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Abstracts

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Growth of aortic annulus in infants with critical aortic stenosis

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Few data exist concerning growth of the aortic annulus after valvotomy in infants with critical aortic stenosis (AS). Possible determinants of growth include initial aortic annular size, residual obstruction, and subsequent aortic regurgitation (AR).

We evaluated aortic growth by 2D echo. In 35 AS patients treated with surgical or balloon valvotomy in the first year of life. All had 2 or more aortic annulus measurements separated by a mean of 2.7 years (0.1-17.5 years) Growth of the aortic annulus was analyzed using Z scores adjusted for age and body surface area obtained from a normal population (Z-score = number of SD from the mean). The aortic annular growth rate (AGR) for each patient was defined as the difference in Z-score for sequential measurements divided by the time interval between measurements in years. Analysis of variance was used to test the effect of initial aortic annular sizes, residual AS, and post-valvotomy AR on growth rate.

The mean AGR for the 35 patients was 2.0±3.7 Z-scores/year, indicating an average increase in annular size of two SD relative to the normal population per year. Pre-intervention aortic annular size measurements (Z_o) were available in 28/35 patients for ANOVA.

Initial Size	n	AGR
Z_o<-2	4	8.8±2.9*
-2 Z_o 0	21	1.6±3.2
Z_o>0	3	-0.3±0.1

The fastest growth rate was seen in infants with the smallest initial annulus diameter (*p=0.0006), with negative growth in those with an initial annulus diameter above the population mean, indicating movement of all patiens toward the mean for the normal population. Faster growth also tended to be associated with less severe residual stenosis and less subsequent regurgitation.

Left ventricular hypertrophy and systolic anterior motion of the mitral valve during dexamethasone therapy for bronchopulmonary dysplasia

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High-dose dexamethasone (dex) is frequently used in premature infants in the treatment of bronchopulmonary dysplasia. Our previous clinical experience has suggested that significant cardiac hypertrophy, tachycardia and blood pressure elevation were frequently observed in patients receiving high-dose dexamethasone. This study was undertaken to prospectively assess the effects of dex on heart rate, blood pressure, and echocardiographic measurements of ventricular septal and wall thicknesses, as well as the incidence of systolic anterior motion of the mitral valve. M-mode echocardiograms were performed sequentially on a weekly basis beginning at 2-4 weeks of age in thirteen respirator-dependent infants [BW 825±64 gms (m ±SE); gestational age 25±0.4 weeks]. Eight neonates who were respirator-dependent (FiO_2 67±9%) received dex therapy (500 g/kg/day tapered progressively over 4-6 weeks). They were compared to five neonates of similar weight and gestational age who were respirator-dependent (FiO_2 38±10%), but did not receive any dexamethasone. Measurements of heart rate and blood pressures revealed that mean arterial pressure was mildly elevated in patients receiving dexamethasone relative both to control infants and baseline levels; heart rates tended to increase during the period of treatment in the dexamethasone-treated infants.

Echocardiographic measurements demonstrated significant increases in left ventricular free wall thickness, ventricular septal thickness and right ventricular free wall thickness by 14 days of therapy in dex-treated infants. The chamber diameters did not change significantly and ejection fraction appeared unaffected. Following 2-4 weeks of therapy, all of the measurements of wall thickness gradually fell to near normal values over approximately six weeks as dexamethasone dose was tapered. Systolic anterior motion was observed in all of the dexamethasone-treated infants at some time during the course of therapy, but was not observed in control infants.

Prolonged, high-dose dexamethasone treatment for bronchopulmonary dysplasia may be associated with the development of a transient form of hypertrophic cardiomyopathy.

Alterations in isolated canine pulmonary arteries during pacing-induced congestive heart failure

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Alterations in myocardial function and systemic vascular reactivity during congestive heart failure CHF) are well established. We examined the effects of pacing-induced CHF (240 b/min) on vascular reactivity of isolated canine pulmonary arteries (PA). In the presence of phenylephrine-induced tone, the receptor-mediated cAMP-dependent relaxation to isoproterenol (ISO) (10 nM-1 μM), arachidonic acid (AA) (10 nM-10 μM) and PGI_2 (10 nM-10 μM) showed >50% depression (P <0.05) during CHF compared to control(c). Relaxation responses to forskolin (a direct stimulator of adenylate cyclase) and RO 20-1724 (a cAMP-specific phosphodiesterase inhibitor) were not altered during CHF. The presence of RO 20-1724 enhanced relaxation to ISO (100 nM) in C by 44±10% and in CHF by only 14±7% (P<0.05). Plasma ANF

level is increased in CHF and it correlates well with clinical status. Since atrial appendages are the main source of ANF and ANF is negatively coupled to adenylate cyclase via C receptors, we studied a group of five dogs in CHF who had undergone atrial appendectomy (CHF+ATR APP) earlier. Relaxation responses to ISO and AA were restored to normal, e.g., relaxation to ISO (1 μM) was 49±5% in C (n=7) vs 21±5% in CHF (n=7) (P<0.05) and it was 49±16% in ATRAPP (n=4) group. Relaxation to AA (10 M); 71±5% (C, n=7), 20±8% (CHF, n=7) (P<0.05), 83±5% (ATRAPP, n=5). We conclude, that during CHF, receptor-mediated cAMP-dependent responses are significantly depressed in PA. Direct stimulation of adenylate cyclase does not appear to be affected and there is no evidence for enhanced degradation of cAMP. Circulating ANF levels appear to modulate cAMP-dependent relaxation responses during CHF.

Effect of inhaled nitric oxide in patients with postoperative pulmonary hypertension

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Nitric Oxide (NO), an endothelium-derived relaxing factor, when administered as a gas, may preferentially relax pulmonary vascular smooth muscle. This is thought to be mediated through a rise in intracellular cGMP levels.

We studied the hemodynamics in nine patients with postoperative pulmonary hypertension following intracardiac repair of a congenital heart defect. Endothelial responsiveness was assessed with a 10^{-6}M infusion of acetylcholine (Ach) into the pulmonary artery over two minutes. Inhaled NO was given at 80 ppm for 15 minutes. Plasma levels of cGMP were measured by radioimmunoassay in response to Ach and NO. Results:

	Baseline	Ach 10^{-6}M	Baseline	NO 80 ppm
BP	65 ± 11.0	60 ± 11.3†	61 ± 10.0	60 ± 8.0
PAp	33 ± 9.4	30 ± 8.5†	34 ± 8.4	26 ± 7.5*
CI	4.2 ± 1.6	4.1 ± 1.6	4.0 ± 1.6	4.0 ± 1.5
SVR	15.3 ± 7.0	14.5 ± 7.2	16.4 ± 8.0	15.2 ± 7.3
PVR	5.8 ± 2.7	5.1 ± 2.5†	6.7 ± 3.7	4.5 ± 2.4*

	cGMP (pmol/m) PA	LA
Pre Ach1	2.2 ± 3.4	11.9 ± 3.4
With Ach	10.6 ± 3.4	11.3 ± 3.9
Pre NO	16.0 ± 3.7	15.4 ± 4.1
With NO	56.2 ± 12.4*	48.3 ± 8.2*
Post NO	24.4 ± 5*	29.2 ± 5.8*

†P<0.05; *p<0.005

The administration of NO produced a significant fall in PVR and PAP with little effect on CI or SVR. The attenuated response to Ach suggests that endothelial dysfunction may exist in these patients postoperatively. NO induced a significant rise in cGMP levels which remained elevated 15 minutes after discontinuation of the inhalation. NO is a selective and effective pulmonary vasodilator which may be of value in the treatment of postoperative pulmonary hypertension in the cardiac patient.

Single tomographic section through heart specimen determined by three-dimensional magnetic resonance imaging

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Tomographic imaging by echocardiography and magnetic resonance imaging (MRI) has introduced the need for knowledge of pathologic anatomy in a format which differs from classical gross anatomy. To provide postmortem heart specimens for study, a technique was devised for tomographic sectioning in planes designed to demonstrate specific anatomical features. Eight postmortem heart specimens with the following abnormalities were fixed by inflation: tetralogy of Fallot (2), double outlet right ventricle (2), common atrioventricular canal, transposition of the great arteries, single ventricle, ventricular septal defect Each specimen was scanned in a 1.5 Tesla Siemens Magnetom using a gradient recall echo 3-dimensional (3D) pulse sequence with a 64 slice partition, repetition time 25 msec, echo time 10 msec, and flip angle 40°. For specimens 6 cm in diameter a small field(orbit) coil was used; for those > 6 cm a spine coil was used. Scanning time was less than 15 min. Following scanning, specimens were infiltrated with paraffin. Images were transferred to a Sun workstation for analysis. From the Volumetric Imaging Display and Analysis (VIDA) software package developed at our institution the oblique sectioning module was used to select a plane which best demonstrated the desired anatomical features for each specimen. Coordinates of the plane were transferred to the volume render module where a 3D shaded surface display of the exterior of the specimen was created. The intersection of the oblique plane of interest with the surface display was shown as a colored stripe. By matching details of each specimen's surface with the line of intersection in the display, the line of intersection was transferred to the specimen. Specimens were then cut on a band saw. In each case, the desired plane of section was achieved. We conclude that 3D MRI can be used to aid in the sectioning of gross anatomical heart specimens to demonstrate pertinent tomographic anatomy.

Initial experience using a new balloon expandable stent to enlarge the branch pulmonary arteries in a neonatal animal model

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Early surgical or balloon angioplasty techniques for hypoplastic branch pulmonary arteries(PA's) in small infants result in better long-term growth; However, both techniques are associated with variable success. The use of a balloon expandable stent that can be enlarged as the infant grows may have obvious advantages. Therefore, we evaluated a newly devised balloon expandable stent specifically designed for newborn babies that could be placed at catheterization. The stent (3.5-5.0 mm x 1.2 cm) was mounted on a balloon catheter and the entire system was delivered through a 5 or 7F sheath. Eight piglets with normal PA's, median age 14.5 days (range 4-22 days), median weight 3.3 Kg (range 2-5.3 Kg) underwent placement of a stent in the branch PA's, the stent was expanded to 1 mm larger than the native PA's diameter. One stent migrated proximally requiring removal and placement of a new stent. Two animals were sacrificed one week later after developing gastrointestinal illness; one animal was sacrificed for leg ischemia. The remaining five animals underwent repeat catheterization at a median interval of 35 days (range 25-54)days), and median weight of 11 Kg (range 10-31 Kg). I Angiography in all five demonstrated patency of the stent, as well as side and distal PA's. All animals were sacrificed by seven weeks. Histology revealed complete endothelialization after four weeks. This study demonstrates that a balloon expandable stent can enlarge and maintain patency of the pulmonary arteries in small animals. This stent procedure may be applicable to small infants with branch pulmonary artery stenoses, obviating the need for early surgical or balloon angioplasty techniques. Moreover, it may augment the growth of the pulmonary arteries.

Recent improvement in outcome using transcatheter embolization for aneurysmal malformation of the vein of Galen: clinical assessment

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Infants with vein of Galen malformation may present with early congestive heart failure (CHF) which is usually fatal if untreated. Early studies using transcatheter embolization in 22 patients had a 50% mortality and a 37% incidence of severe mental retardation in the survivors. A clinical analysis of 11 consecutive patients presenting at N.Y.U. Medical Center from 6/90 to 9/91 was recently conducted. Eight children were male, 10 were diagnosed in the first 3 days of life. All had a bruit over the anterior fontanelle. Ten had a hyperdynamic circulation with 8 requiring treatment of acute CHF. Macrocephaly was present in 6 and 4 had hydrocephaly.

A multi-disciplinary treatment protocol was utilized. There was no mortality and 6 of the children followed for up to 40 months are functionally normal. Two patients have severe neurologic deficits and/or a seizure disorder with one possibly related to the embolization procedure. Some developmental delay was noted in one additional child. Ventriculo-peritoneal shunts were required in four patients for progressive hydrocephalus or to relieve the pressure effects of intraventricular hemorrhage. One child had spontaneous closure of the aneurysmal malformation.

These improvements result, in part, from better general neonatal care and also from specific modifications of the treatment protocol. These latter include earlier diagnosis, newer microcatheters and acrylic polymers, avoidance of digoxin use, restriction of overly aggressive neurosurgical procedures and use of stable central vascular access for total parenteral nutrition. The diagnosis of vein of Galen malformation no longer automatically implies a fatal or dismal prognosis. Continuing improvement in treatment and outcome are to be expected in specialized treatment centers handling the care of infants with this rare defect.

Double umbrella closure of post-myocardial infarction ventricular septal defects

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Ventricular septal rupture is a rare but often fatal complication of acute myocardial infarction (AMI), despite medical/surgical intervention. Between 2/90 and 5/92 we attempted transcatheter closure (TCC) of ventricular septal defects (VSD's) complicating AMI using a Clamshell (C) occluder in 8 patients (pts), age 52-79. Five pts (POST-MI) presented acutely, without surgical intervention all required hemodynamic support and had multi-organ failure. Three other pts survived surgical VSD repair but had significant postoperative residual defects (PORD's), one required hemodynamic support. At catheterization, all pts had pulmonary/systemic blood flow > 1.5 POST MI VSD's were 18-20 mm diameter and were associated with a septal cavity (4) or marked septal thinning (1). PORD's were 8-19 mm diameter. C's were successfully implanted in stable position in all pts (1 pt received 2 devices)—moderate residual angiographic shunting was present in all. Procedural complications included C retrieval (2) with tricuspid leaflet rupture/regurgitation (1), transfusion (3) and acute renal failure (1)—all occurred in pts POST-MI. Survival in all POST-MI pts was short-term (1-90 days), while all pts with PORD's are alive with both hemodynamic and symptomatic improvement (4-31 months). This experience suggests that TCC of post-MI VSD's, limited by available C technology, may provide long-term improvement for pts with post-operative VSD's, but currently offers little long-term benefit in the acute management of unoperated pts with hemodynamic compromise and multiorgan failure.

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