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LETTER TO THE EDITOR

Reply to the Letter to the Editor entitled ''Acute heart failure from noncompaction requiring emergency heart transplantation''

Resposta à Carta ao Editor intitulada ''Insuficiência cardíaca aguda associada a não compactação requerendo transplantação cardíaca emergente''

We appreciate the interest of Josef Finsterer and Sinda Zarrouk-Mahjoub[1] in our recent original article entitled ''Isolated left ventricular noncompaction causing refractory heart failure'',[2] and would like to thank them for their pertinent comments.

First of all, with regard to the concern about the patient's father, we agree that it would be important to have information about his echocardiographic exams or even an autopsy study. However, his father had been treated from disease onset in another hospital and we did not have access to these exams. Besides, the family did not keep the father's previous exams, so it was not possible to analyze them. The family had been contacted and insisted he had no episodes of atrial fibrillation, stroke, cardioembolism, arrhythmias or epilepsy.

Second, concerning the index patient's seizures, these occurred during use of high doses of vasoactive drugs, which could have contributed to atrial fibrillation episodes (recorded on ECG). The seizures really occurred, but only once (with no previous episode in the patient's life), and were considered to be a sign of low cardiac output (LCO) causing cerebral hypoxia due to cerebral hypoperfusion. They were tonic-clonic, lasted about two minutes, and ceased after intravenous benzodiazepine administration; at this time, an electroencephalogram was not recorded. She was then treated prophylactically with phenytoin. An intracardiac thrombus 17 mm×14 mm in size was detected in the left atrium, as reported in the main article.[1] Thrombocytopenia and hepatopathy developed previous to the seizures and were attributed to the use of prophylactic heparin and systemic inflammatory response syndrome (SIRS), in combination with congestive heart failure and LCO.

It is important to note that after heart transplantation and hospital discharge, the patient presented in outpatient care complaining of loss of consciousness, not sudden, but progressively and constantly. Psychological and physical (cerebral magnetic resonance imaging and EEG) investigation revealed nothing. She then underwent tilt testing; a diagnosis was made of vasovagal syndrome and she was advised to make changes in her behavior in order to prevent syncope episodes.

Furthermore, considering chromosomal defects, we understand the role of genetics in this disease[3] and that it is inherited in at least 30-50% of patients, and several genes that cause left ventricular non-compaction (LVNC) have been identified. These genes seem generally to encode sarcomere (contractile apparatus) or cytoskeletal proteins.[4] However, the patient was not investigated for gene mutations and it was presumed to be a congenital abnormality, as reported.

The rest of her family were investigated by echocardiography and screened for similar myocardial changes but no evidence of myocardial hypertrabeculation/non-compaction was found. There were also no clinical or laboratory indications of neuromuscular disease in the index case or any of her relatives, such as easy fatigability, exercise intolerance, muscle wasting or weakness, myotonia or adverse reactions to general anesthesia. We have not screened for chromosomal defects as this is not a routine procedure. We understand the importance of these disorders particularly in acquired hypertrabeculation, and that up to 80% of patients with LVNC suffer from a neuromuscular disorder.[5–7]

Concerning anatomopathology, there were non-specific findings of hypertrophy and mild degeneration of myocardial fibers, with no fibrosis or subendocardial fibroelastosis.

The patient currently has no new symptoms of heart failure and has not developed LVNC in the transplanted heart.

To summarize, we believe that the genesis of isolated LVNC is still not completely understood; further investigation of the disease and of patients is required, and genetic studies in particular should be the aim of future research.

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Conflicts of interest

The authors have no conflicts of interest to declare.

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