gitant vol over a wide range of hemodynamic conditions, correlation equations vary with each set of conditions, suggesting dependence of measurements on factors including pulse rate, mean arterial pressure, and the geometry of the sampling volume. Accurate measurement of regurgitant vol using PWMV is only possible if the exact hemodynamic conditions and correlation equation are known, limiting current clinical use of this technique. Further investigation may reveal how different hemodynamic conditions affect PWMV measurements of flow.

1034-112 Is There an Inheritance Pattern in Congenital Bicuspid Aortic Valve?

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Congenital bicuspid aortic valve (BAV) is the most common congenital valvular malformation with an estimated incidence of about 1%. BAV is generally not regarded as a hereditary condition, although clustering of BAV has been reported in some families. We performed a prospective study to determine the incidence of familial clustering and the inheritance pattern of BAV. Family pedigrees were constructed including all first degree relatives in 30 randomly selected patients who had BAV diagnosed by echocardiography. The mean age of these probands (17 men, 13 women) was 45 years (range 20 to 75). The probands were interviewed with specific questions to ascertain whether any first degree relatives were known to have cardiac disease. Relevant information when available on more distant relatives was also recorded. Of 173 living first degree relatives, 129 (75%) underwent echocardiography. BAV was diagnosed in 14 (11%), and in an additional patient the diagnosis was probable. Other findings included isolated dilated aortic root in 1, dilated aortic root with BAV in 3, and myxomatous mitral valve in 2. Coarctation was not detected. Eleven of the 30 families (37%) had at least 1 family member with BAV. In these 11 families, the male to female ratio of BAV cases was 1. Seven families had 2 generations and 1 family had 3 generations with affected members. Although the inheritance pattern is uncertain, in some families autosomal dominance with variable penetrance appears likely.

In summary, our data shows that familial clustering of BAV is frequently present and autosomal dominance may be the inheritance pattern in some families. Screening for BAV in first degree relatives should be considered.

1034-113 A Modified Flow Convergence Equation for Calculation of Aortic Regurgitant Volume: Studies in a Quantified Animal Model

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The hemispherical flow convergence equation has been shown to significantly overestimate actual aortic regurgitant (AR) volume due to the non-hemispheric shape of the proximal isovelocity contours within the ascending aorta. In 4 sheep with surgically created AR, we examined the accuracy of a new mathematical flow convergence model, a prolate hemispheroid which appears to best fit the elongated convergence contour shape seen in AR. Actual flow volumes were taken as the difference between pulmonary artery flow and aortic flow measured using electromagnetic flow meters zeroed against each other (range: 7–31 cc). At each depth increment proximal to the orifice, instantaneous flow rates were calculated from digital color M-mode velocities; AR volumes were obtained by integrating instantaneous flow rates over the regurgitant period. Calculated AR volumes measured in the region between 1.2 cm and 1.65 cm proximal to the orifice agreed well with actual AR volumes ($y = 0.91x + 0.32$; $r = 0.92$; SEE = 2.57 cc), while underestimating actual flows at locations < 1.2 cm and significantly overestimating at locations > 1.65 cm proximal to the orifice. A prolate hemispheroidal surface area seems to best represent the isovelocity contours at specific sites proximal to an AR orifice. Use of this formula within the flow convergence equation can be easily automated and should allow simple and accurate quantitation of AR.

1034-114 Is Acquired Platelet Dysfunction an Etiologic Factor in Heyde's Syndrome?

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A relation between significant valvular aortic stenosis (AS) and occult gastrointestinal bleeding has been suggested by Heyde (1958). The aim of this study was to investigate the etiology of Heyde's syndrome.

Methods: 284 patients with AS were screened for bleeding episodes, bleeding time (according to Ivy), blood chemistry and by testing fecal hemoglobin (Hemocult^R) on three separate occasions. 131 patients matched by age and gender and without valvular heart disease were chosen as controls (CG). Examination with upper endoscopy and colonoscopy was performed in all patients with positive Hemocult^R. In patients with a prolonged bleeding time (> 570 sec) an extended evaluation of the coagulation system, including the von Willebrand-factor, was performed. Twelve patients with prolonged bleeding time were reexamined after valve replacement.

Results: No significant difference concerning history of bleeding (18% AS vs 20% CG;NS)or positive Hemocult^R (12% AS vs 11% CG;NS) was observed. However, there was a trend towards a higher prevalence of occult bleeding among patients with AS (9/50(18%) vs 2/25(8%)CG;NS). A prolonged bleeding time was significantly more frequent in the patients with AS (7% vs 2% CG;$p < 0.05$). This was considered to be due to platelet dysfunction as the coagulation status, including the von Willebrand-factor, otherwise was normal. The difference was not related to treatment with aspirin or warfarin. Out of the twelve patients, with prolonged preoperative bleeding time, nine (75%) became normalized after valve replacement ($p < 0.001$).

Conclusion: Prolonged bleeding time, due to acquired platelet dysfunction may contribute to the etiology of Heyde's syndrome. Valve replacement might improve this condition.

1034-115 Aortic Valve Replacement With Autogenous Pulmonary Valve

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Between February 1992 and August 1995, 33 children and adults (11 months to 51 years mean 22 years) underwent autologous replacement of the aortic valve using the pulmonary valve. Pulmonary outflow was reconstructed using aortic homografts in 6 and pulmonary homografts in 27. Root replacements were performed in 28 with freehand reconstruction in 5. Follow up extended to 3 years with no early or late mortality. Twenty seven patients (82%) required blood products. Five patients (15%) required mediastinal exploration for bleeding. Four patients developed supraventricular arrhythmias. Mean hospital stay was 7.5 days (range 5 to 23). No valve replacement has required revision. No thromboembolic or endocarditis events have occurred. No patients have required anticoagulation. All patients are NYHA class I. Echocardiographic studies (n = 33) demonstrate no significant neoaortic valve insufficiency or stenosis. Moderate homograft neopulmonary valve insufficiency was seen in 1 patient with 4 having moderate stenosis with calculated gradients of 16 to 30 mmHg.

Pulmonary autograft replacement of the aortic valve is associated with low mortality and morbidity and excellent function of the neoaortic valve. Pulmonary homograft stenosis warrants close follow-up.

1035 Pediatric Cardiology: Selected Topics

Wednesday, March 27, 1996, 3:00 p.m.–5:00 p.m.
Orange County Convention Center, Hall E
Presentation Hour: 3:00 p.m.–4:00 p.m.

1035-116 Convulsive Syncope of the Young: A Vagal Syndrome With Features Distinct From Mixed Vasodepressor/Cardioinhibitory Syncope and From Breath-Holding

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In infants and toddlers, vagal syndromes may manifest in asystole causing convulsions (breath holding spells). No convulsive vagal syndrome is well-documented in older children. Indeed, when tilt testing evaluation discloses asystole in syncope patients, this is often disregarded. To delineate the significance of asystole in children with convulsions we report head-up tilt evaluation of a series of 15 patients *beyond the age of breath-holding* who had convulsive symptomatology and underwent head up tilt testing. *Methods:* Fifteen patients aged 6–19 diagnosed as having convulsions underwent tilt testing involving 1–4 tilts (60°): 1) baseline, 2) esmolol (500 μg/kg load, 50–300 μg/kg/m), 3) esmolol withdrawal & 4) isoproterenol if not contraindicated. The protocol was aborted if long asystole was observed. *Results:* Age at onset was 7.5 ± 1.2 years. Symptoms included convulsions and syncope in 15/15 and presyncope in 8/15. Frequency was 1.9 ± 1/year for convulsions and 4.4 ± 2.5/month for syncope or presyncope. 2 also had history of simple faint, 1 breath-holding, and family history of faints in 6. Generalized motor convulsion was reported in 9, and lesser syndromes were seen in 6. Three had abnormal EEG; 3 received chronic anticonvulsants. Tilt testing documented asystole in 7 (range 7–35 sec, with AV block in 2), mixed in 2, vasodepressor in 5 and