used to supplement the information available from the medical records.

**Results:** 3 patients were pregnant (for the first time) at enrollment, 54 patients were childless 14 pregnancies, which resulted in live birth, were observed in the remaining 22 patients. In addition, 2 abortions (1 elective) were reported in the childless group and 10 (0 elective) in the patients with children. Mean age of the childless patients was 26 (SD 5.74), in patients with children 34 (SD 5.78) years. 5 patients reported fertility problems and only 1 patient was advised against pregnancy by her cardiologist. Maternal complications were: arthralgia (n=6) and heart failure (n=2), in 5 patients and in 6 pregnancies. Caesarian section was performed 13 times (28%), of which 5 on maternal cardiac indication. Mean birth weight was 3064 g (SD 728). 5 births were premature (between 16 and 37 weeks) and 6 children (17%) were small for gestational age (<p10). 3 children died within the first year of life. 2 children (4.3%) had congenital heart disease: 1 child hadToF (died at 10 months). the other had several cardiac malformations in combination with trisome 13.

**Conclusion:** In patients withToF pregnancy is generally well tolerated. However, mater- nal cardiac complications do occur in 13% of pregnancies. The need for Caesarian sec- tion was increased. The incidence of small for gestational age children was increased. Fertility seems not to be compromised.

**POSTER SESSION**

**1114 Pediatric and Congenital Heart Disease**

Monday, March 08, 2004, 3:00 p.m.-5:00 p.m.
Morial Convention Center, Hall G
Presentation Hour: 3:00 p.m.-4:00 p.m.

**1114-199 Genomic Profiles of Left Ventricular and Right Ventricular Hypertrophy in Congenital Heart Disease**

**Background:** Hemodynamic stimuli promote activation of gene expression pathways that mediate ventricular hypertrophy. The LV has a greater ability to tolerate hemody- namic load than the RV. Regulation of this differential response is not known.

**Methods:** Hypertrophied myocardium was obtained at surgery from 9 acyanotic pediatric pts. 4 LVH and 5 RVH (age 0.15-6.7 yrs). Diagnoses included subaortic stenosis, subpul- monary stenosis, and hypertrophic cardiomyopathy. Gene expression (GE) with Affyme- trix DNA microarray gene chips was performed. After log transformation, differences in mean GE between LVH and RVH groups with p value <0.01 was considered significant. Genes with > 2-fold difference were characterized and correlated with severity of obstruction. Immunohistochemical staining was performed to detect myo- cyte apoptosis with TUNEL assay, and fibrosis with trichrome stain in myocardium from 10pts with LVH and/or RVH.

**Results:** There were 253 genes with significant differences in expression between LVH and RVH myocardium samples. 124 genes associated with myocardiop hypertrophy, cytoskeleton, apoptosis, and ion channels were upregulated in LVH. RVH group had 129 genes relatively overexpressed: extracellular matrix components, phospholipase A2 and C, muscle mitochondrial transporters. Fetal gene activation was present in both LVH and RVH, with relative overexpression of ANF and dystrophin in the LV compared to RV (p values <.01). Seventy of obstructive gradients in both groups correlated positively with actin and myosin GE, and negatively with apoptosis related genes (r = 0.8-0.9). ANP GE correlated with LVH and RVH obstruction only. Fibrosis and myocyte apoptosis were present in both LVH and RVH specimens, but was greater in pts with HCM than those with isolated out- flow tract lesions.

**Conclusion:** Myocardium from hypertrophied LV and RV has different genomic profiles. Upregulation of genes in the LVH group that activate adaptive pathways may contribute to the functional advantage observed in the LV vs. the RV with hemodynamic load, espe- cially in pts with congenital heart disease. Further analysis of these candidate genes and downstream signaling effects are warranted.

**1114-200 Increased Incidence of Pulmonary Vein Stenosis in Patients With Atrioventricular Canal: A Multi-Institutional Study**
Henry W. Kogt, Christine Hille, Virgil Larson, Mark C. Johnson, St. Louis Children's Hospital, St. Louis, MO, University of Minnesota, Minneapolis, MN

**Background:** Patients with atrioventricular canal (AVC) appear to be at greater risk for the development of elevated pulmonary vascular resistance (PVR) than patients with other cardiac defects. Review of our experience at a single institution suggested the inci- dence of pulmonary vein stenosis (PVS) may be greater in a subgroup of patients with AVC than in patients with other lesions. We set out to determine if this pattern held true in a larger patient population.

**Methods:** The Pediatric Cardiac Care Consortium data base from 1981 to 2001 (n=90,887) was searched for all patients with a diagnosis of pulmonary vein obstruction (PVO, AVC, or triscusis 13). Patients with AVC were further divided into those with only pulmonary vein obstruction, 13.7% (70 of 509, p<0.0001) and PVS, 12.5% (35 of 280, p=0.0028) as compared with an 8.4% prevalence of AVC in patients without pulmonary vein obstruction (6,661 of 79,656, excluding anomalous pulmonary venous connections). There was no association of Trisomy 21 with pulmonary vein obstruction (p=0.4).

**Conclusion:** Patients with AVC are at greater risk for concomitant pulmonary vein obstruction and PVS than are patients with other congenital cardiac defects. Patients with AVC and evidence of elevated PVR should undergo careful scrutiny of pulmonary venous connections either by echocardiography or cardiac catheterization. Trisomy 21 does not appear to be associated with the increased incidence of pulmonary vein obstruction.

**1114-201 Higher Mortality After Fontan Surgery in Trisomy 21 Syndrome Children**
Mensha Gupta-Mahato, Virgil E. V. Larson, Ronald M. Rosengart, James H. Moller, University of Minnesota, Minneapolis, MN

**Background:** High pulmonary artery pressure (PAP) is associated with increased mor- tality after Fontan surgery. It is not known whether Trisomy 21 syndrome patients have a higher PAP prior to Fontan when compared to the non-Trisomy 21 patients.

**Methods:** We retrospectively evaluated data from our Pediatric Cardiac Care Consor- tium (a data bank for all cardiac surgery cases from participating centers). Of the total Fontan surgeries (n = 1966), 17 were performed on Trisomy 21 patients (Group II). Group I were non-Trisomy 21 patients after Fontan (n = 1949). Of Group II patients, 13 patients (Group III) had had Fontan for unbalanced atrioventricular canal defects. Since it would be difficult to go through charts of such a large population, we selected a control group from the Group I a total of 28 patients (Group IV) with atrioventricular canal defect who were age (at Fontan) and sex matched and who were alive after the surgery. We evaluated the pre-Fontan mean PAP in mmHg, pulmonary vascular resistance in wood units (PVR), length of stay in days at hospital after surgery (LOS), weight in Kg at surgery (WT), Hemoglobin prior to surgery in g/dl (Hb), whether there was prior Glenn surgery or not, whether the Fontan was fenestrated (FEN) or not and whether there was preoperative atrioventricular regurgitation (AVVR) or not.

**Results:** Between Groups I and II there was significantly higher mortality (p = 0.0011) in Group II (n = 617, mean age at surgery 4.8 years) as compared to Group I (n = 204/ 1949, mean age at surgery 6.6 years) without significant difference between the age, Hb, WT and LOS. Between Group III (WT 16, Glenn 85%, AVVR 62%, FEN 54%, PAP 13.8, PVR 2.9, Hb 15.8, LOS 21) and Group IV (WT 22, Glenn 61%, AVVR 57%, FEN 29%, PAP 12.9, PVR 2.9, Hb 15.7, LOS 14) there were no significant differences in the parameters evaluated. Hence, in the selected population of Trisomy 21 who underwent Fontan surgery, PAP and PVR were similar to the non-Trisomy 21 children.

**Conclusion:** The PAP is similar between Trisomy and non-Trisomy patients prior to Fon- tan. Trisomy 21 syndrome is an independent parameter associated with significantly higher risk of mortality (35%) after Fontan surgery as compared to non-Trisomy 21 patients (10%).