Radiofrequency ablation for right-free-wall manifest accessory pathway in a child with asymptomatic Wolff-Parkinson-White syndrome-induced left ventricular dysfunction

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Abstract: A 5-year-old male child with asymptomatic Wolff-Parkinson-White syndrome, cardiac dysfunction and dilated cardiomyopathy was reported. Electrophysiological study revealed a manifest accessory pathway on the right free wall. Heart failure and ventricular remodeling recovered after a successful radiofrequency ablation in the 10-11 o'clock of the tricuspid annulus.

Conclusion: The dyssynchronous movement of the ventricular septum and left ventricular caused by right accessary pathway may be the main electrophysiological mechanism and the prognosis is good after blocking the conduction of accessary pathway.

Keywords: Wolff-Parkinson-White syndrome; Dilated cardiomyopathy; Cardiac dysfunction; Accessory pathway; Radiofrequency ablation.

Introduction

Supraventricular activity passing down through the accessory pathway (AP) to excite part or all of the ventricle in advance called Wolff-Parkinson-White syndrome (WPW), which is often with or without clinical symptoms. However, WPW combined with recurrent or persistent tachyarrhythmia could also cause tachycardia induced

cardiomyopathy (TIC)[1]. Moreover, although some children with dilated cardiomyopathy (DCM) and asymptomatic type B WPW also have been observed, they were mostly diagnosed as idiopathic dilated cardiomyopathy with type B pre-excitation syndrome in the past. As the causal relationship between ventricular pre-excitation and DCM has been discovered in recent years, asymptomatic WPW in children with cardiac dysfunction and DCM has gradually been paid attention to, but its pathogenesis is not specifically clear. Several studies suggested that it was mainly associated with right manifest AP, particularly septum or paraseptal AP. Interestingly, there were still few specific diagnosed cases about right free wall pathways and effectively treatment of ablation having been reported[2-3]. Therefore, present article reports an ablation for right-free-wall manifest accessory pathway in a child with asymptomatic Wolff-Parkinson-White syndrome-induced left ventricular dysfunction.

Case report

A 5-year-old child was hospitalized for three times at the Children's Hospital of Chongqing Medical University due to edema and fatigue. The main diagnosis was acute exacerbation of chronic heart failure, dilated cardiomyopathy, WPW syndrome. The electrocardiogram (ECG) showed type B WPW syndrome (Figure A). Left ventricular end diastolic diameter (LVDd) and left ventricular end-systolic diameter (LVDs) measured by echocardiography demonstrated that the left ventricle significantly enlarged. Echocardiography showed that the left ventricular ejection fraction (LVEF) was 32% and the short-axis fractional shortening (FS) was 15%, indicating cardiac dysfunction. Then, the child discharged because of refusing to receive radiofrequency

ablation after symptoms relieving under receiving digoxin and diuretics during hospitalization. However, she re-hospitalized for aggravating heart failure, with LVEF 28% and FS 13%, even if regular medication and following-up at outpatient was performed.

Given the child coexist asymptomatic type B WPW and DCM, we considered that left ventricular dysfunction was related to the dyssynchronous movement of ventricular septum and left ventricle induced by the right-side AP. After the child was undergone compound intravenous anesthesia, the electrophysiological study (EPS) and radiofrequency ablation are performed and guided by the Carto three-dimensional mapping system (CARTO). Multipolar electrode catheters were introduced through the right femoral vein and positioned in the high right atrium, right ventricle apex, His bundle for recording and stimulation. The real-time rhythm of body surface and intracardiac electrocardiogram were recorded simultaneously. The ablation electrode was initially positioned to map at the right free wall according to ECG and the AV fusion and V wave being obviously pre-delta wave were found at 10-11 o'clock of the tricuspid annulus (Figure B-D). The pre-excitation component disappeared after 2s for the utilization of 40 W energy unit (Figure E) and the ablation endpoint was reached after further application of 160s. After 30 minutes of observation, there was no eccentric conduction and tachycardia episodes being observed under the ventricular pre-procedure stimulation (S1S2) and then the ablation was declared successful.

No complications were observed after one day through the ECG and echocardiography and the ECG showed the disappearance of the date wave. The child

took the medicine for digoxin, diuretics regularly after discharge and returned to the hospital for follow-up after 3 months, with the echocardiography showing the significant increasement in LVEF and improvement of exercise endurance. We concluded that ventricular remodeling for DCM and cardiac dysfunction was associated with the pre-excitation originating from the manifest accessory pathway of the right free wall resulting in the left ventricular myocardial electro-mechanical abnormalities.

Discussion

In the 1970s, heart enlargement and cardiac dysfunction caused by pre-excitation were initially reported. Although DCM may occur in patients with recurrent or persistent tachyarrhythmia mediated by WPW, it also was found in patients with asymptomatic WPW and recovered after blocking the forward function of AP, which suggested that it may be related to pre-excitation [4-5]. Nagai et al. used two-dimensional speckle tracking imaging ultrasound technology and found that △LVEF is positively correlated with △ dyssynchrony index, which further confirmed abnormal excitation-mechanical contraction coupling could lead to ventricular remodeling and left ventricular dysfunction[6]. However, there have been only dozens of cased were reported until now and the incidence of this disease may be underestimated because some patients may had undergone radiofrequency ablation before the development to cardiac dysfunction and other patients are possibly considered to be idiopathic DCM.

New detection technology, speckle tracking echocardiography (STE), equipped higher sensitivity, which could precisely evaluate LV strains, LV dyssynchrony and LV torsion and it has been used to evaluate the features of WPW and confirm the existence

of LV dyssynchrony. Nakatani demonstrated that the brain natriuretic peptide (BNP) level in plasma was positively correlated with the degree of dyssynchrony and proposed a new indicator His-CS delay, which was used together with septal-to-posterior wall motion delay (SPWMD) to assess the degree of LV dyssynchrony[7].

The occurrence of asymptomatic WPW induced left ventricular dysfunction was mainly related to the location of the APs and the degree of pre-excitation. Besides above, the load of pre-excitation and the age of the patients also made an important role in disease development[8]. Although manifest right-side AP, especially the septum or paraseptal AP, was considered the major factor of this disease, still a few cases caused by right free wall APs. Several studies postulated that the right free wall AP has little effect on the part of the ventricular septum and could not significantly affect the synchronization activity of the left ventricle [9-10]. However, Dai et al found that right free wall APs had a greater adverse effect on LV dyssynchrony than septal or paraseptal APs in patients with type B WPW and may induce LV dysfunction, even DCM, if occurred serious dyssynchronous LV contraction and segmental dyskinesia[11]. Moreover, JaeKon et al postulated that basal septal segmental contraction with bulging and delayed free wall activation may lead to significant dyssynchrony in the LV motion and cause ventricular dysfunction[12].

Notably, although there are currently no definite guidelines, the diagnostic criteria must meet the following criteria: 1) The existence of DCM excluding other causes，especially tachycardia cardiomyopathy; 2) Absence of recurrent and sustained

tachyarrhythmias ("asymptomatic" WPW); 3) ECG or EPS proving right-sided AP; 4) Dyssynchronous of ventricular contraction, left ventricle enlargement and LVEF decreasing.

After long-term drug treatment, the patient's condition was not improved significantly. Therefore, considering that the patient's clinical symptoms, exercise tolerance and LVEF improved significantly after radiofrequency ablation treatment and excluding tachycardia cardiomyopathy and other secondary DCM, we believed that the child's DCM and left ventricular dysfunction was induced by asymptomatic WPW. Currently, radiofrequency ablation is still the first choice for almost children who can tolerate surgery due to its high success rate, low complications, and low cost.[1,13] Moreover, alternative pharmacologic therapy such as traditional amiodarone and propafenone instead of RFCA should be chosen for infants with low weight and under 6 months of age who are related to high surgical risk[14,15]. Interestingly, Suzuki et al reported that flecainide was utilized to treat for asymptomatic WPW in an infant with severe left ventricular dyssynchrony and achieved good results and its efficacy and safety in the pediatric population was subsequently confirmed by Cunningham et al. Surprisingly, Kwon et al. even reported a successful RFCA for dyssynchrony-induced dilated cardiomyopathy in a 4.5-month-old infant[16,17].

In short, patients coexist with type B WPW and DCM are recommended to undergo echocardiography to evaluate ventricular wall motion, LV size and LV function. The dyssynchronous movement of the septum and left ventricular caused by pre-excitation may be its most important pathogenesis and RFCA can effectively

improve its prognosis and provide evidence of diagnosis. Therefore, continuous exploration of its pathogenesis, clinical features and diagnosis criteria to help clinicians diagnose as soon as possible and standardize treatment is of great significance for improving the treatment effect.

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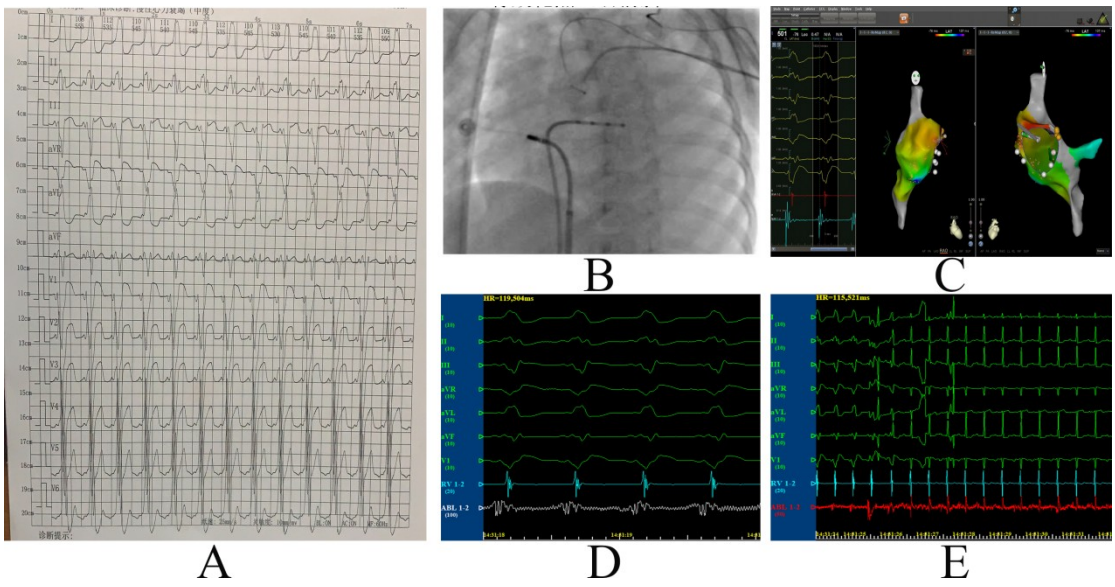

Figure legends

Figure A: The electrocardiogram showed type B WPW syndrome.

Figure B: X-ray appearance of ablation target.

Figure C: Carto three-dimensional mapping of ablation target.

Figure D: Intra-cardiac electrocardiogram image of the target point before ablation (AV fusion and V wave advance).

Figure E: Intra-cardiac electrocardiogram image immediately after successful ablation (A-V separation).