Tetralogy of Fallot with Absent Pulmonary Valve: Evaluation with Magnetic Resonance Imaging

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KAKIZAWA, H., OHNO, T., OZAWA, A., TANAKA, T. and IINUMA, K. *Tetralogy of Fallot with Absent Pulmonary Valve: Evaluation with Magnetic Resonance Imaging*. Tohoku J. Exp. Med., 1997, **182** (1), 35–39 — Two infants with tetralogy of Fallot with absent pulmonary valve were studied with electrocardiographically gated magnetic resonance imaging using a 1.5 T imaging system. In each case, imaging was performed successfully. The magnetic resonance imaging clearly demonstrated a rudimentary pulmonary valve, an aneurysmal pulmonary artery, and compression of the mainstem bronchi. Cine MRI revealed phasic compression of the bronchi by an aneurysmally dilated pulmonary artery. Magnetic resonance imaging produced high quality images and provided all of the anatomic information required for preoperative evaluation. — tetralogy of Fallot; absent pulmonary valve; magnetic resonance imaging
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The syndrome of tetralogy of Fallot (TOF) with absent pulmonary valve in its most severe form is characterized by severe respiratory symptoms in early infancy (McCaughan et al. 1985; Ilbawi et al. 1986; Watterson et al. 1992). Pulmonary annular stenosis, in association with an absent or rudimentary pulmonary valve, causes pulmonary regurgitation, and markedly dilated pulmonary arteries compress the bronchi (Rabinovitch et al. 1982). The treatment is difficult due to airway obstruction, and there is a high infant mortality rate. This complex has been diagnosed by echocardiography and angiography. However, it has been difficult to evaluate the characteristic anatomy of bronchial compression using these methods.

In this report, we describe our experiences using magnetic resonance imaging (MRI) in two patients with TOF with absent pulmonary valve.

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SUBJECTS AND METHODS

Case report

Case 1. This infant boy was born at term, with a birth weight of 3.2 kg, following an uneventful pregnancy. Mild cyanosis and tachypnea were noted from birth. Two-dimensional and Doppler echocardiography at 3 days of age revealed a ventricular septal defect, anterior displacement of the infundibular septum, pulmonary stenosis, a markedly dilated main pulmonary artery, and a rudimentary pulmonary valve, suggestive of TOF with absent pulmonary valve. MRI was performed at 14 days of age, and cardiac catheterization and angiography at 24 days of age, which confirmed this diagnosis. Gradually, he developed dyspnea after 1 month of age and required intubation and ventilation at 40 days of age. Emergency surgery, consisting of a primary repair with a transannular patch and plication of the right pulmonary artery, was performed the following day.

Case 2. This infant boy was born at term, with a birth weight of 2.9 kg, following an uneventful pregnancy. A cardiac murmur and tachypnea were noted from birth. Two-dimensional and Doppler echocardiography performed at 1 day of age revealed TOF with absent pulmonary valve. MRI was performed at 11 days of age. He had developed dyspnea in a supine position by 12 days of age and was sedated in a prone position. Therefore, cardiac catheterization and angiography were postponed, and an emergency surgery, consisting of a primary repair with a transannular patch and plication of the right pulmonary artery, was performed at 17 days of age.

Magnetic resonance imaging

MRI was performed using a 1.5 Tesla system (MAGNETOM, Siemens, München, Germany) with a head coil. Patients were imaged following the administration of a sedative. The sedation regimen consisted of 80 mg/kg of oral chloral hydrate, 30 to 40 min prior to the study. Spin-echo images were acquired with electrocardiographic gating of every heartbeat, so that the repetition time (TR) was determined by each subject's heart rate. The echo time (TE) was 15 msec. A 192×256 matrix was used. Each imaging section was 5 mm thick, and the signals were averaged four times. Axial, coronal, and sagittal imaging planes were acquired. In Case 2, cine MRI was performed in a coronal plane using the technique of fast imaging with steady precession (FISP) scanning with a flip angle of 35° , and a TE of 12 msec.

RESULTS

Images of diagnostic quality were obtained, even though the patients were slightly tachypneic. The electrocardiographic gating was satisfactory. The ventricular septal defects and overriding of the aorta were best visualized on the

sagittal sections in each case. The characteristic anatomies, namely the rudimentary pulmonary valve, the aneurysmal pulmonary artery, and the compression of the mainstem bronchi, were visualized particularly well on the coronal and the axial sections (Fig. 1). In each case, the extent of the bronchial compression could be evaluated with these sections, which was consistent with intraoperative findings. The ligamentum arteriosum was not detected.

Cine MRI in the axial plane demonstrated phasic compression of the bronchi by the markedly dilated pulmonary artery in the patient in Case 2 (Fig. 2).

Fig. 1. Coronal MRI of Case 1, revealing a markedly dilated pulmonary artery (A). The bronchi are compressed bilaterally, especially the right. The right intrapulmonary bronchus is dilated (B). Axial MRI revealing a rudimentary pulmonary valve and an enlarged pulmonary artery (C).

Fig. 2. Axial MRI of Case 2. The bronchus is compressed at the bifurcation site (white arrow) between pulmonary artery and vertebra. Cine MRI revealing the phasic compression of the bronchi by the dilated pulmonary artery. Systolic phase (A). Diastolic phase (B).

DISCUSSION

In this report, we describe for the second time the efficacy of MRI in evaluating the preoperative status of patients with TOF with absent pulmonary valve (Frank et al. 1996). The etiology of this syndrome is unknown. Emmanouilides et al. has proposed the hypothesis that the absence of the ductus arteriosus in fetal life hemodynamically induces the absence of the pulmonary valve (Emmanouilides et al. 1976; Fischer et al. 1984). Momma et al. (1990) have reported that rat models of absent pulmonary valves, induced with bis-diamine, were all associated with an absent ductus arteriosus, supporting the hypothesis by Emmanouilides et al. (1976). MRI supplies anatomic detail of structures not observed with angiography, such as the ligamentum arteriosum (Azarow et al. 1992). In each of our cases, the ligamentum arteriosum was not detected by MRI. The intraoperative diagnosis of an absent ductus arteriosus was made in each case.

The surgical management of severely symptomatic infants with this syndrome has been controversial (McCaughan et al. 1985; Ilbawi et al. 1986; Watterson et al. 1992). The main cause of airway obstruction is dilatation of the central pulmonary artery due to pulmonary regurgitation. Therefore, the aim of the surgery is to control the pulmonary regurgitation and to relieve airway obstruction. Preoperative evaluation is important for confirming both the cardiac structural anomaly and the severity of the bronchial compression. The MRI in this report, particularly the coronal and axial sections, clearly demonstrated a rudimentary pulmonary valve, an aneurysmal pulmonary artery, and compression of the mainstem bronchi. In Case 2, cine MRI revealed phasic compression of the bronchus by the aneurysmally dilated pulmonary artery. No other diagnostic methods can provide such information on both the heart and bronchus, simultaneously.

It is preferable to avoid invasive methods for the preoperative evaluation of such a syndrome. The ventilatory status of the patients must be stabilized prior to surgical intervention. In this regard, Ozkutlu et al. (1992) are of the opinion that the two-dimensional and Doppler echocardiographic methods are the most reliable tests for diagnosing absent pulmonary valve syndrome, and that severely symptomatic infants can be referred for surgery without catheterization. MRI can provide the further details needed to treat patients with TOF with absent pulmonary valve noninvasively. Therefore, our present policy on the preoperative evaluation is to perform MRI as soon as possible before the respiratory status is deteriorating.

Our early experiences indicate that MRI provides excellent visualization of the anatomy of TOF with absent pulmonary valve and bronchial compression. Thus, MRI is a good method for the noninvasive evaluation of patients with such a syndrome.

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