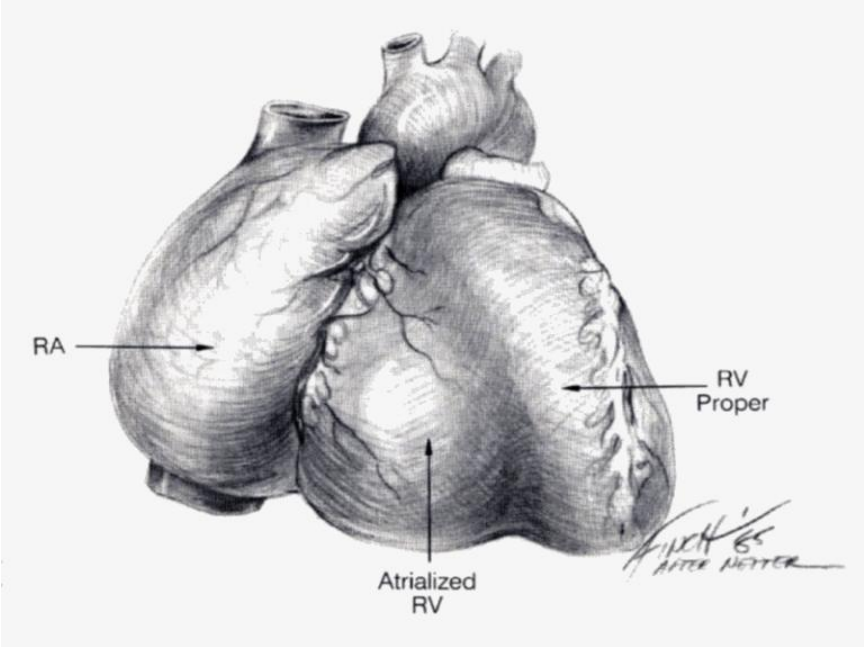
Ebstein's anomaly



History

- 1866 – Dr. Wilhelm Ebstein described cardiac findings of 19 y.o. patient who had died of cyanotic heart disease
- 1950 – Helen Taussig - first clinical syndrome analysis
- 1950's – BT shunt for neonatal Ebstein (functional tricuspid or pulmonary atresia)
- 1954 – Wright, Kirklin – direct closure of ASD for correction of right-to-left shunt (patient survived)

- 1958 – tricuspid valve reconstruction – Hunter & Lillehei – attempt to create competent valve by repositioning of displaced leaflet & excluding atrialized chamber (2 patients – both didn’t survive due to CAVB)

- 1964 - Hardy revived Hunter-Lillehei operation – effective only for mild anomaly; complications: CAVB; RCA injury; RV aneurysm

- Barnard (1963); Lillehei (1967) – tricuspid valve replacement

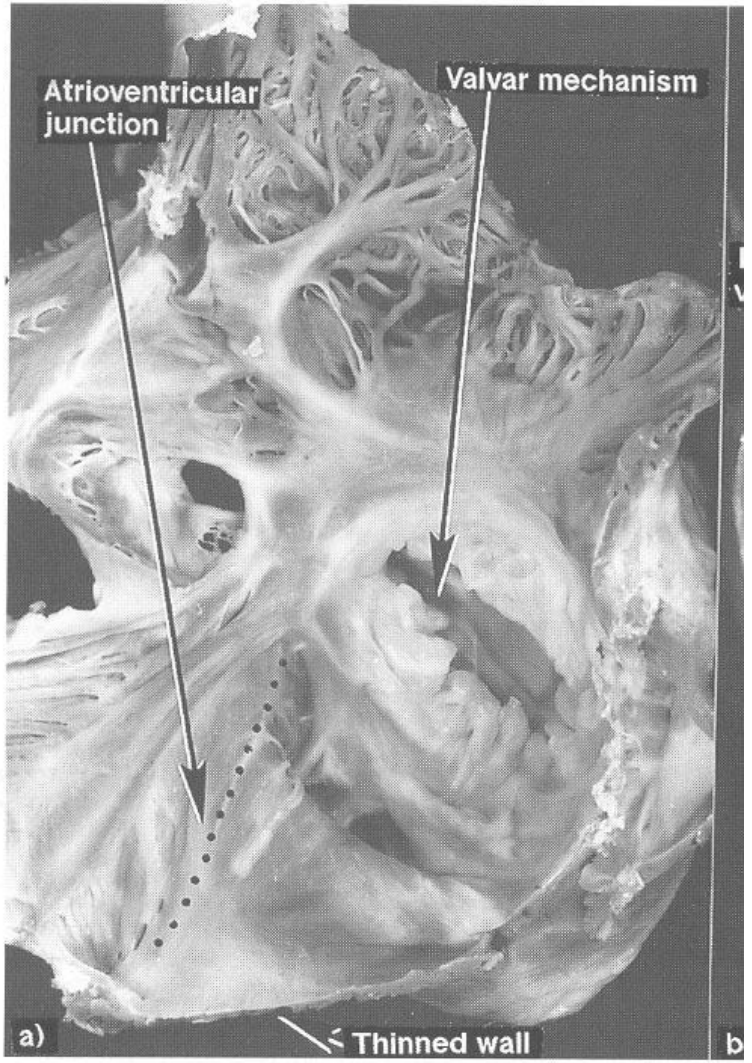
- Danielson (1972); Carpenter (1988) – TV repair based on use of anterior leaflet

- Starnes (1991) – single ventricle palliation of neonatal Ebstein

- Knott-Craig (1994) – biventricular repair of neonatal Ebstein anomaly

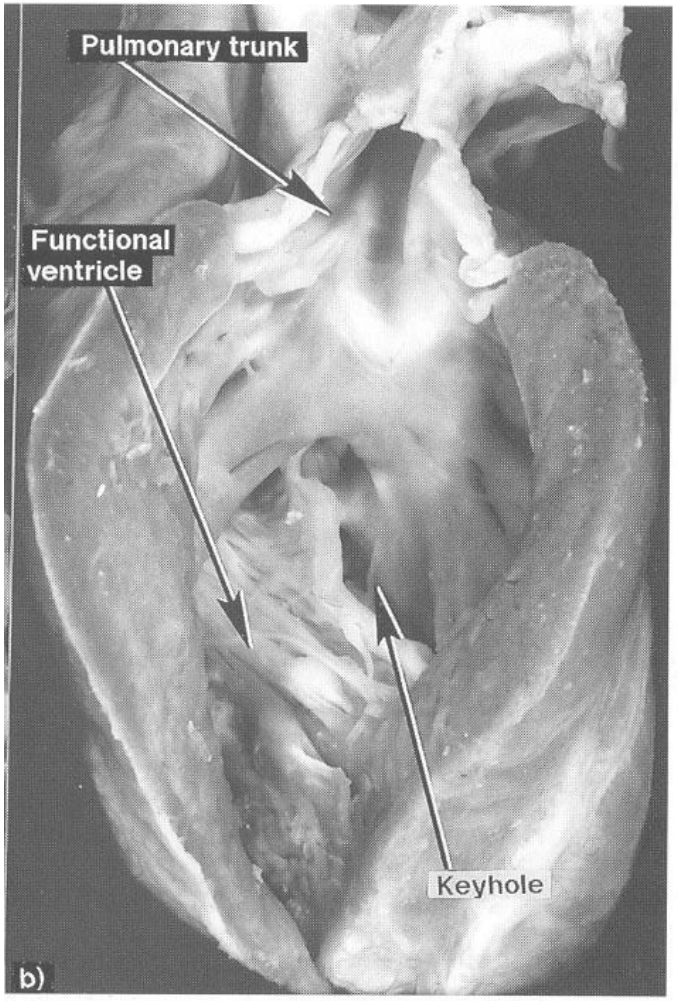
Anatomy

- Distal attachment of the septal & posterior leaflets away from the atrio-ventricular junction
- Plane of closure of the tricuspid valve at the junction of the inlet and apical component of the right ventricle
- Dilatation of the atrio-ventricular junction



Valve pathology is variable from patient to patient

- Variability of location of the valve annular attachment (from inlet to outlet)
- Variability of degree of formation and delamination of the septal and posterior leaflet
- In most cases TV has bifoliate structure with combined antero-superior and posterior leaflet



TV anatomy

TV leaflet malformation:

Septal > Mural (posterior) >Anterior

TV displacement:

maximal – postero-septal comissure

minimal – antero-septal comissure

Anterior leaflet

- Enlarged
- Sail-like
- Thicken and partially muscularized

Anterior leaflet

Leading edge: - free and mobile

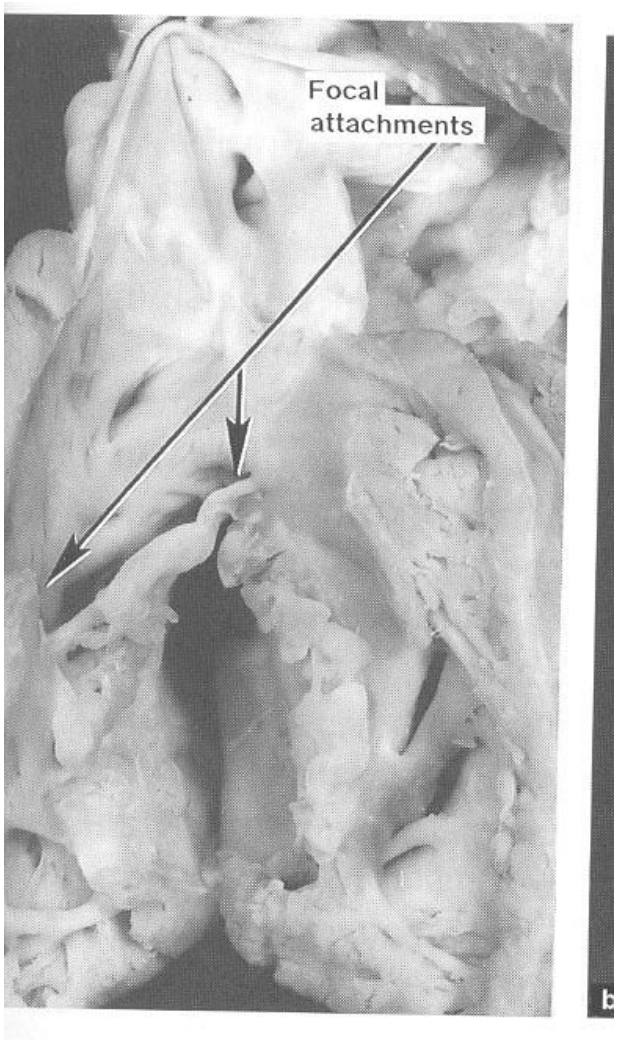
- segmental direct attachment to the myocardium
- linear direct attachment (entire leading edge attached to the myocardium)

Delamination: - partial

- complete

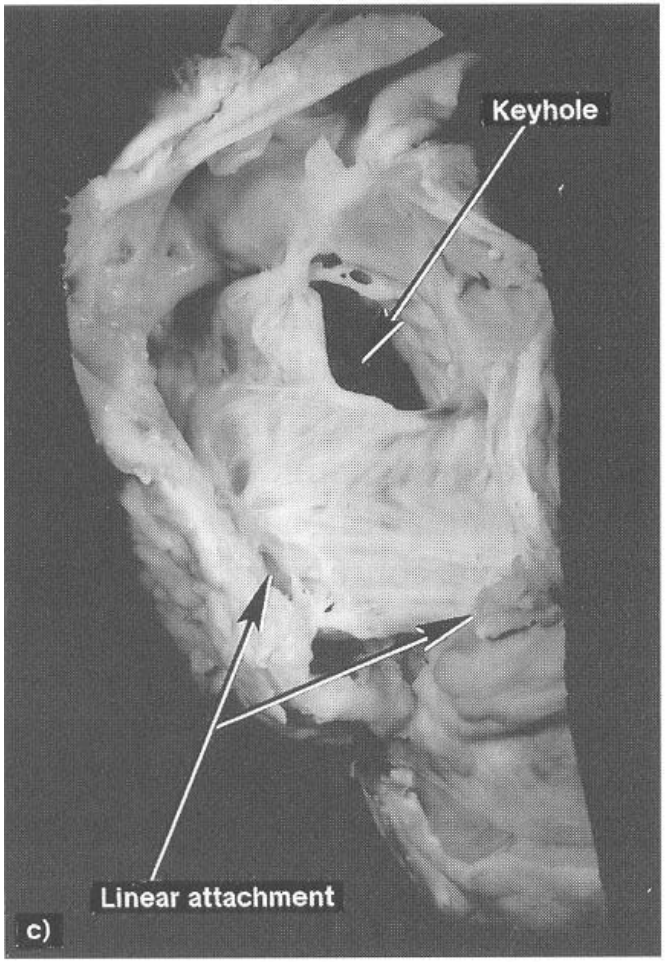
Antero-superior leaflet of the TV

- Attachment of the antero-superior leaflet – focal attachment to the medial and anterior papillary muscle



Antero-superior leaflet of the TV

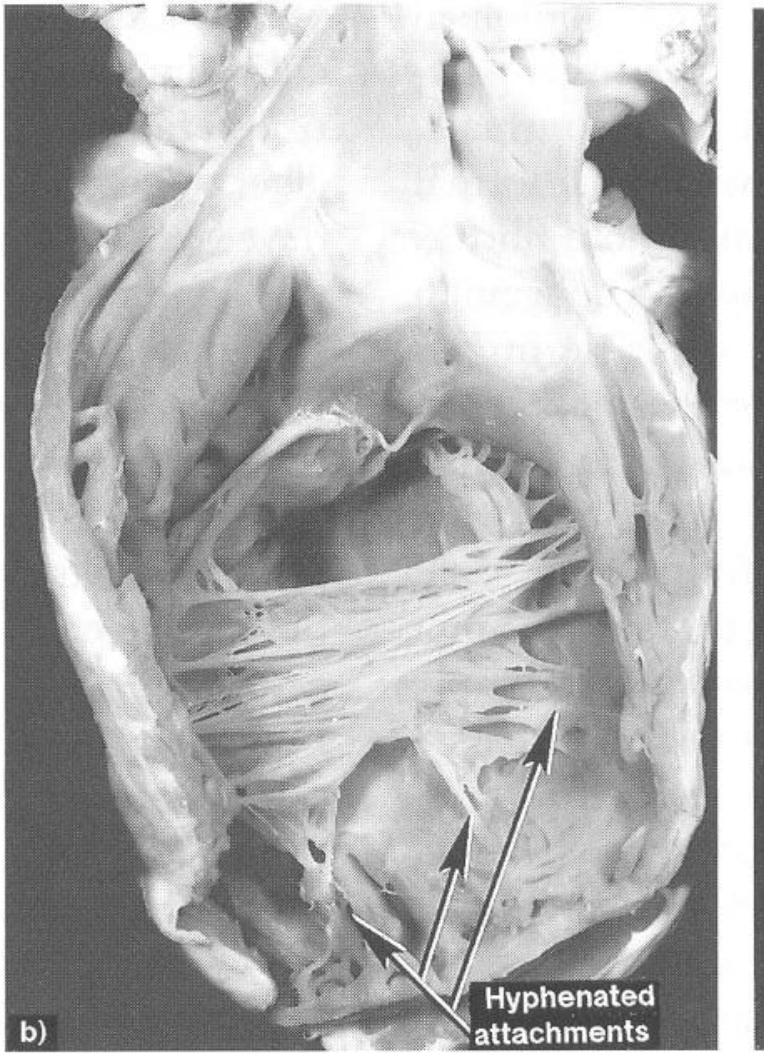
- Attachment of the antero-superior leaflet – entire leading edge of the leaflet is attached linearly to a muscle between inlet and apical component of RV



flet. (a) The heart here has focal attachment. (

Antero-superior leaflet of the TV

- Edge of the anterior leaflet is attached in hyphenated fashion



Right ventricle

- Atrialized RV – inlet part of RV above TV attachment; in symptomatic patient tend to be thin-walled and dilated

- Functional RV – apical and infundibular component: - thinner
  - contain fewer than normal muscular fibers
  - contain more fibrous tissue

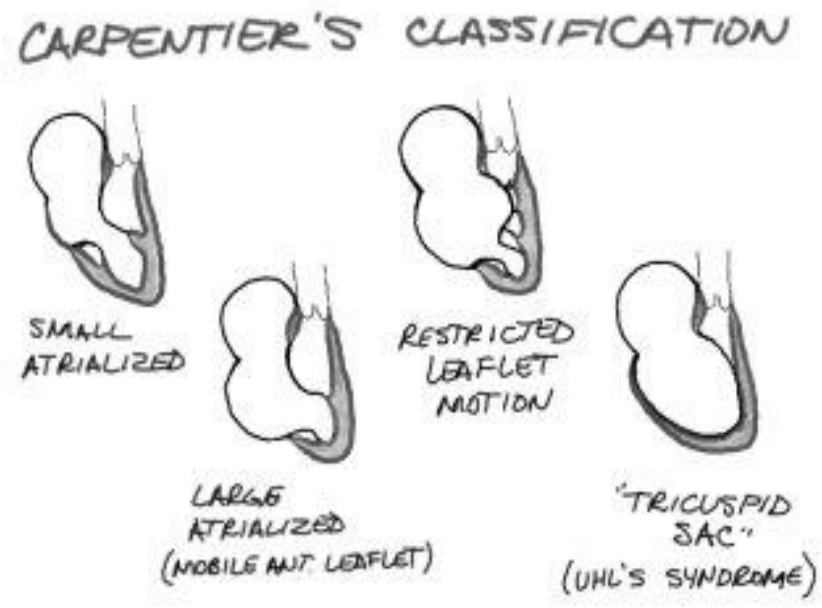
Carpentier classification (1988)

Grade A: mobile anterior leaflet/small contractile atrialized right ventricle

Grade B: mobile anterior leaflet/large, noncontractile atrialized RV

Grade C: tethering of anterior leaflet/large, noncontractile atrialized RV

Grade D: leaflets forming a continuous sac adherent to the right ventricle



Great Ormond Street score

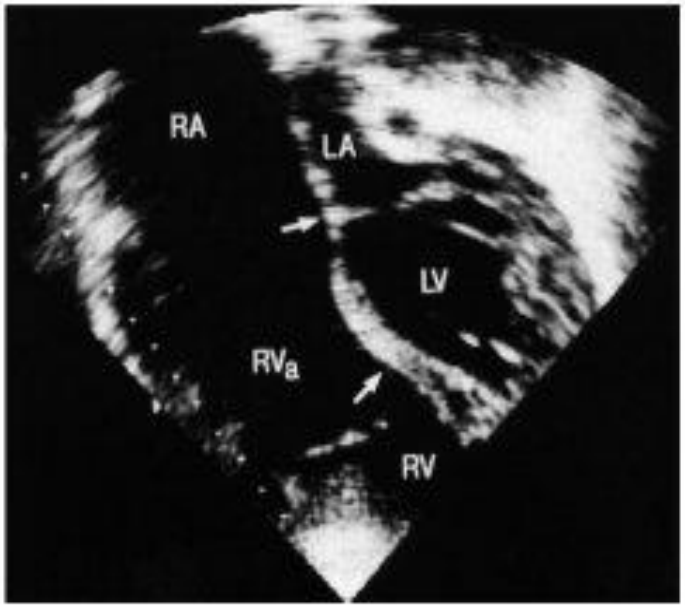
combined area of the right atrium and atrialized portion of the right ventricle divided by the area of functional RV added to the area of the left heart chambers (in diastole)

Grade 1 – ratio less than 0.5

Grade 2 – ratio 0.5 – 0.99

Grade 3 – ratio 1 – 1.49

Grade 4 – greater than 1.5



Physiology

Tricuspid valve:

- regurgitation
- stenosis
- stenosis+regurgitation

Right ventricle:

- functional dysfunction
- Anatomical dysfunction

effective pulmonary blood flow

shunt direction through ASD/PFO

Ebstein's anomaly

Neonatal

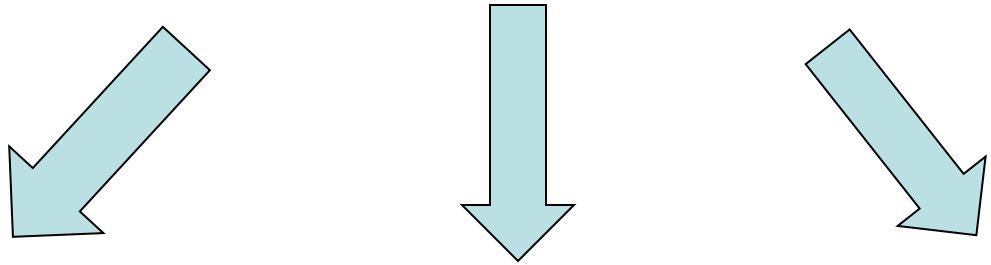
Adult

Age of presentation depends on severity of tricuspid and RV dysfunction

Newborn presentation

- TV dysfunction
- RV dysfunction

- Elevated PVR



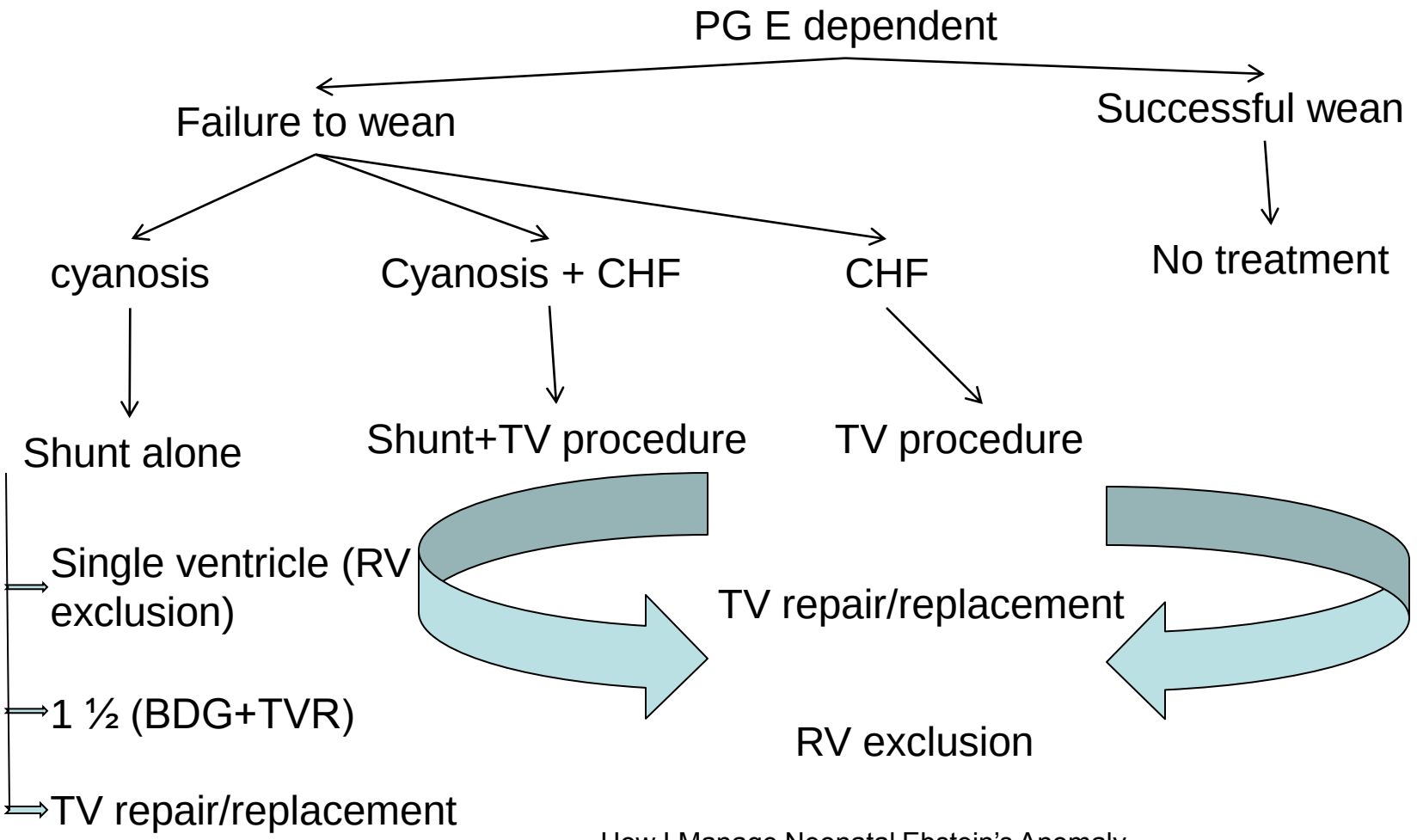
Functional (anatomical?) pulmonic atresia

PDA dependence

Congestive heart failure + RVOTO

Congestive heart failure

Treatment protocol for Ebstein's anomaly in the neonate



How I Manage Neonatal Ebstein's Anomaly
Edward L. Bove, Jennifer C. Hirsch, Richard G. Ohye, and Eric J. Devaney

Univentricular approach (Starnes procedure, 1991)

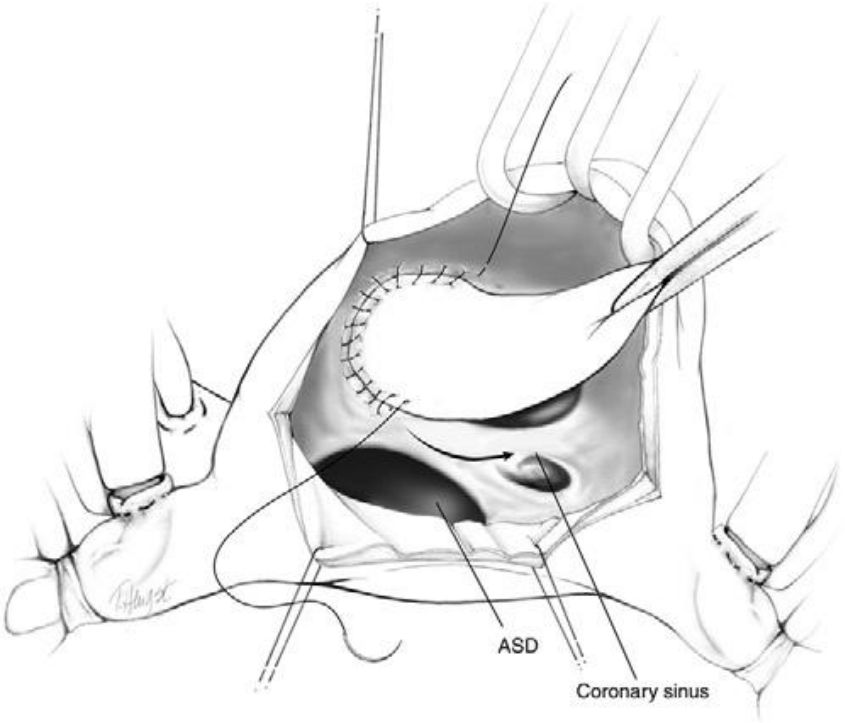
Indication:

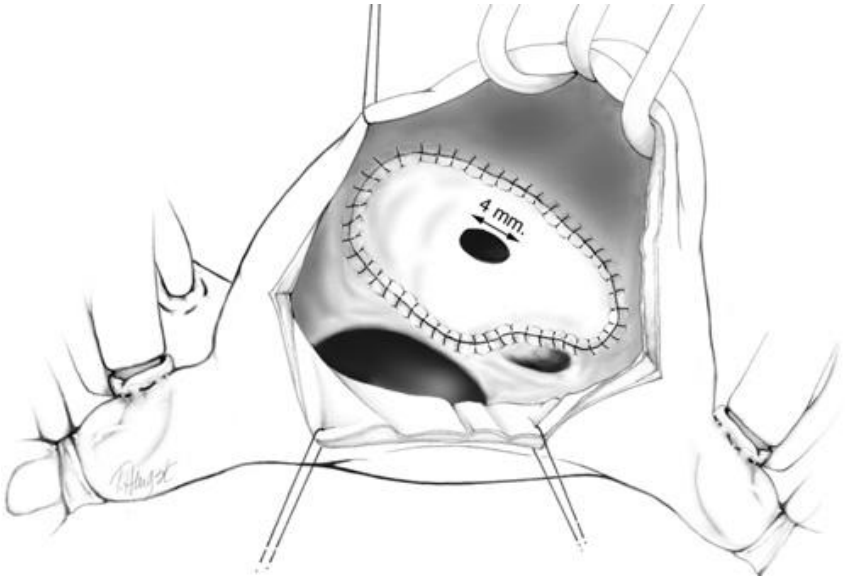
- TV not amenable to repair
- Functional portion of TV is inadequate
- RVOT obstruction

Starnes procedure

- TV closure with fenestrated (4 mm) patch at the anatomic level of the tricuspid annulus.
- Atrial septectomy
- RVOT procedure – only PA division for patient with pulmonary artery insufficiency.
- Reduction atrioplasty
- BT shunt

Starnes procedure





Biventricular repair of neonates and infants

Treatment concept – biventricular repair versus single-ventricle palliation

Biventricular repair (Knott-Craig, 1994) surgical technique:

1. Reduction right atrioplasty (true atrium)

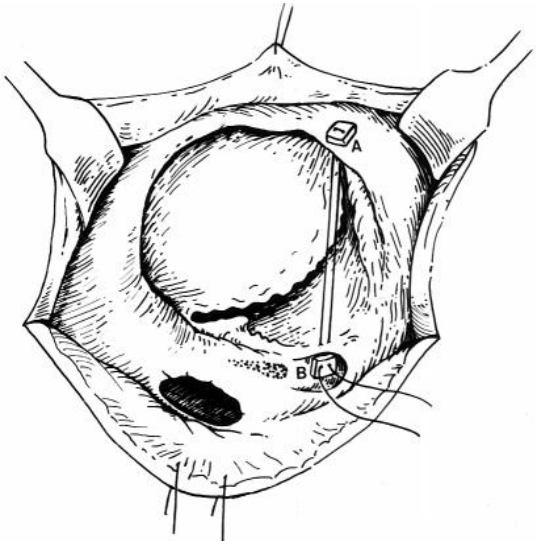
2. TV repair: - reduction annuloplasty (annulus 12-14 mm)
   - construction of monoleaflet TV
   - augmentation of the functional leaflet if deficient

3. ASD closure with 3 - 5 mm fenestration: unload RV; increase cardiac output. Size of the fenestration inversely proportional to the effectiveness of TV repair.

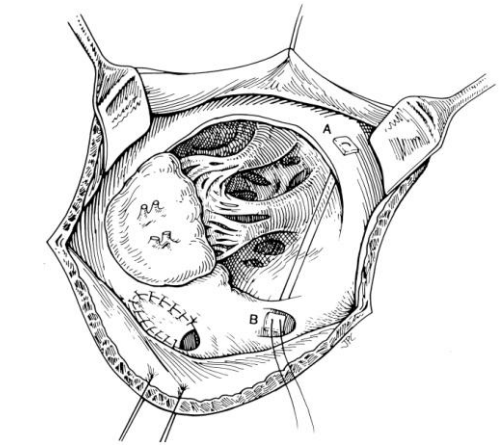
4. Creation of functional RVOT – use a small patch(RVOT – 7-8 mm in neonate);pulmonary insufficiency is very poorly tolerated

5. Patients with suboptimal TV repair – RVOT should be repaired with valve conduit

Anatomical variation of repair

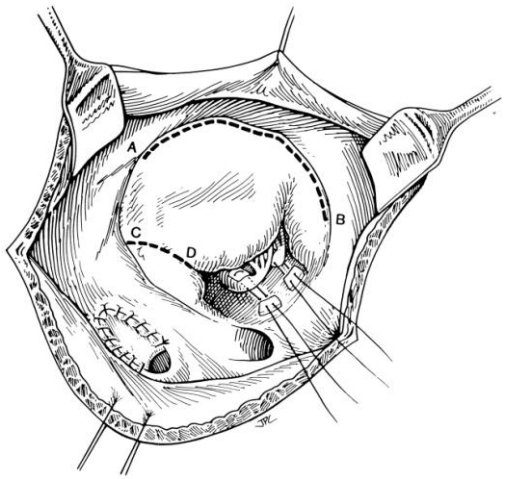


Once detached from the annulus, the anterior leaflet is freed from the underlying muscle ridges at the os infundibuli level, and the annulus is reduced (A to B)

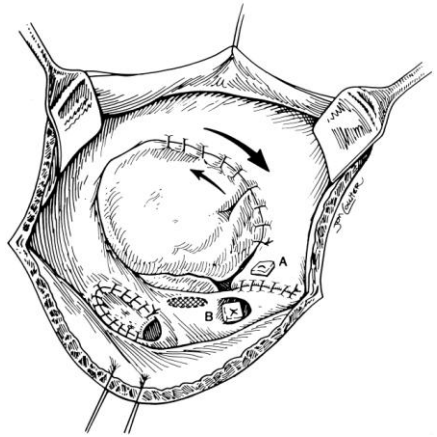


Knott-Craig C. J. et al.; Ann Thorac Surg 2002;73:1786-1793

Fenestrated atrial septal defect closure is shown



When the leaflets are reattached, the leaflets are effectively rotated counterclockwise relative to the annulus, changing the orientation of the orifice to point towards the outflow tract



Knott-Craig C. J. et al.; Ann Thorac Surg 2002;73:1786-1793

Repair of Neonates and Young Infants With Ebstein's Anomaly and Related Disorders

Christopher J. Knott-Craig, MD, Steven P. Goldberg, MD, Edward D. Overholt, MD, Edward V. Colvin, MD, and James K. Kirklin, MD

Department of Thoracic and Cardiovascular Surgery, University of Oklahoma Health Sciences Center, Oklahoma City, Oklahoma, and Division of Cardiothoracic Surgery, University of Alabama at Birmingham, Birmingham, Alabama

Background. Severely symptomatic neonates and young infants with Ebstein's anomaly usually die without surgical intervention. The relative risks and benefits of single-ventricle palliation versus a two-ventricle repair are uncertain. In a recent series, 69% early survival with single-ventricle palliation was reported in 16 neonates with Ebstein's anomaly. Our institutional bias has been to do a two-ventricle repair in all such patients.

Methods. We reviewed our entire surgical experience with a two-ventricle repair in the severely symptomatic neonate (n = 22) and young infant (n = 5). The indications for operation were ventilator dependence, severe cardiac failure, prostaglandin-dependent circulation, and gross cardiomegaly.

Results. Between 1994 and 2006, 27 consecutive patients with Ebstein's anomaly underwent operation. Associated comorbidities included anatomic or functional pulmonary atresia (n = 18), ventricular septal defects (n = 3), small left ventricle (n = 3), hypoplastic branch pulmonary arteries (n = 3), previous cardiac surgery (n = 4), significant intracranial hemorrhage (n = 3), hepatic necrosis and renal insufficiency (n = 3), and malignant tachyarrhythmias (n = 4). Operations consisted of tricuspid valve repair (n = 23) or valve replacement (n = 2), Blalock-Taussig shunt only (n = 1), and bilateral pulmonary arterioplasty with bidirectional Glenn (n = 1). Hospital survival was 74%, and there have been no late deaths during a median follow-up period of 5.4 years (range, 0.2 to 12 years). Three patients required tricuspid valve replacement during the follow-up period. Late arrhythmia requiring medication is present in 1 patient. All patients are currently in New York Heart Association functional class I.

Conclusions. Two-ventricle repair currently has similar early survival compared with single-ventricle palliation. The advantages of a better physiologic repair can be anticipated for a longer follow-up period.

(Ann Thorac Surg 2007;84:587–93)

Ebstein's anomaly (EA) represents a downward, or apical displacement of the posterior and septal leaflets of the tricuspid valve, with a resultant redundant "atrialized" component of the right ventricle situated above the plane of the valve annulus. The anterior leaflet is abnormally broad, often described as saillike, although still based at the true annulus [1, 2]. In the symptomatic neonate with EA, the leading edge of the anterior leaflet is often attached to the free wall of the right ventricle. Associated cardiac anomalies such as pulmonary atresia are common, making a two-ventricle repair technically very difficult [3, 4]. During the past three decades, many repairs have been developed in adults [5] and older children, with variations on the theme of plication of the redundant atrialized right ventricle and utilization of the saillike anterior leaflet in construction of a competent monocuspid valve [6–9]. Symptomatic neonates and very young infants with EA usually present as critically ill, unstable patients, most of whom will die in early infancy without a surgical procedure [7]. In addition, neonates with severe tricuspid regurgitation or gross cardiomegaly, and who are otherwise asymptomatic, are reported to have an associated mortality in infancy of 45% [10, 11]. Single-ventricle palliation of the neonate with EA was first suggested by Starnes and associates in 1991 [12]. We successfully did the first two-ventricle repairs in neonates in 1994, and reported those in 2000. In 2002, Sano and coworkers [13] reported a modified technique of single-ventricle palliation using "ventricular exclusion."

When presented with a critically ill neonate or young infant with EA, the first essential surgical decision that needs to be made is whether a two-ventricle repair is feasible, or whether palliation with embarkation on a single-ventricle pathway is the more reasonable ap-

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Department of Thoracic and Cardiovascular Surgery, University of Oklahoma Health Sciences Center, Oklahoma City, Oklahoma, and Division of Cardiothoracic Surgery, University of Alabama at Birmingham, Birmingham, Alabama

Our institutional bias has been to do a two-ventricle repair in all such patients.

Between 1994 and 2006, 27 consecutive patients with Ebstein's anomaly underwent operation.

Table 1. Associated Comorbidities

Comorbidity	n
Pulmonary atresia (functional 7, anatomic 11)	18
Ventricular septal defect	3
Small left ventricle	3
Hypoplastic branch pulmonary arteries	3
Previous cardiac surgery	4
Grade III or IV intracranial hemorrhage	3
Ischemic hepatic necrosis and renal failure (creatinine >1.5)	3
Malignant tachyarrhythmias	4

Three additional neonates responded well to nitric oxide and prolonged anesthesia, and were ultimately weaned from the ventilator and discharged without surgical intervention

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proach. The relative risks and benefits of single-ventricle palliation compared with two-ventricle repair are uncertain. In 2006 Reemtsen and colleagues [4] updated their results of single-ventricle palliation (closure of the tricuspid orifice with a fenestrated patch, aortopulmonary shunt, and later Fontan completion) in 16 neonates with EA, with 69% early survival. Our institutional preference has been to perform a two-ventricle repair in all neonates and young infants with EA and similar disorders. The results of this approach are presented in this manuscript.

Patients and Methods

Between 1994 and December 2006, we operated on 27 consecutive symptomatic neonates (n = 22) and young infants (n = 5) with EA, at Children's Hospital of Oklahoma and University of Alabama at Birmingham. Approval for this analysis was obtained from the institutional review boards. This series includes 1 neonate operated on in Indianapolis with John Brown, MD, and 1 in San Diego with Glenn Pelletier, MD. Three additional

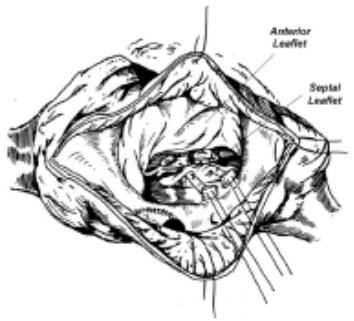

Fig 1. Sobering single-stitch valvuloplasty (papillary muscle of anterior leaflet through tethered septal leaflet).

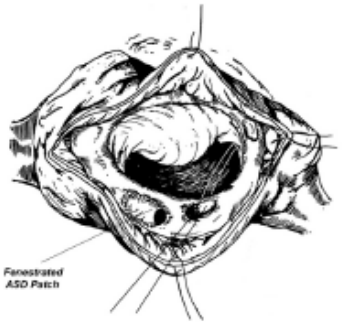

Fig 2. Tricuspid annuloplasty stitch placed in coronary sinus and at location of anteroposterior commissure. (ASD = atrial septal defect.)

neonates responded well to nitric oxide and prolonged anesthesia, and were ultimately weaned from the ventilator and discharged without surgical intervention. One patient subsequently had a successful repair at 8 months of age. There were no neonates with Uhl's anomaly in this series.

The indications for operation were ventilator dependence, prostaglandin-dependent circulation, severe cardiac failure, and gross cardiomegaly. Table 1 describes the associated comorbidities in our patients, most commonly pulmonary atresia (n = 18, 67%), ventricular septal defects (n = 3), small left ventricle (n = 3), hypoplastic branch pulmonary arteries (n = 3), previous cardiac operation (n = 4), significant intracranial hemorrhage (n = 3), ischemic hepatic necrosis and renal insufficiency (creatinine > 1.5; n = 3), and malignant tachyarrhythmias (n = 4). Recent clinical and echocardiography follow-up was complete. Median follow-up was 5.4 years (range, 0.2 to 12 years).

In one patient, the Starnes repair was successfully taken down, and the patient was converted to a two-ventricle repair with tricuspid valve replacement. Another critically ill infant transferred with complete heart block, device closure of an atrial septal defect, and prior tricuspid valve replacement with a tissue prosthesis. After subsequent balloon disruption of the bioprosthesis at the referring institution, the patient underwent successful repeat replacement of the tricuspid valve, removal of the atrial septal defect device, and patch closure of the atrial septum. One patient remained ventilator-dependent despite prior Blalock-Taussig shunt and balloon dilatation of the pulmonary arteries before being referred for a two-ventricle repair. Another patient had four prior sternotomies and three Blalock-Taussig shunts before being referred for repair.

The essential principles in our two-ventricle repair, as

In one patient, the Starnes repair was successfully taken down, and the patient was converted to a two ventricle repair with tricuspid valve replacement.

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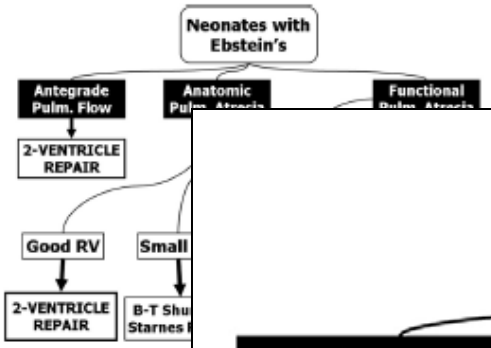

Fig 8. Management algorithm of Ebstein's patients based on associated anomalies and overall stability. (B-T = Blalock-Taussig; LV = left ventricle; MPA = main pulmonary artery; Pulm. = pulmonary; RV = right ventricle.)

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cally ill patient came to us
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gery at 10 days of life, but uni
of the intracranial hemorrhag
repair, the situation was irre
withdrawn.

Comment

Since 1994 we have operated
or young infants, and have
survival rates that are similar
single-ventricle palliation [4].
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a two-ventricle system will
course of a longer follow-up
recent report by Reemtsen a
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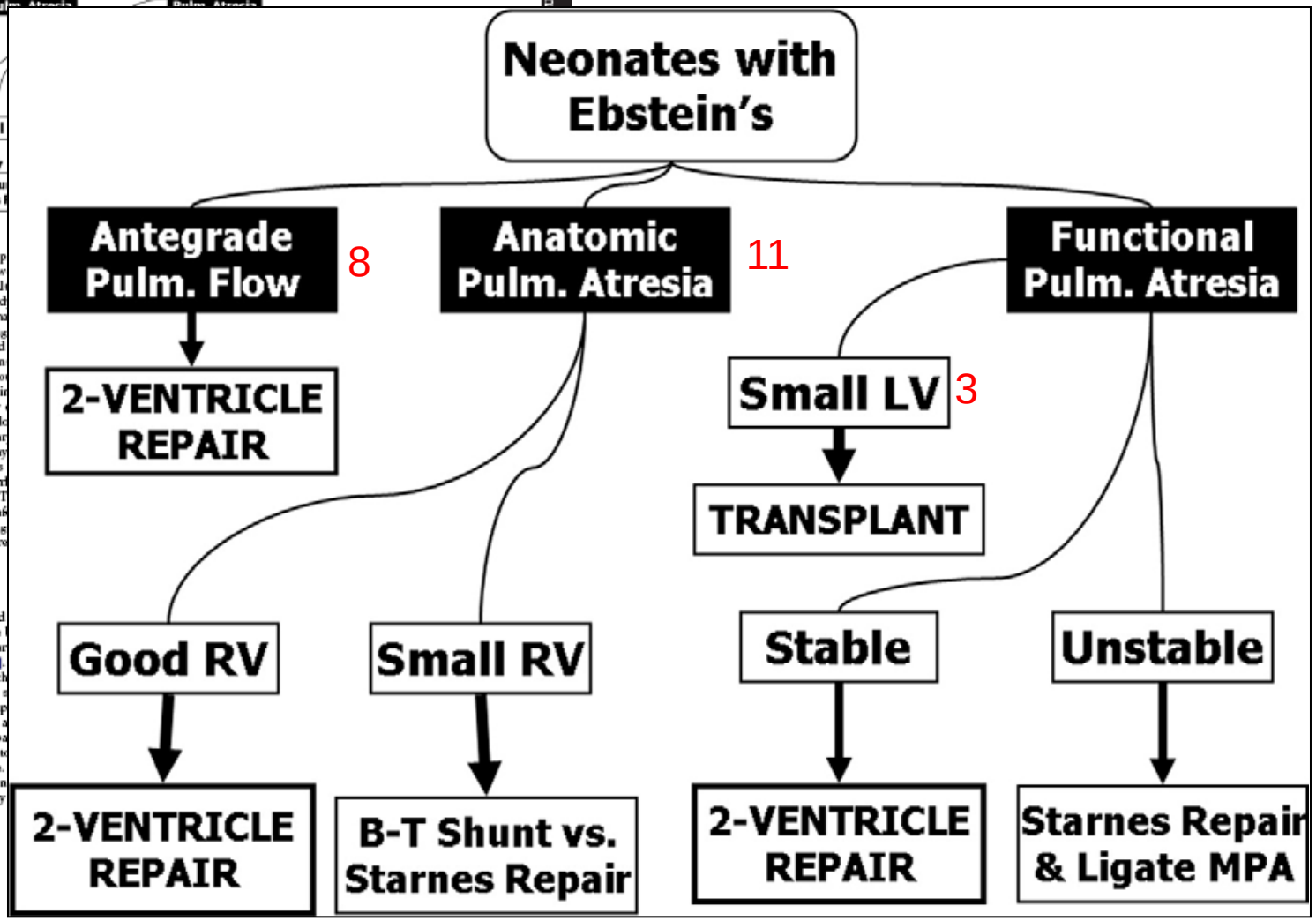

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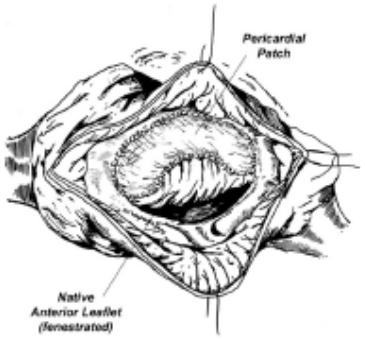


Fig 6. Augmentation of anterior leaflet with an autologous pericardial patch before annuloplasty.

facing septal wall. The annuloplasty stitch is placed with one pledgetted end in the coronary sinus, the other pledgetted end at the location of the commissure between the anterior and (if otherwise present) posterior leaflets (Fig 2). Approximation of the annuloplasty stitch effectively partitions the tricuspid valve orifice into two openings, the "main" orifice and a more "caudal" or "rightward" orifice (Fig 3A). If the valve is then competent, the caudal orifice is closed, plicating the atrialized portion of the right ventricle at the same time as reducing the annular diameter (Fig 3B). A more complex repair is required if the anterior leaflet is dysplastic, poorly fenestrated (restricting right ventricular inflow), or more firmly tethered to the underlying myocardium. In these cases, the leaflet is taken down from the annulus [16, 17], freed up from its underlying attachments (Fig 4), and then reattached to the newly reduced annulus (Fig 5). If the anterior leaflet does not reach the opposing wall, then it can be detached from the annulus and enlarged with an autologous pericardial patch to allow it to bridge the gap (Fig 6). In two of our patients, a modified Blalock-Taussig shunt was added to improve postoperative pulmonary blood flow. In all patients, the placement of a peritoneal dialysis catheter has become very helpful in draining postoperative ascites, aiding diaphragmatic excursion and postoperative pulmonary mechanics.

Postoperatively, the patients initially remain paralyzed, sedated, and ventilated. We use large tidal volumes on the ventilator (12 to 15 mL/kg) because the lungs are usually suboptimal, their full development having been difficult in a chest crowded by cardiomegaly. We institute all measures to reduce elevated pulmonary vascular resistance, including inhaled nitric oxide. Oxygen carrying capacity is optimized by maintaining a hematocrit of at least 45% to 50%, as the oxygen saturation may be low (and tolerated) for the first 3 to 5 days after surgery. We have found isoproterenol infusion (0.05 $\mu g \cdot kg^{-1} \cdot min^{-1}$) to be the most useful inotropic agent in the perioperative period.

Results

In all but 2 patients, two-ventricle repairs were performed. In 1 patient with a small right ventricle and only mild tricuspid regurgitation, but with a prostaglandin-dependent circulation, a Blalock-Taussig shunt only was done. This was taken down 6 months later and a bidirectional Glenn added. Another infant who had three previous Blalock-Taussig shunts elsewhere, before being referred for repair, had extensive bilateral patch augmentation of the pulmonary arteries and a bidirectional Glenn.

Seventy-four percent (20 of 27 patients) survived to hospital discharge. Recent follow-up was achieved in all survivors. The median follow-up was 5.4 years (range, 0.2 to 12 years). There have been no late deaths. All patients are currently in New York Heart Association class I functional status. Three patients have required tricuspid valve replacement during the follow-up period. Bioprostheses were placed 2 to 4 years after their initial operation. Although 4 patients had symptomatic tachyarrhythmias preoperatively, and despite the well-known association of EA with accessory pathway tachydysrhythmia [2, 6], we have had only 1 patient (5%, 1 of 20 patients) manifest a transient tachyarrhythmia requiring medication during the follow-up period.

Of the 7 nonsurvivors, the factors associated with their poor outcome were variable. Of the 11 patients with anatomic pulmonary atresia, 6 died (66%) postoperatively. Three patients required support with an extracorporeal membrane oxygenator postoperatively—1 patient experienced extracorporeal membrane oxygenator pump failure on the third postoperative day; 1 required extracorporeal membrane oxygenator followed by tricuspid valve replacement, the function having been deemed "marginal" by echocardiography; the third extracorpo-

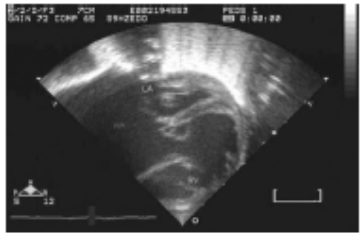


Fig 7. Echocardiogram of a patient with an unacceptably small left ventricle. (LA = left atrium; RA = right atrium; RV = right ventricle.)

Results:

1. In 2 patients BT shunt was added to improve pulmonary blood flow

2. 2 patient underwent single-ventricle repair (25 patient underwent two-ventricle repair)

3. 74% (20 from 27) survive to hospital discharge & no late death

4. 3 patient have required tricuspid valve replacement during follow-up period (5 from 27 patiens – TVR)

5.Of the 11 patients with anatomic pulmonary atresia – 6 died (66%)

6. 3 patients required ECMO support postoperatively (1 required ECMO followed by tricuspid valve replacement). No patient survive to hospital discharge.

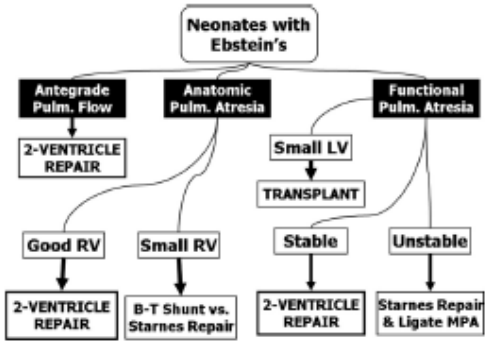


Fig 8. Management algorithm of Ebstein's patients based on associated anomalies and overall stability. (B-T = Blalock-Taussig; LV = left ventricle; MPA = main pulmonary artery; Pulm. = pulmonary; RV = right ventricle.)

real membrane oxygenator patient had hypoplastic pulmonary arteries, and underwent homograft replacement of the pulmonary valve. Although this patient subsequently had excellent hemodynamics, she experienced a severe intracranial hemorrhage and support was withdrawn. No patient requiring extracorporeal membrane oxygenator support survived to hospital discharge. Two patients had small left ventricles preoperatively, and succumbed to low cardiac output after surgery (Fig 7). There was a technical error in 1 patient, involving injury to the right coronary artery during the reduction atrioplasty. After this case, we adopted a policy of mandatory marking of the right coronary artery course with a surgical pen before making any atriotomy. Finally, 1 critically ill patient came to us already having suffered a grade IV intracranial hemorrhage associated with *Staphylococcus aureus* septicemia. This patient underwent surgery at 10 days of life, but unfortunately had a recurrence of the intracranial hemorrhage, and despite a satisfactory repair, the situation was irretrievable and support was withdrawn.

Comment

Since 1994 we have operated on 27 critically ill neonates or young infants, and have been able to achieve early survival rates that are similar to recent reports involving single-ventricle palliation [4]. In our opinion, it remains a feasible, reasonable approach, and the benefits of having a two-ventricle system will surely be realized over the course of a longer follow-up period. In contrast to the recent report by Reemtsen and associates [4] of 16 patients with single-ventricle palliation in whom the tricuspid valve was not amenable to repair, we felt comfortable repairing all but 1 neonate. This patient had prostaglandin-dependent circulation and 2/4 tricuspid regurgitation; we elected to initially place a small aortopulmonary shunt, followed 4 months later by a bidirectional Glenn anastomosis, with an excellent result.

From our experience (74% survival), it seems evident that some neonates may have had a better chance of survival with an alternative operation. This was most evident in patients with anatomic pulmonary atresia, which was associated with a 66% mortality. Many of these patients had small pulmonary arteries; others had diminutive functional right ventricles. As the primary chordae to the anterior leaflet often originate from rudimentary papillary muscles in the infundibulum, these may easily be injured during the construction of a right ventricular outflow tract patch or pulmonary valve replacement, making the tricuspid repair inadequate.

The dilemma remains how to decide which operation is most suitable for each patient. From our experience, we currently feel confident in our ability to repair a neonate who has some prograde flow across the pulmonary valve, albeit while optimizing the pulmonary vascular resistance with inhaled nitric oxide and a low-dose infusion of isoproterenol. Figure 8 represents our management algorithm when approaching a patient with EA, based on their associated anomalies, as well as their overall clinical stability. Patients with pulmonary atresia—representing greater than 60% of the patient group in our series—seem to fall into two general groups: those who are relatively stable on the ventilator, often with gross cardiomegaly, severe tricuspid regurgitation, and a dysplastic (rather than a true EA-like) valve; and those who are very unstable with ongoing progressive metabolic acidosis and functional pulmonary atresia, often with retrograde flow back through the pulmonary valve. The former usually do well with either a two-ventricle repair if they have a decent size functional right ventricle, or just an initial shunt followed by a 1½-ventricle repair at 4 to 6 months of age if they have small functional right ventricles; the latter are probably best served with initial

CARDIOVASCULAR

Patients with pulmonary atresia—representing greater than 60% of the patient group in our series—seem to fall into two general groups:

those who are relatively *stable* on the ventilator, often with gross cardiomegaly, severe tricuspid regurgitation, and a dysplastic (rather than a true EA-like) valve;

and those who are very unstable with ongoing progressive metabolic acidosis and functional pulmonary atresia, often with retrograde flow back through the pulmonary valve.

The 1° group usually do well with either a two-ventricle repair if they have a decent size functional right ventricle, or just an initial shunt followed by a 1½-ventricle repair at 4 to 6 months of age if they have small functional right ventricles

The 2 group are probably best served with Starnes
Single ventricle palliation

Neonatal Ebstein's

- Patients with antegrade pulmonary flow & good size functional RV – biventricular repair
- Patient with pulmonary atresia (functional or anatomical) and cyanosis – shunt in neonatal period followed by definitive repair at 4 – 6 months of age
- Patient with pulmonary atresia, cyanosis and CHF – single ventricle palliation

Ebstein's anomaly-adult patient Indications for operation

- Symptoms of dyspnea or right-sided heart failure (NYHA III-IY)
- Progressive RV dilatation (before significant RV dysfunction)
- Onset / progression of arrhythmias
- Earlier operation if good TV repair is likely
- Cardio-thoracic ratio > 0.65
- Severe, progressive cyanosis
- Reduced LV function
- Associated lesions

Echo assessment of TV

Anterior leaflet:

- At least 50% delamination of anterior leaflet

  Multiple attachments of anterior leaflet (most probably mobilization will be inadequate and coaptation with septum ineffective);

- Free leading edge of anterior leaflet

  Papillary muscle attachment to the leaflet prohibit good repair

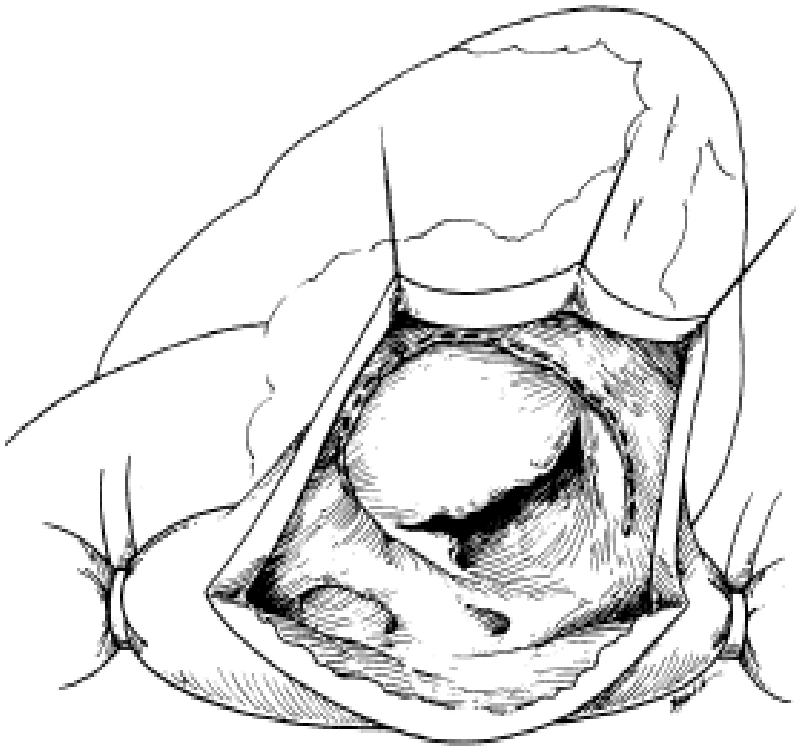
TV displacement:

- If TV doesn't view in the apical 4 chamber view (with a crux in the view) – TV is severe displaced to outflow, functional RV is small – better option replaced the valve

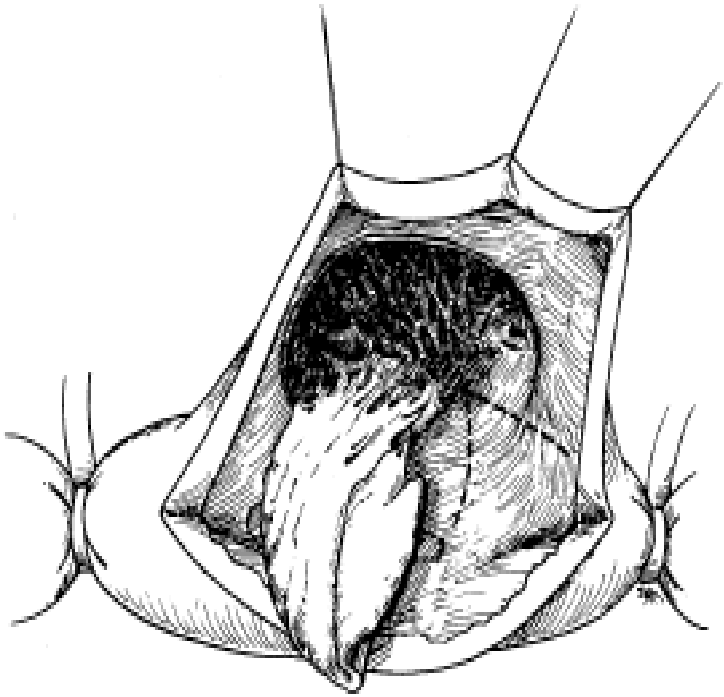
TV - Doppler underestimate TR: - RV pressure is low

- TR direction unusual

Surgical technique

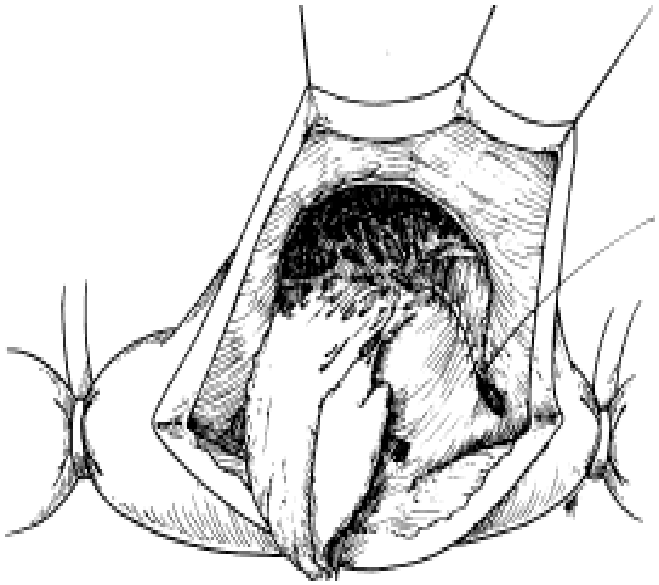


Detachment of the anterior leaflet from the tricuspid annulus.

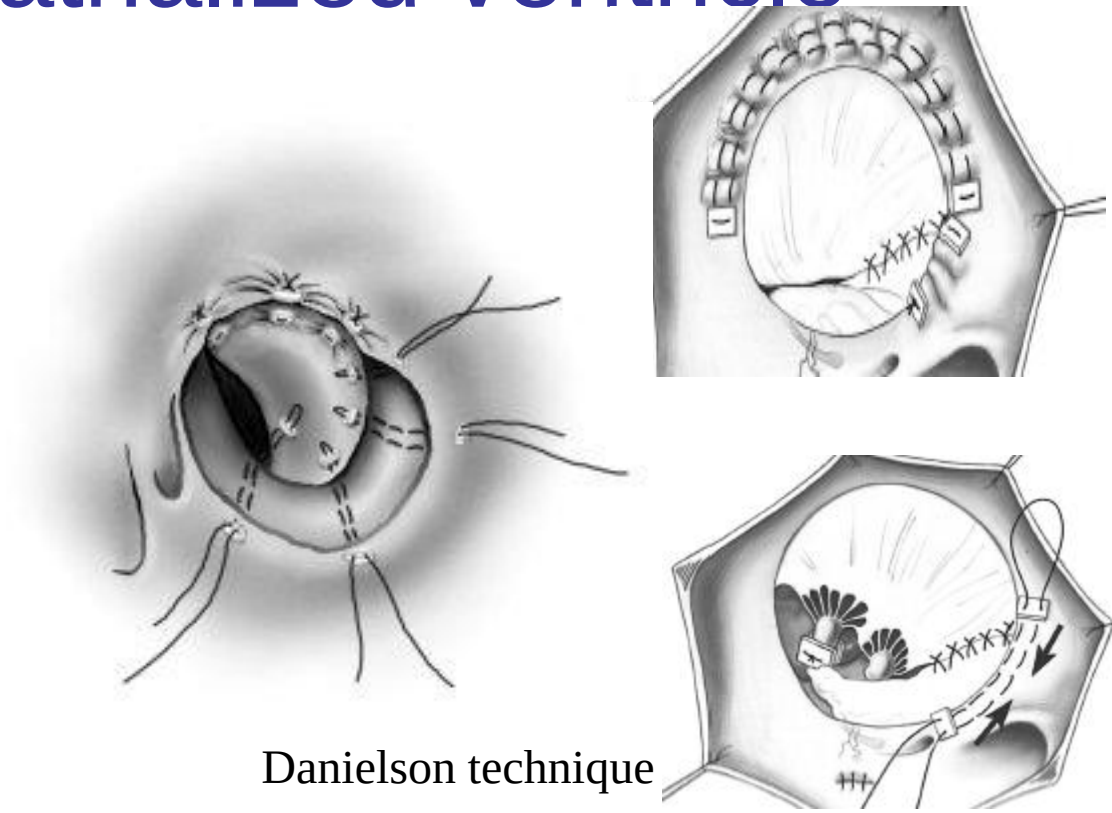


Leaflet fully mobilized. Area of atrialized ventricle to be plicated noted by dotted lines.

Plication of atrialized ventricle



Carpentier technique

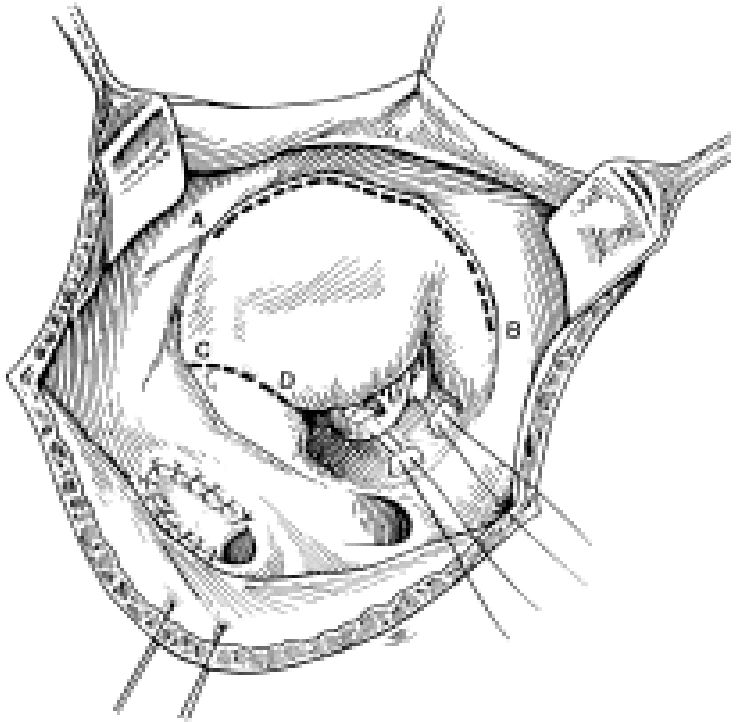


Danielson technique

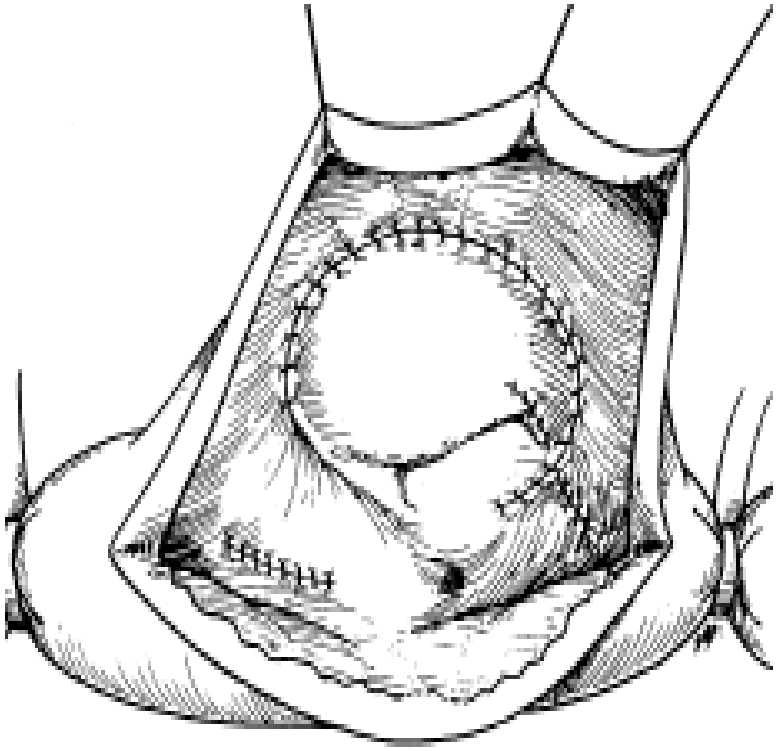
Advantage of RV plication: improve transit of blood through the right side of the heart; lessen compression on the LV

Disadvantage of RV plication: potential compromise of the coronary artery supply to RV; risk of ventricular arrhythmias

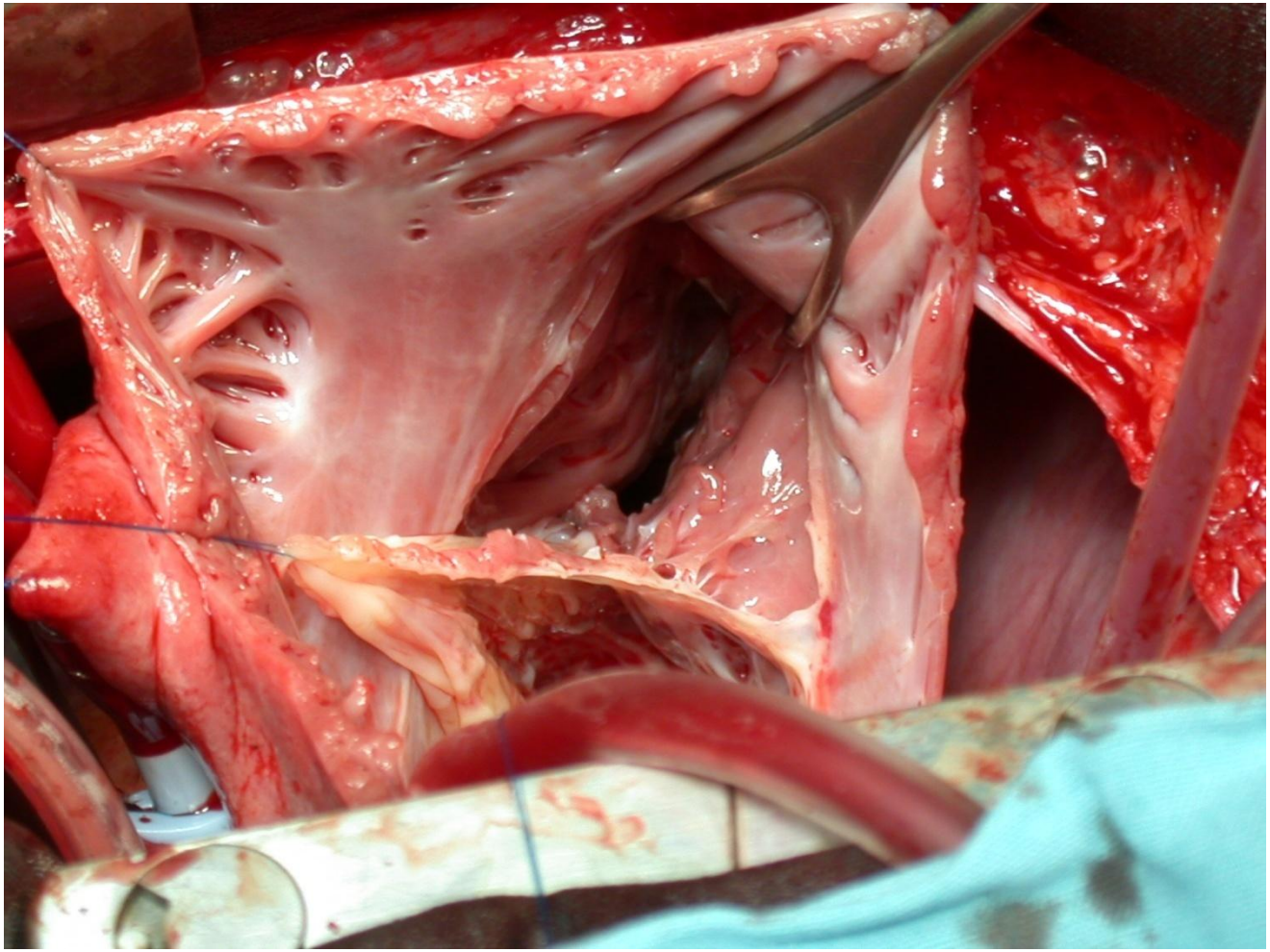
Tricuspid valve repair

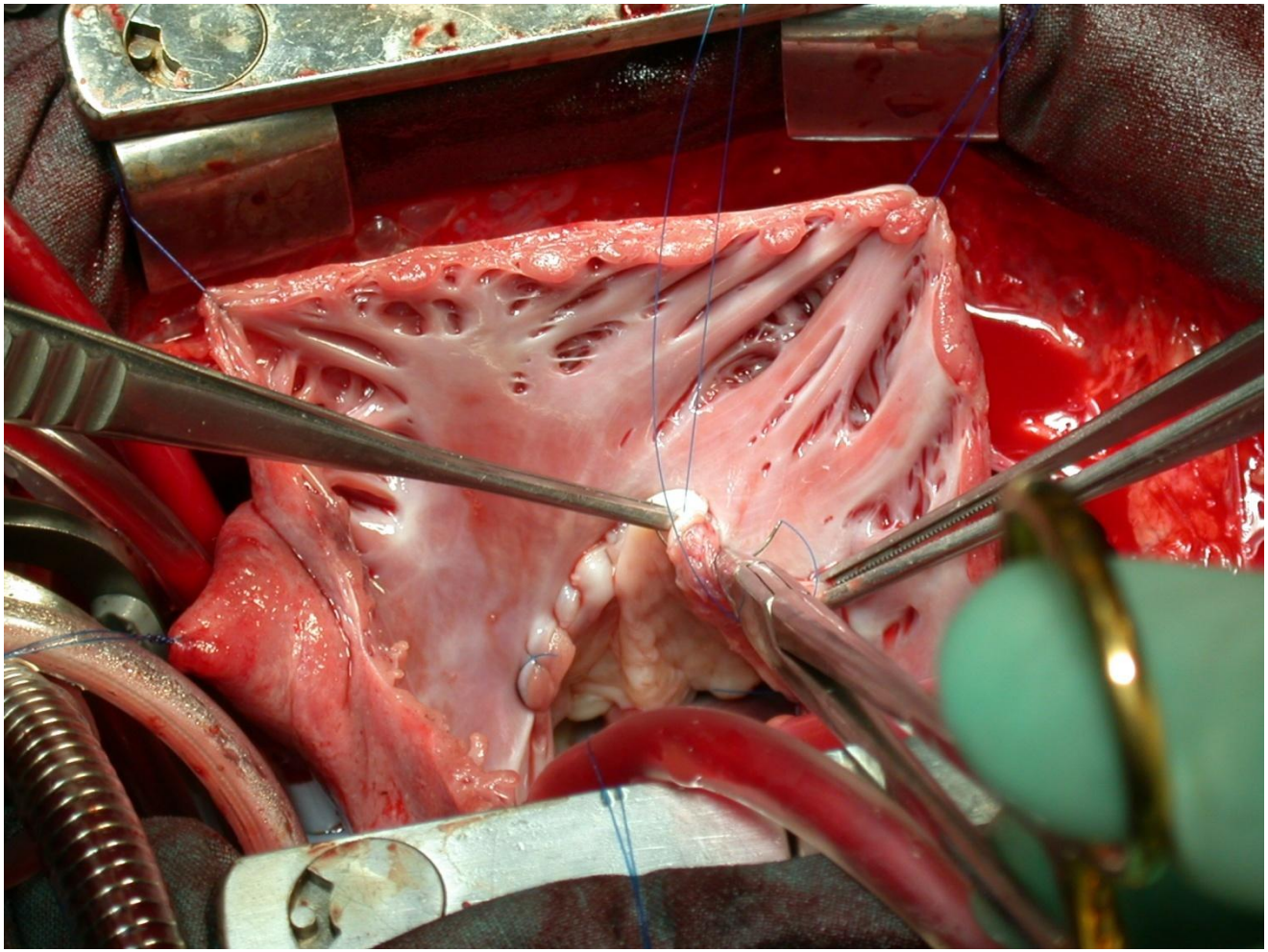


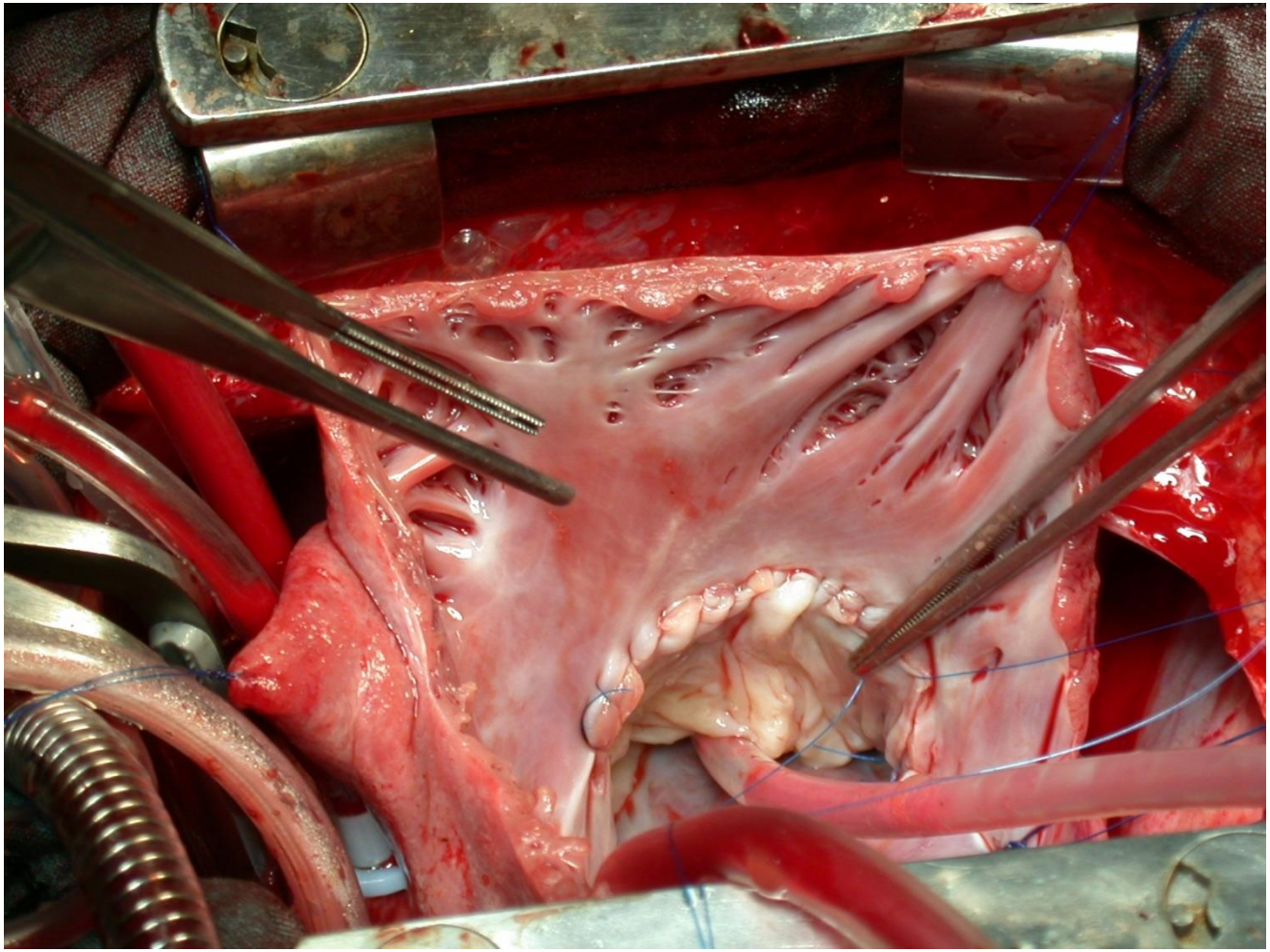
Pledgetted sutures drag the anterior papillary muscle closer to the septal leaflet to improve coaption

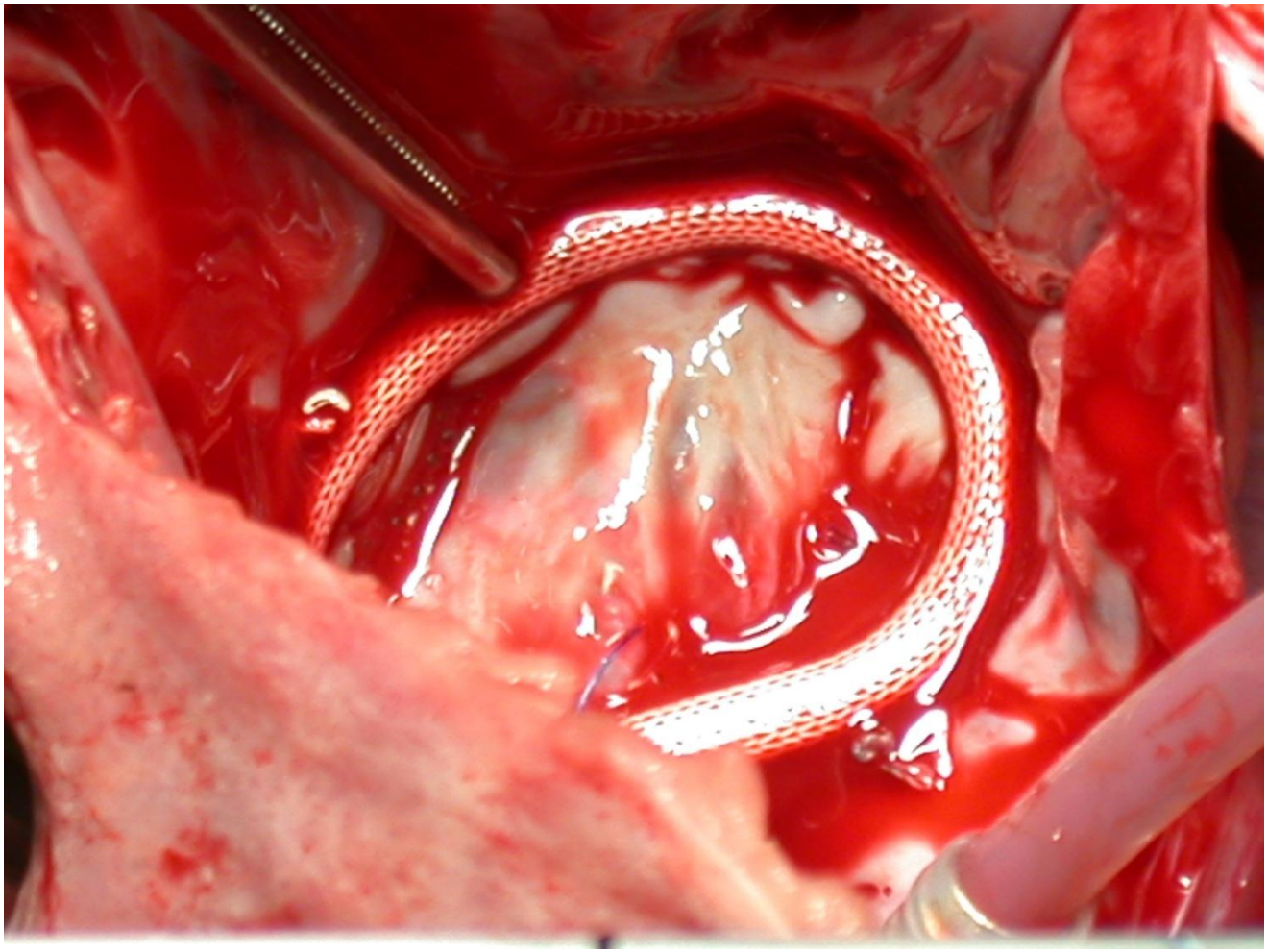


Reconstructed tricuspid valve









1 ½ repair for Ebstein anomaly

- BDG provide pulmonary flow
- BDG decrease RV volume overload
- BDG support LV preload and output
- Functional improvement of TV repair

Hemodinamic conditions for BDG construction

- LV EDP < 15 mm Hg
- Transpulmonary gradient < 10 mm Hg
- Mean pulmonary pressure < 18 – 20 mm Hg

Indication for BDG

Physiological:

- Inadequate RV function (cyanosis +↓NYHA class)
- Cyanosis at rest
- Effort induced cyanosis (low threshold for BDG)
- Intraoperative RA/LA pressure > 1.5

Indication for BDG

Anatomical:

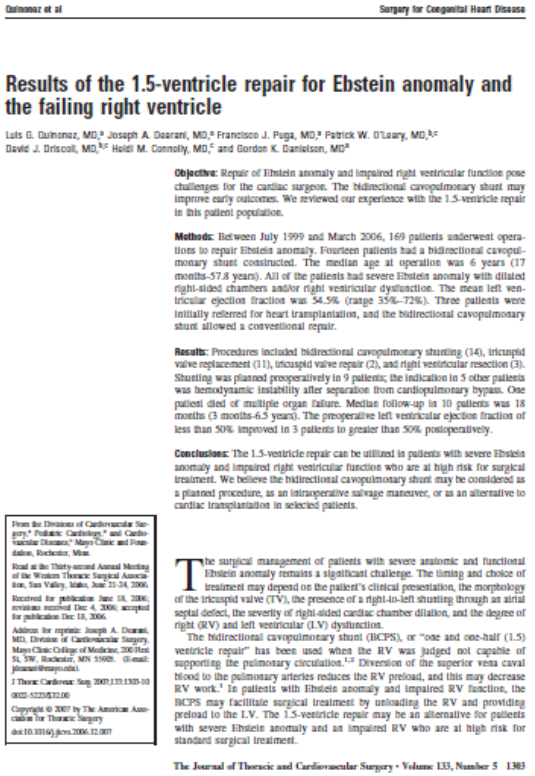
- Small TV orifice/TV stenosis on post-op TEE

- Residual TR

Indication for BDG

Patient with depressed LV function (secondary to RV dysfunction):

- LV EF < 25% - heart transplant
- LV EF – 25 – 35% - BDG + competent TV
- LV EF > 35% - biventricular repair



Quinonez et al — Surgery for Congenital Heart Disease

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

Luis G. Quinonez, MD,[a] Joseph A. Dearani, MD,[a] Francisco J. Puga, MD,[a] Patrick W. O'Leary, MD,[b,c] 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.

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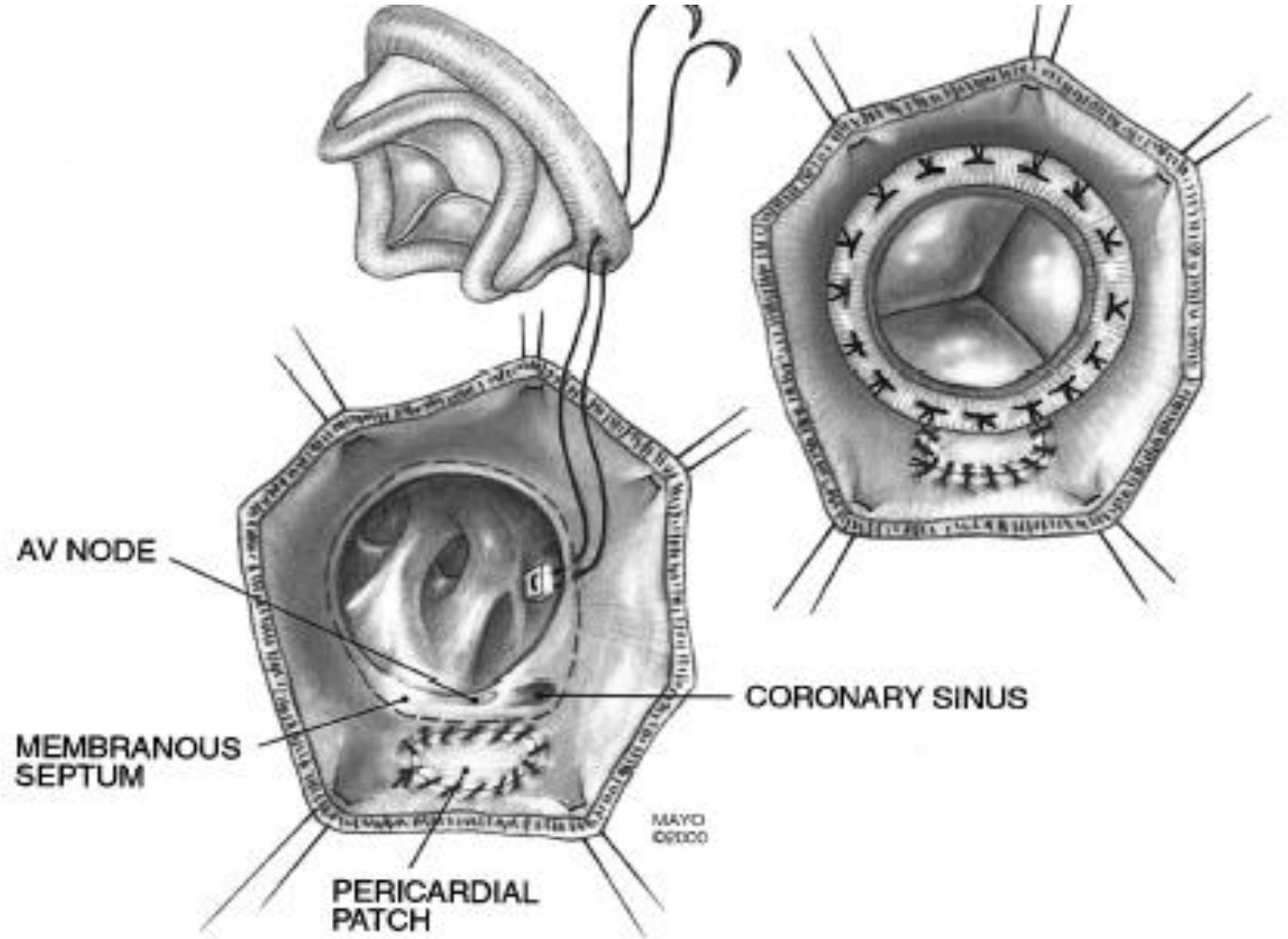
The surgical management of patients with severe anatomic and functional Ebstein anomaly remains a significant challenge. The timing and choice of treatment may depend on the patient's clinical presentation, the morphology of the tricuspid valve (TV), the presence of a right-to-left shunting through an atrial septal defect, the severity of right-sided cardiac chamber dilation, and the degree of right (RV) and left ventricular (LV) dysfunction.

The bidirectional cavopulmonary shunt (BCPS), or "one and one-half (1.5) ventricle repair" has been used when the RV was judged not capable of supporting the pulmonary circulation.[1,2] Diversion of the superior vena caval blood to the pulmonary arteries reduces the RV preload, and this may decrease RV work.[3] In patients with Ebstein anomaly and impaired RV function, the BCPS may facilitate surgical treatment by unloading the RV and providing preload to the LV. The 1.5-ventricle repair may be an alternative for patients with severe Ebstein anomaly and an impaired RV who are at high risk for standard surgical treatment.

CHD

Tricuspid valve replacement



European Congenital Heart Association

150 patients; 13 centers; 1992 – 2005

Median age – 6 y.

80% - type B, C (Carpentier classification)

60% - NYHA III – IY

Procedure: TV replacement – 33%

TV repair – 27%

1 ½ repair (46 pat) (50% - TV replacement;
50% - TV repair) – 26 %

Other – 14%

European Congenital Heart Association

Operative mortality – 13.3% (20 patients):

- TV replacement – 5
- TV repair – 3 pat
- 1 ½ - 7 pat (15%)
- other – 5 pat

Mortality 0% after age 10 y.

Risk factors: age/ palliative surgery associated with younger age.