In this prospective study, 27 consecutive neonates suspected to be suffering from pulmonary atresia and intact ventricular septum underwent detailed two-dimensional echocardiographic examination before cardiac catheterization. Of the 27 neonates 25 had pulmonary atresia and intact ventricular septum and the remaining 2 had "functional pulmonary atresia" secondary to severe Ebstein’s anomaly of the tricuspid valve.

In all 25 neonates with pulmonary atresia and intact ventricular septum, the diagnosis and right ventricular morphology based on the tripartite approach were correctly established by echocardiography. The associated Ebstein’s anomaly in two babies with pulmonary atresia and intact ventricular septum was also correctly identified by echocardiography. Among the five babies who had a sinusoidal-coronary artery communication, echocardiography demonstrated the fistula in one and provided clues for its diagnosis in two others. In the 25 neonates with pulmonary atresia and intact ventricular septum, the echocardiographic dimensions of their tricuspid anulus, right ventricular infundibulum and main pulmonary artery correlated well with the angiocardiographic measurements ($r > 0.8$).

The results of this study suggest that, in the management of neonates with pulmonary atresia and intact ventricular septum, preoperative evaluation by echocardiography is usually sufficient and cardiac catheterization should be reserved for selected cases.


Methods

Echocardiography. Twenty-seven consecutive neonates with suspected pulmonary atresia and intact ventricular septum were studied between January 1982 and June 1987.

Two-dimensional echocardiography was performed with the Advanced Technology Laboratory (Mark V series 300) mechanical scanner fitted with a 5 or 7.5 MHz transducer. All echocardiograms were recorded on videotapes for detailed analysis. With the use of the sequential segmental approach (6), the atrial arrangement and intracardiac and extracardiac anatomy were delineated by the standard echocardiographic views and the diagnosis was established. The right ventricular morphology based on the tripartite approach of Goor and Lillehei (7) and Bull et al. (8) was then studied by a combination of echocardiographic views: the inlet was examined by the apical four chamber view, the trabecular portion beyond the tricuspid valve insertion by the subcostal four chamber view with anterior angulation of the transducer and the outlet (infundibulum) by the high precordial short-axis view. A search was then made for Ebstein’s anomaly and restrictive movement of the tricuspid valve and for sinusoids within the thickened right ventricular myocardium. The anatomy of the proximal coronary arteries was defined from the parasternal short-axis view at the aortic root. With use of either the apical or the subcostal four chamber view, the diameter of the tricuspid anulus at

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end-diastole was measured (9). The diameter of the main pulmonary artery at end-systole and the right ventricular infundibulum at end-diastole was measured immediately distal and proximal to the atretic pulmonary valve, respectively, at the high precordial short-axis cut.

Cardiac catheterization. In all 27 neonates, cardiac catheterization and biplane (anteroposterior and lateral) left and right ventriculography were performed under general anesthesia. By reviewing the cineangiocardiograms, the right ventricular morphology was determined according to the tripartite approach; associated right ventricular anomalies, tricuspid valve dysfunction and sinusoidal-coronary artery communications were identified. The end-diastolic diameter of the tricuspid anulus and right ventricular infundibulum, respectively, were estimated from the biplane right ventriculograms using the known diameter of the catheter as the scale. Similarly, the end-systolic diameter of the main pulmonary artery was measured from the lateral left ventriculogram when the artery was filled by contrast medium passing through the patent ductus arteriosus. The angiocardio­graphic and echocardiographic findings and measurements were then compared.

Statistical analysis was performed with use of the un­paired Student’s t test and linear regression when applicable.

Results

In all 27 neonates, two-dimensional echocardiography disclosed normally connected cardiac chambers, an intact ventricular septum and a diaphragmlike pulmonary valve that failed to open during systole, indicating the diagnosis of pulmonary atresia and intact ventricular septum. Subse­
sequent cardiac catheterization revealed the presence of pulmonary atresia and intact ventricular septum in 25 babies and "functional pulmonary atresia" (10) in the remaining 2, who had severe Ebstein's anomaly. The latter condition was later confirmed at autopsy examination. A description of other relevant observations among the 25 neonates with pulmonary atresia and intact ventricular septum follows.

Right ventricular morphology. This was well defined by both echocardiography and angiography. Twelve babies had a tripartite right ventricle (Fig. 1), 10 had a two-component right ventricle (9 had an absent trabecular portion [Fig. 2] and 1 had an absent infundibular portion [Fig. 3]) and 3 had a one-component right ventricle with an inlet only (Fig. 4).

Associated right ventricular anomalies. Both echocardiography and angiography independently diagnosed associated Ebstein's malformations of the tricuspid valve in two babies with a tripartite right ventricle. Echocardiographically, the displaced proximal attachment of the septal leaflet of the tricuspid valve into the right ventricular cavity was best seen in the apical four chamber view (Fig. 5). Restricted

Figure 3. Echocardiographic-angiographic correlate of a two-component right ventricle with absence of the infundibulum (long segment atretic infundibulum). A, Anteroposterior right ventriculogram showing an inlet (IN) and trabecular portion (TRA) but absence of the infundibulum (white asterisks). B, Echocardiogram (four chamber view) showing an apparently good sized right ventricle (RV). C, The short-axis view at the aortic root (AO) reveals discontinuity (black asterisks) between the main pulmonary artery (MPA) and the right ventricular (RV) cavity. a = aorta; LA = left atrium; LV = left ventricle; RA = right atrium; S = ventricular septum.

Figure 4. Echocardiographic-angiographic correlate of a diminutive right ventricle with an inlet only. A, Anteroposterior right ventriculogram showing a diminutive right ventricle (RV) with an inlet only. Note the small restrictive tricuspid valve (black arrows). B, Echocardiogram (four chamber view) showing the very diminutive right ventricle (RV) with a small restrictive tricuspid valve (TV) (white arrows). C, Echocardiogram (short-axis view at the aortic root [AO]) showing absence of an infundibular cavity (black asterisks) of the very diminutive right ventricle (RV). Abbreviations as in Figure 3.
movement of the tricuspid valve was identified by both imaging techniques in three babies who had a one-component right ventricle. Angiocardiography disclosed a sinusoidal-coronary artery communication in five babies whose right ventricle lacked one or two components. On the other hand, echocardiography demonstrated the coronary artery fistula in only one baby (Fig. 6) and suggested its presence in two others. The echocardiographic features that probably indicate the presence of these abnormal coronary artery communications include dilated proximal coronary arteries, unusual course of a coronary artery (Fig. 7) and irregular spaces (sinusoids) within the "reflectile" myocardium. Coronary artery discontinuity from the aortic root was not observed.

Right ventricular inflow and outflow dimensions. The echocardiographic and angiocardiographic dimensions of the tricuspid anulus, right ventricular infundibulum and main pulmonary artery and their correlations are depicted in Figures 8 through 10. Analysis by linear regression showed that the corresponding echocardiographic and angiographic dimensions correlated well (r > 0.8). The tricuspid annular diameters varied over a wide range. Overall it was larger in babies with a tripartite (range 7.1 to 18.0 mm, mean 10.9) and two-component (range 6.8 to 10.8 mm, mean 9.0) right ventricle than in those with one-component right ventricle (range 5.7 to 6.7 mm, mean 6.3). Although there was no statistical difference in the tricuspid annular diameter between the tripartite and two-component right ventricles as well as the two- and one-component right ventricles, significant difference in tricuspid annular diameter existed between the tripartite and one-component right ventricles (p > 0.01).

The size of the right ventricular infundibulum also exhibited a wide range. It was atretic in all three babies with a
Figure 7. Echocardiographic short-axis view at the aortic root (AO). The right coronary artery (arrows) runs an unusual course into the "reflective" myocardium (m). The patient has a diminutive right ventricle (rv) with an inlet only (white arrows). ra = right atrium.

Figure 8. Correlation of the echocardiographic and angiographic measurements of the tricuspid annular diameter. A, Measurements obtained from the anteroposterior view of the right ventriculogram versus those obtained from the four chamber view of the echocardiogram. B, Measurements obtained from the lateral view of the right ventriculogram versus those obtained from the four chamber view of the echocardiogram.

Figure 9. Correlation of the echocardiographic and angiographic measurements of the size of the right ventricular infundibulum. A, Measurements obtained from the anteroposterior view of the right ventriculogram versus those obtained from the high precordial short-axis view of the echocardiogram. B, Measurements obtained from the lateral view of the right ventriculogram versus those obtained from the high precordial short-axis view of the echocardiogram.

one-component right ventricle and in one baby with a two-component right ventricle. The mean infundibular diameter of the tripartite right ventricles (range 2.2 to 7.9 mm, mean 5.2) was significantly wider than that of the two-component right ventricles (range 2.1 to 6.5 mm, mean 3.8 p < 0.05). All but two babies had a main pulmonary artery diameter >4 mm (range 3.0 to 8.6 mm, mean 6.0). There was no demonstrable relation between the size of the main pulmonary artery and the right ventricular morphology.

Discussion
Pulmonary atresia and intact ventricular septum has a high early and late mortality (2,3,11). It is generally accepted that the dismal outlook for patients with this anomaly may be related to associated right ventricular hypoplasia, tricuspid valve anomaly and the state of the ventricular myocardium (2,3,8). The presence of major sinusoidal-coronary artery communications may predispose to myocardial ischemia and
right ventricular morphology according to the tripartite
the diameter of the tricuspid annulus and the three different

types of right ventricle, but we confirmed that in most cases
(14), failed to demonstrate a significant correlation between
atresia and intact ventricular septum could be based on the
cavity; the trend between groups was statistically significant.
Bull et al. (8) proposed that the management of pulmonary
arteries and irregular spaces.

Echocardiographic assessment. The present study shows
that in neonates with pulmonary atresia and intact ventricu­
lar septum, two-dimensional echocardiography allows accu­
rate diagnosis and sizing of the right ventricular inflow and
outflow orifices. When combined with pulsed Doppler echo­
cardiography, it permits differentiation of functional atresia
from structural pulmonary atresia (13). One limitation of
echocardiography is its low diagnostic yield in detecting
sinusoidal-coronary artery communications. However,
these abnormal communications seldom occur in patients
with a tripartite right ventricle and there are clues that
indicate their existence. The clues include a dilated proximal
corony artery, abnormally large intramyocardial coronary
arteries and irregular spaces.

Clinical implications. In the original study of the tripartite
approach of right ventricular morphology in patients with
pulmonary atresia and intact ventricular septum, Bull et al.
(8) found that patients with the largest tricuspid valve anulus
were in the group in which all components of the ventricular
cavity were present and those with the smallest anulus were
in those groups with only an inlet portion to the ventricular
cavity; the trend between groups was statistically significant.
Bull et al. (8) proposed that the management of pulmonary
atresia and intact ventricular septum could be based on the
right ventricular morphology according to the tripartite
approach. However, in the present study, we, like others
(14), failed to demonstrate a significant correlation between
the diameter of the tricuspid anulus and the three different
types of right ventricle, but we confirmed that in most cases

Conclusions. We suggest that detailed echocardiographic
examination is the method of choice in the evaluation of
pulmonary atresia and intact ventricular septum and that
cardiac catheterization should be reserved for selected
cases, e.g., in cases with suspected major sinusoidal-
coronary artery communications or in the rare situation in
which the pulmonary artery anatomy is not well defined by
echocardiography.

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References
1. Zuberbuhler JR, Anderson RH. Morphological variations in pulmonary
2. Freedom RM. The morphologic variations of pulmonary atresia with
intact ventricular septum: guidelines for surgical intervention. Pediatr
Pulmonary atresia and intact ventricular septum: surgical management
4. Freedom RM, Harrington DP. Contributions of intramyocardial sinusoids
in pulmonary atresia and intact ventricular septum to a right-sided circular
5. Hubbard JF, Girod DA, Caldwell RL, Hurwitz RA, Mahoney LA, Waller
BF. Right ventricular infarction with cardiac rupture in an infant with
pulmonary valve atresia with intact ventricular septum. J Am Coll
6. Shinebourne EA, Macartney FJ, Anderson RH. Sequential chamber
localization—the logical approach to diagnosis in congenital heart dis­
7. Goor DA, Lillehei CW. Congenital malformations of the heart. New
York: Grune and Stratton, 1975:11.
Pulmonary atresia and intact ventricular septum: a revised classification.
9. Gutgesell HP, Bricker JT, Colvin EV, Latson LA, Hawkins EP. Atrio­
ventricular valve annular diameter: two-dimensional echocardiographic-
10. Freedom RM, Calham G, Moe E, Olley PM, Rowe RD. Differentiation
of functional and structural pulmonary atresia: role of aortography. Am J
Cardiol 1978;41:914-20.
intact ventricular septum and critical pulmonary stenosis presenting in the
first month of life; investigation and surgical results. Br Heart J 1973;35:
9-15.
12. Waldman JD, Lamberti JJ, Mathewson JW, George L. Surgical closure of
the tricuspid valve for pulmonary atresia, intact ventricular septum, and

Figure 10. Correlation of the echocardiographic (high precordial short-axis view) and angiographic (lateral view) measurements of
the size of the main pulmonary artery.
right ventricle to coronary artery communications. Pediatr Cardiol 1984;5:221-4.


