Electrophysiology and Surgery Intertwined in Complex Treatment of Ebstein’s Anomaly in Childhood

short title: Electrophysiology and Surgery Intertwined in the Treatment of Ebstein’s Anomaly

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Introduction

Ebstein’s anomaly, a rare and highly variable congenital heart defect (1), still presents a treatment challenge. The currently used cone repair of the tricuspid valve has carried favorable results in suitable patients (2). Arrhythmogenic substrates including accessory pathways (3,4) and right bundle branch block (5) associated with electromechanical ventricular dyssynchrony present additional therapeutic targets. We present a patient with Ebstein’s anomaly of tricuspid valve and Wolff-Parkinson-White syndrome in whom joint electrophysiologic and surgical interventions were used to address all major disease components. This report is unique in highlighting a combination of different treatment modalities for managing severe Ebstein’s anomaly, including catheter pathway mapping and ablation, surgery with cryoablation and right ventricular cardiac resynchronization therapy with optimal results.

Case report

A 4.5 years old girl with Ebstein’s anomaly of the tricuspid valve and Wolff-Parkinson-White syndrome (WPW) has suffered from recurrent poorly tolerated paroxysms of orthodromic atrioventricular reentrant tachycardia despite treatment with Sotalol. Echocardiography showed a type C Ebstein’s anomaly of the tricuspid valve with 3rd degree tricuspid regurgitation and no shunting on the atrial level (Supplemental material, Video 1). Her resting ECG revealed a sinus and low right atrial rhythm with ventricular preexcitation compatible with a right posteroseptal to posterior manifest accessory pathway (Figure 1A). During electrophysiologic study using the EnSite Precision™ 3D cardiac mapping system an orthodromic atrioventricular reentrant tachycardia with right bundle branch block QRS morphology was easily inducible by single atrial extrastimuli (Figure 1B). Initially, successful cryoablation of a right posteroseptal manifest accessory pathway was performed resulting in a change of the preexcitation pattern and separation of local atrial and ventricular electrograms (Figure 1C, D and E). Another right posterior manifest pathway could not be ablated successfully (Figure 1F and G). The distance of approximately 10 mm between both mapping
sites (Figure 1G) is in favor of the presence of two distinct pathways rather than one broad pathway, although this could not be definitely proven. Cryoenergy was used to limit the risk of right coronary artery damage reported during radiofrequency catheter ablation in Ebstein’s anomaly (6) due to the presence of thin ventricular wall at the anatomical tricuspid annulus.

On the subsequent day surgical cryoablation of the remaining accessory pathway followed by the cone repair of tricuspid valve was performed. No further intraoperative mapping was performed. A cryolesion at the anatomical tricuspid annulus extending from the lower rim of the coronary sinus ostium to the inferolateral annulus was applied to cover both mapped pathways. Bidirectional block was proven by atrial and ventricular pacing using the Medtronic CareLink™ 2090 Programmer/Analyzer connected to the temporary atrial and ventricular pacing wires placed on the right atrium and ventricle. Antegrade and retrograde block was seen during adenosine administration. Transesophageal echocardiography showed favorable tricuspid valve function along with severe right ventricular mechanical dyssynchrony attributed the right bundle branch block demasked by pathway interruption (7,8) (Supplemental material, Video 2). Acute right heart failure prevented discontinuation of cardio-pulmonary bypass. Permanent right ventricular cardiac resynchronization therapy (RV-CRT) was implemented by implanting a dual-chamber pacemaker with an epicardial right atrial and right ventricular free wall lead, respectively, programmed to DDD mode with AV delay optimized to achieve complete fusion of the paced and spontaneous ventricular activation and narrow pseudo-normalized QRS complex without signs of right bundle branch block (Figure 2A) (9). This led to major acute improvement of hemodynamics enabling smooth discontinuation of cardio-pulmonary bypass. Coronary artery injury or spasm potentially associated with cryoablation thus seemed unlikely to be the cause of acute post-repair right ventricular dysfunction and coronary angiography was not performed. The patient was extubated 24 hours after surgery and discharged 7 days later.
Postoperative transthoracic echocardiography showed excellent surgical result with synchronous right ventricular contraction and trivial tricuspid valve regurgitation (Supplemental material, Video 3). Immediate resynchronization effect was tested on the 14th postoperative day by switching RV-CRT on and off showing narrow QRS complex and lasting acute improvement of multiple hemodynamic parameters including RV contraction synchrony and efficiency as reflected by decreased RV systolic stretch fraction (10) (Table 1, Figure 2B and C).

During further follow-up of 6 months the girl was asymptomatic with New York Heart Association class 1 functional status, no SVT recurrences and good RV function on continuing RV-CRT.

**Conclusion**

Selected patients with Ebstein’s anomaly of the tricuspid valve will benefit from a complex therapy approach including treatment of the arrhythmogenic substrates and surgical tricuspid valve plasty. Due to difficulties with pathway mapping and ablation typical for the Ebstein’s anomaly and high recurrence rate (11,12,13,14), surgical cryodestruction associated with tricuspid valve repair may be an appealing option if catheter ablation fails. Ablation of manifest right-sided accessory pathway may demask right bundle branch block with subsequent right ventricular electromechanical dyssynchrony amenable to RV-CRT – a so far only anecdotally reported treatment option in Ebstein’s anomaly (9). A complex electrophysiological and surgical therapy approach enables to address all major facets of the disease.
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**Table 1**: Improvement of hemodynamic parameters during acute RV-CRT testing.

<table>
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<tr>
<th>Parameter</th>
<th>RV-CRT off</th>
<th>RV-CRT on</th>
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<tr>
<td>RV filling time [ms]</td>
<td>272</td>
<td>305</td>
</tr>
<tr>
<td>RV maximum +dP/dt [mmHg/s]</td>
<td>233</td>
<td>449</td>
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<tr>
<td>PA velocity time integral [cm]</td>
<td>15.1</td>
<td>16.7</td>
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<td>RV septal to free wall mechanical delay [ms]</td>
<td>97</td>
<td>13</td>
</tr>
<tr>
<td>Pulm. valve closure to peak RV free wall contraction [ms]</td>
<td>57</td>
<td>20</td>
</tr>
<tr>
<td>RV systolic stretch fraction</td>
<td>0.37</td>
<td>0.07</td>
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PA = pulmonary artery, Pulm. = pulmonary, RV = right ventricular, RV-CRT = right ventricular cardiac resynchronization therapy
Figure legends

**Figure 1:** Electrophysiologic study. A. 12-lead ECG prior to ablation. B. Orthodromic atrioventricular reentrant tachycardia with right bundle branch block QRS morphology and QRS complex duration of 120 ms. C. Local electrograms at successful ablation site of the right posteroseptal accessory pathway from the cryo-catheter (FRZ dist and uni) with ventricular signal coinciding with delta wave onset. The strictly negative ventricular signal from the ablation catheter tip (FRZ uni) is in favor of correct placement at the ventricular insertion site of the posteroseptal pathway. D. Local electrograms after posteroseptal pathway ablation (FRZ dist) with clear division of the atrial and ventricular electrogram at the ablation site. E. 12-lead ECG showing different QRS morphology after posteroseptal pathway ablation with a delta wave evident especially in the lead I and V4. F. Local electrograms at the site of the second unsuccessfully ablated right posterior pathway (FRZ dist) with ventricular signal clearly preceding delta wave onset. G. 3D electroanatomical map of the earliest ventricular activation on the tricuspid annulus (delineated by the black line) during sinus rhythm. First manifest pathway could be successfully ablated in the right posteroseptal region (green arrow). The second manifest pathway could be mapped to the posterior tricuspid annulus (blue arrow) and could not be ablated. The distance between the two mapping sites was estimated to be 10 mm (white line) and is in favor of two distinct pathways rather than one broad pathway, although this cannot be definitely proven. FRZ dist, distal bipolar signal from the ablation catheter; FRZ uni, unipolar signal from the ablation catheter tip; CS, signals from the coronary sinus catheter from proximal to distal. Vertical lines depict delta wave onset.

**Figure 2:** A. 12-lead ECG after RV-CRT showing narrow QRS complex (=70 ms) with absence of right bundle branch block. B. Longitudinal strain imaging of the right ventricle during baseline rhythm with right bundle branch block. Significant RV mechanical dyssynchrony is evident with early septal contraction (green arrow) accompanied by RV free wall pre-stretch (yellow arrow) followed by late free wall contraction (red arrow) and septal rebound stretch (orange arrow). Peak basal RV free wall
contraction occurs 57 ms after pulmonary valve closure (PVC - green line) thus not contributing to RV ejection. RV septal to free wall mechanical delay is 97 ms. C. During RV-CRT RV dyssynchrony is abolished with synchronous contraction of the septum and RV free wall. Peak basal RV free contraction occurs almost simultaneously with pulmonary valve closure. RV contraction efficiency is restored.
Figure 2C
Supplemental material: Echocardiographic images. Video 1. Before catheter ablation and surgery. Apical 4-chamber view showing severe Ebstein malformation of the tricuspid valve. Right ventricle is on the left and left ventricle on the right of the picture. Video 2. Intraoperative transesophageal echocardiography after cone repair and right posterior accessory pathway ablation showing RV mechanical discoordination. Right ventricle is on the bottom right side, right atrium on the top left side. Video 3. Apical 4-chamber view 12 days after cone repair and right ventricular cardiac resynchronization therapy displaying the repaired tricuspid valve and synchronous right ventricular contraction. Right ventricle is on the left and left ventricle on the right of the picture.
Key Teaching Points

- Ebstein’s anomaly is a rare and highly variable congenital heart defect presenting with different degrees of severity. The currently used cone repair of the tricuspid valve has carried favorable results in suitable patients.
- Accessory atrioventricular pathways causing atrioventricular reentrant tachycardia are frequent and present an additional therapeutic target.
- Due to difficulties with pathway mapping and ablation typical for the Ebstein’s anomaly and high recurrence rate, surgical cryodestruction associated with tricuspid valve repair may be an appealing option if catheter ablation fails.
- Manifest right-sided accessory pathway ablation may demask right bundle branch block with subsequent right ventricular electromechanical discoordination amenable to right ventricular cardiac resynchronization therapy.