

## Dextrocardia with Situs Inversus Associated with Non-Compaction Cardiomyopathy

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### Introduction

Dextrocardia is a rare condition usually diagnosed incidentally and associated with other congenital anomalies. It is characterized by the position of the heart in the right hemithorax, with its base-apex axis directed to the bottom right. Its incidence associated with situs inversus, in the general population, is 1:10.000<sup>1</sup>. Non-compaction cardiomyopathy (non-compaction CMP), in turn, has an incidence ranging from 0.014 to 1.3%<sup>2</sup>, may occur alone or associated with other congenital defects and results from failure in the process of compaction of myocardial fibers, resulting in the persistence of trabeculations and deep recesses that communicate with the ventricular cavity<sup>2</sup>. A rare association of these two diseases is described. Cases similar to the one reported have not been found<sup>3-5</sup>.

### Case report

A 53-year-old male patient who, in the past 2 years, has presented brief episodes of collapse and presyncope when performing daily activities, with recent worsening in the frequency of symptoms. Regarding the habits of life, the patient was sedentary, smoker and social alcoholic. He sought a general practitioner who requested examinations for initial evaluation. The resting electrocardiogram (ECG) revealed sinus bradycardia (heart rate – HR: 39 bpm), deviation of the electrical axis to the right and right bundle branch block (Figure 1). The exercise test was considered abnormal due to arrhythmia (ventricular extrasystoles with periods of bigeminy during exercise and recovery). 24-hour Holter monitoring showed average heart rate of 37 bpm (minimum of 25 and maximum of 81 bpm) with sinus rhythm interspersed with junctional rhythm. Atrioventricular conduction was within the normal range and intraventricular conduction revealed right bundle branch disorder. The patient presented 11,596 pauses lasting

longer than 2 seconds, rare and isolated monomorphic ventricular extrasystoles and an episode of non-sustained supraventricular tachycardia.

Given the clinical suspicion of sinus node disease, the patient was referred to a cardiologist, who continued the investigation. Transthoracic echocardiography was requested, which revealed dextrocardia (presence of situs inversus, heart positioned in the right hemithorax with apex facing right); images suggestive of trabeculations in the left ventricle (LV); moderate LV diastolic dysfunction with preserved systolic function; left atrial enlargement (57 mm); mild mitral regurgitation.

For diagnostic elucidation and evaluation of cardiomyopathy, CMRI was performed and cine sequences were obtained (balanced steady-state free precession — b-SSFP), anatomy with black-blood turbo spin-echo sequences, with T1 and T2 weighting with and without fat suppression (TSE BB-PD and STIR) and delayed enhancement (turbo-field echo with inversion pulse and recovery, 10 minutes after injection of 0.2 mmol/kg of gadolinium contrast; TE: 6.1; TR 3.0). Besides confirming dextrocardia associated with situs inversus, a slight increase in the left ventricular cavity associated with excessive trabeculations in the middle and apical segments of the inferior, lateral and anterior LV walls was observed. The non-compacted/compacted myocardium relationship was higher 2.3 and the trabecular mass corresponded to 38% of the total left ventricular mass compatible with diagnosis of non-compacted CMP. There were no direct or indirect signs of restriction, thrombus or pericardial abnormalities. There were no areas of fibrosis or infarction detectable in the sequences after gadolinium injection (Figure 2).

Electrophysiological study was performed; programmed ventricular stimulation caused the induction of ventricular fibrillation (two extrasystoles).

Cardioverter defibrillator (CD) was implanted as a normal procedure. Four months after the CD implantation, the patient developed deep vein thrombosis in the left upper limb. The patient was anticoagulated and is currently asymptomatic, with atrial pacing in 100% of the time and no sustained ventricular arrhythmias, treated with amiodarone 200 mg and aspirin 100 mg.

### Keywords

Dextrocardia; Heart Defects, Conenital; Situs Inversus; Magnetic Resonance Spectroscopy; Cardiomyopathies.

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### Discussion

Dextrocardia is a rare congenital malformation characterized by displacement of the heart to the right hemithorax with its base-apex axis oriented to the bottom right<sup>1</sup>. It is caused by factors intrinsic to the heart and there is no relationship with

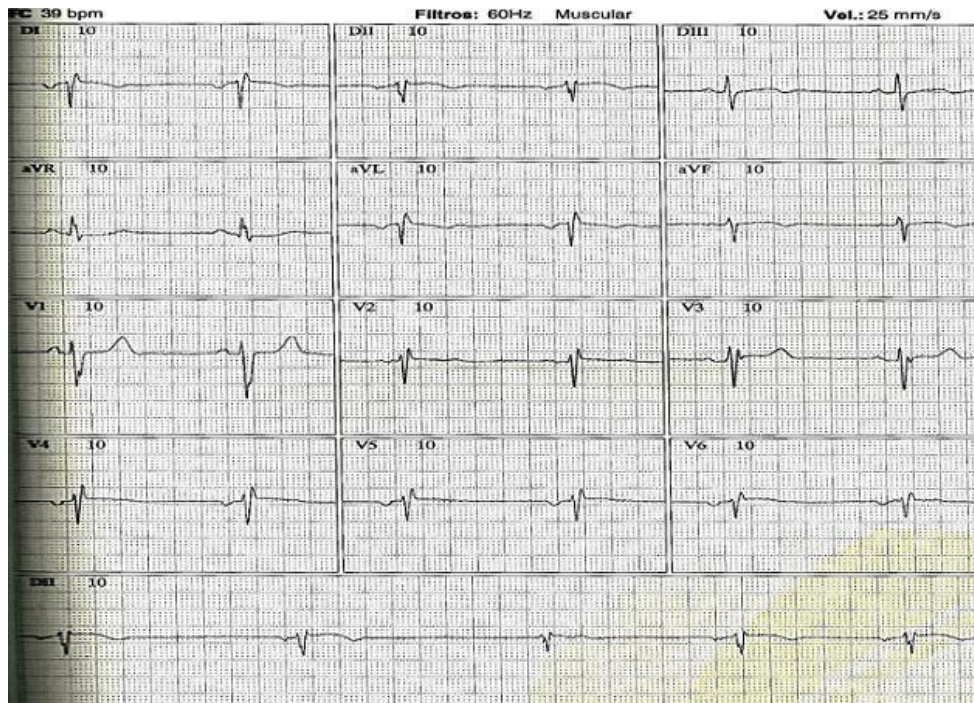


Figure 1 - 12-lead electrocardiogram at rest revealing sinus bradycardia (heart rate: 39 bpm), deviation of the electrical axis to the right and right bundle branch block.

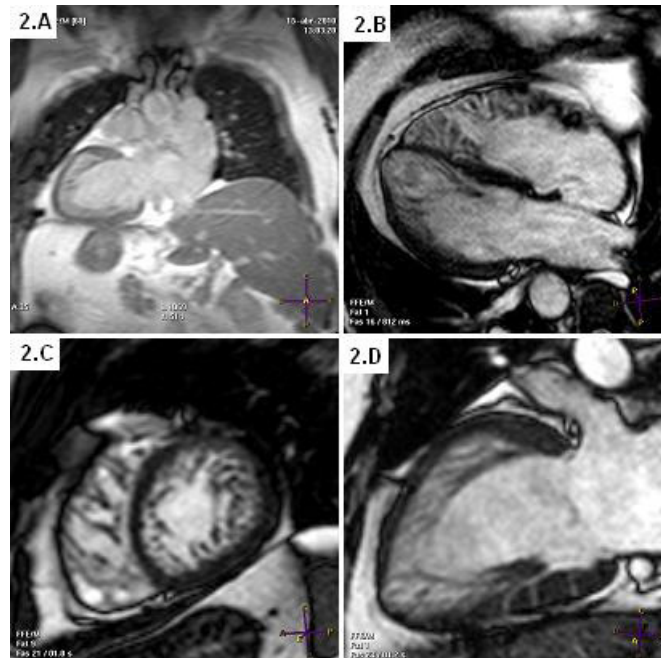


Figure 2 - Turbo field echo sequence demonstrating situs inversus characterized by dextrocardia, and transposition of the abdominal organs (2A). Cine sequences (b-SSFP) in four chambers (2B), two chambers (2C) and short axis (2D), revealing severe trabeculation in middle-apical segments of lower, lateral and anterior walls.

## Case Report

extracardiac abnormalities. It has a variable intracardiac anatomy and is usually associated with other congenital abnormalities, such as defects in the interatrial and interventricular septa, abnormalities of the pulmonary artery and univentricular heart<sup>6</sup>.

Dextrocardia with situs solitus (dextroversion) corresponds to isolated malposition of the heart and other organs in normal position<sup>4-6</sup>. In dextrocardia with situs inversus, there is formation of a mirror image of the heart, great vessels and other organs, preserving the relationship between them. This is an approximate incidence of 1:10.000<sup>1</sup>, while in the former one it is of 1:2.800<sup>6</sup>.

The non-compacted CMP is a rare congenital heart disease with an incidence ranging between 0.014 and 1.3% marked by abnormality of the endomyocardial morphogenesis due to an arrest of compaction of its fibers<sup>2</sup>. It is characterized by the presence of a number of prominent trabeculae with intertrabecular recesses which penetrate deeply through the ventricular myocardium forming a loose network of intertwined muscle fibers. Isolated impairment of LV occurs in 59% of patients and biventricular impairment occurs in 41%. It is presented in sporadic and familial forms. Only in the latter, genes related to the disease were described<sup>7</sup>.

The natural history of the disease is not yet well established. Patients with non-compacted CMP may remain asymptomatic throughout their lives or develop signs and symptoms of heart failure (53%), ventricular tachycardia (41%) or thromboembolic events due to thrombus in the atria or intertrabecular recesses (24%). Sudden arrhythmic death is the leading cause of mortality<sup>2,4</sup>.

Doppler echocardiography is the diagnostic procedure of choice in the evaluation of cardiomyopathies, and the incorporation of new techniques such as tissue Doppler, strain and strain rate imaging, and speckle tracking has been useful in distinguishing normal myocardial trabeculation and non-compacted CMP<sup>2</sup>. Jenni et al<sup>8</sup>, based on observational studies, described with good accuracy, the following criteria for the echocardiographic diagnosis of non-compacted CMP: (a) presence of numerous and prominent trabeculae (at least four in the apical area) with deep intertrabecular recesses; (b) two-layer ventricular wall where the thickness of the non-compacted layer is at least two times greater than the thickness of the epicardial compacted layer; (c) demonstration, through color Doppler, of the presence of blood flow directly from the ventricular cavity into the intertrabecular recesses; (d) common involvement of the middle lateral, lower middle or apical LV areas; and (e) absence of other cardiac abnormalities. However, echocardiography is operator-dependent and still has certain limitations, such as misdiagnosis of non-compacted CMP in normal individuals with fine trabeculations<sup>9</sup>. Thus, especially in cases with inadequate echocardiographic window, the combination of imaging

methods is useful for confirming or excluding the diagnosis of non-compacted CMP. Hence, the subsequent use of CMRI is valuable because it has greater diagnostic accuracy due to three-dimensional cardiac imaging. It has been shown, in a sample of seven patients with non-compacted CMP, that a  $> 2.3$  ratio between the non-compacted myocardial layer and compacted myocardial layer during diastole has 86% sensitivity and 99% specificity for the diagnosis of this pathology<sup>10</sup>. Jacquier et al<sup>10</sup> observed that the presence of trabecular mass corresponding to more than 20% of the total LV mass could distinguish, with satisfactory accuracy, patients with non-compacted CMP of normal controls, with sensitivity and specificity of 93.7%<sup>10</sup>. In this case, the two criteria above were found.

The combined use of two imaging procedures, including echocardiography and CMRI, has been suggested to confirm or exclude the diagnosis of non-compacted CMP.<sup>2</sup> However, it is important to note that the diagnosis of non-compacted CMP should not be solely based on the diagnostic criteria of imaging methods. Kolhi et al<sup>9</sup> showed that about 24% of patients with dilated CMP and 8% of normal controls fulfill diagnostic criteria for non-compacted CMP when the diagnostic criteria of imaging methods are used alone. This must be, therefore, a diagnosis of exclusion.

Friedman et al<sup>4</sup> and Baskurt et al<sup>5</sup> reported cases of association dextroversion and non-compacted CMP in the LV while Grattan et al<sup>3</sup> demonstrated dextroversion associated with biventricular non-compacted CMP. No studies describing the association of dextrocardia with situs inversus and myocardial non-compaction were found. This seems to be the first case described in the literature, showing that rare presentation.

### Author contributions

Conception and design of the research and Acquisition of data: Gonçalves LFG, Souto FMS, Faro FN, Mendonça RC, Oliveira JLM, Sousa ACS; Analysis and interpretation of the data: Gonçalves LFG, Mendonça RC, Oliveira JLM, Sousa ACS; Writing of the manuscript and Critical revision of the manuscript for intellectual content: Gonçalves LFG, Souto FMS, Faro FN, Mendonça RC, Sousa ACS, Oliveira JLM.

### Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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### Study Association

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