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Images in cardiology

Successful ablation of two right atrial tachycardias on either side of the lateral tunnel patch in a patient with double inlet left ventricle and total cavopulmonary connection: Two sites and two mechanisms

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INTRODUCTION

Surgical strategy for patients born with a physiological univentricular heart has evolved from atrio-pulmonary (AP) Fontan (right atrium (RA) to pulmonary artery)[1] to its modification, total cavopulmonary connection (TCPC),[2] with the goal of reduced post-operative incidence of supraventricular tachycardia (SVT).[3–6] However, TCPC patients may also present with symptomatic atrial tachycardia, and the sites of origin can be difficult to access due to the surgical strategy. Owing to recent technological advances in the field of electrophysiology, such as electroanatomic mapping (EAM),[7,8] irrigated-tip catheters[9] and remote magnetic navigation (RMN),[10,11] catheter ablation has become a more relevant and important therapeutic modality for congenital heart disease associated-SVTs. We report on a 33-year-old male, with double inlet left ventricle (DILV) status post TCPC, who underwent catheter ablation of two different atrial tachycardias originating in different parts of the divided RA.

METHODS

The patient has DILV with transposed great arteries, sub-aortic stenosis and pulmonary stenosis. At 12 years of age, he had a Glenn procedure and, three months later, completion of TCPC with a Gore-Tex lateral tunnel. In addition, he underwent redo surgery for resection of high-grade symptomatic subaortic stenosis and ventricular septal defect enlargement. A residual left ventricular outflow tract obstruction (LVOTO) of 25–30 mmHg remained.

From the age of 26 years, he had onset of symptomatic arrhythmia leading to multiple DC cardioversions, as well as two ablations at another centre. The first ablation was in 2010, when an atrial tachycardia (AT) was mapped within the lateral tunnel and ablated. After a second ablation six months later, the patient still experienced arrhythmia recurrences and was referred to our institution. ECGs documented two clinical tachycardias, one AT of cycle length (CL) 340 ms and another of 260 ms.

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Pre-procedural imaging and image processing

The patient underwent preoperative imaging study by cardiovascular magnetic resonance (CMR) including a 3-dimensional, free-breathing, diaphragm navigated balanced steady-state free precession (bSSFP) sequence to image the whole thorax.

Assessments were made for the patency of TCPC pathways, degree and level of LVOTO, ventricular mass, volumes and function, pulmonary, aortic and caval flow (Figure 1). All pre-acquired 3D imaging DICOM data was processed for 3D reconstructions, not only of the heart but also of major veins and the aorta; and images were used during ablation procedures by integration with the mapping information (POLARIS software, Biosense Webster, Brussels, Belgium).

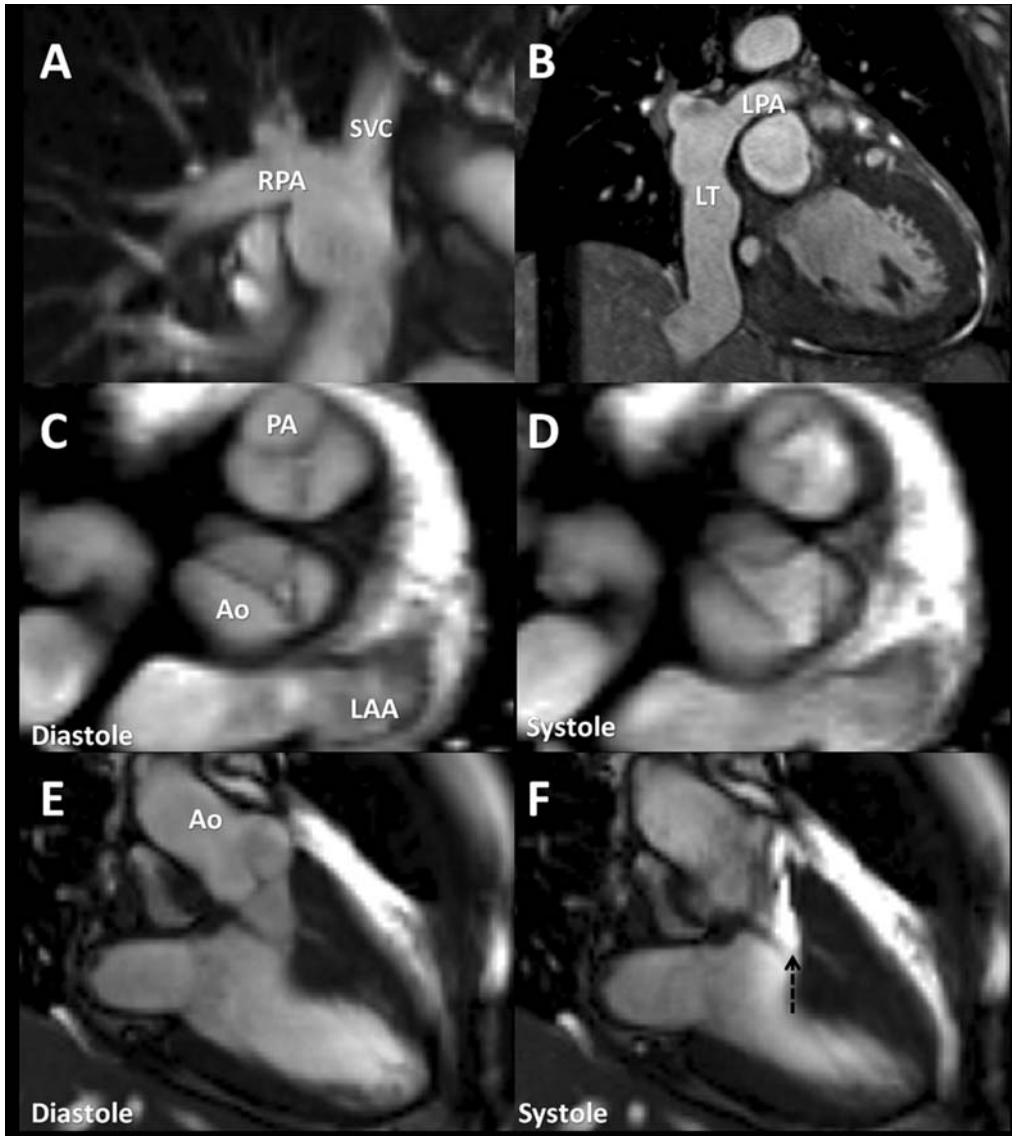

Figure 1. SVC; superior vena cava, RPA; right pulmonary artery, LT; lateral tunnel in total cavopulmonary connection, LPA; left pulmonary artery, PA; pulmonary artery, LAA; left atrial appendage A: Patent Glen anastomosis shown on diastolic still frame from cine image, B: Single frame from unsegmented 3D bSSFP acquisition showing lateral tunnel TCPC, inferior vena cava, left pulmonary artery and proximal right pulmonary artery. Note increased left ventricular wall thickness due to previous and residual LVOTO. C and D show unrestrictive opening of trileaflet aortic valve (systole) with small regurgitant orifice in diastole. E and F show the differing orientation in space of the aorta and aortic valve versus the LVOT and the jet of flow acceleration of LVOTO (dotted arrow).

Electrophysiologic study

After obtaining written informed consent, the procedure was performed under general anaesthesia with continuous invasive arterial blood pressure monitoring in the presence of an experienced cardiac anaesthetist. A decapolar diagnostic catheter (Bard), serving as the timing and pacing reference via the right femoral vein (8 Fr sheath), was positioned in a location with adequate signal amplitude and in a stable position ("hockey-stick" fashion inside the TCPC). The magnetically equipped ablation catheter (Navistar ThermoCool RMT, Biosense Webster, Brussels, Belgium) was introduced via the femoral vein or artery. Intracardiac electrograms were recorded on a recording system (BARD) and all electrogram and mapping information was displayed on the Odyssey platform (Sterotaxis Inc., St. Louis, US). The magnetic navigation system (Niobe II, Sterotaxis Inc., St. Louis, US) was used in conjunction with the cardio drive system. A detailed description of this system has been published previously.[12,13]

Since the patient presented in sinus rhythm at the beginning of the study, programmed stimulation was carried out in order to induce tachycardia. A persistent tachycardia was readily induced, with a CL of 340 ms and predominantly 2:1 AV nodal conduction. Using the arterial access, the magnetic catheter was retrogradely advanced via the aorta and the ventricle to the remainder of RA. Due to the residual stenosis in the subvalvular area, a direct approach was used steering the soft magnetic catheter straight (rather than the usual inverted curve) (Figure 2). Sequential EAM (CARTO 3 RMT, Biosense Webster) was performed during the ongoing tachycardia, which proved to consist of counterclockwise activation of the RA (Figure 3, top panel). After positive entrainment confirmed the EAM-derived diagnosis, a linear lesion connecting the patch of the TCPC to the tricuspid annulus was deployed using irrigated tip radiofrequency current (45 Watts, 30 ml/sec flow, max 120 s). The tachycardia terminated after CL prolongation to stable SR. During the following burst stimulation, a second sustained AT with 260 ms CL was induced with variable 2-3:1 AV nodal conduction. Subsequent mapping of the RA and left atrium demonstrated only bystander activation, which prompted advancing of the magnetic catheter via the venous femoral sheath into the TCPC. Finally, a focal origin high on the crista terminalis was identified with local signals preceding the onset of the surface ECG P wave by ~50 ms. Irrigated tip ablation terminated the tachycardia with a single RF application. Subsequent burst pacing from various sites within the next 45 min failed to induce any further sustained arrhythmia. The overall procedure duration amounted to 158 min with 1 min 48 s of fluoroscopy time (31.1 cGy cm^2).

DISCUSSION

A simple atrial flutter ablation can turn out to be a major challenge in TCPC patients with a no longer directly accessible RA. Perhaps in part due to the additional hurdle of a subaortic stenosis, the previous attempts at remote controlled ablation at a different centre failed to gain retrograde access to the target chamber from the aorta. After the surgical resection and the partial reduction of the LVOTO gradient, the updated 3D imaging allowed the interventionalist to confidently cross the unrestrictive aortic valve, change direction to account for the different plane of the left ventricular outflow tract

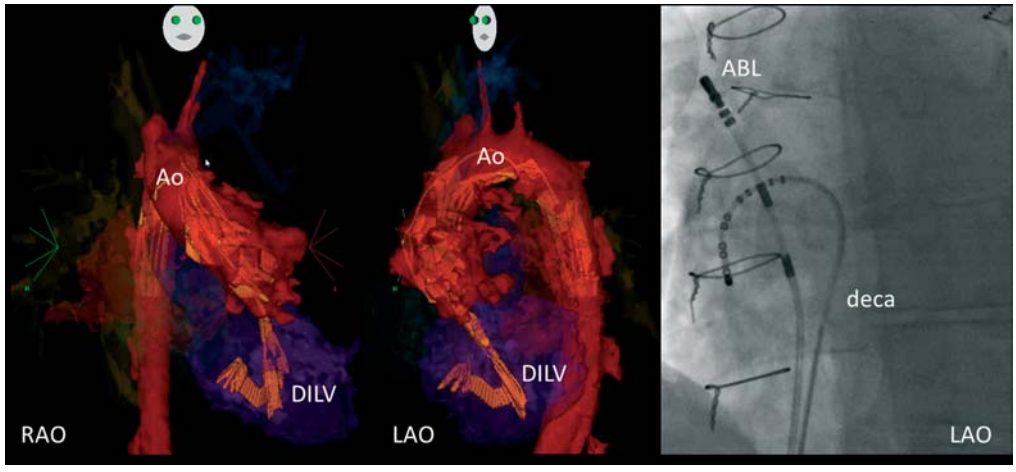

Figure 2. Access through the aortic valve and ventricles depicted using the fast anatomical mapping (FAM) feature highlighting the path of the magnetic ablation catheter (orange). Direct access and position of the ablation catheter at the focal source of tachycardia 2 inside the TCPC (right anterior oblique projection).

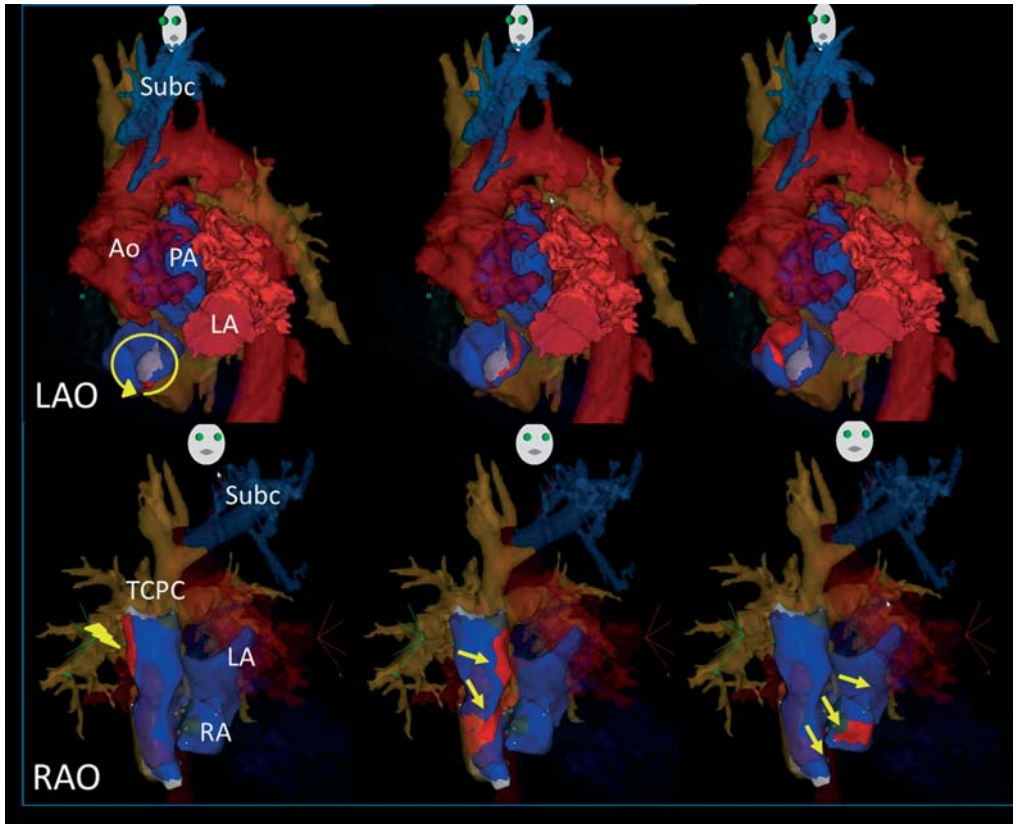

Figure 3. Electroanatomic activation map of the first tachycardia (340 ms CL). Activation propagated around the right-sided atrioventricular valve in a counter-clockwise manner and activation timing fulfilled the entire tachycardia CL. Linear ablation terminated the tachycardia. Electroanatomic activation map of the second tachycardia (260 ms CL). Activation propagated radially from the crista terminalis. Note the late bystander activation of the exclude RA and the LA. Point ablation terminated the tachycardia. LA; left atrium, RA; right atrium.

(LVOT) and confidently manoeuvre the catheter against high velocity LVOT flow. Hence, the remote navigation procedure could be guided safely.

Different arrhythmia substrates (re-entrant versus focal)

The re-entrant circuit is readily explained by the creation of the lateral tunnel (resulting in a scar area opposite to the tricuspid annulus, TA), which thereby creates a readily available substrate for re-entry around the TA. As for cause of the focal arrhythmia originating from the area of the crista terminalis, which is a common area of origin for this type of arrhythmia, one hypothesis could be the stretch of the TCPC lateral tunnel. Yap et al.[14] reported that despite the initial acute procedural success, high recurrence rate supports the concept that diseased myocardium provides further "sources" or substrates of arrhythmia. Whilst some of the focal AT origins were distributed in the areas in the vicinity of surgically-created substrates, such as the RA-RA appendage anastomosis, many focal ATs arose from areas with no relation to previous surgery or previous ablation scars. This suggests that progressive atrial myocardial damage occurs due to persistent haemodynamic overload.[15] Fortunately, in our case this area is easily accessible even without magnetic navigation.

Alternatives to RMN

Transbaffle puncture or re-sternotomy, which can be performed successfully by experienced hands,[16–18] is also an alternative to access the native RA in these patients. In our opinion however, RMN is the least invasive modality which can access difficult sites and with the lowest fluoroscopy exposure. In our adult patient, a previous fenestration was not available as a route of access to atrial tissue excluded from the lateral tunnel by the patch. Should the retrograde approach have failed again due to the residual subaortic gradient, we would have carried out a transbaffle puncture; but that last resort was fortunately avoidable.

CONCLUSION

A TCPC patient had two ATs with different mechanisms, both originating in the RA but on opposite sides of the TCPC patch. Using RMN and image integration with CMR, both arrhythmias were successfully addressed without the need for transbaffle puncture.

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Discloures

Sabine Ernst is a consultant to Biosense Webster and Stereotaxis, Inc.

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