SUNDAY POSTER

1014-155 Aortic Root Growth In Children With Marfan Syndrome

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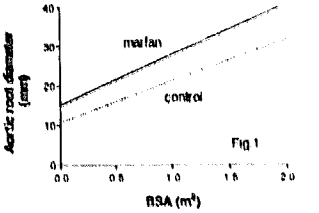
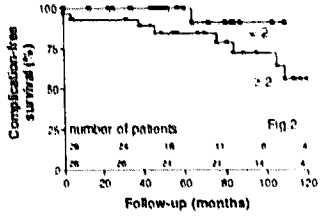
Aim of the Study: To study the clinical significance of aortic root growth on diagnosis and prognosis in children with Marfan syndrome.

Methods: Serial echocardiographic aortic root measurements of 123 subjects (57 Marfan, 66 Control), age 1–20 years, attending Marfan screening from 1983 to 1996, were used to construct individual aortic root growth curves relative to body surface area (BSA). By means of a Fisher discriminant analysis two sets of parameters, describing variation in intercept and slope in each group, were calculated. With the aid of these parameters and individual aortic root growth characteristics a discrimination score per subject was calculated. This score was tested to predict both the diagnosis 'Marfan syndrome' and a 10-year complication free interval for patients with a discrimination score < 2 and ≥22.

Results: The mean aortic root growth in the Marfan group differed significantly from the controls (y = 13.4x ± 14.5 and y = 9.3x ± 12.1 respectively, P < 0.001) (Fig. 1). Sensitivity and specificity of the score to predict the diagnosis Marfan syndrome was 84% and 73% respectively with three serial measurements. During 10-year follow-up the estimated probabilities of no complications were 90% (SE = 9%) for the score <2, and 56% (SE = 13%) for the score ≥2, P = 0.069 (Fig. 2).





Conclusions: Aortic root growth analysis in children may be useful for early identification of Marfan syndrome and risk stratification.

1014-156 Heterogeneous Response In the Aortic Root Elastic Properties to Long Term β-Blockade In Patients With the Marfan Syndrome

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Purpose: This study was undertaken to assess the effect of long-term β-blockade on the Ao root stiffness index and distensibility in patients (pts) with the Marfan Syndrome.

Methods: Ao root stiffness index and distensibility were calculated according to Stefanadi's and Hirai's formulas respectively, using 2D guided M-mode echocardiogram before and after an average of 26 months on atenolol.

Results: Twenty three asymptomatic pts were studied (11 M & 12 F; age 31 ± 14.2 yrs) with a mean follow-up of 4 yrs. The dose of atenolol was individualized (mean 50 ± 12.5 mg). Distensibility increased from 1.85 ± 0.70 to 2.21 ± 0.76 × 10^{-6} cm^2/dynes (p < 0.02) and the stiffness index decreased from 9.68 ± 3.78 to 8.85 ± 3.15 (p = 0.2). Two groups of responses to treatment were identified: 15 pts (65%), the responders, had increased distensibility and decreased stiffness index of the Ao root (p < 0.05); 8 pts (35%), non-responders, had no significant change. Body wt of 200 lb and baseline end-diastolic Ao root diameter >40 mm were significantly associated with no response (p < 0.05). Two pts in the group of non-responders had an echocardiographic progression in the degree of aortic insufficiency but remained clinically asymptomatic..

Conclusions: There was a heterogeneous response in the Ao root elastic properties after long-term treatment with atenolol in asymptomatic pts with Marfan Syndrome. Individualized dose according to Ao root end diastolic diameter and body wt are suggested.

1014-157 Exercise Capacity With Non-obstructive Hypertrophic Cardiomyopathy: Relation to Diastolic Function In Children

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Background: As exercise tolerance in children with hypertrophic cardiomyopathy (HCM) is unknown, we sought to evaluate aerobic capacity and its relations to diastolic function in children.

Methods: Cardiopulmonary exercise testing and echo were performed in 11 asymptomatic patients with HCM with normal systolic function and resting LV outflow gradients of <25 mmHg. Median age at evaluation was 13.5 yr (range, 8.6 to 17 yr). Doppler echo indices of diastolic function were related to maximal oxygen consumption (MVO2) and anaerobic threshold. Diastolic dysfunction (DDFN) was defined as isovolumic relaxation time (IVRT), deceleration time (DT) or a/o mitral diastolic inflow velocity ratio >2 SD's above predicted normal for age.

Results: DDFN was noted in 6 patients (54%), with longer mean IVRT (106 ± 15 msec vs. 69 ± 14 msec without DDFN; p = 0.002). There were no significant differences regarding weight, LVOT gradient, septal thickness or heart rate. Compared to age and sex-matched normals, patients with vs. without DDFN had lower % predicted MVO2 (68 ± 17% vs. 95 ± 7%, p = 0.01) and lower % predicted anaerobic threshold (85 ± 24% vs. 117 ± 14%; p = 0.03). Systolic BP with peak exercise rose less in those with DDFN (23 ± 17 vs. 66 ± 26 mmHg; p = 0.01). Oxygen pulse (stroke volume X avO2 difference/heart rate) was lower in those with vs. without DDFN (8 ± 3 vs. 13 ± 3; p = 0.02).

Conclusion: Diastolic dysfunction is associated with reduced exercise capacity in asymptomatic children with non-obstuctive hypertrophic cardiomyopathy.

1014-158 Etiology and Family History of Pediatric Cardiomyopathy: The Early Pediatric Cardiomyopathy Registry (PCMR) Experience

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Background: To better characterize cardiomyopathy (CM) in children, the NHLBI-sponsored PCMR conducted a study of children with newly diagnosed CM since 1/90.

Methods: From 8/96–8/97, 1093 eligible children were enrolled at 44 centers, representing 730 retrospective cases and 363 prospectively identified cases.

Results: The overall group had a mean age at diagnosis of 5.7 ± 6.0 yrs with a mean of 3.1 ± 2.1 yrs of follow-up. 58% were male, possible reflecting x-linked CM. LV systolic performance was depressed (FS > 2 sd) in 59% of pts. LV dilatation (EDD >2 sd) was noted in 53% of pts. Thickened LV walls (posterior wall thickness >2 sd) was noted in 29% of pts. Only 21% had a known etiology for their CM, a figure that is lower than the 43% reported in a 1993 survey of pediatric cardiologists. Among PCMR children with known etiology, we have observed a broad spectrum of genetic causes: neuromuscular disorders associated with CM (38.5%), isolated CM (18.4%), inborn errors of metabolism (19.5%), malformation syndromes associated with CM (10.3%) and not specified (13.2%). Among 255 pts with supplemental PCMR data, many had first-degree relatives with CM (27%), sudden death (13%), congenital heart defects (9%), arrhythmias (3%), or recognizable genetic syndromes (7%). The incidence of familial disease of potential relevance was higher than expected, occurring in 40% of CM pts. 48% of PCMR pts had an ICU admission for CM and 56% of all pts are on anticongestive therapy. Other therapies reported included 22% antiarrhythmic, 41% ACE inhibitor, 11% calcium antagonist, 11% beta adrenergic antagonist, 19% carnitine, 3% pacemaker, and 12% dietary modification.

Conclusion: Even at centers with interest in pediatric CM, the incidence of CM without a known etiology is surprisingly high. Our data suggests that genetic contributions to pediatric CM are likely to be underestimated.

1014-159 Left Ventricular Noncompaction: Evidence for a Mitochondrial Etiology

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Background: Left ventricular noncompaction (LVNC) is rare type of cardiomyopathy which generally occurs in infants and leads to congestive heart failure. Characteristic echocardiographic findings include deep trabeculations within the left ventricular endocardium, with chamber dilation and/or hypertrophy. Associated cardiac lesions such as ventricular septal defect (VSD) and pulmonic stenosis are common. Though the etiology of LVNC with associated