

Letter to the Editor

Cerebral, Cardiac, or Combined Cause of Syncope in Noncompaction

Josef Finsterer, MD, PhD

Krankenanstalt Rudolfstiftung, Vienna, Austria

*Corresponding Author

Josef Finsterer, MD, PhD

Keywords: dilated cardiomyopathy, noncompaction, syncope, arrhythmia, implantable cardioverter defibrillator.

In a recent article, Glancy *et al.*, reported a 55yo male with dilated cardiomyopathy (dCMP) and left-ventricular hypertrabeculation / noncompaction (LVHT), who was admitted for syncopal episodes. Syncope disappeared after implantation of an implantable cardioverter defibrillator (ICD) (Glancy, D. L. *et al.*, 2017). The study raises a number of comments and concerns.

A patient with a history of recurrent syncope requires neurological work-up. We thus should be informed about the neurological history, the neurologic exam, about the findings on cerebral MRI, electroencephalography, and on carotid ultrasound, which have not been described in the publication (Glancy, D. L. *et al.*, 2017). Even if cardiac causes of syncope are evident, the neurologist must be involved in the work-up of syncope for several reasons: 1. syncope may have an exclusively neurologic cause, 2. Syncope may have both, a neurological and cardiac cause, and 3. a patient with dCMP, heart failure, arrhythmias, and LVHT may be prone to experience cardio-cerebral disease, such as ischemic, strokes, cerebral bleeding, syncope, brain abscess, meningitis, metastasis, dementia, or aneurysm formation (Finsterer, J., & Stoellberger, C. 2016).

LVHT has not been first described by Engberding *et al.*, in 1984 but by Feldt *et al.*, In 1969 at autopsy of a 3 months-old white female with situs inversus and biventricular, bizarre, and spongy myocardium (Finsterer, J. *et al.*, 2017). In 1975, Westwood *et al.*, presented a figure about a case of endocardial fibroelastosis which unambiguously showed biventricular hypertrabeculation (Finsterer, J. *et al.*, 2017). In 1975, the post-natal persistence of a

spongy myocardium, initially recognised during fetal development, was first reported by Dusek *et al.*, (2017). Engberding *et al.*, however, was the first to visualise LVHT on echocardiography.

LVHT is not a genetic disease. Though repeatedly reported in association with genetic disorders, particularly neuromuscular disorders and chromosomal defects, it has never been convincingly confirmed that a certain mutation unambiguously was causative for LVHT. Arguments against a causal relation are that in the majority of the cases, LVHT does not segregate with a mutation in a particular family, that LVHT can be acquired, developing during life, that not all patients carrying a particular mutation may develop LVHT, and that LVHT may occasionally disappear. An argument, however, in favour of a genetic cause is that occasionally LVHT occurs as a familial disease (Finsterer, J. 2009).

Though the presented patient had a history of syncope and cardiac disease, the authors do not explain why this particular patient fainted. Was it due to bradycardia (a heart rate of 37 not necessarily results in syncope), supra- or ventricular arrhythmias (atrial fibrillation, ventricular runs), or low output failure? Which arrhythmias did the ICD record during the 4y of follow-up? Were severe arrhythmias recorded in the absence of a syncope or a shock?

Overall, this interesting case shows that syncope in a patient with LVHT require neurological investigations, that first-degree relatives of an index case with LVHT need to be cardiologically and in case of LVHT also neurologically investigated, and that the cause of syncope needs to be unambiguously

Quick Response Code



Journal homepage:

<http://www.easpublisher.com/easims/>

Article History

Received: 23.10.2019

Accepted: 02.11.2019

Published: 16.11.2019

Copyright © 2019: This is an open-access article distributed under the terms of the Creative Commons Attribution license which permits unrestricted use, distribution, and reproduction in any medium for non commercial use (NonCommercial, or CC-BY-NC) provided the original author and source are credited.

identified. In patients with syncope or episodes of syncopes cardiac, neurologic, and non-cardiac, non-neurologic causes need to be considered.

REFERENCES

1. Glancy, D. L., Helmcke, F. R., & Hoang, A. P. (2017). Bradycardia, Syncope, and Left Ventricular Noncompaction Cardiomyopathy. *The American journal of cardiology*, 120(4), 716-717.
2. Finsterer, J., & Stoellberger, C. (2016). Neurological complications of cardiac disease (heart brain disorders). *Minerva medica*, 107(1), 14-25.
3. Finsterer, J., Stoellberger, C., & Towbin, J. A. (2017). Left ventricular noncompaction cardiomyopathy: cardiac, neuromuscular, and genetic factors. *Nat Rev Cardiol*, 14, 224-237.
4. Finsterer, J. (2009). Cardiogenetics, neurogenetics, and pathogenetics of left ventricular hypertrabeculation/noncompaction. *Pediatric cardiology*, 30(5), 659.