KYACC Meeting
September 10, 2022

Ebstein's Anomaly
Surgical Repair and Outcomes

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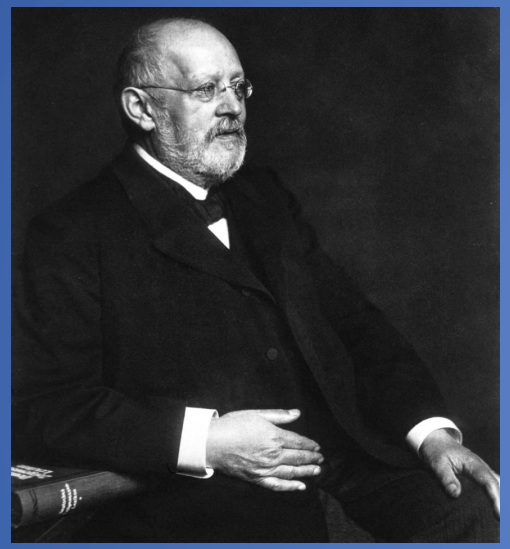
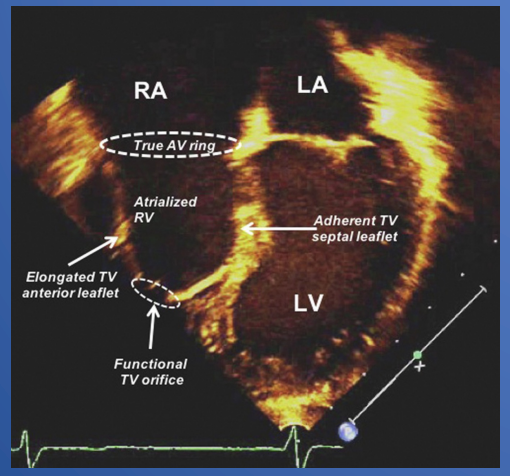
Ebstein's Anomaly





First described in 1866

19 year old male

Cyanosis, dyspnea, palpitations

Autopsy: abnormal tricuspid valve, atrialized RV, dilated RA, PFO

Ebstein's Anomaly

- Rare - < 1% of patients born with congenital heart disease
  - 1/200,000 live births
  - Individual center experience limited, especially for neonates and infants

- Anatomical defect involves both the tricuspid valve and the right ventricle
  - Tricuspid valve
    - Failed delamination of TV leaflets with apically displaced septal and inferior leaflets
    - Redundant anterior leaflet
    - Dilated true right atrioventricular annulus
    - Tricuspid regurgitation – restricted TV leaflet tissue
  - Right ventricle
    - Two chambers – atrialized RV (between displaced TV and true annulus) and functional RV (distal to TV)
    - RV myopathy

Ebstein's Anomaly

Associated defects affect timing and severity of symptoms

- ASD (80-90%)
- Accessory AV conduction pathways (15%)
- Pulmonary atresia or stenosis
- ccTGA
- VSD
- MV anomalies
- LV fibrosis

Ebstein's Anomaly

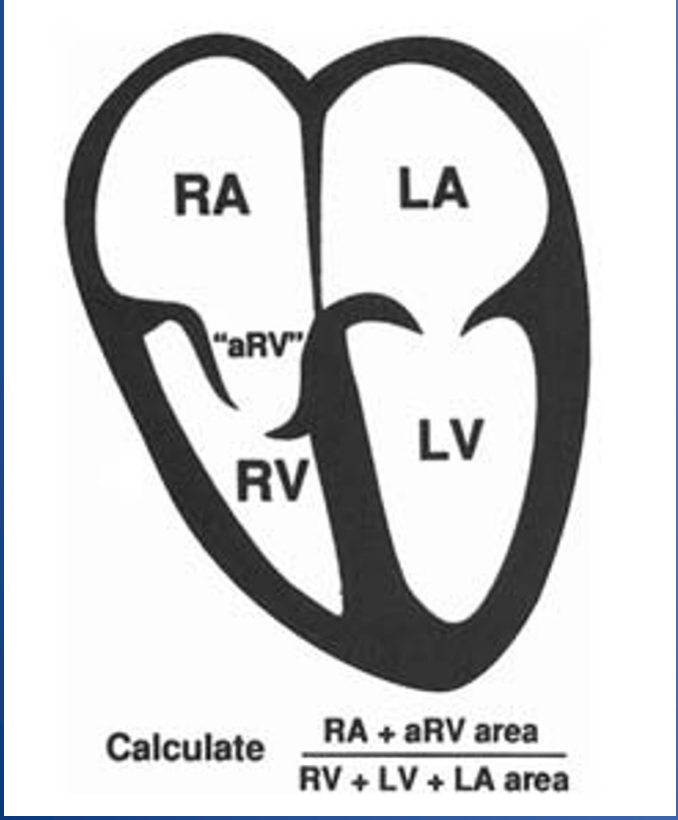
Diversity in anatomy and associated defects

- Results in variability in severity and presentation
  - From a life-threatening condition in the neonate
  - To few to no symptoms into adulthood
  - And in between
- Presentation – essentially two forms
  - Neonate
  - Older children and adults
- Surgery and outcomes differ
  - Neonate
  - Older children and adults

Outcome in Neonates With Ebstein's Anomaly

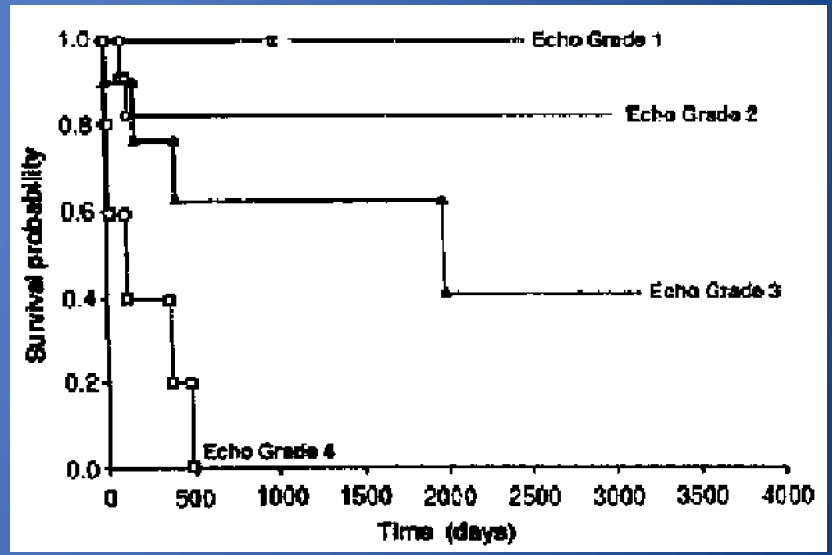
DAVID S. CELERMAJER, MB, BS, MSc, FRACP, SEAMUS CULLEN, MB, CHB, MRCPI,
IAN D. SULLIVAN, MB, CHB, BMEDSC, FRACP, DAVID J. SPIEGELHALTER, PHD,*
RICHARD K. H. WYSE, PHD, JOHN E. DEANFIELD, MB, BCHIR, MRCP
London and Cambridge, England

JACC 19:1041-6, 1992

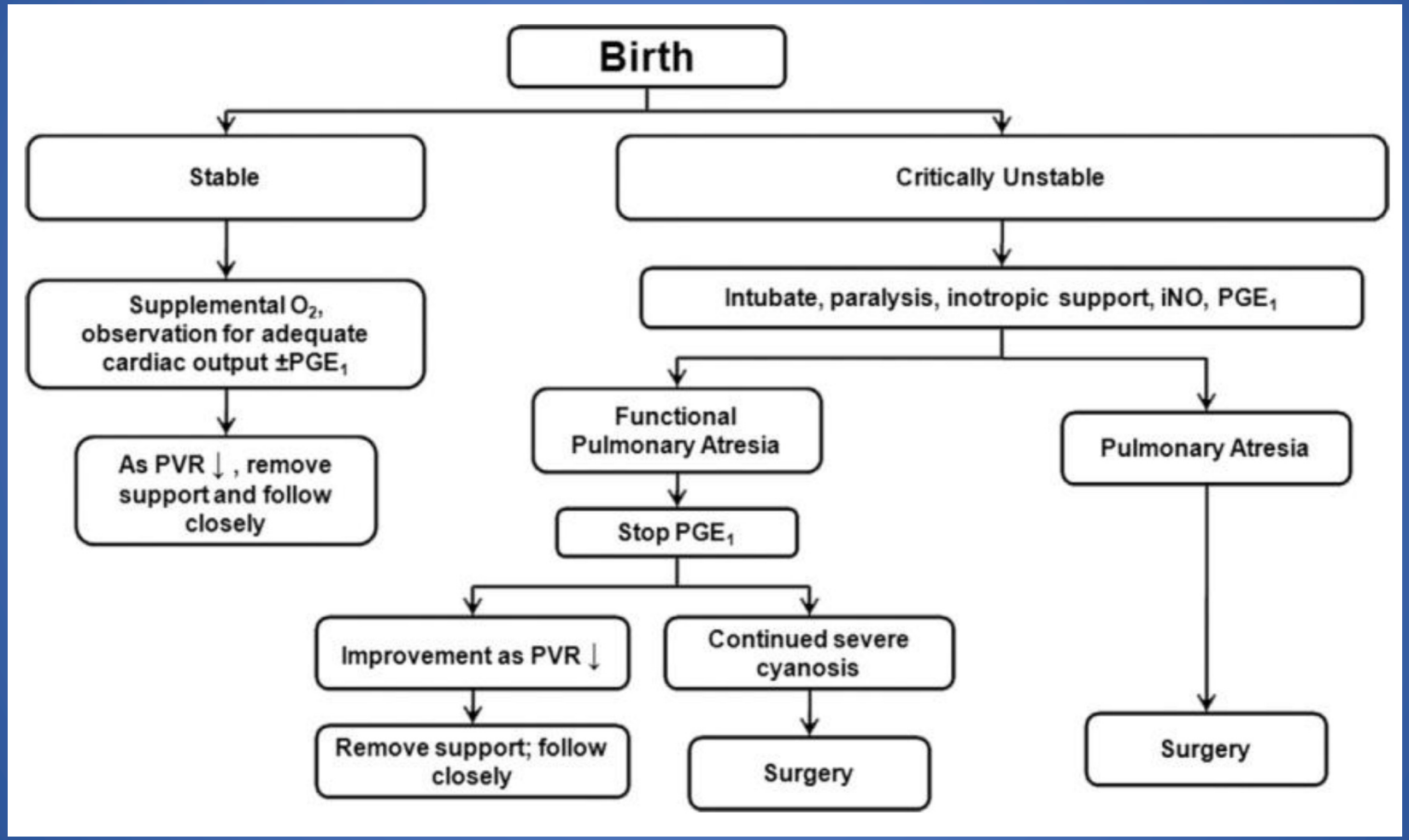


GOSE Score

Grade 1 <0.5
Grade 2 0.5 – 0.99
Grade 3 1.1-1.4
Grade 4 > 1.5



Ebstein's Diagnosed at Birth

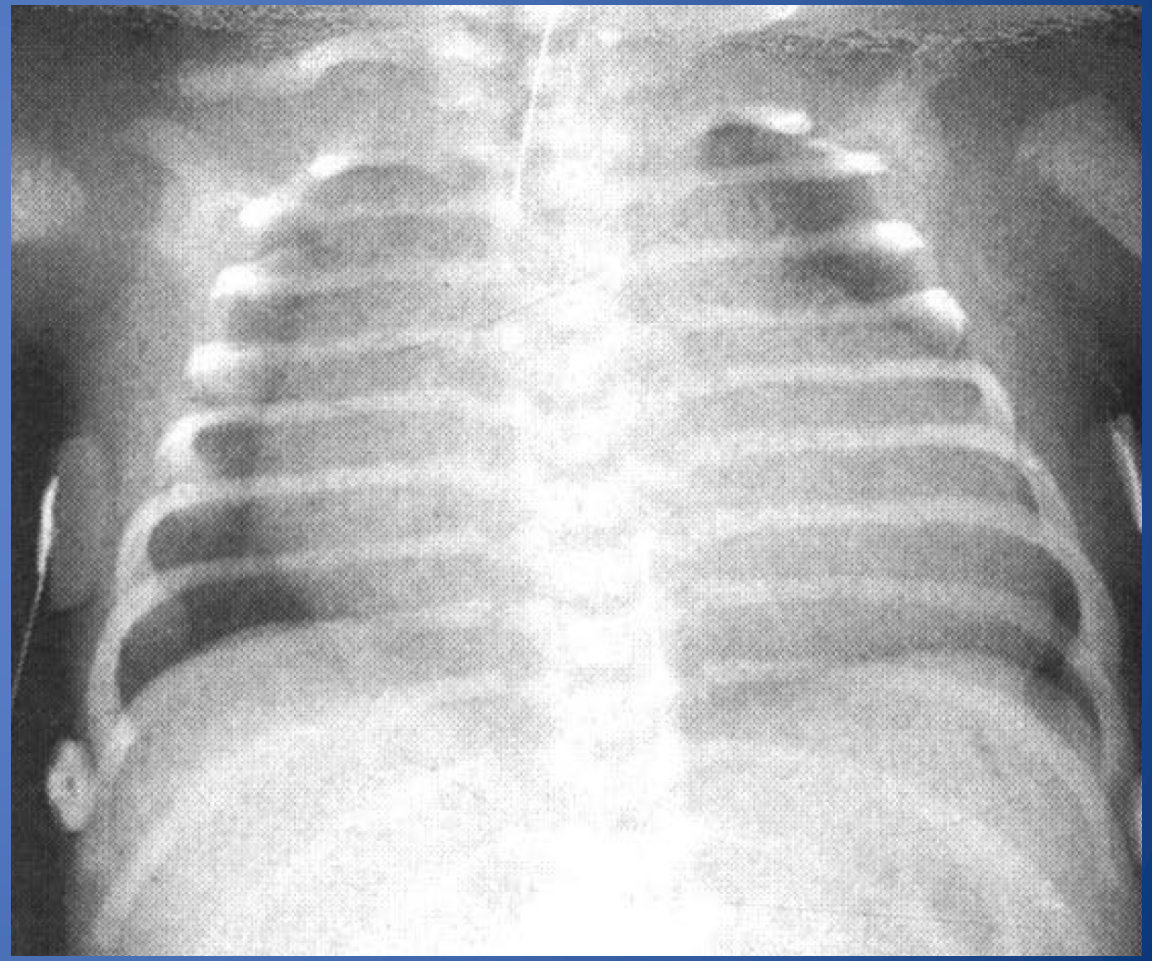


Kumar, Boston, Knott-Craig
SeminThoracCardiovascSurg 2017

Neonatal Ebstein's Anomaly

- Presentation
  - High rate of fetal demise
  - Critical illness apparent soon after birth
  - Enormous heart
  - Cyanotic requiring intubation/ventilation

- Early Management
  - PGE_1 begun
  - Doppler/echo - ?RVOTO?
  - Assess TV anatomy/regurgitation
    RV size/function LV size/function
    GOSE Score



Neonatal Ebstein's Anomaly

Echo evaluation of RVOT

- If RVOTO is anatomic
  - BT shunt or RV-PA reconstruction will be required
- If RVOTO functional
  - Try weaning PGE_1 as PVR falls (consider nitric oxide to help)
    - Best outcome – successful wean without CHF
    - Unsuccessful wean due to cyanosis alone – BT shunt required
    - Unsuccessful wean due to cyanosis and CHF – Tricuspid Valve will need to be addressed

Neonatal Ebstein's Anomaly

Addressing the tricuspid valve

- Close tricuspid valve, open atrial septum, BT shunt – Starnes procedure
  - Converts to single ventricle physiology, subsequent single ventricle pathway to Fontan
- Repair tricuspid valve
  - Not advisable in gravely ill neonate with poor RV function
  - Biventricular repair requires
    - Creation of competent TV based on anterior leaflet
    - Reduction of atrial volume
    - ASD closure leaving small fenestration
    - Repair all associated defects, including pulmonary atresia

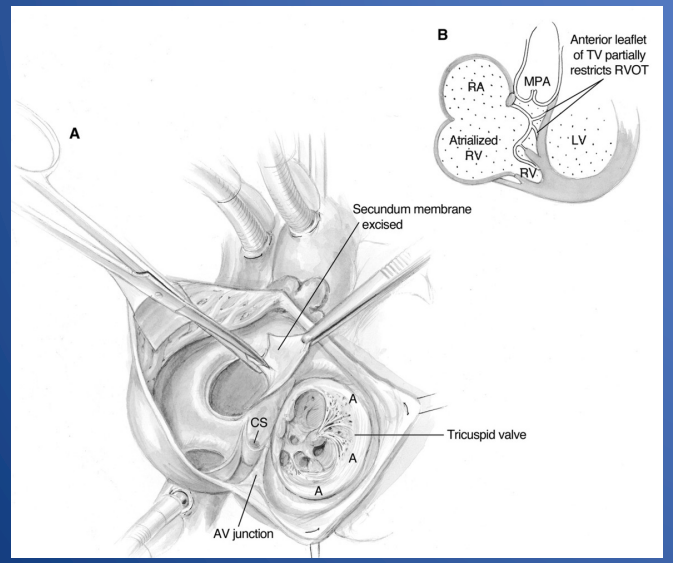
Tricuspid Valve Closure

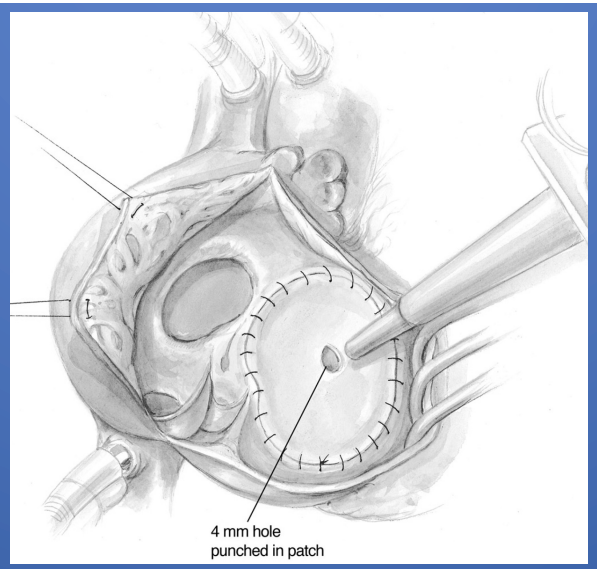
J THORAC CARDIOVASC SURG 1991;101:1082-7

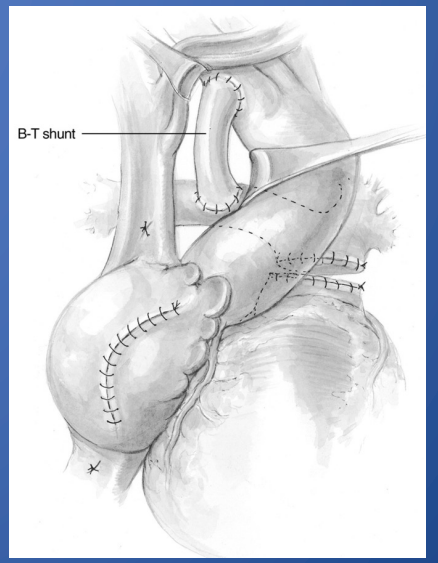
Ebstein's anomaly appearing in the neonate

A new surgical approach

Vaughn A. Starnes, MD,[a] Paul T. Pitlick, MD,[b] Daniel Bernstein, MD,[b]
Michael L. Griffin, MD,[b] Michael Choy, MD,[b] and Norman E. Shumway, MD, PhD,[a]
Stanford and Sacramento, Calif.





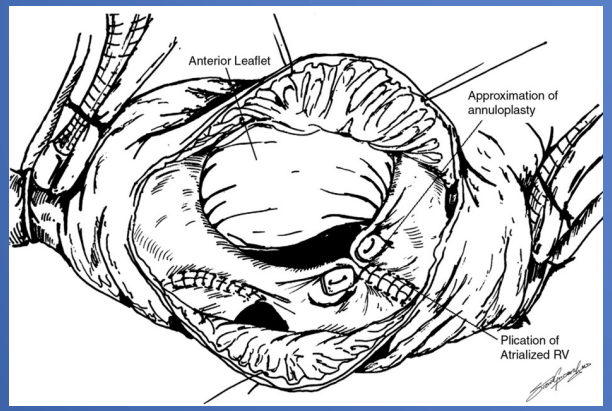


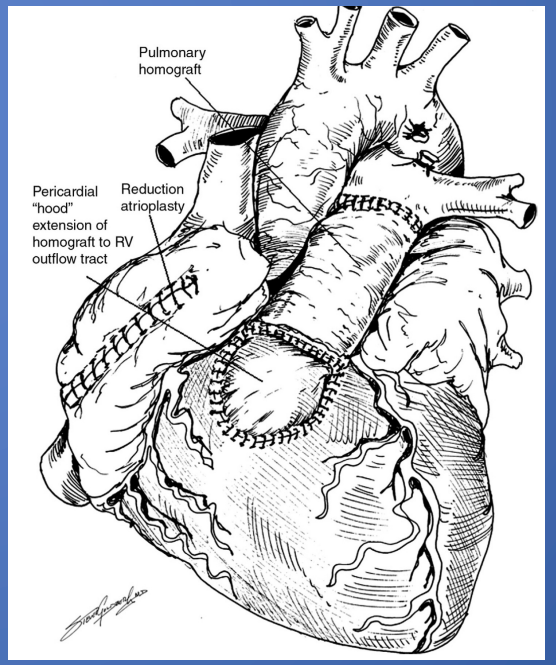
Tricuspid Valve Repair

(Two Ventricle Repair)

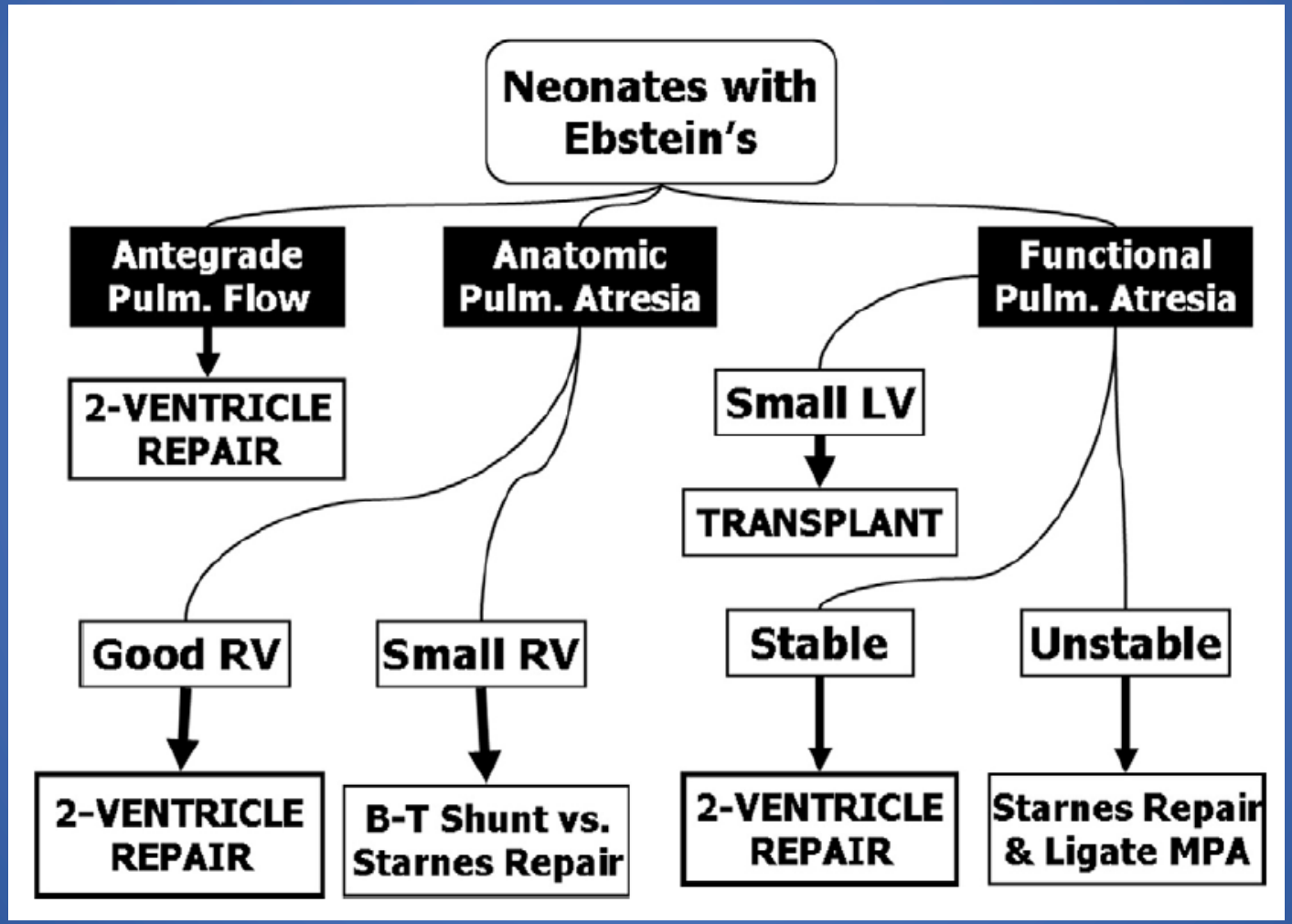
Management of Neonatal Ebstein's Anomaly

Christopher J. Knott-Craig, MD, FACS





Neonatal Ebstein's – Algorithmic Approach



Knott-Craig et al. AnnThorSurg 2007

Surgical Outcomes for Symptomatic Neonates with Ebstein's

Surgical Management and Outcomes of Ebstein Anomaly in Neonates and Infants: A Society of Thoracic Surgeons Congenital Heart Surgery Database Analysis

Kimberly A. Holst, MD, Joseph A. Dearani, MD, Sameh M. Said, MD, Ryan R. Davies, MD, Christian Pizarro, MD, Christopher Knott-Craig, MD, T. K. Susheel Kumar, MD, Vaughn A. Starnes, MD, S. Ram Kumar, MD, PhD, Sara K. Pasquali, MD, MHS, Dylan P. Thibault, MS, James M. Meza, MD, MSc, Kevin D. Hill, MD, MS, Karen Chiswell, PhD, Jeffrey P. Jacobs, MD, and Marshall L. Jacobs, MD

Department of Cardiovascular Surgery, Mayo Clinic, Rochester, Minnesota; Department of Cardiovascular and Thoracic Surgery, UT Southwestern Medical Center and Children's Health, Dallas, Texas; Department of Pediatric Cardiothoracic Surgery, Nemours Children's Health System, Wilmington, Delaware; Department of Pediatric Cardiothoracic Surgery, Le Bonheur Children's Hospital and University of Tennessee Health Science Center, Memphis, Tennessee; Department of Surgery, Keck School of Medicine, University of Southern California, Los Angeles, California; Heart Institute, Children's Hospital Los Angeles, Los Angeles, California; Department of Pediatrics, University of Michigan, Ann Arbor, Michigan; Duke Clinical Research Institute, Duke University School of Medicine, Durham, North Carolina; Division of Cardiovascular and Thoracic Surgery, Department of Surgery, Duke University Medical Center, Durham, North Carolina; Division of Cardiovascular Surgery, Johns Hopkins All Children's Hospital, St. Petersburg, Florida; and Division of Cardiac Surgery, Department of Surgery, Johns Hopkins University School of Medicine, Baltimore, Maryland

AnnThorSurg 106:785-91, 2018

STS Database Neonatal Surgery for Ebstein's

STS Database Neonatal Surgery for Ebstein's

2010 – 2016

255 neonates/95 centers

Median age at operation – 7 days

- Tricuspid valve repair (n = 101) - 40% Mortality 34% Major Morbid/mortality 61%
- Aortopulmonary shunt (n = 52) - 20% Mortality 17% Major Morbid/mortality 42%
- Starnes procedure (n = 24) - 9% Mortality 25% Major Morbid/mortality 54%

Neonate with Ebstein's

- Symptomatic Ebstein's is high risk in the neonate
- Complex treatment algorithm leads to variety of operative procedures
- Mortality with all surgical approaches high - 17% -34%
- Ebstein's in a neonate should be recognized as much greater threat to the patient than Ebstein's in the older child or young adult

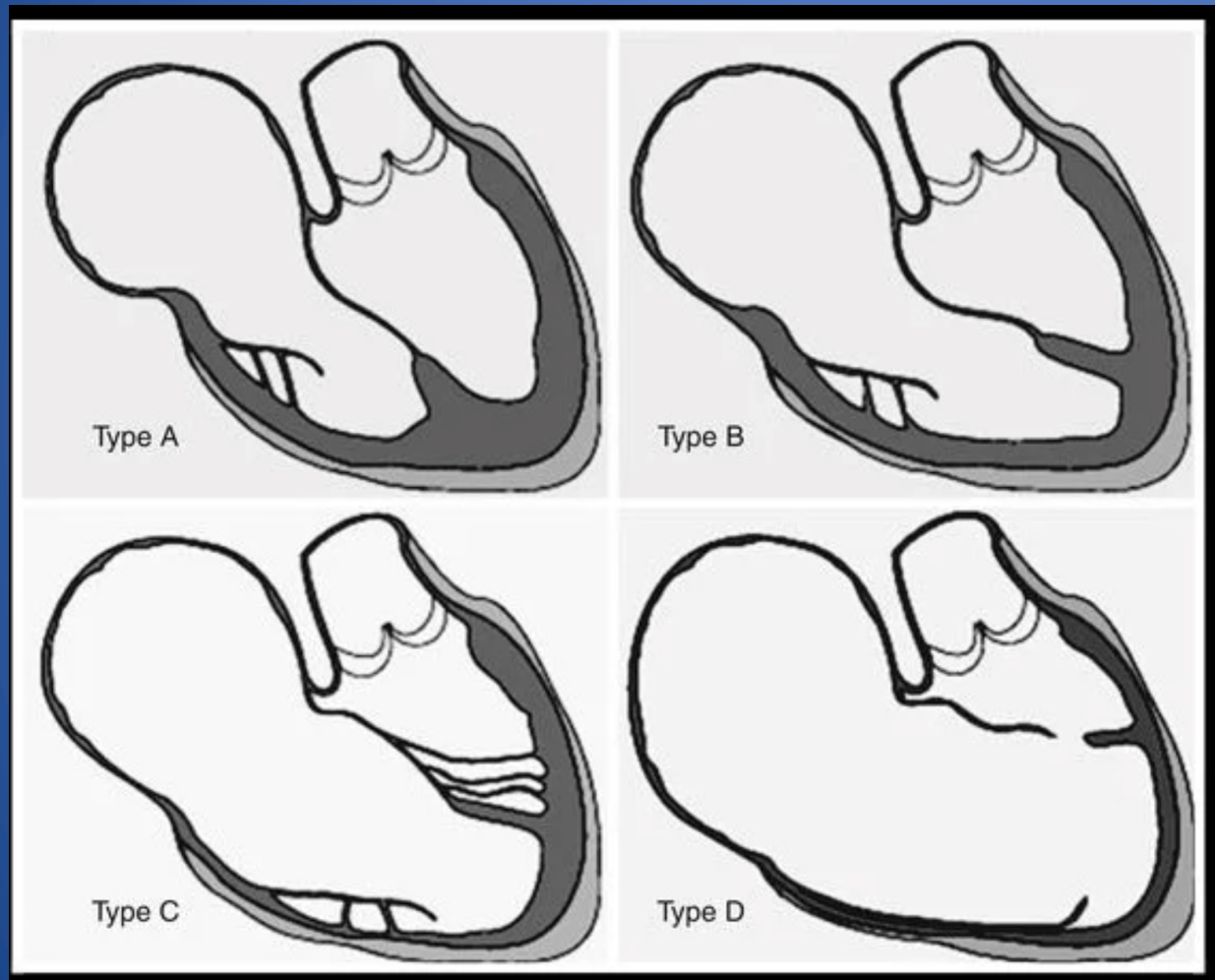
Child/Adult with Ebstein's

- Survived the newborn period with no to minimal symptoms
- Symptoms
  - Dyspnea, Fatigue, Palpitations, Cyanosis
- Diagnosis
  - Echocardiography, ECG, Holter monitor, CXR, exercise stress test
  - Cardiac MRI
  - EP study
  - Cardiac catheterization
- Consider surgery when
  - Quality of life affected
  - Heart enlarging
  - Heart function decreasing

Surgery for Child/Adult with Ebstein's

- Address tricuspid valve regurgitation
  - Repair
  - Replace
- Close atrial septal defect
- Selective cavopulmonary anastomosis
- Maze procedure if indicated for EP situation

Carpentier's Classification of Ebstein's Anomaly



Type A: the volume of the true RV is adequate

Type B: a large atrialized component of the RV exists, but the anterior leaflet of the TV moves freely

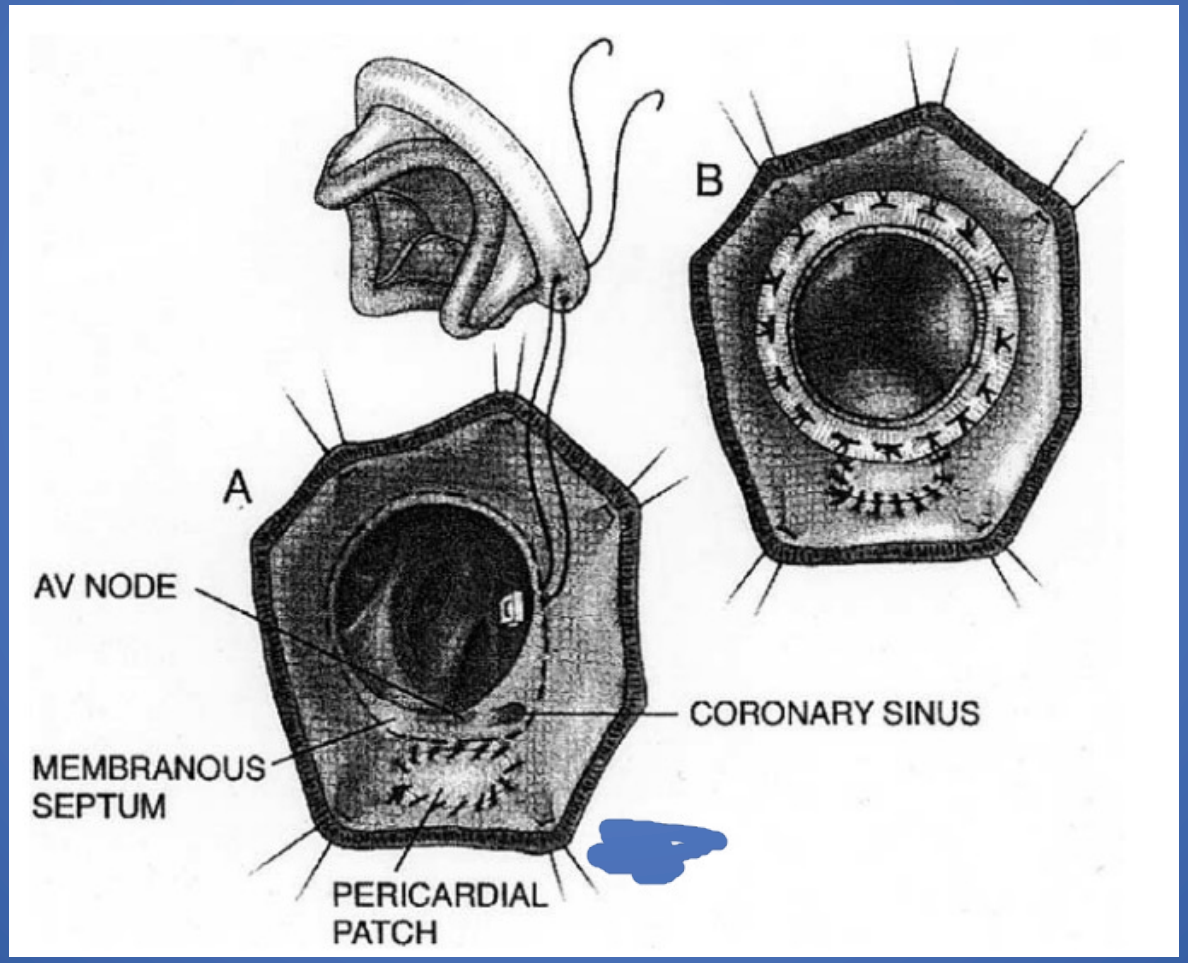
Type C: severe restriction of the anterior leaflet movement that can cause significant RV outflow tract obstruction

Type D: near complete atrialization of the ventricle

Surgery for Child/Adult with Ebstein's

- History of surgical approach to Ebstein's
  - –Replace with prosthesis (bioprosthetic preferred)
  - –Repair by creating monocusp
    - Lillehei/Hardy (1960s)
    - Carpentier (JTCVS 1988)
    - Danielson (JTCVS 1992)
  - –Repair with "Cone" technique
    - da Silva (ArqBrasCardiol 2004)

Tricuspid valve replacement for Ebstein's



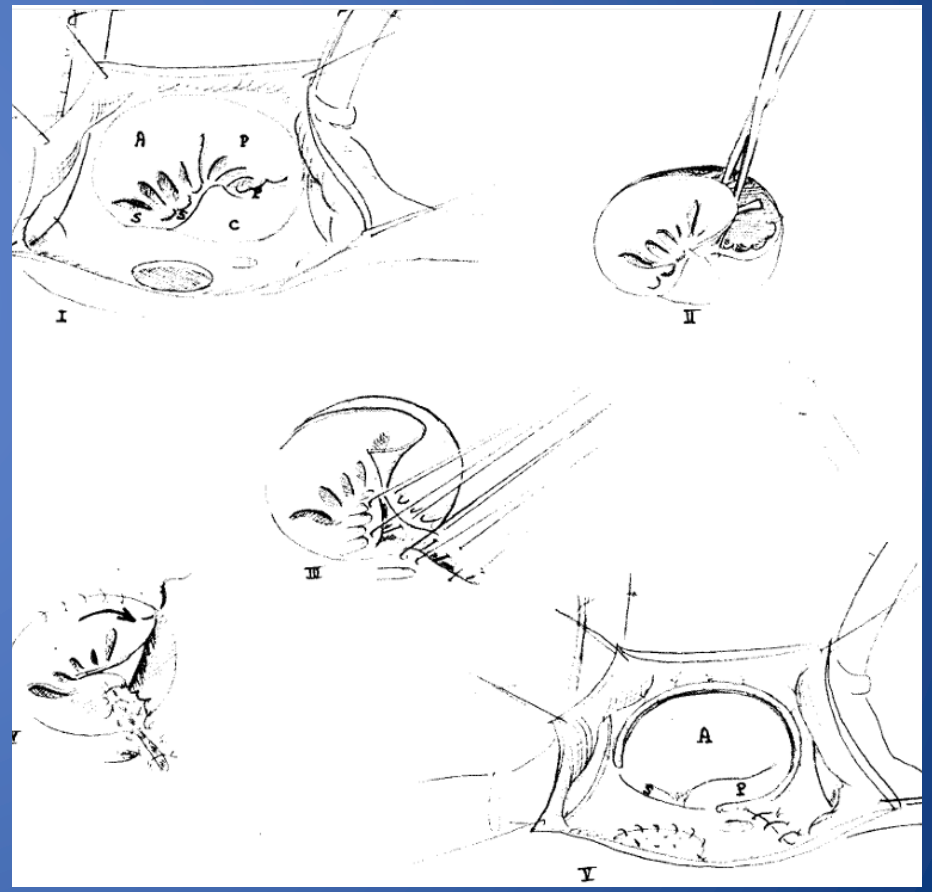
Ebstein's Repair – Carpentier technique

J Thorac Cardiovasc Surg 1988;96:92-101

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

Ebstein's anomaly is a complex malformation involving the tricuspid valve and the right ventricle. Various surgical techniques, either repair or replacement of the abnormal tricuspid valve, have been used with variable results. In an attempt to enlarge the indications of conservative procedures, we developed a technique of repair that comprises the reconstruction of a normally shaped right ventricle and the repositioning of the tricuspid valve at the normal level. In a series of 14 patients operated on between January 1980 and December 1986 in our institution, only one patient with an associated atrioventricular septal defect required a tricuspid valve replacement. All of the other patients, regardless of the complexity of the tricuspid malformation, were able to benefit from this conservative technique. There were two hospital deaths and no late deaths in this series. All surviving patients displayed a marked improvement over their preoperative status with regard to functional class (92% are in class I or II) and rhythm disturbances. Echocardiographic and Doppler studies demonstrated a normal shape of the right ventricle and good tricuspid valve function in all the patients but one.

Alain Carpentier, MD, PhD, Sylvain Chauvaud, MD (by invitation), Loïc Macé, MD (by invitation), John Relland, MD (by invitation), Serban Mihaileanu, MD (by invitation), J. P. Marino, MD (by invitation), Bernard Abry, MD (by invitation), and Pierre Guibourt, MD (by invitation), *Paris, France*

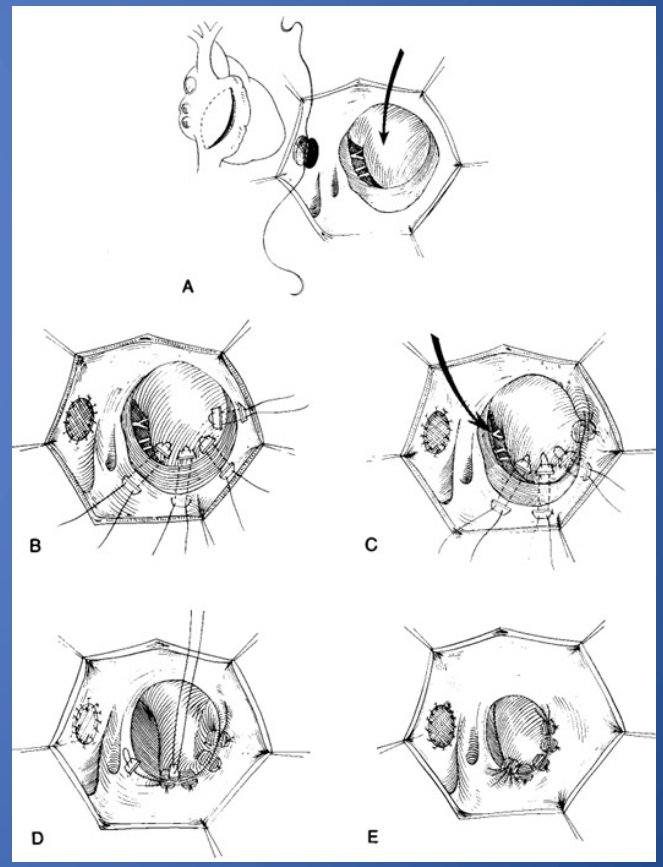


Ebstein's Repair – Danielson technique

Operative treatment of Ebstein's anomaly

From April 1972 to February 12, 1991, 189 patients with Ebstein's anomaly underwent repair. Ages ranged from 11 months to 64 years (median 16 years, mean 19.1 years). In 58.2%, tricuspid valve reconstruction was possible, and in 36.5%, a prosthetic valve, usually a bioprosthesis, was inserted. In 5.3%, a modified Fontan or other procedure was performed. There were 12 hospital deaths (6.3%). All 28 patients who had accessory conduction pathways (Wolff-Parkinson-White syndrome) underwent successful ablation of the pathways as part of the operative treatment. Follow-up was obtained in 151 (85.3%) patients. Of those patients followed up more than 1 year after operation, 92.9% were in New York Heart Association class I or II. There were 10 late deaths: seven cardiac (four sudden), two noncardiac, and one of an unknown cause. Postoperative Doppler echocardiographic assessment showed the atrial septum was intact in all patients and tricuspid valve function was good to excellent in most patients. Four of the 110 patients (3.6%) who underwent valve reconstruction required reoperation 1.4 to 14.1 years later. Postoperative reduction in heart size was usual, atrial arrhythmias were reduced, and late postoperative exercise testing showed a significant improvement in performance: Maximal oxygen consumption increased from a mean of 47% of predicted value before the operation to a mean of 72% after the operation. Nine patients had a total of 12 successful pregnancies with deliveries of normal children. (J THORAC CARDIOVASC SURG 1992;104:1195-1202)

Gordon K. Danielson, MD, David J. Driscoll, MD (by invitation),
Douglas D. Mair, MD (by invitation), Carole A. Warnes, MD (by invitation), and
William C. Oliver, Jr., MD (by invitation), *Rochester, Minn.*



Ebstein's Repair – The Cone Procedure
da Silva technique

Artigo Original

Anomalia de Ebstein. Resultados com a reconstrução cônica da valva tricúspide

José Pedro da Silva, José Francisco Baumgratz, Luciana da Fonseca, Jorge Yussef Afiune, Sônia Meiken Franchi, Lílian Maria Lopes, Daniel Marcelo Silva Magalhães, José Henrique Andrade Vila

São Paulo, SP

Arq Bras Cardiol, volume 82 (nº 3), 212-6, 2004

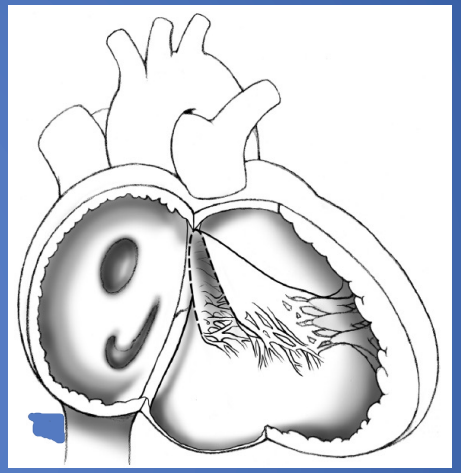
Ebstein's Repair – The Cone Procedure da Silva technique

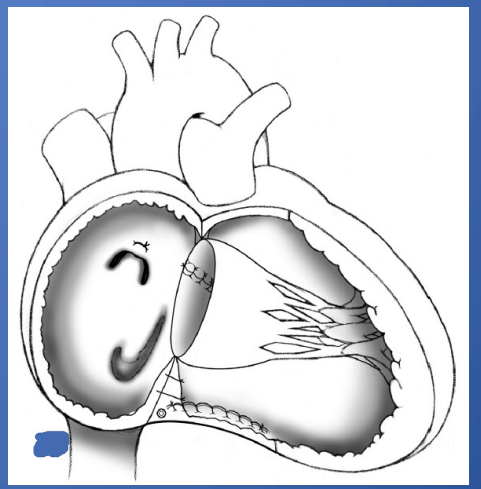
Objective - *To assess the results of a technical modification of tricuspid valvuloplasty in Ebstein's anomaly.*

Methods - *From November 1993 to August 2002, 21 patients with Ebstein's anomaly of the tricuspid valve, with ages ranging from 20 months to 37 years (mean, 23 years), underwent a new technique of tricuspid valvuloplasty. This technique consisted of total or almost total detachment of the anterior tricuspid megaleaflet from the ventricular wall and valvular ring, transforming it into a cone, whose vertex remained fixed in the right ventricular tip, and the base was sutured to the tricuspid ring, after its plication, adjusting it to the size of the base of the cone (tricuspid ring), including the septal region.*

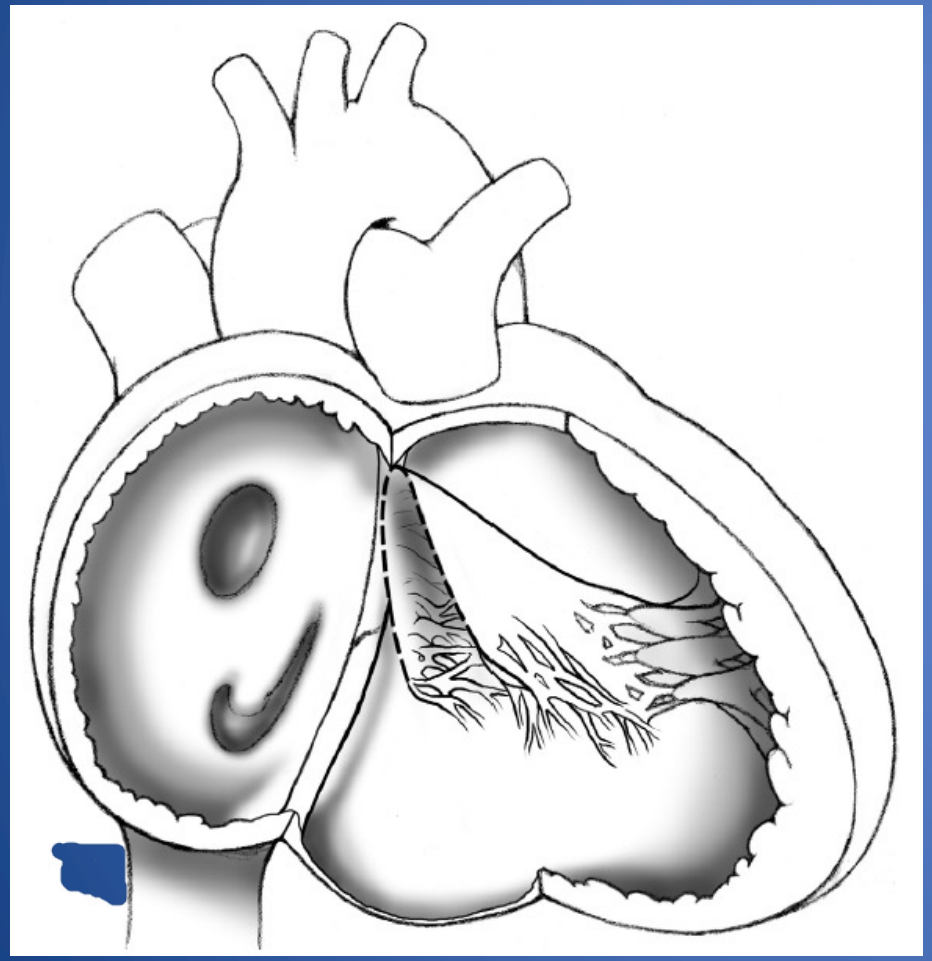
Results - *One (4.7%) patient with cardiomyopathy caused by chronic hypoxia died in the hospital due to low cardiac output. The mean follow-up lasted 4 years, and the recent echocardiograms showed good morphology of the right ventricle and tricuspid valve with mild or minimal insufficiency in 18 patients and moderate insufficiency in 2 patients. In 2 of the 3 patients with an anomalous bundle, it could be located and sectioned during surgery. No atrioventricular block occurred.*

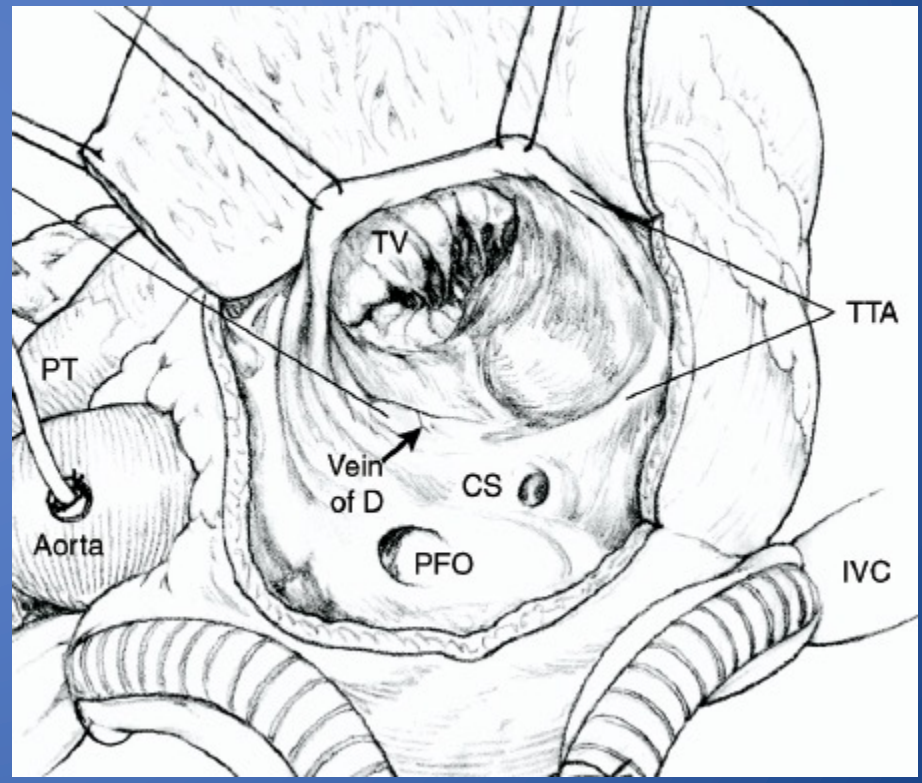
Conclusion - *The technique used was efficient in repairing tricuspid insufficiency and restoring right ventricular morphology, being applicable to all anatomic types of Ebstein's anomaly, except for Carpentier classification type D.*



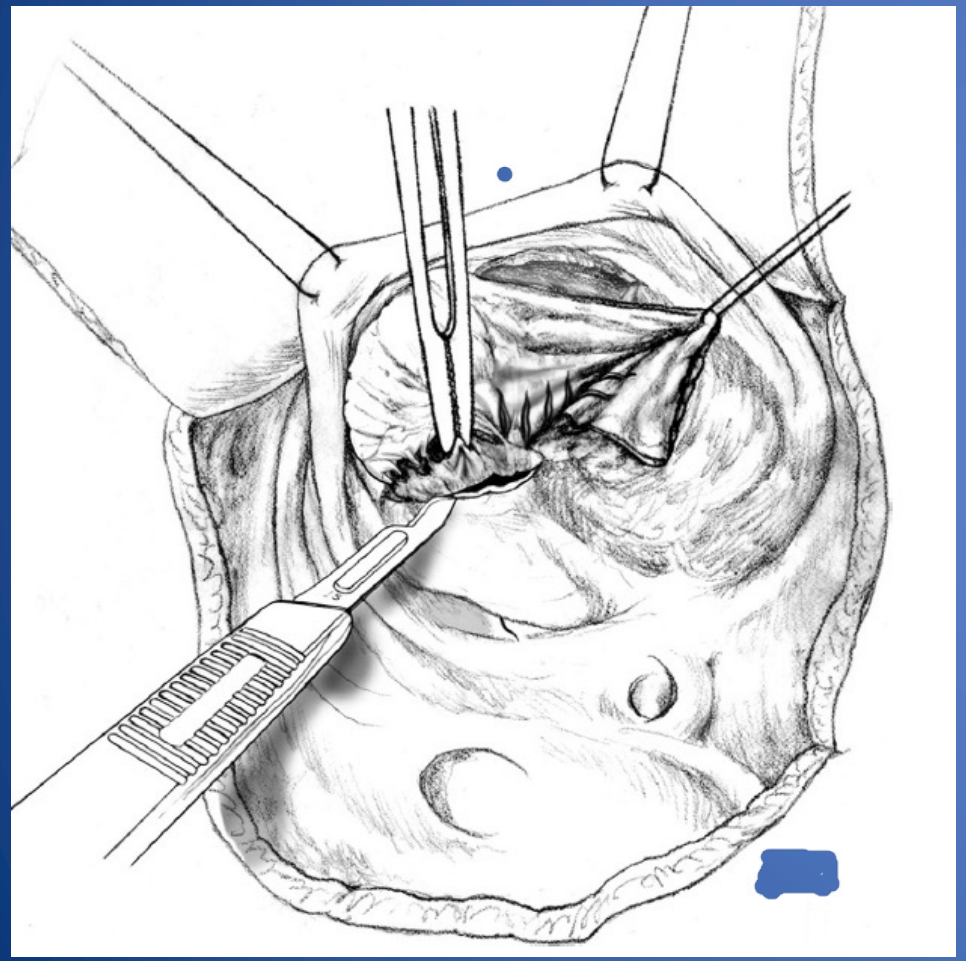


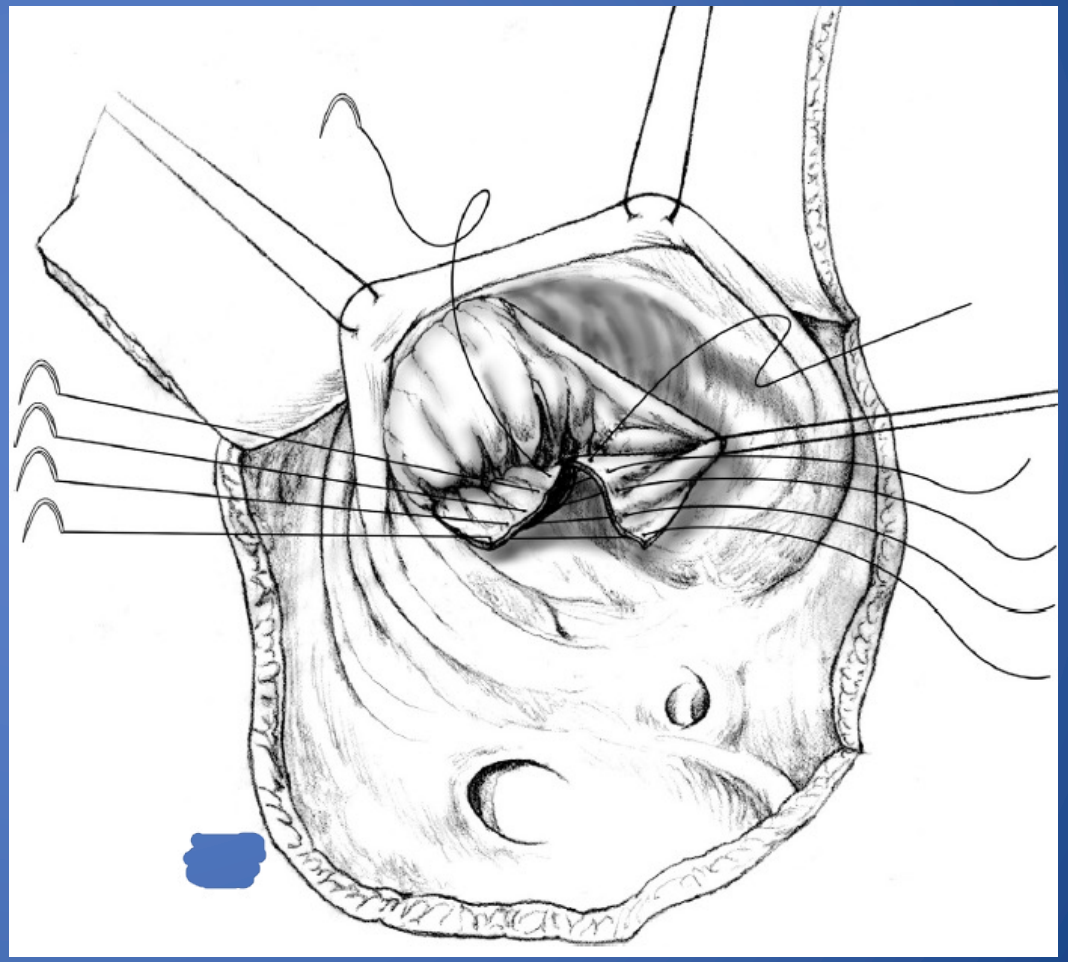
Ebstein's Repair – The Cone Procedure
da Silva technique



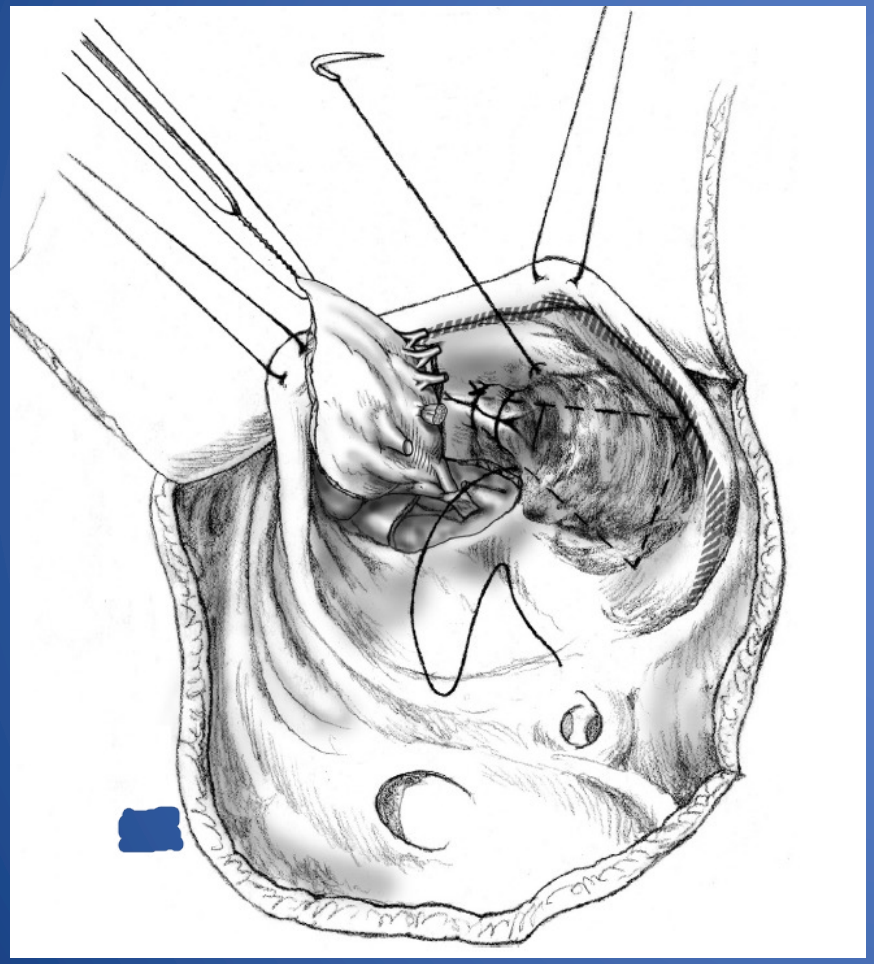


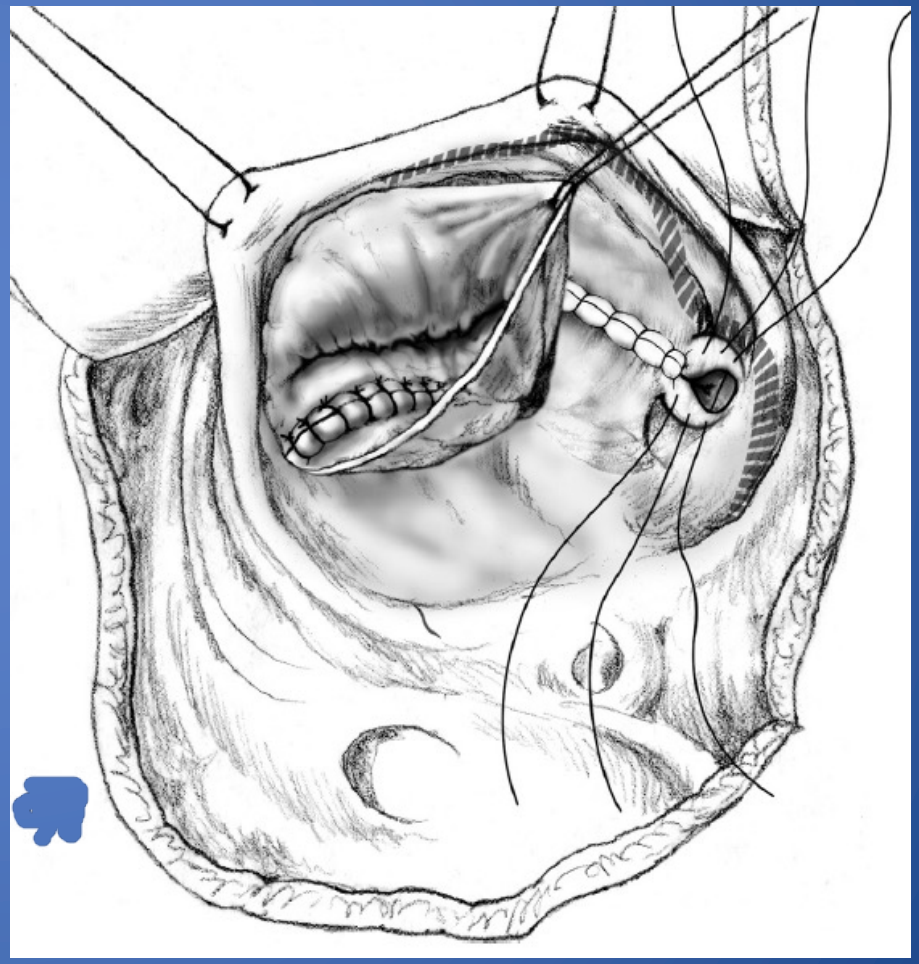
Ebstein's Repair – The Cone Procedure
da Silva technique



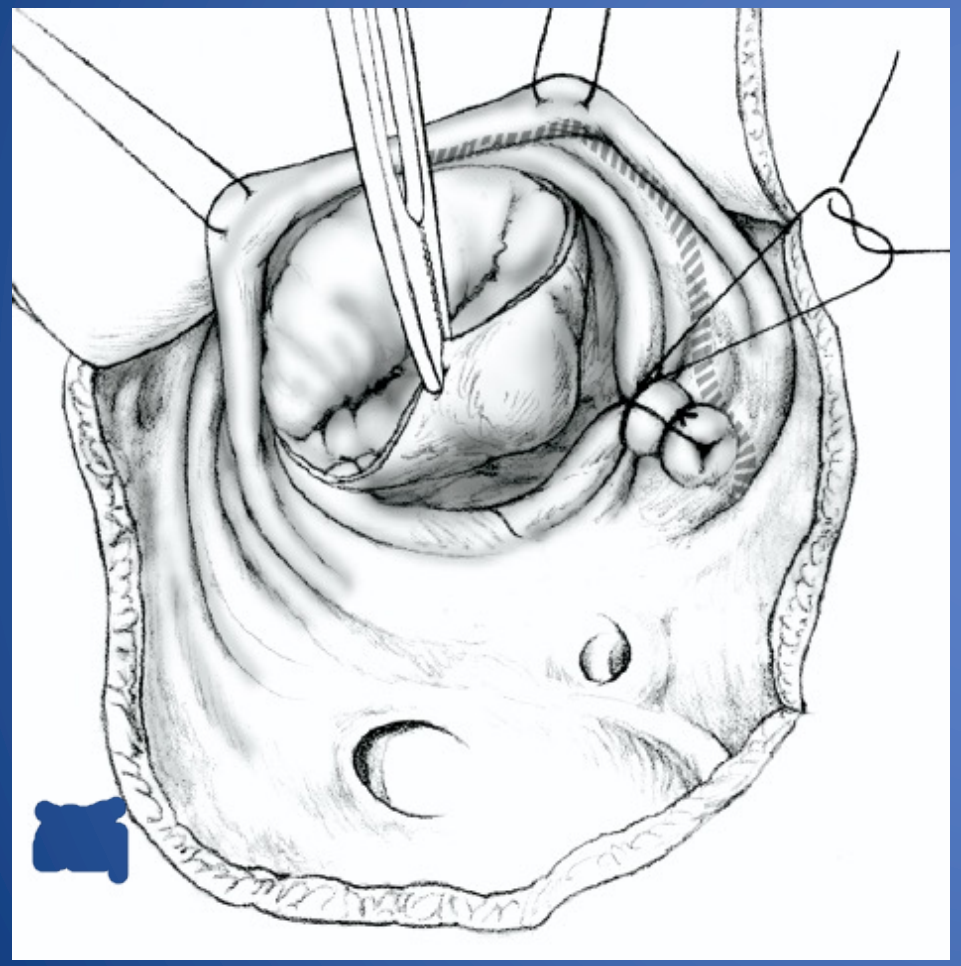


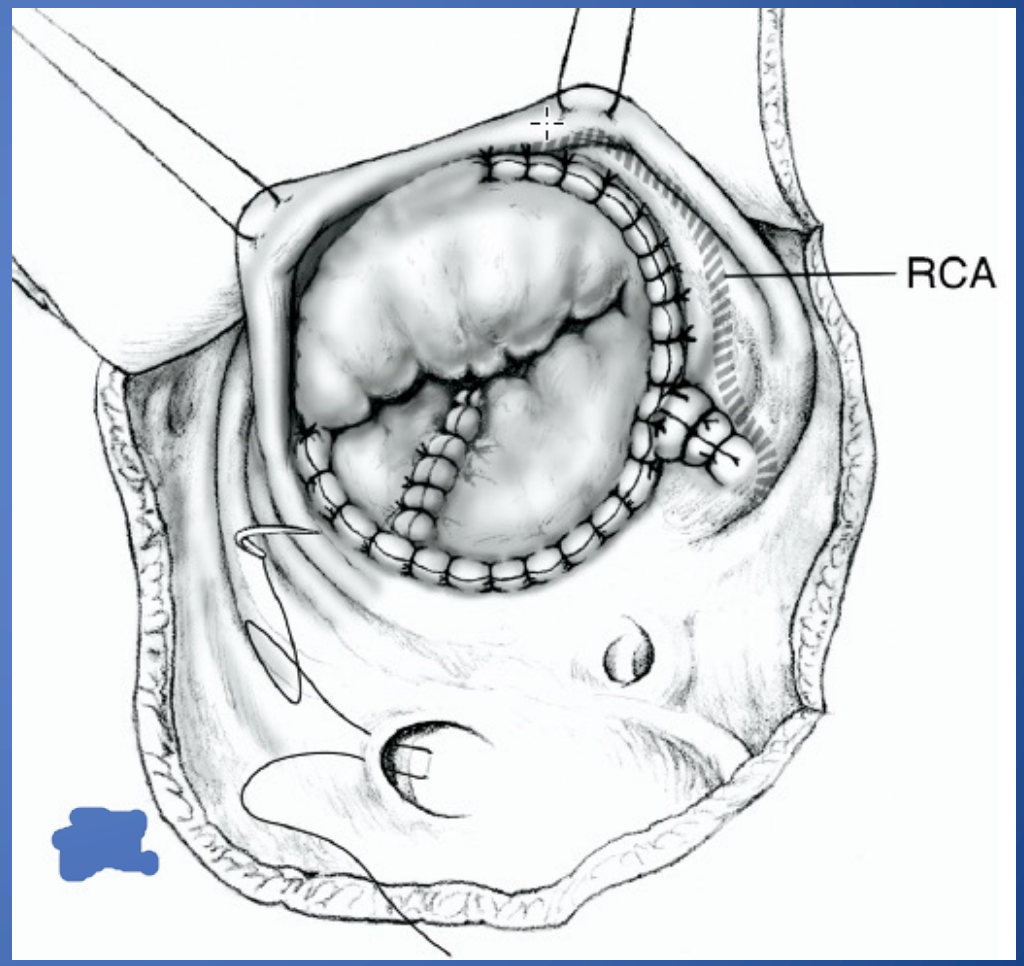
Ebstein's Repair – The Cone Procedure
da Silva technique



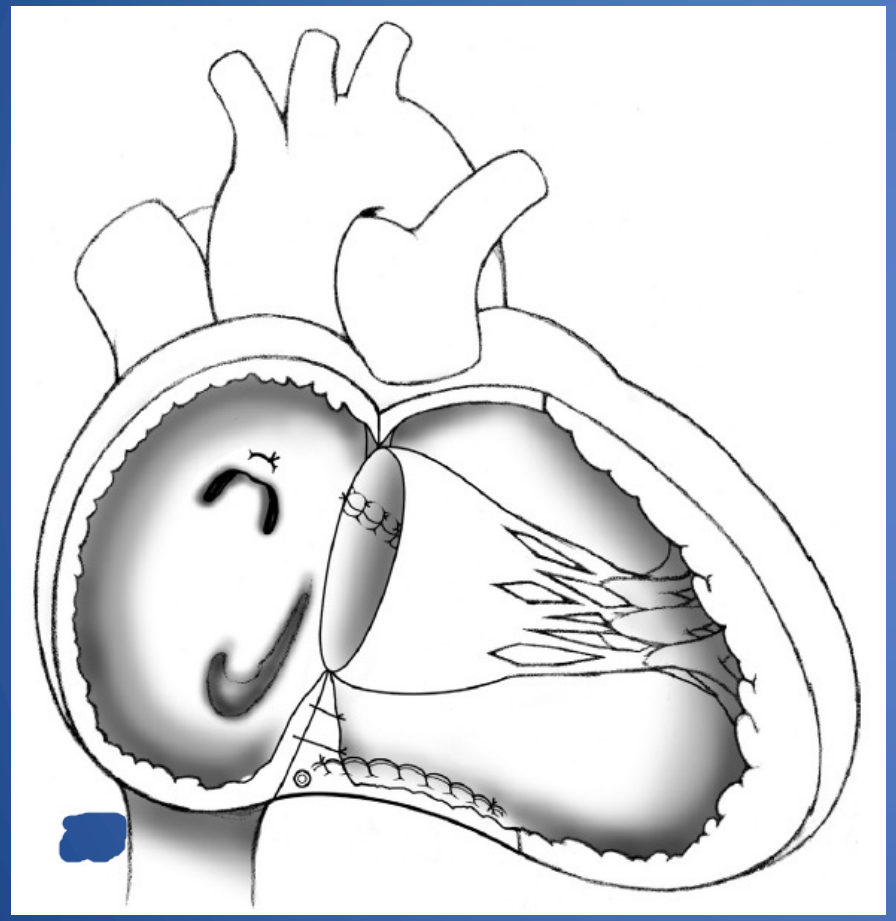


Ebstein's Repair – The Cone Procedure
da Silva technique

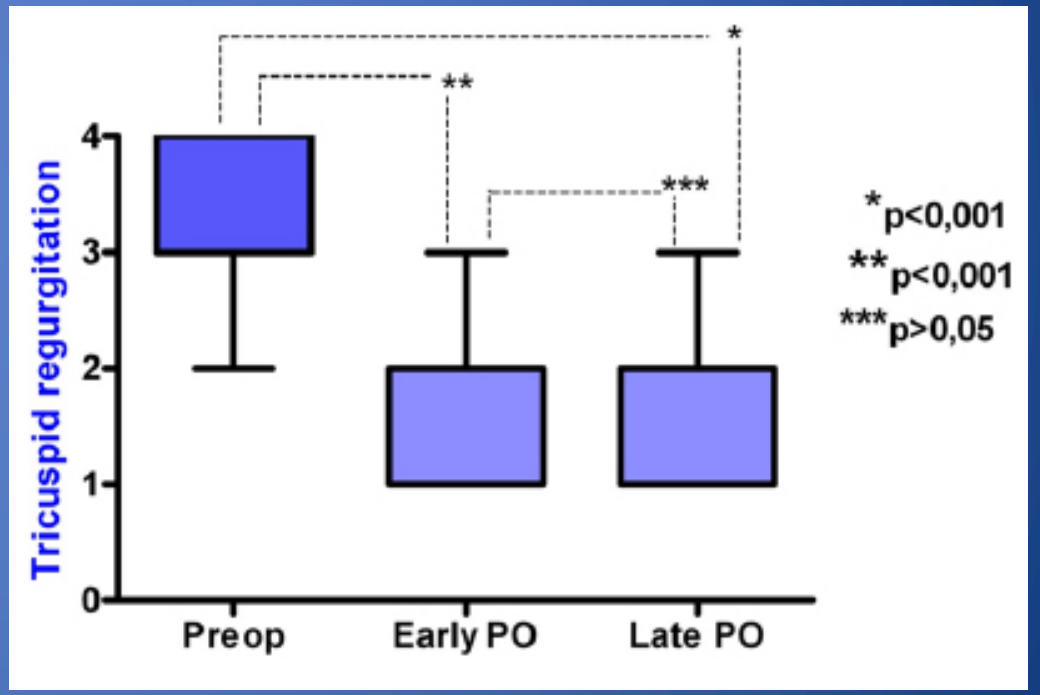




Ebstein's Repair – The Cone Procedure
da Silva technique



Significant and sustained reduction in tricuspid regurgitation



Da Silva, In *Operative Cardiac Surgery*, Spray & Acker 2018

Improving Results of Surgery for Ebstein Anomaly: Where Are We After 235 Cone Repairs?

Kimberly A. Holst, MD, Joseph A. Dearani, MD, Sameh Said, MD, Roxann B. Pike, MD, Heidi M. Connolly, MD, Bryan C. Cannon, MD, Kristen L. Sessions, BS, Megan M. O'Byrne, MA, and Patrick W. O'Leary, MD

Departments of Cardiovascular Surgery, Anesthesiology, Cardiovascular Diseases, and Health Sciences Research, Mayo Clinic, Rochester; Division of Pediatric Cardiology, Mayo Clinic, Rochester; and Mayo Clinical School of Medicine, Mayo Clinic, Rochester, Minnesota

Background. Ebstein anomaly has heterogeneous anatomy and numerous operative techniques are described. Cone repair provides a near anatomic tricuspid valve repair. The purpose of this study was to examine our experience with cone repair.

Methods. Cone repair was performed in 235 consecutive patients with Ebstein anomaly, 134 children (57%) and 101 adults (43%), from June 2007 to October 2015. Median age was 15.6 years (range, 6 months to 73 years). Cone repair was the first operation in 192 patients (82%), the second in 41 (17%), and the third in 2 (1%). Previous tricuspid valve repair had been performed in 27 (12%). Echocardiograms were obtained preoperatively and at hospital dismissal for all patients and for a subgroup of patients at least 6 months after cone repair (n = 81).

Results. Leaflet augmentation was done in 67 patients (28%), Sebening stitch in 57 (24.2%), neochordae in 49 (21%), and annuloplasty band in 158 (67%). Bidirectional cavopulmonary shunt was performed in 46 patients (20%). There was 1 early death (0.4%). Early reoperation was required in 14 patients (5.9%); re-repair was possible in 7 (50%). The majority of early reoperations (11 of 14; 79%) occurred in the first third of the series. Mean follow-up was 3.5 ± 2.5 years. There was sustained reduction in tricuspid regurgitation ($p < 0.0001$), a progressive decline in right ventricle size ($p < 0.0001$), and late increase in right ventricle fractional area change after initial decline ($p < 0.0001$). Freedom from late reoperation was 97.9% at 6 years.

Conclusions. Cone repair is safe, and the learning curve is significant. Sustained reduction in tricuspid regurgitation and favorable changes in the right ventricle at follow-up suggest that cone repair has an advantageous impact on right ventricular remodeling.

(Ann Thorac Surg 2018;105:160–9)

Ebstein's Anomaly

Surgical Repair and Outcomes

- A rare and challenging congenital heart defect
- When surgery is required in the neonate, a variety of surgical procedures must be considered and the risk of death or complications is high
- For patients that present as children or adults, surgical outcomes have become excellent when performed by surgeons experienced with this rare anomaly and the "Cone" technique.

Thank you