

## Operability in transposition of great arteries with ventricular septal defect: A difficult question - is the answer really so simple?

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Children with complete transposition of great arteries with ventricular septal defect (TGA-VSD) show accelerated pulmonary vascular disease (PVD) due to a combination of factors including increased pulmonary blood flow (PBF), elevated pulmonary arterial pressure, hypoxia and endothelial dysfunction. While some degree of PVD can be seen on lung biopsy as early as 2 months, the prevalence and severity of PVD in TGA-VSD rapidly increase by 9-12 months of age.<sup>[1]</sup> The histopathology as well as the hemodynamic characteristics of PVD has been described in both simple left-to-right shunts as well as complex shunts such as TGA. Various methods have been proposed to determine the severity of PVD and the probability of its reversal after elimination of the shunt. These include the use of clinical, echocardiographic and radiographic methods of assessing size of shunt, hemodynamic methods of measuring pulmonary vascular resistance (PVR), reversibility studies using vasodilators, and histological examination of biopsy specimens for studying the extent and grade of PVD. Decision-making is simple at the two ends of the spectrum of PVD in TGA-VSD. None of these methods have proved entirely reliable in predicting the outcome of patients in the middle of the spectrum. Flow and resistance assessment by cardiac catheterization in TGA-VSD, has major technical limitations and sources of error. Structural reversibility of PVD as judged by lung biopsy specimens does not differentiate survivors from nonsurvivors of surgical repair.<sup>[1]</sup> Moreover, operability in the present era does not mean just survival and discharge from hospital. The question is whether the PVD resolves and the PVR returns to near normal levels. In our experience cardiac catheterization done a year after repair of ventricular septal defects or aorta-pulmonary shunts in patients

with preoperatively elevated PVR (> 4.0 units) hardly ever shows a substantial fall in PVR. This is true even if the preoperative study had shown a fall in PVR with oxygen or nitric oxide. Such patients may look clinically well although their response to stress is often abnormal. The long-term prognosis remains guarded in all patients with PVD, and more so in the setting of TGA-VSD. Early repair is the only proven way of achieving satisfactory resolution of pulmonary hypertension in TGA-VSD as well as other shunt lesions.

Bajpai *et al*,<sup>[2]</sup> in this issue of the Journal propose the use of oxygen to test the reactivity of the pulmonary arteriolar bed in children with TGA-VSD and thereby predict the operability of such patients. The authors should be complemented for focusing on a difficult but relevant question. They have rightly referred to the problems of late presentation of TGA-VSD patients in developing countries and the limitations in the use of hemodynamic or histological quantification of PVD.<sup>[1-3]</sup> However their methods and conclusions can be debated. The authors need to define how they measured on echocardiography, parameters like increased pulmonary venous blood flow, increased aortic flow reversal, left to right or right to left shunt across the ventricular septal defect. All septal defect shunt flows are bidirectional depending upon the phase of the cardiac cycle, especially so in patients with TGA-VSD. Pulmonary and systemic blood flow could have been estimated by Doppler interrogation of the ascending aorta and pulmonary artery. Left atrial and left ventricular dimensions can be useful indicators of PBF. While flow measurement by itself is not a totally reliable surrogate for vascular resistance, relative changes in pulmonary and systemic blood flow could give some idea of the changes in pulmonary and systemic vascular resistances as described by the authors.

Some novel Echo-Doppler methods have been proposed recently for the estimation of PVR. Parameters being investigated include the time interval from valve opening to peak velocity in the pulmonary artery, the interval between pulmonary artery peak velocity and peak tricuspid velocity, the right ventricular pressure increase after peak velocity in the pulmonary artery and the

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ratio of the peak tricuspid regurgitation velocity to the right ventricular outflow tract velocity-time integral.<sup>[4-6]</sup> Input impedance of the pulmonary vasculature obtained from pulse-wave Doppler and a single catheter pressure measurement estimates right ventricular afterload (which includes pulmonary arterial compliance), and is reported to be a better predictor of patient outcomes compared with PVR alone.<sup>[7]</sup> A completely noninvasive means of assessing pulmonary arterial compliance using color M-mode Doppler tissue imaging has also been reported.<sup>[8]</sup> Velocity-encoded cine MRI is another promising technique to derive pulmonary artery flow volumes and to compute PVR.<sup>[9]</sup> It would be interesting to see if any of these newer methods or parameters are useful to assess PVD in children with TGA-VSD or with other congenital shunt lesions.

Many pulmonary vasodilators have been used to test the reactivity of PVD. Oxygen is widely available and effective in high concentrations as a pulmonary vasodilator. It is important to obtain observations during the oxygen administration instead of doing a post-oxygen study, as its action ceases almost as soon as it is stopped. It is difficult to administer high levels of oxygen by mask to small babies on a continuous basis for 48 hours. Acute studies using a high inspired concentration of oxygen or inhaled nitric oxide would therefore be necessary to scientifically document the pulmonary vascular reactivity in TGA-VSD. It would also be interesting to see the effects of other pulmonary vasoactive medications such as sildenafil or prostacycline in children with TGA-VSD and pulmonary hypertension.

The use of echocardiography to assess pulmonary vascular reactivity in TGA-VSD as reported is an interesting concept, but only a prospective study in a larger number of patients using refined quantitative echocardiography methods would allow meaningful conclusions. To be useful in a clinical dilemma, the parameter and the outcome should be quantifiable and be tested for sensitivity, specificity and predictive accuracy. PVD is a complex and multifactorial condition with a variable rate of progression. It raises the difficult clinical question of operability and reversibility, and I doubt if it can be solved by too simple an answer.

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