Surgical Repair of Tricuspid Valve Anomalies in Infants and Children

Review

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Discussion Issues

- Ebstein’s anomaly, especially in neonates
- Tricuspid regurgitation in HLHS and single ventricle
- Tricuspid valve in PA+IVS
- Straddling tricuspid valve
- Rare tricuspid valve diseases in infants and children
- Pacemaker induced TVR

Each valve is unique, and each operation is slightly different.
The Mechanisms of Development of Significant AV Valve Insufficiency

Ebstein’s Anomaly

- Occurs in 1–5 of 200,000 live births, accounting for less than 1% of all congenital heart defects.

- Encompasses a wide anatomic spectrum of abnormalities of the tricuspid valve and of the right ventricle including atrialization of parts of the right ventricle due to apical displacement of the septal and posterior tricuspid valve leaflets.

- Additional cardiac anomalies are common

- **Clinical presentation is usually age dependent**

Isolated congenital tricuspid stenosis or regurgitation occurs even more rarely than Ebstein’s anomaly
First description by Wilhelm Ebstein in 19-year-old laborer

Wilhelm Ebstein
1836-1912

Arch Anat Physiol 1866;
7:238–254.

In 1951, Soloff and colleagues first reported a patient in whom the diagnosis was made during life, using cardiac catheterization and angiocardiography.

Carpentier Classification of Ebstein’s Anomaly


**Type A** - The volume of the true right ventricle is adequate

**Type B** - A large atrialized component of the right ventricle exists, but the anterior leaflet of the tricuspid valve moves freely

**Type C** - The anterior leaflet is severely restricted in its movement and may cause significant obstruction of the right ventricular outflow tract

**Type D** - Almost complete atrialization of the ventricle except for a small infundibular component.
Neonatal Ebstein’s Anomaly - Great Ormond Street Echo Score (GOSE)

The ratio of combined end-diastolic area of the right atrium and atrialized RV combined area of the functional RV, LA and LV in a four chamber view

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

Mortality in infants with GOSE Score >1,1-1.5 combined with cyanosis and cardiomegaly approaches 100%

Celermajer et al. JACC 1992;19:1041-6

Jaquiss and Imamura Sem. TCVS 2007;19:358-63
### Development of Surgery for Ebstein’s Anomaly

<table>
<thead>
<tr>
<th>Year</th>
<th>Event</th>
</tr>
</thead>
<tbody>
<tr>
<td>1965</td>
<td>Glenn – Cavo-pulmonary shunt for Ebstein malformation</td>
</tr>
<tr>
<td>1958</td>
<td>Lillehei – Proposed new form of surgical therapy</td>
</tr>
<tr>
<td>1963</td>
<td>Barnard – First TV replacement in patient with Ebstein’s anomaly</td>
</tr>
<tr>
<td>1964</td>
<td>Hardy performed clinically first successful functional repair with exclusion of the atrialized chamber</td>
</tr>
<tr>
<td>1979</td>
<td>Danielson technique</td>
</tr>
<tr>
<td>1998</td>
<td>Carpenter rotation technique + ring</td>
</tr>
<tr>
<td>1991</td>
<td>Quaegebeur technique</td>
</tr>
<tr>
<td>1981</td>
<td>Sebening - stitch</td>
</tr>
<tr>
<td>1986</td>
<td>Starnes operation</td>
</tr>
<tr>
<td>1998</td>
<td>Hetzer technique</td>
</tr>
<tr>
<td>2000</td>
<td>Knott-Craig technique for newborns</td>
</tr>
<tr>
<td>2001</td>
<td>Wu technique</td>
</tr>
<tr>
<td>2002</td>
<td>Sano – right ventricular exclusion</td>
</tr>
<tr>
<td>2004</td>
<td>Friesen technique – posterior annular plication</td>
</tr>
<tr>
<td>2004</td>
<td>Da Silva – Cone repair</td>
</tr>
<tr>
<td>2014</td>
<td>Prifti - Peacock cone technique</td>
</tr>
</tbody>
</table>

### Atrialized Right Ventricle Management

- **Transverse plication** (Hardy 1964; Danielson 1992).
- **No plication** (Sebening 1981; Hetzer 1998).
Circulatory Bypass of the Right Side of the Heart

VI. Shunt between Superior Vena Cava and Distal Right Pulmonary Artery; Report of Clinical Application in Thirty-eight Cases

By William W. L. Glenn, M.D., Nelson K. Ordway, M.D., Norman S. Talner, M.D., and Edward P. Call, Jr., M.D.

Figure 9

Technic of operation for Ebstein’s anomaly with predominant left-to-right shunt through patent foramen ovale. The cavo-pulmonary artery shunt is performed first, followed by closure of the foramen ovale and resection of a part of the wall of the right atrium with the aid of cardiopulmonary bypass.

Circulation 1965;31:172-89
<table>
<thead>
<tr>
<th>Risk Factors for Tricuspid Repair</th>
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</thead>
<tbody>
<tr>
<td>Advanced cardiac insufficiency, cyanosis</td>
</tr>
<tr>
<td>Very small right ventricle</td>
</tr>
<tr>
<td>Carpentier groups C and D</td>
</tr>
<tr>
<td>Restricted cardiac chamber function</td>
</tr>
<tr>
<td>Tachyarrythmia, esp. Atrial fibrillation</td>
</tr>
<tr>
<td>Associated defects</td>
</tr>
<tr>
<td>Newborns in critical condition</td>
</tr>
</tbody>
</table>
Monocusp repair needs a large, mobile anterior leaflet. There were a limited number of patients with acceptable anatomy for this type of repair.
Dis. Chest 1958;33:297-302

C. Walton Lillehei
1918-1999

Figure 4: Shows the triangular piece of atrialized ventricle between the septal leaflet and the true annulus which is to be excluded. Distortion of septal leaflet is seen.—Figure 5: Shows the triangular piece of atrialized ventricular tissue between the posterior leaflet and the true annulus which is to be excluded from heart. R.A. = right atrium, A.L. = anterior leaflet and P.L. = posterior leaflet of tricuspid valve.—Figure 6: The superior border of the displaced posterior leaflet has been approximated to the true annulus fibrosis. The triangular area has been excluded by the plicating stitches.
Surgical correction of Ebstein’s malformation with prosthetic tricuspid valve

Surgery 1963;54:302-8

Ebstein’s malformation is a rare congenital cardiac anomaly in which there is abnormal downward displacement of the septal and/or posterior tricuspid leaflets into the right ventricle. The presence of an atrial septal defect, the frequently severe tricuspid valve incompetence, and often debilitating supraventricular arrhythmias (occasionally associated with Wolff-Parkinson-White syndrome) produce the clinical syndrome in Ebstein’s malformation. In some instances an atrialized right ventricle would not interfere with right ventricular output and correctly concluded that the patient would benefit from replacement of the tricuspid valve.

EBSTEIN'S ANOMALY: A FUNCTIONAL CONCEPT AND SUCCESSFUL DEFINITIVE REPAIR

Kenneth L. Hardy, M.D. (by invitation), Ivan A. May, M.D. (by invitation), Charles A. Webster, M.D. (by invitation), and Kent G. Kimball, M.D. (by invitation), Oakland, Calif.

Hardy et al JTCVS 1964;8:927-940
Danielson Technique


- Plication of the free wall of the atrialized portion
- Posterior tricuspid annuloplasty and right atrial reduction
- Creation of a monocusp valve
Current repair usually involves bringing the anterior papillary muscle(s) toward the ventricular septum, thus facilitating coaptation of the leading edge of the anterior leaflet with the ventricular septum.

“if the anterior tricuspid leaflet was of adequate size. A single mattress suture reinforced by a Teflon felt pledget is placed at point A through the anterior leaflet and then brought to point B between the rudimentary remnants of the septal and posterior tricuspid leaflet. By tying the suture over a second Teflon felt pledget a monocuspid competent valve can be attained”
Carpentier Repair

- Detachment of antero-posterior leaflets
- Longitudinal plication
- Repositioning of leaflets
- Prosthetic ring

Alain F. Carpentier
*1933
B - Detachment of the anterior and posterior tricuspid valve leaflets and their chordal attachments to the ventricular wall.

C - Longitudinal plication of the atrialized portion of the right ventricle.

D - Clockwise spreadout of the anterior and posterior leaflets on the newly created tricuspid valve annulus and direct closure of the atrial septal defects without right atrial reduction.

No prosthetic ring
A New Procedure for Ebstein's Anomaly

Qingyu Wu, MD, and Zhixiong Huang, MD

Fig. 6. A large portion of the atretic ventricular wall is excised along the domed lines.

Fig. 7. The outer edge of the atretic ventricular wall is sutured.

Fig. 8. The tricuspid annuloplasty is completed, and the detached septal and posterior leaflets are reattached to a position just below the natural annulus.

Fig. 10. The whole procedure is completed.
Hetzer Technique Including Sebening stitch

Ebstein’s Anomaly
Repair after 3 weeks of support with Berlin Heart VAD

F. S. - 4 year-old girl
Ebstein’s Anomaly, ASD II, WPW syndrome, cardiac insufficiency and cyanosis

- Heart catheterization, SVT and resuscitation
- Interventional ASD closure
- Ventricular fibrillation, cardiac failure, BVAD
- Stabilization, explantation of BVAD after 3 weeks, Annuloraphy of Ebstein valve, BCPS
- Induction of A-V-Block grade III, Pacemaker
- Stabilized, discharged without neurologic findings
- Good condition at 5 yrs. follow-up
- Catheter ablation of verified accessory bundle
Cone Reconstruction

A - Opened right atrium showing displacement of the tricuspid valve.
B - Detached part of the anterior and posterior leaflet forming a single piece.
C - Clockwise rotation of the posterior leaflet edge to be sutured to the anterior leaflet septal edge and plication of the true tricuspid annulus.
D - Complete valve attachment to the true tricuspid annulus and valved closure of the atrial septal defect.

This technique was conceived bearing in mind Carpentier’s concepts of bringing the TV leaflets to the true tricuspid annulus level and longitudinal plication of the atrialized RV as necessary to restore the right ventricular volume and morphology in Ebstein’s anomaly repair.

da Silva JP, Arq Bras Cardiol 2004;82:212-216
Cone Reconstruction in Ebstein’s Anomaly

The most important aspects of a successful CR include leaflet tissue mobilization and incorporation of even a diminutive septal leaflet.

Anderson et al. Cong Heart Dis. 2014,9:266-71

Figure 1. Cone repair of Ebstein anomaly. The main principle of cone reconstruction includes complete surgical delamination of the tricuspid valve septal leaflet. The mobilized leaflet is then rotated and reattached at the true atrioventricular junction, followed by leaflet-to-leaflet attachment, creating a “leaflet cone.” Subsequently, the atrialized portion of the right ventricle is plicated and patent foramen ovale is repaired (if present).
Cone Reconstruction of the Tricuspid Valve Repair in Ebstein’s Anomaly

YouTube

http://www.youtube.com/watch?v=et4dUuvhyic
Mid-term Result of Cone Reconstruction

- **Da Silva (2012)** - 100 pts, 3 early and 1 late death, no newborns

- **Vogel (2012)** - 19 pts. Few children. Ringed annuloplasty in some patients. No difference from conventional technique (19 pts)

- **Lui (2011)** - 30 pts. Median age 60 mts. 20 pts. additionally received BCPS!

- **Dearani (2011)** - 89 pts. BCPS in 24% of patients, when significant right ventricular dilation and dysfunction were present. 13 reoperations. 6 TV replacements. Ringed annuloplasty (64%) was associated with less moderate TVR

- **Anderson (2014)** – 84 pts. (10±5,9yrs). Selected patients had BCPS. Follow-up 0.8±0.2yrs.) Mild/No TVR -83% of pts. Partial ring annuloplasty had positive effect on TV function

**Therefore, more long-term observations are necessary to determine superiority of cone reconstruction, particularly in children.**

**Cave – growth of tricuspid annulus is a concern!**
Neonatal Ebstein’s Anomaly
42 infants with Ebstein’s anomaly who were symptomatic at a mean age of 6 days.

Survival rates were 69% at 14 days of age, 52% at 1 year, and 37% at 5 years. Thirteen neonates (31%) died within 2 weeks of diagnosis, at a mean age of 5 days.


Survival up to 85 years was reported

Current Trends in the Management of Neonates With Ebstein’s Anomaly

Table 2. In-Hospital Mortality by Management Strategy

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Number</th>
<th>Deaths</th>
<th>Hospital Mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Overall</td>
<td>415</td>
<td>98</td>
<td>24%</td>
</tr>
<tr>
<td>Medical management</td>
<td>257</td>
<td>56</td>
<td>22%</td>
</tr>
<tr>
<td>Interventional cath</td>
<td>29</td>
<td>2</td>
<td>7%</td>
</tr>
<tr>
<td>Interventional cath + surgery</td>
<td>11</td>
<td>7</td>
<td>64%</td>
</tr>
<tr>
<td>Systemic-to-PA shunt</td>
<td>63</td>
<td>17</td>
<td>27%</td>
</tr>
<tr>
<td>TV repair</td>
<td>16</td>
<td>5</td>
<td>31%</td>
</tr>
<tr>
<td>Single-ventricle palliation</td>
<td>36</td>
<td>13</td>
<td>36%</td>
</tr>
<tr>
<td>(Starnes operation)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Transplantation</td>
<td>3</td>
<td>0</td>
<td>0%</td>
</tr>
</tbody>
</table>

Abbreviations: Cath, catheterization; TV, tricuspid valve; PA, pulmonary artery.

Goldberg et al. World Journal for Pediatric and Congenital Heart Surgery 2011;2:554-557
A total of 595 operations on 498 patients with Ebstein’s anomaly were included: 116 on neonates (19%), 122 on infants (21%), 264 on children (44%), and 93 on adults (16%).

Average annual institutional case volumes were low (median, 1 per year; range, 0 to 8 per year).

Neonates had a high rate of palliative procedures: systemic-to-pulmonary artery shunts with or without tricuspid valve closure (43; 37.1%) and tricuspid valve closure (10; 8.6%); Ebstein’s repair or tricuspid valvuloplasty -32%.

In-hospital mortality was high in neonatal patients (23.4%) in comparison with infants (4.1%), children (0.7%), and adults (1.1%).
Mortality in Infants is higher than the US mortality for the Norwood operation.

In a multicenter study from the European Congenital Heart Surgeons Association database, the mean operative mortality for any surgical intervention during the neonatal period was 53.8%.

Sarris et al. J Thorac Cardiovasc Surg 2006;132:50-57
The indications for operation were overt heart failure, cyanosis, and acidosis associated with tricuspid regurgitation, depressed right ventricular function, and severe cardiomegaly.


Starnes Operation versus Repair 1992-2005

16 pts. Mortality 31%;
Fenestrated 20%; Non-fenestrated 67%

9 pts. - BCPC; 3pts. - Fontan operation

Reemtsen et al. JTCVS 2006;132:1285-90
C. J. Knott-Craig Repair of the Neonatal Ebstein’s Anomaly Combination of Danielson and Sebennig Techniques and fenestrated ASD closure

(1) Creation of a competent monocuspid tricuspid valve, based on the anterior leaflet;
(2) Reduction atrioplasty;
(3) Partial closure of the atrial septal defect, leaving behind a small fenestration; and
(4) Simultaneous repair of other cardiac defects.

An important aspect of the repair is to ensure that severe tricuspid regurgitation does not recur during intermittent episodes of pulmonary hypertension. *The Sebening suture is very helpful in this regard.* A pledgetted 4/0 braided suture is placed through a dominant papillary muscle of the anterior leaflet and fixed to the interventricular septum, usually at the site of the laminated septal leaflet. This keeps the free wall of the right ventricle and the anterior leaflet in apposition to the posterior aspect of the tricuspid annulus.
Complete Repair of Ebstein’s Anomaly in Neonates and Young Infants 1994 - 2010

A 16-year follow-up

Single or multiple VSD 4
Anatomic PA 15
Functional PA 7

However, in situations where the anterior leaflet is deficient or where the posterior leaflet is the dominant leaflet, an alternative operative technique, such as the Starnes operation, should be considered.

Surgical Algorithm for Symptomatic Neonates with Ebstein’s Anomaly

Knott-Craig et al. Ann Thorac Surg
2007;84:587-93
1. A large part of the right ventricular free wall is resected. Resultant volume reduction of the right ventricle.

2. The coronary sinus is included in the right atrium.

3. The tricuspid valve orifice is closed, leaving the membranous interventricular septum above the patch to reduce the risk of heart block.

3 children with Ebstein’s anomaly, 5m, 15m and 5 years old.
Cone Reconstruction (CR) for Neonates with Ebstein’s Anomaly

- The role of CR in the neonate remains to be defined. It should be considered in select neonates when the valve anatomy is favorable (hemodynamically stable infants with adequate anterior and septal leaflet tissue).

- The monocusp techniques described by Knott-Craig and the single-ventricle pathway advocated by Starnes for when pulmonary atresia is present are reasonable alternatives in the unstable patient.  

  Anderson et al. Cong Heart Dis 2014;92:66-71

Less than 10 cases of cone repair were reported in infants

Sata-2012; Pizarro-2012; Anderson-2014
Cone Reconstruction in Operated Neonatal Ebstein's Anomaly with severe TV-Regurgitation

Before operation

After cone reconstruction
Non-Ebstein Tricuspid Valve Anomalies in Children

Isolated congenital tricuspid stenosis or regurgitation occurs even more rarely than Ebstein’s anomaly

- Primary annular dilation, prolapse, leaflet undevelopment
- Absent papillary muscle/chordae
- Tricuspid valve dysplasia
- Neonatal tricuspid valve rupture
- Tricuspid regurgitation in PA+IVS
- Tricuspid regurgitation in HLHS
- Tricuspid regurgitation after ToF, VSD repair
- The Gerbode defect
- Tricuspid regurgitation after pacemaker implantation

Uhl's disease is a rare disorder originally described in 1952 in an infant with severe diffuse right ventricular dysfunction with total absence of the myocardium.

Surgical techniques are few and have had variable success.

Uhl's Anomaly is not arrhythmogenic RV dysplasia!

Gerlis BHJ 1993;69:140-50

Sudden death is typical

Tabib Arch Mall Coeur 1992;69:142-50
Uhl’s Anomaly

Treatment strategies

1. Single ventricle strategy: Fontan circulation

2. One and a half ventricle repair combined with partial right ventriculectomy and TV closure + BCPS

   (Yoshii 2001; Azhari 2002; Hoschtilzky 2010)

3. Cardiac transplantation (Ikari et al. 2001)

N.B Long-term survival until adolescence and adulthood with medical therapy is reported (Bewik 1986; Freedom 2004; Hébert 2010)
# Uhl's Anomaly – Surgical Results Before 2002

**Ashari et al. Cardiol Young 2001;12.192-5**

<table>
<thead>
<tr>
<th>Author</th>
<th>Year of publication</th>
<th>Age</th>
<th>Sex</th>
<th>Clinical presentation</th>
<th>Additional anomalies</th>
<th>Operation</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kaul et al.²</td>
<td>1980</td>
<td>4 yrs</td>
<td>M</td>
<td>CHF</td>
<td>Rudimentary pulmonary valve</td>
<td>Glenn shunt</td>
<td>Post op death</td>
</tr>
<tr>
<td>Abe et al.³</td>
<td>1973</td>
<td>17 yrs</td>
<td>F</td>
<td>CHF</td>
<td>Dilated tricuspid annulus</td>
<td>Glenn shunt</td>
<td>Post op death</td>
</tr>
<tr>
<td>Hasegawa et al.⁴</td>
<td>1963</td>
<td>6 yrs</td>
<td>F</td>
<td>Cyanosis + CHF</td>
<td>POF</td>
<td>Glenn shunt</td>
<td>Post op death</td>
</tr>
<tr>
<td>Perrin &amp; Mehrizi¹</td>
<td>1965</td>
<td>6 yrs</td>
<td>M</td>
<td>Cyanosis + CHF</td>
<td>ASD</td>
<td>Potts</td>
<td>Died</td>
</tr>
<tr>
<td>Mitsui et al.³</td>
<td>1966</td>
<td>20 yrs</td>
<td>M</td>
<td>CHF</td>
<td></td>
<td>Glenn shunt</td>
<td>Died</td>
</tr>
<tr>
<td>Shojo et al.³</td>
<td>1968</td>
<td>14 yrs</td>
<td>M</td>
<td>Cyanosis + CHF</td>
<td>Tricuspid plication</td>
<td>Exploratory</td>
<td>Died</td>
</tr>
<tr>
<td>Kinare et al.⁴</td>
<td>1969</td>
<td>5 yrs</td>
<td>F</td>
<td>Cyanosis + CHF</td>
<td>Dilated tricuspid ring</td>
<td></td>
<td>Post op death</td>
</tr>
<tr>
<td>Zuberbühler et al.⁷</td>
<td>1970</td>
<td>17 mos</td>
<td>F</td>
<td>Cyanosis + CHF</td>
<td></td>
<td>Glenn shunt</td>
<td>Intra op death</td>
</tr>
<tr>
<td>Vecht et al.⁸</td>
<td>1979</td>
<td>19 yrs</td>
<td>M</td>
<td>Cyanosis + CHF</td>
<td>ASD</td>
<td>ASD closure</td>
<td>Post op death</td>
</tr>
<tr>
<td>Taussig⁸</td>
<td>1960</td>
<td>6 yrs</td>
<td>M</td>
<td>Cyanosis + CHF</td>
<td>ASD</td>
<td>Potts</td>
<td>Post op death</td>
</tr>
<tr>
<td>Cumming et al.⁸</td>
<td>1965</td>
<td>7 mos</td>
<td>F</td>
<td>Cyanosis + CHF</td>
<td>POF</td>
<td>Glenn shunt</td>
<td>Post op death</td>
</tr>
<tr>
<td>Froment et al.⁸</td>
<td>1968</td>
<td>15 yrs</td>
<td>M</td>
<td>CHF</td>
<td></td>
<td>Exploratory</td>
<td>Post op death</td>
</tr>
<tr>
<td>French et al.⁸</td>
<td>1975</td>
<td>14 yrs</td>
<td>F</td>
<td>Cyanosis + CHF</td>
<td>ASD, deformed tricuspid valve</td>
<td>ASD closure</td>
<td>Survived the operation</td>
</tr>
<tr>
<td>Child et al.⁹</td>
<td>1984</td>
<td>25 yrs</td>
<td>F</td>
<td>CHF</td>
<td></td>
<td>Anterior RV wall resection &amp; replacement Systemic-pulmonary anastomosis</td>
<td>Survived the operation</td>
</tr>
<tr>
<td>Corazza et al.¹⁰</td>
<td>1981</td>
<td>Newborn</td>
<td>M</td>
<td>Cyanosis</td>
<td>Pulmonary atresia, tricuspid hypoplasia, PAD</td>
<td>RV exclusion + Glenn shunt</td>
<td>Post op death</td>
</tr>
<tr>
<td>Present case</td>
<td>2001</td>
<td>5 mos</td>
<td>M</td>
<td>Cyanosis + TE Stroke</td>
<td>POI, tricuspid valve dilation</td>
<td></td>
<td>Alive</td>
</tr>
</tbody>
</table>

Abbreviations: CHF: congestive heart failure; ASD: atrial septal defect; RV: right ventricle; POP: patent oval foramen; TE Stroke: thromboembolic stroke; Post op: post operational; PAD: patent arterial duct

16 pts. 13 deaths – 81.2% mortality
Uhl's Anomaly – RV Exclusion Technique

9 months old; partial RV free wall resection + BCPC

Yoshii et al. JTCVS 2001;122.1026-8

9 months old; RV plication, PA disconnection, atrioseptectomy+BCPC

Hoschtitzky et al. 2010;90:2076-8
Tricuspid Valve in HLHS
The morphologically tricuspid valve in hypoplastic left heart syndrome

Christof Stamm, Robert H. Anderson, Siew Yen Ho *

Bileaflet tricuspid valve (TV) - 12%
Moderately dysplastic TV - 33%
Severely dysplastic TV - 2%

1. The subvalvular apparatus is different with mitral atresia.
2. Dysplasia of the leaflets occurs more often together with mitral stenosis.
3. The number of direct septal attachments was significantly higher in hearts with patent foramen ovale.
Among 74 Norwood survivors, 11 children (15%) with severe TR underwent TV repair at a median interval after the Norwood procedure of nine months with an overall mortality of 18%.

Four patients (36%) underwent reoperation for persistent TR.


Severe TR as a risk factor for mortality in children undergoing palliative procedures for HLHS can be diminished by successful TV procedures

The presence of mitral atresia and longer myocardial ischemia during Norwood operation as incremental risk factors (at the time of diagnosis) for tricuspid intervention

Elmi et al. J Thorac Cardiovasc Surg 2011;142: 1341-1347

Tricuspid Regurgitation in HLHS
Survival of Patients with HLHS Based on Degree of Preoperative Tricuspid Regurgitation.

These patients should be considered for early annuloplasty or tricuspid valve replacement should they have any evidence of hemodynamic dysfunction after their palliative operation.

*Berber et al. AHJ 1988;116:1563-7*
Surgical Repair of Dysplastic or Dysmorphic Tricuspid Valve in HLHS Hearts

A Direct cleft closure along with annuloplasty (functional commissuroplasty) on the antero-inferior commissure.

B Repair of the irregular dysplastic leaflets

C Repair of dysmorphic leaflets by means of the edge-to-edge type repair.

A. The anterior and septal leaflets are sutured together making two effective orifices

B. A con-coapting antero-septal commissure is totally obliterated in addition to the edge-to-edge suture.

Ando and Takahashi
Ann Thorac Surg
2007;84:1571-1576
Options for TV Repair in HLHS

1. Semicircular (Kay, De Vega) or ring anuloplasty

2. Papillary muscle sliding

3. Chordal looping

Best to perform second stage operation

TVR ≥ 2+

4. Papillary muscle head repositioning

5. Artificial chordae
A-V Valve in Single Ventricle Including HLHS

Valve replacement - high incidence of A-V block

Mahle et al. ATS 2001;72:181-6
Atrioventricular Valve Repair in Patients With Single-ventricle Physiology

Honjo et al., Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2011;14:75-84
Tricuspid Valve in Pulmonary Atresia + IVS
The presence of RV-to-coronary-artery connections or a TV Z-score < 2 should caution one against attempting biventricular repair.

**PA+IVS**

1. Heart with three zones present
2. Absence of trabecular portion
3. Absence of infundibular and trabecular portions

**TVR is not rare** because of dysplastic valve tissue

The presence of RV-to-coronary-artery connections or a TV Z-score < 2 should caution one against attempting biventricular repair.

- PA+IVS
- TVR
- Z-score
- Biventricular repair

**References**

- Liava’a et al. EJCTS 2011;40:1406-8
Tripartite TV Z-score > 2.5 – Biventricular repair after radiofrequency RVOT opening

Ebstein type anomaly, small RV, sinusoids
TV Z-score > 4.5 ➔ univentricular strategy

TV Z-score predicting univentricular or biventricular repair of PAIVS

Hanley et al. JTCVS 1993;105:406-23

The presence of RV-to-coronary-artery connections or a TV Z-score < 2 should caution one against attempting biventricular repair

Liava’a et al. EJCTS 2011;40:1406-8
PA IVS Before and After Radiofrequency Membrane Perforation
Repair of Congenital TVR with Artificial Chordae Tendinae (PA+IVS)

Technique of chordal replacement.

4-0 ePTFC suture

Technique of chordal augmentation.

2-0 ePTFC suture

The essence of straddling and overriding of the tricuspid valve is malalignment between the leading edge of the atrial septum and the crest of the muscular ventricular septum

**Valves usually straddle and override at the same time**
Valves usually straddle and override at the same time.
Straddling of A-V Valves

Right ventricular hypoplasia frequently associated with anomalies of the AV valves such as overriding or straddling

- D-TGA with VSD - 19 patients
- Straddling of the TV - 13 patients
- Straddling of the MV - 6 Patients

Surgical Technique for Intraoperative Management of Straddling Tricuspid Valve in TGA.

The chordae or the papillary muscle crossing the VSD into the left ventricular cavity is retracted toward the right ventricle by a hook passed through the native aortic valve. The VSD is closed through the pulmonary valve.

“Curtainlike abnormal tricuspid chordae remain a contraindication to biventricular repair in DORV or TGA”

Tailoring of the myocardial flap in the presence of abnormal attachments of the tricuspid valve on the outlet septum.

Occasionally, abnormalities of the TV or anomalous origin of papillary muscles around the margin of VSD make construction of a satisfactory intracardiac baffle difficult or impossible.

Options:
- TV replacement
- Fontan operation

Becker AE, Anderson RH Cardiac pathology, 1983
Rastelli Correction Versus Fontan Operation

- 34 patients with DORV or D-TGAS with straddling AV valves
- 4 patients were switched to univentricular repair
- 4 early deaths after biventricular repair (13.3%)
- 6 reoperations during follow-up because of subaortic stenosis, MVI, TVI, residual VSD + pacemaker

"The outcome for Rastelli's, in the best-case scenario, is era-independent and 60% survival at 20 years.

I would submit, as a rhetorical question, our Fontan survivals at 20 years are probably about 85% or 90%. Should we, as a society, be exploring doing Fontans on these patients instead of pursuing a biventricular repair?"
Neonatal in Utero Tricuspid Valve Rupture

- * Artificial chordae
- ** Reimplantation of the papillary muscle
- * Annuloplasty (Kay, adjustable De Vega-type)

Causes: Ischemia, Infarction?
No valve displacement

* Anagnostopoulos et al. Ann Thorac Surg 2007;83;1462
** Jaques and Imamura Sem TCVS 2007; 19:258-63
Atrio-Ventricular Rhabdomyoma in 3 day-old Newborn

Emergency Starnes OP, permanent pacemaker. BCPS at 6 months of age.
Pacemaker Lead Induced TVR

Perforation

Said et al. JTCVS 2014; 147:412-19
Thank You