Severe secondary mitral regurgitation due to left ventricular non-compaction cardiomyopathy- a rare cause of heart failure

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Introduction
Left ventricular non-compaction is a rare form of cardiomyopathy charactherized by the presence of a two layered ventricular wall- a thinner epicardial layer and an inner, non-compacted layer with prominent trabeculations associated with deep, intratrabecular recesses that communicate with left ventricle cavity. Clinical manifestations vary in severity, including symptoms of heart failure, thromboembolic events or arrhythmias. Left ventricular (LV) dysfunction leads to tethering of the mitral apparatus and is a cause for secondary significant mitral regurgitation.

Case presentation
We report the case of a 57 year-old female patient, diagnosed with severe mitral regurgitation one year before presentation, with severe heart failure (HF) symptoms, referred to our clinic for the surgical replacement of the mitral valve. Clinical examination revealed no signs of pulmonary or systemic congestion and systolic apical murmur. Blood tests were normal, except for the elevated BNP (552 pg/ml). Electrocardiogram showed sinus rhythm and left ventricular hypertrophy. Coronary angiogram did not identify any coronary artery lesions. Echocardiography revealed mildly dilated left ventricle, but with prominent trabeculations and two distinct myocardial layers with a non-compacted/compacted ratio of 2:1 in the anterior and lateral walls, diagnostic for left ventricular non-compaction cardiomyopathy. LV ejection fraction was 40%, with severe secondary mitral regurgitation due to significant antero-posterior dilation of the mitral ring, with intact mitral leaflets; mild pulmonary hypertension was present.
Magnetic resonance imaging (MRI) identified a two layer antero-lateral myocardium and confirmed the echo diagnosis; there was no evidence of scarring as there was absent late gadolinium enhacement. In the absence of fibrosis on MRI or any arrhythmic events on repeated Holter ECG monitoring, the implantation of a cardiac defibrillator was deferred.
Given the secondary cause for mitral regurgitation (LV dysfunction), specific HF medication with beta blocker and renin-angiotensin-aldosterone blockade was initiated and titrated to optimal doses. With medical treatment the evolution was favourable. Currently, 3 years after the initial diagnosis, 6 minutes walk test revealed good functional capacity (510 m), a BNP value of 104 pg/l, without any worsening of LV systolic function nor progression of pulmonary hypertension.

Conclusion
Left ventricular non-compaction cardiomyopathy is a rare cause of heart failure, but due to advances in imaging modalities and increasing awareness, its prevalence is growing. Its pathogenesis and prognosis largely remain unknown, but early and adequate initiation of neurohormonal medication may be just as essential in order to prevent complications and improve long term prognosis, as for other forms of cardiomyopathy, even in the presence of severe secondary mitral regurgitation.