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

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 recom-

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 trans-}

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
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 sensi-
tivity and specificity of the echocardiographic diagnosis in the
preoperative assessment of those patients has already been inves-
tigated 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 thera-
peutic 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 di-
seases, who have several associated lesions [9,15].

Concomitantly with the reduced number of diagnostic cathe-
terization, we observed a low incidence of flaws in the echocar-
diographic 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'Orosogna et al [23] and may be due
to the difficulty in assessing the extracardiac structures on echo-
cardiography, 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 ex-
tracardiac 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 super-
ior vena cava, mainly when no dilation of the coronary sinus is
observed, which may suggest anomalous systemic venous drainage.

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C - confirmed; F+ - false-positive (when the echocardiogram 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; PAUSD - 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.
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 sub- and postvalvular pulmonary flow. In these cases, we suggest a more detailed evaluation of the relative valvular stenosis due to 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 sub- and postvalvular pulmonary flow.

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. 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., 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. 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., 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. 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. 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...
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 valvar 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.

References