

Very rare association of hypertrophic cardiomyopathy with myocardial crypts, noncompaction of the left ventricle and myocardial bridge in a girl from a family with sudden cardiac deaths

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Objective: To report a particular case of a 14 yo girl admitted in our clinic after her father's sudden cardiac death at the age of 53, a few days after he was diagnosed with hypertrophic cardiomyopathy. In the father's family there were several sudden cardiac deaths (SCD) at a very young age. The patient's cousin was recently diagnosed with hypertrophic cardiomyopathy.

Material and methods: The young girl performed clinical investigations: cardiac examination, ECG, Echocardiography, Holter ECG, angio CT and angio MRI, laboratory tests and genetic examination.

Results: The patient was asymptomatic, with grade II cardiac murmur, normal blood pressure, femoral pulse present. Impressive deep Q waves in limb leads and left ventricular hypertrophy was registered on ECG. Echocardiography revealed concentric hypertrophic cardiomyopathy with deep myocardial crypts in the interventricular septum, well seen after contrast examination, but also noncompaction of the left ventricle. Angio MRI confirmed the lesions, well describing the noncompaction and the crypts and angio CT discovered a tiny myocardial bridge on anterior descending coronary artery at this moment with no obstruction. The familial pedigree demonstrated sudden cardiac deaths in a very young age in father's sister, brother, father and hypertrophic cardiomyopathy diagnosed in father's sister son. Despite that on the girl's Holter ECG there were no PVC, the decision after guidelines was to implant this girl with an ICD (implantable cardioverter defibrillator), to prevent SCD.

Conclusions: Hypertrophic familial cardiomyopathy represent a risk factor for SCD. Myocardial crypts are new features of the disease, described in the literature, but with unknown importance at this moment. Noncompaction cardiomyopathy may induce sudden cardiac deaths due to the complications. Tiny myocardial bridge is irrelevant now. Hypertrophic familial cardiomyopathy with myocardial crypts, associated with noncompaction of the left ventricle and myocardial bridge are very rare associated and till now they were not reported in the literature in this association and when familial SCD exist in the family, increase the risk of SCD of the patient. ICD is the only option to prevent sudden death. Genetic tests were performed and the girl was implanted with a cardiacdefibrillator.