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Case Report
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Para-Hisian Accessory Pathway and Paroxysmal Second Degree Atrio-Ventricular Block in a Child

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Abstract

We report a rare case about 13 years old boy, with a previous history of syncope after painful bodily trauma. His ECG showed ventricular preexcitation and 24 hours ECG Holter showed a paroxysmal second degree atrio-ventricular block type one (Luciani-Wenckeback) during sleeping.

Transcatheter electrophysiologycal study showed a para-Hisian accessory pathway sited 5.1 mm to His area and no arrhythmias-induced.

Keywords: *Accessory pathway – Atrio-ventricular block – pediatric arrhythmias – electrophysiology study*

Abbreviations

AP: Accessory Pathway

AV: Atrio-Ventricular

AVB: Atrioventricular Block

CS: Coronary Sinus

EAM: Electroanatomic Map

EPS: Electrophysiological Study

FAM: Fast Anatomic Map

HR: Heart Rate

RA: Right Atrium

RV: Right Ventricle

AH: Atrial- His Bundle

HV: His-Ventricular

Case Report

We report a rare case about ventricular preexcitation and second degree atrio-ventricular block (AVB) type one (Luciani-Wenckeback) during sleeping. A 13 years old boy was referred at our center for history of two syncopes after painful bodily traumas, never palpitations reported. In another hospital neurological assessment and myocardial enzymes were normal. His ECG showed sinus rhythm at heart rate (HR) of 60 bpm, ventricular preexcitation with an anterograde conducting pathway with positive delta waves in the inferior leads (II, III, and aVF) and the lateral precordial leads (V3 through V6); negative delta waves were present in lead V1 and often in V2. Leads I and aVL have positive delta waves (negative in aVR). Normal QRS and QTc intervals (Fig.1).

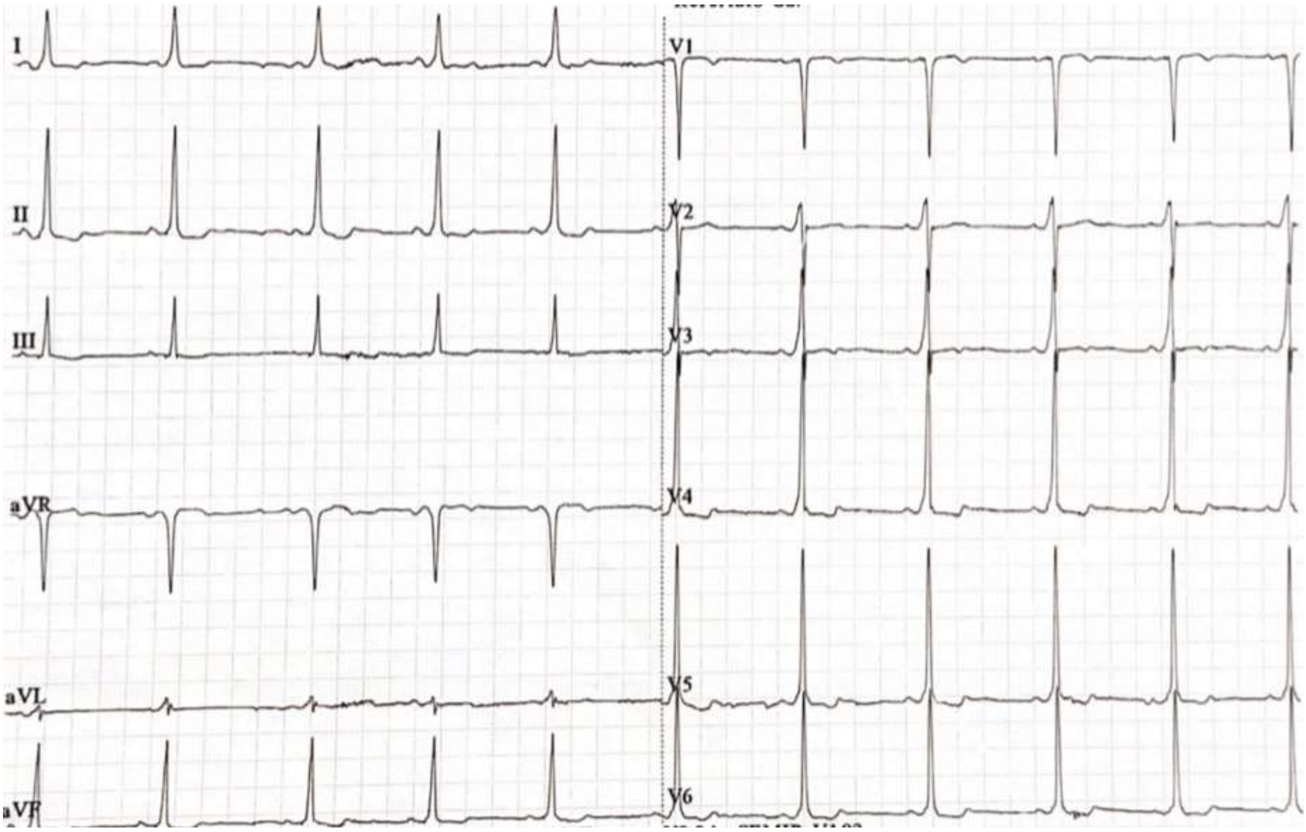

Figure 1 ECG shows sinus rhythm at heart rate of 63 bpm, ventricular preexcitation with an anterograde conducting pathway with positive delta waves in the inferior leads (II, III, and aVF) and the lat-eral precordial leads (V3 through V6); negative delta waves were present in lead V1 and often in V2. Leads I and aVL have positive delta waves (negative in aVR). Normal QRS and QTc intervals.

According to the ECG the ventricular preexcitation seemed to involve an anteroseptal accessory pathway (AP). The ECG Holter 24 h in another hospital showed a paroxysmal second degree AVB type one (Luciani-Wenckeback) during sleeping (Fig. 2) and an uncertain supraventricular tachycardia referred from another hospital.

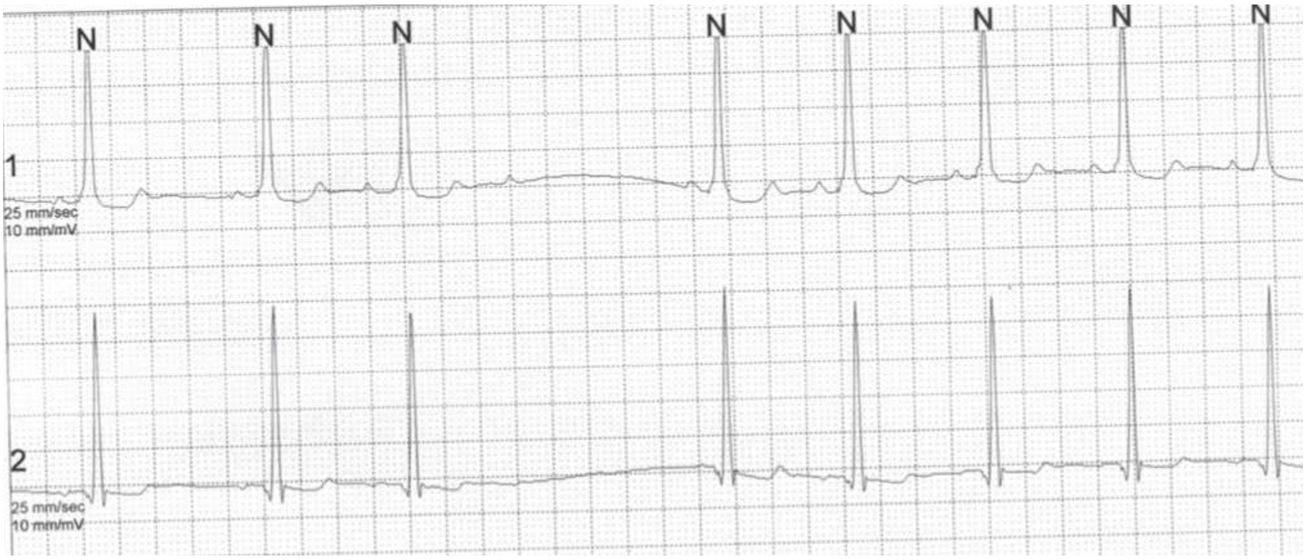

Figure 2 ECG strip shows second degree atrio-ventricular block type 1 during sleeping

The patient was referred to our center and we decided to assess the arrhythmic risk of the AP and the atrio-ventricular (AV) conduction through the transcatheter electrophysiological study (EPS). We de-cided in agreement with family for transcatheter EPS rather than transesophageal EPS to stratify the arrhythmic risk, because of the uncertain supraventricular tachycardia, and to localize the AP as its as-sociation with AVB is very rare.

The echocardiogram showed normal sequential cardiac anatomy, no patent forame ovale, normal ejection fraction, no valvular regurgitation or stenosis, no pericardial effusion.

The procedure was planned as a non-fluoroscopic procedure. Vascular access was obtained without ul-trasonographic guidance. Informed consent was obtained from family.

In view of the paroxysmal second-degree AVB was also evaluate the loop recorder subcutaneous-implantation if a AV nodal disease had been diagnosed.

The procedure was performed under general anesthesia to avoid map shifts caused by patients' movements and endotracheal intubation. The procedure was performed with the guidance of an electroanatomic map (EAM) system CARTO3 (Biosense Webster, Diamond Bar, CA, USA) without using fluoroscopy to reduce the risk of ionizing radiations1-2.

The CARTO3 EAM system, providing the visualization of multiple catheters, was used to navigate the venous system and to perform a right atrium EAM in order to place all the catheters in the heart

Using the Seldinger technique three sheaths were inserted via the right femoral vein: 8 fr for Navistar tip 4 mm catheter, into the right atrium for the EAM, 7 fr for decapolar CS catheter and 6 fr for the quadripolar catheter. A conventional 4 mm quadripolar steerable catheter, with a deflectable tip to facilitate electrophysiological mapping of the heart and to create a matrix (Navistar, Biosense Webster, Inc.) was advanced into the right atrium to obtain 3-dimensional (3D) fast anatomic map (FAM) and to label the anatomic landmarks, including the inferior and superior caval veins, the right atrium (RA), the coronary sinus (CS), His area, and the tricuspid valve annulus.

The reconstruction of the right atrium was obtained by the FAM algorithm. This method continuously records movements of the Navistar catheter. On the basis of volume sampling, a surface reconstruction was built in accordance with the set resolution level. The geometric reconstruction of the right atrium started positioning the Navistar catheter on the superior vena cava. The mapping catheter was then withdrawn from the superior vena cava through the right atrium down to the inferior vena cava, touching the endocardial surface of the lateral, septal, anterior, and posterior walls and tagging specific anatomic landmarks such as the His bundle and the tricuspid ring. The navigation inside the chamber was guided by the integration of the direct visualization of the mapping catheter provided by the CARTO3 system and the analysis of the electrograms recorded from the tip of the roving catheter.

This matrix allowed for the advanced catheter localization of any non–sensor-equipped catheter such as the decapolar catheter advanced into the CS or the quadripolar catheter localized on the His. The His position was identified and marked on the 3D FAM of the RA as a reference.

A decapolar catheter was advanced in the CS and a quadripolar catheter placed in the His area/right ventricle.

The coronary sinus catheter was used as the system reference. To avoid introducing an extra sheath into children, we did not employ a fourth catheter, instead placing the high right atrium catheter in the His bundle when needed. In patients with a baseline sinus rhythm, we performed standard programmed stimulation and burst pacing manoeuvres to induce atrial tachycardia.

Electrophysiological study was performed using standard protocols to assess conduction properties of the AP and arrhythmia inducibility.

The intracardiac recordings showed normal atrial- His bundle (AH) and His-ventricular (HV) intervals.

Early ventricular activation indicative of a para-Hisian AP was noted in the catheter's distal pair (CS 9-10), which had not advanced fully into the coronary sinus, which was sited 5.1 mm to the His area (Fig. 3). Basal cycle length was 740 ms.

Atrial stimulation was initiated with a 600-ms cycle. Wenckeback cycle was calculated after incremental atrial stimulation at 330 ms. Each atrial impulse was always conducted to the ventricle over the AP. Programmed atrial stimulation (600 ms) with single extrastimuli (S1) from 500 ms to 280 ms was performed and a refractory AV nodal period was calculated at 280 ms.

Atrial stimulation with double and triple extrastimuli didn't induce arrhythmias.

The same protocol was repeated with the use of isoproterenol infusion intravenously (8 µg/min) without inducing arrhythmias.

The EPS has been ended as normal nodal conduction and the low arrhythmic risk of the AP. No fluoroscopy was used. The loop recorder has not been implanted because of normal AV conduction and no arrhythmias induced from the EPS.

Based on these data, it was evident that the true mechanism of syncope was vagal stimulation subsequently to painful stimulus. We suspected that the second AVB recorded during sleeping is due to episodes of vagally mediated AVB occurring often during the night.

Syncope in a patient with manifest ventricular preexcitation usually led to indicate a fast conduction to the ventricle over the AP, for this reason the EPS has been conducted to exclude an AP contributing to the mechanism of syncope.

Although this mechanism explains syncope in the most patients with preexcitation, a detailed research for the causal mechanism should be based on the interpretation of the full medical history, considering the second-degree AV block, the painful stimulus and not only on the presence of an AP.

In our literature review, we didn't find cases in pediatric population with similar clinical and electrophysiological characteristics3, few cases have been reported only in adulthood 4-6.

Moret, Schwartz and White wrote an historical manuscript about Wolff-Parkinson-White syndrome coexisting with second degree AV block describing only 8 cases in the 1959 and giving importance and support to their theory of accelerated conduction through the normal pathway 7.

In conclusion, we described a unique case of paroxysmal second degree AVB type one (Luciani-Wenckeback) and para-Hisian AP in a 13 years old boy. The simultaneous presence of second-degree AVB with a para-Hisian AP is rare.

In the baseline intervals of the EPS, we found a normal AH and HV intervals in the His-bundle catheter, leading us to suspect that the paroxysmal second-degree AVB was vagally mediated and not the cause of syncope. In this case, the AP was the preferential way of conduction because during protocol of stimulation the intracavitary electrogram showed always the ventricular preexcitation even in the context of Wenckeback cycle and AV nodal refractory period assessments. The closeness of His area and AP can explain the association of this phenomenon.

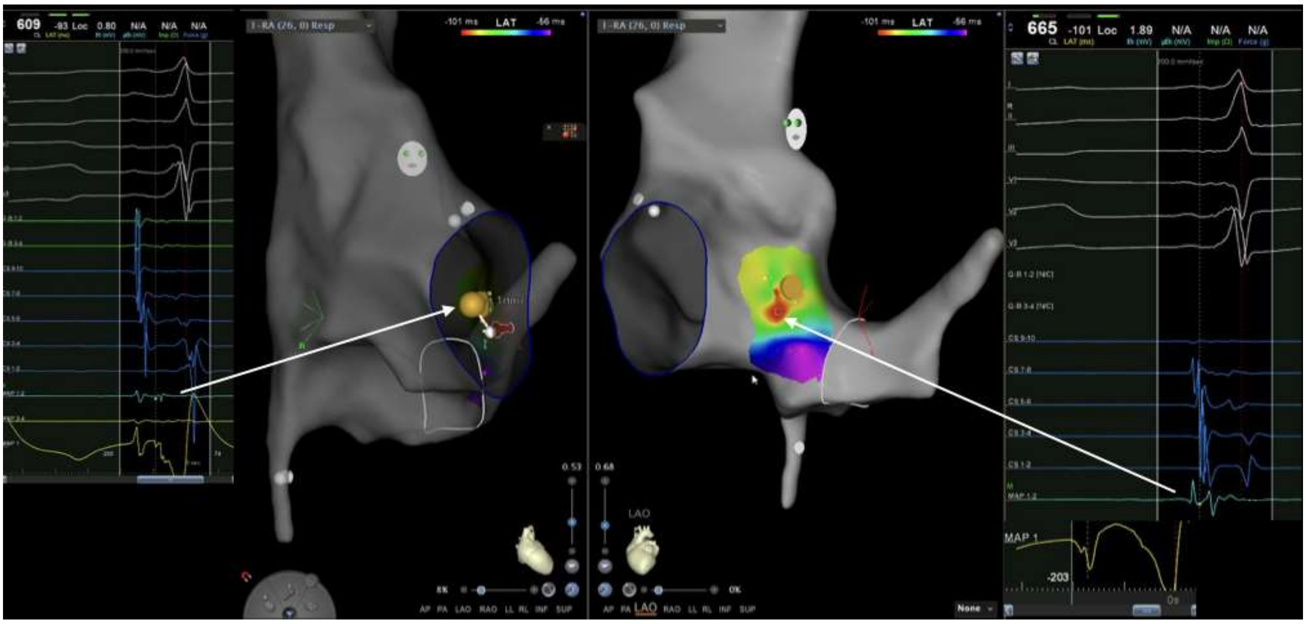

Figure 3 CARTO system shows the right anterior oblique view (on the left panel) and the left anterior oblique view (on the right panel) of the right atrium 3D fast anatomical map with yellow dot—His-bundle potential site and red dot—accessory pathway. The endocardial recording (white arrow on the left) shows His-bundle potential (normal AH - HV intervals) sited 5.1 mm close to the red dot confirming the para-Hisian location of the accessory pathway. On the right panel the endocardial recording (white arrow) shows the accessory pathway potential.

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