

811-4

**Use of Resynchronization Therapy in Pediatric Patients With Congenital Heart Disease and Complete Heart Block**

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**Background:** Biventricular pacing has been shown to be a beneficial therapy in adult patients with ventricular dysfunction and intraventricular conduction delay. However, no pediatric applications for this therapy have been tested. We hypothesized that pediatric congenital heart disease patients with complete heart block and poor ventricular function would benefit from conversion to biventricular pacing from traditional single site atrioventricular (DDD) pacing.

**Patients:** A total of 6 patients with congenital heart disease, poor ventricular function and complete heart block were included in this study. Median age at the time of conversion to biventricular pacing was 11.1 months, ranging from 5.6 to 118 months. Diagnoses included complete atrioventricular canal (1), aortic stenosis/subaortic stenosis (4), anti-Ro, anti-La cardiomyopathy (1). Patients were all treated for heart failure prior to conversion to biventricular pacing, with digoxin (6/6), ACE-inhibitor (6/6), and carvedilol (2/6). Baseline ejection fraction with DDD pacing was 34 ± 12%. Median duration of followup after conversion to biventricular pacing was 2.6 months, ranging from 4 days to 5.8 months.

**Results:** After conversion to biventricular pacing the ejection fraction increased to 57 ± 12%, p<0.004. In all subjects, the ejection fraction improved, with a minimum improvement of 12 EF units. Five patients also showed an improvement in weight for age, p=0.04. Other clinical improvement was noted, with 1 patient removed from the transplant list, and 1 patient tolerating reduction in medical support.

**Conclusions:** Biventricular pacing appears to offer short-term benefit for selected pediatric patients with complete heart block and poor left ventricular function. Further studies looking at the long-term benefit of this therapy in the pediatric population are needed.

Noon

811-5

**Need for Pacemaker Implantation in Patients Late After Atrial Correction for Transposition of the Great Arteries**

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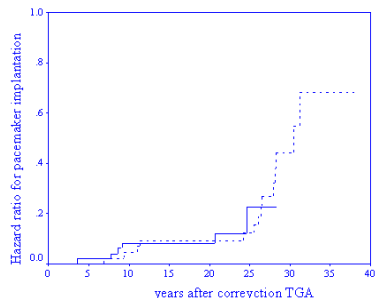
**Introduction** Many patients (pts) with transposition of the great arteries (TGA) who underwent atrial correction according to Mustard (Mu) or Senning (Se) develop rhythm or conduction disturbances requiring pacemaker (pm) implantation. We investigated the need for pm implantation during a 38 years follow up and compared the Mu and Se pts.

**Methods.** 117 pts who underwent surgery between 1961 -1987 according to the Mu (n=57) or Se (n=60) procedure, and survived for more than 30 days after operation, were included. Maximal follow up in the Mu group was 38 y, maximal follow up in the Se group was 28 y. During follow up, 16/57 Mu pts died, and 14/60 Se pts died.

**Results.** After a follow up of 27 y, the number of pacemakerimplantation is identical in both groups (23%). Longer follow up is only available for the Mu group. Between 27 y and 38 y follow up, the number of pm implantations shows a dramatic increase, from 23% to 68%.

**Conclusion.**

After 27 y of follow up, the number of pm is identical in Mu and Se pts. Follow up of the Mu group up to 38 y, shows a dramatic increase in the number of pm.



**1095 Adult Congenital Heart Disease**

Monday, March 08, 2004, Noon-2:00 p.m.

Morial Convention Center, Hall G

Presentation Hour: 1:00 p.m.-2:00 p.m.

1095-199

**Disparity Between Dobutamine Stress and Physical Exercise Magnetic Resonance Imaging in Patients With an Intra-Atrial Correction for Transposition of the Great Arteries**

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**Background** Controversial reports have been published on the hemodynamic response to dobutamine stress and physical exercise in patients with TGA. The purpose of this study was to compare the hemodynamic responses to dobutamine stress and physical exercise in patients with an intra-atrial correction for transposition of the great arteries (TGA).

**Methods** Thirty-nine patients with an intra-atrial correction for TGA and 25 age/sex-matched control subjects underwent either dobutamine stress MRI (15 µg/kg/min) or underwent submaximal physical exercise. End-systolic and end-diastolic right ventricular volumes (ESV;EDV) were determined. In five of these patients, who underwent both stress tests, wall thickening reserve was calculated as systolic wall thickening during stress minus systolic wall thickening at rest.

**Results** The responses were not different between dobutamine and exercise in controls: stroke volume, cardiac output and ejection fraction increased significantly, whereas ESV significantly decreased and EDV was unchanged. In contrast, in patients stroke volume did not increase with either dobutamine or exercise (-8.6% versus 2.9%). Ejection fraction increased significantly with dobutamine but tended to decrease during exercise (16% versus -2.1%, P=0.001). EDV and ESV decreased during dobutamine but were unchanged during exercise (-22% versus 5.0%, P=0.001; -36% versus 9.0%, P=0.01 respectively). Wall thickening reserve was higher with dobutamine than with exercise (0.9 mm versus -0.6 mm, P=0.02).

**Conclusion** This study confirms inadequate stroke volume response during stress in patients with TGA and reveals a disparity between dobutamine and physical exercise. Unchanged ESV, ejection fraction and wall thickening during physical exercise indicate systolic dysfunction, whereas dobutamine showed relatively normal systolic responses with a decreased diastolic filling.

1095-200

**Increased Metabolic Rate as a Mechanism for Cachexia in Adults With Congenital Heart Disease**

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**Background:** Cardiac cachexia is an independent predictor of mortality in the chronic heart failure population, and cachexia has been linked to increased basal metabolic rate in heart failure patients. Approximately 12% of adults with congenital heart disease (ACHD) have cachexia; however causative factors are unknown. Understanding mechanisms underlying cachexia in ACHD is important in the potential development of therapeutic interventions.

**Hypothesis:** We hypothesized that elevated basal metabolic rate is a contributing factor to cachexia in adult patients with congenital heart disease.

**Methods:** Cachexia was defined as a BMI less than 20 and at least one of the following two criteria for body wasting: reduced body fat (<22% for women, <15% for men by bio-impedance) or low percent predicted ideal body weight (<=85%). Twenty cachectic (age 30 ± 10 years; 6 females) and 19 noncachectic (age and gender matched) adults with a variety of congenital heart lesions were studied in the fasting state. Basal metabolic rates were measured for 30 minutes in the supine position by indirect calorimetry (Deltatrac, Datex Ohmeda). Serum was assayed for thyroid function hormones and cortisol levels.

**Results:** A greater proportion of cachectic patients had complex disease than non-cachectic patients (65% vs. 20%, p= 0.01). Measured basal metabolic rate was significantly higher (22.5%) in patients with cachexia (27.2 ± 2.7 kcal/kg) compared to those without cachexia (22.6 ± 3.2 kcal/kg), p<0.0001. Although free T4 and TSH were not different between the two groups, serum cortisol levels were significantly increased in cachectic patients (412 ± 83 nmol/L vs. 273 ± 80 nmol/L), p=0.01.

**Conclusion:** Basal metabolic rate is increased in cachectic adult patients with congenital heart disease. This may be related to increased serum cortisol in the setting of complex congenital heart disease, suggesting a catabolic/anabolic imbalance as a contributing factor to body wasting in these patients. Further studies are warranted to determine if nutritional supplementation can improve prognosis among cachectic adult congenital patients.