

EARLY POSTOPERATIVE TRANSESOPHAGEAL ECHOCARDIOGRAPHIC EVALUATION OF RESULTS OF RIGHT OUTFLOW TRACT RECONSTRUCTION FOR CONGENITAL HEART DISEASE.

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Transesophageal echocardiography (TEE) was performed to evaluate anatomy of RVOT and residual gradients in 27 children (mean age 51 ± 39 months, mean weight 14.7 ± 7.8 kg) using a biplane 11mm probe (Aloka) (n = 22) and a single plane 6.8 mm probe (n = 5) within 24 hours after surgery. 17 children had Tetralogy of Fallot, 4 pulmonary atresia, 3 transposition of the great arteries, and 3 double outlet right ventricle.

Results. Subvalve RVOT was visualized in all using longitudinal (L) plane, and the pulmonary valve and proximal bifurcation by both planes. Measurement of segments of RVOT obtained by L plane and Transverse (T) plane were compared with pre-surgery angiography.

Mean S.D (mm)

	Angio	L Plane	T Plane
Subvalve		14.7±3.9	14.3±3.9
Valve	9.0±5.2	13.8±5.1	13.5±4.7
Supravulve		15.7±3.8	14.9±4.0
LPA	11.3±5.1	11.1±3.5	10.1±3.0
RPA	11.1±4.9	10.0±3.5	8.9±2.6

One patient had residual pulmonary valve stenosis (Doppler velocity 2.3 m/sec). The 2 patients who underwent post-operative cardiac catheterization died. Cardiac catheterization revealed no new anatomical findings in both patients.

Conclusion: Combination of L and T plane is essential for adequate evaluation of RVOT reconstruction. Measurements obtained by either planes for main, right and left pulmonary arteries were similar. Biplane TEE may replace catheterization in post-operative RVOT evaluation.

MANAGEMENT AND PROGNOSIS OF DILATED CARDIOMYOPATHY DETECTED BY FOETAL ECHOCARDIOGRAPHY

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Marked LV dilatation & dysfunction was diagnosed in 7 of 1158 foetal echocardiographies (FE) over 10 years. This represents 6% of structural cardiac abnormalities. FE was first performed at 29-38 weeks (mean=33) following referral for abnormal 4 chamber view(4), foetal hydrops(2) & arrhythmia(1). There was polyhydramnios in 3 cases.

Two foetuses were correctly diagnosed as having critical aortic stenosis, a third had thickened immobile mitral valve, fused foramen ovale (FO) and at autopsy EFE & pulmonary vein stenosis. The 4 remaining foetuses had structurally normal hearts with marked LV dilatation & in one the RV was markedly dilated and there was LV thrombosis. These findings were confirmed postnatally. The foetus with premature closure of FO & one with RV & LV dysfunction developed hydrops, but the other 5 did not & showed normal growth. Spontaneous labour occurred in 6 of 7 cases at mean 36.7 weeks.

Both neonates with critical aortic stenosis survived neonatal valvotomy & 4 of 5 remaining neonates died at 2 hr, 2 & 5 days & 14 months. During the same period, no mid-trimester abortions or late stillbirths with dilated LV were autopsied by our provincial service.

FE can accurately distinguish cardiomyopathy from LV outflow obstruction. In absence of Rt sided disease or premature closure of FO, severe LV dysfunction is well tolerated in utero with normal growth. This suggests aggressive in utero intervention may not be warranted, at least until a better natural history is established.

EVALUATION OF MYOCARDIAL VIABILITY USING POSITRON EMISSION TOMOGRAPHY IN CHILDREN AFTER KAWASAKI DISEASE

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To evaluate myocardial viability in children with coronary artery lesions after Kawasaki disease, we assessed regional myocardial perfusion and metabolism using positron emission tomography (PET) with ^{13}N -ammonia and ^{18}F -deoxyglucose (FDG) at rest in fasting state in 12 patients aged 3 to 14 years. In addition, PET findings were compared with ECG and regional wall motion scored from 0 (normal) to 4 (dyskinesia) on contrast LVgraphy to examine whether these commonly used tests can distinguish viable myocardial regions from non-viable ones. Results: As shown in Table, PET identified 3 groups (A to C) of abnormal myocardial segments. Wall motion score was similar among these 3 groups. Number of segments with neither abnormal Q nor abnormal Q and T waves was not significantly different between viable group (A and B) and non-viable group (C). The duration of exhibiting abnormal Q waves on serial ECGs taken once a month was significantly shorter in viable group than non-viable one (Table).

PET:	Group	viable		
		A(n=4)	B(n=7)	C(n=9)
Perfusion/FDG uptake	/	/	/	/
Wall motion score		1.92±0.99	2.05±0.85	2.19±1.20
Abnormal Q(+Abnormal T)		6(2)/11		6(3)/9
Duration of exhibiting abnormal Q (months)		26.3±15.7		92.4±26.8*

∴ normal, ∴ enhanced, ∴ reduced, ∴ p<0.01 vs viable
Conclusions: Neither regional wall motion nor a single ECG can distinguish viable myocardial regions from non-viable ones. Shorter duration of exhibiting abnormal Q waves on serial ECGs may suggest viable myocardium.

LUNG PERFUSION PATTERNS FOLLOWING BI-DIRECTIONAL CAVO-PULMONARY ANASTOMOSIS: A REFLECTION OF THE PULMONARY VASCULAR BED STATUS.

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To evaluate the pulmonary blood flow (PBF) distribution following bi-directional cavo-pulmonary anastomosis (BCPA) including augmentation of central PA branches and prior to Fontan procedure (FP), perfusion lung scans (LS) were performed on 46 pts who had BCPA at 15 ± 7 (mean \pm SD) months followed by FP at 23 ± 9 months of age. LS were performed immediately prior to NP using Technetium 99m macroaggregated albumin. Relative lung perfusions were determined from geometric mean counts in 5 minute anterior and posterior images in multiple projections. Normal PBF distribution was defined when one lung received at least 45% of the total PBF. The average distribution of the PBF to the left lung (LL) relative to right lung (RL) was 36 ± 11 vs $64 \pm 11\%$. Twelve pts had normal LL/RL perfusion ratio (mean 48/52) with one in-hospital death (8%). Seventeen pts had mildly reduced (35-44% of PBF to LL) LL/RL perfusion ratio (mean 38/62) with 2 deaths (12%). Eleven pts had moderately reduced (25-34% of PBF to LL) LL/RL perfusion ratio (mean 29/71) with 2 deaths (18%). Five pts had severely reduced (\leq 24% of PBF to LL) LL/RL perfusion ratio (mean 17/83) with 1 death (20%).

There was no difference between the 4 groups in age at BCPA, relative size of proximal PA branches (by simultaneous echocardiogram), the site of the previous AO-to-PA shunt (central vs. right-sided) or the AO O₂ sat.% (at simultaneous cardiac catheterization ($83 \pm 5\%$)).

Conclusions: (1) Moderately-to-severely abnormal PBF distributions occurred in 37% of pts reflecting local abnormalities in the pulmonary vascular bed as it was not related to age at BCPA, PA branch size or site of prior shunt. (2) Such PBF abnormalities did not affect AO O₂ sat.% following BCPA, but correlated with early mortality following FP.