Aims: 1-Compare regional LV LS between the 3 types of CA, 2- Determine if they correlate to amyloid deposits; 3- Determine the prognostic value of LV LS.

Methods: 79 patients were prospectively included. 17 segments LV LS was measured. 53 had cardiac MRI with LGE evaluation for each segment. Amyloid deposition was quantified using Congo red staining in the 3 explanted heart following heart transplant (HT). All patients were followed-up. The composite primary endpoint was major adverse cardiac event (MACE) i.e., death, HT, and acute heart failure. MACE predictors were identified by multivariate analysis.

Results: 26 patients had AL, 36 m-TTR and 17 WT-TTR CA. Global (–10±4%) and regional LV LS were similarly impaired in the 3 types of CA. 91% of MRI had LV LGE enhancement. Both LGE and LS analysis showed a basal to apex gradient. Interestingly LS in the basal free wall (mean ± 5±6%) had the best accuracy to predict LGE (AUC=0.97, P<0.002; 95%CI [0.92;0.99]). Amyloid deposits in the 3 HT were more frequent in the basal segments and a strong correlation was found between LGE and LS segments (r=0.72). 64% experienced MACE during the follow-up with median of 13[7;23] months for survivors. Apical LS, NT-proBNP and NYHA III-IV were independent associated with MACE. Cut-off to predict MACE was –14.5% for Apical LS and 4000ng/L for NT-proBNP.

Conclusions: LS abnormalities are similar between CA types and reflect amyloid deposition which predominates in basal LV segments. Impairment of basal free wall LS predict LV MRI LGE. High value of apical LS is an independent predictor of MACE.

0254
Role of 99mTc-HMDP scintigraphy in the diagnosis and follow up of cardiac amyloidosis
Arnault Galat (1), Jean Rosso (2), Aziz Guellich (1), Stephane Rappeneau (1), Soulef Guendouz (1), Claire Marie Tissot (1), Diane Bodez (1), Julien Ternacle (1), Jean-Luc Dubois-Randé (1), Serge Adnot (3), Luc Hittinger (1), Thiibaud Dany (1)
(1) CHU Henri Mondor-APHP, Cardiologie, Créteil, France – (2) CHU Henri Mondor-APHP, Medecine nucléaire, Créteil, France – (3) CHU Henri Mondor-APHP, IMRB INSEMR U955, Créteil, France

Background: Accuracy of 99mTc-HMDP scintigraphy (sHMDP) to diagnose cardiac amyloidosis (CA) and to discriminate between the different etiologies i.e hereditary TTR amyloidosis (hTTR), senile amyloidosis (wt-TTR) and light chain amyloidosis (AL) are unknown.

Methods: 122 patients referred for suspected CA were prospectively studied with sHMDP. HMDP fixation was evaluated by a visual score (0 up to 3). Heart-to-whole-body (H/WB) and heart-to-skull (H/S) ratio were calculated on planar images. Diagnosis of amyloidosis was biopsy proven for all AL. Diagnosis of CA for hTTR and wt-TTR was established by an experts consensus based on echocardiography, cardiac MRI (cardiac wall thickness ≥2mm), genetic testing and biopsy whenever ethically possible.

Results: The mean age was 69 (59; 78), 71% were men; of whom, 14 (11%) were considered having AL, 34 (28%) hTTR, 21 (17%) wt-TTR and 53 (43%) had cardiac hypertrophy from other causes. Of the 61 (50%) patients with CA, 46 (75%) had a HMDP visual scoring ≥1; of whom 11/14 (7%) had AL, 24/26 (92%) hTTR and 21/21 (100%) wt-TTR.

Cardiac HMDP uptake was absent among all patients without CA (n=61). Visual score was stronger in TTR-CA versus AL-CA (p<0.0001). A visual score ≥2 to diagnose TTR-CA had a predictive positive value of 100%. Among CA, H/WB and H/S ratio were significantly higher (p<0.002) in both hTTR (H/WB=3.92±0.93 H/S=2.6±0.9) and wt-TTR (H/WB=4.3±0.95 H/S=2.6±1.12) versus AL (H/WB=2.6±0.61; H/S=1.7±0.53). Eleven patients with myocardial 99mTc-HDPD uptake underwent a second sHMDP with a median (25%; 75%) of 15.1 months (7.3; 16.2) of whom two had an increase in visual score and H/WB ratio. (Image 1, next page)

Conclusion: 99mTc-HMDP scintigraphy is a useful tool to diagnose CA, define its etiology and might be useful for follow-up.

0486
Postmortem genetic testing in a series of 36 young patients after sudden cardiac death
Isabelle Marey (1), Véronique Fressart (2), Caroline Rambaud (3), Estelle Gandjbakhch (1), Elsa Le Boette (1), Céline Bordet (1), Audrey Mallet (1), Geoffrey Lorin De La Grandmaison (3), Pascale Richard (2), Philippe Chartron (1)
(1) CHU La Pitié-Salpêtrière-APHP, Referral Centre for Hereditary Heart Disease, Genetics, Paris, France – (2) CHU La Pitié-Salpêtrière-APHP, Referral Centre for Hereditary Heart Disease, Biochemistry, Paris, France – (3) CHU Hôpital Raymond Poincaré-APHP, Anatomie pathologique, Garches, France

The incidence of sudden cardiac death (SCD) increases with age in parallel with coronary’s diseases’ prevalence. In young persons and athletes, SCD occurs in half of the cases, in the setting of genetically transmitted disorders such as cardiomyopathies. Molecular testing performed after necropsy may help management of families but experience in this area appears very limited. The aim is to report our experience of post mortem molecular testing after SCD and necropsy. We studied 36 patients ≤40 years who died suddenly with a suspected diagnosis of cardiomyopathy, established either after autopsy or known before death, with 6 diluted cardiomyopathy (DCM), 12 hypertrophic (HCM), 2 HCM/DCM, 1 restrictive (CMR), 14 arrythymogenic right ventricular cardiomyopathy (ARVC), 1 HMC and left ventricular noncompaction. Sanger sequencing was performed in most 4-5 frequent genes for a given phenotype. Fifteen mutations have been identified in sarcomeric (11 mutations) or desmosomal (3 mutations) genes. The identification of these mutations had significant impact: assessing right diagnosis in a doubtful case (HCM without LVH), modifying the appropriate diagnosis in another case (HCM and not DCM), confirming a genetic disease even in the absence of affected relatives in the family, providing guidance for genetic counselling and predictive genetic testing in relatives in all situations. Technical, ethical and legal issues may however be encountered and will be discussed. This study is one of rare series of post-mortem molecular testing after SCD. Our findings suggest the feasibility, molecular efficiency and the clinical benefit of the approach in order to improve the management of families. Postmortem molecular testing must take its place in the strategy of family care after SCD, even if a cardiomyopathy is suspected at necropsy, since genetic findings provide additional information useful for the relatives.

0532
Features and outcomes of acute myocarditis in children
Camille Walton, Magali Veyrier, Corinne Ducreu, Mohamed Bakloul, François Sassolas, Loïc Bousset, Olivier Desebre, Roland Henaine, Olivier Metton, Jean Ninet, Sylvie Di Filippo
Hôpital Cardiovasculaire, Cardiologie pédiatrique et congénitale, Lyon, France

This study was to assess features and outcomes of children with acute myocarditis.

Methods: Patients <18y with acute myocarditis (proved by virology and/ or MRI and/ or complete recovery of myocardial function) were included. Clinical data, echocardiographic parameters and outcomes were collected and cases divided in groups I (< 2y), II (2 to 10y) and III (>10y).

Results: 72 patients were included (1983 to 2012), 30males, aged 4±5.1y (median 5y): 43 in group I, 17 in II and 12 in III. Heart failure was present at onset in 57% (82%); 8 cardiac shock (12%), 30 severe HF (44%) were more frequent in I (56%) and II (46%) than in III (17%, p< 0.0001), chest pain (15.5%) was more frequent in III (83%). LVSF at diagnosis was 18.4±9%; 16% and 15% in groups I and II vs 30.5% in III (p= 0.0001). Aortic VTI was 11.4±5.8cm: 8 cm and 11 in groups I and II vs 17 in group III (p< 0.05). Mitral regurgitation was present in 76.5%, pericarditis in 16.4%, thromboembolic events occurred in 5.7%, arrhythmias in 7(10%). Virus was found in 27±37.5% (1 virus in 24, 3 in 3). Nine patients died (13%) within 2months post-diagnosis (2days to 8.6months). 1 was transplanted (3month), 19 have sequelae (27%), 40 recovered (58%), at FU= 5±5.6y. Inotrope was needed –5±6%. 46% experienced MACE during the follow-up with median of 4±5y. 43% had cardiac hypertrophy from other causes. Of the 61 (50%) patients with CA, 46 (75%) had a HMDP visual scoring ≥1; of whom 11/14 (7%) had AL, 24/26 (92%) hTTR and 21/21 (100%) wt-TTR.

Cardiac HMDP uptake was absent among all patients without CA (n=61). Visual score was stronger in TTR-CA versus AL-CA (p<0.0001). A visual score ≥2 to diagnose TTR-CA had a predictive positive value of 100%. Among CA, H/WB and H/S ratio were significantly higher (p<0.002) in both hTTR (H/WB=3.92±0.93 H/S=2.6±0.9) and wt-TTR (H/WB=4.3±0.95 H/S=2.6±1.12) versus AL (H/WB=2.6±0.61; H/S=1.7±0.53). Eleven patients with myocardial 99mTc-HDPD uptake underwent a second sHMDP with a median (25%; 75%) of 15.1 months (7.3; 16.2) of whom two had an increase in visual score and H/WB ratio. (Image 1, next page)

Conclusion: 99mTc-HMDP scintigraphy is a useful tool to diagnose CA, define its etiology and might be useful for follow-up.