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Pre-excitation induced ventricular dysfunction and successful berlin heart explantation after accessory pathway ablation

Utkarsh Kohli, M.D. [a], Karen L. Pumphrey, BSN [b], Aziez Ahmed, M.D. [c], Srikant Das, M.D. [d,*]

[a] Division of Pediatric Cardiology, University of Chicago School of Medicine, Chicago, IL, USA
[b] Arkansas Children's Hospital, Little Rock, AR, USA
[c] Arkansas Children's Hospital, University of Arkansas for Medical Sciences, Little Rock, AR, USA
[d] Department of Pediatrics, Pediatric Electrophysiology, University of Arkansas for Medical Sciences, Little Rock, AR, USA

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ABSTRACT

A 13 kg, 20 month-old, Caucasian girl, presented with cardiomyopathy, biventricular dysfunction and pre-excitation on electrocardiogram. She had normal intracardiac anatomy with severely dilated left ventricle and severely diminished biventricular function (Fig. 1). She was treated with milrinone and epinephrine infusions, mechanical ventilation and listed for heart transplant. She underwent Berlin Heart EXCOR biventricular assist device (BiVAD) placement (30 ml LVAD and 25 ml RVAD pumps). No supraventricular tachycardia (SVT) was inducible or noted during her hospitalization. First ablation attempt without BiVAD support was unsuccessful; however, 18 days post BiVAD implantation, another electrophysiology study and successful radiofrequency ablation of a right anterolateral accessory pathway was performed on BiVAD support. After successful ablation and loss of pre-excitation, the cardiac dysfunction rapidly improved with initial improvement noted as early as 48 h after the successful ablation. Due to recovery of cardiac function, a BiVAD wean protocol was initiated and BiVAD explantation was performed 48 days after the implant (30 days after the successful ablation). To the best of our knowledge, this is the first report of successful BiVAD explantation.

Case report

Ventricular pre-excitation, also known as Wolf-Parkinson-White (WPW) pattern on electrocardiogram (ECG)/WPW syndrome is relatively common and is known to occur in 1–3/1000 live births [1]. It can be asymptomatic or can manifest as supraventricular tachycardia and rarely as syncope or sudden death due to rapid conduction of atrial fibrillation/tachyarrhythmia over the accessory pathway. Another, less common manifestation is ventricular dysfunction [2,3] the pathogenesis of which is poorly understood but is likely due to premature ventricular activation, abnormal ventricular septal movement, left ventricular (LV) dyssynchrony and LV remodeling [4–6].

We report a 20 month-old patient with a right sided accessory pathway and severe ventricular dysfunction requiring biventricular assist device (BiVAD) support which completely resolved after successful ablation of the accessory pathway. This is the first known report of successful explantation of a BiVAD (Berlin Heart EXCOR, Berlin Heart GmbH, Berlin, Germany) in a child.

This 20 month-old Caucasian girl presented with a 6-day history of intermittent fever, cough, labored breathing and poor oral intake. She weighed 13 kg. There was no history of SVT or syncope. No other obvious cause was noted for her cardiomyopathy. Initial evaluation and work-up showed a normal C-reactive protein concentration (<5 mg/l), a negative viral panel, a markedly elevated pro-NT brain type natriuretic peptide concentration (29,800 pg/ml), a negative metabolic work-up, cardiomegaly on chest X-ray, ventricular pre-excitation with a QRS duration of 146 msec on her ECG. Echocardiogram showed normal intracardiac anatomy with severely dilated left ventricle, severely diminished biventricular function and moderate mitral regurgitation. (Figs. 1, 2 and 3).

She was hospitalized to the cardiac intensive care unit and inpatient management included mechanical ventilation, milrinone and epinephrine infusions. She did not have any supraventricular tachycardia during the hospitalization or during the subsequent electrophysiology studies. An electrophysiology study was performed on day 1 of hospitalization without any assist device support in a hemodynamically unstable state despite use of milrinone and epinephrine infusions. The accessory pathway was localized to the right anterior part of the tricuspid valve annulus but attempted radiofrequency ablation was only transiently successful. The procedure was terminated due to hemodynamic instability. Her cardiac function remained poor despite escalation in supportive therapy; she was therefore electively put on BiVAD support (Berlin

* Corresponding author at: Department of Pediatrics, Pediatric Electrophysiology, University of Arkansas for Medical Sciences, Arkansas Children's Hospital, 1 Children's Way, Little Rock, AR 72202, USA.
E-mail address: Sdas@uams.edu (S. Das).

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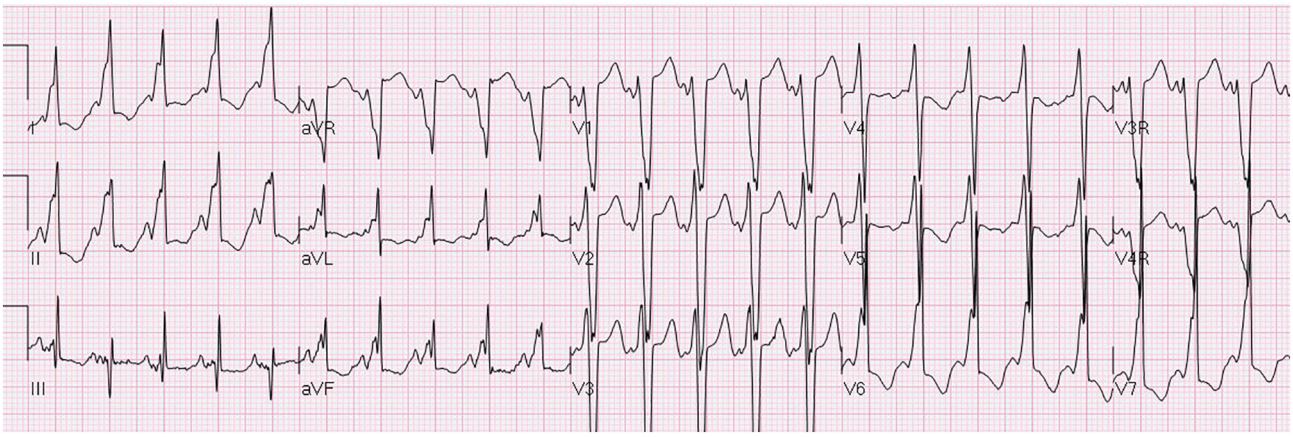

Fig. 1. Pre-excitation through a right-sided accessory pathway.

Heart EXCOR BiVAD, 30 ml LVAD and 25 ml RVAD pumps, two 9 mm inflow cannulas and two 6 mm outflow cannulas) and listed for heart transplant 4 days later. The cardiac dysfunction persisted post-BiVAD implantation; therefore, a repeat electrophysiology study and successful ablation was performed on BiVAD support 18 days after BiVAD implantation. The ablation procedure was performed using only an 8.5 Fr SR-0 sheath (St Jude Medical) in the right femoral vein and a 6 Fr sheath in the left femoral vein. Activation mapping was performed with BlazerTM II (7F/4mm) radiofrequency ablation catheter and modification of the Ensite NavX electrode patches with EnSite™ Velocity™ Cardiac Mapping System. Although there was some difficulty in placement of patches, mapping, movement and positioning of catheter due to small patient size and indwelling BiVAD cannulas, there was loss of delta wave within 7 s of the first attempted lesion. After this successful ablation, there was no return of the delta wave on follow-up. No supraventricular tachycardia (orthodromic or antidromic reentry tachycardia) was inducible during EP studies or noted during her hospital stay. The pathway had an effective refractory period of 280 msec.

After successful ablation and loss of pre-excitation, the cardiac dysfunction rapidly improved with initial improvement noted as early as 48 h after ablation (Figs. 4 and 5). Due to recovery of the heart function, a BiVAD wean protocol was initiated and the Bi-VAD was explanted on Day 52 of hospitalization (48 days after the implant, 30 days after the successful ablation). The explantation was successfully undertaken using a modification and adaptation of the existing Berlin LVAD explantation protocol. This is the first known BiVAD explant in a pediatric patient. The patient was discharged home after 73 days of hospitalization. On follow up at 3 years, there has been no recurrence of delta wave and the cardiac function remains normal.

Discussion

Ventricular dysfunction in the absence of sustained tachycardia is a less common manifestation of ventricular pre-excitation, with only isolated case reports. Most of the reported cases have been associated with a right posteroseptal accessory pathway; [3–9] however, right anterior, right free wall and fasciculo-ventricular pathways have also been implicated. [2, 10, 11] Of note, pre- excitation induced ventricular dysfunction can be reversible after ablation of the pathway [2–13]. Our patient also had a right-sided accessory pathway which is consistent with other reported cases and presented with severe heart failure which required BiVAD support due to biventricular dysfunction.

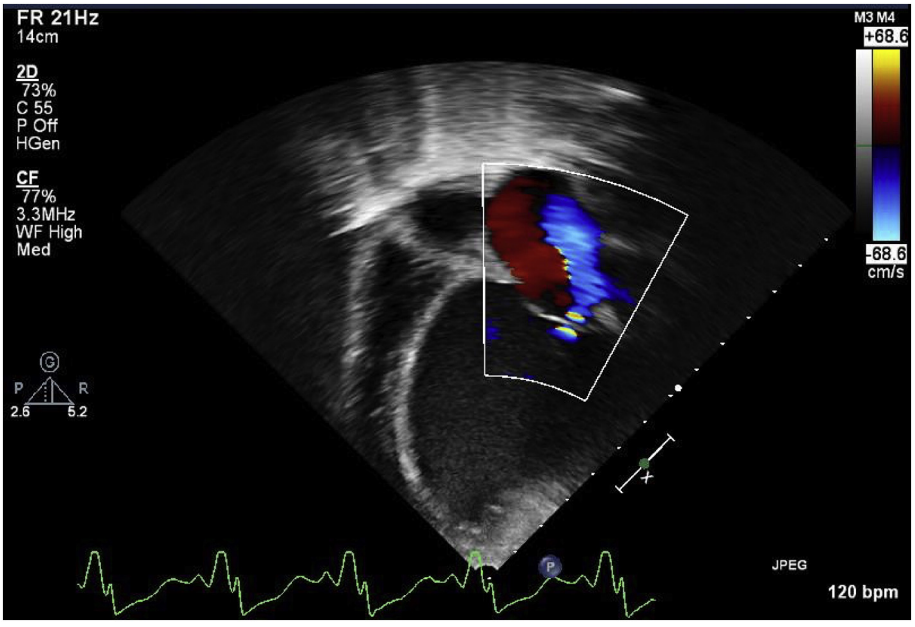

Fig. 2. Severely dilated left ventricle with moderate mitral regurgitation.