Case Report

Pulmonary Damage following Right Ventricular Outflow Tachycardia Ablation in a Child: When Electroanatomical Mapping Isn't Good Enough

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ABSTRACT

A 14-year-old female was referred for severe pulmonary valve insufficiency after undergoing radiofrequency ablation for a right ventricular outflow tract tachycardia that originated in the proximal pulmonary artery at 10 years of age. Clinical records indicated that ablation was guided solely by electrograms and electroanatomical mapping. Due to myocardial tissue extensions, mapping failed to identify the level of the pulmonary valve annulus, which resulted in delivery of energy on the valve proper and into the pulmonary artery. She developed severe pulmonary valve insufficiency and moderate proximal pulmonary artery stenosis necessitating intravascular stent placement 4 years later with an associated transcatheter valve. Although the non-fluoroscopic approach during ablation has gained wide acceptance for use in children, this report highlights the benefits of adjunctive imaging to identify the precise location of the pulmonary valve when ablation therapy is contemplated in the right ventricle outflow tract.

KEYWORDS

Right Ventricular Outflow tachycardia, pulmonary artery, radiofrequency ablation, non-fluoroscopy ablation, electroanatomical mapping

INTRODUCTION

Ventricular tachycardia (VT) is rare in children, especially without underlying cardiac disease.(1, 2) The majority of idiopathic VTs originate in the right ventricular outflow tract (RVOT) and can be effectively treated with radiofrequency (RF) ablation.(1, 3) However, in some circumstances, a unique situation exists in which the arrhythmogenic focus may originate distal to the pulmonary valve. It has been speculated that myocardial remnants in the main pulmonary artery (PA) trunk provide the

arrhythmia substrate.(4-7) To date, there have been no reported major complications associated with RF ablation along the RVOT or within the PA. Although fluoro-less ablations eliminate radiation-related concerns and have been shown to be effective in children for some arrhythmias, reliance solely on electroanatomical mapping is not always advisable. We report a case of a child with the diagnosis of an RVOT tachycardia, treated with RF ablation inadvertently applied inside the PA guided by electroanatomical mapping. The procedure resulted in severe pulmonary valve damage and significant proximal pulmonary artery stenosis requiring intervention.

CASE REPORT

A 14-year-old female was referred for severe pulmonary insufficiency and proximal pulmonary artery stenosis four years after ablation for a RVOT tachycardia. From available records, she had a history of frequent premature ventricular contractions (PVCs) starting at 7 years of age. The inferior QRS axis and the left bundle branch block pattern were compatible with an RVOT location. **(Fig. 1A)** She was initially treated medically, but after three years, symptoms worsened and a 24-hour Holter monitor showed a PVC-burden of more than 35%. During an exercise stress test, ventricular ectopy continued, culminating in a short runs of VT (167 beats/minute) associated with dizziness. The initial echo/Doppler study showed normal biventricular systolic function with a normal pulmonary valve without stenosis or insufficiency. Due to the high PVC-burden and exercise-induced VT, an electrophysiology/ablation study was performed.

From available records, the electrophysiology mapping was performed using the EnSite system (Endocardial Solutions/St Jude Medical, St Paul, MN) and intracardiac electrograms. Programmed stimulation showed normal sinus and atrioventricular nodal functions, with no inducible atrial arrhythmias. However, ventricular ectopic complexes and runs of VT were induced. **(Fig. 1B)** Left ventricular mapping was unremarkable. Activation mapping revealed the arrhythmia location to be along the presumed distal RVOT, proximal to the valve. Pace-mapping of the region effectively localized the ectopic origin with a 12/12 match. RF ablation was then performed at the mapped focus using an 8mm tip Sapphire catheter (St. Jude Medical, St Paul, MN). Fluoroscopy was not used nor

were ventricular/pulmonary artery angiograms. Concomitant echocardiography was not performed. A total of 14 RF ablations with standard myocardial temperatures between 50-70ºC were delivered with effective elimination of all ectopy. (Fig. 2) Post-ablation pacing protocols failed to re-induce any arrhythmias.

The following day, an echocardiography with Doppler study revealed abnormalities of the pulmonary valve and proximal PA with a trace pericardial effusion. The PA appeared narrow and echo-bright at the tip of the pulmonary valve leaflets. Doppler analysis showed pulmonary valve insufficiency. Four years later she remained arrhythmia-free but was referred for an interventional pulmonary artery stent and valve insertion due to progressive fatigue and shortness of breath (NYHA class II-III). A pre-stent echocardiography with Doppler study showed a dilated right ventricle, stenosis in the proximal PA (peak gradient of 51 mmHg) and free pulmonary valve insufficiency. **(Fig. 3).** A pulmonary artery angiogram performed at the time of the interventional study confirmed the site of ablation-induced narrowing to be distal to the valve.

DISCUSSION

RVOT tachycardia, in the pediatric population, typically has an excellent prognosis and is amenable to ablation therapy. A study of children less than 16 years of age in a large catchment area by Roggen et al. reported only 27 patients, with spontaneously occurring episodes of VT (VT incidence of 1.1 episodes/100,000 childhood years).(2) The tachycardia has been shown to be adrenergically-mediated and sensitive to intracellular calcium levels with the substrate mechanism due to cyclic adenosine monophosphate (cAMP)-mediated triggered activity dependent on delayed after depolarizations.(8) To our knowledge, this report is unique in two aspects: young patient age (10 years old) at the time when ablation was performed in the pulmonary artery and the rare association of the arrhythmogenic focus localized in the proximal pulmonary artery due to myocardial extensions. It also highlights a potential complication of RF ablation within the PA and inaccuracies with reliance on electroanatomical mapping.

The mechanism of PA-focus of the "RVOT" arrhythmia is extremely intriguing. Animal studies have shown that the distal myocardial boundary of the outflow tract is not a stable landmark but

moves proximally over the spiraling course of the aorta-pulmonary trunk until the valves develop and septation occurs.(9) The distal myocardial boundary of the outflow tract then continues to regress and the conus septum becomes largely incorporated into the right ventricle. (9) In some individuals, however, there can be incomplete regression resulting in a myocardial sleeve that extends into the pulmonary artery, which may remain connected to the RVOT. (10, 11) Liu et al. demonstrated that myocardial extension in the PA is, in fact, very common (92%) even in normal subjects, although this extension does not always cause arrhythmias. (11) Forty-six percent (11/24 patients of RVOT arrhythmia) had arrhythmic foci located above the pulmonary valve. More recently, the incidence of a PA arrhythmia origin has been reported from 8 to 33% among patients with a left bundle branch block morphology VT. (11, 12) It is important to be aware of this possibility of a PA arrhythmia when VT from RVOT is initially suspected.

Non-fluoroscopy techniques have been effective and safe for ablation of cardiac arrhythmias with elimination of radiation exposure, especially in children. Recent meta-analysis showed that ablations performed with zero or near-zero fluoroscopy showed no difference in efficacy and safety.(13) Recently, ALARA (as-low-as-reasonably achievable) protocols using low radiation dose have shown to have markedly reduced radiation exposure, yet providing equivocal ablation efficacy and procedure safety.(14) However, that may not be the best choice in the RVOT, where electroanatomic mapping may fail to recognize the level of pulmonary valve annulus, typically determined where myocardial voltages suddenly disappear or markedly decrease. In the literature, a cutoff voltage value of 1.9 mV has been used to discriminate between subvalvular and supravalvular pulmonary positions (90% sensitivity, 96% specificity). (15) However, this is based on adult data and the assumption that myocardial signals terminate precisely at the level of the pulmonic valve annulus.(16)

Previous reports have recommended the use of angiography to locate the ablation catheter in relation to the pulmonary valve for RVOT arrhythmias.(4, 5, 7) Transthoracic or intracardiac echocardiography may provide additional real-time guidance to demonstrate the catheter position and pulmonary valve.(11) In this present case, as seen in Figure 2, the region originally labeled "RVOT" was actually the proximal pulmonary artery, ostensibly because electroanatomical mapping failed to

identify the pulmonary valve annulus due to persistence of intracardiac signals. Electrophysiological studies and catheter ablations in children do present some difficulties when compared with older adults. With the current equipment, careful studying of the anatomical location of ablation before and during the ablations using imaging modality like short fluoroscopy times, intracardiac echocardiography (ICE) or angiography as an adjunct to voltage mapping in children is recommended to decrease complications.

Complications associated with RF ablation on PA arrhythmias have not been reported in previous case reports or series. Our report is the first to demonstrate that RF ablation could damage pulmonary valve leaflets and arterial vascular walls when not cautiously performed. In this case, the detrimental effects were detected soon after the procedure and progressed further, leading to eventual referral and intervention. In addition, since the pulmonary artery is a vascular structure and different from the endocardium of the RVOT, the endothelial vascular lining is potentially more susceptible to ablation-induced tissue damages. Because of this, Sekiguchi *et al.* has recommended that RF application should be given only for a limited time with the use of lowered RF energy than if used in the RVOT proper. (7) However, that implies accurate anatomic identification.

In our case, the reason for this complication of the supra-valvular stenosis was secondary to the misidentified location of the arrhythmia focus and, therefore, misguided ablations. In addition, it can be argued that an 8mm catheter tip may not be the most appropriate size to map and ablate with high level of accuracy, especially in the outflow tract of young patients. In addition to this, the high temperature of 70ºC used in the thin musculature of the PA (even though used with the intention of ablating in the RVOT) as well as the high number of ablations (total number of 14 ablations) were probably the reasons for this unfortunate complication in this patient. It can be hypothesized that this is a failure of interpretation of the electroanatomical mapping. This case demonstrates dramatically that utmost care must be taken if RF ablation is performed near the pulmonary valve annulus, especially in the young. Compulsive mapping is crucial to confirm the location of the ablations to drastically limit the number of RF applications, as the RF energy can be quite destructive in the PAs.

CONCLUSION:

Myocardial extensions can occur into the proximal pulmonary artery and arrhythmias arising from such a focus can mimic RVOT tachycardia. Radiofrequency ablation can cause destruction of pulmonary valve leaflets and significant PA stenosis. Before RVOT/PA ablation is performed, it is mandatory to confirm the catheter location in relation to the pulmonary valve by fluoroscopy, angiography and/or echocardiography. Reliance solely on electroanatomic mapping, although accurately localizing arrhythmia foci and avoiding potential radiation-related issues, does not precisely identify valve position and can be counterproductive as well as potentially detrimental.

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FIGURE LEGNEDS

Figure 1: (A) Baseline 15-lead electrocardiogram, showing frequent monomorphic premature ventricular contractions having an inferior QRS axis and left bundle branch block morphology. (B) Intracardiac recording during the electrophysiology study, showing inducible ventricular tachycardia of the same morphology.

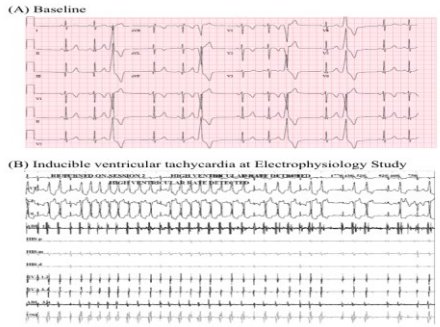

Figure 2: Anteroposterior (left) and lateral (right) views of the EnSite® electroanatomical map obtained at the time of ablation with energy delivery demarcated by dots. The region labeled "RVOT" was erroneously thought to be the right ventricular outflow tract based on activation sequences. As can be seen, the ablation was in the proximal pulmonary artery and not the sub-pulmonary outflow myocardium. SVC: superior vena cava; RA: right atrium; V: ventricle; IVC: inferior vena cava.

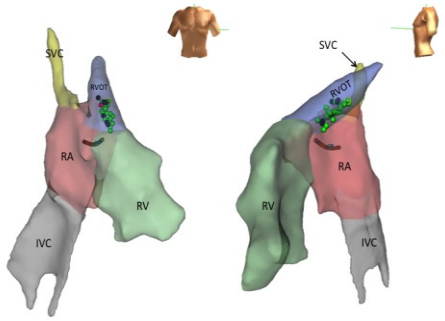

Figure 3: Parasternal echocardiographic long axis view showing the right ventricular outflow tract (RVOT) and main pulmonary artery (MPA). (A) Initial appearance before ablation, illustrating a normal valve and outflow; (B) Comparable view with Doppler taken 4 years later showing increased echogenicity at the valve (arrow) and flow alteration. (C) Comparable view showing a stent placed (arrow) in the area of pulmonary stenosis.