– increased plasma levels of BNP in 45.7% of patients, to value 240 ng/ml increasing cTnI values plasma at 4.34% of cases.

Biological changes were correlated in most cases with clinical manifestations, echo changes induced cardiotoxicity and increase of dispersion QTcQTc intervals.

Conclusions Increased levels of cardiac biomarkers: BNP and cTnI and of the dispersion of QTcQTc intervals in children treated with anthracyclines other drugs with cardiotoxic effects positively correlates with installation of the cardiotoxicity with clinical or infraclinical manifestations, constituting an useful indicator for the cardiotoxicity. Changes in this parameter appeared early than echo changes anticholinergic induced cardiotoxicity and is necessary to systematic monitoring these parameters during and after cytostatic treatment.

The author hereby declares no conflict of interest

0454

Long-term cardiac prognosis and risk stratification in 260 adults presenting with mitochondrial diseases

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Aims To assess the long-term cardiac prognosis of adults with mitochondrial diseases.

Methods and results Between January 2000 and May 2014, we retrospectively included in this study 260 consecutive patients (60% women) ≥18 years of age, (interquartile range [IQR]: 31 to 54), with genetically proven mitochondrial diseases, including 109 with mitochondrial DNA (mtDNA) single large-scale deletions, 64 with the m.3243A>G mutation in MT-TL1, 51 with other mtDNA point mutations, and 36 patients with nuclear genes mutations. Cardiac involvement was present at baseline in 81 patients (30%). Single and multiple variable analyses were performed in search of predictors of major adverse cardiac event (MACE), and hazard ratios (HR) and 95% confidence intervals (CI) were calculated. Over a median follow-up of 7 years [3.6 to 11.7], 27 patients (10%) suffered a MACE, defined as sudden death, death due to heart failure (HF), resuscitated cardiac arrest, 3° degree atrioventricular block, sinus node dysfunction, cardiac transplantation, or hospitalization for management of HF. Patients with single large-scale mtDNA deletions or m.3243A>G mutations had the highest incidence of MACE. By multiple variable analysis, intraventricular conduction block (HR=16.9; 95% CI: 7.2 to 39.4), diabetes (HR=7.0; 95% CI: 2.9 to 17.7), pectinate ventricular complexes (HR=3.6; 95% CI: 1.4 to 9.2) and left ventricular (LV) hypertrophy (HR=2.5; 95% CI: 1.1 to 5.8) were independent predictors of MACE. In patients with 0, 1, and ≥2 risk factors, the incidence of MACE was 1.7, 15 and 42% respectively.

Conclusions Patients with mitochondrial diseases are at high risk of MACE, independently predicted by intraventricular conduction block, diabetes, ventricular prematurity and LV hypertrophy.

The author hereby declares no conflict of interest

0353

Prevalence of hereditary transthyretin cardiac amyloidosis in patients with increase in LV thickness in France

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Methods Between January 2012 and January 2015, we included 76 genetically confirmed Friedreich ataxia patients in Pitié-Salpêtrière Hospital. Clinical examination, ECG, echocardiography and blood samples were obtained.

Patients were aged of 38±12 years, (mean±sd), 50% were male. 4 patients had palpitations, 2 dyspnea and no patients had chest pain. 89% had negative T waves on the ECG. 49% had echocardiographic hypertrophy according to Henry’s nomogram. Patients with hypertrophy were younger: 34±10 years versus 42±14 years, age at onset of the disease was earlier: 15±6 years versus 21±15 years. Interventricular septal wall thickness was 12.9±1.7mm versus 10±2mm, and posterior wall thickness was 11.3±1.5mm versus 9.4±1.1mm. Left ventricle ejection fraction was similar (64%). For patients with hypertrophy, troponin was higher: 22±21 ng/L versus 10±7 ng/L. Plasma NT-proBNP was the same between the 2 groups 104±170 ng/L versus 64±122 ng/L. 5 patients had plasma NT-proBNP ≥300 ng/L, they all had had atrial fibrillation or heart failure.

Plasma high sensitive troponin is a diagnostic marker of hypertrophic cardiomyopathy in Friedreich ataxia’s patients, whereas plasma NT-proBNP is associated with cardiac events and could be a prognostic marker in these patients.

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