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# Atrial Ectopic Tachycardia in a Patient With Marfan Syndrome

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## ABSTRACT

The fibrillin defect central to Marfan syndrome is believed to affect myocardial conduction and predispose affected patients to various arrhythmias, including ventricular tachycardia, atrial fibrillation, and atrioventricular nodal reentry tachycardia. Here we describe an adult Marfan patient with atrial ectopic tachycardia. To our knowledge, this is the first reported case of atrial ectopic tachycardia in the setting of Marfan syndrome.

## CASE REPORT

A 54-year-old woman who was diagnosed with Marfan syndrome at 12 years of age has been followed since diagnosis for mild progressive dilation of her aortic root, mild aortic regurgitation, and trace mitral valve prolapse, for which she was taking atenolol. She presented with the complaint of palpitations that were short-lived and were not associated with exercise. A Holter monitor revealed frequent episodes of nonsustained atrial ectopic tachycardia at a rate of approximately 150 bpm (Figure).

The patient elected to have a radiofrequency catheter ablation under conscious sedation. She was taken to the electrophysiology laboratory for attempted identification of her tachycardia. Her baseline rhythm was sinus with normal intervals. Despite vigorous burst and programmed atrial stimulation with and without administration of isoproterenol, she had no inducible tachycardia, and the procedure was terminated.

Following the electrophysiology study, she was admitted to the hospital and begun on a regimen of sotalol, 80 mg twice a day. She has had no recurrence of tachycardia for the past year as evidenced by symptoms and Holter monitoring.

## DISCUSSION

Marfan syndrome is a systemic disorder of connective tissue resulting from a mutation in the fibrillin-1 (*FBN1*) gene located on chromosome 15q2. Its incidence is approximately 2 to 3 cases per 10,000 individuals. Approximately 75% of cases are familial and inherited in an autosomal dominant fashion, with the remaining 25% resulting from de novo mutations. The disease shows no predilection for race or sex.[1]

The defective synthesis of fibrillin-1, a major structural component of the extracellular matrix, leads to a variety of abnormalities in the skeletal, cardiovascular, ocular, nervous, and pulmonary systems. Signs and symptoms of Marfan syndrome include strikingly disproportionate overgrowth of long bones, scoliosis, arachnodactyly, dolichocephaly, highly arched palate, pes planus, atrioventricular valve dysfunction, aortic aneurysm, myopia, astigmatism, ectopic lentis, dural ectasia, and spontaneous pneumothorax. The diagnosis of Marfan syndrome is a clinical one, best made by physicians experienced with the disorder. Genetic testing is available but remains limited in its ability to establish the diagnosis because of the large number of mutations identified and the fact that mutations of *FBN1* are not unique to Marfan syndrome.[1–3]

Aortic catastrophe has long been considered the most feared cause of early mortality in patients with Marfan syndrome. In these patients, the aorta shows reduced compliance and distensibility. Over time, it stiffens and becomes excessively dilated, and this eventually leads to aneurysm and dissection.[3,4] With

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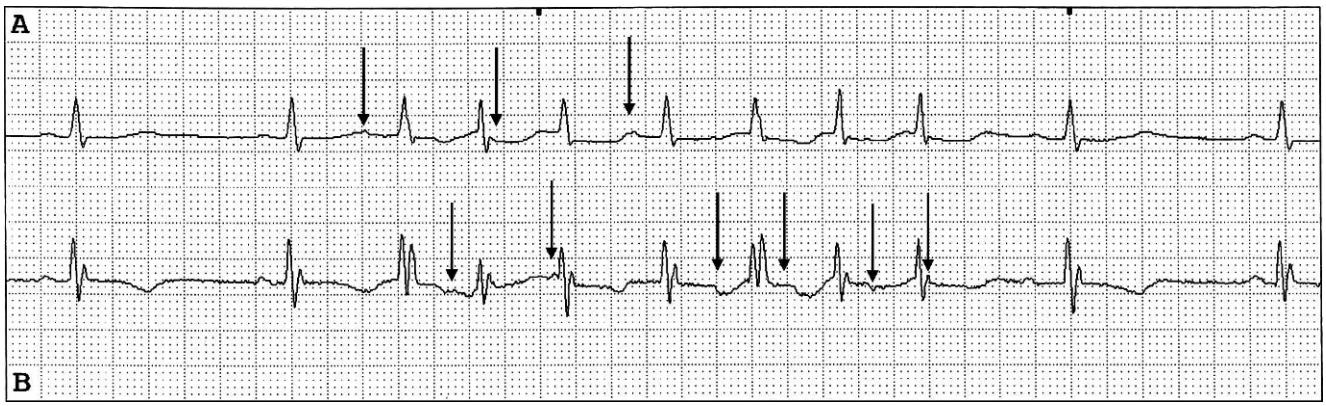


**Figure. Strip of atrial ectopic tachycardia with arrows highlighting atrial contractions.**

current routine monitoring of aortic dilation, however, patients are able to receive prophylactic aortic root replacement and can largely avoid this fate.[1,4] Thus, despite the ubiquitous presence of aortic dilation in the Marfan population, mortality secondary to aortic rupture has greatly diminished. This decrease has increased the awareness of other causes of sudden death in this population, including malignant arrhythmias. Sudden cardiac death attributable to an arrhythmia has a reported incidence of between 4% and 12%, exceeding that of aortic rupture in some studied populations.[4,5] In those patients who have received prophylactic aortic root replacement, sudden death attributable to arrhythmias is second only to rupture of the aortic root as the major cause of long-term mortality.[4,6]

Atrial and ventricular arrhythmias occur with increased incidence in patients with Marfan syndrome for reasons that are poorly understood. It is thought that the fibrillin-1 defect affects myocardial conductivity and predisposes this population to the generation of arrhythmias.[7,8] Marfan patients have a number of structural heart alterations that occur over the course of their disease and predispose to arrhythmias, including aortic dilation, mitral valve prolapse, and left atrial and ventricular enlargements.[5,9] Importantly, ambulatory monitoring studies of patients with Marfan syndrome have indicated that ventricular ectopy occurs with greater incidence than in controls and occurs without the presence of the previously mentioned structural changes.[10] Ambulatory studies have also shown conduction delays and repolarization abnormalities in the Marfan population.[5] Those patients who develop malignant ventricular arrhythmias are more likely to have significant mitral valve prolapse along with repolarization abnormalities.[5,10]

Atrial arrhythmias also occur with increased frequency in patients with Marfan syndrome. Atrial arrhythmias do not carry the same risk of sudden death as ventricular arrhythmias but are still clinically significant. Atrial fibrillation is commonly seen in this population and carries the risk of thromboembolism.[10] We have described here a patient with Marfan syndrome and atrial ectopic tachycardia.

The treatment of atrial arrhythmias in the Marfan population also raises some concerns. Radiofrequency ablation is routinely and safely employed to treat supraventricular tachycardia in the majority of patients. Marfan patients frequently have structural alterations, such as valve disease and intra-atrial scarring from previous surgery, making the procedure inherently more difficult. Of the reports thus far, radiofrequency ablation in this population has been both safe and effective.[5,11]

To our knowledge, this is the first documented case of atrial ectopic tachycardia in a patient with Marfan syndrome. Despite being unable to perform a radiofrequency ablation secondary to the noninducibility of the arrhythmias, the patient has been without episodes of atrial ectopic tachycardia since beginning sotalol treatment.

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