Can MRI Aid Decision-making in the Borderline Small Left Ventricle?

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Disclosures

- None
Outline

• Univentricular vs. biventricular
• Research
• Case example
Univentricular vs. Biventricular
Univentricular vs. Biventricular

Ventricular + valvar dysfunction
Atrial + ventricular arrhythmias
Protein losing enteropathy
Plastic bronchitis
Hepatic and renal dysfunction
↑ Thromboembolic risk
Neurodevelopmental deficits
Psychosocial challenges
Univentricular vs. Biventricular

Overall Survival (n=1,052)

40-Year Follow-up After the Fontan Operation, Pundi KN et al., JACC 2015
Univentricular vs. Biventricular

1V

2V

Ventricular + valvar dysfunction
Pulmonary hypertension
Need for multiple procedures
Neurodevelopmental deficits
Psychosocial challenges
Univentricular vs. Biventricular

- Detailed diagnostic assessment
- Weighing of patient specific factors
- Reversibility
- Multidisciplinary review
- Parental involvement
Univentricular vs. Biventricular

- Challenging decision
- Major consequences
- CMR may enhance decision making
  - Ventricular volumes
  - Blood flow
  - Myocardial fibrosis
Cardiovascular magnetic resonance parameters associated with early transplant-free survival in children with small left hearts following conversion from a univentricular to biventricular circulation

Puja Banka¹,³*, Barbara Schaetzle¹,³,⁵, Rukmini Komarlu¹,³,⁶, Sitaram Emani²,⁴, Tal Geva¹,³ and Andrew J. Powell¹,³
• Retrospective study
• Dx:
  1) Borderline hypoplastic left heart structures
  2) Right-dominant AV canal defect
    – No conotruncal abnormalities
• Uncertainty whether left heart could support a BiV circulation
• All had CMR prior to surgical or cath procedure that converted from a 1→2 ventricle circulation
Borderline Hypoplastic Left Heart
Borderline Hypoplastic Left Heart

- N=22
- Median age at CMR: 39 months (0-82)
- Circulation at CMR
  - 3 prostaglandin
  - 3 stage 1 palliation
  - 15 BDG
  - 1 Fontan
- 17 had a prior procedure to increase LV flow
  - 13 additional source of pulmonary flow
  - 16 ASD restricted
Borderline Hypoplastic Left Heart
Borderline Hypoplastic Left Heart
Borderline Hypoplastic Left Heart

- Median post-op follow-up: 40 mo (4–84)
- 16 with BiV circulation at latest follow-up
- 6 unsuccessful conversions
  - 3 deaths
  - 2 takedowns to 1V circulation
  - 1 heart transplant
Borderline Hypoplastic Left Heart

Imaging parameters associated with transplant-free BiV survival:

- CMR
  - ↑ LVEDVi
  - ↑ LV/RV SV ratio
  - ↑ MV/TV inflow ratio
- Echo
  - ↑ LVEDVi
Borderline Hypoplastic Left Heart

ROC and survival analysis

Survival With Biventricular Circulation

Optimized sensitivity

Optimized specificity
Borderline Hypoplastic Left Heart

• +LGE in 15 of 22
• NOT associated with transplant-free BiV survival
• All 15 had EFE resection
Right-dominant AV Canal
Right-dominant AV Canal

- N=10
- Median age at CMR: 6 mo (2-75)
- Circulation at the time of CMR
  - 2 stage 1 palliation
  - 3 PA band
  - 5 BDG
- All successful biventricular repair at discharge
Right-dominant AV Canal

- Median post-op follow-up: 28 mo (3–51)
- 1 late death (12 mo post-op, MS, LVOTO, resp illness)
- 1 pt underwent MV replacement
- All RVp < ½ systemic
- Too small a sample size for analysis of predictors
- LV EDVi: range 22–39 ml/m²
- LV/RV stroke volume ratio: range 0.19–0.44
Case

2.5 yo with MS and AS s/p stage 1 with a RV-PA conduit, and s/p BDG
Case

AV valve flow
Case

LGE imaging
Case

2.5 yo with MS and AS s/p stage 1 with a RV-PA conduit, and s/p BDG

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Underwent BTS, ASD restriction, and AoV plasty
Case

Underwent BTS, ASD restriction, and AoV plasty
Case
Case

Pre-op

Post-op
# Case

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- Cath and echo data reviewed
- Underwent biventricular conversion
- No subsequent cardiac procedures
Summary

- CMR parameters can guide decision-making regarding 1 vs 2 ventricle repairs
- CMR can assess the impact of procedures designed to rehabilitate the small LV
- Additional research is ongoing to refine patient selection and management strategies