Late Complications of AV Canal

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Surgical Director, Philadelphia Adult Congenital Heart Program

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Perelman School of Medicine University of Pennsylvania

Disclosures

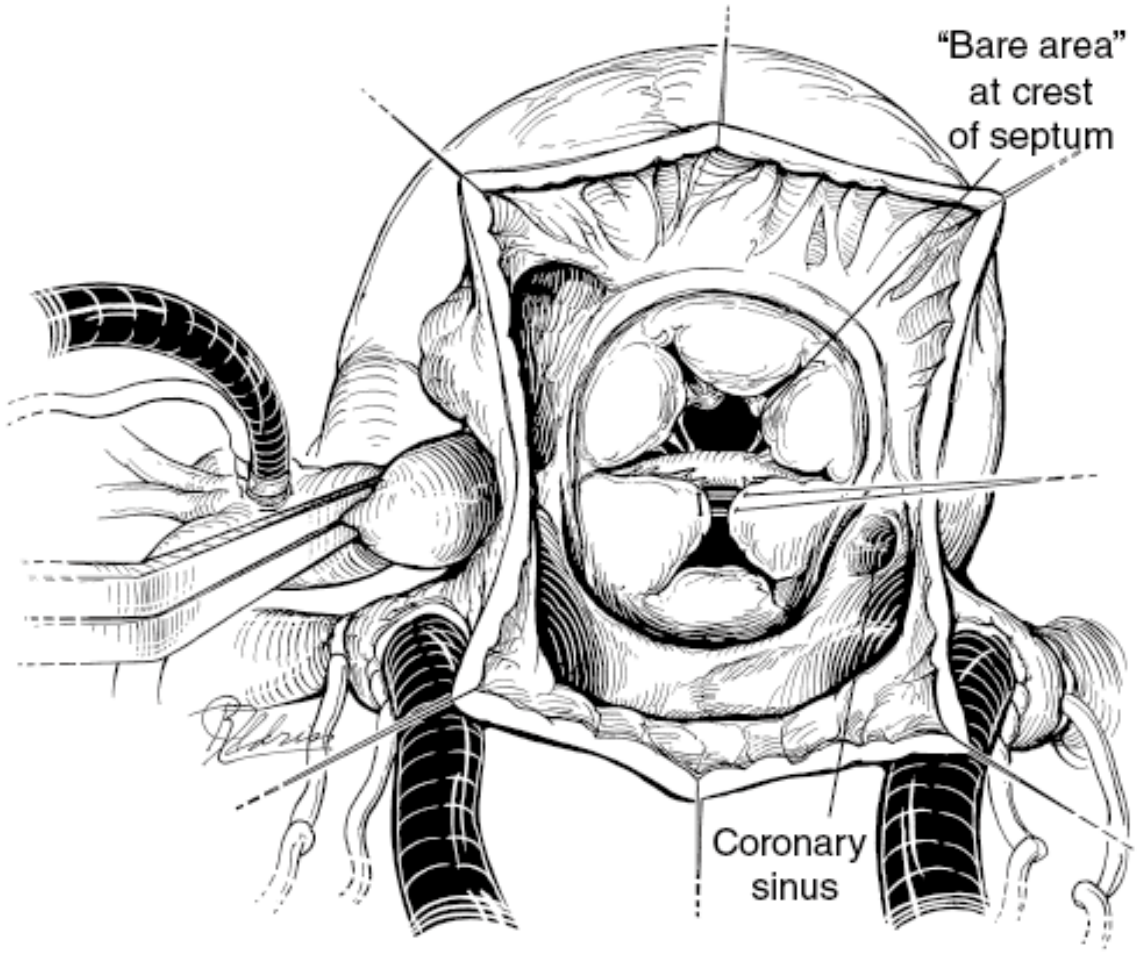
- **None**

Background

- Complex operation
  - Multiple techniques
  - Variable patient population
- Spectrum of presentation
- May not correlate to symptoms

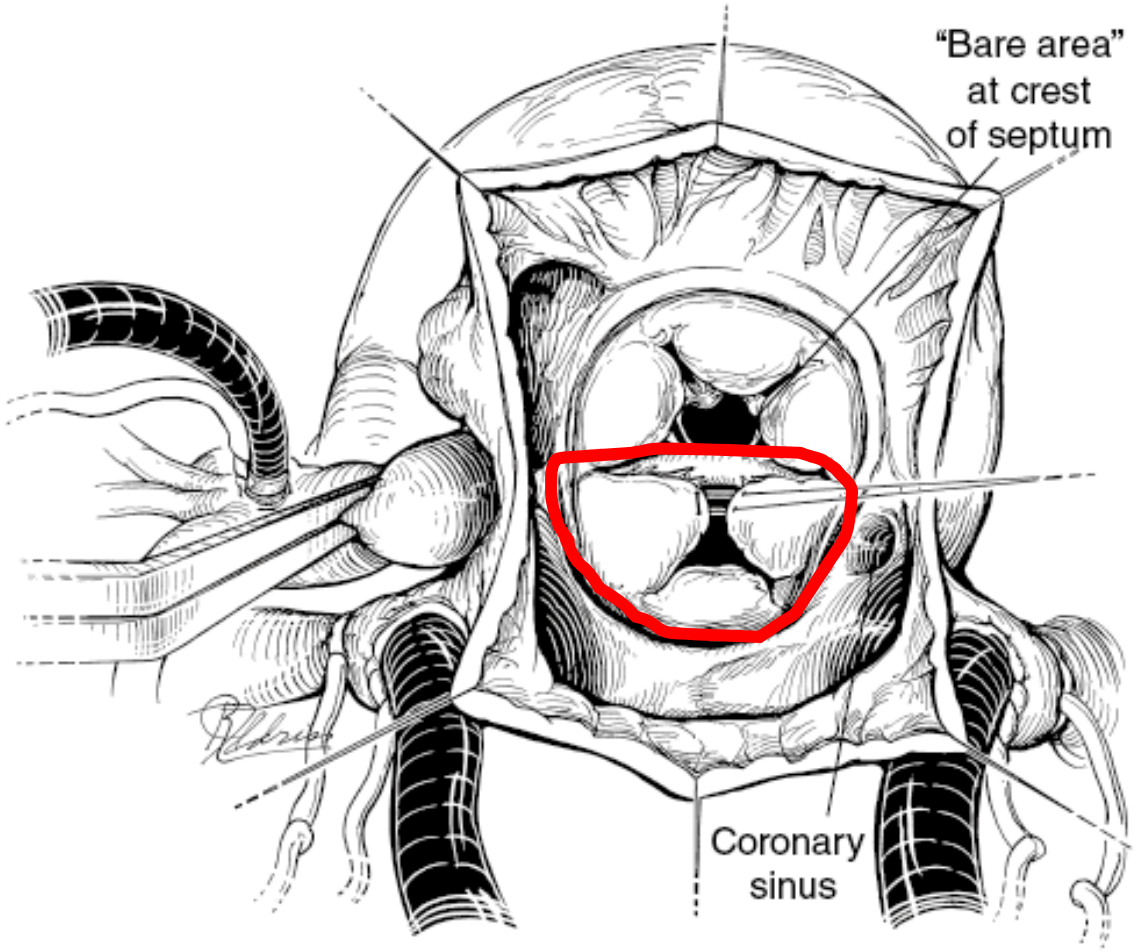
”The Gift that Keeps Giving”

Recurrent Issues:



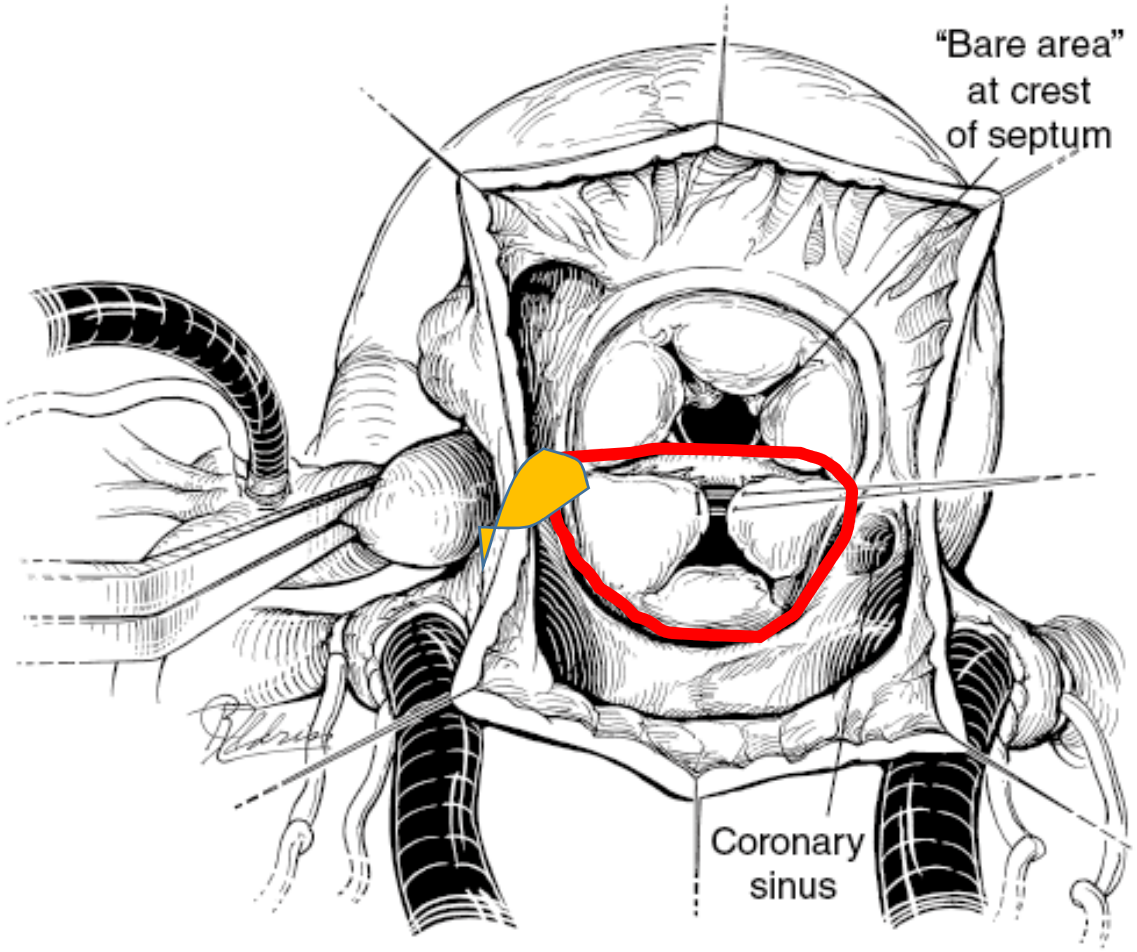
Recurrent Issues:

1. LAVV



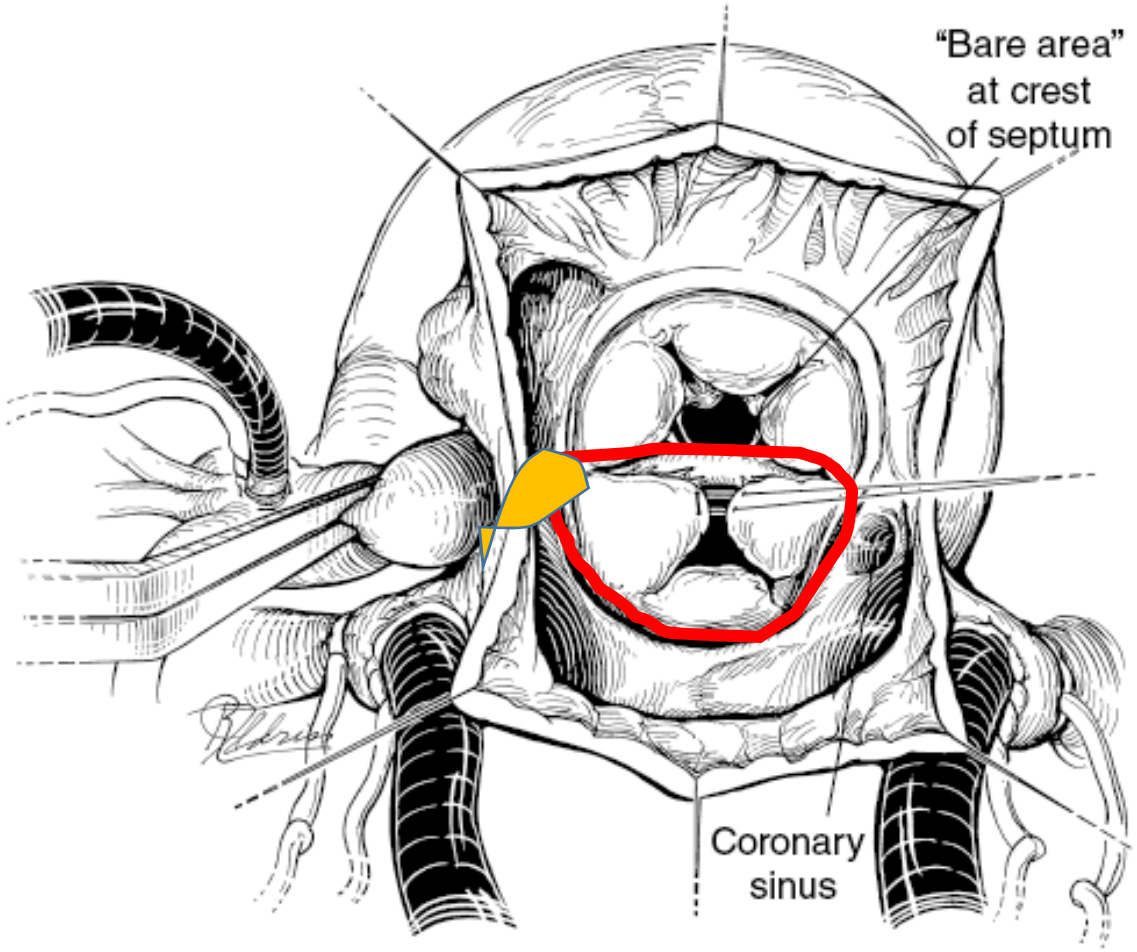
Recurrent Issues:

1. LAVV
2. Subaortic Area



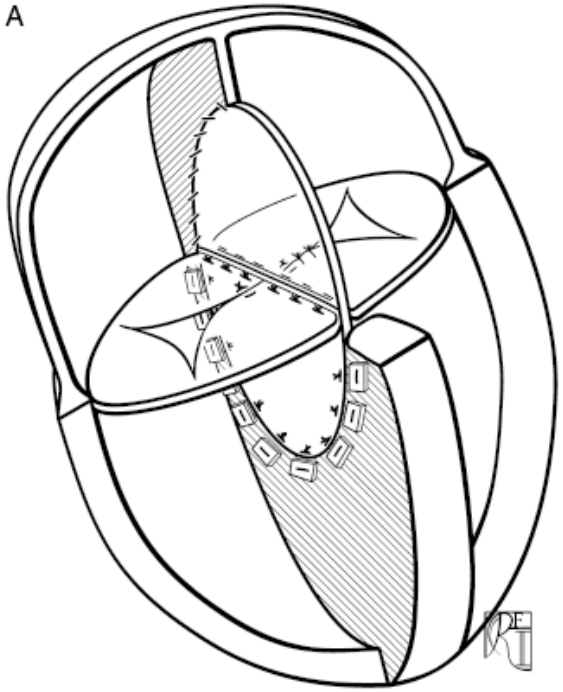
Recurrent Issues:

1. LAVV
2. Subaortic Area
3. RAVV
4. Residual Lesions

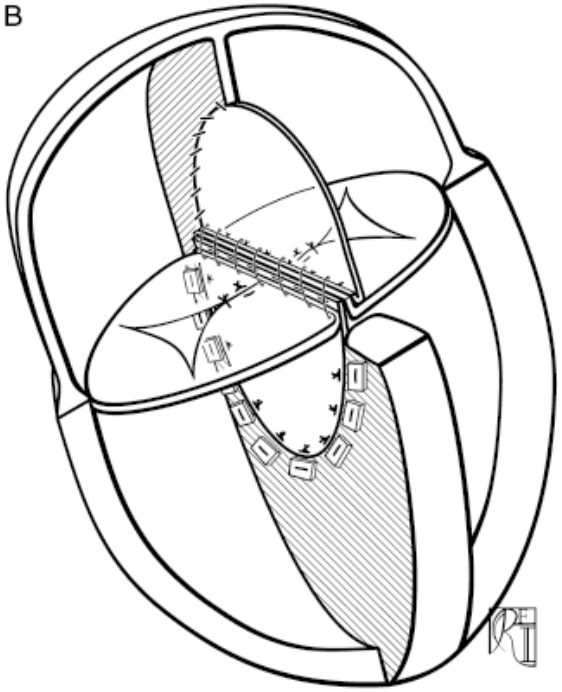


Types of Repairs

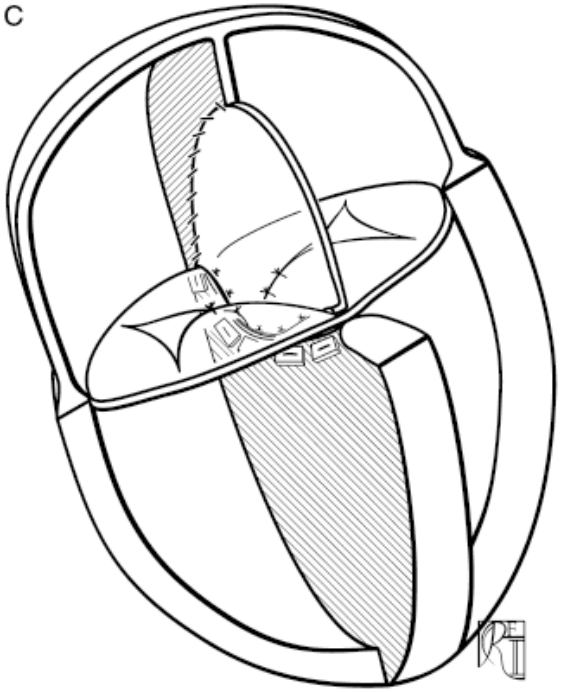
Single



Double



Modified Single



Outcomes: Recent Series

Table 18.1 A comparison summary of selected recent series of AV canal repairs. (NS, not stated.)

Author	Number of patients	Operative mortality	Late mortality	Left AV valve reoperation	Heart block
Modified single-patch technique					
Wilcox [12]	12	1	0	0	0
Nunn [43]	128	2	0	3	0
Jonas [44]	34	0	NS	0	1
Backer [45]	26	1	0	1	0
	200	4 (2%)	0	4 (2%)	1 (0.5%)
Two-patch technique					
Litwin [46]	222	6	6	13	3
Backer [45,19,47]	173	10	2	14	7
Lacour-Gayet [48]	110	4	2	7	3
Masuda [49]	64	2	3	5	NS
Ten Harkel [50]	111	3	3	10	2
Fortuna [51]	209	6	3	15	2
	889	31 (3.5%)	19 (2.1%)	64 (7.2%)	17 (2%)
Classic single-patch technique					
Crawford [52]	88	0	0	9	3
Reddy [53]	72	1	1	2	0
Prifti [54]	190	16	13	23	5
	350	17 (4.8%)	14 (4%)	34 (9.7%)	8 (2.3%)

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Types of Repairs

Single | **Double** | **Modified Single**

But what are the nuances?

Close the Cleft/ZOA – Interrupted vs. Running?

IS THE DEVIL IN THE DETAILS?

Define small left side?

What is the end-diastolic pressure?

Patient acquired risk factors?

Reoperation is a risk for mortality

Long-term outcomes after surgical repair of complete atrioventricular septal defect

Salil Ginde, MD,[a] Janna Lam, BS,[a] Garick D. Hill, MD,[a] Scott Cohen, MD,[b] Ronald K. Woods, MD, PhD,[c] Michael E. Mitchell, MD,[c] James S. Tweddell, MD,[c] and Michael G. Earing, MD[a,b]

198 – 178 – 153 (86%) long term data 17.2 years

TABLE 2. Risk factors for late mortality, by univariate and multivariable proportional hazards analysis

	Univariate analysis		Multivariable analysis	
Variable	*P* value	HR (95% CI)	*P* value	HR (95% CI)
Early surgical era	.001	9.38 (2.56-34.32)	.07	4.34 (0.91-20.71)
Classic 1-patch	.0006	8.36 (2.39-29)	.45	2.17 (0.30-16)
Older age at repair	.02	NA	.42	1.01 (0.96-1.02) per y
Required reoperation	.007	5.4 (1.77-16.6)	.04	3.06 (1.05-8.91)
Down syndrome	.82	1.17 (0.33-4.17)	.88	0.9 (0.24-3.43)
Prior PA band	.29	2.91 (0.40-21.26)	.31	2.87 (0.37-22.04)

TABLE 4. Risk factors for reoperation, by univariate and multivariable proportional hazards analysis

	Univariate analysis		Multivariable analysis	
Variable	*P* value	HR (95% CI)	*P* value	HR (95% CI)
Early surgical era	.04	2.45 (1.06-5.67)	.74	1.29 (0.29-5.62)
Older age at repair	.0001	NA	.6	1.0 (0.97-1.01)
Classic 1-patch	.002	3.8 (1.63-8.88)	.003	3.71 (1.56-8.78)
Non–Down syndrome	.1	2.15 (0.87-5.39)	.27	1.62 (0.68-3.84)
Coarctation of aorta	.16	4.14 (0-22)	.47	2.09 (0.28-15.53)
Prior PA band	.17	0.51 (0.2-1.31)	.7	0.81 (0.28-2.33)

J Thorac Cardiovasc Surg 2015;150:369-74

Survival vs. Freedom from Reoperation

Long-term outcomes after surgical repair of complete atrioventricular septal defect

Salil Ginde, MD,[a] Janna Lam, BS,[a] Garick D. Hill, MD,[a] Scott Cohen, MD,[b] Ronald K. Woods, MD, PhD,[c] Michael E. Mitchell, MD,[c] James S. Tweddell, MD,[c] and Michael G. Earing, MD[a,b]

Operative mortality 10.1%

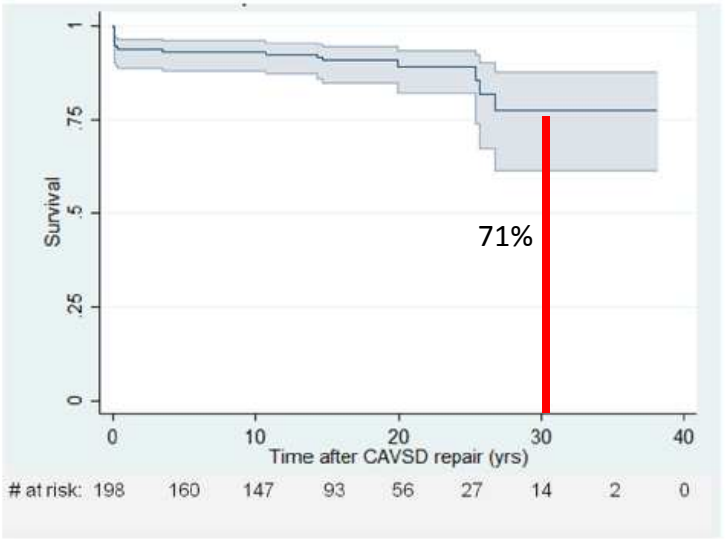

FIGURE 1. Estimated overall survival after CAVSD repair. *CAVSD*, Complete atrioventricular septal defect.

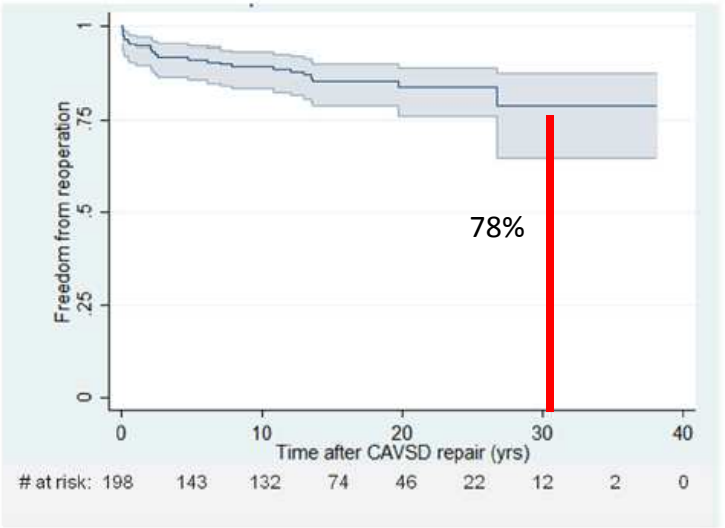

FIGURE 2. Estimated freedom from reoperation after CAVSD repair. *CAVSD*, Complete atrioventricular septal defect.

J Thorac Cardiovasc Surg 2015;150:369-74

Risk factors for reoperation

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TABLE 3. Type and frequency of reoperations performed after complete AV septal defect repair (n = 198)

Indication for reoperation	N (%)
Left AV valve regurgitation	14 (7.1)
LVOT obstruction	7 (3.5)
Residual VSD	7 (3.5)
Residual ASD	1 (0.5)
Native coarctation of the aorta (surgical repair)	1 (0.5)
Vascular ring	1 (0.5)

AV, Atrioventricular; *LVOT*, left ventricular outflow tract; *VSD*, ventricular septal defect; *ASD*, atrial septal defect.

J Thorac Cardiovasc Surg 2015;150:369-74

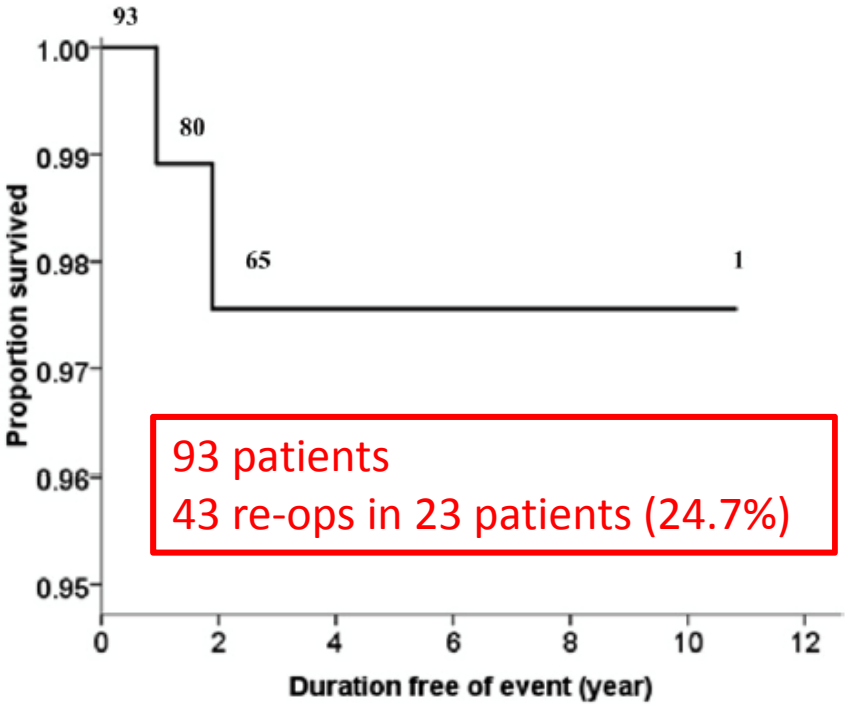
Risk factors for reoperation

Primary Biventricular Repair of Atrioventricular Septal Defects: An Analysis of Reoperations

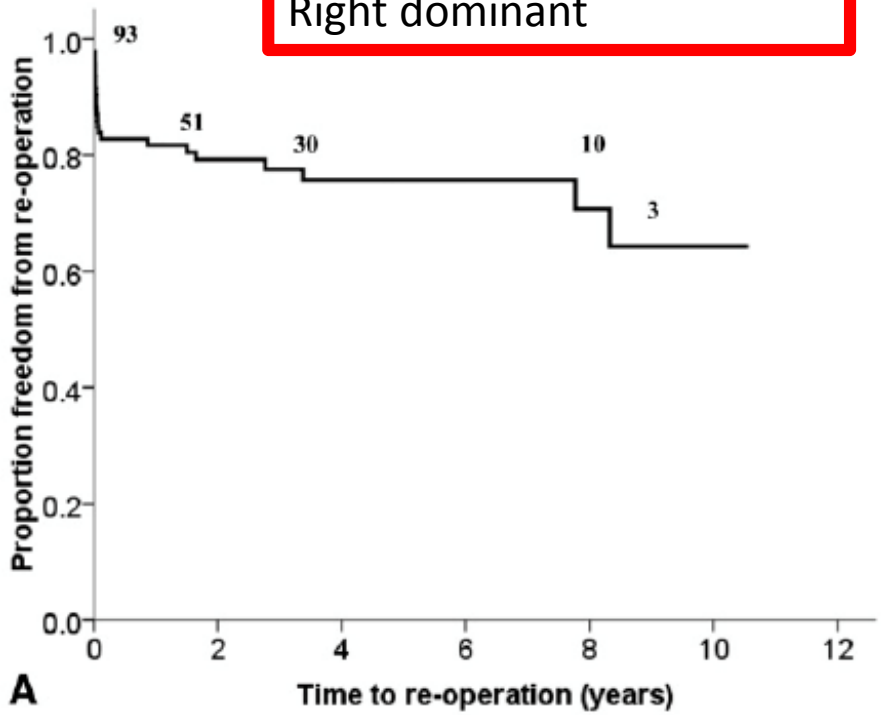
Hunaid A. Vohra, FRCS (CTh), Alicia X.F. Chia, MB BS, Ho Ming Yuen, BS, Joseph J. Vettukattil, MRCP, Gruschen Veldtman, MRCP, James Gnanapragasam, FRCP, Kevin Roman, MRCP, Anthony P. Salmon, FRCP, and Marcus P. Haw, FRCS (CTh)

Wessex Cardiothoracic Centre, Southampton University Hospitals, Southampton, United Kingdom

Predictors of Reoperation:
Down syndrome
Right dominant



93 patients
43 re-ops in 23 patients (24.7%)



Ann Thorac Surg 2010;90:830–8

Indications for reoperation

Primary Biventricular Repair of Atrioventricular Septal Defects: An Analysis of Reoperations

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Ann Thorac Surg 2010;90:830–8

Table 2. Indications for Reoperation After Primary Atrioventricular Septal Defect Repair

	Isolated CAVSD (n = 53)		Complex AVSD[b] (n = 5)		Intermediate AVSD (n = 6)		PAVSD (n = 29)	
Indications[a]	N (%)	No. of Procedures	N (%)	No. of Procedures	N (%)	No. of Procedures	N (%)	No. of Procedures
LAVVR repair	10 (18.9%)	14	1 (20.0%)	2	0 (0%)	0	2 (6.7%)	2
Mitral valve replacement	6 (11.3%)	6	0 (0%)	0	0 (0%)	0	1 (3.4%)	2
Subaortic membrane resection (± LAVVR repair)	1 (1.9%)	2	0 (0%)	0	0 (0%)	0	1 (3.4%)	1
Repair of dehiscence of interatrial or interventricular patch	4 (7.5%)	5	0 (0%)	0	0 (0%)	0	1 (3.4%)	1
Permanent pacemaker	2 (3.8%)	2	2 (40.0%)	2	0 (0%)	0	3 (10.3%)	3
Excision of previous banding site and reconstruction of PA	1 (1.9%)	1	0 (0%)	0	0 (0%)	0	0 (0%)	0
Patch augmentation of PA and pulmonary valve replacement for RVOTO	0 (0%)	0	2 (40.0%)	2	0 (0%)	0	0 (0%)	0

[a] Patients could have multiple indications for reoperation.

[b] Includes complete atrioventricular septal defect (CAVSD) plus tetralogy of Fallot and CAVSD plus double-outlet right ventricle.

Etiology of LAVV Pathology

Reoperations After Repair of Partial Atrioventricular Septal Defect: A 45-Year Single-Center Experience

Ann Thorac Surg 2010;89:1352–9

Table 1. Left Atrioventricular Valve Pathology Encountered in the Operating Room

Pathology[a]	No.	%
Residual ZOA-repair dehiscence	42	44
Residual ZOA	25	26
Previously unrepaired	12	13
Prior repair dehiscence	5	5
Severe annular dilatation	22	23
Dysplastic leaflet tissue	13	14
Abnormal leaflet pliability	11	11
Septation patch dehiscence	10	10
Leaflet prolapsed-tethering	8	8
Annular-leaflet calcification	6	6
Leaflet perforation	4	4
Stenosis from prior repair	3	3
Prosthesis leak-degeneration	2	2

Etiology of LAVV Pathology

Reoperations After Repair of Partial Atrioventricular Septal Defect: A 45-Year Single-Center Experience

Ann Thorac Surg 2010;89:1352–9

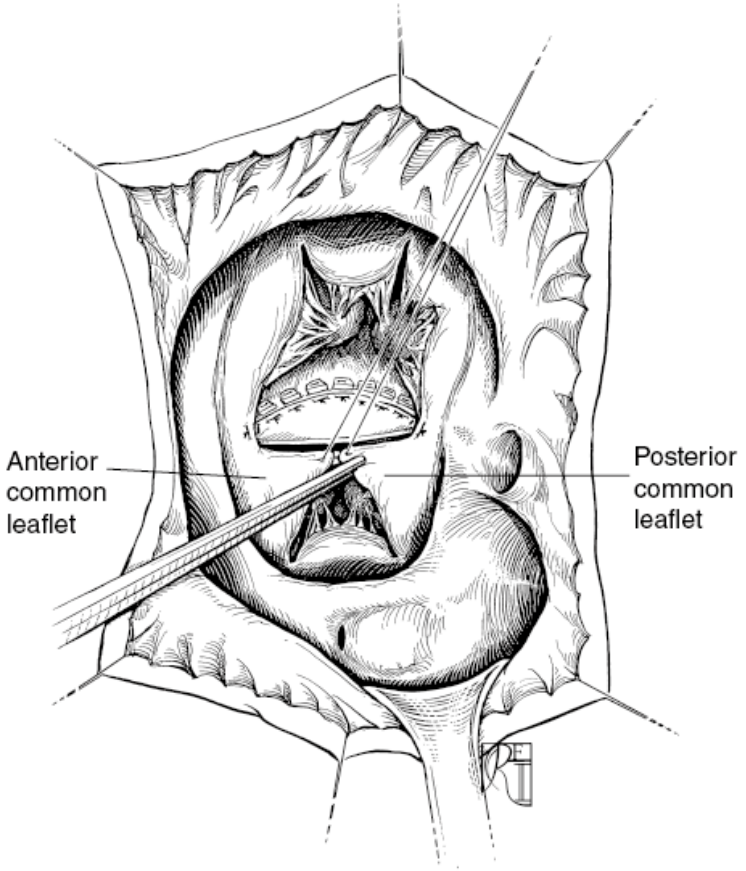
Table 2. Valve Repair Techniques Utilized At First Reoperation

Technique[a]	No.	%
ZOA repair	31	32
Rerepair	19	20
Primary	12	12
Annuloplasty	24	25
Eccentric	14	15
Pursestring	5	5
Ring	5	5
Edge to edge repair	5	5
Leaflet perforation repair	4	4
Papillary muscle splitting	1	1
Accessory orifice closure	1	1

Technique of Repair – Zone of Apposition

<u>Access to the LAVV in a Redo:</u>

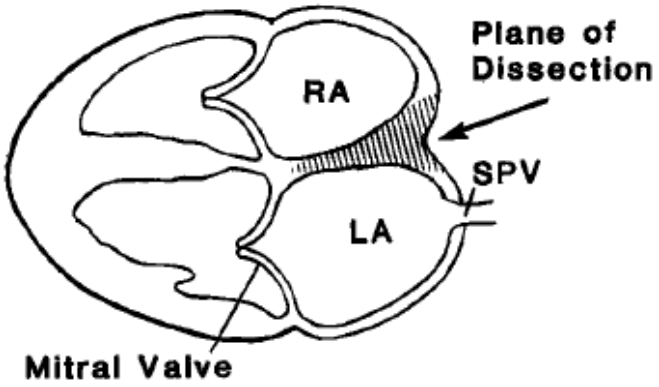
- Trans-septal
  - Be prepared to replace ASD patch

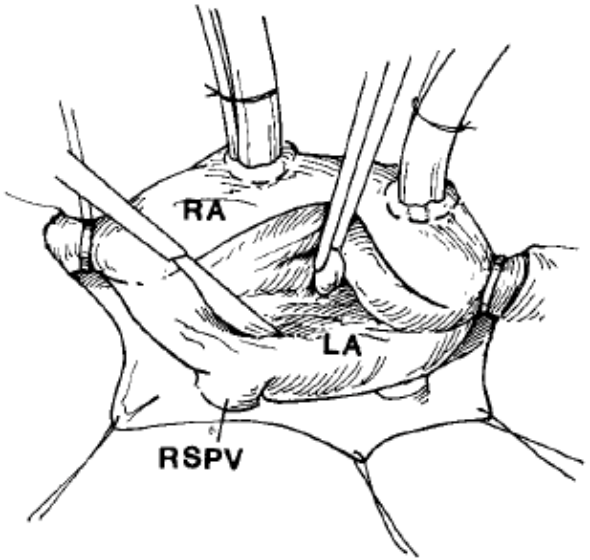


Technique of Repair – Zone of Apposition

Access to the LAVV in a Redo:

- Trans-septal
  - Be prepared to replace ASD patch
- Left Atrial
  - Difficult exposure
  - Sondergaard's groove (1955) is challenging with a previously placed patch



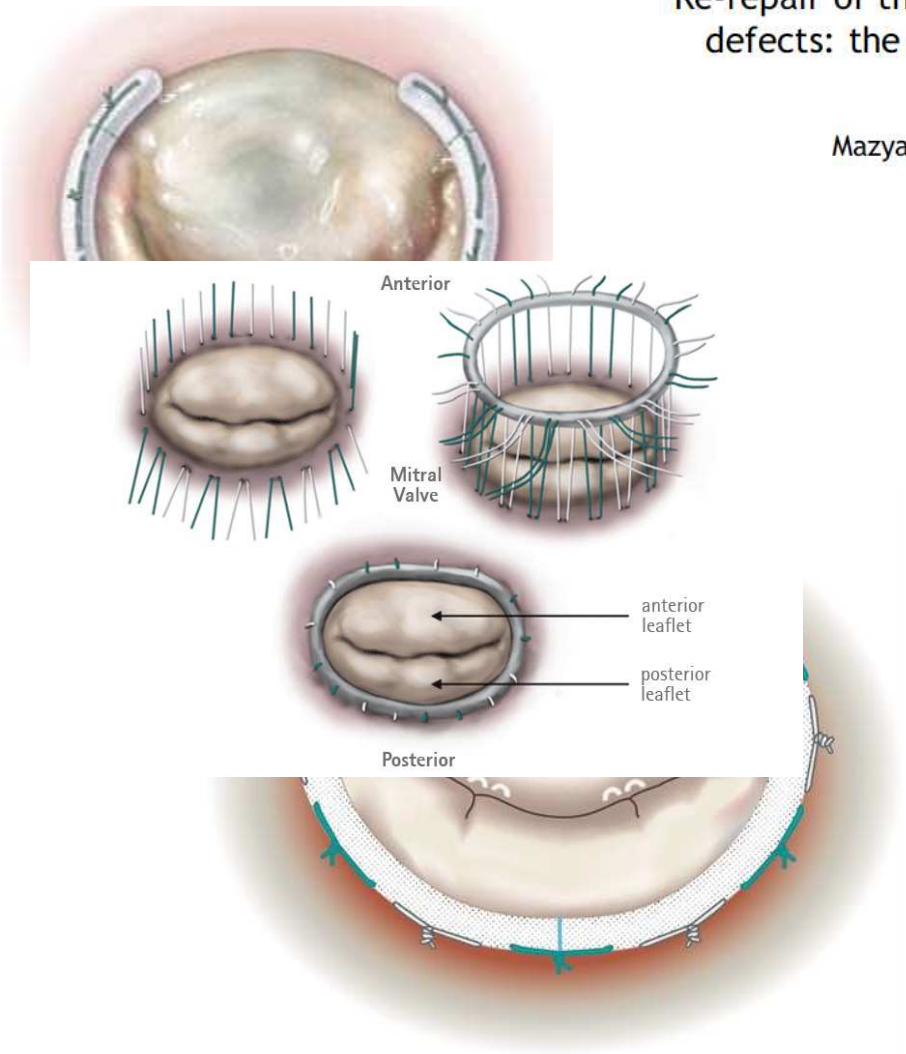


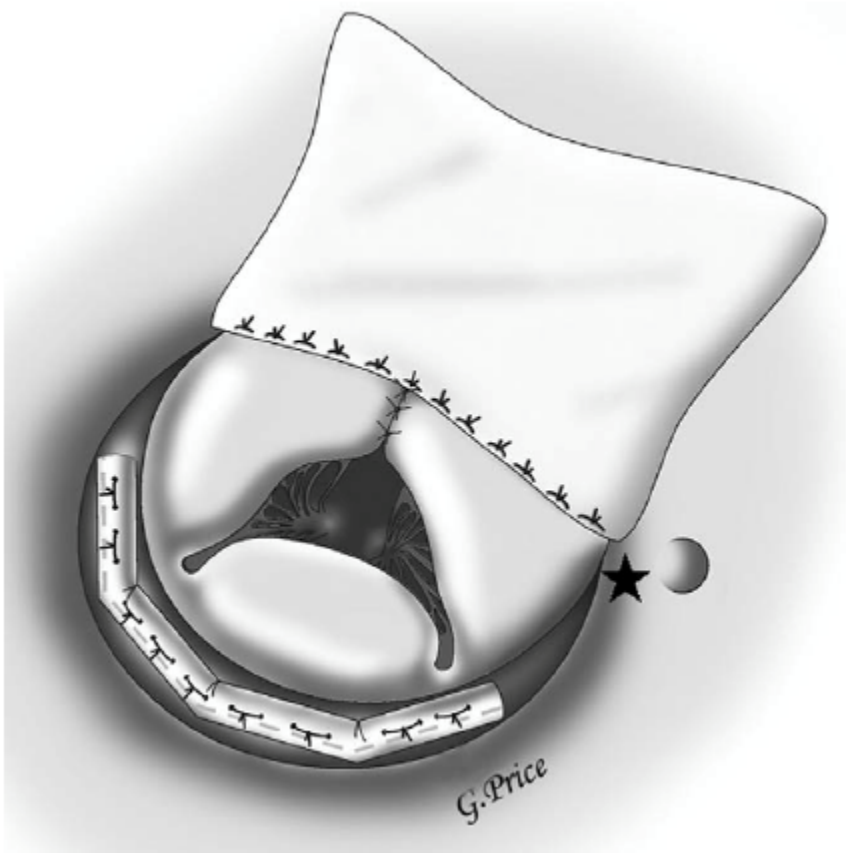
Ann Thorac Surg 1992;54:1186–8.

Technique of Repair – Reduction Annuloplasty

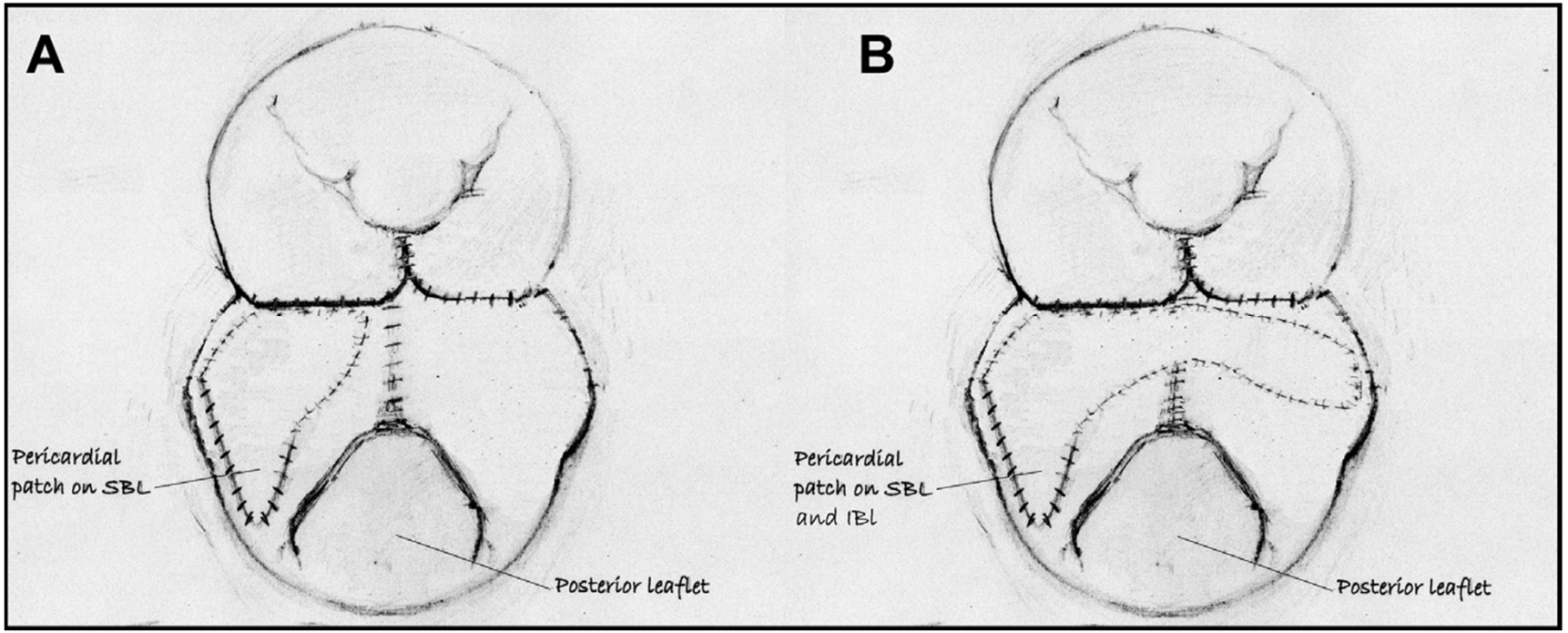
Re-repair of the left atrioventricular valve in atrioventricular septal defects: the morphologic approach to the role of Gore-tex band reduction annuloplasty

Mazyar Kanani*, Victor Tsang, Andrew Cook, Martin Kostolny





Technique of Repair – Insufficient Leaflet

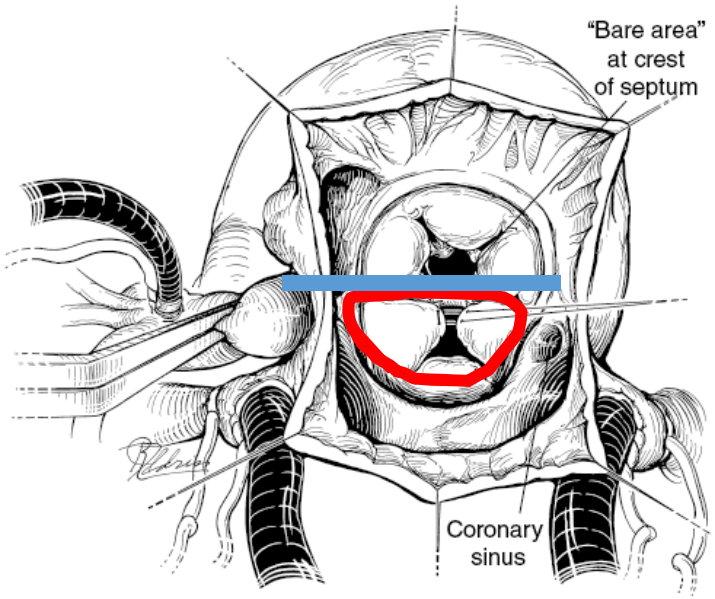


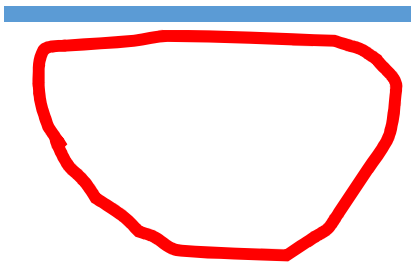
8 patients
50% at reoperation

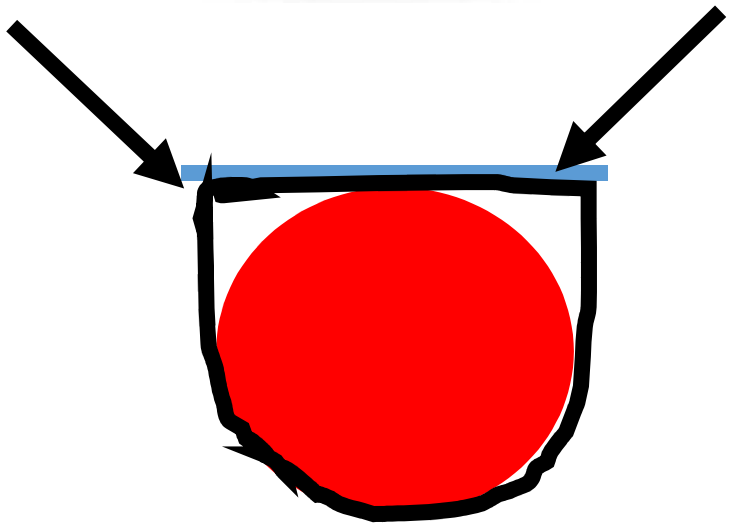
Ann Thorac Surg 2017;104:e101–3

Replacement:

Replacement:

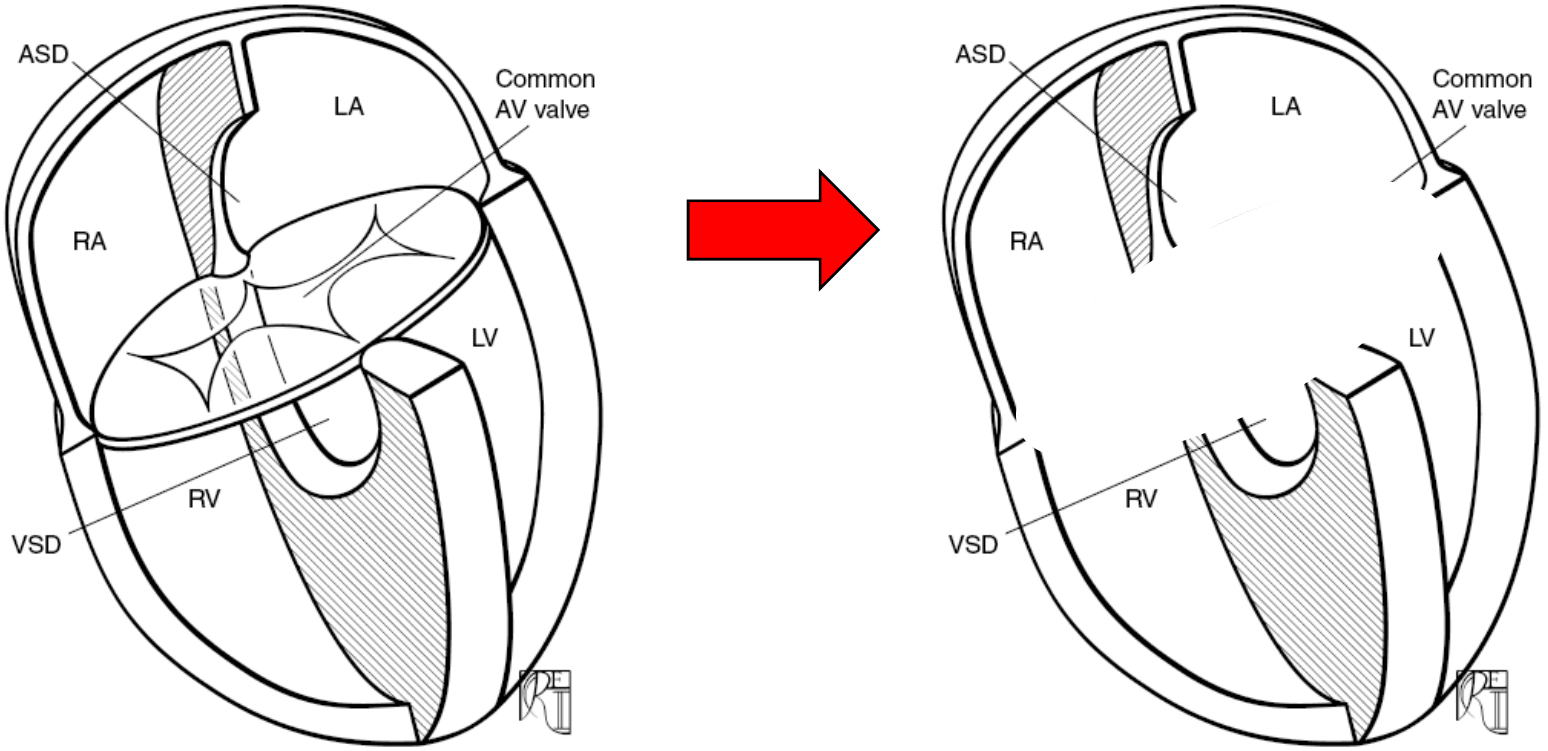




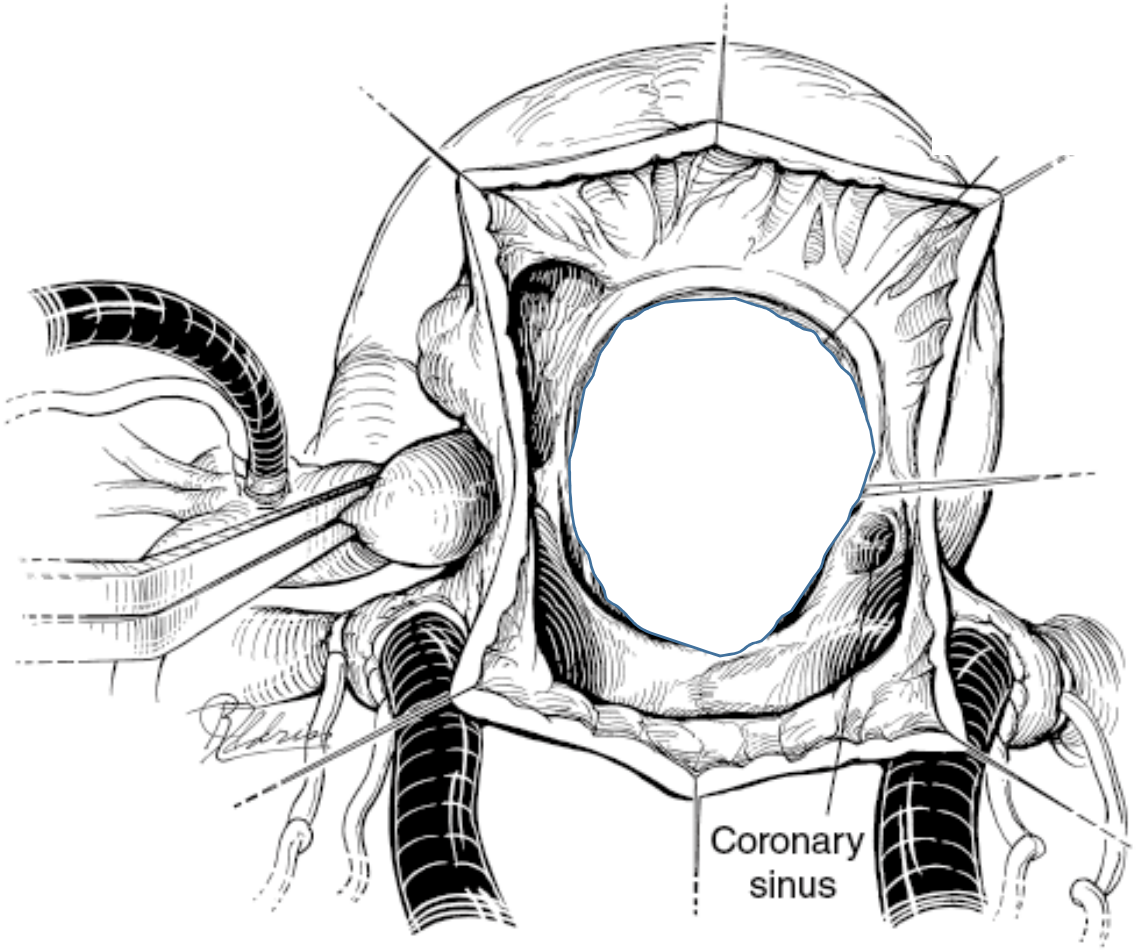


Replacement

Break down a repair



Replacement



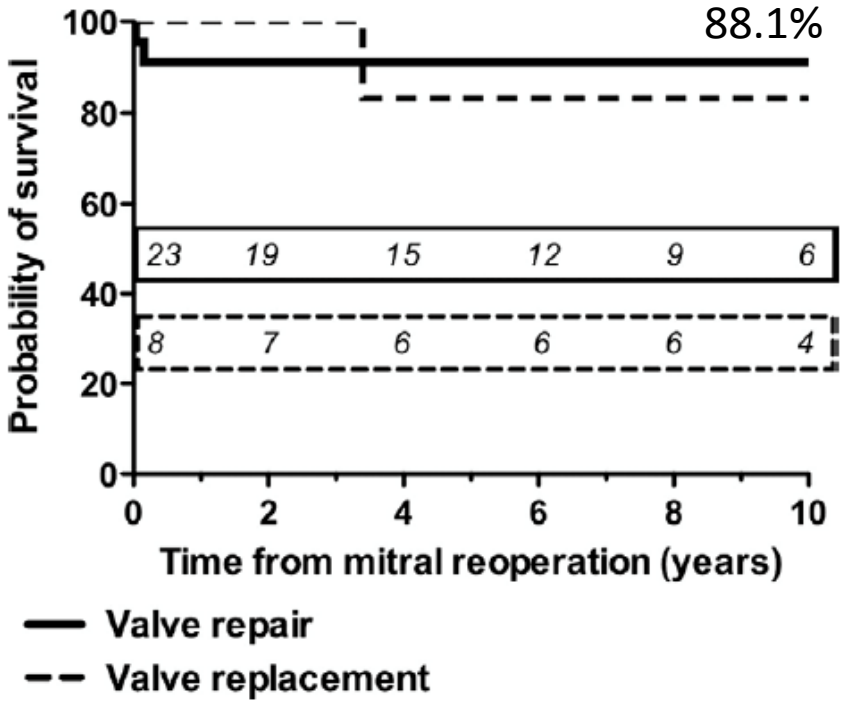
LAVV: Repair or Replace?

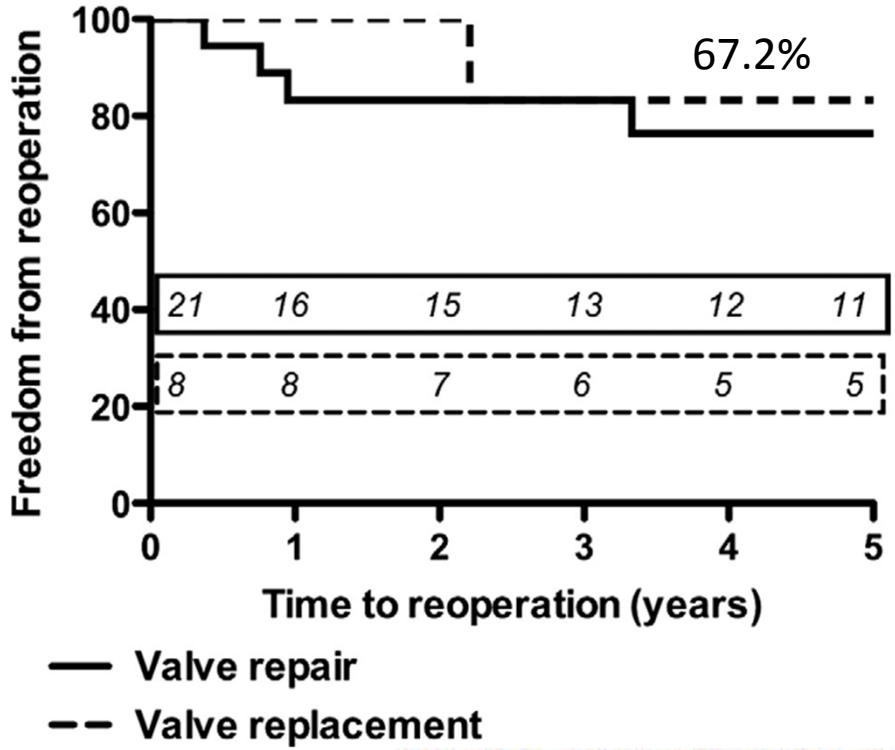
Reoperation for Left Atrioventricular Valve Regurgitation After Atrioventricular Septal Defect Repair

Sunil P. Malhotra, MD, Francois Lacour-Gayet, MD, Max B. Mitchell, MD, David R. Clarke, MD, Marshall L. Dines, BS, and David N. Campbell, MD

Division of Cardiac Surgery, Children's Hospital Heart Institute, and Division of Cardiothoracic Surgery, Children's Hospital, Denver, Colorado

31 PATIENTS
23 Repairs
8 Replacments





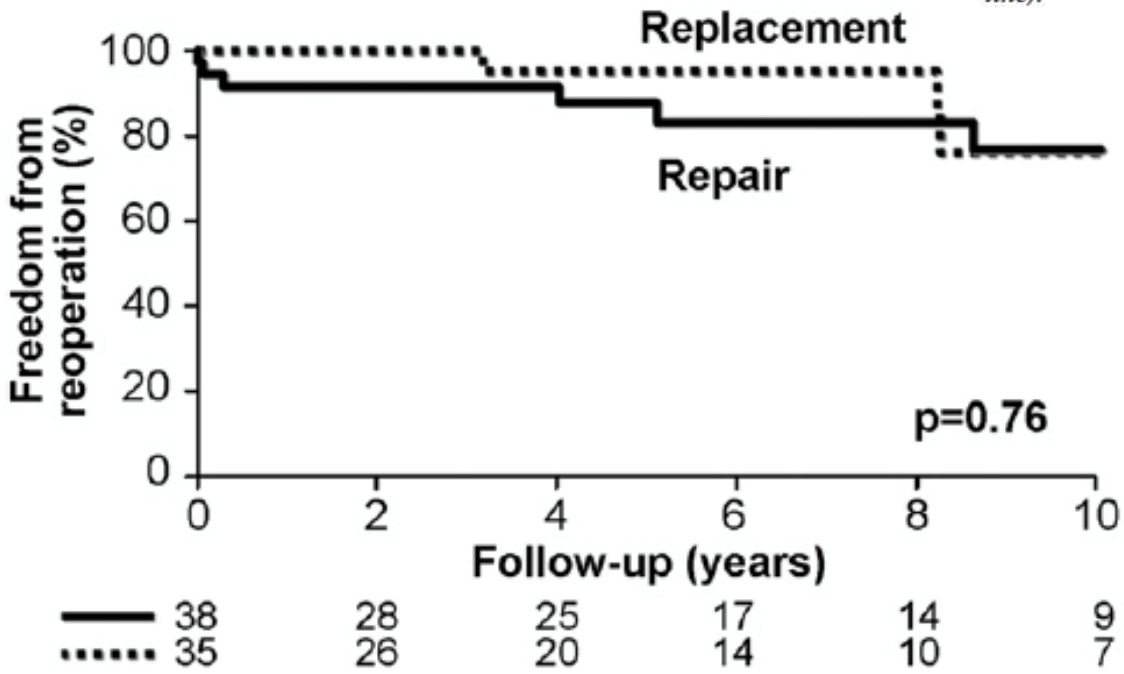
Ann Thorac Surg 2008;86:147–52

LAVV: Repair or Replace?

Reoperations After Repair of Partial Atrioventricular Septal Defect: A 45-Year Single-Center Experience

Ann Thorac Surg 2010;89:1352–9

Fig 5. Actuarial freedom from further reoperation for left atrioventricular valve pathology according to type of procedure performed. There was no significant difference (p = 0.76) between patients who underwent valve repair (solid line) or valve replacement (dashed line).



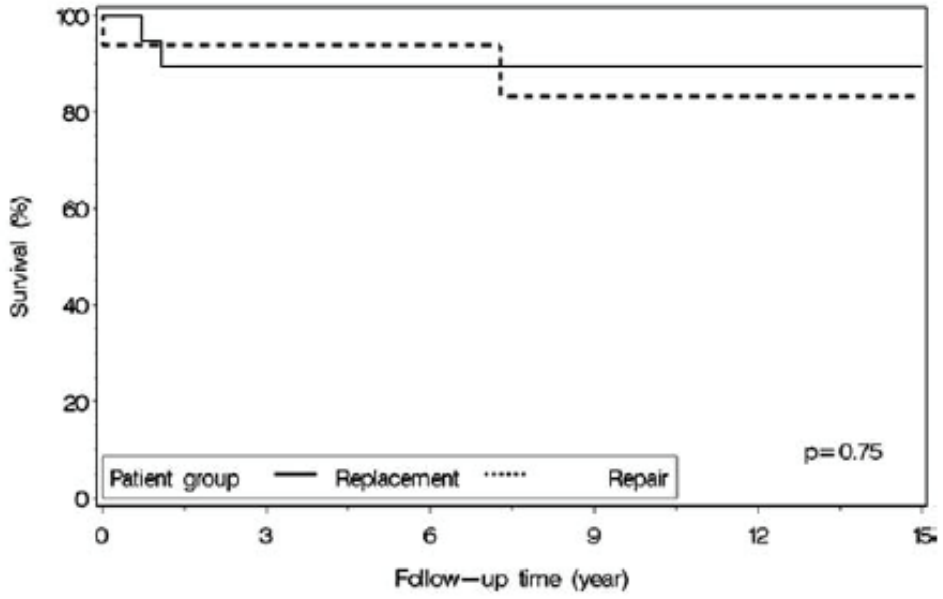
LAVV: Repair or Replace?

Reoperations After Initial Repair of Complete Atrioventricular Septal Defect

John M. Stulak, MD, Harold M. Burkhart, MD, Joseph A. Dearani, MD, Hartzell V. Schaff, MD, Frank Cetta, MD, Roxann D. Barnes, MD, and Francisco J. Puga, MD

Ann Thorac Surg 2009;87:1872–8

Survival after reoperation



Replacement	21	16	15	12	10	9
Repair	16	11	10	6	4	3

LAVV: Repair or Replace?

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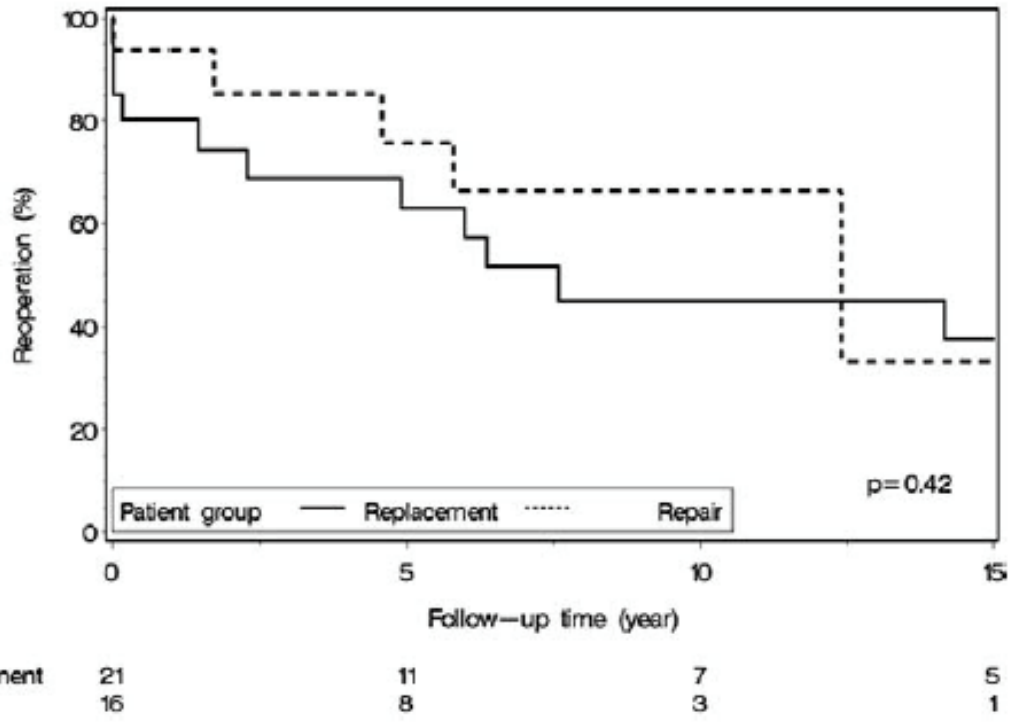
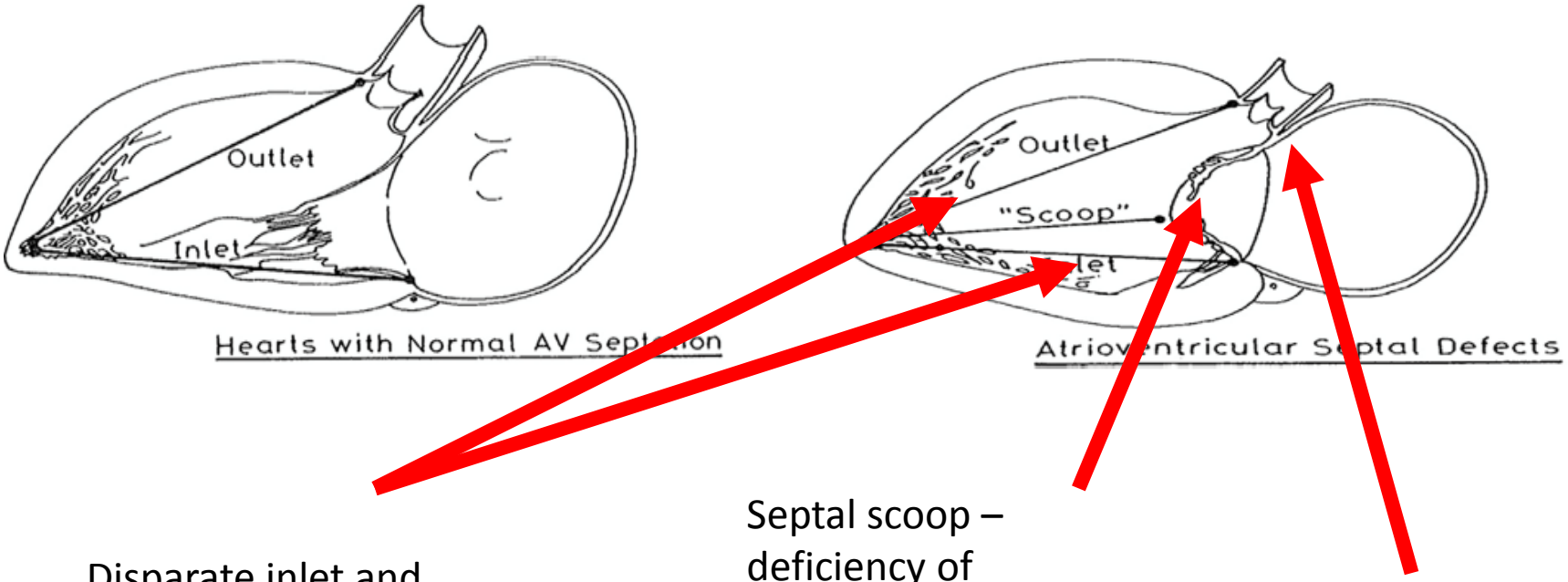
Freedom from further reintervention



42%

LVOTO

Narrow and elongated LVOT



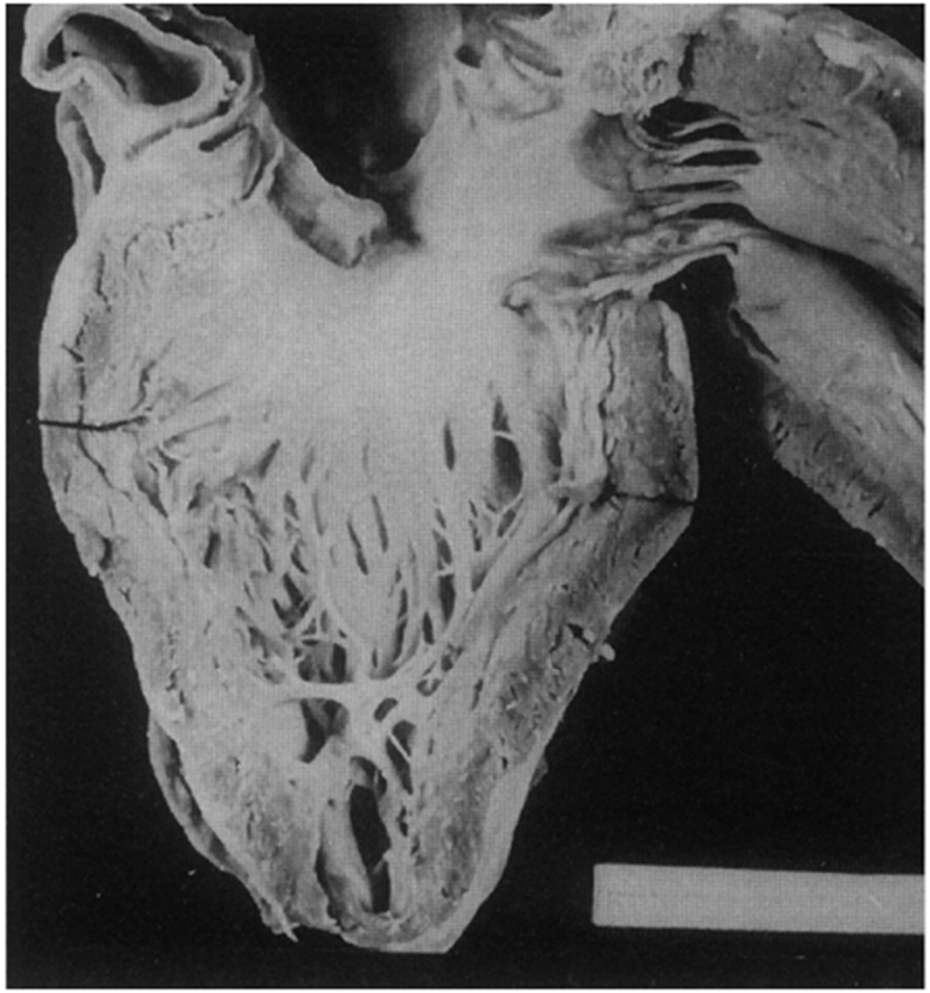
Disparate inlet and outlet septal lengths

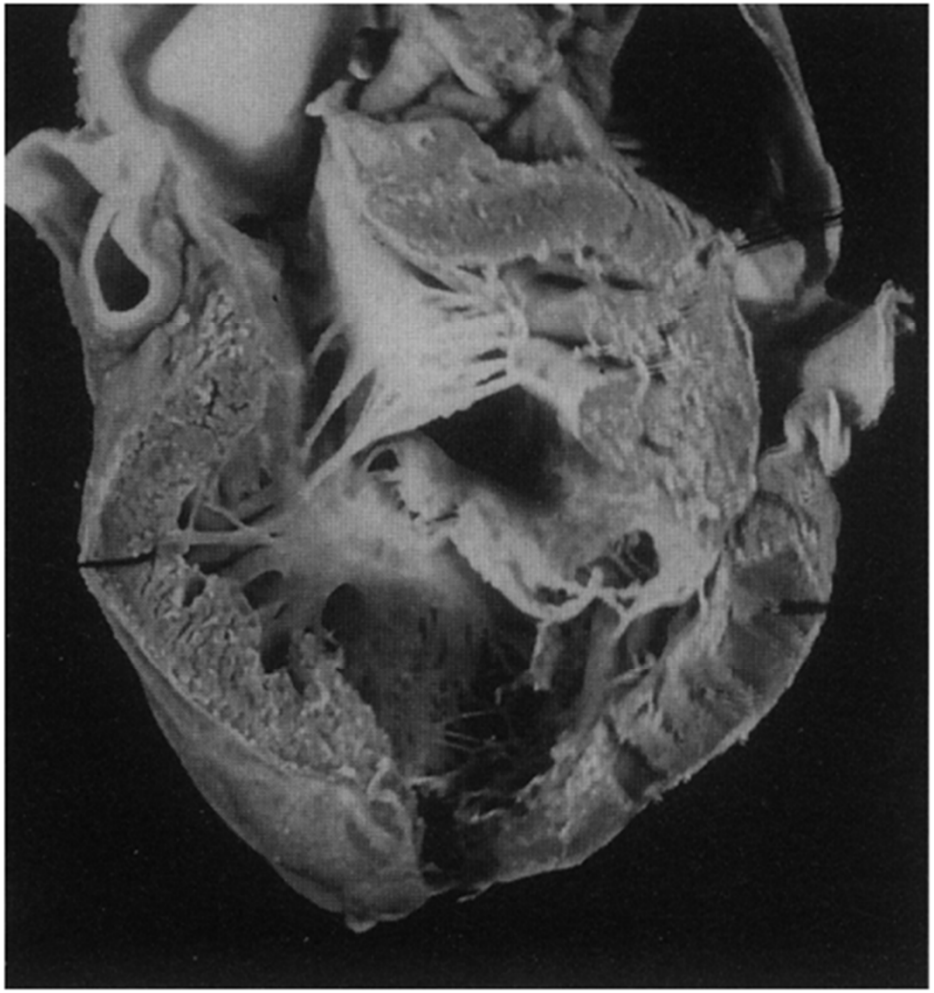
Septal scoop – deficiency of muscular septum

Unwedging of aortic root anterior and rightward

Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 17:43-47

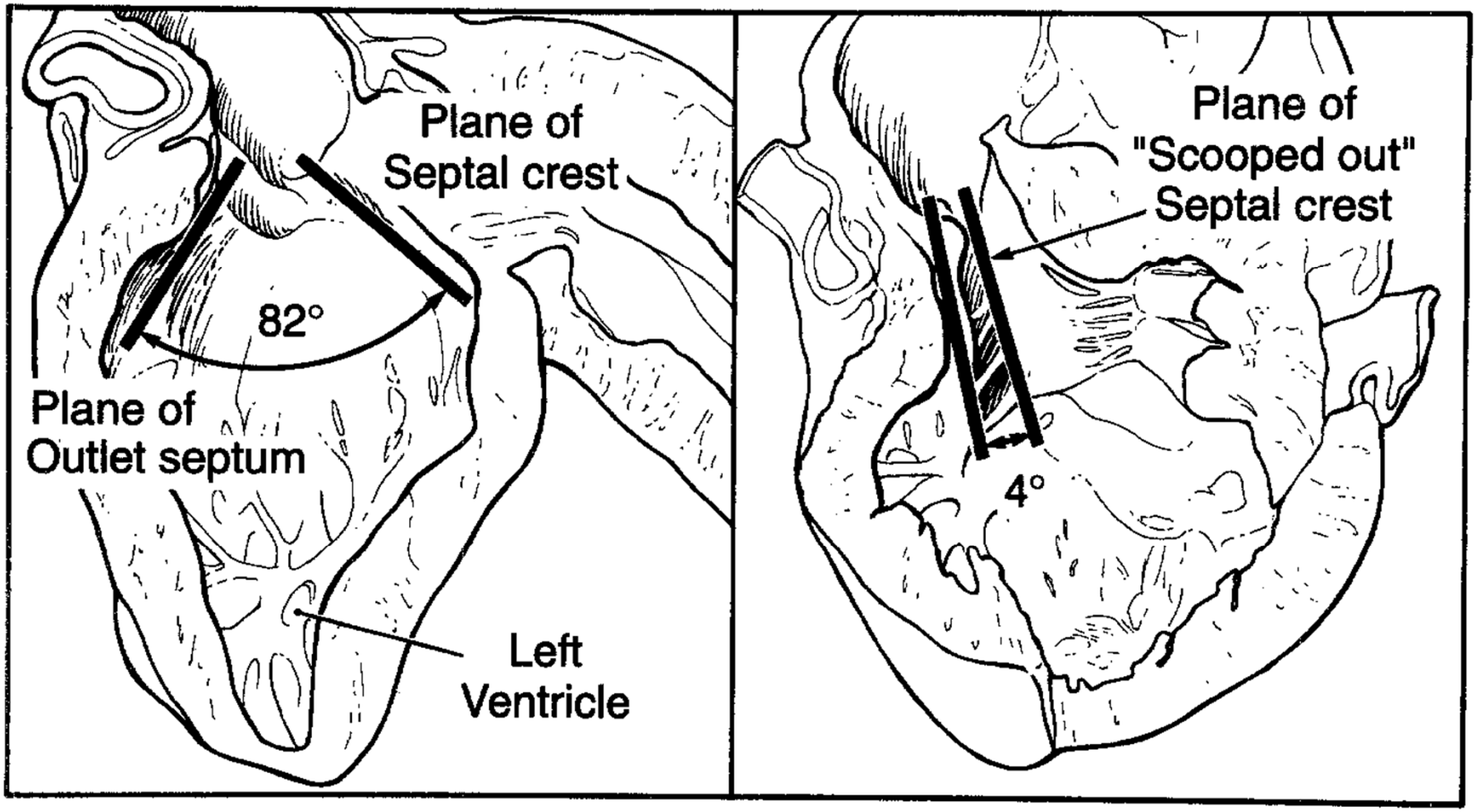
LVOTO





Van Arsdell GS, et al.J Thorac Cardiovasc Surg 1995;110:1534-1541

LVOTO



Van Arsdell GS, et al.J Thorac Cardiovasc Surg 1995;110:1534-1541

Reoperation: LVOTO

- Can occur on multiple levels
- Surgery must address all components
- 0.5-4.5%
- Significant impact on late survival

It is hard to definitively comment on whether the type of primary repair utilized affects the late incidence of left ventricular outflow obstruction. Intuitively, one would think that the placement of a VSD patch may open up the LVOT [left ventricular outflow tract] and lessen the development of late obstruction.

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Risk factors for reoperation

Long-term outcomes after surgical repair of complete atrioventricular septal defect

Salil Ginde, MD,[a] Janna Lam, BS,[a] Garick D. Hill, MD,[a] Scott Cohen, MD,[b] Ronald K. Woods, MD, PhD,[c] Michael E. Mitchell, MD,[c] James S. Tweddell, MD,[c] and Michael G. Earing, MD[a,b]

TABLE 6. Risk factors for need for a reoperation to address left ventricular outflow tract obstruction, by univariate and multivariable proportional hazards analysis

	Univariate analysis		Multivariable analysis	
Variable	***P* value**	**HR (95% CI)**	***P* value**	**HR (95% CI)**
Early surgical era	.15	3.55 (0.76-16.5)	.8	1.36 (0.13-14.49)
Older age at repair	.38	NA	.27	0.97 (0.91-1.03)
Classic 1-patch	.05	5.14 (1.17-22.6)	.10	6.17 (1.24-68.59)
Non–Down syndrome	.37	2.32 (0.59-9.24)	.43	1.81 (0.42-7.72)
Coarctation of aorta	>.98	0 (0-16.22)	>.98	0 (0-∞)
Prior PA band	>.98	0.73 (0-4.8)	.55	0.52 (0.06-4.44)

J Thorac Cardiovasc Surg 2015;150:369-74

LVOTO

Reoperation for Left Ventricular Outflow Tract Obstruction After Repair of Atrioventricular Septal

Components of LVOT Resection in CAVC:

- Complete removal of all fibromuscular material
- Aggressive and extensive muscular resection
- Removal of anomalous secondary and tertiary left AV valve chordae from the LVOT

- Recalcitrant cases require a modified Konno
- John Brown reports apical conduit

Reoperation rate: 15-37%

Van Arsdell GS, et al.J Thorac Cardiovasc Surg 1995;110:1534-1541

Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 17:43-47

LVOTO

Improving Left Ventricular Outflow Tract Obstruction Repair in Common Atrioventricular Canal Defects

Patrick O. Myers, MD, Pedro J. del Nido, MD, Gerald R. Marx, MD, Sitaram Emani, MD, John E. Mayer, Jr, MD, Frank A. Pigula, MD, and Christopher W. Baird, MD

Ann Thorac Surg 2012;94:599–605

Table 3. Operative Techniques of Subaortic Stenosis Relief at Reoperation

Technique	No.
Resection of	
Membrane	41
Submitral accessory tissue or attachments	22
Septal myectomy	27
Modified Konno	5
Aortic valve repair	9
Commissurotomy	3
Leaflet thinning	9

Reoperation for LVOTO

Reoperations After Repair of Partial Atrioventricular Septal Defect: A 45-Year Single-Center Experience

Ann Thorac Surg 2010;89:1352–9

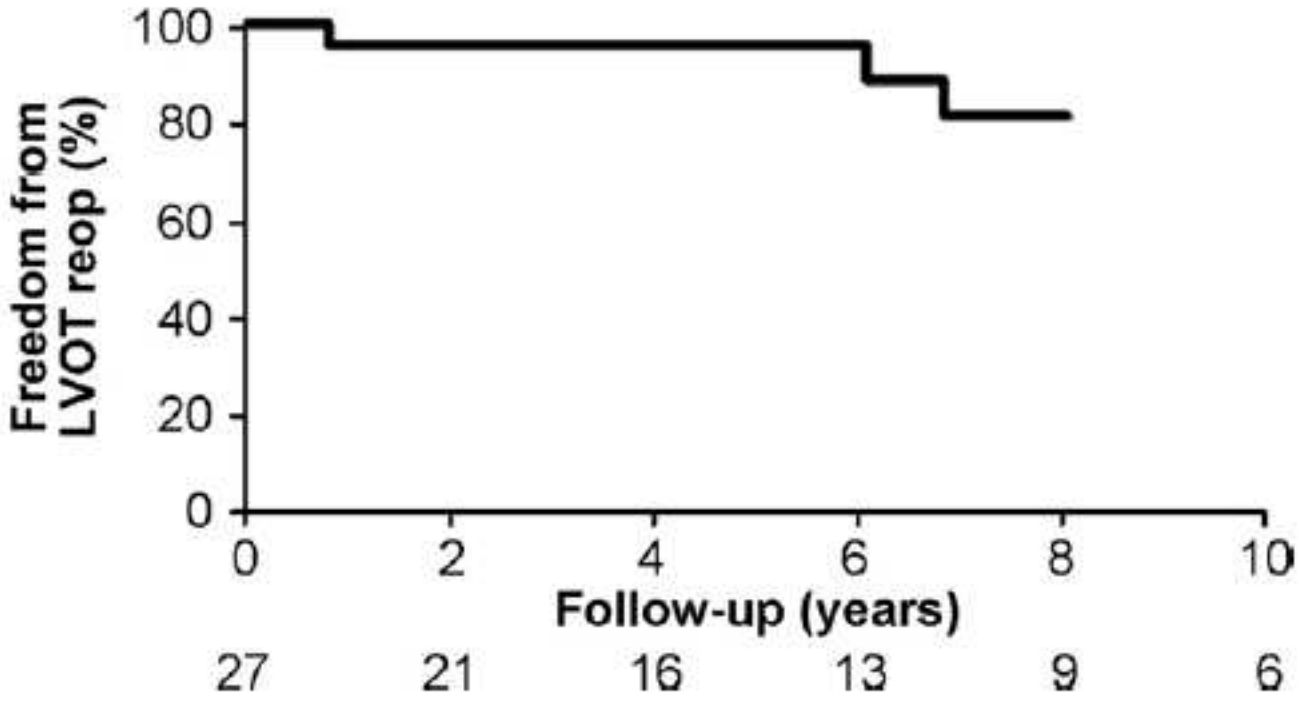

Fig 6. Actuarial freedom from further reoperation for relief of left ventricular outflow tract obstruction.

LVOTO

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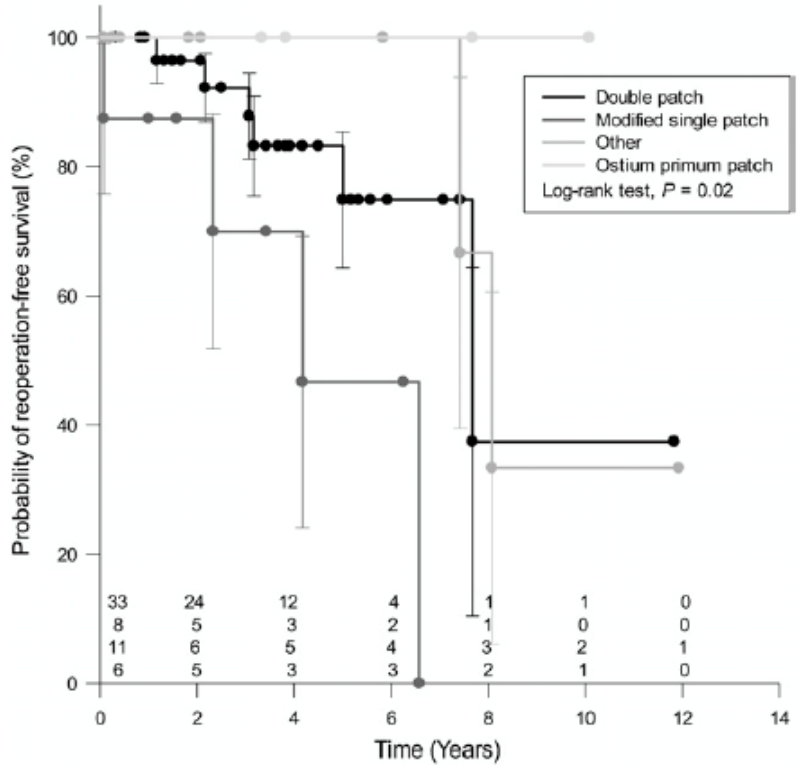

52.5% freedom from LVOT reoperation at 10y

33% Downs
40% Unbalanced

Modified single patch p =0.04

2nd reoperation rate 21%

Conclusions

- High incidence of reoperation
- Need for reoperation affects survival
- Repair techniques are feasible
  - Durability
  - Effectiveness
- If you replace, you may not be done
- Be aggressive about LVOT resection

Conclusions

- These patients require ongoing care
- Better stratification of risk factors at initial operation
- Believe the patient

Thank you!