scheme is proposed to decrease persistent pleural effusions after the Fontan operation.}

Methods: The Gamma regimen consisted of aggressive diuresis utilizing IV furosemide and enteral lactulose, 80% fluid restriction, afterload reduction, supplemental oxygen, and low fat diet. A group of patients with underwent uncomplicated fenestrated Fontan prior to August 2000 (n=20) was used as a control (Group 2). Patient demographics between groups were similar except for surgical approach. Extracardiac Fontana were performed in 90% and 45% of Groups 1 and 2 respectively. Previously our group has shown no statistically significant difference in pleural effusions between extracardiac and intracardiac approaches. One-way ANOVA was done to evaluate length of hospital stay and duration of pleural chest tube placement. Fisher's exact 2-sided test was done for the use of an NPO/TPN strategy and the use of pleural sclerosis.

Results: The standardized medical regimen was well tolerated. The mean length of stay was 8 vs. 16 days for Groups 1 and 2 respectively, p-value <0.01. Pleural chest tubes remained in place for 6 vs. 11 days for Groups 1 and 2 respectively, p-value <0.05. Five patients in Group 2 were made NPO and placed on TPN, four had pleurodesis. No further treatment was necessary for any patient in Group 1.

Conclusions: Initial results suggest that the standardized medical regimen shortened hospitalization and decreased morbidity following the Fontan operation.

Aortic Regurgitation in an Outlet Ventricular Septal Defect Complicated by Right Coronary Cusp Prolapse: Predictors of Prognosis

Hidashi Tomita, Yoshihiko Aoki, Kan-chi Kuroaki, Toshikazu Yaghari, Shigeoyuki Echigo, National Cardiovascular Center, Osaka, Japan, Kurashiki Central Hospital, Kurashiki, Japan.

Background: Aortic regurgitation (AR) is a common complication for outlet ventricular septal defect (o-VSD) complicated by right coronary cusp prolapse (RCCP). However AR usually progresses slowly or remains mild for a long time after the right coronary cusp first prolapses.

Methods: Among 620 patients with o-VSD who underwent the Doppler echocardiography (echo) from January 1985 to May 2001, we reviewed the echo of the aortic valve and associated data in 217 pts. The mean age of patients was 25 ±19 years, that was earlier than in group 3 where AR was first diagnosed at 39±17 years (p<0.01). The aortic valve ring deformed index [ARCDI=(the length of the deformed right coronary cusp/the diameter of the aortic valve), the right coronary cusp imbalance index [R/L=(width of right coronary cusp/width of left coronary cusp)], the right coronary cusp deformity index [ARCDI] the length of the deformed right coronary cusp/lethaperimeter of the right coronary cusp (ARCDI)], the right coronary cusp imbalance index [R/L=(width of right coronary cusp/width of left coronary cusp)], the intercostal level index [IL=(height of right coronary cusp/height of left coronary cusp)] were compared among the 4 groups.

Results: The type of o-VSD (subarterial vs. outlet muscular) in groups 1-4 was 15 vs. 11, 29 vs. 33, 51 vs. 11, and 1 vs. 1 (p=0.01). RCCP was firstly detected at significantly younger age in group 4 (25±15 years) than in group 2 (50±20) and 3 (41±54, p=0.01). In group 4, AR was first diagnosed at 17±19 years, that was earlier than in group 3 where AR was first detected at 28±17 years (p=0.01). Both RCCD (0.4±0.2 vs.0.9) and R/L (0.3±0.09 vs.0.5) were larger than those in groups 2 (0.3±0.01, 0.13±0.05) and 3 (0.0±0.09, 0.13±0.07, p=0.01).

Conclusion: The Overdiagnosis of Marfan Syndrome: Results From a Large Pediatric and Adult Cardiovascular Connective Tissue Disorders Clinic

Elizabeth A. Shank, Curt J. Daniels, Ohio State University, Columbus, Ohio, Children's Hospital, Columbus, Ohio.

Background: Marfan syndrome (MFS) is a connective tissue disorder with significant cardiovascular risk for aortic dilation, aneurysm and dissection. Making the correct diagnosis (dx) of MFS carries significant cardiovascular advantages by decreasing morbidity and mortality. Assigning dx in patients (pts) who do not meet diagnostic criteria (over-dx), has significant psychological effects, leads to unnecessary cardiovascular follow-up and testing, influences insurability, and significantly alters lifestyle. This study was performed to evaluate whether pts previously diagnosed with MFS were accurately diagnosed and to evaluate the factors that lead to over-dx.

Methods: Over the past 12 months, 213 pts (25 male, 86 female) referred to our cardiovascular connective tissue disorders clinic were prospectively evaluated for MFS; age ranged 1 to 62 years. Detailed family history, and clinical criteria-anthropometric measurements, dilated eye exams, radiologic studies, and aortic root measurements (echocardiography) using normal values standardized for aortic body surface area, were used to support the dx of MFS. Results: 44 pts, age range 1 to 42 years (20 male; 14 female) met diagnostic criteria for MFS. Previous MFS dx was confirmed in 24 pts; 19 pts were newly diagnosed; and 1 pt previously without dx was made. RCCP was firstly detected at significantly younger age (25 vs. 19, 25 vs. 17, and 25 vs. 18, p<0.01).

Conclusions: Failure to correctly assign MFS dx as well as over-dx of MFS can lead to significant life-long detrimental consequences. In our study, 35 % of patients with previous dx of MFS did not meet criteria and were over-dx. The over-dx of MFS can lead to significant life-long detrimental consequences. In our study, 35 % of patients with previous dx of MFS did not meet criteria and were over-dx. The over-dx of MFS can lead to significant life-long detrimental consequences. In our study, 35 % of patients with previous dx of MFS did not meet criteria and were over-dx. The over-dx of MFS can lead to significant life-long detrimental consequences. In our study, 35 % of patients with previous dx of MFS did not meet criteria and were over-dx. The over-dx of MFS can lead to significant life-long detrimental consequences. In our study, 35 % of patients with previous dx of MFS did not meet criteria and were over-dx. The over-dx of MFS can lead to significant life-long detrimental consequences.