CASE REPORT

Atrioventricular Nodal Reentrant Tachycardia in a Patient with Superior-Inferior Ventricles and Dextrocardia Treated with Cryoablation

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ABBREVIATIONS
AV = atrioventricular
AVNRT = atrioventricular nodal
    reentrant tachycardia
LSVC = left superior vena cava

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ABSTRACT

A 19-year-old female underwent repair of complex congenital heart disease (atrial and ventricular septal defect with a supero-inferior ventricular relationship) in infancy. Because of recurrent palpitations she underwent an electrophysiology study. Atypical atrioventricular nodal reentrant tachycardia was diagnosed. Catheter ablation was performed successfully using cryothermal energy. The diagnostic and therapeutic approach is discussed.

INTRODUCTION

Symptoms of recurrent tachycardia in patients with repaired congenital heart disease are usually due to intra-atrial reentry or a previously undiagnosed accessory pathway.[1] Atrioventricular (AV) nodal reentrant tachycardia (AVNRT) is an uncommon etiology in this context. The location of the AV node may be atypical in these patients. We report a patient with complex congenital heart disease with unusual presentation of AVNRT, who underwent successful treatment with cryoablation.

CASE REPORT

A 19-year-old female born with complex congenital heart disease and multiple congenital anomalies was evaluated for recurrent palpitations. She was diagnosed with a criss-cross type AV connection, atrial and ventricular septal defect, bilateral superior venae cavae with absent connecting vein, dextrocardia, pulmonary hypertension, exomphalos and diaphragmatic hernia. The extracardiac congenital anomalies were repaired in the neonatal period. At two months of age she underwent repair of her congenital heart defects via a mid-line sternotomy. There was accidental resection of the left superior vena cava (LSVC) which was repaired. Later, a stent was implanted in the LSVC because of anastomotic stenosis. She had recurrent tachycardias since

the age of 4 years and reported episodes of presyncope. She was referred for evaluation with an electrophysiology and hemodynamic study.

During hemodynamic evaluation, the pulmonary arterial pressures were normal and there was no residual intracardiac shunt. There was stenosis at the level of the previous stent in the LSVC with a mean gradient of 5 mmHg. The LSVC drained into the coronary sinus (Fig. 1a). The apex of the heart was rotated to the right, and the ventricles had a superior-inferior orientation (Fig. 1b). During an electrophysiology study, at baseline there was first degree AV block with a PR interval of 366 ms, which subsequently shortened spontaneously to 130 ms. The AH and HV intervals were 90 and 40 ms respectively. The VA conduction was absent during ventricular pacing. Sustained slow-pathway conduction was demonstrated during decremental atrial pacing. Atrial extrastimulus testing during

isoproterenol infusion resulted in AH jump and induction of sustained atypical AVNRT (Fig. 2a) with a cycle length of 452 ms (heart rate 133 bpm) and right bundle branch block (RBBB) morphology (similar to baseline). The AH and HA intervals were 142 and 174 ms respectively (Fig. 2b). Administration of adenosine resulted in termination of tachycardia after VA block.

Cryoablation of the slow pathway was performed based on the earliest retrograde atrial electrogram which was recorded in the proximal coronary sinus (Fig. 3). After 4 cryo-applications in this area the tachycardia was no longer inducible, but the AH jump and a single AV nodal echo were still present. After further cryoapplications on the tricuspid annulus in P2 and M1 locations, the echo beat was no longer present, but the AH jump remained without induction of tachycardia. An attempt of cryoablation at a more midseptal location was in-
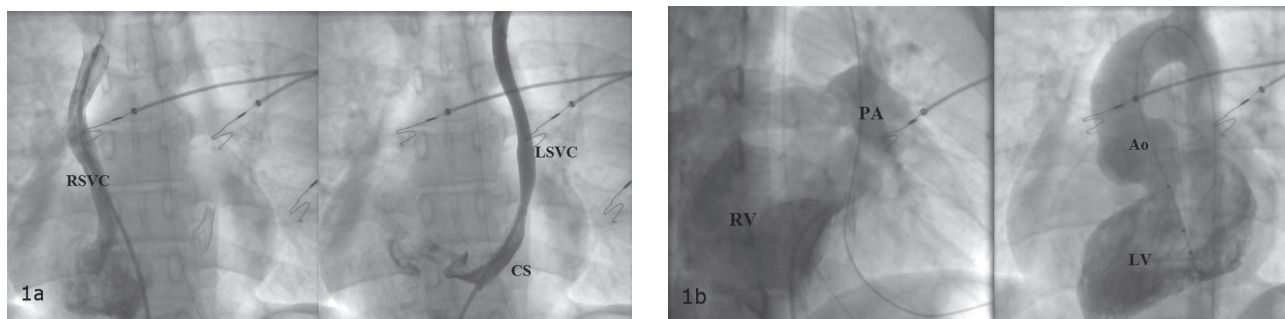

FIGURE 1. In panels 1a, bilateral venae cavae are displayed. The left superior vena cava (right panel) drains into the coronary sinus (CS). There is moderate stenosis above the junction to the coronary sinus. In panels 1b, angiographic demonstration of the superior (RV; left panel)-inferior (LV; right panel) ventricular arrangement with rotation of the cardiac apex to the right (right panel) are depicted. Ao = aorta; CS = coronary sinus; LSVC = left superior vena cava; LV = left ventricle; RSVC = right superior vena cava; PA = pulmonary artery; RV = right ventricle.
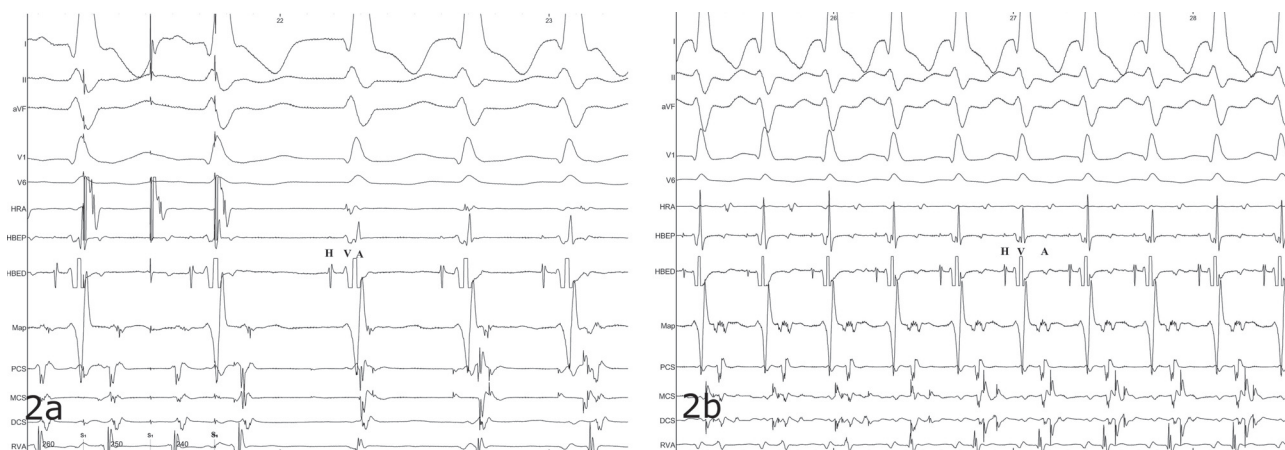
FIGURE 2. Induction of AVNRT with AH jump during programmed atrial extrastimulus testing is illustrated in the left panel (2a). Recording of the stabilized rhythm (right panel, 2b) indicates atypical (slow-slow) AVNRT. AVNRT = atrioventricular nodal reentrant tachycardia.
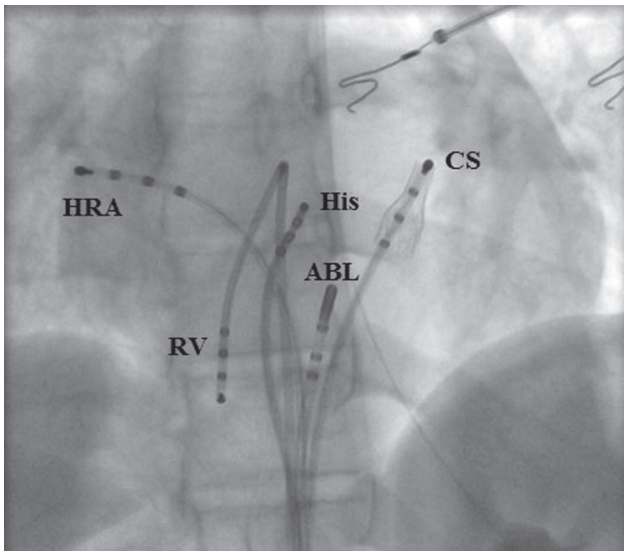
FIGURE 3. Anteroposterior fluoroscopic view of catheter position. The orientation of the catheters is reminiscent of a long axial oblique view. ABL = ablation catheter; CS = coronary sinus; His = His bundle; HRA = high right atrium; RV = right ventricle.
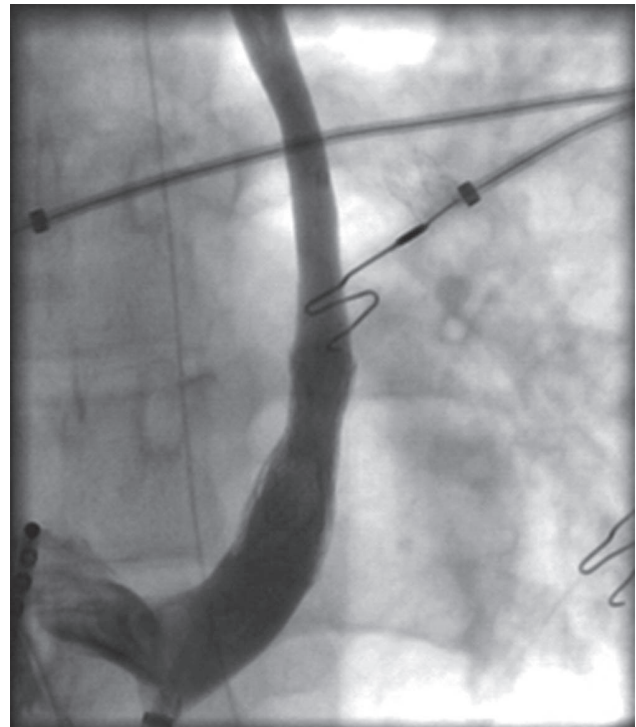
FIGURE 4. Left superior vena cava angiogram after stent implantation with no residual stenosis.

terrupted prematurely because of transient AV block. During the waiting period of the procedure, a Genesis 2910 stent was implanted in the stenosed segment of the LSVC over a 10 mm Opta-Pro balloon (Fig. 4). After the stent implantation, there was no residual gradient. Repeat programmed cardiac stimulation showed no inducible AVNRT despite persistence of slow pathway conduction. Measurement of the intracardiac intervals at the end of the study showed an AH of 108 ms and HV of 58 ms. At this point the procedure was terminated and the patient made an uneventful recovery.

Exercise testing 2 months after the ablation procedure showed a normal heart rate response of 182 bpm (92% predicted) without conduction abnormalities and without tachycardia. Holter monitoring showed sinus rhythm with first degree AV block and rare supraventricular ectopic beats. During follow-up of 7 months after the procedure, the patient reported no episodes of tachycardia.

DISCUSSION

The anatomy of our patient's heart was initially described as a criss-cross AV relationship. Although the two terms are not mutually exclusive, we believe that it is more appropriate to define it as superior-inferior ventricles, since the hallmark of criss-cross heart, i.e. clockwise rotation of the ventricles around their long axes,[2] was not present in our case. As far as we know, this is the first report of a patient with such a malformation diagnosed with AVNRT. Catheter ablation of AVNRT in patients with complex congenital heart disease may be difficult because the anatomy of the AV conduction system may be significantly altered. For example, patients with congenitally corrected transposition of the great arteries (AV and VA discordance) may have two AV nodes, one located anteriorly at the apex of the triangle of Koch and a second located posteriorly close to the coronary sinus ostium.[3] Patients with ostium primum atrial septal defect have a posteriorly located AV node and the bundle of His runs very close to the ostium of the coronary sinus.[4,5] The locations of the fast and slow pathways may occasionally be reversed in patients with ostium primum atrial septal defect.[4] At times the anatomy is even more complex, with two discrete AV nodes and an accessory pathway forming a complex circuit.[6] Although the cardiac anatomy of our patient was complex, the location of the AV conduction system seemed to be relatively straightforward. The main abnormality was the rightward rotation of the heart at the level of the AV junctions, making the anteroposterior fluoroscopic view appear like a left lateral oblique view (Fig. 3). The tachycardia in our patient was atypical, of the so called slow-slow type. We could not demonstrate two different AV nodes, since a single His bundle electrogram was recorded in the usual location. There was no evidence of an accessory pathway, since ventricular pacing resulted in VA

block and administration of adenosine during tachycardia also resulted in VA block and termination of tachycardia. Atrial tachycardia was unlikely, given the induction of tachycardia with programmed atrial stimulation with an AH jump and the termination of tachycardia with adenosine (although the latter is not an absolute criterion).

Treatment of AVNRT with catheter ablation is highly successful. Radiofrequency catheter ablation has a very high success rate, but carries a small but true risk of AV block. This risk may be higher in cases where the anatomy of the AV conduction system is altered by the presence of congenital heart disease. In these cases, the use of cryoablation offers significant advantages, with its excellent safety record, with no case of permanent AV block reported worldwide. In our case, it was very helpful, since we did not know the exact anatomy of the AV node. In fact an attempt to eliminate completely the slow pathway was terminated immediately when transient AV block developed with no permanent sequelae. The success rate of cryoablation is similar to that of radiofrequency energy ablation. A higher recurrence rate has been initially reported,[7] but this seems to be decreasing with experience.[8,9] For this reason many pediatric electrophysiology centers have adopted cryoablation as their primary therapeutic modality in the treatment of AVNRT.

In conclusion, AVNRT was diagnosed in a patient with criss-cross AV relationship after surgical repair of atrial and ventricular septal defect. The tachycardia was successfully treated with cryoablation.

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