

Atrial flutter radiofrequency ablation in the setting of left isomerism and repaired single atrium: First case



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Introduction

Left isomerism is a form of heterotaxy syndrome, characterized by intracardiac defects and caval venous variations, associated with an abnormal arrangement of bronchi and abdominal organs.¹ Left isomerism is defined by symmetrical left anatomy of the atria and bronchi.² Conduction abnormalities and atrial arrhythmias are known manifestations of this condition. However, to the best of our knowledge, atrial flutter is seldom associated with this anomaly.³ We here report for the first time a case of atrial flutter successfully treated by radiofrequency ablation in the setting of left isomerism and repaired single atrium.

Case report

A 26-year-old man diagnosed with left isomerism was hospitalized owing to incessant drug-resistant palpitations. Beta-blockers and flecainide had no effect on the symptoms and the psychological impact was significant, requiring the use of antidepressants.

Cardiac defects associated with left isomerism had been diagnosed antenatally and involved a single atrium and a perimembranous interventricular communication. Surgery had been performed at the age of 2 months and consisted of closing the ventricular septal defect and constructing an atrial septum (Figure 1). The patient had 4 caval veins: a right superior vena cava in a standard position, and a left superior and 2 inferior vena cavae draining into the coronary sinus. The medical follow-up was uneventful, as ventricular functions remained correct and the patient stayed free of symptoms until the onset of persistent palpitations.

A 12-lead electrocardiogram was then performed, followed by a Holter monitoring that enabled diagnosis of

KEY TEACHING POINTS

- Performing a computed tomography scan before radiofrequency ablation in left isomerism is necessary, as vena cava variations impact catheter access to cardiac chambers. Entering the atrium via the coronary sinus limits neither catheter movement nor ablation efficacy.
- Peritricuspid isthmus-dependent atrial flutter can be the first culprit, even in complex cardiomyopathies like atrial isomerism. Ablation of a first loop can enable a second scar-related flutter to emerge.
- Sick sinus syndrome and junctional rhythm with symptomatic retrograde P waves may appear after atrial flutter ablation in left isomerism, requiring the implantation of an AAI pacemaker.

persistent 2/1 atypical atrial flutter (Figure 2). Cardiac anatomy was assessed and the absence of thrombus was confirmed by computed tomography scan prior to the ablation procedure.

Venous access to the right atrium was performed through the right femoral vein with an access to the atrium from the coronary sinus ostium. An electroanatomical mapping system (CARTO®3; Biosense Webster Inc, Irvine, CA) was used and activation mapping revealed a clockwise peritricuspid circuit (295 ms cycle length) that was confirmed by entrainment maneuvers (Figure 3A). Radiofrequency ablation on the septal side of the isthmus between the tricuspid valve and the coronary sinus ostium stopped the tachycardia, allowing a second flutter loop to emerge (280 ms cycle length). The second flutter revolved around a scar on the laterosuperior side of the right atrium and was successfully terminated by radiofrequency ablation (Figure 3B). No steerable sheath was needed, as sufficient access and stability were obtained with the ablation catheter alone. There was no inducible arrhythmia with vigorous stimulation protocols

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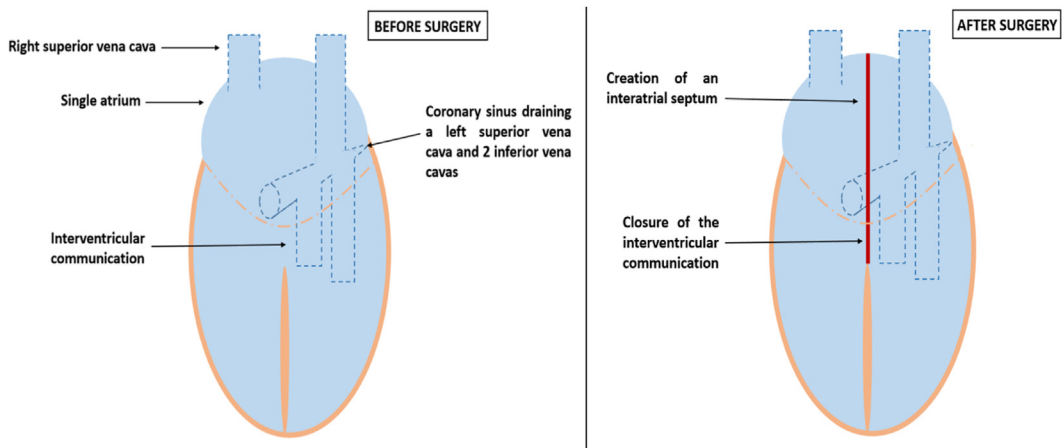


Figure 1 Anatomical scheme of the heart before and after the surgery.

and isoprenaline at the end of the procedure, and no complication arose.

Six months later, the patient developed a symptomatic sick sinus syndrome with junctional rhythm and poorly tolerated retrograde P waves at rest, requiring the implantation of an AAI pacemaker. Henceforth, the patient remained asymptomatic during 36 months follow-up and retrieved a normal lifestyle without antidepressant drugs.

Discussion

Heterotaxic syndromes and particularly left isomerism are associated with a wide range of arrhythmic and conduction diseases owing to congenital conduction tissue abnormalities associated with atriotomy scars consecutive to surgical procedures and hemodynamic overload.³ Atrial tachycardia

and junctional rhythm have been described before in patients with left isomerism; however, atrial flutter has been reported only once and appears as a rare manifestation of the disease.⁴ We here report for the first time a case of double-loop atrial flutter successfully treated by radiofrequency ablation in the setting of left isomerism and repaired single atrium. A pitfall can occur in the diagnosis workup, as the flutter may have an atypical morphology despite being peritricuspid, owing to the atrial cardiomyopathy.

This case report highlights the feasibility and efficacy of atrial flutter radiofrequency ablation in the setting of left isomerism. Despite unusual cardiac anatomy and venous access, catheter stability and maneuvering can be achieved safely. The usefulness of electroanatomical mapping systems, already proven in a wide range of clinical settings, seems of paramount importance when confronted with

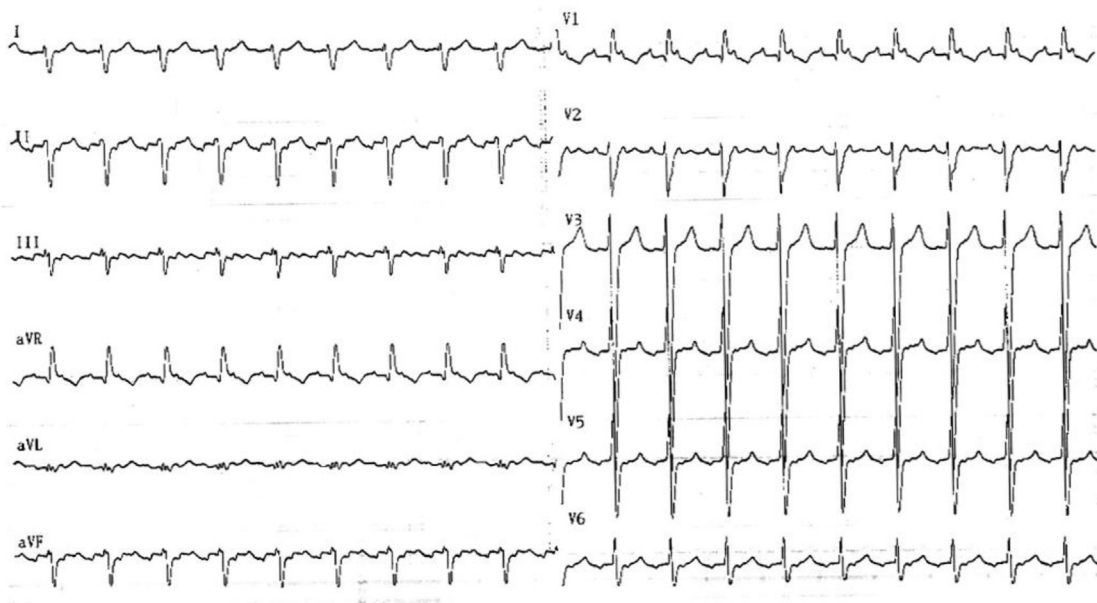


Figure 2 Electrocardiogram showing 2/1 atypical atrial flutter (10 mm/mV, 25 mm/s).

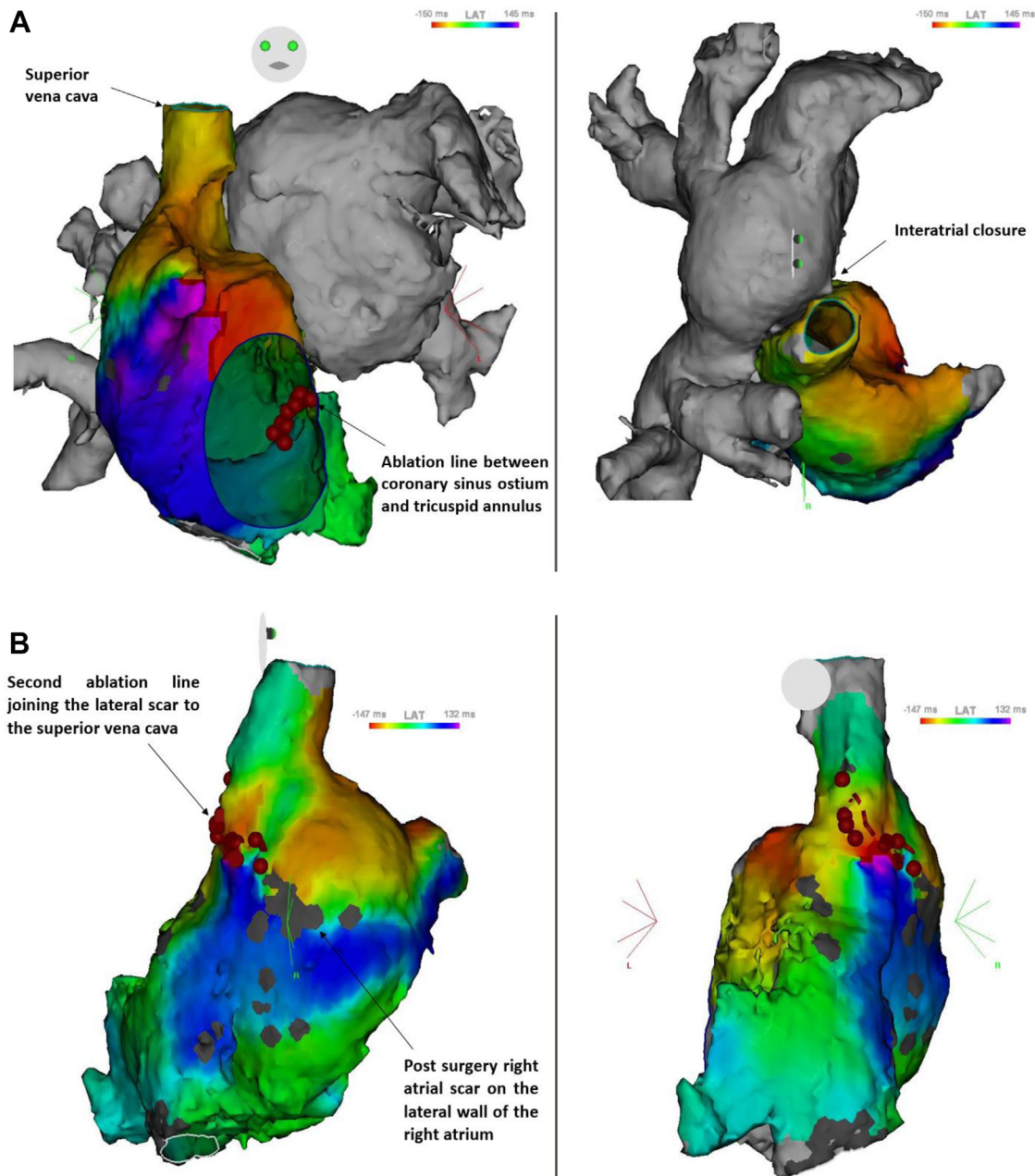


Figure 3 Three-dimensional activation map projected on computed tomography scan views of the atria: red dots represent ablation lesions, gray dots scar tissue. Local activation time (LAT) is indicated by a color scale. **A:** Clockwise peritricuspid loop (anterior and superior views). **B:** Scar-related loop (right lateral and posterior views).

congenital cardiopathy. Indeed, since unusual cardiac and venous anatomy associated with atriotomy scars complexifies reentry circuits, activation and voltage mapping are valuable tools to unveil arrhythmic mechanisms.

Conclusion

We report here a case of atypical atrial flutter in the setting of left isomerism with repaired single atrium, successfully treated by radiofrequency ablation of 2 circuits. Abnormal venous access does not hinder correct catheter mobilization and safety in this specific congenital cardiomyopathy. Electroanatomical mapping is a cornerstone in this complex

anatomical setting where multiple flutter loops can be observed.

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