Successful right anteroseptal manifest accessory pathway cryoablation in a six-month infant with dyssynchrony-induced dilated cardiomyopathy

Pelin Köşger, Fatma Sevinç Şengül[1], Hasan Candaş Kafalı[1], Birsen Uçar, Yakup Ergül[1]
Department of Pediatric Cardiology, Faculty of Medicine, Eskişehir Osmangazi University; Eskişehir-*Turkey*
[1]Department of Pediatric Cardiology/Electrophysiology, Sağlık Bilimleri University, Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Center; İstanbul-*Turkey*

Introduction

Ventricular preexcitation via an accessory pathway (AP) results in an asynchronous spread of ventricular depolarization that may lead to an abnormal regional wall motion and systolic dysfunction in patients with Wolff–Parkinson–White (WPW) syndrome, even without documented supraventricular tachycardia (1-3). It is known that adults and children with preexcitation-associated cardiomyopathy can completely recover following resynchronization

therapy (3-5). However, in infants, a very small number of patients have been treated with resynchronization via catheter ablation and only one case with less than 6 months of radiofrequency ablation was found in the current literature (5, 6). We present our report of one of the youngest patients in the literature, who showed rapid and complete recovery of severe ventricular dyssynchrony and myocardial dysfunction after cryoablation of the right-sided manifest AP. Our case emphasizes that cryoablation can be preferred as a safe method for AP ablation in infants as well.

Case Report

A previously healthy 5-month-old girl was referred to our hospital for a heart murmur. She weighed 8 kg and the physical examination revealed a mild, grade 1-2/6 heart murmur, mild tachypnea, and hepatomegaly. There were typical ventricular preexcitation signs like short PR intervals, wide QRS complexes (130 ms), and delta waves on a 12-lead surface ECG, which were suggestive of an anteroseptal manifest AP (Fig. 1a). The echocardiographic examination showed left ventricular enlargement, dyskinetic movement of basilar septum characterized with rightward systolic bulging, and impaired heart function with a left ventricular end-diastolic diameter of 40 mm (z-score 4.4) and a left ventricular ejection fraction of 34% as calculated by Simpson's method (Fig. 1b). The Holter examination showed no dysrhythmic events. After all the results were thoroughly evaluated, we considered that the LV dyssynchrony caused by preexcitation due to the right anteroseptal AP may have lead to her LV dysfunction. In accordance with the further echocardiographic investigations, measurements for the interventricular mechanical delay (IVMD) and intraventricular septal-to-posterior wall motion delay (SPWMD) were 74 ms and 290 ms respectively, which were consistent with dyssynchrony (Fig. 1c). Due to the confirmed presence of symptomatic dyssynchrony-induced dilated cardiomyopathy, we perfomed an electrophysiologic study.

The patient was intubated and the electrophysiologic study was performed under general anesthesia. The right and left femoral veins were catheterized and an esophageal catheter was also used. Three-dimensional (3-D) mapping (EnSite Velocity system; St. Jude Medical, St. Paul, MN, USA) and limited fluoroscopy were used together for delta mapping, and the earliest location was found within –38 milliseconds in the right anteroseptal/parahisian region. A 6 mm cryocatheter was used for ablation and at the 4th second of the first cryomapping, the AP disappeared (Fig. 2). Four complete cryo-lesions at –80 oC were given at this location/target. There was no complication during the procedure, except for right bundle branch block. After 72 hours post-ablation, the basilar septal hypokinesia and left ventricular function were markedly improved with a shortening fraction of 27%. At the 9-month follow-up, left ventricular functions and dyssynchrony measurements were found to have improved (LV ejection fraction 69%; IVMD=19 ms and SPWMD=5 ms; Fig. 3).

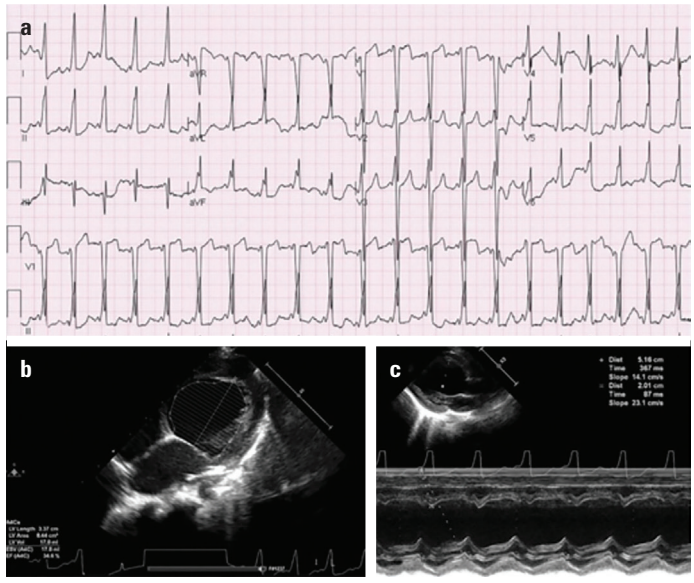

Figure 1. (a) 12-lead surface ECG recorded on admission showing marked delta waves consistent with an anteroseptal manifest AP (WPW preexcitation) (b) Decreased LV systolic function with an EF of 34.6% (measured with Simpson's method in an apical four chamber view) (c) M-Mode echocardiography image showing marked dyssynchrony caused by the ventricular preexcitation of the anteroseptal AP and dyssynchrony measurements

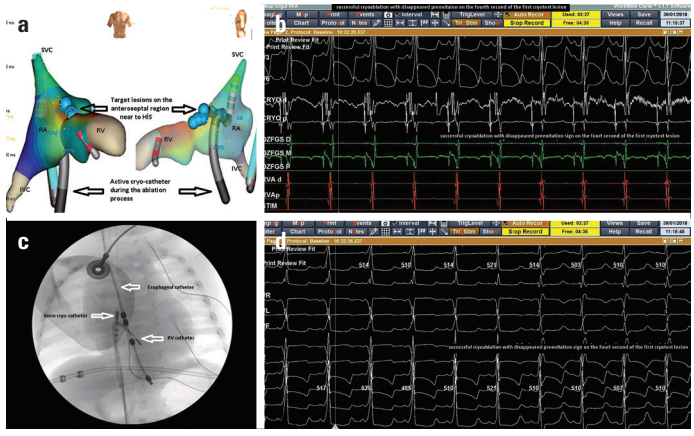

Figure 2. (a) Three-dimensional anatomy of the right atrium and ventricle, with blue dots showing targets on the anteroseptal region and diagnostic catheters in the high right atrium (HRA), esophagus (OZFGS), right ventricle (RVA), and the 6 mm cryocatheter active in the middle. (c) Fluoroscopic image in the left 300 oblique position, with the two diagnostic catheters in the esophagus and the RV and the 6 mm cryocatheter in the anteroseptal region. (b and d) Intracardiac and surface 12-lead electrograms recorded during successful cryoablation showing the disappearance of the preexcitation sign in the 4th second of the first cryotest lesion

IVC - inferior vena cava, RA - right atrium, RV - right ventricle, SVC - superior vena cava

Discussion

The possibility of a causal relationship between ventricular preexcitation and cardiomyopathy without tachycardia in patients with WPW syndrome was first described in a 67-year-old male patient with a diagnosis of WPW in 1998, who showed sig-

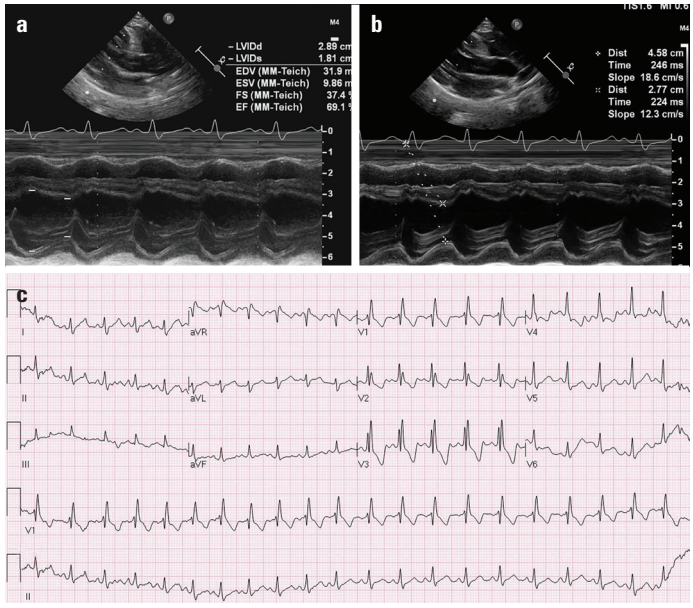

Figure 3. (a) M-mode echocardiography image in parasternal long-axis view showing improved left ventricular systolic function with an EF of 69%. (b) M-mode echocardiography image in parasternal long-axis view showing improved dyssynchrony measures. (c) 12-lead surface ECG, showing no ventricular preexcitation sign and right bundle branch block after successful ablation of the AP

nificant improvement in myocardial function after ablation (7). In 2004, Emmel et al. (8) reported the first such cases in children, where four patients aged 12–156 months of age range were diagnosed with WPW and were considered as having non-tachycardia-induced cardiomyopathy with a left ventricular shortening fraction of 13%–25%. In these cases, a significant improvement in ventricular function followed by a loss of preexcitation with spontaneous or radiofrequency catheter ablation suggests that ventricular preexcitation plays a role in cardiomyopathy without tachycardia (8). Further, the AP located in the right septal region with a longer QRS duration has been reported to be strongly associated with abnormal wall motion and septal dyskinesia both in children and adults (3, 9). In the present case, similar to previous reports, the location of the AP was in the right anteroseptal region and the QRS duration was 130 ms, which was consistent with left bundle branch block (LBBB) morphology.

Abadir et al. (4) reported a significant improvement in dyssynchrony and left ventricular systolic function after catheter ablation of the AP in 16 children aged 14.2±3.7 years. However, in infants, successful radiofrequency ablation has been reported only in two cases, one patient aged 4.5 months and another aged 16 months (5, 6). Furthermore, spontaneous termination of accessory route transmission and pharmacological suppression was reported in a few WPW-diagnosed infants with significant dyssynchrony and cardiomyopathy in the literature (8, 10). Inadequate catheter ablation experience and the increased risk of complications due to low body weight have led to the pharmacological suppression of the AP in infants. Nevertheless, experienced clinicians prefer catheter ablation instead of amiodaron due to the knowledge of drug-resistant cases, frequent relapses of the symptoms of preexcitation, dysfunction following discontinuation of the drug, and long-term drug use causing severe side effects (5, 6). Kwon et al. (6) reported a successful radiofrequency catheter ablation in their 4.5-month-old patient, whose left ventricular function progressively deteriorated and was unresponsive to amiodarone despite the gradually increased dose. This was the first patient aged less than 6 months who was treated for cardiomyopathy caused by WPW-related dyssynchrony in the literature (6). Five years after this, Wu et al. (5) reported that improvement in cardiac functions after radiofrequency ablation began in the second year and full recovery was achieved after 3.5 years in a 16-month-old infant with WPW. Similarly, we preferred catheter ablation to suppress the right anteroseptal AP in our mildly symptomatic 6 month-old patient. In addition to the increased risk of AV block due to radiofrequency ablation in infants and young children, cryoablation was preferred instead of radiofrequency considering the parahisian localization of the AP and preexcitation was successfully suppressed. Unlike Wu et al. (5), we observed improvement in cardiac functions and decrease in dyssynchrony on the third day following ablation.

Conclusion

In conclusion, ventricular preexcitation due to APs, especially those located in the right septal and paraseptal area, may cause ventricular dyssynchrony and cardiomyopathy in infants younger than 6 months of age. Therefore, WPW syndrome should be considered as a reason for preexcitation in infants whose cardiomyopathy is investigated without tachycardia, especially in the presence of prolonged QRS and LBBB morphology. Additionally, with cryoablation performed by the experienced clinicians, there is a chance of successful and safe ablation of the AP and complete restoration of cardiac function in infants younger than 6 months of age.

Informed consent: Written informed consent was obtained from the parents.

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Address for Correspondence: Dr. Pelin Köşger,
Eskişehir Osmangazi Üniversitesi Tıp Fakültesi,
Çocuk Kardiyoloji Bilim Dalı,
Eskişehir-*Türkiye*
Phone: +90 222 239 29 79 - 7440
E-mail: pelinkosger@gmail.com

DOI:10.14744/AnatolJCardiol.2019.93707