($P < 0.05$). Circulatory power was 2073 mmHg.mL^{-1}min^{-1} in group A, 2620 mmHg.mL^{-1}min^{-1} in group B and 4040 mmHg.mL^{-1}min^{-1} in group C ($P < 0.05$). A chronotropic incompetence was observed in 46% of group A and 44% in group B versus 7% in group C ($P < 0.05$). Peak VO_2 was 14 mL.min.Kg^{-1} in group A, 19 mL.min.Kg^{-1} in group B and 20 mL.min.Kg^{-1} in group C ($P < 0.05$). The increase of O_2 pulse during exercise was 2 in group A versus 3 in group B and group C ($P < 0.05$). The VE/VCO_2 slope was increased in group A compared to group B and C (respectively 40 vs. 31 and 30, $P < 0.05$).

Conclusion CPET quantifies and specifies determinants of the poor cardio-circulatory response during exercise in AL amyloidosis patients, including decrease of peak VO2 and low circulatory power suggestive of poor exercise inotropic reserve; a chronotropic incompetence that can be related to cardiac dysautonomia and an increase of VE/VCO2 slope suggestive of exercise pulmonary hypertension.

Disclosure of interest The authors declare that they have no competing interest.

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Targeted panel sequencing in adult patients with left ventricular non-compaction reveals a large genetic heterogeneity

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Introduction Left ventricular non-compaction (LVNC) is a rare cardiomyopathy that may be of genetic origin, however few data are available about the global yield of mutation screening, the spectrum of genes and allelic variations.

Purpose The aim of this prospective study was to better characterize the allelic and genetic spectrum of isolated LVNC in a cohort of 95 unrelated adult patients through the molecular investigation of a custom panel of 107 genes involved in various cardiomyopathies and cardiac arrhythmias.

Results Sixty pathogenic or probably pathogenic variants, including 47 novel ones, were identified in 44 patients (46.3%) including 31 patients (32.6%) with single variant and 13 patients with complex genotypes (13.6%). The most prevalent genes were *TTN*, then *HCN4, MYH7, RYR2, MYH6, and MYBPC3*. The genotype-phenotype correlation and the major clinical outcome enhanced the fact that mutated patients tended to have younger age of diagnosis. Interestingly the mutation yield was significantly higher in youngest patients < 65 years old (42/84, 50%) as compared to oldest patients > 65 years (2/11, 18,2%, $P = 0.02$). The LV mean ejection fraction in patient with a mutation in sarcomeric genes was lower than in patients mutated in non-sarcomeric genes (41.7% vs. 52.7%, $P = 0.05$). The distribution includes 14 genes previously reported in LVNC and 13 additional candidate genes.

Discussion Our results showed that LVNC is basically a genetic disease and support genetic counselling and cardiac screening in relatives. There is a large genetic heterogeneity, with predominant *TTN* mutations and a distribution close to the one observed in dilated cardiomyopathy but with specific genes such as *HCN4*. The prevalence of complex genotypes in these patients is important to notice in the context of genetic counselling. We also identified 13 potential new genes associated with LVNC.

Disclosure of interest The authors declare that they have no competing interest.

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