A newborn male was transferred for severe cyanosis and suspected transposition of the great arteries and ventricular septal defect (VSD). An emergency balloon septostomy was performed. The position and commitment of VSD and great arteries were more precisely defined by echocardiography. There was double-outlet left ventricle (DOLV) with doubly committed VSD, l-malposition of the great arteries, and pulmonary stenosis. The infundibular septum was virtually absent. A modified Blalock–Taussig shunt was inserted before a Rastelli-type corrective surgery was performed at 20 months of age. The VSD was closed, the pulmonary trunk was transected, and a right ventricle–pulmonary artery conduit was placed. Intraoperative findings confirmed the diagnosis. Perioperative assessment was done noninvasively (Figures 1 and 2 and Movies I and II). Mild subaortic stenosis was found and attributed to bulging of the VSD patch. Otherwise, the intracardiac findings were excellent. When the patient was 5 years old, the conduit became stenotic. Again, the intra- and extracardiac anatomy, including the conduit’s aspect and its relation to the sternum, could be seen clearly with the use of noninvasive imaging techniques (Figure 3 and Movie III). The postoperative course has been uneventful. The boy is now 6 years old.

In contrast to double-outlet right ventricle, DOLV is a rarity. After some dispute, its existence was finally verified intraoperatively and in autopsy series. DOLV with doubly committed VSD and l-malposed great arteries comprises fewer than 10% of cases in the largest series. None of these patients had pulmonary stenosis. The anatomic features of this case may not be compatible with traditional pathogenetic explanations of DOLV. DOLV has traditionally been difficult to diagnose accurately. Angiography has often been considered indispensable, and diagnosis has often been left to the surgeon or pathologist. DOLV may thus have been underdiagnosed. Today, cardiac magnetic resonance imaging as an adjunct to echocardiography provides a reliable and sensitive noninvasive diagnostic technique. It allows for comprehensive, detailed intra- and extracardiac assessment independent of echocardiographic windows. Invasive imaging methods should be mostly avoidable.

Disclosures

None.

References

Figure 1. Echocardiography, parasternal long-axis view. Both great arteries are committed to the left ventricle (LV). The aorta (Ao) is malpositioned anteriorly and to the right of the stenotic pulmonary artery (PA). The ventricular septal defect has been closed.

Figure 2. Magnetic resonance imaging, axial scan, gradient echo. The intracardiac anatomy is visualized in detail. There is mild subaortic stenosis. The pulmonary artery (PA) has been transsected. Residual flow is detected across the pulmonary valve. LV indicates left ventricle; Ao, aorta.

Figure 3. Magnetic resonance imaging, sagittal scan, spin echo (gradient echo). The right ventricle (RV)-pulmonary artery conduit is severely stenotic at its proximal anastomosis site (asterisk). Its relationship to the sternum is demonstrated.
Double-Outlet Left Ventricle
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