MRI and Computed Tomography of Cardiac and Pulmonary Complications of Tetralogy of Fallot in Adults

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Abstract: Tetralogy of Fallot (TOF) represents the most common form of cyanotic congenital heart disease, accounting for 6.8% of all congenital heart disease. As surgical techniques and medical management of patients with TOF have improved, most affected patients are reaching adulthood. Though surgical outcomes are favorable (<2% early mortality rate), adults with TOF may experience complications from the long-term sequelae of congenital heart disease and complications related to treatment. We describe common and uncommon findings in adults with TOF, including pulmonary insufficiency, central and peripheral pulmonary artery stenosis and aneurysms, in addition to graft and shunt-related complications. Pulmonary function abnormalities and lung parenchymal imaging findings will be detailed. The diagnostic value of computed tomography and magnetic resonance imaging in adults with complications of TOF will be illustrated.

Key Words: congenital heart disease, tetralogy of Fallot, computed tomography, MRI

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Tetralogy of Fallot (TOF) represents 6.8% of all congenital heart disease and was originally described in 1888 by Fallot after cadaveric examinations of patients who died from cyanotic heart disease.1 It is thought that misalignment of the conal septum in relation to the ventricular endocardial cushion leads to a lack of stimulus for the development of the membranous septum. This results in a misaligned ventricular septal defect (VSD), an overriding aorta, and right ventricular outflow tract (RVOT) obstruction. In experimental models, ablation of neural crest cells produces conotruncal malformations such as TOF. Disease severity and age of presentation depends on the degree of RVOT obstruction.

TOF represents a spectrum of disease and is present in 3 major variants: TOF with pulmonary valve or RVOT stenosis, TOF with pulmonary atresia, and TOF with absent pulmonary valve. TOF with pulmonary atresia is the most severe form and accounts for 20% of the TOF population.2

Since the introduction of surgical palliation in the 1940s and repair in the 1970s, there has developed an increasingly large population of adults living with TOF. The growing population of adults with TOF merits increased attention to their unique problems. The imaging features mirror the evolution in surgical techniques. Radiologists should be familiar with the defects associated with TOF, the types of surgeries performed, and the potential complications. This paper will focus on the value of computed tomography (CT) and magnetic resonance imaging (MRI) in contributing to the care of adults with TOF.

IMAGING MODALITIES

Imaging plays a critical role in the management of adults with TOF and helps physicians decide if and when reoperation is necessary. MRI and CT are performed in this group to address specific clinical questions. MRI is noninvasive and does not require ionizing radiation. It is excellent at depicting cardiac anatomy, extracardiac vascular structures, and cardiac function, and permits flow quantitation by using velocity-encoded cine sequences. MRI is useful in measuring the pulmonary regurgitant fraction, provides information regarding the ratio of pulmonary and systemic blood flow through a VSD, the differential blood flow through both the left and right pulmonary arteries, and permits calculation of the size and function of the ventricles (Fig. 1).3,4 In addition, MRI can provide inferential information useful in estimating the pressure gradient across a stenotic lesion in a pulmonary artery. Disadvantages of MRI include the fact that it is time-consuming, requires patient cooperation, and is contraindicated in patients with pacemakers, and that contrast material cannot be administered in those with renal failure.5

CT has the advantage of short scanning times and wide availability. Ionizing radiation and the need for iodinated contrast material are disadvantages of CT. Electrocardiogram (ECG)-gated CT is the modality of choice for demonstrating major coronary artery anomalies6 and also provides valuable information regarding intracardiac anatomy and associated defects. MRI and CT are both useful in providing detailed information regarding pulmonary artery anatomy, the presence of collaterals,7 and pulmonary artery stenosis or aneurysms.8

SURGICAL TREATMENT OF TOF

Surgical palliation for TOF began in the 1940s with the performance of the first Blalock-Taussig shunt (Table 1).9 Without surgery, the average lifespan is 12 years, with only 3% of patients living past age 40. The
current trend at most centers is to perform a complete repair on infants between 3 to 6 months of age. Survival after repair today exceeds 95% and results in an improved quality of life. Surgery for TOF offers initial symptomatic relief and a favorable prognosis; however, long-term complications do arise.5

Primary surgical repair includes closure of the VSD and relief of the RVOT obstruction. A minority of patients have inadequate pulmonary artery anatomy or coronary artery anomalies that may require a staged procedure. Palliative surgery for TOF using a Blalock-Taussig shunt was frequently performed in the past (Fig. 2), but is only used today if the anatomy is not favorable for early complete repair.

Patients with pulmonary atresia represent a more complicated population. In these patients, the pulmonary arteries can range from normal in size to complete atresia of the central pulmonary arteries. In some cases of pulmonary atresia, the lungs are supplied by systemic collaterals from the aorta or its branches, called multiple aortopulmonary collateral arteries. These may represent the only blood supply to portions of the lung or may be redundant to existing pulmonary arteries.

TABLE 1. Types of Surgery for Tetralogy of Fallot

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infundibulectomy</td>
<td>Relieves the RVOT obstruction by excision of the infundibular muscle, especially the hypertrophied ventricular septal myocardium. Performed for less severe pulmonary stenosis.</td>
</tr>
<tr>
<td>Transannular patch</td>
<td>Relieves the RVOT obstruction by placement of a patch across the opened outflow tract. Performed when there is marked obstruction of the pulmonary valve annulus.</td>
</tr>
<tr>
<td>Right-ventricular-to-pulmonary-artery conduit</td>
<td>A valved conduit (porcine or synthetic) is inserted into the anterior wall of the right ventricle and connected to the pulmonary arteries.</td>
</tr>
<tr>
<td>Blalock-Taussig shunt</td>
<td>Creation of a subclavian to pulmonary artery shunt via a Gore-Tex tube or direct subclavian artery anastomosis. The BT shunt is a palliative procedure.</td>
</tr>
<tr>
<td>Unifocalization</td>
<td>This procedure involves redirecting the collaterals into a single vessel which is used to establish blood supply to the lung. Performed in patients with pulmonary atresia and extensive systemic to pulmonary collaterals.</td>
</tr>
</tbody>
</table>

BT indicates Blalock-Taussig; RVOT, right ventricular outflow tract.
In patients with extensive collaterals, a procedure called unifocalization may be performed.\textsuperscript{10} During unifocalization, the collateral vessels are redirected into a single vessel that establishes a new pulmonary arterial circulation either through a shunt, conduit, or direct anastomosis to the right ventricle (RV). Severely hypoplastic or atretic central pulmonary arteries present a persistent dilemma that may preclude definitive repair.

**PULMONARY PARENCHYMAL AND VASCULAR ABNORMALITIES RELATED TO TOF**

Lung function has been shown to be negatively affected in patients with TOF even after repair (Table 2).\textsuperscript{11,12} The finding of decreased pulmonary function in patients with TOF is related to diminished pulmonary parenchymal perfusion. In addition to RVOT obstruction, patients with TOF may have stenosis of the central or peripheral pulmonary arteries. Pulmonary artery stenosis has been shown to increase RV end-systolic pressure, leading to decreased exercise tolerance.\textsuperscript{13} In patients with TOF and pulmonary atresia, lung segments supplied exclusively by aortopulmonary collaterals without moderate peripheral pulmonic stenosis can develop pulmonary hypertension due to the exposure of systemic arterial pressure.

The CT appearance of the lung parenchyma depends on the pattern of collateral formation. Regions of the lung may be adequately perfused, underperfused, or overperfused. On CT, the lungs have a mosaic perfusion pattern related to the variability in regional lung perfusion and pulmonary hypertension (Fig. 3). Large systemic collaterals

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**TABLE 2. Complications of Tetralogy of Fallot in Adults Related to Longstanding Disease and Its Treatment**

<table>
<thead>
<tr>
<th>Effects on the lungs</th>
<th>Pulmonary regurgitation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reduced lung function due to diminished parenchymal perfusion</td>
<td>Complication of surgery</td>
</tr>
<tr>
<td>Regional pulmonary hypertension due to exposure to systemic pressure by MAPCAS</td>
<td>Increases risk of RV failure, ventricular arrhythmias, sudden death</td>
</tr>
<tr>
<td>Pulmonary artery aneurysm</td>
<td>Aneurysm of RV outflow tract patch</td>
</tr>
<tr>
<td>Due to absence of the pulmonary valve</td>
<td>May be due to areas of distal stenosis leading to pressure overload</td>
</tr>
<tr>
<td>Residual RV outflow tract stenosis</td>
<td>Tricuspid insufficiency</td>
</tr>
<tr>
<td>A common complication after infundibulectomy</td>
<td>Occurs in approximately 10% of patients with TOF</td>
</tr>
<tr>
<td>Residual VSD</td>
<td>Left ventricular dysfunction</td>
</tr>
<tr>
<td>Occur when the patch is placed on the trabeculated portion of the right ventricle</td>
<td>May be caused by persistent hypoxemia</td>
</tr>
</tbody>
</table>

MAPCAS indicates multiple aortopulmonary collateral arteries; RV, right ventricle; TOF, tetralogy of Fallot; VSD, ventricular septal defect.
are often directly visible in patients with pulmonary atresia, but their presence may also be indirectly inferred by CT findings including serrated pleural thickening and subpleural parenchymal bands.  

In addition to being a component of TOF, peripheral pulmonary artery stenosis can also be a postoperative complication related to insertion of a Blalock-Taussig shunt with stenosis at the anastomotic site. Patients with significant stenosis postoperatively may need reoperation or stenting. Pulmonary regurgitation and RV dilation after transannular patch repair can result in elongation and rotation of a previously normal left pulmonary artery resulting in kinking, a functional equivalent to pulmonary artery stenosis. On imaging, the kinked pulmonary artery has an acute angle at its origin. MRI and CT are both excellent modalities to assess the central and peripheral pulmonary arteries and provide detailed information essential for surgical planning (Fig. 4).

**PULMONARY ARTERY ANEURYSM**

Aneurysmally dilated central pulmonary arteries (Fig. 5) can develop in a subset of patients with TOF, usually those with absence of the pulmonary valve. The exact cause of pulmonary artery aneurysms in TOF is unknown. It has been postulated that absence of the patent ductus arteriosus plays a role. Unusual aneurysms of aortopulmonary collaterals occasionally occur and may be related to Blalock-Taussig shunt placement. CT and MRI clearly depict pulmonary artery aneurysms. MRI also provides detailed information about vessel-specific flow states.  

![Figure 3](image1.png)

**FIGURE 3.** A 61-year-old woman after remote repair of tetralogy of Fallot. A, Contrast-enhanced chest CT shows postrepair distortion of the pulmonary valve (arrow). B, Coronal reconstructed images from a contrast-enhanced CT shows a thrombosed, partially calcified, right-sided Blalock-Taussig shunt (arrows). The left pulmonary artery is aneurysmal (thick white arrows). C, Lung windows show mosaic attenuation with patchy and asymmetric pulmonary blood flow, more prominent on the left. The right lung is smaller than the left. CT indicates computed tomography.

![Figure 4](image2.png)

**FIGURE 4.** A 40-year-old woman with tetralogy of Fallot and pulmonary atresia previously treated in childhood by connecting a large systemic collateral to the left pulmonary artery. A, Maximum intensity projection coronal image from a contrast-enhanced MRA demonstrates small confluent central pulmonary arteries (white arrowhead) discontinuous from the right ventricular outflow tract. There is a right aortic arch with mirror-image branching (black arrows). The left pulmonary artery is larger than the right and was surgically connected to a left-sided systemic collateral which has a high-grade stenosis (thick white arrow). B, Maximum-intensity projection, short-axis image from a contrast-enhanced MRA demonstrates a subaortic ventricular septal defect with a large overriding aorta (thick white arrow) receiving the entire cardiac output. The collateral to left pulmonary artery anastomosis is tortuous and has a long-segment, high-grade stenosis (thin white arrows). C, Three-dimensional image from an MRA shows multiple aortopulmonary collateral arteries (MAPCAS) of varying sizes arising from the descending thoracic aorta (arrows). MRI indicates magnetic resonance angiography.
RVOT COMPLICATIONS

Infundibulectomy, transannular patch, and RV-to-pulmonary-artery conduit are surgical procedures to correct the RVOT obstruction in TOF (Table 1) (Fig. 6). Pulmonary regurgitation is present in nearly all patients surgically treated for TOF and is most common in patients treated with a transannular patch. Chronic pulmonary regurgitation has been shown to cause RV dilation and failure, ventricular arrhythmias, and an increased risk of sudden death.5

Debate still exists as to when the pulmonary valve should be replaced after TOF repair. Previous studies evaluating individuals with TOF who underwent pulmonary valve replacement showed decreased RV end-diastolic and end-systolic volumes, with conflicting results regarding improvement of RV systolic function. Pulmonary and tricuspid insufficiency also improved, as did arrhythmias.15–19

Clinical indications that favor replacement of the pulmonary valve include the onset of right heart failure symptoms in a previously stable patient and the onset of tricuspid insufficiency due to RV dilatation and arrhythmia. Studies have shown that it is beneficial to replace the pulmonary valve before RV function deteriorates and patients become symptomatic. In adults with TOF who underwent pulmonary valve replacement, there was a substantial reduction of RV volume if the pulmonary valve was replaced before RV end-diastolic volume was >170 mL/m² or RV end-systolic volume was >85 mL/m² (Table 3). Elevation

FIGURE 5. A 52-year-old man after repair of tetralogy of Fallot. A, Contrast-enhanced chest CT demonstrates an abnormally thickened pulmonary valve (black arrow) and a large left pulmonary artery aneurysm (white arrowheads). B, Oblique maximum-intensity projection image from an ECG-gated cardiac CTA shows the left anterior descending coronary artery arising from the right sinus of Valsalva (arrows), sharing a short common origin with the right coronary artery, and coursing anterior to the right ventricular outflow tract. C, Three-dimensional reconstruction from an ECG-gated cardiac CTA shows the left anterior descending coronary artery (arrows) arising from the right sinus of Valsalva and coursing anterior to the right ventricular outflow tract. Ao indicates aorta; CT, computed tomography; CTA, computed tomography angiography; ECG, electrocardiogram; PA, pulmonary artery.

FIGURE 6. A 22-year-old woman after repair of tetralogy of Fallot who developed a recurrent ventricular septal defect (VSD), pulmonary and tricuspid insufficiency, and right heart failure. A, Contrast-enhanced CT shows deformity and calcification of the right ventricular outflow patch (arrows). There is a right aortic arch with right descending aorta (thick white arrow). There are coils to the left of the descending aorta from prior embolization of MAPCAS. B, Contrast-enhanced CT demonstrated dehiscence of the subaortic VSD patch (arrow) with resultant left to right shunt. The right atrium and right ventricle are dilated. CT indicates computed tomography; MAPCAS, multiple aortopulmonary collateral arteries.
of the RV to left ventricular end-diastolic volume ratio also improves after pulmonary valve replacement. MRI is the best tool to measure these volumes. Until tested in a clinical trial, these results may serve as a guideline to the performance and timing of reoperation.15 In those treated with RV-to-pulmonary-artery conduits, stenosis or regurgitation of the conduit may occur, usually due to degeneration of the valve of the conduit. Residual or postoperative RVOT stenosis is a common complication following infundibulectomy.20

Aneurysms of the RVOT patch may develop and may be due to areas of distal stenosis leading to pressure overload (Fig. 7). Other proposed causes include problems with the patch material, scarring and fibrosis, and turbulent flow. The RVOT will often be dyskinetic postoperatively, especially in patients with a transannular patch. MRI and CT are excellent at depicting the morphology of the RVOT. The advantage of MRI is its ability to quantitate pulmonary regurgitation or stenosis using velocity-encoded sequences. The aneurysms and areas of stenosis are well demonstrated in a plane parallel to the RVOT. Cine MRI will show reduced diameter of a stenotic RVOT. MRI and ECG-gated CT allow for observation of wall motion abnormalities and calculation of RV volume and ejection fraction.

TRICUSPID INSUFFICIENCY

Tricuspid insufficiency is a complication in approximately 10% of patients with TOF.21 Factors contributing to tricuspid insufficiency include dilatation of the RV and tricuspid annulus and tricuspid valve injury during VSD repair.22 Tricuspid insufficiency can be quantitated with standard 2-dimensional echocardiography, but velocity-encoded cine MRI can also be performed, which may show flow through the tricuspid valve during systole. On axial spin-echo imaging, an enlarged right atrium is consistent with tricuspid insufficiency.

RESIDUAL VSD

Residual VSDs may exist after surgery but are a rare complication. They are thought to occur when the patch is placed on the trabeculated portion of the RV, leading to an area of communication through the intertrabeculated spaces between the left and right ventricles.23 Usually echocardiography is adequate to assess for the presence of a residual VSD. CT and MRI also reliably depict even small VSDs.2 Cine MRI provides information regarding the ratio of pulmonary and systemic blood flow through a VSD and will demonstrate the direction of flow as a jet traversing the region of the patch.

VENTRICULAR DYSFUNCTION, FIBROSIS, AND MYOCARDIAL INFARCTION

Right ventricular and left ventricular dysfunction can complicate TOF in adults via a number of mechanisms (Fig. 8). Ventricular fibrosis is a complication of long-standing TOF that contributes to right ventricular and left ventricular dysfunction and worsening symptoms of dyspnea and exercise intolerance.24

Left ventricular dysfunction can develop as a complication of hypoxemia, with the severity of dysfunction correlated with the severity of hypoxemia. Patients who have undergone surgical repair of TOF after 2 years of age have been shown to develop left ventricular dysfunction in the presence of an increased afterload, a predictor for sudden death.25

Though conventional 2-dimensional echocardiography can provide information about ventricular dysfunction, MRI and ECG-gated CT allow for observation of wall motion abnormalities, calculation of ventricular volumes, and ejection fraction. Coronary artery anomalies are common in TOF, occurring in up to 36% of patients. The most common anomaly is an anomalous left anterior descending coronary artery arising from the right coronary artery or right sinus of valsalva and coursing anterior to the RVOT.26 The anomalous coronary artery must be identified preoperatively or it may be inadvertently damaged during repair. ECG-gated CT is the modality of choice for demonstrating major coronary artery anomalies and is useful for preoperative planning.27 with a sensitivity approaching 80% to 90%.

### TABLE 3. Indications for Pulmonary Valve Replacement

<table>
<thead>
<tr>
<th>Clinical</th>
<th>Imaging</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right ventricular failure</td>
<td>Before right ventricular end-diastolic volume</td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>&gt;170 mL/m²</td>
</tr>
<tr>
<td>Tricuspid insufficiency</td>
<td>or</td>
</tr>
<tr>
<td></td>
<td>End-systolic volume &gt; 85 mL/m²</td>
</tr>
<tr>
<td></td>
<td>Elevated right to left ventricular end diastolic ratio (exact cutoff uncertain)</td>
</tr>
</tbody>
</table>

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**FIGURE 7.** A 22-year-old woman with tetralogy of Fallot after repair. Short-axis cine image shows postrepair deformity and dilatation of the RV outflow tract. A jet of pulmonary insufficiency is present (arrows). RV indicates right ventricle.
FIGURE 8. A 21-year-old man with tetralogy of Fallot with pulmonary atresia after a Blalock-Taussig shunt in infancy. A, Black-blood axial MRI shows marked cardiomegaly with biventricular dilatation and hypertrophy. There is a large subaortic VSD (thick white arrow) and a right aortic arch with a right descending aorta (arrow). B, Oblique sagittal reconstruction from a contrast-enhanced ECG-gated CT shows a large overriding aorta (thick white arrows) receiving the entire cardiac output. There is a large subaortic VSD. Marked cardiomegaly with biventricular dilatation and hypertrophy is present. Multiple aortopulmonary collateral arteries (MAPCAS) arise from the descending aorta (black arrows). C, Oblique reconstruction from a contrast-enhanced ECG-gated CT depicts the hypoplastic main pulmonary artery (thin white arrow) separated from the hypoplastic right ventricular outflow tract (black arrow) by the atretic pulmonary valve. There is a patent left Blalock-Taussig shunt (thick white arrow). CT indicates computed tomography; ECG, electrocardiogram; MRI, magnetic resonance imaging; VSD, ventricular septal defect.

CONCLUSIONS
In conclusion, adults with TOF, the most common cyanotic congenital heart disease, suffer from cardiac and pulmonary complications of longstanding disease and complications related to treatment. MRI and CT provide anatomic and physiologic information that is integral to guiding management in this complex population.

REFERENCES


