552 Strain echocardiography in hypertrophic cardiomyopathy. Relation to risk factors of sudden cardiac death.

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Echocardiography has a major role in evaluation of hypertrophic cardiomyopathy. New tissue Doppler-based measurements could provide better insights into myocardial regional characterization and provide supplemental information in risk stratification. We aimed to study the relation between strain measurements and established sudden death risk factors.

Methods: 36 (aged 47±15; 27) male consecutive patients with hypertrophic cardiomyopathy underwent echocardiography with Doppler-derived strain and strain rate measurements, stress test, Holter monitoring, and clinical examination. Results: Maximal left ventricular wall thickness (MLVWT) was 20.1±4.8 mm. 11 (31%) had significant LVOT obstruction. 31% had abnormal blood pressure response during exercise test. 69% were in NYHA functional class I. 16% had related to impaired septal strain, figure 1 (p=0.002). Septal strain was associated with NSVT on Holter, severe LVOT obstruction, and MLVWT. On multivariate analysis, severe LVOT obstruction and MLVWT remained associated. Number of risk factors was related to impaired septal strain, figure 1 (p<0.001). Septal strain correlated to left atrial dimension (r=0.40, p=0.03).

Conclusions: Doppler-derived strain imaging relates to clinical and echocardiographic risk factors of sudden death in hypertrophic cardiomyopathy.

553 Differentiation of transient left ventricular apical ballooning from anterior myocardial infarction.

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Background: Transient left ventricular (LV) apical ballooning (AB) is characterized by chest pain, ECG changes and LV apical akinesia mimicking acute myocardial infarction in the absence of coronary artery disease. Since most AB patients are diagnosed by angiography, the echocardiographic features have not been well described.

Methods and Results: Among 1512 patients with an acute coronary syndrome (ACS) undergoing coronary angiography over a 5-year period, 30 AB patients (2% of total) were identified (age 69±14, median 62 years). In all patients transthoracic echocardiography (TTE), coronary angiography, left ventricular function, strain imaging and follow-up of complications such as intraventricular pressure gradients and LV thrombus were measured by 3D echo and Doppler imaging. All subjects had intrinsic ventricular rhythm. Values were expressed as mean±SD.

Results: the right ventricle had slightly reduced systolic function on 3D echocardiography. The results of the tissue Doppler analysis are shown in the table.

Conclusions: Systolic strain is reduced in the mitral and apical segments of the RV free wall, even before RV involvement is apparent by 2D and 3D echo and before symptoms occur. ARVD may be localized and the variation in the measurements calls for some caution in the interpretation of the findings.

554 Coronary microvascular dysfunction in hypertrophic cardiomyopathy: Doppler echocardiography study.

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Background: microvascular dysfunction, reflected by an inadequate increase in myocardial blood flow in response to dipyridamole infusion, is a recognized feature of hypertrophic cardiomyopathy (HCM) and may have prognostic impact. Coronary flow reserve (CFR) can be assessed on left anterior descending coronary artery (LAD) noninvasively with Doppler 2D-echocardiography.

Aim: to prospectively evaluate a cohort of patients with HCM after they had undergone quantitative assessment of CFR on LAD with ultrasound.

Methods: 53 patients (New York Heart Association class I or II) with HCM were followed for a mean (±SD) of 33±15 months after dipyridamole (0.84 mg/kg over 10')-Doppler echocardiography.

Results: CFR on LAD was normal (≥2.0) in 36 and abnormal (≤2.0) in 15 patients. Eighteen events occurred during follow-up: 7 left atrium dilations, 5 atrial fibrillations, 3 hospitalizations for unstable angina, 1 cardioverter-defibrillator implantation, 1 pacemaker implantation and 1 sudden death. The event-free survival was significantly higher in patients with normal than in patients with abnormal CFR (p<0.0001) (Figure). With a Cox analysis, abnormal CFR on LAD (HR=8.7, 95% CI=2.7-27.4; p=0.0001) and interventricular septal thickness at end-diastole (HR=1.42; 95% CI=1.14-1.78; p=0.002) were independent prognostic indicators.

555 Concealed amyloid heart disease in patients with the clinical features of hypertrophic non obstructive cardiomyopathy (HNCM): invasive and echocardiographic investigations.

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Concealed myocardial storage disorders (MSD) may completely mimic the clinical picture of HCM. This holds especially true for cardiac Fabry disease (FD) where the clinical presentation of amyloid heart disease in between 6% and 8% of unselected consecutive pts. The diagnosis of concealed MSD is of considerable prognostic and therapeutic consequence based on recent advances in molecular biology and recent advances in AHD. To date systematic investigations in large patient collectives concerning the frequency of concealed AHD in HCM are lacking.

Methods: We performed for the first time right ventricular endomyocardial catheter biopsy (EMCB) in a large group of 300 consecutive pts with HCM (mean age 55 years, range 14 to 90 years; left ventricular hypertrophy with septal wall ≥ 15 mm). In all pts transonic echocardiography (TTE), coronary angiography, left ventricular angiogram and ECMB (approximately 4 tissue samples per pt) with light (LM) and electron microscopic (EM) evaluation were performed. All pts were showed akinesia of the entire LV apical area including septal, anterior, inferior and lateral segments. A transient LV pressure gradient (10-45 mm Hg) was present only in 4 AB patients and resolved within 4 days. Normalization of LV function was seen only in AB patients following a sequence of akinesia, hypokinesia and early recovery. Two AB patients developed a LV thrombus and stroke despite therapeutic anticoagulation. Normalization of the ECG occurred only in AB patients and always later (91±114 days) than echocardiographically documented resolution of LV dysfunction (20±18 days).

Conclusion: By demonstrating akinesia of the entire apical area echocardiography aids in differentiating apical ballooning from AMI. Serial echo is valuable for detection and follow-up of complications such as intraventricular pressure gradients and LV thrombus.