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
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

Decrease flow through the right heart

effective pulmonary blood flow

shunt direction through ASD/PFO
Ebstein’s anomaly

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

PG E dependent

Failure to wean

- cyanosis
  - Shunt alone
- Cyanosis + CHF
  - Shunt+TV procedure
- CHF
  - TV procedure

Successful wean

No treatment

CHF

Single ventricle (RV exclusion)

1 ½ (BDG+TVR)

TV repair/replacement

RV exclusion

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).

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

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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 shunts 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.

Ebstein’s anomaly (EA) represents a downward, or apical displacement of the posterior and septal leaflets of the tricuspid valve, with a resultant redundant “restrictal” 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 sail-like leaflets and utilization of the sail-like anterior leaflets 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 embolization on a single-ventricle pathway is the more reasonable approach...
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.
Three additional neonates responded well to nitric oxide and prolonged anesthesia, and were ultimately weaned from the ventilator and discharged without surgical intervention.

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 bioprostheses 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.
Neonates with Ebstein’s

Antegrade Pulm. Flow

2-VENTRICLE REPAIR

Good RV

2-VENTRICLE REPAIR

Small RV

B-T Shunt vs. Starnes Repair

Anatomic Pulm. Atresia

Small LV

TRANSPLANT

Stable

2-VENTRICLE REPAIR

Unstable

Starnes Repair & Ligate MPA

Functional Pulm. Atresia

Good RV

Starnes Repair & Ligate MPA

Comment

Since 1994 we have operated on young infants, and have survival rates that are similar to those seen in single-ventricle palliation [4]. Feasible, reasonable approach a two-ventricle system will require a longer follow-up. Our results are similar to those of others with single-ventricle palliation. A critical issue is whether or not the right ventricle can be repaired, and if it cannot, the situation is dire.
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 patients – 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.
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 + \(\downarrow\) 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
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 \( \frac{1}{2} \) repair (46 pat) (50% - TV replacement; 50% - TV repair) – 26 %
Other – 14%
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.