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Right heart involvement in patients with laminopathies

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Introduction Laminopathies are rare genetic diseases, with various clinical manifestations. Cardiac involvement may include conduction system disease, supraventricular and ventricular arrhythmias, and dilated cardiomyopathy. In other cardiac diseases, right ventricular dysfunction has been associated with an impaired prognosis.

Objective To study the prevalence of right ventricular involvement in laminopathies and its influence on patient outcome.

Methods and results We retrospectively included 138 patients (age=37 [25-49], female=58%) with LMNA gene mutations. At baseline, conduction system disease was present in 36% of patients, supraventricular arrhythmias in 25%, non-sustained and sustained ventricular tachyarrhythmias (VT) in 7 and 6%, left ventricular dysfunction in 42%. The association of right ventricular systolic dysfunction and clinical signs of right heart failure was observed in 18 patients (13%) including 9 who presented with isolated right ventricular without left ventricular systolic dysfunction. Over a 13.5 median follow-up duration, 47 patients (34%) developed major cardiac adverse events, including 23 with end-stage heart failure, 28 with sustained VT, and 26 with 2nd degree Mobitz II or 3rd degree AV blocks. Right ventricular involvement was significantly associated with truncating mutations (44% vs. 22%, $p=0.04$), higher global mortality (44% vs. 17%, $p<0.001$), end-stage heart failure (56% vs. 11%, $p<0.001$), sustained ventricular tachycardia (56% vs. 17%, $p<0.001$), supraventricular tachyarrhythmias (94% vs. 49%, $p<0.001$), and sinus node dysfunction (33% vs. 7%, $p<0.001$).

Conclusion Patients with laminopathies are at risk for right ventricular dysfunction, which may be present without any left ventricular systolic dysfunction and frequently leads to end-stage heart failure.

The author hereby declares no conflict of interest

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Atrial flutter in myotonic dystrophy type 1: patient characteristics and clinical outcome

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Background The characteristics of DM1 patients with atrial flutter (AFL) and their clinical outcome are unknown.

Methods We retrospectively included the patients ≥ 18 years of age with DM1 who were admitted in our institutions with AFL between January 2000 and September 2013 and analyzed their clinical outcome. We compared the incidence of AFL recurrences in patients who were treated with versus without radiofrequency (RF) ablation. Single and multiple variable analyses were performed to identify predictors of AFL recurrences.

Results We included 60 consecutive patients (age=41 \pm 13 years, male=34), including 55 with persistent, 2 with paroxysmal, and 3 permanent AFL. Over a 53 \pm 28 months mean follow-up duration, AFL recurrence occurred in 12 patients (24%), atrial fibrillation in 13 (26%), ischemic stroke in 2 (3%), cerebral hemorrhage in 1 (2%) and sinus node dysfunction in 4 (7%). Patients treated by RF ablation were significantly more frequently free of AFL recurrences by Kaplan Meier analysis (95% vs. 61%, HR=0.17, 95% CI 0.08 to

0.97, $p=0.04$). By multivariate analyses, RF ablation was the only parameter significantly associated with absence of AFL recurrence ($p=0.01$).

Conclusions Patients with DM1 presenting with AFL are exposed to stroke, severe sinus node dysfunction and cerebral hemorrhage. RF catheter ablation is associated with a lower risk for AFL recurrences and may limit iatrogenic complications associated with pharmacological treatments.

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Predictive factors of left ventricular recovery in acute heart failure revealing reduced left ventricular ejection fraction

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Aims Evaluate the factors presents at diagnosis associated to left ventricular recovery in acute heart failure (AHF) revealing reduced left ventricular ejection fraction (LVEF).

Background A lot of paperwork have studied the left ventricular evolution in chronic heart failure but not in AHF and indistinctly of LVEF. We don't know the predictive factors of left ventricular recovery in a specific population of acute heart failure revealing reduced LVEF.

Methods We conducted a prospective and observational longitudinal study with retrospective data collection based on a cohort of patients with acute heart failure revealing reduced LVEF ($\leq 45\%$), in the emergency department and cardiac intensive care unit (ED and CCIU) of the University Hospital of Lille, between November 1st, 2011 and May 31st, 2013. The primary endpoint was the improvement of LVEF or left ventricular reverse remodeling.

Results Among the 111 patients included, 57% of patients benefited from an improvement of LVEF and 62% from a reverse remodeling in the 51% of patients with a dilated left ventricle. The improvement of LVEF was associated in univariate analysis to non ischaemic etiology ($p=0.002$) and greater utilization of betablockers ($p=0.076$) and angiotensin-converting enzyme ($p=0.0337$) and in multivariate analysis, independently to younger patients ($p=0.0325$) and presentation by cardiogenic shock ($p=0.0484$). The same predictive factors were found for reverse remodeling.

Conclusion The knowledge of the prevalence and the early predictive factors of the left ventricular recovery can be useful to guide the follow-up and non-pharmacological management of heart failure with reduced LVEF.

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Predictive factors of tolerance to carvedilol in systolic heart failure

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Introduction Heart failure experiencing exponential growth in Algeria. Among the recommended therapeutic, the b blockers are largely under prescribed. The objective of this study was to determine predicting factors of tolerance to carvedilol in systolic heart failure.

Patients and Methods Carvedilol has been introduced in 100 consecutive patients hospitalized with systolic heart failure. Clinical parameters, the BNP levels and echocardiographic indices were measured blind to admission to the first and second week.

Results The average age of patients was 66 \pm 1 year. The FE: 31 \pm 1%, the NYHA 2.8 \pm 0.1, BNP 620 \pm 48pg/mL, E/A: 1 8 \pm 0.3, TDE 160 \pm 7, E/E', 9.1 \pm 0.3, PAPS: 36 \pm 0.4, VCI: 15+1mm. An acute decompensation episode occurred in 21patients and b blocker was arrested at 9 of them without the possibility of reintroduction. In univariate analysis, the values on admission NYHA stage, the plasma level of BNP, the TDE and reports E/A and E/E'