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Case Report

An unusual type of accessory pathway in tricuspid atresia

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ABSTRACT

The occurrence of pre-excitation in tricuspid atresia (TA) is slightly more common than that in normal children. The accessory pathway (AP), when it occurs in the setting of congenital atrioventricular valvar disease, is usually ipsilateral to the side of the abnormal valve. This report describes a patient with TA who had pre-excitation due to a left-sided AP that masked and modified the typical electrocardiographic features. The electrophysiological study confirmed an epicardial left posterior AP that was successfully ablated with radiofrequency energy, through the coronary sinus. Left-sided APs including epicardial ones may rarely be seen in TA and can potentially cause difficulties due to lack of vascular access to the heart after the Fontan surgery if arrhythmias occur. They are amenable to successful radiofrequency ablation and need to be dressed prior to Fontan surgery.

<**Learning objective:** Pre-excitation may become manifest during follow-up even if minimal at initial presentation. Accessory pathways may occur in the left side of the heart in tricuspid atresia, in contrast to the usual notion that APs occur ipsilateral to the side of the abnormal valve. Electrophysiological study may be useful prior to Fontan-type procedures as this helps in identifying and to prognosticate pre-excitation so that ablation of the pathway can be performed prior to the Fontan procedure.>

Introduction

The incidence of pre-excitation in tricuspid atresia (TA) is 0.29–1.3% [1] which is slightly more common than in other children (0.05–0.31%). This is of paramount importance in these patients in whom Fontan type palliation is usually indicated. TA also has a characteristic electrocardiographic (ECG) pattern among various cyanotic congenital heart diseases (CHD). This case report underscores several atypical features about accessory pathway (AP) and ECG pattern in a patient with TA that has important clinical implications.

Case report

An 8-year-old girl with TA, non-restrictive ventricular septal defect, severe pulmonic stenosis, and normally related great vessels had undergone central aortopulmonary shunt surgery at 1 month of age. Subsequently, at the age of 9 months, she underwent division of the central shunt, bidirectional Glenn procedure (BDG), and atrial septectomy along with augmentation of the right pulmonary artery with a pericardial patch. ECG at that time was showing short PR interval and minimal pre-excitation (delta wave evident only in leads I and aVL).

Two months' post-BDG, she was found to have stenosis of the superior venacaval (SVC) anastomotic site with a mean gradient of 6 mm of mercury which was treated with balloon dilatation. During the same procedure, venovenous collateral from innominate vein was detected and coiled successfully. Subsequently, she remained asymptomatic with a saturation of 85–90% by pulse oximetry and has never had any palpitations. Recently she underwent a catheter study prior to the planned total cavopulmonary connection procedure (TCPC). Pulmonary pressures were normal and two major arteriovenous collaterals were identified and coiled. This time, however, her ECG showed short PR interval, delta wave, and deep S wave in lead V6 with no features of left ventricular (LV) dominance in precordial leads (Fig. 1A). As she was planned to undergo a TCPC, it was decided to undertake an electrophysiological study (EPS) before. Basal cycle length was 590 ms. Recording the His potential was difficult and unstable and HV interval was 20 ms. Incremental rate atrial pacing showed 1:1 pre-excited atrioventricular conduction up to 250 ms.

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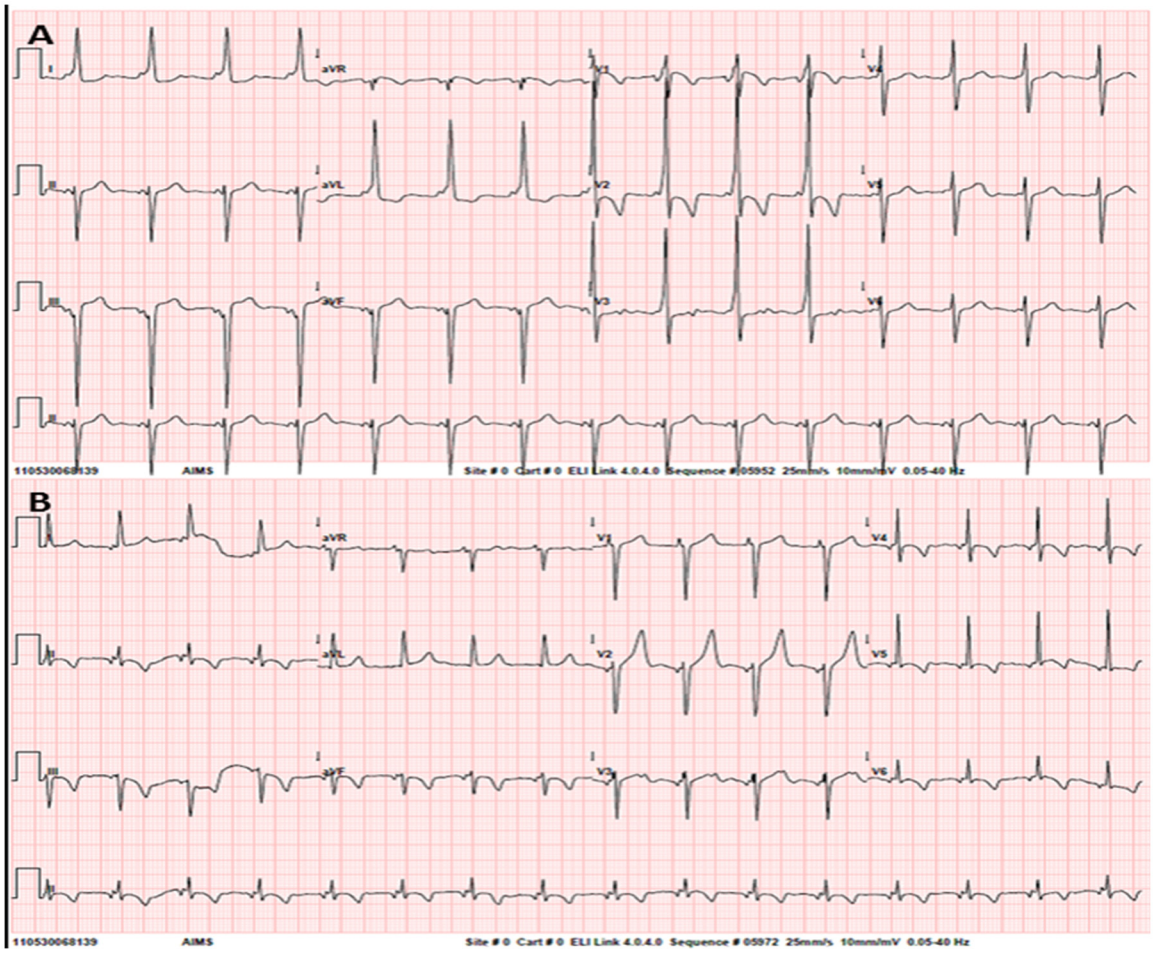

Fig. 1. (A) Twelve-lead electrocardiogram (ECG) showing short PR interval, delta wave and deep S waves in lead V6. The typical precordial pattern of tricuspid atresia is masked and the LV forces are inconspicuous. (B) Twelve-lead ECG after radiofrequency ablation shows a change in precordial pattern and LV forces typical of tricuspid atresia have been evident. 'Memory T wave inversions' are also to be seen.

Programmed stimulation showed antegrade effective refractory period (ERP) of the pathway to be 500–240 ms. Ventricular pacing showed eccentric non-decremental VA conduction with the earliest A in the coronary sinus (CS) 5, 6 electrodes. There was no inducible tachycardia. Mapping of the mitral annulus showed no areas where local V preceded the earliest delta wave. Mapping in the proximal CS (Fig. 2) showed a fusion of AV signal and local V preceded the earliest delta wave by 10 ms, suggesting a left posteroseptal epicardial AP (Fig. 3A). Radiofrequency ablation (RFA) was done here with 20 W power at 50 °C and resulted in local AV separation and loss of pre-excitation in 6 s (Fig. 3B). Ablation was continued in this region for a total duration of 120 s. There was no evidence of AP conduction at 30 min post-ablation. ECG after RFA showed LV dominance typical of TA, a slight reduction in the frontal QRS axis, and diffuse T wave inversions (Fig. 1B). There was ectopic atrial rhythm as evidenced by abnormal P wave axis and the AH interval was 50 ms.

Discussion

This case opens several interesting avenues for discussion. The implications of pre-excitation in TA, the occurrence of a mimicking pattern, its role in masking the typical ECG pattern, and the phenomenon of 'memory T waves' are all worth scrutiny. More important is the unusual occurrence of accessory pathways contralateral to the atretic valve and its epicardial location. There are several reports of accessory atrioventricular pathways in patients with TA and previously surgical division of AP during Fontan procedure has been performed in these patients [2]. Currently, RFA is successfully performed in this scenario [3]. Pre-excitation in patients with TA has major connotations. Arrhythmias are a well-known cause of failure of the Fontan circulation. Atrial arrhythmias that can occur after Fontan can result in a fast ventricular rate in the presence of an AP. Endovascular access to the heart is hampered after Fontan procedure making mapping and ablation of the APs more complex. Puncture through the baffle is needed for this. The pre-excitation may become manifest for the first time only after the Fontan procedure, when there is a change in the conduction properties of atrial tissue, AV nodal tissue, and the sinus node function [4–6]. Our patient had no evidence of pre-excitation at the time of presentation, and it was noted only during evaluation prior to TCPC procedure. She never had any palpitations or documentation of tachycardia. An EPS was performed to find out the refractory period of the pathway, which is important in prognostication of asymptomatic pre-excitation. As the pathway had relatively short ERP, ablation was carried out even though there was no inducible tachycardia considering the difficulties of performing the same after TCPC procedure. It is noteworthy that this patient had only subtle features of pre-excitation in initial ECGs, and pre-excitation

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