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EPIDEMIOLOGIC ASPECTS OF KAWASAKI SYNDROME IN MID-MANHATTAN AND IN JAPAN. Nunzia S. Fatica, Fukiko Ichida, Mary Allen Engle, Kathryn H. Ehlers, Arthur A. Klein, Aaron R. Levin, John E. O'Loughlin, Michael S. Snyder. Cornell University Medical College-The New York Hospital, Pediatric Cardiology, New York, NY

We compared epidemiologic and clinical features of Kawasaki Syndrome (KS) in 110 pts aged 2 wks to 12 9/12 yrs and seen Jan '80 to Aug '86 with large series from Japan. Similarities: age below 5 yrs (72%), sex (61% male), urban and suburban dwelling almost entirely, recurrence rate (2%), myocardial infarction (1%), and death rate (1%). Incidence of coronary aneurysms (18%) was associated in both areas with duration of fever ( $\geq 14$  days) and in NY with Oriental race and height of platelet count. Yearly seasonal incidence was highest in springtime, as in Japan during epidemics.

Dissimilarities: Heterogeneity in NY (50% Caucasian, 18% Black, 16% Hispanic, 16% Asian); mean age in NY ( $3\frac{1}{2}$  yrs) higher than in Japan ( $1\frac{1}{2}$  yrs); 32% <2 yrs in NY vs 50-60% in Japan; higher incidence of clinically apparent acute myocarditis (8%), but lower incidence of late cardiac failure (1%) and valvular regurgitation (2%) in NY.

Personal interview with 60 families revealed use of rug shampoo within 1 month of onset in 12 (20%) and 1 case of recurrence after 2nd shampoo. This was the only family with 2 siblings affected, 1 wk apart.

We conclude that KS in a heterogeneous Manhattan population resembles KS in Japan except for age distribution. In NY it occurred more often and more severely in Asians than in non-Asians.

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ERYTHROPOIETIN TITERS IN CHILDREN WITH CYANOTIC CONGENITAL HEART DISEASE  
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Previous reports have suggested that patients with polycythemia secondary to congenital heart disease have elevated serum erythropoietin titers (EPO). To further define the predictors of EPO in these patients, we measured hemoglobin concentrations (Hb), PaO<sub>2</sub>, aortic oxygen saturation (SAT O<sub>2</sub>), iron storage variables, red cell 2,3-DPG, and serum EPO in 17 children less than 8 years of age with SAT O<sub>2</sub> < 90%. Patients were divided into 2 groups, those with increased EPO (> 30 mpu/ml) and normal EPO. Results are shown in the table. Two patients were iron deficient and both had increased EPO. For children under 8 yrs of age, elevated EPO titers were associated with severe hypoxemia and elevated red cell 2,3-DPG. However, we found children with moderate hypoxemia and iron sufficiency to have normal EPO titers at Hb levels below those which caused significant hyperviscosity.

TABLE  
Results given in Mean  $\pm$  S.D.

	N	EPO mpu/dl	Hb gm/dl	SAT O <sub>2</sub> %	PaO <sub>2</sub> mmHg	DPG $\mu$ mol/ml
High EPO	5	47.3 $\pm$ 8.54	18.2 $\pm$ 2.6	68 $\pm$ 12	36 $\pm$ 7	3.21 $\pm$ 0.73
Low EPO	12	9.3 $\pm$ 6.4	16.4 $\pm$ 2.1	80 $\pm$ 6	48 $\pm$ 8	2.45 $\pm$ 0.35

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DOSE REQUIREMENTS FOR MAXIMAL EFFECTS OF PROPRANOLOL IN PEDIATRIC PATIENTS. Daphne T. Hsu, Daniel T. Williams, Henrietta Wolland, Welton M. Gersony, Columbia University College of Physicians and Surgeons, Department of Pediatrics, New York City, New York.

The maximal effective dose of propranolol (P) has not been well-defined in pediatric patients (pts.). Extremely high doses of P have been recommended for the treatment of SVT and for palliation of tetralogy of Fallot, but neither maximal nor optimal dosages have clearly been determined. We studied a group of 7 pts. (mean age 12.3 $\pm$ 3.2 yrs.) who were treated on a research protocol for uncontrolled rage outbursts with P doses far beyond those generally recommended. Following a baseline period, P was administered in a stepwise fashion, reaching maximal doses of 200-490 mg TID (12-25.1 mg/kg/day) and achieving peak P levels of 300-691 ng/ml. At maximal doses, the mean decrease from baseline in HR was 14.5%, in SBP was 10.1%, and in DBP was 13.9% (p<0.001). No significant adverse symptoms occurred. The minimum P dose significantly affecting resting HR and BP ( $\geq 1$  S.D. below baseline) varied in individual pts.; the minimum P dose that produced maximal effects on resting HR and BP was often well above previously recommended dosage (table). The P dose required to produce maximal effects on resting HR and BP was variable, but averaged 1.7-2.2 times greater than usually recommended. Further increases in dosages were well tolerated and produced no adverse effects on HR or BP. The current maximal P dosage recommendations may require modification.

	DOSE(mg/kg/day)	RANGE	MEAN
Recommended		1.0-5.0	3.0
Sign. effect	HR	0.3-4.8	3.0
	SBP	0.3-8.4	2.7
	DBP	0.3-2.7	1.3
Max. effect	HR	2.7-10.1	6.5
	SBP	3.6- 8.4	5.8
	DBP	0.5-10.6	5.2

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CARDIAC SIZE AND FUNCTION IN SCHOOL ATHLETES AT REST AND AFTER EXERCISE. Tarek S. Husayni, Juan Longhi, Marc Puczynski, Regina Smarto, and Anthony F. Cutilletta, Loyola University Stritch School of Medicine, Department of Pediatrics, Maywood, Illinois.

To determine the effect of high school athletics on cardiac structure and function, we performed 2-D directed M-Mode echos on students involved in isometric (IM) and isotonic (IT) sports and compared them to non-athletic controls (C). Measurements were made at rest and immediately after stage 4 treadmill exercise. Twenty students were included in each group. LV mass was greater in both IM and IT compared to C (C 111 $\pm$ 4, IM 132 $\pm$ 3, IT 130 $\pm$ 4 g/M<sup>2</sup>, p<0.05). In IT LV diastole volume (LVDV) was increased with normal wall thickness/radius ratio (t/r), whereas, in IM, LVDV was normal but, t/r was increased. Stroke index (SI) was increased in IT (C 44 $\pm$ 1.7, IM 46 $\pm$ 1.6, IT 52.3 $\pm$ 2.0 ml/M<sup>2</sup>, p<0.02). Cardiac index (CI) and shortening fractions were similar among the three groups. After exercise LVDV and systolic volume decreased in both C and IT but, remained unchanged in IM. CI increased with exercise in all groups. Although resting systolic (S), diastolic (D), and mean (M), blood pressures (BP) were similar in all groups, after exercise both C and IT had an increase in SBP, (C 39%, IT 32%) and a fall in DBP, (C-26%, IT-27%) MBP remained unchanged. In IM, SBP rose less, 22%, and DBP fell less, -18%. These data suggest that students engaged in ordinary school sports can develop either eccentric or concentric LVH depending upon the type of sport. The IM athlete appears to have a thicker, less compliant LV and demonstrates less change in blood pressure with exercise.

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ALTERATIONS OF ENERGY METABOLISM IN HYPERTROPHIED HEARTS DURING ANOXIA: A P-31 NUCLEAR MAGNETIC RESONANCE STUDY. Mark D. Jacobstein, Pierre G. Carlier, Mark H. Cohen, Majeed Bhat and Thomas A. Gerken. (Spon. by Jerome Liebman). CWRU School of Medicine, Department of Pediatrics, Cleveland

The metabolic effect of anoxia on hypertrophied myocardium was investigated using P-31 NMR spectroscopy. Hearts from 18 mo. SHR (hypertrophied, n=7) and age-matched WKY (control, n=6) rats were mounted on a modified Langendorff apparatus, paced at 240/min and perfused with 36°C phosphate-free, glucose-containing buffer bubbled with oxygen or nitrogen (anoxia). Left ventricular pressure (LVP), LV dP/dt and perfusion pressure were continuously recorded while consecutive 3-min spectra were collected. This allowed temporal assessment of myocardial phosphate levels [incl. inorganic phosphate (P<sub>i</sub>), creatine phosphate (CP) and ATP] during baseline conditions, anoxia and recovery. Anoxia was maintained until a 70% fall in LVP occurred. Compared to the WKY, SHR rats had higher in vivo BP (163 vs 104; p<.001) and cardiac hypertrophy (heart/body weight = 5.3 vs 3.5 mg/g; p<.001). During baseline perfusion, SHR hearts had higher resistance (9.8 vs 5.9 mmHg/cc/min/g; p=.003) but no significant difference in LVP or dP/dt. SHR hearts demonstrated a faster fall in high-energy phosphates and LVP during anoxia (SHR fell to 30% baseline LVP in 8.5 vs 13.0 mins; p=.018). Throughout the protocol SHR hearts had lower CP (3.8 vs 5.6  $\mu$ moles/g; p=.008), ATP (3.2 vs 3.9  $\mu$ moles/g; p=.047) and CP/P<sub>i</sub> ratio (1.5 vs 2.5; p=.043). We conclude that chronically hypertrophied hearts have: (1) less ATP/g and CP/g than age-matched controls, and (2) a faster decline in mechanical and metabolic function during anoxia.

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EVALUATION OF UNIVENTRICULAR ATRIOVENTRICULAR CONNECTION BY NMR IMAGING.

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Twelve patients (pts), ages 7 mos. to 23 yrs. (mean 11 yrs.), with known univentricular atrioventricular (AV) connection ("Single ventricle") were evaluated by ECG-gated magnetic resonance imaging (MRI). Multiple sections were obtained in transverse, coronal and oblique planes using a 0.3T or 1.0T magnet and SE 30 pulsing techniques. MRI studies were evaluated using a segmental approach prior to reviewing the echocardiograms and angiograms which were available on all pts. MRI accurately depicted: 1) cardiovisceral and atrial situs 2) presence of splenic tissue 3) systemic and pulmonary venous connections 4) the mode of AV connection (i.e. absent right, double inlet, etc.) 5) the morphology and spatial relationships of the main and rudimentary ventricular chambers (6) the ventriculoarterial (VA) connections and 7) surgical procedures (i.e. shunts, PA bands). Seven pts had absent right AV connection, 4 had double inlet anatomy and 1 had an absent left AV connection. The absent AV connection was true atresia in 6 with invagination to the cardiac crux of fat-containing sulcus tissue which appears bright on MRI and an imperforate valve in 1. In 11 pts, the rudimentary chamber could be visualized. VA discordance was demonstrated in 2 pts with absent right AV connection, in all 5 with double inlet LV's and the 1 with absent left AV connection. We conclude that MRI accurately depicts cardiac morphology in pts with univentricular AV connection, often providing information not available with other imaging techniques.