Atrial flutter ablation in a patient with Marfanoid syndrome and anomalous cavotricuspid isthmus

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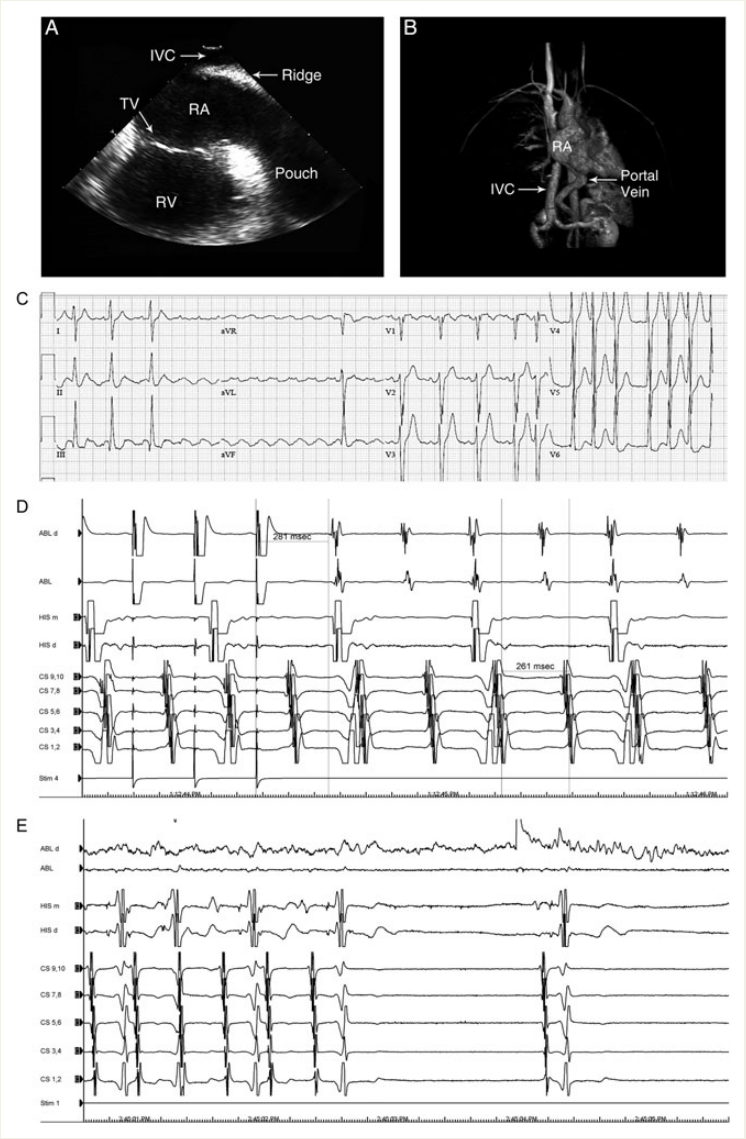

Figure 1 (*A*) Intracardiac ultrasound image of patient's RA and RV. A large 'pouch' was seen along the medial CTI, near the TV and adjacent to the CS. What was believed to be a prominent Eustachian ridge was present near the IVC. (*B*) Cardiac magnetic resonance imaging of the RA, showing an anomalous venous structure (portal vein) entering the floor of the RA. The putative prominent Eustachian ridge was in fact due to a high entry of the IVC into the lateral RA. (*C*) Twelve-lead electrocardiogram of typical atrial flutter. (*D*) Entrainment from the ablator (ABL), which was placed at the CTI, confirmed that the CTI was in the circuit (post-pacing interval—tachycardia cycle length was $<$30 ms). CS, coronary sinus; HIS, His bundle; ABL, Ablator; d, distal; m, mid. (*E*) Atrial flutter termination during radiofrequency ablation. CS, coronary sinus; HIS, His bundle; ABL, Ablator; d, distal; m, mid.

An 18-year-old man with a Marfanoid connective tissue disorder presented with counterclockwise typical flutter. During ablation, an enormous 'pouch' was visualized on intracardiac echocardiography. Ablation medial to the pouch terminated the flutter with bidirectional block. A cardiac magnetic resonance imaging revealed that the 'pouch' was an atrial anastomosis from the right atrial floor to an anomalous vein from the liver.

Clinical history

An 18-year-old man, with a Marfanoid connective tissue disorder presented with atrial flutter with 1 : 1 conduction. He was referred for electrophysiology study and ablation.

Multipolar catheters were placed in the right atrium (RA), His-bundle region, right ventricle (RV), and coronary sinus (CS). At baseline, the patient was in atrial flutter (*Figure 1C*). Entrainment from the lateral RA isthmus, proximal CS, and the cavotricuspid isthmus (CTI), showed that these areas were within the tachycardia circuit and thus confirmed the presence of typical counterclockwise CTI-dependent atrial flutter (*Figure 1D*).

On intracardiac echocardiography (ICE; AcuNav, Biosense Webster), an enormous putative 'pouch' was visualized along the medial CTI, near the tricuspid valve (TV), and adjacent to the CS (*Figure 1A* and *Video*). Using a 3.5 mm Thermocool ablation catheter (Biosense Webster), ablation lateral to this pouch slowed the atrial flutter but did not terminate it (ablation settings: power control, 30–50 W, titrated to 10% impedance decreases and electrogram attenuation, with a cut-off temperature of 40°C). Ablation medial to the pouch, essentially encircling the pouch, was able to terminate the atrial flutter (*Figure 1E*) with bidirectional block. The patient has remained arrhythmia-free. A cardiac magnetic resonance imaging (*Figure 1B*) showed that the atrial 'pouch' was, in fact, an atrial anastomosis from the RA floor to an anomalous 'portal' vein from the liver (arrow). The prominent ridge seen on ICE was due to a high entry of the inferior vena cava (IVC) into the lateral RA.

Commentary

Common cardiac manifestations of Marfan's syndrome include aortic dilation and aneurysmal formation leading to aortic dissection or rupture. Other cardiac anomalies include mitral valve prolapse, calcification of the mitral annulus, dilation of the pulmonary artery, and rarely left ventricular dysfunction. To our knowledge, structural abnormalities of the atrium and venous anomalies have not been described. The pathophysiology underpinning Marfan's syndrome involves mutations of the gene encoding fibrillin-1, a protein essential to extracellular matrix formation, and leads to tissue weakness.[1]

To date, only several cases of atrial flutter and Marfan's syndrome have been reported,[2] and none have defined RA anatomy by ICE. This is an unusual case of a flutter circuit involving an abnormal CTI deformed by an anastomosis to an anomalous 'portal' vein. Because this scenario had not been previously encountered or recognized, our management of the CTI flutter ablation was inferred from other cases in which we identify prominent pouches. Pouches can sometimes be present in the CTI and can introduce challenges to ablation if they are particularly prominent. Because of poor blood flow, ablation near or within pouches can result in poor power delivery, sharp impedance increases, and possible coagulum formation.[3] Intracardiac echocardiography was a valuable tool for ablation in this case and should be considered in those patients with connective tissue disorders, as they may be predisposed to aberrant cardiac anatomy.

Conflict of interest: none declared.

References

1. Cañadas V, Vilacosta I, Bruna I, Fuster V. Marfan syndrome. Part 1: pathophysiology and diagnosis. *Nat Rev Cardiol* 2010;**7**:256–65.
2. Halawa A, Brahmbhatt V, Fahrig SA. Initial description of radiofrequency catheter ablation as treatment for atrial flutter in Marfan's syndrome: a case report and literature review. *J Interv Card Electrophysiol* 2007;**19**:37–40.
3. Asirvatham SJ. Correlative anatomy and electrophysiology for the interventional electrophysiologist: right atrial flutter. *J Cardiovasc Electrophysiol* 2009;**20**:113–22.