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## Unusual cause of cardiomyopathy in a young woman

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## ABSTRACT

Dual atrioventricular nodal nonreentrant tachycardia (DAVNNT) is a rare form of supraventricular tachycardia. In some patients, the presence of a dual pathway physiology results in two paths in the atrioventricular (AV) node with different conduction velocities. An atrial impulse arriving at the AV node may unfold and travel along these two pathways simultaneously, causing two ventricular activations. Thus, the ventricular rate will be twice the atrial rate. DAVNNT is less common than AVNRT, but its frequency may be underestimated.

The ECG is crucial to suspect the diagnosis. At first glance it looks like an irregular tachycardia, but a more careful look shows a rhythmic pattern. A sinus P wave followed by two QRS complexes (narrow or wide) should raise suspicion of this arrhythmia.

It is often unnoticed by the patient, and ventricular dysfunction due to tachycardiomyopathy is not uncommon. The response of DAVNNT to medication, including beta-blockers, flecainide, and amiodarone is very poor or absent, so the treatment of choice is slow pathway ablation. We report a Case of cardiomyopathy caused by this entity.

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## 1. Case report

A 44-year-old woman presented to hospital with dyspnoea on minimal exertion and orthopnoea, of about 4 weeks' duration. She had not noticed palpitations. She smoked about 10 cigarettes per day. She had no other history of interest and did not take regular medication. Chest X-ray showed cardiomegaly and signs of heart failure. Echocardiogram showed dilatation of the four chambers and severe biventricular dysfunction. An MRI confirmed the findings: the left ventricular end-systolic volume was 177 ml and the ejection fraction was 21%; no enhancement in the myocardium of both ventricles. On coronary angiography, the coronary arteries were normal.

Telemetry showed frequent bursts of non-regular tachycardia, unnoticed by the patient, with frequencies ranging from 110 to 180 bpm. The ECG (Fig. 1) revealed an "irregular" narrow complex tachycardia at about 120 bpm and evidence of left ventricular (LV) enlargement. Dual atrioventricular nodal nonreentrant tachycardia

(DAVNNT) was suspected. An electrophysiological study was performed (Fig. 2A), which showed double nodal conduction of each sinus beat, with two AH intervals (short and long) repeating rhythmically. There was no ventriculoatrial (VA) retrograde conduction (Fig. 2B). A radiofrequency application in the slow pathway area interrupted this dual response, leaving only conduction through the shorter AH interval. This confirmed the diagnosis of DAVNNT.

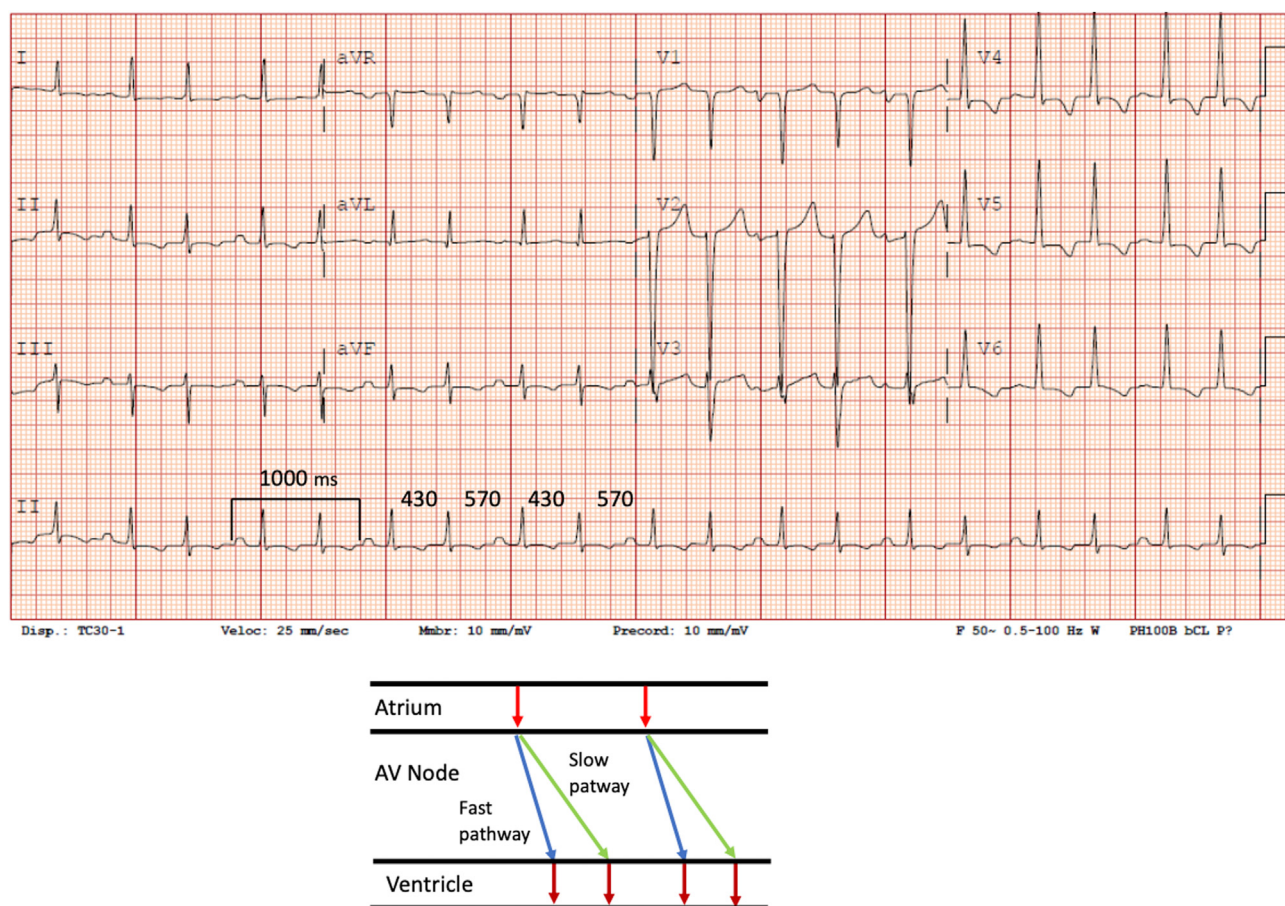
After six months, the patient was asymptomatic. The ECG (Fig. 3) showed sinus rhythm with a PR interval of 150 ms and LV enlargement data had disappeared. On chest X-ray (Fig. 4A and B), the size of the cardiac silhouette had normalized. The echocardiogram showed non-dilated cavities and the ejection fraction of both ventricles was normal (Fig. 4C and D).

In some patients, functional dissociation of the atrioventricular (AV) node may result in two conduction pathways, known as the fast and slow pathways. A sinus impulse arriving at the AV node may "go down" one of these pathways and, upon reaching the end of the pathway, "go up" the other pathway into the atrium, while continuing through the His-Purkinje system into the ventricle. This is the mechanism of the most common paroxysmal supraventricular tachycardia, the AVNRT. But on rare occasions, under certain circumstances, a sinus beat, on reaching the AV node, can be

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**Fig. 1.** The ECG shows sinus rhythm at 60 bpm (1000 ms). Each P wave is followed by two QRS complexes. The mean ventricular rate is 120 bpm. The R-R intervals reveal a fixed sequence of 430 and 570 ms. There is alternation in QRS morphology, due to variation in the length of the R-R intervals, leading to changes in the ventricular activation pattern. There is evidence of left ventricular enlargement.

directed by both fast and slow pathways simultaneously and cause two ventricular depolarisations.

The phenomenon of double ventricular response to a single sinus beat (double fire) was first described in 1975 [1]. Since then no more than 100 cases of DAVNNT have been reported [2]. And although it is a rare arrhythmia, it may be more common than previously thought.

The 12-lead ECG is the key test for suspecting and diagnosing DAVNNT [3]. The most typical finding is a sinus P wave followed by two QRS complexes. These show a regularly irregular sequence and the R-R intervals vary in a predictable and recurrent pattern.

DAVNNT has been confused with many other arrhythmias. One of the most common misdiagnoses is that of atrial fibrillation (AF) [4]. AV conduction 1:2 is often intermittent, and the ECG at first glance may appear to be AF. The rare condition of a nodal reentrant tachycardia with 2:1 retrograde block can be ruled out because in the latter entity the P waves are not of sinus morphology (the impulse travels from the AV node to the atrium). More frequent is the diagnostic doubt with atrial extrasystoles; it is necessary to explore the presence of a possible atrial depolarisation preceding the “second” QRS. The most complicated differential diagnosis is with the rare event of junctional extrasystoles in bigeminism. In DAVNNT each P wave is linked to the next two QRS with constant PR1 and PR2 intervals, whereas due to the influence of junctional extrasystole on atrioventricular conduction, in the latter entity, PR1

and PR2 intervals generally vary from beat to beat.

If one or both QRS of DAVNNT are wide (i.e. aberrancy), it may also be mistaken for ventricular tachycardia or ventricular extrasystoles in bigeminism [5].

In the electrophysiological study, our patient presented the two classic conditions described for the appearance of this type of arrhythmia: 1) Retrograde unidirectional block of the slow pathway, otherwise the impulse that has gone down the fast pathway could go up the slow pathway and collide with the previous impulse and be extinguished; in our Case, there was no retrograde VA conduction, and 2) A very marked difference between the conduction time of the fast and slow pathways (AH2-AH1 390 ms, in this case), so that when the impulse conducted through the slow pathway reaches the common AV nodal pathway and the His-Purkinje system, these are not refractory.

Failure to perceive palpitations, or delayed diagnosis, has not infrequently led these patients to progress to tachycardia-induced dilated cardiomyopathy [6].

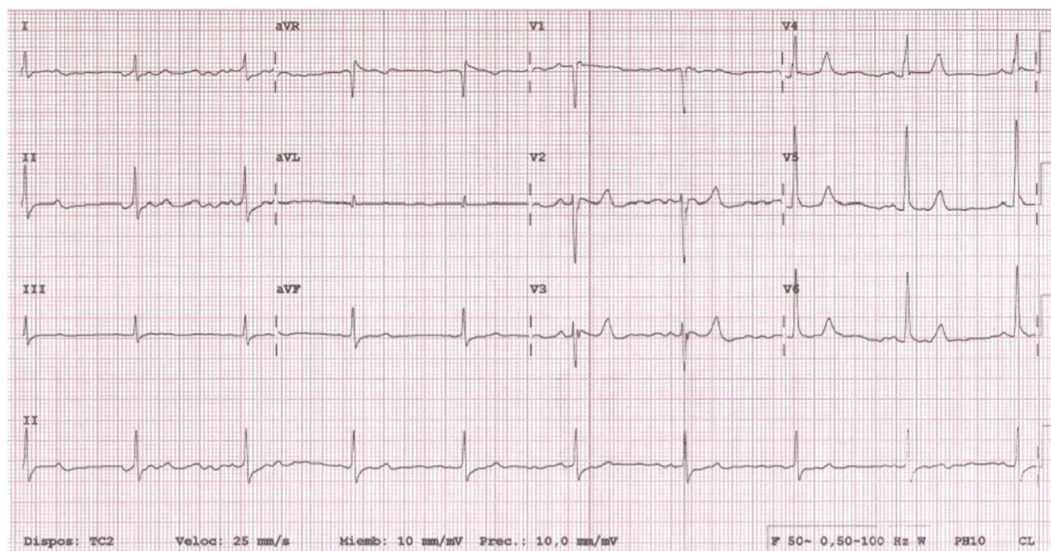
The response of DAVNNT to medication, including beta-blockers, flecainide, and amiodarone is very poor or absent [7], so the treatment of choice is slow pathway ablation.

DAVNNT should therefore be suspected in patients presenting with a regularly irregular tachycardia, with visible P waves of sinus morphology, with narrow or wide QRS. The presence of ventricular dysfunction is not uncommon.

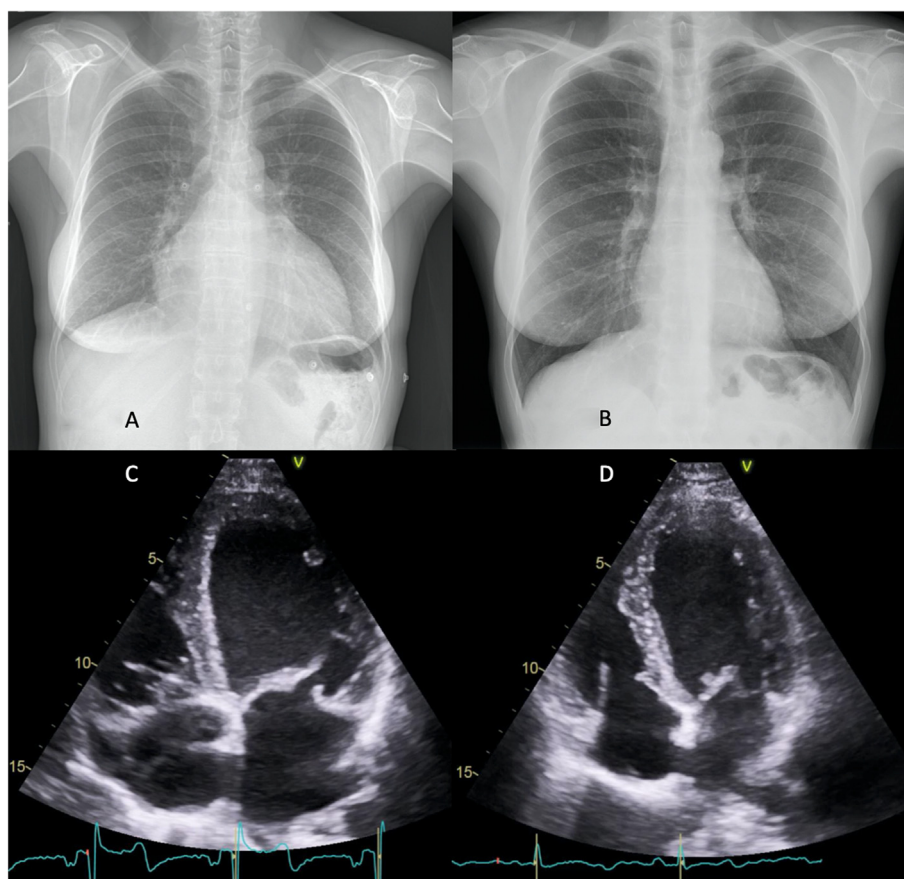


**Fig. 2.** A) Electrophysiological study. It is observed that each atrial activation (A), is followed by two hisian activations (H1, H2) and two ventricular activations (V1, V2). The intervals AH1 and AH2 remain constant. The difference between the conduction times of the fast and slow pathways (H1–H2 interval) is 390 ms. HRA; right atrium; His p, m, d: proximal, middle and distal His; RVd: right ventricle; CS p, 4, 3, 2, d: coronary sinus from proximal to distal. B) Absence of retrograde VA conduction during pacing from RV apex.





**Fig. 3.** ECG 6 months after ablation. Sinus rhythm with PR interval of 150 ms. Signs of left ventricular enlargement have disappeared.



**Fig. 4.** A) Chest X-ray on admission reveals cardiomegaly and signs of heart failure. B) Six months after ablation, the X-ray shows no alterations. C) Initial echocardiogram showed dilatation of the four chambers and severe biventricular dysfunction. D) Six months after ablation, the echocardiogram has normalized.

## Authors' contributions

Jose Amador Rubio Caballero: Writing-Original Draft and Supervision; Cecilia Marco Quirós and Verónica Artiaga de la Barrera: Visualization; Victoria Espejo Bares: Writing-Review & Editing.

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## Declaration of competing interest

None.

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