

# Fontan Outcomes Network Case Review Conference

*Should this patient have a Fontan?  
Implication of AV valve Interventions*

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*March 15, 2022*

*Mayo Clinic*

*Summary Slides*



# A case of a 6-year-old was presented

- Unbalanced AVSD, DORV, asplenia
- S/p DKS, atrial septectomy, BT shunt
- S/p left BD Glenn, AV valve repair
- S/p redo AV repair
- S/p common AVV replacement 23 mm St Jude mechanical valve
- S/p common AVV replacement 25 mm porcine bioprosthesis (Sep 2019)

# Summary of Discussion

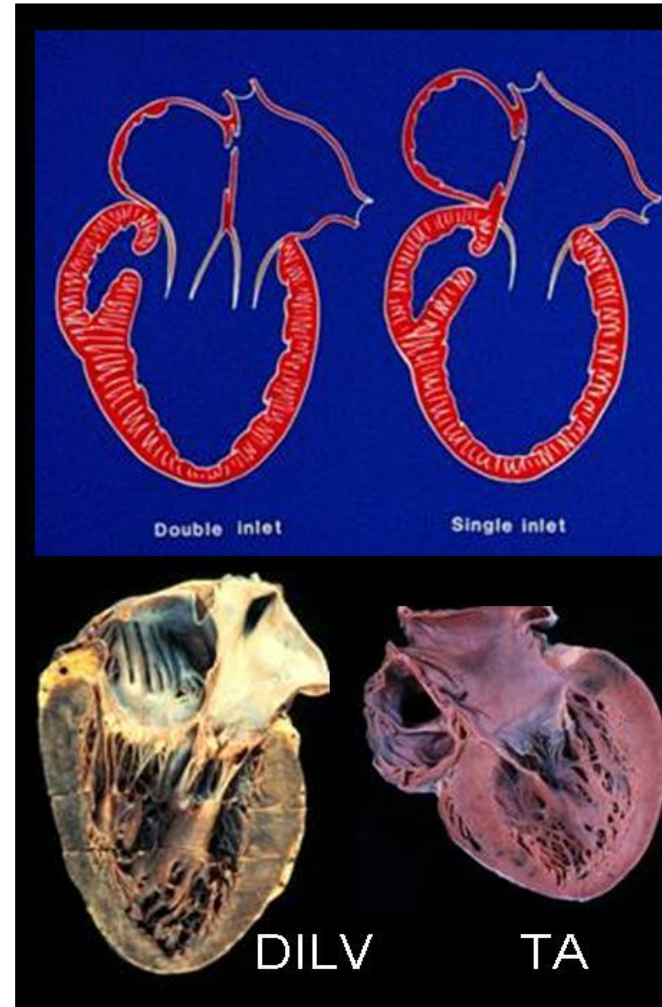
When to intervene on the AV valve in patients with functional single ventricle?

- *After Glenn ...before Fontan ?*
- *Repair vs. replacement ?*
  - *Repair technique depends on AV valve morphology*
- Patient/family need to be capable of managing warfarin issues
- If no Fontan, then what ?
  - Shunt vs. transplant

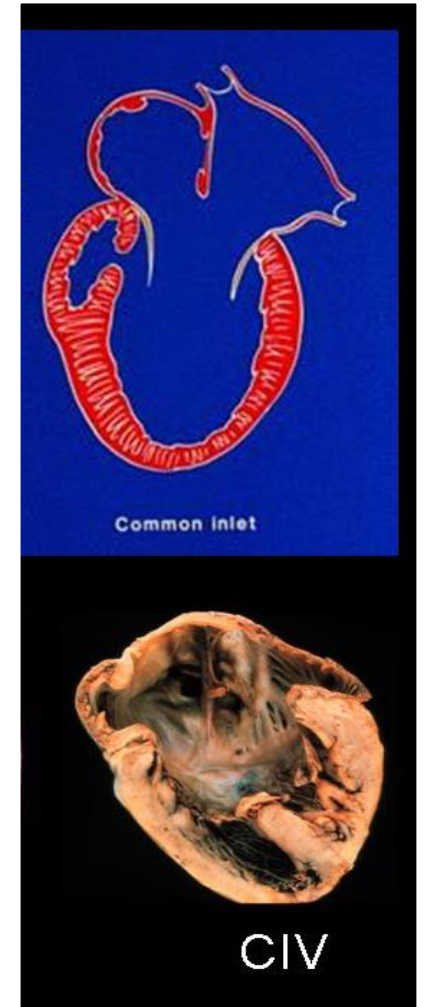
# All AV valves are not the same ...



Anatomically TV



Anatomically MV



Complex, often unbalanced  
Common AV Valve

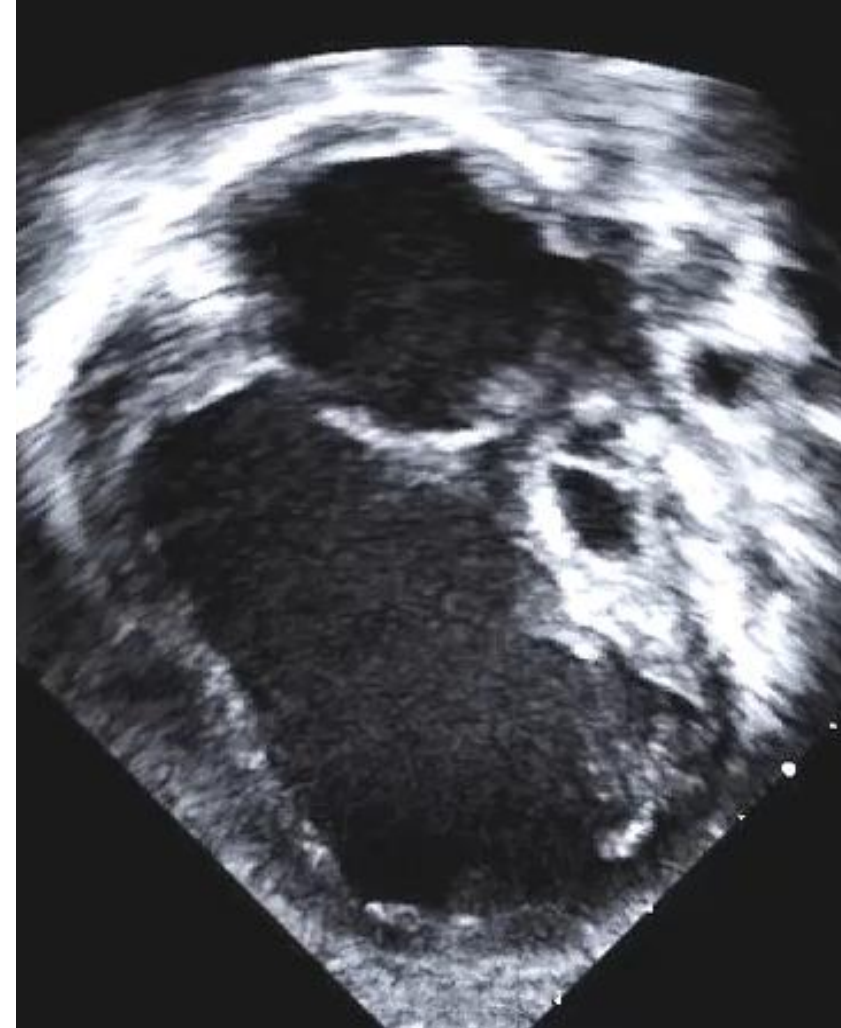
# “Mitral” Morphology AV Connections

- Tricuspid Atresia
  - 2 “robust” papillary supports for each valve
  - No septal attachments
  - 360° supporting fibrous annulus
- “Less” prone to annular dilation and regurgitation



# “Tricuspid” Morphology AV Connections

- HLHS
  - Multiple small papillary muscle supports
  - Septal attachments
  - Incomplete (not 360°) supporting fibrous annulus
- “More” prone to dilation and regurgitation than valves with Mitral morphology



# Common Inlet Ventricles

- Complex Valve Anatomy
  - Abnormal AV valve leaflets
  - Multiple Regurgitant Origins
  - Anatomic similarities to common valves in unbalanced AVSD
  - Prone to dilation and regurgitation
- Challenging to Repair
- Relative Frequency of AVV regurg.
- Common AVV > HLHS > DILV > Tri. At.



# Atrioventricular Valve Failure in Fontan Palliation



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## ABSTRACT

**BACKGROUND** Atrioventricular valve failure (moderate or greater regurgitation, or valve operation) is a risk factor for adverse outcomes in patients undergoing Fontan palliation.

**OBJECTIVES** This study determined the incidence of atrioventricular valve failure and its clinical impact on patients undergoing Fontan palliation.

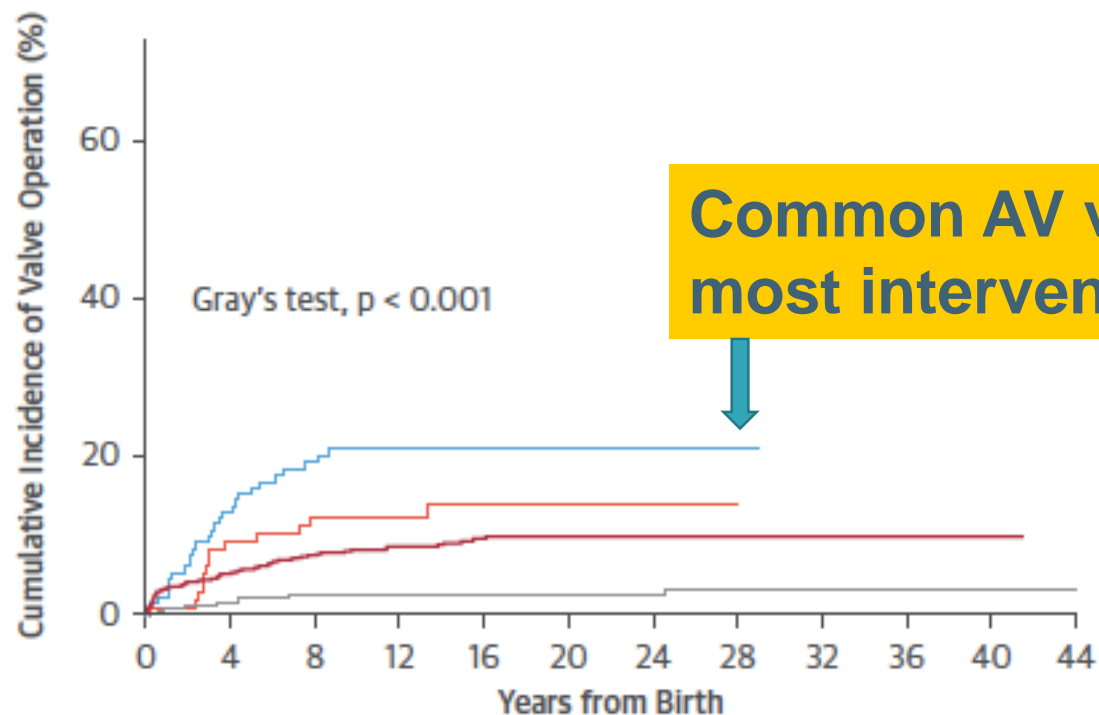
**METHODS** A retrospective cohort longitudinal, bi-national, population-based registry.

**RESULTS** A total of 1,468 patients who underwent Fontan palliation were identified; complete follow-up data were available for 1,199 patients. Six hundred eighty-six patients had 2 atrioventricular valves, 286 had a single mitral valve, 130 had a common atrioventricular valve, and 97 had a single tricuspid valve. A total of 132 repairs were performed in 110 patients, and 15 replacements were performed in 13 patients. The cumulative incidence of atrioventricular valve failure at 25 years of age for patients with a common atrioventricular, single tricuspid, single mitral, and 2 atrioventricular valves was 56% (95% confidence interval [CI]: 46% to 67%), 46% (95% CI: 31% to 61%), 8% (95% CI: 4% to 12%), and 26% (95% CI: 21% to 30%), respectively. In patients without valve failure, freedom from Fontan failure at 10 and 20 years post-Fontan palliation was 91% (95% CI: 89% to 93%) and 77% (95% CI: 73% to 81%), respectively, compared with 77% (95% CI: 69% to 85%) and 54% (95% CI: 42% to 68%), respectively, in patients with valve failure (hazard ratio: 2.43; 95% CI: 1.74 to 3.39;  $p < 0.001$ ).

**CONCLUSIONS** Atrioventricular valve failure occurs frequently in patients undergoing Fontan palliation. Patients with valve failure are twice as likely to have their Fontan circulation fail than those without valve failure. (J Am Coll Cardiol 2019;73:810–22) © 2019 by the American College of Cardiology Foundation.

Cumulative incidence of AV valve failure at 25 yrs of age ... 56%



**B**

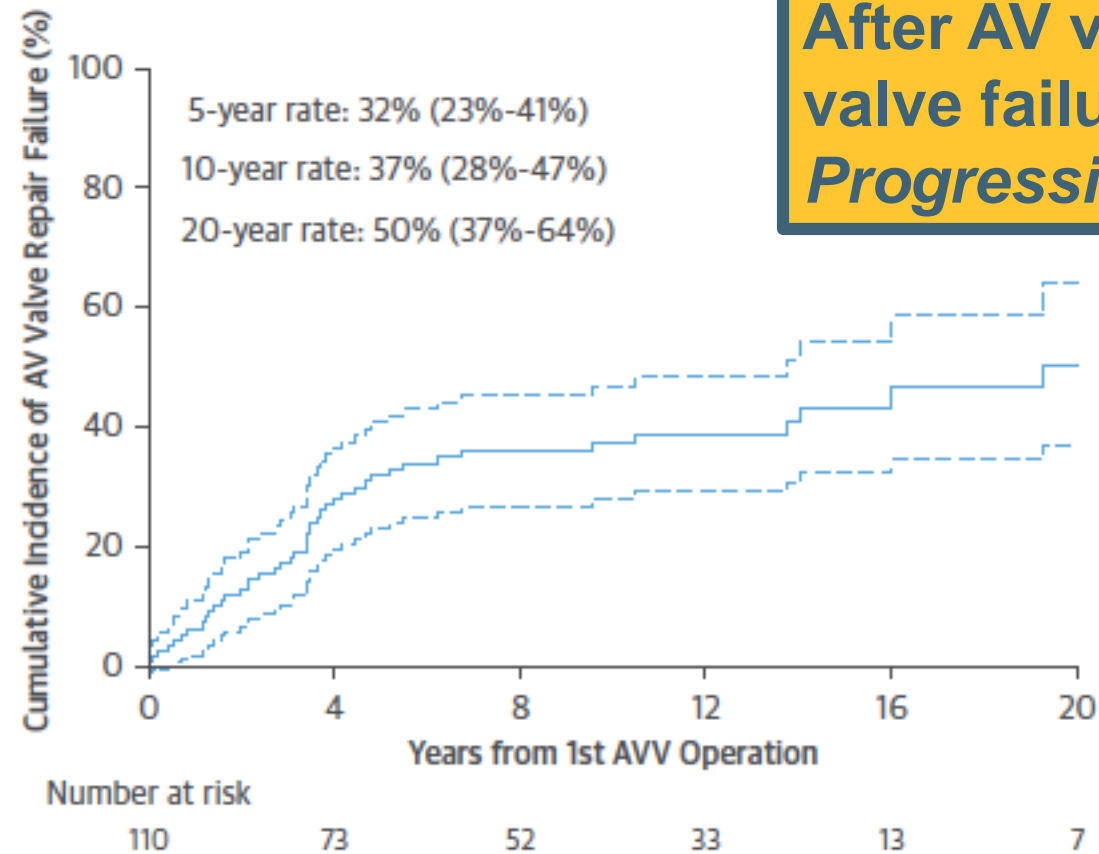
Number at risk

Common AV valve	130	112	96	72	50	37	25	12				
Mitral atresia	97	88	78	58	45	26	20	10				
Tricuspid atresia	286	279	261	226	174	134	95	56	38	24	14	11
Two AV valves	686	646	573	428	303	210	138	75	48	26	13	

— Common AV Valve — Mitral Atresia — Tricuspid Atresia — Two AV Valves

**(A)** Cumulative incidence curve for estimated incidence of atrioventricular valve intervention (repair or replacement) in patients who underwent Fontan palliation and **(B)** cumulative incidence curve for estimated incidence of atrioventricular valve intervention (repair or replacement) stratified by valvular morphology for patients who underwent Fontan palliation. **Dashed lines** denote 95% confidence intervals.

**FIGURE 3** Durability of AV Valve Repair

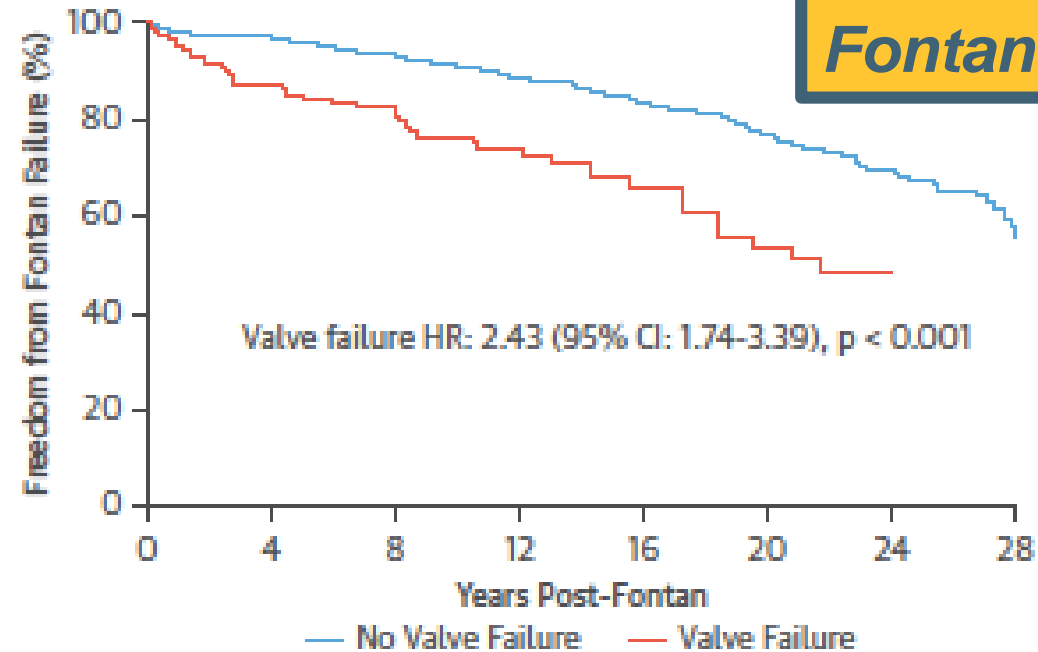


After AV valve repair ...  
valve failure is a  
*Progressive* disease

Cumulative incidence curve for estimated incidence of atrioventricular (AV) valve repair failure (re-repair or replacement or recurrent moderate or greater regurgitation) following initial atrioventricular valve repair in patients who underwent Fontan palliation. **Dashed lines** denote 95% confidence intervals.

**FIGURE 5 AV Valve Function and Fontan Failure**

**Bad AV valves, developed  
*Fontan failure* sooner**



**Number at risk**

No valve failure	1,093	911	650	462	322	199	94	31
Valve failure	102	102	82	49	24	27	10	

Time-varying covariate Kaplan-Meier curve for estimated freedom from Fontan failure in patients with AV valve failure (**orange line**) and in patients without AV valve failure (**blue line**). HR = hazard ratio; other abbreviation as in Figure 3.

- 1167 in Mayo Fontan database
- 153 AV valve repair/replacement

***Valve intervention At or After Fontan ...***

**Increased risk for death or transplant**

**TABLE 1. Strategy for management of the bad atrioventricular valve in the single-ventricle patient\***

Mechanism of mitral morphology

- Structural – chordal, cleft, leaflet (minor) → repair > replace  
marked dysplasia → replace > repair
- Functional – annular/ventricular dilation → repair ≈ replace

Mechanism of regurgitation for tricuspid and common atrioventricular valve morphology

- Structural – chordal, cleft, leaflet (minor) → repair ≈ replace  
marked dysplasia → replace > repair
- Functional – annular/ventricular dilation → replace ≈ repair

Valve morphology

- mitral → repair > replace
- tricuspid → replace ≈ repair
- common → replace ≈ repair

Ventricular size/function

- normal/normal → repair > replace
- RV (dominant) dilation/normal → replace ≈ repair
- LV (dominant) dilation/normal → repair ≈ replace
- Univentricular, unbalanced normal/normal → replace > repair
- Dilation/reduced function → replace vs Tx

# Summary

For many patients with single ventricle physiology issues with the atrioventricular (AV) valve are best dealt with prior to Fontan

- Unfortunately, this involves an extra surgery
- Some of these valves can be repaired, others replaced
- If the AV valve has more than moderate leakage, the long-term outcome after Fontan is not good
- Some of these patients may do better long-term with a shunt or a transplant
  
- Care needs to be individualized since the anatomy of these hearts and the AV valves is quite variable