Reprint of Aortic Coarctation in Infants: Results of a Tailored Surgical Approach in the Modern Era

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Background: The surgical repair of aortic coarctation in infants has evolved over time, largely in response to high rates of residual or recurrent coarctation. This review evaluates our current approach utilizing extended and/or transposition anastomosis without prosthetic material to enhance all areas of aortic arch hypoplasia.

Methods: The Michigan Congenital Heart Center Surgical database was reviewed for infants who underwent isolated repair of aortic coarctation from January 1, 1990 to January 1, 2000. Patients who underwent simultaneous repair of other lesions and those with significant associated lesions were excluded.

Results: Eighty-three infants underwent surgical repair of isolated coarctation during this decade. Median age at repair was 21 days (range 3-363 days). Repair was performed in the first week in 72 patients while the remaining 11 underwent median sternotomy with circulatory arrest because of severe transverse arch hypoplasia. There were 2 deaths: 1 due to pulmonary hypertension in a patient with alveolar capillary dysplasia and 1 late death of pneumonia in a patient with non-cardiac anomalies. Neither had residual coarctation. Technique related complications of bronchial compression, chylothorax, and vocal cord paralysis were noted in 4 patients. At discharge, 14 patients had a gradient ≥ 15 mmHg by both physical exam and echocardiography. Follow-up data were available for 65 patients (78%) with mean follow-up duration 4.3 years (SD ± 1.1 years); 1 patient received surgery for hypertension after 1 month, and 3 underwent balloon angioplasty within 7 months of initial repair. The remaining 60 patients are asymptomatic, with no age-dependent epigastric aneurysms, and have aortic arch gradients < 15 mmHg. Thus far, this low recurrence rate approaches that reported with other techniques in older children.

Pulmonary Position Cryopreserved Homografts: Durability in Ross and Non-Ross Patients

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Actuarial 5-year survival rates for right ventricle to pulmonary artery (RV-PA) homografts are reported to range between 49% and 81%. It is not known whether there is a difference in homograft durability when utilized for right ventricular outflow tract (RVOT) disease or for the Ross operation for pulmonary autograft replacement of the aortic valve. To evaluate outcome and risk factors for implant failure in patients who have undergone pulmonary position homograft placement for RVOT obstruction or the Ross operation, the records of all patients receiving an RV-PA homograft at this institution from January 1990 to January 2001 were reviewed. 148 consecutive patients were included in the study (65 Ross and 83 RVOT repairs). Adequate data was available on 136 (92%). The median follow-up time was 3.8 years (1-11 years). Right ventricular homograft survival after Ross and non-Ross operations at 5 years was 85% and 56% respectively (p<0.05). Potential differences in 5 years for homograft failure analyzed: age at operation, diagnosis, type of surgery, homograft type (aortic vs. pulmonary) and width of homograft (mm). On univariate analysis, young age, homograft width, and non-Ross operation were risk factors for homograft failure (p<0.01). On multivariate analysis: smaller homograft width was the single predictor of graft failure (p<0.01). Outcomes were compared for 5 age groups: 10 years (n=99). Homograft failure rates were 35%, 20%, and 9% for the 2 groups respectively. For non-Ross patients: failure rates were: 43%, 25% and 14% and for Ross patients: 12.5%, 5% and 0%.

Conclusions: Ross patients had significantly longer homograft survival than RVOT disease patients. 2. Smaller homografts placed in younger patients required earlier intervention.

Pulmonary Position homografts inserted in Ross patients at young ages (<10 yrs) lasted longer than similar aged non-Ross patients: whereas patients older than 10 yrs at the time of operation had similar failure rates.

Surgical Correction of Tetralogy of Fallot in Adults in the Current Era

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BACKGROUND: Information on the surgical results in adult uncorrected Tetralogy of Fallot (TOF) patient is scanty. METHODS: In this retrospective study we sought to (1) examine the results of TOF surgery at our institution and (2) review the surgical outcome, (3) analyze the frequency and determinants of late deaths and mid-term (mean 34 ± 14 years) provide long-term survival data.

RESULTS: From 1980 to 12000, a total of 221 patients had surgical correction of TOF in our center. Twenty-four patients older than 18 years were identified on clinic (mean 34 ± 14 years). Eleven patients (47%) had immigrated to the United States. Reasons for delayed surgical correction in the immigrants included a lack of surgical facilities in their home countries and/or non-diagnosis. In the permanent residents, definitive surgical correction had not been undertaken due to obstetrical pollution after palliative procedure (1941-1947), non-compliance with medical advice (3/14, 21.4%) and institutionalization for Downs syndrome (2/14, 14.3%). Surgical corrections were successful in all patients.

Three patients (12.5%) developed conduction disturbances after operation. Seven patients (29.2%) developed clinically significant atrial arrhythmias and 1 patient (4.2%) developed repeated episodes of non-sustained ventricular tachycardia in their early post-operative period. Two of the 7 atrial arrhythmias were paroxysmal, the remaining 5 were persistent and required treatment. Univariate analysis showed previous surgical palliation (p = 0.028) and a higher Gp:Qs ratio (p = 0.045) were significantly associated with early postoperative atrial arrhythmia. One patient (4%) died during the hospitalization. On the basis of available survival data in 21 patients, the probability of survival at 5, 10, 15 and 20 years post-operatively were 95%, 95%, 76%, and 76%, respectively.

CONCLUSIONS: Successful late surgical repair of TOF in these patients can be achieved with low surgical mortality and favorable long term survival in patients with experience in dealing with adult congenital heart disease. Atrial arrhythmias and conduction disturbance are the most common source of early postoperative morbidity in this group of patients.
Doppler echocardiography provides observations that could be of assistance during AV defect repair.

**POSTER SESSION**

**1094 Pediatric Cardiology I**

Monday, March 31, 2003, 9:00 a.m.-11:00 a.m.
McCormick Place, Hall A
Presentation Hour: 9:00 a.m.-10:00 a.m.

**1094-155 Acute Hemodynamic Effects of New Inotropic Agent(Coforsin Darapate) in Pediatric Patients With Left to Right Shunt Disease**

Motomi Iemura, Teiji Akagi, Yasashi Mizumoto, Yoko Sugahara, Wakako Himano, Yasuji Komine, Masahito Ishii, Toyoyuki Matsunaga, Kiyohide Urayama, Kurume University, Kurume, Japan

**Background:** Coforsin darapate(COF) is a new positive inotropic agent which having vasodilative effects. These effects are mediated by an increase in intracellular c-AMP concentration caused by the stimulating action of COF on adenylate cyclase, and not through beta-adrenoreceptor. Such characteristics may have advantage in treatment of left to right(LR) shunt disease.

**Objective:** To evaluate the hemodynamic effects of COF, 15 pediatric patients(4.8±2.5 yrs) with LR shunt disease(VSD:ASD=7:8) and 10 controls underwent COF(0.5ug/kg/min) infusion, challenge examination during catheterization, and compared to dopamine(DOA) infusion(1.5ug/kg/min)

**Result:** In the control group, COF significantly increased of cardiac index (2.79±0.54 vs 2.90±0.54) as same efficacy as DOA(10ug). Hemodynamic effects in LR shunt group was shown in table. No significant hemodynamic responses were observed between ASD and VSD patients in our study protocol.

**Conclusion:** These data suggested that COF had similar positive inotropic effect as DOA(10ug) without increasing of shunt ratio in patients with LR shunt disease. Such clinical features may play an important role for treatment of congestive heart failure in this setting.

Hemodynamic results

<table>
<thead>
<tr>
<th></th>
<th>Control</th>
<th>DOA 5u</th>
<th>DOA 10u</th>
<th>Cof 0.5u</th>
</tr>
</thead>
<tbody>
<tr>
<td>HR</td>
<td>100±6.4</td>
<td>102.2±5.9</td>
<td>110.4±7.8</td>
<td>110.7±7.1</td>
</tr>
<tr>
<td>PA(m)</td>
<td>35.7±0.4</td>
<td>40.2±0.5</td>
<td>44.2±0.6</td>
<td>2.7±0.3</td>
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<tr>
<td>ccwp</td>
<td>82.9±0.9</td>
<td>85.2±0.9</td>
<td>7.9±0.9</td>
<td>6.3±0.8</td>
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<tr>
<td>PCWP</td>
<td>22.4±2.7</td>
<td>25.2±2.9</td>
<td>30.7±2.5</td>
<td>22.8±4.1</td>
</tr>
<tr>
<td>Ao(m)</td>
<td>73.5±2.0</td>
<td>73.9±2.3</td>
<td>64.9±2.7</td>
<td>78.2±3.1</td>
</tr>
<tr>
<td>Qp/Qs</td>
<td>2.35±0.26</td>
<td>2.39±0.25</td>
<td>3.68±0.80</td>
<td>2.04±0.18</td>
</tr>
<tr>
<td>RvRS</td>
<td>0.13±0.05</td>
<td>0.11±0.02</td>
<td>0.12±0.02</td>
<td>0.11±0.02</td>
</tr>
</tbody>
</table>

*p < 0.05 versus control

**1094-156 Myocardial Acceleration During Isovolumic Contraction as a New Index of Right Ventricular Contractile Function and its Relation to Pulmonary Regurgitation in Patients After Repair of Tetralogy of Fallot**

Manatomo Toyono, Kenji Harada, Masahiro Tamura, Fumio Yamamoto, Akita University School of Medicine, Akita, Japan

**Background:** Myocardial acceleration during isovolumic contraction (IVA) calculated from tissue Doppler imaging (TDI) is a new noninvasive index of right ventricular (RV) contractile function that is unaffected by the shape of the ventricle and loading conditions. In this study, we assessed the utility of IVA to measure RV contractile function in patients after repair of tetralogy of Fallot (TOF).

**Methods:** We examined 18 TOF patients (6.2±2.4 yrs) with significant PR but no significant RV outflow obstruction (gradients <20mmHg) and 27 age-matched healthy children. Using TDI, Peak myocardial velocities during isovolumic contraction (IVV) and systolic ejection (S) were measured at the base of RV free wall from an apical four-chamber view. IVA was calculated by dividing IVV by the time interval from onset of the wave during isovolumic contraction to the time at peak velocity of this wave. The grading of the degree of PR (mild, moderate, and severe) was based on color Doppler findings.

**Results:** TDI measurements are shown in the table. S, IVV, and IVA in each TOF patient group were significantly lower than those in controls. However, S and IVA in the TOF patients remained constant regardless of severity of PR. There was a stepwise decrease in IVA from mild to severe PR. There was also a significant relationship between IVA and degree of PR(r=-0.82, p<0.0001).

Conclusion: IVA may be an effective means of assessing RV contractile function in the TOF patients with various degree of PR.

<table>
<thead>
<tr>
<th>Control</th>
<th>Mid (n = 5)</th>
<th>Moderate (n = 7)</th>
<th>Severe (n = 6)</th>
</tr>
</thead>
<tbody>
<tr>
<td>S (cm/sec)</td>
<td>11.7±2.7</td>
<td>7.0±0.7*</td>
<td>6.6±1.6</td>
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<tr>
<td>IVV (cm/sec)</td>
<td>8.1±1.6</td>
<td>6.1±1.0*</td>
<td>5.9±0.6*</td>
</tr>
<tr>
<td>IVA (cm/sec)</td>
<td>258±39</td>
<td>192±5</td>
<td>155±22*</td>
</tr>
</tbody>
</table>

*p < 0.01 vs control, tp < 0.01 vs mild PR, #p < 0.05 vs moderate PR

**1094-157 Right Ventricular to Coronary Artery Communications in Pulmonary Atresia: A New Anatomic-Functional Classification and its Utility in Predicting Immediate and Long-Term Outcome**


**Background:** The poor outcome in patients(pts) with pulmonary atresia(PAT) and right ventricle to coronary artery communications(RVCC) may be related to the fact that a significant portion of the left ventricular myocardium is dependant upon perfusion from these communications. Current definition of RVCC is qualitative and subjective. Therefore, we propose a new classification scheme for RVCC and test its utility in predicting the outcome.

**Methods:** Charts of pts with PAT and RVCC between 1974-2002 were reviewed. The demographic, hemodynamic, angiographic, number and type of operations, immediate outcome and the latest follow-up data were collected. The RV Aortic angiograms were analyzed for RVCC patterns and were classified as types 0, 1A, 1B, 2, and 3. Type 1A/fant/rare filling of the secondary branches with no filling of the main coronary arteries. Type 1B/retrograde filling of one of more main coronary arteries with out any luminal stenosis,irregularities, and with no retrograde filling of the ascending aorta(AA). Type 2:1A type 1 with luminal dilatation and irregularities, with retrograde filling of the AA.type 3: type 2 with stenosis in one or more main coronary arteries.type 4: Type 2 or 3 with interruptions in one or more main coronary arteries. Results: Of 28 pts with PAT and RVCC, 26 had angiograms for analysis. We found RVCC type 0 in 3 pts,type 1 in 2 type 2 in 7 type 3 in 7, and type 4 in 5 pts. Two pts died before any surgical intervention and 26 pts had a shunt procedure. There were 2 post-operative deaths and 9 late deaths. The remaining 16 pts are alive and had further operations(biventricular repair in 11, 15 ventricular repair in 1,Fontan in 10, Glenn in 1 and shunt in 2 pts). Actual survival including in hospital and late deaths was 66%,40%, and 25% at 5,10, and 20 years respectively. Mortality was associated with RVCC types 3 and 4(p<0.05).

**Conclusions:** The long-term outcome of pts with PAT and RVCC is poor with a significant correlation between the types of RVCC and mortality. Left ventricular perfusion imaging of pts with RVCC types 3 and 4 may help develop specific treatment strategies and better risk stratification.

**1094-158 Occult Myocardial Injury in Otherwise Healthy Newborns**

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**Hypothesis:** Since the perinatal period is a time of the greatest pediatric myocardial stress, we proposed that the neonatal myocardium will sustain a greater injury from a similar insult than an older myocardium, demonstrating clinically occult myocardial injury.

**Methods:** Prospective study to identify whether otherwise healthy newborns without known cardiac disease had evidence of myocardial injury. The Electrolyte Tropinone STAT Immunoassay identified cTnT values of >0.01 ng/ml as elevated. C-reactive protein (hsCRP) was measured by N High Sensitivity CRP assay, and CK-MB was assayed by ACS:180 CK-MB assay (>5.5 ng/ml elevated).

**Results:** Consent was obtained on 27 neonates (median bw 3.3 kg; median gestational age 39.7 weeks, 95% AOA, median LOO 5 days). Median umbilical cord cTnT was 0.006 mg/ml (0.005-0.020 mg/ml); median neonatal serum cTnT was 0.068 ng/ml (0.005-0.437 ng/ml). Serum cTnT was elevated in 76% of patients (Pis) with umbilical cord samples with concentrations above these cut-offs, occult myocardial injury was excluded in the elevated cTnT were also had cTnT elevations. Myocardial injury was more common in otherwise healthy neonates than in older such children (6%), and was associated with lower 1 minute Apgar scores (r=-0.85, p<0.001), a younger gestational age (r=-0.53, p<0.04), increased IUGR (p=0.52, p=0.04), non-white race (r=0.0085), and elevated hsCRP (r=0.79, p=0.0008), a marker of generalized inflammation. Elevated hsCRP correlated

**JACC**

March 19, 2003