J Korean Med Sci 2002; 17: 463-7
ISSN 1011-8934

Pediatric Cardiac Surgery with Echocardiographic Diagnosis Alone

The diagnostic accuracy of echocardiography alone and the safety of cardiac surgery using this diagnostic approach were retrospectively assessed in 111 children operated for congenital heart defects (CHD) during a 3.5-yr period ending in October 2001. Preoperative diagnosis was compared with the intraoperative findings obtained by surgical inspection. Perioperative death was defined as death within 30 days postoperatively. Of the patients, 70% were operated on in infancy. Seventy-six percent (84 of 111) underwent surgery after echocardiographic diagnosis alone. A high percentage of patients with patent ductus arteriosus (100%), partial atrioventricular canal (100%), coarctation of the aorta (89%), ventricular septal defect (86%), atrial septal defect (85%), and total anomalous pulmonary venous connection (75%) was operated without prior catheterization. Diagnostic errors occurred in 2.4% (2 of 84) of patients with echocardiography only and in 7.4% of patients with catheterization. No error in either group was related to surgical morbidity or mortality. There were five (6.0%) perioperative deaths in the echocardiography group and two (7.4%) in the catheterization group, with no difference in the mortality between the groups. In conclusion, many patients with CHD can be accurately diagnosed by echocardiography alone, and can safely undergo surgery without catheterization, not increasing the overall risk.

Key Words : *Echocardiography; Heart Defects, Congenital; Thoracic Surgery*

Sejung Sohn, Hae Soon Kim, Jae Jin Han*

Departments of Pediatrics and Pediatric Cardiac Surgery*, Ewha Womans University College of Medicine and Ewha University Mokdong Hospital, Seoul, Korea

Received : 5 February 2002
Accepted : 2 April 2002

Address for correspondence
Sejung Sohn, M.D.
Department of Pediatrics, Ewha Womans University Mokdong Hospital, 911-1 Mok-dong, Yangchon-gu, Seoul 158-710, Korea
Tel : +82.2-650-5579, Fax : +82.2-653-3718
E-mail : sohn@ewha.ac.kr

INTRODUCTION

Accurate preoperative morphologic and functional diagnosis is of vital importance for surgical repair of congenital heart defects (CHD). Before the era of echocardiography, only cardiac catheterization and angiography made it possible to derive adequate information for cardiac repair. However, cardiac catheterization, especially in neonates and small infants, carries a risk. Recent technical advances in echocardiography provides the information needed for a safe cardiac surgery in many patients without invasive catheterization (1-6). Initially, this exclusively echocardiography-based diagnostic approach has been applied to simple cardiac defects (7-10) and functional single ventricle defects requring palliative surgery (11, 12); in recent years, even complex lesions were proven to be adequately defined for surgical repair by non-invasive echocardiographic evaluation alone (13-18).

Accordingly, we intended to evaluate our institution's approach regarding non-invasive preoperative diagnostic work-up based on echocardiography alone in patients undergoing cardiac surgery. The purpose of this study was to determine the accuracy of preoperative diagnosis by echocardiography alone, and to assess the safety of cardiac surgery using this diagnostic approach in children with CHD.

MATERIALS AND METHODS

We reviewed retrospectively the records of all pediatric patients who underwent either palliative or corrective cardiac surgery at our institution during a period of 3.5 yr ending in October 2001. Included were all children undergoing open heart surgery as well as patients with ductal closure, coarctation repair, and palliation with a Blalock-Taussig shunt. Excluded were patients >15 yr of age who underwent corrective repair of CHD, and patients undergoing banding of a pulmonary artery.

All patients had undergone comprehensive preoperative evaluation, including physical examination, 12-lead electrocardiogram, and two-dimensional echocardiography with spectral and color-flow Doppler. Echocardiograms were done by one of two cardiologists and repeated by the other for cross-check when echocardiography alone was thought as sufficient to define the cardiac lesions for surgical treatment. Otherwise cardiac catheterization was performed for further detailed delineation of the cardiac lesions. The decision to perform a diagnostic catheterization was made by the attending cardiologist.

For the purpose of analysis, the patients were classified into two groups: those undergoing preoperative echocardiography

alone (echo group) and those who had preoperative cardiac catheterization in addition to echocardiography (cath group). For the diagnostic classification, when there were two or more separate defects, they were classified according to the more significant and severe defect.

Diagnostic accuracy was assessed by comparing the preoperative diagnosis made by echocardiography and/or catheterization with the intraoperative findings obtained by surgical inspection.

Perioperative mortality was defined as death within the first 30 postoperative days or before hospital discharge.

Statistics

Comparisons between the groups were assessed by chi-square tests. A p value of <0.05 was considered significant. Data were analyzed with SPSS for Windows V10.0 (SPSS Inc., Chicago, IL, U.S.A.).

RESULTS

During the study period, a total of 111 pediatric patients underwent either palliative or corrective cardiac surgery at our institution. The patients ranged in age from 2 days to 15 yr (median, 4 months). Of the patients 37 (33%) were neonates and 41 (37%) were in their first year of life. Table 1 shows the distribution of cardiac defects in the study population, with the respective proportions of children who underwent preoperative echocardiography alone.

Table 1. Distribution of cardiac defects and respective proportions of patients with echocardiographic diagnosis

Diagnosis	Total No. of Patients	No. of Infants (<1 yr)	Echo only n (%)	No. of Children (>1 yr)	Echo only n (%)
ASD	13	2	2 (100)	11	9 (82)
PDA	26	22	22 (100)	4	4 (100)
VSD	22	17	14 (82)	5	5 (100)
CAVSD	2	1	–	1	–
PAVSD	3	1	1 (100)	2	2 (100)
COA	9	8	8 (100)	1	–
TAPVR	4	4	3 (75)	–	–
TGA	3	3	1 (33)	–	–
TOF	5	2	–	3	–
DORV	6	5	3 (60)	1	–
BT shunt	9	8	6 (75)	1	1 (100)
Others*	9	5	3 (60)	4	–
Total	111	78	63 (81)	33	21 (64)

ASD, atrial septal defect; BT, Blalock-Taussig; CAVSD, complete atrioventricular septal defect; COA, coarctation of the aorta; DORV, double outlet right ventricle; PAVSD, partial atrioventricular septal defect; PDA, patent ductus arteriosus; TAPVR, total anomalous pulmonary venous return; TGA, transposition of the great arteries; TOF, tetalogy of Fallot; VSD, ventricular septal defect.
*Others include cor triatriatum (n=1), double outlet left ventricle (n=1), hypoplastic left heart syndrome (n=1), subaortic stenosis (n=1), tricuspid atresia (n=2), truncus arteriosus (n=2), and vascular ring (n=1).

Of 111 patients, 84 (76%) had echocardiography as the only diagnostic modality and the remaining 27 had both echocardiography and catheterization. In the group of neonates and infants (<1 yr of age), the proportion of patients diagnosed by echocardiography appeared to be higher than in the group of older children (81% vs 64%, p=0.09). A high percentage of patients with patent ductus arteriosus (100%), partial atrioventricular canal (100%), coarctation of the aorta (89%), ventricular septal defect (86%), atrial septal defect (85%), and total anomalous pulmonary venous connection (75%) was referred for surgery without catheterization.

Diagnostic errors were identified at the time of surgery in 2 of the 84 echo group patients (2.4%) and in 2 of the 27 cath group patients (7.4%), with no significant difference between the groups (p=0.53). No error in either group was directly related to surgical morbidity or mortality. No instances arose in which catheterization corrected an echocardiographic error in patients with either a single defect or a combination of defects. Errors are listed in Table 2. Of the four patients with transposition of the great arteries or Taus-

Table 2. Diagnostic errors

Diagnosis	Operative finding	Consequence
Echo group		
DORV (Taussig-Bing)	High take-off LCA	None
DORV (Taussig-Bing)	Dual LAD	None
Cath group		
CAVSD (unbalanced)	Single papillary muscle	None
Vascular ring	Ductal diverticulum Ligamentum arteriosum	None

LAD, left anterior descending artery; LCA, left coronary artery; other abbreviations as in Table 1.

Table 3. Cases with perioperative mortality

Diagnosis	Age at Operation	Name of Operation	Cause of Death
Echo group			
PA/IVS	2 days	BT shunt + Pulmonary valvotomy	DIC, Sepsis
HLHS	5 days	Norwood operation	Sepsis
TAPVR	17 days	Total correction	CPB weaning failure
DORV (Taussig-Bing)	6 days	Norwood operation	DIC, Sepsis
DORV (Taussig-Bing)	13 days	Jatene operation	Mediastinitis
Cath group			
CAVSD (unbalanced)	4 months	Biventricular repair	Low cardiac output
TOF	9 months	Total correction	Mediastinitis

CPB, cardiopulmonary bypass; DIC, disseminated intravascular coagulopathy; HLHS, hypoplastic left heart syndrome; PA/IVS, pulmonary atresia with intact ventricular septum; other abbreviations as in Table 1.

sig-Bing anomaly undergoing the arterial switch operation, the coronary pattern was correctly determined in three. In one patient with Taussig-Bing anomaly, repeat echocardiogram suggested the presence of an unusual coronary artery pattern, which was not detected at the first echocardiogram. Although the coronary anatomy was not definitely diagnosed, this suggestion was proven to be helpful for an uneventful operation. This coronary artery pattern proved dual left anterior descending artery. In a patient with vascular ring, echocardiogram and aortic angiogram showed a type of vascular ring such as right aortic arch with mirror-image branching accompanied with a ventricular septal defect, but missed left ligamentum arteriosum originating from the descending aorta as a large diverticulum.

There were seven perioperative deaths: five (6.0%) in the echo group, and two (7.4%) in the cath group. There was no difference in the perioperative mortality between the two groups (p=0.84). They are summarized in Table 3. In no case was the mortality affected by a diagnostic error. Deaths were mainly caused by sepsis or mediastinitis except in two cases: one patient, in the echo group, with obstructive total anomalous pulmonary venous drainage died from weaning failure of cardiac bypass. In the cath group, one patient with Down's syndrome was diagnosed as having right-dominant unbalanced atrioventricular canal, with the preoperative indexed potential left ventricular volume of 17 mL/m^2. Biventricular repair was attempted according to the suggestion that preoperative indexed potential left ventricular volume of >15 mL/m^2 allows biventricular repair (19), however, he succumbed to low cardiac output after operation.

DISCUSSION

There remains little debate that in simple shunt lesions (atrial septal defect, patent ductus arteriosus and ventricular septal defect) cardiac catheterization provides no advantage over echocardiography in defining the anatomy of these defects, and echocardiography is adequate as the only diagnostic modality (7-10). Cardiac catheterization might nowadays only be justified with the intention to close such shunts by interventional procedures. However, there is controversy over whether catheterization is routinely necessary for preoperative diagnosis of more complex lesions (1-6, 13-18, 20-23). Although certain complex defects such as functionally univentricular heart or pulmonary atresia with aortopulmonary collaterals invariably require preoperative catheterization, echocardiography alone may be sufficient to obtain the anatomic and functional information needed for surgical repair in patients with most major CHD. Indeed, owing in part to increasing confidence in echocardiography, the practice of cardiac surgery in patients with major lesions without catheterization is becoming increasingly common at many centers. As a result of this changing trends in the approach to preoperative diagnosis of CHD, 65 to 82% of patients undergo cardiac surgery after preoperative diagnosis by echocardiography alone (15-18).

This study demonstrates a high reliability of non-invasive preoperative diagnosis by echocardiography enables us to do cardiac surgery in a majority of patients without catheterization, not increasing the overall risk. Several studies have addressed this issue and similar conclusions have been drawn (15-18). Although this study had a relatively small number of total subjects compared to the others, the proportion of the group of neonates and infants in the study population was higher and nonetheless the outcome of cardiac surgery using this exclusively echocardiography-based diagnostic approach was comparable.

The incidence of diagnostic errors in this study was lower than in the published results (15-18), and they were considered of little importance as they did not affect the operative procedure or surgical outcome. It may be partially attributed to repeat echocardiographic examination for cross-check. As in our experience, repeat echocardiogram may offer the additional or missed findings not seen at the first study. Thus, cross-check examination is considered necessary to increase the accuracy of preoperative diagnosis by echocardiography alone.

Accurate preoperative evaluation of coronary anatomy is essential for a successful surgical repair in patients with certain forms of CHD, such as transposition of the great arteries or tetralogy of Fallot. In these patients, the intracardiac anatomy was adequately shown by echocardiography. The coronary anatomy could be shown with a high accuracy by echocardiography (24-26). In this study the coronary pattern was correctly predicted in most patients with transposition of the great arteries or Taussig-Bing anaomaly undergoing the arterial switch operation, although in one newborn, an unusual coronary anatomy was not diagnosed preoperatively. Nonetheless, because the coronary artery anatomy is identified at the time of surgery, and because the arterial switch procedure is performed in all forms of complex coronary anatomy at our institution, this error did not alter the surgical treatment. Our institutional practice is to define the coronary anatomy by echocardiography in patients with transposition of the great arteries. Catheterization is performed only for balloon septostomy, not for coronary angiography. If the echocardiographic diagnosis is equivocal, the surgeon is alerted and is prompted to carefully examine the coronary arteries.

Regarding the surgical mortality, it was not affected significantly by the method of diagnosis, but appeared to be highly related to postoperative infection in most cases.

There are some limitations in this study. As a retrospective nature, the patients were not randomly assigned to the two groups of diagnostic methods. In addition, the range of indication for catheterization within each group of defects was variable between the attending cardiologists, in some cases

reflecting physicians' different levels of comfort with echocardiographic diagnosis. Another limitation is a relatively small number of subjects studied with over half of them having simple defects, probably minimizing the diagnostic error of echocardiography. Finally, it may not be possible to generalize our results to other institutions. Each institution must have its own approach, as determined by physicians' experience and their consensus.

Despite the limitations, this study shows that many patients with CHD can be accurately diagnosed by echocardiography alone, and can safely undergo cardiac surgery without catheterization, not increasing the overall risk.

REFERENCES

1. Stark J, Smallhorn J, Huhta J, de Leval M, Macartney FJ, Rees PG, Taylor JFN. *Surgery for congenital heart defects diagnosed with cross-sectional echocardiography. Circulation 1983; 68(suppl II): 129-38.*
2. Huhta JC, Glasow P, Murphy DJ Jr, Gutgesell HP, Ott DA, McNamara DG, Smith EO. *Surgery without catherization for congenital heart defects: management of 100 patients. J Am Coll Cardiol 1987; 9: 823-9.*
3. Krabill KA, Ring WS, Foker JE, Braunlin EA, Einzig S, Berry JM, Bass JL. *Echocardiographic versus cardiac catheterization diagnosis of infants with congenital heart disease requiring cardiac surgery. Am J Cardiol 1987; 60: 351-4.*
4. Sreeram N, Colli AM, Monro JL, Shore DF, Lamb RK, Fong LV, Smyllie JH, Keeton BR, Sutherland GR. *Changing role of non-invasive investigation in the preoperative assessment of congenital heart disease: a nine year experience. Br Heart J 1990; 63: 345-9.*
5. Marino B, Corno A, Carotti A, Pasquini L, Giannico S, Guccione P, Bevilacqua M, de Simone G, Marcelletti C. *Pediatric cardiac surgery guided by echocardiography. Established indications and new trends. Scan J Thorac Cardiovasc Surg 1990; 24: 197-201.*
6. Sharma S, Anand R, Kanter KR, Williams WH, Dooley KJ, Jones DW, Vincent RN. *The usefulness of echocardiography in the surgical management of infants with congenital heart disease. Clin Cardiol 1992; 15: 891-7.*
7. Freed MD, Nadas AS, Norwood WI, Castaneda AR. *Is routine preoperative cardiac catheterization necessary before repair of secundum and sinus venosus atrial septal defects? J Am Coll Cardiol 1984; 4: 333-6.*
8. Shub C, Tajik AJ, Seward JB, Hagler DJ, Danielson GK. *Surgical repair of uncomplicated atrial septal defect without "routine" preoperative cardiac catheterization. J Am Coll Cardiol 1985; 6: 49-54.*
9. Lipshultz SE, Sanders SP, Mayer JE, Colan SD, Lock JE. *Are routine preoperative cardiac catheterization and angiography necessary before repair of ostium primum atrial septal defect? J Am Coll Cardiol 1988; 11: 373-8.*
10. Carotti A, Marino B, Bevilacqua M, Marcelletti C, Rossi E, Santoro G, de Simone G, Pasquini L. *Primary repair of isolated ventricular septal defect in infancy guided by echocardiography. Am J Cardiol 1997; 79: 1498-501.*
11. Ueda K, Nojima K, Saito A, Nakano H, Yokota M, Muraoka R. *Modified Blalock-Taussig shunt operation without cardiac catheterization: two-dimensional echocardiographic preoperative assessment in cyanotic infants. Am J Cardiol 1984; 54: 1296-9.*
12. Marino B, Corno A, Pasquini L, Guccione P, Carta MG, Ballerini L, de Simone G, Marcelletti C. *Indication for systemic-pulmonary artery shunts guided by two-dimensional and Doppler echocardiography: criteria for patients selection. Ann Thorac Surg 1987; 44: 495-8.*
13. Zellers TM, Zehr R, Weinstein E, Leonard S, Ring WS, Nikaidoh H. *Two-dimensional and Doppler echocardiography alone can adequately define preoperative anatomy and hemodynamic status before repair of complete atrioventricular septal defect in infants <1 year old. J Am Coll Cardiol 1994; 24: 1565-70.*
14. Santoro G, Marino B, Di Carlo D, Formigari R, de Zorzi A, Mazzera E, Rinelli G, Marcelletti C, de Simone G, Pasquini L. *Echocardiographically guided repair of tetralogy of Fallot. Am J Cardiol 1994; 73: 808-11.*
15. Marek J, Skovranek J, Hucin B, Chaloupecky V, Tax P, Reich O, Samanek M. *Seven-year experience of noninvasive preoperative diagnostics in children with congenital heart defects: comprehensive analysis of 2,788 consecutive patients. Cardiology 1995; 86: 488-95.*
16. Tworetzky W, McElhinney DB, Brook MM, Reddy VM, Hanley FL, Silverman NH. *Echocardiographic diagnosis alone for the complete repair of major congenital heart defects. J Am Coll Cardiol 1999; 33: 228-33.*
17. Pfammatter JP, Berdat PA, Carrel TP, Stocker FP. *Pediatric open heart operations without diagnostic cardiac catheterization. Ann Thorac Surg 1999; 68: 532-6.*
18. Pfammatter JP, Berdat P, Hammerli M, Carrel T. *Pediatric cardiac surgery after exclusively echocardiography-based diagnostic work-up. Int J Cardiol 2000; 74: 185-90.*
19. Van Son JA, Phoon CK, Silverman NH, Haas GS. *Predicting feasibility of biventricular repair of right-dominant unbalanced atrioventricular canal. Ann Thorac Surg 1997; 63: 1657-63.*
20. Van der Velde ME, Parness IA, Colan SD, Spevak PJ, Lock JE, Mayer JE Jr, Sanders SP. *Two-dimensional echocardiography in the pre- and postoperative management of totally anomalous pulmonary venous connection. J Am Coll Cardiol 1991; 18: 1746-51.*
21. George B, DiSessa TG, Williams R, Friedman WF, Laks H. *Coarctation repair without cardiac catheterization in infants. Am Heart J 1987; 114: 1421-5.*
22. Glasow PF, Huhta JC, Yoon GY, Murphy DJ Jr, Danford DA, Ott DA. *Surgery without angiography for neonates with aortic arch obstruction. Int J Cardiol 1988; 18: 417-25.*
23. Bash SE, Huhta JC, Vick GW III, Gutgesell HP, Ott DA. *Hypoplastic left heart syndrome: is echocardiography accurate enough to guide surgical palliation? J Am Coll Cardiol 1986; 7: 610-6.*
24. Pasquini L, Sanders SP, Parness IA, Wernovsky G, Mayer JE Jr, van der Velde ME, Spevak PJ, Colan SD. *Coronary echocardiography in 406 patients with d-loop transposition of the great arteries. J Am Coll Cardiol 1994; 24: 763-8.*
25. Pasquini L, Parness IA, Colan SD, Wernovsky G, Mayer JE, Sanders

SP. *Diagnosis of intramural coronary artery in transposition of the great arteries using two-dimensional echocardiography. Circulation 1993; 88: 1136-41.*

26. Need LR, Powell AJ, del Nido P, Geva T. *Coronary echocardiography in tetralogy of Fallot: diagnostic accuracy, resource utilization and surgical implications over 13 years. J Am Coll Cardiol 2000; 36: 1371-7.*