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Letter to the Editor

Cardiac and extra-cardiac abnormalities associated with noncompaction

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In their eminent article, Saraiva et al. reported about a 34-yo-male with heart-failure, AV-block-I, and left-bundle-branch-block (LBBB), reduced systolic function and left-ventricular hypertrabeculation / noncompaction (LVHT) [1]. We have the following comments and concerns.

The authors claim that the patient had LBBB. Assuming that the registration speed was 25mm/s, QRS-duration is borderline in figures 1 and 2. There is no AV-block I. Deviation of the electrical axis to +30° and -145° is not reconstructible.

Though ST-elevation in V1-V3 could be attributed to the assumed LBBB it would be interesting if the patient had coronary-heart-disease. Did he complain about angina or were CK and troponin-values ever elevated? Did he have a history of myocardial infarction? Did he undergo stress-testing, myocardial scintigraphy, or coronary angiography?

Which is the cause of dilative cardiomyopathy and heart-failure? Was there any indication for chronic alcoholism, coronary-heart-disease? borreliosis, viral infection, storage-disease, or neuromuscular disorder (NMD)?

The patient had a viral respiratory infection. Did he develop concomitant myocarditis or pulmonary embolism? Was there acute right-heart-strain on clinical examination, ECG, or echocardiography?

According to which definition was LVHT diagnosed? Öchslin's, Stöllberger's or Chin's criteria [2-4]? Does figure 2 show an apical or cross-sectional view? Which value had the ratio noncompaction / compacted layer?

LVHT is located in the apex and the interventricular septum. In our series of >100 LVHT-patients none showed affection of the medial or basal septum. How to explain the occurrence of septal involvement?

Was the patient ever investigated for NMD or other rare hereditary disease, like dystrophinopathy, dystrobrevinopathy, myotonic dystrophy, zaspopathy, myoadenylate-deaminase-deficiency, Charcot-Marie-Tooth-disease, mitochondrial disorder, Barth- syndrome, Friedreich-ataxia, Pompe's disease, Turner-syndrome, Ohtahara-syndrome, Roifman-syndrome, Noonan-syndrome, nail-patella-syndrome, Melnick-needles-syndrome, Melnick-Fraser-syndrome,

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MIDAS-syndrome, DiGeorge-syndrome, congenital adrenal hyperplasia, distal 4q-trisomy and distal 1q-monosomy, chromosome 1q43-deletion, distal chromosome 5q-deletion, trisomy 11, or trisomy 13 [5]?

Was the history positive for stroke/embolism, occasionally associated with LVHT? Which therapy was given, which were the results on the long-term follow-up, and which the outcome?
Rarely, LVHT develops during life. Were previous echocardiograms reviewed for LVHT? Did LVHT change concerning morphology and extent over time?

Were first degree family members investigated for LVHT? Since LVHT is suggestive of hereditary disease a family screening is strongly recommended to eventually clarify the nature of LVHT and associated underlying other disorders.

Overall, LVHT is usually not exclusively confined to the heart why more comprehensive investigations are mandatory in LVHT. If additional cardiac and extra-cardiac co-morbidity is not fully elucidated, the pathogenesis of LVHT will remain elusive.

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