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Catheter ablation of atrial arrhythmias in a patient with surgically corrected congenital heart disease and inferior vena cava interruption

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A 15 year old girl who underwent surgical correction of ventricular septal defect and patent ductus arteriosus ligation in childhood presented with atrial tachycardia of crista terminalis origin and counterclockwise atrial flutter. She also had associated interruption of inferior vena cava which continued as azygous vein and left superior vena cava which drained via coronary sinus into the right atrium. She underwent radiofrequency ablation of both the tachycardias via internal jugular vein and azygous vein approach using 3D electroanatomical mapping system.

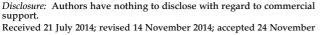
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Keywords: Interrupted inferior vena cava, Atrial flutter, Radiofrequency ablation, Drug refractory arrhythmias, 3D mapping

Introduction

C atheter ablation of atrial arrhythmias is usually performed via the femoral vein. Interruption of the inferior vena cava (IVC) poses technical challenges in the performance of the procedure. We report a case with operated congenital heart disease having drug-resistant atrial arrhythmias with IVC interruption. Internal jugular vein and azygos vein were used for access with 3D electroanatomical mapping to successfully ablate the arrhythmias. A 15-year-old girl born with a subaortic ventricular septal defect (VSD), patent ductus arteriosus (PDA) with left-to-right shunt, pulmonary artery hypertension, interrupted IVC continuing as azygos vein and left superior vena cava (LSVC) draining via coronary sinus (CS) into the right atrium. She underwent transatrial Gore-Tex patch closure of the VSD and PDA ligation at the age of two years. She also had an imperforate anus with a rectovesical fistula which was surgically repaired in infancy.

She presented with history of recurrent palpitations since the age of ten. Her electrocardiograms



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(ECG) showed regular narrow QRS tachycardia at the rate of 250–300 beats per minute, suggestive of atrial tachycardia (Fig. 1A) and atrial flutter (Fig. 1B). She was drug refractory and had breakthrough episodes on amiodarone, beta blockers, verapamil and digoxin with hemodynamic instability, which once required cardioversion.

In view of a past history of cardiac surgery during early childhood, scar related arrhythmias were a possibility; hence, the patient was taken up for electrophysiology study (EP) with 3D St. Jude Ensite velocity mapping system. Due to IVC interruption with azygos continuation and LSVC draining to CS, there was a technical difficulty in placing the catheters. Three diagnostic EP catheters were positioned into the high right atrium, right ventricle and CS via azygos vein from the right femoral vein (Fig. 2). EP study revealed atrial tachycardia from the superior crista terminalis. Mapping and ablation of this tachyarrhythmia by transfemoral route was unsuccessful, and internal jugular vein approach was then taken. Superior crista terminalis of right atrium was mapped for the site of earliest activation. At a site which was 50 ms earlier than the P wave on surface ECG, radiofrequency energy of 50 W, 50 °C

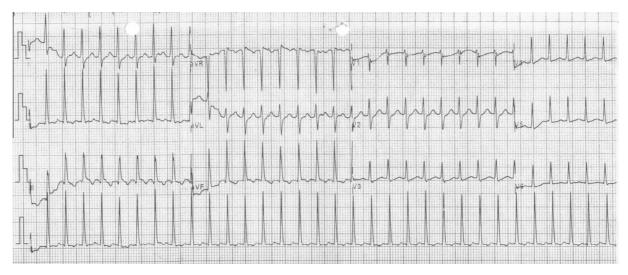


Figure 1A. ECG showing narrow QRS tachycardia suggestive of either atrial tachycardia with varying block or atrial fibrillation.

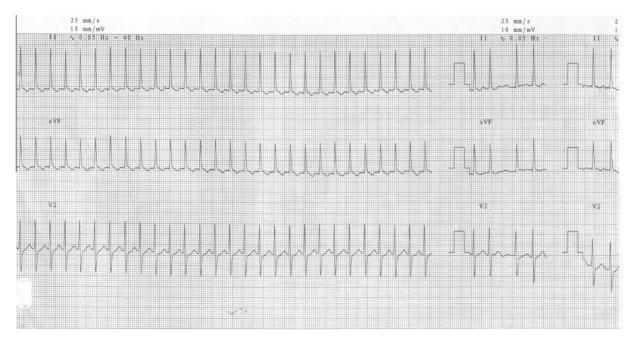


Figure 1B. ECG showing regular narrow QRS tachycardia at rate of 250–300 beats per minute suggestive of atrial flutter.

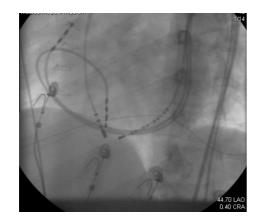


Figure 2. Fluoroscopy showing three diagnostic electrophysiology study catheters placed across LSVC to right atrium, right ventricle and coronary sinus, and mapping catheter through right IJV into cavotricuspid isthmus.

for 60–120 s, with a St. Jude irrigated Cool Path catheter was administered and the tachycardia was terminated (Fig. 3A). Following this, another irregular tachycardia was induced. The activation and propagation map was suggestive of typical counterclockwise atrial flutter. Linear lesions across cavotricuspid isthmus at 50 W, 50 °C for 120–240 s resulted in termination of atrial flutter and restoration of stable sinus rhythm (Fig. 3B). After ablation of this arrhythmia, no more arrhythmia could be induced with vigorous stimulation protocols and with isoprenaline. There were no periprocedural complications. Patient is not on any antiarrhythmic drugs. She has been on a

clinical follow-up of two years and has had no recurrence.

Discussion

IVC interruption or congenital stenosis is a rare congenital anomaly in the general population (<0.1%) [1]. Anomalies of IVC and associated abnormalities include retrocaval ureter, renal vein collar, left sided IVC, bilateral IVC, unilateral or bilateral azygos continuation of the IVC, circumaortic left renal vein, retroaortic left renal vein, and the absence of infrarenal IVC or entire IVC [2,3]. Its incidence is 0.6% in patients with congenital heart disease (cyanotic and acyanotic) and dextrocardia [13,14]. Catheter ablation of cardiac arrhythmias is usually performed via the femoral vein approach. Congenital or acquired abnormalities of the IVC pose technical challenges in accessing the ablation site. Alternative approaches via internal jugular vein [4–7], subclavian vein [8,9], and transhepatic [10] approach have been described. There are case reports of radiofrequency ablation of atrioventricular nodal reentrant tachycardia [15], accessory pathways [16], and atrial fibrillation [17] via superior approach in patients with IVC anomalies. There are reports of atrial flutter ablation via azygos vein approach [18]. There are case reports of radiofrequency ablation of atrial arrhythmias in patients with congenital heart diseases and IVC anomalies [11]

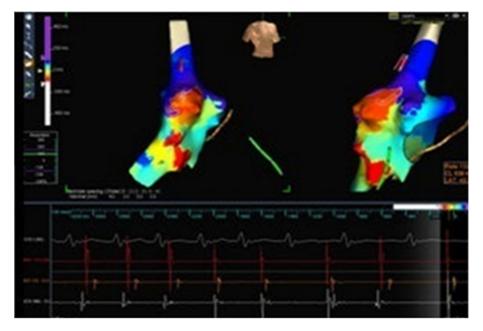


Figure 3A. 3D electroanatomical mapping system showing ablation catheter with good ablation signals at superior crista and termination of tachycardia.

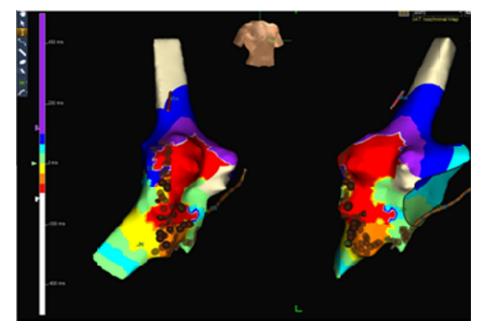


Figure 3B. 3D electroanatomical mapping system with right anterior oblique and left anterior oblique views showing ablation on the cavotricuspid isthmus.

using 3D electroanatomical mapping with Carto-Sound [12].

Conclusion

Arrhythmias in the presence of IVC interruption could be successfully ablated with the aid of 3D electroanatomical mapping. Prior cardiac surgery does not always lead to scar related arrhythmias and an attempt at ablation should always be made with a view to permanent cure. Further, IVC interruption may be technically challenging, but it does not preclude radiofrequency ablation as one of the therapeutic modalities.

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