OBJECTIVE: To investigate differences in cardiac manifestations of patients affected by laminopathy, according to the presence or absence of neuromuscular involvement at presentation.

METHODS: We prospectively analyzed 40 consecutive patients with a diagnosis of laminopathy followed at a single centre between 1998 and 2017. Additionally, reports of clinical evaluations and tests prior to referral at our centre were retrospectively evaluated.

RESULTS: Clinical onset was cardiac in 26 cases and neuromuscular in 14. Patients with neuromuscular presentation experienced first symptoms earlier in life (11 vs 39 years; $p < 0.0001$) and developed atrial fibrillation/flutter (AF) and required pacemaker implantation at a younger age (28 vs 41 years [$p = 0.013$] and 30 vs 44 years [$p = 0.086$] respectively), despite a similar overall prevalence of AF (57% vs 65%; $p = 0.735$) and atrio-ventricular (A-V) block (50% vs 65%; $p = 0.500$). Those with a neuromuscular presentation developed a cardiomyopathy less frequently (43% vs 73%; $p = 0.089$) and had a lower rate of sustained ventricular tachyarrhythmias (7% vs 23%; $p = 0.387$). In patients with neuromuscular onset rhythm disturbances occurred usually before evidence of cardiomyopathy. Despite these differences, the need for heart transplantation and median age at intervention were similar in the two groups (29% vs 23% [$p = 0.717$] and 43 vs 46 years [$p = 0.593$] respectively).

CONCLUSIONS: In patients with laminopathy, the type of disease onset was a marker for a different natural history. Specifically, patients with neuromuscular presentation had an earlier cardiac involvement, characterized by a linear and progressive evolution from rhythm disorders (AF and/or A-V block) to cardiomyopathy.

KEYWORDS: Atrial fibrillation; Bradyarrhythmias; Emerin; Familial cardiomyopathies; Lamin; Neuromuscular
disorders; Ventricular tachycardias

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