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Cryo Ablation of AVNRT in A Child with L-Transposition of Great Arteries (L-TGA); Where are the AV Node Slow Pathway Inputs?

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Abstract

Atrio-Ventricular Re-entrant Tachycardia (AVNRT) is a relatively uncommon form of supraventricular tachycardia in children under 10 years of age. It is particularly rare in patients with L-Transposition of the Great Arteries (L-TGA), especially within the pediatric demographic. In this report, we describe a case involving a child with concurrent Atrio-Ventricular Re-entrant Tachycardia (AVRT) and AVNRT. The AVRT, which involved an infero-septal accessory pathway along the left-sided AV (tricuspid) valve annulus, was successfully ablated using radiofrequency energy. The AVNRT was addressed with cryoablation energy applied to an unusual location along the right antero-superior septum, which typically does not receive slow pathway inputs from the AV node.

Introduction

Atrio-Ventricular Nodal Re-entrant Tachycardia (AVNRT) is a relatively infrequent form of supraventricular tachycardia in children under 10 years of age. In the context of L-Transposition of the Great Arteries (L-TGA), AVNRT is particularly rare, with only a few cases and small series reported [1-3]. Accessory pathways are more commonly observed in patients with L-TGA, exhibiting an incidence of 1.36%, compared to a 0.3% incidence in patients without L-TGA [3,4]. This report details the case of a four-year-old child with L-TGA who presented with AVNRT and Orthodromic Reciprocating Tachycardia (ORT) via a concealed left infero-septal accessory pathway. The ORT was successfully ablated using radiofrequency energy, while AVNRT was treated with cryoablation, targeting a modification of the slow pathway in the right antero-lateral region.

Case report

The patient was a 4-year-old boy with a history of L-transposition of the great arteries, a ventricular septal defect, and a secundum-type atrial septal defect. Despite antiarrhythmic therapy with propranolol and digoxin, he experienced frequent episodes of Supraventricular Tachycardia (SVT). During these episodes, surface Electrocardiograms (ECGs) revealed narrow complex tachycardia at 180-200 bpm without discernible P

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waves. An Electrophysiology (EP) study was conducted under general anesthesia. The placement of electrophysiology catheters included a decapolar catheter in the coronary sinus, a quadripolar catheter at the morphologic left ventricular apex (right-sided), and another quadripolar catheter in the His region. The coronary sinus was notably well-formed, contrasting with the often-underdeveloped sinuses seen in this condition. His bundle potentials were detected at the antero-septal aspect of the right (mitral) AV valve annulus, mirroring the typical location in structurally normal hearts.

Baseline sinus rhythm exhibited a non-pre-excited Cycle Length (CL) of 577 ms, an AH interval of 89 ms, an HV interval of 37 ms, and a QRS duration of 91 ms. Antegrade conduction showed normal decremental properties, initially without evidence of dual AV node physiology. In contrast, retrograde conduction was non-decremental, did not block with adenosine, and the earliest atrial activation occurred at the proximal coronary sinus, well before His activation. The introduction of single atrial extra stimuli consistently triggered sustained orthodromic reciprocating tachycardia with a CL of 335 ms and a VA interval of 80 ms, with an activation pattern identical to that during ventricular pacing (AH interval at 216 ms, and HV at 41 ms).

Atrial activation during this tachycardia and during ventricular pacing was traced to the infero-septal aspect of the (left-sided) tricuspid annulus. Radiofrequency energy application at this site via an antegrade approach across the atrial septal defect successfully eliminated the accessory pathway conduction, rendering the tachycardia non-inducible.

Subsequently, retrograde conduction became decremental and concentric, blocking with adenosine. Further testing with single atrial extra stimuli revealed dual AV node physiology with a clear AH "jump" and the induction of typical AVNRT post-jump, characterized by a CL of 409 ms, a VA interval of 66 ms (earliest on the His catheter), an AH interval of 302 ms, and an HV interval of 42 ms.

Introduction of His-refractory ventricular premature beats during this tachycardia did not advance atrial activation, confirming the absence of a retrograde accessory pathway. Ventricular extra stimuli consistently induced atypical AVNRT, characterized by a CL of 391 ms, a VA interval of 150 ms (earliest on the His catheter), an AH interval of 208 ms, and an HV interval of 37 ms. Adenosine administration during this tachycardia led to gradual VA prolongation before the termination of the SVT.

The RF ablation catheter was then swapped for a 6 mm tip cryoablation catheter. AV node cryo-modification initially targeted the outer third to mid-portion of Koch's triangle. Cryoenergy applications were performed in sinus rhythm with pacing protocols conducted post-freeze. The procedure was adjusted for persistence of AVNRT inducibility or fast pathway effects. After observing lack of efficacy in the middle of Koch's triangle, ablation was targeted to the areas around the coronary sinus ostium and then anterior and superior to the His signals, successfully eliminating both AVNRT inducibility and slow pathway conduction. Subsequent return of slow pathway conduction and AVNRT led to the exchange of the 6 mm tip for an 8 mm tip cryocatheter. Further cryoablation with the larger catheter at the same location effectively eliminated the slow pathway conduction, rendering no further tachyarrhythmias inducible. After the procedure propranolol and digoxin were discontinues. At a follow-up duration of up to 1 year, no recurrence of arrhythmia was noted.

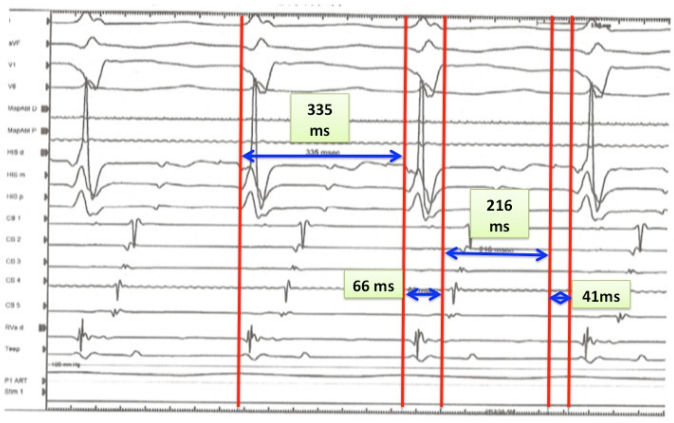

Figure 1: Orthodromic Reciprocating Tachycardia. Orthodromic reciprocating tachycardia with cycle length of 355 msec, VA interval 80 msec (earliest atrial activation on CS5 catheter, clearly preceding the His atrial signal), AH interval 216 msec, and HV interval 41 msec. This tachycardia was successfully eliminated with application of radio-frequency energy along the infero-septal aspect of left AV (tricuspid) valve annulus.

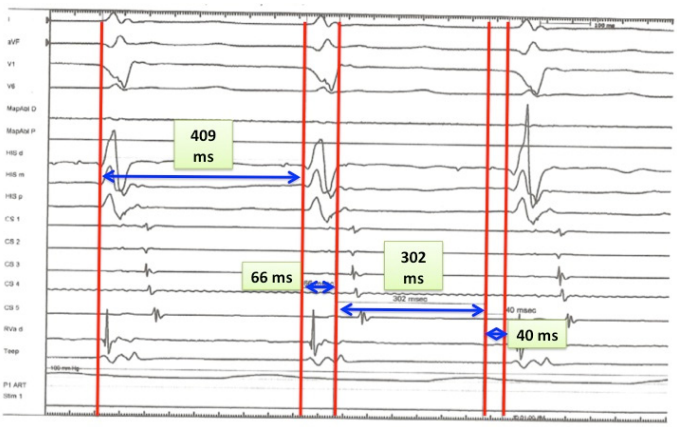

Figure 2: Typical Atrioventricular Nodal Reentrant Tachycardia. Typical AV node re-entrant tachycardia with cycle length 409 msec, VA interval of 66 msec with earliest atrial activation noted on the His catheter, AH interval 302 msec, and HV interval 40 msec. Both typical and atypical AVNRT were eliminated with cryoablation at the right antero-superior septum.

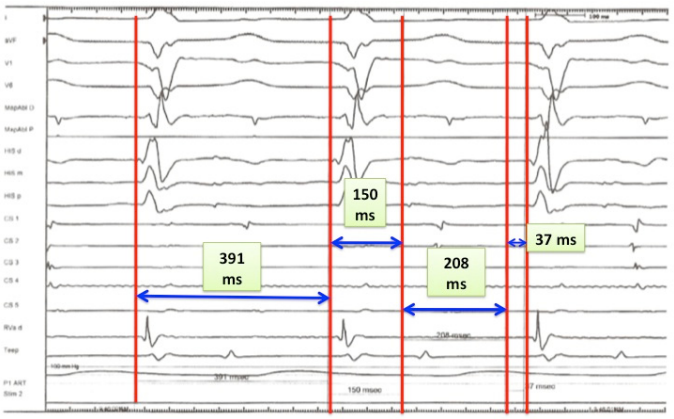

Figure 3: Atypical Atrioventricular Nodal Reentrant Tachycardia. Atypical AV node re-entrant tachycardia with cycle length 391 msec, VA interval 150 msec with earliest atrial activation noted on the His catheter, AH interval 208 msec and the HV interval 37 msec.

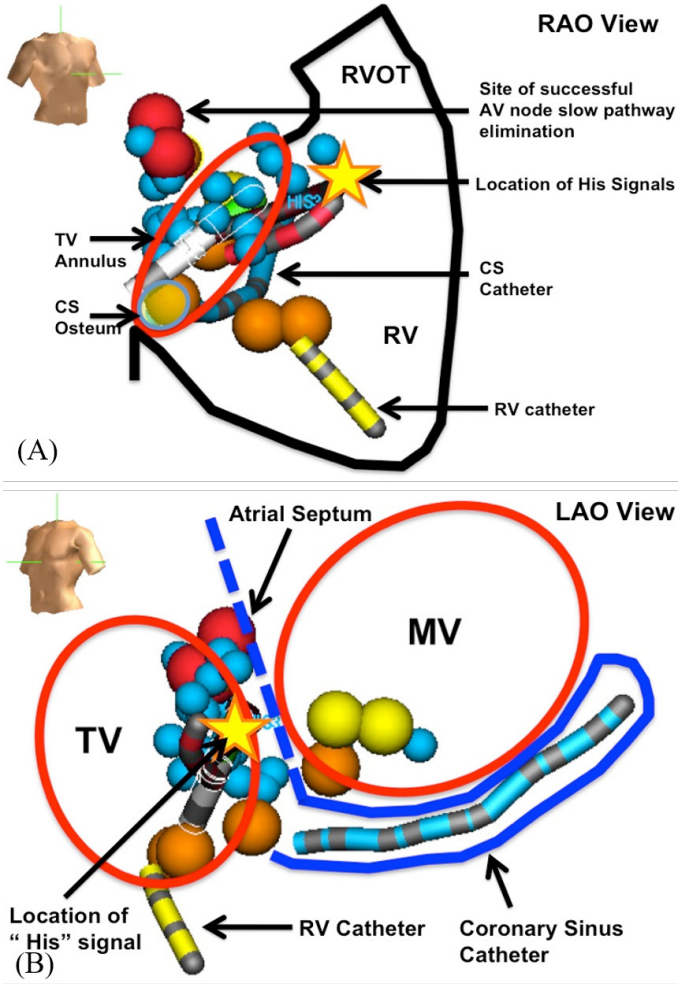

Figure 4: (A & B) Virtual 3D- ablation spots as demonstrated by the Ensite Nav-X electroanatomic recording system. Image A represents the analogous RAO 30^{0} fluoroscopy projection and the image B represents 60 degrees analogous LAO fluoroscopic projection. Anatomic landmarks have been drawn to demonstrate approximate locations of the anatomic structures. The red catheter demonstrates the quadripolar catheter positioned in the His location. The yellow catheter is a quadripolar catheter positioned in the right ventricle and the blue catheter is the decapolar catheter positioned in the coronary sinus. The white catheter represents the cryo ablation catheter positioned superior to the His catheter. The red dots represent the final two cryo freeze applications performed with an 8 mm tip cryo ablation catheter that resulted in elimination of AVNRT, thereby confirming the antero-superior location of slow-pathway input.
Abbreviations: RV: Right Ventricle; RVOT: Right Ventricular Outflow Tract; TV: Tricuspid Valve; CS: Coronary Sinus.

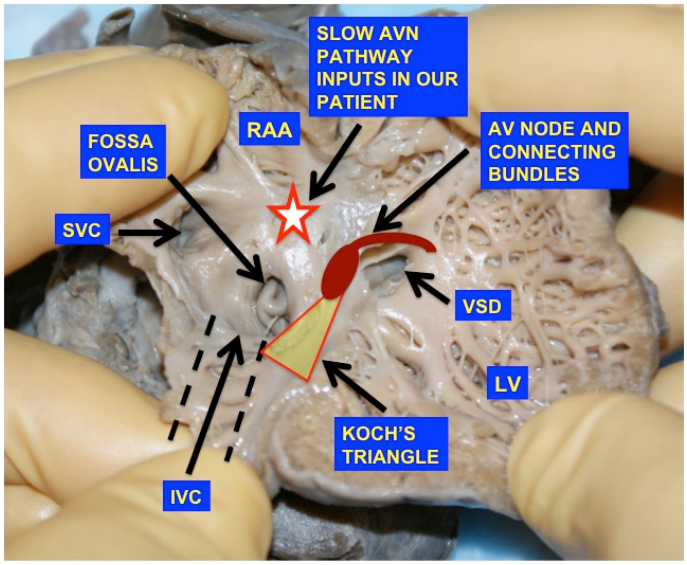

Figure 5: This image demonstrates the anatomic features of a pediatric specimen with L-transposition of the great arteries, The image is displayed with Right Atrium (RA) and Left Ventricle (LV) splayed open. The patient also has an associated ventricular septal defect similar to our patient. The AV node is normally located at the apex of the Koch's triangle and the connecting bundles assume a course over the superior margin of Ventricular Septal Defect (VSD). The slow pathway inputs that are normally located at the outer thirds of Koch's triangle, were noted to be located superior and slightly anterior to the usual location of AV node (Star).

Discussion

AVNRT in a child with L-TGA is rare. AVNRT is an uncommon arrhythmia in pediatric age group accounting for fewer than 13% of all supra-ventricular tachycardia in pediatrics [5] and is extremely rare in L-TGA.

Patients with L-TGA and AVNRT represent an ablation challenge due to unusual anatomy of the atrio-ventricular conduction system. Presence of two AV nodes (both anatomic and physiologic) has been described in patients with L-TGA [6-10]. Anderson et.al. have demonstrated that an anterior connecting AV node is always present and is located anteriorly in the right atrium at the lateral junction of pulmonary and mitral (right AV) valve [6]. The anteriorly situated bundle descends into the morphologic left (right sided) ventricle and encircles the antero-lateral quadrant of pulmonary outflow tract before descending on the anterior septum and bifurcating [6]. In patients with associated ventricular septal defects such as ours, conducting tissues were related to the anterior quadrants of defects [6]. The posterior AV node, if present is usually present in addition to the anterior AV node and not in isolation. Posterior AV node is located in the usual position at the apex of Koch's triangle; however, it is usually not connected to the ventricle. [6,7].

The precise locations of the fast and slow pathway inputs in patients with L-TGA remain poorly defined. In the first documented case of AVNRT in an adult with L-TGA, described by Tada et al., the fast pathway inputs were located at the anterior portion of the left-sided AV annulus, while the intermediate and slow pathways were positioned at the antero-septal portion of the right-sided AV annulus, which seems to be consistent with findings in our patient [1].

Their patient had multiple pathway inputs (slow, intermediate, and fast) to the AV node and radiofrequency ablation at the right antero-septal location resulted in elimination of intermediate and slow pathways [1]. In another report involving an adult with L-TGA and AVNRT, Eisenberger et al. employed electroanatomic mapping to demonstrate that a single AV node and the bundle of His were located in the normal position at the apex of the triangle of Koch, and they identified multiple pathway inputs to the AV node (slow, intermediate, and fast). Radiofrequency ablation at the right postero-septal area successfully eliminated the slow and intermediate pathway conduction [2].

Our patient also demonstrated multiple inputs into the AV node, providing a substrate for both typical (slow-fast) and atypical (slow-slow) AVNRT mechanisms.

Cryoablation in children is considered to be a safe and effective modality for AVNRT ablation [11-13]. Due to the uncertain anatomic location of the slow pathway input, we opted for AV node modification using the conventional anatomic approach,

beginning in the outer third to the middle of Koch's triangle, and subsequently ablating in and around the coronary sinus ostium, before moving superiorly towards the His signal location. Success was achieved anterior and superior to the His signal potential, eliminating tachycardia inducibility. Similar to the case described by Tada et al., ablation near the anterior AV node eliminated AVNRT inducibility as well as slow pathway conduction (Figure 5) [1].

Our experience and review of previously reported ablations for AVNRT in the setting of L-TGA suggest that the locations of slow pathway inputs to the AV node are variable, and ablation at both antero-septal and postero-septal locations along the right AV valve has been successful. The enhanced safety of cryoablation technology proved to be advantageous in our patient.

Patients with L-TGA can experience a variety of cardiac arrhythmias, including various degrees of heart block, Atrio-Ventricular Re-entrant (AVRT) mechanism supraventricular tachycardia involving left-sided accessory pathways, sick sinus syndrome, atrial flutter, and ventricular tachycardia [10]. In addition to AVNRT, our patient also had Orthodromic Reciprocating Tachycardia (ORT) involving a concealed left infero-septal accessory pathway along the tricuspid (left AV) valve. Successful elimination of this accessory pathway was performed using radio-frequency energy. To the best of our knowledge, the co-existence of ORT and AVNRT in a patient with L-TGA has also not been previously reported.

Conclusion

Patients with L-Transposition of the Great Arteries (L-TGA) may present with co-existing Atrioventricular Re-entrant Tachycardia (AVRT) and Atrioventricular Nodal Re-entrant Tachycardia (AVNRT). Multiple pathway inputs (fast, intermediate, slow) to the AV node may be encountered in these patients. The locations of slow and intermediate pathway inputs are variable, typically situated in the superior or antero-septal regions along the right AV valve in individuals with L-TGA. Cryoablation has proven to be an effective method for AV node modification and slow pathway elimination in this with L-TGA.

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