Editorial Comment

Doppler Color Flow Imaging and Determination of Pulmonary Blood Supply in Infants With Pulmonary Atresia With Ventricular Septal Defect*

DONALD J. HAGLER, MD, FACC
Rochester, Minnesota

To date, little has appeared in the cardiology literature concerning the echocardiographic diagnosis of pulmonary atresia with ventricular septal defect (1–3). However, several investigators (4,5) have described the clinical or surgical management of distressed hypoxic infants with this condition on the basis of two-dimensional echocardiography alone. High resolution (7.5 and 10 MHz) two-dimensional imaging has greatly improved diagnostic accuracy, allowing precise identification of tiny central pulmonary arteries (internal diameter ≥2 mm) even in premature infants. Clearly, many infants with pulmonary atresia with ventricular septal defect and central confluent pulmonary arteries, 3 mm in diameter, supplied by a patent ductus arteriosus have undergone palliative surgical creation of a shunt on the basis of two-dimensional echocardiographic findings alone.

Unresolved questions have focused primarily on recognition and clear demonstration of confluent nonstenotic and fully distributed pulmonary arteries with the unequivocal diagnosis of pulmonary atresia and with the exclusion of other significant associated anomalies (e.g., anomalous pulmonary venous connection).

The present study. The work by Smyllie et al. (6) in this issue of the Journal presents an outstanding series of infants with pulmonary atresia with ventricular septal defect. The investigators purposely excluded infants with complex diseases such as atrioventricular (AV) canal and situs abnormalities to avoid complicating features of anomalous pulmonary venous drainage. The select group represents one of the largest series for a prospective assessment of the lesion comparing the findings of high resolution two-dimensional echocardiographic imaging with the additive findings obtained with Doppler color flow imaging. The results indicate that Doppler color flow imaging substantially improved the noninvasive echocardiographic evaluation of pulmonary atresia with ventricular septal defect. It appeared to have improved the results of conventional two-dimensional imaging in 32 infants by correctly identifying additionally 1) four infants with confirmed diagnosis of pulmonary atresia (versus severe stenosis), 2) three infants with central pulmonary arteries, 3) five infants with usable confluent pulmonary arteries, and 4) two infants with central pulmonary artery stenosis. Undoubtedly, this represents a substantial improvement in noninvasive recognition of important central pulmonary artery anatomy that otherwise would have required angiography before palliative surgery.

One of the earlier recognized benefits of Doppler color flow imaging has been the remarkable rapidity with which correct diagnosis and recognition of anatomy and associated anomalies can be attained. In this lesion also—pulmonary atresia with ventricular septal defect—Doppler color flow imaging appears to allow very rapid detection and clear recognition of the presence of central confluent pulmonary arteries. One of the certain benefits should be this improved vessel recognition and definition based on typical Doppler color flow patterns. Previous two-dimensional echocardiographic experience allowed demonstration of small central vessels but often failed to allow complete depiction of confluent arteries.

What areas remain unclear? Some aspects of this well designed study leave unanswered questions. Much of the previous two-dimensional echocardiographic experience suggest that the high resolution imaging in the past did allow precise demonstration of very small (2 to 3 mm internal diameter) confluent pulmonary arteries in infants such that palliative surgery could be performed without invasive study. In their study, Smyllie et al. (6) seemed to have greater difficulty with two-dimensional imaging alone. It is difficult in such comparative studies to be truly objective (even with "blinding") and there may have been a definite bias for better recognition with color flow imaging. The impact of color flow imaging on improved recognition may be less obvious than the authors imply, but clearly we should appreciate an easier recognition and more confident demonstration of a confluence.

The authors seem somewhat conservative in detection of central pulmonary arteries. Six infants had hypoplastic central pulmonary arteries (diameter ≤3 mm) that were not recognized by either two-dimensional or Doppler color flow imaging. Some investigators may still consider such central pulmonary arteries clinically important and acceptable for a palliative surgical right ventricular outflow reconstruction. Some of the authors’ imaging problems may be related to...
their use of earlier available color flow instrumentation, which only allowed color flow imaging with relatively low resolution two-dimensional imaging (3.75 MHz).

Additionally, the authors (6) admit that their color flow technique did not provide improved noninvasive delineation of the status of multiple systemic pulmonary collateral vessels in patients with nonconfluent pulmonary arteries. The authors did imply that the color flow technique provided improved recognition of central pulmonary artery stenosis in two patients, which again may reflect inadequate two-dimensional imaging or resolution.

It should also be clearly recognized that these observations may be pertinent only in infants who have a strong central communication with the pulmonary artery confluence such as a ductus arteriosus. Indeed, six infants with stenotic distal systemic pulmonary collateral vessels supplying a central confluence were not recognized with color flow imaging. Therefore, older children or adults with such anatomy may not benefit from the addition of this technique.

Future applications. This study suggests that newer technology providing high resolution two-dimensional echocardiographic imaging (7.0 MHz) with added digital expansion capabilities in combination with dual frequency (5.0 MHz) Doppler color flow imaging capability will substantially improve noninvasive management of infants with pulmonary atresia with ventricular septal defect. For infant studies, technical development should continue to emphasize improved resolution of two-dimensional imaging. With such capability, several of the remaining problems experienced in this study may be resolved. One should expect improved resolution of tortuous ducts or systemic pulmonary collateral vessels and better recognition of nonconfluence. However, most physicians would agree with the authors (6) that, for the foreseeable future, the delineation of systemic pulmonary collateral vessels and nonconfluent or maldistributed pulmonary arteries (multifocal origin) will remain in the domain of angiography.

References