What to Do for a 75 Year Old Man of Severe Ebstein Anomaly with Decreased RV Function, Atrial Fibrillation

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The authors have no financial conflicts of interest to disclose concerning the presentation.
Ebstein Anomaly
Ebstein’s Anomaly

Severe Ebstein’s Anomaly

Displacement of Septal Leaflet

Shiina A, JACC 1984:356-70
Ebstein Anomaly
Ebstein Anomaly
Ebstein Anomaly
Ebstein Anomaly
Ebstein Anomaly
Ebstein Anomaly
Ebstein Anomaly
Anatomic Assessment

Tricuspid valve leaflets

- Apical displacement of septal/posterior leaflet (> 8 mm/m² or 20 mm in adults)
- Tethering, elongation, absence of the leaflets
- Leaflet fenestrations (anterior leaflet)
- Accessory leaflet tissue (muscular shelf)
  - Between septal and anterior leaflets
- Annulus dilatation
Anatomic Assessment

omite Right ventricle

- Atrialized right ventricle
- Right ventricular dilatation
  - Between annulus and posterior leaflet
- Posterior aneurysm/RVOT aneurysm
  - 20%

mite Left ventricle

- Size and shape of the left ventricle
- Mitral valve prolapse
  - Elongated chordae
Functional Assessment

Tricuspid valve

- Tricuspid regurgitation or stenosis
- Pressure gradient across the tricuspid valve
- Severity of tricuspid regurgitation
- Right ventricular inflow tract obstruction
Functional Assessment

- **Right ventricle**
  - Progression of chamber enlargement
  - Right ventricular function

- **Left ventricle**
  - Left ventricular function
  - Mitral regurgitation (severity)
Ebstein Anomaly
Ebstein Anomaly
Ebstein Anomaly

FR 12Hz
13cm

Full Volume
3D 52%
3D 50dB
CF
50%
2.7MHz

Delay 0ms

JPEG
64 bpm
Ebstein Anomaly
Severe Ebstein Anomaly
Carpentier Classification

The volume of the true RV is adequate
A large atrialized component of the RV exists, but the anterior leaflet of the TV moves freely

A

The anterior leaflet is severely restricted in its movement and may cause significant obstruction of the RVOT
Almost complete atrialization of the RV except for a small infundibular component

B

C

D
Ebstein Anomaly
Ebstein Anomaly

FR 58Hz
12cm

2D
55%
C 48
P Low
HGen

JPEG
59 bpm
Aneurysmal Dilatation of RVOT

Shiina A, JACC 1984:356-70
Anatomy & Functional Disability

Shiina A, JACC 1984:356-70

<table>
<thead>
<tr>
<th>2-D echo features</th>
<th>Index Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Septal leaflet</td>
<td>1</td>
</tr>
<tr>
<td>displacement ≥ 25 mm/m²</td>
<td>1</td>
</tr>
<tr>
<td>Anterior leaflet</td>
<td>1</td>
</tr>
<tr>
<td>displacement of free edge</td>
<td>1</td>
</tr>
<tr>
<td>restricted motion</td>
<td>1</td>
</tr>
<tr>
<td>severe prolapse</td>
<td>1</td>
</tr>
<tr>
<td>Intracardiac cavities</td>
<td>1</td>
</tr>
<tr>
<td>aneurysmal RVOT</td>
<td>1</td>
</tr>
<tr>
<td>aRV/RV ≥ 50%</td>
<td>1</td>
</tr>
<tr>
<td>tricuspid anulus ≥ 45 mm/m²</td>
<td>1</td>
</tr>
<tr>
<td>right atrium ≥ 80 mm/m²</td>
<td>1</td>
</tr>
</tbody>
</table>

TOTAL 10

Severity Index

0 1 2 3 4 5 6 7

mild (I) moderate (II) severe (III-IV)

p=0.017

p=0.007

Ratio between atrialized right ventricle and whole right ventricle

Functional capacity (NYHA)

100
90
80
70
60
50
40
30
20
10
0

Shiina A, JACC 1984:356-70
Surgery vs. Observation
50 YO / M

- Alleged CHD, DOE Fc I
50 YO / M

FR 39Hz
19cm

2D
75%
C 50
P Low
HPen

JPEG
70 bpm
50 YO / M
Associated anomalies

- Patent foramen ovale/atrial septal defect
- RV inflow and outflow tract obstruction
- VSD
- Accessory conduction pathway (WPW syndrome), increasing risk of atrial tachycardia
- PS
- TOF
- CoA
- Mitral valve abnormalities
Surgeons with training and expertise in CHD should perform tricuspid valve repair or replacement with concomitant closure of an ASD, when present, for patients with Ebstein’s anomaly with the following indications:

- **Symptoms** or deteriorating exercise capacity. *(Level of Evidence: B)*
- **Cyanosis** (oxygen saturation less than 90%). *(Level of Evidence: B)*
- **Paradoxical embolism.** *(Level of Evidence: B)*
- Progressive **cardiomegaly** on chest x-ray. *(Level of Evidence: B)*
- Progressive **RV** dilation or reduction of RV systolic function. *(Level of Evidence: B)*

*Circulation. 2008;118:2395-2451*
## Surgical Intervention

<table>
<thead>
<tr>
<th>Indications</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Indications for surgery</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Surgical repair should be performed in patients with more than moderate TR and symptoms (NYHA class &gt;II or arrhythmias) or deteriorating exercise capacity measured by CPET</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>• If there is also an indication for tricuspid valve surgery, then ASD/PFO closure should be performed surgically at the time of valve repair</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>• Surgical repair should be considered regardless of symptoms in patients with progressive right heart dilation or reduction of RV systolic function and/or progressive cardiomegaly on chest X-ray</td>
<td>IIa</td>
<td>C</td>
</tr>
</tbody>
</table>

EHJ 2010;31:2915–2957
Surgery vs. Observation

Decreased RV function
Surgery vs. Observation

Decreased RV function

75 years old
Natural Course in Exercise Capacity

< 18 years old

≥ 18 years old

Table I. Demographics

<table>
<thead>
<tr>
<th>Characteristic</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Male gender, n (%)</td>
<td>13 (57)</td>
</tr>
<tr>
<td>Age at first CPX (y), median (range)</td>
<td>17.9 (8.1-52.5)</td>
</tr>
<tr>
<td>Time from first to last CPX (y), median (range)</td>
<td>3.3 (0.6-7.3)</td>
</tr>
<tr>
<td>No. of CPX</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>12 (52%)</td>
</tr>
<tr>
<td>3</td>
<td>6 (26%)</td>
</tr>
<tr>
<td>4</td>
<td>3 (13%)</td>
</tr>
<tr>
<td>5</td>
<td>2 (9%)</td>
</tr>
<tr>
<td>Mean ± SD</td>
<td>2.8 ± 1.0</td>
</tr>
<tr>
<td>Severe tricuspid regurgitation*</td>
<td>9 (39%)</td>
</tr>
<tr>
<td>Atrial level right to left shunting †</td>
<td>7 (30%)</td>
</tr>
<tr>
<td>ES grade(^1,10)</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>7 (32%)</td>
</tr>
<tr>
<td>2</td>
<td>10 (45%)</td>
</tr>
<tr>
<td>3</td>
<td>4 (18%)</td>
</tr>
<tr>
<td>4</td>
<td>1 (5%)</td>
</tr>
</tbody>
</table>

N = 23 patients; 64 exercise tests.  
* Based on echocardiogram within 6 months of the first exercise test.  
† Oxygen saturation at rest or with exercise ≤93%.

Am Heart J 2012;163:486-91
Natural Course in Exercise Capacity

Table V. Average rate of change per year for $\text{VO}_2\%$ and $\text{O}_2\text{ pulse}\%$

<table>
<thead>
<tr>
<th></th>
<th>$\text{VO}_2%$: change/y</th>
<th></th>
<th>$\text{O}_2\text{ pulse}%$: change/y</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean ± SD</td>
<td>$P^*$</td>
<td>$P^\dagger$</td>
</tr>
<tr>
<td>Overall (n = 23)</td>
<td>$-1.87 \pm 8.04$</td>
<td>.05</td>
<td></td>
</tr>
<tr>
<td>Age at initial CPX</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age &lt;18 y (n = 12)</td>
<td>$-3.04 \pm 6.78$</td>
<td>.01</td>
<td>.15</td>
</tr>
<tr>
<td>Age ≥18 y (n = 11)</td>
<td>$-0.43 \pm 8.79$</td>
<td>.77</td>
<td></td>
</tr>
<tr>
<td>Tricuspid regurgitation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mild or moderate (n = 14)</td>
<td>$-1.59 \pm 8.85$</td>
<td>.18</td>
<td>.57</td>
</tr>
<tr>
<td>Severe (n = 9)</td>
<td>$-2.67 \pm 6.92$</td>
<td>.11</td>
<td></td>
</tr>
<tr>
<td>ES grade</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 (n = 7)</td>
<td>$-1.91 \pm 9.87$</td>
<td>.36</td>
<td>.82</td>
</tr>
<tr>
<td>2 (n = 10)</td>
<td>$-3.43 \pm 7.94$</td>
<td>.04</td>
<td></td>
</tr>
<tr>
<td>3 or 4 (n = 5)</td>
<td>$0.02 \pm 4.23$</td>
<td>.99</td>
<td></td>
</tr>
</tbody>
</table>

Overall and by age, severity of tricuspid regurgitation, and ES grade. Other abbreviations as in Table III.

* $P$ value from 1 sample $t$ test comparing mean slope to 0.

† $P$ value from 2 sample $t$ test comparing mean slopes in the groups.

Am Heart J 2012;163:486-91
21 patients with Ebstein anomaly (between 6 and 59 years of age)

<table>
<thead>
<tr>
<th>TABLE 1. Changes in CPET: Comparing Baseline Testing with Follow-up After Surgical Intervention</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td><strong>Sex</strong></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td><strong>Age</strong></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td><strong>Tricuspid regurgitation</strong></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td><strong>RER</strong></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td><strong>SpO₂† (%)</strong></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td><strong>Peak Vo₂</strong></td>
</tr>
<tr>
<td><strong>Ve/Vco₂</strong></td>
</tr>
<tr>
<td></td>
</tr>
</tbody>
</table>

Baseline test  | Follow-up  | P values* |
---------------|------------|-----------|
5/16           | 5/16       |           |
24.5 (13.7 – 43.6) | 26.7 (14.8 – 44.9) |           |
0 / 2 / 16 / 3 | 9 / 10 / 2 / 0 | < .001    |
1.07 (1.03 – 1.13) | 1.07 (1.03 – 1.13) | .501      |
98 (95 – 99)    | 99 (97 – 100)  | .004      |
92 (84 – 98)    | 97 (95 – 100)  | .010      |
21.0 (17.6 – 23.5) | 20.7 (18.8 – 27.9) | .009      |
68.4 (52.2 – 83.3) | 77.3 (57.2 – 91.4) | .009      |
32.5 (30.3 – 43.8) | 29.3 (27.8 – 33.1) | .001      |

CPET, Cardiopulmonary exercise test; RER, respiratory exchange ratio. *2-sided Wilcoxon test. †SpO₂ could be assessed in only 20 patients.
21 patients with Ebstein anomaly (between 6 and 59 years of age)

**TABLE 2. Changes in CPET variables during follow-up by patients undergoing primary surgery and patients being reoperated**

<table>
<thead>
<tr>
<th></th>
<th>Primary surgery (group 1)</th>
<th>Reoperation (group 2)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n = 14, Median (Q1-Q3)</td>
<td>n = 7, Median (Q1-Q3)</td>
</tr>
<tr>
<td>Sex</td>
<td>3/11</td>
<td>2/5</td>
</tr>
<tr>
<td>Age</td>
<td>18.3 (10.8–46.3)</td>
<td>31.1 (21.1–41.3)</td>
</tr>
<tr>
<td>TR</td>
<td>I/II/III/IV</td>
<td></td>
</tr>
<tr>
<td>Peak</td>
<td>1.04 (1.01–1.09)</td>
<td>1.12 (1.08–1.23)</td>
</tr>
<tr>
<td>Rest</td>
<td>95 (92–98)</td>
<td>99 (98–100)</td>
</tr>
<tr>
<td>Peak</td>
<td>88 (78–94)</td>
<td>99 (98–100)</td>
</tr>
<tr>
<td>peak V̇O₂ (mL/min/kg)</td>
<td>21.0 (17.8–22.1)</td>
<td>20.3 (16.4–24.1)</td>
</tr>
<tr>
<td>% predicted</td>
<td>68.8 (51.0–83.0)</td>
<td>56.4 (53.1–86.7)</td>
</tr>
<tr>
<td>V̇E/V̇CO₂ Slope (Slope)</td>
<td>32.3 (30.8–50.5)</td>
<td>32.5 (30.0–36)</td>
</tr>
</tbody>
</table>

*CPET, Cardiopulmonary exercise test; RER, respiratory exchange ratio; TR, tricuspid regurgitation. *2-sided Wilcoxon test. †Chi-squared test comparing the sex distribution of the 2 subgroups. ‡2-sided Mann-Whitney-U test comparing the age of the 2 subgroups at baseline test. §SpO₂ saturation could be assessed in only 20 patients.
Functional Status after Surgery

Functional Status After Operation for Ebstein Anomaly

The Mayo Clinic Experience

Morgan L. Brown, MD,* Joseph A. Dearani, MD, FACC,* Gordon K. Danielson, MD, FACC,* Frank Cetta, MD, FACC,†‡ Heidi M. Connolly, MD, FACC,‡ Carole A. Warnes, MD, FACC,†‡ Zhuo Li, MS,§ David O. Hodge, MS,§ David J. Driscoll, MD, FACC†

Rochester, Minnesota

Objectives The objective of this study was to review the long-term functional outcome of patients with Ebstein anomaly who had cardiac operation at our institution.

Background Ebstein anomaly is a spectrum of tricuspid valvular and right ventricular dysplasia. Many patients will require operation in an attempt to improve quality of life.

Methods From April 1, 1972, to January 1, 2006, 539 patients with Ebstein anomaly underwent 604 cardiac operations at the Mayo Clinic in Rochester, Minnesota. Patient records were reviewed, and all patients known to still be alive were mailed a medical questionnaire or contacted by telephone.

Results At the initial operation at our institution, the mean age of the patients was 24 years (range 8 days to 79 years) and 53% were female patients. Survival at 5, 10, 15, and 20 years was 94%, 90%, 86%, and 76%, respectively. Survival free of late reoperation was 86%, 74%, 62%, and 46% at 5, 10, 15, and 20 years, respectively. Surveys were returned by 285 of 448 (64%) patients known to be alive at the time of this study. Two hundred thirty-seven (83%) patients were in New York Heart Association functional class I or II, and 34% were taking no cardiac medication. One hundred three patients (36%) reported an incident of atrial fibrillation or flutter, 5 patients (2%) reported having had endocarditis, and 1 patient (<1%) reported having a stroke. There were 275 pregnancies among 82 women. The recurrence of congenital heart disease was reported in 9 of 232 (3.9%) liveborn children.

Conclusions Patients have good long-term survival and functional outcomes after undergoing surgery for Ebstein anomaly. Atrial arrhythmias are common both before and after surgery. Many patients have had one or more successful pregnancies with a low-recurrence risk of congenital heart disease. (J Am Coll Cardiol 2008;52:460–6) © 2008 by the American College of Cardiology Foundation
Functional Status after Surgery

Health Status Questionnaires
NYHA functional class
- I (43%)
- II (40%)
- III (12%)
- IV (4%)

J Am Coll Cardiol 2008;52:460–6
Functional Status after Surgery

Figure 3
Self-Reported Activity Scale After Operation for Ebstein Anomaly

Table 3
Self-Reported Exercise Tolerance

<table>
<thead>
<tr>
<th>My Ability of Exercise Relative to Peers Is</th>
<th>n = 285</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Much greater</td>
<td>7</td>
<td>2.5</td>
</tr>
<tr>
<td>Slightly greater</td>
<td>29</td>
<td>10.2</td>
</tr>
<tr>
<td>About the same</td>
<td>105</td>
<td>36.8</td>
</tr>
<tr>
<td>Slightly less</td>
<td>89</td>
<td>31.2</td>
</tr>
<tr>
<td>Much less</td>
<td>43</td>
<td>15.1</td>
</tr>
<tr>
<td>I am unable to exercise</td>
<td>5</td>
<td>1.8</td>
</tr>
<tr>
<td>Unknown</td>
<td>7</td>
<td>2.5</td>
</tr>
</tbody>
</table>
539 Patients with mean [range] age at surgery 24 years [8 days to 79 years]

Early mortality: 4.8%

Survival after Surgery

JTCVS 2008;135:1120-36
Reoperation after Surgery

A

Survival free of late reoperation (%)

- TV repair <12
- TV replacement <12

P<0.001

TV repair <12
44
TV replacement <12
105
27
20
39
61

Years
0 5 10 15

B

Survival free of late reoperation (%)

- TV repair ≥12
- TV replacement ≥12

P=0.77

TV repair ≥12
129
TV replacement ≥12
217
90
63
39

0 5 10 15

Late survival free of reoperation (%)

0 20 40 60 80 100

Years
508
307
166
91
37

JTCVS 2008;135:1120-36
11 patients were > 60 years of age

Table 1  Clinical Characteristics

Patient characteristics (n = 81)
- Age at diagnosis, yrs: 43 ± 19
- Women: 51 (63)

Pre-procedural characteristics (n = 89)
- Patient age at surgery, yrs: 59 ± 8
- New York Heart Association functional class
  - I/II: 13 (15)
  - III/IV: 76 (85)
- Dyspnea: 74 (83)
- Palpitations: 69 (78)
- Edema: 30 (34)
- Cyanosis: 21 (24)
- Stroke/TIA: 21 (24)
- Dizziness: 16 (18)
- Right-sided heart failure: 13 (15)
- Syncope: 7 (8)
- Clubbing: 6 (7)

Values are mean ± SD or n (%).
TIA = transient ischemic attack.

Table 2  Surgical Procedures Performed (N = 89)

<table>
<thead>
<tr>
<th>Procedure Type</th>
<th>No. of Procedures (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tricuspid valve operation</td>
<td></td>
</tr>
<tr>
<td>Repair</td>
<td>22 (25)</td>
</tr>
<tr>
<td>Replacement</td>
<td>65 (73)</td>
</tr>
<tr>
<td>None</td>
<td>2 (2)</td>
</tr>
<tr>
<td>ASD/PFO closure</td>
<td>58 (65)</td>
</tr>
<tr>
<td>Right reduction atrioplasty</td>
<td>41 (46)</td>
</tr>
<tr>
<td>Anterior right pericardectomy</td>
<td>18 (20)</td>
</tr>
<tr>
<td>Plication of atrialized RV</td>
<td>12 (13)</td>
</tr>
<tr>
<td>Right-sided maze procedure</td>
<td>18 (20)</td>
</tr>
<tr>
<td>Ablation of accessory pathway</td>
<td>9 (10)</td>
</tr>
<tr>
<td>CABG</td>
<td>9 (10)</td>
</tr>
<tr>
<td>Mitral valve surgery</td>
<td></td>
</tr>
<tr>
<td>Repair</td>
<td>6 (7)</td>
</tr>
<tr>
<td>Replacement</td>
<td>4 (5)</td>
</tr>
<tr>
<td>None</td>
<td>2 (2)</td>
</tr>
<tr>
<td>Bidirectional Glenn operation</td>
<td>2 (2)</td>
</tr>
<tr>
<td>Aortic root surgery</td>
<td>2 (2)</td>
</tr>
<tr>
<td>Repair of PS</td>
<td>1 (1)</td>
</tr>
<tr>
<td>Permanent pacing</td>
<td>1 (1)</td>
</tr>
</tbody>
</table>

ASD = atrial septal defect; CABG = coronary artery bypass grafting; PFO = patent foramen ovale; PS = pulmonary stenosis; RV = right ventricle.

J Am Coll Cardiol 2012;59:2101–6
For the 71 patients with clinical follow-up, improvement in functional class occurred in 63 patients (89%). Postoperatively, only 8 patients remained in functional class III or IV (11%).

*J Am Coll Cardiol* 2012;59:2101–6
## Table 4

Univariate Predictors of Death During Follow-up

<table>
<thead>
<tr>
<th>Parameter</th>
<th>HR (95% CI)</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>No post-operative improvement</td>
<td>10.0 (3.01–33.4)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Pre-operative history of heart failure</td>
<td>4.42 (1.42–13.7)</td>
<td>0.01</td>
</tr>
<tr>
<td>Pre-operative LVEF &lt;50%</td>
<td>3.59 (1.20–10.7)</td>
<td>0.02</td>
</tr>
<tr>
<td>Diabetes mellitus</td>
<td>6.76 (1.43–31.9)</td>
<td>0.02</td>
</tr>
<tr>
<td>Pulmonary hypertension</td>
<td>1.21 (0.36–4.08)</td>
<td>0.75</td>
</tr>
<tr>
<td>History of atrial fibrillation</td>
<td>0.95 (0.26–3.46)</td>
<td>0.94</td>
</tr>
<tr>
<td>Tricuspid valve replacement</td>
<td>1.63 (0.44–5.99)</td>
<td>0.46</td>
</tr>
<tr>
<td>Male</td>
<td>1.96 (0.65–5.89)</td>
<td>0.23</td>
</tr>
<tr>
<td>Age at surgery, per 10 yrs</td>
<td>2.53 (1.28–5.00)</td>
<td>0.007</td>
</tr>
</tbody>
</table>

Cl = confidence interval; HR = hazard ratio; LVEF = left ventricular ejection fraction.

**J Am Coll Cardiol 2012;59:2101–6**
Surgery + Maze operation
Vs.
Catheter ablation
Catheter Intervention

**Class I**
- Adults with Ebstein’s anomaly should have catheterization performed at centers with expertise in catheterization and management of such patients. (*Level of Evidence: C*)

**Class IIa**
- Catheter ablation can be beneficial for treatment of recurrent supraventricular tachycardia in *some patients* with Ebstein’s anomaly. (*Level of Evidence: B*)

*Circulation. 2008;118:2395-2451*
# Catheter Intervention

## Indications for catheter intervention

- Patients with relevant arrhythmias should undergo electrophysiologic testing, followed by ablation therapy, if feasible, or surgical treatment of the arrhythmias in the case of planned heart surgery.  

- In the case of documented systemic embolism probably caused by paradoxical embolism, isolated device closure of ASD/PFO should be considered.

- If cyanosis (oxygen saturation at rest <90%) is the leading problem, isolated device closure of ASD/PFO may be considered but requires careful evaluation before intervention (see text).

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*EHJ 2010;31:2915–2957*
Accessory Pathway Mediated SVT

- Localization of accessory pathways is often challenging & > 50% of patients have multiple accessory pathways.

- The success rate for catheter ablation is lower in Ebstein anomaly (≤81%) patients compared with patients with structurally normal hearts (≥95%).

- Rarely, right coronary artery stenosis has been seen following catheter ablation.
Ebstein Surgery & Maze

- APMT (n = 49)
  - No Sg for AP (n = 4)
  - RFA* (n = 6)
  - Combined† (n = 11)
  - Surgical division (n = 26)
  - Surgical cryoablation (n = 4)

- AVNRT (n = 10)
  - No Sg for AVNRT (n = 3)
  - Perinodal cryoablation (n = 7)
  - Combined† (n = 11)

- AFI/F (n = 70)
  - No Sg for AFI/F (n = 22)
  - Right-sided maze (n = 38)
  - Cryoablation of atrial isthmus (n = 10)

References:

JTCVS 2004;128:826-33
Circulation. 2015;131:1110-1118
Ebstein Surgery & Maze

Recurrence of atrial fibrillation

- 6/14 (43.0%) without Maze op
- 12/48 (25.6%) with Maze or cryoablation

\[ \text{JTCVS 2004;128:826-33} \]
\[ \text{Circulation. 2015;131:1110-1118} \]
86 patients who had corrective surgery and concomitant maze procedure

Table 5. Late Outcomes According to Type of Preoperative Arrhythmia

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Paroxysmal</th>
<th>Persistent</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>No AFL/F + no AAM</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RSM</td>
<td>88%</td>
<td>71%</td>
<td>0.17</td>
</tr>
<tr>
<td>BAM</td>
<td>77%</td>
<td>86%</td>
<td>0.08</td>
</tr>
<tr>
<td>Freedom from warfarin</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RSM</td>
<td>83%</td>
<td>86%</td>
<td>0.87</td>
</tr>
<tr>
<td>BAM</td>
<td>75%</td>
<td>71%</td>
<td>0.86</td>
</tr>
</tbody>
</table>

* Probability value for column comparison.

AAM = antiarrhythmic medications; AFL/F = atrial flutter or fibrillation; BAM = bialtrial maze; RSM = right-sided maze.

The absence of LAMF after the Cox maze III procedure is as high as 39% and has been demonstrated to persist up to 56 months.

Circulation. 2015;131:1110-1118
What to Do for a 75 Year Old Man of Severe Ebstein Anomaly with Decreased RV Function, Atrial Fibrillation

Surgery + Maze operation Anticoagulation

Earlier surgery should have been considered.
Thank you for your attention.