FACTS ABOUT AND UNCERTAINTIES OF LEFT VENTRICULAR HYPERTRABECULATION / NONCOMPACATION

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To the Editor:

The interesting review by Towbin et al. [1] about left-ventricular hypertrabeculation / noncompaction (LVHT) raises concerns.

Concerning the history of LVHT, Grant et al. did not provide the first description of LVHT [2]. In his patho-anatomic description intertrabecular spaces communicated freely with coronary vessels including arteries, veins, and capillaries [2]. More likely, LVHT was first described by Feldt et al. in 1969 or Westwood et al. in 1975 [2].

We do not agree that there are different forms of LVHT. LVHT may be associated with other congenital or acquired cardiac abnormalities and may be complicated by heart failure, thrombus-formation / embolism, arrhythmias, or sudden cardiac death. There is no “benign” LVHT since over time or suddenly LVHT may change from a “silent” abnormality to a problem with increased morbidity and mortality [3].

The authors’ fade-out that LVHT is associated with neuromuscular disorders (NMDs) or chromosomal defects in the majority of cases [4]. Though the association between LVHT and NMDs is unsolved, the NMD aspect must be recognised since it is of prognostic relevance [3].

Currently, there is no proof for a causal link between any mutation and LVHT. More arguments exist for LVHT to represent a myocardial reaction to hemodynamic, contraction, vascular, conduction, or autonomic alterations than for the genetic hypothesis. LVHT may not only be associated with mutations in the TAZ, NKK2-5, MYH7, LDB3/ZASP, ACTC1, TNNT2, MYBPC3, TPM1, TNNI3, LMNA, and SCN5A genes but also in the dystrophin, DMPK, α-DTNA, RYR1, ITGA7, MYH7B, LAMP2, GAA, GBEI, MADD, SDH, COL7A1, MMACHC, PMP22, FXN, β-globin, PLEC1, HCN4, and GLA genes, and mtDNA genes [5].

Finally, the authors did not mention that LVHT may be acquired after birth, such as in NMDs, athletes, or pregnant females, challenging the congenital hypothesis of LVHT [6].

References :


[5.] Finsterer J, Zarrouk-Mahjoub S. Considerations about the genetics of left ventricular hypertrabeculation / noncompaction. Cardiol Young 2015;(in press).