Surgical Techniques for the repair of Ebsteins’ Anomaly

University of Ulsan, College of Medicine
Asan Medical Center

Yun, Tae-Jin
Surgical algorithm (AMC)

Ebstein’s anomaly

- Sizable, Freely mobile AL
  - Functional aRV
    - TVP Without aRV Plication ± BCPS
  - Dysfunctional aRV
    - TVP With aRV Plication ± BCPS

- Small, Restrictive AL
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- Tricuspid Sac anomaly
  - Fontan Op + Tricuspid sac Obliteration
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Tricuspid Sac anomaly
- Fontan Op + Tricuspid sac Obliteration
Typical scenario

- M / 45, Carpentier type B
- Functional class III
- Op finding: Hugely dilated RA
  - Large, thin-walled aRV
  - Small functional RV

- Operation
  - TVP (cone procedure)
    - High CVP, RV dysfunction, severe TR on Echo
    - Addition of TAP without success
    - TVR with bioprosthesis

- Rocky postoperative course with RV dysfunction
Ebstein anomaly: Myths

- TV repair is technically feasible in all Ebstein anomaly patients!

- Surgical outcome of each surgical technique is excellent with minimal residual TR!

- Excellent early surgical outcome can lead to excellent long term outcome!
Surgical outcome of Ebstein Anomaly
(Chen et al, JTCVS 2004)

- Children’s hospital of New York
- Age: 2 m – 46.8 yrs (median: 8.2 yrs)
- Carpentier technique
- Residual moderate to severe TR: 13/25(52%)
- Reoperation (3) : TVR(2), Addition of BCPS (1)
- Late death: 2 patients with severe residual TR
Repair of Ebstein anomaly is tricky!

- Wide spectrum of disease
- Difficult to define morphology
- Limited experience
- No standardized surgical strategy
- Various surgical techniques
Ebstein Anomaly as a Spectrum

C. type

A  B  C  D
Ebstein Anomaly as a Spectrum

C. type

A  B  C  D

Manifestation

Late  Early
Ebstein Anomaly as a Spectrum

C. type
A B C D

Manifestation
Late
Early

Surgical Tx.
TVP TVR Fontan
Ebstein Anomaly as a Spectrum

C. type
A  B  C  D

Manifestation
Late  Early

Surgical Tx.
TVP  TVR  Fontan

Outcome
Risk of residual TR
Risk of RV dysfunction
Delineation of TV leaflets

1. Commissures in systole
2. Papillary muscles

⇒ Difficult in Ebstein Anomaly
Ebstein anomaly:

Delamination failure of TV leaflets

→ Downward displacement of valve mechanism
Degree of Delamination failure
Degree of Delamination failure
Degree of Delamination failure
Effective valve orifice in 23 specimens
(Schreiber et al, JTCVS 1999)
Tricuspid Regurgitation?

A: f RV → aRV
B: aRV → RA
Carpentier Classification
(Carpenter, 1988)
Celermajer classification
(Celermajer, 1992)

Area ratio \((AR) = \frac{RA \text{ area} + aRV \text{ area}}{f \text{ RV area} + LV \text{ area} + LA \text{ area}}\)

Grade I : \(AR < 0.5\)
Grade II : \(0.5 \leq AR \leq 0.99\)
Grade III : \(1.0 \leq AR \leq 1.49\)
Grade IV : \(1.5 \leq AR\)
Celermajer index changes!

Postnatal day 0
Area idex = 1.79

Postnatal day 3
Area idex = 1.5

Postnatal day 21
Area idex = 0.96

Postnatal 6 m (post 1 ½ repair)
TV repair techniques

- Danielson technique
- Carpentier technique
- Cone procedure
- ‘Wu’ technique
- Hetzer technique
Danielson Repair

![Diagram showing surgical repair process]
Danielson Repair
Danielson Repair
## Operative series of Ebstein Anomaly

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* Clinical reports from the ‘Hospital Broussais  † Clinical reports from ‘Mayo Clinic’
Carpentier repair (1)
Carpentier repair (2)
Carpentier repair (3)
Carpentier repair (4)
Advantages of vertical plication of aRV

Transverse (Horizontal) Plication

Vertical (Longitudinal) Plication
Limitations of Carpentier technique

1. Tethering of AP leaflets
2. Tricuspid sac anomaly
Cone procedure
Wu technique (Wu et al, ATS 2004)
Wu technique
(Wu et al, ATS 2004)

Ebstein’s anomaly

Giant RA

RCA
Hetzer repair

- No plication of aRV
- Incorporation of aRV to f RV
- Allowance of multiple trial and error
- Drawbacks
  1. Requires well developed AL
  2. Applicable to mild disease
  3. Risk of heart block
Hetzer repair
(Hetzer et al, JTCVS1998)
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Hetzer repair
-Sebening stitch-
21-year-old man with Ebstein’s anomaly

- Pre-op chest PA
- Functional class III
- Severe TR: TR jet area 25 cm²
- Association of Atrial flutter
aRV function after Hetzer repair

M/21, Ebstein anomaly (A)
Pre-operative
aRV function after Hetzer repair

M/21, Ebstein anomaly (A)
Immediate Post-operative
aRV function after Hetzer repair

M/21, Ebstein anomaly (A)
Post-operative 14 months
21-year-old man with Ebstein’s anomaly
aRV function after Hetzer repair

F/25, Ebstein anomaly (B)

Pre-operative
aRV function after Hetzer repair

F/25, Ebstein anomaly (B)

Immediate postoperative
AMC experience of Hetzer repair

- Original Hetzer technique
- Modified Hetzer technique
Effective valve orifice in 23 specimens (Schreiber et al, JTCVS 1999)
AMC experience of Hetzer Technique

- Duration: 2002-2013
- Patient No.: 27
- Male / Female: 7 / 20
- Age at operation: 1 m – 57 y (median: 29 y)
- Carpentier type: A (7), B (13), C (3), unknown (4)
- Pre-op desaturation (<95%): 7/27
- Associated lesion
  - AF-Af (7), AVNRT (1), WPW (1), ASD (9), RPA atresia (1)
AMC experience of Hetzer Technique

- Hetzer technique: Original (6), Modified (21)
- Associated Procedure
  - BCS (23), Arrhythmia Op (8), ASD closure (9)
- CPB / ACC: 119 min / 54 min
- aRV plication: 1/27
- aRV Obliteration: 2/27
- RA reduction: 11/27
- Reoperation: 1/27
AMC experience of Hetzer Technique

- ICU stay / Hospital stay: 2.6 day / 12.6 day
- No early or late death
- F/U: 1 m – 124.8 m (median: 55 m)
- NYHA functional class ≤ II: 22/27
- TR on Immediate post-op TTE
  - < II (18/27)
  - III (6/27)
  - IV (3/27): TR jet area 46 cm$^2$ → 22 cm$^2$
CT ratio on Chest x-ray

Post op months

pre op  0  1month  6month  12month  24month
Freedom of significant TR or re-operation

96%
92%

Postoperative months
Surgical algorithm (AMC)

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Tvr in Ebstein anomaly

- 20 – 80% of entire cohort

- Excellent long term outcome of porcine bioprosthetic valve (Mayo Clinic)

- Issues in surgical technique:
  1) above coronary sinus vs. under coronary sinus
  2) Combined procedure: plication of aRV
Ebstein Anomaly as a Spectrum

C. type

A
B
C
D

Surgical Tx.

TVP

TVP

TVR

Fontan

Outcome

Risk of residual TR

Risk of RV dysfunction
Overly aggressive TVP

C. type

A  B  C  D

Surgical Tx.

TVP  TVR

Outcome

Risk of residual TR

Risk of RV dysfunction
Over-reliance on TVR

C. type

A  B  C  D

Surgical Tx.

TVP  TVR  Fontan

Risk of residual TR

Risk of RV dysfunction

Outcome
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TVR with Bioprosthesis
(Mayo clinic, 1998)
TVR with Bioprosthesis in young age
(Mayo clinic, 1998)
TVR in Ebstein anomaly
1 ½ repair in Ebstein anomaly
- Theoretical advantages -

- Increased capability of valve repair
- Decreased early and late mortality
- Decreased risk of re-operation
- Better functional class
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1 ½ repair in Ebstein Anomaly

C. type
- A
- B
- C
- D

I. Surgery Without BCPS
- TVP
- TVR
- Fontan

II. Surgery With BCPS
- TVP
- TVR
- Fontan

BCPS
1 ½ repair in Ebstein Anomaly

C. type
A B C D

I. Surgery Without BCPS
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Risk of Residual TR

I II
### 1 ½ repair in Ebstein Anomaly

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#### I. Surgery Without BCPS

- TVP
- TVR
- Fontan

#### II. Surgery With BCPS

- TVP
- TVR
- Fontan

#### Risk of RV dysfunction

- I
- II
One and a Half Ventricle Repair in Adults: Postoperative Hemodynamic Assessment Using Phase-Contrast Magnetic Resonance Imaging

Jin Woo Chung, MD, Hyun Woo Goo, MD, Yu-Mi Im, MS Hong-Ju Shin, MD, Won Kyoung Jhang, MD, Jae-Kon Ko, MD, and Tae-Jin Yun, MD, PhD

Division of Pediatric Cardiac Surgery, Department of Radiology, and Division of Pediatric Cardiology, Asan Medical Center, University of Ulsan, Seoul, Republic of Korea

Background. One and a half ventricle repair (1½ repair) strategy has been used for patients with a hypoplastic or dysfunctional right ventricle (RV), or both. We sought to assess the postoperative hemodynamics of 1½ repair using phase-contrast magnetic resonance imaging (PC-MRI).

Methods. Ten adults, 9 with Ebstein’s anomaly and 1 with tricuspid stenosis, underwent 1½ repair (median age at operation, 42.4 years). The ayzygos vein was left open in all patients on 1½ repair to prevent severe postoperative central venous hypertension. Postoperative PC-MRI studies were performed to measure blood flow to the ascending aorta (Q_{Asc-Ao}), the main pulmonary artery (Q_{MPA}), the superior vena cava (SVC) (Q_{SVC}), and the branch pulmonary arteries and veins. From these values, blood flow to the upper compartment of the body (Q_{UC}), right ventricular volume unloading effect (Q_{MPA}/Q_{Asc-Ao}), proportion of blood flow to the upper compartment of the body (Q_{UC}/Q_{Asc-Ao}), and venous return to arterial forward flow ratio of the upper compartment of the body (Q_{SVC}/Q_{UC}) were calculated. Two patients also underwent preoperative PC-MRI.

Results. On PC-MRI, Q_{MPA}/Q_{Asc-Ao}, Q_{UC}/Q_{Asc-Ao}, and Q_{SVC}/Q_{UC} were 0.58 to 0.84 (median, 0.67), 0.19 to 0.36 (median, 0.27), and 0.47 to 1.57 (median, 0.93, lower than 1.0 in 7 patients), respectively. In 2 patients who had preoperative and postoperative PC-MRI, Q_{UC}/Q_{Asc-Ao} decreased from 0.26, 0.32 to 0.21, 0.28, respectively.

Conclusions. After 1½ repair, right ventricular volume unloading was effective in all patients, but intercaval collateral veins (i.e., Q_{SVC}/Q_{UC} < 1) appeared to develop in most of the patients. Furthermore blood flow to the upper compartment of the body appeared to diminish, presumably due to postoperative elevation of central venous pressure.

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1) RV volume unloading
   \[ \frac{Q_{MPA}}{Q_{a-Ao}} \]

2) Collateral flow to the lungs
   \[ Q_{Coll} = Q_{L-Coll} + Q_{R-Coll} \]
   \[ Q_{L-Coll} = Q_{LPA} - Q_{LPV} \]
   \[ Q_{R-Coll} = Q_{RPA} - Q_{RPV} \]

3) Flow to the upper body
   \[ Q_{UC} = Q_{a-Ao} - Q_{Coll} - Q_{d-Ao} \]

4) SVC-IVC Collateral flow
   \[ Q_{SVC-IVC} = Q_{UC} - Q_{SVC} \]
Table 3. Comparison of preoperative and postoperative PC-MRI data in patient 9 and 10 (cc/beat)

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PC-MRI: phase-contrast velocity mapping magnetic resonance imaging; Pre-op: preoperative; Post-op: postoperative 3 months in patient 9 and 2 months in patient 10; $Q_{asc-Ao}$: ascending aortic blood flow; $Q_{desc-Ao}$: descending aortic blood flow; $Q_{coll}$: aortopulmonary collateral blood flow; $Q_{UC}$: blood flow to the upper compartment of the body; $Q_{MPA}$: blood flow of the main pulmonary artery.
Ebstein’s anomaly

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- Tricuspid Sac anomaly
  - Fontan Op + Tricuspid sac Obliteration

Surgical algorithm (AMC)
Fontan op in Ebstein anomaly

• 10% of entire cohort

• Role of Fontan operation in old age: ?

• Issues in surgical technique:
  1) Extracardiac Fontan
  2) Tricuspid sac obliteration
Ebstein Anomaly as a Spectrum

C. type
A B C D

Manifestation
Late Early

Surgical Tx.
TVP TVR Fontan

Risk of residual TR

Outcome
Ebstein patients who needs Fontan: 10%
1. Neonatal stenotic Ebstein
2. High mortality of Carpentier group
Techniques of RV exclusion

Patch closure of TV annulus (Starnes Op)

Suture obliteration of aRV (Yun et al, JTCVS 2006)
Summary

- Fontan track: 10% of entire cohort
- TVR for Ebstein’s anomaly is a risky operation, especially in patients with severe disease.
- Hetzer procedure is technically feasible, and leads to excellent longterm outcome
- Addition of BCS is beneficial upon TVP/TVR.