QTc RESPONSE TO ATRIAL PACING IN CONGENITAL LONG QT SYNDROME.

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A number of patients may present with symptoms and arrhythmias suggestive of congenital long QT syndrome (LQTS) but have only borderline or mildly prolonged corrected QT interval (QTc) on resting electrocardiogram The purpose of this study was to determine if atrial pacing at the time of electrophysiologic study (EPS) could be used to differentiate those pts with LQTS from normals. Using Bazett's fromula, the maximal QTc was determined on resting ECGs at presentation and during atrial pacing at multiple cycle lengths from 600 -300ms.Twenty -seven pts had LQTS diagnosed by clincal and electrocardiographic criteria: QTc > .45s, torsades de pointes or complex ventricular arrhythmias, and syncope or cardiac arrest associated with exercise or emotional stresses. The normal group was comprised of 15 pts with no intrinsic heart disease and no pre-excitation who were studied for evaluation of supraventricular arrhythmias. No pts were on medication at the time of EPS. The maximal QTc (mean ± SD) values are shown below;

	ECG RESTING	EPS PACED
LQTS	.45s ± .06	.54s ± .06
Normals	.40s ± .02	.43s ± .01
p	< 0.03	< 0.0001

While only 10 LQTS pts had QTc > .45s on presentation, all 27 had QTc > .46s in response to atrial pacing. None of the normals had QTc prolongation (>.45) on either resting ECG or with pacing. The maximal QTc occurred at a similar paced cycle lengths in the LQTS and the normals (460 ±90 ms vs 463 ± 96ms with a median of 450 ms in both). There was a wider range of QTc response to pacing seen in the LQTS compared with the normals (0.46-0.64 s vs. 0.4-0.45 s).
Conclusions: Atrial pacing can differentiate between pts with and without LQTS and guide therapeutic decisions in these pts at high risk for ventricular arrhythmia and sudden death.

LATE POSTOPERATIVE ARRHYTHMIAS FOLLOWING REPAIR OF TETRALOGY OF FALLOT : COMPARISON BETWEEN TRANS-ATRIAL AND TRANS-VENTRICULAR APPROACHES.

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Onehundred and five patients (pts) who were discharged following intracardiac repair of Fallot's Tetralogy, between September 1980 and August 1988, were followed for at least 2 years. The purpose of this study is to compare the incidence and severity of late postoperative ventricular arrhythmias between two surgical approaches. Group 1 included 71 pts, aged 0.2 to 61 years (mean 6.8 years) using the classical RV approach. Group 2 consisted of 34 pts, aged 0.6 to 39 years (mean 7.9 years), repaired through the RA. Follow-up ranged from 33 to 114 months (mean 72.9 months) in group 1: 64 pts are in functional class (FC) I, and 2 pts required reoperation (with 1 late death). There were no late deaths or reoperations in group 2, and all 34 pts are in FC I, during a mean follow-up of 45 months (range 24 to 72 months). In group 1, six pts (8.4%) have palpitations, and premature ventricular complexes (PVC) on surface EKG. Severe ventricular arrhythmias (≥ Lown grade 2) were present in 20 of 66 pts (30.3%) who underwent 24-hour ambulatory monitoring, and in 9 of 43 pts (20.9%) during treadmill exercise tests. Postoperative catheterization in 32 pts showed elevated RV systolic pressure (≥ 60 mmHg) in 2 pts. Sustained ventricular tachycardia was reproduced in electrophysiologic studies in 4 pts, and the site of reentry was in the ventriculotomy scar. In group 2, none have palpitations, or PVC on EKG. 24-hour ambulatory monitoring in 34 pts, and treadmill exercise test in 20 pts demonstrated no severe ventricular arrhythmias. In summary, the right atrial approach significantly ($p<0.001$) reduces the incidence and severity of late postoperative arrhythmias after repair of Fallot's Tetralogy.

DUAL CHAMBERED PACING IN CHILDREN: THE OPTIMAL LEAD CONFIGURATION - UNIPOLAR OR BIPOLAR?

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Cardiac sensing with bipolar pacing leads (BI) has been suggested to be superior to the unipolar mode (UNI). However, little is known of the differences in chronic capture thresholds between the two modes. Our purpose was to determine the optimum method of dual chambered cardiac pacing in children.

Twenty-four hour ECGs and a voltage (V) - pulse width strength duration relationship were performed in pacemaker patients (PTS) during follow-up. Atrial endocardial leads had been implanted in 142 PTS (UNI - 100, and BI - 42); ventricular endocardial leads in 147 PTS (UNI - 99, and BI - 48). All pacing leads were actively fixed.

We found: 1) atrial oversensing occurred less commonly with BI sensing (5%) than UNI (21%), $p< 0.01$. In the UNI mode, atrial oversensing events usually occurred < 3/day and resulted in pauses between 1-2 secs. 2) Ventricular oversensing occurred equally in the two modes (12 vs 8.5 %, p=NS). 3) Atrial pacing threshold (0.5 msec) was lower with UNI leads (1.9 +/- 0.8 vs 3.4 +/- 2.8 V, $p< 0.05$). 4) Similarly, ventricular pacing threshold (0.5 msec) was lower with UNI leads (1.8 +/- 0.9 vs 2.4 +/- 0.8 V, $p< 0.01$).

Pacemaker longevity was predicted using the voltage strength duration relationship and measured lead impedance. Assumed was 100% ventricular pacing at a rate of 100/min, 2X voltage safety margin, 50% atrial pacing, and battery capacity = 1.8 amp-h. Predicted pacemaker longevity in the atrial and ventricular UNI paced mode (8.5 yrs) exceeded the BI paced mode (5.8 yrs).

Our results suggest that if pacemaker longevity is of primary importance then atrial and ventricular pacing should be performed in the unipolar mode.

RESISTANT SUPRAVENTRICULAR TACHYCARDIA IN CHILDREN - A ROLE FOR CATHETER FULGURATION?

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The management of resistant supraventricular dysrhythmias (RSVT) in children is challenging, particularly when arrhythmia surgery (Sx) and medical therapy (Mx) have failed. We performed direct current catheter fulguration (Abl) on 9 children (pts), mean age 10 years (3.5-14.9 yrs). There were two groups: a) 6 pts had RSVT associated with an accessory connection (AC) - 5/6 had failed prior Sx to interrupt the AC. b) 2 pts with atrial ectopic tachycardia (AET) and 1 pt with atrial flutter (AF) had cardiomyopathy, 2/3 following corrective Sx for congenital heart disease. A total of 11 attempts at Abl were performed, delivering a mean of 2.8 shocks /attempt, mean 3.8 joules/kg/shock. In each case, the energy was delivered using a standard platinum-tipped electrophysiology catheter as the cathode.
Results: a) 5/6 pts with AC had successful Abl and are SVT and medication-free (mean follow up 1.4 yrs). One pt failed both catheter and subsequent Sx Abl. b) 2/3 pts with AET had successful planned His bundle Abl and are symptom-free following pacemaker insertion (mean follow up 0.8 yrs). Abl of an AF focus failed in 1 pt. There were no major complications or deaths.
Conclusions: Pending further technologic refinements, catheter fulguration for RSVT can be safely and reliably used in children, particularly when arrhythmia Sx has failed or when poor myocardial function precludes surgery or aggressive antiarrhythmic medication.