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Brief Report

Radiofrequency ablation of a left-sided atrioventricular pathway in a patient with Marfan syndrome

Anji T. Yetman, Joel Temple, Christopher C. Erickson

Department of Cardiology, Arkansas Children's Hospital, University of Arkansas For Medical Sciences, Little Rock, AR, USA

Abstract Supraventricular tachycardia is a documented feature of Marfan syndrome. The safety and efficacy of radiofrequency ablation in this population of patients, however, has not been reported. We report on the successful use of radiofrequency ablation utilizing a trans-septal approach for the treatment of supraventricular tachycardia produced by an accessory muscular atrioventricular connection in a patient with Marfan syndrome.

Keywords: Wolff-Parkinson-White syndrome; supraventricular tachycardia; interventional catheterization

Isolated patients with atrial arrhythmias, including atrial fibrillation and atrioventricular tachycardia, have been documented in the setting of Marfan syndrome.[1–4] As far as we are aware, however, there is no data on the overall prevalence of such arrhythmias. While initially thought to affect primarily only the aortic root, the fibrillin defect, intrinsic to Marfan syndrome, is now thought to affect myocardial structure and performance.[5] It has been postulated that the fibrillin defect may also be responsible for altered atrioventricular conduction in this group of patients.[1] Rare instances of delayed atrial–His intervals have been reported, and complete heart block has been noted.[6] The mechanism of such alterations in conduction remains unknown. While radiofrequency ablation is employed commonly for treatment of supraventricular tachycardia in children in general, as far as we know this procedure has not previously been used in a child with Marfan syndrome. The safety and efficacy of the procedure remain unknown. We report here the successful use of radiofrequency ablation in a child with orthodromic reciprocating tachycardia and a concealed accessory bypass tract.

Correspondence to: Dr Anji Yetman, Department of Cardiology, Arkansas Children's Hospital, 800 Marshall Street, Little Rock, AR 72202, USA. Tel: 501 320 1479; Fax: 501 320 3665; E-mail: YetmanAnjiT@uams.edu

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Case report

A 12-year-old boy was followed since the age of 2 years with a diagnosis of narrow complex tachycardia, albeit with no evidence of pre-excitation on his surface electrocardiogram. He was managed with digoxin, which provided reasonably good control until he reached the age of 10 years. At this time, breakthrough episodes of tachycardia became more frequent. He was then managed with beta-blockade, but had persistent episodes of breakthrough, and was unable to tolerate the medication without developing unacceptable symptoms of fatigue. During the course of follow-up, the patient was diagnosed with Marfan syndrome according to the revised criterions established by de Paepe and his colleagues, the so-called Gent criterions.[7] Clinical features included tall stature, an abnormal ratio of the upper to the lower body segment, arachnodactyly, ectopic lenses, and dilation of the aortic root. Because of persistent episodes of supraventricular tachycardia, the family requested radiofrequency ablation.

The child was taken to the cardiac catheterization laboratory, and an electrophysiologic study performed. There was baseline sinus rhythm, with no pre-excitation. Baseline intervals were normal, including a normal His–Ventricular interval of 41 ms. There was no evidence of dual atrioventricular nodal physiology. Ventricular–atrial conduction was eccentric and nondecremental, suggesting retrograde conduction

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over an accessory pathway. Ventricular premature stimulation advanced the atrial electrogram without advancing the His spike, and without affecting the sequence of retrograde atrial activation, confirming the presence of an accessory pathway. The mitral annulus was mapped using a trans-septal approach. The earliest retrograde atrial activation was located in left anterolateral position, where the atrial and ventricular signals were fused. A radio frequency lesion was delivered, with instant disappearance of ventricular–atrial conduction. There was no return of ventricular-atrial conduction during one hour of observation.

Because of intolerance to beta-blockers, the patient was maintained on an inhibitor of angiotensin-converting enzyme as prophylaxis against progressive dilation of the aortic root and dissection. He has had no recurrence of tachycardia in the 6 months following the procedure. Follow-up 24-h monitoring has revealed no episodes of supraventricular tachycardia, atrioventricular nodal block, or disturbances of interventricular conduction.

Discussion

Atrial arrhythmias are known to occur in both children[2] and adults[3] with Marfan syndrome. Despite such reports, we could find no instance of radiofrequency ablation being used to treat such arrhythmias in this group of patients. While radiofrequency ablation is recognized as a safe and effective procedure for treatment of atrioventricular tachycardia in children,[8] concern over the effect of a structural abnormality in fibrillin on both properties of conduction and myocardial integrity in patients with Marfan syndrome may raise theoretical issues as to the safety of this procedure. If the structural integrity of the myocardium is affected, as it is in the aortic root, concern regarding cardiac perforation may prohibit such a procedure. Electrophysiologic study prior to radiofrequency ablation in our patient confirmed the presence of normal intracardiac intervals. Radiofrequency ablation occurred without untoward event. We have shown, therefore, that radiofrequency ablation can be a safe and effective option for treating patients with Marfan syndrome in whom supraventricular tachycardia is the consequence of existence of an accessory muscular atrioventricular connection. The long-term effectiveness of the procedure, and its effect on myocardial structure, remain to be seen.

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