



## EDITORIAL COMMENT

# Tetralogy of Fallot after repair: A heritage of modern cardiac surgery



## Tetralogia de Fallot após reparação: uma herança da cirurgia cardíaca moderna

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Repair of tetralogy of Fallot (TOF) is one of the greatest achievements of congenital cardiac surgery. The first surgical palliation was in 1944 and repair was introduced 10 years later, more than 60 years ago, before the first coronary artery bypass surgery.<sup>1-3</sup> This was followed by remarkable advances in the many areas that contribute to good surgical results, which have enabled many patients not only to survive, but to live largely normal family and working lives.

In an internal review at our institution 10 years ago, 85% of TOF patients with ages ranging from 18 to 63 years, most of them operated long before, had good functional capacity. This is corroborated by the paper by Cruz et al. in this issue of the *Journal*,<sup>4</sup> in which 90% of patients are reported as asymptomatic. Our review included several women who became mothers. One of them is now a physician working as a general practitioner in the Portuguese hinterland. I myself worked as a fellow in Chicago with a resident pediatric cardiologist who had undergone TOF repair.

Even so, not all patients do well. The paper by Cruz et al. draws our attention again to the problems that may arise in

relation with a dilated ascending aorta (AAo). Cardiologists in charge of patients after TOF repair should be aware of this possibility; they actually need to be aware of quite a significant number of possible negative developments.

Indeed, personal attention to each individual person remains a major aspect of clinical medicine. Regardless of how important averages, standard deviations and percentiles may be, the subject of medical practice is still that person sitting in front me with his/her own and unique condition.

He needs that I understand what he suffers from, even if he is not able to provide me with a full medical history. I will recall some of the problems he may have.

Did he start by having palliative surgery, or did he have a primary repair? (You may get help by looking at his chest. Does he have a lateral thoracotomy scar? Or a sternotomy scar, or both? This might be indicative, although current central palliative shunts are often done via a sternotomy). If he had a Blalock-Taussig anastomosis, was this a modified or a classic anastomosis? Was the subclavian artery sacrificed? Can blood pressure be measured reliably in the ipsilateral arm? Did he suffer from complications of palliation? Could he have diaphragmatic palsy due to phrenic nerve injury? Could he have pulmonary artery branch stenosis? Did he have, or does he need a ventilation/perfusion scan? What type of corrective surgery was performed? Did he have

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a transannular patch? or a conduit? or an infundibular resection with pulmonary valvotomy? Does he have a right ventricular outflow aneurysm? Did he have surgical atrioventricular block? Does he have, or need, a pacemaker? Did he have major collaterals? Were they ligated? embolized? Does he have a residual left-to-right shunt? a residual collateral? a residual surgical anastomosis? a residual ventricular septal defect (VSD)? Is there pulmonary valve regurgitation? Is the right ventricle dilated? How severely? Is he in danger of ventricular arrhythmias? Does he need a pulmonary valve replacement? Should this be done surgically or percutaneously? Has he had regular echocardiographic studies? Holter recordings? exercise tolerance tests? Should he undergo cardiovascular magnetic resonance imaging (MRI)? or computed tomography? To look at what?

The purpose of these comments is not to address each and all of these questions. But both the pediatric cardiologist in charge of operated congenital patients and the adult cardiologist with expertise in congenital heart disease need to be familiar with all these issues.

In the paper by Cruz et al. the authors address again the possibility of dilatation of the AAO becoming problematic with time. AAO dilatation in TOF is a well-known occurrence. As Cruz et al. point out, it has been postulated that long-standing volume overload of the aorta before TOF repair may be responsible for aortic dilatation. They also mention that a wide range of prevalence of aortic dilatation in TOF has been published. The morphologic features of TOF, which are present since intrauterine life, stimulate aortic development beyond normal, as all right-to-left blood shunting at the ventricular level increases aortic blood flow and promotes aortic growth.

According to the findings of Cruz et al., this appears to be more likely to happen in men who underwent repair later in life, and may eventually lead to aortic valve regurgitation. Left ventricular mass index is the only factor independently associated with AAO dilatation in this study. Aortic valve regurgitation is more common in patients with pulmonary valve atresia with VSD, a patient group the authors decided not to include. This decision could be viewed as arguable, since pulmonary valve atresia with VSD and TOF may be no more than different degrees of the same basic disease. The condition is in fact also often called 'TOF with pulmonary atresia'. In addition, it is not certain that groups 1 and 2 (without and with AAO dilatation, respectively) in Cruz et al.'s study represent anything more than variations

in severity, or time-related differences between patients belonging to the same group. Nevertheless, the study has the merit of stressing the importance of progressive AAO dilatation and the risk of Ao valve incompetence. It is reassuring to note that aortic dissection is rare. It is difficult to ascertain whether the possible intrinsic histological abnormalities of the aortic wall, to which the authors refer,<sup>5</sup> are a cause or a consequence of the disease.

As the authors point out, there could be some institutional bias in their study resulting from its single-center nature. Still, this large prospective cardiovascular MRI study is relevant and suggests that the new imaging techniques are becoming widespread and an ever more useful tool.

Cruz et al. consider that AAO dilatation following TOF repair is best assessed by cardiovascular MRI, due to its accuracy and lack of ionizing radiation, and they suggest that this should be done at three-year intervals. This is perhaps one of the most interesting take-home messages of this paper.

In dealing with the most common form of cyanotic congenital heart disease it is essential to remember that most patients with TOF repair do well. But, as in many other fields of medicine, physicians need to be aware of the negative effects that may develop as a result from sequelae, residues and complications, or simply as a late consequence of the nature of the original disease. Only by doing so will they be able to provide the personal and compassionate attention that each individual patient expects from his physician.

## Conflicts of interest

The author has no conflicts of interest to declare.

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