Artículo original

# Accuracy and utility of preoperative transthoracic echocardiography alone in the surgical management of congenital heart disease

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**Resumen**

Hay un número creciente de pacientes con cardiopatía congénita que se operan sólo con la información que proporciona la ecocardiografía transtorácica, sin necesidad de cateterismo cardiaco. Se evaluó la utilidad de la ecocardiografía preoperatoria en un grupo no seleccionado de pacientes sometidos a cirugía para una cardiopatía congénita en un centro de tercer nivel. Se revisaron en forma retrospectiva los datos de 363 pacientes consecutivos sometidos a cirugía correctora o paliativa de cardiopatía congénita en 1998 en el Texas Children's Hospital. El 49 por ciento de los pacientes eran acianóticos y 51 por ciento, cianóticos. Se compararon los diagnósticos ecocardiográficos preoperatorios con los hallazgos quirúrgicos y se valoró el impacto de las discrepancias diagnósticas. Se definieron las discrepancias diagnósticas en mayores, si hubo necesidad de modificar el abordaje quirúrgico; menores si no hubo impacto sobre la cirugía. Hubo en total 16 (4.4%) discrepancias menores y tres (0.1%) mayores en las 363 operaciones. De las 232 cirugías realizadas en base de datos ecocardiográficos exclusivamente (64% de todos los casos), hubo siete discrepancias diagnósticas menores (3.0%) y una mayor (0.4%). El 74 por ciento de las cirugías iniciales y el 68 por ciento de las cirugías definitivas se realizaron sobre la base exclusiva de la ecocardiografía. Los neonatos fueron los pacientes que más se operaron en tal forma (83 por ciento) de todo el grupo. Se estimó que ningún paciente tuvo resultado adverso por ninguna descrepancia diagnóstica ecocardiográfica. La ecocardiografía por sí sola es confiable y segura en el diagnóstico de la mayoría de los pacientes con cardiopatía congénita, incluso los casos complejos con múltiples lesiones.

**Palabras clave**: Ecocardiografía exclusivamente, diagnóstico preoperatorio, cardiopatía congénita.

**Abstract**

Increasing numbers of patients undergo surgery for congenital heart disease (CHD) based on transthoracic echocardiographic data alone, without cardiac catheterization. We sought to evaluate the utility of preoperative echocardiography in an unselected group of patients undergoing surgery for congenital heart disease in a large, tertiary care setting. Preoperative data from 363 consecutive patients who underwent definitive or palliative surgeries for congenital heart disease in 1998 at Texas Children's Hospital were retrospectively reviewed. Forty-nine percent of the patients had acyanotic and 51% had cyanotic congenital heart disease. Preoperative echocardiographic diagnoses were compared to operative findings and the impact of diagnostic discrepancies was evaluated. Diagnostic discrepancies were defined as either major if the surgical approach was altered, or minor if there was no impact on surgery. A total of 16 minor (4.4%) and 3 major (0.1%) diagnostic discrepancies were noted in the 363 surgeries. Of the 232 surgeries performed based on echocardiographic data alone (64% of all cases), there were 7 minor (3.0%) and 1 major (0.4%) diagnostic discrepancies. Seventy-four percent of initial surgeries and 68% of definitive surgeries were performed based on echocardiography alone. Neonates had the highest echocardiography alone rate (83%) of all age groups. No patients were judged to have had a significant negative outcome due to any echocardiographic diagnostic discrepancies. Echocardiography alone is reliable and accurate for pre-operative diagnosis in the majority of patients with congenital heart disease, including complex cases with multiple lesions.

**Key words**: Echocardiography alone, preoperative diagnosis, congenital heart disease

## Introduction

With advances in echocardiographic techniques, including 2-dimensional imaging and pulse and color-flow Doppler, noninvasive assessment of much of the anatomic and hemodynamic data previously only available from cardiac catheterization is now possible in many patients with congenital heart disease. As such, noninvasive diagnosis via transthoracic echocardiography has replaced preoperative evaluation by cardiac catheterization for most relatively simple forms of CHD at many centers. In addition, an increasing number of centers are now using echocardiography alone for preoperative diagnosis of increasingly complex CHD, including neonates and patients with multiple cardiac lesions [1-8]. The purpose of this study was to evaluate the utility of transthoracic echocardiography alone in the preoperative evaluation of an unselected group

of patients undergoing cardiac surgery in a large tertiary care setting. We compared preoperative echocardiographic results with surgical findings to evaluate the accuracy of pre-operative diagnosis by echocardiography alone, as well as to assess the clinical impact of diagnostic discrepancies.

## Methods

### Patients

The records of 363 consecutive patients with congenital heart disease who underwent definitive or palliative surgery in 1998 at Texas Children's Hospital were retrospectively reviewed. All patients had preoperative evaluation including physical examination, chest roentgenogram, 12-lead electrocardiogram and echocardiogram. A total of 232/363 patients (64%) had surgery without catheterization, including 8 who underwent catheterization for bedside balloon atrial septostomy only (without additional invasive diagnostic evaluation). The remaining 131/363 (36%) had both echocardiography and catheterization as part of their preoperative diagnostic evaluation.

### Echocardiography

Studies were performed by experienced pediatric sonographers and/or pediatric echocardiographers on commercially available echocardiographic scanners utilizing appropriate frequency transducers (2.5-8 MHz) for body size. Images were recorded on super VHS videotape. Studies were viewed live in real-time and the recorded images were also reviewed by experienced pediatric echocardiographers prior to final diagnosis. Studies were performed in a systematic fashion from parasternal long- and short-axis, apical, subcostal and suprasternal notch views, allowing complete segmental analysis of cardiac anatomy. Two-dimensional, M-mode, color flow Doppler and pulse and continuous wave Doppler were performed in all patients. Oral chloral hydrate and/or intranasal midazolam were used for sedation when necessary.

Echocardiographic diagnostic accuracy was evaluated by comparing the pre-operative echocardiographic diagnoses with surgical findings as documented in operative procedure notes. Diagnostic discrepancies were defined as either major if the surgical approach was altered or the surgical risk was increased, or minor if there was no significant impact on surgery and outcome.

### Statistical analysis

SPSS statistics software (SPSS PLUS, SPSS INC., Chicago, IL) was used to perform data entry and statistical analysis. For all analyses, a p-value $<0.05$ was considered statistically significant.

## Results

Of the 363 surgical patients, 223 (61%) were male and 140 (39%) were female. Patients ranged in age from 1 day to 40 years (mean 3 years), including 75 neonates (21%) and 211 patients less than 1 year old (58%). Definitive surgery was performed in 277/363 patients (76%) and the remaining 86/363 patients (24%) had palliative surgery. Most patients underwent their first surgery (285/363, 79%) and/or had procedures requiring cardiopulmonary bypass (307/363 pts, 85%). Patients were evenly distributed with respect to physiology with 179 patients (49%) having acyanotic CHD (left to right shunts, systemic outflow obstruction, normal or increased pulmonary blood flow) and 184 (51%) having cyanotic CHD (right to left shunts, decreased pulmonary blood flow, complex/multiple cardiac lesions with intracardiac mixing).

A total of 232/363 surgeries (64%) were performed based on echocardiography alone. Table 1 shows the percentages of patients undergoing surgery based on echocardiography alone for different age ranges. Neonates underwent surgery based on echocardiography alone most frequently (83% of patients). The percentage of patients undergoing surgery based only on echocardiography gradually decreased with age up to 4 years, with an increase to 75% in the 4-7 year age group. The percentage of patients undergoing surgery based on echocardiography alone decreased to 53% in patients over 7 years of age.

Table 2 shows the diagnostic categories for patients undergoing surgery based on echocardiography alone. A significantly higher percentage of patients in the acyanotic

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CHD group underwent surgery based on echocardiography alone than the cyanotic group (79% vs. 50%, P<0.01).

**Table 1.** Age Distribution of Surgeries Based on ECHO Alone

| *Age group* | *Total number of surgeries* | *Percentage of surgeries based on echocardiography alone* |
|---|---|---|
| 0-28 day | 75 | 83% (62/75) |
| 1-6 month | 101 | 68% (69/101) |
| 7-12 month | 35 | 51% (18/35) |
| 1-3 years | 64 | 45% (29/64) |
| 4-7 years | 35 | 75% (28/35) |
| >7 years | 53 | 53% (28/53) |
| Total | 363 | 64% (232/363) |

Table 3 demonstrates the diagnostic discrepancies between echocardiography and surgical findings. Of the 232 patients (64%) who underwent surgery without catheterization there were 8 diagnostic discrepancies compared to intraoperative surgical findings, 1 major (0.4%) and 7 minor (3%), resulting in an accuracy rate of 96.6% for patients undergoing surgery based on echocardiography alone. Both echocardiography and cardiac catheterization were performed preoperatively in 131/363 patients (36%). There were 2 major (0.4%) and 9 minor (6.8%) diagnostic discrepancies based on echocardiographic diagnoses in patients who had both echocardiography and catheterization. Primary indications for catheterization included evaluation of pulmonary artery resistance/anatomy or aortopulmonary collateral arteries (n=87; 66%), interventional procedures (n=31; 24%), diagnostic questions not defined adequately by echocardiography (n=7, 5.4%), and evaluation of coronary artery or pulmonary venous anatomy (n=6; 4.6%). Catheterization correctly identified the 2 major diagnostic discrepancies not diagnosed by echocardiography (1 pulmonary artery sling misdiagnosed as a retroesophageal subclavian artery by echocardiography, but correctly diagnosed by MRI and catheterization, and 1 patient status post balloon dilatation of coarctation of the aorta diagnosed as recurrent coarctation was found to have an aneurysm at coarctation site by catheterization with surgery identifying aneurysm and dissection). Catheterization identified 6 out of 9 minor diagnostic discrepancies incorrectly diagnosed by echocardiography.

Overall, of the 363 surgeries performed, 344 pts (94.8%) had preoperative echocardiographic diagnoses consistent with intraoperative surgical findings. There was no significant difference in the incidence of diagnostic discrepancy between patients evaluated preoperatively with echocardiography alone and those who underwent additional cardiac catheterization. Of the total of 19 diagnostic discrepancies between preoperative echocardiography and surgical findings, 16/363 (4.4%) were minor and 3/363 (0.1%) were major (Table 3). Mortality was 3.3% (12/363 patients) within 30 days of operation. There were no diagnostic discrepancies among those 12 patients.

**Table 2.** Diagnoses and Percentage of Patient Undergoing Surgery Based on Echocardiography Alone

| *Diagnosis* | *Number of patients* | *Echocardiography alone (%)* | |
|---|---|---|---|
| | | *First surgery* | *Repeat surgery* |
| **Acyanotic CHD** | 179 (49%) | 84% | 36% |
| Left to right shunt | 110 | 86% | 33% |
| Outflow tract obstruction | 58 | 81% | 40% |
| Miscellaneous | 11 | 83% | 33% |
| **Cyanotic CHD** | 184(51%) | 62% | 22% |
| TGA | 38 | 61% | 30% |
| TOF | 43 | 61% | 43% |
| Tri Atresia | 10 | 29% | 0% |
| PA | 35 | 50% | 41% |
| TRUNCUS | 10 | 86% | 67% |
| DORV | 15 | 63% | 14% |
| HLHS | 18 | 100% | 17% |
| Other* | 16 | 58% | 25% |

TGA=transposition of the great arteries; TOF= Tetralogy of Fallot; Tri Atresia=tricuspid atresia; PA=pulmonary atresia; Truncus=truncus arteriosus; DORV=double outlet right ventricle;HLHS=hypoplastic left heart syndrome.
*Other includes: double inlet left ventricle (DILV), total anomalous pulmonary venous return (TAPVR), single ventricle, Ebstein's anomaly.

**Table 3.** Diagnostic Discrepancies Between Echocardiography and Surgical Findings

| *Echocardiography* | *CATH* | *Surgical findings* |
|---|---|---|
| **Echocardiography alone (232 pts, 64%)** | | |
| *Major (1/232, 0.4%)* | | |
| TGA, right aortic arch | Not done | TGA, double aortic arch |
| *Minor (7/232, 3%)* | | |
| CAVC | Not done | CAVC with small secundum ASD |
| CAVC, Rastelli type A | Not done | CAVC, Rastelli type B |
| CAVC, TGA, mesocardia, common atria with atrial situs ambiguous | Not done | CAVC,TGA, atrial inversion |
| Status post CAVC, cleft mitral valve | Not done | States post CAVC, flail mitral valve |
| Subaortic stenosis | Not done | Subaortic stenosis, mild supravalve aortic stenosis |
| COA with hypoplastic aortic arch | Not done | COA with hypoplastic aortic arch, PDA |
| TOF, hypoplastic branch pulmonary arteries | Not done | TOF, hypoplastic branch pulmonary arteries small aortopulmonary collateral artery |
| **Echocardiography plus catheterization (131 pts, 36%)** | | |
| *Major (2/131, 0.4%)* | | |
| Retroesophageal subclavian artery | PA sling | PA sling |
| Status post COA balloon dilation no aneurysm seen | Aneurysm at COA site | Aneurysm and dissection at balloon site |
| *Minor (9/131, 6.8%)* | | |
| Status post PA banding for TGA with normal coronary artery | Single CA | Yacob B CA |
| DORV, DCRV, malposition of great arteries, branch PA stenosis | DORV, DILV, L-malposition of great arteries | DILV,hypoplastic subaortic chamber, ASD, mild PS, L-malposition of great arteries |
| L-malposition of great arteries, ventricular inversion, COA | L-TGA | L-TGA |
| Corrected TGA, PS, LPA not seen | LPA stenosis | LPA stenosis |
| TOF, D-TGA, CAVC, DILV | DILV, PA | D-TGA, CAVC |
| D-TGA, DORV, COA coronary arteries not defined | D-TGA,COA, right CA from LAD | D-TGA, DORV, COA,Yacoub E coronaries |
| PA, single RV, TAPVR, RPA stenosis | PA, single RV, TAPVR RPA stenosis | PA, single RV, PAPVR, RPA stenosis |
| CAVC | CAVC, secundum ASD | CAVC, secundum ASD, PDA |
| Complex dextrocardia, unable to see branch pulmonary artery | Branch PA stenosis | Left pulmonary artery stenosis, small PDA |

CAVC=complete atrioventricular canal defect; ASD=atrial septal defect; COA=coarctation of aorta; PDA=patent ductus arteriosus; PA=pulmonary artery; CA=coronary artery; DCRV=double chamber right ventricle; RV =right ventricle; PAPVR= partial anomalous pulmonary venous return; PS=pulmonary stenosis; LAD=left anterior descending coronary; RPA=right pulmonary artery; LPA= left pulmonary artery; other abbreviations as in Table 2.

Of the 285 patients undergoing their initial surgical procedure, 212 (74%) had surgery based on echocardiography alone with 1 major (0.4%) and 5 minor (1.8%) diagnostic discrepancies. Of 78 patients undergoing repeat surgery, 22 (28%) had surgery based on echocardiography alone with no major and 6 minor (7.7%) diagnostic discrepancies. There was a statistically significant difference in echocardiography alone rates between patients undergoing their initial surgery and those undergoing repeat surgery (74% vs. 28%, $P<0.01$). Eighty percent of first time surgeries were definitive, compared to 64% of repeat surgeries ($p<0.05$). Of the 277/363 patients undergoing definitive surgeries, 189 (68%) had preoperative diagnosis by echocardiography alone with 1 major (0.5%) and 4 minor (2.1%) diagnostic discrepancies.

## Discussion

Transthoracic echocardiography can provide detailed information on cardiac structure, function and hemodynamics even in complex cardiac malformations. Increasing numbers of patients, including those with complex structural disease, undergo surgery for congenital heart disease based on echocardiographic data alone. As our experience shows, the majority of the patients with congenital heart disease requiring intervention can undergo cardiac surgery, both palliative and definitive, safely and successfully based on echocardiographic data alone. This appears to be particularly true in the neonatal population, a group with typically very good acoustic windows, but at higher risk for complications due to cardiac catheterization. Additional noninvasive imaging modalities not evaluated in the current study, such as magnetic resonance imaging (MRI) and intraoperative techniques such as transesophageal echocardiography, can also provide additional diagnostic information in questionable cases, thus obviating the need for cardiac catheterization in all but a select subset of patients. Eliminating unnecessary diagnostic catheterization is particularly important in preserving vascular access in patients who will be undergoing interventional procedures in the future.

In our patients, all three major diagnostic discrepancies involved the aortic arch or branch pulmonary arteries (double aortic arch, pulmonary artery sling, and aneurysm and dissection at coarctation site), all of which may be difficult to accurately diagnose by echocardiography, although previously published reports showed that these diagnoses can be made by echocardiography [9-12] and certainly by MRI [13]. Review of the data for these 3 patients suggests that the primary reasons for missing these diagnoses includes inexperience of the staff interpreter and/or technically limited acoustic windows. The 16 minor diagnostic discrepancies identified included coronary artery abnormalities, missed small secundum ASD and PDA, pulmonary artery anomalies including branch pulmonary artery stenosis and small aortopulmonary collateral artery, complete atrioventricular Rastelli type, flail mitral valve leaflet and supra-valve aortic stenosis. Most of these errors are also avoidable and/or not critical for surgery; however the presence and anatomy of aortopulmonary collateral arteries, anomalies of the distal branch pulmonary arteries and coronary arteries remain both important for operation, as well as sometimes difficult to define by transthoracic echocardiography. Previous published studies have found similar type and frequency of missed diagnoses [14-16]. Again, alternate non-invasive and/or intraoperative imaging strategies can often overcome the limitations of transthoracic echocardiography in the problem areas noted above.

The limitations of this study include its retrospective nature and limitations in defining diagnostic discrepancies based on operative findings as recorded in the operative note only.

In summary, echocardiography alone for pre-operative diagnosis of congenital heart disease is reliable and accurate in the vast majority of patients. Care should be taken to ensure complete evaluation of the cardiac anatomy in patients in whom surgery is contemplated based on echocardiographic data alone. The limitations imposed by less-than-ideal acoustic windows also must be recognized, so that additional imaging modalities (MRI and/or catheterization) can be applied judiciously as needed in selected patients. In addition, a detailed review of the echocardiographic data by an experienced pediatric echocardiographer is recommended for any patient undergoing surgery based on this data alone.

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