Noninvasive Quantitative Evaluation of the Morphology of the Major Pulmonary Artery Branches in Cyanotic Congenital Heart Disease
Angiocardiographic and Echocardiographic Correlative Study

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Background Precise noninvasive evaluation of pulmonary artery (PA) morphology is extremely important for medical and surgical management of patients with cyanotic heart disease. In this study, the accuracy of two-dimensional echocardiography combined with color Doppler flow mapping to assess the size, stenosis, and atresia of the major PA branches was examined using a new parasternal approach.

Methods and Results With the use of right and left high parasternal windows, we visualized each of the major portions along the right (R-PA) and left (L-PA) pulmonary arteries in 45 of the 47 examinations (96%) in 38 patients with cyanotic heart disease. The patients were between 13 days and 20 years old (mean age, 2.9 years). The internal diameters of the major PA branches were measured at three points along the R-PA (the proximal, mid, and distal portions) and at the proximal and distal portions on the L-PA in systole by both two-dimensional echocardiography and angiography. In addition, the diameter of the stenosis in the PA branch was measured. These PA values as determined by two-dimensional echocardiography correlated well with those obtained by angiography (r= 95 to 97). By two-dimensional echocardiography with color Doppler flow mapping, 17 of 19 lesions with stenoses or atresia of the major PA branches were predicted as defined by angiography (sensitivity, 89.5%; specificity, 100%). Differences between the distal parts of the L-PA and R-PA of >30% in diameter were determined by angiography in 15 examinations and by two-dimensional echocardiography in 12 examinations (sensitivity, 80%; specificity, 97.4%).

Conclusions Our technique permits noninvasive evaluation of the size, stenoses, and atresia of the major portions of the PA branches in patients with cyanotic heart disease both before and after surgery. (Circulation. 1994;89:1306-1316.)

Key Words • echocardiography • pulmonary arteries • stenoses • Doppler

Hypoplasia, stenosis, or atresia of the major pulmonary artery (PA) branches associated with cyanotic congenital heart disease is not uncommon and hampers the normal development of the lungs, making future total or radical correction more difficult.1-6 These changes in the PA morphology can be acquired after birth as a result of dactal constriction, systemic-to-pulmonary shunting, and central pulmonary banding procedures; they may also be congenital in some cases with pulmonary atresia and ventricular septal defect.6-10 These stenotic lesions may progress with time. Traditionally, angiography has been required to evaluate the anatomy of the PA. Therefore, a noninvasive method of accurately assessing PA morphology and the effects on the PA of palliative surgical procedures is needed. Previous echocardiographic studies of major PA branches have usually been performed using a suprasternal view.11-15 Several investigators showed that the left PA (L-PA) often was seen less easily than the right PA (R-PA).12-14 Furthermore, few studies have been concerned with assessment of the size or stenosis of the PA branches as measured by two-dimensional echocardiography.12-16 We therefore (1) attempted to find a better technique for imaging the R-PA and L-PA and (2) evaluated the accuracy of two-dimensional echocardiography combined with color Doppler flow mapping in determining the size, stenosis, and atresia of the major PA branches.

Patients
Between June 1991 and March 1993, 37 patients with cyanotic heart disease and obstructed pulmonary flow before radical surgery and 3 patients with a complex intracardiac anomaly who had undergone central PA banding were included in this study. Two patients were excluded from the analysis because absent central PA and bilateral ductus arteriosus were found by echocardiography and angiography and subsequently confirmed by surgery. The main diagnosis of cardiac anomaly in 38 patients (21 male and 17 female patients) are given in the Table. Four patients had dextrocardia, situs inversus, or both.

During this study, before or after palliative operation 8 patients underwent two or three follow-up examinations by both echocardiography and angiography that emphasized R-PA and L-PA growth: systemic-to-pulmonary arterial shunt operation only (n=4), shunt operation and enlargement of segmental PA stenosis (n=2), PA banding (n=1), and Brock’s procedure and shunt operation (n=1). At the time of 47 examinations in 38 patients, their ages ranged from 13 days to...
Main Diagnosis in 38 Patients

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tetralogy of Fallot</td>
<td>13</td>
</tr>
<tr>
<td>Univentricular heart</td>
<td>7</td>
</tr>
<tr>
<td>Pulmonary atresia (VSD)</td>
<td>5</td>
</tr>
<tr>
<td>Double-outlet right ventricle</td>
<td>4</td>
</tr>
<tr>
<td>Pulmonary atresia (IVS)</td>
<td>3</td>
</tr>
<tr>
<td>Common atrioventricular canal</td>
<td>3</td>
</tr>
<tr>
<td>Transposition of the great arteries</td>
<td>1</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>1</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>38</td>
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VSD indicates ventricular septal defect; IVS, intact ventricular septum.

Procedures for Imaging the L-PA

Approach 1-L

The transducer was placed over the left subclavicular or high parasternal regions and was rotated slightly counterclockwise. Then, the fanned ultrasonic beam was directed toward the patient’s left, inferiorly and posteriorly to obtain the long-axis plane of the L-PA. In this view, L-PA and its junction of the main PA were imaged (Fig 1, A). On the patient’s left, the first branch was seen.

Approach 2-L

This technique has been used to delineate both the ductus arteriosus and the L-PA.7 In brief, the transducer was placed in left intercostal space 2 or 3, rotated clockwise, and directed inferiorly. On this plane, the L-PA and main PA junction was clearly imaged, and the L-PA was visualized in an orientation that makes the flow parallel to the Doppler cursor. This approach was used to obtain the blood flow profile when approach 1-L failed to detect it.

Furthermore, some modification of the approach is necessary when the orientation of the R-PA and L-PA took an unusual course, such as the mirror-image branching. For imaging of the R-PA, the transducer was located at a position similar to that of approach 1-R, rotated counterclockwise, and directed toward the patient’s left, posterosuperiorly (Fig 1, B). For imaging of the L-PA, the transducer was positioned at the high left parasternal region and directed posterosuperiorly without any rotation (Fig 1, B).

Angiography

All patients underwent diagnostic cardiac catheterization and biplane cineangiography. Angiographic images of the PA were obtained after injection into the pulmonary trunk, aorta, and ventricle and within ductus arteriosus or surgical shunts. Pulmonary vein wedge angiography also was used to determine whether atresia of major PA branches was present. Most angiograms for evaluation of PA branch size and morphology were obtained in anteroposterior views with or without slight cranial angulation.18 This angiographic image of the major PA branches corresponds most closely to the echocardiographic image of the R-PA obtained by approach 1-R and that of the L-PA by approach 1-L. When stenosis at the origin of the R-PA or L-PA was not clearly imaged by the anteroposterior projection, its presence was confirmed by the steep left anterior oblique view with caudal angulation to demonstrate the origin of the R-PA and L-PA.19

Assessment of the Size and Morphology of the Major PA Branches

In the present study, the PA confluence was defined when the R-PA and L-PA were in continuity at the midline. PA branch stenosis was defined as when a localized narrowing area was found within the PA branches and the smallest diameter of the narrow part was ≤50% of the largest diameter of the ipsilateral artery.4,7,12 The term “atresia of major PA branch” was applied to obstructions where a sharply demarcated obstruction was found without flow directing to the distal PA and was confirmed by pulmonary vein wedge angiography.7 Differences in diameter between the distal parts of the R-PA and L-PA of >30% were defined as imbalance of PA growth.7 In both two-dimensional echocardiograms, usually examined with approaches 1-R and 1-L and angiography, the internal diameters were measured not only just before the takeoff of the first branch but also at the proximal portion of the R-PA and L-PA and at the midportion of the R-PA (Fig 1). When the first branch was not clearly visualized on the echocardiogram, the internal diameters were measured where the R-PA passed across the superior vena cava for the R-PA and at the most distal portion of the segment with a clear endovascular surface for the L-PA. All echocardiographic...
Fig 1. A, Two-dimensional echocardiograms showing normally oriented right (R-PA; A₁) and left pulmonary arteries (L-PA; A₂) in a patient with acyanotic tetralogy of Fallot. B, Two-dimensional echocardiograms showing mirror-image branching in a patient with dextrocardia, situs inversus, and double-outlet right ventricle. B₁ indicates R-PA; B₂, L-PA. The internal diameters of major pulmonary branches were measured at the proximal, mid, and distal portions along the R-PA (smaller A, B, and C) and the proximal and distal portions on the L-PA (smaller A and B) in systole. Note the different courses of the R-PA and L-PA between A and B. F indicates first lobar branch; M-PA, main pulmonary artery; AO, aorta; SVC, superior vena cava; LA, left atrium; S, superior; I, inferior; L, left; and R, right.
measurements were made with calipers from the video screen in systolic frame to allow for the largest diameter using the inner edge–to–inner edge method. Angiographic measurements of the R-PA and L-PA were made directly from the anteroposterior image, with correction for magnification made with a 1-cm calibration grid. When the anteroposterior projection was not used to image the PA, these measurements were made using the known diameter of the angiographic catheter as a reference value. If an area of localized narrowing within the PA branch was present, the smallest diameter was measured on both two-dimensional echocardiograms and angiograms. Then this measurement was used as a value of corresponding segments to the proximal, mid, and distal portions.

**Statistical Analysis**

The diameters of the R-PA and L-PA as determined echocardiographically were compared with the values obtained angiographically by linear regression analyses, and the SEE was determined. Sensitivity and specificity of the two-dimensional echocardiographic imaging technique to detect stenosis or atresia and imbalance of the R-PA and L-PA sizes were determined from the percentage of the number of examinations with true-positive and true-negative results. To evaluate the interobserver variability, two investigators separately measured all five points of the PA branches from 10 patients who were selected randomly.

**Results**

Satisfactory echocardiograms, which were available for analysis of the major PA branches, were obtained in 45 examinations (96%) of 36 patients, with two patients excluded who had hyperinflation of the lungs. Among these 45 examinations, the first branch of the R-PA or L-PA was clearly imaged in 41 (91%) and 35 (78%), respectively.

**Detection of the Stenosis or Atresia of the R-PA or L-PA**

Seventeen of the 19 lesions of PA branch stenosis or atresia could be predicted correctly by the use of color flow two-dimensional echocardiography as defined by angiography. The positions of these 17 lesions determined by echocardiography corresponded well with those determined by angiography. The proximal part of the R-PA was stenotic in 5 patients, and that of the L-PA was stenotic in 6 patients and atretic in 4 patients, although 1 patient with tetralogy of Fallot had stenoses at the proximal portions of both the R-PA and the L-PA. In addition, stenosis in the mid or distal parts of the R-PA was seen in 3 patients who had undergone previous right Blalock-Taussig shunt operation. One patient in whom L-PA stenosis was not defined by echocardiography was 1 of 2 patients with unsatisfactory echocardiograms. Another false-negative result was produced because the diameter of the smallest part of the R-PA was measured as 70% of the maximal part of the R-PA on the echocardiogram. On the other hand, all patients without stenosis or atresia by angiography were correctly diagnosed as being without stenosis or atresia with the use of two-dimensional echocardiography combined with Doppler color flow mapping. Thus, the sensitivity and specificity of our echocardiographic technique in detecting the stenosis or atresia in the PA branches were 89.5% and 100%, respectively.

**Measurement of PA Branch Size**

The mean interobserver difference for echocardiographic measurements was 3.9% for the three points of the R-PA (range, 3.4% to 4.2%) and 3.9% for the two points of the L-PA (range, 3.6% and 4.3%). On angiographic analysis, measurement of eight portions of the R-PA and eight portions of L-PA was not possible because of overlapping images between these portions on the major branch and the main PA or patent ductus arteriosus. In addition, only measurements of three segments on the R-PA could be made in 4 patients with atresia on the proximal portion of the L-PA. Fig 2 shows the relation between the echocardiographic and angiographic measurements of each point of the PA branches. The correlation coefficients for the proximal, mid, and distal portions of the R-PA were .97, .96, and .95, and those of the proximal and distal portions of the L-PA were .97 and .95, respectively. The values obtained by echocardiography were slightly smaller than those obtained by angiography, and underestimation by either >15% or >2 mm in diameter occurred in 10 of the 127 (8%) measurements for the R-PA and 7 of 74 measurements (9.5%) for the L-PA.

**Detection of Imbalance in the R-PA and L-PA Sizes**

Imbalance in the R-PA and L-PA sizes was found on the angiograms in 15 examinations of 13 patients. In 9 examinations of 7 patients, the distal portion of PA branch at the side of the stenosis was smaller than the contralateral artery (Fig 3). In the other 6 patients without segmental stenosis, PA banding or Blalock-Taussig shunt operation had been performed previously. Two-dimensional echocardiography correctly revealed 12 of these findings (80% sensitivity), and only one of the 38 examinations was misinterpreted as a small L-PA (97.4% specificity).

**Follow-up Studies of the Morphology of the Major PA Branches**

In 8 patients, the mean interval between the first and last echocardiographic and angiographic examinations was 8 months (range, 2 to 16 months). In 3 patients, the PA branch stenosis could be correctly predicted by echocardiography before first palliative surgery (Fig 4). The stenotic segment with the equine pericardium was enlarged in 2 patients with severe stenosis (Figs 4 and 5). One patient was found to have restenosis of the same portion 6 months later by echocardiography and subsequently confirmed by angiography (Figs 5 and 6). In one patient with L-PA stenosis who underwent only right Blalock-Taussig shunting, the R-PA increased in size with time, but the L-PA remained small on the echocardiogram and was subsequently confirmed by angiography (Fig 3). Six other patients showed an adequate increase in the sizes of both PA branches after palliative surgery on serial echocardiograms, whereas the distal portion of R-PA was markedly larger than the proximal portion of R-PA in one patient (Fig 7). These findings correlated well with those obtained by angiography.

**Discussion**

We found that the R-PA and L-PA could be adequately determined by high parasternal echocardiography in patients with various forms of cyanotic congenital
heart disease, although the L-PA was visualized less frequently by the suprasternal approach. It was interesting that most of the R-PA and L-PA were clearly imaged by a similar technique or slight modification of the position and direction of transducer regardless of the form of the heart disease. However, to obtain adequate imaging of the major PA branches with a quite different course such as the mirror-image branches, the approach required some modification. In this regard, special attention should be directed to patients with situs inversus, dextrocardia, or both. In our experience, if the examiner recognized the possibility of a different course of the PA branch in these conditions, these were no major problems.

**Role of Color Doppler Flow Mapping**

Color Doppler echocardiography was quite useful in rapid detection and recognition of the presence of central PA, with the exclusion of other vessels such as systemic or pulmonary veins, as suggested previously. Smylie et al also pointed out the limited value of the color flow technique in differentiating nonconfluent PA from major aortopulmonary collateral vessels. Unfortunately, during the study period, we could not examine any patients with this category of disease, although with our technique the absence of central PA and bilateral ductus arteriosus could be predicted correctly in two patients.

We found the color flow technique to be a sensitive and useful method of identifying a stenosis and atresia of the major PA branches. Color signals were not clearly visualized in some patients, probably because of either nonparallel orientation of the vessel in relation to the ultrasonic beam or severely decreased blood flow. However, when present, color flow channel narrowing served to enhance the level of confidence with which the diagnosis of a stenosis or atresia was made. A similar finding has been reported in identification and measurements of the region of coarctation in patients with coarctation of the aorta.

On the other hand, the severity of intravascular stenosis is commonly assessed by estimating pressure gradients across the stenosis by using continuous wave Doppler echocardiography. All patients in this study had pulmo-
FIG 3. Two-dimensional echocardiograms and angiogram in a patient with transposition of the great arteries, ventricular septal defect, and pulmonary valvar and infundibular stenoses. Top, Localized stenosis (arrow) and hypoplasia of the left pulmonary artery (L-PA). Middle, Internal diameters of each portion of the right pulmonary artery (R-PA) were markedly larger than those of the L-PA. This panel shows the imbalance of the R-PA and L-PA sizes. Bottom, Cineangiogram (anteroposterior projection) shows the localized stenosis (arrow) and hypoplasia of the L-PA. Note the imbalance of the R-PA and L-PA sizes. PL-SVC indicates persistent left superior vena cava; AO, aorta; F, first lobar branch; smaller A, B, and C, proximal, mid, and distal portions.
FIG 4. Color Doppler two-dimensional echocardiograms of a patient with univentricular heart and pulmonary valvar and infundibular stenoses examined at 3 months of age (top) and at 6 months of age after enlargement of the stenotic segment in the right pulmonary artery (R-PA) and left Blalock-Taussig shunt operation (bottom). Top, Localized reflectile echo density (arrow) and hypoplasia of the right pulmonary artery. Bottom, Color flow image shows enlarged R-PA and clear lower lobe branch (L). SVC indicates superior vena cava; AO, aorta; and LA, left atrium.
Fig 5. Color Doppler two-dimensional echocardiograms from same patient as shown in Fig 4, examined at 15 months of age. Top, Restenosis (long arrow) within the proximal right pulmonary artery (R-PA). Note mosaic pattern of color flow signal (short arrow) at stenotic area. Bottom, Dilated left pulmonary artery (L-PA) clearly shown. AO indicates aorta; and F, first lobar branch.
Fig 6. Cineangiograms from same patient as in Figs 4 and 5 examined at 6 months of age (top) and at 15 months of age (bottom). Top, Similar size of right (R-PA) and left (L-PA) pulmonary arteries after enlargement of the R-PA. This angiogram was obtained in the anteroposterior view with cranial angulation to image clearly the proximal parts. Bottom angiogram (anteroposterior projection) shows localized stenosis (arrow) and hypoplasia of the R-PA. M-PA indicates main pulmonary artery.
nary obstruction with or without systemic-to-pulmonary shunt or PA banding. In these situations, the severity of the stenosis appeared to be assessed more accurately by the diameter of stenotic area than by the pressure gradients across the obstruction.

**Evaluation of the PA Branch Size**

The size of the major PA branches is currently estimated from the diameter of the R-PA and L-PA before the origin of the first lobar branch by angiography. Previous reports and our findings have indicated that not all parts of the R-PA or L-PA were necessarily of the same size. In some patients, the proximal portion of the PA branch was considerably smaller than the portion just before the first lobar branch, even though the localized stenosis was not present in the PA branches. This observation indicates that the measurement of the proximal portion of the PA branches that is easily imaged with the routine echocardiographic view does not always reflect the size of their distal portion. In addition, Fontan et al measured the size of the major PA branches not only at the immediate prebranching point but also at the origin of the R-PA and L-PA and at the midportion of the R-PA on the cineangiogram, and they suggested that these multiple measurements might give a better idea of resistance to flow throughout the R-PA and L-PA. Our study, which demonstrated excellent correlation between two-dimensional echocardiographic and angiographic measurements of five points within the major PA branches, is the first report of echocardiographic evaluation of PA branch size on the basis of these considerations.

**Study Limitations**

The major problem in the echocardiographic assessment of the PA branch relates to the availability of a good acoustic window. In our experience, depending on the cross-sectional plane used, the left or right decubitus position helps to eliminate the problems attendant with parasternal examinations. In addition, when older children have poor acoustic windows because of an expanded upper lung segment that is blocking the transmission of sound waves, we often could obtain a good image using the right or left middle intercostal spaces as the acoustic windows.

Second, as shown previously, the PA branch size was slightly smaller than that obtained by angiography. Our study found echocardiographic underestimation of either >15% or >2-mm diameter in 8.5% of all measurements. There could be errors in the calculation of the magnification factor, which could lead to imprecise cineangiographic measurements of lumen size. However, the values obtained by echocardiography could be smaller than those obtained at surgery. Based on the present and previous studies, echocardiographic underestimation was most likely caused when the echo plane transected and was oblique or off center of the vessels. Thus, the transducer should be angled by the scanning technique to obtain as large a vessel diameter as possible. Another limitation of the echocardiographic measurement of PA branch size was that we used an inner edge-to-inner edge method of measurement rather than the leading edge method because we believed that inclusion of the leading vessel wall would overestimate the true vessel diameter. Improved resolution of echocardiographic imaging would lessen this problem.

**Clinical Implications**

Recent reports have recommended early reconstruction of stenotic or nonconfluent PA to avoid the imbalance of PA growth and an increase in pulmonary vascular resistance. Because of the unpredictability...
of PA growth with time, a noninvasive method of making the measurements would be useful in following PA growth. Results of the present study suggest that echocardiography is a useful noninvasive technique that helps to monitor the results of and complications resulting from palliative shunt and PA banding procedures. These follow-up examinations would facilitate the timing of subsequent angiographic assessment and surgical procedures in these patients. Furthermore, this technique complements cardiac catheterization and angiography by reducing the number of invasive procedures and aiding in the precatheterization planning of injection sites by angiography in patients with complex cyanotic heart disease.

In conclusion, our echocardiographic method provides useful information regarding size, stenosis, and atresia of the major portions of PA branches in patients with cyanotic heart disease and encourages us to perform serial evaluation of PA growth both before and after surgery.

Acknowledgments

This work was supported by a grant from Kitasato University Foundation and a grant-in-aid for general scientific research from the Ministry of Education, Science, and Culture of Japan. We gratefully acknowledge the technical assistance of Mr Yoshiyuki Makita and Miss Keiko Watanabe as well as the assistance of Miss Mami Hiraishi in the preparation of the manuscript.

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_Circulation_. 1994;89:1306-1316
doi: 10.1161/01.CIR.89.3.1306

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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