Rest and exercise pulmonary hypertension in hypertrophic cardiomyopathy

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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 (≥50mm 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 (β=−0.15, P=0.021), LV obstruction tract (LVOT) peak gradient (β=0.22, P=0.001) and left atrial volume (β=0.39, P=0.0001). Multivariate resting correlates of exercise PASP were PASP (β=0.28, P=0.001) and mitral regurgitation (MR) grade (β=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=0.001), and a worse survival at 4 years (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.

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 determine 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 diagnoses. 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.

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 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.

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.
Six-month-old OF mice weighed 19.3% more than NF mice. Left ventricular fractional shortening (LVFS) and ejection fraction (LVEF, 51.4% vs. 57.6%, p<0.001) were decreased in OF mice. Left ventricular internal diameter in diastole (LVIDd) was significantly greater in OF mice. After one month of moderate CR, body weight of NF-CR and OF-CR was reduced by 12.6% as compared to their respective AL-group. LVEF increased in OF-CR (60.2% vs. 49.4%, p<0.001) and became comparable to this of NF-AL, while LVIDd was also normalized, whereas no difference between NF-CR and NF-AL were observed. After 30min of global ischemia, hearts isolated from OF-CR mice showed smaller infarct size than this of others groups.

Our study suggests that while short-term moderate CR could diminish body weight in both NF and OF groups of mice, CR was able to improve cardiac function and susceptibility to myocardial ischemia-reperfusion injury only in OF, reversing the deleterious influence of postnatal programming by early overnutrition.

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Right ventricular systolic function in heart failure, a Moroccan experience
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Background: The right ventricle (RV) has received less attention than the left in heart failure patients probably because morbidity and mortality associated with left ventricle disease is clinically more apparent. In our study, we tried to prove the importance to characterize the prevalence and clinical significance of right ventricular (RV) systolic dysfunction (RVD) in patients with heart failure.

Methods and Results: We studied 1613 patients with chronic heart failure at the HF Registry of the university hospital Ibn Rochd Casablanca, during a follow up of 6 years. RVF (RV function) were determined by echocardiography. RV dysfunction defined by (S'VD <10cm / s and TAPSE <16mm). The primary endpoint was the occurrence of acute heart failure decompensation (AHFD). RESULTS: RVD was present in 117 patient (7,1%), this group had an average age of 64 years, they were more likely to be men (sex ratio: 2:1), to have atrial fibrillation, and chronic diuretic therapy. At echo, patients with RVF had slightly lower LVEF (75% lower LVEF and 25% preserved LVEF), worse diastolic dysfunction, lower blood pressure and cardiac output, higher pulmonary artery systolic pressure (PASP) 23%, and more severe RV enlargement and tricuspid valve regurgitation. Patients with RV dysfunction had 11.54% of AHFD occurrence. The association of both RVD and pulmonary hypertension increased the risk of AHFD (14.26%), while patients with normal RVF had only 1.48% to develop AHFD. Adjusting for age, sex, PASP and comorbidities, RVD defined by TAPSE and echo Doppler tissue imaging, was associated with higher risk of HF decompensation.

Conclusions: In our community, RVD is common in HF patients, associated with clinical and echocardiographic evidence of more advanced HF and predictive of poorer outcomes so it had independent prognostic utility. We should give more attention to the RVF in our systematic assessment of HF patients.

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Right ventricular dysfunction in patients presenting with an idiopathic dilated cardiomyopathy: predictive factors and prognostic value
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Purpose: Right ventricular (RV) dysfunction is a predictor of impaired prognosis in patients presenting with an idiopathic dilated cardiomyopathy (DCM). We intended to determine which parameters were associated with RV dysfunction and to analyze the prognostic role of RV dysfunction independently of the level of LV dysfunction (LVEF and end-diastolic diameter).

Methods: 136 consecutive patients (73% men, mean age 59.0±13.2 years) with idiopathic DCM (LVEF ≤45%) were retrospectively enrolled. 34 patients (25%, group 1) presented with RV dysfunction defined as TAPSE ≤15mm (tricuspid annular plane systolic excursion) and 102 (group 2) had normal RV function. Patients were followed for a mean time of 2.7±1.1 years for the occurrence of any major adverse cardiac event (MACE: cardiac death and hospitalization, heart transplantation).

Results: Mean LVEF was 27.5±8.7%. Mean TAPSE was 18.6±5.4mm with an interquartile range of 15-21.8mm. Multivariate predictors of RV dysfunction were LV outflow tract TVI (OR 0.8 (95% CI 0.7-0.9), p=0.003) and E-wave deceleration time ≤145ms (OR 4.1 (95% CI 1.3-12.8), p=0.017).

Conclusions: RV dysfunction defined by TAPSE ≥15mm is associated with a systolic (aortic TVI) and a diastolic (E-wave DT) LV function parameters. RV dysfunction in patients presenting an idiopathic DCM is an additional and independent prognostic factor independent of the level of LV dysfunction and dilatation.