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CASE REPORT

INTERMEDIATE

CLINICAL CASE

An Extremely Rare Congenital Muscle Bundle Crossing the Right Atrial Cavity

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ABSTRACT

Muscle bundles in the right atrium are an extremely rare congenital anomaly. We report the case of a patient with 2 atrial septal defects and a large muscle bundle crossing the right atrium. Only 3 comparable cases have previously been published. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2022;4:128–132)

A 16-year-old white woman presented with recent onset of palpitations and vertigo for several minutes, mostly occurring during and shortly after exercise. She reported a self-measured heart rate of 190 beats/min during these episodes. She did not describe dyspnea or chest pain. Several years earlier, she had experienced 3 syncopal episodes, which appeared to be of neurocardiogenic nature. Otherwise, her medical history was unremarkable. She did not describe nicotine or alcohol use or drug abuse.

LEARNING OBJECTIVES

- To be aware of rare congenital anomalies like a muscle bundle freely crossing the right atrium.
- To understand the higher risk for supraventricular tachycardia in patients with congenital malformations of the right atrium.
- To appreciate the value of multidisciplinary discussion before closure of complex atrial septal defects.
- To consider combinations of different device types to close multiple atrial septal defects.

PHYSICAL EXAMINATION

Her heart rate was 80 beats/min, and her blood pressure was 107/61 mm Hg. No clinical signs of heart failure were present. Cardiac auscultation revealed fixed splitting of the second heart sound and a systolic murmur at the left sternal border.

DIFFERENTIAL DIAGNOSIS

Considering the reported symptoms and the results of physical examination, we suspected supraventricular tachycardia, possibly triggered by an atrial septal defect (ASD). As a differential diagnosis, ventricular tachycardia or tachycardia in the context of pre-excitation could not be excluded at this point.

INVESTIGATIONS

An electrocardiogram revealed a sinus rhythm of 79 beats/min with incomplete right bundle branch block

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and a corrected QT interval of 458 ms, without further abnormalities. During 7-day Holter monitoring, narrow complex tachycardias were detected (**Figure 1**), lasting for several minutes and correlating with the reported symptoms. Exercise stress testing demonstrated normal physical performance without any symptoms or arrhythmia.

The transthoracic echocardiography findings supported the suspected diagnosis of an ASD (Video 1), exhibiting a dilated right atrium (RA) and dilated right ventricle, with normal systolic function (right ventricle end-diastolic area index 13.5 cm^2/m^2, end-diastolic volume index 3D 81 ml/m^2, fractional area change 50%, tricuspid annular systolic excursion 30 mm, tissue Doppler S' 15 cm/s, right ventricular ejection fraction 3D 46%). Surprisingly, an unclear tissue structure crossing the RA was detected.

Transesophageal echocardiography (TEE) revealed 2 separate ASDs type II with predominant left-to-right-shunting: a cribriform ASD (9 × 9 mm) in direct proximity to the persistent foramen ovale and a larger ASD (20 × 13 mm) in the inferior septum (**Figure 2,** Video 2). In addition, a large contracting bundle was identified, extending from the inferior vena cava to the crista terminalis and freely suspended in the cavity of the RA (**Figure 3,** Videos 3 and 4).

An electrophysiology study revealed typical atrioventricular node re-entry tachycardia with a cycle length of 340 ms.

MANAGEMENT

As a first step, the patient underwent successful slow-pathway ablation. After heart team discussion, device closure of the 2 ASDs was planned. To enable good interleaving of the 2 devices and soft contact to the aortic root, the inferior ASD was closed with a 16-mm Amplatzer Septal Occluder, and the larger defect was treated from the contralateral groin with a 30-mm Gore Cardioform ASD Occluder (**Figure 4**). The large muscle bundle did not hinder positioning and handling of the catheters or devices. After detachment of the 2 occluders, a small (hemodynamically irrelevant) residual shunt was observed between the aorta and the superior device. The patient was discharged with aspirin 100 mg daily for 12 months and clopidogrel 75 mg for 6 months.

DISCUSSION

Muscle bundles crossing the RA are an extremely rare congenital anomaly apparently associated with ASD. Only 3 cases have been reported so far: 2 in Indian patients[1,2] and 1 in a white patient in the United States.[3] In general, atrial congenital defects are associated with arrhythmias, commonly supraventricular tachycardias.[4,5]

Congenital abnormalities, like atrial malformations, are often associated with a structural

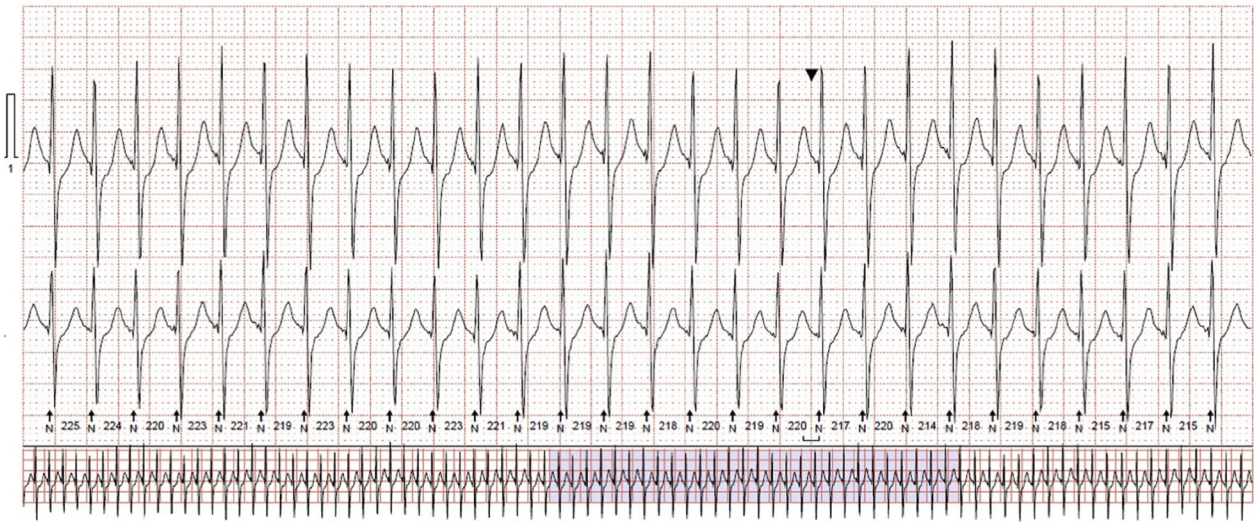
ABBREVIATIONS AND ACRONYMS

ASD = atrial septal defect

RA = right atrium

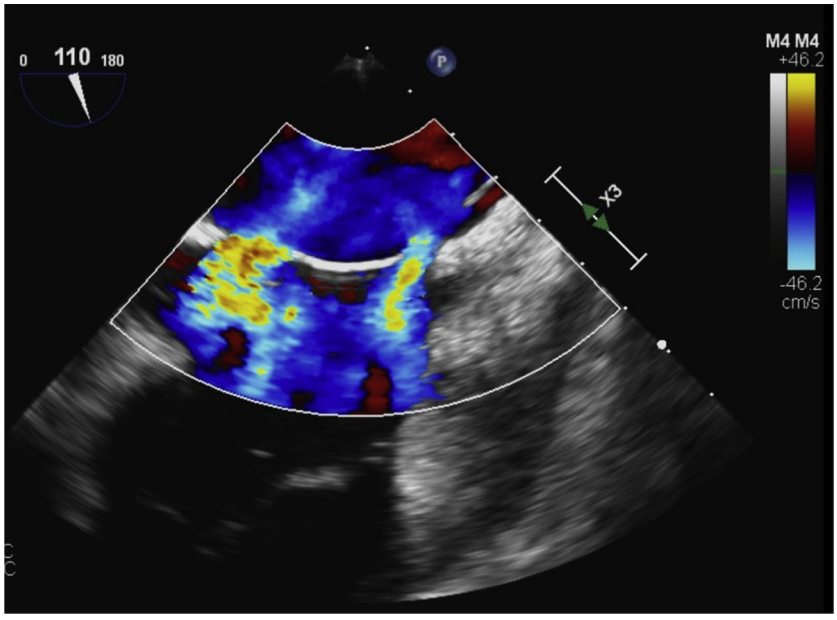
TEE = transesophageal echocardiography

FIGURE 1 Holter Tracing



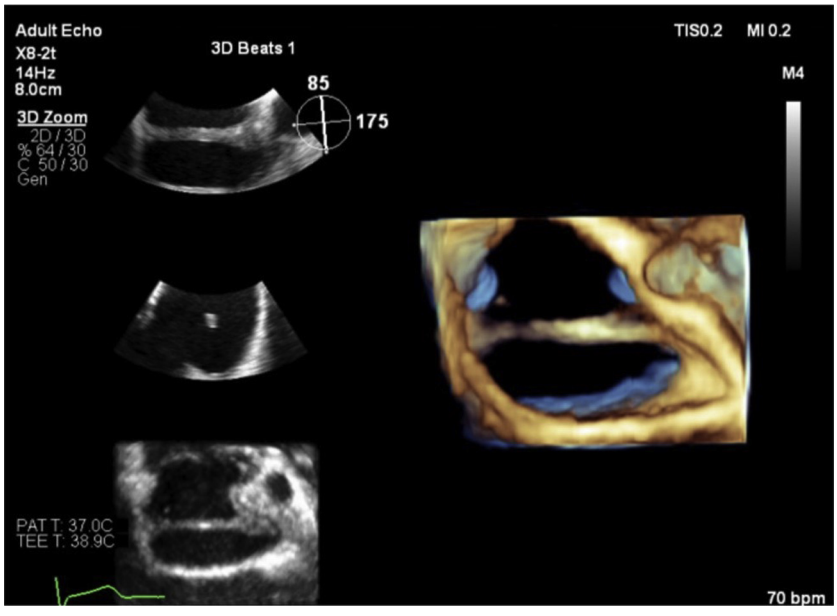
Two-lead Holter tracing with narrow-complex tachycardia of 220 beats/min.

FIGURE 2 Transesophageal Color Doppler of the Interatrial Septum



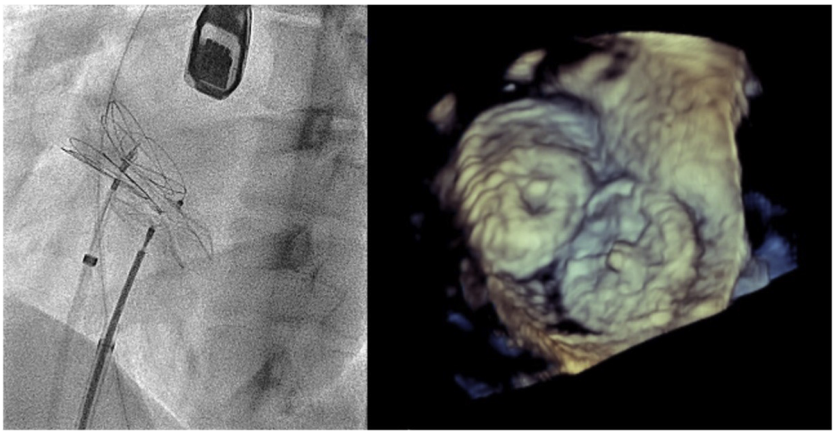
Transesophageal echocardiographic color Doppler image with 2 separate atrial septal defects type II: a cribriform atrial septal defect (9 × 9 mm) in direct proximity to the persistent foramen ovale and a larger defect (20 × 13 mm) in the inferior septum.

FIGURE 3 Transesophageal 3D Image of the Right Atrial Bundle



3D transesophageal echocardiographic reconstruction of a large contracting bundle, extending from the inferior vena cava to the crista terminalis and freely suspended in the cavity of the right atrium.

FIGURE 4 Intraprocedural Images of Atrial Septal Defect Occluders

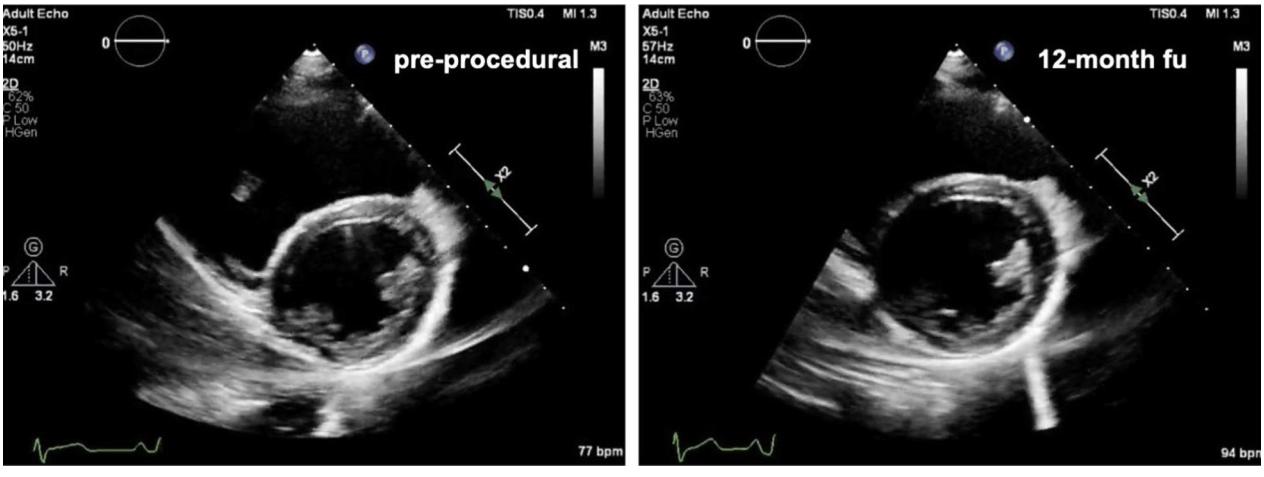


(Left) Fluoroscopic view showing a 30-mm Gore Cardioform atrial septal defect occluder in the superior atrial septal defect and a 16-mm Amplatzer septal occluder in the inferior atrial septal defects, before detachment. **(Right)** 3D transesophageal echocardiography image of the 2 occluders after detachment, view from the left atrium.

remodeling, which may contribute to supraventricular arrythmias. A chronic left-to-right shunt associated with an ASD leads to increased volume load and geometric remodeling, particularly of the RA, possibly including increased fibrotic burden.[6] The chronic volume overload furthermore leads to electrical remodeling that may precipitate through altered intra-atrial conduction and eventually the development of arrhythmias.[7] Besides, atrial muscle bundles may cause additional re-entry pathways through the bundle itself. Therefore, it seems conceivable that the increased risk for rhythm disturbances is not explained by the ASD alone, but that the muscle bundles per se also represent an arrhythmogenic substrate.

Because of the complexity of the RA anatomy, a multidisciplinary discussion involving a congenital heart disease specialist and cardiac surgeons is crucial. Furthermore, thorough imaging of all structures by TEE allows detailed preprocedural planning.[8] The exact size and position of anomalies such as the observed muscular band need to be

FIGURE 5 Transthoracic Images of Right Ventricular Size



Transthoracic echocardiographic short-axis views at the level of the papillary muscles, before and 12 months after the procedure, depicting reduction of right ventricle size. fu = follow-up.

understood to ensure proper positioning of the devices.[1,9] Furthermore, adequate evaluation for embryonic abnormalities such as cor triatriatum dexter or Chiari network should be considered. In the literature, thrombotic formations, catheter entrapment, and, especially in the case of cor triatriatum dexter, device dislodgement have been described.[10,11]

In our patient, the muscle bundle did not obstruct the access or hinder correct implantation.

FOLLOW-UP

At the 6- and 12-month follow-up visits, the patient was free of symptoms. The right heart cavities were normal in size (right ventricle end-diastolic area 10.8 cm^2/m^2, RAVI 25 ml/m^2) (**Figure 5**). TEE still showed the small residual shunt between the aorta and the superior device. Holter monitoring revealed no residual arrythmia.

CONCLUSIONS

We describe an extremely rare congenital anomaly with a muscle bundle crossing the RA cavity, associated with 2 ASDs type II. The bundle did not hamper successful percutaneous closure of the defects.

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Dr. Wolfrum has served as a proctor for Biosensors. Dr. Kobza has served as a consultant for Medtronic; and has received institutional research grants from Abbott, Boston Scientific, Biotronik, Biosense-Webster, Medtronic, and Sis-Medical. Dr. Kretschmar has received consulting fees from Medtronic, Carag AG, and Abbott. Dr. Toggweiler has served as a proctor and consultant for Abbott, Biosensors, Boston Scientific, and Medtronic; as a consultant for Shockwave, Teleflex, Medira, atHeart, and Veosource; has received institutional research grants from Boston Scientific and Fumedica AG; and has held equity in Hi-D Imaging. Dr. Stämpfli has received consulting and speaker fees from Alnylam, Amgen, AstraZeneca, Bayer, Bristol-Myers Squibb, Pfizer, and Takeda. All other authors have reported that he has no relationships relevant to the contents of this paper to disclose.

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KEY WORDS atrial septal defect, congenital heart defect, imaging

APPENDIX For supplemental videos, please see the online version of this paper.