1023-101 **Predictive Ability of Color M-Mode and Doppler Tissue Imaging Compared to Traditional Measures of Diastolic Dysfunction in Children**

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Background: There is no reliable echocardiographic method of evaluating diastolic function in children. Two new modalities, Color M-mode (CMM) and Doppler tissue imaging (DTI), are used in adults. We evaluated their ability to identify patients with abnormal cardiac function and compared them to traditional methods.
Methods: Measurements were made in two groups: 1) Normal - referred to rule out disease with a normal echocardiogram [n = 27 (14 male), mean age = 8.1yr (range: 0.05 - 17)], and 2) Cardiomyopathy - diagnosis of dilated, hypertrophic or restrictive cardiomyopathy [n = 22 (14 male), mean age = 13.9 yr (range: 1.6 - 19.6)]. Echocardiographic measurements included: mitral E and A waves, deceleration time, pulmonary vein S, D, and A reversal waves, isovolumic relaxation time, CMM derived propagation velocity (Vp) of the mitral inflow, and DTI derived early diastolic velocity (Em) of the four margins of the mitral annulus. Receiver operating characteristic analysis was performed on all measurements.
Results:

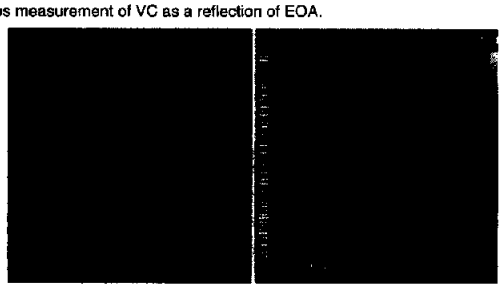
Measure	Area under curve	Standard error
E wave	0.685	0.076
A wave	0.591	0.082
Deceleration time	0.328	0.079
S wave	0.476	0.084
D wave	0.658	0.079
A reversal wave	0.307	0.079
IVRT	0.507	0.090
Vp	0.826	0.061
Em septal	0.924	0.039
Em anterior	0.895	0.046
Em lateral	0.891	0.047
Em posterior	0.878	0.050

Conclusions: All Em measurements and Vp have very strong predictive abilities. Other measures have mild-to-no predictive ability to separate healthy from abnormal patients. Both CMM and DTI are promising adjuncts to traditional echocardiographic methods of evaluating diastolic function. Future studies should include establishment of age-adjusted normal data and correlation with clinical and hemodynamic measure of cardiac dysfunction.

1023-102 **Digital 3-D Velocity Reconstructions Allow Extraction of the Vena Contracta (VC) Area for Flow Rate Computation: An In Vitro Study Using Raw Scanline Transfer and Reconstruction of 3-D Data**

Yoshiki Mori, Paul R. Detmer, Xiang-Ning Li, Roy W. Martin, Antoinette Kenny, Susan L. Martin, David J. Sahn, *Oregon Health & Science University, Portland, Oregon, ATL Ultrasound, Bothell, Washington.*

Methods: Our study imaged proximal jet flows from 3 orifices (rectangular, circular and triangular, each 0.24 cm²) with an ATL 3000 using a 5MHz multiplane probe with stepper function controlled by the scanner and a connection allowing transfer of raw scanline digital velocity data to an SGI workstation. By interrogating jet flow in the in vitro model (stroke volumes 10-60 cc/beat measured by an ultrasonic flowmeter) parallel to the direction of propagation and analyzing it in a cross-section selected for smallest diameter and flattest velocity profile, a high velocity core in the VC region could be identified. **Results:** After rotational acquisition at 5° spacing, 3D digital color VC regions extracted and measured as cross-sectional areas matched the fluid dynamically determined effective orifice area (EOA) for the modeled valve defects (p = NS, mean difference + 0.06 cm²). Also, when VC area was integrated with the CW Doppler velocity time integral, a high correlation with both actual peak instantaneous flow rate 3.7-11.4 L/min (r = 0.98)(slight overestimation) and stroke volume/beat (r = 0.98) was obtained. VC areas had extreme similarity to the orifices' shapes. **Conclusions:** The ability to analyze cross-sectional digital velocity data from proximal jet flow fields from 3D datasets obtained parallel to flow enhances measurement of VC as a reflection of EOA.



POSTER SESSION

1047 Diagnosis and Management of Congenital Heart Disease

Sunday, March 17, 2002, Noon-2:00 p.m.
Georgia World Congress Center, Hall G
Presentation Hour: 1:00 p.m.-2:00 p.m.

1047-97 **Effect of Vasodilator Therapy on Left Atrioventricular Valve Regurgitation After Atrioventricular Septal Defect Repair**

Reenu S. Eapen, Claudio Ramaciotti, Matthew S. Lemler, Daniel Stromberg, *University of Texas, Southwestern Medical Center, Dallas, Texas.*

Background: Postoperative left atrioventricular valve (LAVV) regurgitation after atrioventricular septal defect (AVSD) repair is a significant risk factor for morbidity and mortality. We performed a prospective study to determine whether vasodilator therapy with Nitroprusside changes the severity of LAVV regurgitation after AVSD repair.
Methods: Postoperative patients received a Nitroprusside infusion at 0.5mcg/kg/min, titrated to a maximum dose of 2mcg/kg/min in an attempt to decrease systolic blood pressure by 15%. The vena contracta of the LAVV regurgitant jet, and regurgitant fraction across the LAVV were measured before and during Nitroprusside administration by transthoracic echocardiography. All measurements were made off line in blinded fashion. Cardiac output and systemic vascular resistance (SVR) were evaluated during the study periods using arterial and venous blood gases.
Results: Of the 17 patients studied, 6 patients achieved both a decrease in systolic blood pressure and a significant reduction of SVR (> 10%) during Nitroprusside infusion. These patients had a significant increase in their cardiac output (15.6%, p=0.002), decrease in the vena contracta of the regurgitant jet in the apical four chamber view (29.5%, p=0.03) and a decrease in the LAVV regurgitant fraction (35.9%, p=0.025). Those patients who did not demonstrate a decrease in SVR, whether or not accompanied by a decrease in blood pressure (via decreased stroke volume, n=5), did not experience a reduction in LAVV regurgitation.
Conclusions: 1. Vasodilator administration which diminishes SVR augments cardiac output and decreases LAVV regurgitation after AVSD repair. 2. Vasodilation that solely achieves a reduction in blood pressure without an accompanying decrease in SVR does not reduce LAVV regurgitation in postoperative AVSD patients.

1047-98 **Carvedilol for the Treatment of Congestive Heart Failure in Children With Cardiomyopathy**

Paolo Rusconi, Esmail Redha, Jennifer R. Marin, Maria Rossique-Gonzalez, Ming-Lon Young, Grace S. Wolff, *University of Miami School of Medicine, Miami, Florida, Jackson Memorial Hospital, Miami, Florida.*

Background: Carvedilol (CAR) has proven to be effective in the treatment of congestive heart failure (CHF) in adults. However there is paucity of data on the use of beta-blockers, their safety and efficacy in the treatment of CHF in children. Our goal is to assess safety and efficacy of CAR in the treatment of CHF in children.
Methods: Retrospective analysis of clinical data of 22 children with cardiomyopathy (CMP) and CHF who received CAR after conventional treatment with Digoxin, Angiotensin Converting Enzyme Inhibitor and diuretics. Change in clinical status before and after CAR treatment was assessed using New York Heart Association (NYHA) class. Echocardiogram was performed before starting CAR and at 3-6 months interval to monitor ejection fraction (EF) and sphericity index (ratio between left ventricle transverse diameter and left ventricle longitudinal diameter in diastole and multiplied by 100). CAR was started (mean initial dose=0.1 mg/Kg/day in two divided doses), at a median of 10.6 months after the diagnosis of CMP was made. CAR was increased weekly to an average maximum dose of 0.8 mg/Kg/day (0.25-1.2).
Results: There were 13 males, median age: 8.9 years (0.5-18). CMP etiology: idiopathic 11, myocarditis 5, muscular dystrophy 3, others 3. NYHA classification was ≥2 in all patients; 8 children were in class 4. All patients had EF ≤40%. In 2 patients CAR was stopped after 1 week because of worsening CHF in one and asthma in the other. Mean follow up was 19.5 months (2.7-37). Four patients died 1 awaiting transplant and 3 with muscular dystrophy. Three patients underwent heart transplant. One child developed first and Mobitz I second degree atrioventricular block. EF improved from 27±7% before CAR was started to 39±12% (mean improvement 12.5%, p< 0.001). The average sphericity index decreased from 85±9% to 79±11% (mean change 9.4%, p<0.005). NYHA class improved from 3.15 to 1.95 (p<0.001).
Conclusion: When used in combination with standard therapy CAR is well tolerated and seems to improve clinical status and cardiac function of children with CHF. Control studies with a larger number of patients are required to determine if CAR improves mortality and reduce hospitalization in this age group.

1047-99 **A Medical Strategy to Address Persistent Chest Tube Drainage After the Fontan Operation**

Joseph R. Cava, Raymond T. Fedderly, Sarah M. Bevandic, Michelle Steltzer, Patrick J. Phelan, Kan N. Hor, James S. Tweddell, S. Bert Litwin, *Medical College of Wisconsin, Milwaukee, Wisconsin, Children's Hospital of Wisconsin, Milwaukee, Wisconsin.*

Background: Surgical modifications have dramatically improved survival following the Fontan operation; however, morbidity related to persistent pleural effusion continues to be a significant problem. Total parenteral nutrition (TPN) along with eliminating feedings (NPO) and pleural sclerosis have been used to treat persistent drainage. Risk factors for persistent pleural drainage have been previously reported, although medical management in this population of patients varies widely. A standardized medical management

scheme is proposed to decrease persistent pleural effusions after the Fontan operation.
Methods: From August 2000 to July 2001, 20 patients who underwent an uncomplicated fenestrated Fontan procedure were placed on a standardized medical regimen (Group 1). The regimen consisted of aggressive diuresis utilizing IV furosemide and enteral aldactazide, 80% fluid restriction, afterload reduction, supplemental oxygen, and low fat diet. A group of patients who 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 Fontans 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 9 vs. 15 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. Neither 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.

1047-100 The Overdiagnosis of Marfan Syndrome: Results From a Large Pediatric and Adult Cardiovascular Connective Tissue Disorders Clinic

Elizabeth A. Sparks, 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 this 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 (127 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 age and body surface area, were used to support the dx of MFS. *Results:* 44 pts, age range 1 to 42 years (30 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 MFS dx had dural ectasia on MRI, fulfilling MFS dx criteria. 13/37 pts (35%) with previous MFS dx did not meet sufficient diagnostic criteria: 4 of the 13 had a first-degree relative with MFS and 9 were over-dx based on skeletal features alone.The MFS dx was removed in the over-dx group. *Conclusion:* 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 correct dx of MFS relies on a combination of family history data *plus* methodical clinical evaluation of ocular, skeletal, cardiovascular, and neurologic systems and should be performed in an experienced center.

1047-101 Aortic Regurgitation in an Outlet Ventricular Septal Defect Complicated by Right Coronary Cusp Prolapse: Predictors of Prognosis

Hideshi Tomita, Yoshio Arakaki, Ken-ichi Kurosaki, Toshikatsu Yaglhara, Shigeyuki Echigo, *National Cardiovascular Center, Osaka, Japan, Kurashiki Central Hospital, Kurashiki, Japan.*

Background
Early surgery to prevent progressive aortic regurgitation (AR) is commonly recommended for an 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 525 patients with an o-VSD who underwent the Doppler echocardiography (echo) from January 1985 to May 2001, we reviewed the echo of the aortic valve and associated AR in139 patients. Patients in groups 1-3 did not undergo surgery beyond 18 years old. Group 1 consists of 26 patients without any deformity of the aortic cusp. Group 2 includes 56 patients who did not have AR despite typical RCCP. In 38 patients with RCCP and AR, AR was not progressive and subclinical (Group 3). Nineteen patients underwent surgical treatment because of moderate to severe AR (Group 4). The right coronary cusp deformity index [RCCD=(the length of the deformed right coronary cusp/the diameter of the aortic valve ring)] and the right cusp imbalance index [R/L = (width of right coronary cusp/width 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, 22 vs. 34, 25 vs. 13, and 18 vs. 1 ($p<0.01$). RCCP was firstly detected at significantly younger age in group 4 (6±5 years) than in group 2 (15±8) and 3 (14±5, $p<0.01$). In group 4, AR was first diagnosed at 17±19 years, that was earlier than in group 3 where AR was first detected at 26±17 years ($p<0.01$). Both RCCD (0.40±0.20) and R/L (1.39±0.04) in group 4 were larger than those in groups 2 (0.31±0.09, 1.15±0.10) and 3 (0.26±0.09, 1.13±0.07, $p<0.01$).
Conclusion
Subarterial VSD, early occurrence of RCCP with AR, large RCCD and R/L were predictors of progressive serious AR. However, AR remains mild and not progressive beyond 18 years in the considerable number of patients.

1047-102 Delayed Diagnosis of Kawasaki Disease in the Gamma Globulin Era: How Often and Why? Are the Criteria Too Strict?

Umakumaran Ponniah, Frank C. Smith, Nader H. Atallah-Yunes, Rae-Ellen W. Kavey, Craig J. Byrum, Daniel A. Kveselis, Winston E. Gaum, *SUNY-Upstate Medical University, Syracuse, New York.*

Since gamma globulin therapy (IVGG) is most effective during the first 10 days of illness in reducing the risk of coronary artery aneurysms (CAA) in children with Kawasaki Disease (KD) we retrospectively reviewed all cases of KD from 1978-2001 to determine the following: 1) Is KD diagnosed late (>10 days after onset of illness) as often in the IVGG era (defined as after 8/87) as in the pre-IVGG era (before 8/87)? 2) Are pts diagnosed more often with < 5 criteria in the IVGG era than pre-IVGG and do they have CAA more often than those with ³5 criteria? 3) Is the late diagnosis of KD due to poor detection of KD by physicians in pts with ³ 5 criteria or due to delayed appearance of criteria? Complete data were available in 217/285 pts. The mean time to diagnosis (Dx) was evaluated in pts with definite KD (those with ³5 criteria or with < 5 criteria and CAA, n=173). Number of pts with < 5 vs pts with ³ 5 criteria was determined in 217 pts. Results: Mean time to Dx was shorter in the IVGG era than in the pre-IVGG era (7.7d±4.3 vs 10.2d±7.9; p=.0135), but 33% of pts pre-IVGG and 22% in the IVGG group were diagnosed late (p=NS). Number of pts with <5 criteria was not significantly different between the groups (23% in IVGG group versus 24% pre-IVGG; p=NS). Incidence of CAA was similar in pts with <5 criteria (13.7%) and pts with ³ 5 criteria (10.2%; p=NS). Of the 30 pts in the IVGG era diagnosed late, 13(43%) developed the 5th criterion late (peeling), 5(17%) had <5 criteria with CAA (atypical KD),and 12(40%) were diagnosed late with ³ 5 criteria. CAA were seen in 23% of pts in the late diagnosis group. Conclusion: Although KD is diagnosed sooner in the IVGG era, the number of cases diagnosed after 10 days remains high. 40% of late cases were diagnosed late in spite of fulfilling ³ 5 criteria; however, the other 60% were delayed either because the 5th criterion (peeling) did not appear until after 10 days of illness or because suspicion of KD in spite of < 5 criteria led to the echo diagnosis of CAA. These data emphasize the need for a high index of suspicion of KD in children with <5 criteria and for continued education about the importance of prompt Dx. The results suggest that strict adherance to current criteria prevents the timely diagnosis of KD in a significant number of cases.

POSTER SESSION
1072 Adult Congenital Heart Disease
Sunday, March 17, 2002, 3:00 p.m.-5:00 p.m.
Georgia World Congress Center, Hall G
Presentation Hour: 4:00 p.m.-5:00 p.m.

1072-97 Should We Use 2-D Echo to Assess RV Form and Function in Adults With Repaired Tetralogy of Fallot?

Candice Silversides, Gruschen R. Veldtman, Gary D. Webb, Judith Therrien, Claudia Quammie, Dara-May Rowlins, Naeem Merchant, Annette Flynne, Min-Jin Jung, Samuel Siu, Brian W. McCrindle, *University of Toronto Congenital Cardiac Centre for Adults, Toronto, Ontario, Canada.*

Background. 2-D echo assessment of RV form & function may be inaccurate because of the non-geometric shape of the RV. Methods. We prospectively recruited & evaluated 65 consecutive patients >18 years with standardized clinical assessment, 2-D echo, & MRI. One observer graded the RV visually as follows (echo): 1= normal function or enddiastolic volume (EDV), 2 = mild dysfunction or enlargement, 3 = moderate dysfunction or enlargement; 4 = severe dysfunction or enlargement. Echo was compared with MRI. Results. Median age was 31 years (range 18 to 63). 35 (54%) were female. 25 (39%) patients had transannular patch repair. Conclusions. Echo assessment of the RV gives important initial information, but may be inaccurate. Different echo grades of RVEF & RVEDV frequently overlap when compared to MRI quantification.

Table 2. RVEF assessment

RVEF echo grading	Number with both tests	MRI-RVEF for each echo grade, Mean ±SD	MRI range of RVEF for each echo grade, %
1	17	45±8	28-56
2	25	39±10	12-51
3	11	31±11	17-45

Table 1. RVEDV assessment

RVEDV echo grading	Number with both tests	MRI-RVEDV for each echo grade, Mean±SD (mls)	MRI RVEDV range for each echo grade (mls)
1	7	134±64	50-270
2	19	174±58	50-285
3	19	211±63	77-354
4	7	228±81	114-360