MRI EVALUATION OF THE SYSTEMIC-PULMONARY ARTERIAL SHUNTS IN PATIENTS WITH PULMONARY ARTERY STENOSIS OR COMPLEX PULMONARY ARTERY ATRESIA

R.L. Dumitru, R. Capsa, I. Lupescu

Department of Radiology and Medical Imaging, Fundeni Clinical Institute
"Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

Abstract Introduction and Purpose. To evaluate the role of MRI in the study of the pulmonary artery (PA) and the system of aorto-pulmonary collateral vessels, in patients with tetralogy of Fallot or different forms of variants.

Methods. Retrospective study covering the period 1 January 2001 - June 1, 2013. We have studied the clinical and imaging data of 32 consecutive patients (diagnosed with tetralogy of Fallot or different forms of variant pulmonary stenosis), sent for MRI examination. The goal of the MRI examination was to depict the anatomy of the pulmonary vascular system and to image the aortopulmonary collaterals. Patients followed a protocol that included transthoracic echocardiography (TTE) or transoesophageal echocardiography (TEE) and angiocardiology/ selective pulmonary artery catheterization imaging data was compared with intraoperative results in all cases that required surgery.

Results: After the MRI findings, we classified patients in several groups according to the diagnosis: tetralogy of Fallot with pulmonary atresia (n = 14), tetralogy of Fallot with pulmonary stenosis (n = 10), trilogy of Fallot (n = 4), pentalogy of Fallot (n = 3) and single ventricle involving pulmonary artery stenosis. (n = 1). Morphology and size of central pulmonary arteries, sources of the major aortopulmonary collateral vessels, Blalock Taussig shunt patency and the postoperative appearance of the pulmonary circulation were assessed.

Conclusion: MRI can provide accurate information in assessing systemic-pulmonary shunts in patients with complex pulmonary artery stenosis or atresia and it is a useful tool in the preoperative management and subsequent follow-up.

Keywords: Cardiac, Cardiovascular system, Pulmonary vessels, MR, MR-Angiography, Hemodynamics / Flow dynamics

Introduction

Pulmonary valve stenosis is an usual anatomic feature of most cyanotic congenital heart diseases (CHD). In these cases, the central pulmonary arteries might be hypoplastic or even absent; furthermore, the pulmonary vascular bed might be fed through multiple sources, including multiple

aorto-pulmonary collateral vessels or surgically placed shunts.

Since ultrasound examination is often technically limited and arterial catheterization is an invasive procedure, this study evaluates the efficiency of magnetic resonance imaging (MRI) as a noninvasive tool in diagnosis of pulmonary artery anatomy (PA) and aorto-pulmonary shunts, informations which are

Address for correspondence: R.L. Dumitru, MD

Department of Radiology and Medical Imaging, Fundeni Clinical Institute, Bucharest, Romania, e-mail: radu.dumitru@gmail.com

crucial in determining surgical management and followup of the patients with complex congenital heart disease, involving stenosis or pulmonary atresia.

Purpose

Our main purpose was to evaluate the role of MRI in the study of the PA and the system of aorto-pulmonary collateral vessels and secondary to depict the particular anatomy in patients that reached adulthood, but did not require surgical treatment.

Materials and Methods

Study methodology: retrospective study covering the period 1 January 2004 - June 1, 2013.

Inclusion criteria: We have studied the clinical and imaging data of 32 patients (diagnosed with tetralogy of Fallot or different forms of pulmonary stenosis), sent for MRI examination in the Department of Radiology and Medical Imaging of Fundeni Clinical Institute.

The goal of the MRI examination was to depict the anatomy of the pulmonary vascular system and to image the aortopulmonary collaterals. We have excluded patients with advanced renal disfunction ($GFR < 15 \text{ mL/min/1.73 m}^2$).

Imaging protocol: Patients followed a protocol that also included transthoracic echocardiography (TTE) or transoesophageal echocardiography (TEE) and angiocardiology/selective pulmonary artery catheterization; imaging data was compared with intraoperative results in all cases that required surgery.

All the MRI studies were performed on a GE Signa Horizon 1,5 T machine, using a TORO phased array coil and respiratory and cardiac gating (MRI safe carbon electrodes). In all studies we used administration of a paramagnetic contrast agent. For the morphological assesment of the heart we used SE sequences in at least 2 planes (perpendicular and parallel to the interventricular septum) with slice thickness of 10 mm and spacing of 2 mm. For the angiographic evaluation we used gadolinium enhanced (Gd iv 0,2 mL/kg, flow 3 ml/s) 3DMRA sequences with bolus detection (SMARTPREP-GE); after the acquisition of the source images, multiplanar reconstructions and maximum intensity projection (MIP) and virtual rendering (VR) were performed. For the

Table 1 - Demographic and cardiac diagnostic date in the study group

References: D. L. Radu; Radiology and Medical Imaging Department, Fundeni Clinical Institute, Bucharest, Romania

Sex, male/female	12/20
Median age at MRI (range)	22 (11-36 yrs)
Cardiac diagnoses	
Tetralogy of Fallot with pulmonary atresia	14
Tetralogy of Fallot with pulmonary stenosis	9
Trilogy of Fallot	3
Pentalogy of Fallot	4
Status post palliative Blalock Taussig shunt	3
Status post definitive surgical correction	2

evaluation of the right ventricle (RV) mass, volume and ejection fraction cine MRI sequences were performed.

Results

We included 32 patients, with a mean age of 24 years and a male/female ratio of 1:3 (table 1).

After the MRI exam was performed, we split the initial group in several categories, coresponding to the MRI features observed:

- tetralogy of Fallot with pulmonary atresia (n= 11),
- tetralogy of Fallot with pulmonary stenosis (n= 9),
- trilogy of Fallot (n= 4),
- pentalogy of Fallot (n= 3) (fig 1).

Figure 1 - Origin of the systemic-pulmonary shunts according to the level of the aorta and associated anomalies.

References: Radiology and Medical Imaging Department, Fundeni Clinical Institute, Bucharest, Romania

Figure 2 - 3D MRA. MIP reconstruction oblique coronal view in a 25 year old patient with Tetralogy of Fallot, showing spontaneous pulmonary revascularization with multiple systemic- pulmonary collateral vessels at a parietal thoracic level. PA- pulmonary artery, Ao-Aorta, arrow- collateral vessels

References: Radiology and Medical Imaging Department, Fundeni Clinical Institute, Bucharest, Romania

Figure 3 - Oblique coronal SE sequence and MIP oblique sagittal reformat (fig 5). 19 year old patient with tetralogy of Fallot and palliative correction with Blalock Taussig shunt which is partially occluded and with an aneurysmal aspect (arrow). LA- left atrium, LV- left ventricle.

References: Radiology and Medical Imaging Department, Fundeni Clinical Institute, Bucharest, Romania

Five patients had undergone previous shunt surgery (3 with paliative Blalock-Taussig shunt - (fig. 3, fig. 4) and 2 with definitive surgical correction). The rest of the patients had spontaneous pulmonary vascularisation, with systemic-pulmonary collaterals.

The aortopulmonary shunts were well demonstrated, with clear delineation of the spatial relationship of the shunts to both adjacent vascular and non vascular structures. In all cases, MRA confirmed the presence or absence of central pulmonary arteries (24 (72%) of 32 patients had central pulmonary arteries).

The shunts were classified either as individual

vessels (2.5 mm diameter) or as multiple smaller vessels that tended to arise in a cluster (each vessel typically 2.5 mm diameter) - (fig. 2). The majority of shunt vessels arose from the descending aorta (63.6% of single aortopulmonary shunts and 66.7% of multiple smaller vessels). In most cases, it was possible to tell whether they supplied a single lobe or entire lung (30% drained into the right upper lobe, and 18.1% supplied the right pulmonary artery).

The most common origin of the systemic-pulmonary shunt was at the level of the descending aorta (63% in patients with tetralogy of Fallot) - (fig. 5).

Figure 4 - MIP oblique sagittal reformat - same patient as figure 4.

References: Radiology and Medical Imaging Department, Fundeni Clinical Institute, Bucharest, Romania

Figure 5 - MIP reconstruction. 21 year old patient with Tetralogy of Fallot and pulmonary stenosis.

Spontaneous pulmonary revascularization through multiple aorto-pulmonary collateral arteries at the parietal thoracic level (arrow).

References: Radiology and Medical Imaging Department, Fundeni Clinical Institute, Bucharest, Romania

Figure 6 - Axial SE sequence and 3D FSPGR post Gd iv. The absence of the pulmonary trunk at the supraventricular level. There is a small vestigial pulmonary artery trunk which is connected to the ascending aorta and then it divides into two hypoplastic branches that provide the vascularization of both lungs. In the oblique coronal reformat, aorto-pulmonary collateral vessels between the descending aorta and inferior lobar branches (Ao- aorta, PV- pulmonary veins, arrow - right pulmonary artery)

References: Radiology and Medical Imaging Department, Fundeni Clinical Institute, Bucharest, Romania

Discussion

This study assessed the value of MRI both with and without Gd administration in the morphologic assessment of the aortopulmonary shunts in a predominantly adult population. Early detection and treatment are associated with a better outcome but require accurate demonstration of the anatomic distribution of the anomalous vessels, which is also important for serial evaluation [1].

Both the initial diagnosis and the management during follow-up of congenital heart disease depend on an accurate depiction of cardiac anatomy and function [1,2]. It is often difficult to obtain sufficient information with transthoracic echocardiography in adolescents and adults because they have larger chests and hearts and because of the natural reduction of the ultrasound (US) window. Special problems occur in patients who have undergone midline thoracotomy and have acquired precordial fibrotic tissue [1,3].

Several groups have reported their findings in detecting cardio vascular anomalies using MRA, but in contrast to the present study (9), these were either in predominantly pediatric populations or were retrospective studies [4,5,6,7]. Geva et al [4] demonstrated the accuracy of MRA in the delineation of all sources of pulmonary blood supply in patients with complex pulmonary stenosis and atresia compared with diagnostic catheterization with x-ray angiography. Although some adult patients were included in their series, the median age range was 4.7 years, unlike the present study. In a prospective study, Sanjay et al. [5] showed that contrast-enhanced 3D MRA is a safe, rapid, noninvasive and robust method to detect vascular anomalies in adult patients with CHD with aortopulmonary collateral vessels. In this particular study the mean age was closer to our own study group (31 years) and it depicted the origin and distribution of aortopulmonary collateral vessels, compared with classical angiography. A degree of pulmonary regurgitation is present in nearly all patients and can be accurately quantified serially with MR imaging flow studies [8].

Conclusion

MRI can provide accurate information in assessing systemic-pulmonary shunts in patients with complex pulmonary artery stenosis or atresia and it is a necessary tool in the preoperative management and subsequent followup.

Furthermore, we showed that MR is valuable imaging method in the follow-up of adult patients and presurgical planing in the same category of patients.

Although MRI is an indispensable and non-invasive diagnostic tool, careful correlation with the echocardiography and angiographic is recommended to be performed in each case.

References

1. Uta C. Hoppe, Birgit Dedenchs, Hans J. Deutsch, Peter Theissen, Harald Schicha, Udo Sechtem-Congenital Heart Disease in Adults and Adolescents: Comparative Value of Transthoracic and Transesophageal Echocardiography and MR Imaging;Radiology 1998;669-677
2. Dominique Didier, Charles B. Higgins, Madeleine R. Fisher, Luci Osaki, RNNorman H. Silverman, Melvin D. Cheitlin-Congenital Heart Disease: Gated MRI in 72 Patients-Radiology 1986, 158:227-235
3. Greutmann M, Tobler D, Biaggi P, Mah ML, Crean A, Oechslin EN, Silversides CK.-Echocardiography for assessment of right ventricular volumes revisited: a cardiac magnetic resonance comparison study in adults with repaired tetralogy of Fallot. J Am Soc Echocardiogr. 2010 Sep;23(9):905-11
4. Sanjay K. Prasad, Nikolaos Soukias, Timothy Hornung, Mohammed Khan, Dudley J. Pennell, Michael A. Gatzoulis, and Raad H. Mohiaddin- Role of Magnetic Resonance Angiography in the Diagnosis of Major Aortopulmonary Collateral Arteries and Partial Anomalous Pulmonary Venous Drainage Circulation 109: 207-214
5. Geva T, Greil GF, Marshall AC, et al.- Gadolinium-enhanced 3-dimensional magnetic resonance angiography of pulmonary blood supply in patients with complex pulmonary stenosis or atresia: comparison with x-ray angiography. Circulation. 2002; 106: 473-47
6. Holmqvist C, Hochbergs P, Bjorkhem G, et al.- Pre-operative evaluation with MR in tetralogy of Fallot and pulmonary atresia with ventricular septal defect. Acta Radiologica. 2001; 42:63-66
7. Boechat MI, Ratib O,Williams PL, et al. Cardiac MR Imaging and MR Angiography for Assessment of Complex Tetralogy of Fallot and Pulmonary Atresia1 RadioGraphics 2005; 25:1535-1546.
8. Babar JL, Jones RG, et al. Application of MR Imaging in Assessment and Follow-up of Congenital Heart Disease in Adults. RadioGraphics 2010, 10.1148/rg.e40.
9. Dumitru L. Radu, Lesanu R, Capsa RA, Lupescu GI, Georgescu S. MRI evaluation of the systemic-pulmonary arterial shunts in patients with pulmonary artery stenosis or complex pulmonary artery atresia, ECR, 2011, doi 10.1594/ecr2011/C-2028.