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Echocardiographic Assessment of Pulmonary Blood Supply in Patients With Pulmonary Atresia and Ventricular Septal Defect

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Objectives. **The purpose of this study was to determine the role of high resolution two-dimensional echocardiographic imaging and color flow Doppler study in assessing the pulmonary blood supply in patients with pulmonary atresia and ventricular septal defect (VSD).**

Background. **Although echocardiography is a well established technique for assessing central pulmonary arteries in pulmonary atresia VSD and for determining the presence or absence of a patent arterial duct, few data are available on its role in patients whose source of blood supply is from collateral vessels.**

Methods. **Forty-two patients aged a few hours to 19 months (mean 29 days) were prospectively assessed by high resolution echocardiography, including color flow Doppler study, during a 4-year period ending in 1994, before any intervention other than intravenous administration of prostaglandins. Angiographic confirmation was available in 29 patients, including 18 (95%) of 19 with aortopulmonary collateral channels.**

Results. **A patent arterial duct was correctly identified as the sole source of pulmonary blood supply in 23 patients, whereas aortopulmonary collateral channels were detected in 19, with one of these having a small patent arterial duct and collateral channels. The patent arterial duct originated from the undersurface of the aorta in 16 (67%) of 24 patients and from the base of the brachiocephalic trunk in 7 (33%) of 24. All patients with a patent ductus as the sole source of pulmonary blood supply had confluent pulmonary arteries. Nonconfluent pulmonary arteries were present in six patients, with all but one having aortopulmonary collateral channels as the sole source of pulmonary flow. Aortopulmonary collateral channels were direct in 17 (89%) of 19 patients, whereas in 2 (11%) of 19, both direct and indirect collateral channels were present. Color flow Doppler study was accurate in determining the presence or absence, the side and the origin of the collateral channels in all patients, with the correct number being determined in 12 (67%) of 18. "Wash-in" to the hilar pulmonary arteries (retrograde color flow) was seen in 12 (92%) of 13 patients with collateral channels and confluent pulmonary arteries. Failure to identify a tiny central pulmonary artery occurred in one patient.**

Conclusions. **High resolution imaging and color flow Doppler study provide good appreciation of the source of pulmonary blood supply in neonates and young infants with pulmonary atresia VSD.**

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The designation pulmonary atresia and ventricular septal defect (VSD) implies a biventricular heart, concordant atrioventricular (AV) connections, a large VSD, an overriding aorta and no direct flow from the ventricular mass into the pulmonary arteries (1). Echocardiography has had a major impact on the preoperative evaluation of patients with this defect. In our opinion, this technique provides enough data for the surgeon to construct a systemic to pulmonary artery shunt (1–3) in patients with clear confluent pulmonary arteries ≥3 mm that receive their blood supply from a patent arterial duct.

Until recently, the assessment of aortopulmonary collateral channels by echocardiography has been difficult (4). Using color flow Doppler study, Smyllie et al. (2) detected some collateral channels but found it impossible to reliably distinguish a tortuous ductus from multiple collateral vessels. It has been suggested (5) that newer technology providing high resolution two-dimensional echocardiographic imaging in combination with dual frequency color Doppler study will improve noninvasive diagnosis of infants with pulmonary atresia VSD. This study was designed to determine the role of high resolution two-dimensional imaging and color flow Doppler study in assessing pulmonary blood arterial supply in patients with pulmonary atresia VSD.

Methods

Patients. Forty-two infants with pulmonary atresia VSD aged a few hours to 19 months (mean age 29 days) were prospectively diagnosed by echocardiography at our institution over a 4-year period ending in September 1994. All patients were first evaluated by echocardiography before any intervention other than intravenous administration of prostaglandins.

Angiographic evaluation was performed in 29 patients, 0 to 360 days (mean 70) after initial diagnosis by echocardiography.

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Abbreviations and Acronyms
AV = atrioventricular
VSD = ventricular septal defect

Of these 29 patients, 19 had collateral channels, and 18 of these 19 (95%), underwent cardiac catheterization 1 to 69 days (mean 26) after echocardiographic diagnosis.

Echocardiography. The studies were performed with either an ATL Ultramark 9 ultrasound unit, using a 7.5-MHz mechanical transducer for imaging and a 5-MHz phased array transducer for two-dimensional echocardiographic imaging and color Doppler study, a Hewlett-Packard SONOS 1500 equipped with a dual 7.5- and 5-MHz transducer for imaging and color Doppler study or an Acuson 128 XP using a 7.5-MHz transducer for imaging and color Doppler study.

The echocardiographic examination focused on the following issues (1): 1) intracardiac anatomy; 2) confluence of the pulmonary arteries; 3) size of the pulmonary arteries; and 4) Source of pulmonary blood supply: a) aortopulmonary collateral channels (origin, side and number); b) patent arterial duct (side and site of origin); c) side of the aortic arch; and d) branching pattern of the brachiocephalic vessels.

Pulmonary arteries. The confluence and size of the pulmonary arteries were assessed from the frontal plane in a high parasternal or suprasternal position (6) by using a combination of two-dimensional imaging and color flow Doppler study. If confluent pulmonary arteries were found, the direction of the color flow was used to deduce the main source of the pulmonary blood. In patients with a large patent arterial duct (Fig. 1) the flow was expected to be anterograde in both pulmonary arteries. If retrograde flow originating at the hilum (wash-in) was identified, aortopulmonary collateral channels were presumed to be the main source of blood supply to the ipsilateral lung (Fig. 2).

Collateral channels. The origin of aortopulmonary collateral channels was determined by pulsed Doppler study and color flow mapping of the coronary arteries, ascending aorta, brachiocephalic vessels and descending thoracic and abdominal aorta. Collateral channels were suspected if continuous flow was detected in any of the preceding areas, with the diastolic phase expected to be anterograde proximal and retrograde distal to the origin of the collateral channels.

The ascending aorta, aortic arch and proximal descending aorta were studied from the long axis of the aortic arch from the suprasternal or high parasternal positions. The descending aorta was also evaluated from the subcostal long axis of the thoracic and abdominal aorta.

With use of a high parasternal or suprasternal position in the frontal plane, color flow interrogation of the hilum of the lungs was also undertaken to detect continuous flow, signaling the entry of collateral channels into the hilar pulmonary arteries. It was necessary to angle the transducer beam posteriorly to interrogate this area, thus avoiding confusion with pulmonary venous flow.

Ductus arteriosus. The patent arterial duct was assessed from the suprasternal or high parasternal view and followed to its junction with the mediastinal pulmonary arteries.

Angiography. During the 1st 3 years of the study, by policy, patients with pulmonary atresia and VSD and a large patent arterial duct supplying confluent pulmonary arteries without obvious collateral channels received a systemic to pulmonary artery shunt plus or minus ligation of the patent ductus, relying only on echocardiographic evaluation. Cardiac catheterization was performed later, before complete repair or before a second systemic to pulmonary artery shunt.

During the last year, it has been the policy at our institution to perform a primary repair in those patients with confluent

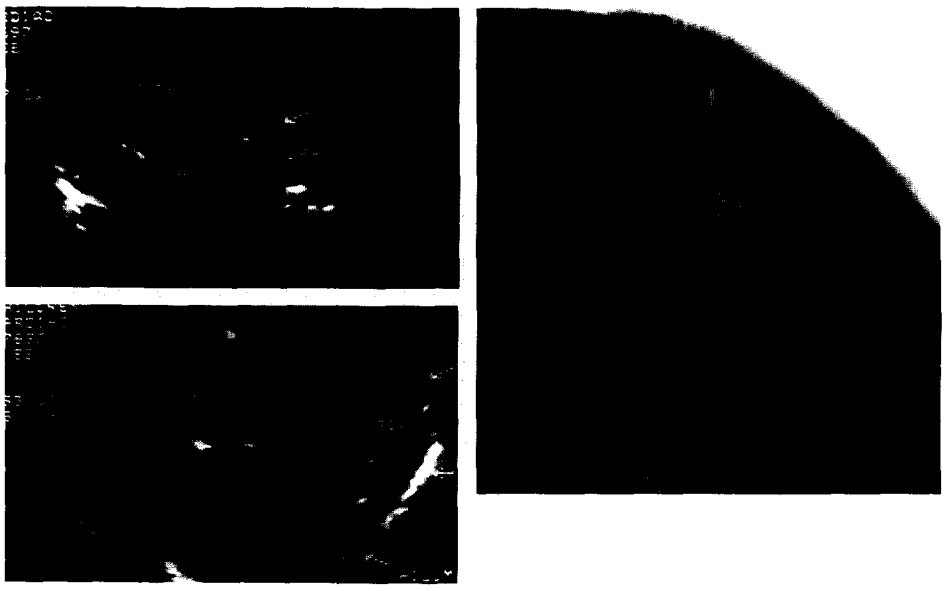

Figure 1. Mediastinal pulmonary arteries in the setting of pulmonary atresia, ventricular septal defect and pulmonary blood supply by way of a patent arterial duct. The duct arises from the base of the left innominate artery and courses within the mediastinum, proximal to the hilum, as outlined by the **arrows** in the **lower left panel,** and connects to a mediastinal pulmonary artery. There is mild left pulmonary artery stenosis. LPA = left pulmonary artery; PDA = patent arterial duct; RPA = right pulmonary artery.

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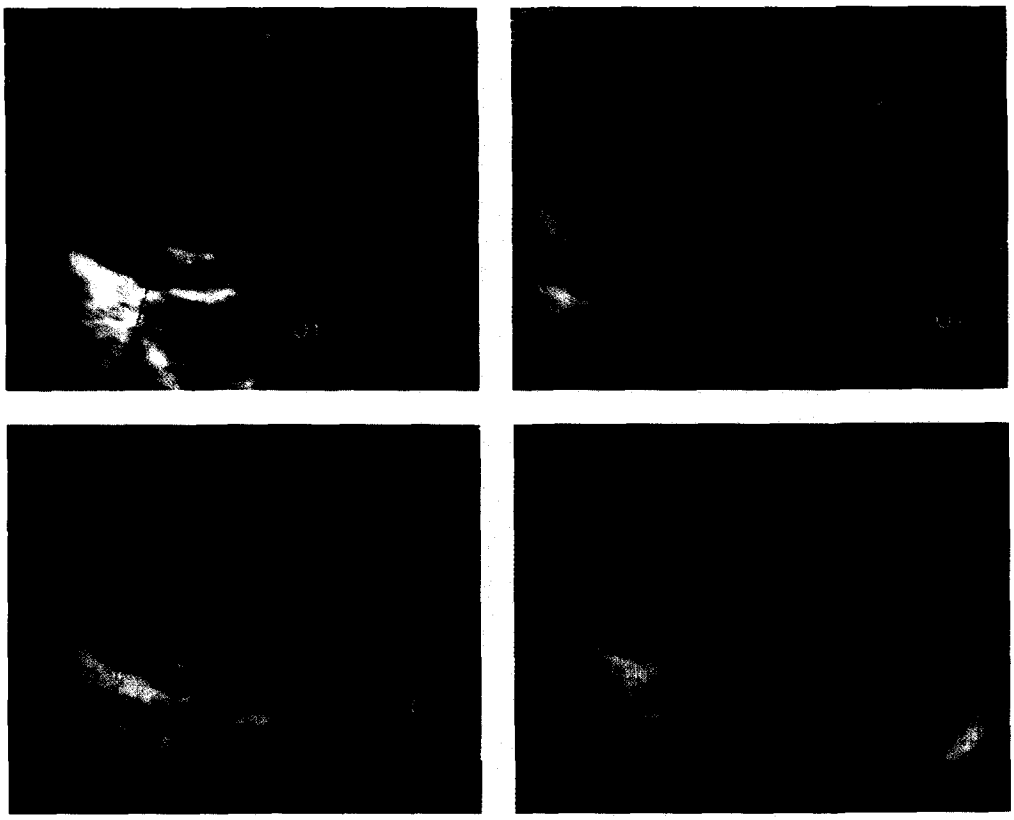

Figure 2. Suprasternal views from a patient with pulmonary atresia, ventricular septal defect, confluent pulmonary arteries and aortopulmonary collateral channels. The **upper left panel** demonstrates "wash-in" to a mediastinal left pulmonary artery from an aortopulmonary collateral vessel (COL). The turbulent flow within the hilar region at the site of connection of the collateral vessel with the mediastinal pulmonary artery results in retrograde flow into the left pulmonary artery, as outlined by the red flow toward the transducer. The **upper right panel** shows anterograde laminar blue flow down the right pulmonary artery, with flow originating from the left-sided hilar collateral connection. Note the turbulent flow from an additional right-sided collateral vessel, which supplies a different segment of the right lung. There appears to be no connection between the right-sided collateral vessels and the mediastinal pulmonary artery. The **lower left panel** shows what appears to be a single vessel arising from the descending aorta, which could easily be mistaken for a patent arterial duct. The **lower right panel,** which uses the addition of color flow, reveals two vessels, indicating that this is most likely collateral flow. This hypotheses is then confirmed by the **upper panels,** which demonstrate that the site of connection is at the hilum and not with the mediastinal pulmonary arteries. AO = aorta; LA = left atrium; lpa = left pulmonary artery.

pulmonary arteries that are deemed to be of adequate size and are supplied by a patent arterial duct. As a result, all patients have undergone, before repair, an initial echocardiographic evaluation followed by cardiac catheterization.

The angiographic investigations included detailed assessment of the pulmonary arteries and their blood supply, including selective collateral injections and pulmonary vein wedge angiography when deemed necessary. The surgical, autopsy or angiographic findings were used alone or in combination to judge the accuracy of the echocardiographic evaluation.

Statistical analysis. Descriptive analysis was used, incorporating mean values ± SD for measurements and percentages for describing numbers of cases.

Results

Intracardiac anatomy. Two-dimensional imaging alone accurately diagnosed the intracardiac anatomy in all patients.

Pulmonary arteries. Thirty-six patients had confluent pulmonary arteries (Fig. 3). In 28 (78%) of 36 patients, the confluence was identified by two-dimensional imaging alone, whereas in 7 (19%) of 36 color Doppler study was necessary to identify the confluence. It was not possible to identify the pulmonary arteries in 1 (3%) of 36 patients with aortopulmonary collateral channels and tiny right and left confluent pulmonary arteries measuring 1.0 and 1.5 mm, respectively, on angiographic study.

In patients with ductal supply and confluent pulmonary arteries, the respective right and left pulmonary artery measurements were as follows: right pulmonary artery mean 4.2 mm (95% confidence limits [CI] 3.7 to 4.6); left pulmonary artery 4.3 mm (95% CI 2.8 to 7.0). In those with confluent pulmonary arteries and absence of a patent arterial duct, the right pulmonary artery mean size was 2.4 mm (95% CI 1.9 to 2.8), with a left pulmonary artery mean size of 2.4 mm (95% CI 1.4 to 4.1).

Pulmonary blood flow supply (Fig. 3). The main source of pulmonary blood flow supply was correctly deduced by a

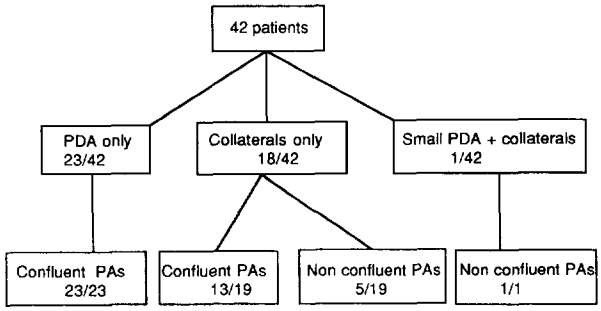

Figure 3. Source of pulmonary supply and pulmonary confluence in 42 patients with pulmonary atresia and ventricular septal defect. PAs = pulmonary arteries; PDA = patent arterial duct.

combination of imaging and color flow Doppler study in all 42 patients as detailed next.

Patent arterial duct. A patent arterial duct was present in 24 (57%) of 42 patients (Fig. 3) with its presence or absence and origin correctly identified in all by two-dimensional imaging and color Doppler study. A large ductus was the sole source of pulmonary blood flow in 23 (64%) of 36 patients with confluent pulmonary arteries measuring ≥2.8 mm in all. In one patient (Patient 8, Table 1) a combination of a patent arterial duct and collateral channels were noted. Color flow study demonstrated continuous flow, commencing within the mediastinal pulmonary arteries and continuing distally in an anterograde direction toward both hilar regions. The patent arterial duct originated from the undersurface of the aorta in 16 (67%) of 24 patients and from the base of the brachiocephalic trunk in 7 (33%) of 24.

Aortopulmonary collateral channels. Aortopulmonary collateral vessels were the sole source of blood supply in 18 (43%) of 42 patients (Fig. 3), with one other patient having a patent arterial duct to one lung and aortopulmonary collateral channels to the other. Color flow Doppler study was accurate in determining the presence or absence and the origin of the collateral channels in all, with the correct number being identified in 12 (67%) of 18 patients who underwent angiographic evaluation (Table 1). In the one other patient direct collateral channels could be identified; however, the child died before angiography was performed and no autopsy was available. In one patient, a direct collateral channel diagnosed by echocardiography and confirmed to be a collateral vessel during operation was misinterpreted as patent arterial duct by angiography. In another patient two collateral channels could be seen by color flow study; however, only one could be seen terminating in the left lung, and the site of connection of the second one was unclear (Table 1, Patient 12).

The collateral vessels were direct in 17 (89%) of 19 patients and mixed (direct and indirect) in 2 (11%) of 19. In these two, the direct collateral channels originated from the descending aorta and the indirect from the brachiocephalic trunk (Fig. 4). In patients seen to have an indirect collateral channel, only a small portion of the vessel was visualized within the mediastinum as the remainder was outside the field of vision, being shielded by the lung (Fig. 4).

Differentiation between an aortopulmonary collateral channel and a patent arterial duct was not based on the flow pattern within the vessel, which may be laminar or turbulent, depending on relative pressure differences. Likewise, it was not based on the site of origin of the vessel, as in some cases an aortopulmonary collateral channel may arise from a similar site to a patent ductus arterious. Rather, it was based in part on the presence of multiple vessels arising from the aorta, in conjunction with the known pattern of connection of aortopulmonary collateral arteries, that is, into the hilum of the lung, rather than into the mediastinal pulmonary arteries. Therefore, in the setting of an aortopulmonary collateral channel, the flow into a mediastinal pulmonary artery confluence was retrograde (so called wash-in) and not anterograde as in patients with a patent arterial duct (Fig. 2).

Indeed, wash-in to small confluent pulmonary arteries was seen in 12 (92%) of 13 patients with collateral channels and confluent pulmonary arteries. In these patients the collateral vessel could be identified at its site of entry into the hilar pulmonary arteries, with invariably low velocity flow washing into the mediastinal pulmonary artery on that side. The flow pattern in the other pulmonary artery was then anterograde toward the hilum, unless another aortopulmonary collateral channel or patent arterial duct connected with the mediastinal pulmonary artery on that side, providing competitive flow in the mediastinal pulmonary artery. We failed to visualize wash-in in one patient in whom tiny pulmonary arteries were only identified by angiography.

Table 1. Number of Collateral Channels: Echocardiography Versus Angiocardiography

	Echocardiography		Angiocardiography		
Case No.	Right Side	Left Side	Right Side	Left Side	Correct No. by Echocardiography
1	2	2	>3	>3	No
2	1	1	1	1	Yes
3	1	1	2	1	No
4	1	0	1	0	Yes
5	2	0	2	0	Yes
6	1	2	2	2	No
7	1	1	1	1	Yes
8*	0	2	0	2	Yes
9	2	0	2	2	No
10	2	0	2	0	Yes
11	1	1	1	1	Yes
12	1	1	1	1	†
13	2	0	2	0	Yes
14	2	1	2	1	Yes
15	0	2	0	2	Yes
16	1	1	1	1	Yes
17	1	0	2	2	No
18	1	0	1	0	Yes

*Patient 8 had a patent arterial duct in conjunction with collateral channels. Echocardiography identified the correct number of collateral channels in Patient 12, but the side of connection of one channel was unclear.

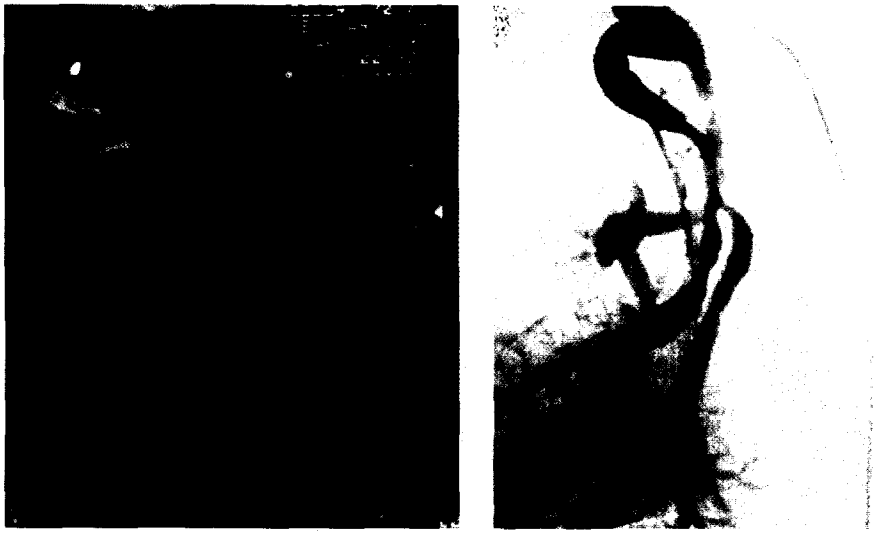

Figure 4. Suprasternal view in pulmonary atresia ventricular septal defect, with the right lung supplied by indirect collateral channels, arising from a brachiocephalic vessel. Only a small component of the collateral vessel is seen as it traverses the mediastinum, with the connection within the hilum being shielded by lung parenchyma in this patient. This situation differs from that of patients with a patent arterial duct, where the site of connection occurs within the mediastinum and hence is invariably visible. ao = aorta; col = collateral vessel.

In this series there were no patients with bilateral patent arterial ducts and nonconfluent pulmonary arteries.

Aortic arch side and branching. A right aortic arch was demonstrated in 17 (40%) of 42 patients, with mirror image branching being present in all but 1 patient with an aberrant left subclavian artery. An aberrant origin of the right subclavian artery from the descending aorta was present in two patients with a left aortic arch. The echocardiographic assessment of the aortic arch proved to be correct in all cases.

Discussion

This study demonstrates that high resolution two-dimensional imaging and color flow Doppler study provide accurate and detailed information in these patients with pulmonary atresia and ventricular septal defect. It is apparent that in most cases it is possible to determine that the pulmonary arteries are confluent and to locate their source of pulmonary blood supply.

Differentiating an aortopulmonary collateral channel from a patent arterial duct. This task is simplified if the examiner has a sound understanding of the various sites of origin and termination of a patent arterial duct or an aortopulmonary collateral channel. First, a patent arterial duct is either single or bilateral, arising from the base of the first brachiocephalic branch of the aorta or from the underside of the arch, finally terminating in a mediastinal pulmonary artery. In contrast, aortopulmonary collateral vessels may be single or multiple, arising from a variety of sites, with final termination within the hilum of the lung.

The presence of multiple vessels confirms the presence of aortopulmonary collateral channels; however, when they are single or double (one on either side), they may be confused with a patent arterial duct, particularly as the sites of origin may be similar. The flow patterns within the vessel may not be helpful, because both a patent arterial duct and an aortopulmonary collateral channel may have either laminar or turbulent flow, depending on the relative reductions in pressure along their length.

Therefore, the echocardiographer should pay strict attention not only to the number of vessels and their sites of origin, but more importantly to the relative directions of flow in the pulmonary arteries, both centrally and peripherally within the hilum, as these provide valuable clues as to the source of blood supply. "Wash-in" to a hilar pulmonary artery, particularly in the presence of confluent pulmonary arteries, indicates collateral supply and should result in careful interrogation of those sites that may be giving rise to either direct or indirect collateral vessels. In our experience, when the mediastinal pulmonary arteries are small, this wash-in tends to be of low velocity, as there are invariably significant stenoses between the mediastinal and hilar pulmonary arteries, resulting in a reduction of blood flow to the mediastinal vessels. Turbulent flow may be seen in the presence of greater flow; however, the direction of flow is still retrograde from hilum toward the mediastinal pulmonary arteries.

In patients with confluent mediastinal pulmonary arteries and absence of a patent arterial duct, anterograde flow in one pulmonary artery is indicative of predominant collateral flow to the contralateral lung (Fig. 2). This pattern will be different in patients with collateral vessels terminating in both hilar regions, with the potential for retrograde flow in both. Whether or not the anterograde flow generated by the contralateral pulmonary artery masks the ipsilateral retrograde flow from the other collateral channel will depend on the relative amount of flow from each channel.

Although no patients with bilateral patent arterial ducts and nonconfluent pulmonary arteries were encountered during this study, our previous experience with this entity indicates that there is the potential for confusion between two single aortopulmonary collateral channels arising from exactly the same sites as bilateral patent arterial ducts; that is, one from the undersurface of the aortic arch and the other from the base of the first branch of the aorta. Therefore, unlike Smyllie et al.

(2), we believe that it is only in this setting that confusion between a collateral vessel and a patent arterial duct might cause a problem. In patients with confluent pulmonary arteries it should be, and thus far has been, possible to differentiate a collateral channel from a patent arterial duct if attention is paid to the site of connection of the vessel and the various flow patterns described previously.

Finally, differentiation between an aortopulmonary collateral channel and a patent arterial duct on the same side may be possible in this setting, particularly if the pulmonary arteries are confluent, as in general they do not supply the same segment of lung. Therefore, the flow in the segment of pulmonary artery connected to the patent arterial duct will be anterograde, with the potential for competitive flow from the collateral channel if they are interconnected.

Aortic runoff. Increased diastolic flow in the aorta also indicates runoff and should result in further exploration to determine the source. In patients with collateral channels that arise proximally from the coronary arteries or brachiocephalic vessels, the runoff will be retrograde in diastole above the upper descending aorta, whereas those with collateral channels arising from the upper descending aorta have diastolic runoff in the vicinity of the collateral channels. Flow in the descending aorta below these channels will be anterograde in systole and retrograde in diastole. Persistent anterograde diastolic flow beyond this point indicates that collateral vessels probably lie distal to that site. The limitation of diastolic runoff for all the preceding scenarios relates to the amount of flow from the collateral vessels.

Missed collateral vessels. The lack of agreement between angiography and color flow Doppler study in 12 (67%) of cases as to the precise number of collateral vessels may be due in part to the complex nature of aortopulmonary collateral channels, with tortuous overlapping vessels that frequently arise from one side of the aorta and then course in front of the descending aorta.

Current management strategies. Given this current information, it might be possible to modify the management strategies in patients with pulmonary artresia VSD. If confluent pulmonary arteries of adequate size are identified, with the sole source of blood supply arising from a patent arterial duct and no recognizable aortopulmonary collateral channels, then it might seem reasonable to consider performing repair in these patients without prior angiography, particularly if no other risk factors are identified. If the pulmonary arteries appear too small for complete repair, or if aortopulmonary collateral channels are identified, then angiography is advisable before further intervention. However, echocardiography should make it possible to postpone further investigation during the neonatal period in patients with significant aortopulmonary collateral channels, as unifocalization is usually deferred for a few months. Similarly, our study confirms previous findings that a systemic to pulmonary artery shunt can be constructed on the basis of echocardiographic findings in those centers where this is the preferred mode of management in the newborn period.

Further limitations of echocardiographic assessment. Age is an obvious limitation, as excellent suprasternal and subcostal images are invariably available only in the neonate and young infant. Fortunately, the majority of patients with pulmonary atresia VSD present during this time period, when excellent imaging is possible.

The inability to identify more distal pulmonary arteries is a major limitation of echocardiography. However, it is unusual to have significant distal stenosis in the presence of confluent central pulmonary arteries of adequate size. Stenosis of a pulmonary artery at its site of connection with a collateral vessel within the hilum of the lung also cannot be detected. Although in many cases the number and origin of collateral channels can be correctly identified, it is not possible to determine how many of these channels enter a particular lung segment.

Conclusions. A combination of high resolution two-dimensional echocardiographic imaging and color flow Doppler study permits good appreciation of both the mediastinal pulmonary arteries as well as their blood supply in patients with pulmonary atresia VSD. This technique is a valuable adjunct to cardiac catheterization and can aid in the decision process in patients with this condition.

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