

## The Role of Imaging in Adult Tetralogy of Fallot Patients

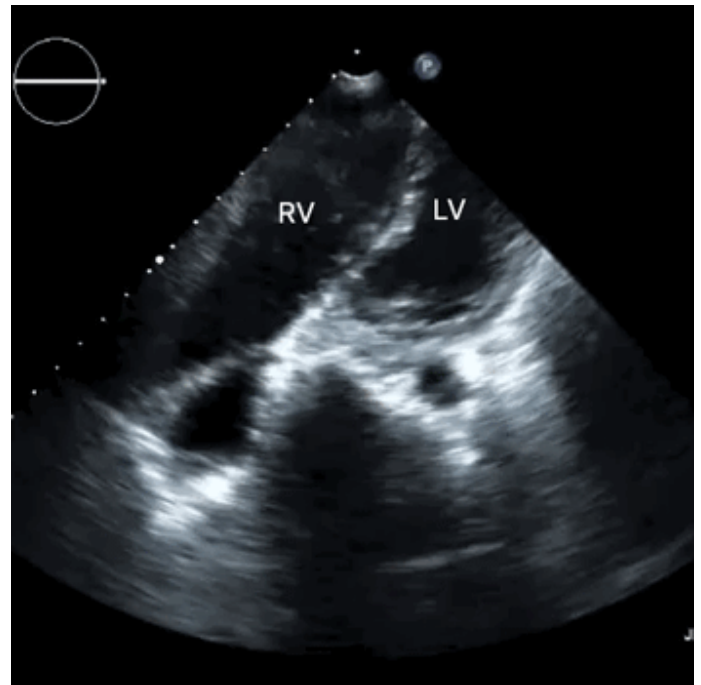
- As many as 1,250,000 adults in the US have congenital heart disease (CHD); of these, Tetralogy of Fallot (TOF) is one of the most common lesions.
- Most cases of TOF are diagnosed and initially treated in childhood; patients should be followed longitudinally, as new sequelae or residual lesions often present in adulthood, most commonly pulmonary regurgitation (PR).
- Several imaging modalities—cardiac ultrasound (echocardiography), cardiac MRI, and cardiac CT—are used in both surveillance and pre-surgical planning for adult TOF.

The past several decades have seen a dramatic rise in the number of adults in the US with congenital heart disease (CHD). According to some estimates, the US patient population is increasing by 5 percent every year, with an estimated 1,250,000 patients as of 2012. The increasing incidence is due at least in part to medical and surgical advances in the 1960s and 1970s that led to improved survival for children with CHD. Occurring in roughly five of every 10,000 newborns, Tetralogy of Fallot is one of the most common congenital defects, affecting up to 0.5 per 1,000 live births. These numbers underscore the importance of subspecialty monitoring of adults, particularly those who have undergone repair for CHD.

### Pulmonary Regurgitation in Tetralogy of Fallot Patients

Adult TOF can include a range of presentations and consists of cases diagnosed and treated in childhood with sequelae or residual lesions in adulthood, cases diagnosed in childhood but not treated, and new cases diagnosed in adulthood. In each case, diagnosis and treatment can be complicated by age-related factors such as symptoms of acquired heart disease, adding to the need for advanced imaging in managing patients.

TOF is typically treated with surgical repair when the diagnosis is made in infancy or childhood. In the modern era, rarely do patients present unrepaired in adulthood. While survival after surgical repair is generally excellent, patients may suffer late complications. Studies show a three-fold increase in mortality in the third decade post intervention, and 14% of patients manifest significant impairment over several years. To improve long-term outcomes, TOF patients must be monitored through adolescence and adulthood.



**Figure 1. Cardiac ultrasound (echocardiography)**  
A horizontal long-axis view of the heart demonstrates enlargement of the right ventricle (RV). The left ventricle is partially imaged in this view. While this patient has relatively clear views of the right ventricle, often portions of the right ventricle are obscured, and ultrasound cannot completely visualize the entire ventricle in order to perform volume measurements; in this case, the right ventricular dilatation and symptoms prompted further investigation with cardiac MRI.

One of the most prevalent complications in adult TOF referrals is pulmonary regurgitation (PR), due to a leaky or absent pulmonary valve (which may have been surgically removed to alleviate narrowing). Over time, PR can result in right ventricular dilation and dysfunction and create co-morbidities including deteriorating exercise tolerance, arrhythmias and even sudden cardiac death.

Pulmonary valve replacement is often recommended in cases of adult TOF patients with PR. Options include surgical pulmonary valve replacement and, more recently, transcatheter-based percutaneous pulmonary valve replacement. Both approaches require cardiac ultrasound (echocardiography) for screening and follow up, cardiac MRI for targeted, detailed surveillance and may require cardiac CT for pre-procedure planning.

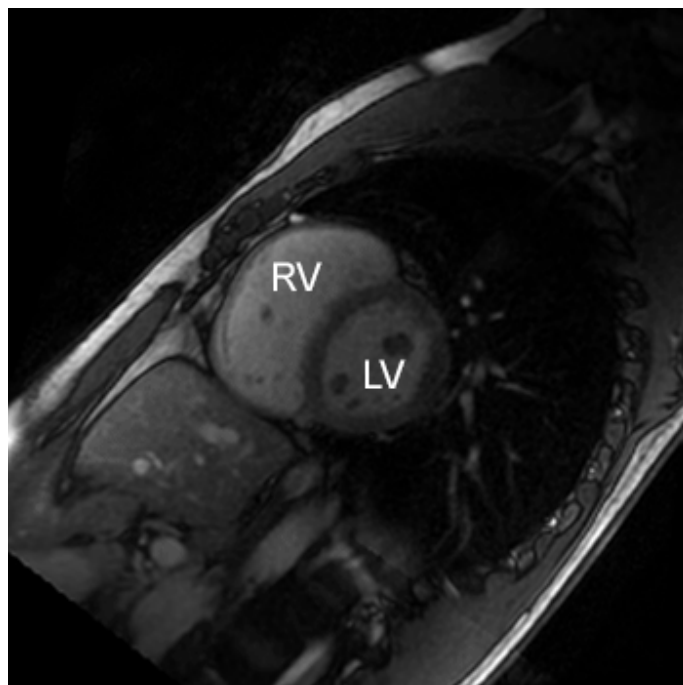
### Cardiac Ultrasound (Echocardiography)

Ultrasound is used as the initial screening test for PR in adult TOF patients and is effective as a first-line modality for qualitative assessment of the right ventricle. This method allows real-time visualization of the heart wall in motion, recorded as single images and video clips. However, obtaining comprehensive images of the right ventricle with ultrasound can be challenging due to its unique pyramid-shaped geometry and its location in the chest behind the sternum. Lung and bone block the sound waves, leaving a difficult acoustic window for assessment in many patients. Comparisons of the accuracy of right-heart parameters measured with both ultrasound and MRI in adult-repaired TOF patients have generally shown the benefits of MRI over ultrasound. Studies have shown that systolic function measures by ultrasound do not provide a complete and accurate measure of right ventricular ejection fraction as compared to cardiac MRI. Additionally, it may be challenging to accurately quantify PR echocardiographically; PR may be more precisely quantified by cardiac MRI.

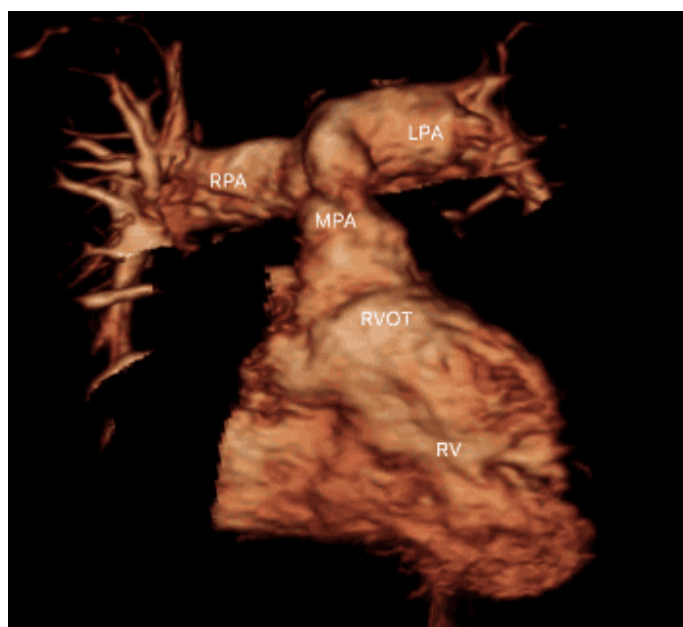
### Cardiac MRI

Cardiac MRI is used as a second-line, complementary modality for the evaluation of PR in adult TOF patients who have undergone repair. Introduced in the late 1990s, cardiac MRI is now considered the gold standard for assessment of right ventricle size and function. However, because it uses a powerful superconducting magnetic field, it can pose challenges and contraindications for patients with metallic implants, including some pacemakers and certain other implants.

In recent years, the development of two-dimensional (2D) and four-dimensional (4D) flow-velocity MRI has added measures of blood flow volume and velocity to the unrestricted spatial planes that MRI allows. A key metric is the direct assessment of ventricular volumes of the



**Figure 2. Cardiac MRI** A stack of short axis views of the right ventricle (RV) and left ventricle (LV) allow complete visualization of all portions of both ventricles, and thus allow quantitative measures including end-diastolic and end-systolic volume measurements.



**Figure 3. Cardiac MRI Angiography (MRA)** An angiographic reconstruction of a contrast-enhanced MRA depicts the pyramidal shape of the right ventricle (RV) as well as the muscular infundibulum known as the right ventricular outflow tract (RVOT), which in this case supply blood to the main pulmonary artery (MPA) and its branches, the right and left pulmonary arteries (RPA, LPA). This patient's pulmonary valve had been excised years earlier during an augmentation procedure, resulting in a stenotic segment in the main pulmonary artery.

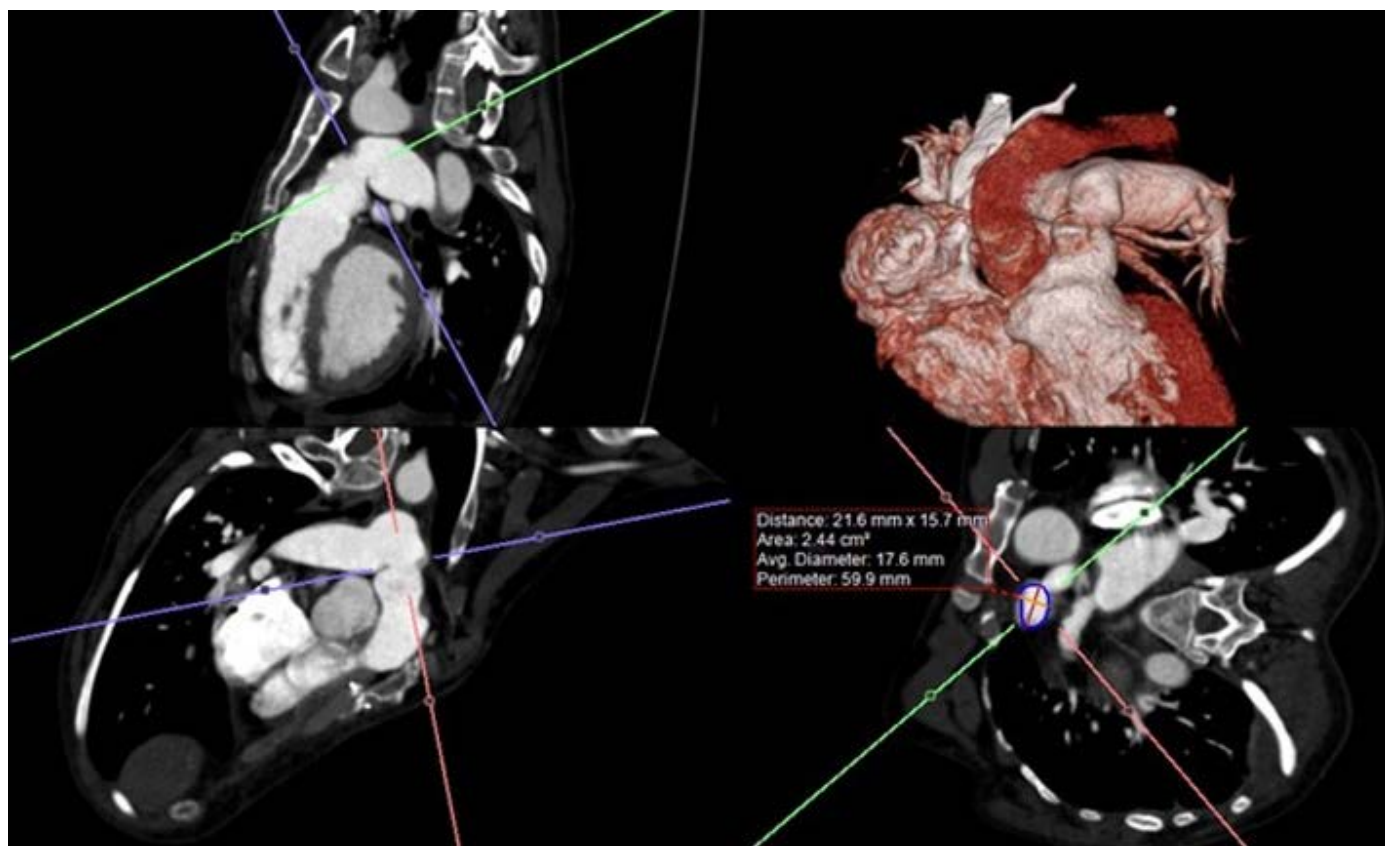
ventricular chamber (whereas ultrasound only measures blood flow velocities). The volumes of blood in the ventricle (*i.e.*, end-diastolic right ventricular volume), as well as the proportion of blood pumped per beat (*i.e.*, ejection fraction) best predict the appropriate timing for valve repair or replacement. This assessment is accomplished via careful measurement of multiphase datasets and processing of velocity-encoded images. Researchers are currently exploring the possibility of using information derived from 4D flow datasets in patients who have undergone repair for TOF to facilitate efficacious determinations of right ventricle dilation and function.

## Cardiac CT

Cardiac computed tomography (CT) and CT angiography (CTA) can be used to screen for PR in adult TOF patients when cardiac MRI is contraindicated or when additional, complementary information (such as coronary artery anatomy) is needed. CT is also used for pre-procedural planning prior to percutaneous catheter-based valve deployment. Minimally invasive transcatheter-based pulmonary valve replacement requires detailed valve area anatomy to properly size the device prosthesis.

CTA provides superior spatial resolution compared to cardiac ultrasound and cardiac MRI. It allows three-dimensional (3D) visualization, which unlike MRI or ultrasound, is not performed and acquired in real time, but rather depends on workstation post-processing of image datasets. For example, CT can precisely measure the pulmonary annulus (a 3-D structure) on which both the type and size of an implanted valve depends. More accurate sizing of pulmonary outflow anatomy is associated with decreased post-operative complications.

Because they require ionizing radiation, CT and CTA are reserved for adult TOF patients with abnormal first- or second-line screening at high risk of PR and those with a favorable anatomic profile for intervention.



**Figure 4. Cardiovascular CT Angiography (CTA)** Multiplanar-reformatted images and a volume-rendered image (top right) were performed to evaluate the stenotic segment of the main pulmonary artery. The bottom right image demonstrates the minimal luminal area, which is used for sizing of potential minimally-invasive valve placement or surgical planning, based on individual patients' anatomy. The corresponding long-axis views (left-sided panels) depict the stenotic segment for precise localization. CTA has superior spatial resolution which allows the most precise measurements for planning.

## Scheduling

Echocardiography, cardiac MRI and cardiac CT are performed on the main campus of Massachusetts General Hospital in Boston. Endocardiography is also performed at Mass General Imaging - Waltham and Mass General/ North Shore Center for Outpatient Care. Cardiac CT is also performed at Mass General Imaging - Waltham. Appointments can be made through Epic (inside the Partners network) or [Physician Gateway](#) (outside the Partners network) by calling 617-726-8396. The office number for the MGH Adult Congenital Heart Disease Program is 617-643-7024 or 617-643-7024.

## Further Information

For further information on the role of CT and MRI imaging in the care of adult patients with TOF, please contact [Brian B. Ghoshhajra, MD, MBA](#), Division of Cardiovascular Imaging, Department of Radiology, Massachusetts General Hospital, at 617-643-0239.

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*Janet Cochrane Miller, D. Phil., Co-Author*

*Gary Boas, Co-Author*

*Raul N. Uppot, M.D., Editor*

