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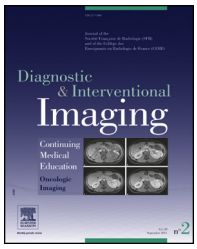

CONTINUING EDUCATION PROGRAM: FOCUS...

Imaging of postoperative tetralogy of Fallot repair

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Abstract Over the last years, the surgical techniques used to repair Tetralogy of Fallot as well as the cross-sectional cardiac imaging techniques have substantially improved. Now, the survival rate after surgical repair is more than 90% at 40 years old. A follow-up is needed and the imaging evaluation should be guided by the surgical techniques used. This article reviews the most common surgical procedures for a complete repair, the associated anatomic and hemodynamic complications and the role of cardiac imaging, mainly magnetic resonance imaging.

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease (CHD) with an incidence of approximately 0.5/1000 live births (5 to 7% of CHDs). Without surgical intervention, most patients die during childhood with a rate of survival of 66% at one year of age, 11% at 20 years and 3% at 40 years. Since the introduction of total surgical repair in 1955, the long-term prognosis for these patients has improved but life expectancy is still reduced by comparison with a normal age-matched population [1]. Total repair within the first months of life is now currently feasible with a mortality rate lower than 2% [2,3]. Nevertheless, complications, residual lesions and sequelae are frequent.

Adult patients with CHD have now outnumbered those under the age of 18 years [4]. Treatment and follow-up of this growing number of TOF patients require the expertise of a multidisciplinary team. The radiologist plays an important role, interpreting many different imaging modalities, thus requiring full understanding of TOF anatomical features, surgical approaches and potential postoperative complications.

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Echocardiography still remains the main diagnostic tool for the evaluation of pre and postoperative CHD. Because of numerous technical limitations, transthoracic echocardiography can fail to assess adequately hemodynamic or anatomic information. Among noninvasive imaging modalities, computed tomography (CT) angiography allows to evaluate cardiovascular structures with an excellent spatial resolution in a very short examination time. Magnetic resonance imaging (MRI) provides a better image quality of intracardiac anatomy, unlimited choice of imaging planes, accurate flow quantification and ventricular function evaluation without the use of ionizing radiation, and can thus be repeated without concerns for radiation toxicity. Chest X-ray still keeps an appropriate place in some cases [5].

The goal of this article is to briefly describe the anatomical features of TOF and the surgical strategies used in patients with this condition, and then review and illustrate the surgical approaches and the postoperative complications through the different imaging modalities.

Anatomic features of TOF

Embryologically, TOF results from underdevelopment of the subpulmonary infundibulum or from the anterocephalad deviation of the outlet septum according to Van Praagh or Anderson theory respectively [6,7]. This misalignment of conal septum leads to four distinct anatomic characteristic features, which include pulmonary outflow tract obstruction, ventricular septal defect (VSD), overriding aortic root and right ventricular hypertrophy. The level of pulmonary obstruction may occur at one or more of the following heart structures: infundibulum (right ventricular outflow tract (RVOT)), pulmonary valve, main pulmonary artery (PA) and/or branch pulmonary arteries. A large spectrum of diseases exists, ranging from mild RVOT obstruction to severe obstruction in the presence of TOF with pulmonary atresia. The natural history of TOF is dictated by the degree of pulmonary outflow tract obstruction.

Surgical strategy

After a diagnosis of TOF in the perinatal period, surgical options include palliation followed by definitive repair or the most frequent, primary (first intention) complete repair. Complete repair is usually performed from 3 to 11 months of age. The modified Blalock-Taussig shunt is the most frequent palliative form of surgery. It is performed when hypoplasia of the PA is severe, the newborn weight is inferior to 2 kg or/and the surgical risk is high. This procedure reduces the clinical cyanosis and enables the growth of the PAs.

Complete repair

A complete TOF repair consists in a VSD closure and in the relief of any pulmonary outflow tract obstruction. The level and degree of pulmonary obstruction will determine the surgical technique [8].

If the pulmonary annulus is hypoplastic, a longitudinal incision is performed through the main PA, pulmonary annulus into the RV infundibulum and reconstructed with a transannular patch, resulting in pulmonary valve regurgitation and RVOT dyskinesia. Cardiac MRI will show denudation of the anterior RVOT wall and bulging of the RVOT and main PA, findings that represent the patch (Fig. 1). As expected, a patch does not contract during the cardiac cycle and appears as an akinetic region on cine images. Delayed contrast-enhanced images can show extensive areas of enhancement due to both fibrotic tissues enveloping the patch and adjacent areas of fibrosis [9].

In patients with an adequate size pulmonary annulus, efforts will be made to spare the pulmonary valve by performing a pulmonary valvuloplasty. In those cases, the main PA is enlarged in addition to either a subvalvular muscular resection or a limited infundibulectomy and the closure of the VSD [10]. Cardiac MRI appearance of the heart after infundibulectomy consists of an intact RVOT myocardium, with little or no evidence of fibrosis on delayed contrast images. On cine MR images, normal RVOT contractility and

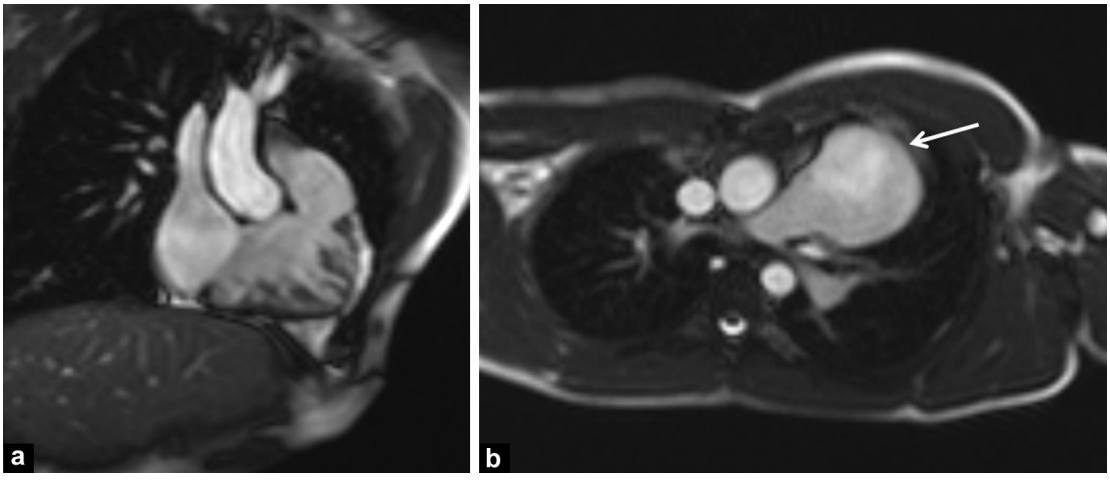

Figure 1. Complete repair of TOF with transannular patch at 6 months of age; a: coronal cine TrueFISP MRI view demonstrates dilatation of the RVOT at the site of transannular patch at 9 y.o.; b: axial oblique cine TrueFISP MRI view shows bulging and anterior denudation of RVOT (arrow) without any residual pulmonary valve.