

## Topic 21 – Heart failure, cardiomyopathy – A

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### 0045

#### Rest and exercise pulmonary hypertension in hypertrophic cardiomyopathy

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**Background:** Heart failure (HF) symptoms, related to pulmonary capillary hypertension, are frequent in hypertrophic cardiomyopathy (HCM). Pulmonary hypertension (PH) is the consequence of left ventricular (LV) diastolic pressure elevation and/or functional mitral regurgitation. We aimed to evaluate determinants and prognostic significance of rest and exercise PH in HCM.

**Methods and Results:** We included 235 patients referred for clinical work-up of HCM. Rest pulmonary artery systolic pressure (PASP) was measurable in 214 (91%) patients (48±16 years, 161 males). A symptom-limited semi-supine bicycle exercise was carried out in 188 patients. PASP was measurable during exercise in 108 patients (57%). Resting PH (≥60mm Hg) was present in 56 patients (26.2%) and exercise PH (≥60mm Hg) in 38 patients (35.2%). Multivariate correlates of rest PASP were sinus rhythm ( $\beta = -0.15$ ,  $P=0.021$ ), LV obstruction tract (LVOT) peak gradient ( $\beta = 0.22$ ,  $P=0.001$ ) and left atrial volume ( $\beta = 0.39$ ,  $P<0.0001$ ). Multivariate resting correlates of exercise PASP were PASP ( $\beta=0.28$ ,  $p=0.001$ ) and mitral regurgitation (MR) grade ( $\beta=0.48$ ,  $P<0.0001$ ). LVOT peak gradient emerged as an independent correlate of exercise PASP when MR was excluded. Patients with rest PH had a worse event-free survival at 4 years (24.8 ± 8.8 vs 66.2 ± 5.2 %,  $P < 0.0001$ ), survival without HF (55.6 ± 10.5 vs 81.8 ± 4.3% ,  $P = 0.005$ ), and overall survival (84.2 ± 7.1 vs 97.1 ± 1.7 %,  $P = 0.001$ ). Patients with exercise PH had also a worse event-free survival (47.7 ± 9.5 vs 65.7 ± 7.5%,  $P = 0.007$ ) and a worse survival without HF (64.1 ± 9.4 vs 83.4 ± 5.5%,  $P = 0.016$ ). By contrast, there was no difference regarding overall survival ( $p = 0.49$ ).

**Conclusion:** In patients with HCM, the main determinants of rest PH are sinus rhythm, LVOT peak gradient and left atrium volume. Determinants of exercise PH are rest PASP, grade of MR and rest LVOT gradient. Rest and exercise PH predict a poor outcome in HCM.

### 0066

#### Clinical outcomes of childhood hypertrophic cardiomyopathy associated with RASopathy: the Necker Sick Children Hospital experience

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**Background:** Because it represents few patients and a heterogeneous group of disorders, the natural outcomes for children with hypertrophic cardiomyopathy (HCM) are unclear; even more with regard to RASopathies. The aim of this study was to determinate outcomes of children with hypertrophic cardiomyopathy associated with RASopathy according to their therapeutic strategy.

**Methods and Results:** Patients in this study were identified between December 1980 and December 2012 in the Congenital and Pediatric Cardiology Unit of Necker Sick Children Hospital in Paris (France). The RASopathy cohort consisted of 81 children with clinical phenotypic diagnostic and echocardiographic hypertrophy cardiomyopathy. The median age at diagnosis of cardiomyopathy was 11,5 +/- 8 months without expecting 9 antenatal diag-

noses. 46% and 75% of all RASopathy children were diagnosed with cardiomyopathy respectively before 1 months and 1 year. The duration of median follow-up was 8,2 +/- 7,7 years after date of birth. 35% of the study cohort required at least 1 surgical procedure.

82% of deaths occurred before 1 year of life in children with RASopathy and HCM. Risk factors for mortality in the first year of life were antenatal diagnosis (HR=3,38), surgery before 1 year of life (HR=3,38), HCM diagnosed before 2 months of life (HR=2,85), and male (HR=1,21). All deaths after 1 year of life were due to sudden death. 4 children had regressive hypertrophy. At the latest follow-up, 37% (n=11/28) children survived.

**Summary:** The outcome of children with hypertrophic cardiomyopathy due to RASopathy is frightful and early with 80% of survey at 1 year of life. Main risk factors seem to be antenatal diagnosis, surgery before 1 year of life, and HCM diagnosed before 2 months of life.

### 0241

#### Does Ramadan fasting has any effects on hemoglobin, glucose and renal function in patients with chronic heart failure?

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**Objective:** The effects of Islamic fasting on physiologic functions in normal conditions have been considered in different studies and different topics. This study was to determine the quantitative changes of hemoglobin, glucose and renal function during Ramadan fasting in patients with chronic heart failure.

**Methods:** Fifty-three outpatients with chronic heart failure were followed in 2 stages: one week before the start of Ramadan and then on the last day of Ramadan by proper blood sampling in suitable time of day.

**Results:** There were 31 (58.5%) males and 22 (41.5%) females with a mean age of 60±11.06 years. Nineteen patients (35.8%) had mild left ventricular systolic dysfunction, 29 patients (54.7%) had moderate systolic dysfunction and 5 patients (9.4%) had severe systolic dysfunction. There were no significant changes in the NYHA Class nor there were any significant changes in the Canadian Cardiac Society (CCS) class in patients with coronary artery disease. We found non significant decrease in hemoglobin and blood glucose level ( $P=0.76$ ) and ( $P=0.16$ ) respectively, and non significant increase in creatinine ( $P=0.07$ ) before and at the end of Ramadan in either males or females.

**Conclusion:** The conclusions from this study can not be extrapolated to patients with worse functional classes or those who are unstable. It is clear that more work should be carried out to evaluate the impact of fasting on cardiac patients with higher NYHA class, and discover the significance of Ramadan fasting in patients with heart disease.

### 0053

#### Short-term moderate diet restriction in adulthood can reverse alterations of cardiac function induced by postnatal overfeeding in mice

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Postnatal overfeeding (OF) in rodents induces early programming of cardio-metabolic risk: permanent moderate increase in body weight, metabolic disorders and progressive alterations of cardiac function in adulthood. Our aim was to determine whether moderate diet restriction, performed at the time where these disorders are acquired, could restore cardiac function and ameliorate post-ischemic recovery.

Immediately after birth, litters of C57BL/6 mice were either maintained at 9 (normal-fed group, NF), or reduced to 3 in order to induce OF. At weaning, mice of both groups received a standard diet ad libitum (AL). At 6 months of age, half of the NF mice and OF mice were assigned to a moderate 20% calorie restriction (CR, NF-CR, OF-CR) for one month, while the other half of mice continued to eat AL (NF-AL, OF-AL). Cardiac function was followed using echocardiography and, at 7 months, the sensitivity to ischemia-reperfusion injury was evaluated in isolated perfused hearts.