Ablation of accessory pathway from right atrial appendage to anatomic left ventricle in L-transposition of the great arteries

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Introduction

A 24-year-old female patient with a history of congenitally corrected transposition of the great arteries (ccTGA) and Wolff-Parkinson-White (WPW) syndrome status post multiple ablations presented with recurrent palpitations from a narrow complex tachycardia at 200 beats/min. She was found to have a rare accessory pathway from the right atrial appendage (RAA) to the morphologic left ventricle, which was successfully ablated.

Case report

History

We present a 24-year-old female patient who was seen at the adult electrophysiology clinic for recurrent palpitations, which were increasing in frequency. These episodes correlated with a narrow complex tachycardia with heart rate around 180–200 beats per minute. She was tried on different antiarrhythmic agents, including sotalol and a combination of flecainide and bisoprolol, without improvement. She had numerous hospital presentations for her symptomatic supraventricular tachycardia (SVT).

Past medical history

The patient had a past history of ccTGA (also known as L-transposition of the great arteries [L-TGA] or ventricular inversion), remote ligation of patent ductus arteriosus, WPW syndrome (Figure 1A) status post multiple ablations at various facilities, and morbid obesity. On prior ablations, radiofrequency applications at the site of shortest AV on the mitral annulus were ineffective at even temporary loss of accessory pathway conduction.

KEYWORDS Accessory pathway; Right atrial appendage; L-transposition of the great vessels; Radiofrequency ablation; Wolff-Parkinson-White syndrome; Supraventricular tachycardia

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KEY TEACHING POINTS

- The use of tools such as the remote navigation catheter should be encouraged, as they increase the likelihood of detecting elusive pathways, while allowing for safe mapping.
- Understanding the embryology of an accessory pathway is important in comprehending its anatomy, which can assist with tracing the pathway and isolating it for ablation when usual approaches fail.
- Rare accessory pathways are found in normal healthy hearts and congenital heart disease alike, but the latter situations pose challenges owing to difficult anatomy.

Investigations

An updated coronary computed tomography angiogram was obtained to assess her cardiovascular anatomy. A prominent RAA was noted. A transesophageal echocardiogram was performed prior to the start of the case and revealed the morphologic left ventricle on the right side and communicating with the pulmonary trunk with a left ventricular ejection fraction >55%, and the morphologic right ventricle on the left side communicating with the aorta. Right ventricle fractional area change was normal at 34%.

Procedure

The electrophysiology study and ablation were done under general anesthesia. Access was obtained via the right femoral vein, left femoral vein, and left subclavian vein. Baseline rhythm was sinus with cycle length 920 ms. The P-delta interval was 60 ms, QRS was 175, and QT was 420 ms. During the electrophysiology study, a multipolar catheter (PentaRay; Biosense Webster, Irvine, CA) was advanced to the right atrium to create a 3-dimensional electroanatomic map with the assistance of the CARTO mapping system (Biosense

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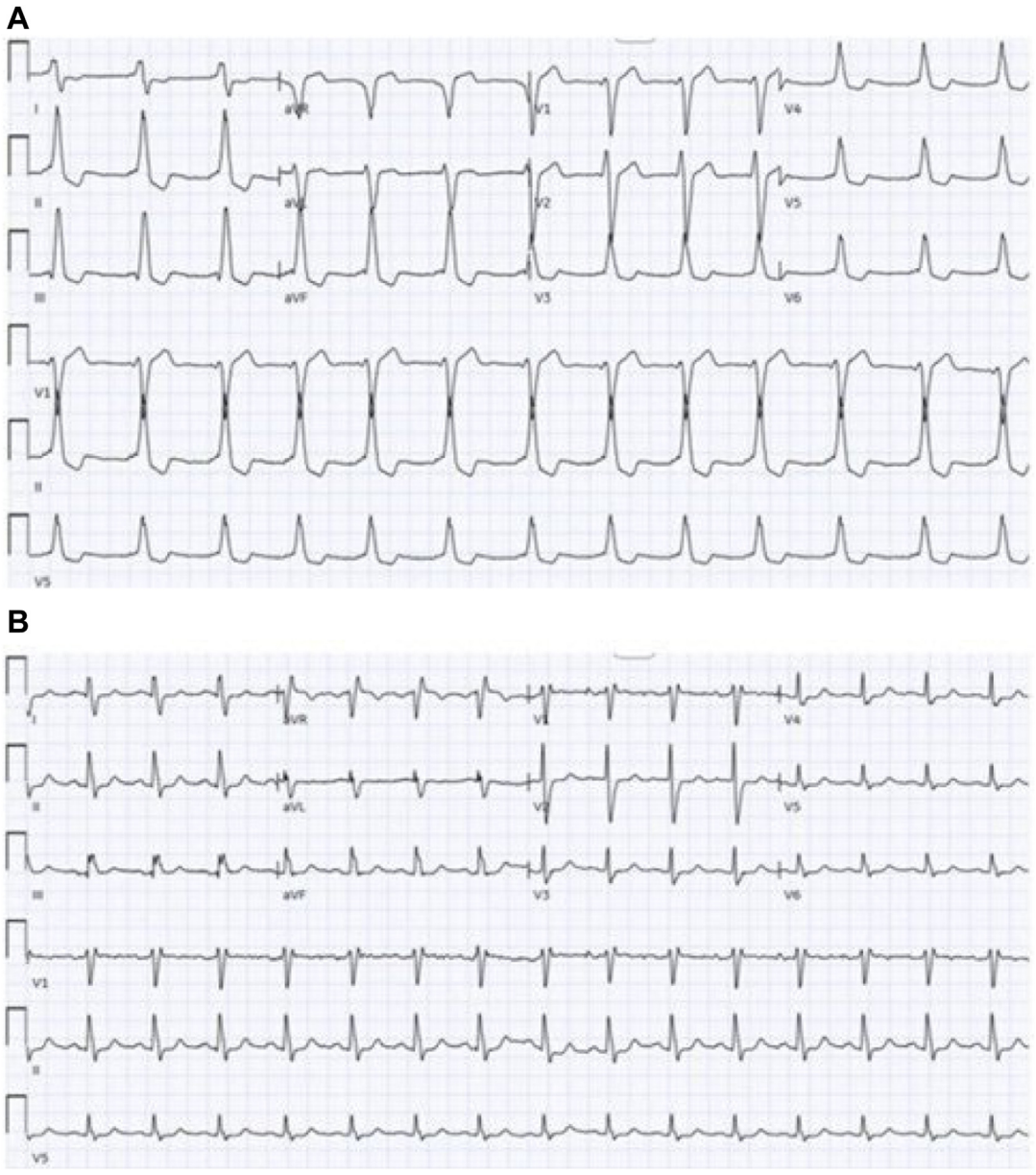

Figure 1 Electrocardiograms. **A:** Preablation. **B:** Postablation.

Webster). Despite extensive mapping, a His signal was not recorded. This inability to find the His had hampered the attempts to ablate by previous operators, as ablation in the anterior mitral annulus may result in heart block in patients with ccTGA. Other ablators had been less circumspect about this potential pitfall and had ablated the area without adverse effects on conduction. A steerable duodecapolar catheter (Biosense Webster) was advanced into the coronary sinus (CS) from the subclavian 7F sheath and left atrial signal was recorded. Ventriculoatrial (VA) conduction was nondecremental and concentric. VA conduction was present down to ventricular effective refractory period of 400/230 ms. VA 1:1 conduction could not be determined owing to initiation of a narrow complex SVT with every attempt at incremental pacing. Pre-excitation was intermittent clinically and patient was not pre-excited at time of study after induction of anesthesia (which had been observed with this patient previously). Nonsustained atrial fibrillation was consistently induced with incremental atrial pacing. The shortest R-R during atrial fibrillation was 270 ms. Because of this, atrioventricular (AV) node block cycle length 1:1 conduction cycle could not be determined. Postablation, AV node 1:1 was $<$280 ms. The patient developed hemodynamic instability when paced faster, so a more exact determination was not made. Antegrade conduction was decremental. AV node effective refractory period was 400/260 ms. Antegrade accessory pathway conduction was felt to be poor based on intermittent pre-excitation. The patient was not pre-excited at the time of our study, so antegrade accessory pathway effective refractory period was not measured. Catheter manipulation

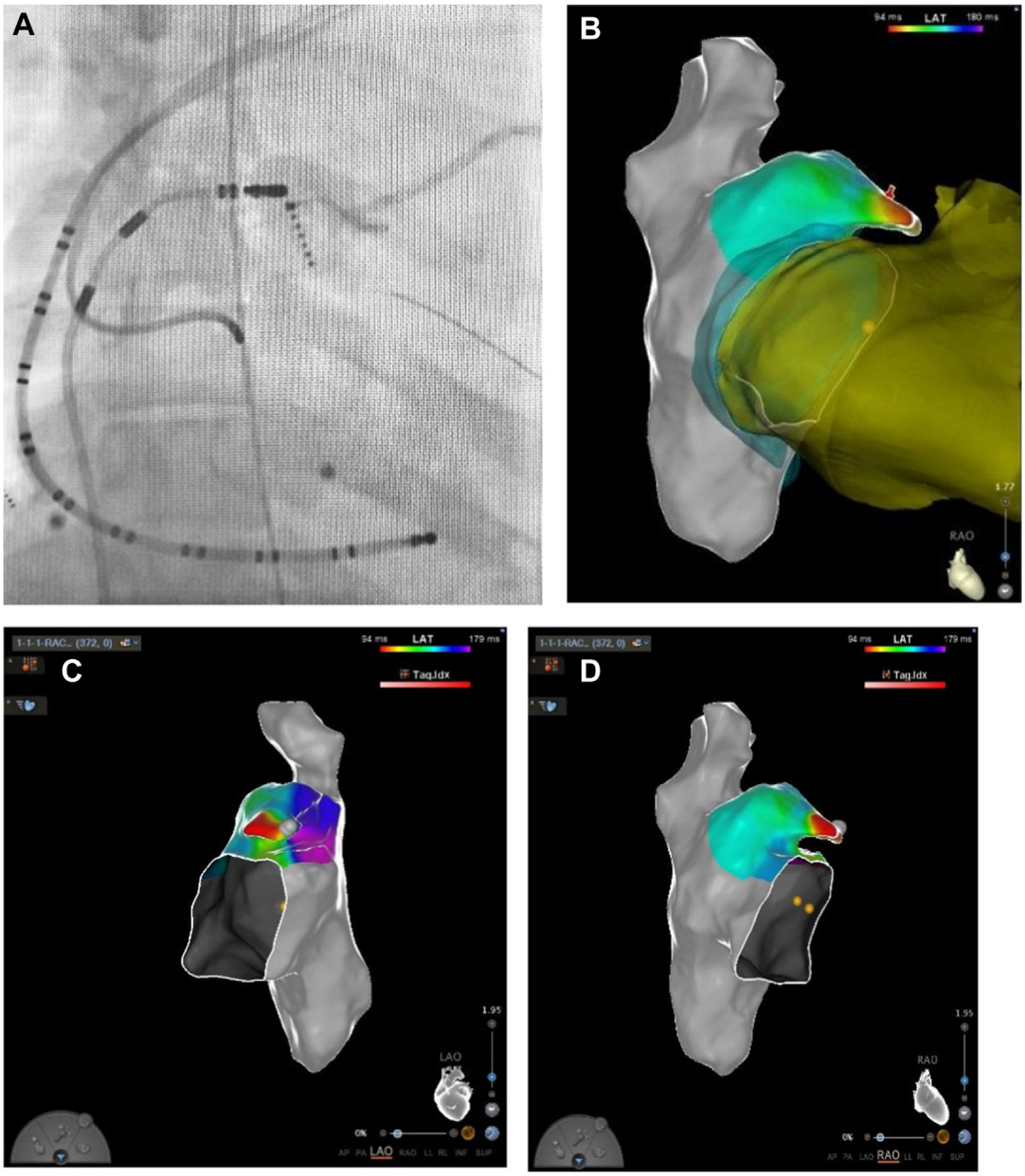

Figure 2 **A:** Anterior fluoroscopic view showing catheter positions at successful ablation site. **B–D:** CARTO map showing earliest activation at the appendage in anteroposterior (**B**), left anterior oblique (**C**), and right anterior oblique (**D**) views. Yellow dots represent ventricular pacing location.

within the right atrium frequently induced tachycardia requiring rapid overdrive pacing to terminate owing to hemodynamic instability. The tachycardia cycle length was 340 ms with a VA time measured to the high right atrium of 85 ms, VA to proximal CS was 150 ms and VA to distal CS was 175 ms. A quadripolar catheter (Biosense Webster) was advanced into the base of the morphological left ventricle where straight pacing was performed at 600 ms, revealing earliest atrial activation at the 12-o'clock to 2-o'clock position on the mitral valve annulus. An irrigated ablation catheter (NaviStar RMT ThermoCool; Biosense Webster) was set up to use with a stereotaxis sheath (St. Louis, MO) and then advanced to the right atrium, where ablation was attempted with earliest activation preceding the onset of the earliest P wave on the 20-pole catheter by 10–30 ms during ventricular pacing. This was unsuccessful, and further maneuvering toward the RAA revealed an early signal within it. There, the signal was found to be early by 70 ms (Figure 2). Initial radiofrequency energy delivered at low power was unsuccessful and fusion of atrial and ventricular signals remained. When power was increased to 40 W, with a 40°C maximum temperature, application of energy was successful at separating the atrial and ventricular signals (Figure 3). The catheter remained in the vicinity but was withdrawn 2 mm to avoid catheter suppression during a 30-minute monitoring period. Neither was the arrhythmia was inducible nor was there evidence of return of the pathway. No atrioventricular block or other complications occurred during the procedure. The pre-

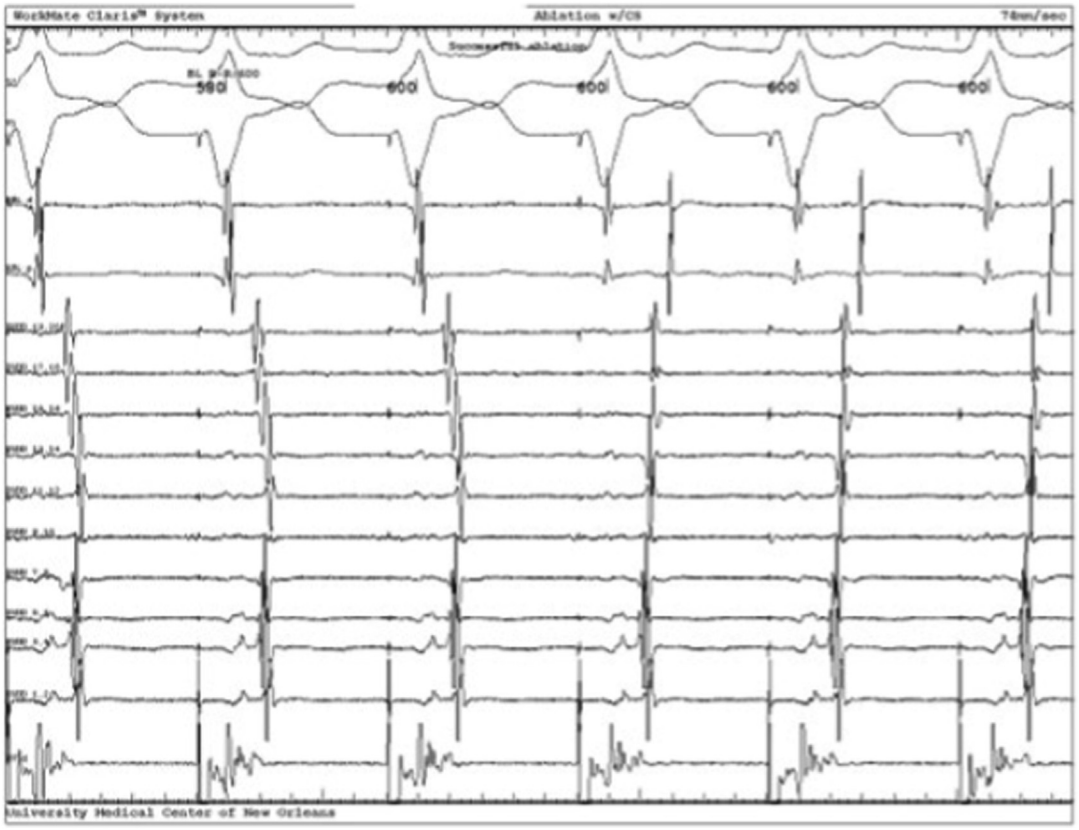

Figure 3 Radiofrequency ablation delivery with separation of ventriculoatrial signals. Change in atrial activation can be seen on 20 pole. The 20 pole is inserted via the left subclavian with poles 19-20 at the junction of the superior vena cava and the right atrium and poles 1-2 in the distal coronary sinus. Therefore, the catheter reflects activation along the lateral right atrial wall, cavotricuspid isthmus, and coronary sinus.

and postablation electrocardiograms are displayed in Figure 1. The patient was discharged home the following day.

Follow-up

On follow-up a few weeks afterward, the patient reported no recurrence of arrhythmias. Her electrocardiogram revealed absence of a delta wave and normalization of PR interval.

Discussion

We present a case with rare and interesting anatomy and pathology, and we believe this is the first of its kind that has been reported. This patient was found to have an accessory pathway extending from the RAA to the anatomic left ventricle, a finding that has not been reported previously, to the best of our knowledge. Additionally, we also present the first successful ablation of this rare pathway in the RAA.

We believe this pathway had an endocardial origin with an epicardial course. Use of the irrigated ablation catheter allowed safe mapping of the appendage, and we believe this is what brought success when previous attempts at ablation at other institutions failed. Intracardiac echocardiography was not used this this case owing to limited access.

Accessory pathways have been reported between the left atrial appendage and left ventricle, and between the RAA and the right ventricle. In the pediatric population, concurrent occurrence of L-TGA and WPW syndrome is not uncommon. In these cases, the accessory pathway is usually in the posteroseptal region.

There have been case reports on right atrial diverticula associated with WPW syndrome.[1,2] Hocini and colleagues[3] reported a case of a 14-year old girl with a "bifid" RAA, thought to be from a giant right appendage and a smaller diverticulum leading to life-threatening arrhythmias from an accessory pathway connecting the appendage and the right ventricle. There are several reports of accessory pathways from the RAA to right ventricle or the left atrial appendage to the left ventricle,[4–7] but to our knowledge this is the only case of an accessory pathway between the RAA and the left ventricle. There are several rare and unique cases reported in L-TGA with accessory pathways, and ours is another addition to a long list of unique presentations.

Understanding the embryology of an accessory pathway is important in comprehending its anatomy. While the atria are normally mostly separated from the ventricles by the annulus fibrosis, most accessory pathways (aberrant myocardial bundles) occur on the AV annulus and are generally exterior or epicardial to the annulus.[8,9] It is important to be aware of unusual accessory pathway insertion locations when mapping at conventional locations has failed.

Conclusion

This is a rare presentation of a patient with SVT originating from an accessory pathway extending from the RAA to the anatomic left ventricle, and a report of our successful ablation of this rare pathway.

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