

CPB time, anatomic group, QRS duration, change in QRS duration, or baseline CI.

**Conclusions:** Multisite ventricular pacing results in improved CI and BP following surgery for CHD. In some pts, significant hemodynamic improvement with pacing facilitated weaning from CPB. Multisite pacing should be considered for adjunctive therapy of post-operative cardiac dysfunction.

Noon

815-5

### Prospective Assessment After Pediatric Cardiac Ablation: Initial Results of the Multicenter Study

George F. Van Hare, Dorit Carmelli, J. Philip Saul, Ronn E. Tanel, Peter S. Fischbach, Ronald J. Kanter, Michael S. Schaffer, Stanford University, Palo Alto, CA

In 1998, a prospective multicenter study of children undergoing radiofrequency ablation (RFA) of either an accessory pathway or atrioventricular node reentry (AVNRT) was begun. 461 patients aged 0-16 yrs were enrolled by 23 US centers, and followed for 2 years. During the study period, 1832 pts undergoing RFA were reported by 31 centers to a separate registry. Data are available at 2 months post ablation with 98% follow-up (f/u) for the enrolled group. Results with respect to success and recurrence are summarized in the table below.

The overall initial success rate was 95%. Complications due to electrophysiology study occurred in 4% and due to ablation in 4% of patients, with no deaths reported. Recurrence occurred in 8% of patients at 2 months. The most important predictor of recurrence was pathway site ( $p < .0001$ ), with right freewall and septal locations being most likely to recur. In addition, the use of power rather than temperature mode was associated with recurrence ( $p = .0077$ , Chi-square), but age and weight were not.

**Conclusions:** In this prospectively enrolled cohort of pediatric patients with nearly complete follow-up at 2 months, initial success rates for ablation in children are similar to those reported previously by both adult and pediatric single centers. There is a relatively high incidence of recurrence by 2 months, seen primarily in patients with right-sided accessory pathways. With 98% f/u, recurrence rates are likely to be more reliable and less biased than those from prior retrospective studies.

Success and recurrence rates

| Substrate/pathway location | Total patients | Successes | Success rate (%) | Recurrence rate (%) |
|----------------------------|----------------|-----------|------------------|---------------------|
| Right freewall             | 53             | 49        | 92%              | 17%                 |
| Right septal               | 69             | 63        | 91%              | 15%                 |
| Left septal                | 20             | 19        | 95%              | 5%                  |
| Left freewall              | 209            | 206       | 99%              | 6%                  |
| AVNRT                      | 128            | 118       | 92%              | 3%                  |
| Mahaim                     | 2              | 2         | 100%             | 50%                 |
| Overall                    | 481            | 457       | 95%              | 8%                  |

## POSTER SESSION

### 1119 Fetal Cardiology

Monday, March 31, 2003, Noon-2:00 p.m.

McCormick Place, Hall A

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

1119-155

### The Fetus With Hypoplastic Left Heart Syndrome: Risk Factors and Outcomes

Cyrus Samai, Carlen A. Gomez, Mark W. Russell, David A. Parra, Achi Ludomirsky, University of Michigan, Ann Arbor, MI

Prenatal diagnosis of hypoplastic left heart syndrome (HLHS) has increased significantly over the past decade. At our institution, approximately 50% of infants presenting with HLHS are diagnosed *in utero*. Fetal diagnosis serves to allow parental planning and implementation of a multidisciplinary approach to manage these high-risk patients. However, the effect of fetal diagnosis on patient outcome has been debatable. The purpose of this study is to assess outcomes and to identify the incidence of risk factors associated with survival in fetuses with HLHS.

**Methods:** A retrospective review was performed of the prenatal and postnatal medical records for all patients with a diagnosis of HLHS followed in our fetal cardiology program from June 1998 through December 2001. Outcomes were assessed, and an analysis of risk factors was performed. Risk factors included obstructed pulmonary venous return, right ventricular dysfunction, other associated cardiac defects, low birth weight/prematurity, and non-cardiovascular anomalies.

**Results:** The patient population consisted of 76 fetuses with a diagnosis of HLHS. The overall survival to hospital discharge was 57% (43/76). Of the 33 patients that died, 39% (13/33) expired prior to surgical intervention (including 4 with *in utero* demise and one elective termination). Sixty-three infants underwent surgical palliation (2 with a Ross-Konno operation, and 61 with a Norwood procedure). Survival to hospital discharge in those palliated was 68% (43/63).

Fifty percent of all fetuses (38/76) had at least one risk factor; however, only 16 of these 38 fetuses were identified as high risk prenatally. Presence of 1 risk factor resulted in 56% mortality, and the presence of 2 or more risk factors resulted in 67% mortality.

Patients with no risk factors undergoing surgical palliation had 80% survival.

**Conclusion:** Fetuses with a prenatal diagnosis of HLHS have a high incidence of risk factors. Many of the additional abnormalities can be recognized *in utero* and help to determine prognosis. Nonetheless, prenatal counseling should address the potential for risk factors that may not be detected before birth.

1119-156

### Diagnosis and Outcome of Dextrocardia in the Fetus

Rebecca Walmsley, Takashi Hishitani, George G. Sandor, Kenneth Lim, Walter J. Duncan, Francine Tessier, Duncan F. Farquharson, James E. Potts, Children's and Women's Health Centre of B.C., Vancouver, BC, Canada

**Background:** The incidence of cardiac malformations and the accuracy of fetal echocardiography in the diagnosis of dextrocardia are not known.

**Methods:** We retrospectively reviewed 5,539 fetal echocardiograms (FE) performed at our institution between 1979 and 2001 to identify all cases of fetal dextrocardia. Prenatal and postnatal diagnoses were compared and the outcomes reviewed. **Results:** Dextrocardia was defined as the heart's position being in the right hemithorax. 82 cases were identified by FE. Of these, 30 were referred for FE following diagnosis of dextrocardia on general ultrasound, while 52 were referred for other reasons. 44 were classified as "primary" and 38 as "secondary". Among primary cases, 9 had situs solitus (all with complex cardiac malformations); 18 had situs inversus (16 with no cardiac anomaly); and 17 had situs ambiguus/isomerism (all with complex cardiac malformations). Causes of secondary dextrocardia were diaphragmatic hernia (31% had cardiac malformations), cystadenomatoid malformation, and pleural effusion. The FE diagnosis of major cardiac malformations was correct in 87% of cases. In 7% of those incompletely or inaccurately diagnosed, the outcome would have been affected. The pregnancy was terminated in 19 cases, 6 of these had no cardiac defects; 6 cases aborted spontaneously; 11 cases were offered compassionate care post-natally or died perinatally; 14 operative procedures were performed (1 Rastelli repair, 1 coarctation repair with pulmonary artery banding, 5 completed Fontans, 1 VSD closure, 1 BT shunt alone (died post-operatively), 2 pacemaker insertions and 2 ligation of PDA, 1 case had surgery for AV discordance). Between 1990-96 the incidence of congenital heart disease, primary-, and secondary dextrocardia was 0.92%, 0.064%, and 0.034%, respectively (calculated using 26,725 scans over the same period). **Conclusions:** The incidence of each type of dextrocardia differs from postnatal series. Complex cardiac malformations were found to frequently co-exist. The rates of termination, spontaneous abortion and postnatal death are high in this patient group. Fetal echocardiography is accurate and counselling of parents should be specific.

1119-157

### Prenatal Growth of Tricuspid Valve and Right Ventricle Determines the Evolution and Postnatal Outcomes of Tricuspid Atresia

Gautam K. Singh, Ian C. Balfour, Su-chiung Chen, Barbara Ferdman, Saadeh Jureidini, Andrew C. Fiore, P. S. Rao, Saint Louis University School of Medicine, St. Louis, MO, University of Texas-Houston Medical School, Houston, TX

**Background:** Tricuspid atresia (TA) with normally related great arteries is postulated to develop from mal-alignment between atrial and ventricular loop (Van Praagh). Based on observations we hypothesized that TA may evolve prenatally from tricuspid stenosis (TS) without mal-alignment and sought to identify the prenatal morphological determinants of postnatal outcomes. **Methods:** Ten fetuses diagnosed with TS without atrio-ventricular mal-alignment at 21.3  $\pm$  2.7 wks were prospectively studied up to infancy by serial echocardiography to develop growth curves of cardiac segments and compare them with data from 51 normal fetuses and infants. **Results:** TS fetuses had z-scores of  $-1.5 \pm 0.1$  for tricuspid valve (TV),  $-1.6 \pm 0.2$  for right ventricle end diastolic dimensions (RVEDD) and  $-1.4 \pm 0.2$  for right ventricle length/heart length (RVL/HL) at diagnosis. Right heart structures' growth rates were reduced ( $p < .01$ ). Six TS fetuses with intact septum and TV z-score  $< -1.6$  developed TA, right ventricular tripartite hypoplasia (RVEDD, RVL/HL and outflow z-score  $< -2$ ), reverse ductal flow, and pulmonary atresia by 31 wks. Four TS fetuses with ventricular septal defects and TV z-score  $< -1.5$  also developed TA and right ventricle inlet and sinus hypoplasia but with normal outflow tract, pulmonary and ductal flow by 32 wks. Ventricular septal defects in 2 TS fetuses later became restrictive resulting in reverse ductal flow and small pulmonary arteries. Z-scores of TV and RVEDD at diagnosis and their growth rates correlated strongly ( $r > 0.8$ ) with postnatal RVEDD. Postnatally, 2 TA infants with unrestrictive ventricular septal defect had duct-independent whereas other 8 had duct-dependent pulmonary circulation, the latter requiring palliative shunt before staged cavo-pulmonary connection. **Conclusions:** A subset of TA may evolve from TS without prerequisite atrio-ventricular mal-alignment. Fetuses with TV and RVEDD z-score  $< -1.5$  in 2nd trimester are likely to present postnatally as TA with RV hypoplasia, which are important determinants of clinical outcome. Development of reverse ductal flow and restrictive ventricular septal defect in fetal TS predicts duct-dependent postnatal pulmonary circulation.

1119-162

### Fetal Rhabdomyoma: Prenatal Diagnosis, Clinical Outcome, and Incidence of Associated Tuberos Sclerosis Complex

Rima S. Bader, David Chitayat, Edmond Kelly, Greg Ryan, Jeffrey Smallhorn, Lisa K. Hornberger, The Hospital for Sick Children, Toronto, ON, Canada, Mount Sinai Hospital, Toronto, ON, Canada

**Objectives:** We reviewed our institution's experience with fetal cardiac rhabdomyoma (CR) to document the clinical outcome and the incidence of tuberous sclerosis complex (TSC) associated with this condition, information which is critical for accurate prenatal counseling. We also compared our findings to patients diagnosed with CR after birth over the same period of time. **Methods:** Medical records of all patients diagnosed prenatally or postnatally from January 1990 to June 2002 were reviewed.