Dextroversion and Noncompaction

Josef Finsterer[1] e Claudia Stöllberger[2]

Krankenanstalt Rudolfstiftung[1], 2° Departamento Médico, Krankenanstalt Rudolfstiftung[2], Viena, Áustria

Dear Editor,

With interest we read the article by Gonçalves et al[1] about a patient with dextrocardia and left-ventricular hypertrabeculation/noncompaction (LVHT)[1]. We have the following comments and concerns.

Dextrocardia in association with LVHT has been repeatedly reported[2].

The exact pathomechanism of LVHT has not been discovered. Though the failing compaction process may play a role, there are also patients in whom LVHT developed during lifetime (acquired LVHT).

Which was the cause of collapses and presyncope? Did the patient undergo cerebral magnetic resonance imaging (MRI) to rule out cardioembolic stroke originating from the inter-trabecular spaces, a complication repeatedly reported in LVHT? Which were the results of carotid ultrasound investigations?

The patient required implantation of an implantable cardioverter defibrillator (ICD) because of inducible ventricular fibrillation. Did the ICD ever discharge during follow-up?

LVHT is frequently associated with neuromuscular disorders (NMDs). Was the patient ever seen by a myologist to rule out a NMD? Neuromuscular disorders, such as periodic paralysis or Marden-Walker syndrome, were associated with dextrocardia.

LVHT is frequently familial. Were other family members investigated for LVHT? Was LVHT found in any of the first-degree relatives? Did any of the family members develop cardiac symptoms?

The patient received gadolinium for cardiac MRI. Did radiologists observe late enhancement, which may indicate myocardial fibrosis? Myocardial late enhancement can be typically found in dystrophic NMD patients with myocardial affection[3].

The diagnostic criterion of at least four LV trabeculations on echocardiography for diagnosing LVHT on echocardiography has not been provided by Jenny et al[4] but by Stöllberger et al[5].

The figure of 24% of LVHT patients to experience embolic stroke from inter-trabecular thrombi is high and not referenced.

Though cardiac MRI is useful to detect LVHT, it is not diagnostic in each case. Vice-versa echocardiography may not always detect LVHT, most frequently due to poor image quality, due to marked LV dilatation, or due to severe myocardial thickening[6].

There are a number of sporadic LVHT cases in which mutations in single genes, such as the SCNA5 or G4.5 gene, have been reported, suggesting that not only familial LVHT is associated with mutated genes.

Overall, LVHT remains an enigmatic condition with several practical implications for the management of these patients. In LVHT it is essential that all clinical and scientific aspects are considered not only to improve the outcome of these patients but also to contribute to the clarification of the pathogenetic background.

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Keywords

Cardiomyopathies / pathology; Heart Failure; Stroke; Dextrocardia.

Mailing Address: Josef Finsterer •
Postfach 20. Postal Code 1180, Vienna, Austria, Europe
E-mail: fifigs1@yahoo.de
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Reply

In response to the letter, we agree that noncompacted cardiomyopathy is an enigmatic condition with several pathophysiological aspects not completely elucidated, making its characterization by the use of the major diagnostic tools, echocardiography and cardiac magnetic resonance imaging, difficult. Thus, as highlighted in the case report, the use of both methods has been postulated as often required to either confirm or exclude the diagnosis of that condition.

a) The patient underwent neither neurological assessment nor cerebral magnetic resonance imaging, because the episodes of lipothymia and presyncope were attributed to bradyarrhythmia, and the patient became asymptomatic after cardioverter defibrillator implantation. Duplex carotid ultrasound showed no significant atherosclerotic lesion;

b) One single inappropriate discharge was identified in the presence of atrial flutter;

c) The patient has no symptoms of neuromuscular disorders; however, he has not been examined by a myologist;

d) There is no report of cardiac symptoms of other family members, although, despite the high interest of the assisting team, no systematic cardiological investigation has been conducted in all first-degree relatives;

e) As reported in the article, after gadolinium administration, no late enhancement, indicating fibrosis, was identified;

f) The occurrence of thromboembolic events, due to thrombi from the atria and intertrabecular recesses, in up to 24% of patients with noncompacted myocardiopathy has been reported by Friedman et al[1] and Baskurt et al[2], references cited in the case report, as well as by Oechslin et al[3].

Sincerely,

Fernanda Maria Silveira Souto
Joselina Luzia Menezes Oliveira
Antônio Carlos Sobral Sousa

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