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Outcomes of patients with left ventricular noncompaction and preserved ejection fraction

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BACKGROUND

Left ventricular noncompaction (LVNC) has a wide phenotypic expression. Prognosis of patients with preserved ejection fraction (pEF) remains uncertain.

PURPOSE

To describe the characteristics and natural history of this subgroup of patients.

METHODS

LVNC patients were included in a multicentric registry. Those with pEF (LVEF > 50%) were considered for the analysis.

RESULTS

491 LVNC pts from 10 Spanish centres were recruited from 2000 to 2018. 239 (49%) had baseline pEF. Compared to those with reduced EF (rEF), they were younger, with no differences in gender and had less comorbidities (Table 1). Mean LVEF was 62% (SD 8). 18 pts (9% of the available CMR) had fibrosis even though LV volumes and LVEF were normal.

Family screening was completed in 199 pts, being positive in 113 (57%). Genetic testing was performed in 146 index cases, being positive in 80 (55%): ACTC1 (40), MYH7 (17), TTN (8), HCN4 (6) and other individual variants.

During a median follow-up of 4.9 years (IQR 2.1-7.3), there was a significant decrease in LVEF: last LVEF was 30-40% in 5 pts (2%) and 40-50% in 21 (9%) ($p = 0.01$ compared to baseline LVEF). 6 pts (2.5%) died during follow-up, only 1 of cardiovascular cause. 9 patients (4%) presented heart failure (HF) and 25 (10.5%) ventricular tachycardia or fibrillation (VT/VF). All cardiovascular outcomes were less frequent compared to rEF (Image 1, all $p < 0.05$). In multivariate analysis (including demographic, imaging, genetic and family aggregation parameters) the only predictor for HF was change in LVEF (OR 0.89, mean LVEF at the event 47%, $p = 0.01$ compared to no HF). Fibrosis was not associated with VT/VF.

CONCLUSIONS: Patients with LVNC and pEF have an overall excellent prognosis, which is markedly better than those with rEF. However, there is progressive decrease in LVEF, associated with heart failure, and moderate risk of life threatening arrhythmias. Therefore, periodic follow-up should be promoted.

Table 1

	LVNC pEF (n = 239)	LVNC rEF (n = 252)	p
Men, n (%)	131 (55)	146 (58)	0.65
Median age at diagnosis (IQR) - yr	38 (23-54)	58 (42-72)	0.01
Median follow up (IQR) - yr	4.9 (2.1-7.3)	3.9 (1.4-7.9)	0.04
QRS (SD) - ms	93 (18)	117 (32)	0.01
LGE, n (%)	18 (9)	52 (30)	0.01

Abstract P1442 Figure. Image 1

