

Operative Modifications for Ebstein Anomaly

Han K Park

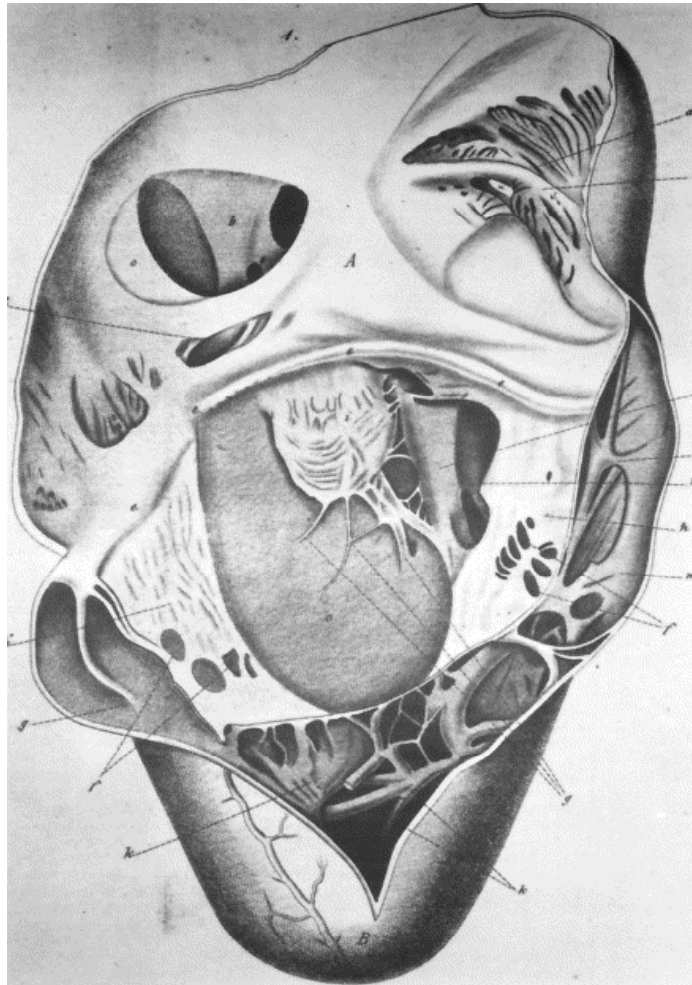
Department of Cardiovascular Surgery
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Wilhelm Ebstein



Ebstein's Anomaly

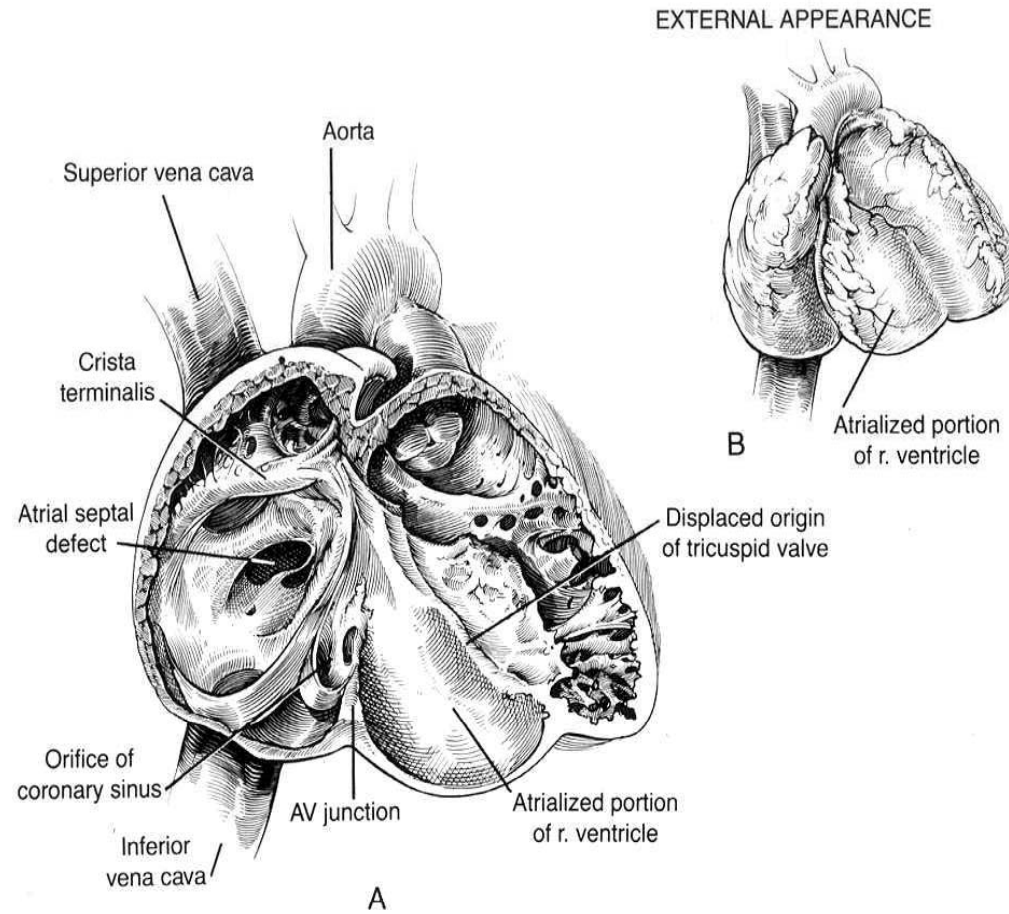
Ebstein's Anomaly



Characteristics of Ebstein's Anomaly

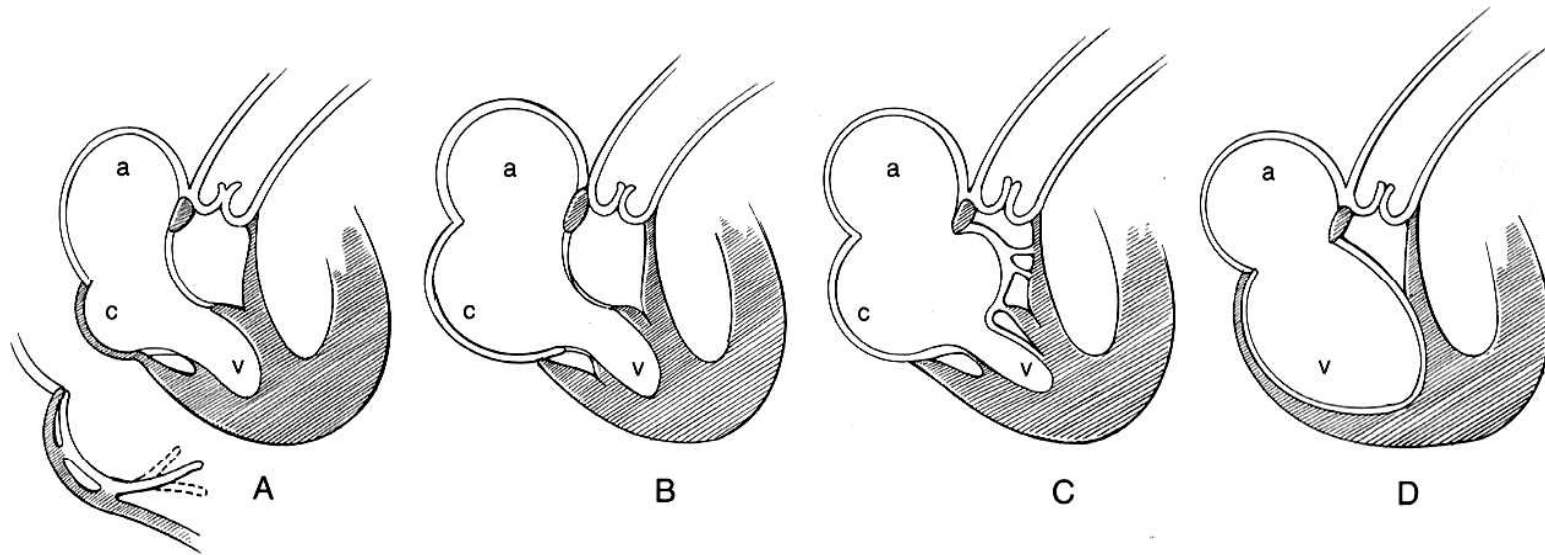
- Adherence of the septal and posterior leaflets to the underlying myocardium (failure of delamination)
- Downward displacement of the functional annulus (septal > posterior > anterior)
- Dilatation of the "atrialized portion of the RV, with various degrees of hypertrophy and thinning of the wall
- Redundancy, fenestration, and tethering of the anterior leaflet
- Dilatation of the right atrioventricular junction (true tricuspid annulus)

Pathologic Anatomy



Pathologic Anatomy

Carpentier's Anatomic type of Ebstein's Anomaly



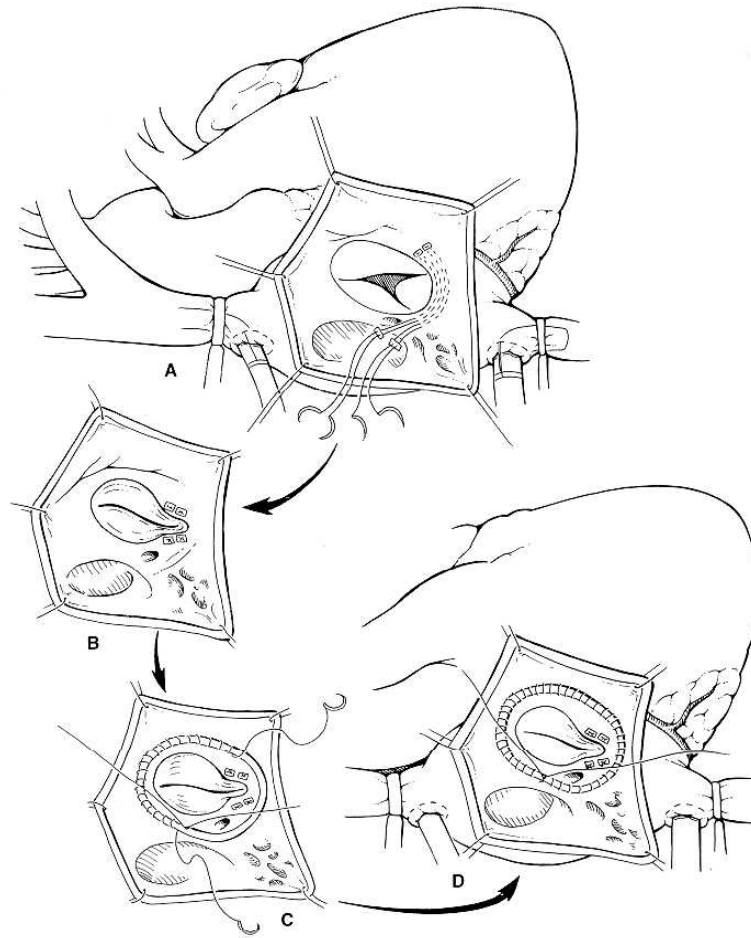
Surgical Repair

Barnard and Schrire

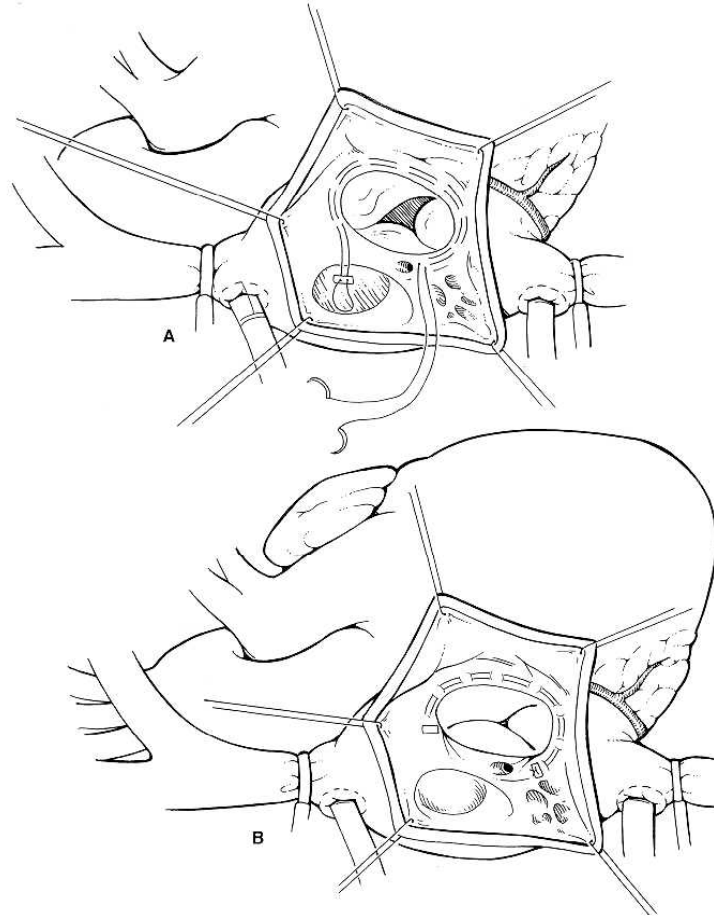
Surgical correction of Ebstein's
malformation with prosthetic valve

Surgery 1963;54:302

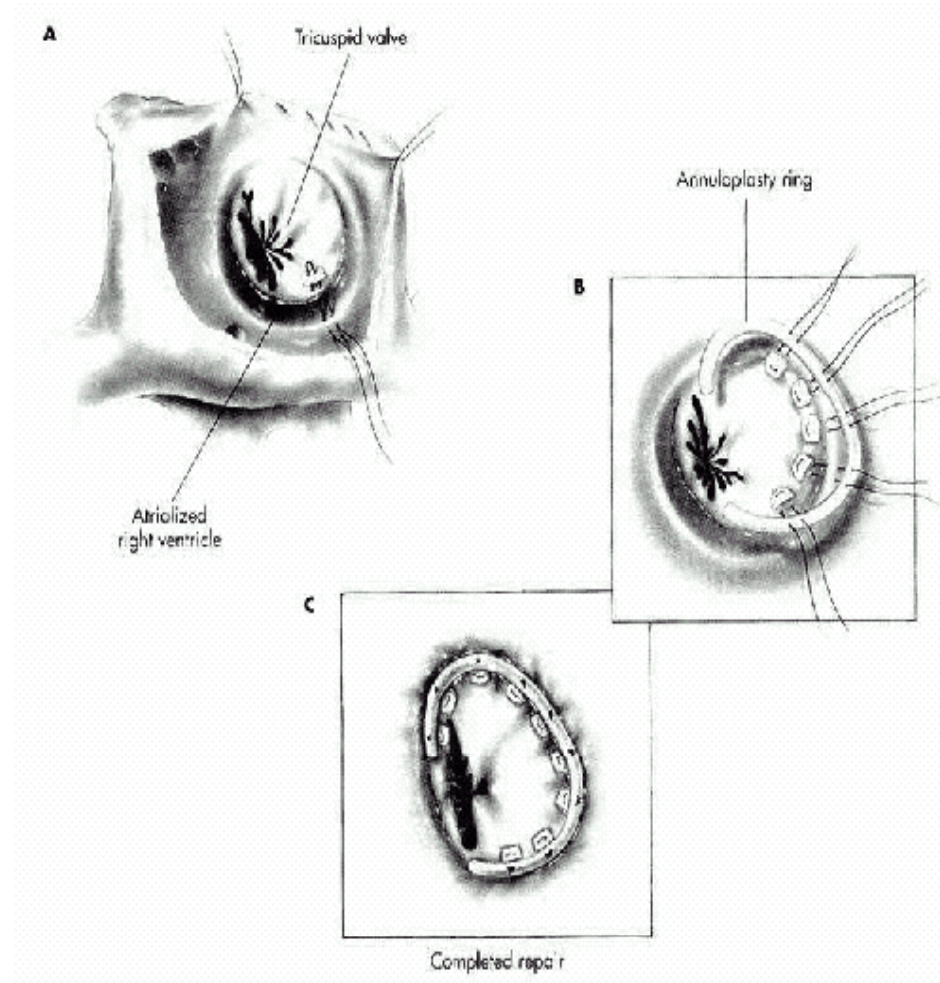
Kay Annuloplasty with Ring



De Vega Annuloplasty



Carpentier Ring Annuloplasty



Operative Management

1. EP mapping for localization of accessory conduction pathways in patients with ventricular pre-excitation
2. Closure of any atrial septal communications
3. Correction of previously placed shunts and any associated anomalies such as VSD, PS, and PDA
4. Performance of any indicated anti-arrhythmia procedures such as surgical division of accessory conduction pathways, cryoablation of AVNRT, or right side maze procedure
5. Consideration of plication of the atrialized RV
6. Reconstruction of TV when feasible, or valve replacement
7. Right reduction atrioplasty

Lellehei CW, Kalke BR, Carlson RG

Evolution of corrective surgery for Ebstein's
anomaly

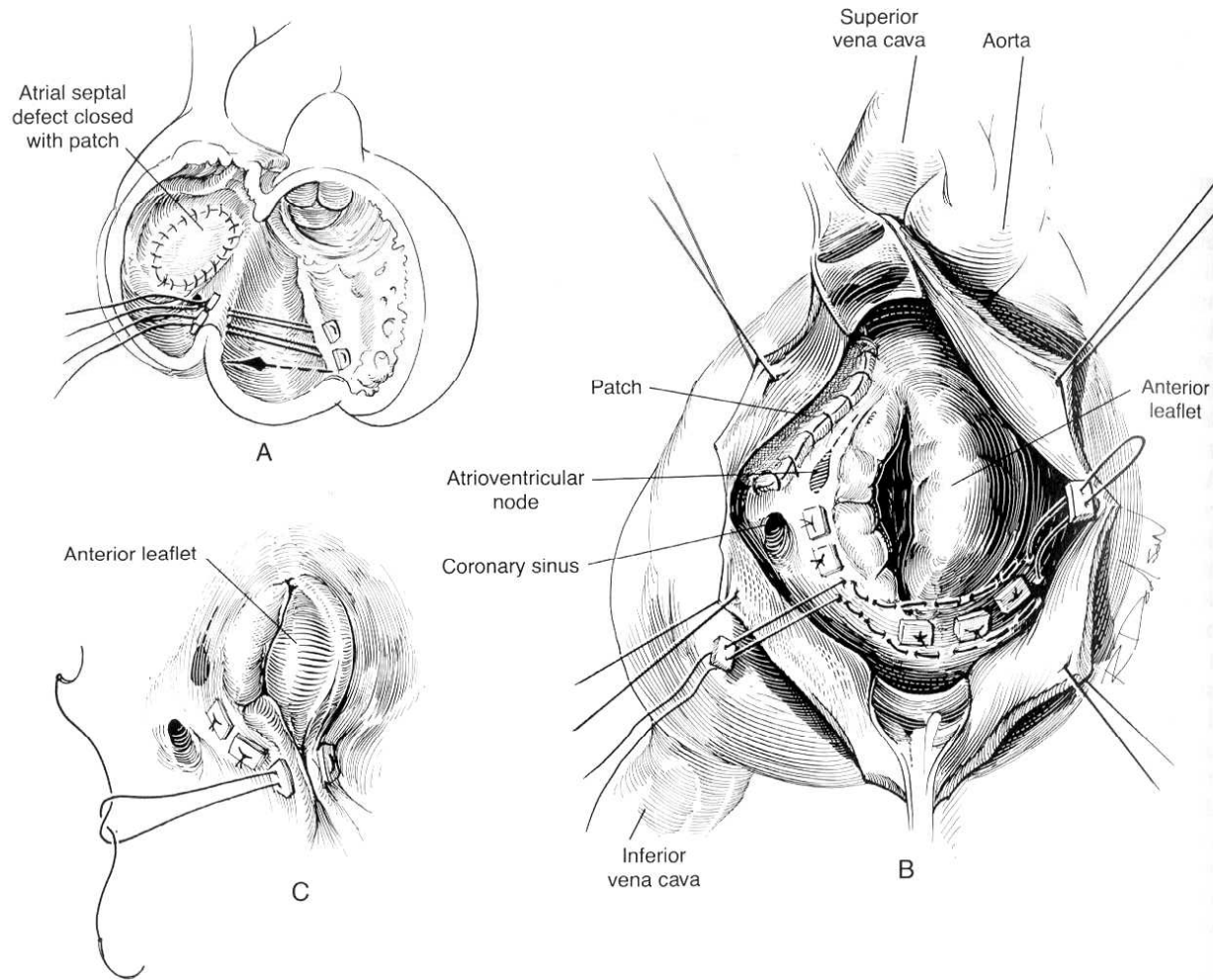
Circulation 1967;35:111

Hardy KL, Roe BB

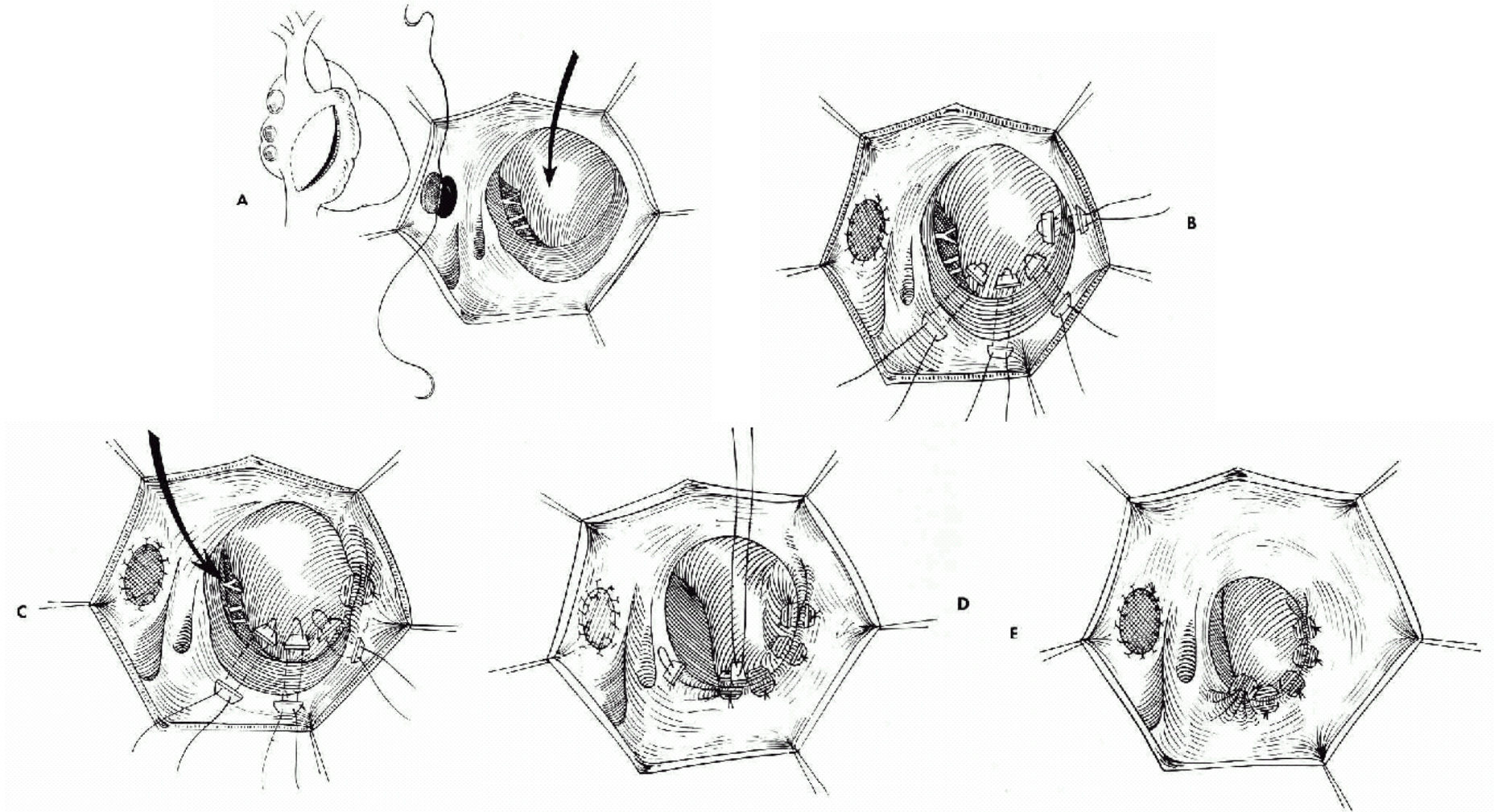
Ebstein's anomaly: Further experience with
definitive repair

J Throac Cardiovasc Surg 1969;58:553

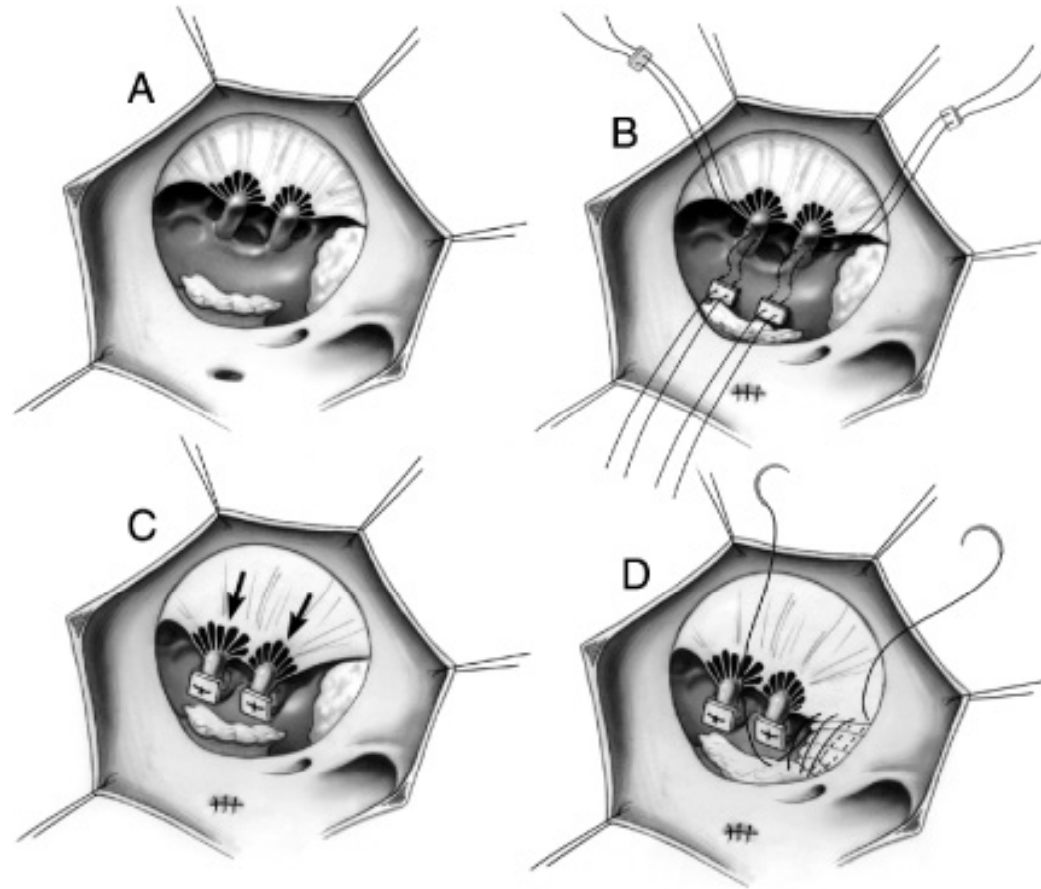
Danielson Repair of Ebstein's Anomaly



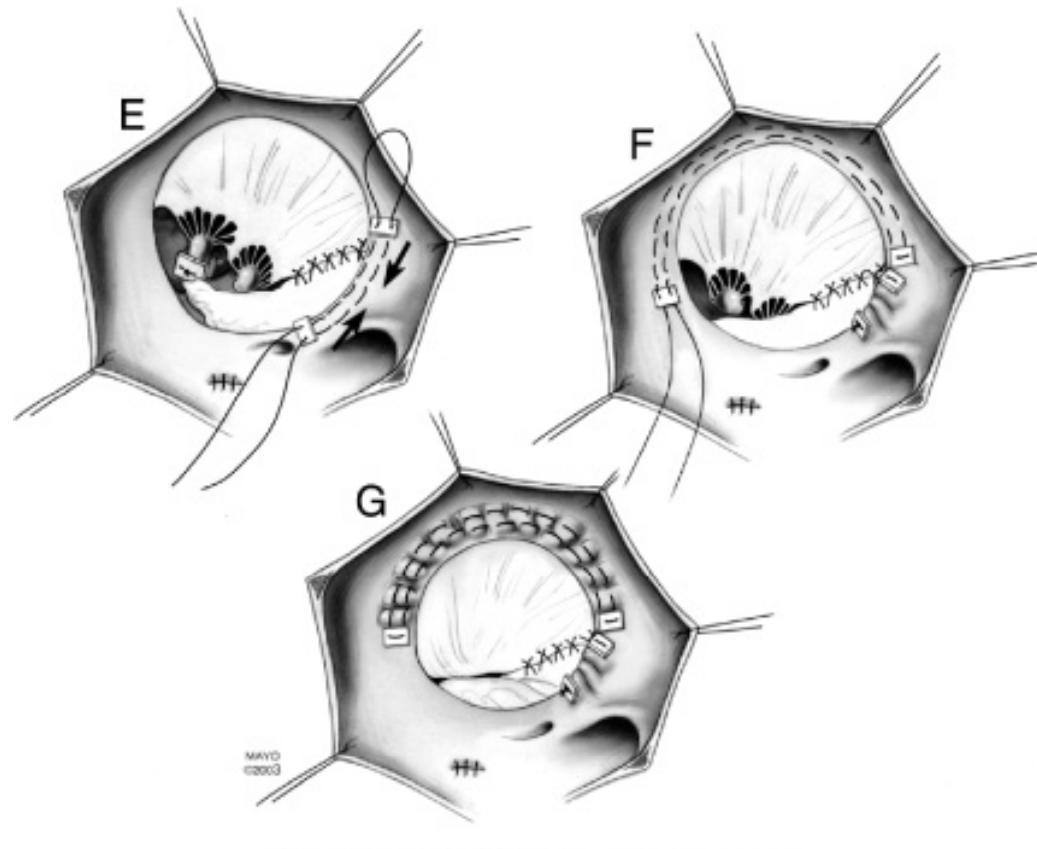
Danielson Repair



Danielson Repair



Danielson Repair



Danielson Repair

- 1972 ~ 2005
- 540 consecutive patients (at Mayo Clinic)
- Age: median 20 yr (2 mo ~ 79 yr)
- TV reconstruction 34.4%
- TV replacement: 65.6%
- Early death: 5.4%
- Late death: 7.6% follow up 7.1 (~25) yrs

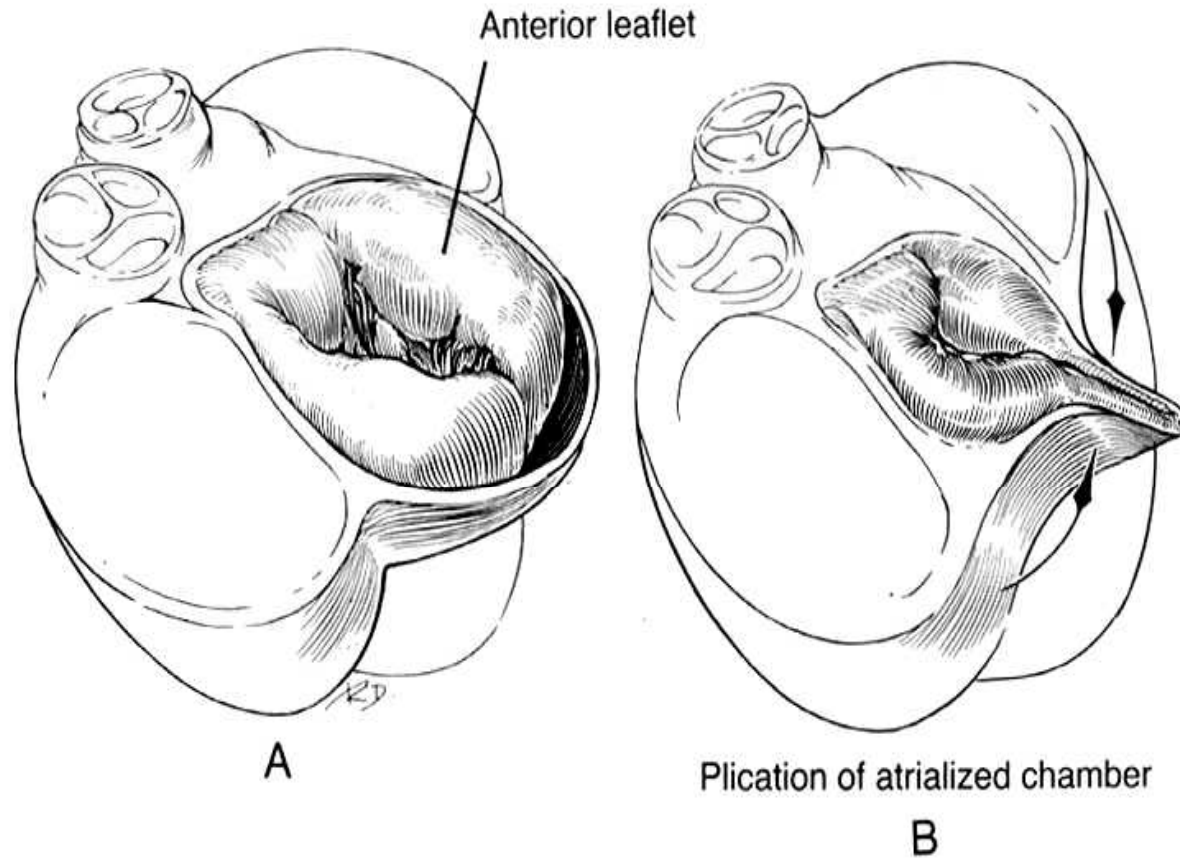
A new reconstructive operation for Ebstein's anomaly of the tricuspid valve

Carpentier et al.

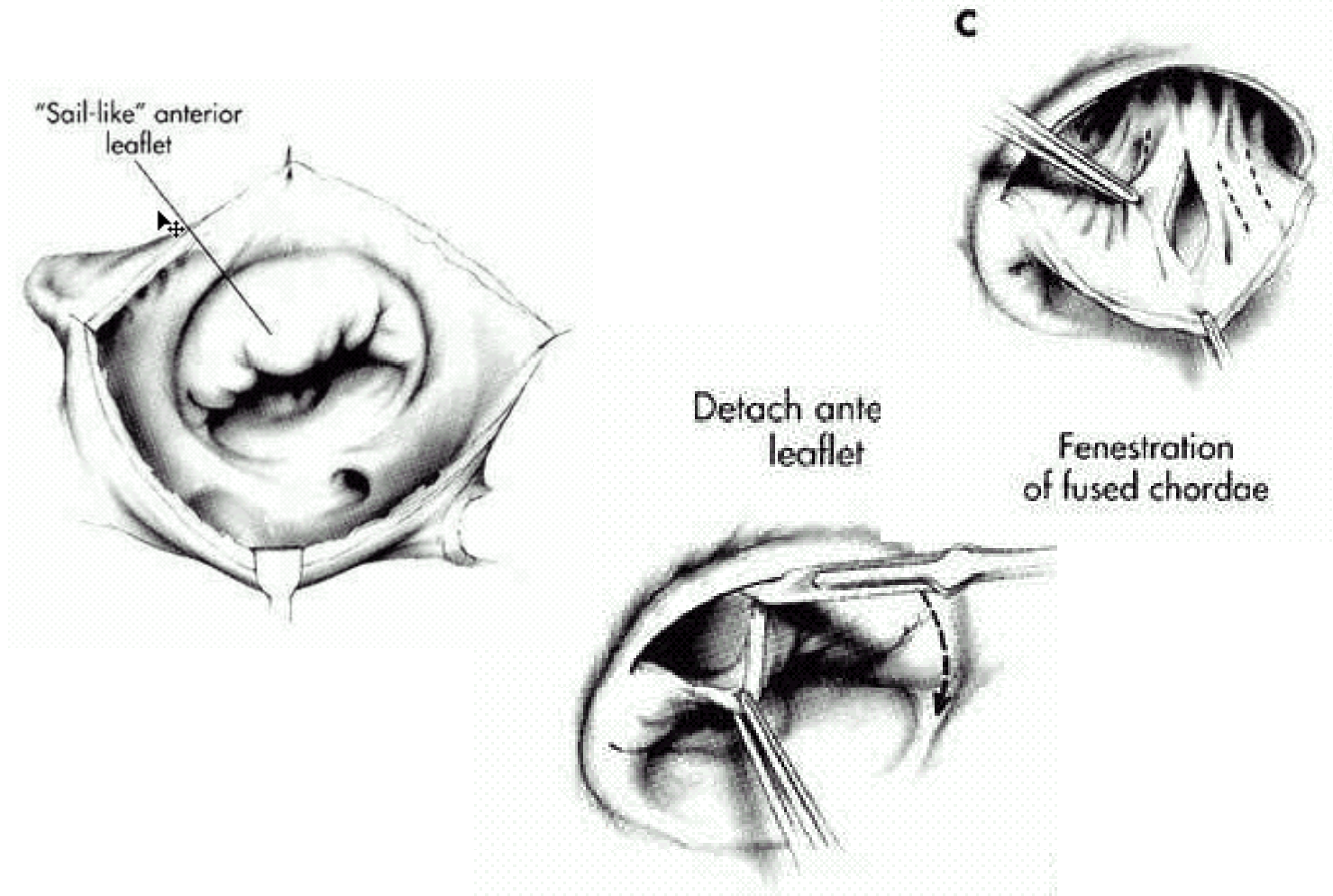
J Thorac Cardiovas Surg 1988;96:92

- Temporary detachment of anterior leaflet
- Longitudinal plication of the atrialized RV
- Reposition of the anterior and posterior leaflet to cover the orifice area at the normal level
- Remodeling and reinforcement of the tricuspid annulus with a prosthetic ring

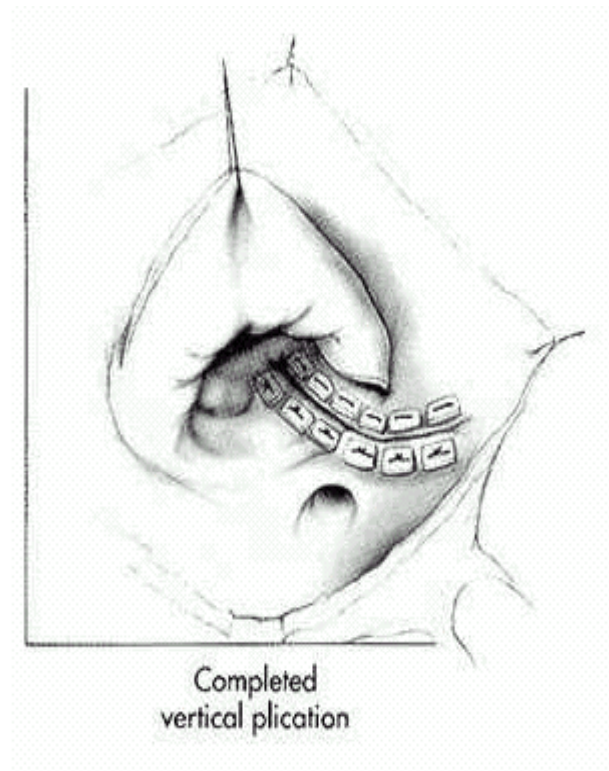
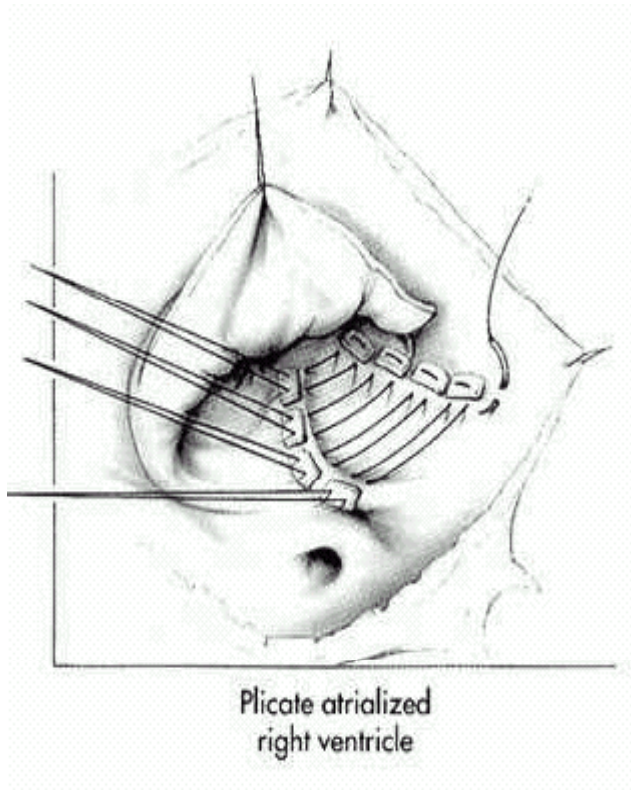
Carpentier Repair



Carpentier's Repair

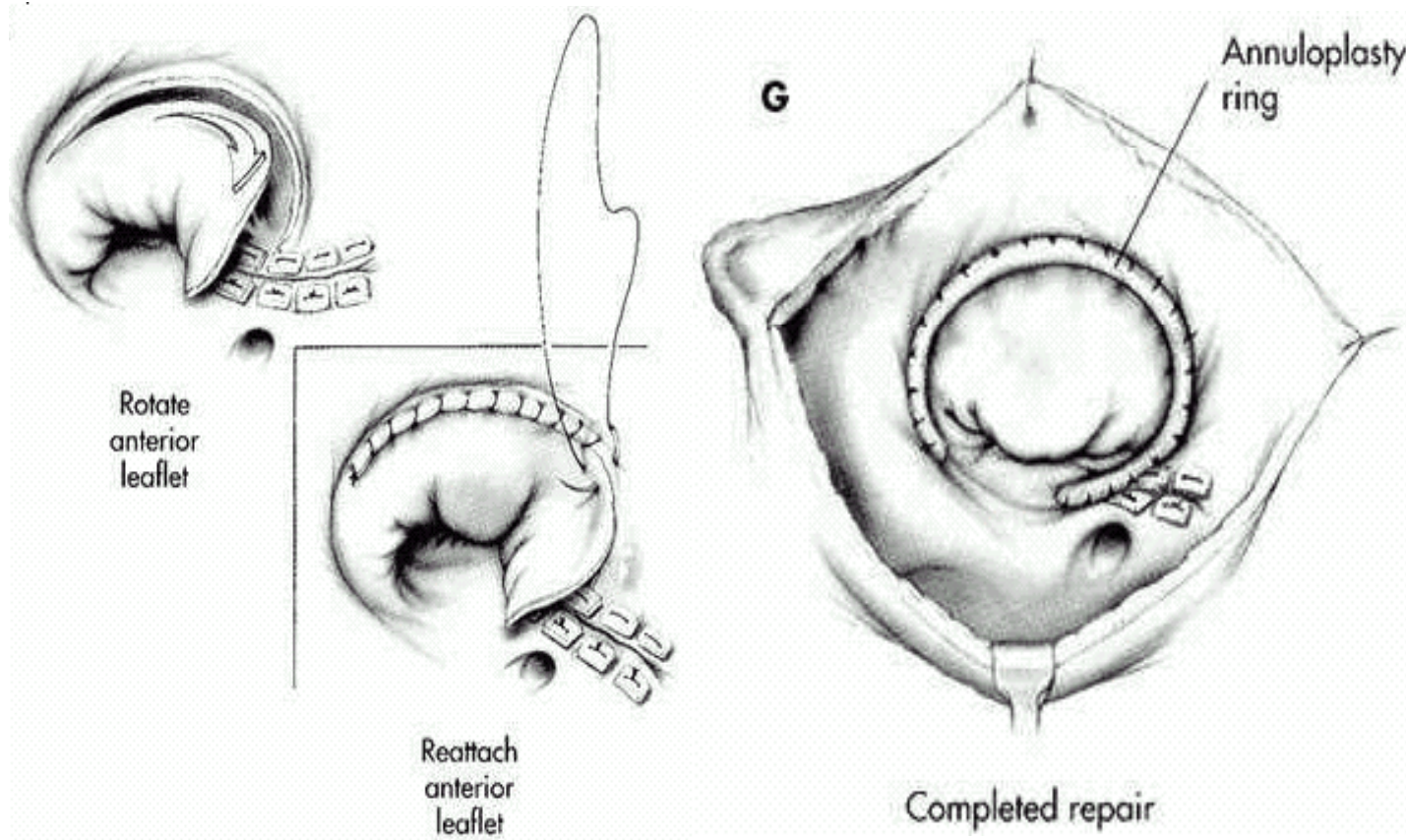


Carpentier's Sliding TV Annuloplasty

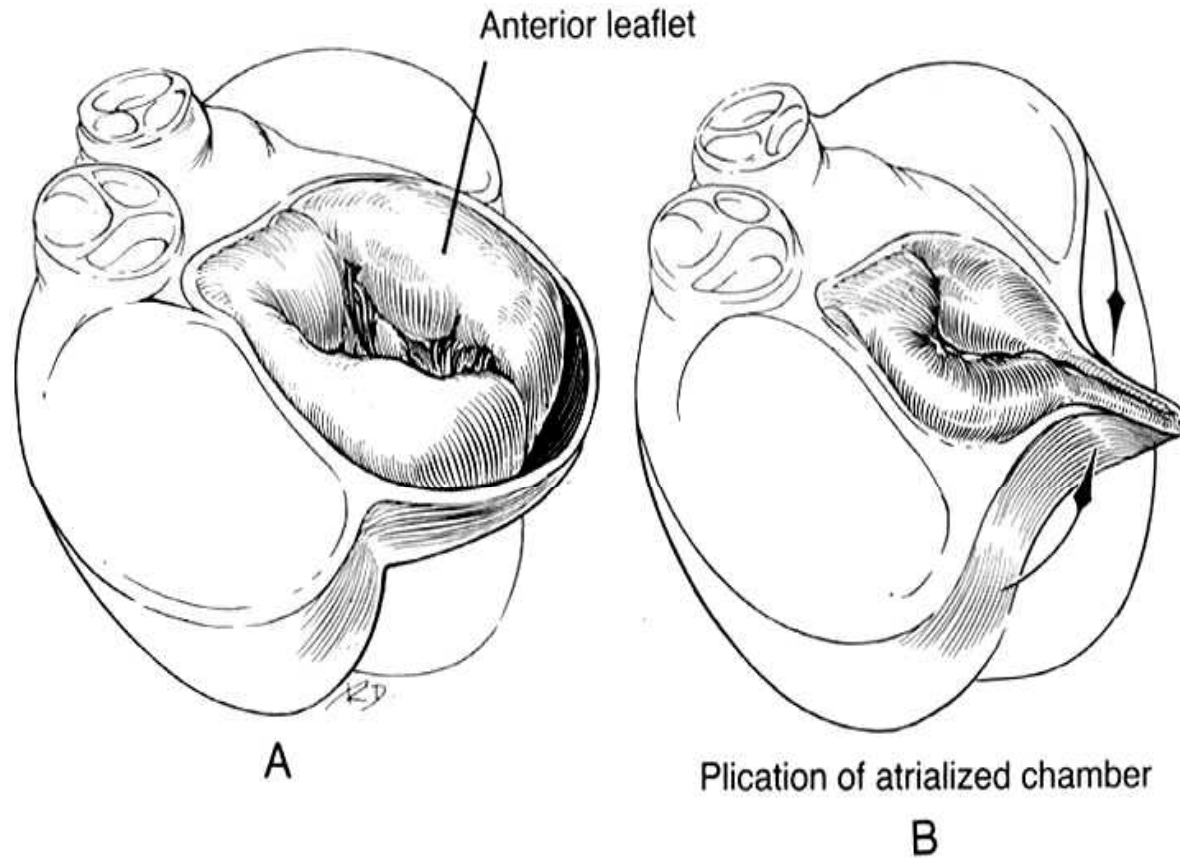


Carpentier's Repair

Sliding TV Annuloplasty with C ring



Carpentier Repair



Ebstein's anomaly: repair based on functional analysis

Carpentier et al.

Eur J Cardiothorac Surg 2003;23:525

- Preserved RV geometry
- Late problems due to devitalized tricuspid valve tissue related to reattachment?

A new reconstructive operation for Ebstein's anomaly of the tricuspid valve

Carpentier et al.

J Thorac Cardiovas Surg 2003;23:525

- 1980 ~2002
- 191 patients (Age: 24.7 ± 15 years)
- Early mortality: 9% (18 pts)
- Survival at 20 years: $82 \pm 5\%$
- Reoperation at 20 years: $88.6 \pm 3.5\%$
- Reoperation 2.6 yrs later due to TR (14), TS (2)

A MODIFIED REPAIR TECHNIQUE FOR TRICUSPID INCOMPETENCE IN EBSTEIN'S ANOMALY

Roland Hetzer, MD, PhD
Nicole Nagdyman, MD
Peter Ewert, MD
Yu Guo Weng, MD
Vladimir Alexi-Meskhisvili, MD, PhD
Felix Berger, MD
Miralem Pasic, MD, PhD
Peter E. Lange, MD, PhD

Objective: A modified technique for tricuspid valve repair in Ebstein's anomaly restructures the valve mechanism at the level of the true tricuspid annulus by using the most mobile leaflet for valve closure without plication of the atrialized chamber. Midterm results of this therapeutic approach for patients with Ebstein's anomaly and tricuspid valve incompetence are reported. **Methods:** Between October 1988 and April 1997, the incompetent tricuspid valve was repaired with our technique in 19 patients (12 female, 7 male; 2 to 54 years, mean 21 years). The indication for operation was congestive heart failure of various degrees in all patients. Tricuspid incompetence was grade II in two patients, grade III in 14, and grade IV in three. Associated congenital malformations were simultaneously repaired (interatrial communication in 18, ventricular septal defect in two, pulmonary stenosis in two, mitral valve prolapse in one). Follow-up ranged between 10 and 103 months (median 28 months) and was complete for all patients. **Results:** There were no operative deaths. One patient with active endocarditis and pulmonary abscess died 2 months after the operation of recurrent sepsis; there were no late deaths. During follow-up, New York Heart Association functional class improved from 2.8 before the operation to 1.9 without recurrent cyanosis, and tricuspid incompetence decreased from a mean grade of 3.1 to one of 0.9, without any echocardiographic deterioration of the tricuspid valve function or right ventricular dilation. **Conclusions:** Our technique allows tricuspid valve repair in patients with Ebstein's anomaly, even in cases usually reserved for primary valve replacement, without late functional deterioration. (J Thorac Cardiovasc Surg 1998;115:857-68)

A modified repair technique for tricuspid incompetence
in Ebstein's anomaly

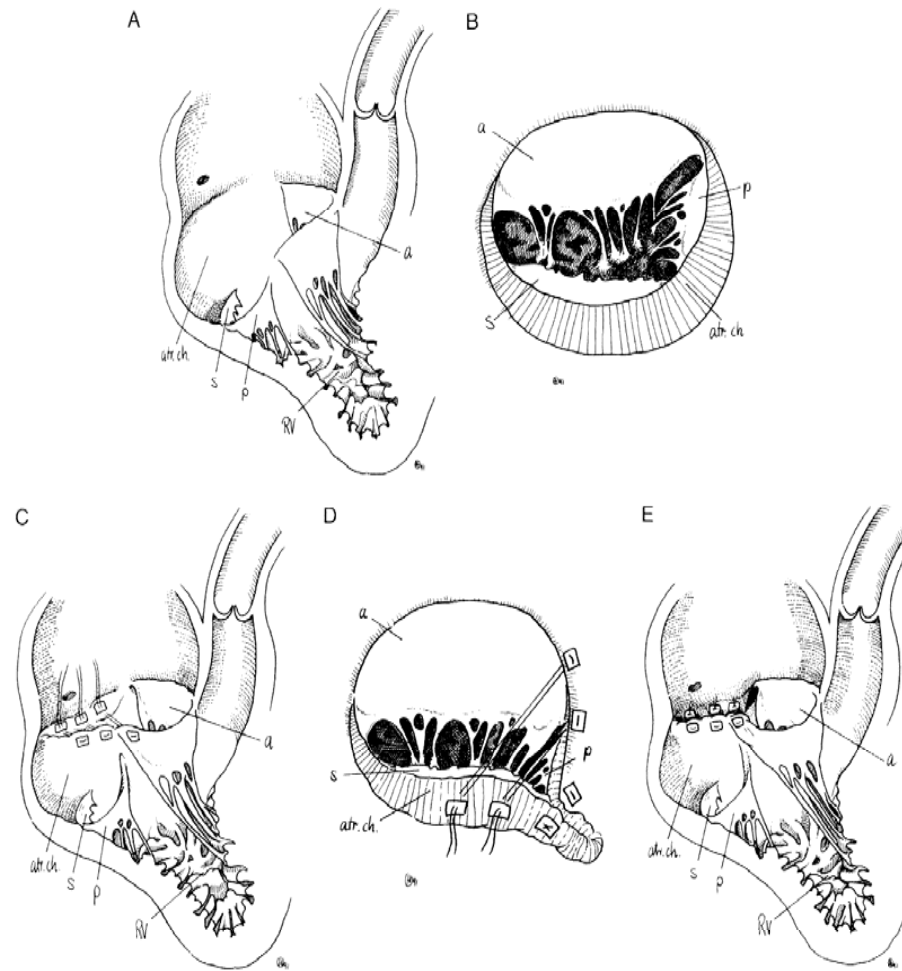
Hetezer et al

J Throac Cardiovasc Surg 1998;115:857

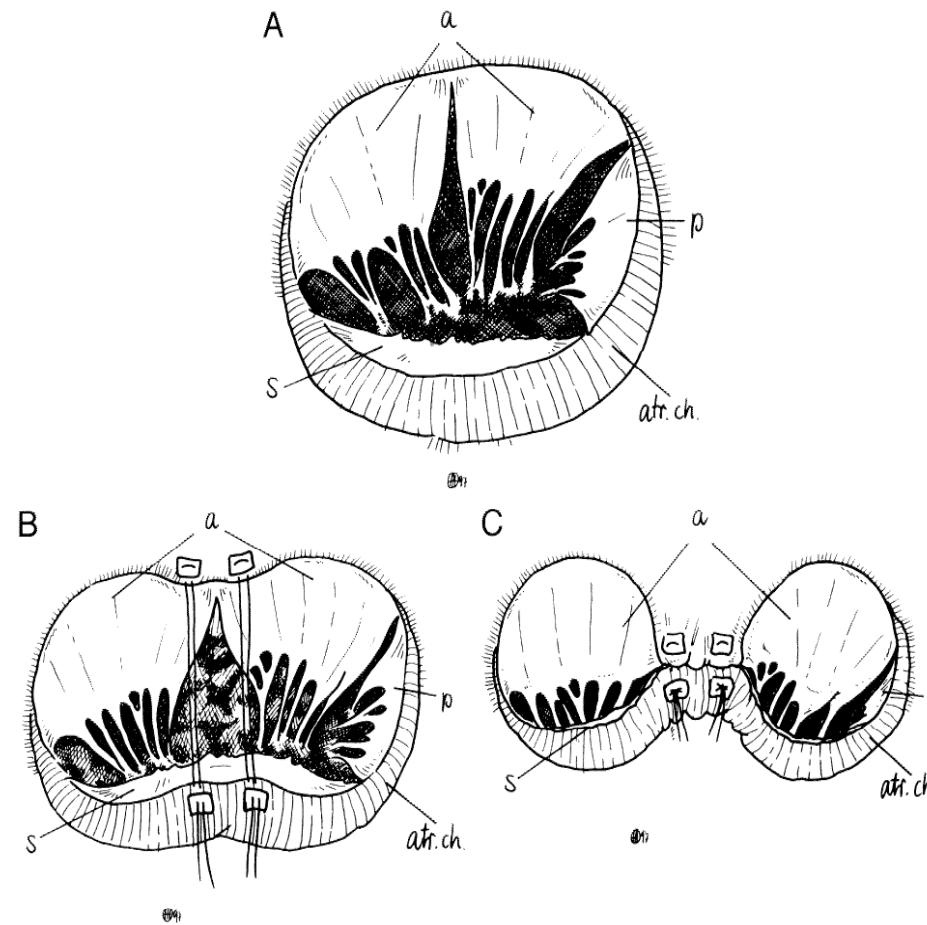
- Principle

Reconstruct the valve mechanism at the level of the true tricuspid annulus by using the most mobile leaflet for valve closure without plication of the atrialized chamber

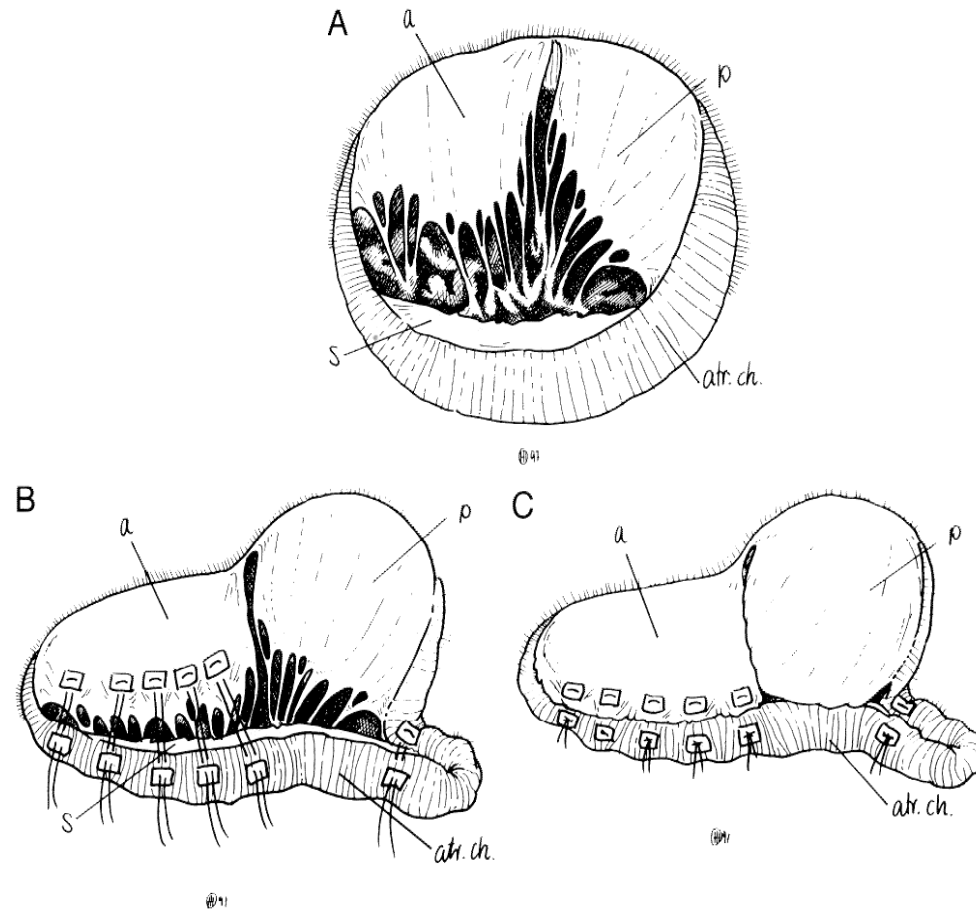
Modified Repair Technique for Tricuspid Incompetence in Ebstein's Anomaly



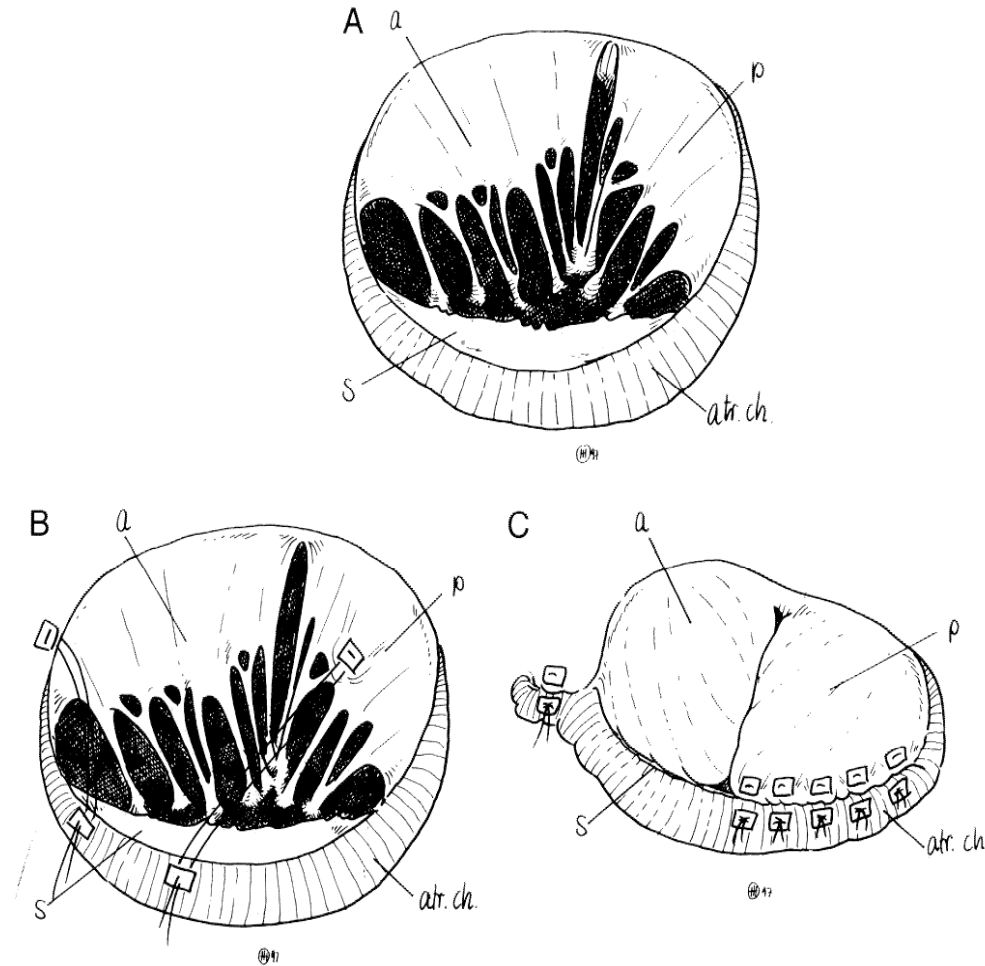
A modified repair technique for Tricuspid Incompetence in Ebstein's Anomaly



A modified Repair Technique for Tricuspid Incompetence in Ebstein's Anomaly



A modified repair technique for Tricuspid Incompetence in Ebstein's Anomaly



A modified repair technique for tricuspid incompetence in Ebstein's anomaly

Hetezer et al

J Throac Cardiovasc Surg 1998;115:857

- 1988-1997
- 19 patients (mean age: 22 yr, 2~54 yr)
- No operative death
- Follow up median 28 (10 ~ 103) mo
- TR mean 3.1 → 0.9

A modified repair technique for tricuspid
incompetence in Ebstein's anomaly

Hetezer et al

J Throac Cardiovasc Surg 1998;115:857

- Simple
- Not plicate aRV: Adequate RV volume

Ventricularization of the Atrialized Chamber: A Concept of Ebstein's Anomaly Repair

Michael V. Ullmann, MD, Sabine Born, MD, Christian Sebening, MD, Matthias Gorenflo, MD, Herbert E. Ulmer, MD, and Siegfried Hagl, MD

Departments of Cardiac Surgery and Pediatric Cardiology, University of Heidelberg, Heidelberg, Germany

Background. We report results of a technique of Ebstein's anomaly repair by creating a predominantly monocuspid valve with simultaneous ventricularization of the atrialized right ventricular (aRV) chamber.

Methods. Between March 1993 and April 2003, Ebstein's anomaly repair by valvuloplasty with combined ventricularization was performed in 23 patients aged 13.6 (4.1–52.6) years presenting with tricuspid valve regurgitation (TVR) (I°, n = 1; II°, n = 3; III°, n = 13; IV°, n = 6). Valvuloplasty consisted of creating a predominantly monocuspid valve at the level of the anatomical atrioventricular junction resulting in a ventricularization of the atrialized chamber. Postoperatively all survivors were examined regularly with an actual prospective evaluation.

Results. One early death (4.4%) occurred and was caused by right heart failure. Follow-up was 4.6 (0.5–10.9) years. Important recurrent atrioventricular valve regurgi-

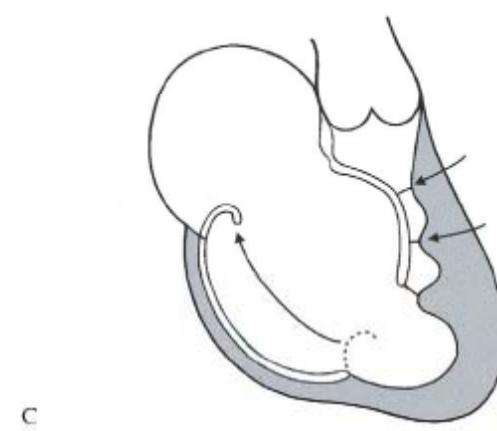
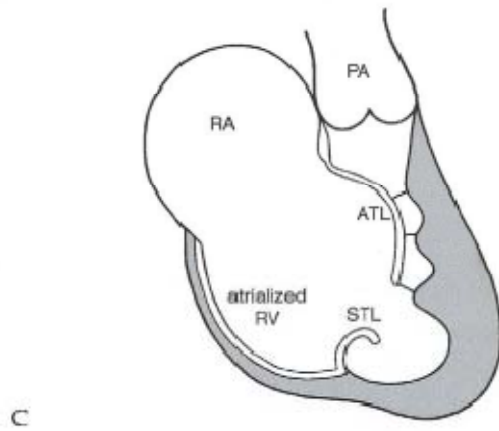
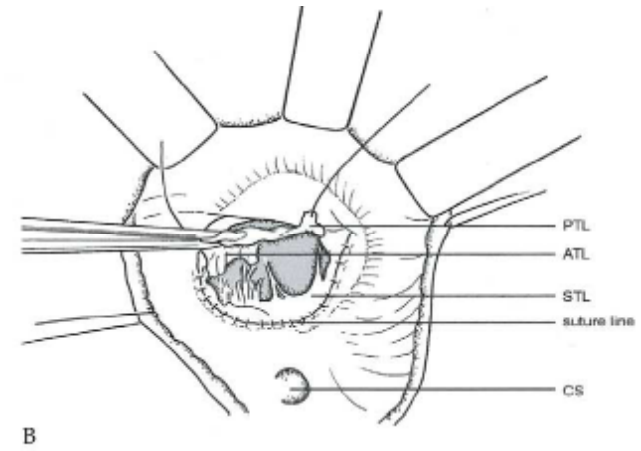
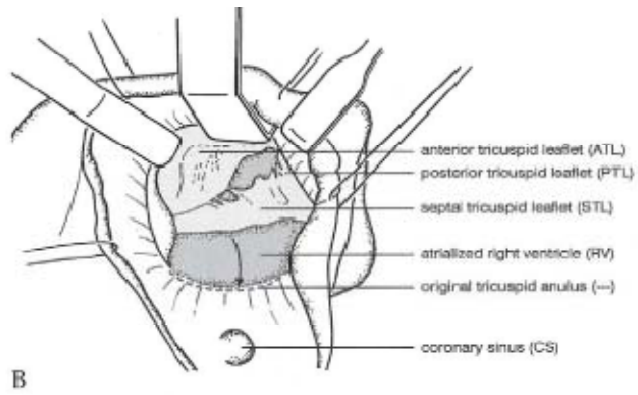
tation caused by rupture of fixation sutures occurred in 3 patients (13%), necessitating reintervention at 3 (0.03–4) months (revalvuloplasty, n = 2; TV replacement, n = 1). One patient presenting with hypoplastic right ventricle with consecutive right heart failure underwent creation of a total cavopulmonary connection at 10 months. At present all patients are doing well. Actual echocardiographic examination revealed significant improvement of right atrioventricular valve regurgitation ($p < 0.0001$) and favorable restoration of RV geometry and function.

Conclusions. This technique of Ebstein's anomaly repair with ventricularization of the atrialized chamber provides excellent results regarding right atrioventricular valve function and leads to a favorable restoration of RV geometry and function.

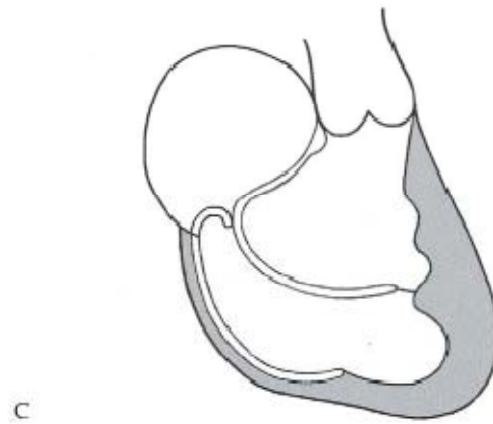
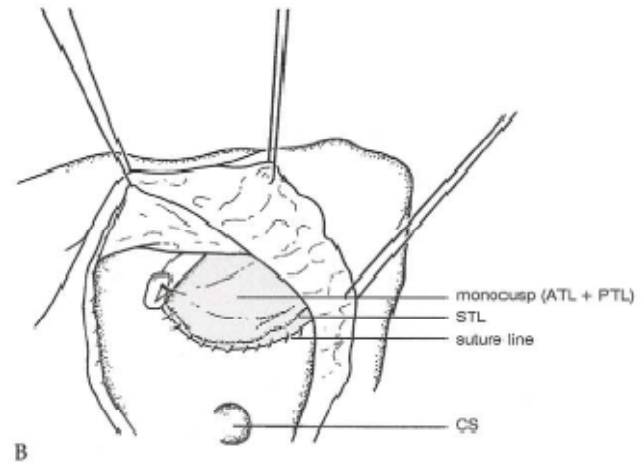
(Ann Thorac Surg 2004;78:918–25)

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Ebstein's Anomaly



Ebstein's Anomaly



Ventricularization of the Atrialized chamber: A Concept of Ebstein's Anomaly Repair

Ullmann et al

Ann Throac Surg 2004;78:918

- 1993 ~ 2003
- 23 patients
- Early death: 4.4%
- Follow up 4.6 years
 - 3 reoperation due to rupture of fixation suture
 - 1 TCPC

A New Procedure for Ebstein's Anomaly

Qingyu Wu, MD, and Zhixiong Huang, MD

Department of Cardiovascular Surgery, Cardiovascular Institute, Fu Wai Hospital, Chinese Academy of Medical Science, Peking Union Medical College, Beijing, China

Background. A new procedure for correction of Ebstein's anomaly that restores to near normal the anatomic and physiologic function of the tricuspid valve and the right ventricle is reported.

Methods. Between December 1997 and September 2002, 34 consecutive patients with Ebstein's anomaly underwent this new procedure. There were 13 male and 21 female patients aged 9 months to 48 years (mean, 17 years). Tricuspid incompetence was moderate in 12 patients and severe in 22. Our repair technique is as follows: the displaced posterior leaflet with some chordae tendineae and corresponding papillary muscle are detached from the annulus and ventricular wall, respectively. The leaflet is then reattached to the native posterior annulus with reimplantation of the papillary muscle. The displaced septal leaflet is treated in the same manner. Most of the atrialized portion of the ventricular wall is excised; the tricuspid annulus is plicated. In 8 of the

patients the septal leaflet was severely hypoplastic and necessitated creation of a new leaflet using autologous pericardium.

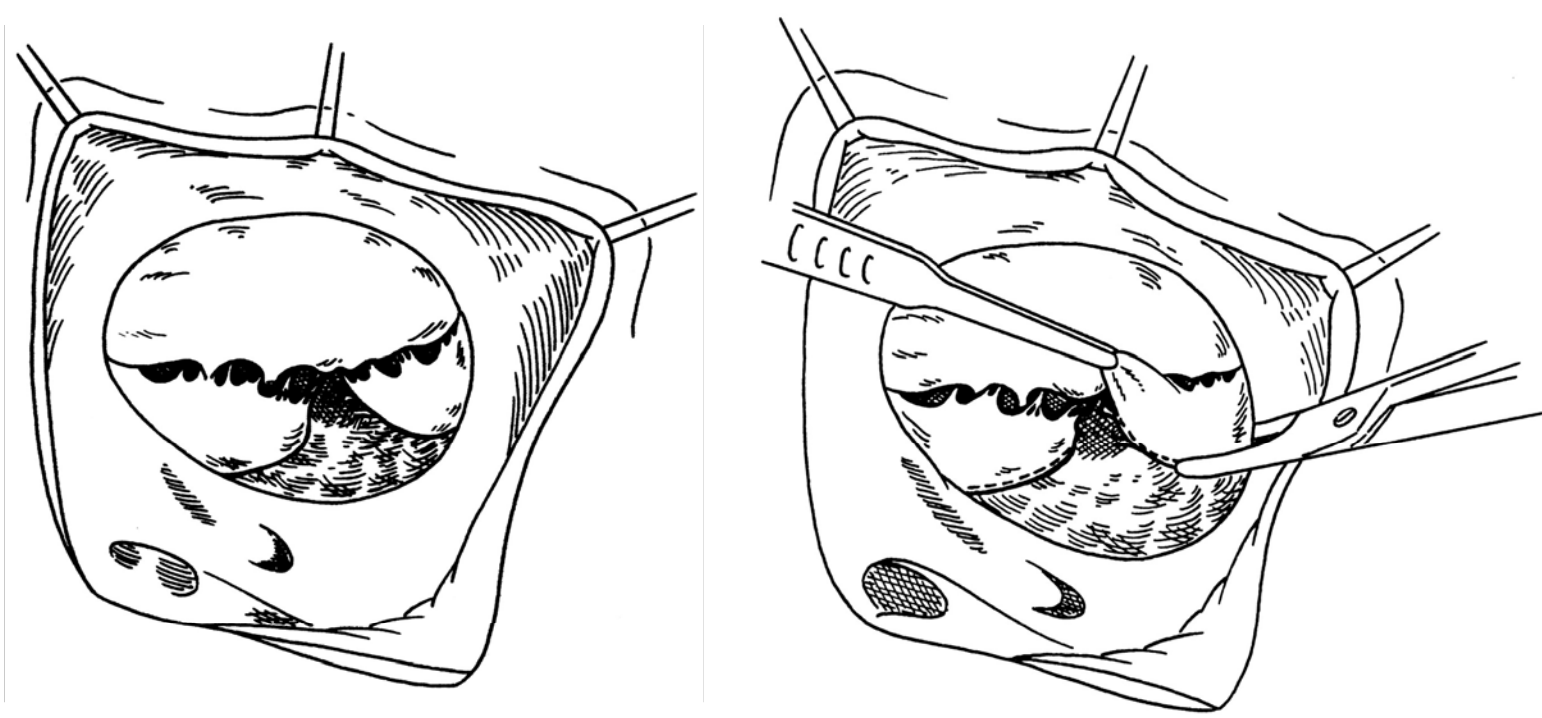
Results. All patients survived and recovered uneventfully. Postoperative echocardiography showed that tricuspid incompetence disappeared in 29 patients and was mild in 5. Right ventricular size decreased significantly with complete disappearance of the atrialized segment. Follow-up of patients ranged from 1 to 55 months (mean, 25 months), with 9 patients having more than 3 years of follow-up. They are doing well and their exercise tolerance improved to normal.

Conclusions. This new procedure anatomically corrects Ebstein's anomaly with the satisfactory early and mid-term results.

(Ann Thorac Surg 2004;77:470-6)

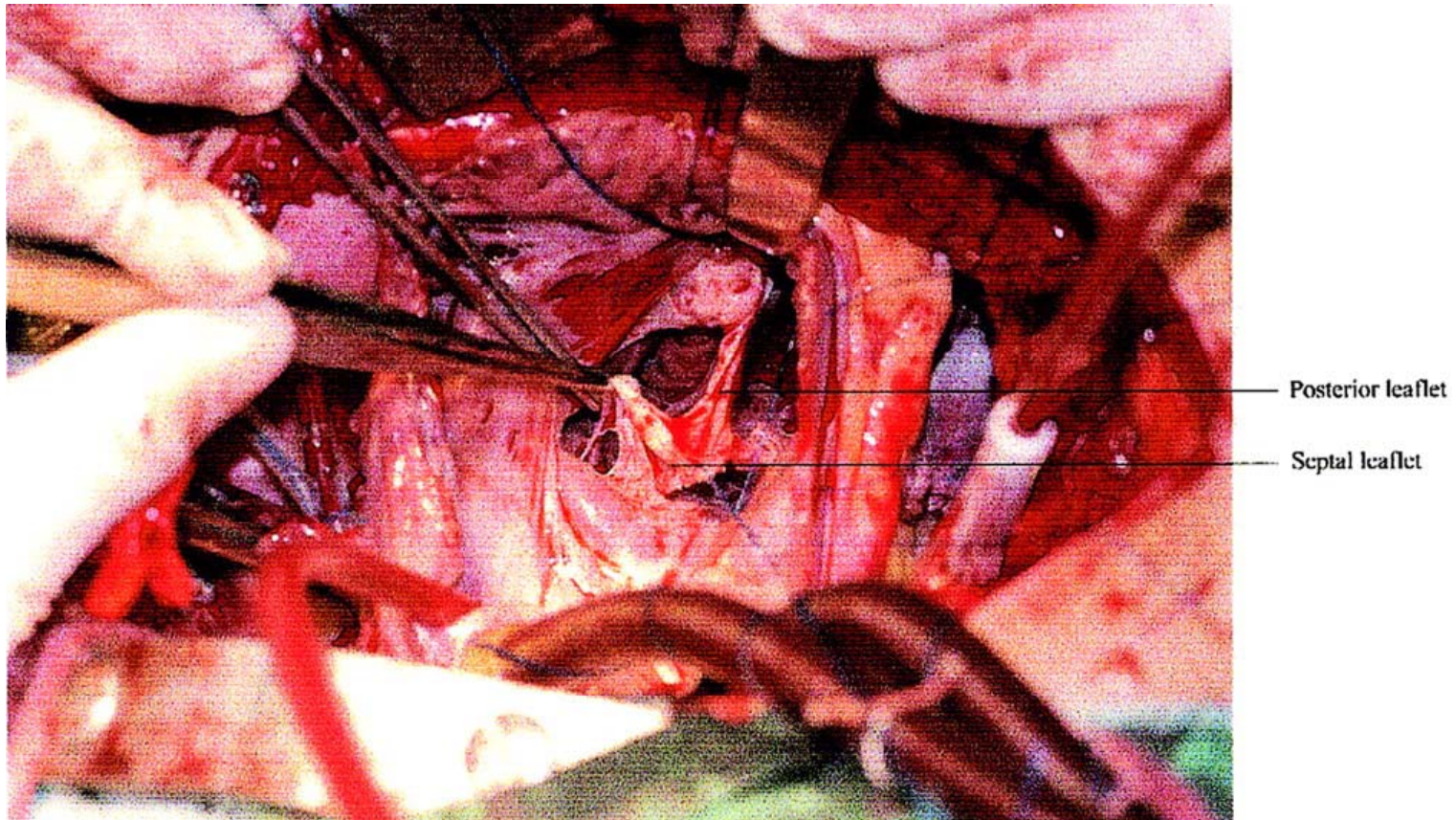
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Wu Repair



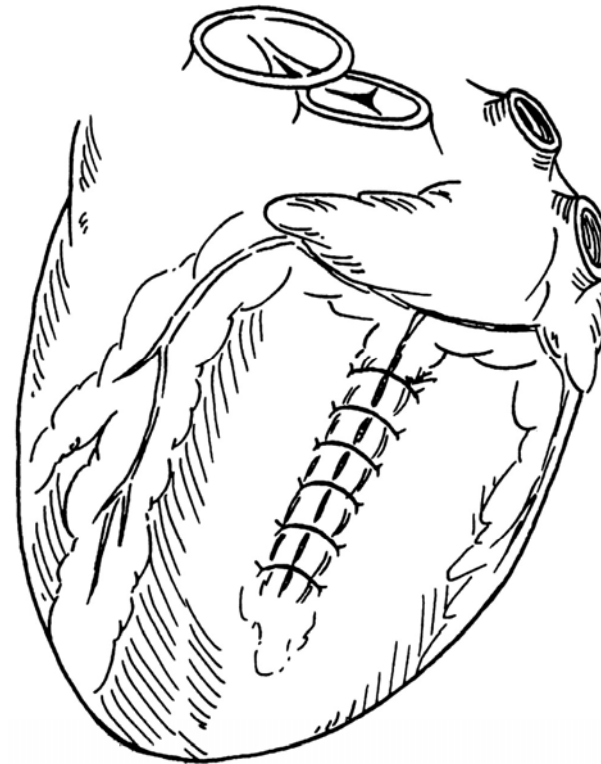
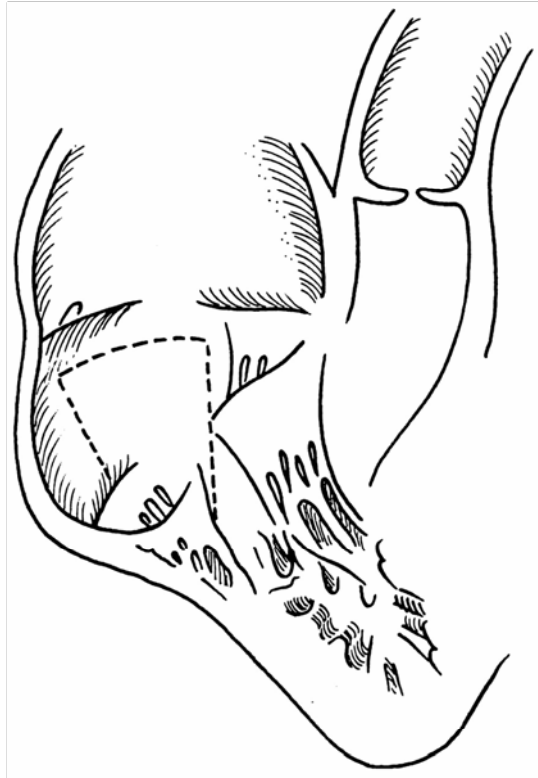
Wu Q. et al.; Ann Thorac Surg 2004;77:470-476

Wu Repair



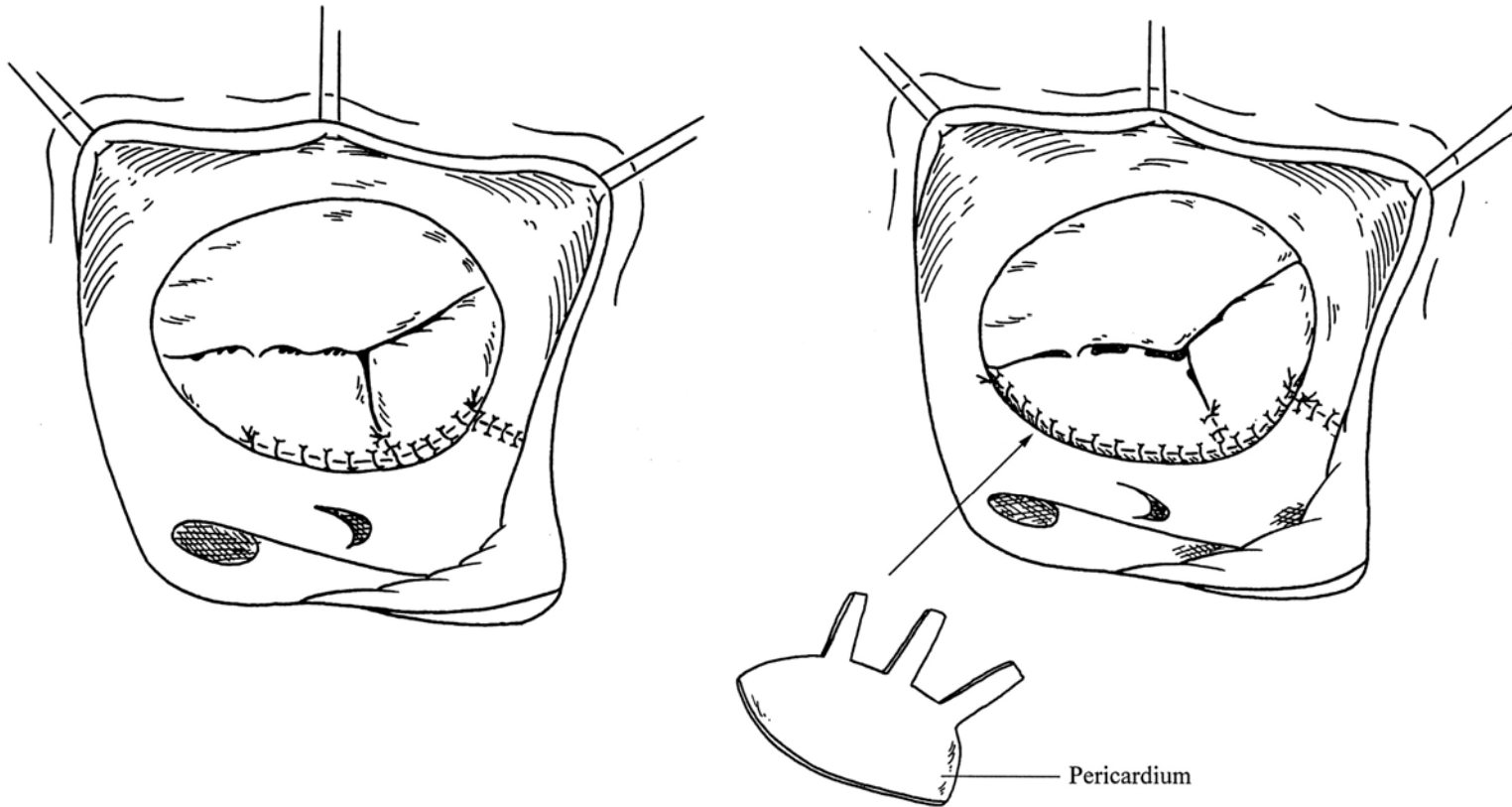
Wu Q. et al.; Ann Thorac Surg 2004;77:470-476

Wu Repair



Wu Q. et al.; Ann Thorac Surg 2004;77:470-476

Wu Repair



Wu Q. et al.; Ann Thorac Surg 2004;77:470-476

A new procedure for Ebstein's Anomaly

Wu et al

Ann Thorac Surg 2004;77:470

- Atrialized ventricular wall is excised longitudinally
 - RV geometry is restored
 - the atrialized ventricular chamber is completely obliterated
 - load of the RV is alleviated
 - RV cavity dimensions are restored to near normal, which is beneficial for the recovery of RV function

A new procedure for Ebstein's Anomaly

Wu et al

Ann Thorac Surg 2004;77:470

- Displaced leaflets with their partial subvalvular structures are detached from their abnormal positions and reattached to a position near the true tricuspid annulus

Improve the function and durability of "the new leaflets"

A new procedure for Ebstein's Anomaly

Wu et al

Ann Thorac Surg 2004;77:470

- Less likelihood for tricuspid annulus to enlarge after annuloplasty
the effective size-reduction of the tricuspid annulus and the restoration of the valve leaflets to a near normal position, leading to competence of valve function and eventual decrease of the RA and RV pressures

A new procedure for Ebstein's Anomaly

Wu et al

Ann Thorac Surg 2004;77:470

- 1997 ~ 2002
- 34 patients
- Age: 17 years (9 mo ~ 48 yr)
- Carpentier classification A: 2, B: 7, C: 25
- TR moderate: 12 severe: 22
- No surgical mortality
- Follow up: 25 (1 ~ 55) mo
- TR absent: 28, mild: 3, moderate: 3

Early and midterm results in anatomic repair of Ebstein anomaly

Wu et al

J Thorac Cardiovasc Surg 2007;134:1438

- 1997~2006
- 83 patients
- TR moderate 22 severe 56
- Age: 17.9 ± 7.2 (9 mo 52 years)
- 1½ repair 1, TCPC: 4
- No surgical mortality, major morbidity
- Follow up: 46 ± 12.5 mo
- TR: absent 54, mild 17, moderate 3, severe 3

Posterior Annular Plication: Tricuspid Valve Repair in Ebstein's Anomaly

Camille L. Hancock Friesen, MD, Robert Chen, MD, Jonathan G. Howlett, MD, and David B. Ross, MD

Divisions of Cardiac Surgery and Pediatric Cardiology, IWK Health Centre, Dalhousie University, and Division of Cardiology, QEII Health Sciences Centre, Halifax, Nova Scotia, Canada

Background. Nonneonatal Ebstein's anomaly is a rare congenital anomaly for which numerous operative techniques have been described to repair the aberrant tricuspid valve, exclude the atrialized right ventricle, and address right heart dysfunction. We reviewed short-term outcomes in surgical patients treated by a simplified operative technique.

Methods. Our operative approach to Ebstein's anomaly is approximation of the anteroposterior commissure to the remnant septal leaflet with closure of the cul-de-sac longitudinally. Plication of the atrialized right ventricle (1 patient) and a bidirectional cavopulmonary connection (2 patients) were performed only if necessary. All patients were followed postoperatively by their cardiac surgeon and cardiologist.

Results. Seven patients with a mean age of 39 years (range, 3.6 to 63.8 years) underwent repair. Preoperatively

all patients had 4+ tricuspid valve regurgitation and were New York Heart Association class III. Mean postoperative hospital stay was 7 ± 2 days (range, 4 to 11 days). Mean follow-up is 42 ± 18 months (range, 7 to 58 months). At last follow-up 5 patients are New York Heart Association class I and average tricuspid valve regurgitation is mild.

Conclusions. Plication of the posterior annulus without plication of the atrialized right ventricle, resiting the tricuspid valve, or performing prophylactic cavopulmonary connection appears to be a reasonable operative approach to nonneonatal Ebstein's anomaly. Long-term follow-up of this cohort is necessary to determine the durability of such a surgical approach.

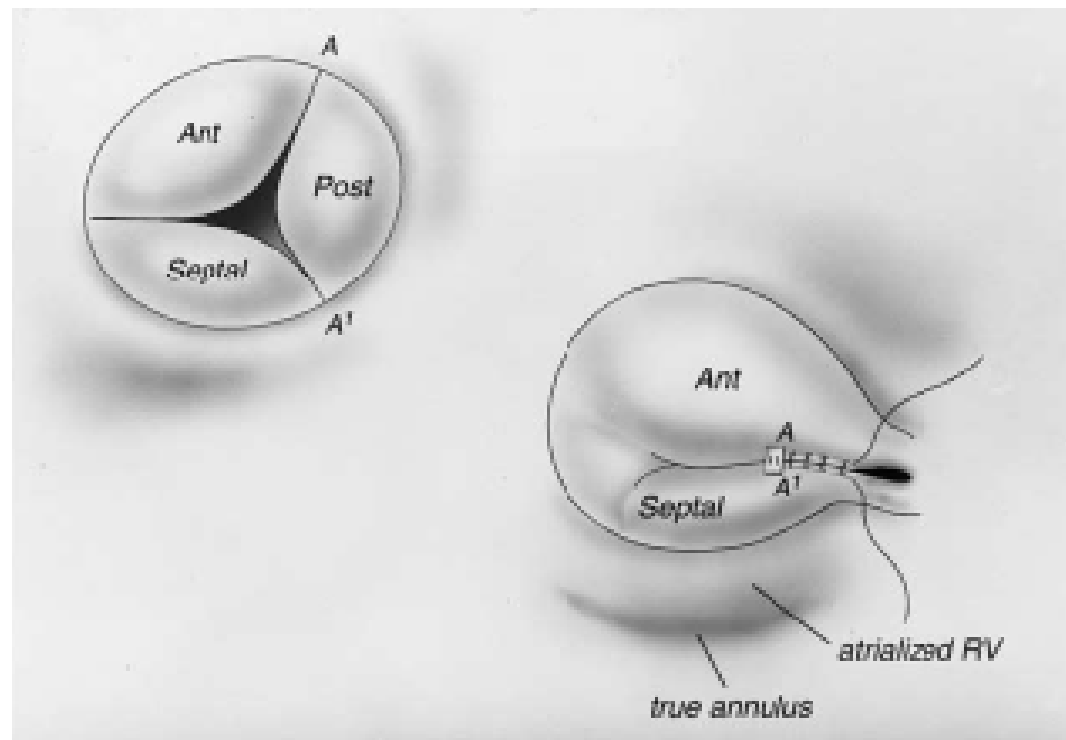
(Ann Thorac Surg 2004;77:2167-71)

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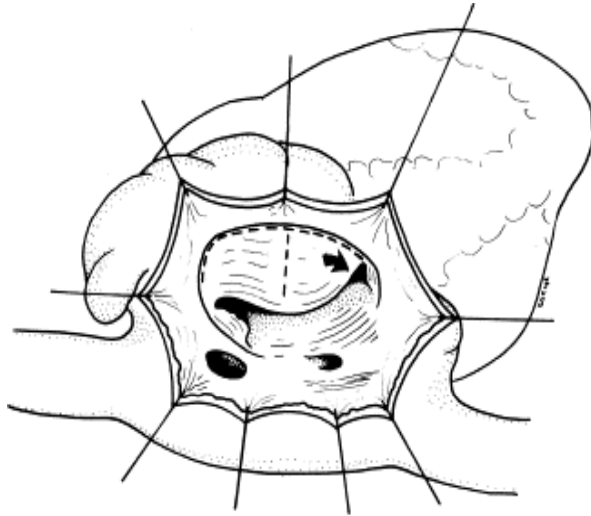
Posterior annular plication: Tricuspid valve repair in Ebstein's anomaly

Hancock et al.

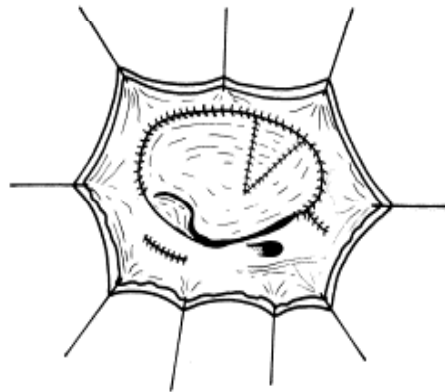
Ann Thorac Surg 2004;77:2167



Pericardial patch augmentation of Anterior tricuspid leaflet



A

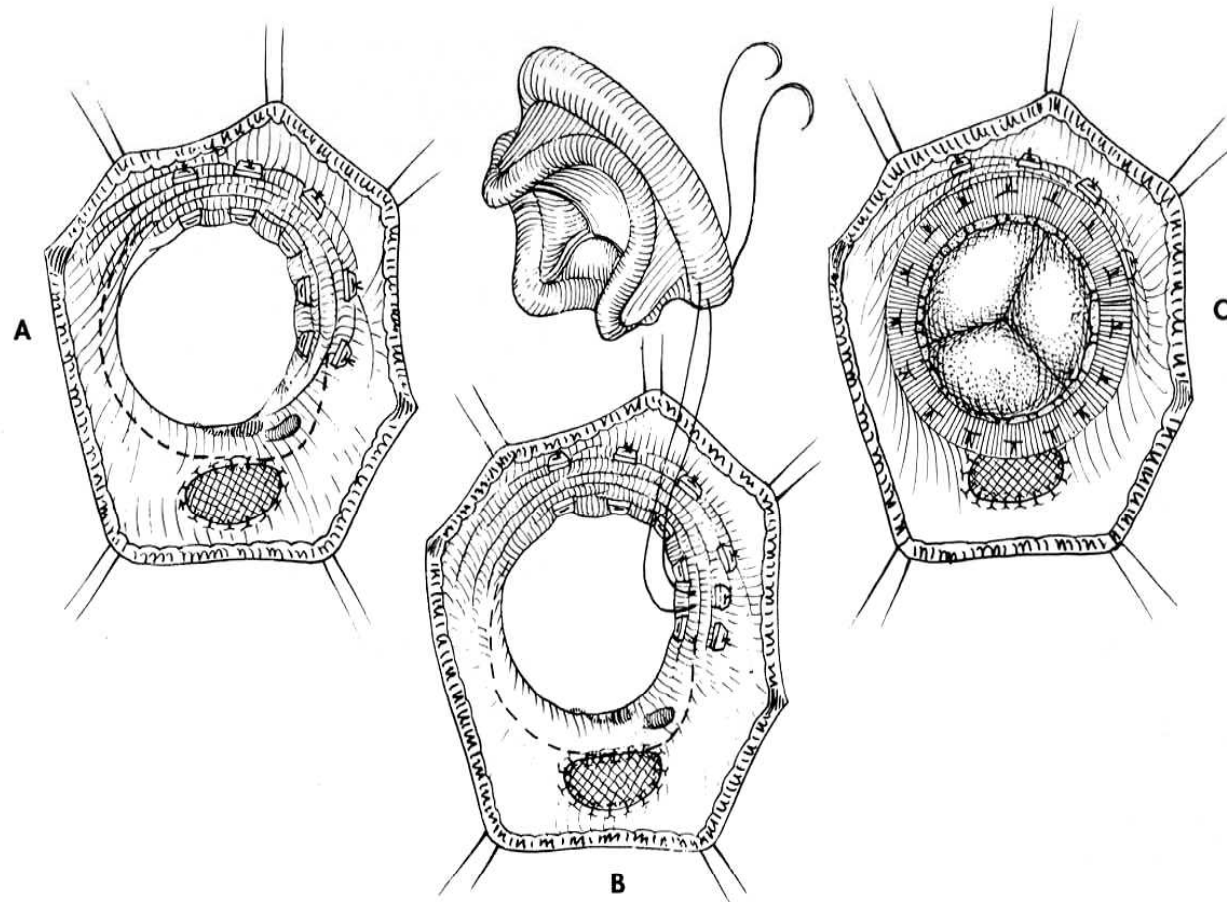


B

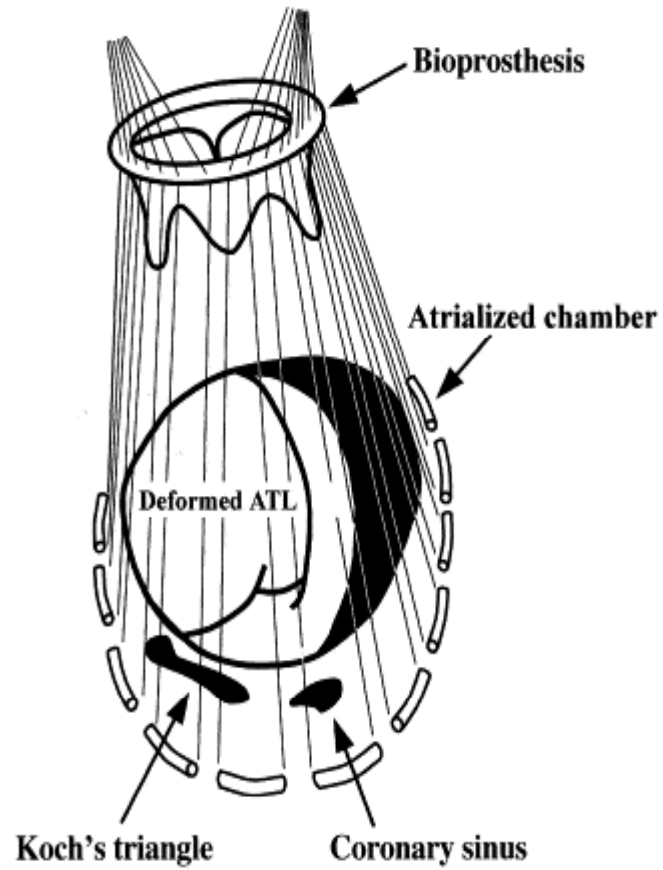
Pericardial patch augmentation of the anterior leaflet of the tricuspid valve in Ebstein's anomaly.

Valve Replacement

Technique for TVR in Ebstein's Anomaly



Tricuspid Valve Supra-annular Implantation



Late Results of Bioprosthetic Tricuspid Valve Replacement in Ebstein's Anomaly

H. Tarik Kiziltan, MD, David A. Theodoro, MD, Carole A. Warnes, MD,
Patrick W. O'Leary, MD, Betty J. Anderson, RN, and Gordon K. Danielson, MD

Division of Thoracic and Cardiovascular Surgery, Section of Pediatric Cardiology, and Division of Cardiovascular Diseases and Internal Medicine, Mayo Clinic and Mayo Foundation, Rochester, Minnesota

Background. Historically, porcine bioprosthetic valves have poor durability in pediatric patients; nearly half will require replacement within 5 years. However, our early experience with patients having Ebstein's anomaly suggests that tricuspid bioprostheses in this anomaly might have better durability.

Methods. One hundred fifty-eight patients who received a primary tricuspid bioprosthesis because of tricuspid valve anatomy unsuitable for repair between April 1972 and January 1997 were reviewed. Results were analyzed and Kaplan-Meier curves were constructed to estimate patient survival and probability of remaining free of reoperation.

Results. Follow-up of 149 patients (94.3%) who sur-

vived 30 days ranged up to 17.8 years (mean, 4.5 years). Ten-year survival was $92.5\% \pm 2.5\%$ (SE), 129 late survivors (92.1%) were in New York Heart Association class I or II, and 93.6% were free of anticoagulation. Freedom from bioprosthesis replacement was $97.5\% \pm 1.9\%$ at 5 years and $80.6\% \pm 7.6\%$ at 10 and 15 years.

Conclusions. Bioprosthesis durability in the tricuspid position in patients with Ebstein's anomaly compares very favorably with bioprosthesis durability in other cardiac valve positions, especially for pediatric patients, and also compares favorably with tricuspid bioprosthesis durability in patients with other diagnoses.

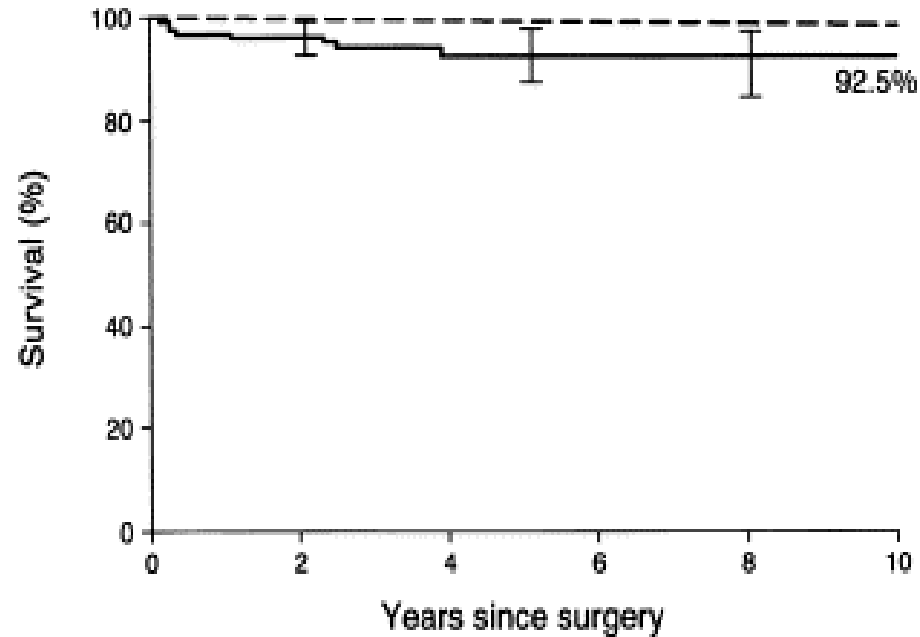
(Ann Thorac Surg 1998;66:1539-45)

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Late Results of Bioprosthetic TVR in Ebstein's Anomaly

Kiziltan et al

Ann Thorac Surg 1998;66:1539-1544

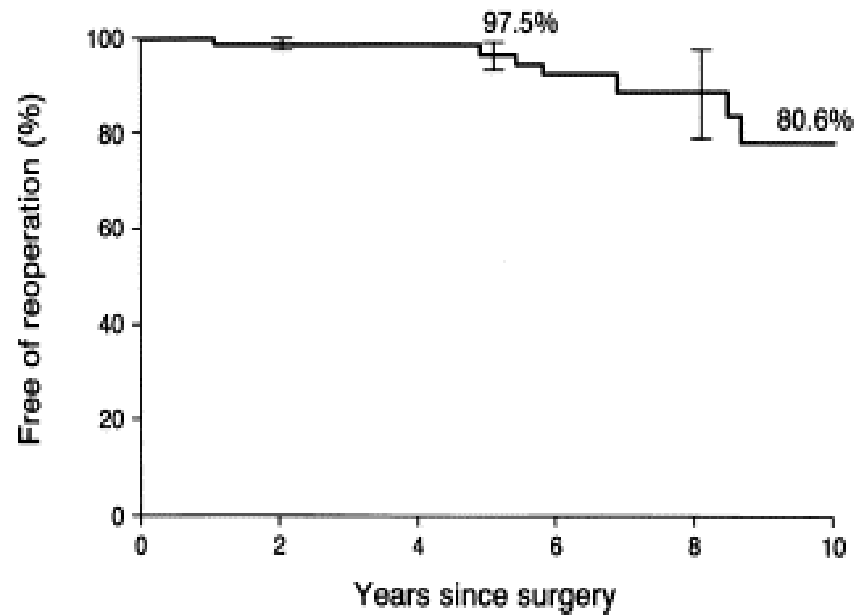


Late survival of the early (hospital) survivors.

Late Results of Bioprosthetic TVR in Ebstein's Anomaly

Kiziltan et al

Ann Thorac Surg 1998;66:1539-1544

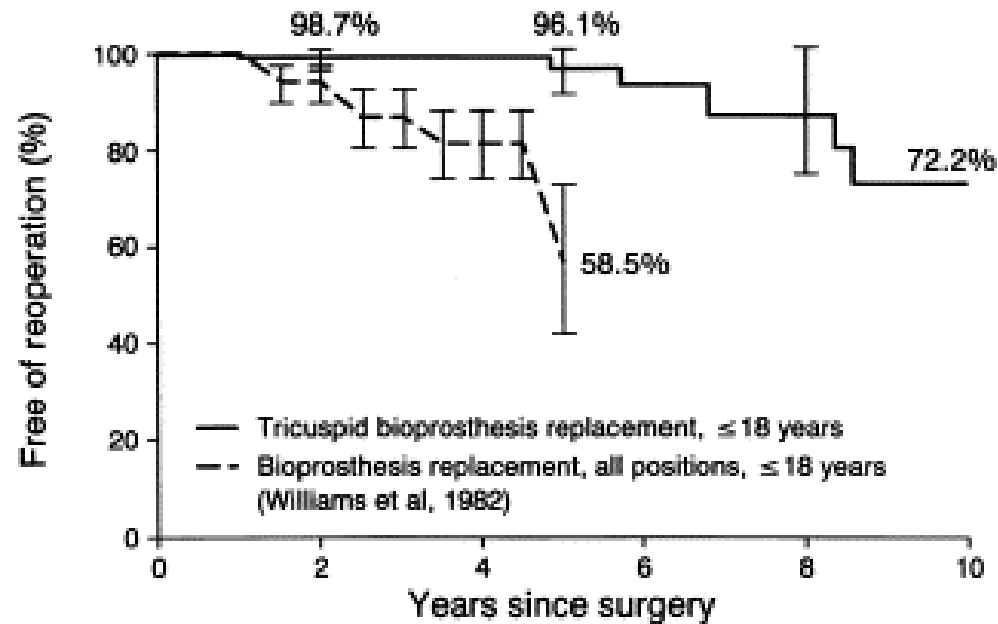


Freedom from Reoperation

Late Results of Bioprosthetic TVR in Ebstein's Anomaly

Kiziltan et al

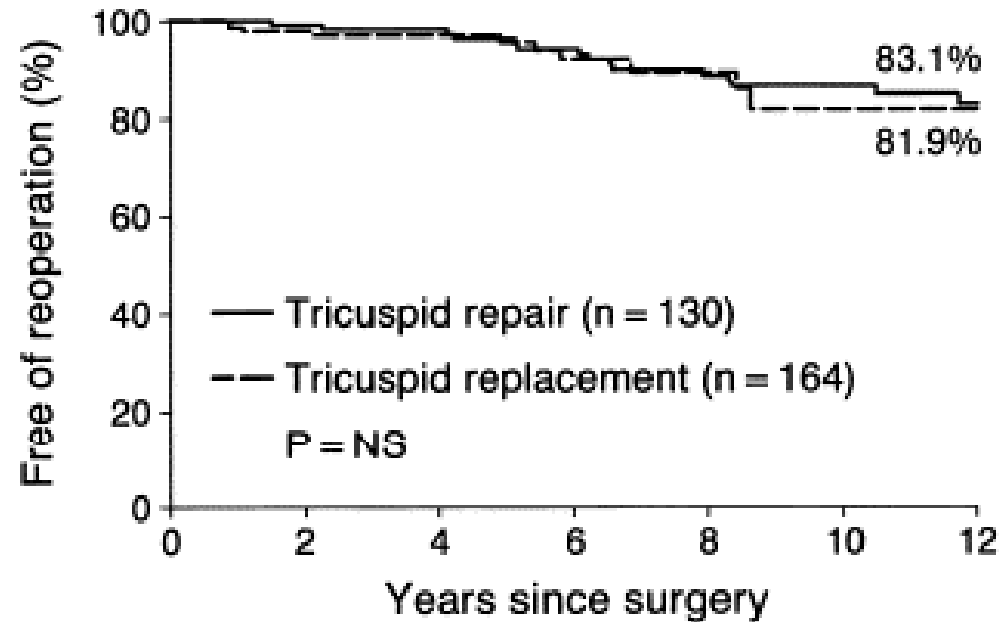
Ann Thorac Surg 1998;66:1539-1544



Late Results of Bioprosthetic TVR in Ebstein's Anomaly

Kiziltan et al

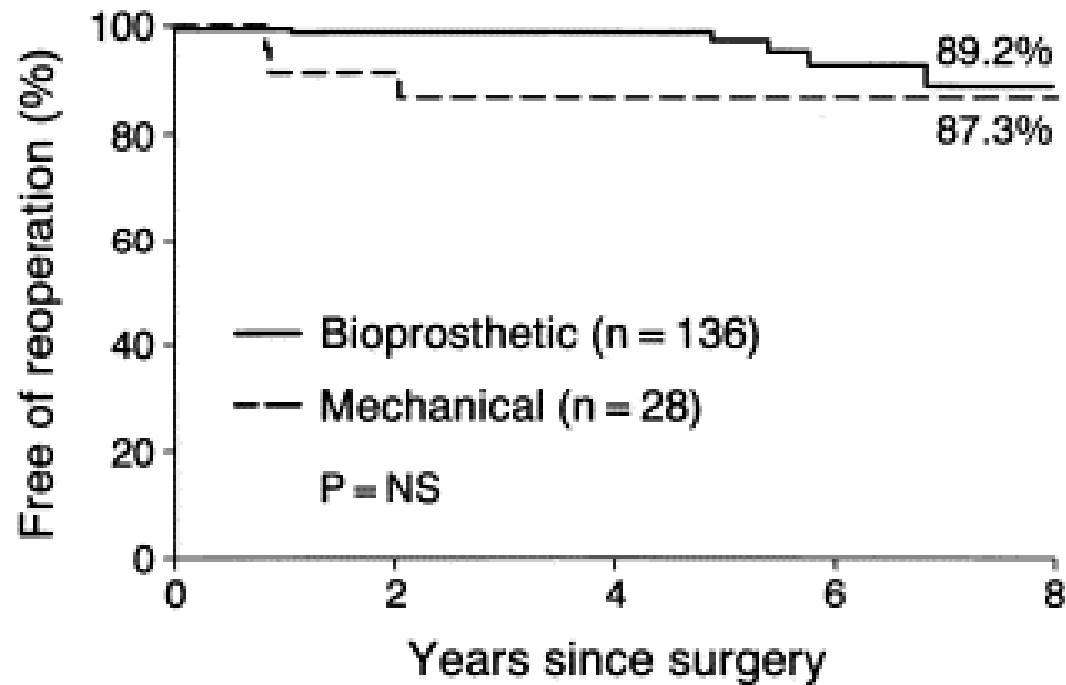
Ann Thorac Surg 1998;66:1539-1544



Late Results of Bioprosthetic TVR in Ebstein's Anomaly

Kiziltan et al

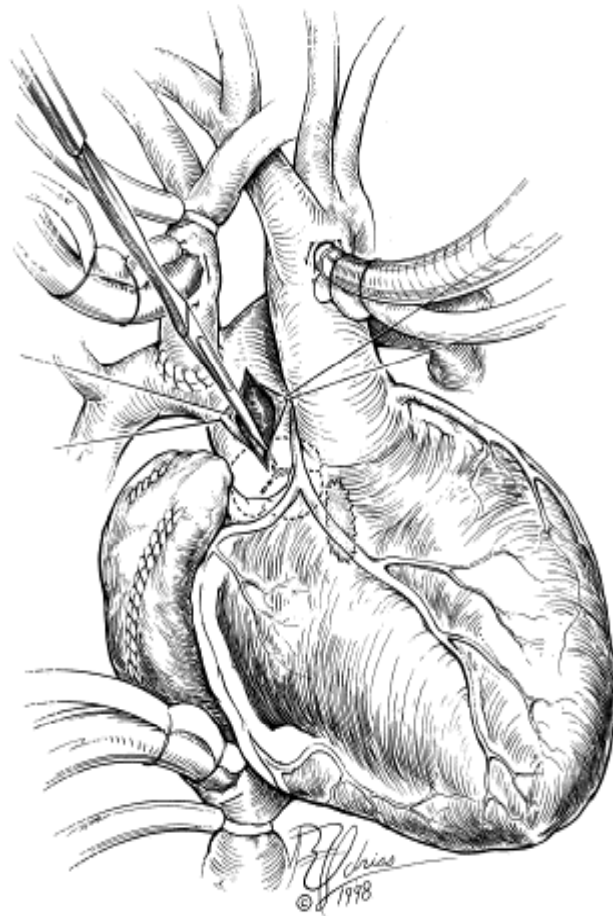
Ann Thorac Surg 1998;66:1539-1544



Cavopulmonary Connection

Ebstein's Anomaly

Bidirectional Glenn shunt in association with congenital heart repairs: the 1 + 1/2 ventricular repair





Bi-directional cavopulmonary shunt associated with ventriculo and valvuloplasty in Ebstein's anomaly: benefits in high risk patients¹

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Abstract

Objective: The prognosis for surgical repair of Ebstein's anomaly depends on the tricuspid valve repair and on the right ventricular function. In order to decrease the preload of the compromised right ventricle, a bi-directional cavopulmonary shunt was added to the intracardiac repair. **Methods:** Among 113 patients operated on for Ebstein's anomaly at our institution from 1980–1997, a cohort of 60 patients was selected for high risk for postoperative complications. Patients presented with one or more several criteria: massive tricuspid valve dysfunction, extended atrialized right ventricle, poor right ventricular contractility, or long standing atrial fibrillation. After prior informed consent, this cohort was divided into two groups. Both groups had similar preoperative clinical patterns: Group I (45 patients), surgical technique included longitudinal right ventricular plication and tricuspid valve valvuloplasty; Group II (15 patients), where the surgical technique was similar to Group I except a bi-directional cavopulmonary shunt was added at the end of the procedure. **Results:** Operative mortality was 24% (11/45) in Group I and 0% (0/15) in Group II ($P < 0.05$). The survival at 5 years was $66.1 \pm 14\%$ in Group I and $80 \pm 16\%$ in Group II (not significant). Reoperation rate was 11% (5/45) in Group I and 0% (0/15) in Group II. No deleterious effects of the bi-directional cavopulmonary shunts were observed clinically. Residual tricuspid valve insufficiency rate was 26% in both groups. However, patients with the bi-directional cavopulmonary shunt had a better tolerance and have not needed reoperations to date. **Conclu-**

BCPS associated with Ventriculo & Valvuloplasty in Ebstein's anomaly

- 113 pts operated on for Ebstein anomaly (1980-1997)
- 60 patients with high risk
 - Massive tricuspid valve dysfunction
 - Extended atrialized RV
 - Poor RV contractility
 - Long standing atrial fibrillation
- Group I (45)
 - Longitudinal RV plication + Tricuspid valvuloplasty
- Group II (15)
 - Longitudinal RV plication + Tricuspid valvuloplasty + BPCS

BCPS associated with Ventriculo & Valvuloplasty in Ebstein's anomaly

	Group I	Group II
Number of patients	45	15
Age (years \pm SD)	22 \pm 14	25 \pm 15
Functional class NYHA	<i>n</i> (%)	<i>n</i> (%)
1	1 (2)	0 (0)
2	14 (31)	1 (6)
3	21 (47)	7 (47)
4	9 (20)	7 (47)
Cyanosis	29 (64)	10 (67)
C/TR	0.64 \pm 0.07	0.69 \pm 0.06
TV insufficiency grade	<i>n</i> (%)	<i>n</i> (%)
1	1 (2)	0 (0)
2	1 (2)	0 (0)
3	22 (49)	5 (33)
4	21 (47)	10 (67)
TV stenosis	14 (31)	3 (20)
Rhythm disturbances	12 (27)	5 (33)
Associated anomalies	27 (60)	11 (73)

BCPS associated with Ventriculo & Valvuloplasty in Ebstein's anomaly

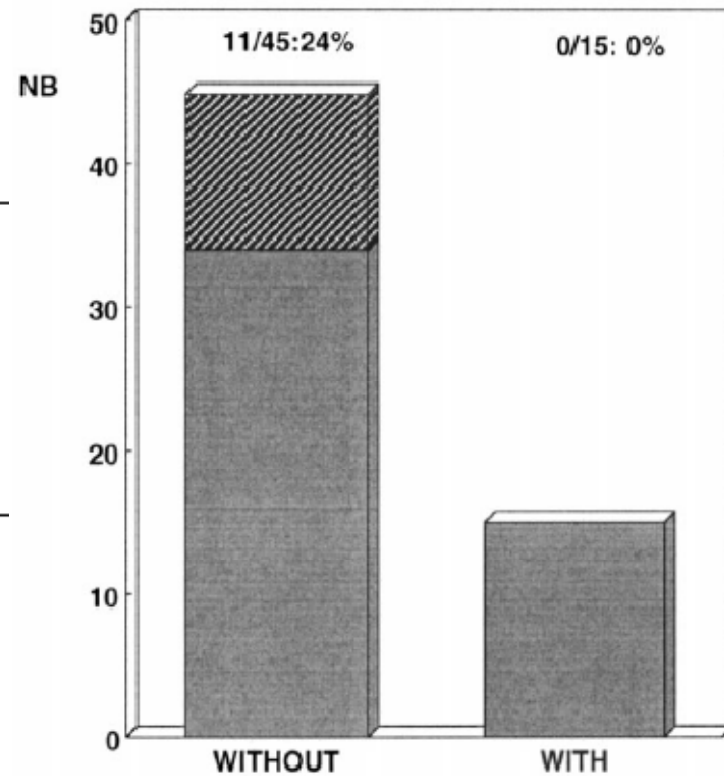
Causes of operative mortality

Group I (11/45)

RV failure	5
LV failure	1
Rhythm disturb	2
Infection	2
MOF	1

Group II (0/15)

RV: right ventricle, LV: left ventricle, MOF: multi organ failure.



BCPS associated with Ventriculo & Valvuloplasty in Ebstein's anomaly

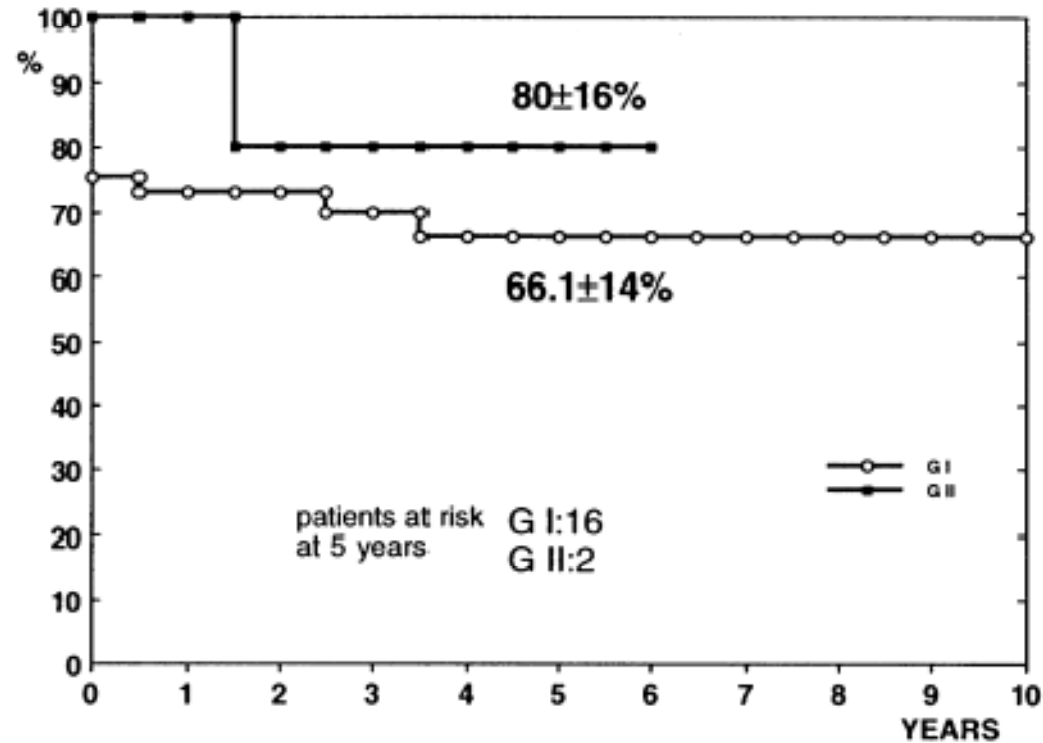


Fig. 4. Actuarial survival. The difference between group I and II is not statistically significant.

BCPS associated with Ventriculo & Valvuloplasty in Ebstein's anomaly

Grades of postoperative tricuspid valve insufficiency (assessed on echo Doppler) in Group I and in Group II

Grade of TV	Insufficiency	1	2	3	4
Group I	<i>n</i> : 34	4	21	6	3
	%	11.7	61.7	17.6	8.8
Group II	<i>n</i> : 15	3	8	3	1
	%	20	53.3	20	6.6

Postoperative functional class (NYHA) in Group I and in Group II

NYHA	Class	1	2	3	4
Group I	<i>n</i> : 34	13	16	4	1
	%	38.2	47	11.7	2.9
Group II	<i>n</i> : 15	11	4	0	0
	%	73.3	26.6		

BCPS associated with Ventriculo & Valvuloplasty in Ebstein's anomaly

Advantage

- Diverts 30~40% of systemic venous return from RV
- ↓ work index of the compromised RV
- direct preloading of the LV
- avoids the negative effect of a distended RV on the compliance and diastolic function of the LV
- Prevent postoperative ventricular dilatation
- ↓ Operative mortality
- ↓ incidence of reoperation
- Better tolerance of residual TV dysfunction

Indication

beneficial for those with large functional RV of poor systolic function



Ebstein's anomaly: repair based on functional analysis[☆]

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Abstract

Objective: 'Classical' repair of Ebstein's anomaly is usually performed with transverse plication of the atrialized chamber. However, the anterior leaflet has restricted motion which is an important factor of the tricuspid valve insufficiency. We studied the long term results of mobilization of the anterior leaflet associated with longitudinal plication of the right ventricle. **Methods:** From 1980 to July 2002, 191 patients (mean age 24.4 ± 15 years (1–65)) were operated on. Anterior leaflet function was assessed on pre-op echocardiography and on surgical examination. Conservative surgery was possible in 187 patients (98%) and included mobilization of the anterior leaflet, longitudinal plication of the right ventricle and prosthetic annuloplasty in adults. Bidirectional cavo-pulmonary shunt was associated in 60 patients. Four patients had valve replacement. **Results:** Hospital mortality occurred in 18 patients: 9% (95%CL: 6–15%) due to right ventricle (RV) failure in nine patients. Mean follow-up was 6.4 years (0.07–22). Actuarial survival was 82% at 20 years. Tricuspid valve insufficiency was 1 or 2+ in 80% of the cases. Reoperation occurred in 8% (16 patients). A successful second repair was obtained in ten patients. Electron beam computerized tomography (20 patients) demonstrated improved left ventricle ejection fraction 56–66% ($P < 0.05$). Supraventricular tachycardia and pre-excitation syndromes were reduced from 23 to 5%. **Conclusion:** Conservative surgery is indicated for all symptomatic patients. The incidence of valve repair is high when leaflet mobilization is performed. Valve replacement can be avoided in most cases. Functional and hemodynamic results are excellent.

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Keywords: Ebstein's anomaly; Tricuspid valve

Cavopulmonary Connection

Ebstein's anomaly: repair based on functional analysis

Chauvau et al. EJCTS 2003;23:525-31

191 patients

conservative surgery: 187 pts (BCPS 60 pts, 34%)

valve replacement: 4 pts

↑ LV EF

adjunction of CP connection improves the preload on the LV in patient who would otherwise have a difficult postoperative course.

Results of the 1.5-ventricle repair for Ebstein anomaly and the failing right ventricle

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David J. Driscoll, MD,^{b,c} Heidi M. Connolly, MD,^c and Gordon K. Danielson, MD^a

Objective: Repair of Ebstein anomaly and impaired right ventricular function pose challenges for the cardiac surgeon. The bidirectional cavopulmonary shunt may improve early outcomes. We reviewed our experience with the 1.5-ventricle repair in this patient population.

Methods: Between July 1999 and March 2006, 169 patients underwent operations to repair Ebstein anomaly. Fourteen patients had a bidirectional cavopulmonary shunt constructed. The median age at operation was 6 years (17 months-57.8 years). All of the patients had severe Ebstein anomaly with dilated right-sided chambers and/or right ventricular dysfunction. The mean left ventricular ejection fraction was 54.5% (range 35%–72%). Three patients were initially referred for heart transplantation, and the bidirectional cavopulmonary shunt allowed a conventional repair.

Results: Procedures included bidirectional cavopulmonary shunting (14), tricuspid valve replacement (11), tricuspid valve repair (2), and right ventricular resection (3). Shunting was planned preoperatively in 9 patients; the indication in 5 other patients was hemodynamic instability after separation from cardiopulmonary bypass. One patient died of multiple organ failure. Median follow-up in 10 patients was 18 months (3 months-6.5 years). The preoperative left ventricular ejection fraction of less than 50% improved in 3 patients to greater than 50% postoperatively.

Conclusions: The 1.5-ventricle repair can be utilized in patients with severe Ebstein anomaly and impaired right ventricular function who are at high risk for surgical treatment. We believe the bidirectional cavopulmonary shunt may be considered as a planned procedure, as an intraoperative salvage maneuver, or as an alternative to cardiac transplantation in selected patients.

Results of the 1.5-ventricle repair for Ebstein Anomaly and the failing right ventricle

Quinoez et al. EJCTS 2007;133:1303

1999~2006

169 pts underwent operation for Ebstein Operation

BCPS 14 pts

Severe Ebstein anomaly with dilated right-sided chambers and/or
RV dysfunction

BCPS + TVR(11), TV repair (2), RV resection (3)

LV EF 54.5% (35-72%)

Surgical Mortality: 1

Ebstein's Anomaly

TABLE 1. Patient demographics, clinical presentations, and primary operations

Patient	Age	Clinical presentation	Primary procedure(s)	Other procedures
1	17 mo	Biventricular failure; shock; atrial flutter	TVR 27 mm	PFO closure; isthmus cryoablation; right reduction atrioplasty; BCPS; ECMO; open chest
2	18 mo	Cyanosis; decreased stamina	TVR 21 mm; BCPS	ASD closure; interruption MBTS; dilatation LPA; resection subpulmonary tissue
3	23 mo	Asymptomatic ASD	ASD closure	
4	46 mo	Decreased activity and energy	TV re-repair; BCPS	RV resection; right reduction atrioplasty
5	57 mo	Cyanosis; decreased stamina	TVR 25 mm; BCPS	ASD closure; right reduction atrioplasty
6	59 mo	Progressive fatigue	TVR 27 mm	Right reduction atrioplasty; BCPS
7	70 mo	Cyanosis; moderate aortic stenosis	Failed TV repair; TVR 25 mm; aortic valvotomy	ASD closure; right reduction atrioplasty; BCPS; open chest
8	73 mo	Asymptomatic RV enlargement	TV repair; BCPS	Right reduction atrioplasty
9	100 mo	Cyanosis	TVR 25 mm; BCPS	ASD closure; RV resection; right reduction atrioplasty
10	154 mo	Fatigue; cyanosis	TVR 35 mm	ASD closure; right reduction atrioplasty; BCPS; open chest
11	23.2 y	Dyspnea; atrial arrhythmias; transplantation waiting list	TVR 29 mm; BCPS	Modified maze
12	48 y	Dyspnea; atrial arrhythmias; transplantation assessment	TVR 31 mm	BCPS; modified maze; RV resection; right reduction atrioplasty
13	56.2 y	Stroke with ASD	ASD closure; TVR 33 mm	Open chest
14	57.8 y	Dyspnea; cyanosis; atrial fibrillation; transplantation assessment	TVR 31 mm	BCPS; PFO closure; modified maze; right reduction atrioplasty; IABP; open chest

ASD, Atrial septal defect; *BCPS*, bidirectional cavopulmonary shunt; *ECMO*, extracorporeal membrane oxygenation; *IABP*, intra-aortic balloon pump; *LPA*, left pulmonary artery; *MBTS*, modified Blalock-Taussig shunt; *PFO*, patent foramen ovale; *RV*, right ventricular; *TV*, tricuspid valve; *TVR*, tricuspid valve replacement.

Ebstein's Anomaly

TABLE 2. Postoperative course and follow-up after hospital discharge

Patient	Course	Hospital LOS	Follow-up
1	ECMO; operative thrombectomy of TV; delayed sternal closure	28 d	None
2	Uneventful	6 d	78 mo
3	Uneventful	6 d	47 mo
4	Uneventful	4 d	None
5	Uneventful	6 d	29 mo
6	Uneventful	5 d	10 d
7	Open chest; ECMO; conversion to Fontan circulation; Fontan takedown; central aortopulmonary shunt; bleeding; dialysis; withdrawal of support	13 d	Death from MOF with fungal sepsis
8	Uneventful	5 d	24 d
9	Uneventful	8 d	29 d
10	Delayed chest closure; anticoagulation for decreased TV prosthesis leaflet mobility	11 d	21 mo
11	Uneventful	7 d	8 mo
12	IABP	12 d	5 mo
13	Bleeding; RV failure; BCPS; IABP; delayed sternal closure	11 d	None
14	IABP; delayed sternal closure; complete heart block; epicardial lead placement	15 d	3 mo

BCPS, Bidirectional cavopulmonary shunt; *ECMO*, extracorporeal membrane oxygenation; *IABP*, intra-aortic balloon pump; *LOS*, length of stay; *MOF*, multiple organ failure; *RV*, right ventricular; *TV*, tricuspid valve.

Results of the 1.5-ventricle repair for Ebstein Anomaly and the failing right ventricle

Quinoez et al.

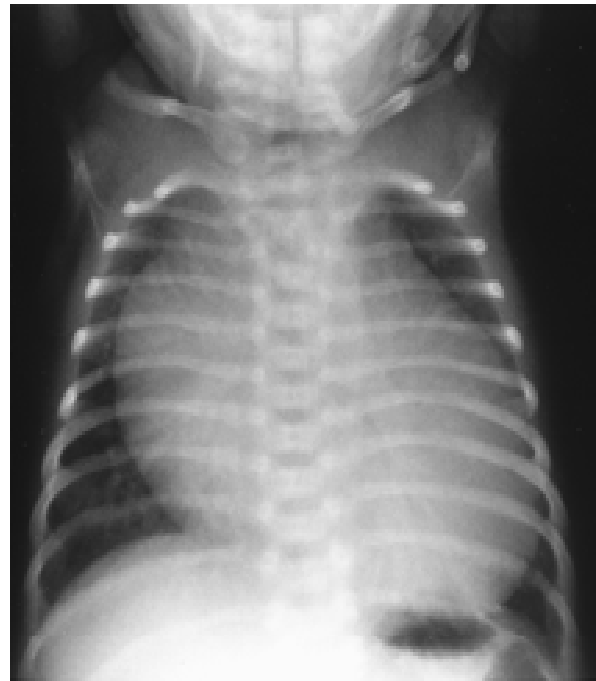
EJCTS 2007;133:1303

Conclusions: The 1.5-ventricle repair can be utilized in patients with severe Ebstein anomaly and impaired right ventricular function who are at high risk for surgical treatment. We believe the bidirectional cavopulmonary shunt may be considered as a planned procedure, as an intraoperative salvage maneuver, or as an alternative to cardiac transplantation in selected patients.

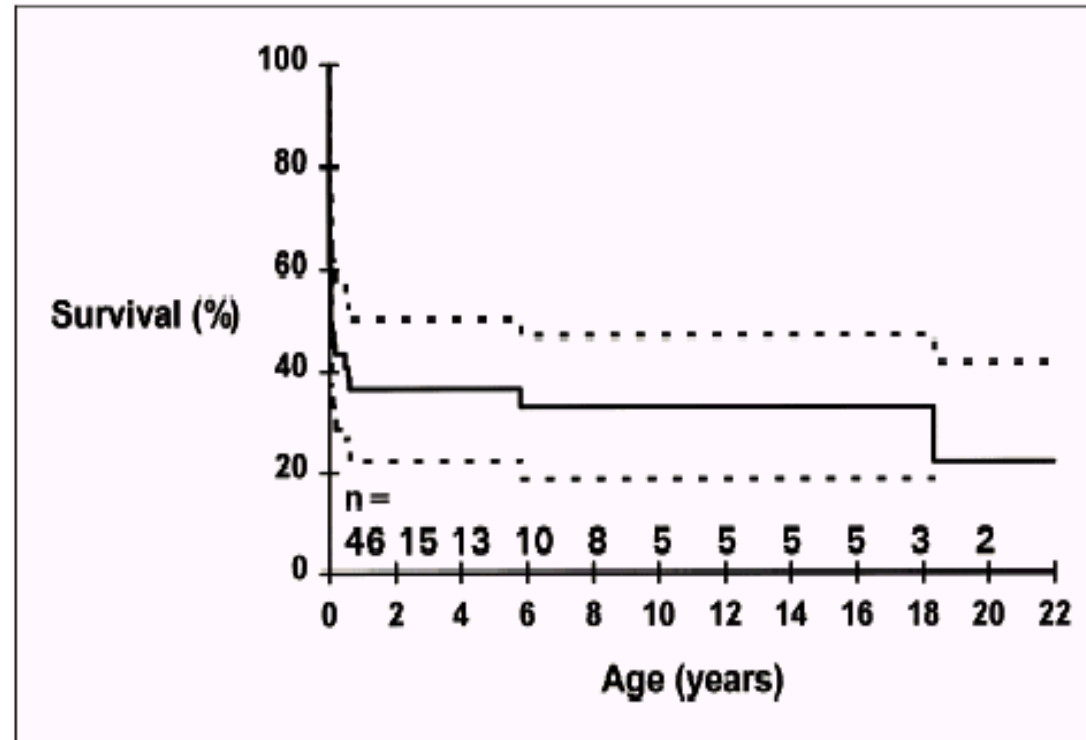
Repair of Ebstein anomaly for Symptomatic Neonates

Ebstein's Anomaly

Neonatal Ebstein's Anomaly



Outcome in Cyanotic Neonate With Ebstein's Anomaly



Kaplan-Meier survival curve indicating percent survival increasing age

Indicators of Poor clinical outcome

- Functional or anatomic pulmonary atresia
- Combined cardiac anatomy
 - Pulmonary Atresia, subpulmonic stenosis, valvular PS, PDA, VSD, ASD
- Cyanosis, low SaO₂
- Severe TR
- High CT ratio
- Decreased pulmonary blood flow on initial CXR
- Acidosis
- Poor RV function

Symptomatic Neonate with Ebstein's Anomaly

- Try to bring the infant through the first week of life by reducing pulmonary vascular resistance with NO or other pulmonary vasodilator
- If it is not successful, surgery

Surgery for Neonates

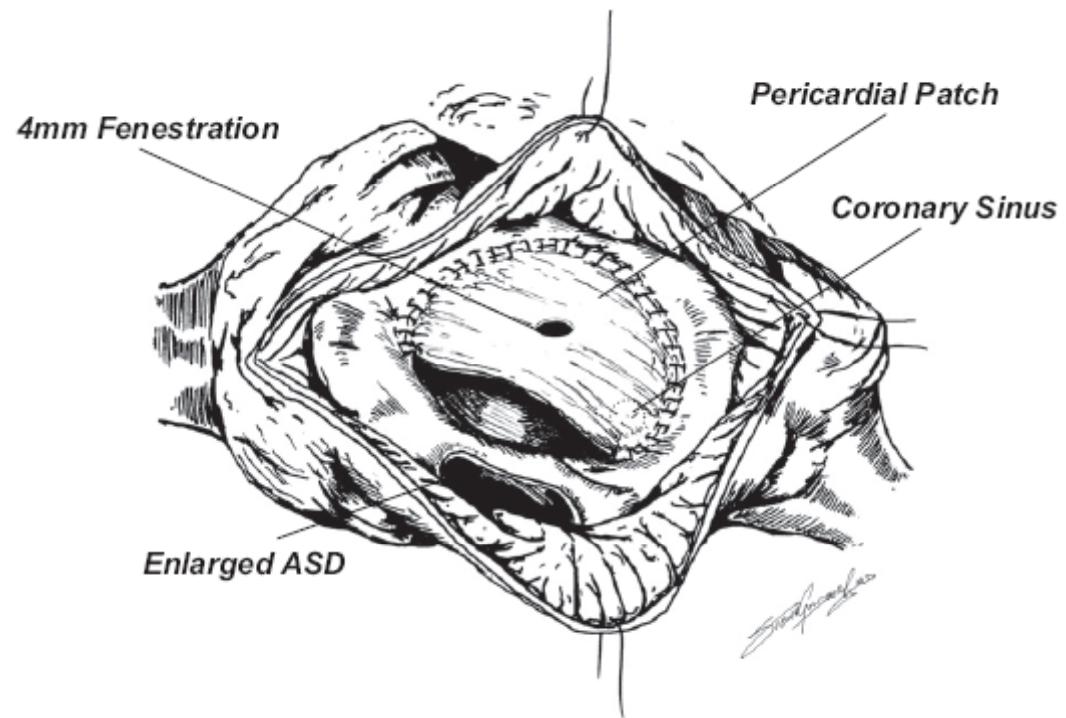
Ebstein's anomaly appearing in the neonate

Starnes et al.

J Thorac Cardiovasc Surg 1991;101:1082

- Single ventricle palliation for severely ill neonates
 - Pericardial patch closure of the tricuspid orifice with a 4 mm fenestration in the patch
 - Atrial septectomy
 - Creation of a 4 mm PTFE aorto-pulmonary shunt
 - Ligation of MPA if pulmonary regurgitation is present
- Fontan completion later in life

Ebstein's Anomaly



- Hospital survival around 70% in the neonatal population

Surgery for Neonates

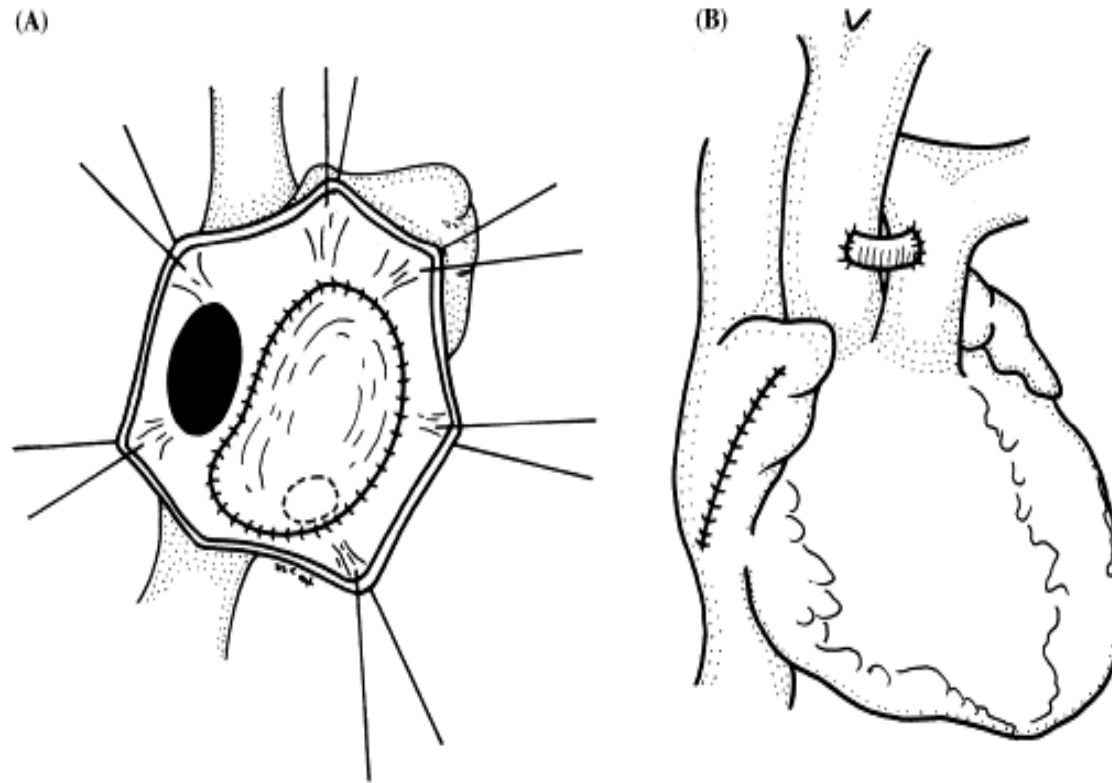
Conversion of complex neonatal Ebstein's anomaly into functional tricuspid or pulmonary atresia

Van Son et al.

Eur J Cardiovasc Surg 1998;13:280-5

- Early univentricular repair within the first week of life
- 5 symptomatic patients newborns
- No early mortality
- Fontan completion later in life

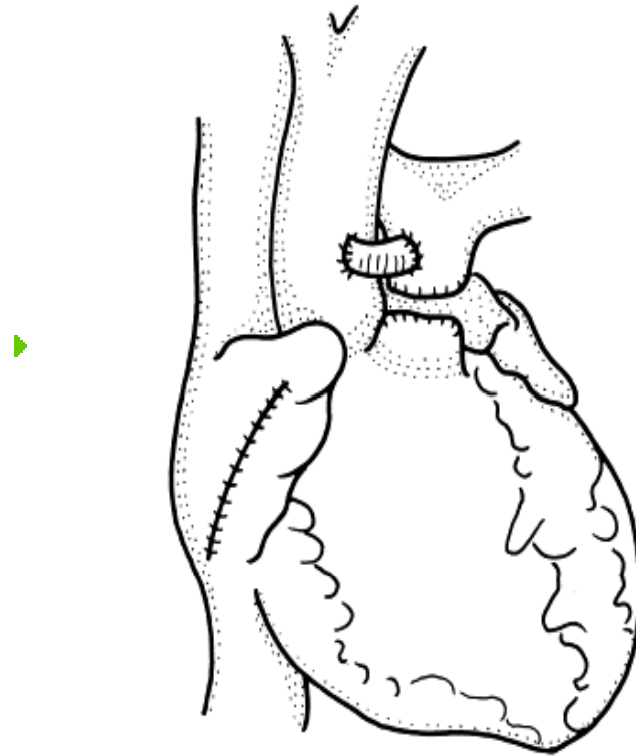
Surgery for Neonates



Conversion of complex neonatal Ebstein's anomaly into functional tricuspid or pulmonary atresia

Eur J Cardiovasc Surg 1998;13:280-5

Surgery for Neonates



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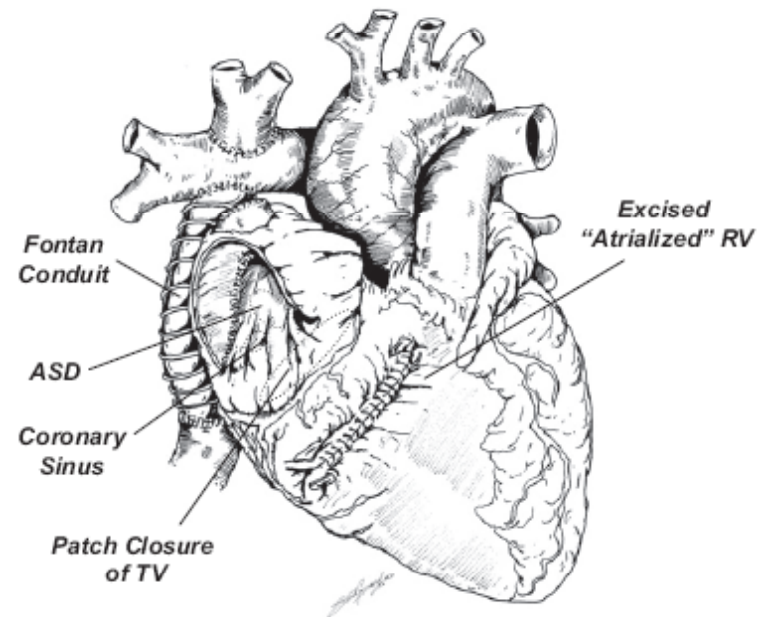
RV Exclusion

Total right ventricular exclusion procedure: An operation for isolated congestive right ventricular failure.

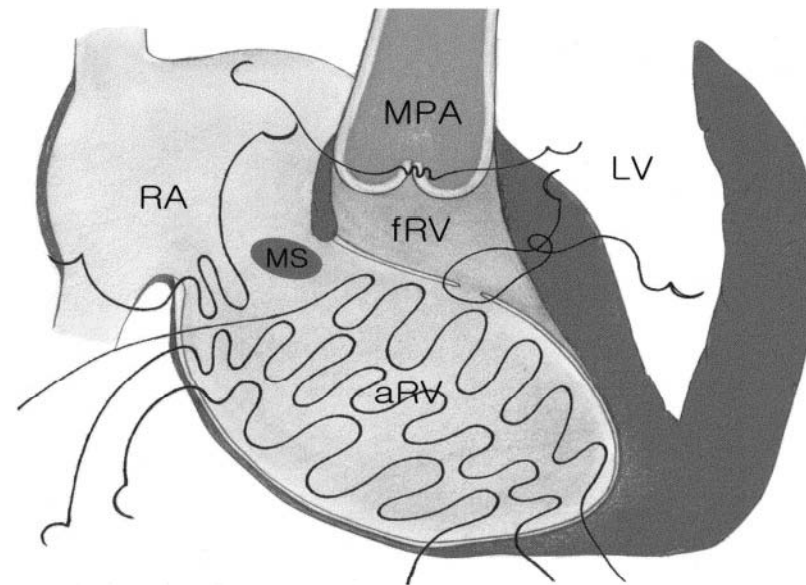
Sano et al.

J Thorac Cardiovasc Surg 2002;123:640

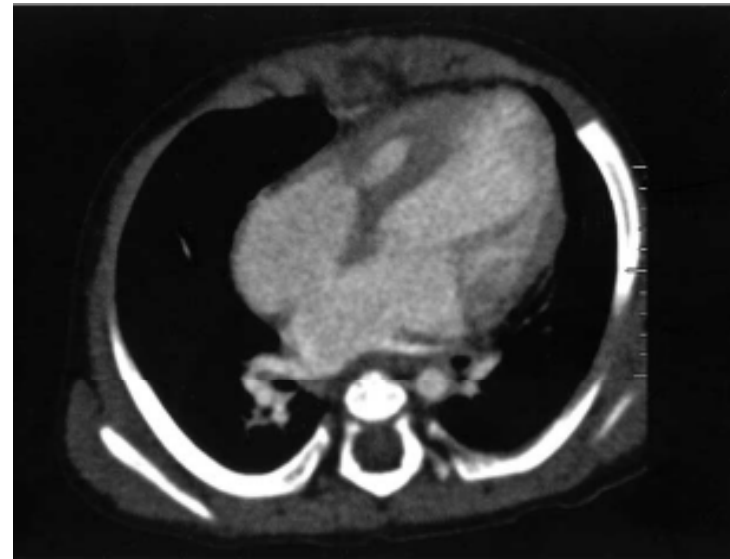
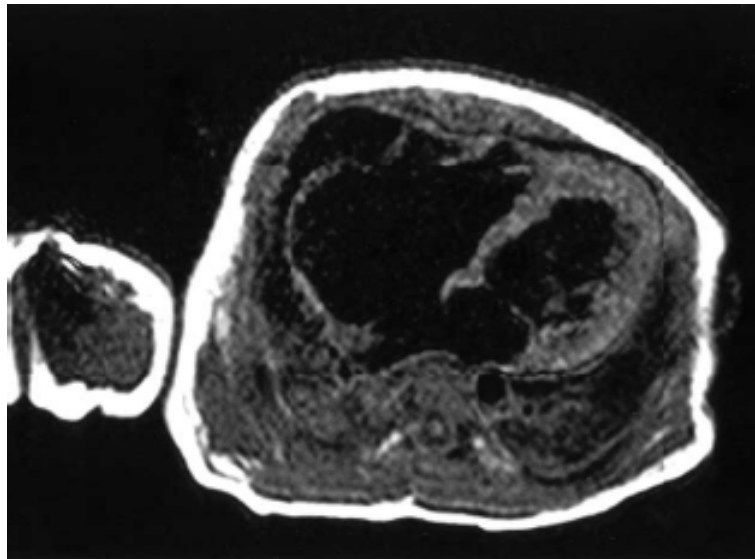
- PTFE patch closure of the TV orifice
- Baffling of the coronary sinus flow through an ASD by way of a folded portion of RA wall



Neonatal stenotic Ebstein's anomaly: A novel technique of right ventricular exclusion



Neonatal stenotic Ebstein's anomaly: A novel technique of right ventricular exclusion



Bi-ventricular Repair for Neonates

Repair of Ebstein's Anomaly in the Symptomatic Neonate: An Evolution of Technique With 7-Year Follow-Up

Christopher J. Knott-Craig, MD, Edward D. Overholt, MD, Kent E. Ward, MD,
Jeremy M. Ringewald, MD, Sherri S. Baker, MD, and Jerry D. Razook, MD

Sections of Thoracic and Cardiovascular Surgery and Pediatric Cardiology, Children's Hospital at Oklahoma University Medical Center, Oklahoma City, Oklahoma

Ann Thorac Surg 2002;73:1786

Bi-ventricular Repair for Neonates

Repair of Ebstein's anomaly in the symptomatic neonate:
An evolution of technique with 7-year follow-up

Knott-Craig et al.

Ann Thorac Surg 2002;73:1786

Principles of Repair

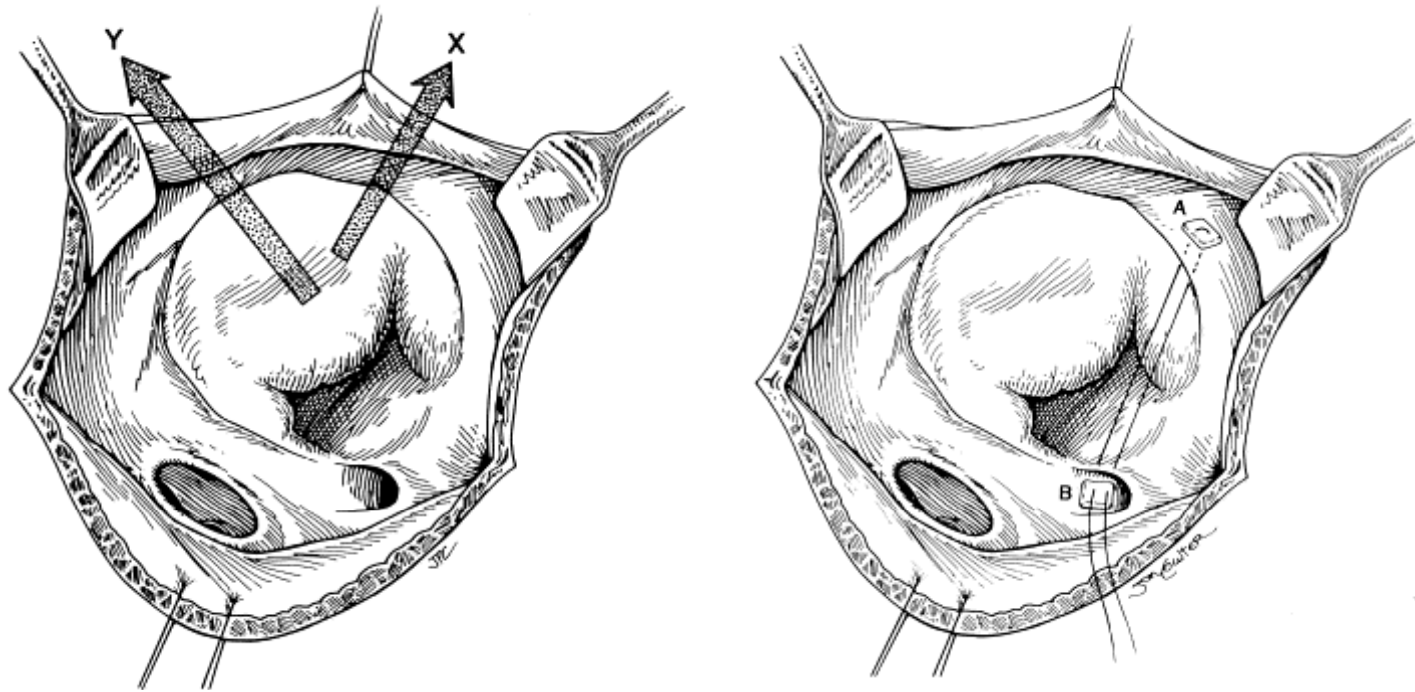
Creation of a competent tricuspid valve, based on the anterior leaflet

Reducing the RA volume

Closing the ASD, leaving a small fenestration

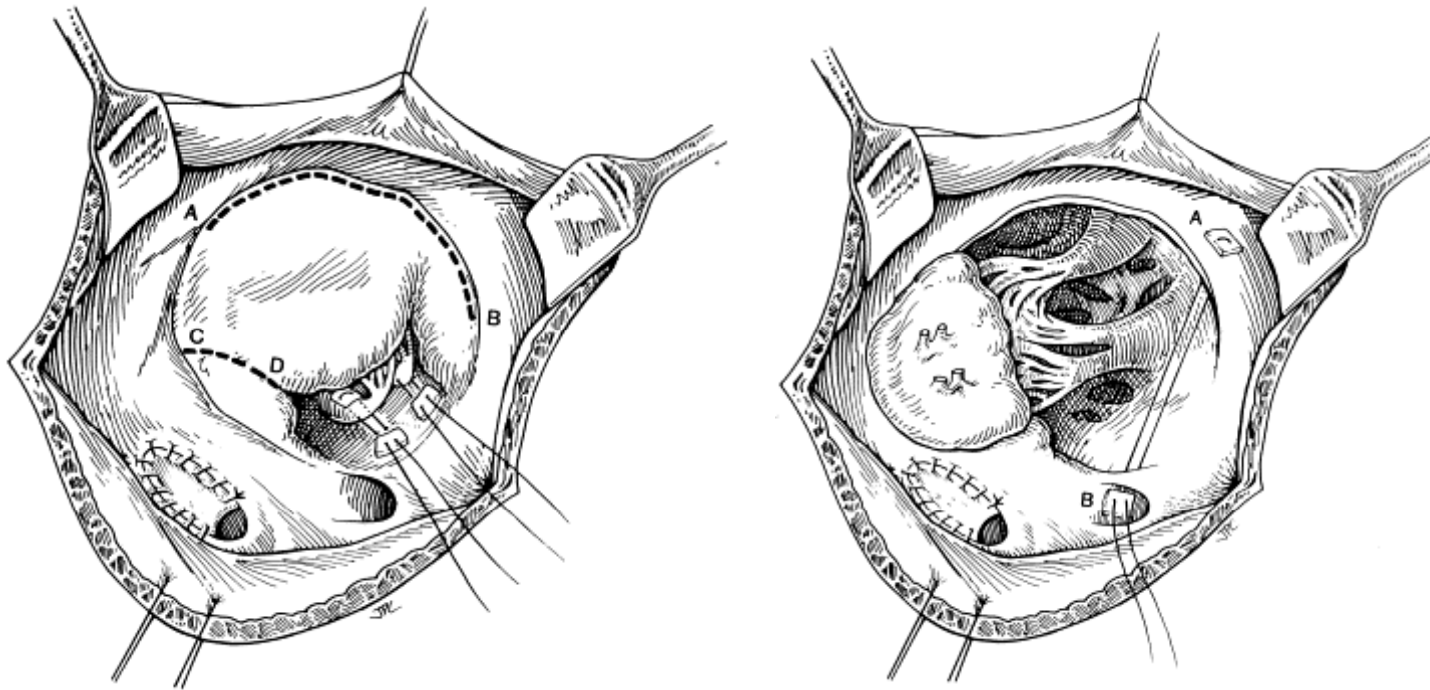
Repairing all associated defects, including pulmonary atresia

Ebstein's Anomaly



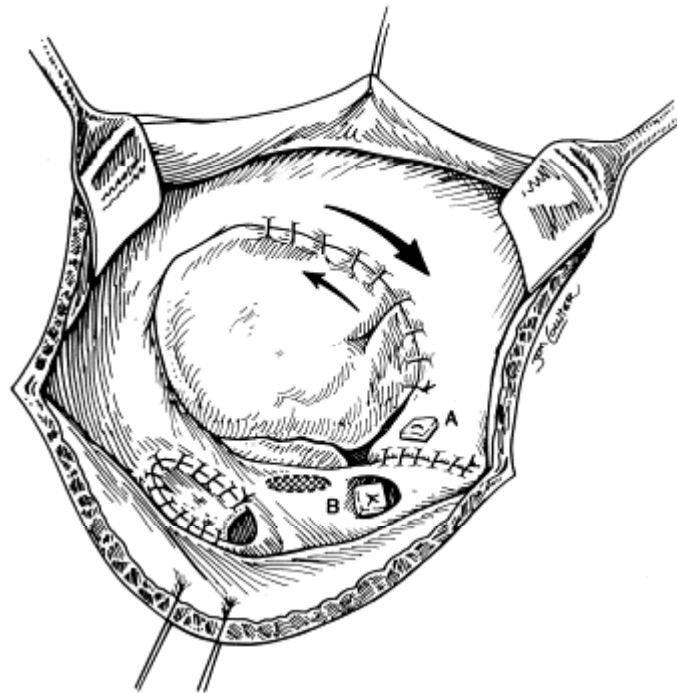
Knott-Craig et al. Ann Thorac Surg 2002;73:1786

Ebstein's Anomaly



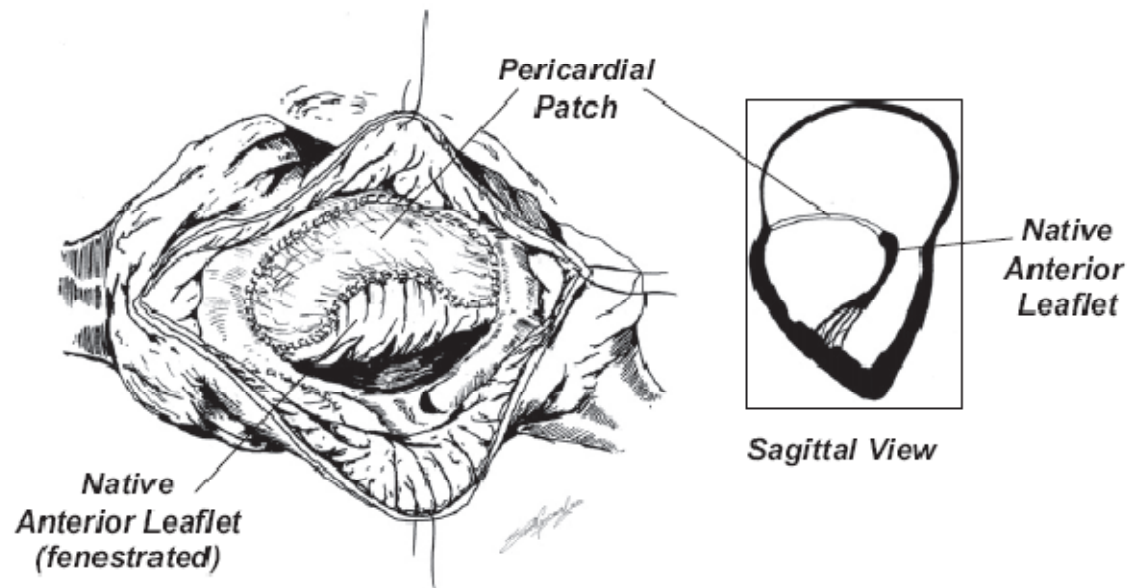
Knott-Craig et al. Ann Thorac Surg 2002;73:1786

Ebstein's Anomaly



Knott-Craig et al. Ann Thorac Surg 2002;73:1786

Ebstein's Anomaly



Knott-Craig et al. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2007;10:112

Repair of Ebstein Anomaly in Symptomatic Neonate

	Wt	Age/Sex	Repair	Outcome
1	3.2 kg	19d M	Danielson, VSD closure	Alive, 7 y/o
2	2.9 kg	16d M	Danielson, PV dilation	Alive, 7 y/o
3	3.1 kg	6d M	De Vega, RVOT patch	Alive, 7 y/o
4	2.8 kg	2d M	De Vega	Alive, 2 y/o
5	3.8 kg	2.5m F	Danielson, TVR	Alive, 17 m/o
6	2.1 kg	7d F	Danielson, De Vega, PVR	Died, Coronary injury (?)
7	3.1 kg	5d F	Danielson, De Vega	Alive, 15 m/o
8	6.4 kg	4m M	Chauvaud, Hemi-Fontan	Alive, 1 y/o

Knott-Craig et al. Ann Thorac Surg 2002;73:1786

Bi-ventricular Repair for Symptomatic Neonates

Management of Neonatal Ebstein's Anomaly

Knott-Craig et al.

Semin Thorac Cardiovasc Surg Pediatric Card Surg Ann 2007;10:112

1994 ~

22 symptomatic neonate and young infants

16 hospital survivors (73%)

No late death

Reoperation: Hemi-Fontan, TVR (2)

Arrhythmia

- WPW Syndrome (10~30%)
- Atrial ectopic tachycardia
- Atrial flutter / fibrillation
- Atrial reentry tachycardia
- AVNRT: Perinodal cryoablation after institution of CPB

Arrhythmia

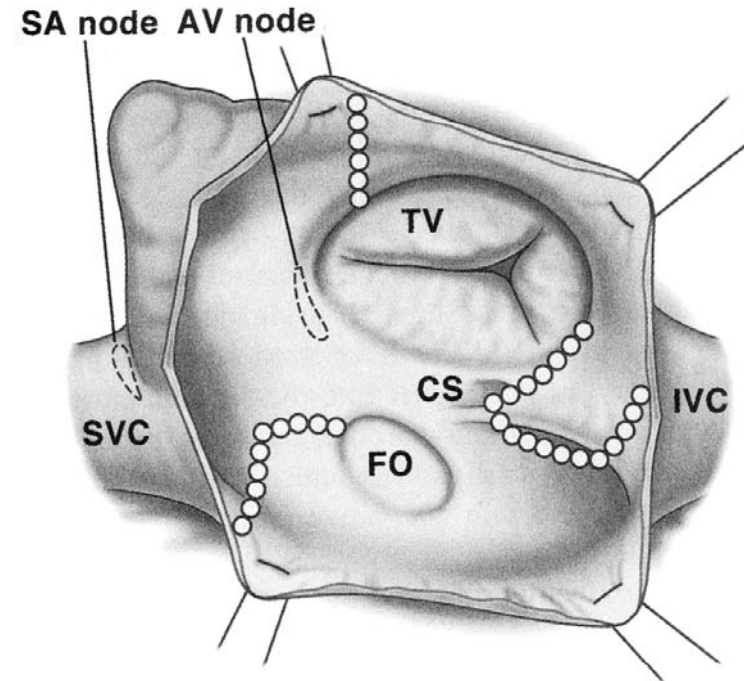
- WPW syndrome
 - Previous history of supraventricular tachycardia
 - ECG finding of pre-excitation syndrome
 - Electrophysiologic study
 - Surgical ablation
 - RF catheter ablation

Atrial tachyarrhythmia

Atrial fibrillation / flutter
(paroxysmal / chronic)

Right-side maze

Cryoablation procedure
with include RA
isthmus



Different Surgical Techniques for TV repair in Ebstein's Anomaly

Technique	Plication of atrialized chamber	Detachment of leaflet	Annuloplasty
Hardy	Transverse	No	No
Danielson	Transverse	No	Yes
Carpentier	Longitudinal	Sliding plasty	Ring suggested
Quaegebeur	Longitudinal	Sliding plasty	No
Sebening	None	No	No
Hetzer	None	Rarely required	Partial annulorrhapy
Wu	Excision	Reattached to normal annulus	Yes

Surgical Repair for Ebstein's Anomaly

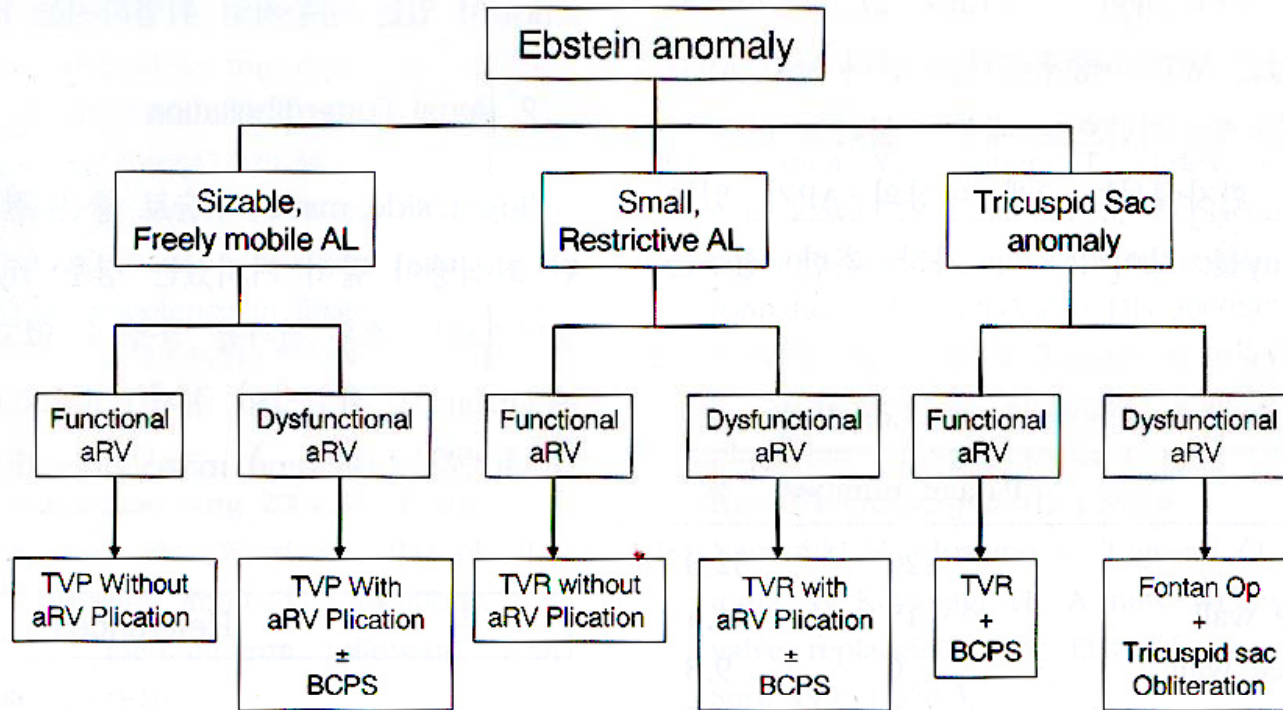
- Endless anatomic variability of Ebstein anomaly.
- Limitations and incomplete satisfaction with each of the repair method available
- The best way for the surgeon to optimize his or her ability to repair the Ebstein valve is to be familiar with all of the available techniques and to selectively apply principles and portions of the different techniques to the specific anatomy encountered in the operating room.

Ebstein's Anomaly

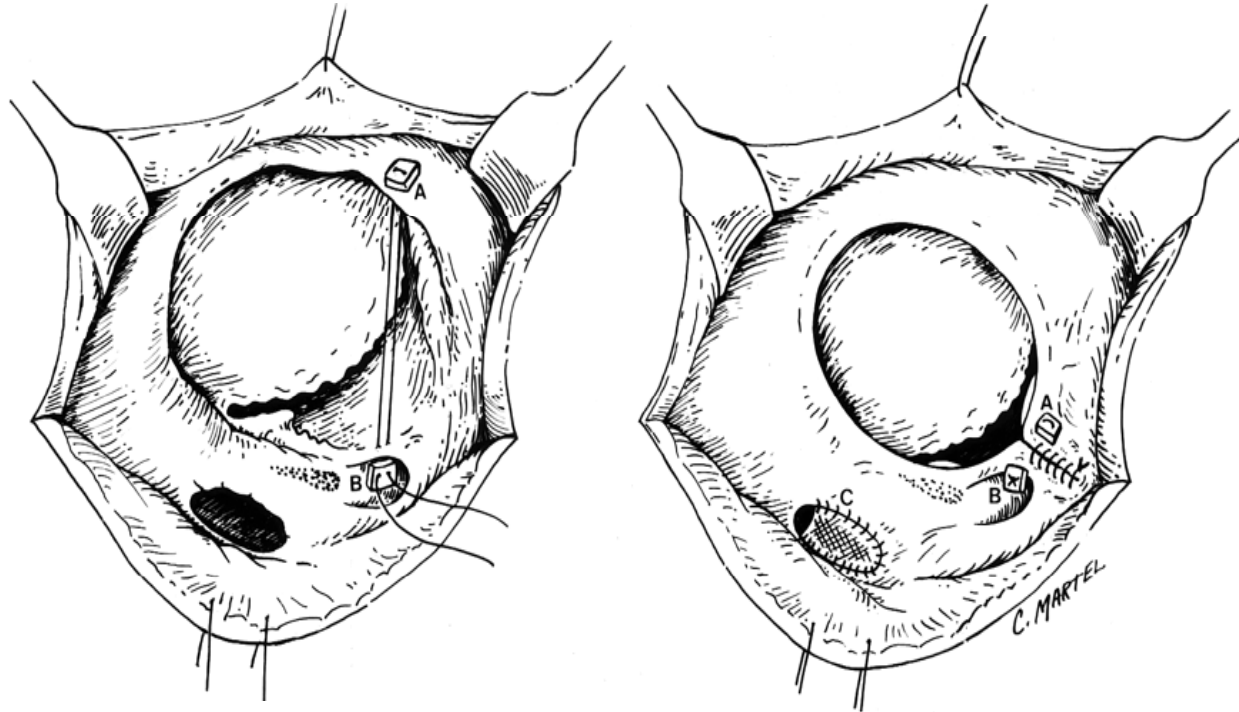
Different Surgical Techniques for TV repair in Ebstein's Anomaly

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Surgical Options for Ebstein Anomaly



Ebstein's Anomaly



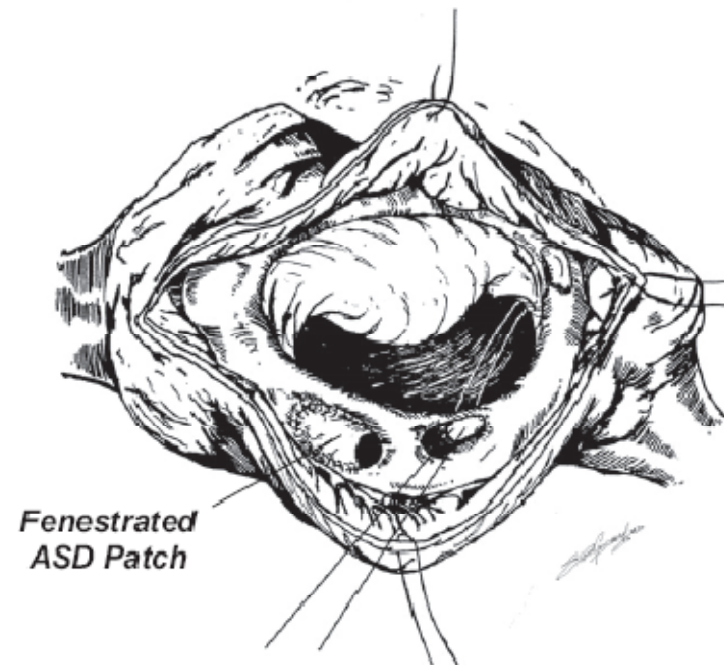
Technique of repair in neonates with Ebstein's anomaly.

Ann Thorac Surg 2000;69:1505-1510

Combined Anomaly

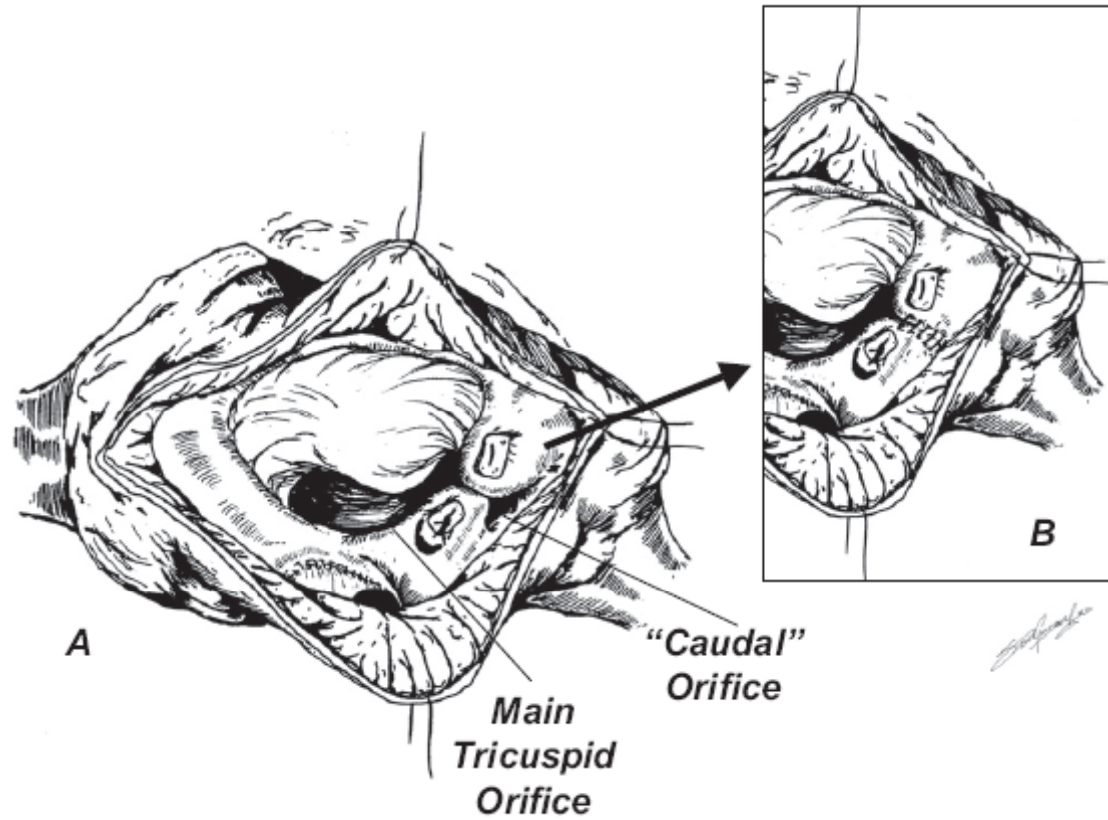
- ASD, PFO
- Pulmonary stenosis
- Pulmonary atresia
- VSD
- AVSD
- TOF
- Ventricular pre-excitation (WPW) syndrome

Ebstein's Anomaly



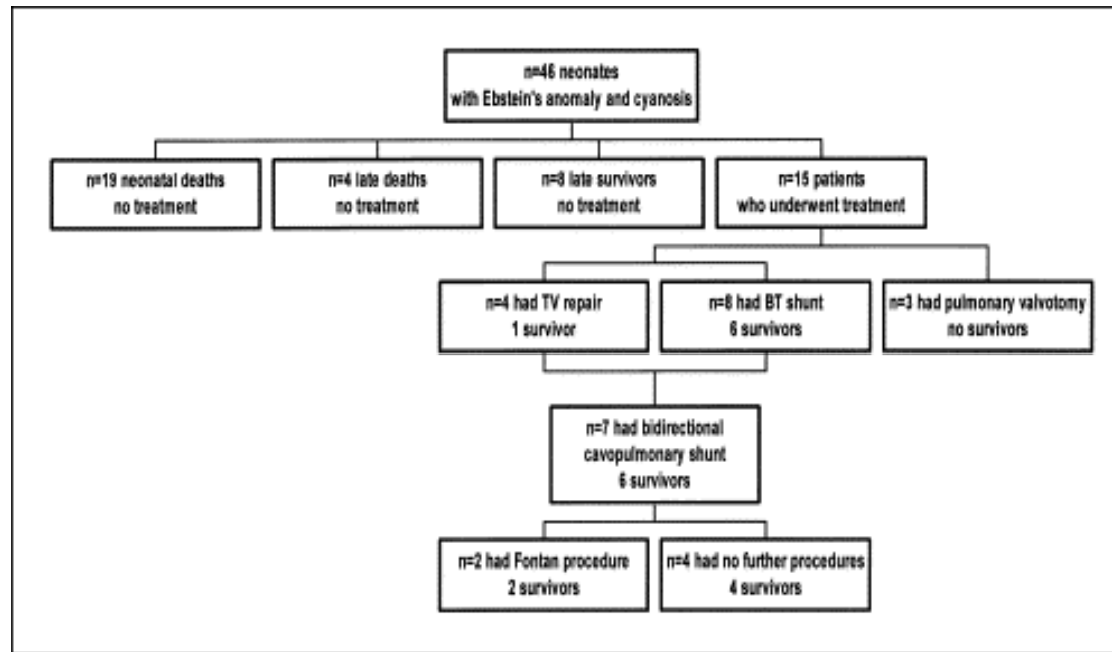
Knott-Craig et al. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2007;10:112

Ebstein's Anomaly



Knott-Craig et al. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2007;10:112

Outcome in Cyanotic Neonate With Ebstein's Anomaly



Outcome of neonates with Ebstein's anomaly presenting with cyanosis.

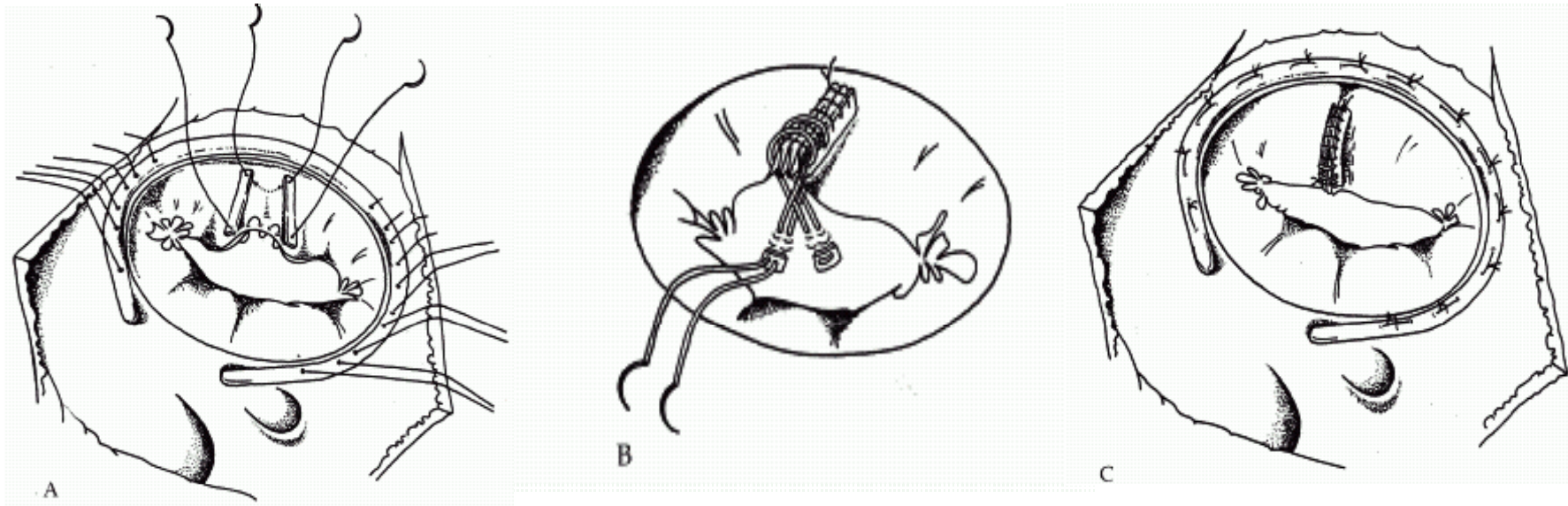
Ebstein's Anomaly

- Ebstein's anomaly is a rare congenital heart defect reported to occur in 0.2% of live births.
- First described by Wilhelm Ebstein in 1866 19 year old laborer -
- Failure of delamination of leaflets
- Whitish and thickened myocardium

Surgical Repair

- In 1959, repair of the tricuspid valve in two patients: patients died.
- Barnard and Schrire. Surgical correction of Ebstein's malformation with prosthetic valve. *Surgery* 1963;54:302
- 1964 Hardy TV repair

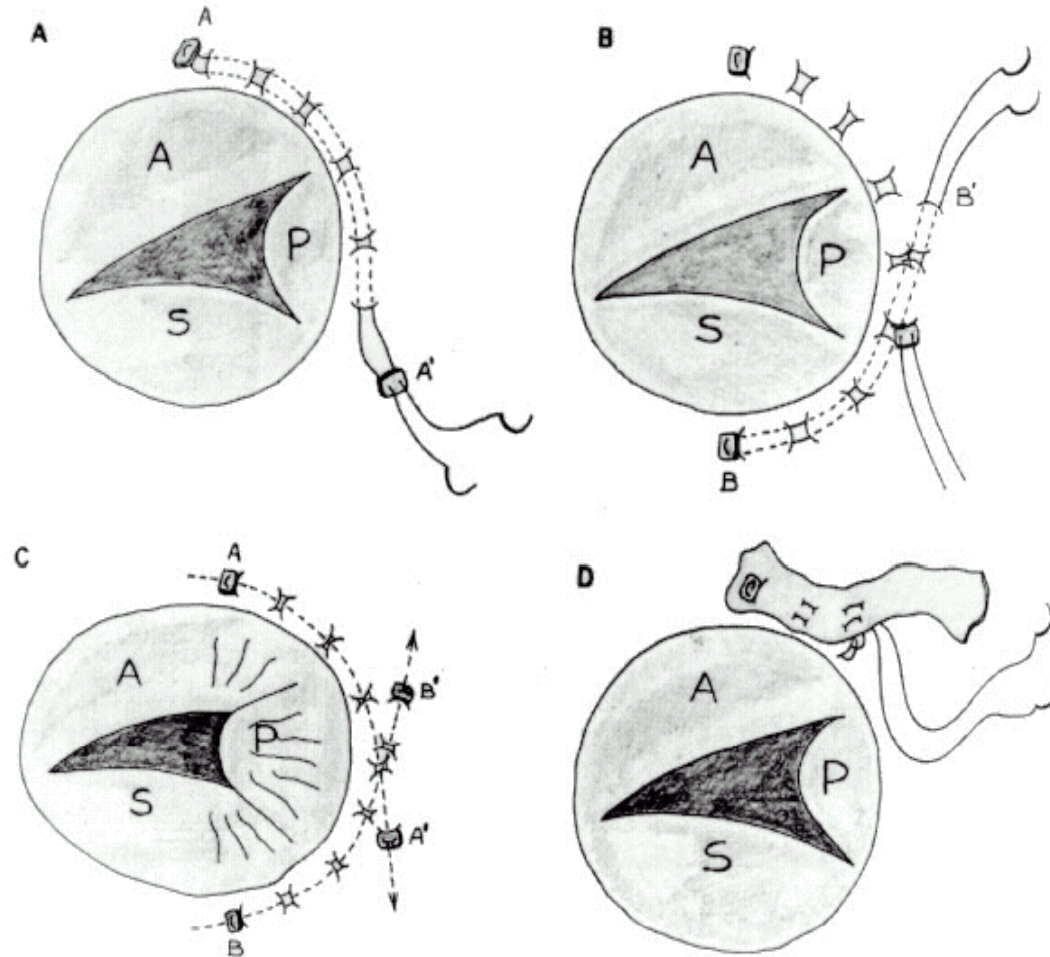
Surgical Management of Isolated Congenital Tricuspid Regurgitation



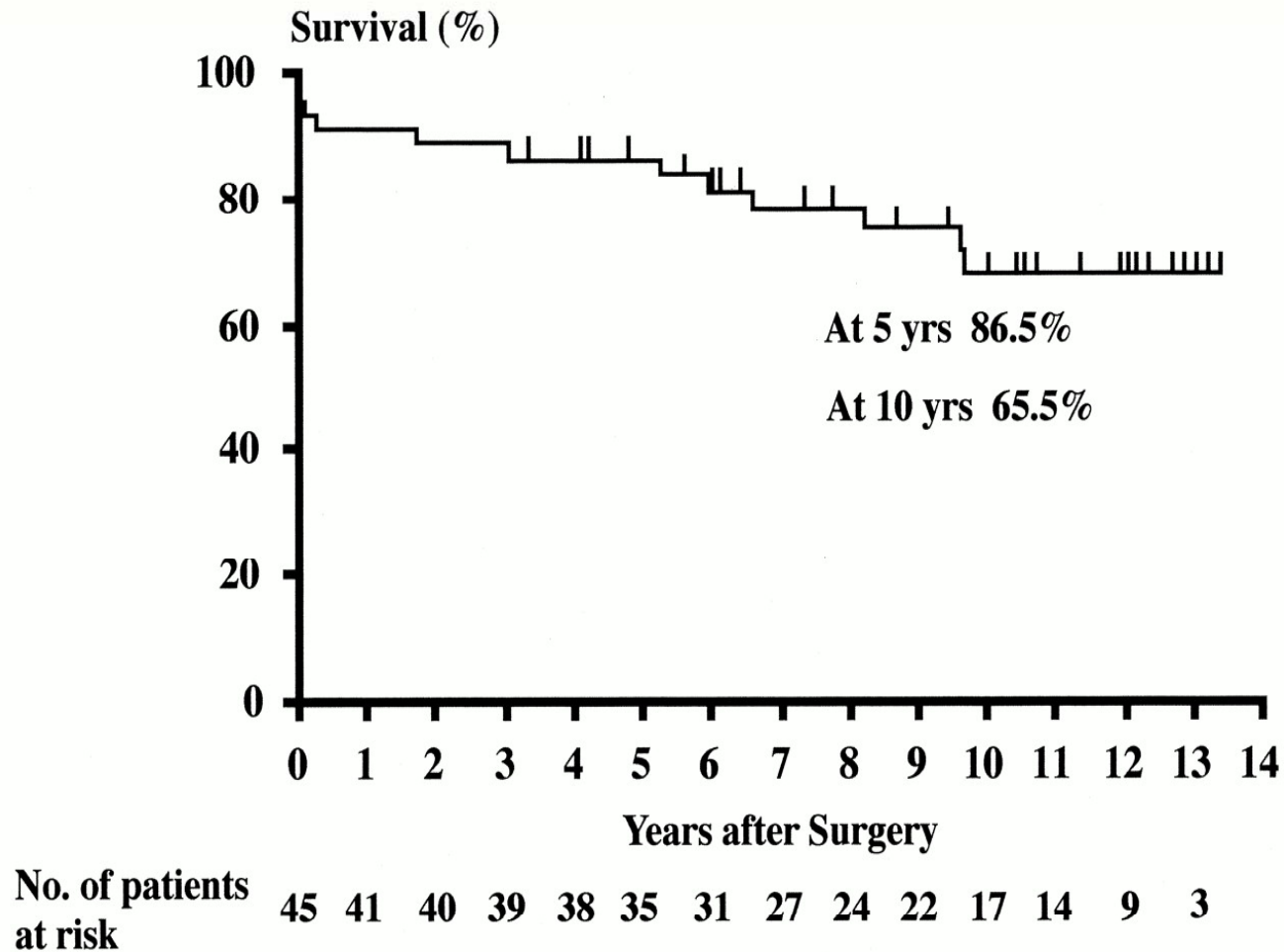
(a) Gathering plasty of anterior-posterior leaflet complex with continuous suture of 5-0 Prolene reinforced by autologous pericardial strip.

(b) Implantation of artificial chordae of expanded polytetrafluoroethylene suture. (c) Completed procedures with Carpentier-Edwards ring, gathering valvuloplasty, and artificial chordae.

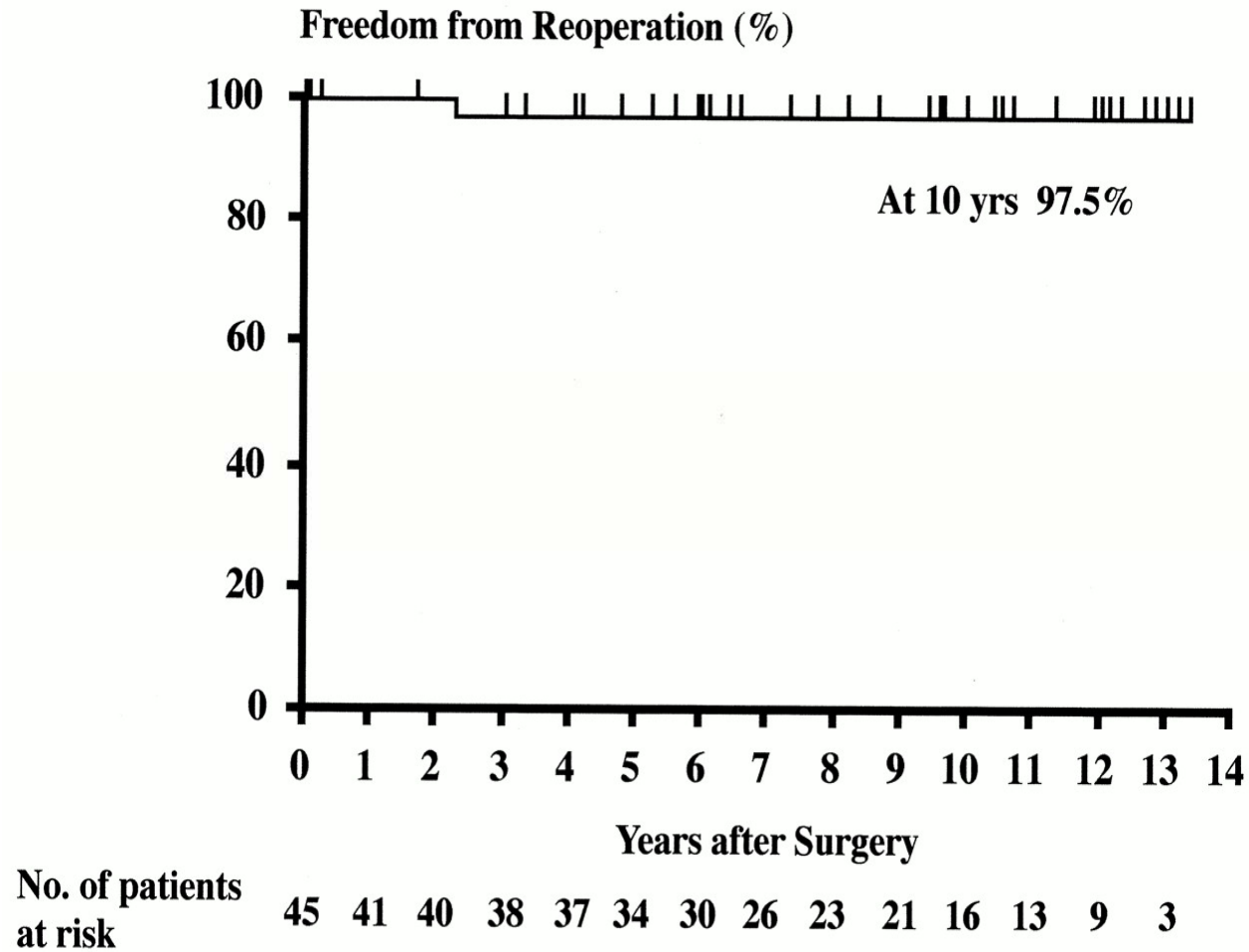
Correction of Functional Tricuspid Insufficiency by Means of A Limited Posterior Crossed Suture Annuloplasty



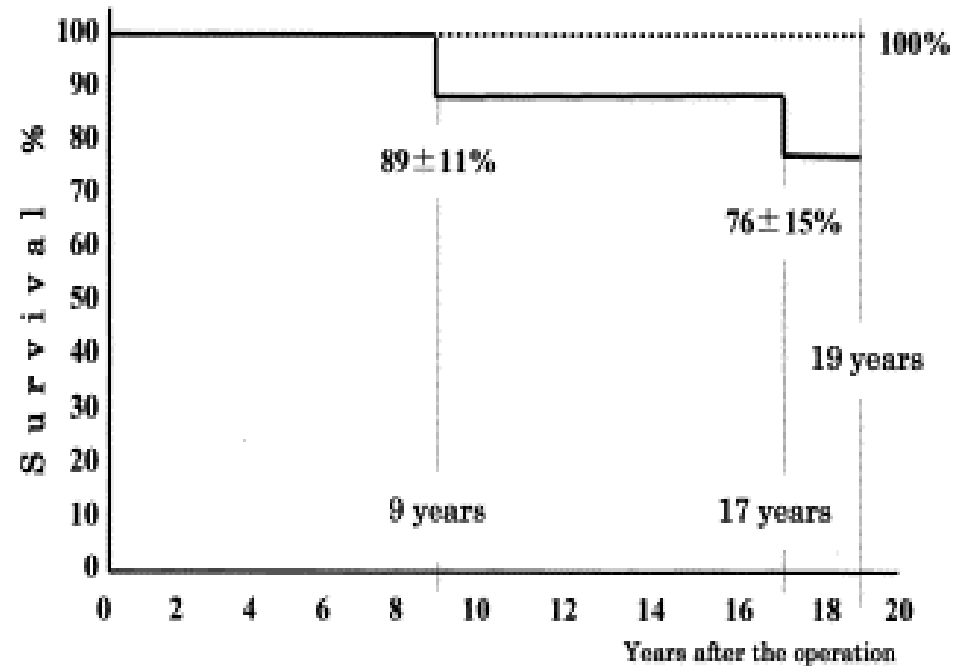
Long-term follow-up after Carpentier-Edwards ring annuloplasty for tricuspid regurgitation



Long-term follow-up after Carpentier-Edwards ring annuloplasty for tricuspid regurgitation

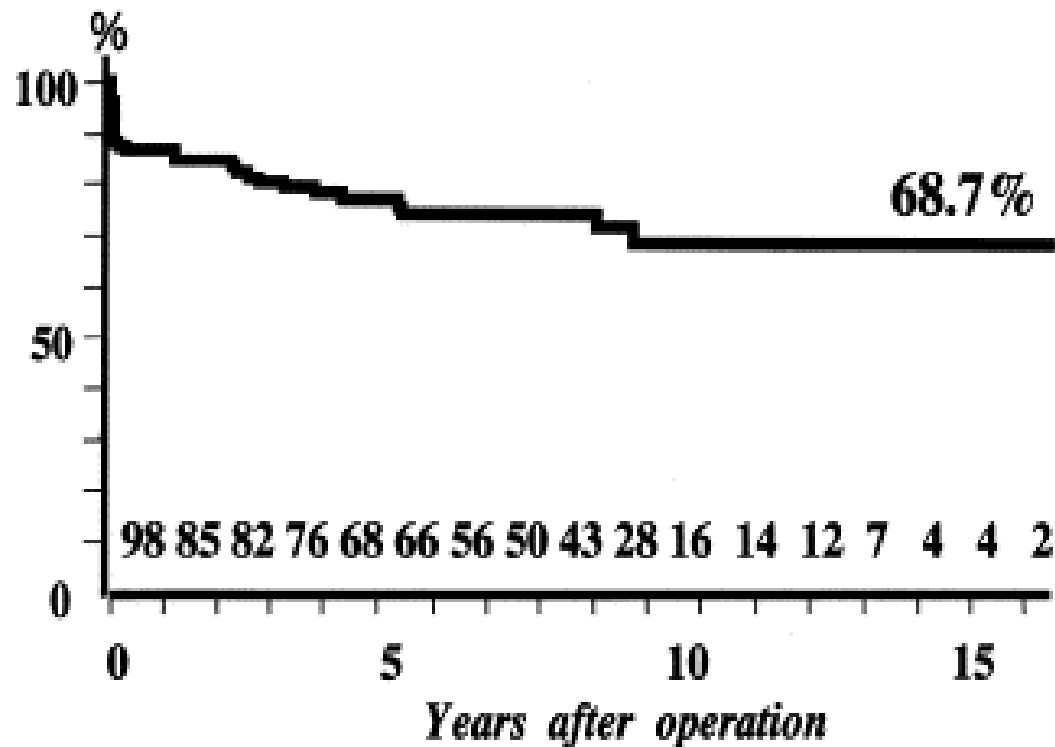


Tricuspid valve supra-annular implantation



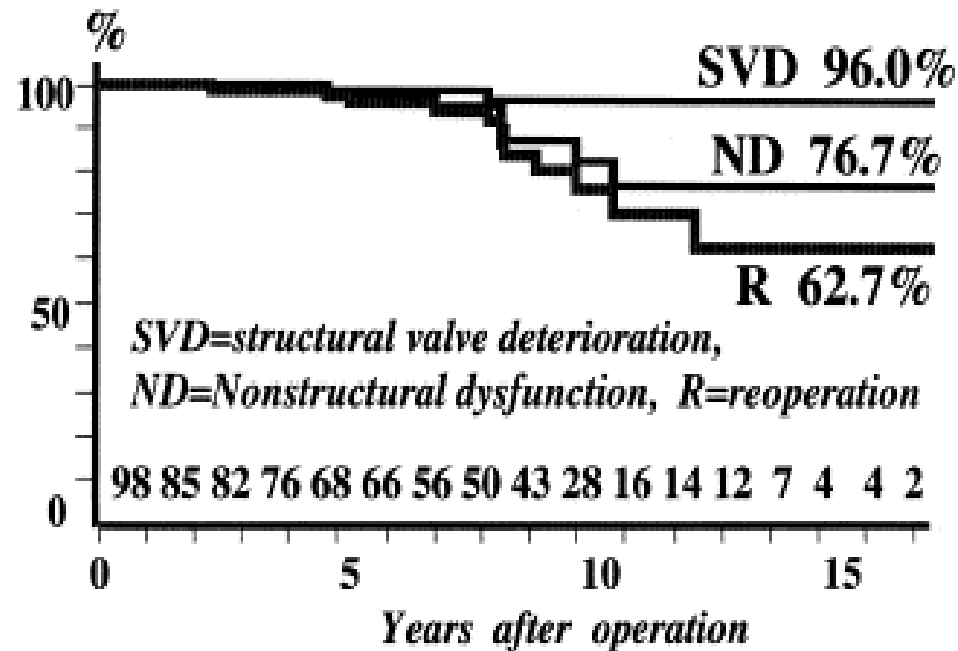
The actuarial survival rate, freedom from structural valve deterioration, and reoperation at 19 years after the operation.

TVR with Bioprosthesis: long-term results and cause of valve dysfunction



Actuarial survival curve, including operative deaths *Ann Thorac Surg* 2001;71:105-109

TVR with bioprostheses: long-term results and causes of valve dysfunction



Actuarial rates for freedom from reoperation, structural valve deterioration, and nonstructural dysfunction.

Two Dimensional Echocardiography (Four Chamber View)

