● 151 PACEMAKER THERAPY FOR SICK SINUS SYNDROME IN CHILDREN

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We evaluated our recent experience with pacemaker (PM) treatment of sick sinus syndrome (SSS) in pediatric patients (pts). In the past 5 years, 21 pts underwent implantation of permanent PM for SSS: 12 had had prior surgery for associated structural heart disease (SHD), 5 had no SHD; 2 had drug-induced SSS, 1 had unoperated SHD, 1 had cardiomyopathy. The median age was 8 yrs (range 2-21 yrs). Twenty pts had syncope or marked exercise intolerance. Each pt had an intracardiac electrophysiologic study and each had an abnormal corrected sinus node recovery time (mean 687 ± 53msec SEM). Supraventricular tachycardia (SVT) was documented in 15 pts: 7 atrial muscle reentry, 4 sinus node reentry, 2 Kent bundle reentry, 1 AV node reentry, 1 atrial ectopic focus. Ten pts had normal AV conduction and received an epicardial atrial PM. Eleven pts had abnormal AV conduction and received an epicardial ventricular PM. No pt has died during mean followup of 31 mos (range 6-63 mos). Symptoms due to bradycardia were relieved in each pt. SVT has become easier to control in 11/15 pts; SVT worsened in both pts with Kent bundles. One atrial PM and 2 ventricular PMs failed to sense, requiring revision. No PM has failed to capture and no battery has yet depleted. We conclude: pacing for SSS in pediatric pts reliably relieves symptoms of bradycardia, facilitates treatment of SVT and may prevent sudden death.

152 TWO DIMENSIONAL ECHOCARDIOGRAPHIC (2DE) ASSESSMENT OF RIGHT VENTRICULAR VOLUMES (RVV) AND EJECTION FRACTION (EF).

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The ability of M-mode and 2DE to measure RVV and EF was assessed in 15 children without right ventricular outflow tract obstruction (RVOTO) (Group I). M-mode measurement of end diastolic dimension (RVEDD) was performed in the usual fashion. From the apical 4 chamber 2DE view the long axis of the right ventricle (RVLA) was measured from the apex to mid tricuspid valve, and area planed (RVA). From the anterior-posterior (AP) and lateral (lat) angiographic (angio) views the right ventricular body (RVB) longest length was measured, area planed, and volumes calculated using a Simpson's Rule algorithm. Data were compared by linear regression analysis (r=correlation coefficient, a=slope). RVEDD by M-mode correlated poorly with angio data. The 2DE obtained RVLA length correlated well with the maximum angio obtained length from either the AP or Lat views at both end systole (ES) and end diastole (ED). 2DE RVA also correlated with the planed area obtained from either AP or Lat angio projections. To determine the relationship between RVB volume (RVBV) and total right ventricular volume (TRVV), these volumes were separately assessed in 30 other children without RVOTO (Group II). A constant relationship existed between RVBV and TRVV such that RVBV/TRVV=0.75. By 2DE RVBV was determined from the apical 4 chamber view by an area-length algorithm assuming an ellipsoid model. Accurate 2DE assessment of TRVV and EF in Group I patients could be achieved by the equation, Volume = 1.34 X echo calculated RVBV; (EDV r=0.92, a=0.98; ESV r=0.84, a=0.95; EF r=0.80, a=1.19). Therefore, 2DE is able to accurately measure RVV and EF in patients without RVOTO.

153 TREATMENT OF CARDIAC ARRHYTHMIAS WITH DISOPYRAMIDE (NORPACE)

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Disopyramide (D) has been reported to be an effective antiarrhythmic agent in adults; little experience is available using D in pediatric patients. We have treated eleven patients, ranging in age from 10-18 years (median-13 years) with D for ventricular (8), and supraventricular (3)arrhythmias. Pharmacokinetic data was obtained in selected patients.Two patients were given D intravenously prior to beginning oral therapy. The $T_{\frac{1}{2}}$ of elimination was 4.8 hours. D was primarily excreted unchanged in the urine in the first 24 hours. The oral dosage varied from 100-150mgm every 6 hours, 6-14mg/kg/day, (median 11mg/kg/day). Peak plasma D levels occurred between 2-4 hours after an oral dose and ranged from 3.0-3.5mg%. Of the 8 patients treated with D for ventricular arrhythmias, 6 had excellent control of the arrhythmia. Four of these patients had previously had poor results with Quinidine. In 2 patients D had to be discontinued;one because of GI side effects and the second due to D enhancement of periodic paralysis. Two patients had atrial flutter (AF). In one the AF was converted to sinus rhythm and in the second there was no effect on the AF. A third patient with recurrent supraventricular tachycardia D had to be discontinued because of its anti-cholinergic effects. In conclusion, D appears to be a safe and effective alternative antiarrhythmic agent in pediatric patients, particularly for the treatment of recurrent ventricular arrhythmias.

154 SINUS NODE RECOVERY TIMES FOLLOWING REPAIR OF TRANSPOSITION OF THE GREAT ARTERIES. COMPARISON BETWEEN MUSTARD AND SENNING OPERATION.

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In order to determine the effect of interatrial baffle repair for transposition of the great arteries(TGA) on sinus node function, we compared percent sinus node recovery times(%SNRT) in 44 TGA pts, 16 before and 28 one year after operation(op). Ten pts had Mustard(M) repair and 18 had Senning's(S) op. The 16 preop pts served as controls. Results are shown as mean %SNRT, ±SD (n) (*$p < 0.05$ compared to control).

Cycle Length(CL)	Preop (16)	Mustard (10)	Senning (18)
667 msec	116(1)	140±23(2)	124±36(2)
500	101±19(3)	142±22(6)*	123±22(9)
400	120±18(13)	133±16(10)	130±12(15)
333	115±12(15)	120±33(9)	134±25(17)*
286	115±9(13)	147±56(5)	133±36(16)

The postop mean %SNRT were slightly longer than preop and no significant differences between M and S groups occurred. Six of 10 M pts and 9 of 18 S pts had prolonged %SNRT (> 2 SD above mean preop)at one or more CL. One pt from each group had sick sinus syndrome and prolonged %SNRT (236%, 244%, $p < 0.05$).

These data indicate that there is no advantage of one operation over the other regarding alterations in sinus node function. A significantly prolonged %SNRT may identify those at risk for developing serious arrhythmia.

● 155 ABNORMALITIES OF BREATHING CONTROL AND AIRWAY MAINTENANCE IN INFANTS AND CHILDREN AS A CAUSE OF COR PULMONALE.

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Respiratory control abnormalities may result in cor pulmonale. This report summarizes the clinical history, diagnostic evaluation, treatment and outcome of 15 infants and children presenting with cor pulmonale subsequently found to be due to sleep-dependent hypoventilation. Thirteen patients had cardiomegaly and RVH while two had only severe RVH. Four infants with Central Hypoventilation Syndrome (CHS) had severe sleep-dependent asphyxia and resultant acute respiratory failure; all were ultimately treated with phrenic nerve pacing. One patient with Alveolar Hypoventilation Syndrome, a less severe ventilatory deficit than CHS, presented with severe pulmonary hypertension and ultimately died despite symptomatic relief with respiratory stimulants. Ten patients presented with obstructive sleep apnea and secondary asphyxia. Four children underwent T&A with normalization of symptoms in one, clinical improvement in two and no clinical improvement in another. This unimproved patient and the six remaining obstructive sleep apnea children improved dramatically after tracheostomy to bypass the sleep-dependent airway obstruction; none presently have evidence of cor pulmonale. In summary, early recognition and appropriate treatment of sleep-dependent airway obstruction will eliminate sleep hypoventilation and associated sleep deprivation symptoms, and reverse the cor pulmonale.

156 EFFECTS OF COLD CARDIOPLEGIA IN TETRALOGY OF FALLOT REPAIR.

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The value of cold cardioplegia was studied in 196 successive patients receiving three different methods of myocardial preservation during intracardiac repair of Tetralogy of Fallot. Group A (n=100)were repaired normothermic using intermittent aortic arch clamping (IAAC). Group B (n=43) received moderate hypothermia (28°C) and IAAC. Group C (n=53) were given moderate hypothermia and cold cardioplegia fluid. There was no significant difference in sex, type of right ventricular outflow tract obstruction or pre-op hemoglobin between groups. More Group A patients had a previous shunt ($p < .01$) and had higher pre-op systemic saturations ($p < .01$). Group B patients tended to be older and larger at repair though these patients had significantly higher incidence of early death (within 30 days post-op),($p < .01$). Aortic cross clamp time was significantly longer in C patients than other groups ($p < .01$). Post-op pressures in the right ventricle ($p < .01$) and pulmonary artery ($p < .01$) were lower in C patients. The major difference between groups occurred in comparing duration of inotropic support. Group A patients required 33 + 2.9 hours,Group B 39 + 4.3 hours. Significantly, Group C patients required only 12.5 + 2.9 hours ($p < .001$) of dopamine or isoproterenol to maintain clinically normal cardiac output in the post-operative period. In conclusion, cold cardioplegia offers markedly better myocardial preservation than previous methods, allowing longer operative time with less compromise of cardiac output in the post-operative period.