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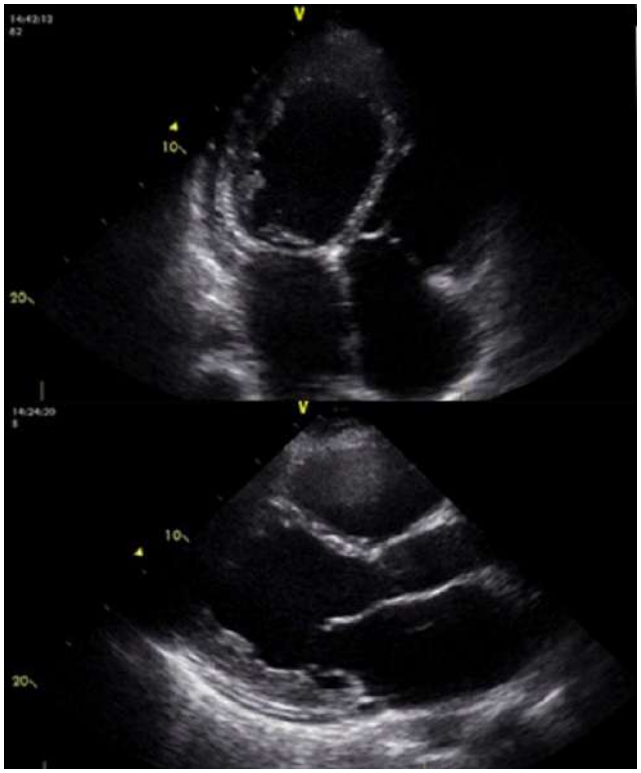
A sporadic or a familial case of left ventricular noncompaction?

D Bursacovschi[1]; R Negru-Cemortan[1]
[1]*Institute of Cardiology, Chisinau, Moldova (Republic of)*

Introduction: Left ventricular noncompaction (LVNC) is a rare congenital disorder that can be sporadic, but familial recurrence and associated dysmorphism has been reported. Diagnosis is challenging and often with a predilection for overdiagnosis. Progressive heart failure, ventricular arrhythmias and systemic embolic events are the classical triad of complications.

Clinical case description: We report a case of a 41-year-old male patient with signs of congestive heart failure during the previous 2 months. He had no chest pain, palpitations, or syncope. Physical examination revealed bilateral crackles in the lower-lung fields and edema of the lower extremities. The electrocardiogram showed a sinus rhyth, 85 beats/min and a few premature ventricular contractions. Ambulatory ECG (Holter) monitoring (24h) registered frequent premature ventricular contractions. Transthoracic echocardiography (TTE) showed a severe dilatation of all heart chambers, a severe reduced ejection fraction (EF 25%), diffuse hypokinesia and a lot of trabeculae located on the posterolateral wall of the left ventricle (LV) and apex (Picture 1, 2). The blood flow into the intertrabecular recesses. The Jenni (2001) criteria for the diagnose of LVNC where positive. The lateral wall of right ventricle was highly trabeculated and the contraction was poor. Third degree LV diastolic disfunction, moderate regurgitation of mitral and tricuspid valves, severe signs of pulmonary hypertension. The CMR analysis showed non-compact myocardium at the level of left and right ventricles, the ratio of non-compacted myocardium to compact myocardium was 5:1 at the postero-lateral wall and of 8:1 at the apex of the LV. From first-degree relatives, patient's father has no cardiac pathologies. Patient's mother was diagnosed with dilatative cardimyopathy (DCM) 7 years ago. The remodeled LV examined through repetead TTE made it difficult to exclude DCM. Further, we examened the patient's daughter (14 years old, asymptomatic), through TTE we identified small trabeculation localized on posterolateral wall and apex of the LV. The girl's CMR ratio of non-compacted myocardium to compact myocardium at the end of systole was 1:2.5 and the mass of trabeculation was 33% with a slightly decreased EF (47%). Patient's brother and nephew showed no cardiac abnormalities. Medical management of the patient included: β-blockers, ACEI, a loop diuretic, spironolactone, digoxin and warfarin. We obtained a significant improvement in dyspnea and a reduction of peripheral congestion at one month follow-up.

Questions: LVNC is a distinct cardiomyopathy or a morphological expression of different cardiac diseases? Still no gold standard exists for diagnosis and the applied criteria lack specificity, especially in children. So there is a large diagnostic 'grey area' between LVNC, DCM and even normal myocardium. It is unknown whether the prognosis could be improved by early diagnosis and treatment.



Picture 1