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Tetralogy of Fallot with Left Heart Hypoplasia, Total Anomalous Pulmonary Venous Return, and Right Lung Hypoplasia: Role of Magnetic Resonance Imaging

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Abstract. We report a rare case of tetralogy of Fallot with total anomalous pulmonary venous return, left heart hypoplasia, right lung hypoplasia, and left ocular–mandibular synchinesia (Marcus-Gunn phenomenon), correctly diagnosed by cardiovascular magnetic resonance imaging and successfully operated by modified Glenn anastomosis.

Key words: Total anomalous pulmonary venous return — Tetralogy of Fallot — Cardiovascular MRI

Tetralogy of Fallot (TOF) associated with total anomalous pulmonary venous return (TAPVR) is rare (0.25% of TOF) [2, 4]. In one case, TAPVR was a surgical unexpected finding despite preoperative cardiac catheterization [3]. The purpose of this case report is highlight to the capability of magnetic resonance imaging (MRI) to delineate pulmonary veins anomaly more accurately than x-ray angiography.

Case Report

A full-term Caucasian girl, born by normal delivery with birth weight of 3.260 kg, was admitted to a neonatal intensive care unit because of cyanosis and reduced right lung expansion. On chest x-ray the right lung was almost completely opacified and the mediastinum deviated to the right.

A diagnosis of TOF with persistent left caval vein draining into the coronary sinus was made on two-dimensional echocardiography. The left atrium and left ventricle were very small. The aortic valve and the ascending aorta overriding the ventricular septal defect (VSD) were slightly dilated as expected in TOF, and TAPVR was suspected. Because of a hypoxic spell, at 1 month of age she was started on propranolol (3 mg/kg/day). At 3 months of age, her physical examination showed good general conditions, weight 5.8 kg, mild cyanosis at rest (transcutaneous saturation 80% on room air), no hepatomegaly, and lung auscultation was normal bilaterally. On cardiac auscultation, there was a 3/6 ejection systolic murmur on the midprecordial area. Cardiac catheterization confirmed the diagnosis of TOF, TAPVR, to the right atrium (Fig. 1), right aortic arch and normal-size pulmonary artery branches. However, angiography failed to fully demonstrate the connection between the pulmonary veins and the atria. To better delineate the anatomy of the anomalous pulmonary venous return and to calculate left ventricular volumes, cardiovascular MRI was performed at 6 months of age (weight, 7.5 kg) under general anesthesia. A GE Signa/CV 1.5 T magnet was used.

The exam showed the following: an unusual anomalous pulmonary venous return consisting in two left pulmonary veins and a single right pulmonary vein (upper) draining into a common collector located posterior to the heart close to the opening of the inferior caval vein (Figs. 2 and 3); left superior caval vein draining into a huge coronary sinus; a very small left atrium and a hypoplastic left ventricle (Fig. 4) (telediastolic volume = 28 cc/ms; z-value, −3.2); a normal aortic valve and good size pulmonary artery branches.

At 7 months of age, the patient underwent cardiac surgery and the MRI findings were confirmed. The pulmonary collector had a horizontal posterior course for approximately 1 cm and then was joined by two left pulmonary veins and a single right pulmonary vein. The small left atrium and the huge coronary sinus made unfeasible the anastomosis between the pulmonary venous collector and the left atrium. Therefore, a repair of the anomalous venous return and the TOF was avoided and a bicavobilateral pulmonary anastomosis was constructed leaving an antegrade flow through the native severely stenotic pulmonary artery.

The postoperative course was uneventful. The patient was weaned from the ventilator and discharged from the hospital on postoperatively days 2 and 13, respectively. She is on diuretic treatment. Her transcutaneous saturation on room air is approximately 90%.

Discussion

Cardiovascular MRI, because it is relatively noninvasive and has a high probability of delineating cardiac and vessel anatomy, is being increasingly used in pediatric cardiology [1]. This case demonstrates how crucial the MRI study was in detecting the unusual

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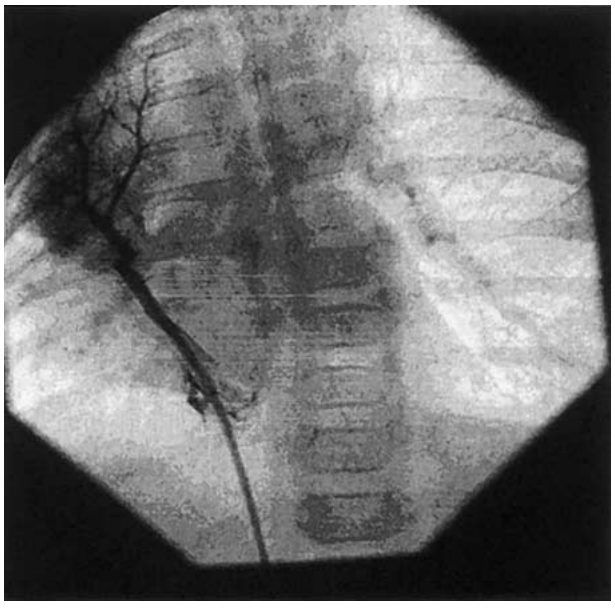

Fig. 1. X-ray angiography AP projection. Injection in the right pulmonary vein reached from the inferior vena cava through the pulmonary veins collector.

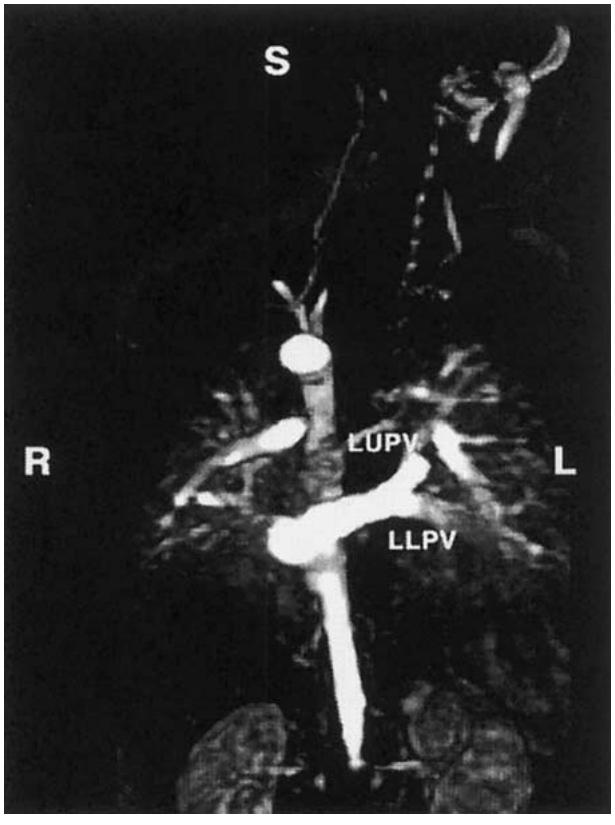

Fig. 2. MR angiography. *LLPV*, left lower pulmonary vein; *LUPV*, left upper pulmonary vein.

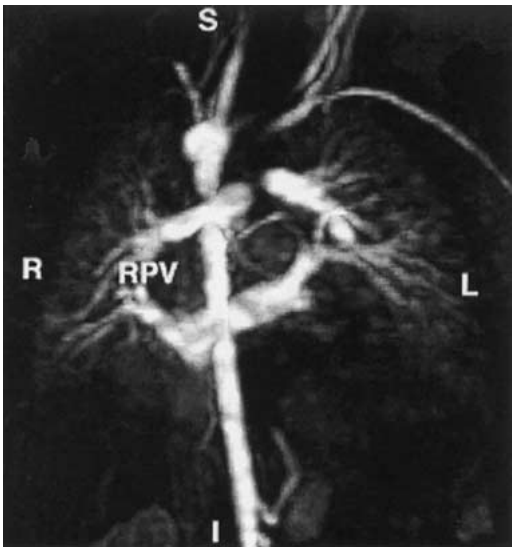

Fig. 3. MR angiography. *RPV*, right pulmonary vein.

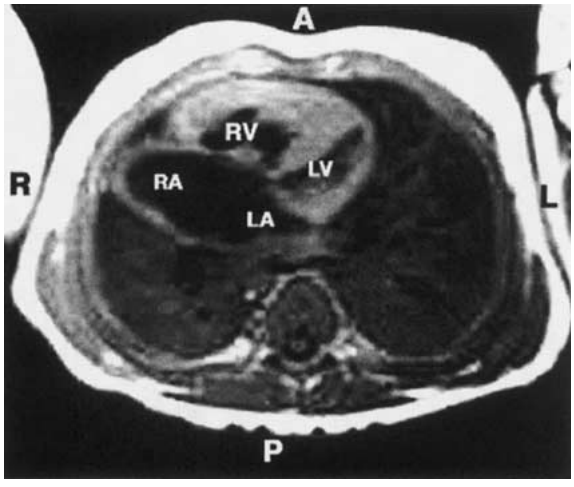

Fig. 4. FSE (black blood): axial cut. Note the small left ventricle and the displacement of the heart to the right. *LA*, left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle.

pulmonary veins. In fact, MRI demonstrated much more clearly than cardiac catheterization the anotomy of the pulmonary veins and the position of the common collector relative to the inferior caval vein, the coronary sinus, and the very small left atrium, which eliminated the possibility of creating an anastomosis between the pulmonary collector and the left atrium. Furthermore, MRI provided a more accurate estimation of left ventricular volume than echocardiography, which was inaccurate due to the extremely enlarged right ventricle.

From a surgical stand point, we believe a bicavo bilateral pulmonary anastomosisa was a reasonable and less risky option than complete correction with regard to the hypoplastic left atrium and ventricle,

particularly the relationship between the pulmonary venous collector, the coronary sinus, and the left atrium.

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References

1. Greil GF, Powell AJ, Gildein HP, Geva T (2002) Gadolinium-enhanced three-dimensional magnetic resonance angiography of pulmonary and systemic venous anomalies. *J am Coll Cardiol 16*:335–341
2. Muster AJ, Paul MH, Nikaidoh H (1973) Tetralogy of Fallot associated with total anomalous pulmonary venous drainage. *Chest 64*:323
3. Neema PK, Krishnamanohar SR, Rathod RC (2002) Tetralogy of Fallot with total anomalous pulmonary venous connection: pathophysiology and management. *J Cardiothorac Vasc Anesth 16*:211–213
4. Redington AN, Raine J, Shinebourne EA, Rigby ML (1990) Tetralogy of Fallot with anomalous pulmonary venous connections: a rare but clinically important association. *Br Heart J 64*:325