Original Article

The Role of Echocardiography as an Isolated Method for Indicating Surgery in Patients With Congenital Heart Disease

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Objective

To determine the diagnostic accuracy of echocardiography for indicating surgery without catheterization in patients with congenital heart disease through a prospective analysis and comparison of the echocardiographic diagnosis with the intraoperative findings, or invasive study, or both.

Methods

From February 2000 to January 2001, 493 patients with congenital heart diseases indicated for surgery were followed up. They underwent echocardiography with color-flow mapping for a therapeutic decision. The results were compared with the findings of surgery or catheterization, or both, when the latter were performed for diagnostic reasons.

Results

Of the patients studied, 94.3% (465 cases) underwent congenital heart disease correction based only on echocardiographic findings, without a diagnostic catheterization. The invasive study was performed for diagnostic reasons in 28 (5.6%) patients, the surgical treatment was performed in more than 95% of the patients, and therapeutic catheterization was performed in 3.6%. The echocardiographic findings were confirmed in 464 (94.1%) patients, which showed the high accuracy of the method. False-positive findings occurred in 8 (1.6%) patients, and false-negative findings in 39 (7.9%). According to surgeons, no diagnostic error led to complications or adversely affected the surgical results.

Conclusion

Echocardiography proved to be a sensitive and safe method for indicating surgery, making catheterization, often, unnecessary. The invasive study was restricted to diagnostic or therapeutic uses.

Key words

echocardiography, noninvasive diagnosis, congenital heart disease

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The preoperative diagnosis of congenital heart disease from both the functional and anatomical points of view should be performed to provide enough data to allow the surgeon to plan the surgical approach, avoiding unpleasant surprises during the procedure. Some years ago, adequate assessment was only possible by use of cardiac catheterization, an invasive method with greater risk for patients, mainly children; in addition, several complications related to that procedure have been reported [1-5].

Advances in the field of imaging have allowed the development of increasingly more sophisticated devices that have improved the quality of the images obtained, allowing a detailed assessment of the heart and great vessels. In several services, echocardiography has become the first-line approach for preoperative study, catheterization being reserved for diagnostic backup. However in the literature, controversy still exists about the indication for surgery with no invasive study for all patients with complex cardiac defects [6-15], an approach adopted by some institutions only for the simple defects, such as isolated lesions with intracardiac shunt and heart diseases with single ventricle physiology that require palliative surgery.

This study aimed at analyzing the accuracy of echocardiography in the preoperative assessment of patients with congenital heart diseases, by comparing the echocardiographic diagnosis with intraoperative or catheterization findings, or both.

Methods

From February 2000 to January 2001, of 2,004 patients studied through echocardiography with color-flow mapping in our service, 493 were indicated for corrective or palliative surgical treatment and were prospectively followed up during the in-hospital phase. The results of preoperative echocardiography were compared with the intraoperative anatomic description developed by the attending surgeon. Patients with previous cardiac surgery were excluded from the study.

The preoperative investigation comprised 2-dimensional echocardiography with analysis of intracardiac flows by using pulsed and continuous Doppler and color-flow mapping. The following devices were used for performing the echocardiographic examinations: HDI 5000 (Advanced Technology Laboratories, Bothell, WA, USA) and Toshiba Powervision 6000 and 140 (Toshiba, Tokyo, Japan) with 2.5-, 5.0-, and 7.5-MHz transducers. In pediatric

patients, sedation, when necessary, was performed through the oral or rectal route with 8% chloral hydrate at the dosages recommended for the pediatric age group.

The transthoracic echocardiographic study was performed by pediatric cardiologists and comprised the following: evaluation of the thoracic and abdominal situs; determination of the presence of 2 atria and 2 ventricles, superior vena cava, inferior vena cava, pulmonary veins, and their respective drainage sites; assessment of the integrity of the atrial and ventricular septa, of the morphology and location of the atrioventricular valves, right and left ventricular outflow tract, arterial ventricular connection, morphology of the pulmonary artery and branches, and morphology of the aortic arch. Doppler and color-flow mapping were used to assess the flows in the pulmonary artery, its branches, and the aorta, as well as the intracavitary and transvalvular flows, in addition to researching septal defects. Our study did not aim at a comparison with transesophageal echocardiography as a diagnostic method, although that examination was used as a complementary technique in adult patients with atrial septal defects and an inadequate transthoracic echocardiographic window.

Catheterization was performed when echocardiography did not clarify all anatomic details of the heart disease and of diagnostic complementation or when discordance existed between the clinical and echocardiographic findings. The adult patients aged < 40 years underwent surgery without catheterization when no doubts existed on echocardiography.

The patients were analyzed in regard to the distribution of their congenital heart diseases and classified as having an isolated lesion (only one defect) or as having a major lesion (associated with 2 or more defects). In the cases of associated lesions, the major lesion was established according to the greatest severity or hemodynamic repercussion.

Accuracy of the transthoracic echocardiography was determined by comparing the data obtained on the preoperative echocardiography and the findings of the surgery and catheterization, when the latter was performed. The divergences found were divided into false-positive (when the echocardiogram showed a finding not confirmed on surgery or catheterization, or both) and false-negative (when the echocardiogram did not show a feature observed on catheterization or surgery, or both). The errors were subdivided into major errors, when they led to a significant modification in the surgical procedure or affected the surgical risk or postoperative evolution, and minor errors, when the preceding aspects were not observed.

For the description of quantitative variables, the median and standard deviation were used; for qualitative variables, the absolute and relative frequencies were used, and the values were expressed as proportions. For comparing the groups, the Student *t* test was used in the independent continuous samples. For the samples expressed as proportions, the chi-square test with Yates correction and the Fisher exact test were chosen. The results whose descriptive level (P) was lower than 0.05 were considered statistically significant.

Results

Of the 493 patients prospectively followed up during the in-hospital phase, 447 (90.6%) underwent cardiac surgery without catheterization, and 46 required invasive study. The patient's ages ranged from one day to 64 years (mean, 5 years and 110 days; median, 365 days). The female sex accounted for 48% of the sample and the male sex for 52%.

The echocardiographic finding was confirmed neither on surgery, nor on catheterization in 8 patients (false-positive, 1.6%) as follows: 1) 3 patients had atrial septal defect: one patient had an associated significant enlargement in the right chambers and an increase in tricuspid regurgitation due to valvular dysplasia, and 2 patients had associated partial anomalous pulmonary venous drainage; 2) 2 patients had systemic-pulmonary collaterals in tetralogy of Fallot; 3) one patient had mild pulmonary valvular stenosis; 4) one patient had type I truncus arteriosus comunis; and 5) one patient had pulmonary atresia with a ventricular septal defect.

In 39 patients, the echocardiogram did not show some aspect observed on catheterization or surgery, or both, (false-negative, 7.9%), persistence of the ductus arteriosus (18) being the most common in diseases with pulmonary hypertension or increased blood flow, such as ventricular septal defect, coarctation of the aorta, and atrioventricular septal defect. The following were not also diagnosed: coronary artery anomaly (4); persistence of the left superior vena cava (4); anomalous pulmonary venous drainage (3); coarctation of the aorta (2); pulmonary atresia (2); pulmonary valvular stenosis (1); aortic supravalvular stenosis (1); aortic subvalvular stenosis (1); anomalous right ventricular band (1); double-outlet right ventricle (1); and presence of systemic-pulmonary collaterals (1). The comparison of the false-positive results in simple and complex diseases was not statistically significant, but that of the false-negative results was statistically significant ($P < 0.004$). Table I shows the totally correct results, the false-positive and false-negative results related to each heart disease, as well as the number of patients in whom catheterization was performed in each disease. Table II shows the false-negative and false-positive results (discrepancies) identified for each type of congenital heart disease diagnosed on echocardiography, both in the isolated and associated form.

The accuracy of the echocardiographic diagnosis was determined by comparison with the results of catheterization, surgical or autopsy findings, or both. The echocardiographic diagnosis was completely correct in 464 (94.1%) patients. The accuracy rate for the major associated lesions on the echocardiogram was greater than that for the lesions considered in isolation, and this difference was statistically significant ($P < 0.001$). In our hospital, the accuracy rate of the method was 94.1%. According to the surgeons, the diagnostic errors neither led to complications nor had an adverse effect on the surgical results.

Diagnostic catheterization was performed in 28 (5.6%) patients and complemented the investigation more frequently in more complex diseases; however, no statistical significance was observed when that frequency was compared with the number of times catheterization was performed in less complex diseases, such as atrial septal defects, for assessing pulmonary pressure (5 cases); for assessing pulmonary pressure and the presence of coronary heart diseases in adults > 40 years (2 cases); and due to age > 40 years (2 cases). Systolic pressure in the pulmonary artery in these patients ranged from 60 mmHg to 100 mmHg.

Eighteen (3.6%) patients underwent invasive hemodynamic procedures with therapeutic finality as follows: Amplatzer prosthesis

placement in atrial septal defect (2); coil placement in persistent ductus arteriosus (1); and pulmonary valvuloplasty (7). In 8 patients, balloon catheterization was performed for the atrial septostomy procedure [transposition of the great arteries (4), tricuspid atresia (2), pulmonary atresia with intact ventricular septum (1), and tetralogy of Fallot with pulmonary atresia (1)].

Discussion

In this study, the echocardiogram proved to be highly reliable as a method of preoperative assessment of patients with congenital heart disease undergoing their first surgical procedure. The sensitivity and specificity of the echocardiographic diagnosis in the preoperative assessment of those patients has already been investigated in some centers of cardiac surgery and echocardiography, and similarly to the findings in our study, a significant concordance was observed between the echocardiographic diagnosis and the surgical findings or the findings of catheterization, when performed, or both [16-18]. However, most studies cited in the literature do not separately consider the patients undergoing the surgical procedure first, including also the patients undergoing surgery for correction of the univentricular type, in which catheterization is frequently required for preoperative assessment of pulmonary pressure.

Catheterization has been progressively restricted to therapeutic use, and several studies in the literature have confirmed satisfactory results with fewer risks for the patients with closure of atrial septal defects, patent ductus arteriosus, dilation of the pulmonary valve, coarctation of the aorta, and recoarctation of the aorta [19-21]. Jarrar et al [22] reported a rate of pulmonary restenosis after a 10-year follow-up of only 4.8% in patients undergoing valvuloplasty with a balloon catheter. Diagnostic catheterization is more frequently performed in patients with complex heart diseases, who have several associated lesions [9,15].

Concomitantly with the reduced number of diagnostic catheterization, we observed a low incidence of flaws in the echocardiographic diagnosis. The most frequent causes of diagnostic error or incomplete diagnosis occurred due to the lack of adequate detection of the extracardiac structures, mainly the presence of patent ductus arteriosus in patients with ventricular septal defect, atrioventricular septal defect, and coarctation of the aorta. This has already been described by D'Orsogna et al [23] and may be due to the difficulty in assessing the extracardiac structures on echocardiography, because such structures have similar echodensities.

In the cases of ductus arteriosus, the low flow between the aorta and the pulmonary trunk occurs due to equalization of the pressures between those chambers due to pulmonary hypertension or increased flow, which may hinder its visualization. Because the suprasternal section is mandatory in routine echocardiographic examination of congenital heart diseases, the assessment of extracardiac structures through that window is known to be difficult, mainly in small children. That limitation, undoubtedly contributes to the incomplete diagnosis in patients with persistent left superior vena cava, mainly when no dilation of the coronary sinus is observed, which may suggest anomalous systemic venous drainage.

Table I - Distribution of the congenital heart diseases according to the echocardiographic diagnosis

Lesion (N)	Echo diagnosis isolated lesion			Isolated (N)	Echo diagnosis major lesion			Major (N) Echo + cathet	
	C	F+	F-		C	F+	F-		
ASD (79)	52	1	7	60	18	1	—	19	9
VSD (96)	50	—	13	63	33	—	—	33	1
PDA (35)	34	—	1	35	—	—	—	—	1
TAVSD (23)	13	—	2	15	8	—	—	8	1
PAVSD (10)	7	—	—	7	3	—	—	3	—
TAPVD (4)	4	—	—	4	—	—	—	—	—
PAPVD (8)	6	—	—	6	1	1	—	2	1
Anom Coron aa. (5)	5	—	—	5	—	—	—	—	3
AoS (13)	8	—	2	10	3	—	—	3	—
CoAo (29)	6	—	3	9	20	—	—	20	1
Interrup. (3)	—	—	—	—	3	—	—	3	—
HLHS (11)	10	—	—	10	1	—	—	1	—
HRHS/TA (17)	14	1	1	16	1	—	—	1	5
Ebstein (3)	3	—	—	3	—	—	—	—	—
PS (14)	10	1	—	11	3	—	—	3	7
PA w/ intact VS (7)	6	—	—	6	1	—	—	1	2
T4F (72)	34	2	8	44	27	—	1	28	5
TGA (27)	18	—	—	18	9	—	—	9	4
CTGA (2)	1	—	—	1	1	—	—	1	—
Truncus (8)	7	1	—	8	—	—	—	—	1
DVSVD (5)	1	—	—	1	4	—	—	4	1
SV (8)	2	—	—	2	6	—	—	6	1
Complex (5)	4	—	1	5	—	—	—	—	2
Miscellaneous (9)	9	—	—	9	—	—	—	—	1
Total (493)	304	6	38	348	142	2	1	145	46

C - confirmed; F+ - false-positive (when the echocardiographic finding was confirmed neither on surgery nor on catheterization); F - false-negative (when the echocardiogram did not show a finding observed on catheterization or on surgery, or both); ASD - atrial septal defect; VSD - ventricular septal defect; PDA - persistent ductus arteriosus; TAVSD - total atrioventricular septal defect; PAVSD - partial atrioventricular septal defect; TAPVD - total anomalous pulmonary venous drainage; PAPVD - partial anomalous pulmonary venous drainage; Anom Coron aa. - anomalous coronary arteries; AoS - aortic stenosis; CoAo - coarctation of the aorta; Interrup. - interruption of the aortic arch; HLHS - hypoplastic left heart syndrome; HRHS/TA - hypoplastic right heart syndrome or tricuspid atresia, or both; PS - pulmonary stenosis; PA w/ intact VS - pulmonary atresia with intact ventricular septum; T4F - tetralogy of Fallot; TGA - transposition of the great arteries; CTGA - corrected transposition of the great arteries; DORV - double-outlet right ventricle; SV - single ventricle.

Table II - Distribution of the isolated and major defects and their respective discrepancies

Isolated/major lesion (N)	False negative	False positive
ASD (79)	PVS (1), TAPVD (2), PAPVD (1), Supra AoVS (1), LSVC (2)	PVS (1)
VSD (96)	PDA (12), RV band (1)	PVS (1)
PDA (35)	CoAo (1)	—
TAVSD (23)	LSVC (1), PDA (1)	—
PAVSD (10)	—	—
TAPVD (4)	—	—
PAPVD (8)	—	ASD (1)
Anom Coron aa. (5)	—	—
Coronárias (5)		
AoS (v,sub,supra) (13)	PDA (1), CoAo (1)	—
CoAo (29)	PDA (3)	—
Interrup (3)	—	—
HLHS (11)	—	—
HRHS (17)	Collaterals (1)	CAT+collat (1)
Ebstein (3)	—	—
PS (v,sub,supra) (14)	—	supra plum stenosis (1)
PA com septo íntegro (7)	—	—
T4F (72)	PDA (1), PA (2), coronary anomaly (4), DORV (1), subAo stenosis (1)	Collaterals (2)
TGA (27)	—	—
CTGA (2)	—	—
Truncus (8)	—	Classification-Type I (1)
DORV (5)	—	—
SV (8)	—	—
Complex (5)	LSVC (1)	—
Miscellaneous (9)	—	—

ASD - atrial septal defect; PVS - pulmonary valve stenosis; TAPVD - total anomalous pulmonary venous drainage; PAPVD - partial anomalous pulmonary venous drainage; supra AoV S - supra aortic valve stenosis; LSVC - persistent left superior vena cava; VSD - ventricular septal defect; PDA - persistent ductus arteriosus; RV- right ventricle; CoAo - coarctation of the aorta; TAVSD - total atrioventricular septal defect; PAVSD - partial atrioventricular septal defect; AoS (v, sub, supra) - aortic stenosis (valvular, subvalvular, supravalvular); HLHS - hypoplastic left heart syndrome; HRHS - hypoplastic right heart syndrome; PS - pulmonary stenosis; PA - pulmonary atresia; T4F - tetralogy of Fallot; DORV - double-outlet right ventricle; TGA - transposition of the great arteries; CTGA - corrected transposition of the great arteries; SV - single ventricle; CAT - common arterial trunk.

We found some false-positive and false-negative results in the assessment of valvular stenoses in heart diseases with increased pulmonary flow, such as atrial and ventricular septal defects. Those findings were considered secondary to an over- or underestimated assessment of the relative valvular stenosis due to increased pulmonary flow. In these cases, we suggest a more detailed evaluation of the valvular morphology for the correct differential diagnosis, as well as the analysis of the sub- and postvalvular pulmonary flow, which will appear with different velocities.

Assessment of the pulmonary valve may also be hindered in the cases of tetralogy of Fallot with a significant obstruction of the right ventricular outflow tract, in which the pulmonary valve is usually very hypoplastic [24]. In that case, the analysis with the pulsed Doppler located right after the pulmonary valve could help to differentiate the functional pulmonary atresia from the anatomic pulmonary atresia. Still regarding tetralogy of Fallot, the presence of subaortic stenosis, as an associated lesion, has already been reported by Snider et al [25], and it may be present as a thin membrane or fibromuscular ring. However, due to the presence of a ventricular septal defect, the transvalvular gradient may not be perceived on Doppler, even when very significant, and the 2-dimensional mode assessment acquires extreme importance.

To conclude the discussion about the false-negative and false-positive results found in tetralogy of Fallot, the literature is still controversial about the differentiation between tetralogy of Fallot and double-outlet right ventricle [26]. Some authors consider the presence of complete bilateral double infundibulum essential for the diagnosis of double-outlet right ventricle. We agree with Tynan et al [27], who consider that that criterion hinders the diagnosis of double-outlet right ventricle with tetralogy of Fallot. They chose to define as double-outlet right ventricle the cases in which more than 50% of the circumference of the aorta relates to the right ventricle, independently of the nature of the structures that support it [28]. Thus, some examples of double-outlet right ventricle also have tetralogy of Fallot. We believe that that differentiation is interesting for surgical planning without invasive study, because the extension of the patch used to close a ventricular septal defect is greater in the cases of double-outlet right ventricle of the "Fallot type," with a greater risk of postoperative obstruction of the left ventricular outflow tract. Knowing that the long parasternal axis is the most recommended for assessing the degree of aortic override, one patient had double-outlet right ventricle of the "Fallot type" interpreted as classic tetralogy of Fallot. The patient had a significant chest deformity that did not allow the adequate performance of that section and the correct alignment in regard to the septum, underestimating its override.

Assessment of the coronary arteries proved to be adequate on echocardiography, because, in 27 patients with transposition of the great arteries, no coronary anomaly was missed, and, in 72 patients with tetralogy of Fallot, only 4 (5.5%) false-negative results occurred. We recommend the systematic assessment of the coronary arteries during the echocardiographic examination, including the use of color Doppler, because that anomaly may be present in up to 31% of the patients with tetralogy of Fallot, whose most common form is represented by a dilated right coronary artery (indirect signal) from which the anterior descending artery originates, crossing anteriorly the right ventricular outflow tract [29]. In the 4 patients with false-negative results in our case series, the diameter of the right coronary artery was normal, and that valuable indirect signal that would raise that hypothesis was not observed.

A false-positive and a false-negative diagnosis occurred in the patients with truncus arteriosus. In the false-positive case, the pulmonary artery was not defined because it was very hypoplastic, and the pulmonary arteries seemed to originate directly from the aorta, considered a common arterial trunk. During the surgical act, pulmonary atresia with important hypoplasia of the proximal pulmonary artery and ventricular septal defect were observed. In the false-negative case, an error occurred in the classification of a type III truncus, which was interpreted as type I. The intraoperative findings showed 2 individualized pulmonary arteries with a long left pulmonary artery and a much smaller right pulmonary artery. The error may be explained by the fact that the right pulmonary artery was not visualized, and the long left branch was interpreted as the pulmonary trunk. That difficulty occurs in type III truncus because the pulmonary arteries are not usually visualized together in the same parasternal plane, the origin of one pulmonary

artery being seen in the short parasternal axis and the origin of the other pulmonary artery being more difficult to observe in the suprasternal plane [30].

The diagnostic impression of atrial septal defects may often occur in adult patients due to the difficulty of the echocardiographic window, because the atrial septum is located very far away from the transducer, resulting in lack of definition of the tissue of the oval foramen, which is much thinner than the rest of the septum. Two of the false-positive results in our case series occurred in association with partial anomalous pulmonary venous drainage, which may be explained by the fact that most partial drainages are associated with atrial septal defect, which may have influenced the echocardiographer when interpreting the dubious image. The third unconfirmed diagnosis of atrial septal defect occurred in a young female patient who had an image suggestive of septal discontinuity associated with important tricuspid regurgitation and right chamber dilation. The surgery revealed isolated tricuspid valve dysplasia corrected with valvuloplasty and reduction of the valvular ring.

In a patient with ventricular septal defect, an anomalous right ventricular band was not identified, being interpreted as dynamic reactive hypertrophy, also known as jet lesion.

The results of this study showed that the surgery could be performed without previous invasive study, even in more complex cases, because the echocardiography proved to be a highly sensitive and specific method. The technological evolution has allowed a constant improvement in the quality of the echocardiographic image and increasing reliability as a diagnostic preoperative tool, avoiding the performance of cardiac catheterization, which increases the risks and costs for the patient and health system. Based on these findings, we concluded that echocardiography may be used as a safe and sensitive method for the preoperative assessment of patients with congenital heart diseases, catheterization being reserved for cases with specific indications.

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