Case

- **15-year-old boy with bicuspid AV**
- **Severe AR with moderate AS**

Ross vs. AVR (or AVP)

AMC case

- 14-year-old boy with bicuspid AV
- Severe AS with mild AR
- Body size

  Bwt: 55 kg, Ht: 154 cm, BSA: 1.53 m^2
- Echocardiography

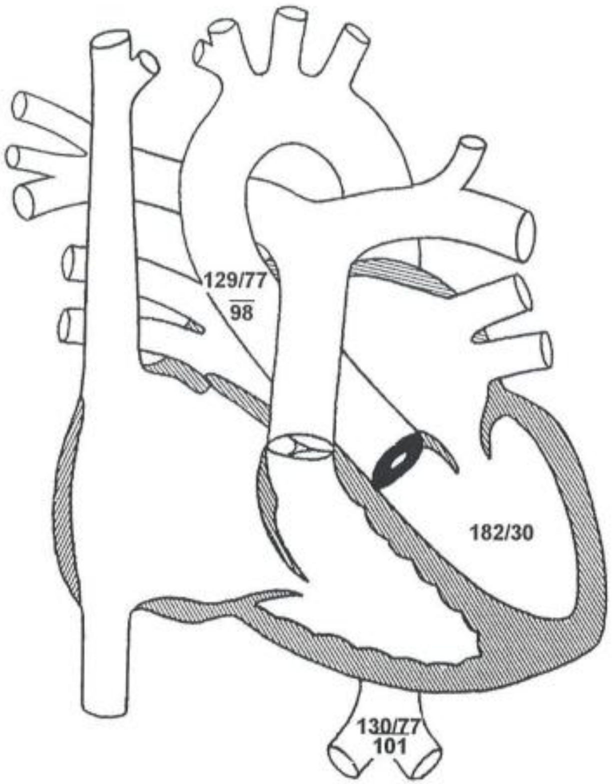
  AS: velocity 4 m/sec, Mild AR (II/IV)

  AV annulus: 20 mm, PV annulus: 21 mm

  Mild LVH

Asan Medical Center

Department of Pediatric Cardiology
Cardiac Catheterization Laboratory



Arrows indicate catheter course.

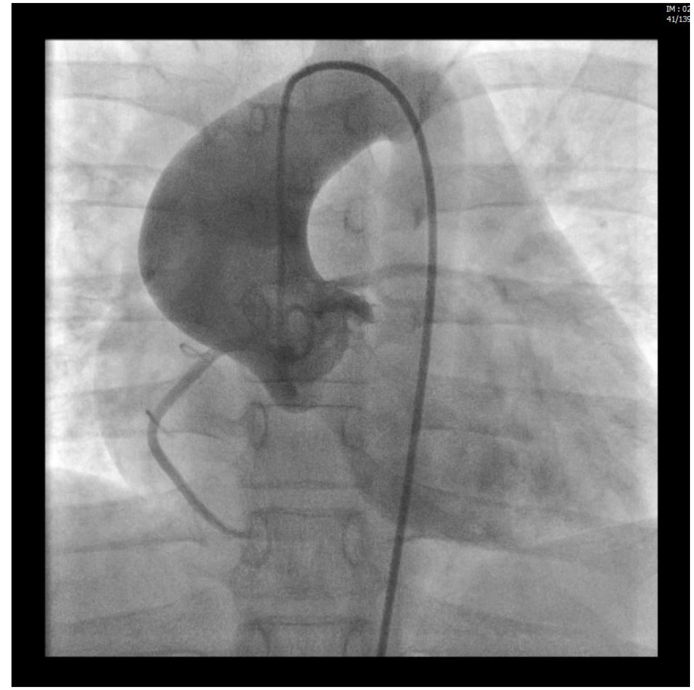
Diagnoses / Procedures

BAV, AS

Comments

Angiography
LV(AP/LAT)
aortic valve stenosis, doming(+)
post stenotic dilatation, 38mm
aortic regurgitation, moderate

Pressure
LV-Ao 185-125 = 60mmHg



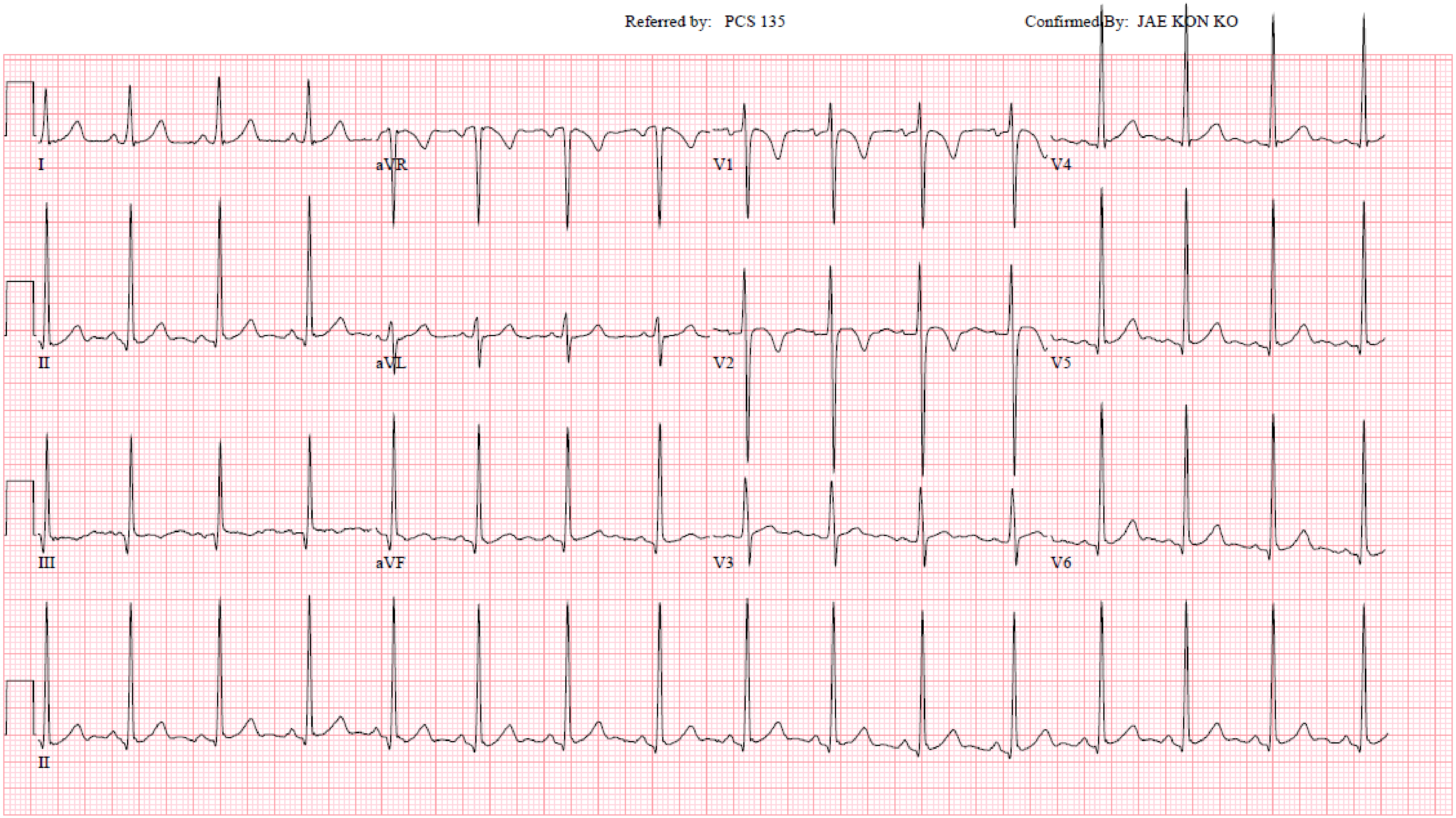
Preoperative EKG

ID:24924739 08-JUL-2013 15:05:26 AMC CARDIOVASCULAR CENTER

22-MAR-2000 (13 yr)	Vent. rate	92	BPM	** ** ** ** * Pediatric ECG Analysis * ** ** ** **
Male	PR interval	132	ms	Normal sinus rhythm
	QRS duration	90	ms	Left ventricular hypertrophy
Room:	QT/QTc	356/440	ms	Borderline Prolonged QT , may be secondary to QRS abnormality
Loc:127	P-R-T axes	34 66	33	Read by Sun-bong, Kim

Technician: C M N
Test ind:11491

Referred by: PCS 135 Confirmed By: JAE KON KO



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Surgical options

- **Aortic valve repair**
  1) **Definitive Treatment**
  2) **Bridge procedure for later AVR**

- **Aortic valve replacement**
  1) **Autograft PV (Ross procedure)**
  2) **Bioprosthetic valve or homograft**
  3) **Mechanical valve**

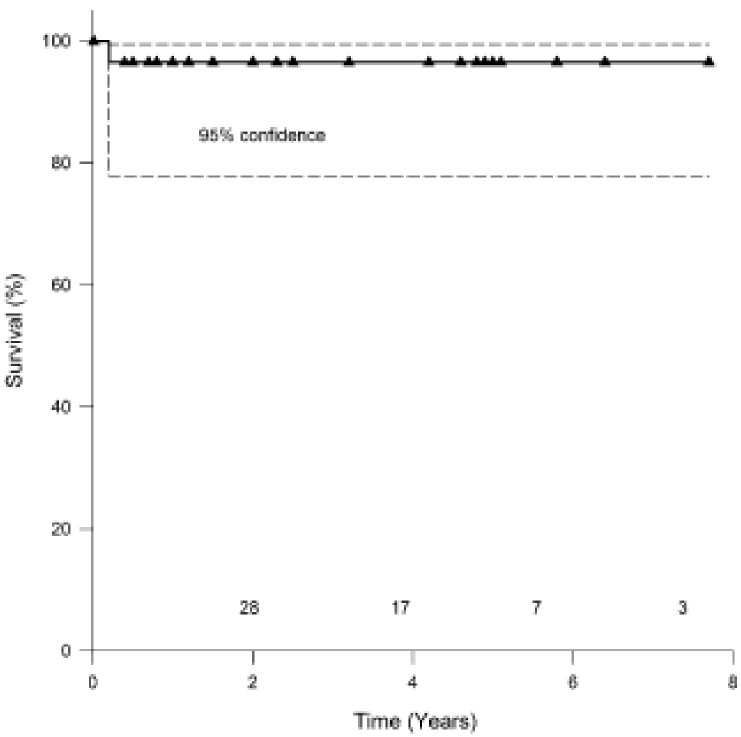
Pediatr Cardiol (2016) 37:84–89
DOI 10.1007/s00246-015-1243-0

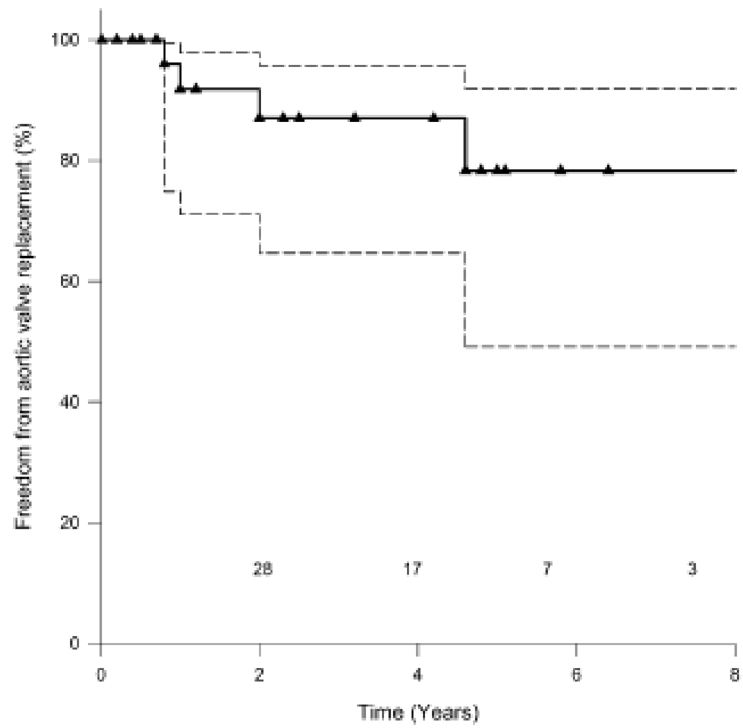
CrossMark

ORIGINAL ARTICLE

Progression of Aortic Regurgitation After Different Repair Techniques for Congenital Aortic Valve Stenosis

Fabian A. Kari[1] · Johannes Kroll[1] · Jan Kiss[1] · Carolin Hess[1] · Brigitte Stiller[2] · Matthias Siepe[1] · Friedhelm Beyersdorf[1]





Outcomes and Associated Risk Factors for Aortic Valve Replacement in 160 Children

A Competing-Risks Analysis

Tara Karamlou, MD; Karen Jang, MS; William G. Williams, MD; Christopher A. Caldarone, MD; Glen Van Arsdell, MD; John G. Coles, MD; Brian W. McCrindle, MD, MPH

Background**—**We sought to define patient characteristics, outcomes, and associated risk factors after aortic valve replacement (AVR) in children.

Methods and Results**—**Clinical records from children undergoing AVR from 1974 to 2004 at our institution were reviewed. Competing-risks methodology determined the time-related prevalence of 3 mutually exclusive end states: death, repeated replacement, and survival without subsequent AVR and their associated risk factors. Longitudinal echocardiographic data were analyzed by mixed linear-regression models. Children (n=160) underwent 198 AVRs, with 33 having >1. Competing-risks analysis predicted that 10 years from the initial AVR, 19% had died without subsequent AVR, 34% underwent a second AVR, and 47% remained alive without replacement. Risk factors for death without a second AVR included lower weight ($P<0.001$) and younger age at AVR ($P=0.04$), performance of aortic arch reconstruction together with AVR ($P=0.03$), and nonautograft use ($P=0.03$). Risk factors for a second AVR included earlier operation year ($P=0.04$) and implantation of a bioprosthetic or homograft valve ($P=0.004$). Analysis of serial echocardiographic measurements showed that pulmonary autograft use was associated with slower progression of peak aortic gradient ($P=0.002$), smaller left ventricular dimension ($P=0.04$), and decreased prevalence of aortic regurgitation ($P=0.04$).

Conclusions**—**Mortality and repeated valve replacement are common after initial AVR in children, especially in younger patients and those with bioprosthetic or homograft valves. Pulmonary autograft use is associated with decreased mortality, slower gradient progression, and smaller left ventricular dimension. (*Circulation*. **2005;112:3462-3469.**)

Key Words: heart defects, congenital ■ follow-up studies ■ pediatrics ■ aortic valve ■ valvuloplasty

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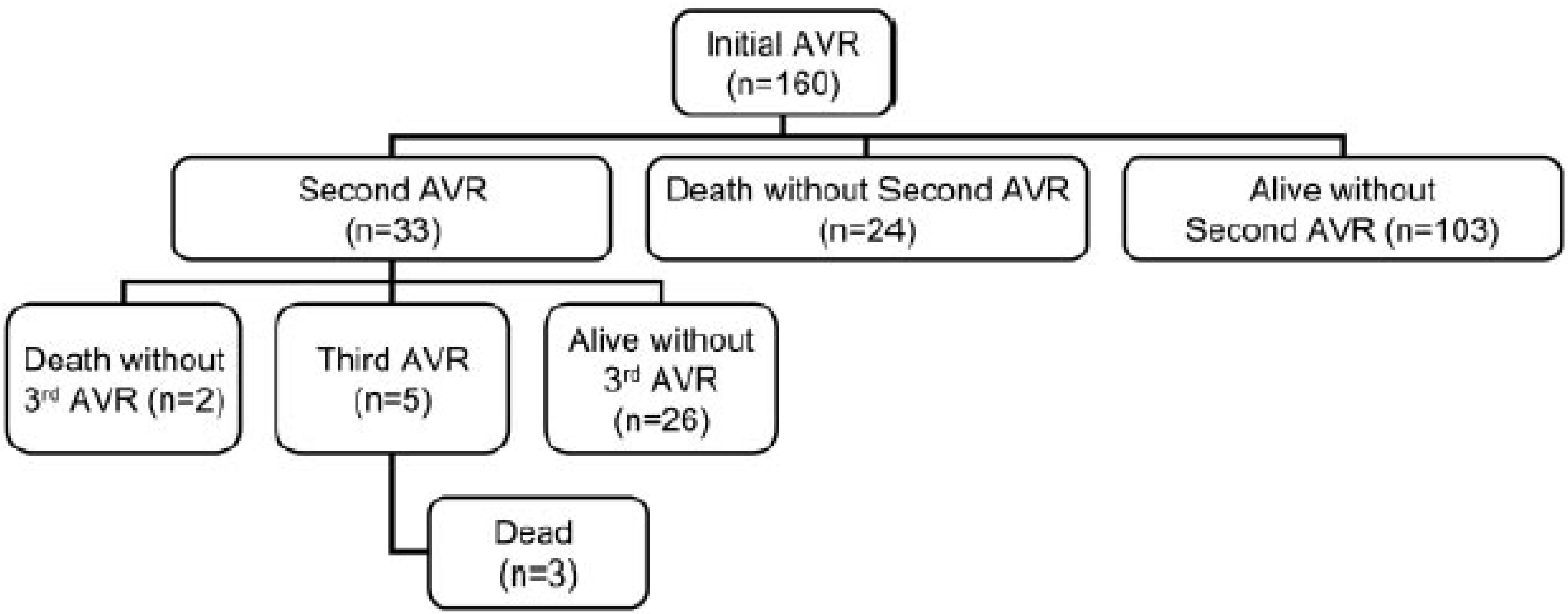

TABLE 1. Patient Characteristics (n=160)

Variable	Value	No. Missing
Demographic characteristics		
Sex, female:male, n:n	59:101	0
Noncardiac anomaly	20 (13)	0
Previous LVOT procedure	100 (63)	0
Open valvotomy	39 (24)	0
Open valvuloplasty	9 (6)	0
Balloon valvotomy	15 (9)	0
Ventricular septal defect (associated with aortic insufficiency) closure	7 (4)	0
Subaortic resection	6 (4)	0
Other	24 (15)	0
Morphological characteristics		
Etiology of aortic valve disease		
Congenital	133 (84)	1
Rheumatic	10 (6)	1
Ascending aortic aneurysm	8 (5)	1
Endocarditis	3 (2)	1
Predominant valve physiology		
Stenosis	33 (21)	3
Insufficiency	72 (46)	3
Combined stenosis+insufficiency	52 (33)	3
Major associated cardiac anomalies		
None: isolated aortic disease	77 (48)	1
Mitral valve anomaly	36 (23)	1
Ventricular septal defect	11 (7)	1
Coarctation of the aorta	7 (4)	1
Truncus arteriosus	6 (4)	1
Shone's syndrome	6 (4)	1
Atrial septal defect	4 (3)	1
Atrioventricular septal defect	3 (2)	1
Double-outlet right ventricle	1 (1)	1
Transposition of the great arteries	1 (1)	1

Values are n (%) unless otherwise noted.

TABLE 2. Procedural Characteristics at Initial and Subsequent AVRs Combined (n=198 Procedures in 160 Patients)

Variable	n	Missing	Value
Median age at AVR, y (range)	198	0	12 (0 to 28)
Median weight at AVR, kg (range)	186	12	40 (0 to 180)
Mean duration of cardiopulmonary bypass, min, ±SD	182	16	166±55
Mean duration of aortic cross clamping, min, ±SD	173	25	124±72
Type of AV replacement, n (%)	185	13	
Bjork-Shiley			45 (24)
St. Jude Medical			35 (19)
Pulmonary autograft			31 (17)
Allograft			30 (16)
Hancock			15 (8)
Ionescu-Shiley			10 (5)
Carbomedics			9 (5)
Carpentier-Edwards			5 (3)
Other mechanical			5 (3)
Mean size of AV implant, mm, ±SD	186	12	23±4
Bjork-Shiley			23±3
St. Jude Medical			23±4
Pulmonary autograft			20±6
Allograft			19±3
Hancock			24±4
Ionescu-Shiley			21±4
Carbomedics			22±4
Carpentier-Edwards			25±2
Other mechanical			23±3
Mean *Z*-score of AV prosthesis, ±SD	186	12	4±2

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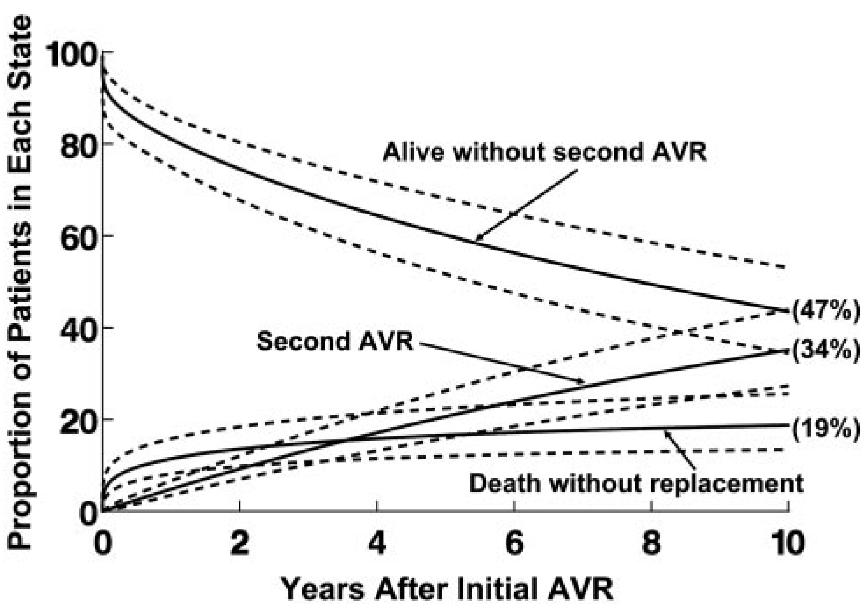

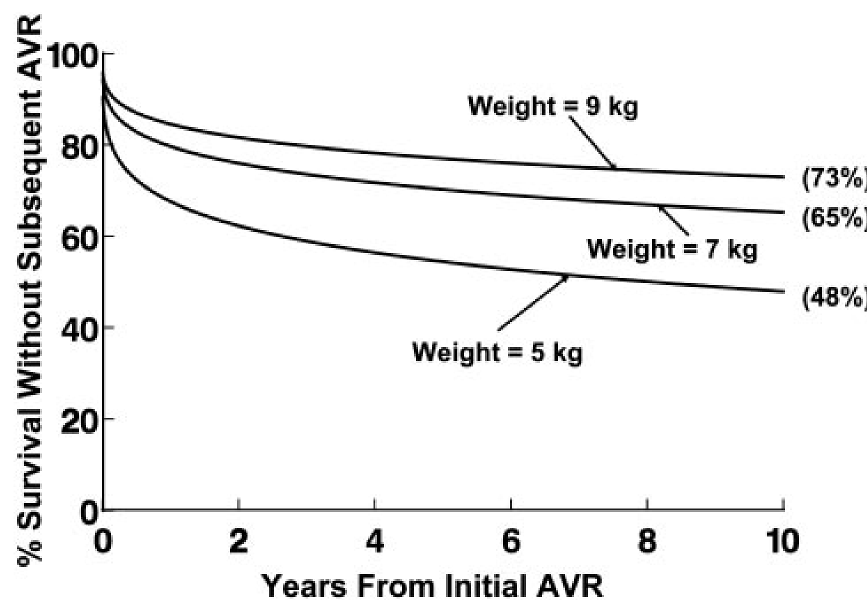

Pediatric Cardiology

Outcomes and Associated Risk Factors for Aortic Valve Replacement in 160 Children

A Competing-Risks Analysis

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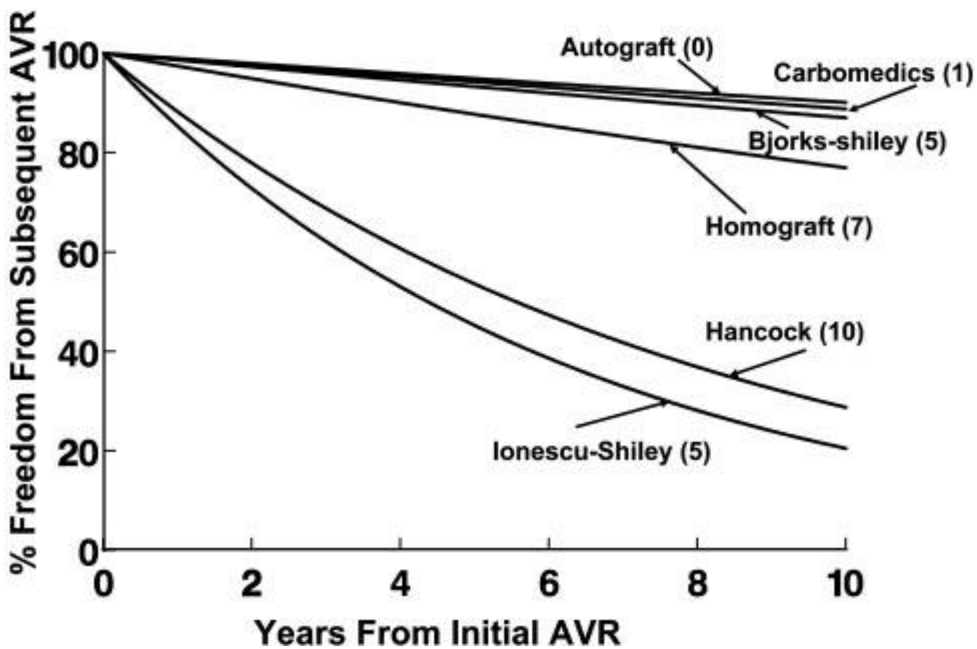

Figure 5. Freedom from reoperation after initial AVR stratified by prosthetic type. A multivariable equation was constructed for remaining alive after initial AVR without subsequent valve replacement according to the original competing risk model and forcing all valve types into the equation. The resulting model was then solved for a hypothetical 10-year-old patient of 40 kg undergoing operation in 1990. The autograft has superior longevity, whereas the tissue valves and allografts have considerably worse durability. The numbers in parentheses represent the total number of AVR episodes for each prosthesis type.

Mechanical valves versus the Ross procedure for aortic valve replacement in children: Propensity-adjusted comparison of long-term outcomes

Bahaaldin Alsoufi, MD,[a] Zohair Al-Halees, MD,[a] Cedric Manlhiot, BSc,[b] Brian W. McCrindle, MD, MPH,[b] Mamdouh Al-Ahmadi, MD,[a] Ahmed Sallehuddin, MD,[a] Charles C. Canver, MD,[a] Ziad Bulbul, MD,[a] Mansoor Joufan, MD,[a] and Bahaa Fadel, MD[a]

CHD

Objective: We aimed to identify characteristics differentiating children undergoing aortic valve replacement by using mechanical prostheses versus the Ross procedure and to compare survival and the need for aortic valve reoperation after each procedure.

Methods: From 1983 to 2004, 346 children underwent aortic valve replacement (215 underwent the Ross procedure and 131 underwent placement of a mechanical prosthesis). Factors associated with procedure choice were used to construct a propensity score for use as a covariate in regression models to adjust for potential confounding by indication.

Results: Patients undergoing the Ross procedure were younger, more likely to have a congenital cause, and less likely to have a rheumatic or connective tissue cause. They had a lower frequency of regurgitation, required more annular enlargement, and had less concomitant cardiac surgery. Competing-risk analysis showed that 16 years after aortic valve replacement, 20% of patients had died without subsequent aortic valve replacement, 25% underwent second aortic valve replacement, and 55% remained alive without further replacement. After propensity adjustment, factors associated with early-phase death included mechanical valves and a nonrheumatic cause. Mechanical valves were also associated with constant-phase mortality. Repeated aortic valve replacement was associated with the Ross procedure and a rheumatic cause. Both factors were also associated with all-cause cardiac reoperation. In children receiving mechanical prostheses, younger age and smaller valve size were significant risk factors for death. Freedom from homograft replacement after the Ross procedure was 82% at 16 years of follow-up.

Conclusion: Results from this study showed good outcomes and an acceptable complication rate with both valve choices. Given the significantly increased risk of early and late death in younger children receiving smaller mechanical valves, the Ross procedure confers survival advantage in this age group at the expense of increased reoperation risk, especially in patients with a rheumatic cause.

The Journal of Thoracic and Cardiovascular Surgery c February 2009

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TABLE 1. Patients' characteristics

	Cohort (n = 346)	Mechanical prosthesis (n = 131 [38%])	Ross procedure (n = 215 [62%])	*P* value
Sex (male)	260 (75%)	109 (83%)	151 (70%)	.007
Mean age at surgical intervention (y)	12.4 ± 4.4	14.0 ± 3.8	11.4 ± 4.6	<.0001
Median year at presentation	1998 (1983–2005)	1996 (1983–2005)	1999 (1991-2004)	<.0001
Cause				
Rheumatic	201 (58%)	97 (74%)	104 (48%)	<.0001
Congenital	116 (34%)	15 (11%)	101 (47%)	<.0001
Connective tissue	14 (4%)	12 (9%)	2 (1%)	<.0001
Endocarditis	15 (4%)	7 (5%)	8 (4%)	.59
Hemodynamic manifestation				
Stenosis	44 (13%)	8 (6%)	36 (17%)	.005
Regurgitation	224 (65%)	115 (88%)	109 (49%)	<.0001
Mixed	78 (22%)	8 (6%)	70 (33%)	<.0001
Previous percutaneous intervention	20 (6%)	0 (0%)	20 (9%)	<.0001
Previous cardiac operation	97 (28%)	41 (31%)	56 (26%)	.33
One previous operation	89 (26%)	36 (27%)	53 (25%)	
Two previous operations	8 (2%)	5 (4%)	3 (1%)	
Concomitant cardiac operation	147 (42%)	97 (74%)	50 (23%)	<.0001
Arch repair	5 (1%)	4 (3%)	1 (<1%)	.07
Left ventricular outflow tract enlargement	35 (16%)	3 (2%)	32 (15%)	<0.0001

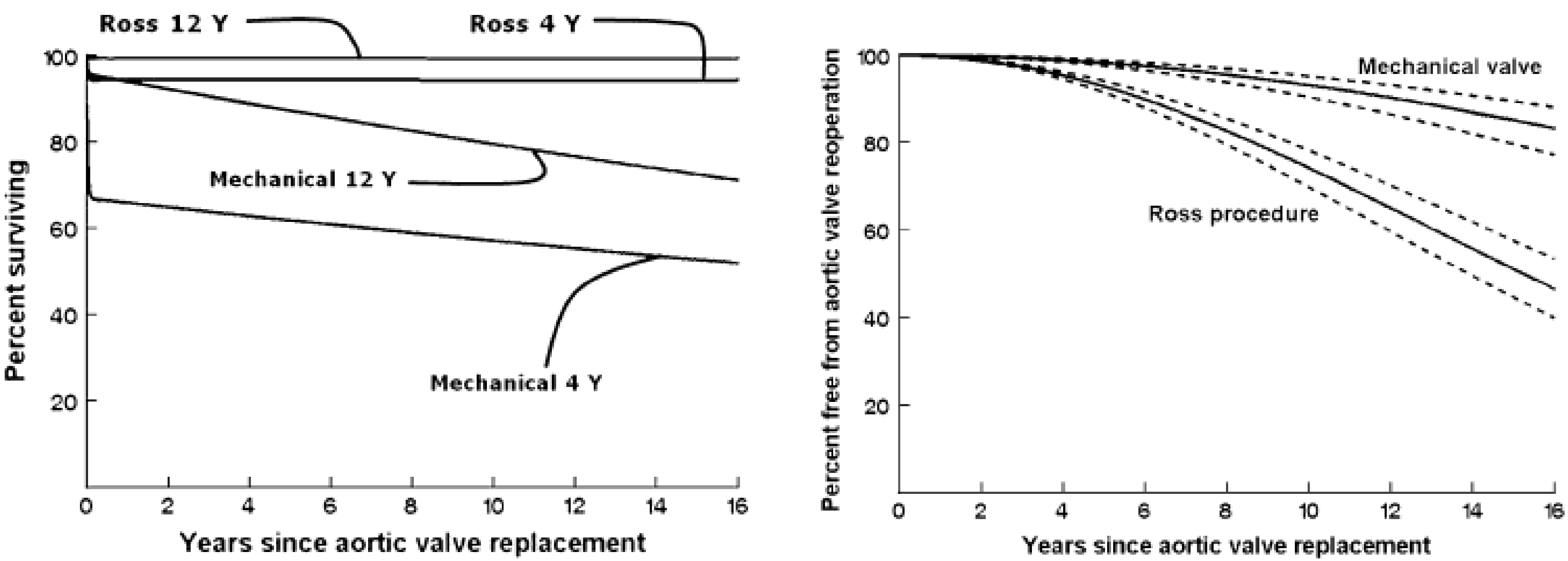
Mechanical valves versus the Ross procedure for aortic valve replacement in children: Propensity-adjusted comparison of long-term outcomes

TABLE 2. Incremental risk factors for time-related transition from initial AVR to either death or a second AVR

	Unadjusted		Propensity adjusted	
	Estimate ± SE	*P* value	Estimate ± SE	*P* value
Death without subsequent AVR: Early phase				
Valve type (mechanical vs Ross)	2.05 ± 0.57	.0004	2.71 ± 0.70	.0001
Cause other than rheumatic	1.75 ± 0.77	.03	2.10 ± 0.69	.003
Younger age at surgical intervention	0.12 ± 0.05	.03		
Death without subsequent AVR: Constant phase				
Valve type (mechanical vs Ross)	1.60 ± 0.58	.007	1.72 ± 0.65	.009
Concomitant operations	2.74 ± 1.01	.007		
Survival to a subsequent AVR: Late phase				
Valve type (Ross vs mechanical)	2.72 ± 0.48	<.0001	2.47 ± 0.47	<.0001
Rheumatic cause	1.92 ± 0.62	.002	2.09 ± 0.61	.0006
Concomitant cardiac surgery	0.91 ± 0.38	.02		

AVR, Aortic valve replacement; *SE*, standard error.

Mechanical valves versus the Ross procedure for aortic valve replacement in children: Propensity-adjusted comparison of long-term outcomes



Mechanical valves versus the Ross procedure for aortic valve replacement in children: Propensity-adjusted comparison of long-term outcomes

Operative Details

Detailed steps of the surgical intervention will not be discussed in this section; however, some important information will be briefly listed.4 Midline sternotomy was performed, and standard cardiopulmonary bypass and myocardial protection techniques were used in all cases. For AVR with a mechanical prosthesis, the valve was secured to the annulus with multiple mattressed sutures, with the pledgets placed at the ventricular aspect of the annulus. For the Ross procedure, the pulmonary autograft was implanted as a full root, with coronary transfer in all cases. The autograft muscle cuff was trimmed with sutures placed almost directly at the autograft annulus. The proximal suture line was performed with running polypropylene sutures. The neoaortic annulus was not reinforced in these patients, so as not to limit growth. In patients with left ventricular outflow tract (LVOT) obstruction, a modified Ross–Konno technique was used. Part of the septum was cored out to completely open up the LVOT without creating a large ventricular septal defect. In those patients the fibrous annulus of the aortic valve was often divided, and the cut was partially taken down to the septum. In our current practice, in patients with a dilated aortic annulus of greater than 29 to 30 mm or with an aortic annulus 2 to 3 mm larger than the pulmonary valve annulus, the Ross procedure is not considered, and we rarely use any aortic annulus reduction techniques. The distal suture line was occasionally reinforced with Teflon felt in patients with a dilated ascending aorta. The immediate postoperative results were assessed in all patients in the operating room by means of transesophageal echocardiographic analysis.

AMC case

- **14-year-old boy with bicuspid AV**
- **Severe AS with mild AR**
- **Body size**

Bwt: 55 kg, Ht: 154 cm, BSA: 1.53 m^2

- **Echocardiography**

AS: velocity 4 m/sec, Mild AR (II/IV)

AV annulus: 20 mm, PV annulus: 21 mm

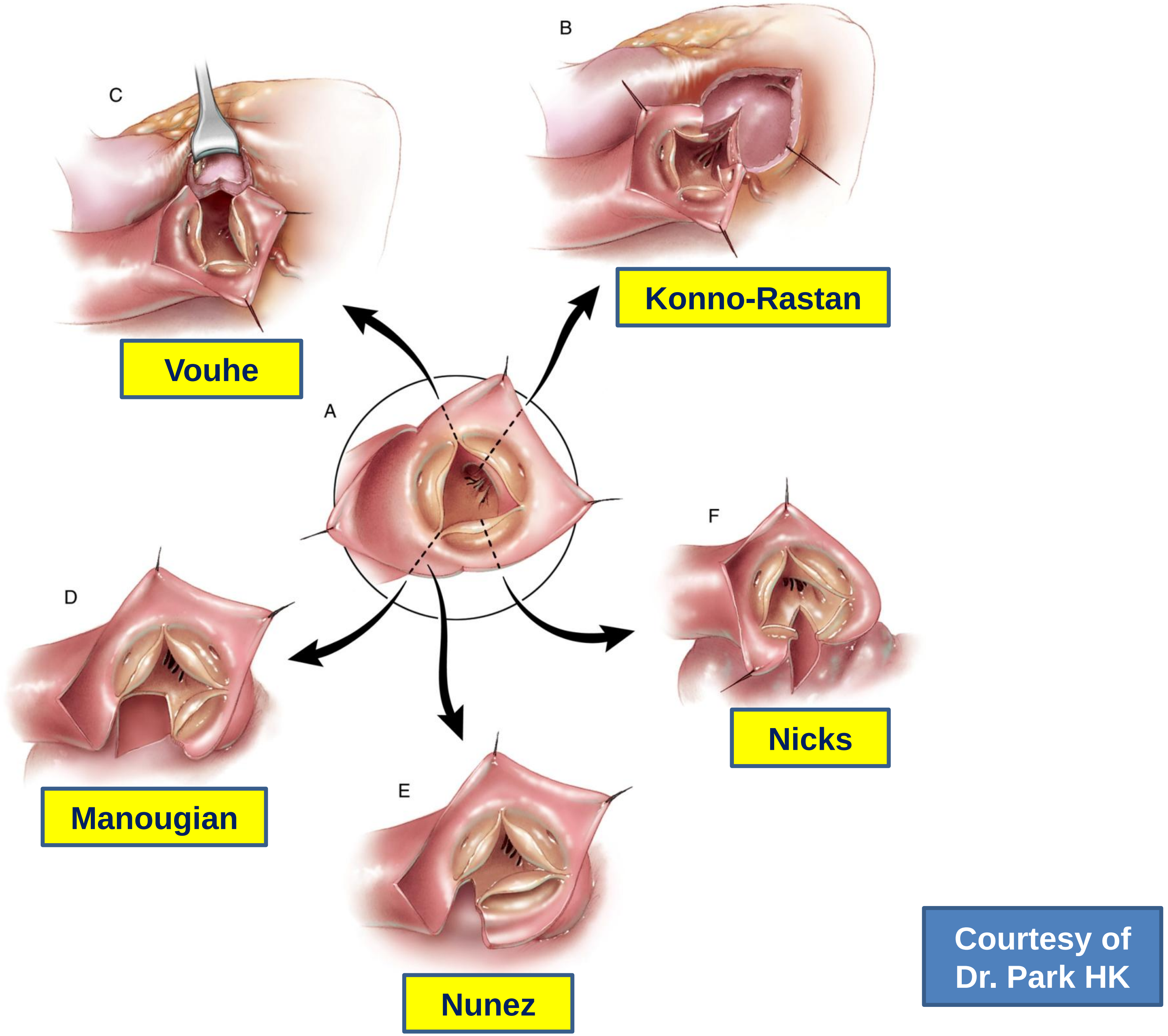
Mild LVH

Operation

- AVR with AV annulus enlargement

1) AV annular enlargement using bovine pericardium (Manougian)

2) AVR with Saint-Jude supra-annular Regent valve (21 mm)


B
C
Konno-Rastan
Vouhe
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Manougian
Nunez
Courtesy of
Dr. Park HK

Postoperative EKG

ID:24924739 26-JAN-2016 13:59:21 AMC CARDIOVASCULAR CENTER

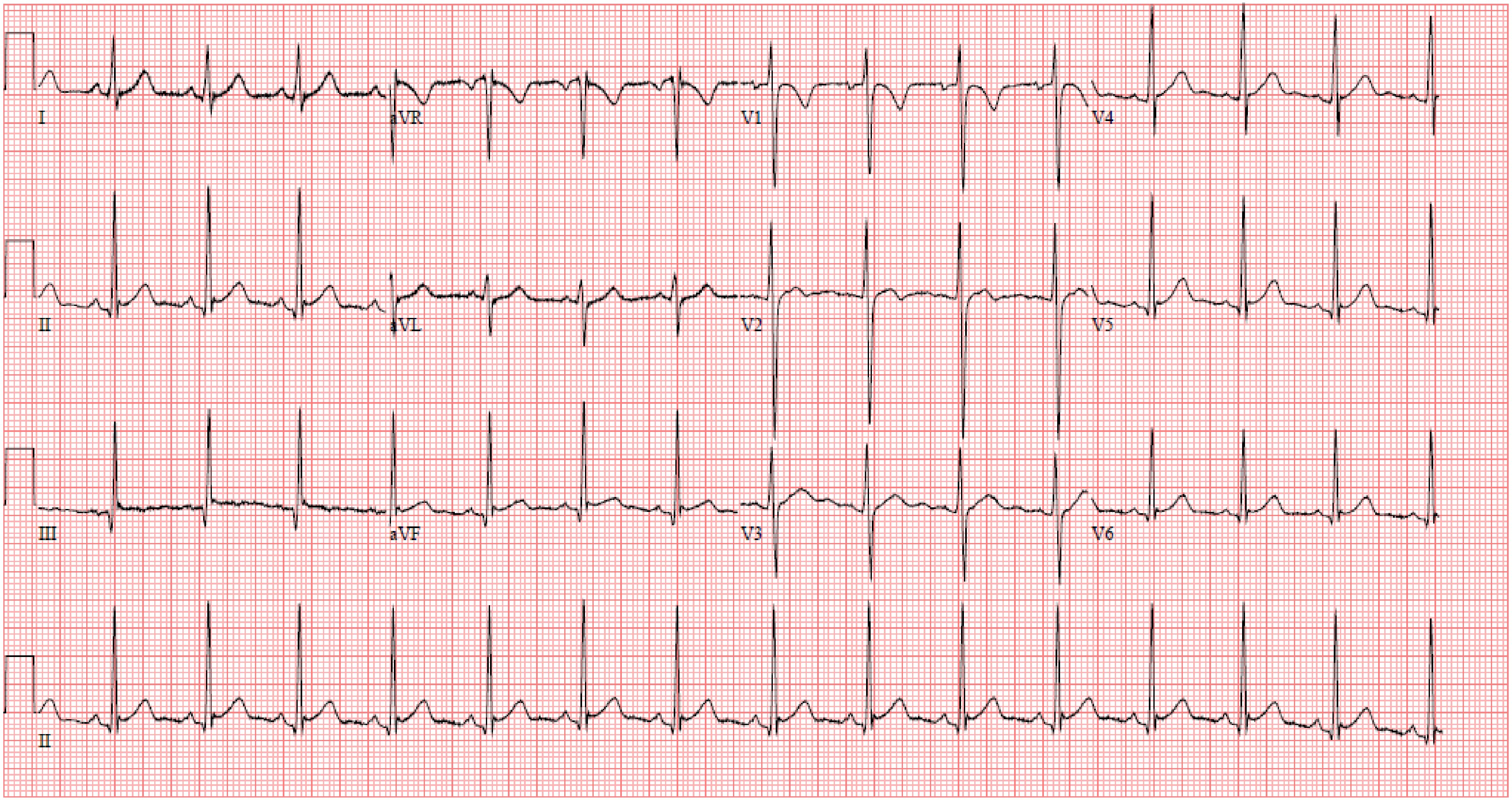
22-MAR-2000 (15 yr)	Vent. rate	90	BPM
Male	PR interval	140	ms
	QRS duration	90	ms
Room:	QT/QTc	360/440	ms
Loc:127	P-R-T axes	36 70	38

** ** ** ** * Pediatric ECG Analysis * ** ** ** **
Normal sinus rhythm
Borderline Prolonged QT

Technician: K M K
Test ind:1889

Referred by: PCD PC

Confirmed By: YOUNG HWEU KIM



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Echocardiographic findings

	Preoperative	Postoperative (18m)
LVOT velocity	4 m/sec	2.1 m/sec
AR	II / IV	-
MR	trace	trace
TR	trace	trace
LVIDd	48 mm	45 mm
LVIDs	30 mm	20 mm
LVEF	68 %	85 %
LVDEV index	74 ml/m^2	56 ml/m^2
LV mass index	85 g/m^2	73 g/m^2

Case

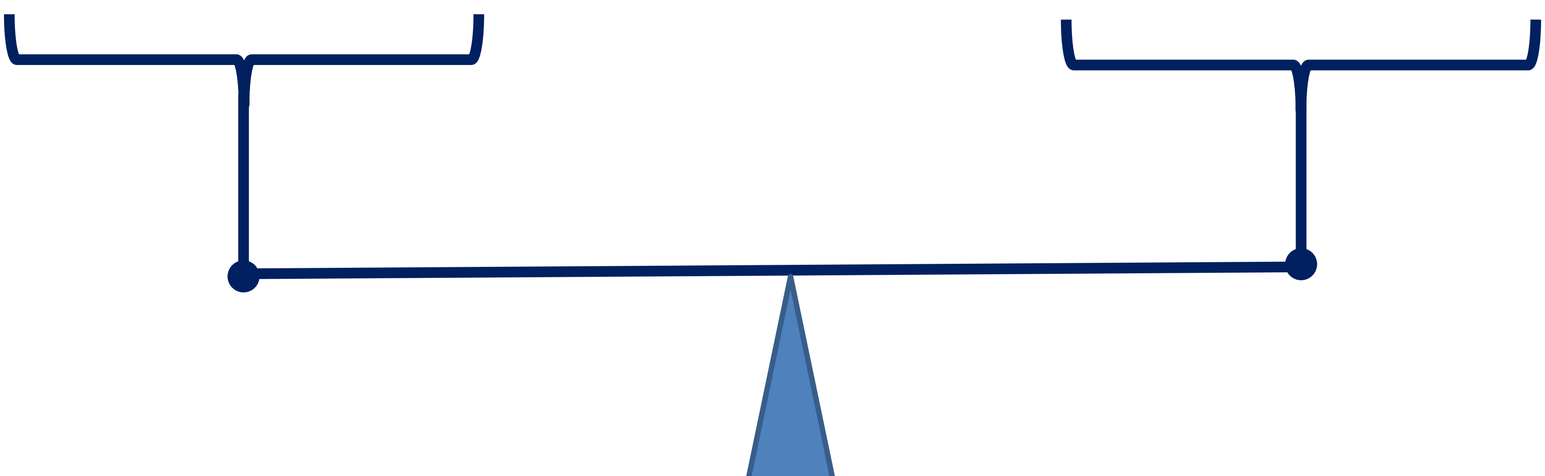
- **15-year-old boy with bicuspid AV**
- **Severe AR with moderate AS**

AVR with mechanical valve

Pitfalls of each option

Mechanical AVR

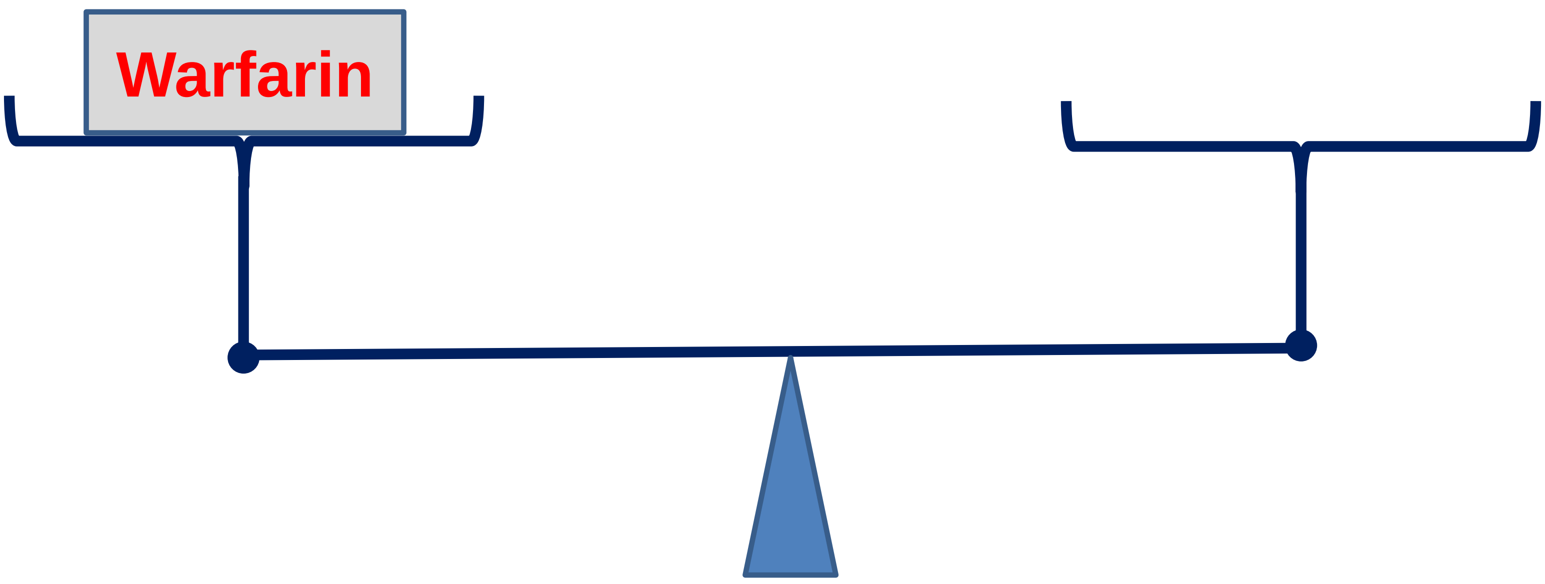
Ross procedure



Pitfalls of each option

Mechanical AVR

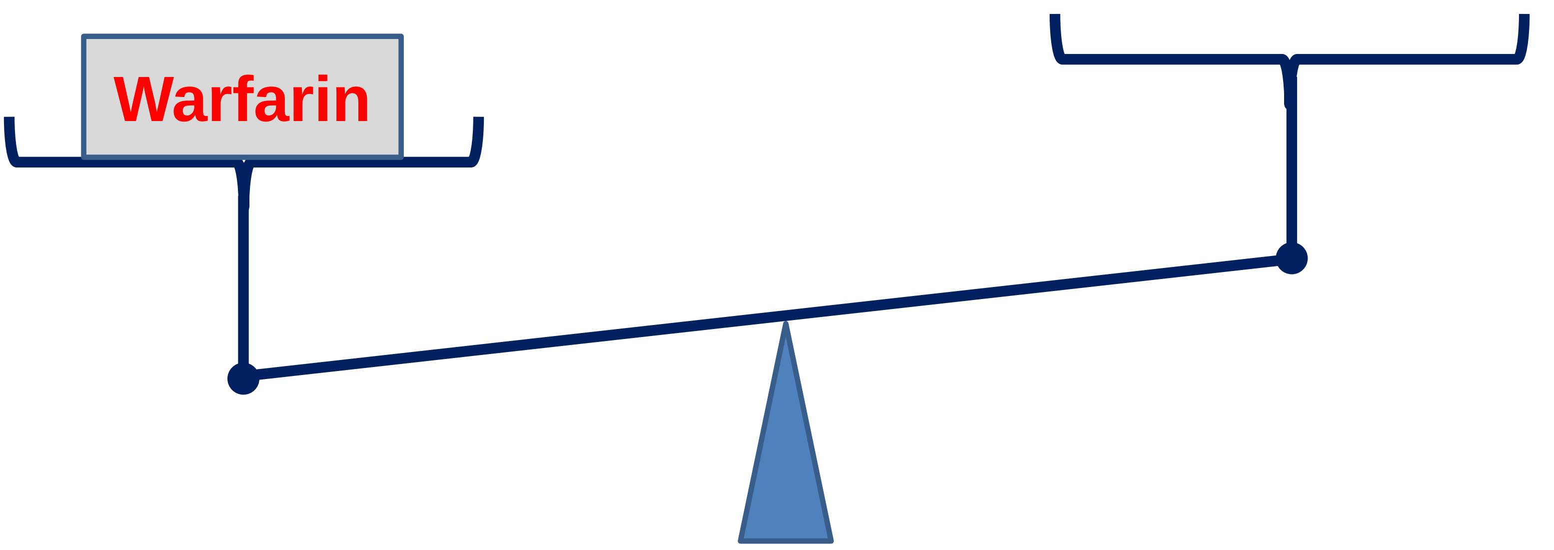
Ross procedure



Pitfalls of each option

Mechanical AVR

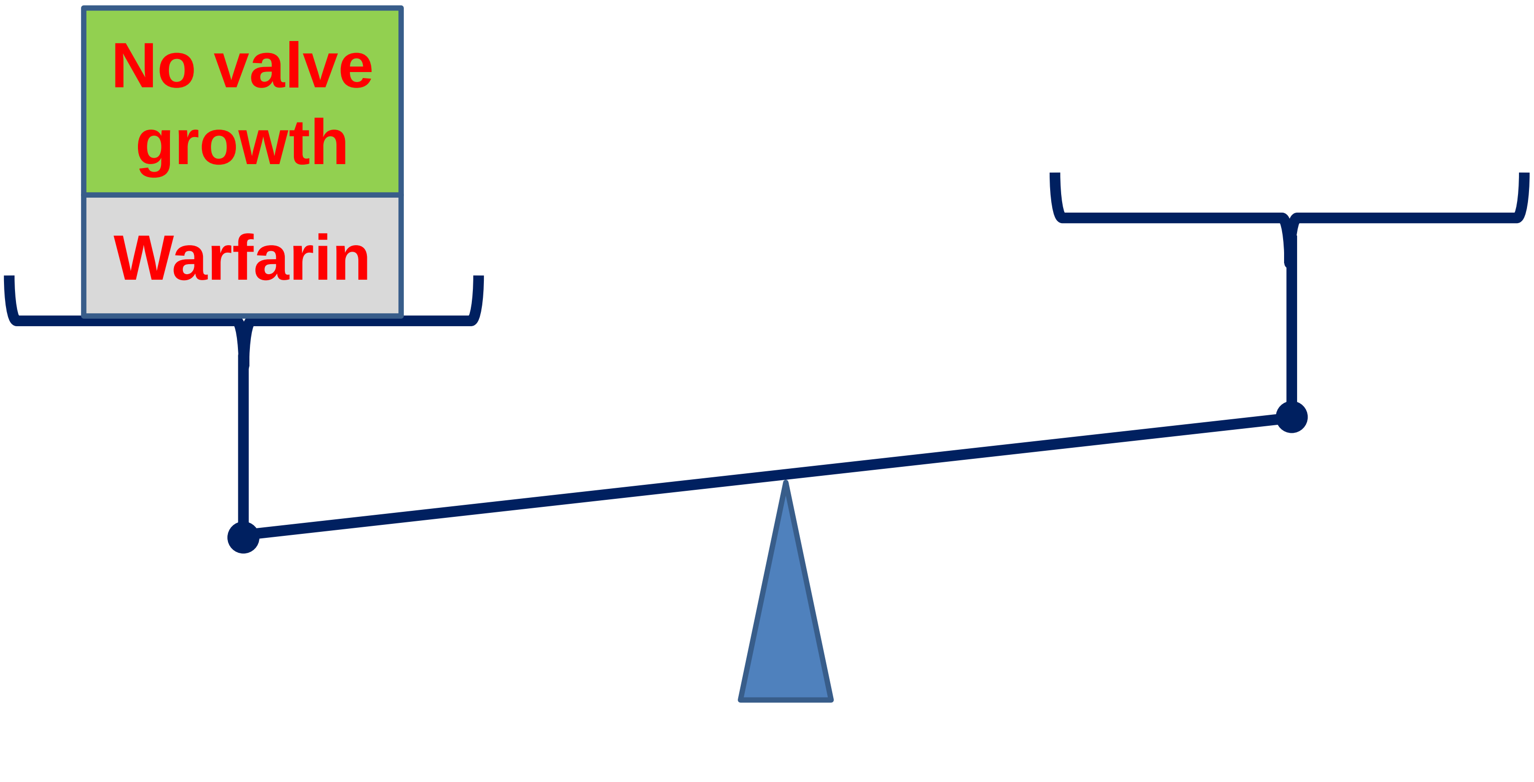
Ross procedure



Pitfalls of each option

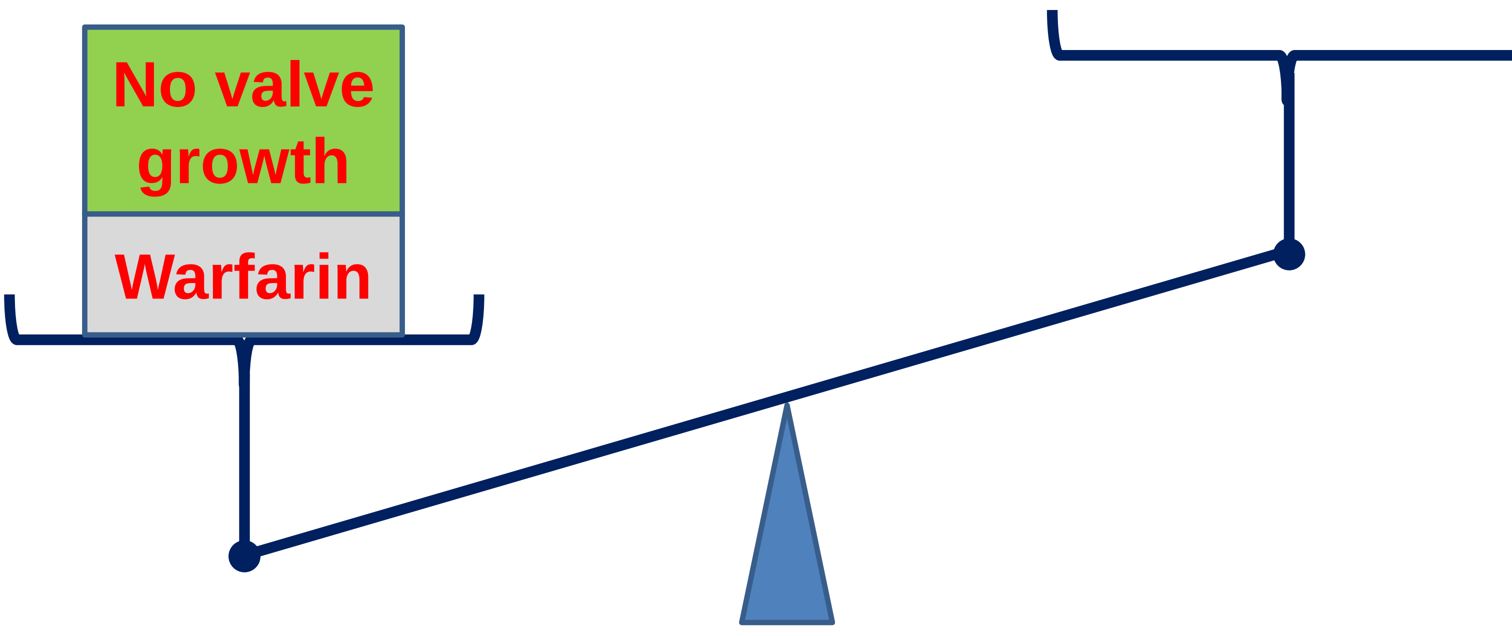
Mechanical AVR

Ross procedure

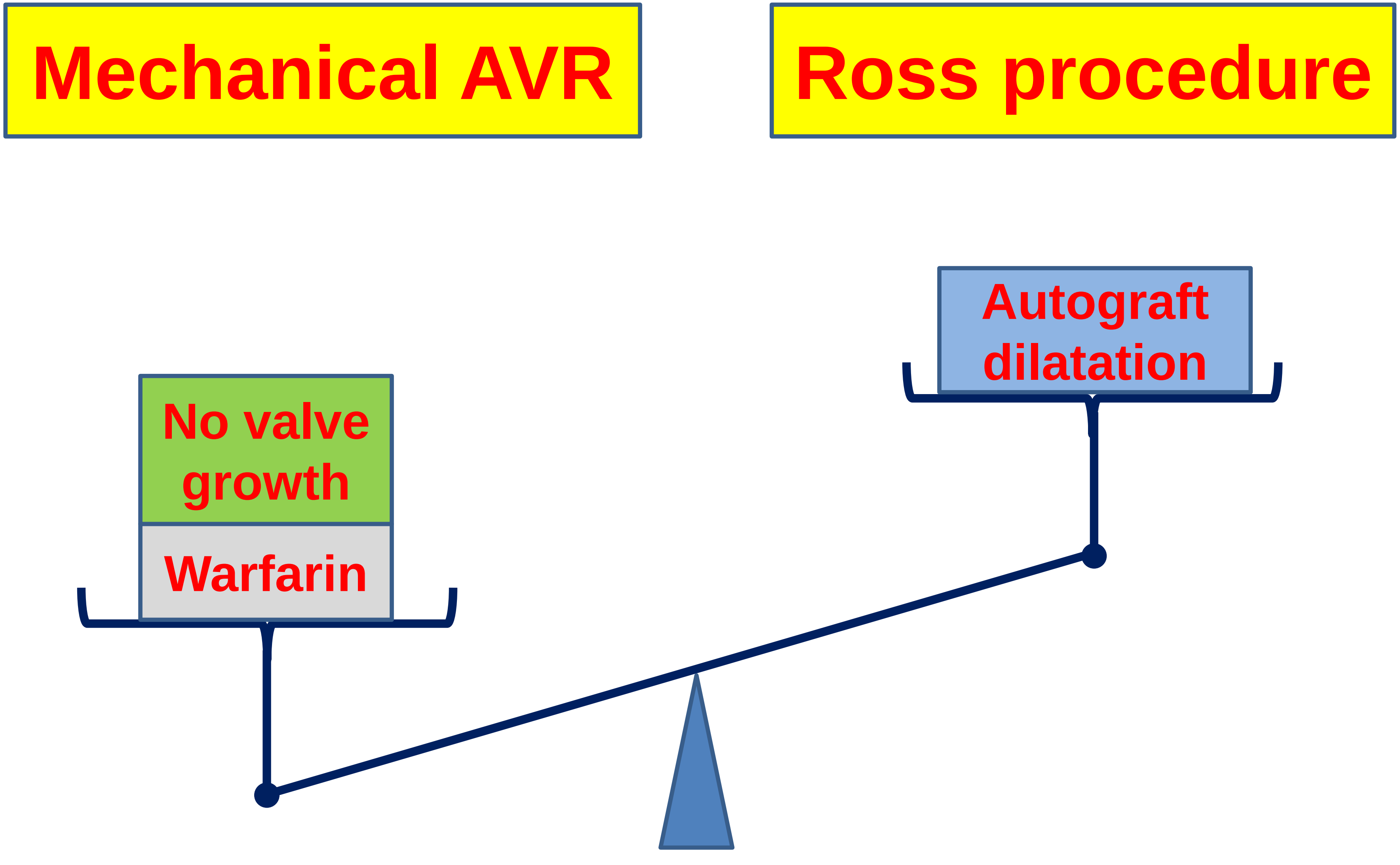


Pitfalls of each option

Mechanical AVR | **Ross procedure**

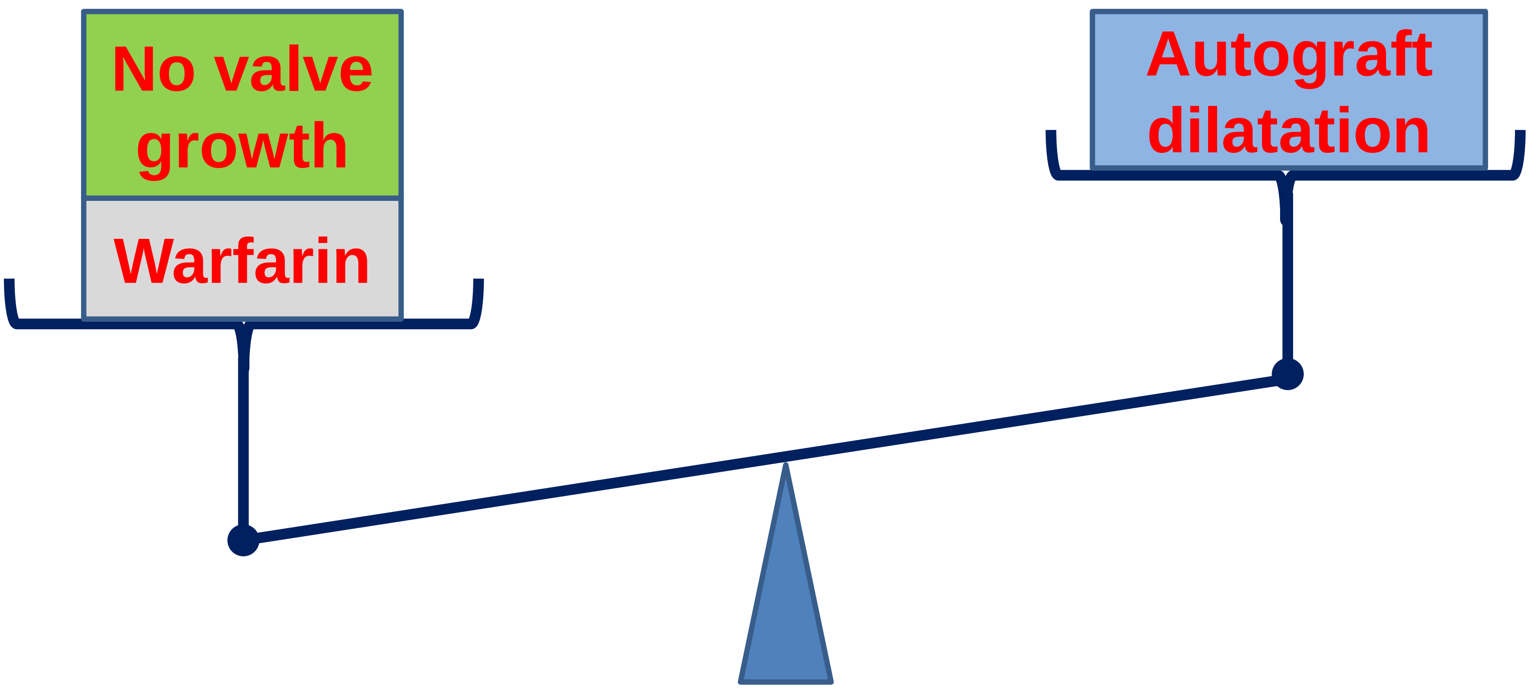


Pitfalls of each option



Pitfalls of each option

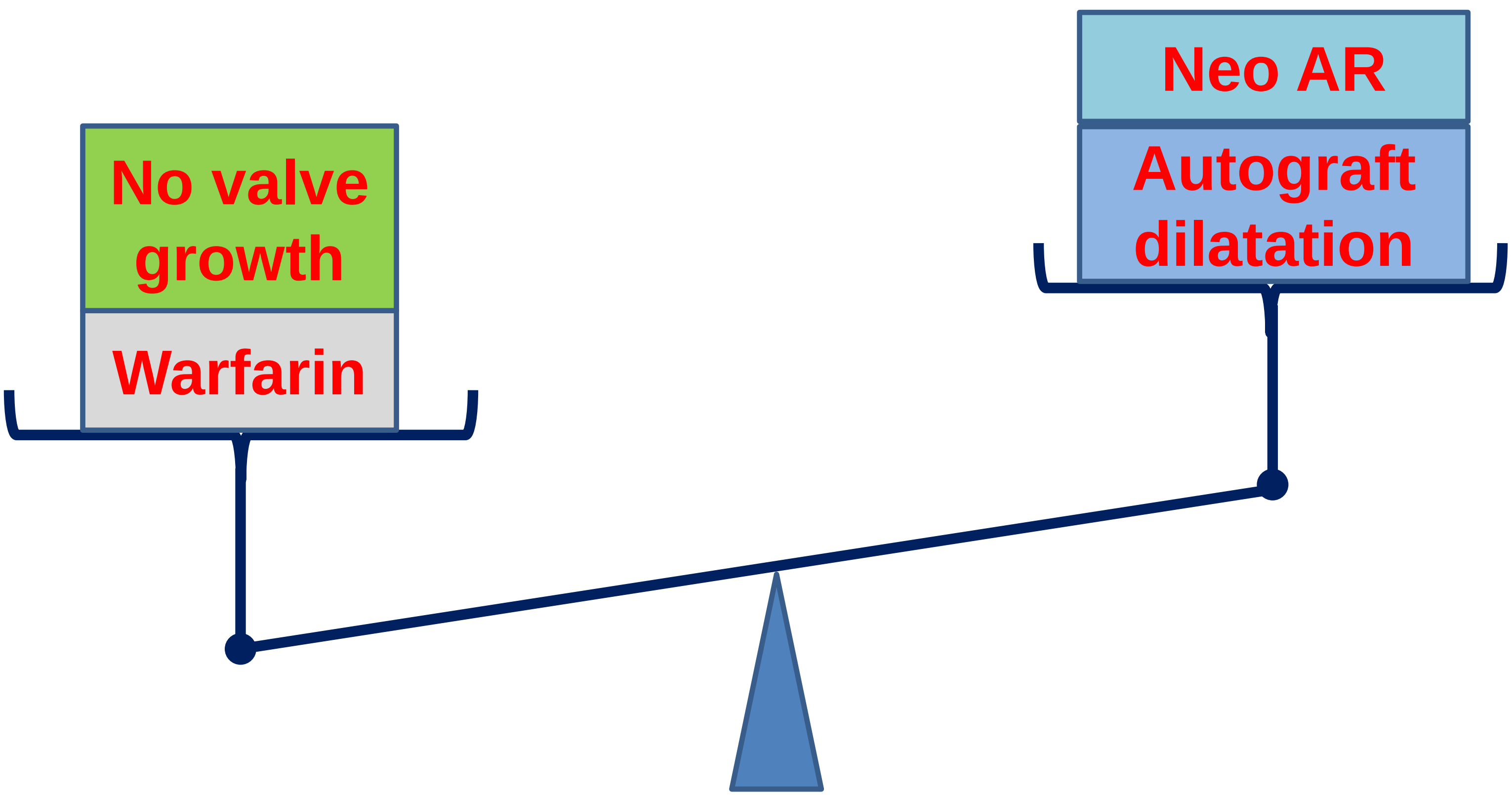
Mechanical AVR | **Ross procedure**



Pitfalls of each option

Mechanical AVR

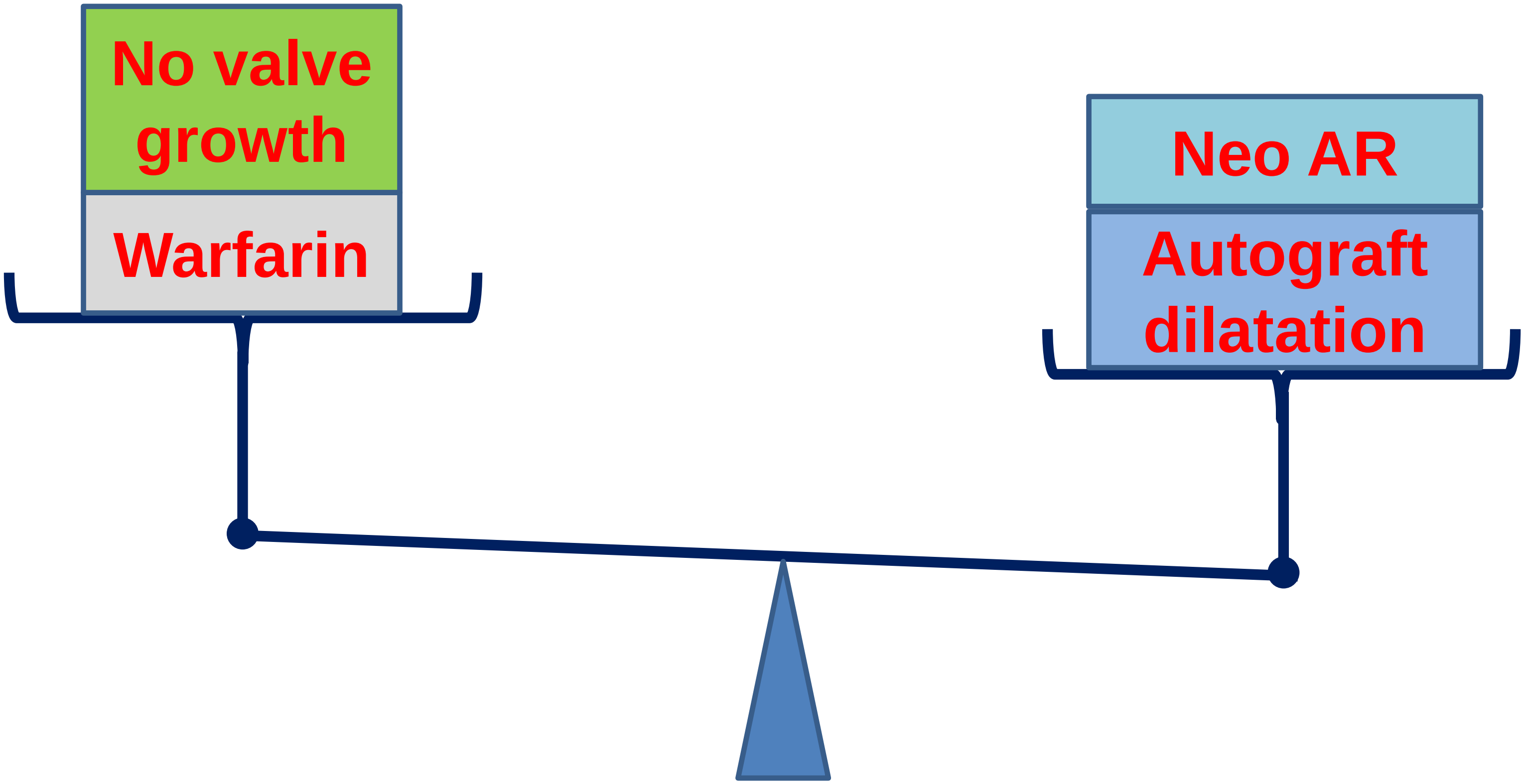
Ross procedure



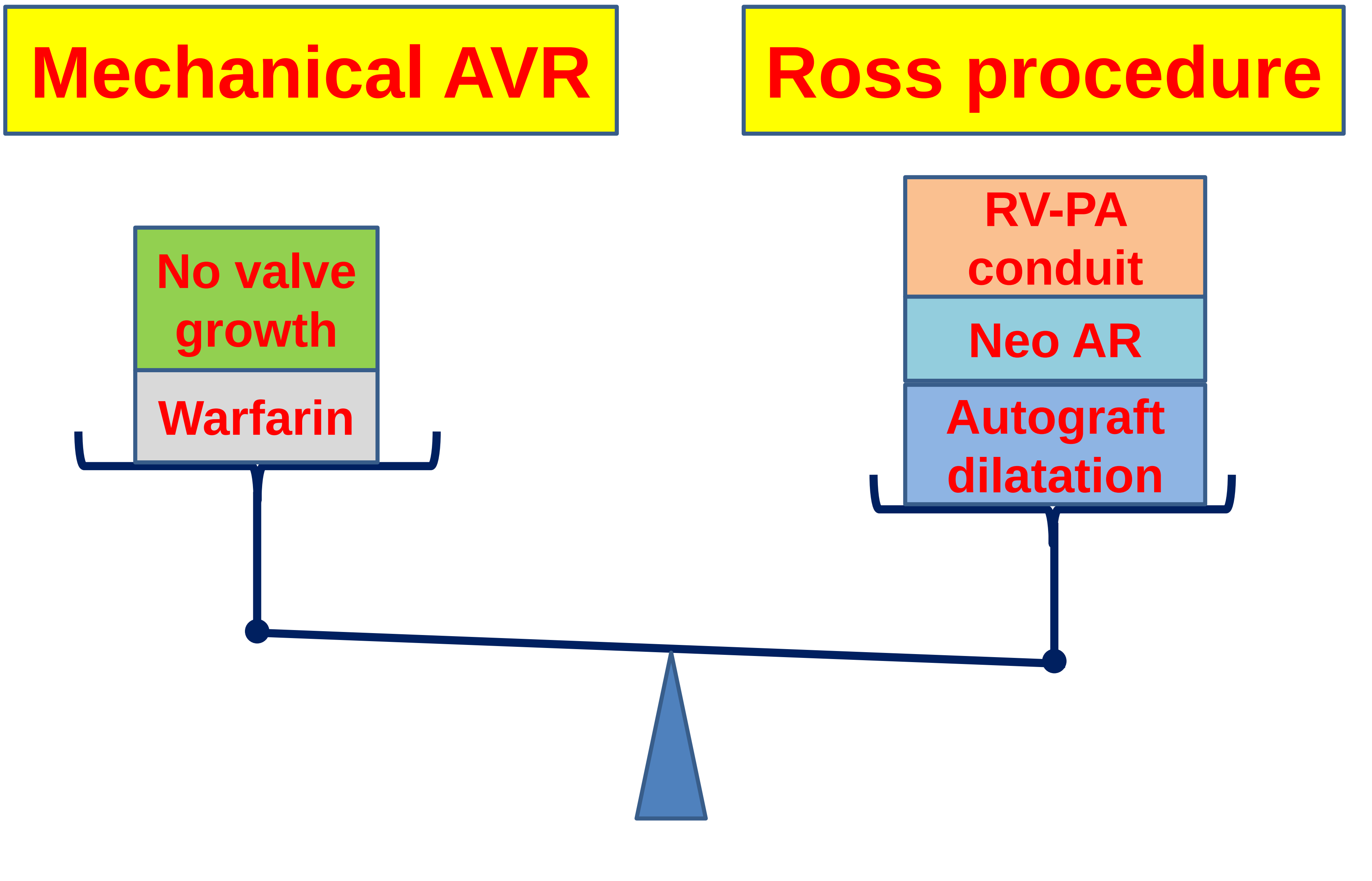
Pitfalls of each option

Mechanical AVR

Ross procedure



Pitfalls of each option



Pitfalls of each option