INNOVATIVE TECHNIQUES

COMPLEX CASE STUDY

Ablation of a Focal Atrial Tachycardia in a Patient with Univentricular Heart Palliated with Total Cavopulmonary Connection

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ABSTRACT. We describe a case of an atrial tachycardia in a patient with univentricular palliation with total cavopulmonary connection (TCPC) ablated within the extracardiac conduit. The pulmonary vein (PV) atrium was accessed via a novel technique: transcatheter puncture guided by intracardiac ultrasound with pre-dilation of the puncture site avoiding the step of balloon septoplasty as it has been previously reported. On later follow-up, he presented with a different atrial tachycardia, a large clot in the conduit, and modest improvement of his ventricular function. This case also illustrates the complexity of managing adult patients with complex congenital heart disease.

KEYWORDS. atrial tachycardia, catheter ablation, cardiac mapping, intracardiac ultrasound, adult, congenital heart disease.

Introduction

A 24-year-old man presented to the emergency room (ER) with recurrent palpitations and pre-syncope. A 12-lead electrocardiogram (ECG) in the ER revealed an atrial tachycardia with positive p waves in inferior and precordial leads, and negatives in AVL with 2:1 AV conduction (Figure 1a). He had five electrical cardioversions in 2 months for this arrhythmia and subsequently failed β-blockers, calcium channel blockers, and amiodarone. His original diagnosis was double inlet ventricle, right ventricular hypoplasia, atrial septal defect, d-transposition of great vessels, and severe pulmonary stenosis. He underwent total cavopulmonary connection (TCPC) via an extra cardiac conduit and ligation of a prior bidirectional Blalock–Taussing shunt at 8 years of age. A computed tomography scan of his chest with contrast is shown in Figure 1b. Because he did not have a history of congestive heart failure or an atrial clot on transesophageal echocardiogram (TEE), consideration was made for percutaneous catheter ablation instead of a surgical approach. The options for ablation included percutaneous ablation via transseptal conduit puncture versus direct transthoracic access ablation. Since he did not have atrial thrombus on TEE and a percutaneous approach was more desirable, we decided to proceed with a percutaneous approach.

Catheter ablation procedure

After written informed consent was obtained and the patient was under general anesthesia, the following catheters were placed in the heart: a duo-decapolar catheter (Blazer Dx-20, Boston Scientific, Minneapolis, MN) was placed from the common femoral artery retrograde into the ventricle and looped back into the anatomical left (pulmonary vein (PV)) atrium. A decapolar catheter and an 8F intracardiac ultrasound catheter (ICE) were positioned within the baffle (Viewflex, St Jude Medical, St. Paul, MN) from the right femoral vein. The patient came to the laboratory in tachycardia. To precisely map this tachycardia the stiff Gore-Tex extracardiac conduit was punctured
to access the PV atrium. Briefly an 8.5F Agilis sheath (St Jude Medical) and a Brokenbrough (BRK-1) transseptal needle were placed superiorly within the extracardiac Fontan conduit and pulled down until the tip of the dilator was at the level of the PV atrium. Tenting of the conduit’s wall with the needle was observed on ICE, but the needle did not cross by applying gentle pressure (Figure 1c). Electrical cautery set at 20 watts of power in cut mode was applied to the end of the BRK-1 needle in unipolar fashion to successfully enter the PV atrium. A 0.14 guidewire was placed through the BRK-1 needle and looped back into the PV atrium (Figure 1d). However, the 8.5F sheath did not advance over the wire. An 8.5F dilator was advanced back and forth into the PV atrium several times over the wire. Then the 8.5F sheath and dilator were exchanged for an 8F Mullins that crossed the conduit with slight resistance (Figure 1d). The patient’s hemodynamics were stable, and no effusion/hemothorax was noted on ICE.

A 4-mm-tip cool-tip irrigated catheter (St Jude Medical) was placed in the PV atrium through the Mullins sheath for mapping and ablation. Intravenous heparin with an activated clotting time of 300 s was maintained during the procedure. Manipulation of the ablation catheter within the PV atrium was relatively easy. Electroanatomical activation mapping (NavX, St Jude Medical) of this tachycardia showed a centripetal map with the earliest activation site at the roof of the PV atrium (Figure 2a, b). This tachycardia could not be entrained. Several applications at the earliest PV atrial site failed. After extensive mapping within the PV atrium the ablation catheter was placed within the extracardiac conduit. Careful mapping revealed an early site at the edge of the conduit where it connected to the PA in which the atrial electrograms preceded the P wave by 43 ms (Figure 2c). Ablation at this site eliminated the arrhythmia within 5 s. Owing to the high incidence of typical flutter in patients with repaired congenital heart
Cavotricuspid isthmus ablation was performed with ablation at the PV atrium and the extracardiac conduit confirming bidirectional block at the end of the procedure in a conventional fashion. Programmed stimulation post ablation did not induce any other arrhythmia. There was a small intracardiac shunt right to left depicted on the ICE without hemodynamic consequences at the end of the procedure. He was discharged home 2 days post procedure in good condition.

Clinical follow-up

He was arrhythmia free for 3 months but presented to the ER in heart failure and atrial tachycardia with a slower cycle length and different P wave morphology to the one ablated. A large clot on the conduit seen on TEE precluded us from performing cardioversion (Figure 2b). He admitted to not taking warfarin. He converted spontaneously to sinus rhythm 14 h later and was placed on amiodarone and unfractionated heparin, and was switched to warfarin before discharge.

At 6 months’ follow-up he was in sinus rhythm with mild improvement of his LV function (now 35% from 20%). There was still a clot on the conduit with evidence of subclinical pulmonary embolism on a chest CT. There was no worsening of the atrial shunt.

At 9 months’ follow-up he was admitted to the hospital in SR with fever, hemoptysis, and cavitary lung lesions on a chest CT and diagnosed with necrotizing pneumonia. He

Figure 2: Panel A: A heart shell from the RAO projection generated with Ensite Navix is shown. A = atrium; Ao = aorta; IVC = inferior vena cava; V = ventricle. Panel B: electroanatomical activation map of atrial tachycardia from the left posterior lateral view; white and purple depict early and late atrial sites respectively. Panel C: Surface ECG leads (I,II,AVF,V1), intracardiac electrograms from the duodecapolar catheter (1-2 to 7-8) and the ablation catheter (MAP-p and MAP-d) at the successful ablation site are shown.
was discharged home in a stable condition after aggressive treatment with antibiotics.

Discussion

Surgically univentricular palliation has undergone several modifications since the initial Fontan procedure. Total atrial PV connection (TAPV) and lately TCPC are the most common contemporary variation of the Fontan technique with a decrease in post-procedure atrial dilation and subsequently atrial arrhythmias.\(^1\) Despite these surgical modifications, supraventricular tachycardia (SVTs) still remains a frequent finding (16–40%) and contributes to ongoing morbidity and mortality.\(^2\) The presence of suture lines, anatomical barriers, i.e. crista terminalis, vein orifices plus atrial dilatation, and fibrosis that occur over time set the perfect “milieu” for re-entrant tachycardia in this population.

Focal atrial tachycardias are less characterized and tend to be anatomically distributed around the appendage, crista terminalis, cava orifices, surgical conduit, and the PVs. In order to achieve successful catheter ablation of SVTs in patients with TCPC, access to the surgically excluded atrium from the systemic circulation is frequently needed. Considering that manipulation of the ablation catheter through the tortuous retrograde aortic approach is at the least challenging and in most cases insufficient for ablation of atrial tachycardia, different ways to enter the PV atrium have been developed. Direct transthoracic percutaneous puncture of the atrium without sternotomy has been reported with successful outcomes.\(^3,4\) Risk of post-procedure tamponade, pneumothorax, and hemothorax was substantial, mandating rapid surgical availability. A hybrid procedure, in which the surgeon enters the chest, performs an atriotomy on the beating heart, and helps with sheath entry into the atrium through a purse string incision, has also been described.\(^5\) Poor catheter maneuverability and the need for constant light traction on the pursing string were described to be limiting factors. Dave et al.\(^6\) recently reported access of the PV atrium via the conduit using a transseptal needle and radiofrequency (RF). This case is quite similar to our case with the difference being that they performed balloon angioplasty after puncturing the baffle. In our case dilation of the puncture site was done by up-sizing dilators.

Manipulation of the ablation catheter was easy and there was no acute or late significant baffle–PV atrium shunting that required device closure acutely and on 9 months’ follow-up.

Considering improved life expectancy in patients with congenital heart disease that are surgically palliated, the incidence of atrial arrhythmias, often poorly tolerated, is expected to increase. Our technique, though challenging,
offers an easier and much simpler approach than surgery to ablate these patients. The lack of need for balloon angioplasty makes this procedure even more attractive for the electrophysiologist already familiar with trans-septal puncture. In addition, we provided a longer follow-up of up to 10 months without the development of any negative hemodynamic consequences from the baffle puncture. Nonetheless, further studies are needed to establish the safety and feasibility of this procedure. Because of its ease and the fact that successful ablation of this tachycardia was within the baffle, we believe that detailed mapping of the extracardiac conduit should be performed first before entering the PV atrium. This case report also emphasizes the need for anticoagulation before and after the procedure as well as the importance of the patient’s compliance with medications.

Summary

Successful ablation of an atrial tachycardia in a patient with univentricular heart palliated with TCPC ablated within the baffle is reported. Transconduit puncture and RF ablation with dilation of the puncture site with several passes of larger dilators were used to access the PV atrium. On later follow-up, he presented with a different atrial tachycardia, a large clot (Figure 3) in the conduit from anticoagulation non-compliance, and modest improvement of his ventricular function. This case also illustrates the complexity of managing the adult patient with repaired congenital heart disease in which catheter ablation is often useful but nonetheless a palliative procedure.

References