Electrophysiology and surgery intertwined in complex treatment of Ebstein's anomaly in childhood

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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 electrophysiological 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 (RV-CRT), with optimal results.

Case report

A 4.5-year-old girl with Ebstein's anomaly of the tricuspid valve and Wolff-Parkinson-White syndrome 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 third-degree tricuspid regurgitation and no shunting on the atrial level (Supplemental Video 1). Her resting electrocardiogram revealed a sinus and low right atrial rhythm with ventricular preexcitation compatible with a right posteroseptal-to-posterior manifest accessory pathway (Figure 1A). During electrophysiological 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–1E). Another right posterior manifest pathway could not be ablated successfully (Figure 1F and 1G). The distance of approximately 10 mm between both mapping sites (Figure 1G) is in favor of the presence of 2 distinct pathways rather than 1 broad pathway,

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.
- Owing 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.

KEYWORDS Ebstein's anomaly; Wolff-Parkinson-White syndrome; Cone repair; Cryoablation; Right ventricular cardiac resynchronization therapy; Accessory pathway ablation; Congenital heart disease
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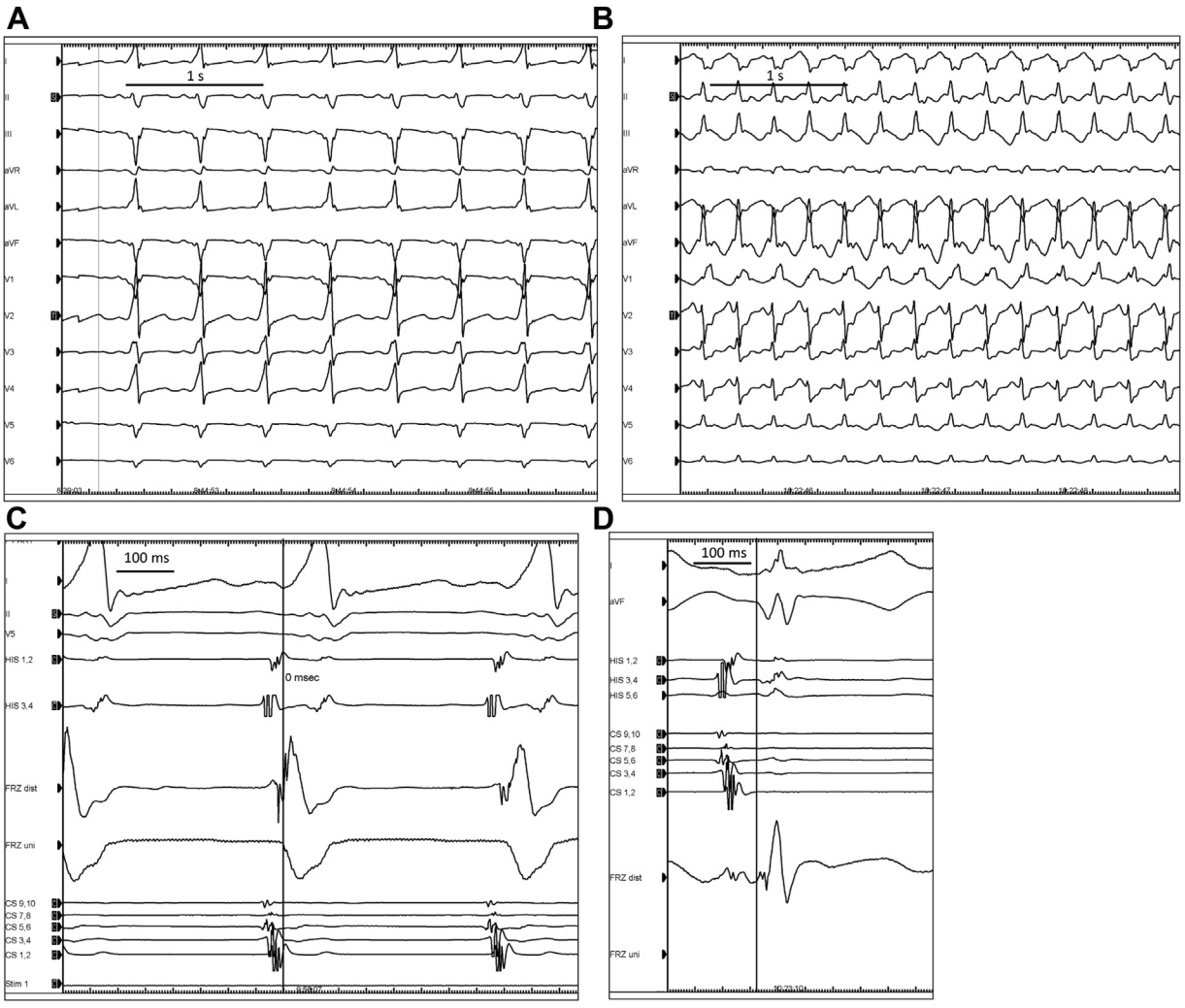

Figure 1 Electrophysiological study. **A:** Twelve-lead electrocardiogram (ECG) prior to ablation. **B:** Orthodromic atrioventricular reentrant tachycardia with right bundle branch block QRS morphology and QRS complex duration of 120 ms. **C:** Local electrograms at successful ablation site of the right 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:** Twelve-lead ECG showing different QRS morphology after posteroseptal pathway ablation with a delta wave evident especially in leads I and V_4. **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:** Three-dimensional 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 2 mapping sites was estimated to be 10 mm (*white line*) and is in favor of 2 distinct pathways rather than 1 broad pathway, although this cannot be definitely proven. FRZ dist indicates 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.

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] owing 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 RV mechanical dyssynchrony attributed to the right bundle branch block demasked by pathway interruption[7,8] (Supplemental Video 2). Acute right heart failure prevented discontinuation of cardiopulmonary bypass. Permanent RV-CRT was implemented by implanting a dual-chamber pacemaker with an epicardial right atrial and

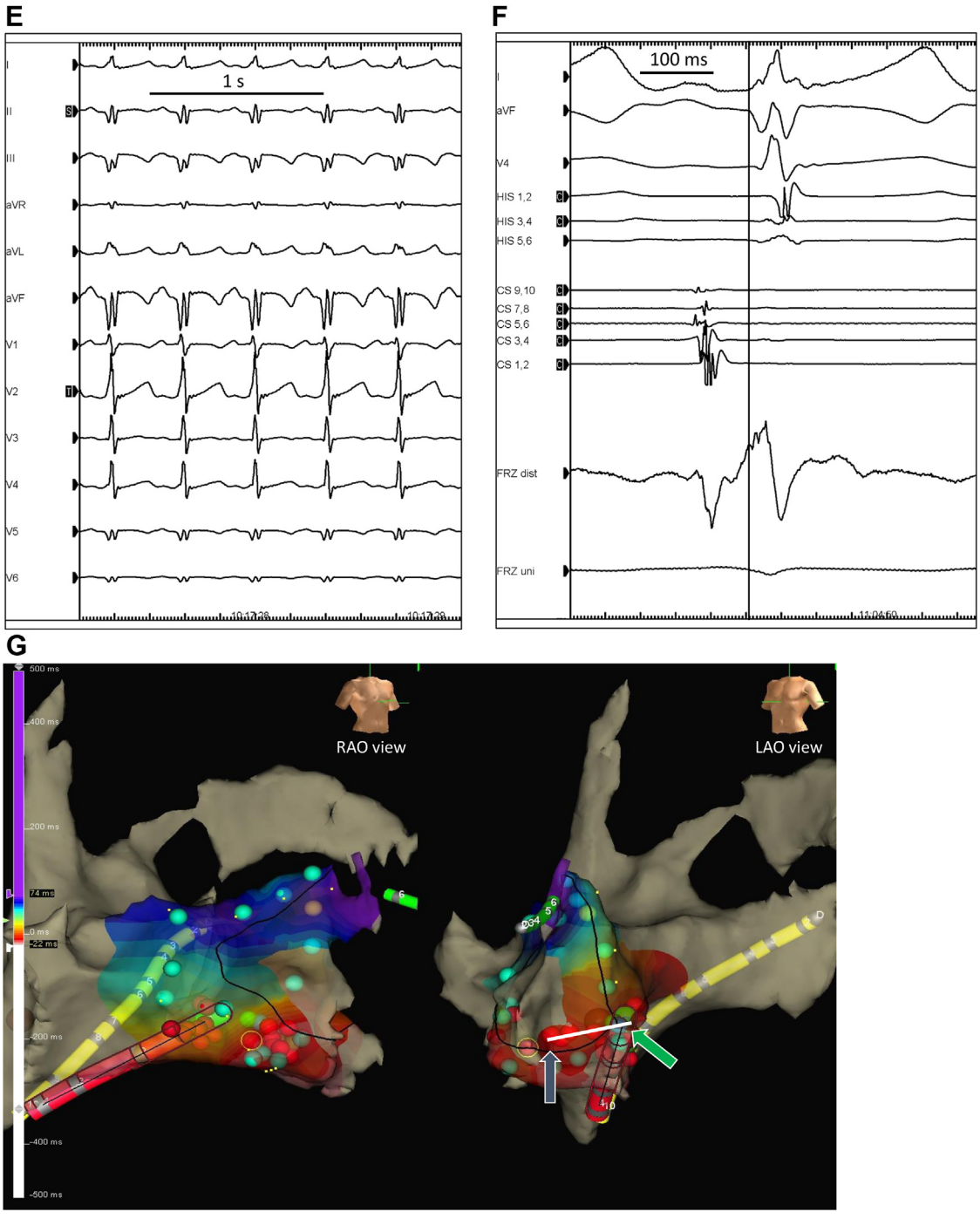

Figure 1 Continued

RV 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 cardiopulmonary bypass. Coronary artery injury or spasm potentially associated with cryoablation thus seemed unlikely to be the cause of acute postrepair RV 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 RV contraction and trivial tricuspid valve regurgitation (Supplemental 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

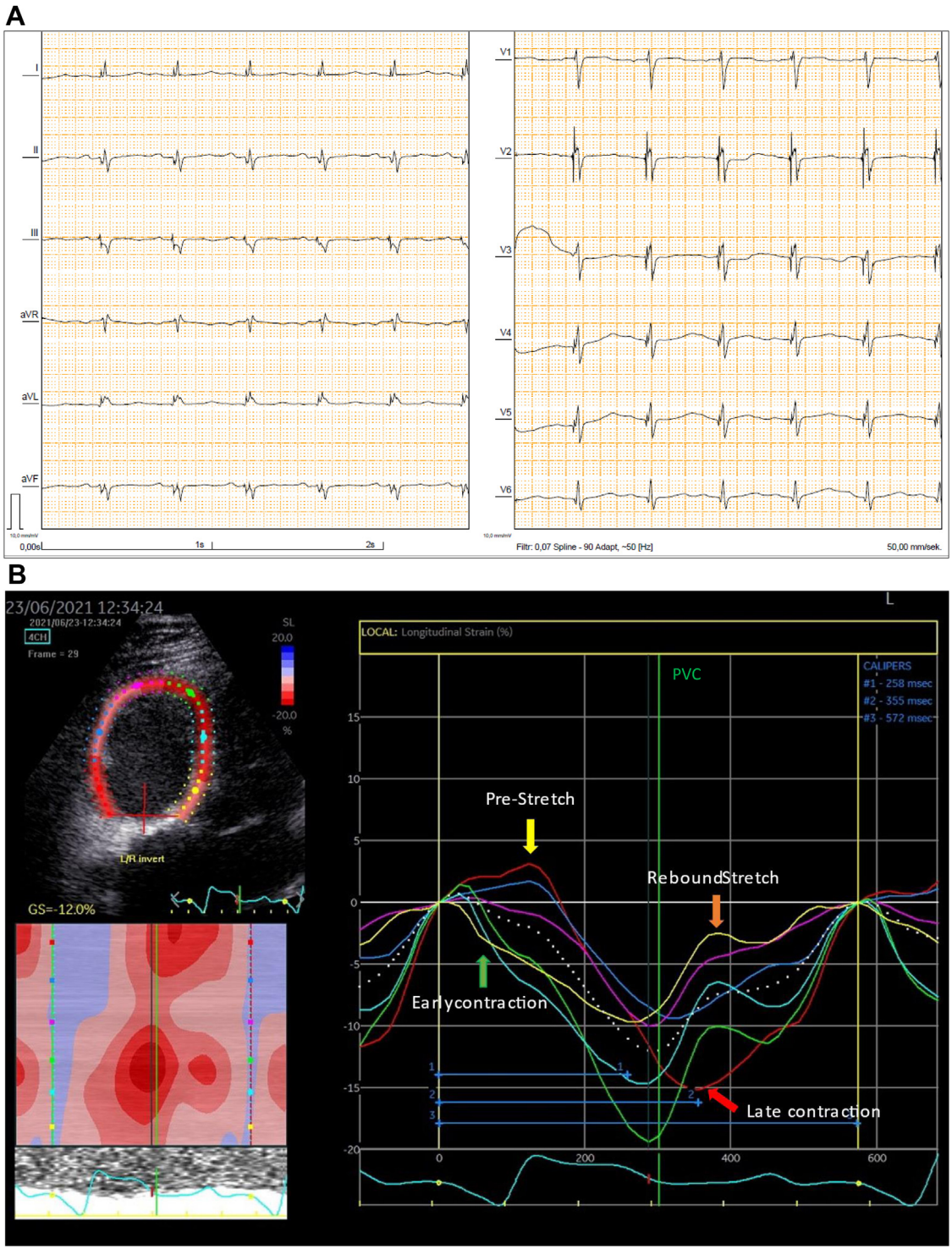

Figure 2 **A:** Twelve-lead electrocardiogram after right ventricular cardiac resynchronization therapy (RV-CRT) showing narrow QRS complex (=70 ms) with absence of right bundle branch block. **B:** Longitudinal strain imaging of the right ventricle during baseline rhythm with right bundle branch block. 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.

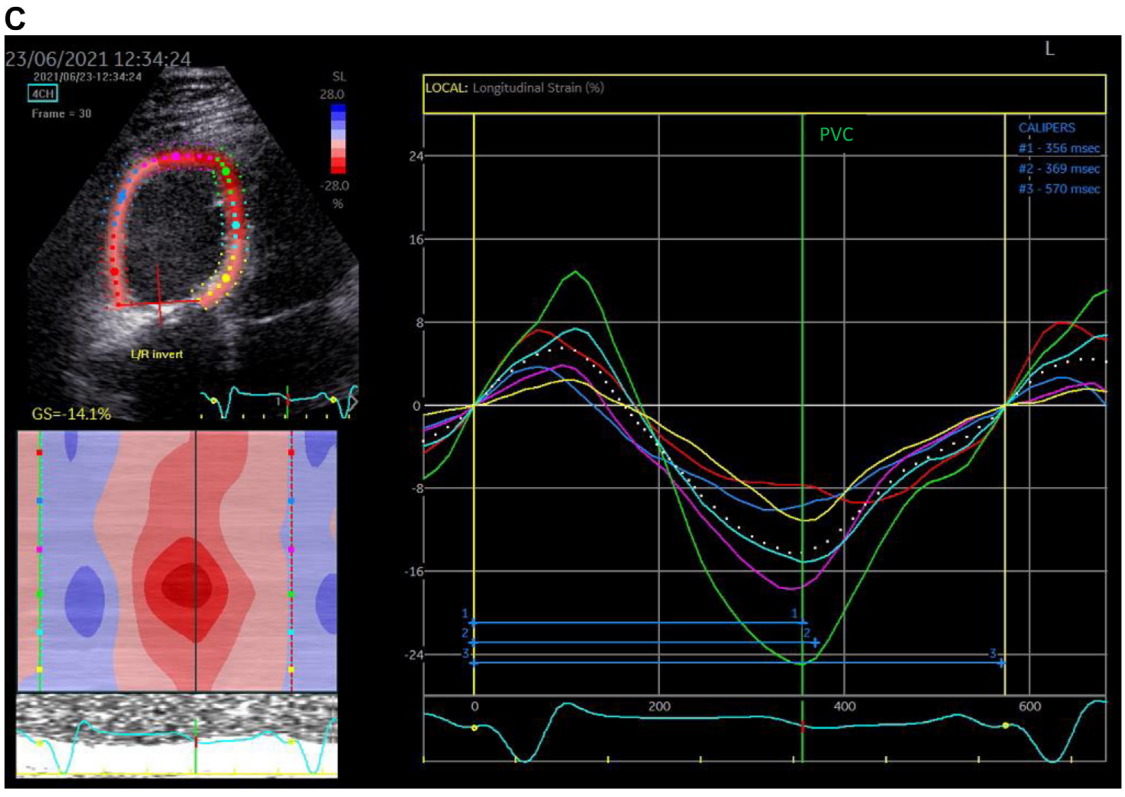

Figure 2 Continued

reflected by decreased RV systolic stretch fraction[10] (Table 1, Figure 2B and 2C).

During further follow-up of 6 months, the girl was asymptomatic, with New York Heart Association class 1 functional status, no supraventricular tachycardia 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. Owing to difficulties with pathway mapping and ablation typical for the Ebstein's anomaly and high recurrence rate,[11–14] 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 RV 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.

Table 1 Improvement of hemodynamic parameters during acute right ventricular cardiac resynchronization therapy testing

	RV-CRT off	RV-CRT on
RV filling time [ms] rowhead	272	305
RV maximum +dP/dt [mm Hg/s] rowhead	233	449
PA velocity time integral [cm] rowhead	15.1	16.7
RV septal-to–free wall mechanical delay [ms] rowhead	97	13
Pulm valve closure to peak RV free wall contraction [ms] rowhead	57	20
RV systolic stretch fraction rowhead	0.37	0.07

PA = pulmonary artery; Pulm = pulmonary; RV = right ventricular; RV-CRT = right ventricular cardiac resynchronization therapy.

Appendix

Supplementary data

Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2022.09.014.

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