

Aims and objectives

Left ventricular non-compaction (LVNC) is a cardiomyopathy characterized by a thin, compacted epicardial layer and an extremely thick endocardial layer with prominent trabeculation and deep recesses that communicate with the left ventricular cavity but not with the coronary circulation. The prevalence of this disorder in the general population is estimated between 0,5-0,25% for year. Left ventricular non-compaction is morphologically characterized by numerous prominent trabeculations and deep intertrabecular recesses. It is also known as “spongy myocardium” or “persistent embryonic myocardium”, and it is probably secondary to an arrest in the normal process of myocardial compaction during fetal life. In this retrospective study we want to test the diagnostic accuracy of cardiovascular magnetic resonance (CMR) imaging in differentiating the left ventricle non-compaction disease from the hypertrabeculated left ventricular of β -Thalassemia Major (β - TM). The cardiovascular magnetic resonance is used to differentiate the left ventricle non-compaction from other pathological and physiological conditions characterized by a prominent left ventricular trabeculae but β – Thalassemia Major, in whom hypertrabeculated left ventricle has also been described, has not been considered so far as a potential differential diagnosis.

Methods and materials

We retrospectively analyzed the cardiovascular magnetic resonance cine images of 10 patients with a previously diagnosed left ventricle non-compaction and 38 patients with β - Thalassemia Major. Two cardiovascular magnetic resonance diagnostic criteria were applied at the end-diastole: a ratio of non-compacted to compacted myocardium (NC/C ratio) >2.5 at a segmental level and a percentage of trabeculated left ventricular mass $>20\%$ of global left ventricular mass.

Results



Fig. 1: Jacquier criteria: trabeculated mass $> 20\%$ of the global mass.

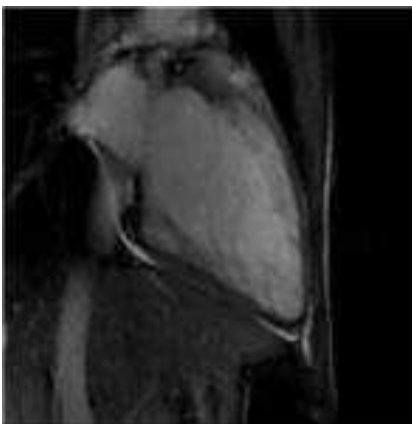


Fig. 2: Petersen criteria: maximal NC/C $> 2,3$.

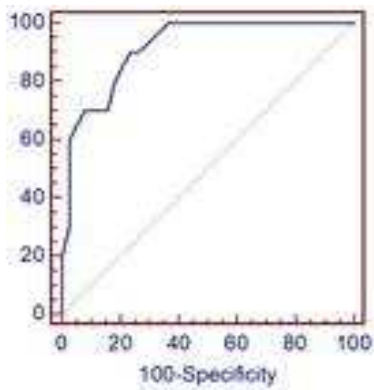


Fig. 3: NC/C ratio.

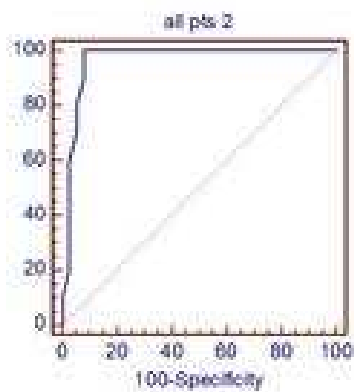


Fig. 4: Percentage of trabeculated LV mass.

The fifty percent of the β - Thalassemia Major patients had at least one positive Non Compacted/Compacted ratio segment. Although the areas of non-compaction defined by the Non Compacted/Compacted ratio were less frequent in the β - Thalassemia Major than in left ventricle non-compaction patients (7% vs. 37% of overall myocardial segments, $P < 0.0001$), they had a similar distribution within the left ventricle (predominant at the apex and postero-lateral wall, uncommon at the septum) which precluded a differential diagnosis. A Non Compacted/Compacted ratio of >2.5 showed a low specificity (58%) to distinguish the left ventricle non-compaction from the β - Thalassemia Major whereas a trabeculated left ventricular mass $>20\%$ was more accurate (sensitivity 100%, specificity 87%). The best specificity (92%) was obtained with a trabeculated left ventricular mass percentage of $>26\%$.

Conclusions

The differentiation of left ventricle non-compaction from the β - Thalassemia Major patients, probably may depend on the selected cardiovascular magnetic resonance criteria. In this study of population, the percentage of the trabeculated left ventricular mass showed to be better than the Non Compacted/Compacted ratio.

References

- 1) *Jacquier A, Thuny F, Jop B et al. Measurement of trabeculated left ventricular mass using cardiac magnetic resonance imaging in diagnosis of left ventricular non-compaction. Eur Heart J 2010; 31: 1098-104*
- 2) *Petersen SE, Selvanayagam JB, Wiesmann F et al. Left ventricular non-compaction. Insight from cardiovascular magnetic resonance imaging. J Am Coll Cardiol 2005; 46:101-5.*