Results: 1351 patients were studied, the median age was 63 years, 367 (27%) are diabetics. Overall, it exists similarity of the parameters studied between the 2 groups (D) and (ND) in terms of morphology and hemodynamics (end diastolic left ventricle volume; interventricular septum size; pulmonal arterial systolic pressure; left atrial volume...). But, we have found important differences of 3 parameters: Ejection fraction of left ventricle is higher (48.5% than 35%) in diabetics group. We found more segmental kinetic disorders (76% than 50%) and more diastolic dysfunction with higher filling pressures (51% than 34%) in diabetic population.

Conclusions: In our study, we have found more diastolic dysfunction and more segmental kinetic disorders but ejection fraction of left ventricle is higher.

089

Expanding the phenotype associated with a desmoplakin dominant mutation: Carvajal/Naxos syndrome associated with leukonychia and oligodentia

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We report on a family with a rare form of cardiac and non-cardiac disease that shed new lights on the clinical spectrum associated with desmosomal gene mutations, usually related to arrhythmogenic right ventricular cardiomyopathy.

The 29-year-old patient presented with woolly hair from infancy and palmoplantar keratoderma, leukonychia (hands and feet) and oligodontia (agenesis of 10 teeth). Global cardiac biventricular involvement was present on echocardiography and MRI. Late potentials were present on signal-average ECG and ventricular premature beats (PVB) on Holter-ECG (>10/24h). A single-chamber prophylactic implantable cardioverter-defibrillator was implanted.

The son of the patient had palmoplantar keratoderma, woolly hair, oligodontia (agenesis of 10 teeth) and brittle nails. At 10 years of age, a diagnosis of acute myocarditis was made (chest pain, cardiac troponin elevation, multiple isolated PVB, typical sign on MIBI). RV infundibulum was dilated on Echocardiography. Evolution was unremarkable after medical treatment.

A heterozygous missense mutation (c.1691C>T, p. Thr564Ile) was identified in the desmoplakin gene (both in father and son). No additional mutation was identified in other desmosomal genes.

Carvajal/Naxos syndromes are usually recessive and characterized by wooly hair, palmoplantar keratoderma and a cardiomyopathy. We report on a family with unique features associated with a new mutation of desmoplakin gene. First, mode of inheritance is autosomal dominant and not recessive. Second, clinical expression is associating not only the usual Carvajal/Naxos syndrome but also oligodontia and leukonychia. This report (i) extends the phenotype associated with desmosomal gene mutations, (ii) demonstrates that leukonychia and oligodontia may be associated with severe cardiomyopathy and maybe therefore require systematic cardiac examination, (iii) suggests that desmoplakin gene is involved in the normal and abnormal development of teeth and nails.

090

Could heart rate predict duration of hospitalizations for patients admitted for acute pericarditis?

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Purpose: Acute pericarditis is rather frequent. Annual incidence is estimated to 27.7 new cases per 100,000 inhabitants in Europe. About 5% of all non-ischemic chest pains admitted at emergencies could be pericarditis. Most of the patients are young patients, with a significant cost to society, particularly as regards hospitalizations. Indeed, pericarditis represents 1% of all hospitalizations in department of cardiology. It could be very interesting if clinical presentation and especially heart rate could help predict duration of hospitalizations.

Methods: Between March 2007 and February 2010, we conducted a retrospective study concerning all patients admitted in our center for acute pericarditis. Diagnosis criteria included 2 among the 4 following: typical chest pain, friction rub, pericardial effusion on echocardiography, or typical ECG findings. We evaluated hospital events (heart failure, acute pains, death) and biology during hospitalization (CRP on admission, on days 1, 2, 3, and especially peak).

At one month, clinical events were recorded through phone calls when not noticed in clinical settings.

Results: We included 73 patients. Mean age was 41.0 y (CI 95% 37.2-44.8) and mean hospitalization duration was 3.5 d (2.5-4.5). Heart rate on admission was 88 bpm (83.6-92.4) and 71.8 bpm (68.9-74.7) on discharge. CRP peak was strongly correlated with heart rate (r=0.54; p=0.0001) and with hospitalization duration (r=0.8; p=0.007). Finally, we found a positive correlation between heart rate on admission and duration of hospitalizations (r=0.226; p=0.06). Fever was scarcely observed (21%), and was not correlated with heart rate and with CRP.

Conclusion: In acute pericarditis, cardiac frequency at admission is correlated with hospitalization duration, and could be a new prognostic marker. This point deserves to be explored, in order to reduce hospitalization duration.

091

Cardiac sarcoidosis, myocardial spect contribution

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Background: Sarcoidosis is a multiorgan systemic disease rare. The cardiac involvement is common, but only 3-5% of them are symptomatic up to sudden death. The prognosis depends on early and potential reversibility of lesions treated. Myocardial SPECT (TSM) and especially magnetic resonance (NMR) are examinations of choice to objectify sarcoid lesions.

Objective: The aim of this study is to assess the prevalence of possible myocardial sarcodosique and monitored by an evolutionary TSM MIBI

Methods: For this, we have, in a prospective study after a clinical examination, electrical and echocardiographic sought, the TSM MIBI rest, fixing defects after myocardial reversible injection of a vasodilator dipyridamole, 31 consecutive patients addressed to the pulmonology department mediasinal-pulmonary sarcoidosis.

Results: There are 22 men and 09 whose average age was 38±11 years. Six patients (19%) had problems with perfusmillens extent of tracer uptake of 3.8±1.7 segments of 17. On the ECG, there is a supraventricular tachycardia,
one case of atrioventricular block of first degree, two cases of ventricular extrasystoles bigéménées. The echocardiographies are normal except for a case of diastolic dysfunction. The standard therapeutic SST in 5 patients.

The TSM MIBI such as NMR is a good diagnostic examination and scalable for a proper and early treatment. Available in our institution, it enabled us to diagnose 19% of cases of myocardial sarcoid lesions.

092

Prognosis value of QRS duration in patients with heart disease and syncope

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Background: Patients with heart disease (HD) and syncope are at high risk of sudden death. Implantable defibrillator (ICD) is recommended in patients with unexplained syncope and left ventricular ejection fraction (LVEF) <30% or in patients with LVEF >30% and inducible ventricular tachycardia (VT).

Aim: The purpose of the study was to evaluate the prognostic significance of QRS duration measurement in patients with HD and syncope.

Methods: 528 patients, 89 women and 439 men, mean age 65±12 years, were admitted for syncope. All of them had an HD, either ischemic HD (n=382) or left ventricular impairment of other origin (n=115). Holter monitoring, electrophysiological study and head-up tilt test were systematic. Filtered QRS duration was measured at signal-averaged ECG (Fidelity 2000 of Cardionics) (filter 40 Hz, noise level <0.6 µV). The patients were followed from 3 months up to 18 years (mean 5 ±4 years).

Results: Mean LVEF was 40±14%. Cardiac defibrillator was implanted in 73 patients. 30 patients died suddenly, 75 died from heart failure or were transplanted (n=9). Remaining patients are alive or died from non cardiac death (n= 8). The last group differed from group who died suddenly by an higher LVEF (42±14% vs 32±13) (p<0.0001) and a shorter QRS duration (125±34 msec vs 144±31) (p<0.026). They tended to be older (65±12 years vs 61±13) (p<0.09).

The alive group differed also from group who died from heart failure by an higher LVEF (42±14% vs 32±13) (p< 0.00001) and a shorter QRS duration (125±34 msec vs 141±31) (p<0.0033). They tended to be younger (65±12 years vs 67±10) (p<0.08). Patients who died suddenly and those who died from heart failure had similar LVEF and QRS duration but patients who died suddenly are younger than patients who died from heart failure (p<0.01).

Conclusions: Low LVEF is a classical risk of worse prognosis in patients with HD and syncope. A longer QRS duration is also a noninvasive and simple test of worse prognosis. A QRS duration more than 125 msec had a sensitivity of 73% and a specificity of 64% to predict cardiac mortality.

093

Associations between clinical characteristics and NT-proBNP in heart failure with preserved ejection fraction: data from the KaRen study

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Purpose: In heart failure with reduced ejection fraction, N-terminal pro brain natriuretic peptide (NT-proBNP) is useful for diagnosis and prognosis, and is higher with older age, female gender, renal failure and lower in obesity. In heart failure with preserved ejection fraction (HFPEF), NT-proBNP is also useful for diagnosis and prognosis, but clinical correlates are less well established.

Methods: KaRen is a multicenter prospective registry of HFPEF. Inclusion criteria are acute presentation with Framingham symptoms and signs of heart failure together with NT-proBNP >300 ng/L and LVEF >=45%. The association between clinical characteristics and log NT-proBNP was assessed with univariable linear regression, with backward step-wise selection based on p<0.05.

Results: We studied 400 patients, mean (SD) age 77 (9) years, 56% women. The table lists variables independently associated with log NT-proBNP, their baseline values and Beta coefficients.

Conclusions: In HFPEF, NT-proBNP is determined mainly by NYHA class and clinical signs of heart failure. Clinical signs are important predictor of severity of disease. As in reduced EF, NT-proBNP is inversely associated with BMI and GFR, but unlike in reduced ejection fraction, it is not associated with age or gender.

Table – Independent associations with log NT-proBNP

<table>
<thead>
<tr>
<th>Variable</th>
<th>BMI</th>
<th>GFR</th>
<th>NYHA I / II / III / IV pulmonary rales</th>
<th>peripheral edema</th>
<th>hepatomegaly</th>
<th>pleural effusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean (SD) or n (of 400)</td>
<td>29±6</td>
<td>63±28</td>
<td>2 / 28 / 159 / 211</td>
<td>304</td>
<td>277</td>
<td>21</td>
</tr>
<tr>
<td>Beta</td>
<td>-0.39±0.01</td>
<td>-0.009±0.002</td>
<td>0.22±0.07</td>
<td>0.22±0.10</td>
<td>0.27±0.09</td>
<td>0.45±0.19</td>
</tr>
<tr>
<td>p</td>
<td>&lt; 0.0001</td>
<td>&lt; 0.0001</td>
<td>0.001</td>
<td>0.032</td>
<td>0.004</td>
<td>0.02</td>
</tr>
</tbody>
</table>

BMI, body mass index; GFR, glomerular filtration rate; NYHA, New York Heart Association Class.