Complex 3D Mapping in a Young Adult with D-TGA, Atrial Tachycardia, and Limited Venous Access: A Case Report

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Introduction
Continuing advancements in the diagnosis and treatment of pediatric cardiac abnormalities have translated into a rapidly expanding population of adults living with congenital heart disease (CHD). Of those infants born with cardiovascular anomalies, approximately 85% will survive into adulthood, and current conservative estimates put the number of US adult CHD patients at 800,000. Arrhythmias complicate the care of many of these patients and serve as a major source of morbidity. Compounding this arrhythmia burden is the increased prevalence of systemic venous anomalies that make access for ablation therapies more difficult. Lastly, the need for repeat procedures, often at relatively young ages, places the patients at higher risk for the long-term negative sequelae of radiation exposure.

We present the case of a 19-year-old male whose clinical presentation is a summation of these compounding cardiac concerns: he is an adult CHD patient with repaired dextro transposition of the great arteries (D-TGA), supraventricular arrhythmias, and extensive venous abnormalities that complicated access to ablation therapy. The application of cryothermal ablative techniques, unique venous access methods, and three-dimensional (3D) NavX mapping system (St. Jude, Minneapolis, MN) helped to overcome these medical challenges and enhanced the positive clinical outcome that was observed.

Clinic history
A 19-year-old male with D-TGA with a ventricular septal defect (VSD) with a history of arterial switch procedure and VSD closure presented to the Pediatric and Adult Congenital Heart Arrhythmia Clinic after an episode of supraventricular tachycardia.

He had a history of congenital heart disease including coarctation of the aorta, D-TGA with a VSD, patent foramen ovale (PFO), and patent ductus arteriosus (PDA) with supravalvular pulmonic stenosis. His initial surgery was coarctation repair, PDA ligation and pulmonary artery (PA) banding within the first week of life. At 5 months of age he underwent an arterial switch procedure with closure of a small subpulmonic VSD, suture closure of the PFO, and a PA band take down. Owing to difficult vascular access at the time of surgery, he had a left femoral artery cut down and was noted to have a thrombosed left femoral vein.
At 4 years of age he underwent a repeat surgical repair for worsening supravalvular neopulmonary stenosis, at which time he also underwent a balloon angioplasty for aortic coarctation restenosis.

At age 19 years he presented to the arrhythmia clinic with palpitations and a brief syncopal episode. The palpitations typically lasted 1–5 min and were associated with chest pain and intermittent shortness of breath. His baseline electrocardiogram showed sinus rhythm at 60 bpm with right bundle branch block (RBBB). An event monitor of a symptomatic episode documented a wide QRS complex (similar to his baseline RBBB morphology) tachycardia at a rate of 220 bpm. Diltiazem was started, but there continued to be breakthrough episodes. Therefore he elected to undergo an electrophysiology (EP) study and ablation procedure.

Given the history of difficult vascular access, a venous duplex ultrasound was performed of the femoral vessels prior to the EP study. This was interpreted as showing patent femoral veins and arteries bilaterally.

**Electrophysiology procedure**

The patient was brought to the EP laboratory in a fasting state. He underwent general endotracheal anesthesia. 3D NavX patches (St. Jude) were placed to allow 3D mapping. The patient was prepped and draped in the usual fashion. The right femoral venous access was easily obtained without the use of fluoroscopy. The left femoral vein access was difficult, without the ability to advance a wire. Subsequently, contrast was injected to both the left and the right femoral veins, which showed no direct communication to the inferior vena cava (IVC) in either vessel, although there was an adequate size distal right femoral (Figure 1a,b).

Owing to bilateral femoral venous occlusion, access was obtained in the right brachial vein (6.5 French sheath) and a right internal jugular vein (7.5 French sheath). Without radiation a 6 Fr steerable octapolar catheter was advanced via the right internal jugular into the heart. Right atrial geometry was obtained after a marker was placed at the IVC and superior vena cava (SVC) using the presence or absence of intracardiac signals to assist with localization of the right atrium (RA). The steerable catheter was then positioned within the coronary sinus and a His/right ventricular apex (His/RVA) catheter was placed through the right brachial vein and advanced into the RVA. A 5 Fr quadropolar catheter was placed in the esophagus to obtain atrial electrograms.

Even with high output pacing, there was no atrial capture from the esophageal lead. Therefore pacing thresholds were established in the coronary sinus and RVA. A baseline EP study was obtained. On isoproterenol there were short bursts of an atrial-driven arrhythmia with a cycle length of 210 ms, suggestive of atrial ectopic tachycardia or intra-atrial re-entrant tachycardia. Adenosine did not terminate the arrhythmia. Using the coronary sinus catheter, maneuvered with 3D NavX guidance, entrainment was not demonstrated. The earliest atrial activation of the arrhythmia was near the sinus node in the lateral SVC–RA junction, consistent with atrial ectopic tachycardia. Given the proximity to the sinus node and phrenic nerve a Freezor Xtra 6-mm-tip cryoablation catheter (Medtronic, Minneapolis, MN) was used to map the earliest site using 3D NavX. The octapolar coronary sinus position was saved on the 3D NavX map. This catheter was exchanged for the ablation catheter via the right internal jugular sheath because of limited venous access. High output pacing was performed, which captured the phrenic nerve in a location slightly lateral to the earliest activation (Figure 2 and Figure 3). Using the Freezor Xtra catheter, there was successful termination of the ectopic beats shortly after cryotherapy initiation.

![Figure 1: Right (a) and left (b) femoral venous occlusions demonstrated by contrast injection.](image-url)
However, after approximately 1 min of cryoenergy application there was evidence of slowing of the sinus node; therefore, the cryotherapy lesion was discontinued. Additional lesions were placed in closed proximity to the successful site. High output pacing from the ablation catheter was done prior to energy application, and sinus node function was monitored continuously during the ablation. If phrenic nerve stimulation was noted at the potential ablation site, a lesion was not placed at that location. If sinus slowing was noted, the energy application was terminated immediately. In all, eight cryoenergy lesions (4–5 min duration) were placed. Post-ablation EP testing showed no arrhythmia, echo beats or atrioventricular jump. The total fluoroscopy time was 1 min 8 s, the majority of which was used with initial venous access to confirm bilateral femoral venous obstruction. The procedure time was 3 h and 40 min. The patient has had no recurrence of the arrhythmia after 8 months of follow-up.

**Discussion**

With the progressive improvement in the treatment of pediatric congenital heart disease, the population of adults with congenital heart disease is continuing to grow. This, in turn, is increasing the demand for cardiologists who are competent to deal with the unique concerns of this expanding patient population. The case presented here is unique in that it involves the summation of several distinctive attributes. These include the methods required for appropriate venous access, the application of 3D electroanatomic imaging, and the requirement for cryoablative techniques in an adult with CHD.

Venous access for uncomplicated ablative therapies typically requires the utilization of the left and right femoral veins and/or the right internal jugular. The vascular anomalies and venous occlusions observed in some adult CHD patients, however, necessitates the implementation of less traditional methods of access. This can include brachial, subclavian, and transhepatic venous access and occasionally via the femoral or even the brachial or radial arteries. The bilateral femoral venous occlusion necessitated the right brachial and internal jugular veins to be utilized in this patient. The brachial venous approach is used with reasonable frequency by pediatric interventional cardiologists due to limitations imposed by patient anatomy (such as in a bidirectional cavopulmonary anastomosis), but it is not typically considered for EP studies. Nevertheless, it can be considered when venous access is limited. When
using this approach care must be taken to access the
medial brachial vessels, which drain into the axillary
and subsequently the subclavian vein. This allows a
more direct approach to the heart than the more lateral
antecubital vessels. Lateral vessels within the brachial
fossa are more likely to drain via the shoulder, which
may not allow catheter advancement or manipulation.

Three-dimensional mapping methods allow for precise
electroanatomic mapping of the endocardium while limit-
ing fluoroscopy and improving catheter localization for
both initial and repeat energy applications. With the 3D
NavX system any diagnostic EP or ablation catheter can
be used to create the 3D geometry. Adopting technology
such as 3D NavX can also assist with the localization of
important anatomic structures: in our patient’s case, the
sinus node and phrenic nerve. This technique has been
described in pediatric and young adult patients with more
complex arrhythmia substrates. Using these techniques,
the radiation time for this patient’s procedure was limited
to just over 1 min. Minimizing radiation exposure is
especially important to patients who are relatively young
because of concern for long-term sequelae, notable of
which being neoplasm development.

Traditional ablation therapy utilizes radiofrequen-
cy energy; however, the improved safety profile of
cryothermal ablation makes the utilization of this
technique increasingly common and the treatment of
choice for many patients with complex arrhythmia
substrate. Of particular interest is the attribute of lesion
reversibility that comes with the use of cryotherapy. In
this particular case, the proximity of our patient’s atrial
ectopic tachycardia to the sinus node and right phrenic
nerve presented increased risk of injury to those
structures, therefore prompting the use of cryothermal
energy.

**Conclusion**

This case highlights the impact of multiple technologies
as well as unique access strategies often required when
treating CHD patients with arrhythmias. When appro-
aching an adult with CHD, one should consider the
use of 3D mapping for improved localization of the
arrhythmia, identifying vital structures, and minimizing
radiation exposure to the patient. Unique vascular access
may need to be considered, and can still be utilized with
3D mapping techniques. Owing to the varied anatomy
in many patients with CHD, cryotherapy should be
considered when the arrhythmia substrate is located
near an area with increased likelihood of collateral
damage to normal structures.

Figure 3: Right lateral and anterior-posterior views of the electroanatomic three-dimensional (3D) map using 3D NavX. The yellow lesions are the successful site. Visualized catheters include the esophageal (Eso) catheter in orange, the combined His/ RVA (right ventricular apex) catheter (His labeled green, RV blue) and ablation catheter (Abl). There is a shadow of a
cryocatheter in the posterior superior vena cava at a location of phrenic nerve capture with pacing. Phrenic nerve capture was
noted just posterior to the successful site, and the sinus node was just anterior to the successful site.
References


