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Ablation of post-operative atrial flutter in the presence of interrupted IVC

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1. Case

A 27-year-old male with operated congenital heart disease presented with palpitations. Review of records showed that he had undergone intracardiac biventricular repair for double outlet right ventricle with subaortic ventricular septal defect and pulmonary stenosis 17 years ago. Notes also mentioned persistent left superior vena cava (SVC) draining to coronary sinus (CS) and interrupted inferior vena cava (IVC) with azygous continuation. He had atrial flutter with fast ventricular rate early after surgery, which was treated with cardioversion and oral amiodarone for a year. He was lost to follow up three years after surgery until the current admission.

Electrocardiogram at presentation showed a regular, wide complex tachycardia at 220 beats per minute with a right bundle branch block morphology and right axis deviation. Intravenous diltiazem established variable atrioventricular block and unmasked flutter waves which were positive in inferior and lateral leads and negative in lead V1 (Fig. 1). Echocardiography confirmed the presence of persistent left SVC, interruption of the IVC and drainage of the hepatic veins into the right sided atrium. Ultrasonography showed polysplenia, confirming the diagnosis of heterotaxy syndrome in the form of left isomerism. There was no residual intracardiac shunt, peak gradient across right ventricular outflow tract was 18 mm Hg and mild tricuspid regurgitation was present with estimated right ventricular systolic pressure of 30 mm Hg.

The patient was taken up for electrophysiology study and ablation with an electroanatomical mapping system (CARTO 3, Biosense Webster). Venography with contrast injection from IVC, left arm and right arm confirmed azygous continuation of IVC and bilateral SVC with a small bridging vein. A decapolar catheter was positioned in the CS through a left axillary vein puncture. The mapping catheter (Navistar Thermocool, Biosense Webster) was placed through a long, deflectable sheath (Agilis, St Jude), which was introduced from the right femoral vein and placed in the right sided atrium (RSA) through the azygous vein and SVC (Fig. 2). Activation mapping of the RSA was done during flutter at a cycle length of 250 ms. Window of interest was selected to encompass the whole cycle length with a CS electrogram as reference. A line of double potentials was seen along the lateral atrium corresponding to the atriotomy scar. At the lower end of this line there were low voltage areas and fragmented, long electrogram spanning more than 40% of the cycle length. Activation proceeded caudo-cranially posterior to this line and cranio-caudally anterior to this line (Fig. 3). Entrainment from the cavotricuspid isthmus, lateral atrium anterior to the line and lateral atrium posterior to the line showed PPI − TCL of 64, 20 and 10 msec respectively, confirming re-entry around the atriotomy and non-involvement of the isthmus. Ablation was done along the inferior border of the atriotomy scar. Flutter terminated during the second radiofrequency ablation and linear ablation was continued to connect this to the hepatic vein. Post ablation, the decapolar catheter was positioned along the lateral atrium across the line and pacing from below the line demonstrated conduction block (Fig. 4). Atrial burst pacing and up to three atrial extrastimuli did not induce any arrhythmia.

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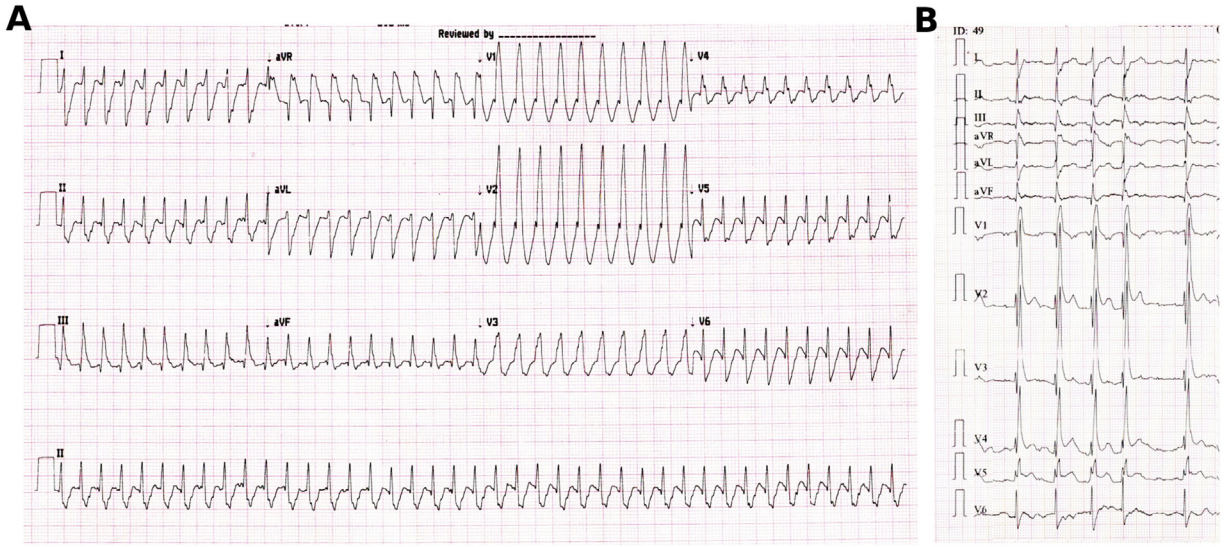

Fig. 1. ECG at presentation.
Panel A shows the 12-lead electrocardiogram at presentation. In panel B, atrial flutter is unmasked after administration of intravenous Diltiazem.

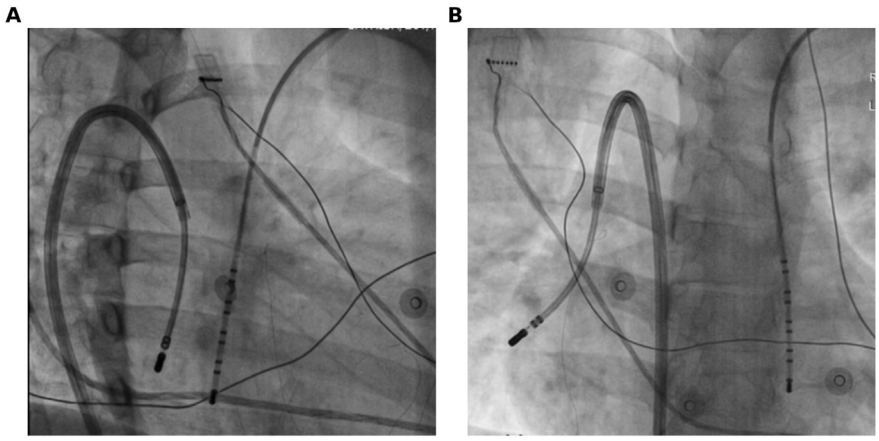

Fig. 2. Catheter positions in RAO and LAO views.
RAO (panel A) and LAO (panel B) views show the mapping catheter in the right sided atrium and the decapolar catheter within the coronary sinus. The decapolar catheter has been introduced from the left axillary vein through the left SVC. The long sheath can be seen extending from the azygous vein into the SVC and right sided atrium through which the mapping catheter is introduced.

2. Discussion

Atrial arrhythmias are an important source of morbidity in patients with repaired congenital heart disease. These are usually due to macro-reentry and often involve the cavotricuspid isthmus, but may also be non-isthmus dependent, when the atriotomy scar may serve as an anatomical obstacle sustaining the circuit [1]. It has been suggested that extending atriotomy incisions to anatomical obstacles of electrical isolation during the initial operative procedure itself may avoid development of these re-entry circuits [2]. These arrhythmias are difficult to manage medically, but respond well to ablation, especially when they are non-septal in location [3]. Activation mapping remains the key to the characterization of these arrhythmias and is combined with identification of lines of block and entrainment to identify critical isthmuses [4]. Because of the frequent involvement of the cavotricuspid isthmus in reentrant circuits, consideration may be given to ablate this even when the primary flutter is not isthmus dependent, even though we elected not to do that in this patient.

Interruption of the IVC with azygous continuation is a rare anomaly and can pose significant problems in obtaining access to the right atrium during electrophysiology procedures. Such patients often require ablation from a superior approach [5,6]. In our patient, we were able to use the femoral route with the catheter coursing from the IVC through the azygous vein to the SVC. Using a long sheath helped manipulate the catheter despite this course. The inferior approach has the advantages of less risk of access complications, less radiation exposure and more operator comfort compared to a superior approach.

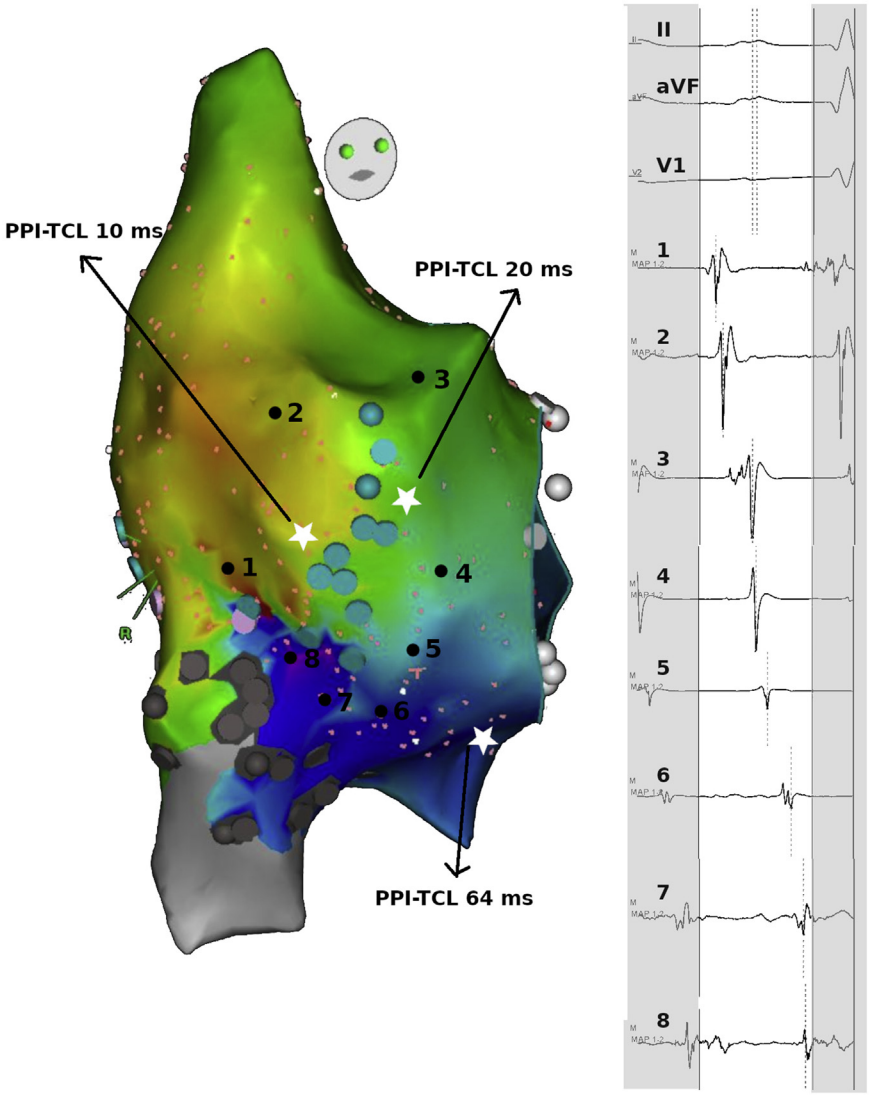

Fig. 3. Activation map during flutter.
Activation map constructed during the flutter in a modified RAO view shows the reentrant circuit on the atrial free wall. Electrograms from several locations along the circuit are shown on the right. The series of blue circles mark locations where double potentials were recorded and indicate the line of conduction block along the atriotomy scar. The white stars indicate the points from which entrainment was done and the measured post pacing intervals are indicated.

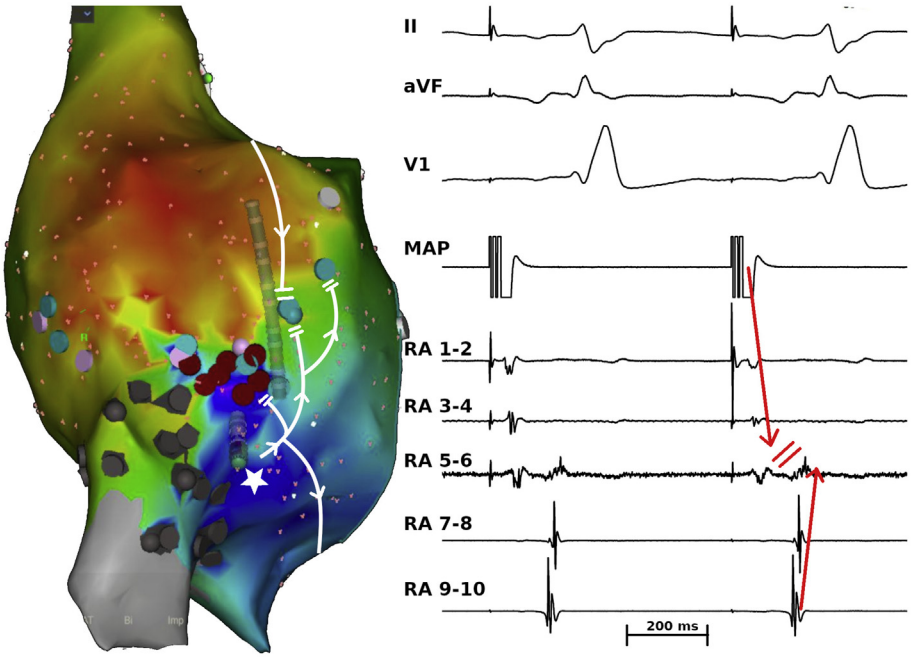

Fig. 4. Conduction block across the line after ablation.
After ablation, the decapolar catheter has been repositioned along the lateral wall of the right sided atrium across the line and the mapping catheter is positioned below the line. In panel A, position of both the catheters may be seen on a voltage map obtained during the flutter. Blue circles indicate double potentials while the red circles indicate ablation lesions. Electrograms recorded while pacing from the mapping catheter are shown in panel B. Panel C is a schematic illustration of the demonstration of conduction block. Activation of the electrodes located above the line of block happens in a craniocaudal direction confirming block.
HV = Hepatic Vein, TA = Tricuspid Annulus, Atr = Atriotomy scar, Abl = Ablation line.

Disclosures

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