

Summer 8-12-2021

Long Term Outcome of Childhood Bicuspid Aortic Valve

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Recommended Citation

Kotula, Anna and Yetman, Angela, "Long Term Outcome of Childhood Bicuspid Aortic Valve" (2021).
Posters: 2021 Summer Undergraduate Research Program. 62.
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BACKGROUND

- Bicuspid aortic valve (BAV) with aortic stenosis (AS) often presents in childhood necessitating palliation with a balloon or surgical valvotomy
- Subsequent, more definitive operations, namely aortic valve repair or replacement may be required.
- Comparative data on the morbidity and mortality associated with each definitive repair in a contemporaneous cohort is lacking

OBJECTIVES

- We sought to examine long term outcomes in a cohort of adult patients (age > 18 years) who presented during early childhood with BAV and AS and required intervention prior to 18 years.

METHODOLOGY

- IRB approved retrospective chart review was performed and children with BAV and AS +/- aortic insufficiency identified.
- Patients with Shone's complex, or other cardiac defects aside from isolated aortic coarctation (CoA) were excluded
- Data reviewed to assess for demographic and surgical independent predictors of adverse outcome (stroke, death, heart failure requiring transplant or mechanical support), bacterial endocarditis or need for repeat aortic valve surgery

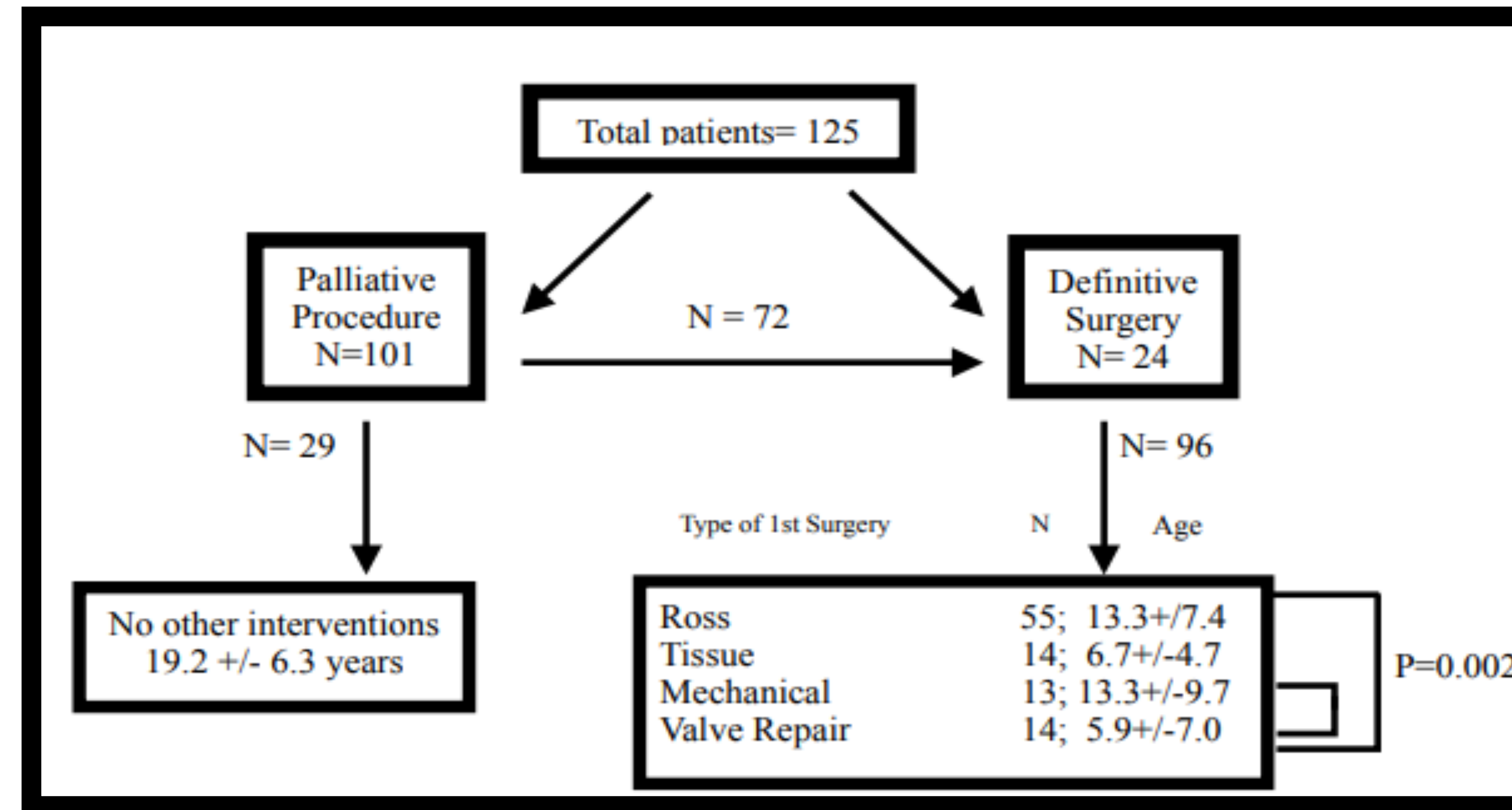
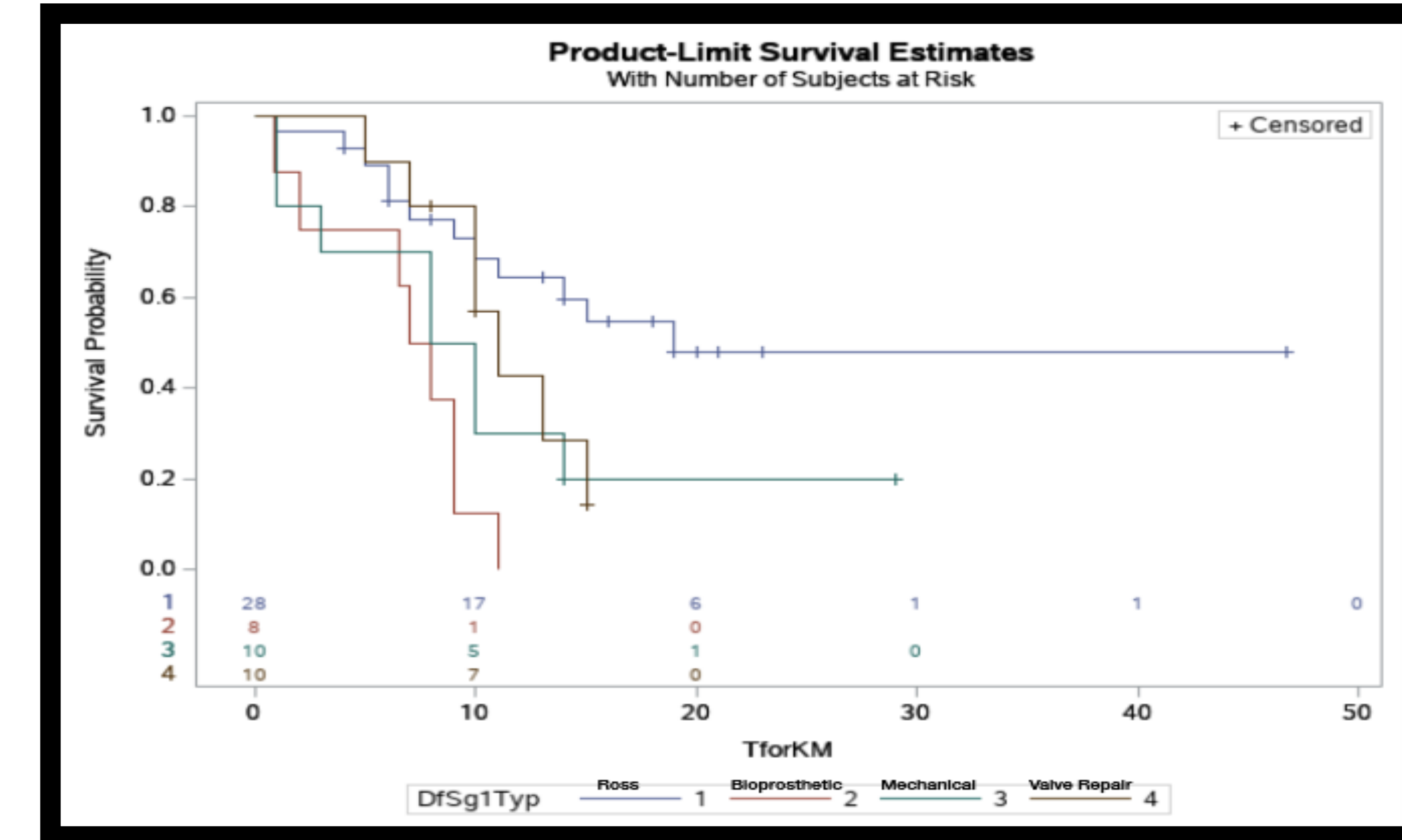
RESULTS

- At last follow-up patients were age 29.0 ± 8.0 with a follow-up duration of 23.4 ± 8.9 since first aortic valve intervention
- After 161 total valvuloplasties, patients underwent a total of 167 definitive aortic valve/ascending aortic procedures with a median number of lifetime aortic/aortic valve procedures of 3 (1-7).
- Mean longest surgery-free interval was 14 ± 8.7 years
- 7 patients developed SBE; Risk of endocarditis was higher in patients with a pacemaker (p=0.04) and those with more aortic valve surgeries (p=0.02) but did not relate to the type of aortic surgery; Ross 1.5 %, Tissue 8.3% Mechanical 7.0 % and valve repair 2.2% (p=0.11)

RESULTS

- Unplanned surgeries on the left ventricular outflow tract were required in 48/95 (51%) patients including 22/55 (40%) patients with initial Ross procedures, 12/14 (86%) with initial tissue valve*, 6/13 (46%) with initial mechanical valve and 8/14 (57%) with initial valve repair (p=0.02*)

Procedure Type	# of Procedures
Balloon Valvuloplasty	145
Surgical Valvotomy	35
Konno	15
Valve Repair	19
Mechanical Valve	28
Ross/covered Ross	60/5
Tissue Valve	24
OHT/VAD	3



Variable	Patient Characteristics N= 125
Age at 1st Valvuloplasty (months)	5 (0-188)
Age at 1st Repair/Replacement (years)	12.7 ± 7.0
Gender (M/F)	77/48
Patients with adverse outcome	15/125 patients (12%)
Genetic syndrome	10/125 patients (8%)
Coarctation	20/125 patients (16%)
Arrhythmia	37/125 patients (29.6%)
Pacemaker	12/125 patients (9.6%)
Cardiac Medications	22/125 patients (17.6%)
Cardiac Complications	32/125 patients (25.6%)

Independent Variables	P	Odds Ratio of Adverse Outcome	95% CL Odds Ratio
Konno	0.002	6.1	1.8-21.1
Number of surgeries	0.03	1.60	1.03-2.5
Pacemaker	<0.0001	19.9	5.2-77.2
Coarctation	0.2	2.4	0.7-8.5
Gender	0.2	2.5	0.7-9.5
Age at Definitive Surgery	0.2	0.95	0.9-1.03
Ross	0.03	0.3	0.08-0.97
Tissue Valve	0.8	1.2	0.24-6.08
Mechanical Valve	0.001	5.3	1.8-15.8
Valve Repair	0.5	0.5	0.06-3.8

CONCLUSIONS

- Repeat surgical procedures are very common in all surgical interventions for BAV with AS in childhood
- Greater #, but not type of, or age at, aortic valve surgery is associated with a higher incidence of surgical complications & SBE
- On long-term followup the odds of a composite adverse outcome were less with a Ross procedure