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ABSTRACTS

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Mode of death in cardiac amyloidosis

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Introduction Cardiac amyloidosis (CA) is a severe disease, often lethal. Little is known about cause of death during the natural course of CA. Identification of cause of death during CA is of particular interest to design clinical trials and identify therapeutic targets in this pathology.

We aimed to describe mode of death in CA patients during long-term follow-up, according to the type of amyloidosis.

Material and methods All patients consecutively referred to the French referral center for light-chain (AL), transthyretin hereditary (hTTR) or wild-type (WT-TTR) CA were consecutively included between 2010 and 2016 were included. Mode of death was centrally adjudicated by two blinded clinical committees, using multiple sources, and classified as cardiovascular (including worsening heart failure, sudden death, stroke) or non-cardiovascular (1).

Results From the 565 patients included, 187 patients had AL amyloidosis, 220 had hTTR and 158 had WT-TTR amyloidosis. One hundred thirty-nine patients (25%) died during a follow-up of 864 patients-years, with median follow-up in survivors 14.6 months (IQR 3.7—33.9). One-year survival was 67% in AL patients, 92% in hTTR patients and 89% in WT-TTR patients (logrank < 0.0001). Among 139 deaths during follow-up, 86 (62%) resulted from cardiovascular causes, mainly worsening heart failure (58/86, 67%) and sudden death (20/86, 23%). Thirty-four deaths were from non-cardiovascular causes (24%), mostly from infection (27/34, 79%).

Discussion and Conclusions Mortality is high during natural course of cardiac amyloidosis, significantly different according to CA type. Main causes of death were cardiovascular (mostly worsening heart failure and sudden death), opening room for optimal prevention and management.

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

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Cardio-circulatory exercise response in AL amyloidosis and comparison with hypertrophic cardiomyopathy

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Introduction Cardiopulmonary exercise test (CPET) can help the physician in understanding mechanisms that underlie patients exercise limitation. We aimed to characterize exercise response in AL amyloidosis through CPET and to compare it with hypertrophic sarcomeric cardiomyopathy (HCM).

Methods We prospectively included 49 patients: 24 with cardiac amyloidosis (group A), 10 with amyloidosis without cardiac involvement (group B) and 15 HCM (group C). All were clinically stable and underwent clinical examination, ECG, echocardiography, cardiac MRI and CPET. CPET assessed systolic blood pressure (SBP) as well as heart rate (HR) responses, O_2 pulse response, peak VO_2, circulatory power ($SBPxVO_2$) and VE/VCO_2slope.

Results The SBP response was 25 mmHg in group A from rest to peak exercise, 19 mmHg in group B and 49 mmHg in group C