleeper, L. R. Freud, A. C. Marshall, M. E. Godfrey, M. Drogosz, T. Lafranchi, C. B. Benson, L. E. Wilkins-Haug, W. Tworetzky

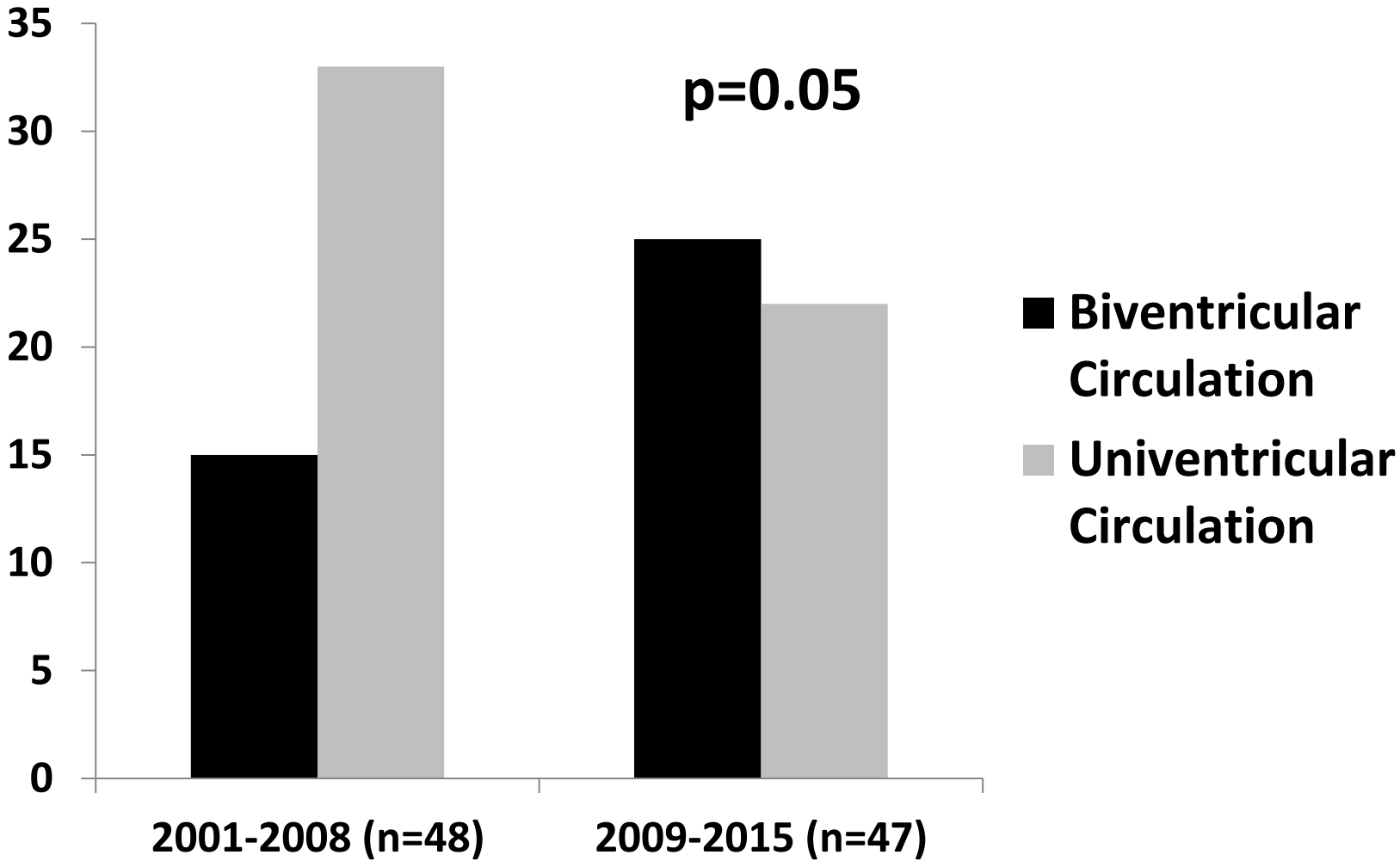
First published: 22 May 2017 | https://doi.org/10.1002/uog.17530 | Citations: 42

Improvement in Technical Success



Friedman et. Al. UOG 2017

Postnatal Outcome in Liveborns after Technically Successful FAV



Friedman et. Al. UOG 2017

What about the other patients.

- Borderline left heart
  - Options for creative surgeries
- HLHS
  - Standard Fontan

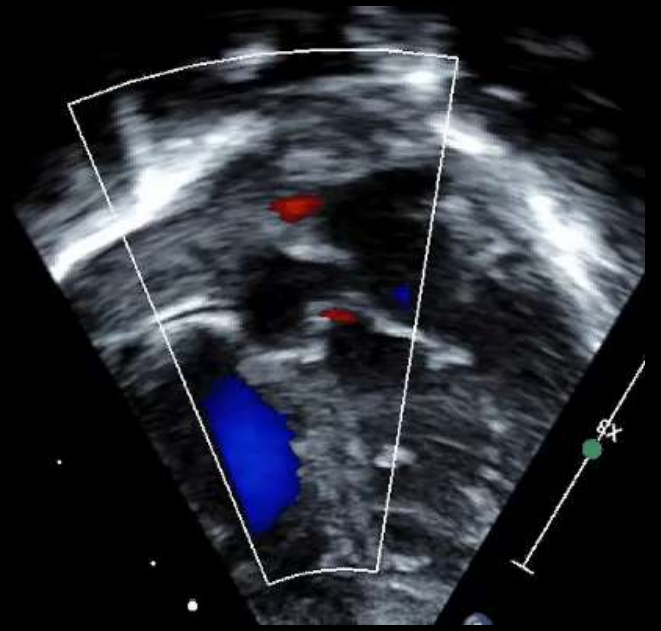
After All The Doom and Gloom
Aortic Stenosis “failures”
Novel approach to “Borderline Left Heart”

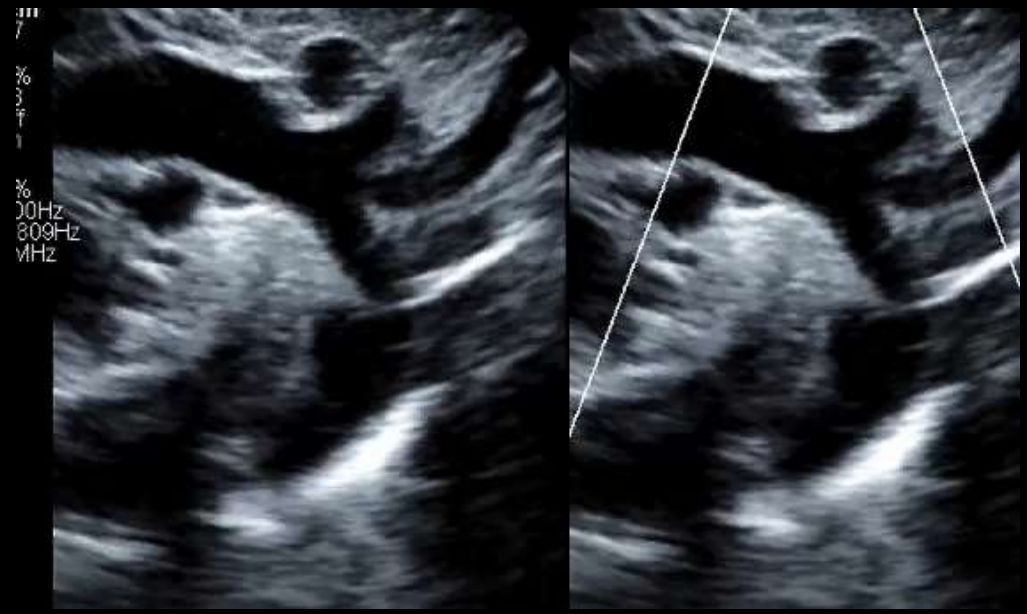
- Staged surgery - conversion from HLHS/BLH to biventricular repair
- Hybrid stage 1 – surgically placed external branch PA bands and PDA stent
- Trans-catheter stage 1 – internal branch PA bands and PDA stent
- Reverse one and a half ventricle repair
- Reverse biventricular repair

s/p Fetal Intervention at 23 w for Aortic Stenosis with Evolving HLHS

Pre-FCI 23 weeks

Neonatal Echo
Borderline LH
HLHS





s/p FCI at 23 weeks, neonatal borderline left heart

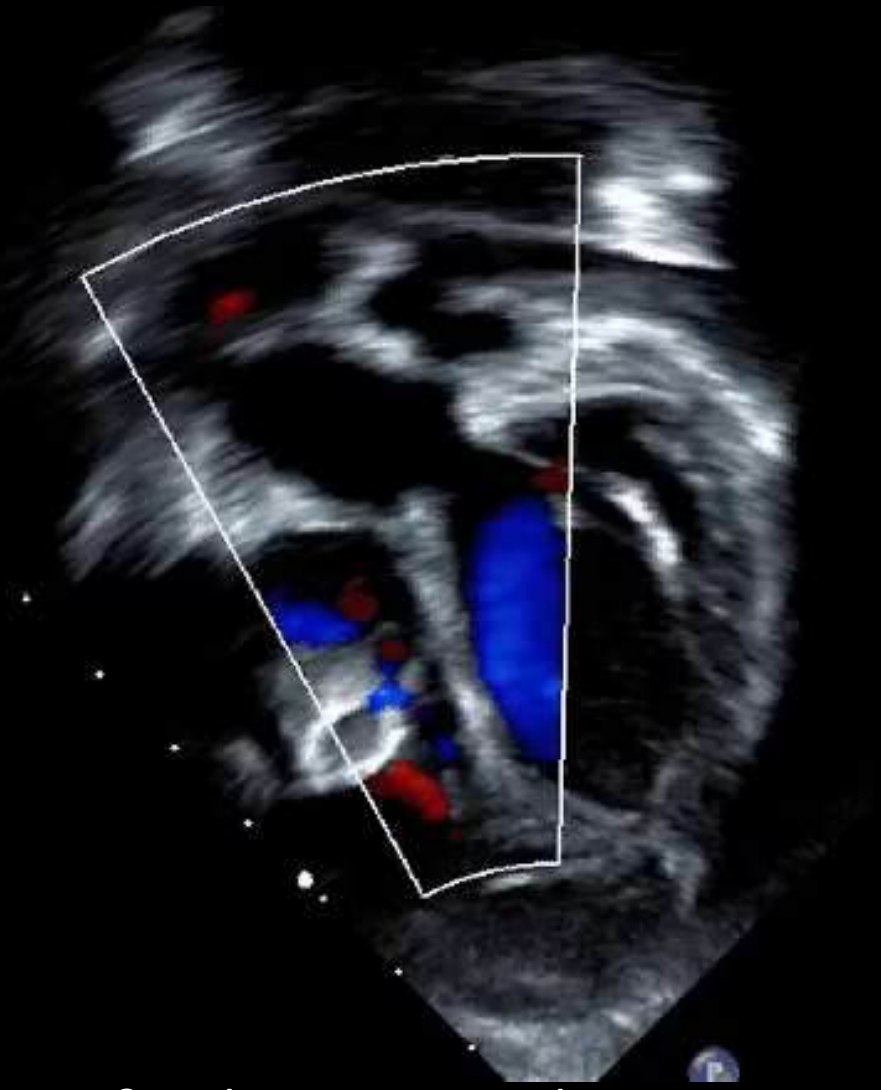
- Modified stage 1 Sano
- Mitral and aortic valvotomy
- Mildly restrictive atrial septum

- Stage 2
- Glenn / SVC to RPA and larger Sano conduit to LPA (increased pulmonary blood flow and LV preload) “super Glenn”
- Endocardial fibroelastosis resection

- Wait for left heart structure to grow

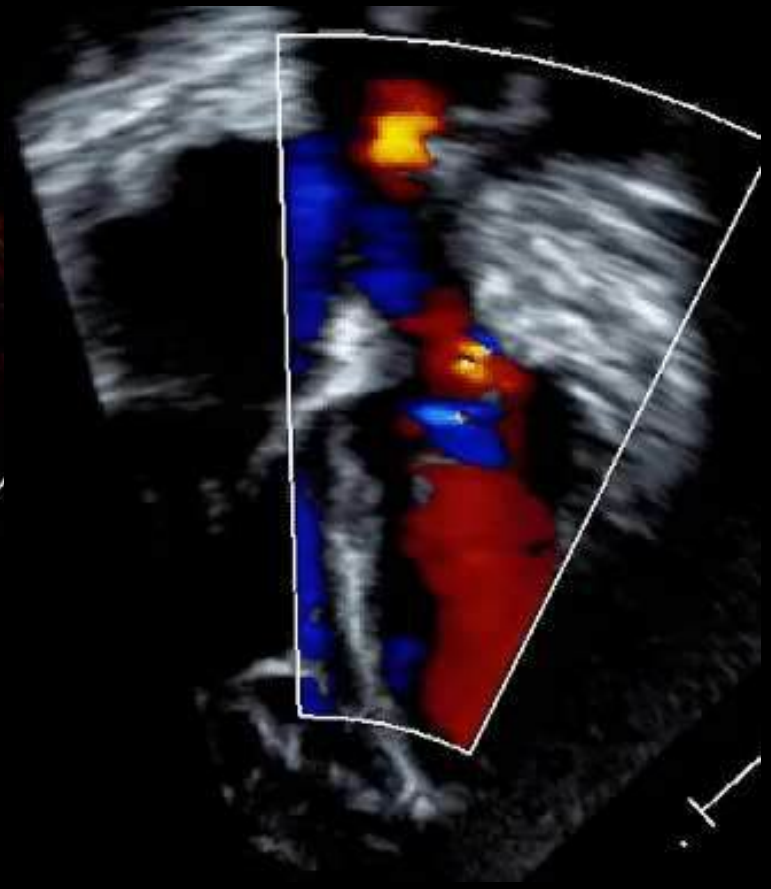
- Around 3 years of age
  - Fontan? (with substantial LV output)
  - Biventricular repair?
  - Other options?

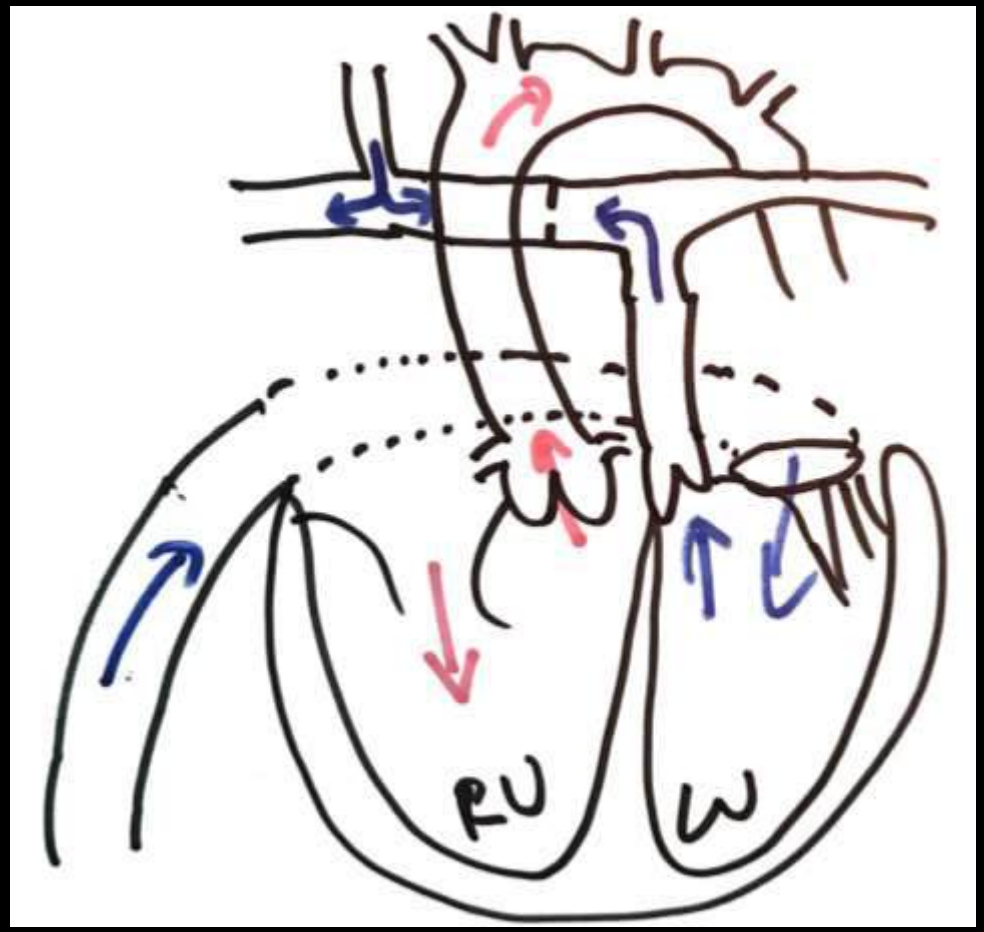
s/p modified stage 1 and 2 for Borderline Left Heart HLHS



Elevated LVEDP. Not a candidate for biventricular repair
Use the LV as a sub-pulmonary ventricle??

Age 3 years. s/p Reverse 1 ½ ventricle repair
Hemi-Mustard (IVC to mitral valve), sub-pulmonary LV (LV to LPA)





Case Reports › Ann Thorac Surg. 2020 Dec;110(6):e529-e530.
doi: 10.1016/j.athoracsur.2020.04.071. Epub 2020 Jun 3.

1.5–Ventricle Repair Using Left Ventricle as the Subpulmonary Ventricle

Corinne W Tan [1], Kevin G Friedman [2], Wayne Tworetzky [2], Pedro J Del Nido [3], Christopher W Baird [3]

Affiliations + expand

PMID: 32504613 DOI: 10.1016/j.athoracsur.2020.04.071

› Semin Thorac Cardiovasc Surg. 2022 Sep 28;S1043-0679(22)00226-X.
doi: 10.1053/j.semtcvs.2022.09.009. Online ahead of print.

Early Experience With Reverse Double Switch Operation for the Borderline Left Heart

Brandi Braud Scully [1], Eric N Feins [2], Wayne Tworetzky [3], Sunil Ghelani [3], Rebecca Beroukhim [3], Pedro J Del Nido [2], Sitaram M Emani [2]

Affiliations + expand

PMID: 36180012 DOI: 10.1053/j.semtcvs.2022.09.009

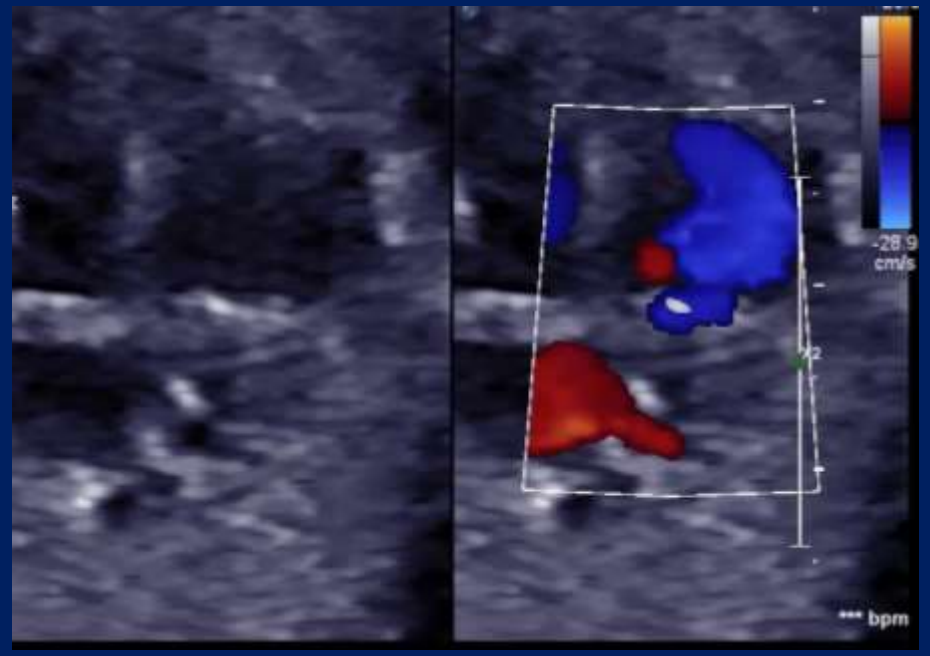
Modified 3rd Stage for HLHS/Borderline LV
Reverse 1 ½ Ventricle Circulation

Theoretical advantages

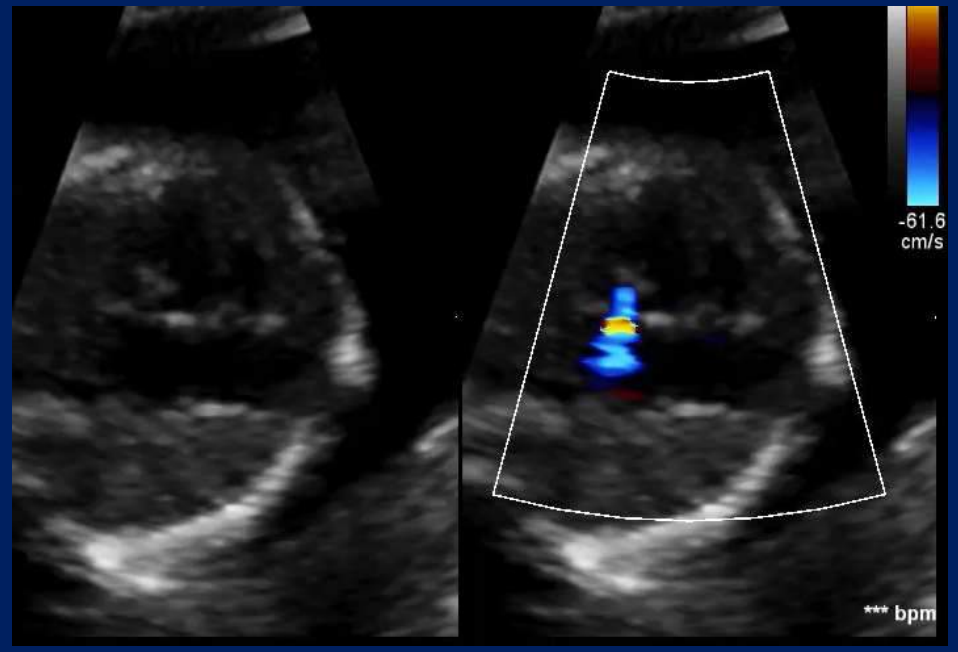
1. Phasic IVC flow
2. Augmented pulmonary blood flow
3. Pulsatile flow in the PAs
4. Long term vascular health 1 ½ vs. Fontan???

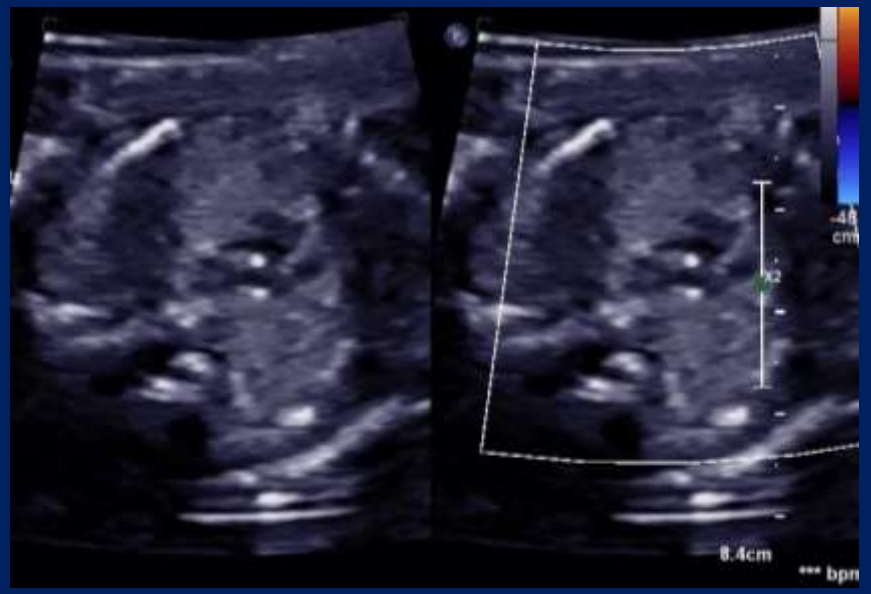
Contraindications

1. High LVEDP
2. Severe LV systolic and diastolic dysfunction
3. MS/MR



22w





FCI for AS

Cannula to LV

Wire across AOV

FCI for AS

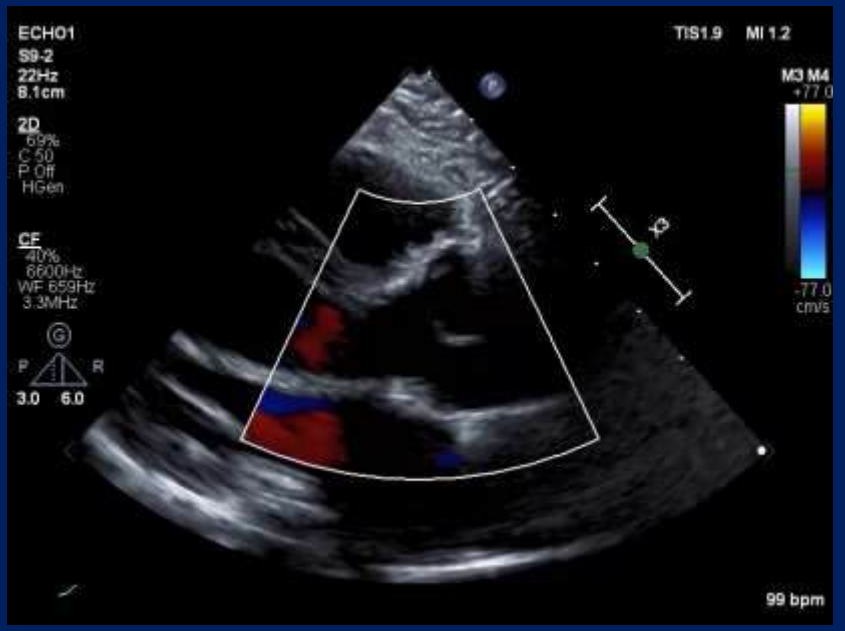
Balloon dilation

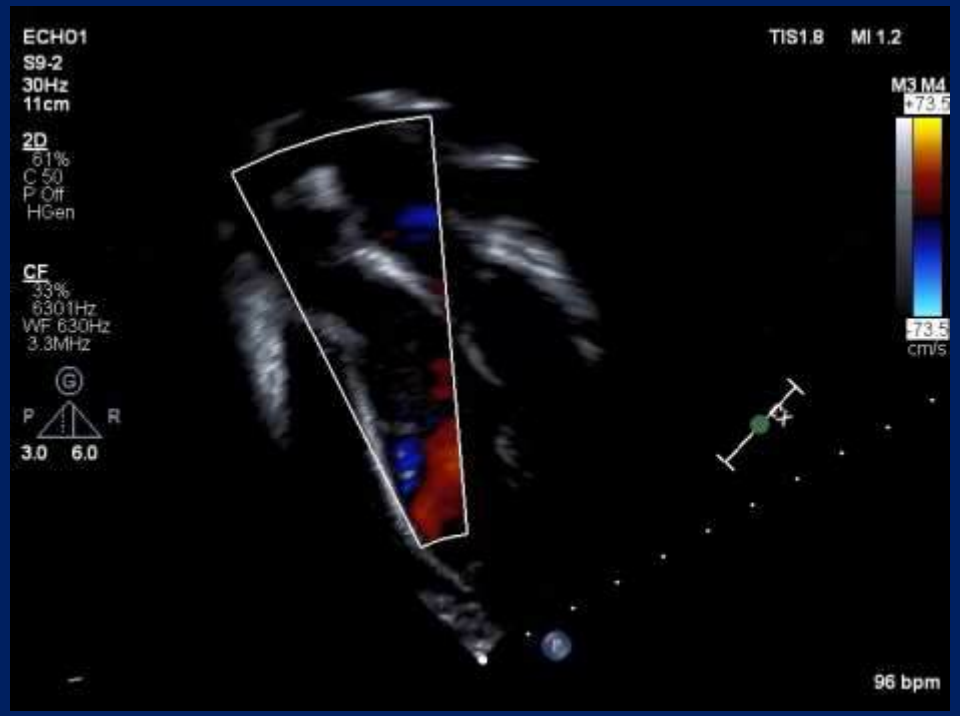
Post BD Flow

Infant Course

- Neonatal BD of AOV
- SubAS resection, AOV plasty
- High LVEDP
- Mixed AOV disease
- Ross?
- Or…?

FCI for AS
3 yrs
Aov Tx





In Utero Intervention for Vein of Galen / Cerebral AVM

Darren B. Orbach

MD, PhD

Chief, Neurointerventional Radiology; Co-Director, Cerebrovascular Surgery and Interventions Center; Sage Schermerhorn Chair in Image-Guided Therapy

Associate Professor of Radiology, Harvard Medical School

Request an Appointment

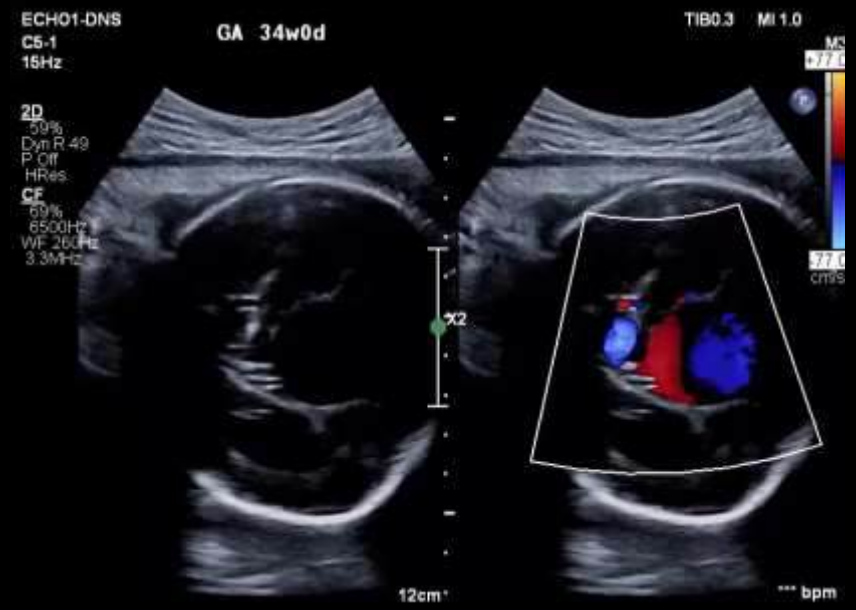
View Research Profile

Vein of Galen / Cerebral AVM

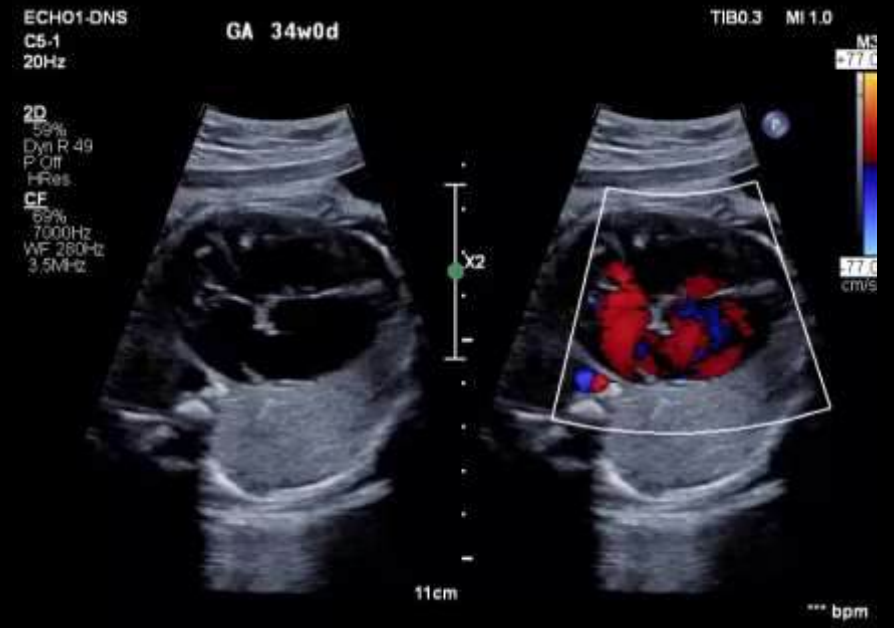
- In Fetal Life
- Enlarge throughout gestation
- Often detected late
- High cardiac output
- Brain damage

- Postnatal
- Systemic runoff
- Pulmonary hypertension
- High output failure
- Brain damage

Vein of Galen Pre In Utero Procedure

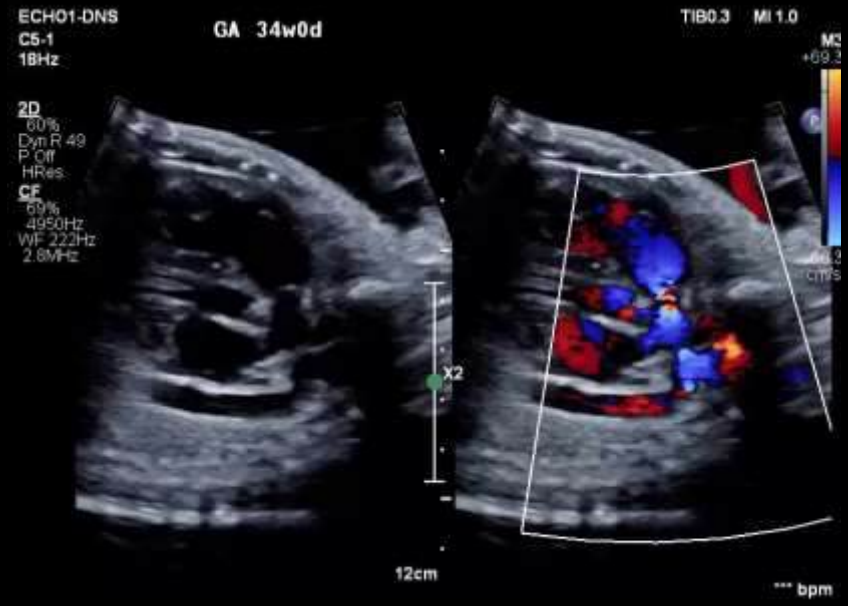


Cerebral AVM



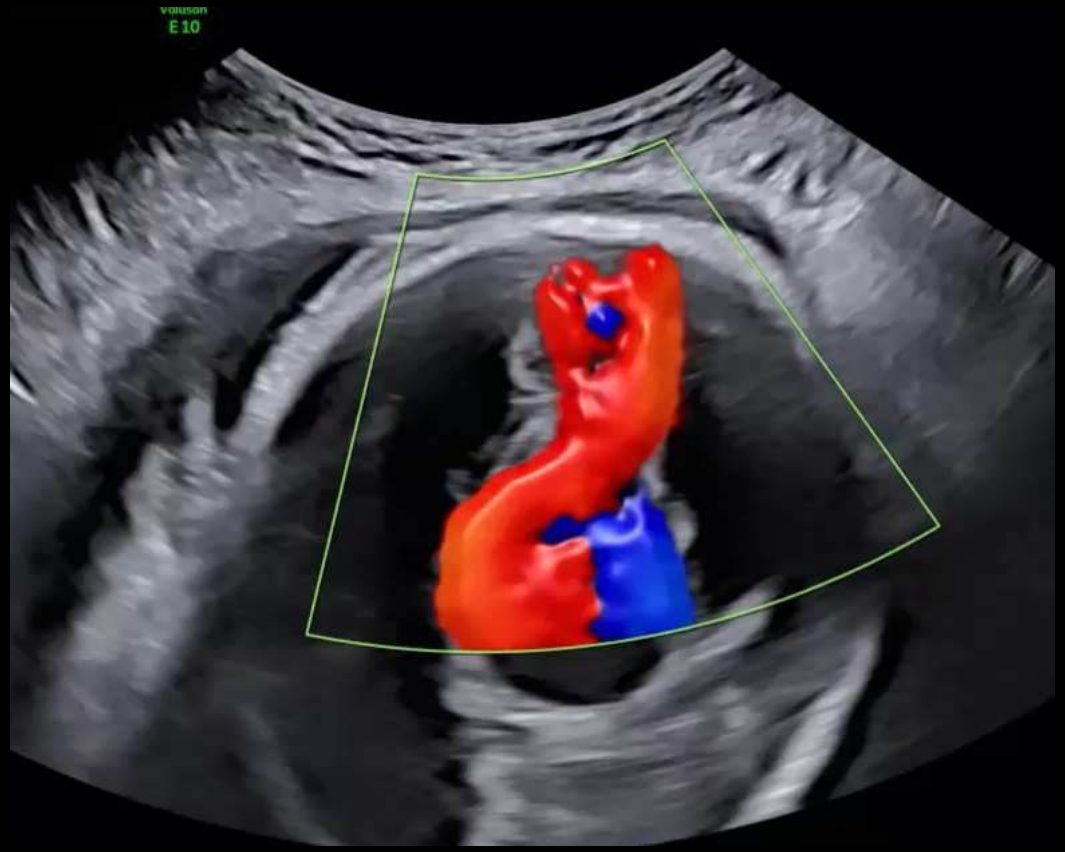
Cardiomegaly

Dilated SVC



Retrograde Arch Flow

Vein of Galen – In Utero Procedure



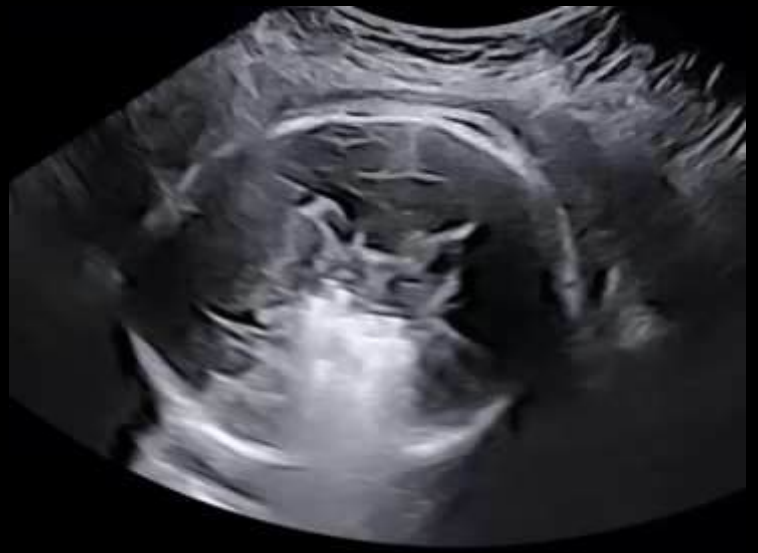
Vein of Galen- In Utero Procedure

Cannula to defect

Initial Coils

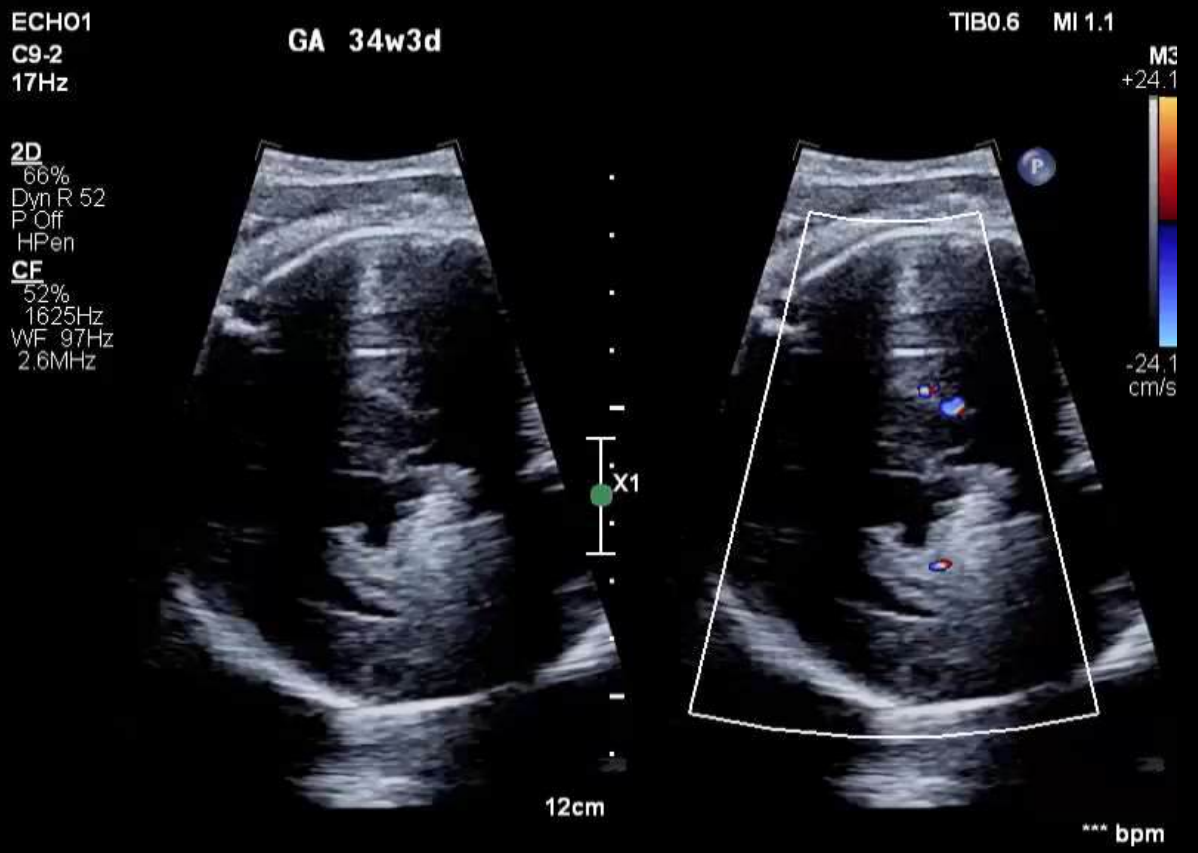
Cannula in defect

Final Coils

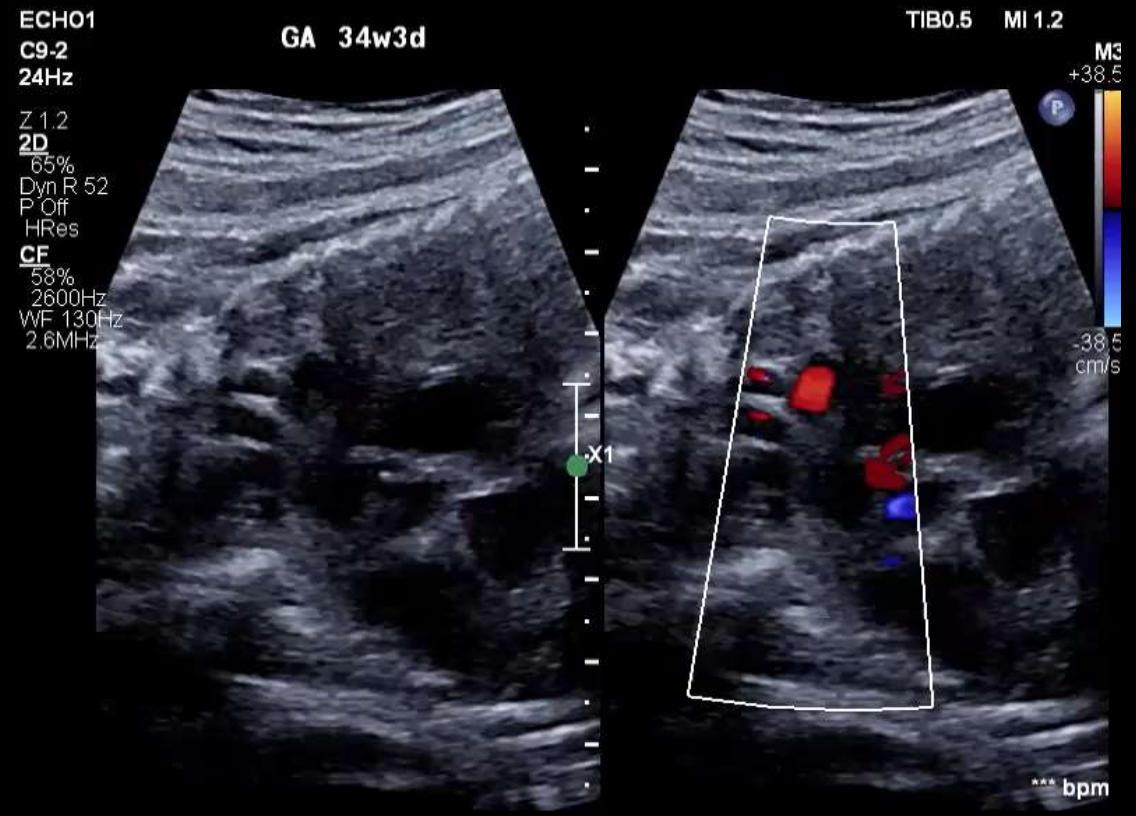


Vein of Galen – In Utero Procedure – Next Day

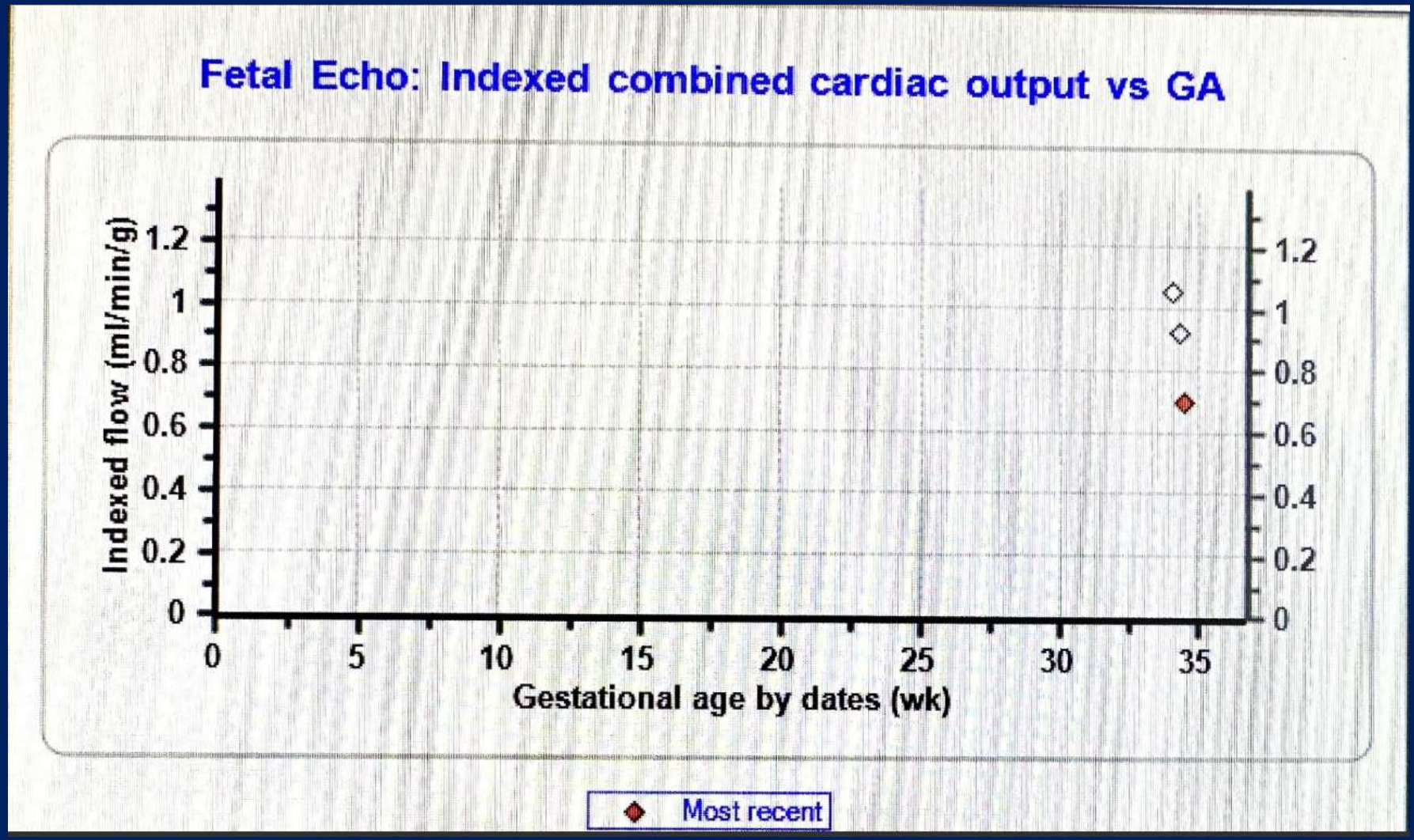
Coils in brain AVM



Improved arch flow



Post Fetal Intervention for Vein of Galen Combined Ventricular Output



Vein of Galen Fetal Procedure

Stroke

CURRENT ISSUE | ARCHIVE | JOURNAL INFO

RESEARCH ARTICLE | Originally Published 4 May 2023 |

Check for updates

Transuterine Ultrasound-Guided Fetal Embolization of Vein of Galen Malformation, Eliminating Postnatal Pathophysiology

Darren B. Orbach, MD, PhD, Louise E. Wilkins-Haug, MD, PhD, Carol B. Benson, MD, Wayne Tworetzky, MB, ChB, Shivani D. Rangwala, MD, Stephanie H. Guseh, MD, Nicole K. Gately, RN, Jeffrey N. Stout, PhD, Arielle Mizrahi-Arnaud, MD, and Alfred P. See, MD

AUTHOR INFO & AFFILIATIONS

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Vein of Galen Fetal Procedure
No postnatal procedure needed

CNN Health Life, But Better Fitness Food Sleep Mindfulness Relationships Watch

Doctors performed brain surgery on a baby before she was born and now she's thriving

By Nadia Kounang and Amanda Sealy, CNN

5 minute read · Updated 9:21 AM EDT, Thu May 4, 2023

Discussion Generation

- Should we expand indications for fetal intervention for AS with evolving HLHS?
- Create neonatal borderline left heart (rather than allowing clear HLHS)
- Expand the surgical options
- Left heart rehabilitation
  - Trans-catheter stage 1
  - Conversion to a biventricular circulation (LV diastolic dysfunction)
  - Reverse biventricular circulation (essentially corrected TGA)
  - Reverse 1 ½ ventricle circulation with a sub-pulmonary LV
- Is that better than Fontan long-term?

Summary

- Fetal AS has a spectrum of severity, progression and outcome
- Fetal intervention is performed when features of evolving HLHS
- Majority of biventricular require some surgery, many require a Ross
- Long term fate of the LV is unknown (concern for diastolic dysfunction)
- HLHS with a Fontan is way worse

Thank You!

Fetal (Neonatal) Cardiac Intervention Team 2000-2025

- Louise Wilkins-Haug, Stef Guseh and Carol Benson, Mary Frates (Obstetrics/Radiology)
- Donna Morash, Ginny Silva, Terra Lafranchi Miah Newman (Nursing)
- Linda Bulich, Arielle Mizrachi, Roland Brusseau (Fetal Anesthesia)
- BWH Maternal Anesthesia Team
- Wayne Tworetzky Kevin Friedman and Colleagues (Echo)
- Cardiac Anesthesia and CICU teams
- Audrey Marshall James Lock Diego Porras Ryan Callahan Jesse Esch Brian Quinn(Cath)
- Fellows, Sonographers
- Pedro Del Nido Sitaram Emani (Cardiac Surgery)
- Referring MFMs and Pediatric Cardiology Colleagues
- Patients

Thank You!

- Questions?

Fetal AS

- N=189
- Technically successful and BiV from birth n=75
- 5/75 cardiac transplant
- Mortality 3/75 (1 post transplant)
- Ross ~25%
- BiV conversion n=8
- Fetal demise 5-10%
- HLHS with expected mortality and HLHS to transplant n=14

Aortic stenosis with evolving HLHS

- Patient selection/rejection (all have features of evolving HLHS)
  - Good: higher LV pressure, normal size LV, minimal EFE
  - Bad: Low LV pressure, EFE, short globular LV
- Procedure
  - Patience, teamwork, communication
  - Fetal position
  - General anesthesia – Epidural only (2004 on)
  - Occasional mini laparotomy – Percutaneous (2004 on)
  - Resuscitation: less frequent, immediate
  - Improved technical success
- Postnatal Novel Surgeries
  - LV rehab with 1V to 2V conversion
  - Using the borderline LV for the pulmonary circulation (instead of Fontan)
  - Valve transplant