EP CASE EXPRESS

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

Russell Heath, Joseph Kay, and Duy Thai Nguyen*

Electrophysiology, Cardiology Division, University of Colorado, Anschutz Medical Campus, 12401 E. 17th Avenue, B-132, Aurora, CO 80045, USA

* Corresponding author. Tel: +1 720 848 0758; fax: +1 720 848 0475, E-mail: duy.t.nguyen@ucdenver.edu

An 18-year-old man, with a Marfanoid connective tissue disorder, presented with atrial flutter (Panel C). He was referred for electrophysiology study and ablation. Entrainment manoeuvres confirmed the presence of typical counterclockwise cavotricuspid isthmus (CTI)-dependent atrial flutter (Panel D). On intracardiac echocardiography (ICE; AcuNav, Biosense Webster), an enormous pouch was visualized along the medial CTI, near the tricuspid valve, and adjacent to the coronary sinus (Panel A and Video).

Using a 3.5 mm Thermocool catheter (Biosense Webster), ablation lateral and medial to the pouch terminated the atrial flutter with bidirectional block (Panel E). A cardiac magnetic resonance imaging (Panel B) showed that the pouch was, in fact, an atrial anastomosis from the right atrial (RA) floor to an anomalous ‘portal’ vein from the liver (arrow).

Cardiac manifestations of Marfan’s syndrome include aortic dilation and aneurysmal formation, leading to aortic dissection or rupture. To our knowledge, structural abnormalities of the atrium and venous anomalies have not been described. Only several cases of atrial flutter and Marfan’s syndrome have been reported, and none have defined RA anatomy by ICE. Intracardiac echocardiography was a valuable tool for ablation in this case and should be considered in patients with connective tissue disorders, as they may be predisposed to aberrant cardiac anatomy.

The full-length version of this report can be viewed at: http://www.escardio.org/communities/EHRA/publications/ep-case-reports/Documents/Atrial-flutter-ablation-in-a-patient.pdf.

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