ATRIAL NATRIURETIC PEPTIDE RELEASE IN HYPERTROPHIC CARDIOMYOPATHY (HCM): ASSOCIATION WITH ABNORMAL PERIPHERAL VASCULAR RESPONSES

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Sudden death is common in HCM. Exercise hypotension is seen in 1/3 of pts and is due to an inappropriate fall in systemic vascular resistance occurring at high work loads. Abnormal peripheral vasodilation correlates with exercise hypotension and is associated with younger age and a family history of sudden death. Atrial natriuretic peptide (ANP), is released from the atria in response to stretch and has potent peripheral vasodilator properties. Pts with HCM may secrete large amounts of ANP during exercise. To examine the role of ANP in abnormal peripheral vasodilation, 16 pts (9 males), aged 17-64, mean 44 yrs, underwent symptom limited treadmill exercise testing. Peripheral venous ANP levels were measured at rest and at peak exercise. Peripheral vascular responses were assessed by forearm plethysmography at rest and peak of symptom limited supine bicycle exercise. 8 pts demonstrated abnormal peripheral vasodilation (<12% decrease in forearm blood flow (FBF) from resting), and 8 pts had a normal FBF response. The resting and peak ANP were similar in the 2 groups; however, the percentage change in ANP was significantly greater in those with peripheral vasodilation ($p<0.05$).

	Normal FBF response (SEM) (pmol/l)	Vasodilator response (SEM) (pmol/l)
Resting ANP	7.2 (2.6)	4.8 (1.8)
Peak ex ANP	13.5 (5.0)	12.3 (2.7)
% increase ANP	82.6 (18.4)	231.8 (64.6)

Change in ANP did not correlate with other clinical or hemodynamic indices. These results suggest that ANP release during exercise may be important in the initiation of hemodynamic collapse in pts with HCM.

ASSESSMENT OF MYOCARDIAL DISARRAY IN PATIENTS WITH NOONAN'S SYNDROME AND LEFT VENTRICULAR HYPERTROPHY

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Patients with Noonan's syndrome (NS) have been reported to have echocardiographic features of hypertrophic cardiomyopathy (HCM). The presence of characteristic histological features of HCM in NS has not been assessed. We studied the hearts of 7 necropsy patients with NS and echocardiographic LVH who died suddenly or in heart failure (age 2 weeks to 10 years; 6 were male). These were compared to age-matched control children dying of non-cardiac causes. In all cases, a transverse cut of both ventricles was fixed and stained with H&E, and the presence or absence of disarray evaluated with an image analyser at a constant magnification of x 16. The ventricular septum was evaluated counting 200 fields for each of the Noonan patients and 100 for the controls. The presence of disarray was significantly higher in the septum of NS (26.14 *vs* 3.83%, $p<0.001$). The same procedure was carried out for the left ventricular free wall, screening 100 fields for each of the Noonan patients and controls. The frequency of disarray in the LV free wall was also significantly higher in NS (22.33 *vs* 3.33%, $p<0.001$). Thus, NS with LVH shows a significantly higher incidence of myocardial disarray than age-matched controls. Similar post-mortem evaluation of children with HCM has not been reported, however, the extent of disarray seen in NS is similar to that reported in adults with HCM. Whether the presence of echocardiographic and histological features of HCM in patients with NS represents autosomal dominant familial HCM remains to be determined by pedigree analysis and ultimately by identification of the molecular defect.

DISTINCTION BETWEEN ATHLETE HEART AND HYPERTROPHIC CARDIOMYOPATHY BY QUANTITATIVE ASSESSMENT OF REGIONAL ULTRASONIC BACKSCATTER

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We have previously shown that integrated backscatter (IB) index is higher in pts with hypertrophic cardiomyopathy (HCM) than in control subjects. To ascertain whether IB value is useful in the distinction between HCM and athletes with myocardial hypertrophy, we evaluated with conventional echo-Doppler technique 10 HCM asymptomatic pts and 10 elite athletes (weight lifters). An on-line radio frequency analysis (by means of a prototype implemented in our Institute) was also performed to obtain quantitative operator independent measurements of the IB signal of the ventricular septum and the posterior wall. The integrated values of the radio frequency signal were normalized for the pericardial interface and expressed in percent (IB%). HCM pts and athletes showed comparable values LV end-diastolic diameter (46 ± 7, mean ± SD, vs 53 ± 4 mm), septal (17 ± 1 vs 15 ± 1 mm) and posterior wall (10 ± 1 vs 12 ± 1 mm) end-diastolic thickness. On the contrary, IB% values were significantly higher in HCM vs athlethes for both the septum (55 ± 14 vs 23 ± 8%, $p<.01$) and the posterior wall (40 ± 12 vs 12 ± 3, $p<.01$).
We conclude that quantitative assessment of ultrasonic reflectivity may be of value in distinguishing physiologic LV hypertrophy due to athletic training from pathologic LV hypertrophy due to HCM.

Thursday, March 7, 1991
8:30AM–10:00AM, Room 264, West Concourse
Cardiovascular Disease in Older Persons: Clinical Studies

8:30

MULTIVESSEL CORONARY ANGIOPLASTY IN THE ELDERLY: 10 YEAR EXPERIENCE.

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Bypass surgery (CABS) is associated with increased morbidity and mortality in the elderly. Accordingly, we examined the outcome of 1373 Pts, 65 (71±5) years and older, who underwent 1640 multivessel coronary angioplasty (MV PTCA) procedures over 10 years. 224 (14%) Pts had an ejection fraction ≤40% and 412 (25%) had prior CABS. A mean of 3.5 lesions were dilated per Pt with an angiographic success rate of 96%. Complete revascularization was achieved in 857 (52%) Pts. 52 (3.2%) Pts experienced a major complication. There were 27 (1.6%) deaths, 24 (1.4%) myocardial infarctions, and 14 (0.8%) emergent CABS procedures. Independent predictors of mortality were ejection fraction ≤40% ($p\leq0.001$), 3 vessel disease ($p\leq0.01$), female gender ($p\leq0.02$), and procedure date before 1985 ($p\leq0.05$). 1023 Pts have been followed ≥1 year (33±21 months). 162 (16%) Pts underwent CABS, 81 (8%) had a myocardial infarction and 156 (15%) died during follow-up. 371 (36%) Pts required repeat PTCA. Actuarial survival was 92%, 86% and 78% at 1, 3 and 5 years. Improved survival was observed in Pts with 2 vessel disease ($p\leq0.008$) and in those completely revascularized ($p\leq0.001$). Conclusions: 1) MV PTCA in the elderly has a high primary success rate and low risk. 2) complete revascularization improves long-term survival. MV PTCA is a safe and effective alternative to CABS in the elderly.