STS/EACTS Latin America Cardiovascular Surgery Conference

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Surgery For Ebstein Anomaly

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The Society of Thoracic Surgeons

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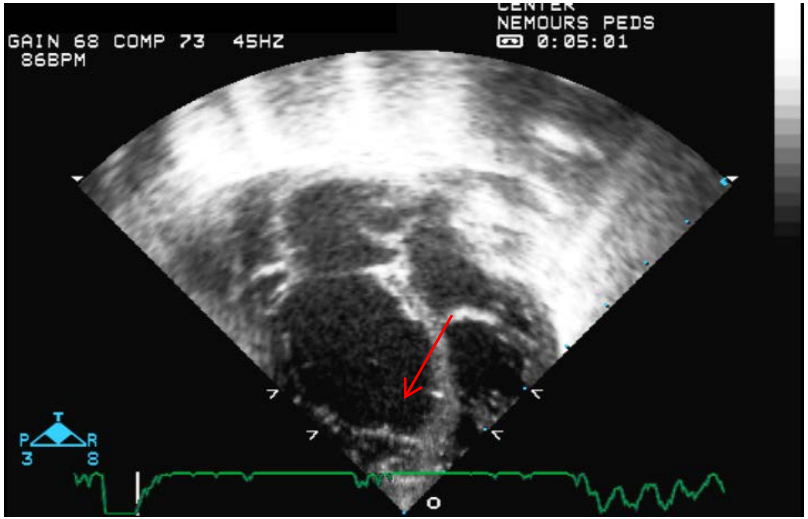
- No disclosures

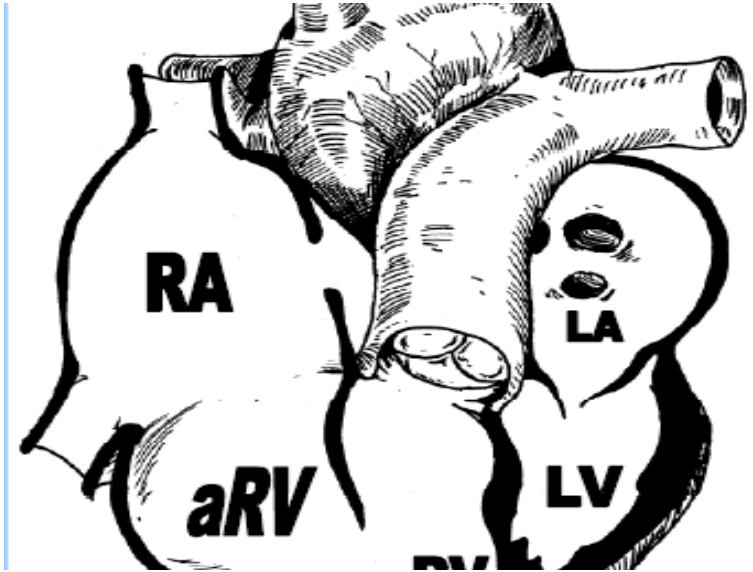
Ebstein Anomaly

- Morphology first described by Ebstein in 1866
- TV leaflets variably adherent to RV myocardiaum
- Spectrum of disease
- Clinically documented by Taussig in 1950
- Rare
- Bimodal age at presentation



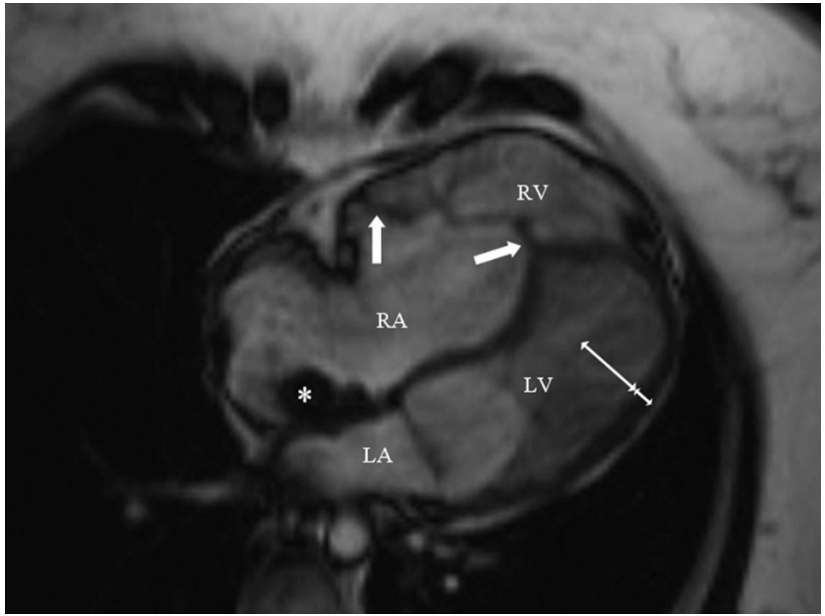
Neonatal Ebstein's: Predictors of death

- Cardiothoracic ratio greater than 0.85 (100% fatal)
- Echocardiography score grade 4/4 (>1.5:1; 100% fatal)
- Echocardiography score grade 3/4 (>1.1:1) and cyanosis (100% fatal)
- Severe tricuspid regurgitation (mostly fatal)
- Echocardiography score grade 3/4 (>1.1:1; 45% fatal in infancy)



Celermajer, DS. J Am Coll Cardiol 1992;19:1041-1046
Pavlova, M. Am Heart J 1998;135:1081-1085
Yetman, AT. Am J Cardiol 1998;81:749-754

MRI evaluation



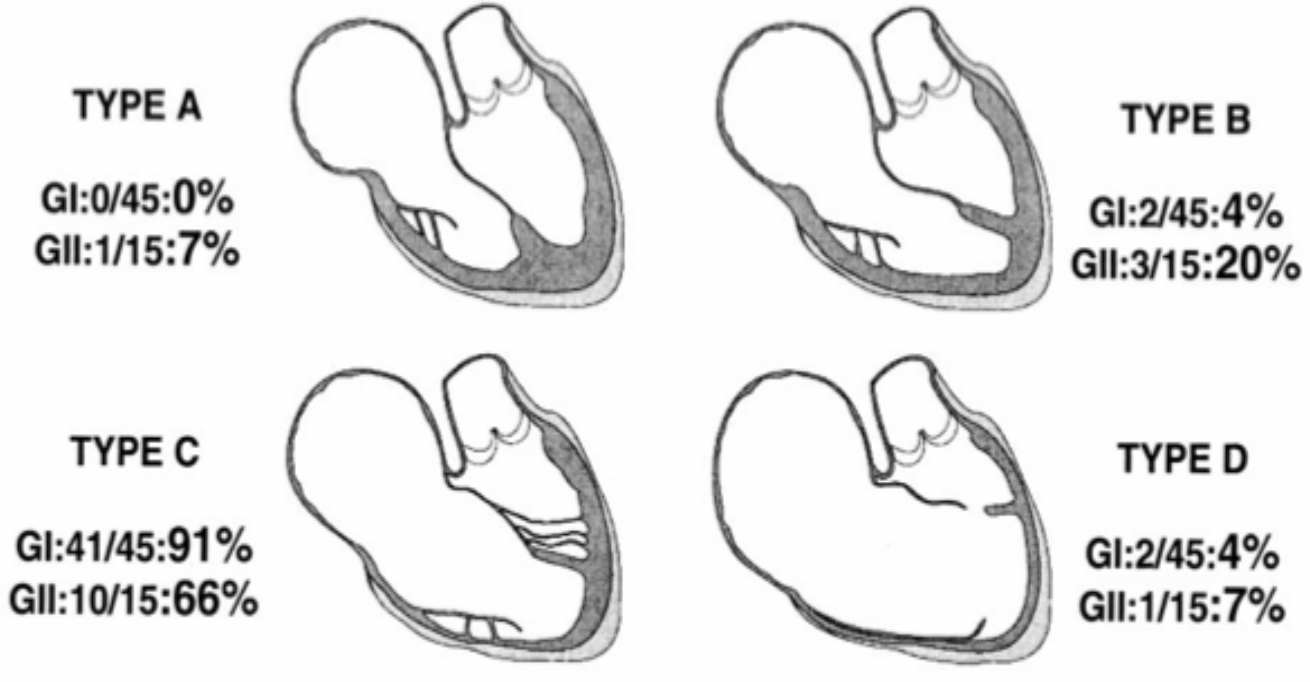
Improved functional assessment
RV and RA volumes
RV function
Delayed Enhancement

Objectively assessment
over time

Indications for repair

- Symptoms
- Deteriorating exercise capacity
- Heart failure (NYHA class III-IV)
- Cyanosis
- Paradoxical embolism
- Progressive RV/RA enlargement
- New onset arrhythmias

Carpentier classification

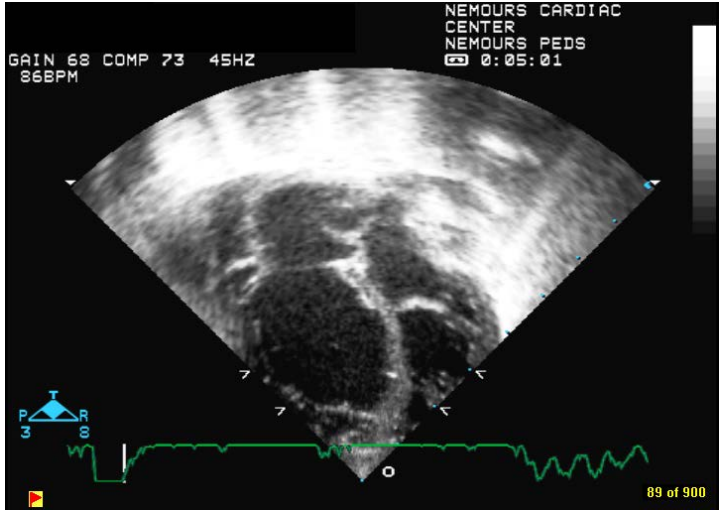


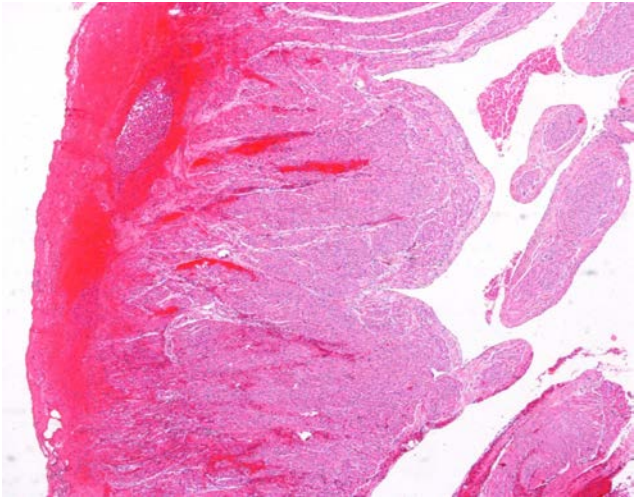
Anatomy is highly variable

Anatomic variability continues to be a challenge for the surgeon

Ventricular dysfunction

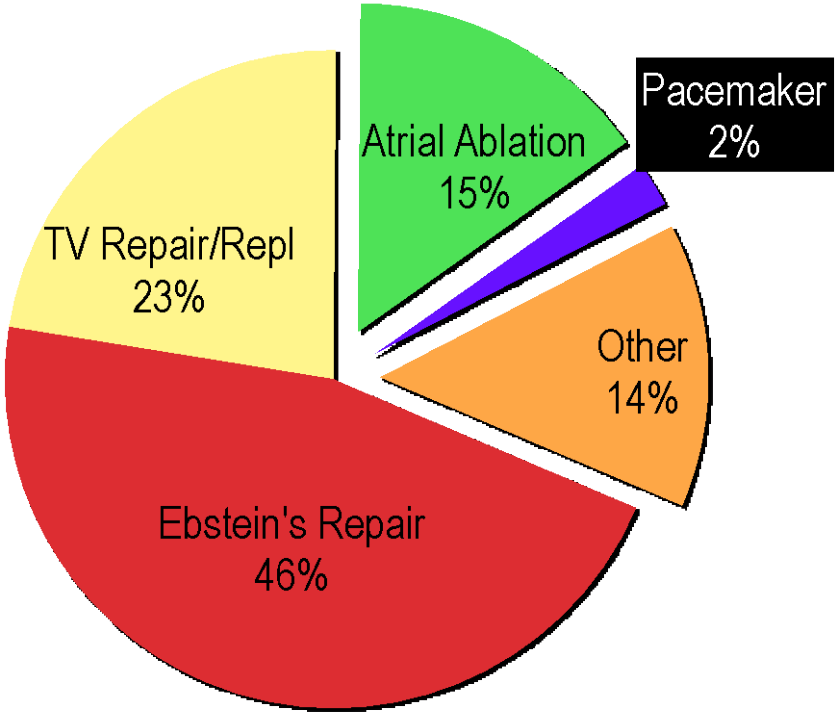
- Ineffective LV filling
  - due to “to and fro” flow into the right ventricle / right atrium
  - Increase right atrial capacitance
- “Pancaked” Left ventricle





RV myopathy
Thinned out and fibrotic muscle

STS database: common interventions



STS database analysis. Davies et al. Richard E Clark paper 2015

Center Experience

- 82 centers
- Median annual experience 1 case/yr
  - (IQR 0.5-1.8)
- Highest volume center: 8.3 cases/yr

- Neonates + infants
  - 63 centers
  - Median annual experience 0.5 cases/yr
    - (IQR 0.1-1.0, maximum 5.2)

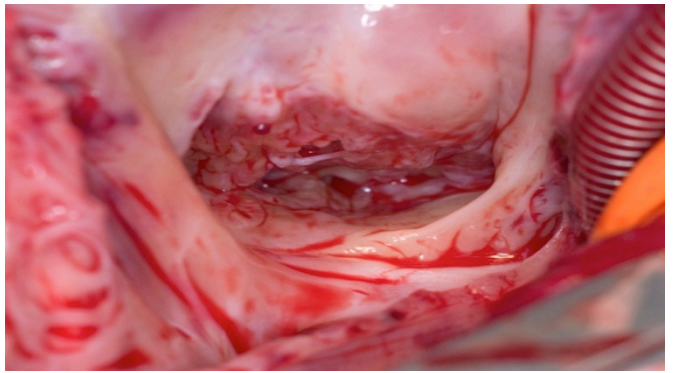
Arrhythmias are common among adult Ebstein anomaly

Preoperative Risk Factor	N (%)
Any Preoperative Risk Factor	29/89 (32.6%)
Acidosis	0/89 (0.0%)
Arrhythmia	19/89 (21.4%)
Shock	0/89 (0.0%)
Mechanical Ventilation	0/89 (0.0%)
Previous Cardiac Procedure	32/93 (34.4%)

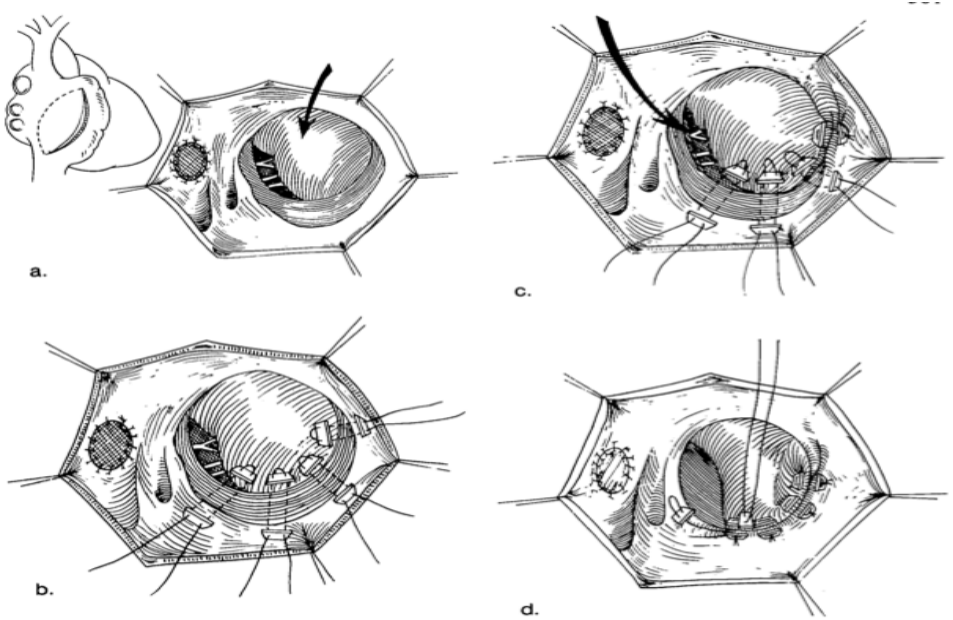
STS database analysis. Davies et al. Richard E Clark paper 2015

Surgical considerations

- Mechanism of TR, # of jets, location, valve morphology
- Delamination and tethering of each leaflet
- Leaflet edges, fenestrations
- Annular dilatation
- Size and function of the RV
- Ventricular septum and LV function
- Age of the patient



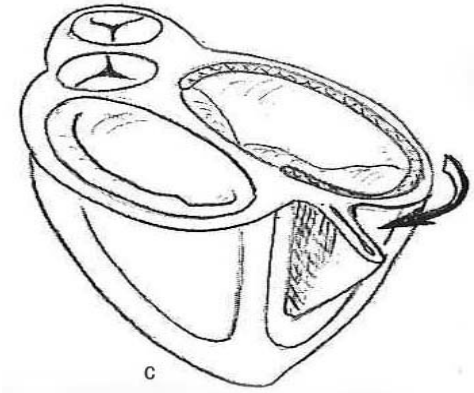
Danielson repair



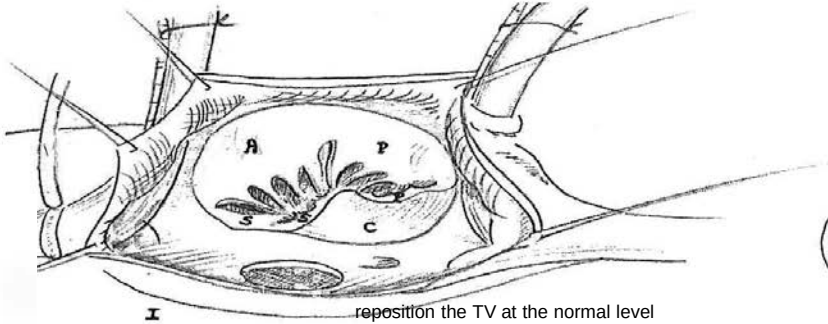
1972-1982
42 pts
Monocusp
Horizontal plication
No sutures on the IVS
81% repair
2 pts (5%) had a Glenn
7.1% mortality

Ann Thorac Surg.1982;196:499-504

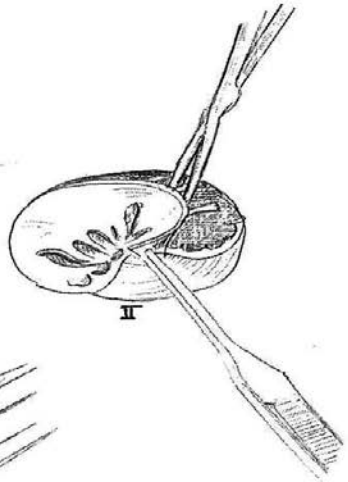
Carpentier Technique



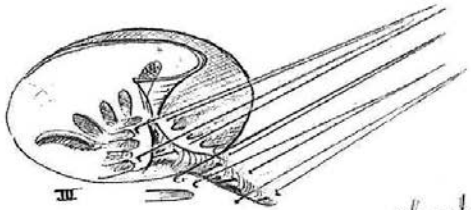
reconstruct a normal shaped RV
longitudinal plication of atrialized RV

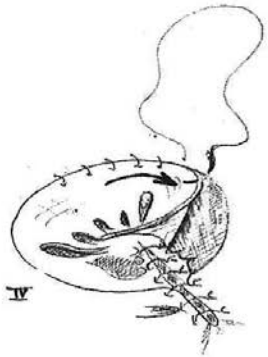


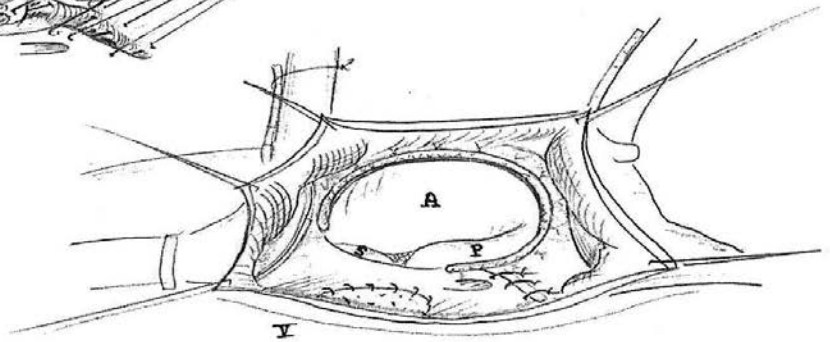
reposition the TV at the normal level



Monocuspid or bicuspid







Surgery for Ebstein's Anomaly: The Clinical and Echocardiographic Evaluation of a New Technique

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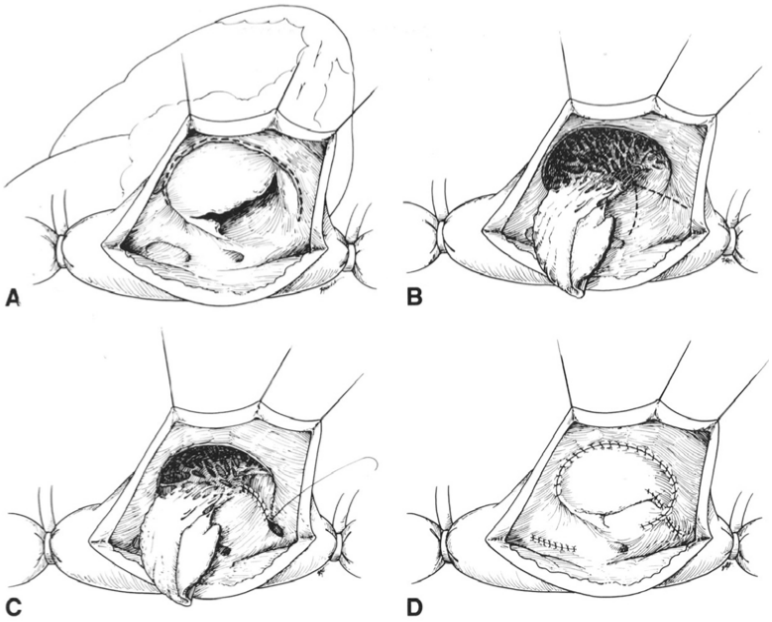
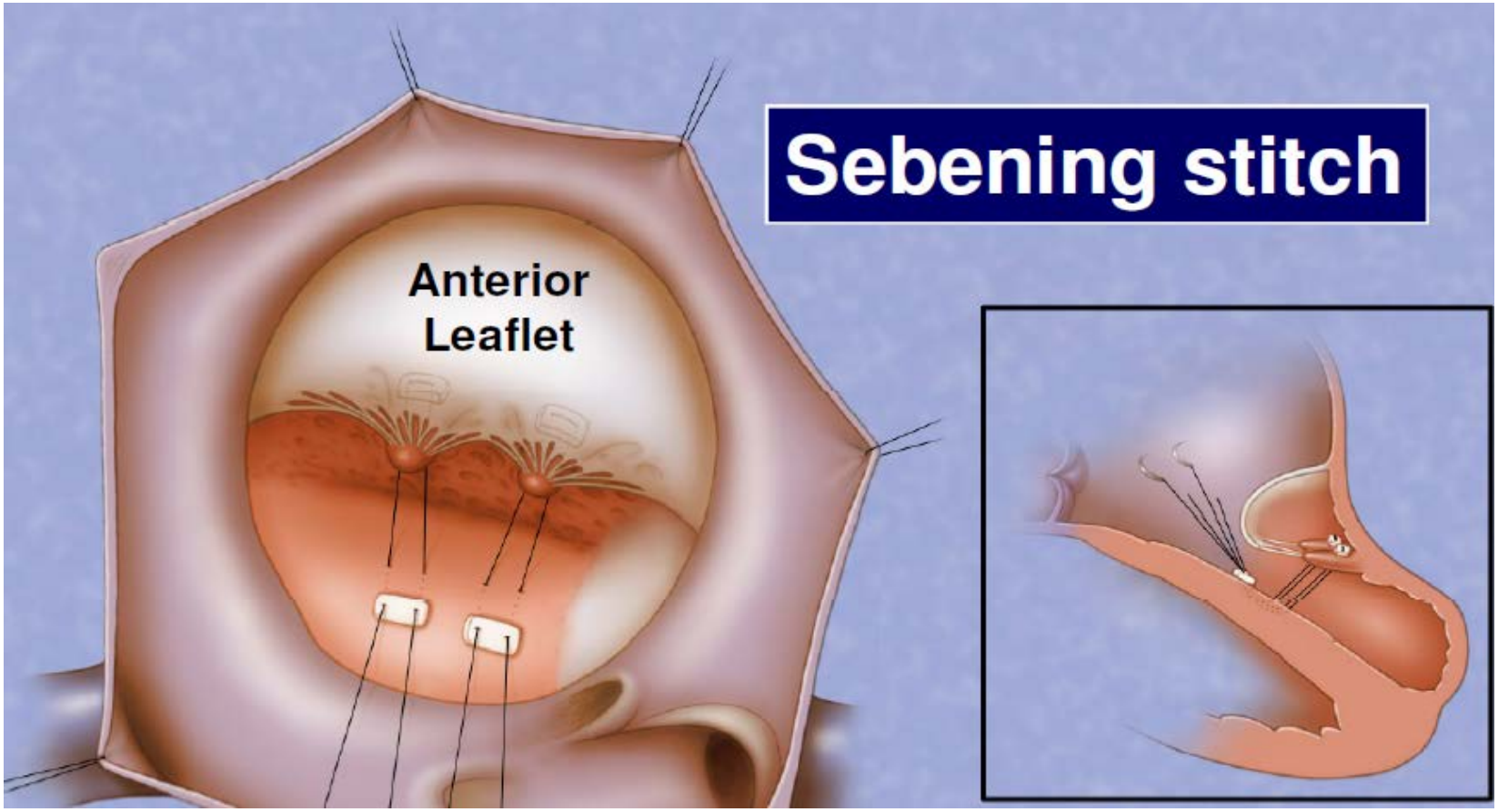
Rotterdam, The Netherlands



1988-1990
10 pts (4-44 yrs)
9 repairs
Bi leaflet valve
Vertical plication
No atrial reduction
No annuloplasty ring
No transection of PM
No heart block
No mortality
f/u 11.7 mo

JACC. 1991;17: 722-8

Sebening stitch



Common elements of TV repair

Basic strategy

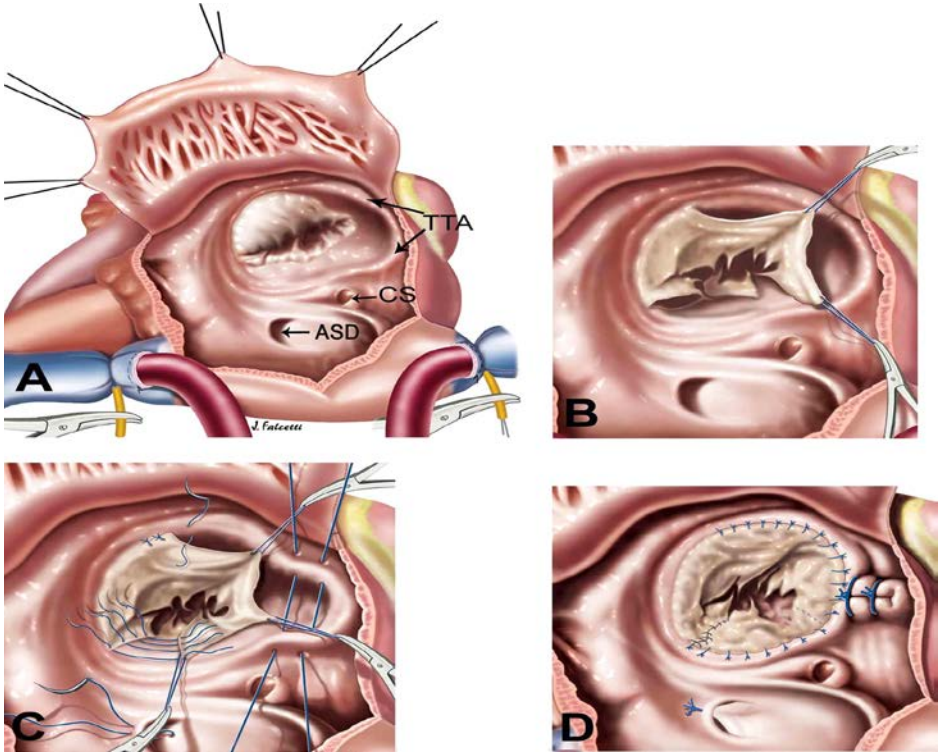
- Monocusp repair (allows coaptation against the ventricular septum)
  - based on anterior leaflet
  - degree of a RV and annular dilatation
  - tethering of the anterior leaflet
- Sebening stitch

Failures

- Due to focus on the annulus not on delamination
- Attempts to repair with poor valve anatomy
  - marked linear attachment
  - marked leaflet muscularization
  - complete absence of septal leaflet.
- Massive annular dilatation
- Older age
- Pulmonary HTN

The cone reconstruction of the tricuspid valve in Ebstein's anomaly. The operation: early and midterm results

José Pedro da Silva, MD[a,*], José Francisco Baumgratz, MD[b], Luciana da Fonseca, MD[b], Sônia Meiken Franchi, MD[a], Lilian Maria Lopes, MD[b], Gláucia Maria P. Tavares, MD[a], Andressa Mussi Soares, MD[a], Luiz Felipe Moreira, MD[a], Miguel Barbero-Marcial, MD[a]



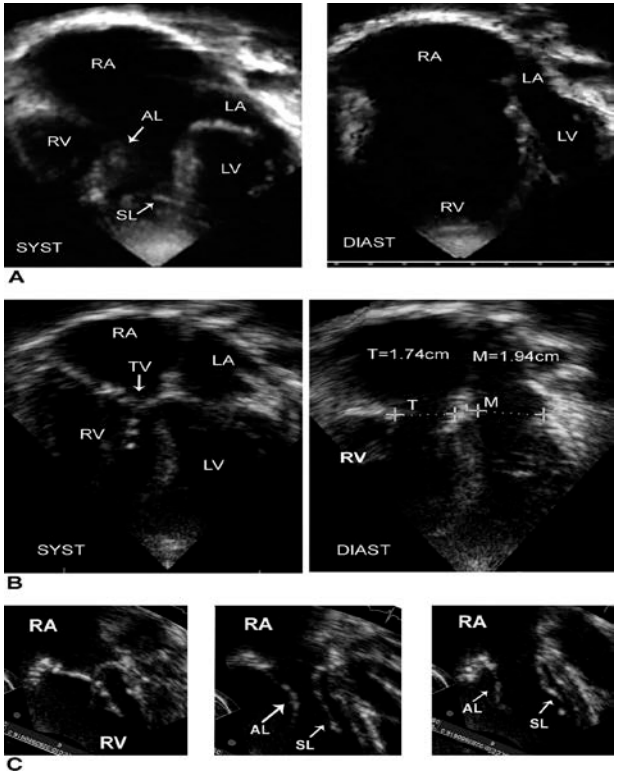
J Thorac Cardiovasc Surg 2007;133:215-223

Postoperative Outcome

One (2.5%) hospital death
and 1 late death

↓ in TR grade from
3.6 ± 0.5 to 1.2 ± 0.5
(*P* < .0001).

At a mean follow-up of 4 years:
Functional class (NYHA)
improved from 2.6 ± 0.7 to
1.2 ± 0.4 (*P* < .0001).



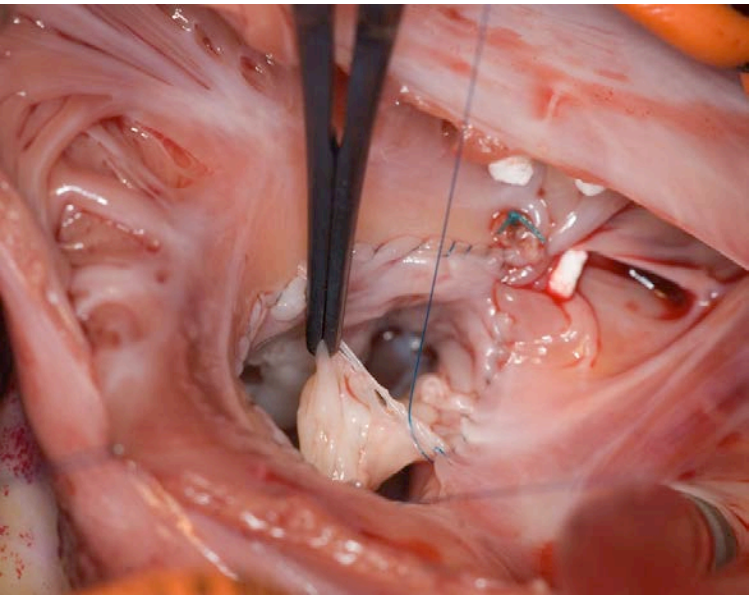
da Silva J. P. et al.; J Thorac Cardiovasc Surg 2007;133:215-223

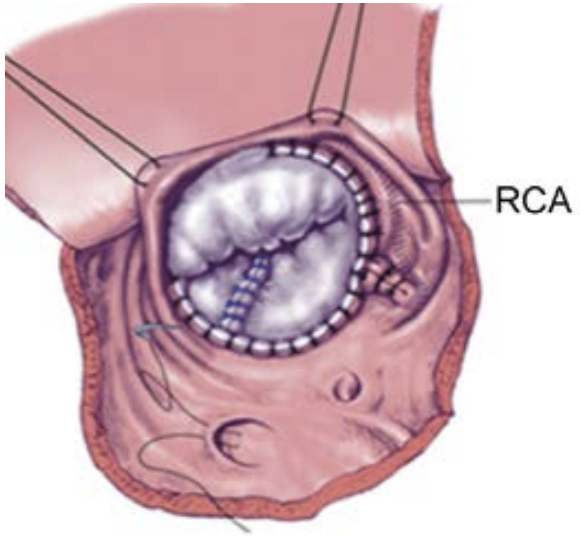
Valve inspection

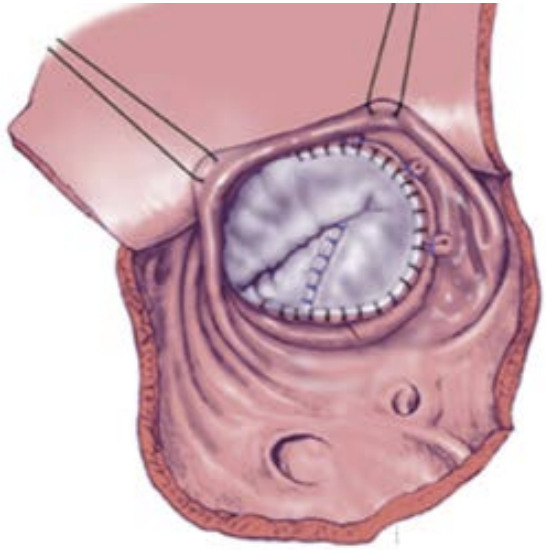
Leaflet dettachement

Mobilization of all leaflet tissue (septal)

Annular reduction

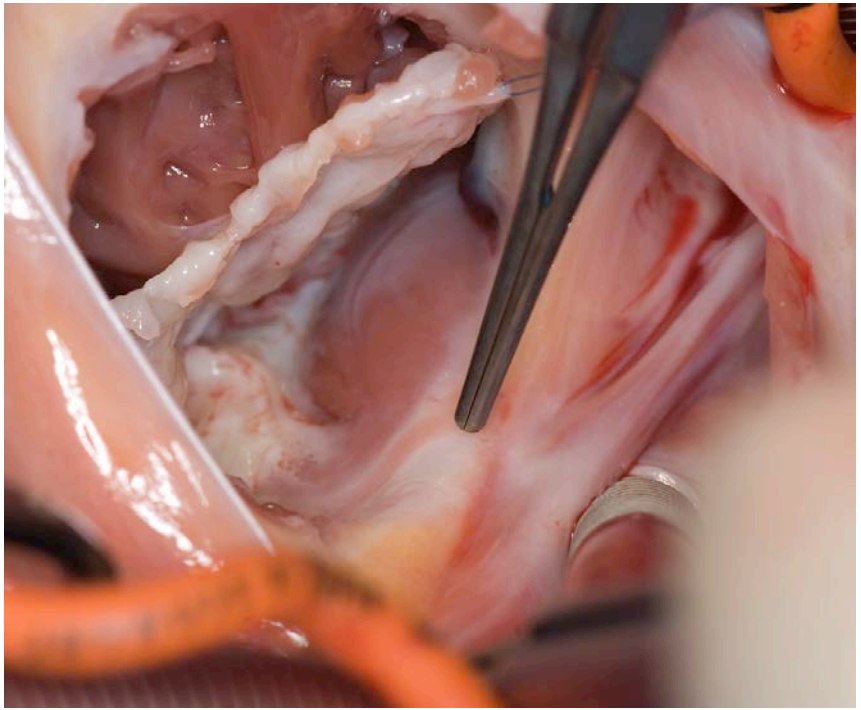


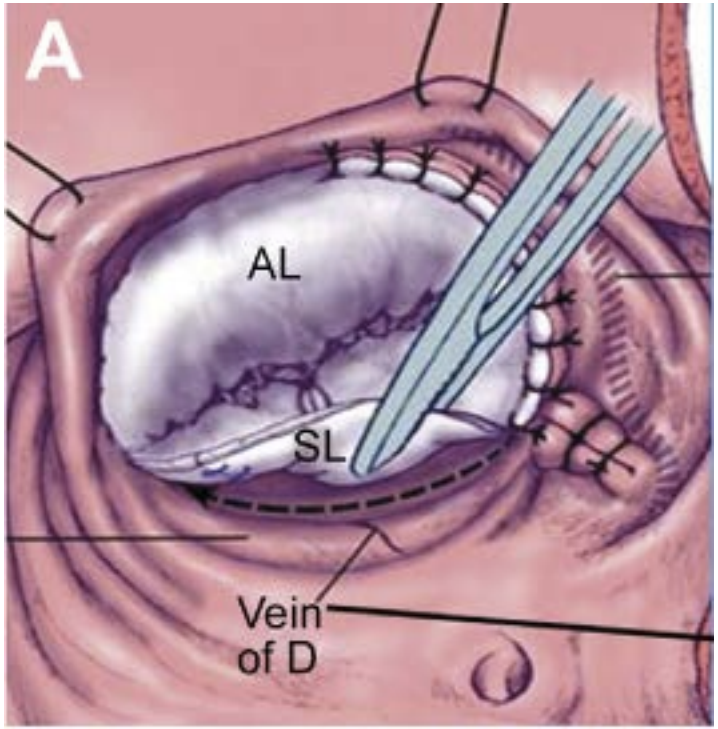




Ann Thorac Surg 2013;95:220 – 8

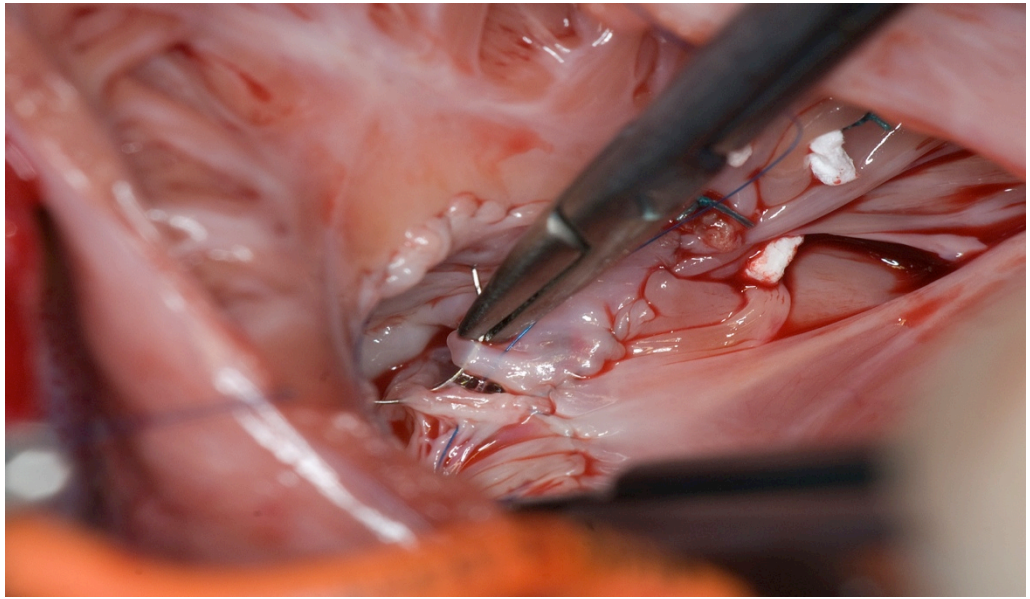
Reimplantation at the annular level



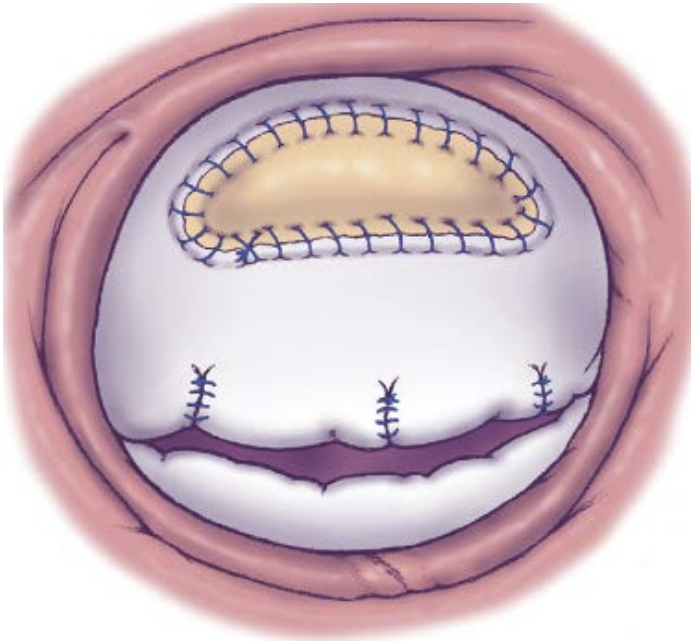


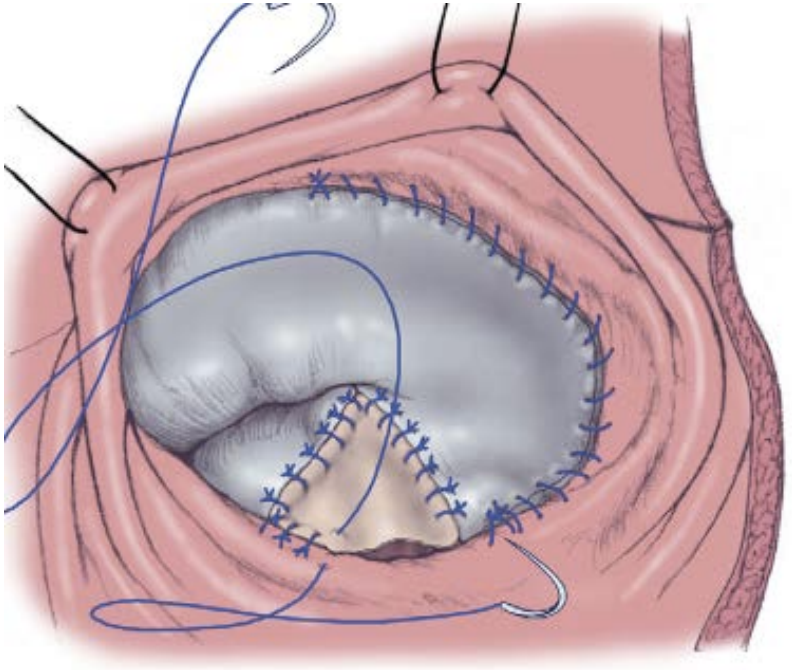
Ann Thorac Surg 2013;95:220 – 8

Incorporation of septal leaflet



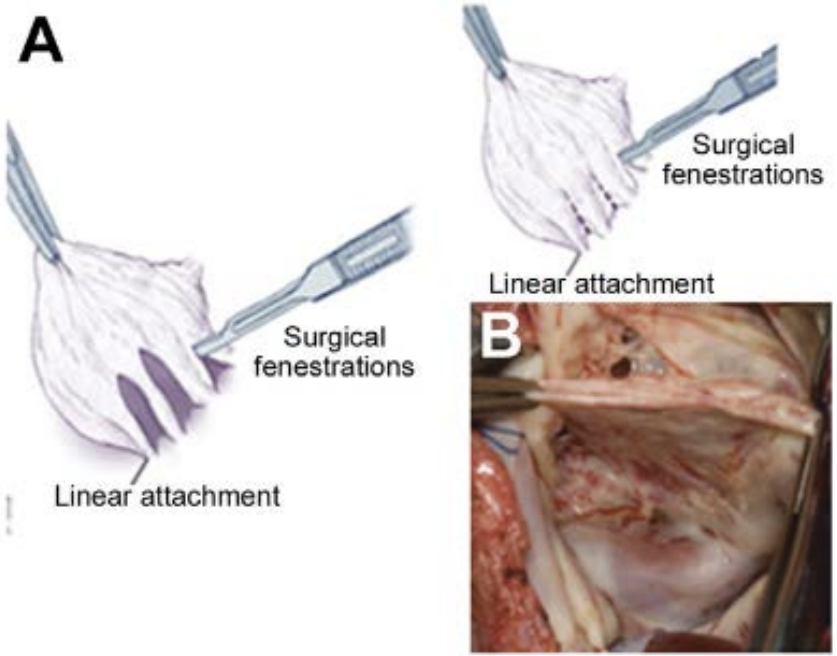
Leaflet augmentation





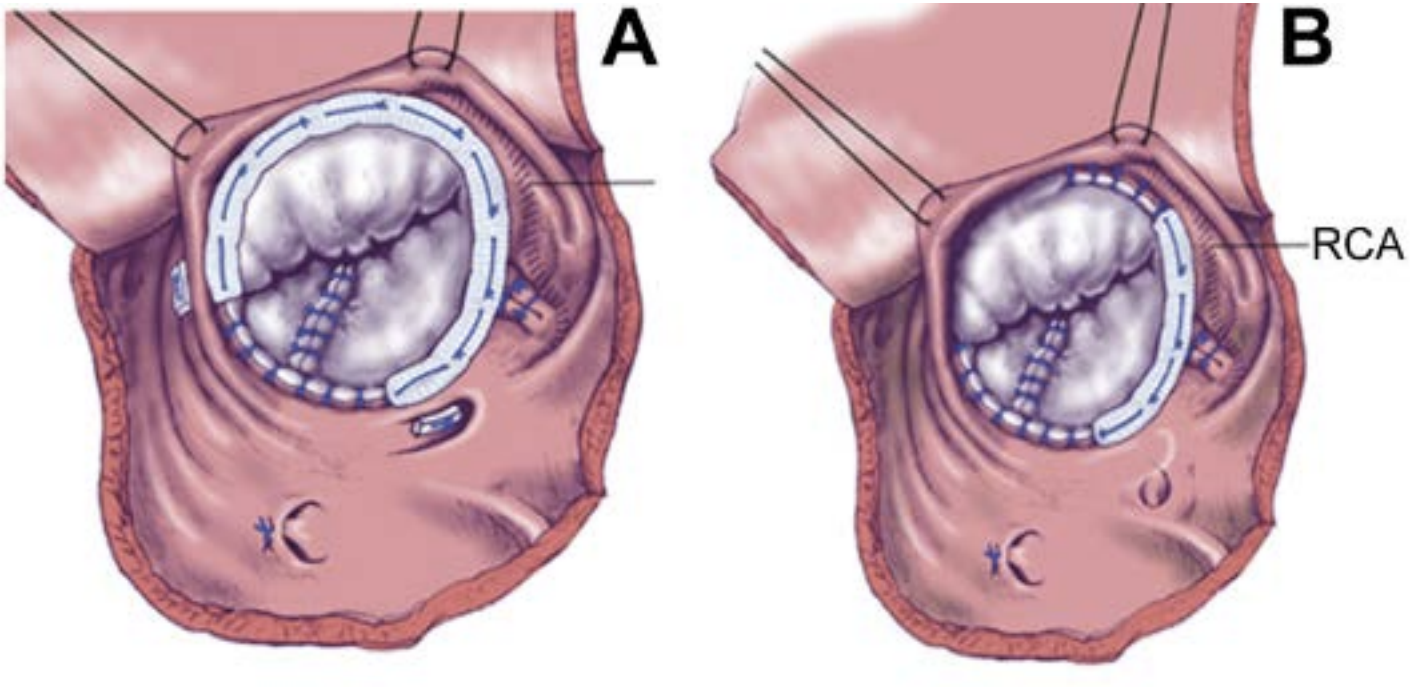
Ann Thorac Surg 2013;95:220 – 8

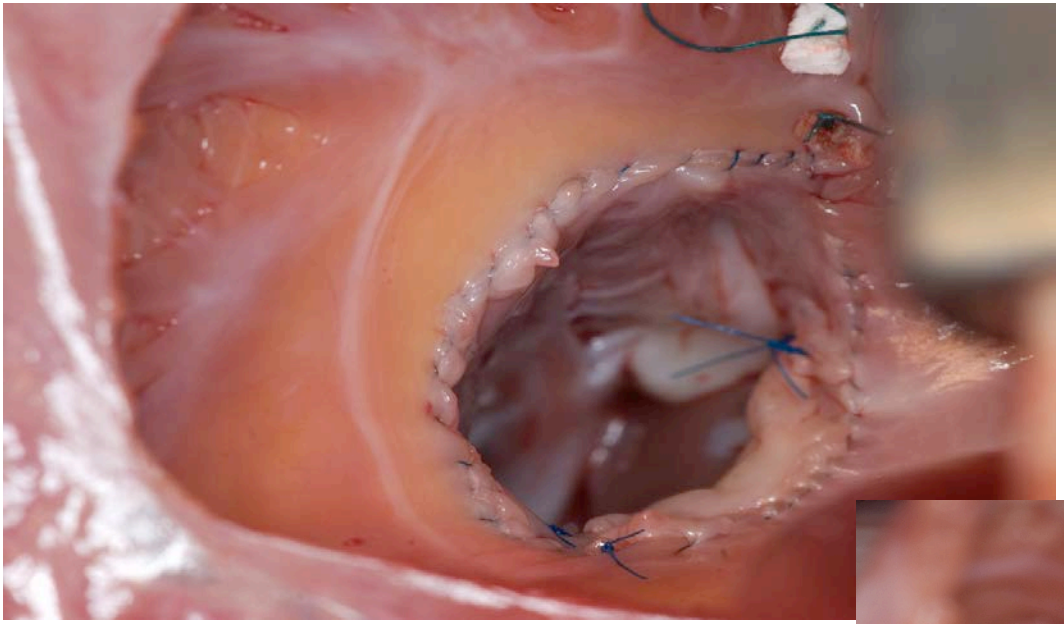
Creation of neochordae (fenestrations)



Ann Thorac Surg 2013;95:220 – 8

Annuloplasty





Circumferential leaflet coverage

Leads to improved competency

Surgical technique

- Circumferential leaflet tissue repair (360)
- Valve anchored at the true annulus
- Ringed annuloplasty
- Leaflet augmentation
- Autologous chordae
- Bidirectional Glenn

Anatomic Repair of Ebstein's Malformation: Lessons Learned With Cone Reconstruction

Joseph A. Dearani, MD, Sameh M. Said, MD, Patrick W. O'Leary, MD, Harold M. Burkhart, MD, Roxann D. Barnes, MD, and Frank Cetta, MD

- Commonly annular reduction is necessary/Use pledgeted suture
- RCA runs in the right AV groove. Avoid kinking or occlusion of the RCA
- Liberal use of the Sebening stitch. Avoid dimple on the RV free wall (patch augmentation of the anterior leaflet)
- Plication of the RV should be confined to the smooth non trabeculated inferior wall. Avoid the IVS (PDArt)
- Decision about the feasibility of a good durable repair should be made early.
- Repair rate for adults is ~80%

Important considerations

- Aortic occlusion and cardioplegic arrest for precise suture placement, avoidance of coronary or conduction tissue injury, and minimizing motion trauma during suture placement.
- The time required to perform EA repair can be lengthy, even in experienced hands, and the consequences of prolonged operation in a patient with depressed ventricular function may not be prudent.

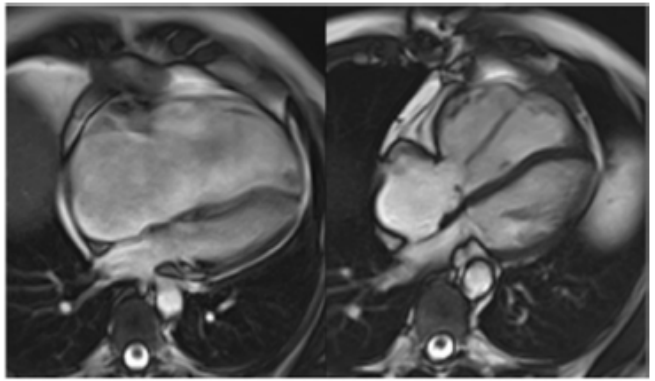
Cone reconstruction for Ebstein's anomaly: Patient outcomes, biventricular function, and cardiopulmonary exercise capacity

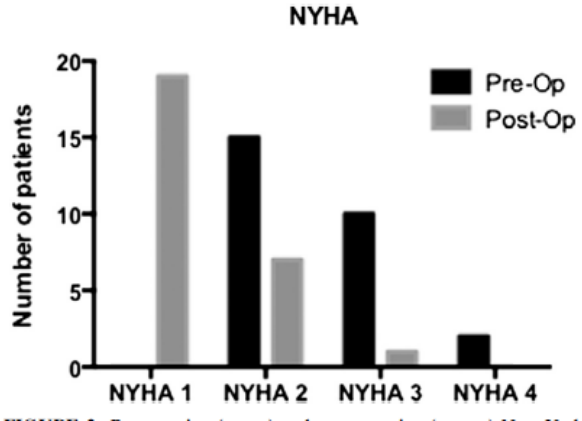
Michael Ibrahim, MD, PhD,[a,b] Victor T. Tsang, MD, FRCS,[a,b,c] Maryanne Caruana, MD,[d] Marina L. Hughes, DPhil, FRACP,[d,e] Synetta Jenkyns, BD,[e] Elodie Perdreau, MD,[e] Alessandro Giardini, MD,[c,e] and Jan Marek, MD, PhD[c,e]

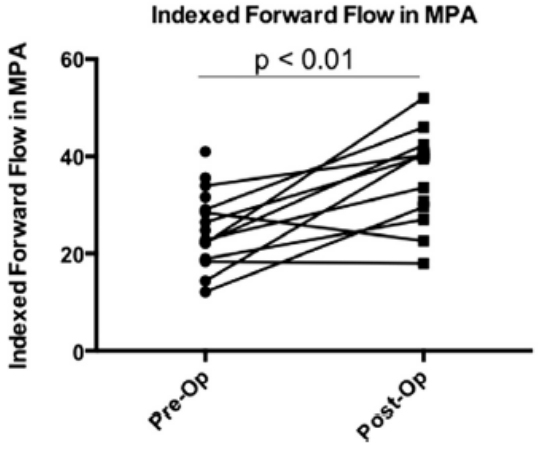
TABLE 2. Cardiac magnetic resonance imaging analysis of effect of cone reconstruction on right ventricle (RV) and left ventricle (LV) volumes

	Preoperative	Postoperative	*P* value
RV end-systolic volume	112.1 ± 80.6	91.0 ± 45.3	.54
RV end-diastolic volume	166 ± 66.3	145.9 ± 56.1	.24
LV end-systolic volume	19.8 ± 8.9	22.4 ± 10.4	.25
LV end-diastolic volume	49.4 ± 14.4	60.14 ± 14.5	**.006**

All values are indexed for body surface area. Boldface type indicates statistical significance. *RV*, Right ventricle; *LV*, left ventricle.







Summary

- Ebstein anomaly exhibits a broad range of anatomic features commonly leading to severe TR
- Due to morphologic variability, no perfect repair for all patients with EA
- The Cone reconstruction is the only technique that provides circumferential leaflet coverage and is highly effective to restore valve competency.
- Reduced right ventricular function continues to be a challenge, as is the need for reoperation for recurrent tricuspid regurgitation
- Steep learning curve requires institutional and surgeon experience for optimal outcomes
- Improved results should influence timing of repair.
- When severe RV dysfunction is present, TVR should be considered early.

Muchas Gracias!

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September 21-22, 2017 | Cartagena, Colombia

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Thank You

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Nemours. Alfred I. duPont Hospital for Children

- Ebstein anomaly accounts for 1% of all congenital heart disease. It is a right ventricular myopathy with failure of tricuspid valve delamination and highly variable tricuspid valve morphology that usually results in severe regurgitation. It is the only congenital heart lesion that has a range of clinical presentations, from the severely symptomatic neonate to an asymptomatic adult. Neonatal operation has high operative mortality, whereas operation performed beyond infancy and into adulthood has low operative mortality. Late survival and quality of life for hospital survivors are excellent for the majority of patients in all age brackets. Atrial tachyarrhythmias are the most common late complication. There have been more techniques of tricuspid repair reported in the literature than any other congenital or acquired cardiac lesion. This is largely due to the infinite anatomic variability encountered with this anomaly. The cone reconstruction of Ebstein anomaly can achieve near anatomic restoration of the tricuspid valve anatomy. Early and intermediate results with these repairs are promising. Reduced right ventricular function continues to be a challenge for some patients, as is the need for reoperation for recurrent tricuspid regurgitation. The purpose of this article is to outline the current standard of care for diagnosis and treatment of Ebstein anomaly and describe innovative strategies to address poor right ventricular function and associated right-sided heart failure.

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Subtitle

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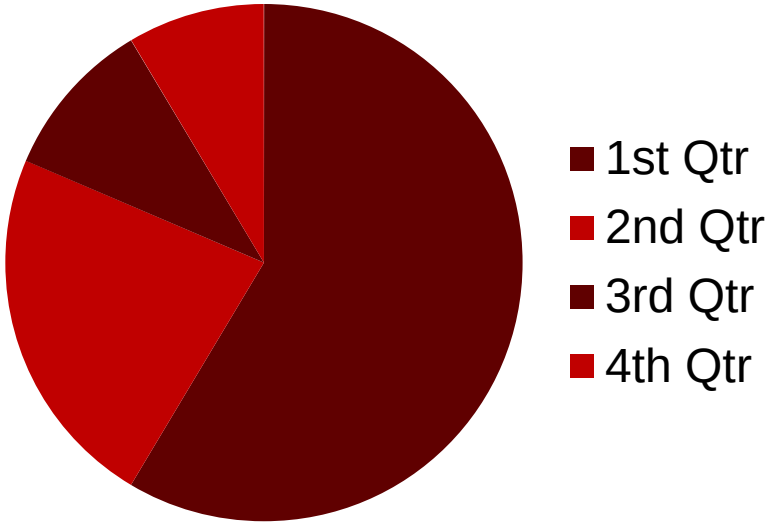
- Bullet number two
  - Sub-bullet number three
  - Sub-bullet number four
- Bullet number five

Description of Graph or Table



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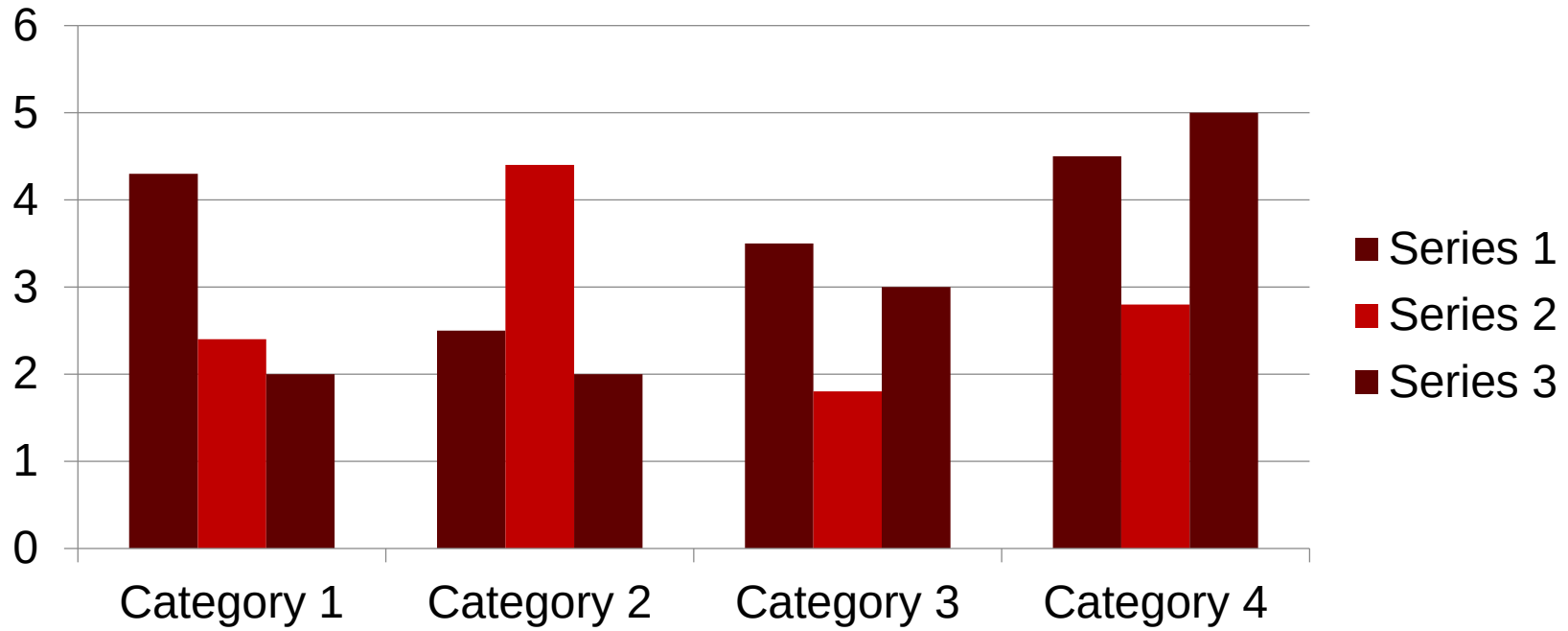

SLIDE TITLE HEADLINE

Bi-directional cavopulmonary shunt associated with ventriculo and valvuloplasty in Ebstein's anomaly: benefits in high risk patients[1]

S. Chauvaud*, J.F. Fuzellier, A. Berrebi, P. Lajos, J.P. Marino, S. Mihaileanu, A. Carpentier

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Received 8 December 1997; revised version received 9 February 1998; accepted 16 February 1998

- RV unloading, decreases RV dilatation
- Improves LV/RV interaction
- Increases effective LV filling (Cardiac output)
- Allows tighter annuloplasty
- Not feasible in neonates and youg infants

Chaveaud et al. EJCTS 1998;13: 514-19