tion fractions were measured by 3D echocardiography (Philips SONOS 7500®). Regional right ventricular systolic velocities and strain were measured by Tissue Doppler Imaging (GE VIVID 7<sup>®</sup>). 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. Conclusion: Systolic strain is reduced in the mid 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.

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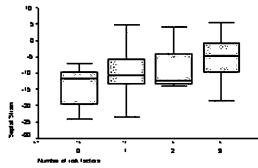
# Strain echocardiography in hypertrophic cardiomyopathy. Relation to risk factors of sudden cardiac death

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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 stablished 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 exhaustive clinical evaluation

Results: Maximal left ventricular wall thickness (MLVWT) was 20.1±4.8 mm. 11 (31%) had significant LVOT obstruction. 31% had abnormal blood pressure re-sponse during exercise test. 69% were in NYHA functional class I. 16% had nonsustained ventricular tachycardia (NSVT) on Holter. 42% had none, 33% one, and 25% had >1 risk factors for sudden death. 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=0.002). Septal strain correlated to left atrial dimension (r:0.40, p=0.03)



Figure

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

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### 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%) were identified (26 f, 4 m, age 71 $\pm$ 10). Prevalence was 5.1% (26/509) in females and 0.4% (4/1003) in males. AB patients were compared to 10 consections utive patients with anterior myocardial infarction (AMI) and similar findings on LV angiography. There was no difference regarding age, symptoms or ejection frac-tion. A triggering event was present in 1/10 AMI vs 28/30 AB patients (p<0.001) consisting of emotional (n=20)±physical stress (n=20). In AB patients levels of CK (188±84 vs 1550±3180 U/l) and troponin | (7.0±4.3 ng/ml vs 174±190 ng/ml) were significantly lower than in AMI. ST-segment elevation (28/30 vs 10/10) and T-wave inversion (30/30 vs 7/10) was similar in both groups. Q waves, present in 10/30 AB patients only in the acute phase, resolved during follow-up but persisted in 9/10 AMI patients (p<0.001). Compared to AMI with a more asymmetric wall motion abnormality, all AB patients undergoing echocardiography in the acute phase

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 relaxation. Two AB patients developed a LV thrombus and stroke despite thera-peutic anticoagulation. Normalization of the ECG occurred only in AB patients and always later (91±114 days) than echocardiographically documented resolution of LV dysfunction (20±19 days).

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

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#### Coronary microvascular dysfunction in hypertrophic cardiomyopathy: Doppler echocardiography study

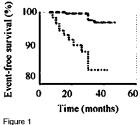
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Background: microvascular dysfunction, reflected by an inadequate increase in myocardial blood flow in response to dipyridamole infusion, is a recognized feature of hypertrophic cardiomyopathy (HCM) and may have prognostic impact. Coronary flow reserve (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 under-gone 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 38 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% Cl=1.14-1.78; p=0.02) were independent prognostic indicators.



Conclusions: in patients with HCM, the ultrasound-based assessment of the degree of microvascular dysfunction is a strong, independent predictor of clinical deterioration. Severe microvascular dysfunction is often present in patients with mild or no symptoms and may precede clinical deterioration by years

#### 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 mimick the clinical picture of HNCM. This holds especially true for cardiac Fabry disease (FD) and Amyloid heart disease (AHD). Cardiac FD may mimick the clinical features of HNCM 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 enzyme replacement therapy of FD and therapeu-tic advances in AHD. To date systematic investigations in large patient collectives concerning the frequency of concealed AHD in HNCM are lacking.

Methods: We performed for the first time right ventricular endomyocardial catheter biopsy (EMCB) in a large group of 300 consecutive pts with HNCM (mean age 55 years, range 14 to 90 years; left ventricular hypertrophy with septal wall =/> 15 mm). In all pts transthoracic echocardiography (TTE), coronary angiography, left ventricular angiogram and EMCB (approximately 4 tissue samples per pt) with light (LM) and electron microscopic (EM) evaluation were performed. All pts were