Echocardiogram Can Replace Catheterisation in The Management of Congenital Heart Diseases

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The technology of echocardiography has grown by leaps and bonds and has become an essential part of diagnosis and surgical / non surgical management of congenital heart diseases (CHD). So much so that it has almost threatened the supremacy of catheterization and angiogram, which is considered as a gold standard for diagnosis especially of complex congenital heart diseases. Today 2-D echo along with colour doppler, transoesophageal echo, fetal echo, intracardiac echo and 3-D echo have made the work of an interventional cardiologist and cardiac surgeons so much more easy. In many centers, cross-sectional echocardiography has replaced cardiac catheterization and angiography as the primary diagnostic tool, and cardiac operations are planned and executed on the basis of echocardiography. The transoesophageal approach has now been extended to very small infants with smaller multiplane probes used in infants of 2 to 3 kgs and this intra-operative approach has dramatically reduced the need for immediate post-operative reoperation, morbidity and mortality. Fetal echocardiography has provided a new frontier for diagnosis, counseling and treatment of CHD.

As the information that one gets from echo with segmental approach is so complete, that people started questioning whether costly, invasive and at times risky investigations like catheterization, angiogram and MRI angiogram in small and sick children is necessary? The introduction and development in echo and Doppler modalities enable definitive diagnosis of CHD without cardiac catheterization.[1] The introduction of each new echocardiographic technique was associated with a significant fall in the total catheter operation ratio compared with the preceding period.[2] Hence, in many centres all over the world, the patients with congenital heart diseases go for surgery/ intervention with only echo diagnosis without catheterization. Marino et al reported in 1990, cardiac surgery in 602 children without preceding cardiac catheterization, diagnosis being based on clinical findings and 2-D echo and Doppler.[3] There was major error in 2.5% and minor error in 2% of cases. They gave the new trends and established indications – the malformations most suitable for non-bypass surgery without catheterization seem to be those with reduced pulmonary blood flow requiring systemic pulmonary artery shunt, coarctation of aorta (COA) and patent ductus arteriosus (PDA). For open heart surgery without invasive investigations, atrial septal defect (Fig. 1 a, b), pulmonary stenosis, aortic and subaortic, supra- aortic stenosis, cardiac tumors (Fig. 2) and isolated valve disorders like Ebstein's anomaly are 'classic' candidates. Recent experience indicated that 'selected' cases of completed AV canal defect, tetralogy of Fallot, truncus arteriosus, total anomalous pulmonary venous connection (TAPVC) (Fig. 3) and transposition of the great arteries (Fig. 4 a, b) may safely undergo primary repair with Echo evaluation, without cardiac catheterization.

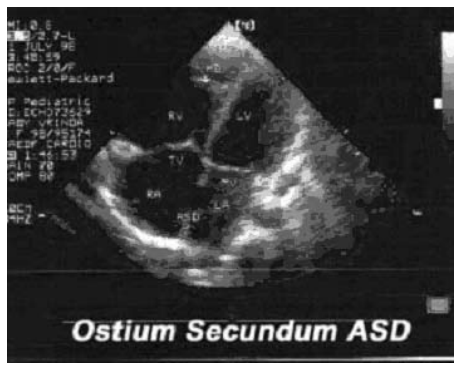


Fig. 1a: Ostium secondum ASD

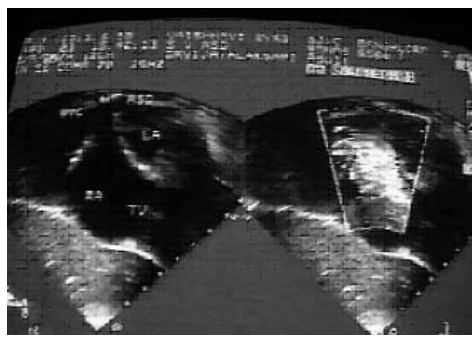


Fig. 1b: Sinus Venosus type ASD

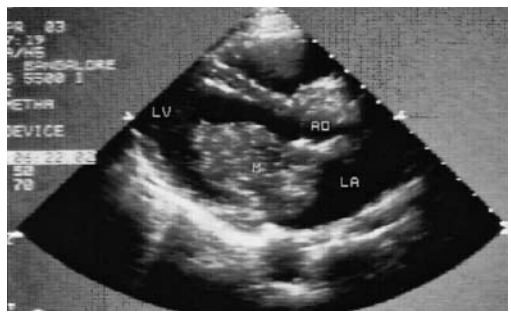


Fig. 2: Left Atrial Myxoma

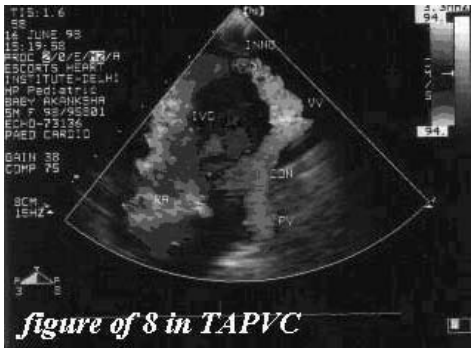


Fig. 3: TAPVC -figure of eight appearance shown in echo.
Vertical vein, horizontal vein draining into SVC & RA junction

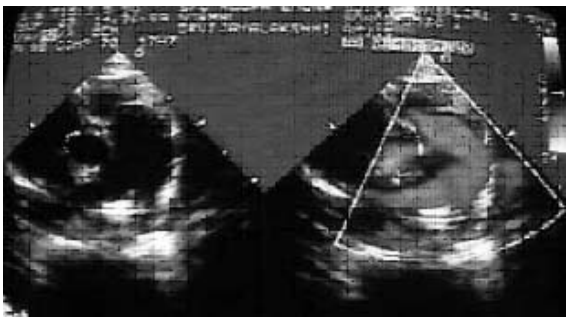


Fig. 4a: Normal relation of great arteries

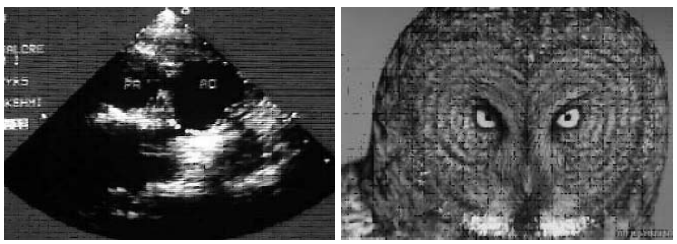


Fig. 4b: In TGA, the great arteries typically appear as "double circles" (owl's eyes) and aorta is anterior and to the right of the main pulmonary artery

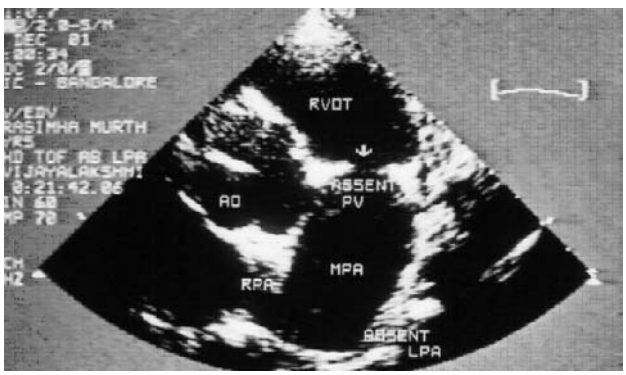
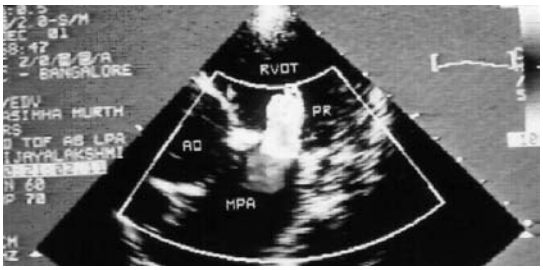


Fig. 5a: TOF with absent PV and LPA

Catheterization is a must when cases of AV canal defects are associated with pulmonary hypertension (PH) or when TAPVC is infradiaphragmatic, which is usually associated with PH or when haemodynamic vice is predicted. Cardiac catheterization remains mandatory for patients with tetralogy of Fallot.[4] Based on the excellent diagnostic correlation between echocardiography and angiocardiography, a policy of echo-guided primary repair of uncomplicated selected cases of tetralogy of Fallot was introduced



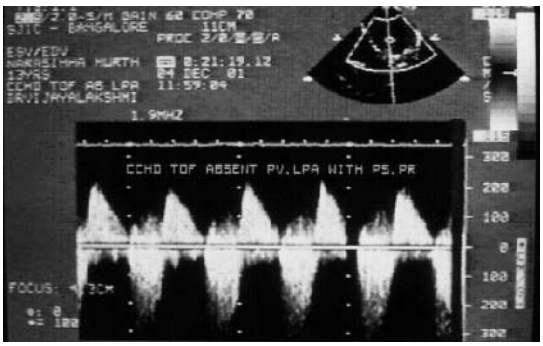


Fig. 5b, c: TOF with PS and PR in short axis and colour Doppler

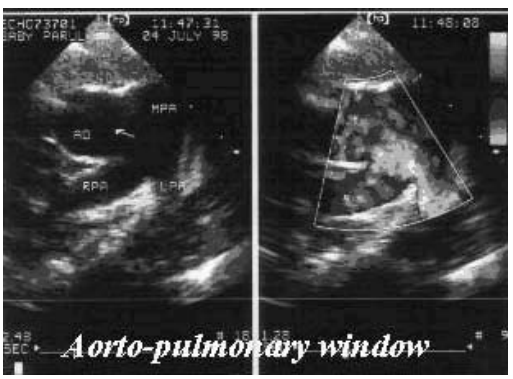


Fig. 6: Aorto-pulmonary window

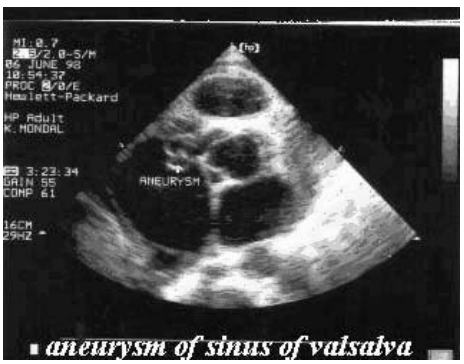


Fig. 7: Aneurysm of sinus of Valsalva

at Ospedale Bambina Gesa in Italy. Echocardiography showed an overall sensitivity of 100% and specificity of 85% for detection of associated malformations. In other words, echo cannot replace catheterization in cases of TOF with collaterals anomalous coronary arteries, absent pulmonary valve right or left pulmonary arteries (Fig. 5 a, b, c) or associated supravalvar pulmonary artery stenosis or poor pulmonary artery tree, requiring altered strategy for surgery. But in fact with careful application of mind, Echo can detect these conditions accurately.

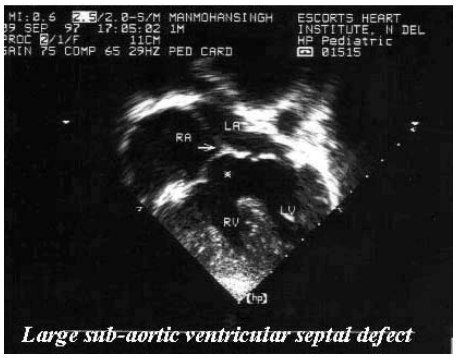


Fig. 8a: Subaortic VSD

Fig. 8b: Mulitple VSDs

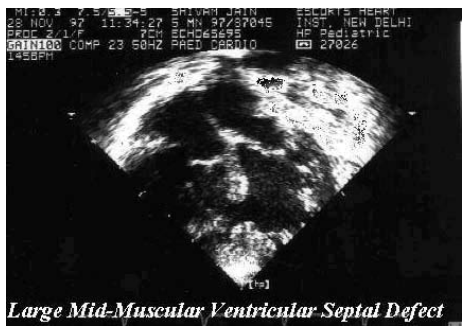
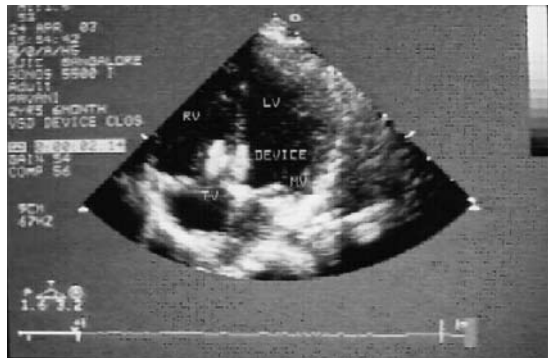


Fig. 8c: Mid-muscular VSD

Atrial septal defect (ASD) is the most common CHD which was catheterized in the past. [5] Today the precise diagnosis of type, number of atrial septal defects (ASDs), septal aneurysm rims around the defect and suitability for device closure can be made out by TTE or TEE. In fact echo plays a major role in balloon sizing of ASD and positioning of AMPLATZER device during the procedure. Even associated partial anomalous venous connection can be detected by echo. Nowadays TEE and intracardiac echo are being used for transcatheter procedures and in the post-surgical period.

In the past, patients with continuous murmur were taken up for surgical ligation and were closed on not finding the PDA on the operation table. Today echo can differentiate PDA, aorto-pulmonary window (Fig. 6), Coronary AV fistula and RSOV (Fig. 7). The catheterization is required only to know the pulmonary artery pressure and pulmonary vascular resistance.

Echo can detect the site, size number of VSDs, septal aneurysm, associated lesions (Fig. 8 a,b,c) and also after closure procedures (Fig. 9 a, b). Cardiac catheterization is commonly performed before repair of ventricular septal defect (VSD) in infancy. But the



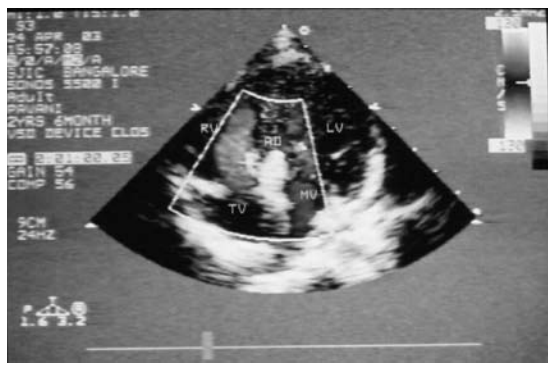


Fig. 9 a, b: Device across perimembanous VSD, colour Doppler shows no residual shunt, no MR/TR

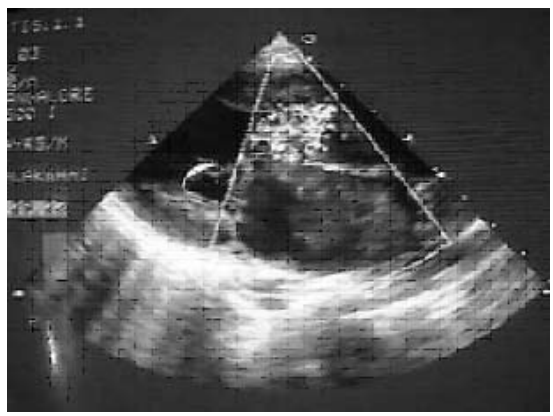


Fig. 10: VSD with aortic regurgitation due to cusp prolapse

sensitivity and specificity of echocardiography in the detection of additional VSDs was 60% and 99% compared with 53% and 97% for cardiac catheterization. Hence, conclusion drawn was routine preoperative cardiac catheterization for infants with a primary diagnosis of VSD is probably no longer justified. [6] The pulmonary artery pressure estimated by echo gives an estimate of the "size" of the VSD, as large VSDs are associated with high pulmonary artery pressure and vice versa. But calculation of flow in ASD or VSD by echo can be erroneous. Catheterization is essential to calculate pulmonary vascular resistance in cases of PH. The echo plays an important role in an early detection of prolapse of aortic cusp, causing aortic regurgitation (Fig. 10).

The patients with heterotaxy syndrome frequently have complex congenital cardiac and non-cardiac malformations requiring detailed diagnostic evaluation by noninvasive as well as invasive imaging. Catheterization data were necessary only to determine pulmonary vascular resistance before Fontan operation.[7]

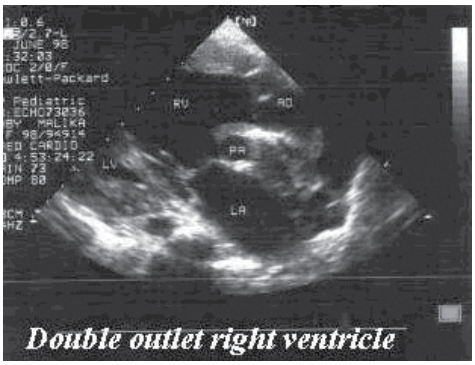


Fig. 11: Double outlet RV

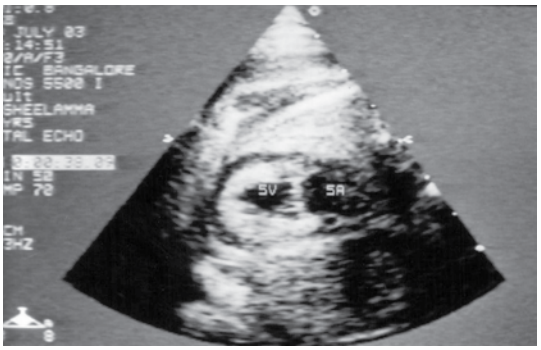


Fig. 13: Fetal echo showing single ventricle

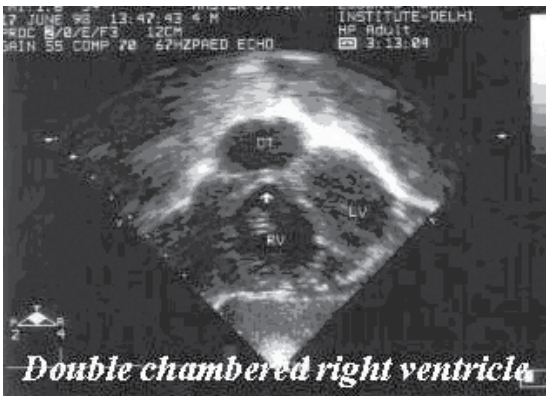


Fig. 12: Double chambered right ventricle

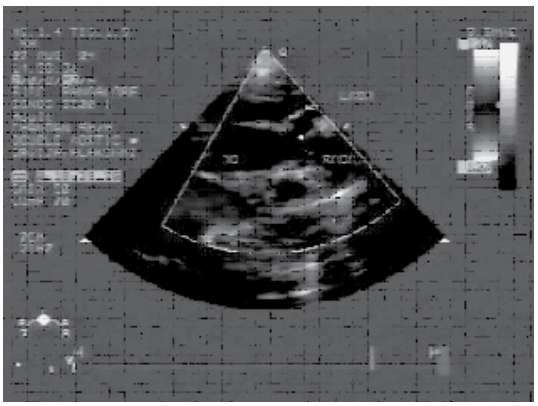


Fig. 14: Double aortic arch

Several studies have shown that non-invasive diagnosis of some infants with congenital heart disease can be performed without catheterization and it reduces hospital costs, decreases total number of catheterizations performed and influences the structure of training programs.[8-10] But some feel combined clinical and non-invasive echo evaluation results in accurate diagnostic information adequate for formulation of appropriate management strategies in the majority of patients but in many individuals with heart disease still require evaluation with catheterization and angiogram for complete diagnosis.[11] In fact echo helps not only before surgery but TEE is used during surgery to get the best surgical results. The study performed to evaluate accuracy of exclusively non-invasive preoperative diagnostic work up based on echocardiography and to assess the safety of cardiac surgery show that diagnostic errors occurred in 3.9% of catheterized patients and in 6.9% of patients with echocardiography only.[12,13] The major diagnostic errors (resulting in prolongation of cardiac bypass time) were observed in equal frequency in both groups (1.7% in echo group and 1.6% in catheterized group). The outcome showed that diagnostic cardiac catheterization could be avoided in majority of cases. Hence, echocardiography combined with Doppler studies is now the most informative diagnostic modality and its usefulness is being expanded by transesophageal studies. The study performed to assess the feasibility, additional diagnostic value and potential applications of biplane transesophageal echocardiography in neonates, infants and children, showed that longitudinal plane provided superior visualization of both right and left ventricular outflow tracts, the inter-atrial septum, the main pulmonary artery, ascending aorta and right coronary artery. In 16% of patients the longitudinal plane provided completely new diagnostic information that was not obtained with combined transthoracic and transverse plane transesophageal echocardiography. The longitudinal plane is complimentary to transverse plane, but cannot substitute it. [14]

Recent developments in the field of pediatric cardiac surgery and pediatric cardiology, especially non-surgical transcatheter management have changed the pre-operative or pre-procedure diagnostic work up of the cardiac malformations. Excellent, accurate diagnosis can nowadays be achieved in most of the patients without invasive cardiac catheterization and based exclusively on echo findings alone.[15] The double outlet right ventricle (Fig. 11), Double chambered RV (Fig. 12), transposition of great arteries, ASD, single ventricle even in foetus (Fig. 13), corrected transposition of great vessels and tricuspid atresia can be accurately diagnosed by echocardiography.[16] The defects in which coronary arterial anatomy may complicate repair are TOF, truncus arteriosus, transposition of great arteries and double aortic arch (Fig. 14) can also be evaluated by echo. However, intra-operative evaluation of coronary artery is imperative to avoid complications.[17]

In the neonates and children, echo window is excellent and gives every details of the heart, whereas catheterization and angiography under general anesthesia carry high incidence of morbidity and rarely mortality in very sick children. Hence, non-invasive echo which is cost-effective, safe, reproduceable has replaced invasive cardiac catheterization in decision making in congenital heart disease in many cases.