P5273

Trabecular complexity as a subclinical structural alteration in Fabry cardiomyopathy: a cardiac magnetic resonance study

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Background: Heart involvement represents the main cause of death in Fabry Disease (FD), thus its early detection is important to define the optimal therapeutic strategy. Recently, a disproportionate increase in myocardial trabeculation has been described in FD by cardiac magnetic resonance (CMR), even in early (prehypertrophic) stage of the disease. In addition, CMR with T1 mapping can identity the presence of myocardial sphingolipid storage (causing lowering of native T1 values) in more than 50% of FD patients with no LVH. However, it is not clear whether a relationship exists between trabecular complexity and sphingolipid storage in FD.

Aim: To explore the association between myocardial trabecular complexity, quantified by endocardial border fractal analysis, and sphingolipid storage, described by CMR T1 mapping, in different stages of Fabry cardiomyopathy.

Methods: Study population included 60 subjects: 15 FD patients with no detectable signs of cardiac involvement (no LVH, normal T1; 2 M, age 30.6±14; Group 1); 15 FD patients with early sphingolipid storage (no LVH, low T1; 9 M, age 33±9.6; Group 2); 15 FD patients with LVH (11 M, age 53.5±9.6; Group 3); 15 healthy controls (9 M, age 34±10). Patients and controls underwent CMR with T1 mapping; disease severity was quantified using Mainz Severity Score Index (MSSI). Myocardial trabecular fractal dimension was evaluated, blinded to patients'characteristics, on short axis cine images using the Image J dedicated plug-in FracLac, deriving the following parameters: total, basal, mid-ventricular and apical fractal dimensions.

Results: Total fractal dimension was higher in all Fabry groups compared to controls. Indeed, a gradient of total fractal dimension was observed, with this parameter gradually increasing from healthy controls to Groups 3 (1.27±0.02 in controls vs 1.29±0.02 in Group 1 vs 1.30±0.02 in Group 2 vs 1.34±0.02 in Group 3; $p<0.001$) (Figure 1A). Interestingly, both total and basal fractal dimensions were significantly higher in Group 1 compared to controls (1.27±0.02 vs 1.29±0.02, p=0.044 and 1.26±0.04 vs 1.30±0.03; p=0.007, respectively). Moreover, considering the total population, fractal dimension showed significant correlations with: i) T1 values (r=–0.567; $p<0.001$ - Figure 1B); ii) LV mass (r=0.674, $p<0.001$); iii) trabecular mass expressed as percentage of global LV mass (r=0.611; $p<0.001$); iv) MSSI (r=0.535; $p<0.001$).

Conclusion: Cardiac involvement in FD is characterized by a progressive increase in fractal dimension of endocardial trabeculae (Figure 1C). Both total and basal myocardial trabeculation are increased in Fabry patients even before the presence of detectable sphingolipid storage, thus representing a very early sign of cardiac involvement.

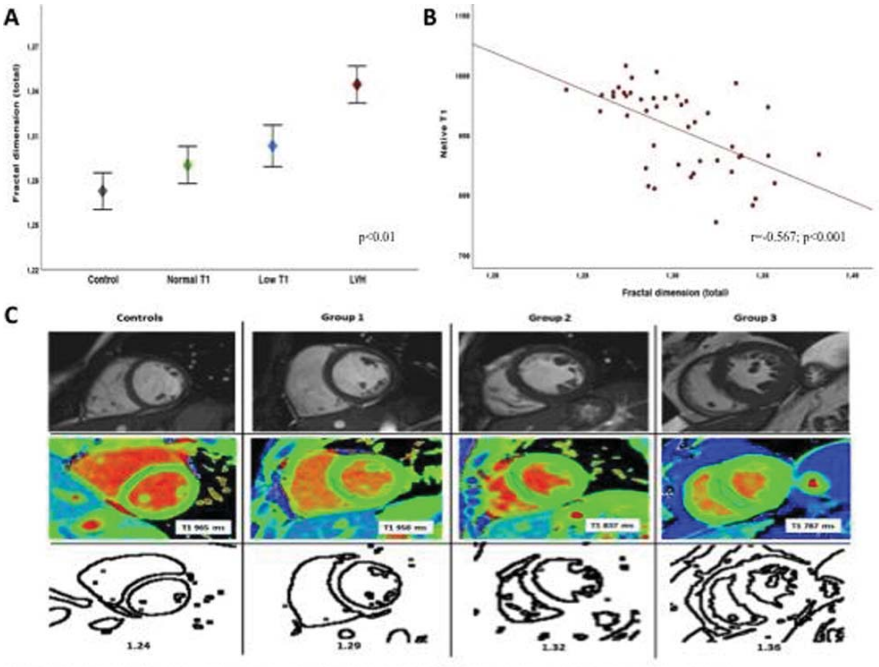

Figure 1. Progressive increase in trabecular fractal dimension from healthy control to Group 3.
Total myocardial trabeculae complexity quantified by endomyocardial border fractal analysis in different stages of Fabry cardiomyopathy and healthy controls (**A**). Correlation between fractal dimension of endocardial trabeculae and native T1 values in study population (**B**). Cine images (upper panels), T1 maps (middle panels) and trabecular fractal dimension (lower panels) of 4 subjects, one from each Groups. Moving from Controls to Group 3 (left to right), progressive increase in fractal dimension together with lowering of T1 values can be observed (**C**).