BETA ADRENERGIC RECEPTOR (BAR) BINDING CHARACTERISTICS AND ADENYLATE CYCLASE (AC) ACTIVITY IN THE PULMONARY ARTERY (PA) AND THORACIC AORTA (AO).

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In vascular smooth muscle, stimulation of BAR activates AC, leading to increased cAMP production and smooth muscle relaxation. The mechanisms involved in BAR stimulation and AC activation may differ in the pulmonary vs. systemic circulation. We examined BAR binding characteristics and AC activity in the PA and AO of adult male Sprague-Dawley rats. In each experiment, membrane preparations were made from tissue pooled from 5 rats. BAR number at saturation (Bmax) and the dissociation constant (Kd) were determined in Scatchard analyses with (1251) iodocyanopindolol, 10 to 100 pM. Alprenolol, 10⁻⁵M, was used to assess nonspecific binding. Basal AC activity and that stimulated by isoproterenol (Iso, 10⁻⁵M) were measured by radioimmunoassay for cAMP. Six experiments were performed. We found that BAR Bmax was greater in PA than in AO (403±70 vs 235±18 pmoles/mg protein, M±SEM, p<0.05), and that BAR Kd was lower (i.e., affinity was higher) in PA compared to AO (38±5 vs 61±9 pM, p<0.05). Basal AC activity was similar in PA and AO (1.5±0.4 vs 1.2±0.2 pmoles cAMP/mg protein/min), as was Iso-stimulated activity (3.1±1.8 vs 2.2±0.8). We conclude that BAR density and affinity are greater in PA than in AO in the adult male rat. Findings of similar basal and beta-stimulated AC activity may indicate that the coupling of BAR to AC differs in the two vascular beds. Further characterization of BAR binding and coupling to AC in PA smooth muscle will enhance our understanding of the regulation of pulmonary vascular tone.

RIGHT VENTRICULAR OUTFLOW TRACT ANEURYSM FOLLOWING TETRALOGY OF FALLOT REPAIR. Michael S. Snyder, Aaron R. Levin, Mary Allen Engle. Cornell University Medical College-The New York Hospital, Pediatric Cardiology, New York, NY

Aneurysms (AN) of right ventricular outflow tract (RVOT) following tetralogy of Fallot (TOF) repair were analyzed in 53 pts. Pulmonary-to-aortic root ratio, subpulmonic chamber size and supravalvar anatomy were assessed on pre-op angiograms, and operative reports were reviewed for the extent of RVOT reconstruction. AN size, judged from angiograms obtained 2-57 mos following surgery, was graded 1-4+. Large AN (3-4+), comprising more than 1/3 of total RV area on end-systolic lateral images, were found in 20 pts (38%), 3 of whom also had marked cardiomegaly (4+). Moderate AN (2+) were found in 14 pts (26%) and small RVOT AN (1+) in 19 pts (36%). We excluded 5 pts with a large residual VSD (Qp:Qs>1.8): 4/20 3-4+ pts and 1/33 pts with 1-2+ AN. Pre-op studies on the 3-4+ pts showed smaller pulmonary-to-aortic root diameters (.48±.05 vs .59±.06, p<.05), more tight supravalvar stenoses (38% vs 9%) and a lower incidence of appreciable subpulmonic chambers (31% vs 58%) than in other pts. Age at repair (0.8-12.8 yrs; mean 4.3±1 yr) was not different for those with large vs small AN (4.4±1.5 vs 3.3±1.2). In pts with 3-4+ AN, 15/16 (93%) required pulmonary valvotomy and RVOT patch, compared to 21/33 (63%) of the 1-2+ group. RV/LV pressure ratios were higher in the 3-4+ pts at post-op cath (.65±.12 vs .42±.05, p<.01). Multiple levels of pre-op severe stenosis leading to incomplete relief of outflow obstruction were the chief factors leading to the development of large RVOT aneurysms after TOF repair.

CHILDREN WITH COMPENSATED AORTIC STENOSIS MAINTAIN ENHANCED EJECTION PERFORMANCE DURING EXERCISE.

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It has been shown that ejection phase indices are elevated in children with aortic stenosis (AS) at rest. We performed two-dimensional and M-mode echocardiography in 14 patients with AS and in 7 normal (NL) children at rest in the upright position, and at 50% and 75% of predicted maximal workload (PMW) during upright bicycle exercise. The two groups were compared by student's t tests and were not significantly different at the .05 level with respect to ht., wt., BSA, race and sex. There were no significant differences between the two groups in heart rate, % MVO2, or cardiac index. We also found no difference in LV diastolic dimension between the two groups at rest, at 50% and at 75% PMW. LV systolic dimension was significantly smaller at all levels in AS. LV systolic wall thickness was increased in AS. Rate corrected velocity of fiber shortening (VCFc) was significantly higher at rest in patients with AS, and remained so at both levels of exercise. The stepwise increase in VCFc was significant in both NL and AS from rest to 50%, and from 50% to 75% PMW; however, the incremental increase in VCFc from rest to exercise was not significantly different between the two groups. We conclude that patients with AS maintain enhanced ejection performance at 50% and 75% of PMW as evidenced by increased VCFc. A failure to increase VCFc during exercise in patients with aortic stenosis may imply decompensation and improve timing of surgical intervention.

MYOCARDIAL INFARCTION IN CHILDREN

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Myocardial infarction (MI) is an uncommonly suspected cause of morbidity in children. This retrospective case series study was undertaken to ascertain the incidence and etiologies of MI in the pediatric population as diagnosed by ECG, and to develop a protocol for prospective study and therapy. All ECGs reported at Texas Children's Hospital from 1954-1986 were reviewed. Over 100,000 ECGs on 54,605 patients were obtained. There were 1,013,210 pediatric admissions and 31,305 pts referred to the section of pediatric cardiology. Seventy-two pts had ECGs read as consistent with MI and multiple etiologies noted. These included: anomalous left coronary artery (20%), Kawasaki disease (14%), myocarditis (13%), neontatal critical aortic stenosis (11%), ventricular tumor (5.5%), dilategd cardiomyopathy (4%), pulmonary atresia-intact ventricular septum (4%), birth asphyxia (4%), aortic thrombosis (4%), muscular dystrophy (2.5%), coaractation of aorta (2.5%), rheumatic carditis (2.5%), TAPVR (2.5%), and chest trauma (2.5%), rhe positive ECGs represent 0.13% of ECGs at TCH during this period, 0.23% of cardiology referrals, and 0.007% of pediatric admissions. Conclusion: 1) A variety of etiologies of MI are observed in a pediatric population. 2) Congenital coronary artery anomalies was the leading cause of childhood MI in this series, accounting for 20% of cases. 3) MI may be underdiagnosed in children and this series, because of retrospective study design, may represent a minimal incidence.

THE CONTINUING OUTBREAK OF ACUTE RHEUMATIC FEVER IN THE INTERMOUNTAIN WEST. L. George Veasy, Susan E. Wiedmeier, Edward L. Kaplan and Harry R. Hill. Primary Children's Medical Center and the University of Utah School of Medicine, Departments of Pediatrics and Pathology, Salt Lake City and the University of Minnesota, Minneapolis.

The incidence of acute rheumatic fever (ARF) has shown a steady decline in the United States. Attack rates in most recent studies have averaged 0.5 per 100,000 population leading to the concept that the disease has "virtually disappeared from the U.S." Over the past 23 month period, a total of 98 cases of ARF occurred in the Intermountain West surrounding Salt Lake City, Utah. This represents an eight fold increase in the annual incidence of the disease over the past decade. The patients presented with classic signs of ARF including carditis (90%), polyarthritis (57%), and chorea (31%). Doppler echocardiography proved to be a valuable adjunct in the diagnosis of cardiac involvement in ARF, especially in patients with chorea. The attack rate among 5 to 17 year old individuals in Salt Lake County was 18.1 per 100,000 in 1985. Only one third of the patients surveyed gave a history of a preceding respiratory infection. Culture surveys in two Utah communities where index cases occurred failed to reveal an increased preva-lence of group A streptococcal carriage (15%). Examination of group A organisms from these surveys and from patients and their siblings revealed mucoid M-type 18 and M-type 3 strains. These findings indicate that acute rheumatic fever is still a health problem in the U.S. and suggest that continued attention be directed towards the recognition and appropriate therapy of acute streptococcal infections.

RECOVERY HEART RATE DIFFERENCES IN MALES AND FEMALES AFTER MAXIMIM VOLINIARY EXERCISE. Reginald L. Washington, Jeff C. Van Qurdy, Robert R. Wolfe. Sponsored by: James Todd, The Children's Hospital Department of Pediatrics Division of Cardiology, University of Colorado School of Medicine, Division of Pediatric Cardiology, Danver, Colorado.

One hundred and fifty one unconditioned normal children aged 7yr 6 months to 12 yr 9 months were exercised to exhaustion utilizing graded progressive bicycle ergometry (Dames Protocol). There were 70 females and 81 males. Heart rate, blood pressure, and ventilatory gases were measured. Ventilatory anerobic threshhold (MMI), Quetelet index (QI) of ponderosity, and time of exercise after the VMI were calculated for all subjects. Baseline heartrate (HMR), heart rate at maximum exercise (MaxMR), and 1 minute recovery heart rate (HMR) for males (M) and females (F) are shown in the table, with standard deviation.

	EHR	MaxHR	RHR
M	99.3 (+ 20.4)	193.2(+ 11.4)	140.9 (+ 19.2)
F	98.8(+ 14.6)	196.9(+ 19.2)	153.4 (+ 15.5)

The only significant (P < .001) intersex difference found was the recovery heart rate (NFR) at 1 minute, females having higher 1 minute recovery heart rates than males. This could not be explained by differences in age, proderosity (II), Oxygen debt (time beyond NFM), or cardiquilmonary condition (NFM). Recovery heart rate at one minute has been used as a standard indicator of overall physical fitness. In the age group studied, females returned to baseline at a slower rate than did age and HSA metched males. This is not related to (II, time at AT Max heart rate, VO₂ max, or VAT. Recovery heart rates should not be used as an index of physical fitness unless compared to age-garder specific normal values.