Atrioventricular Valve Repair in Patients with Single Ventricle Physiology: Impact of Ventricular Function and Morphology, Valve Morphology, and Mechanism of Insufficiency on Outcomes

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Disclosure

Authors have no disclosures
Single Ventricle and AV Valve Insufficiency

• Background

Contraindication for Fontan operation in the past

Still a risk factor for death or failure of cavopulmonary circulation

• Repair of AV Valve insufficiency may alter the risk for poor outcome
Hypothesis

**Primary Hypothesis**
Repair modifies the natural history of functional single ventricle physiology with AV valve regurgitation

**Secondary Hypothesis**
Survival Impacted by: AV valve morphology  
Mechanism of insufficiency  
Ventricular morphology and function
1998 to 2008 review

1. Blinded re-evaluation of echocardiograms
   Primary Outcome: death or transplantation
   Secondary Outcomes:
   - Re-operation
   - AV Valve Function
   - Ventricular Function

2. A case match control for primary outcome
Patient Population

57 (13.5%) AV valve repair
422 functional single ventricle patients

Median age: 6.8 months
(31+/-46 months 8 days to 17.4 years old)

Median weight: 6.9 kg
(11.3+/-11 kg, 2.1-58 kg)
Diagnosis/Morphology

N=57

Diagnosis

Dominant ventricle

Undetermined
3%

Left ventricle
14%

Right ventricle
83%

AV valve

Common AV valve
28%

Mitral
4%

Undetermined
2%

Tricuspid
66%
Timing of AV valve repair

Patients (n)

- Stage I: 2.5%
- Stage II: 60%
- Between Stage II and III: 17.5%
- Fontan: 17.5%
- After Fontan: 2.5%
Preoperative echo findings

Insufficiency

Ventricular dilatation

Mild to Moderate

Moderate

Severe

None

Ventricular dysfunction

Mild

Moderate

Severe

None
AV valve abnormalities

- Annular dilatation: 70%
- Prolapse: 50%
- Dysplasia: 50%
- Cleft: 20%
- Chordal elongation: 40%
- Restriction: 50%
Primary mechanism of AV valve insufficiency

- Dysplasia: 35%
- Restriction: 2%
- Prolapse: 46%
- Cleft: 2%
- Annular dilatation: 15%
Repair techniques

- **Annuloplasty** (localized, 96%, ring 4%)
- **Commissuroplasty**
- **Suture repair of leaflet dysplasia**
- **Edge-to-edge repair** (Alfieri’s stitch)
- **Cleft closure**

Percentage of the patients
Overall survival
Entire cohort (n=57)

Freedom from death or transplant

Median follow-up: 59 months (1-128 months)

Cum Survival

Months from valve repair

1 year: 79%
3 year: 69%

57 43 34 28 15 4
AV Valve Repair Result

Grade: 2.1+/−0.5 vs. 0.84+/−0.6
P=0.0001

Grade

3
Severe

2
Moderate

1
Mild to Moderate

2
Moderate

1
Mild

0
None or trivial

(12%)
Factors Impacting Survival
Valve and Ventricular Function

Freedom from death/transplant

- Successful repair/normal function (n=41, 74%)
- Failed repair/normal function (n=5, 8%)
- Failed repair/poor function (n=9, 15%)
- Successful repair/poor function (n=2, 3%)

Log rank: P=0.02

Failed repair ≥ Moderate AVVR
Poor function ≥ moderately reduced
Death or Transplant

Impact of Diagnosis and Morphology

Diagnosis

- AVSD
- HLHS
- TA/DILV
- DORV

Ventricular morphology

- Indeterminate
- LV
- RV

Log rank: P=0.4

Months from valve repair

AV valve morphology

- Mitral valve
- Tricuspid valve
- Common AV valve

Log rank: P=0.8

Months from valve repair

Impact of Diagnosis and Morphology

Mitral valve

Log rank: P=0.8

Common AV valve

Months from valve repair
Competing risk outcomes

Alive/no re-valve repair
Dead
Alive after transplant
Alive after re-valve repair

3 years
Primary Outcome
Predictors for death/transplant

• Multivariate logistic regression model

  Increased indexed AV valve annulus size \( p=0.05 \)
  Cardiopulmonary bypass time \( p=0.04 \)
  Post-repair ventricular function \( p=0.02 \)

• Cox regression model

  Annular dilatation as the primary mechanism \( p=0.02 \)
  Cardiopulmonary bypass time \( p=0.02 \)
  Post-repair ventricular function \( p=0.02 \)
  Post-repair residual AV valve insufficiency \( p=0.03 \)
Secondary Outcome
Re-valve repair or replacement

N=10 (17.5%)
(Median 21 months)

Re-repair: n=6
Replacement: n=4

Freedom from death/transplant
Overall (n=10)

1 year survival: 70%

Freedom from death/transplant
Re-repair vs. replacement

Log-rank: p=0.6
Predictors for re-repair/replacement

- Multivariate logistic regression model

  - Young age at repair \( p=0.05 \)
  - Small body surface area \( p=0.03 \)
  - Increased indexed AV valve annulus size \( p=0.002 \)
  - Increased ventricular dimension \( p=0.01 \)
  - Leaflet dysplasia \( p=0.03 \)
  - Post-repair residual AV valve insufficiency \( p=0.05 \)

\( n=49 \)
Case match control study

n=50

Freedom from death/transplant

Hazard function

Matching criteria:
- Age
- Body weight, BSA,
- Diagnosis
- Ventricular/AV valve morphology
- Ventricular function

Valve repair group

Case match control

Log-rank: p=0.015
Impact of Valve and Ventricular Function

Group 1: Successful repair/normal function

- Valve repair group (n=35) (successful repair/normal function)
- Case match control (n=35)

Log rank: p=0.36

Group 2: Successful repair/poor function

- Valve repair group (n=9) (successful repair/poor function)
- Case match control (n=9)

Log rank: p=0.03
Summary

AV valve insufficiency was anatomic in 85%

AV valve repair was successful in 88%

Good repair with preserved function provided equivalent survival to case match controls.
Summary

Predictors of Failure
- Poor Function
- Dilated annulus and ventricle
- Long bypass
- Residual AV valve insufficiency

Strategies designed to preserve ventricular function and geometry may alter the natural history

Early successful repair may alter late outcome