

# Atypical twin atrioventricular nodal re-entrant tachycardia in a congenitally corrected transposition of the great arteries patient with a surgical repair of a ventricular septal defect: a case report

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## Background

Twin atrioventricular (AV) nodal tachycardia is a rare mechanism of supraventricular arrhythmia, only seen in some specific congenital heart defects (CHD). It consists of a re-entrant circuit between two distinct AV nodes (anterior and inferior). Since both nodes have antegrade and retrograde conduction, there is usually two QRS morphologies in sinus rhythm.

## Case summary

This case is about an atypical twin AV nodal tachycardia in a 15 years old patient with congenitally corrected transposition of the great arteries and previous history of a ventricular septal defect repair. The surgical closure was probably responsible of a poor antegrade conduction over the inferior AV node, which was responsible for a unique QRS morphology. He finally received a catheter ablation of the inferior AV node. He remained asymptomatic without anti-arrhythmic drugs at 8-months post-ablation.

## Discussion

Twin AV nodal re-entrant tachycardia is a rare phenomenon. The presentation and electrophysiological study can be both atypical due to previous surgical repair. The operator should be aware of specific CHD where twin AV nodal re-entrant tachycardias are expected.

## Learning points:

- Previous ventricular septal defect (VSD) surgical repair can modify electrophysiological properties of the conduction system and create an atypical presentation of a twin atrioventricular (AV) nodal tachycardia
- Twin AV nodes re-entrant tachycardia must be sought in case of tachycardia and AV discordance (with situs solitus or inversus), mal-aligned AV septal defect, and right or left atrial isomerism.
- Both anatomical and electrophysiological approaches are needed for a successful catheter ablation in complex congenital heart defects.

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**Keywords**

Case report • Congenital heart disease • Twin AV nodes • Congenitally corrected-TGA • Catheter ablation • Twin atrioventricular nodal re-entrant tachycardia

**Introduction**

In 1913, Mönckeberg *et al.*<sup>1</sup> reported the first pathological evidence for the existence of twin atrioventricular (AV) nodes in a patient with AV discordance and a double-outlet right ventricle. A macrore-entrant circuit in which both AV nodes are obligatory components, i.e., twin AV nodal re-entrant tachycardia, has since been described in a few case reports and small case series.<sup>2–4</sup> The most common associated congenital heart defects are AV discordance (with situs solitus or inversus), malaligned AV septal defect, and right or left atrial isomerism. We describe a successful catheter ablation of twin AV nodal re-entrant tachycardia in a patient with congenitally corrected transposition of the great arteries (cc-TGA) and a previous surgically repaired ventricular septal defect (VSD).

**Timeline**

06/2005	Birth [antenatal diagnosis of congenitally corrected transposition of the great arteries and ventricular septal defect (VSD)]
09/2005	Pulmonary artery banding
08/2008	VSD surgical repair
01/2018	Palpitations
03/2020	Palpitations and dizziness, first electrocardiogram documentation
03/2020	Appointment with an electrophysiologist
04/2020	Admission via emergency room for incessant tachycardia
04/2020	Electrophysiological study and catheter ablation of twin atrioventricular nodal tachycardia
03/2021	Free from arrhythmia without antiarrhythmic drugs

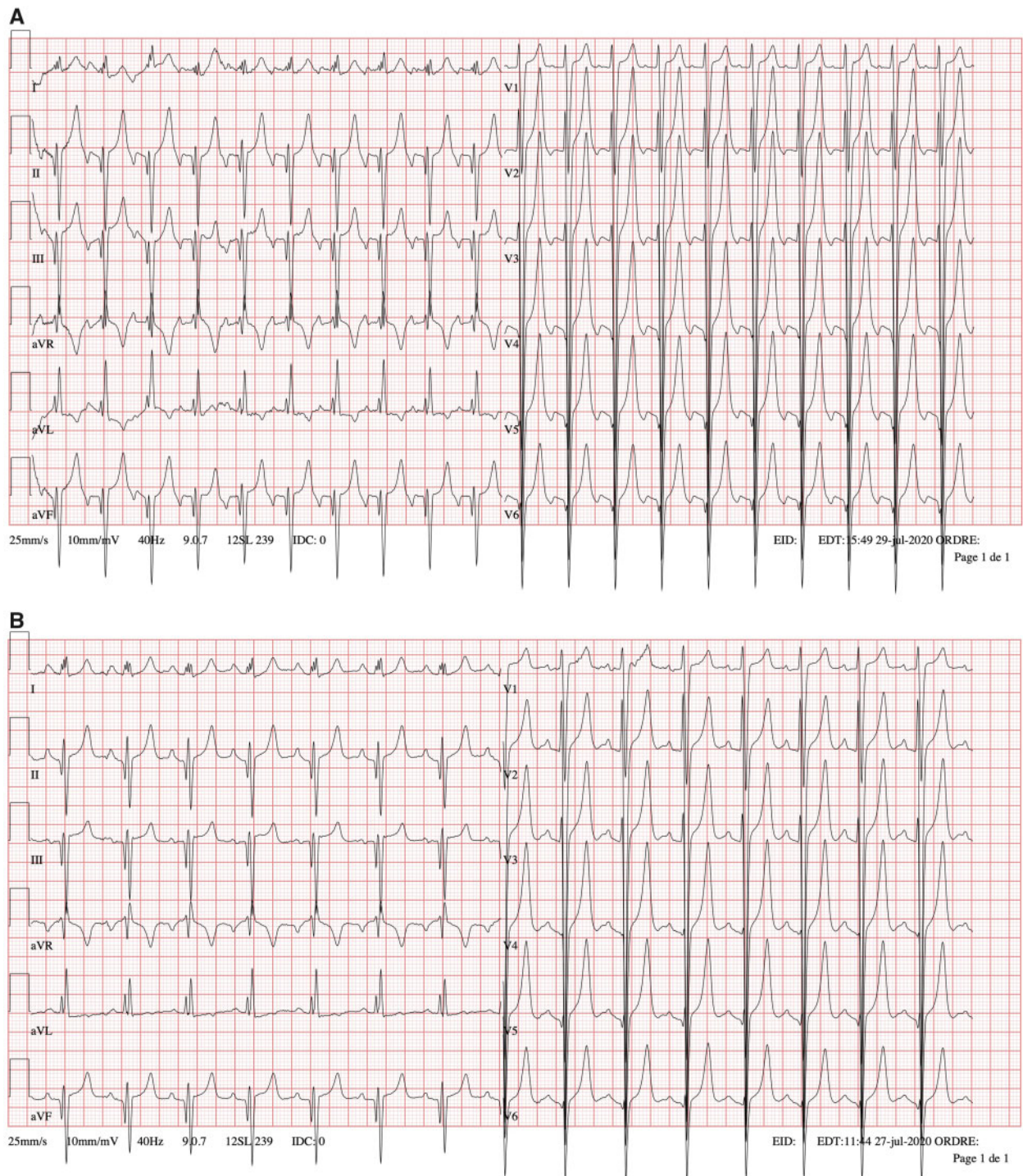
**Case presentation**

A 15-year-old teenager with complex congenital heart disease was referred for catheter ablation after presenting to the emergency room with incessant palpitations and dizziness. A regular narrow complex tachycardia with the frequency of 125 b.p.m. with a long RP' interval and negative P wave in the inferior leads was terminated upon administration of intravenous adenosine but immediately restarted (*Figure 1A*). The electrocardiogram (ECG) in sinus rhythm did not show any change in the QRS axis, while P waves became positive in the inferior leads, demonstrating an atrial activation from the sinus node (*Figure 1B*). Over the preceding 2 years, the patient

complained of incessant rapid and regular palpitations of sudden onset. He was born with *situs solitus* cc-TGA, mild sub-pulmonary stenosis, and a perimembranous VSD. He underwent a pulmonary artery banding at the age of 3 months, and a surgical VSD closure with a patch at the age of 3 years. He was suffering from residual pulmonary arterial hypertension and hypoxia (85–90%) due to a residual right–left shunt through the VSD. Apart from arrhythmia episodes, the patient had a Class II New York Heart Association status, with a Stage 5/6 systolic murmur. The ejection fraction of his systemic right ventricle was estimated as 45% by transthoracic ultrasound. He did not receive antiarrhythmic drugs prior to the admission. Integrating the heart defect condition and ECG data, three differential diagnoses were considered: a twin AV nodal re-entrant tachycardia, an atrial tachycardia (AT), and an atypical (fast–slow) atrioventricular nodal re-entrant tachycardia (AVNRT).

Under conscious sedation, an electrophysiological study was performed with three-dimensional electroanatomic mapping (CARTO 3, Biosense Webster, Johnson & Johnson, CA, USA) and cardiac computed tomography (CT) image integration to better understand the complex anatomy and to determine the location of the AV nodal path. A 6-Fr decapolar reference catheter (Livewire, St. Jude Medical/Abbott, Menlo Park, CA, USA) was inserted through the right femoral vein into the lateral right atrium wall in order to get a reference of atrial signals. After a meticulous electroanatomical mapping of the right atrium in sinus rhythm with a multipolar catheter (Pentaray, Biosense Webster, Johnson & Johnson, CA, USA), two distinct AV nodes signals were recorded on the superior and inferior sides of the VSD. Considering that twin AV nodal re-entrant tachycardia was plausible, a 6-Fr quadripolar reference catheter (Livewire, St. Jude Medical/Abbott, Menlo Park, CA, USA) was placed on the anterior side of the VSD to record signals from the superior node. The 8-Fr irrigated radiofrequency ablation catheter (Smart touch SF, Thermocool, Biosense Webster) was then placed on the inferior side of the VSD to record signals from the inferior node (*Figure 2*) and start electrophysiological study.

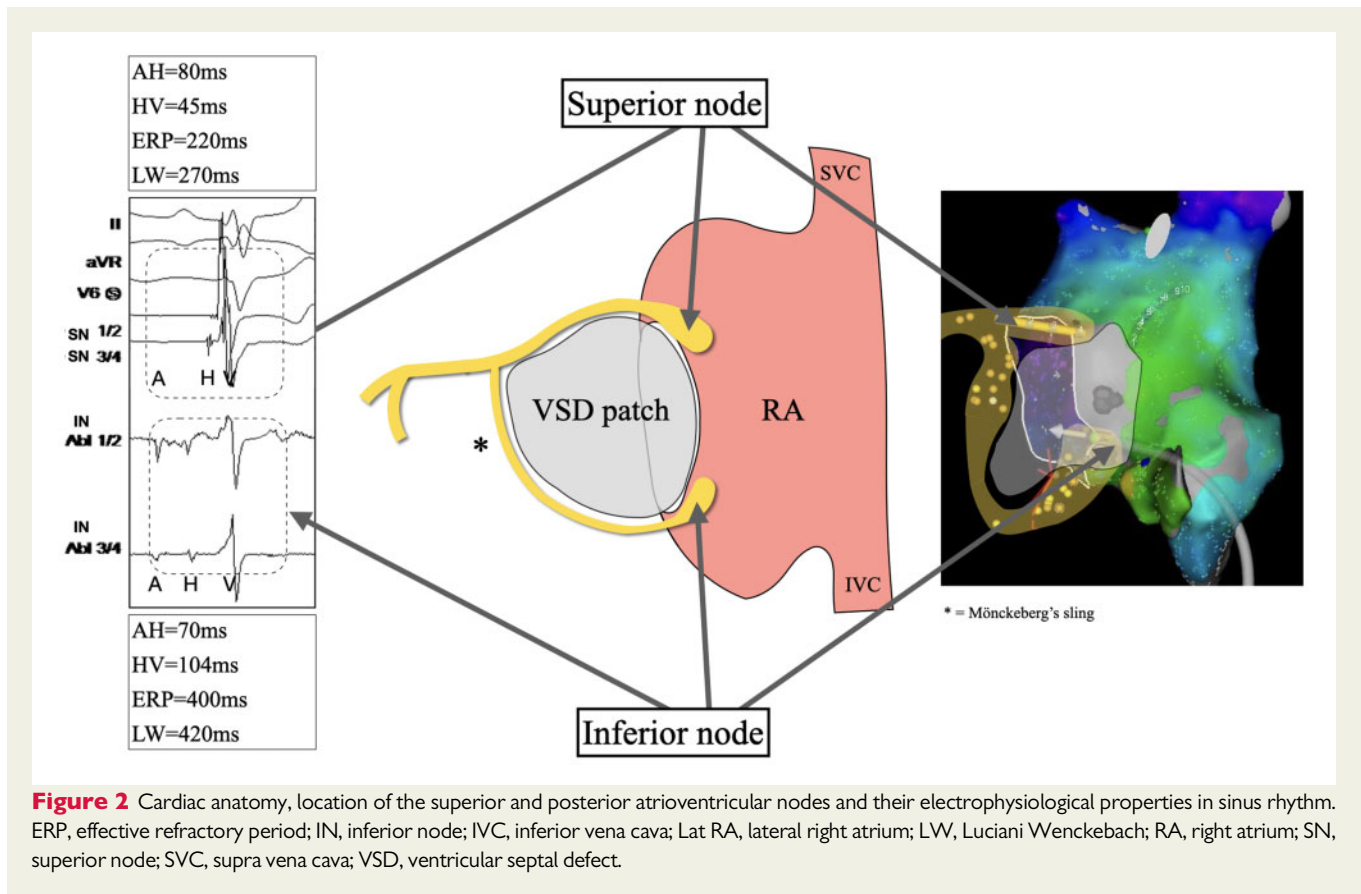
Electrophysiological testing confirmed the presence of two distinct AV nodes, i.e., superior and inferior, each with decremental properties, discrete His-bundle electrograms, but only a single pattern of ventricular activation in sinus rhythm. The superior AV conduction system was associated with an His to Ventricle (HV) interval of 45 ms, an AV node effective refractory period of 220 ms at a drive train of 600 ms, and an antegrade Wenckebach cycle length of 270 ms. Corresponding values for the inferior AV conduction system were less effective: HV 104 ms, effective refractory period of 400 ms at a drive train of 600 ms, and Wenckebach cycle length 420 ms. A sling of conduction tissue, so-called Mönckeberg's sling, usually connects the two AV conduction systems along with the VSD, as schematically portrayed in *Figure 2*. Expected signals along the sling characterized by high-frequency potentials that preceded local ventricular activation were not catchable, as a patch of VSD covered this region.



**Figure 1** Twelve leads electrocardiogram during tachycardia (A) and sinus rhythm (B). The electrocardiogram during tachycardia (A) and in sinus rhythm have the same QRS axis. The long RP' interval with negative P wave in the inferior leads is in favour of an inferior atrial activation. The trace in sinus rhythm (B) did not show any change in QRS axis, while P waves became positive in the inferior leads.

Sustained supraventricular tachycardia was easily induced by atrial extra stimuli without isoproterenol infusion. During tachycardia, entrainment mapping at the high output from superior and inferior AV nodal pathway sites confirmed that they were both components of

the circuit, ruled out the differential diagnoses of an atypical fast-slow AVNRT and atrial tachycardia. Electroanatomic mapping of the clinical tachycardia with image integration is shown in [Figure 3](#). The twin AV nodal re-entrant tachycardia coursed antegrade across the



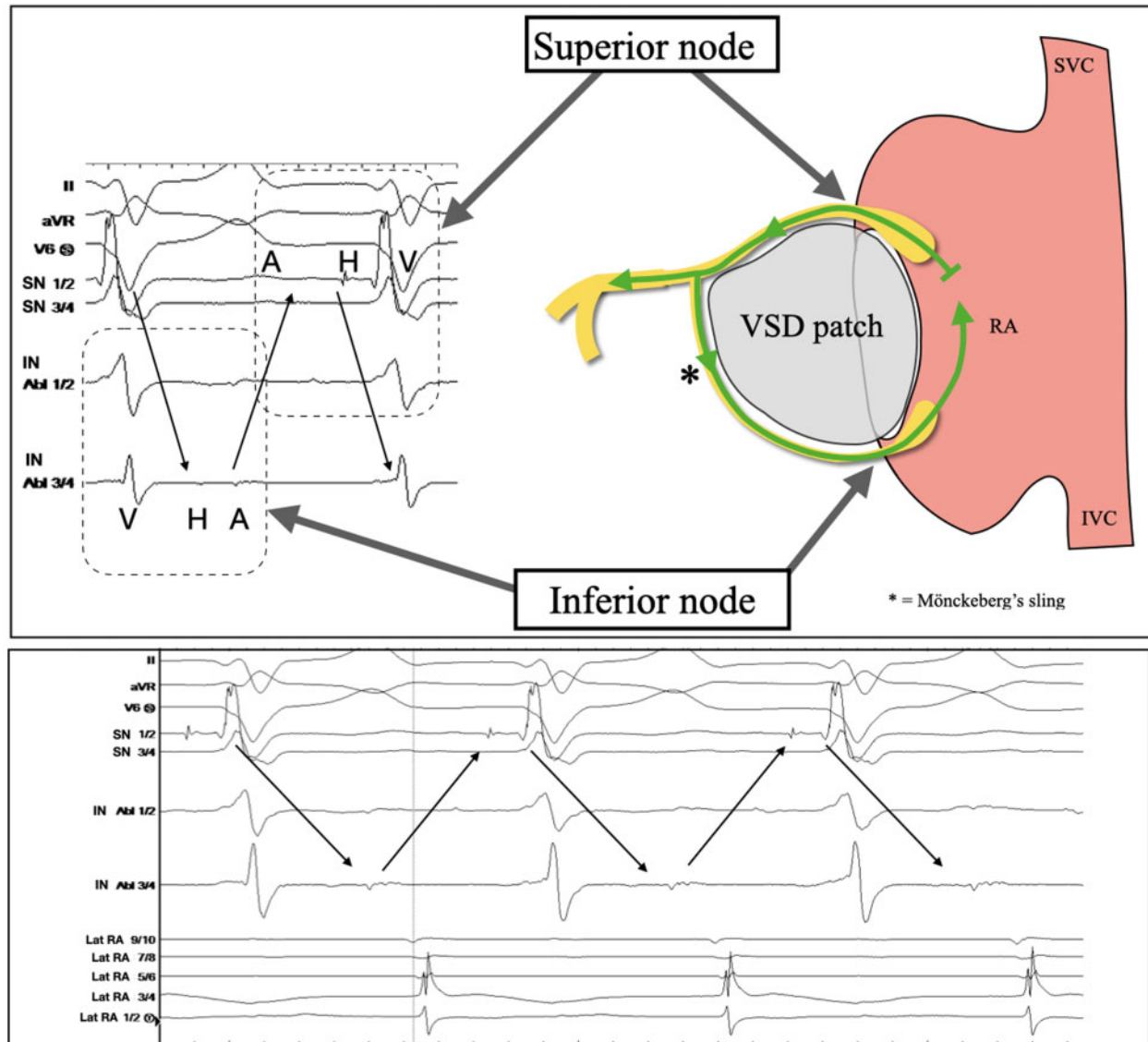
superior AV nodal pathway, down Mönckeberg's sling, and retrograde through the inferior AV node. In tachycardia, the antegrade superior node conduction system was associated with an Atrium to His (AH) interval of 70 ms and an HV interval of 47 ms. Corresponding retrograde values for the inferior AV conduction system were: Ventricle to His (VH) 133 ms and His to Atrium (HA) 100 ms. The inferior AV node was targeted during tachycardia with irrigated radiofrequency ablation considering its less robust conduction properties. Successful elimination of the inferior AV node was achieved in two steps: RF applications were started on the right side of the VSD but only induced a prolongation of the VA interval. Then, shots were performed over a residual shunt, on the left side of the VSD, resulting in a termination of the tachycardia while the axis from the ECG in sinus rhythm remains similar (Figures 1 and 4). During a 30-min observation period on and off isoproterenol, no tachycardia was inducible. Ventricular stimulation demonstrated absence of retrograde conduction via the remaining superior AV node, while its antegrade conduction properties were comparable. At 8 months of follow-up, the patient remains arrhythmia free with no antiarrhythmic drugs.

## Discussion

Here, we present a rare case of a twin AV nodal re-entrant tachycardia in a patient with cc-TGA, sub-pulmonary stenosis, and a prior

surgical repair of a VSD. While most patients with such arrhythmia have a very complex cardiac anatomy, our case was still challenging since the circuit between the two nodes was persisting despite the stitching of a patch to close the VSD that was probably responsible of a poor antegrade conduction through the inferior AV node. The presence of two distinct nodes is the consequence of embryological sequelae. In *in situs solitus* form of cc-TGA, there is a malalignment gap between the ventricular septum and the atrial septum. The gap in between the two structures is filled by a large membranous septum or a VSD. It is responsible of an anterior AV node dislodgement, below the orifice of the right appendage while the regularly situated posterior AV node is hypoplastic and cannot connect with any AV bundle. In cases such as ours, where the pulmonary outflow tract is narrowed, septal malalignment is reduced. It can create a sling of AV conduction system connected to both anteriorly and posteriorly situated AV nodes, and thus re-entrant circuit between the two structures.<sup>5</sup>

In our case, electrophysiological features include (i) poor antegrade conduction through the inferior node, and a normal antegrade conduction through the superior node, responsible for only one QRS morphology in sinus rhythm instead of two (ii) discrete His bundle electrograms recorded at two separate anatomic locations associated with a similar QRS morphology, (iii) decremental conduction over each AV nodal pathway with a stable HV interval, (iv) inducible

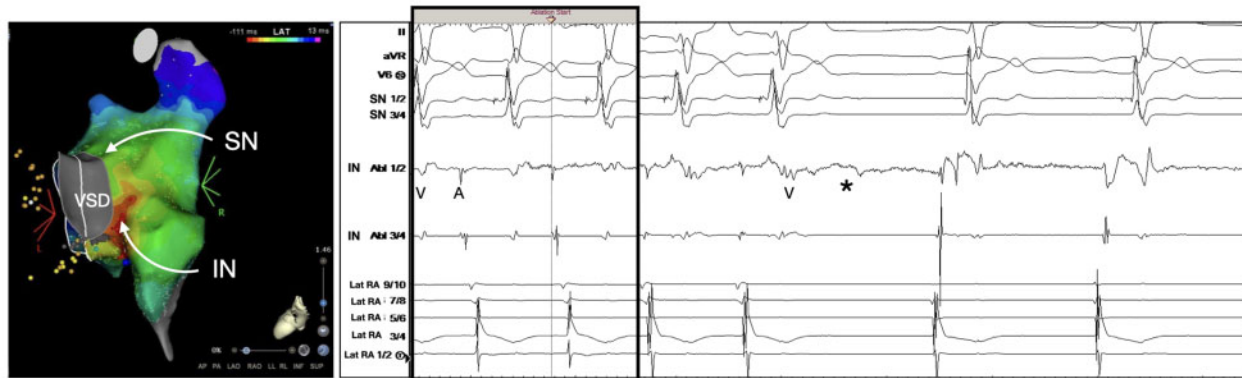


**Figure 3** Twin atrioventricular nodal re-entrant tachycardia and electrophysiological components of the two nodes. The superior panel describes detailed electrophysiological activation of the superior and inferior atrioventricular nodes in tachycardia. The inferior panel depicted the EGM pattern of the re-entry. IN, inferior node; IVC, inferior vena cava; Lat RA, lateral right atrium; RA, right atrium; SN, superior node; SVC, supra vena cava; VSD, ventricular septal defect.

tachycardia with antegrade conduction over the superior AV nodal pathway and a good enough retrograde conduction across the inferior AV nodal pathway, ensuring the circuit and incessant tachycardia, (v) entrainment mapping demonstrating that both AV nodal pathways are within the tachycardia circuit, and (vi) abolition of the tachycardia circuit by ablation of the inferior AV node.

In addition to the detailed characterization of the rare phenomenon of twin AV nodes, and the connection linking the two conduction systems, the case highlights some of the complexities encountered in catheter ablation procedures in patients with

congenital heart disease.<sup>6</sup> These may include formidably large chambers of interest, with difficulties ensuring optimal catheter contact and transmural lesions. Distorted anatomies and acute angles may limit access. Occasionally, arrhythmia substrates are concealed beneath patches or prosthetic material. In this case, VSD closure was probably responsible for a poor conduction over the inferior AV node and thus, for a single QRS morphology instead of two. A degenerative or innate origin is less probable. Still, all components of the circuit, including both nodes and the Mönckeberg's sling were good enough to create re-entrant circuit and incessant arrhythmia.



**Figure 4** Termination of the tachycardia during catheter ablation of the inferior atrioventricular node. The left panel is an activation map during tachycardia (Carto3 system, Biosense Webster), highlighting atrial activation through the inferior node. The first local activation time (LAT) dots of the atrial signals are coloured in red, as the red colour in both the histogram and the colour scale is the colour representing the local EGM signals that anticipates the more the signal recorded with the reference catheter. The right panel depicts the electrophysiological activation in tachycardia. Signals recorded just before the ablation starts are framed. The tachycardia termination is preceded from a VA prolongation. It ends with a V signal that demonstrates an interruption of the inferior node conduction. IN, inferior node; Lat RA, lateral right atrium; SN, superior node; VSD, ventricular septal defect.

Interestingly, VA connections of the inferior node were targeted at multiple sites, from the right and left sides of the VSD, through the residual shunt of the patch.

Twin AV nodal re-entrant tachycardia is a rare supraventricular arrhythmia. Usually, electrophysiological features include two distinct AV nodes with discrete His bundle electrograms, decremental conduction, and separate non-preexcited QRS morphologies; a sling of tissue connecting the two AV conduction systems; and tachycardia that courses antegrade by one AV nodal pathway, across Mönckeberg's sling, and retrograde via the second AV nodal pathway. In our case, previous surgical VSD closure did not prevail on conduction properties of the circuit but was probably responsible for only a single QRS morphology in sinus rhythm. Catheter ablation, which consists of targeting the most fragile AV node, was effectively achieved after accurate mapping of the circuit.

## Lead author biography



Francis Bessière works as an electrophysiologist specialized in pediatrics and congenital heart diseases. After his training, at the Montreal Heart Institute he joined the Louis Pradel cardiovascular hospital (Hospices Civils de Lyon) as an assistant professor. His field of research and expertise aims to develop new technics for treating complex arrhythmias.

## Supplementary material

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

**Slide sets:** A fully edited slide set detailing these cases and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The authors confirm that written consent for submission and publication of this case report has been obtained from the patient, in line with the COPE guidelines.

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