

Is Sildenafil a Therapeutic Option for Noncompaction?

Josef Finsterer and Claudia Stöllberger

Krankenanstalt Rudolfstiftung, Vienna, Austria

To the editor

With interest we read the article by Redondo et al¹ about a 6yo male with heart failure and left-ventricular hypertrabeculation/noncompaction (LVHT)¹. We have the following comments and concerns.

Though LVHT is congenital in the majority of the cases, there are some cases in which LVHT is not present on previous echocardiographies (acquired LVHT)². In such cases, LVHT may be truly absent or hidden. Hidden LVHT may occur if there is severe myocardial thickening or severe dilatation.

We do not agree that pulmonary embolism is a major complication of LVHT¹. LVHT is an abnormality of the left ventricle. Thus, LVHT is frequently associated with ischemic stroke or systemic embolism due to emboli originating from the intertrabecular recessus of the left ventricular myocardium.

LVHT is not directly attributable to a single genetic defect. It is rather a secondary reaction to primary affection of the heart by a genetic or chromosomal defect³. Typically, it does not occur in each carrier of a mutation, but only in some rare cases, without knowing the cause of this selectivity.

The presented case is interesting for the beneficial effect of sildenafil for heart failure. Did sildenafil also have a beneficial effect on the trabeculations? Did the amount of trabeculae

regress? Did the ratio non-compacted layer to compacted layer decrease during therapy?

The authors mention that LVHT was first described in 1932 on autopsy. It would be interesting to know the reference of this observation, since there is ongoing debate about the first description of LVHT.

LVHT is frequently associated with neuromuscular disorders (NMDs). Was the patient ever seen by a myologist to rule out a NMD? Did the patient present with features of a NMD, such as weakness, wasting, fatigue, exercise intolerance, or double vision? Was the family history positive for NMD?

Occasionally, LVHT is associated with pulmonary hypertension⁴. Was the pressure of the pulmonary artery measured, and, if increased, did sildenafil also have a beneficial effect on this parameter, since it is also given in this indication?

We do not agree that LVHT in adults occurs only in the absence of other concomitant structural cardiac diseases. In adults, LVHT has been described in association with Ebstein's anomaly or aortic root dilatation⁵.

Overall, there is a need to thoroughly investigate patients with LVHT, not only for cardiac disease but also for extra-cardiac manifestations. A comprehensive appreciation is necessary to optimize the management of LVHT patients.

Keywords

Heart Defects, Congenital; Heart Failure; Cardiomyopathy, Dilated/complications; Phosphodiesterase Inhibitors/therapeutic use.

Mailing Address: Josef Finsterer •

Postfach 20, Vienna. 1180, Austria

Email: ffigs1@yahoo.de

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