A 25-year-old male with a history of repaired total anomalous pulmonary venous (PV) connection and persistent atrial fibrillation (AF) refractory to antiarrhythmic medication was referred for percutaneous catheter ablation. At 10 weeks of age, he underwent surgical repair for total anomalous PV connection to the coronary sinus (cardiac variant) involving deroofing of the coronary sinus and closure of the atrial septal defect. At 19 years of age, he developed symptoms of breathlessness and palpitations and was found to be in persistent AF. He underwent a cardioversion and maintained sinus rhythm for 3 months before undergoing a second cardioversion for recurrent symptomatic persistent AF. He maintained sinus rhythm for 18 months on sotalol 120 mg BID before representing with increased breathlessness on exertion. Exercise echocardiography demonstrated an increase in pulmonary arterial pressures from 34 at rest to 60 mm Hg at peak exercise together with the initiation of AF. In view of this, he underwent a cardiac MRI, which demonstrated all 4 PVs draining into a dilated PV confluence (unroofed coronary sinus), which was connected to the left atrium by a small orifice measuring 13 by 15 mm. This was associated with restricted and turbulent flow between the 2 chambers, thought to be responsible for the significant dilatation of the PV confluence, and may have provided the substrate for the initiation and maintenance of AF. He remained in AF and was rate controlled with β-blockers. At 23 years of age, he underwent an operation to relieve the obstruction between the 2 chambers. Access to the right atrium was made through the original incision from the superior to the inferior vena cava and to the left atrium through the interatrial septum. The connection between the PV confluence and left atrium was confirmed to be 15 mm in both directions. The coronary sinus was cut back further to enlarge the opening between the 2 chambers. Although initial plans were for a MAZE procedure, the surgeon felt the anatomy was difficult and was complicated further by extensive adhesions and, therefore, this was not undertaken.

After his operation, the patient was commenced on amiodarone, cardioverted, and maintained sinus rhythm for 1 year before developing symptomatic persistent AF when he was referred for catheter ablation.

Under general anesthesia, a single transseptal puncture facilitated access to the left atrium but only limited access to the circumference of the PV confluence could be achieved because of difficulty in catheter manipulation. A circumferential mapping catheter confirmed the presence of rapid atrial activity within the venous confluence (Figure); however, electric isolation could not be achieved. Therefore, he was commenced on amiodarone, and a further procedure was planned after a preprocedural computed tomographic scan for integration with Carto 3 (CARTO-MERGE, Biosense Webster). The computed tomography image demonstrated all 4 PVs draining into the dilated venous confluence, which was connected to a small left atrium (Figure).

The repeat procedure was performed in sinus rhythm. A single transseptal puncture facilitated left atrial access with a 3.5-mm Thermocool ablation catheter and Agilis sheath (St. Jude Medical, Minneapolis, MN). Image integration provided precise visualization of the anastomosis between the PV confluence chamber and the left atrium proper, such that electric isolation of the entire venous confluence from the left atrium could be performed and confirmed with a LASSO mapping catheter (Figure). Nine months after his procedure, he has had no further episodes of AF and has discontinued his amiodarone.

Although there are case reports of patients with partial anomalous PV drainage undergoing catheter ablation, this is the first report of successful catheter ablation of AF in a patient with total anomalous PV connection. These patients are more commonly at risk for the development of atrial tachycardias, with surgical scars and barriers creating conduction characteristics sufficient to sustain a reentry circuit. Reports of AF in this patient group are rare because of the lack of muscular sleeves around anomalous PVs and the absence of significant...
atrial dilatation when repaired early. Nevertheless, we found electric potentials within the anomalous PVs in this patient with the cardiac variant of total anomalous PV connection, together with evidence of rapid atrial activity within the PV confluence during AF. We hypothesized that the obstruction between the PV confluence and left atrium caused significant dilatation and stretch, which created a favorable substrate for AF initiation and maintenance and, therefore, provided the appropriate target for a catheter ablation strategy. This case highlights the importance of image integration for ablation procedures in patients with complex atrial anatomy and supports a confluence isolation strategy for management of AF in patients with repaired cardiac variant total anomalous PV drainage.

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Disclosures

None.

References


KEY WORDS: anomalous pulmonary venous □ atrial fibrillation □ catheter ablation

Figure. A and B, Cardiac computed tomography (CT)—posterior and lateral views: all 4 pulmonary veins are shown draining into a dilated pulmonary venous confluence (unroofed coronary sinus), which drains into the left atrium. C, CARTO and CT integration with representative electrograms during atrial fibrillation (AF) recorded from the left atrium and pulmonary venous confluence from the first procedure D, CARTO image showing ablation lesions around the connection between the pulmonary venous confluence and left atrium, which resulted in isolation of this chamber from the left atrium.
Catheter Ablation for Persistent Atrial Fibrillation in a Patient With Previous Repair of Total Anomalous Pulmonary Venous Connection
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