IMPROVEMENT OF CARDIAC FUNCTION AFTER RIGHT SIDED ACCESSORY PATHWAY CATHETER ABLATION IN A CHILD WITH DILATED CARDIOMIOPATHY

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INTRODUCTION

Ventricular pre-excitation can be asymptomatic or can manifest as supraventricular tachycardia[1,2]. Another cause of morbidity is accessory pathways mediated ventricular dysfunction due to abnormal ventricular septal movement[3-9]. We present a case of cardiac resynchronization by RFCA to a right-sided accessory pathway (AP) in a pediatric patient with a genetically determined DCM.

CASE REPORT

A 12 months old infant showed a paradoxical splitting of the second heart sound at a routine pediatric checkup. Her 12-lead ECG showed wide QRS complexes with delta wave (**figure 1A**). Severe LV dysfunction was detected by transthoracic echocardiography, with a dilated LV and an ejection fraction of 37% (**figure 2A**). She was asymptomatic, and multiple holter ECGs were negative for tachyarrhythmia. Viral, metabolic, autoinflammatory work-up tested negative; a coronary angiography ruled out coronary arteries anomalies. Genetic testing showed an heterozygous mutation in NEXN gene (shared with her mother). She underwent medical therapy with enalapril and spironolactone, ineffective.

figure 1A

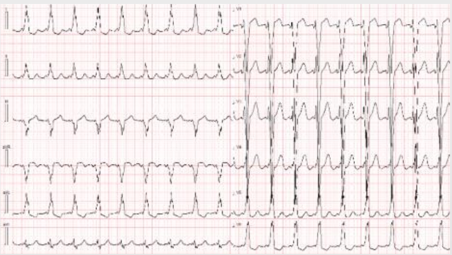
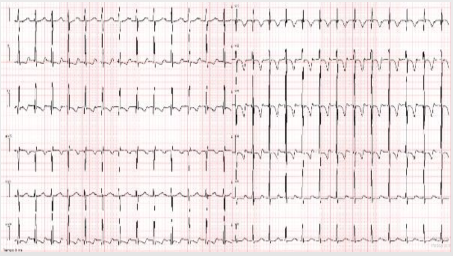


figure 1B



Considering that ventricular pre-excitation may have worsened her LV dysfunction, we performed an electrophysiological study (EPS) and RFCA at the age of 6 years old. The EPS showed a right antero-lateral AP with a refractory period of 350 ms and without induction of tachyarrhythmia; the RFCA, performed with a zero-fluoroscopy approach, was effective in eliminating the AP. In the 12-lead ECG performed the next day a first degree A-V block was noted (**figure 1B**). This finding was consistent with a previous A-V conduction delay, masked by ventricular pre-excitation, as RFCA was performed without affecting His bundle.

A two years follow-up showed absence of the delta wave, improvement of the LVEF up to 60% (**figure 2B**), and recovery of basal septum dissynchrony (**figure 3**).

figure 2A

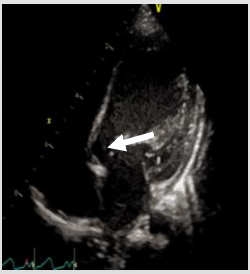
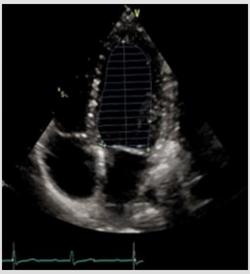


figure 2B



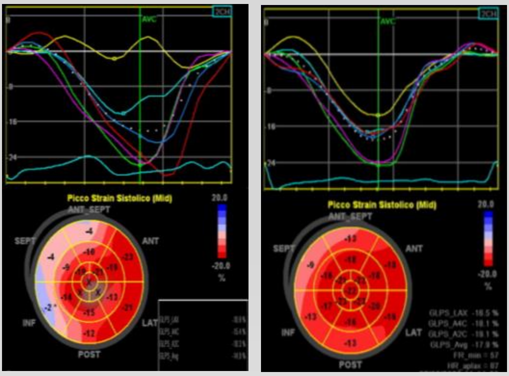


figure 3: *longitudinal 2D strain in apical 2-chamber view before (on the left) and 4 months after RFCA (on the right).*

DISCUSSION and CONCLUSIONS

Premature ventricular activation generates asynchronous ventricular wall motion especially with septal APS[10]. With septal APs there is in fact a short conduction time from the sinus node to the atrial insertion of the septal AP. This "earlier" pre-excitation induces a basal septum segmental contraction which will be followed only later by the remaining LV myocardium activation[6]. In the present case, a possible explanation of such septal dissynchrony with a right free wall AP might be found in the long PR interval at the 12-leads ECG post RFCA[11]. In fact, if a right free wall AP coexists with a conduction delay in the normal A-V system, there is a "relative" pre-excitation of the basal septum provided by the AP; the delayed conduction via the normal A-V system will thus activate later the remaining LV myocardium, contributing to LV dissynchrony.

The finding of a severely reduced LV function in patients with WPW syndrome requires a thorough examination of the underlying causes, as in most of the pediatric patients with WPW syndrome an asynchronous septal motion doesn't affect LV performance. Although catheter ablation of APs in patients asymptomatic for tachyarrythmia is still object of debate[2], in selected patients, in which pre-excitation is likely to disarrange ventricular synchrony, EPS and RFCA may be indicated.

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