The Value of Doppler Color Flow Mapping in Determining Pulmonary Blood Supply in Infants With Pulmonary Atresia With Ventricular Septal Defect

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Thirty-two neonates and infants with pulmonary atresia with ventricular septal defect were initially investigated with cross-sectional and spectral Doppler echocardiography and Doppler color flow mapping. All 32 had subsequent correlative angiography. This demonstrated that 24 infants had adequate-sized right and left pulmonary arteries (19 confluent, 5 nonconfluent). Of the five infants with nonconfluent pulmonary arteries, four had bilateral ductus arteriosus and one had a single left-sided ductus with anomalous origin of the right pulmonary artery from the ascending aorta. Nineteen infants had confluent pulmonary arteries, all of which were supplied by a single ductus. Eight infants had complete absence of or inadequate pulmonary arteries; all had multiple aortopulmonary collateral vessels arising from the descending aorta.

The presence of adequate-sized right and left pulmonary arteries was correctly predicted in 21 of 24 infants by cross-sectional echocardiography alone and in all 24 by Doppler color flow mapping. Confluence of the right and left pulmonary arteries was predicted by cross-sectional imaging in 14 of the 19 infants in whom it occurred, and by Doppler color flow mapping in all 19 infants.

The precise definition of the pulmonary blood supply was correctly predicted by Doppler color flow mapping in 16 of the 19 infants with confluent pulmonary arteries and a single ductus. However, in three infants in this group, Doppler color flow mapping made a false diagnosis of multiple aortopulmonary collateral vessels. In the eight infants with inadequate pulmonary arteries, Doppler color flow mapping correctly predicted the presence of two or more aortopulmonary collateral vessels, but it was unreliable in predicting the multifocal pulmonary blood supply in four of five patients with nonconfluent pulmonary arteries.

In summary, Doppler color flow mapping improved the noninvasive evaluation of pulmonary atresia with ventricular septal defect. It consistently identified adequate-sized confluent pulmonary arteries supplied by a single ductus. In such cases, systemic pulmonary shunting can be performed without prior angiography. However, when Doppler color flow mapping suggested a multifocal pulmonary blood supply, the morphology was too complex to allow accurate ultrasound definition and angiography remained the essential diagnostic technique.

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Table 1. Morphology of the Pulmonary Artery and Pulmonary Blood Supply as Defined by Angiography in 32 Neonates and Infants With Pulmonary Atresia With Ventricular Septal Defect

<table>
<thead>
<tr>
<th>32 Neonates and infants</th>
</tr>
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<tbody>
<tr>
<td>2 Absent/6 Minuscule PAs</td>
</tr>
<tr>
<td>Adequate R and L PAs</td>
</tr>
<tr>
<td>Nonconfluent RPA and LPA</td>
</tr>
<tr>
<td>Confluent RPA and LPA</td>
</tr>
<tr>
<td>Multiple ao-pulm. collats.</td>
</tr>
<tr>
<td>Bilateral ductus</td>
</tr>
<tr>
<td>RPA from Asc Ao</td>
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<tr>
<td>LPA from ductus</td>
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<tr>
<td>Single ductus</td>
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Asc Ao = ascending aorta; ao-pulm. collats = aortopulmonary collateral vessels; L = left; LPA = left pulmonary artery; PAs = pulmonary arteries; R = right; RPA = right pulmonary artery.

appropriate systemic/pulmonary shunt can be performed (2). Primary corrective surgery is not normally performed in this age group (3). Infants with absent pulmonary arteries and adequate pulmonary blood flow because of multiple aortopulmonary collateral vessels do not normally require either a prostaglandin infusion or a palliative systemic/pulmonary shunt.

Thus, the optimal diagnostic technique that would allow appropriate medical and surgical management decisions in pulmonary atresia with ventricular septal defect must first identify the intracardiac anatomy, and then define the pulmonary blood supply. The latter has traditionally been accomplished with angiography, but with recent improvements in cross-sectional echocardiography and the introduction of spectral Doppler display and Doppler color flow mapping, an alternative diagnostic approach may now be available. To assess the value of Doppler color flow mapping and define its place in decision-making in the management of pulmonary atresia with ventricular septal defect, a prospective correlative study was performed in which the echocardiographic findings at initial presentation were compared with the results of subsequent angiography.

Methods

Study patients. Thirty-two consecutive neonates and infants presenting de novo to the Wessex Paediatric Cardiac Unit over a 2 year period with pulmonary atresia with ventricular septal defect as an isolated lesion were investigated by cross-sectional and spectral Doppler echocardiography and Doppler color flow mapping. Infants with pulmonary atresia associated with other forms of complex congenital heart disease (such as atrioventricular septal defect) were excluded from the study. Infants were entered into the study after the initial ultrasound studies suggested a diagnosis of pulmonary atresia with ventricular septal defect, and the information was correlated with that derived from subsequent angiography, which was performed in all infants. Informed consent as approved by the local institutional ethical committee was obtained from the parents of all infants.

The patients’ age at the time of the ultrasound study ranged from a preterm infant of 31 weeks’ gestation to 3 months (mean 3.2 weeks). The morphology of the pulmonary blood supply encountered in this series as defined by angiography is illustrated in Table 1. Twenty-four patients were judged to have adequate-sized right and left pulmonary arteries, 19 being confluent and 5 nonconfluent. For the purposes of this study, we defined an adequate-sized pulmonary artery as one of sufficient size to allow the construction of a systemic/pulmonary shunt. Our previous surgical experience suggested that a minimal internal diameter of >3 mm measured at the proximal part of either right or left pulmonary artery, as determined by echocardiography, was the smallest size of pulmonary artery compatible with a good surgical result. In this series, the shunt normally constructed was a modified Blalock-Taussig anastomosis (4). Of the 19 infants with confluent pulmonary arteries, all had a single ductus arteriosus and 2 had a central pulmonary artery stenosis. Of the five infants with nonconfluent pulmonary arteries, four had bilateral ductus and the other had a single ductus supplying the left pulmonary artery, with anomalous origin of the right pulmonary artery from the ascending aorta. Eight infants were defined to have minuscule (n = 6) or absent (n = 2) pulmonary arteries, all of whom had multiple collateral vessels arising from the descending aorta.
Ultrasound studies. All 32 infants had complete ultrasound studies, which included cross-sectional echocardiography, spectral Doppler (both pulsed and continuous wave) display and Doppler color flow mapping, as the initial investigation. Cross-sectional imaging was performed in the standard manner using an ATL Ultramark 4 mechanical sector scanner with 5.0 and 7.5 MHz transducers. Cross-sectional imaging, spectral Doppler display and Doppler color flow mapping studies were also performed using a Toshiba SSH-65A ultrasound system. Doppler color flow mapping studies were performed using the velocity/variance mode and applying appropriate gain and filter settings.

Ultrasound studies were used to assess 1) the intracardiac anatomy, with particular emphasis on differentiating pulmonary valve atresia (either absent connection or imperforate membrane) from severe infundibular or valve stenosis, or both; 2) the presence or absence of the main, left and right pulmonary arteries and, if present, whether they were confluent or not; 3) the pulmonary blood supply (that is, ductus versus multiple aortopulmonary collateral vessels); 4) the presence of any central pulmonary artery stenosis; and 5) the aortic arch morphology, including definition of the brachiocephalic branch pattern.

The differentiation of pulmonary valve atresia from severe right ventricular outflow obstruction may be difficult using cross-sectional imaging alone (5,6). We confirmed valve atresia with continuous wave Doppler ultrasound and Doppler color flow mapping by demonstrating the absence of high velocity turbulent flow in the right ventricular outflow tract or main pulmonary artery and the presence of continuous flow in the pulmonary vessels. The presence of central pulmonary arteries was shown using a combination of cross-sectional imaging and Doppler color flow mapping. Doppler color flow mapping was able to demonstrate flow (which was laminar in the majority of cases) within normally positioned intrapericardial pulmonary arteries, thus confirming their presence. Confluence of the central pulmonary arteries was predicted by demonstrating the “sea gull” appearance, which on both Doppler color flow mapping and angiography (7) characterizes the normal junction of the left and right pulmonary arteries. The presence of a ductus was demonstrated either by the direct cross-sectional imaging of the characteristic vessel joining the descending aorta to the pulmonary artery (8) or by visualizing a single turbulent mosaic jet on the color flow map (8) entering the right or (more commonly) the left pulmonary artery on its superior aspect. Multiple aortopulmonary collateral vessels were identified by Doppler color flow mapping using a suprasternal transducer position and by demonstrating multiple areas of turbulent flow in the paraaortic area usually originating from the descending aorta (Fig. 1). The diagnosis of central pulmonary artery stenosis was made with color flow mapping by demonstrating a discrete area of turbulent flow within the vessel distal to and separate from the site of any turbulent flow associated with the entry of a ductus. The aortic arch morphology (that is, right- or left-sided) was assessed in the standard manner using suprasternal cross-sectional imaging (10). The brachiocephalic branch pattern was also assessed from the suprasternal position to determine the origin of both subclavian arteries and to exclude anomalous origin of the right subclavian artery from the descending aorta.

Angiography. Angiography was performed to further define the pulmonary blood supply in 18 patients immediately after the ultrasound study, with 9 proceeding to emergency systemic/pulmonary shunting. The remaining 14 who underwent systemic/pulmonary shunting on the basis of the ultrasound information had elective angiography in the late postoperative period to confirm the growth and development of normal pulmonary arteries.

To keep the dose of contrast agent used in these neonates and infants to a minimum, initial angiography was normally confined to 1) a right ventriculogram to confirm the diagnosis of pulmonary atresia with a ventricular septal defect, and 2) aortography to delineate the pulmonary blood supply. Selective injections into a ductus or an aortopulmonary collateral vessel were also made in the majority of patients. Biplane cineangiography was routinely performed in the frontal and lateral projections.

Results

Pulmonary valve atresia (Table 2). The diagnosis of valve atresia by cross-sectional imaging alone was accurate in 28 infants (88%). In four infants, the diagnosis was deemed to be unreliable because cross-sectional imaging alone was unable to differentiate pulmonary atresia with ventricular septal defect from severe tetralogy of Fallot. The addition of continuous wave Doppler ultrasound and Doppler color flow mapping made an accurate diagnosis of valve atresia in all 32 infants.

Pulmonary artery morphology (Table 2). Adequate-sized right and left pulmonary arteries were correctly identified by cross-sectional imaging in 21 (88%) of the 24 infants in whom they were present. In three infants, the pulmonary arteries could not be adequately visualized because of hyperinflation of the lungs, which reduced image quality. The addition of continuous wave or pulsed Doppler ultrasound did not provide extra diagnostic information. Doppler color flow mapping, however, demonstrated areas of flow within central vessels, confirming the presence of normally positioned right and left pulmonary arteries in all 24 infants.

In eight infants, cross-sectional imaging plus Doppler color flow mapping could not detect the presence of any intrapericardial pulmonary arteries. However, angiography demonstrated extremely small, hypoplastic, confluent pulmonary arteries in six of the infants who received their blood supply through a stenotic communication with an aortopul-
Figure 1. Multiple aortopulmonary collateral vessels in an infant with pulmonary atresia with ventricular septal defect and absent pulmonary arteries. A. Color flow map recorded from a suprasternal transducer position which visualizes the aortic (Ao) arch and descending (DESC) thoracic aorta. There are two areas of color “mosaic” (arrow), which represent turbulent flow within two separate aortopulmonary collateral (col) vessels (1 and 2). These collateral (COLLAT) vessels originate from the descending aorta (D Ao) just below the left subclavian artery (LSA). B. Diagrammatic representation of the color flow map. The areas of color mosaic are depicted as stippled areas. LSA = left subclavian artery.

Figure 2. Demonstration of how a single tortuous ductus may be misinterpreted as multiple aortopulmonary collateral vessels when the ductus is transected in multiple planes by Doppler color flow mapping. A. Color flow map from a suprasternal transducer position of the aortic arch and descending aorta (Desc Ao). There are two areas of color “mosaic” (arrows). These were interpreted as turbulent flow within two separate aortopulmonary collateral vessels, as diagrammatically illustrated in B. However, the subsequent angiogram demonstrated the correct morphology, which was a tortuous ductus represented schematically in C (see text). INN V = innominate vein; lca = left carotid artery; other abbreviations as in Figure 1.

by demonstrating a continuity of flow throughout the central pulmonary vessels. There were no false-positive diagnosis of confluence in the five infants with nonconfluent pulmonary arteries on either cross-sectional imaging alone or with additional Doppler color flow mapping. In infants with nonconfluent pulmonary arteries, the areas of flow that represented the right and left pulmonary arteries were clearly noncontinuous and separated by an area in which no flow took place.

Two infants with confluent pulmonary arteries had a central pulmonary artery stenosis, which could not be visualized by cross-sectional imaging, but which was demon-
strated by a discrete area of turbulence (not related to the entry point of the ductus) within the right or left pulmonary artery on Doppler color flow mapping.

Pulmonary blood supply (Table 3). Cross-sectional imaging alone did not visualize the complete morphology of the multiple aortopulmonary collateral vessels where they were present and was, therefore, not considered a useful technique for defining the pulmonary blood supply. Doppler color flow mapping correctly identified a unifocal blood supply from a single ductus in 16 of the 19 infants with confluent pulmonary arteries. In the other three infants with this anatomy, Doppler color flow mapping made a false diagnosis of multiple aortopulmonary collateral vessels. In all three infants, using the suprasternal transducer position, Doppler color flow mapping had demonstrated multiple areas of turbulence in the paraaortic area suggestive of collateral vessels (Fig. 2A and B) but subsequent angiography demonstrated the presence of a single tortuous ductus arteriosus (Fig. 2C).

Doppler color flow mapping also suggested the presence of two or more collateral vessels arising from the descending aorta in a further eight infants (Fig. 1), all of whom had minute or absent central pulmonary arteries, and thus the correct morphologic diagnosis was made. However, Doppler color flow mapping failed to define the precise number and site of origin of the collateral vessels and the lung segments they supplied.

Among the five infants with nonconfluent pulmonary arteries, Doppler color flow mapping could only define the precise pulmonary blood supply in one infant who had anomalous origin of the right pulmonary artery from the ascending aorta and ductal supply to the left. In the remaining four patients with bilateral ductus, Doppler color flow mapping could only demonstrate the left-sided duct. There were no cases with multiple aortopulmonary collateral vessels in which Doppler color flow mapping made a false diagnosis of a single, unifocal pulmonary blood supply.

Aortic arch morphology. The position of the aortic arch as determined by angiography was correctly predicted by

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**Table 2. Diagnostic Accuracy of the Pulmonary Artery and Aortic Arch Morphology of Each Ultrasound Imaging Mode as Correlated With Angiography**

<table>
<thead>
<tr>
<th></th>
<th>CSE Alone</th>
<th>CSE Plus Spectral Doppler</th>
<th>DCFM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary atresia (n = 32)</td>
<td>28</td>
<td>32</td>
<td>32</td>
</tr>
<tr>
<td>Adequate PA size (n = 24)</td>
<td>21</td>
<td>21</td>
<td>24</td>
</tr>
<tr>
<td>Confluent PAs (n = 19)</td>
<td>14</td>
<td>14</td>
<td>19</td>
</tr>
<tr>
<td>Nonconfluent PAs (n = 5)</td>
<td>3</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>PA stenosis (n = 2)</td>
<td>0</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Aortic arch morphology (n = 32)</td>
<td>30</td>
<td>30</td>
<td>32</td>
</tr>
</tbody>
</table>

CSE = cross-sectional echocardiography; DCFM = Doppler color flow mapping; PAs = pulmonary artery(ies).

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**Table 3. Diagnostic Accuracy of Doppler Color Flow Mapping in Determining the Pulmonary Blood Supply as Compared With Angiography**

<table>
<thead>
<tr>
<th></th>
<th>Single Ductus</th>
<th>Bilateral Ductus</th>
<th>Multifocal Blood Supply by Collateral Vessels</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adequate size, confluent PAs (n = 19)</td>
<td>16/19</td>
<td>0/0</td>
<td>3/0*</td>
</tr>
<tr>
<td>Adequate size, nonconfluent PAs (n = 5)</td>
<td>–</td>
<td>0/4</td>
<td>1/1†</td>
</tr>
<tr>
<td>Absent or minuscule PAs (n = 8)</td>
<td>–</td>
<td>0/0</td>
<td>8/8</td>
</tr>
</tbody>
</table>

*Three false diagnoses of a multifocal blood supply; †this patient is entered here for convenience; right pulmonary artery supply was direct from the ascending aorta and not from collateral vessels. PAs = pulmonary arteries.

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Discussion

This study demonstrates that the addition of Doppler color flow mapping to the other ultrasound imaging modes provides considerably more information than the combination of cross-sectional echocardiography and spectral Doppler display alone in the definition of the configuration of pulmonary atresia with ventricular septal defect in neonates and infants. The differentiation of pulmonary valve atresia...
from tetralogy of Fallot was aided by the use of both continuous wave Doppler ultrasound and Doppler color flow mapping in four infants in this series. Cross-sectional imaging alone may not consistently distinguish severe right ventricular outflow obstruction from atresia (5,6).

**Definition of pulmonary artery morphology.** The recognition of adequate-sized central pulmonary arteries, considered to be of sufficient size to allow the construction of a systemic/pulmonary artery shunt, and the prediction of their confluence was improved by Doppler color flow mapping compared with cross-sectional imaging alone. The additional use of spectral Doppler display was of no extra benefit in this context. The benefits inherent in such use of Doppler color flow mapping as an adjunct to the recognition of vascular structures is well understood because color flow information is obtained and processed separately from the cross-sectional echocardiographic information and later overlaid onto the cross-sectional image. Cross-sectional images of the pulmonary arteries obtained in patients with pulmonary atresia with ventricular septal defect are often suboptimal because of either poor image quality or as a result of hyperinflation of the lungs, which makes it difficult to see whether structures around the heart are vascular or not. By demonstrating color-encoded flow within these structures, Doppler color flow mapping allows the anatomic and morphologic correlates to be visualized. This reasoning also applies when trying to visualize the aortic arch morphology when the suprasternal cross-sectional images are of poor quality.

An adequate-sized pulmonary artery was defined as one large enough to accept a systemic/pulmonary shunt. We realize that this definition was arbitrary and based on the clinical and surgical experience of this unit. In this series, the internal diameter of the pulmonary arteries considered to be adequate was always >3 mm, as determined by echocardiography at the initial investigation. Central pulmonary arteries of smaller diameter were not visualized by any of the ultrasound imaging modes. Such small vessels were present in six infants, all of whom had coexisting multiple aortopulmonary collateral vessels. No infant was encountered in this series who had severely hypoplastic pulmonary arteries without associated aortopulmonary collateral vessels. The reasons for the failure of Doppler color flow mapping to identify these hypoplastic central pulmonary arteries are either that they were too small for the inherent lateral resolution problems associated with the technique or that turbulent flow detected in the area of the pulmonary arteries was mistaken for an aortopulmonary collateral vessel. All these infants had multiple aortopulmonary collateral vessels and, therefore, form part of the subgroup that the results of this study suggest should be fully investigated by both Doppler color flow mapping and angiography to allow appropriate management decisions to be made. Although not encountered in this series, we would expect that it would be virtually impossible for Doppler color flow mapping to differentiate nonconfluent distal pulmonary arteries from aortopulmonary collateral vessels.

**Definition of the morphology of the pulmonary blood supply.** The definition of the precise nature of the pulmonary blood supply in neonates and infants with complex pulmonary atresia with ventricular septal defect is not possible in more than a few cases using cross-sectional echocardiography alone. Our initial experience in this series confirmed the problems inherent in the use of cross-sectional imaging alone and suggested that Doppler color flow mapping might prove to be a useful adjunct in consistently differentiating ductal blood supply from multiple aortopulmonary collateral vessels. However, subsequent experience demonstrated that when multiple areas of turbulent mosaic flow were visualized arising from the descending aorta, it was not possible to reliably distinguish a tortuous ductus from multiple collateral vessels (Fig. 2). Presumably, this appearance was caused by the ultrasound beam sectioning the tortuous ductus in multiple planes, thus giving the appearance of turbulent flow within multiple vessels. In cases where this situation is suspected, cardiac catheterization remains the investigation of choice. However, in this series when only a single area of turbulent flow was detected, communicating between the descending aorta and confluent pulmonary arteries, the presence of a single ductal supply was reliably predicted. Although theoretically, a single ductus might not be distinguishable from a single aortopulmonary collateral vessel, there are no reported cases of a unifocal blood supply to the lungs from a single collateral vessel. This means that in this series, cross-sectional echocardiography plus Doppler color flow mapping identified 16 of the 19 infants (50% of the total group) with adequate-sized confluent central pulmonary arteries with unifocal blood supply from a single patent ductus arteriosus and, therefore, a correct decision could be made on the timing of surgery and the side on which to perform the thoracotomy. It is now our policy to routinely refer such neonates and infants for a palliative shunt procedure without recourse to prior angiography.

**Conclusions.** Doppler color flow mapping enhances the noninvasive evaluation of the pulmonary artery morphology and the pulmonary blood supply in patients with pulmonary atresia with ventricular septal defect. In infants with confluent pulmonary arteries and a unifocal blood supply demonstrated by Doppler color flow mapping, the presence of a single ductus can be reliably predicted and the appropriate medical and surgical decisions can be made. In cases where Doppler color flow mapping suggested a multifocal blood supply, irrespective of the pulmonary artery morphology (confluent, nonconfluent or absent), angiography was essential.
We thank Tineke van de Kolk for her secretarial assistance.

References


