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CASE REPORT
Adult congenital heart disease

Collaborative work in a complex case of Fontan for treating intra-atrial reentrant tachycardia and severe aortic stenosis: a case report

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Background Intra-atrial reentrant tachycardia (IART) is a frequent arrhythmia in patients with Fontan circulation. Although its supraventricular origin, such arrhythmia can be poorly tolerated as it leads to haemodynamic impairment. Concomitant assessment of pressure/volume overload of cardiac chambers due to valvular disease or residual shunts is necessary.

Case summary We report the case of a 33-year-old male with Fontan extracardiac conduit, suffering from IART with initial poor haemodynamic tolerance. He had a medical history of pulmonary atresia with intact ventricular septum and Type 0 bicuspid aortic valve, with a total of four cardiac surgeries. Echocardiography demonstrated a severe impairment of the univentricular ejection fraction and a critical aortic stenosis. Given the limited medical treatment options of the arrhythmia and the risks of another heart surgery, both IART ablation and transcatheter aortic valve replacement (TAVR) were performed during the same procedure. The IART critical isthmus located in the antero-lateral region of the extracardiac conduit was effectively treated with radiofrequency. Rapid pacing during TAVR was provided by a catheter placed in the unique ventricle via a transconduit puncture. The aortic valve was deployed with minimal para-valvular regurgitation and a satisfactory transvalvular gradient. At follow-up, the univentricular ejection fraction normalized and no arrhythmic episode was recorded in absence of anti-arrhythmic drugs.

Discussion This case highlights the need of a collaborative approach for treating complex cases of adult congenital heart disease, suffering from both electrophysiological and haemodynamic disorders. This combination offered an elegant and safest solution for treating concomitantly a life-threatening arrhythmia and an aortic stenosis.

Keywords Catheter ablation • Fontan • TAVR • IART • Bicuspid aortic valve • Case report

ESC Curriculum 4.2 Aortic stenosis • 5.5 Supraventricular tachycardia • 5.4 Atrial flutter • 9.7 Adult congenital heart disease • 2.1 Imaging modalities

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Learning points

- Pressure overload in cardiac chambers due to valvular stenosis may act as a trigger for IART in Fontan circulation and should be considered for treating the patient.
- TAVR could be a reasonable approach in complex case of congenital heart disease.
- Combined procedure for treating simultaneously the arrhythmia and the valvular disease is feasible.

Introduction

Since its introduction in 1968, the Fontan operation has become the surgical procedure of choice in patients with single ventricle anatomy. Almost 20% of patients receiving Fontan surgery will develop a supraventricular arrythmia during their lifetime.[1] The prevalence of the arrhythmia depends on the type of Fontan technique (historical Fontan vs. intracardiac lateral tunnel or extracardiac conduit).[2] Arrhythmias are typically represented by intra-atrial reentrant tachycardia (IART), the anatomical substrates being areas with pre-existing intra-atrial conduction abnormalities, suture lines, scar tissue, and prosthetic material.[3] Despite their supraventricular origin, such arrhythmias can be poorly tolerated in univentricular circulation. Their management may be challenging, as it requires a comprehensive treatment of the arrhythmia itself, but also evaluation of other parameters such as ventricular function, valvular disease, and residual shunt. We describe a successful IART ablation and concomitant transcatheter aortic valve replacement (TAVR) in a patient with extracardiac Fontan circulation.

Timeline

07/1987	Birth. Post-natal diagnosis of pulmonary atresia with intact ventricular septum and bicuspid aortic valve.
07/1987	First heart surgery: left De Leval anastomosis (systemic cavo-pulmonary shunt); non-functional 1 year later.
05/1988	Second heart surgery: right Blalock Taussig Thomas shunt.
06/1988	Balloon atrial septostomy (Rashkind procedure).
03/1989	Creation of an inter-atrial communication.
02/1992	Third heart surgery: creation of a total cavo-pulmonary anastomosis; suppression of right Blalock Taussig Thomas shunt.
11/1994	Fourth heart surgery: intervention on the cavo-pulmonary anastomosis; final Fontan circuit with an extracardiac conduit.
1994–2020	No particular medical interventions.
11/2020	Admission via emergency room for first episode of arrythmia with haemodynamic impairment.
11/2020	Discovery of impaired univentricular ejection fraction and severe aortic stenosis on transthoracic echocardiography (TTE).
01/2021	Electrophysiological study and catheter ablation of IART. TAVR.
04/2021	Cardiac magnetic resonance (CMR) showing normalized ejection fraction and residual para-valvular leak. Flecainide stopped.
07/2022	Free of arrhythmia without anti-arrhythmic drugs.

Case presentation

A 33-year-old male with complex congenital heart disease was referred for catheter ablation after presenting to the emergency room for palpitations and dizziness. He had a medical history of pulmonary atresia with intact ventricular septum and Type 0 bicuspid aortic valve. He underwent four previous palliative and corrective heart surgeries, the last one being a complete cavo-pulmonary Fontan circulation with extracardiac conduit at the age of 7 years. His follow-up was erratic. The per-critical electrocardiogram showed a narrow QRS complex tachycardia with a frequency of 260 b.p.m. and no discernable *P* waves. The decrease in heart rate after Amiodarone infusion allowed a better visualization of the atrial activity, with negative waves in the inferior limb leads and positive waves in Leads V1 and V2 (cycle 310 ms). A diagnosis of IART was made (*Figure 1*). The arrythmia was terminated by electrical cardioversion, given the poor haemodynamic tolerance (blood pressure 82/56 mmHg, NYHA IV status, 87% O_2 saturation on air). On TTE, there was an impaired ejection fraction of the unique systemic ventricle estimated at 35%, a probably severe but underestimated aortic stenosis with a mean gradient of 32 mmHg and a dilation of the ascending aorta (52 mm). An anti-arrhythmic (Amiodarone 200 mg daily) and anti-coagulant (Apixaban 5 mg twice a day) treatment was subsequently started. At 1-month follow-up, TTE demonstrated an improvement of the univentricular ejection fraction up to 55% and confirmed the severity of the aortic stenosis, with a mean gradient of 47 mmHg and a 4.2 m/s Vmax (*Figure 2*). Considering the limited options for medical treatment of the arrhythmia, a relatively favourable anatomy (the aortic annulus area was 7.2 cm^2 with moderate calcifications of a Type 0 bicuspid aortic valve, sino-tubular junction was measured at 40 mm and the distance between annulus and coronary ostia was 16 mm) and the prohibitive risks of another heart surgery (Euroscore II estimated at 18.1% and complex congenital heart disease), a decision was made to perform a radiofrequency arrhythmia ablation combined with a transcatheter aortic valve implantation with on-site surgical team back-up (*Figure 3*) considering the risk of valve embolization.

Under conscious sedation, an electrophysiological study was performed, with three-dimensional electro-anatomic mapping (CARTO 3, Biosense Webster, Johnson & Johnson, CA, USA) and integration of cardiac computed tomography images. After transconduit puncture, a meticulous mapping of both atria in sinus rhythm was realized using a multipolar catheter (Pentaray, Biosense Webster, Johnson & Johnson, CA, USA). The IART was easily triggered, with the same tachycardia cycle length of 310 ms. Activation mapping of both atria and the extracardiac conduit was needed to completely understand the reentrant circuit. The critical isthmus of the tachycardia was localized in the antero-lateral region of the extracardiac conduit. A 35 Watts radiofrequency ablation led to termination of the tachycardia (*Figure 4*). The IART was no longer inducible with burst pacing at 200 ms and Isoprenaline infusion.

A left transfemoral access was used for the TAVR procedure. After a careful assessment of the distance between the aortic annulus and the coronary ostia and invasive measurements demonstrating a 56 mmHg mean gradient, a 29 mm Edwards Sapien valve (Edwards Lifesciences, Irvine, CA, USA) was deployed with 3 cc extra-volume above the nominal value (*Figure 5*) and no prior valve dilatation to prevent prosthesis embolization. The rapid ventricular pacing during aortic valve implantation was provided by a catheter placed in the unique systemic ventricle

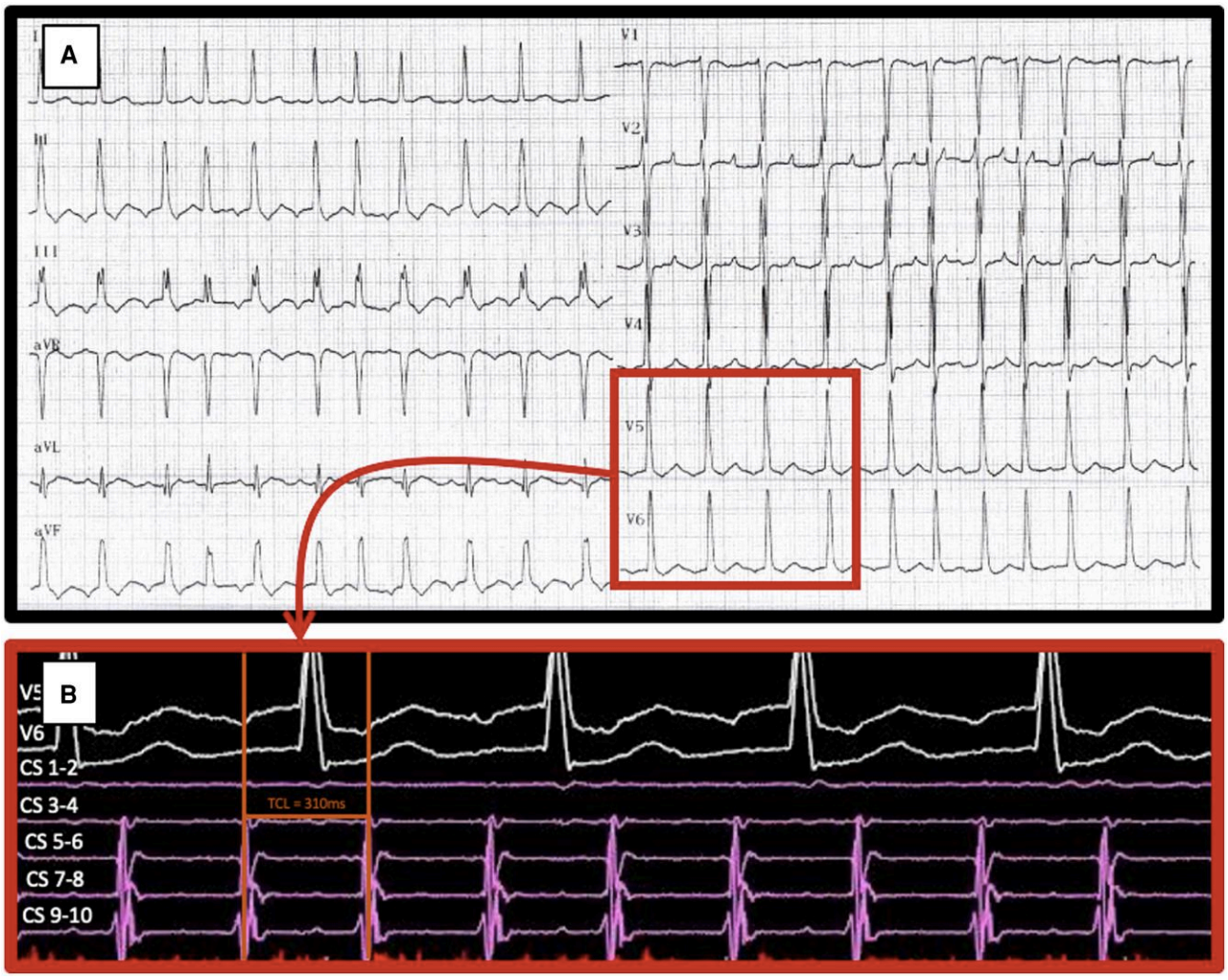


Figure 1 12-Leads electrocardiogram (*A*) showing an intra-atrial reentrant tachycardia with variable atrioventricular conduction. The tachycardia cycle length is 310 ms on the intracardiac electrogram (*B*).

via a transconduit puncture. There was a mild para-valvular aortic regurgitation on angiography (1.5/4) and a mean transvalvular gradient of 8 mmHg on TTE following the aortic valve implantation.

Anti-arrhythmic (Amiodarone replaced by Flecainide 100 mg daily) and anti-coagulant treatment were given following the IART/TAVR combined procedure. At 3-month follow-up, the patient was arrhythmia free with NYHA 1 status, NT-proBNP 92 ng/L ($n < 300$) and a VO_2 max at 21.5 mL/kg—120 W. CMR showed a 55% ventricular ejection fraction, with a non-compaction aspect. Two little spots of fibrosis were visible on late gadolinium enhancement sequence, located on the interventricular septum and the inferolateral wall of the ventricle (*Figure 6*). The flecainide was stopped, and anti-coagulation therapy continued. Angiotensin-converting enzyme inhibitors were introduced but poorly tolerated and finally stopped. The follow-up is now 1.5 years, the patient being asymptomatic, without further arrhythmic episodes.

Discussion

This case highlights the potential benefits of a collaborative approach for treating patients with complex congenital heart disease, suffering from both electrophysiological and haemodynamic disorders. Our patient had an IART and severe aortic stenosis in a context of extracardiac tunnel Fontan circulation.

Such patients are prone to develop IARTs as early complication, although less frequent in intracardiac or extracardiac tunnel than with classic/historical Fontan. Sustained arrhythmia with associated rapid atrioventricular conduction represents a medical emergency, as it often leads to haemodynamic impairment. Prompt treatment by electrical cardioversion is recommended. Although Amiodarone may be effective in preventing arrhythmia recurrence, the long-term side effects limit its use in clinical practice. Sotalol is another medical option. A low threshold of arrhythmia ablation should be encouraged, despite the technical difficulties due to the cardiac anatomy.[4] In our patient, the young age, the clinical presentation with haemodynamic intolerance and the tachycardia induced cardiomyopathy were in favour of radiofrequency ablation.

An arrhythmic event in patients with Fontan circulation should prompt haemodynamic evaluation. In our patient, there was a strong recommendation of aortic valve replacement, possibly associated with ascending aorta surgery.[5] Nonetheless, given the high surgical risk associated with four previous heart surgeries, TAVR was considered an acceptable solution despite the young age. This decision was

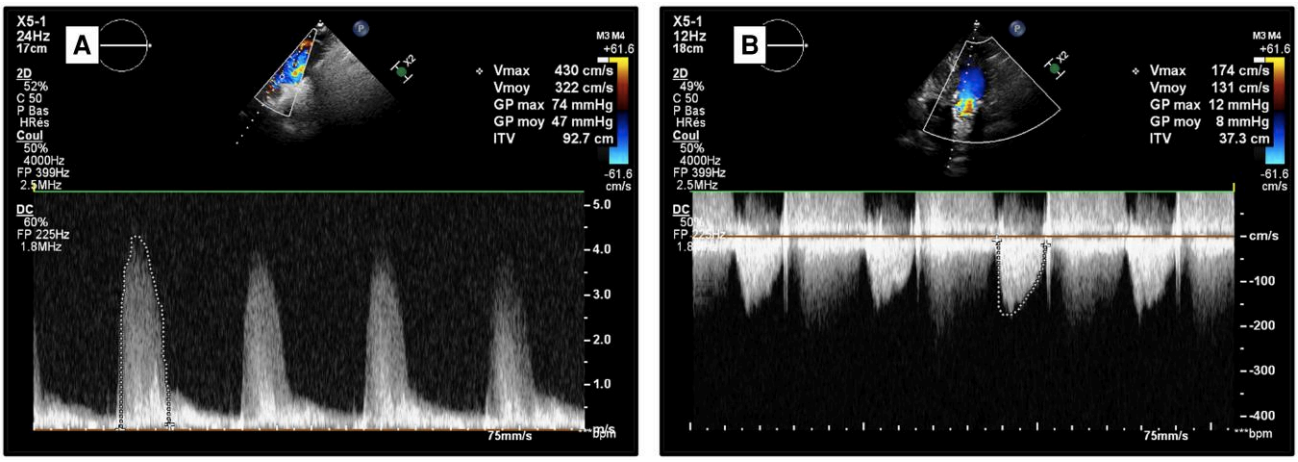


Figure 2 Ultrasound acquisitions before the transcatheter aortic valve replacement showing severe aortic stenosis with a mean gradient at 47 mmHg —suprasternal view (*A*) dropping to 8 mmHg after the transcatheter aortic valve replacement—apical five-chamber view (*B*).

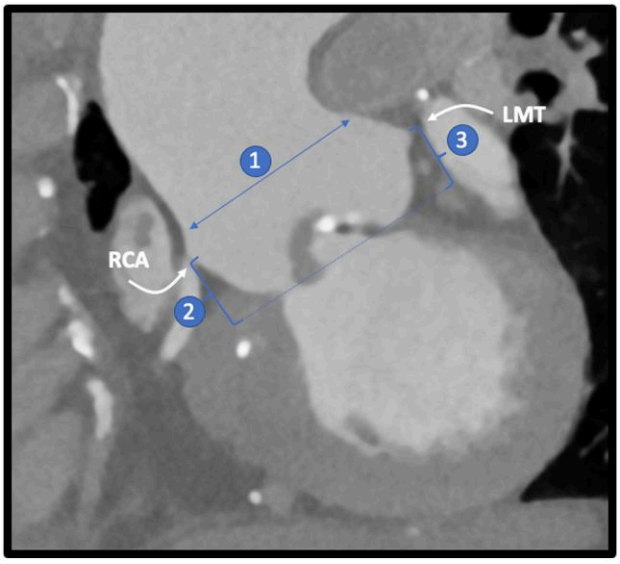


Figure 3 Cardiac computed tomography with geometry analysis of the aortic valve and roof in long axis showing few calcifications on the leaflets with a 40 mm a sino-tubular junction[1] and a 16 mm distance between annulus and coronary ostia.[2,3]

further supported by the favourable anatomy of the aortic root and valve and the recent results of TAVR on bicuspid valve. There is limited evidence regarding the safety and efficacy of TAVR in cases of bicuspid aortic valve, especially in young patients,[6] but recent publications demonstrated results similar to TAVR on tricuspid valve, with durability similar to surgical replacement. Concerns are usually linked to the dilated ascending aorta, the abnormal valve geometry and the risk of valve mispositioning and malfunction, with higher rates of moderate/severe para-valvular leak.[7] There was only a mild para-valvular aortic regurgitation and a non-significant mean transvalvular gradient in our case.

Cases of second transcatheter aortic valve deployment (TAV-in-TAV procedure) are described[8] and may be indicated in the future in this patient given the limited patency of transcatheter valve on long-term. Currently, there are no clear indications and the outcome of patients with two deployed prostheses is poorly known. Nonetheless, this could remain a valid option in our patient in case of aortic prosthesis dysfunction as the valve has a 29 mm diameter. Finally, despite a good result at distance of this hybrid procedure, the patient would probably be eligible for heart transplant in the future.

Figure 4 Intra-atrial reentrant tachycardia activation map with critical isthmus on the lateral wall of the extracardiac Fontan conduit (*A*). The green spot corresponds to the first radiofrequency ablation that terminated the tachycardia. Cardiac computed tomography three-dimensional reconstruction of the corresponding Fontan extracardiac conduit (*B*).

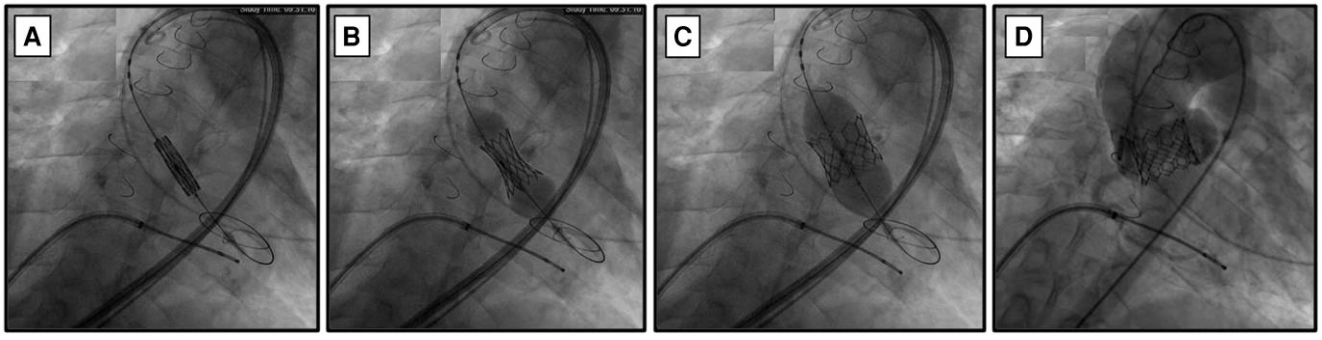


Figure 5 Different steps of the transcatheter aortic valve replacement with final angiography, showing a correct deployment of the valve and proper coronary perfusion (*A–D*).

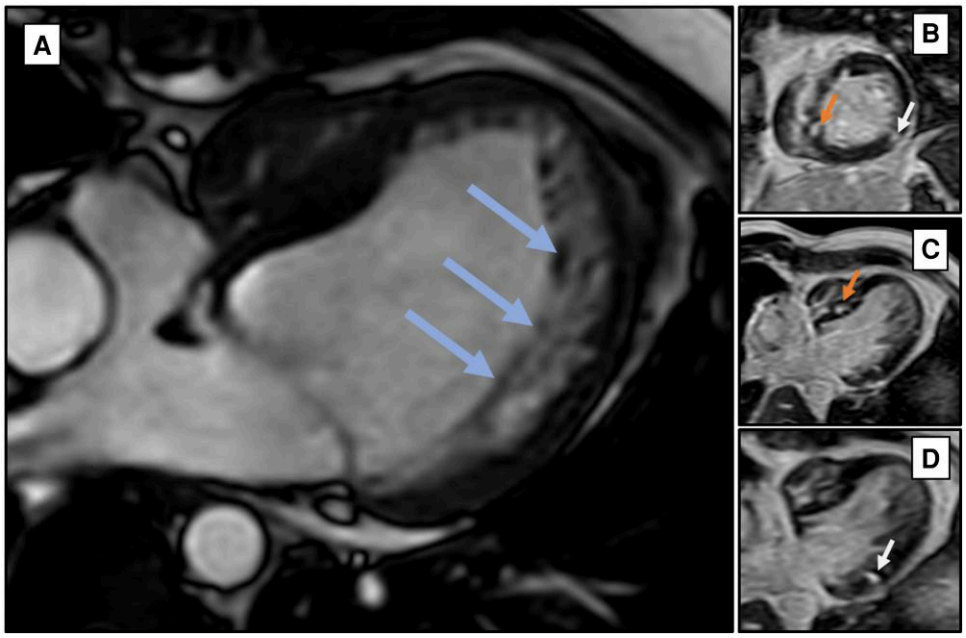
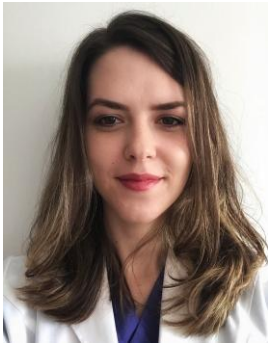


Figure 6 Cardiac magnetic resonance showing the non-compacted aspect of the ventricle (arrows) in long axis (*A*), and fibrosis spots in the infero-septal (arrows) (*B*, *C*) and infero-lateral (arrows) (*B*, *D*) portions of the ventricle in short and long axis-late gadolinium enhancement sequence.

Lead author biography



Dr Camelia Acatrinei is a senior resident in cardiology at Hospices Civils de Lyon, involved in electrophysiology and pacing. She is particularly involved in cardiac imaging to elaborate complex cases and plans to take a fellowship position within the next months.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

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