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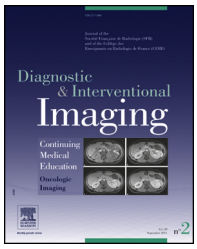

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Tetralogy of Fallot: Preoperative assessment with MR and CT imaging

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Abstract Tetralogy of Fallot has a broad anatomical spectrum. In mild forms of the condition the obstruction is only located in the right ventricular infundibulum, whereas in severe forms the pulmonary valve is atretic, the pulmonary arteries are absent and the lung is supplied by aorto-pulmonary collateral arteries. Surgical management differs from conventional surgery in the former situation, whereas in the latter it is complex and requires reconstruction of the pulmonary arteries (unifocalization) carried out in more than one stage and with a high morbidity rate. The key factors to establish before corrective surgery are the levels and degree of obstruction of the right ventricular outflow tract, the development of the pulmonary arteries and the presence of collateral arteries. The main role of magnetic resonance imaging along with that of computed tomography angiography are discussed and illustrated.

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease (CHD) and represents 5% to 7% of all CHD. TOF has an estimated incidence of 0.5/1000 live births. It is also the most frequent complex CHD in adulthood. Complete correction involves closing the ventricular septal defect and broadening the pulmonary outflow tract. ''Timing'', type of broadening and prognosis of surgery are determined by the level of obstruction of the pulmonary outflow tract and the patient's clinical condition.

In pediatric patients, particularly in newborns, transthoracic echocardiography provides a good assessment of the preoperative cardiac anatomy. In cases in which echocardiography is insufficient, however, imaging, mainly with magnetic resonance imaging (MRI),

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represents a less invasive alternative to cardiac catheterization and provides the requisite information for diagnosis and surgical planning [1—6]. In order to assist the multidisciplinary team in the process of deciding on treatment, radiologists must have a good knowledge of the disease and corrective surgical techniques for TOF, particularly as this is one of the heart diseases for which complementary imaging is most often requested.

The goal of this article was fourfold. First, we wanted to describe and illustrate the anatomical spectrum of TOF and its related abnormalities. Second, we wished to clarify the information to be obtained from computed tomography (CT) and/or MRI before surgery and offer a practical approach to the preoperative assessment of TOF. Third, we aimed to review the differential diagnoses of heart disease with right ventricular outflow tract (RVOT) obstruction and concomitant ventricular septal defect. Finally, we wanted to briefly discuss the place of palliative forms of surgery and their complications.

Anatomical description

Embryologically, according to Van Praagh's theory, TOF is secondary to an antero-superior deviation of the conal septum (Fig. 1). This misalignment results in four characteristic anatomical findings [7]. The classical anatomical description of TOF given by Etienne-Louis Arthur Fallot, is an obstruction of the pulmonary outflow tract, a ventricular septal defect (VSD) due to misalignment, an overriding aorta and right ventricular hypertrophy. Stenosis of the infundibulum or right ventricular outflow tract (RVOT) is an integral part of the condition and has a dynamic component. It is usually more pronounced in the proximal region (Figs. 2 and 3) although the pulmonary obstruction may occur at one or more levels in addition to the infundibulum. The pulmonary valve and main pulmonary artery and/or its branches can also be obstructed at various levels. The pulmonary valve is often abnormal with a varying degree of hypoplasia of the valve ring, and thickened, fused leaflets producing valve or stenosis atresia. The proximal pulmonary arteries may be atretic, hypoplastic or stenotic (Fig. 4).

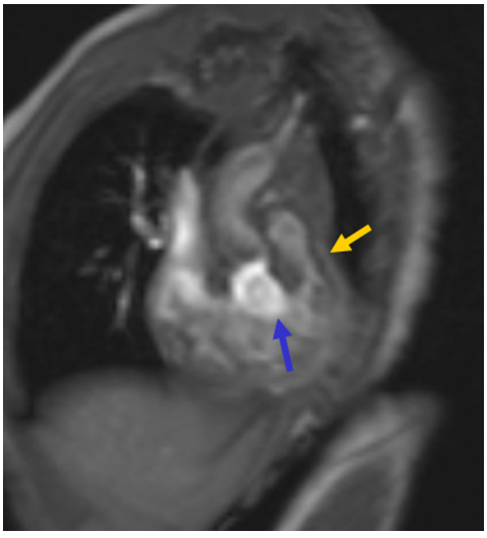

Figure 1. Classical tetralogy of Fallot with an obstructive infundibular component on cardiac MR examination. 2D-Flash Cine MR image in coronal oblique view shows antero-superior displacement of the conal septum causing moderate stenosis of the right ventricular outflow tract, which is more pronounced in the proximal region (yellow arrow) and a VSD due to misalignment (blue arrow).

There is therefore a broad spectrum of abnormalities, ranging from mild obstruction of the RVOT to severe obstruction with pulmonary valve atresia (Figs. 5 and 6), this latter situation being present in 7% of TOF. In severe cases, pulmonary flow is maintained by a patent ductus arteriosus when the main pulmonary artery and its branches are intact and/or by major aorto-pulmonary collateral arteries (MAPCAs) when the pulmonary arteries are absent or very hypoplastic. The MAPCAs are present from birth and are alternatives to the systemic pulmonary arterial supply. They usually arise from the descending thoracic aorta but may also take their origins from the subclavian or coronary arteries and the abdominal aorta (Fig. 7). Anastomosis with the intra-pulmonary arteries usually occurs close to the hilum and intimal stenosis may be found at this level and/or more proximally. It is essential, however, to distinguish the MAPCAs from the bronchial arteries which develop in hypoxemia and which tend to increase in size, whereas the MAPCAs remain of the same diameter (Fig. 8).

A rare form of TOF, in which the pulmonary valve leaflets is rudimentary, creating both valvular stenosis and regurgitation, is observed in 2.5% of cases [8,9]. The main pulmonary artery and its branches are secondarily massively dilated (Fig. 9).

Associated abnormalities

Anatomical abnormalities

Coronary artery abnormalities are associated with TOF in 5% of cases [8]. The most common is a right coronary artery arising from the left anterior descending (LAD) artery following an anterior path to the right ventricular infundibulum or a proeminent conal branch of the right coronary artery also passing in front of the infundibulum (Fig. 10). These findings change the surgical approach, involving either the introduction of a conduit between the right ventricle and the pulmonary arteries (RV—PA conduit) rather than right ventricular infundibulectomy because of the possible associated myocardial ischemic damage. Approximately 25% of cases have a right aortic arch, usually in a mirror image [8]. An atrial septal defect (ASD) may be seen in 5% of cases and a left superior vena cava in 11% of cases. Coexistence of TOF and a common atrio-ventricular channel is also seen in patients with trisomy 21.

Genetic abnormalities

TOF is multifactorial in origin. Up to 25% of patients with TOF have a chromosomal abnormality, usually trisomy 21 or a 22q11 deletion. The 22q11 deletion is more common in cases of TOF with a right aorta and/or pulmonary atresia or stenosis and can be associated with an absent thymus (Fig. 11) [9,10].