Tetralogy of Fallot
General Principles of Management

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KEYWORDS

• Tetralogy of Fallot • Adults with congenital heart disease • Pulmonary regurgitation
• Pulmonary valve replacement • Arrhythmia • Pregnancy

KEY POINTS

- Tetralogy of Fallot (TOF) is one of the most common diagnoses encountered when caring for adults with congenital heart disease.
- Key issues for follow-up and surveillance include residual right ventricular outflow tract (RVOT) disease, right ventricular dilation or dysfunction, heart failure, and arrhythmia.
- Most adults with repaired TOF require pulmonary valve replacement (PVR). Indications for PVR are evolving, and transcatheter pulmonary valves (TPVs) have emerged as an alternative to surgery for some patients.
- Arrhythmias are prevalent in patients with repaired TOF, and sudden death does occur. Risk stratification is complex and often requires expert consultation.
- Pregnancy is generally well tolerated in women with uncomplicated repaired TOF; however, individual risk-stratification is indicated.

INTRODUCTION

TOF is the most common cyanotic congenital heart lesion, affecting 3% to 10% of all babies born with congenital heart disease.[1–3] Historically, it was the first complex cardiac lesion to be palliated surgically.[4] During the ensuing six decades, advances in surgical technique and perioperative management have resulted in excellent survival rates into adulthood. Assessment of life expectancy after TOF repair remains limited by the few patients currently in their sixth and seventh decade of life, but in several large series, the 30- to 40-year survival rate has been reported at 85% to 90%.[5–8] Morbidities such as arrhythmia and heart failure are common, however, and many patients require reintervention in adulthood. Postoperative TOF therefore requires lifelong care and is one of the most common diagnoses encountered by practitioners caring for adults with congenital heart disease.

ANATOMY AND INITIAL SURGICAL REPAIR

Developmentally, TOF occurs when the conal or infundibular portion of the ventricular septum is displaced anteriorly into the RVOT. This displacement produces (1) a large ventricular septal defect

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(VSD) and (2) obstruction to right ventricular outflow at the infundibular, valvar, or supravalvar levels (Fig. 1). The degree of RVOT obstruction is highly variable, ranging from very mild (the so-called pink tetralogy) to complete pulmonary valve atresia with diminutive or absent branch pulmonary arteries.

Surgical repair of TOF consists of VSD closure and relief of RVOT obstruction to the greatest extent possible. This is usually accomplished as a primary repair in infancy. In the era when neonatal cardiopulmonary bypass was not readily available, however, a staged approach was used. Patients requiring augmentation of their pulmonary blood flow in infancy received a systemic to pulmonary shunt, followed by complete repair at an older age. Today's adult congenital heart disease (ACHD) practitioner encounters both of these histories in clinical practice.

ANATOMIC SEQUELAE OF REPAIRED TETRALOGY OF FALLOT

Hemodynamically significant residual VSDs are uncommon in the adult with repaired TOF. By contrast, the vast majority of patients have residual RVOT disease. Patients whose RVOT obstruction was initially mild may have been treated with a surgical pulmonary valvotomy and augmentation of the infundibulum, thereby sparing the pulmonary valve annulus but leaving the potential for recurrent stenosis. Conversely, those with significant pulmonary annular hypoplasia typically require a transannular patch at the time of initial repair. This technique disrupts the valve architecture, providing good relief of obstruction but resulting in significant pulmonary regurgitation (PR). Finally, in a minority of patients, a right ventricle (RV) to pulmonary artery conduit is required, either because of complete pulmonary valve atresia or aberrant coronary anatomy that precludes an incision in the infundibulum. These conduits are not durable and often develop hemodynamically significant stenosis and/or regurgitation after one to two decades (or less). Knowledge of the individual surgical history is critical to the care of the postoperative patient, and therefore, review of original operative notes is recommended whenever possible.

GENERAL PRINCIPLES OF OUTPATIENT SURVEILLANCE

The 2008 American College of Cardiology (ACC)/American Heart Association (AHA) Guidelines for Management of Adults with Congenital Heart Disease propose a general framework for outpatient surveillance of the adult with repaired TOF.[7] Office visits with an ACHD physician are recommended at least annually, with a focus on identifying and managing the commonly encountered complications outlined in Box 1. Guidelines for frequency of imaging and testing are based on expert consensus, but a typical framework includes, at minimum, annual physical examination and electrocardiography with echocardiography as indicated.[9]

The physical examination is a useful starting point in elucidating residual anatomic lesions or associated conditions. Findings that may be encountered in patients with TOF are presented in Table 1. In patients with significant RVOT disease, careful attention to the jugular venous waveform is imperative to evaluate for volume overload

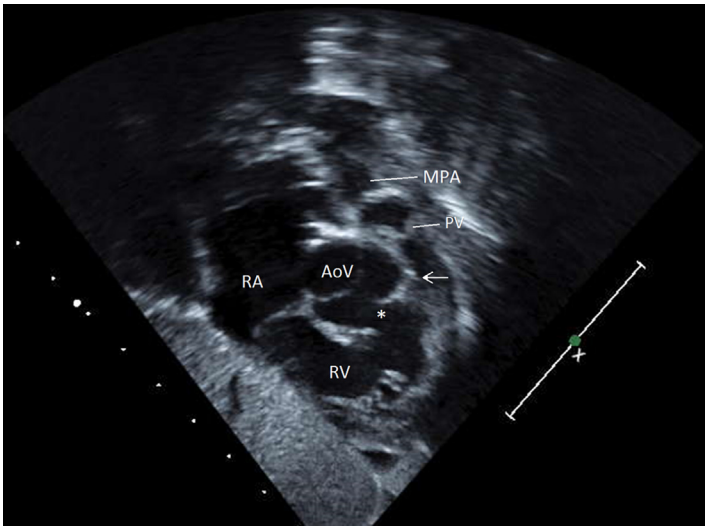

Fig. 1. Subcostal oblique view with anterior angulation demonstrating anteriorly malaligned conal septum (*arrow*) with resulting ventricular septal defect (*asterisk*) and hypoplastic pulmonary valve. AoV, aortic valve; MPA, main pulmonary artery; PV, pulmonary valve; RA, right atrium; RV, right ventricle.