nary 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 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 (≥36mmHg) was present in 56 patients (26.2%) and exercise PH (≥60mmHg) in 38 patients (35.2%). Mutivariate 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<0.0001), 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.

The author hereby declares no conflict of interest

0198

Global and regional echocardiographic strain to assess early phase of hypertrophic cardiomyopathy due to sarcomeric mutations

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Objective Hypertrophic cardiomyopathy (HCM) is a genetic disease with delayed cardiac expression. Our objective was to characterize global and regional LV myocardial strain by two-dimensional imaging in sarcomeric HCM families and hypothesized that early systolic dysfunction, before hypertrophic stage, may be diagnosed by this technique.

Methods and results We analyzed 81 adults: HCM patients with LV hypertrophy (LVH+, n=38), mutation carriers without LV hypertrophy (LVH-/G+, n=20), and normal control subjects (n=23). We calculated global longitudinal strain (GLS), regional peak longitudinal strain and the Echo/TDI score (combination of 3 parameters about remodeling and pulse TDI). Age, sex ratio and body surface area were not significantly different between groups. Maximal 2D wall thickness of left ventricle was 10.1±1.6mm in LVH-/G+ and not different from controls (9.9±1.2mm). We observed that LV GLS was not different in LVH-/G+ as compared to controls (–21.6%±3.2 vs –23.5%±3.3) but was reduced in HCM patients (–15.3%±4.5) although a normal ejection fraction. Interestingly, regional peak longitudinal strain was similar in LVH-/G+ and controls except antero-septo-basal segment strain that was decreased in LVH-/G+ as compared to controls (–15.6%±7.2 vs –20.0%±3.9, p=0.025). A cut-off of –16% for abnormal strain of antero-septo-basal segment identified LVH-/G+ subjects with a sensitivity of 47% and a specificity of 90%. The Echo/TDI score was different in LVH-/G+ as compared to controls (p=0.0008) and sensitivity of previous defined cut-off was 83% for identification of LVH-/G+. All LVH-/G+ subjects with abnormal regional strain, except one, had abnormal Echo/TDI score.

Conclusion We observed that regional longitudinal strain, but not global strain, was significantly reduced at early stage of HCM. This tool may be useful for clinical evaluation of relatives in daily practice, but does not provide significant additional information as compared to the Echo/TDI score.

The author hereby declares no conflict of interest

0203

Pregnancy in women with a cardiomyopathy: outcomes and predictors from a retrospective cohort

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Background and objective Pregnancies in women with cardiomyopathies are considered at high risk for complications. However we lack data to characterize their natural history and to predict the outcome. Our aim was to evaluate the prevalence and predictors of acute cardiac and obstetrical events in pregnant women with a cardiomyopathy, excluding peripartum cardiomyopathy.

Methods and results The monocentric retrospective survey includes 43 pregnancies in 36 women with dilated cardiomyopathy (DCM, n=10), hypertrophic cardiomyopathy (HCM, n=28), arrhythmogenic right ventricular cardiomyopathy (ARVC, n=3), tachycardia-induced-cardiomyopathy (TCM, n=1) and non-compacted left ventricle (NCLV, n=1). There were 1 fetal death (with maternal cardiac death), 10 (23%) preterm deliveries and 11 (26%) low neonatal birth weights. There were major maternal cardiovascular events in 14 pregnancies including 6 acute heart failures (5 DCM, 1HCM), and 3 cardiac deaths, which occurred in cases that did not follow our usual multidisciplinary protocol (1 pregnancy denial with DCM, 1 undiagnosed HCM, 1 DCM recognized near term). CARPREG score was predictive of maternal cardiac events that occurred in 67%, 33% and 25% of pregnancies with CARPREG scores of 2, 1 and 0 respectively. However major complications (heart failure, asymptomatic degradation of LVEF, and symptomatic aggravation of pulmonary artery hypertension) occurred in 3 women with no known risk factors. LV ejection fraction alone, gradient in HCM, ZAHARA or WHO scores were less discriminant than CARPREG for maternal outcome.

Conclusion Pregnancy in women with a Cardiomyopathy is a high risk period for both major cardiac and fetal events. The worst complications occurred in cases that did not benefit from a multidisciplinary team management. The highest rate of events was observed in DCM patients but significant risk was also associated with HCM. Prediction of risk with CAPREG scoring is appropriate but need to be improved.

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0545

3 years experiences of a heart failure management unit in Bordeaux: results and prognosis factors

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Background Heart failure is the leading cause of hospital admissions and economic burden. In accordance with guidelines, a dedicated heart failure unit has been created in Bordeaux (France) in May 2011.

Aim Evaluate the impact of a heart failure management unit with ambulatory sector on readmission rate and mortality; and identify prognosis parameters at diagnosis and 6 months after.

Methods Descriptive study of mortality and heart-failure-related readmission rates after a first admission for heart failure, from May 2011 to July 2014 (inclusion stopped in July 2013). Student's test and Cox's regression have been performed.

Results Since the creation of this unit, its activity has been growing by 30% in the first year and 10%, in the second. 421 patients with less than 40% left ventricular ejection fraction (LVEF) have been included; 181 had been treated before May 2011 while 240 were news patients treated during the study. Average age was 57±14 years old and average LVEF was 28%±7%.

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