Balloon Aortic Valvuloplasty in the Fetus

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The Concept...

- AS may evolve into HLHS in fetal life

- ↑ Stenosis ➔ LV dysfunction ➔ Flow reversal at FO and aortic arch ➔ Poor LV growth

- FAV - promote forward flow and LV growth

- Selection criteria - identify those likely to benefit
Historical Context

• First performed 1989 (Reported 1991)\(^1\)
• 1989 – 1997: 12 cases performed\(^2\)
  • 6 Centers
  • Third trimester
  • Successful balloon placement in 7
  • 1 survived beyond newborn period
• Boston data 2004\(^3\)
  • Fetal aortic valvuloplasty offered to 24
  • Attempted in 20 with technical success in 14

2. Am J Cardiol. 2000;85:1230-33
Historical Context

Flow Chart of All Patients Evaluated for Fetal Aortic Valvuloplasty

Patients Evaluated
n=24

In-utero Valvuloplasty
n=20

Technical Success
n=14
- Liveborn 2 ventricle heart
  n=3
- Live born HLHS
  n=6
- In utero demise
  n=2
- Still in utero
  n=3

Technical Failure
n=6
- Liveborn HLHS
  n=3

Termination
n=1
- Live born HLHS
  n=3

Declined Procedure
n=4

Premature or in utero demise
n=2

(Circulation. 2004;110:2125-2131.)
Current Outcomes

Attempted fetal aortic valvuloplasty
n=100

Fetal demise, n=11
Termination of pregnancy, n=1

Live-born patients
n=88

HLHS from birth
n=57
Comfort care, n=1
Died from sepsis, n=1
Died post-transplant, n=1

Stage 1 surgery
n=54

Biventricular from birth
n=31

BIVENTRICULAR CIRCULATION
n=38
(43% of live-born)

HLHS
n=47

(Circulation. 2014;130:638-645.)
Guidelines

AHA Scientific Statement

Diagnosis and Treatment of Fetal Cardiac Disease
A Scientific Statement From the American Heart Association

Endorsed by the American Society of Echocardiography and Pediatric and Congenital Electrophysiology Society

The American Institute of Ultrasound in Medicine supports the value and findings of the statement.*

The Society of Maternal Fetal Medicine supports the statement’s review of the subject matter and believe it is consistent with its existing clinical guidelines.†

Mary T. Donofrio, MD, Chair; Anita J. Moon-Grady, MD; Lisa K. Hornberger, MD; Joshua A. Copel, MD; Mark S. Sklansky, MD; Alfred Abuhamad, MD; Bettina F. Cuneo, MD; James C. Huhta, MD; Richard A. Jonas, MD; Anita Krishnan, MD; Stephanie Lacey, DO; Wesley Lee, MD; Erik C. Michelfelder, Sr, MD; Gwen R. Rempel, RN; Norman H. Silverman, MD, DSc, FAHA; Thomas L. Spray, MD, FAHA; Janette F. Strasburger, MD; Wayne Tworetzky, MD; Jack Rychik MD; on behalf of the American Heart Association Adults With Congenital Heart Disease Joint Committee of the Council on Cardiovascular Disease in the Young and Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and Council on Cardiovascular and Stroke Nursing

(Circulation. 2014;129:2183-2242.)
Selection Guidelines

• Favorable anatomy (no atresia)
• Minimal subvalvar obstruction
• Evidence for process of evolving AS
  • Depressed LV function and flow abnormalities
    – Retrograde/bidirectional flow in the transverse arch
    – Monophasic inflow across the mitral valve
    – L-R flow across the atrial septum
    – Bidirectional flow across pulmonary veins

• Favorable factors for 2-ventricle repair:
  – LV long axis z-score >-2
  – LV – >10mmHg across AV or 15mmHg on MR
  – Mitral valve diameter z-score > -3
Starting a Program

Research Article
The Learning Curve for a Fetal Cardiac Intervention Team

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• Dedicated multidisciplinary team (MFM, Intervention, Fetal Echocardiography)
• Animal model
• Equipment
Edited Case: Fetal Aortic Valvuloplasty

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Fetal Aortic Valvuloplasty: Edited Case

Clinical data

- Fetus at 27-28 weeks gestation
- Estimated weight: 1 kg
- Mom: 35 year-old female, 2\textsuperscript{nd} gestation
- First child non-dysmorphic
- No US signs of chromosomal abnormality
- Associated malformation: pelvic renal dilatation
Clinical data

- Cardiac diagnosis @ 27-28 weeks
  - Critical AS
  - Impending HLHS
Fetal Aortic Valvuloplasty: Edited Case

Follow up

• Mom was discharged home following day with no complications

• Fetus
  – Active, normal heart rate, better LV fxn
  – Moderate pleural effusion
Fetal Aortic Valvuloplasty: Edited Case

After 3 days (June 17, 2011)
Fetal Aortic Valvuloplasty: Edited Case

Neonatal Period

- DOL 3: Balloon aortic valvuloplasty
- DOL 10: Hybrid (PA banding + PDA stenting)
- DOL 16: Atrial septostomy
Follow up in infancy

- Serial echos showed improving LV fxn
- ASD ~ 4mm; PSG AoV ~ 30 mmHg; no AI
- No gradient across the ductal stent or RAA
- PA bands with ~ 50-60 mmHg gradient
Follow up in infancy

- LV, MV and AoV of adequate size
- At 9 months of age - referred to Boston Children’s Hospital for possible BV repair
Cath @ 9 Months: Pre LV Overhaul

Courtesy: Lisa Bergensen
LV overhaul: EFE resection
BV Circulation: TEE after CPB

Courtesy: Pedro del Nido and Tal Geva
Post Surgical LV Overhaul and BV Repair

Follow up

• 10 days in hospital
• Uneventful course
• Currently living in the northeast of Brazil
• Asymptomatic, on Propranolol
• Normal systolic and diastolic LV fxn
• PIG across AoV < 20 mmHg
• No gradient within PAs; Normal RVSP
• ASD 2 mm
Conclusions

• Evolving with improving technical success

• Very specialized process (Limited centers)

• Post-natal interventions likely

• Longer-term follow-up data
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