**Innovative Collections**

**Complex Case Study**

**Atypical Atrial Flutter after Surgical Correction of Anomalous Pulmonary Venous Return**

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**Abstract.** We report the case of a patient who had recurrent atypical atrial flutter (AFL) despite aggressive medical therapy status following surgical correction of a partial anomalous left pulmonary venous return to the coronary sinus (PAPVR). The surgery opened the roof of the coronary sinus (CS) to the left atrium (LA) to ensure a large communication between the CS and the LA, closed the CS ostium and atrial septal defect (ASD) with a pericardial patch, and undertook right pulmonary vein isolation with cryoenergy. The patient was brought to the electrophysiology (EP) laboratory, and transeptal (TS) puncture was performed through the ASD patch followed by successful mapping and ablation of the AFL circuit between the anterior superior mitral valve annulus and the right inferior pulmonary vein (PV) os.

**Keywords.** Atrial Flutter, Catheter Ablation, Congenital Heart Disease.

**Introduction**

Partial anomalous pulmonary venous return (PAPVR) is defined as a left-to-right shunt where one or more, but not all, pulmonary veins (PVs) drain into a systemic vein or the right atrium (RA). The overall incidence of PAPVR is approximately 0.5% .

Anomalous right-sided PVs can drain into the superior vena cava (SVC), RA, inferior vena cava, azygos vein, hepatic vein, or portal vein. Anomalous left-sided PVs can drain into the left brachiocephalic vein, coronary sinus (CS), and hemiazygos vein. The severity of clinical signs and symptoms is related to the degree of left-to-right shunting and the presence of other associated cardiac and pulmonary defects. Dyspnea, fatigue, exercise intolerance, palpitations, syncope, atrial arrhythmias, right heart failure, and pulmonary hypertension may occur .

Other than sinus node dysfunction , atrial tachyarrhythmia in general and atrial flutter (AFL) in particular are very common and associated with hemodynamic deterioration, increased risk of thromboembolism, and cardiac death . In these patients, treatment with β-blockers, digoxin, and other antiarrhythmic agents, such as amiodarone, is often unsuccessful and accompanied by side effects . Thus, curative percutaneous catheter ablation has evolved as an alternative treatment modality for this type of arrhythmia.

**Case report**

A 54-year-old man presented with a history of recurrent symptomatic AFL. Five years prior to presentation, he underwent surgical correction of a PAPVR and right PV isolation for preoperative atrial fibrillation. His congenital left upper and lower PVs formed a common trunk that emptied into the CS. The surgeons opened the roof of the CS to the left atrium (LA) and performed a pericardial patch closure of his CS ostium and atrial septal defect (ASD). He underwent concomitant intraoperative cryoisolation of his right PVs. After surgery, the patient underwent uncomplicated implantation of a dual-chamber pacemaker due to complete heart block. Despite the surgical correction and ablation therapies, he suffered recurrent symptomatic atypical AFL, which was persistent despite multiple cardioversions and antiarrhythmic drug therapy.

After written informed consent was obtained, the patient was brought to the electrophysiology (EP) laboratory for...
endocardial mapping and curative ablation. Baseline transesophageal echo (TEE) and computed tomography (CT) angiogram of the PVs were obtained 1 day before the procedure. The TEE showed no evidence of LA thrombus, and the CTA PVs confirmed the findings of fusion between the left superior and inferior PVs with drainage into the CS (Figure 1).

The patient underwent an EP study with a baseline rhythm of AFL CL of 270 ms. AFL demonstrated poor entrainment mapping from the RA, including the region of the crista terminalis and the cavotricuspid isthmus (CTI). An intracardiac ultrasound echo catheter (ICE) was subsequently advanced into the RA and demonstrated that the interatrial septal patch repair from the prior surgery appeared thin and amenable to transeptal (TS) puncture.

Under ICE guidance and via an 8.5 F steerable sheath, a single TS puncture was performed. A second access to the LA was obtained through a retained guide wire approach with advancement of an additional 8-F sheath for the circular mapping catheter and 8.5-Fr steerable sheath for the ablation catheter. Unfractionated heparin was administered to achieve an activated clotting time of >300 s as soon as the needle was introduced into the LA. Initially, an electroanatomic shell of the LA and PVs was constructed using a three-dimensional mapping system, with a simultaneous LA timing activation map (Figure 3). The postpacing interval with the circular mapping catheter was equivalent to the tachycardia cycle length anterior to the base of the LA appendage. The earliest LA activation along this region of entrainment was found anterior, near the mitral valve (MV) along with fractionated electrograms near the superior edge of the pericardial patch (Figure 2). A linear lesion set was created between the anterior superior MV annulus and the right inferior PV os (Figure 3). During the course of ablation along this line, there was slowing of the AFL cycle length with subsequent AFL termination and restoration of sinus bradycardia (Figure 4). Further testing demonstrated no other inducible atrial fibrillation or AFL with burst atrial pacing, atrial extra stimuli, and isoproterenol challenge up to 20 mcg/min infusion.

Discussion

The prevalence of atrial arrhythmia in patients with repaired congenital heart disease (CHD) is approximately 15%, which is nearly three times higher than in the general population5. The high prevalence of atrial arrhythmias in patients with surgically repaired CHD can be due to diffuse pathophysiologic changes of atrial muscles, progressive atrial myopathy due to chronic volume overload of the RA, or right atriotomy or other

Figure 1: CT of the left atrium in the LAO view showing fused left inferior and superior PVs that form a common trunk dumping into the CS. Abbreviations: CS: coronary sinus; LAO: left anterior oblique; LSPV: left superior pulmonary vein; LIPV left inferior pulmonary vein; PV: pulmonary vein.
Figure 2: Intracardiac tracing showing atrial flutter with postpacing intervals equivalent to tachycardia cycle length with some alteration of the endocardial activation pattern and early fractionated electrograms signals on the ablation catheter. Abbreviations: ABL: ablation catheter; La 1-20: electrodes on the circular mapping catheter, CS: coronary sinus catheter.

Figure 3: AP view of the left atria shows a flutter line created between the anterior superior mitral valve annulus and the right superior pulmonary vein os, pink dot shows the lesion that terminated the flutter. Abbreviations: AP: anterior-posterior; MV: mitral valve; RIPV: right inferior pulmonary vein; RSPV: right superior pulmonary vein.
incisional scars\textsuperscript{7}. The majority of these patients will require extensive EP evaluation and treatment because they are highly symptomatic and are often medically refractory to treatment.

The most common mechanisms of late AFL were found to be CTI dependent. Other mechanisms in these patients include non-CTI dependent and scar-related AFLs\textsuperscript{8,9}. Therefore, during EP evaluation, the role of the CTI as the underlying mechanism of the AFL should be defined early in the catheter mapping procedure. After excluding CTI dependency, defining the other mechanisms of AFL by entrainment maneuvers in addition to three-dimensional activation electroanatomical mapping appears crucial to localize and ablate the flutter circuits in this type of patient\textsuperscript{10}.

In this patient, who presented with an atypical AFL morphology on his baseline electrocardiogram, it is unclear whether the atypical flutter was a residual or incomplete concomitant cryoablation of the LA or whether this was an intrinsic arrhythmia. The decision was made to access the LA, despite the presence of ASD patch closure, due to failed entrainment mapping from the RA, including the CTI. The TS access was performed smoothly with the application of some force on the dilator, and the sheath was advanced into the LA without the need to employ a cutting balloon. This case demonstrates that TS puncture can be safely performed in the presence of previous ASD closure patch under ICE guidance and fluoroscopy. This was previously reported in patients with pericardial patch, dacron patch, and the Amplatzer closure device\textsuperscript{11,12,13}. Thickness, shape, and the presence of calcifications or aneurysmal dilatation should be assessed on ICE and fluoroscopy before accessing the ASD patches in these patients to avoid embolization and complications.

Conclusions

Patients with AFL after repair of CHD can present with a high symptom burden and are often refractory to medication. These patients can be treated successfully with catheter-based ablation despite the anatomic complexity involved.

References