A 6-month-old child with progressive cyanosis was referred to our service for assessment. Transthoracic echocardiography confirmed the diagnosis of transposition of the great arteries, a very large ventricular septal defect (VSD), and subpulmonary stenosis. The atrial septum was intact. The transcutaneous oxygen saturation levels measured 40% to 50% in room air with a hemoglobin value of 19 g/dL. Conventional treatment with an arterial switch and VSD closure was not possible, because it would result in neonatal obstruction because of the significant subpulmonary stenosis. Additionally, the size of the VSD was such that achieving adequate septation of the ventricles remained questionable. To aid surgical planning, a cardiac magnetic resonance scan was performed to determine whether the low transcutaneous oxygen saturation levels were due to poor mixing of blood or limited pulmonary blood flow.

The scan was performed on a commercial 1.5T scanner (Achieva; Philips Healthcare, Best, The Netherlands) with a 2-channel coil. The child was awake and self-ventilating with qualitative information. Quantitative assessment demonstrated adequate pulmonary blood flow, Qp:Qs 1.4:1.0, indicating that this was not the limiting factor for oxygenation. Through visualization of the pathline data (Figure 2, Movie I in the online-only Data Supplement), very little mixing of blood between the 2 ventricles was seen. Thus, unfavorable streaming conditions were responsible for the low oxygen saturations.

Transposition of the great arteries is characterized by ventriculoarterial discordance. The aorta arises from the right ventricle, and the pulmonary artery arises from the left ventricle. The presence of a VSD is a common association. An intracardiac shunt permits mixing of the 2 parallel streams of oxygenated and deoxygenated blood. In the absence of a shunt, once there is closure of the ductus arteriosus or restriction of the atrial septal communication, the mixing of blood is unable to occur, leading to progressive cyanosis and death. Although the presence of a large VSD may provide a substrate for intracardiac shunting, this can be limited owing to unfavorable streaming of flow within the ventricle. This phenomenon has been described in diagnostic catheter studies, but it has not been visually demonstrated.

The preferred method of surgical repair for transposition of the great arteries with VSD is an arterial switch and VSD closure. Alternatively, where there is subpulmonary stenosis, a Rastelli procedure in which the left ventricle is baffled to the aorta through the VSD and a right ventricle to the pulmonary artery conduit can be performed. This patient had complex anatomy contraindicating either of these options. Determining unfavorable streaming as the cause of the low transcutaneous oxygen saturations indicated that any intervention to increase pulmonary blood flow would not improve the patient’s condition. A decision was made to improve the mixing of blood by performing an atrial septectomy. The septectomy was successfully performed the following day with a resultant improvement in transcutaneous oxygen saturation levels to 70% to 80% in room air.

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Two-dimensional phase-contrast sequences quantify flow without giving information on the intricacies of flow patterns. Accelerated 4D flow techniques offer the opportunity to acquire both of these parameters in a timely fashion and are applicable in a clinical setting.

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Disclosures
None.

References

Figure 1. MR angiogram overlaid over axial SSFP image. There is transposition of the great arteries with the aorta arising anteriorly from the right ventricle and the pulmonary artery arising from the left ventricle. MR indicates magnetic resonance; and SSFP, steady-state free-precession.

Figure 2. Qualitative analysis of pathline data. Oxygenated blood (red) enters the right ventricle and predominantly flows into the pulmonary artery. Deoxygenated blood is shown in blue. Despite the presence of a large VSD, there is little mixing of blood, explaining the low transcutaneous oxygen levels. VSD indicates ventricular septal defect.
Streaming in Transposition of the Great Arteries by Using Cardiac Magnetic Resonance Imaging
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**Movie Legend**

**Movie 1.** 4D flow analysis overlaid over axial cine images in a child with transposition of the great arteries. The aorta arises anteriorly from the right ventricle and the pulmonary artery from the left ventricle. The atrial septum is intact but there is a large VSD. Despite the large VSD pathline analysis demonstrates that limited mixing of blood is responsible for the low transcutaneous oxygen saturation levels. There is adequate pulmonary blood flow. Best viewed with Windows Media Player.