Review

Essentials of Imaging the Repaired Tetralogy of Fallot Patient

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ABSTRACT

Tetralogy of Fallot is one of the commoner forms of congenital heart disease (CHD) with an incidence estimated at around 1 in 3000 live births. Its prevalence is roughly 5%-10% of all forms of CHD. Although it is a cyanotic disorder at presentation, early surgical repair has led to survival to adulthood in the overwhelming majority of cases. However—as with most repaired CHD—such patients are fixed, not cured, and will inevitably come to the attention of the imaging specialist. This article attempts to describe and summarize the main forms of repaired tetralogy that may be encountered in tertiary hospital imaging practice. The anatomy of different forms of tetralogy is reviewed, and relevant pathophysiology is discussed. Reference is made to 22q11 syndrome, which is often missed or not considered by even specialist congenital physicians, and the alerting features of this condition are reviewed. The benefits and challenges of computed tomography and magnetic resonance imaging in the congenital population are covered, and there is discussion regarding the various approaches to imaging available using these techniques.

RÉSUMÉ

La tétralogie de Fallot est l'une des formes plus courantes de cardiopathie congénitale, son incidence étant d'environ 1 naissance vivante sur 3000. Sa prévalence est d'approximativement 5 à 10 % de toutes les formes de cardiopathie congénitale. Bien qu'il s'agisse d'une maladie à présentation cyanogène, sa correction chirurgicale dans les premiers stades permet le prolongement de la survie jusqu'à l'âge adulte dans la très grande majorité des cas. Les patients qui bénéficient de ce traitement, comme c'est le cas de la plupart de ceux atteints d'une cardiopathie congénitale réparée chirurgicalement, sont cependant traités, et non guéris. Aussi, la maladie apparaîtra inévitablement à l'imagerie. Le présent article tente de décrire et de résumer les principales formes de tétralogie prises en charge par chirurgie qui pourraient ressortir dans le cadre d'examens d'imagerie en contexte de soins tertiaires. Il en examine les différentes particularités anatomiques et discute des aspects physiopathologiques pertinents. Il fait également référence au syndrome de délétion 22q11 qui est souvent négligé, même par les spécialistes en maladies congénitales, et explore les caractéristiques alarmantes de ce syndrome. Il passe en revue les bienfaits que la tomodensitométrie et l'imagerie par résonance magnétique peuvent offrir à la population de patients présentant une maladie congénitale ainsi que les difficultés qui peuvent survenir lors de ces examens. Enfin, il se penche sur les diverses méthodes d'imagerie qu'il est possible d'utiliser avec ces techniques.

Initial Anatomy Dictates the Later Problems

The intracardiac anatomy of the patient with tetralogy at birth determines not only the nature of the surgical repair but also, in consequence, many of the problems that the patient will later face.[1] There are 4 primary lesions that comprise the anatomic tetrad. The outlet portion of the ventricular septum is displaced both anteriorly and superiorly, and—together with hypertrophy of the septoparietal trabeculations and the conal muscle—this leads to infundibular obstruction and the presence of a nonrestrictive ventricular septal defect (VSD)

(Fig. 2).[2] The pulmonary annulus may be small or frankly hypoplastic. The pulmonary valve itself is often abnormally thickened and may be bicuspid. Many of the causes of right heart obstruction can occur in tandem (Table 1). Other common anatomic findings in tetralogy are summarized in Table 2.

Surgical correction of the defects of tetralogy of Fallot (ToF) is always necessary.[3] Historically, patients with tetralogy could not be repaired while infants due to their small size. It was standard practice to treat cyanosis by the placement of a systemic to pulmonary artery shunt, usually the Blalock-Thomas-Taussig shunt, which had the secondary advantage of promoting central pulmonary artery growth due to increased blood flow[4] (Fig. 5). Once the child had reached an adequate size, usually somewhere between the ages of 3 and 5 years, this shunt would be surgically tied off (or occluded) and
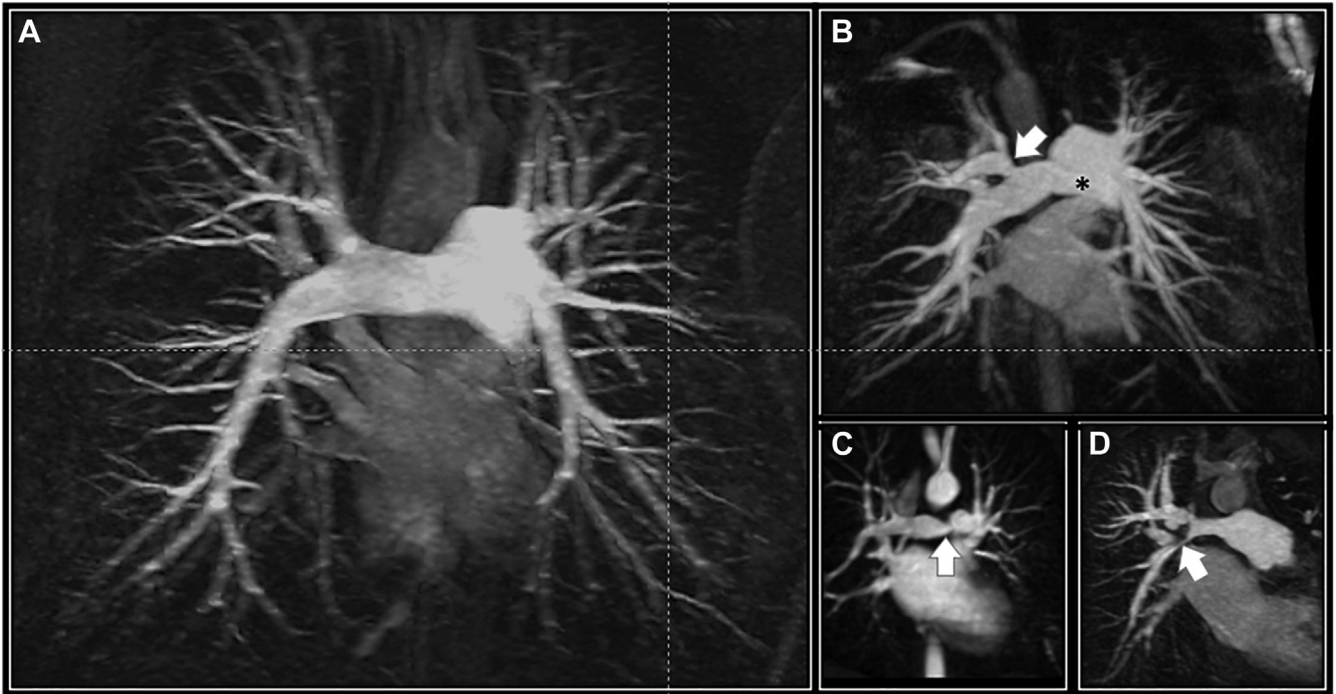
Figure 1. Normal and abnormal pulmonary arterial tree on magnetic resonance angiography (MRA). (**A**) Normal MRA demonstrating an absence of central or peripheral pulmonary arterial stenoses. Compare these normal appearances with (**B**) ostial right pulmonary artery (RPA) stenosis (**asterisk**) and right upper lobe ostial branch stenosis (**arrow**). (**C**) Tight ostial RPA stenosis (**arrow**). (**D**) Right lower lobe tubular stenosis (**arrow**).

a complete intracardiac repair performed. Nowadays, repair usually occurs within the first 6 months of life without the intermediate stage of a systemic to pulmonary shunt. Nevertheless, the imager will still occasionally come across remnants of these shunts in older adult patients with tetralogy.

Right-sided obstruction is dealt with by cutting away excess infundibular muscle and opening up the outflow tract with an artificial patch. In former days, it was common to extend the patch through the pulmonary annulus in an attempt to open this up more completely also. However, this has the effect of corrupting normal pulmonary valve coaptation and inevitably results in some degree of pulmonary regurgitation (PR). This is the principal lesion leading to right ventricular (RV) enlargement and dysfunction in adult life
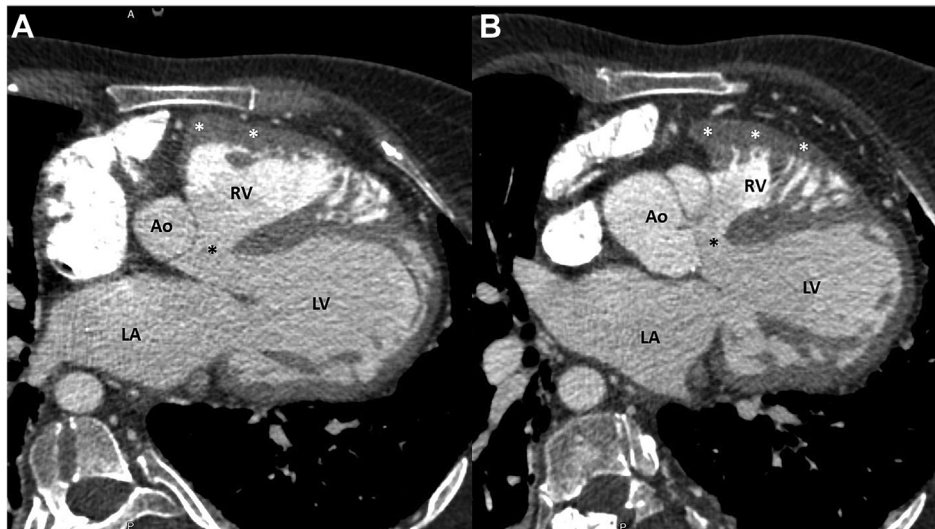

Figure 2. Unrepaired tetralogy of Fallot. Sequential axial slices from a gated cardiac CT study demonstrating 3 of the 4 features of tetralogy, including large subaortic VSD (**asterisk**), overriding aorta (Ao), and right ventricular (RV) hypertrophy (**white asterisks**). CT, computed tomography; LA, left atrium; LV, left ventricle; VSD, ventricular septal defect.
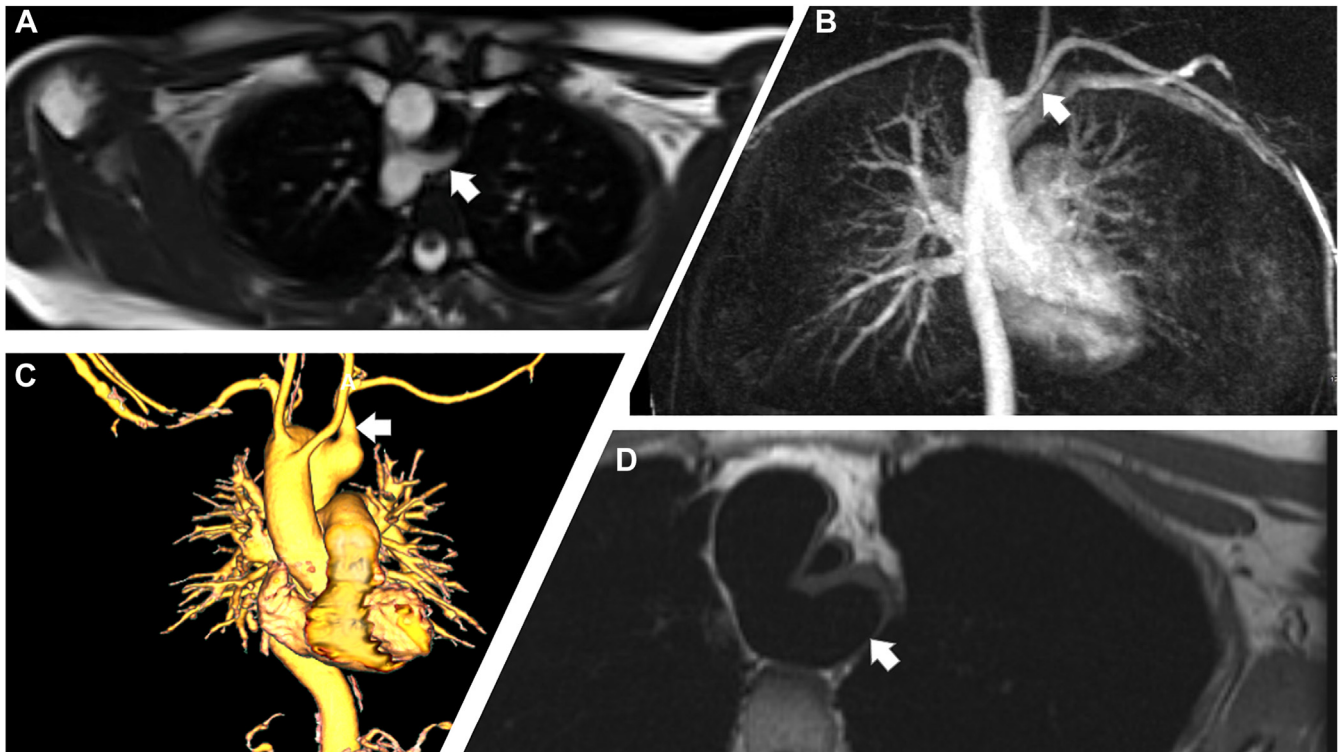
Figure 3. Examples from different patients with right aortic arch with the aberrant left subclavian artery on (**A**) axial cine, (**B**) maximum intensity projection MR angiogram, (**C**) volume-rendered MR angiogram, and (**D**) axial black blood. Note that, when a right aortic arch is present in a patient with tetralogy, there is an increased chance of 22q11.2 deletion syndrome (see Table 3), and this should be highlighted to the referring clinician so that genetic testing can be discussed with the patient. MR, magnetic resonance.

and is commonly seen in the older adult population with tetralogy. Younger patients may have been spared this or, if transannular patching was unavoidable, may have been deliberately left with a degree of obstruction through under-resection of RV muscle bundles. The aim here is to promote mild right ventricular hypertrophy in the ventricle, which is then perhaps more resistant to RV dilatation from the inevitable PR. A small proportion of infants will not be suitable for right ventricular outflow tract (RVOT) patching due to the presence of an anomalous or dominant coronary branch that crosses the RVOT. In this setting a tube graft (conduit) is sewn between the anterior aspect of the RV and the bifurcation of the pulmonary arteries (PAs)—the so-called RV-to-PA conduit.
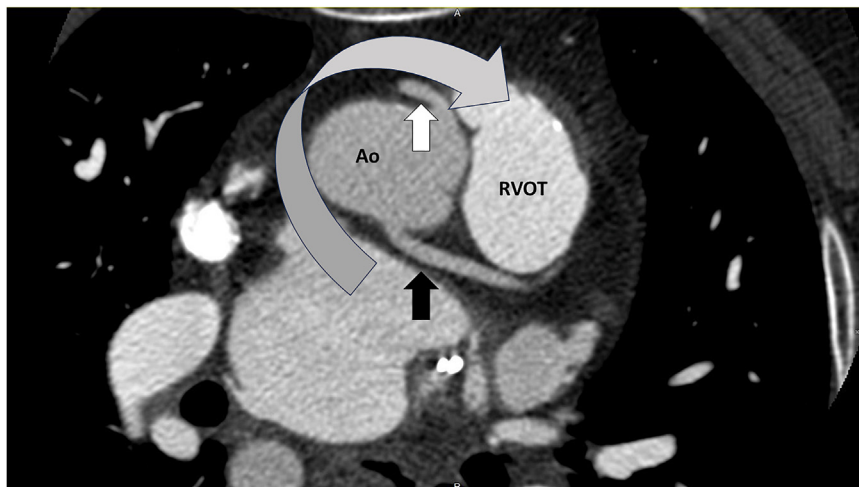

Figure 4. Clockwise rotation of aortic root in a patient with tetralogy. Both the right coronary (**white arrow**) and left main coronary artery (**black arrow**) arise anatomically from the correct part of the aortic root, but the entire root has effectively turned a little clockwise (**large grey arrow**) during development. Ao, aorta; RVOT, right ventricular outflow tract.

Table 1. Causes and frequency of right heart obstruction in tetralogy of Fallot

Levels of right heart obstruction in tetralogy				
Infundibular	Infundibular and valvar	Valvar and annular	Diffuse hypoplasia of RVOT	Only valvar or annular level
27%	26%	16%	27%	2%-5%

RVOT, right ventricular outflow tract.

What Are Our Tools?

Let us now consider the tools at our disposal for imaging the population with tetralogy. In most cases, a single imaging modality will not suffice, and judicious use of 2 or even 3 approaches may be necessary. Table 4 lists the relative strengths of the available techniques. In general terms, echo and cardiac magnetic resonance (CMR) are the workhorses of congenital imaging. However, there may be specific situations in which cardiac computed tomography (CT) is preferred including claustrophobia, the presence of devices, the presence of stainless steel embolization coils, or the need for unequivocal coronary imaging.

Another important aspect to consider is the ability of the patient to cooperate during the test. While echo is rarely problematic, adults with syndromic disability including Downs and 22q11 deletion may find it difficult to remain motionless and cooperate with breathing instructions in the CMR scanner. In this situation, the choice is between a CMR performed under general anaesthetic or a quick cardiac CT scan (usually well tolerated). Very often we choose the latter, especially when most of the necessary flow information has already been acquired by echo.

Haemodynamics is the major Achilles heel of most noninvasive imaging. Although various echo parameters may provide approximations of intracardiac pressures, cardiac catheterization is usually required for confirmation of stenosis severity within conduits, as well as right and left heart pressures where there is evidence of clinical heart failure. Again, a proportion of patients may require this to be done under anaesthesia.

What Are the Common Residual "Late" Problems in Adults With ToF?

Adult patients with tetralogy will inevitably encounter late complications at some point in their lifetime.[5] As mentioned above, the single biggest problem in repaired tetralogy is the near-universal presence of PR.[6,7] For a long time this was regarded as unimportant and well tolerated. Also, our methods of assessing both PR and its effects on the RV were relatively poor until the routine introduction of CMR imaging into clinical practice roughly 20 years ago. We now appreciate that slow dilatation of the RV is seen in almost all patients with free PR, and that over time this can levad to not only dysfunction of the RV and clinical right-sided heart failure but also a reduction in left heart function through the mechanism of ventricular interdependence.[8] We generally measure both the absolute volume of PR and PR as a percentage of the total forward flow across the RVOT (Fig. 6). Although sometimes regarded as interchangeable, it is the former that is more predictive of subsequent RV enlargement.[9]

Table 2. Beyond the tetrad; other common anatomic findings in tetralogy of Fallot (ToF)

Lesion	Comment
Narrow/short pulmonary trunk	Worse when outflow tract is severely hypoplastic
Central stenoses of branch pulmonary arteries (Fig. 1)	Relatively common. May cause differential flow to the lungs
Additional muscular VSDs	Seen in 3%-15% of patients
Right-sided aortic arch in 25% (Fig. 3)	Usually mirror image branching
Aortic root rotation (Fig. 4)	Often clockwise rotated
Aortic root dilatation	May be present from birth
	Often stable rather than progressive in adult life
Coronary artery anomalies	• LAD may arise from RCA and cross infundibulum (2%-14%)
	• Single right or left coronary artery
	• Large conus branch crosses infundibulum
22q11.2 deletion syndrome (qv)	Also known as Di George syndrome
	Many possible issues (see Table 3)
Trisomy 21 (in approximately 5% of patients with ToF)	With AV canal defects

AV, atrioventricular; LAD, left anterior descending coronary artery; RCA, right coronary artery; VSD, ventricular septal defect.
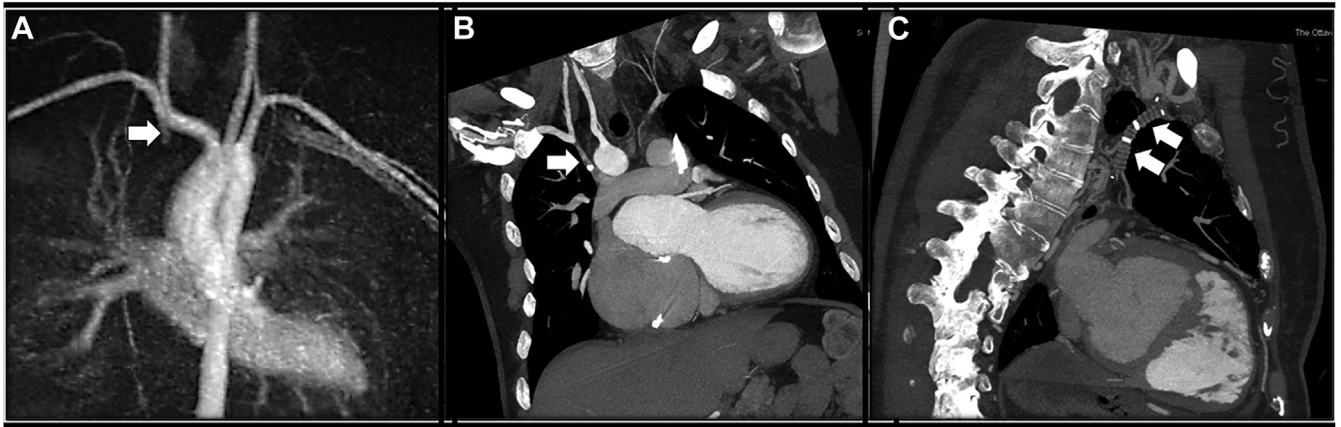
Figure 5. Variety of appearances of prior BTT shunts. (**A**) MR angiogram dynamic frame showing a vertical tiny vessel (**arrow**) descending from the left subclavian artery; this is all that remains of an otherwise occluded modified BTT shunt. (**B**) Coronal view from a cardiac CT showing this time the full length of an occluded BTT shunt running from the subclavian artery superiorly to the pulmonary artery inferiorly. (**C**) Nice example of an occluded BTT shunt on coronal oblique MIP from a CT angiogram. In this example, the ridges of the Gore-Tex material can be appreciated. BTT shunts used to be performed when a child was too young for total repair in infancy and then closed later when full repair was performed. In the current era, most patients will have a total repair in early life with no need for a shunt of this sort; however, the imaging physician will still encounter them in older adults—hence their demonstration here. BTT, Blalock-Thomas-Taussig; CT, computed tomography; MIP, maximum intensity projection.

As mentioned earlier, some patients may have residual RVOT obstruction that was surgically "engineered" to promote resistance to ventricular dilatation from PR.[10] However, others may have inadvertent obstruction due to conduit calcification or degeneration (Fig. 7).

Yet others may develop obstruction if endocarditis supervenes, with vegetations forming on the pulmonary valve. Endocarditis also seems to occur more frequently in patients whose conduits have been previously stented with a Melody valve.[11-13] Echo sometimes struggles to precisely locate the level of obstruction (valvar vs subvalvar), and here 3-dimensional techniques such as CMR and CT have a distinct advantage (Fig. 8). Dual energy CT may have particular advantages for overcoming metal streak artefact while managing to preserve tissue contrast.[14]

Another common issue is that of RVOT pseudoaneurysm. Less commonly seen now, these are usually present in older patients who underwent a repair that included a large transannular patch. Thus, the pseudoaneurysm is of patch tissue rather than the native RVOT wall itself. Large pseudoaneurysms are mechanically disadvantageous, and as they do not contract, they will also appear to reduce overall ejection fraction even when the contractile properties of the residual muscular ventricle are intact.[15-17] Areas of intersection between patch material and native tissue may also include areas of myocardial disruption that can be triggers for ventricular arrhythmia.[18,19] Although these cannot be imaged directly, the extent of the patch is usually apparent both from non-contractility and dyskinetic systolic motion, as well as potential enhancement postgadolinium due to an enveloping cover of fibrous tissue (Fig. 9).

Conduits pose particular challenges and inevitably require replacement at some point in the patient's unnatural history. In the younger age group, it is inevitable that a child will outgrow the conduit as (s)he ages. Conduits placed into adult patients may be appropriately sized at the time of insertion, but they almost invariably distort and calcify as they age and most will require replacement within 1-2 decades. They are often difficult for echo to image well because calcification is common and may lead to acoustic shadowing. Furthermore, gradients derived from echo are less accurate in conduits than across valves and may lead to overestimation compared with catheter-derived measurements. CMR is useful for helping to show the level of obstruction and for providing some measurement of conduit size at various levels.[20] However, cardiac CT provides optimum assessment given its high isotropic spatial resolution, ability to identify calcium, and fidelity for the coronary arteries.[21] This last is essential when conduit stenting is under consideration to ensure that the coronary arteries are sufficiently distant to avoid compression at the time of stent deployment.

Table 3. Clinical features of 22q11.2 deletion syndrome*

Cardiac abnormalities especially tetralogy of Fallot
Immunodeficiency and recurrent infections
Distinctive facial features
Submucosal cleft palate leading to nasal sounding speech, feeding difficulties
High arched palate and bifid uvula
Hypocalcaemia (may cause seizures)
Hypoparathyroidism
Thrombocytopenia
Gastrointestinal problems
Hearing loss
Mild short stature and/or spinal abnormalities (less common)
Developmental arrest with mild intellectual disability and difficulty with:
- Reading
- Mathematical-related tasks
- General problem solving
Attention deficit hyperactivity disorder
Autism spectrum disorder

* Not all patients have all features.

Table 4. Comparison of the relative strengths of commonly used imaging modalities

	Echo	Nuclear	CT	CMR	CATH
Spatial resolution	++	+	++++	++	++++
Temporal resolution	++++	+	++	+++	++++
Suitability in obese	++	+	++++	++++	+++
Multiplanar reformats possible	−	++	++++	+++	−
3D reconstructions possible	−	++	++++	+++	−
Limited by metal	++	+++	+	+++	−
Limited by calcium	+	+	+	++	−
Speed of assessment	++	++	++++	+	+++
Requires exogenous contrast injection	−	++++	+++	+	++++
Availability	++++	++++	++++	++	++++
Claustrophobia	−	+	++	+++	−
Radiation	−	+++	+/++	−	++/+++
Myocardial characterization	+	++	+++	++++	+/−
Lung assessment	−	+/−	++++	++	+/−
Thoracic vasculature assessment	++	−	++++	++++	+++
Valvular assessment	+++	−	+	++++	+

CATH, catheterization; CMR, cardiac magnetic resonance; CT, computed tomography.

The central PAs are commonly abnormal in patients with tetralogy. They may be small and underdeveloped or, more often, are a reasonably normal size but with stenoses that occur at their origins from the main pulmonary artery.[22] In many cases, these stenoses are relatively mild in degree and cause only relatively minor degrees of flow asymmetry between the right and left lung. It is important to measure this by phase contrast flow mapping of each branch PA. Relative flow distributions can alternatively be measured reliably with nuclear perfusion techniques (Fig. 10). This may be a useful technique when used to assess the success of unifocalization of major aortopulmonary collateral (APC) arteries in tetralogy with pulmonary atresia.[23]

Flow ratios of 45:55 or 60:40 are common and do not need intervention to equalize flow. Ratios of 70:30, or especially 80:20 or worse, may trigger a need to consider whether pulmonary enlargement can be achieved by intervention or surgery. Contrast-enhanced angiography, whether by CT or CMR, is useful to fully appreciate the asymmetry and complexity of stenoses—especially where intervention is considered. After intervention, CT is preferable to CMR due to unavoidable metal artefacts (Fig. 11).
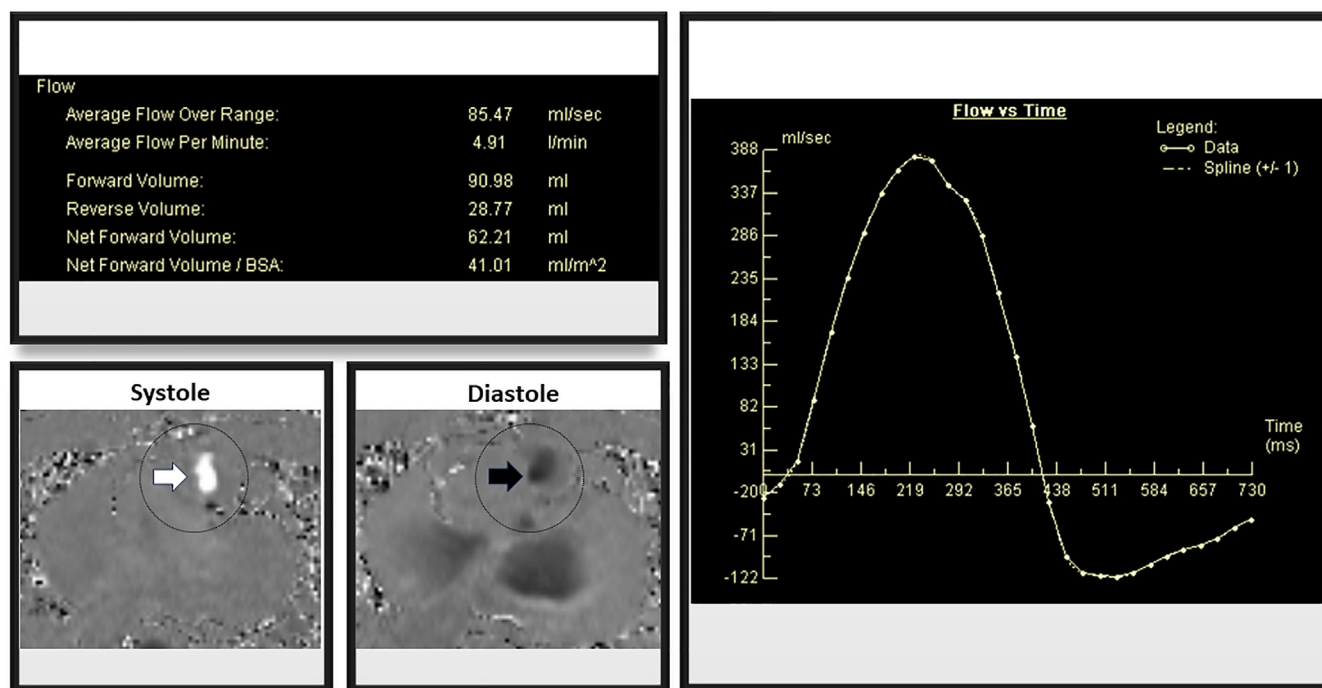

Figure 6. Demonstration of phase contrast imaging in a tetralogy patient with moderate pulmonary regurgitation. The right ventricular outflow tract is imaged just below the pulmonary valve in this example. Forward systolic flow is encoded white (**white arrow**), whereas diastolic backward flow appears black (**black arrow**). By integrating flow across the cardiac cycle, a flow curve can be produced from which measures of forward and backward flow are derived.
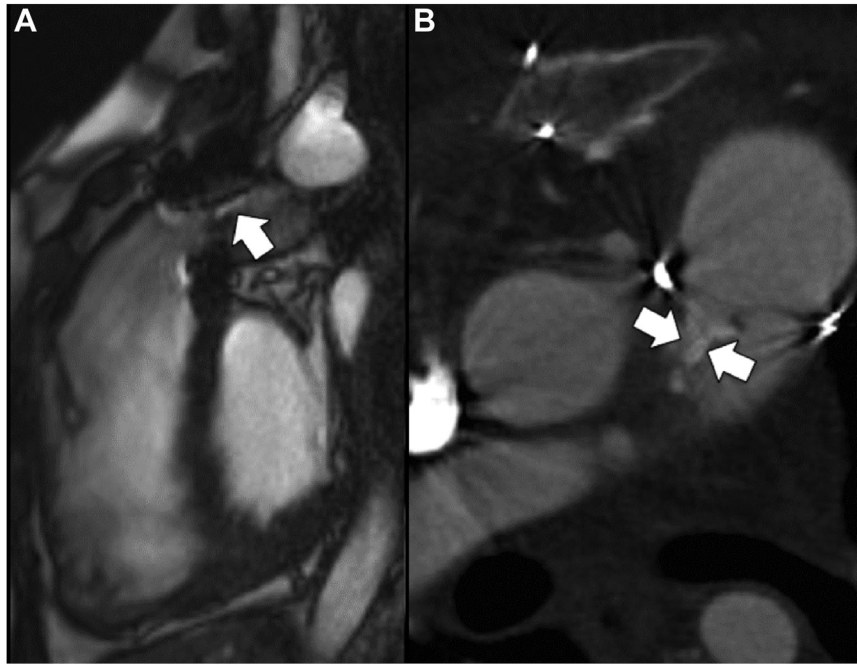
Figure 7. Outflow tract obstruction across a homograft conduit in tetralogy of Fallot identified by CMR and CCT. This patient with prior insertion of a pulmonary valve homograft conduit was noted to have increased velocities across the RVOT at echocardiography, but the exact level of obstruction could not be determined with certainty. (**A**) Sagittal cine SSFP CMR image demonstrates flow acceleration (**arrow**) that appears to be arising just above the level of the conduit valve. (**B**) Axial cardiac CT slice confirms that the obstruction is at valve level and is due to dystrophic calcification of both valve leaflets (**arrows**). CCT, cardiac computed tomography; CMR, cardiovascular magnetic resonance; RVOT, right ventricular outflow tract; SSFP, steady-state free precession.

Although aortopathy is commonly seen, especially in adult patients with tetralogy, it is rarely an issue requiring treatment, even in cases where the aortic root is larger than the threshold for surgery in other aortic expansion conditions.

Epidemiologic data suggest that thoracic aortic dissection is an uncommon complication in tetralogy with risk factors including age >60 (by far the strongest risk factor), male sex, and incompletely controlled hypertension.[24]

Figure 8. Subacute bacterial endocarditis complications in a patient with tetralogy. (**A**) There is evidence of soft tissue vegetation (**white arrow**) adjacent to the pulmonary valve leaflet. Evidence of local infection is seen with the presence of a loculated anterior pericardial effusion (**asterisks**) with thickened enhancing margins. (**B**) Disseminated spread of infection shown by the presence of low attenuation abscesses (**white arrows**) within the chest wall muscles.
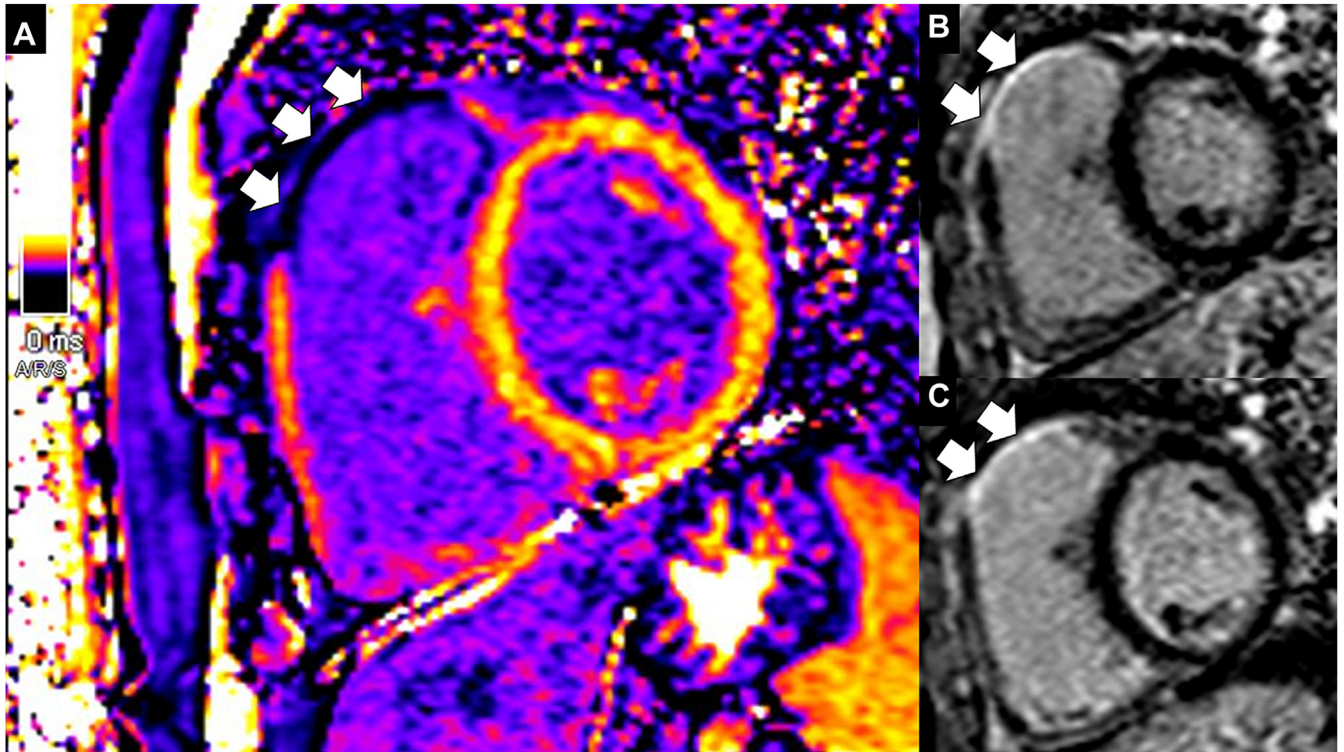
Figure 9. RVOT patch fibrosis on postcontrast imaging. (**A**) Postcontrast short axis T1 map with markedly reduced T1 (avid gadolinium uptake) in the region of the RVOT patch (**arrows**). (**B, C**) Standard late gadolinium enhancement images demonstrating similar diffuse contrast uptake (**arrows**) in the region of the patch. Note that patch material itself does not absorb contrast, but that the enhancement is presumed to be due to dense fibrosis that forms on the inner surface of the patch material. RVOT, right ventricular outflow tract.

Another late complication is aortic regurgitation. This may be due to abnormal aortic tissues or to the presence of aortic root dilatation from right to left shunting, if long-standing.[25]

Endocarditis can also be responsible. Therefore, imaging of the aortic root and aortic valve is an important component of any CMR protocol for tetralogy.
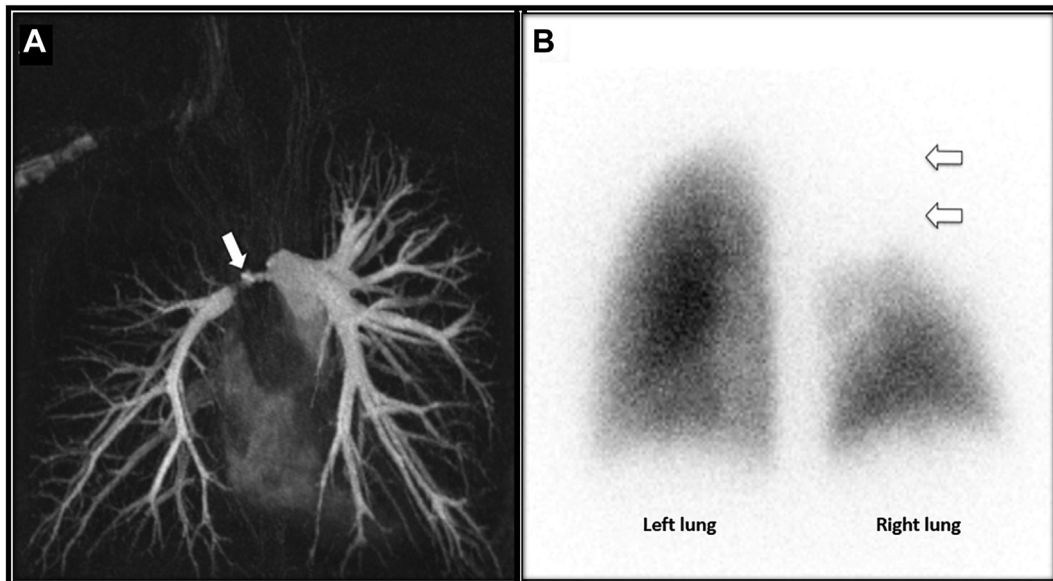

Figure 10. Differential lung perfusion in a patient with tetralogy of Fallot. (**A**) Coronal view of a magnetic resonance (MR) angiogram timed to display the pulmonary arterial tree. The right pulmonary artery appears irregular and abnormal (**arrow**) due to the presence of a metallic stent, which causes local distortion of the magnetic field and results in artefact. As a result MR flow measurements cannot be made here. (**B**) Quantitative lung perfusion (**rear view**) in the same patient demonstrates a lobar perfusion defect in the right upper lobe (**arrows**). Overall distribution of pulmonary flow was measured as 30% to the right lung and 70% to the left side.
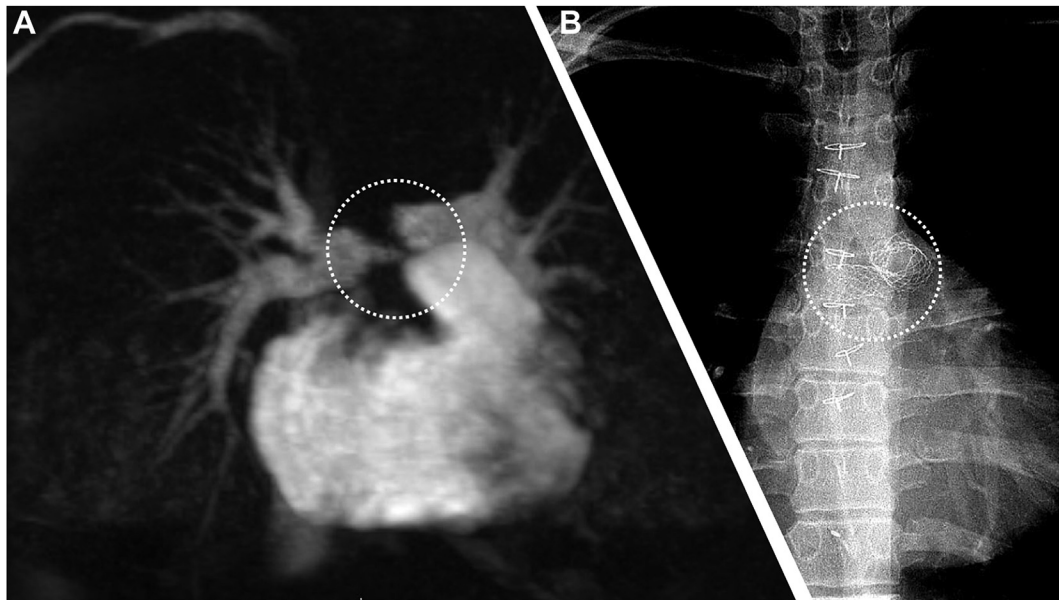
Figure 11. Effect of metal on visibility in CMR. Tetralogy patient with bilateral pulmonary artery stents. (**A**) Coronal MR dynamic angiogram in the pulmonary arterial phase. Note the odd appearance of both central pulmonary artery ostia (**dotted circle**). These do not look like typical stenoses, and the suspicion should be that the appearances are artefactual. (**B**) This is confirmed on plain CXR (shown here with the mediastinum sharpened for clarity), where bilateral PA stents (**dotted circle**) are visible. Metal usually causes local signal void on CMR. There is no good way around this, and cardiac CT was required in this case to confirm stent patency. CMR, cardiac magnetic resonance; CT, computed tomography; CXR, chest x-ray; MR, magnetic resonance; PA, pulmonary artery.

Other issues to consider include whether or not there may be a residual VSD leak. If so, this is usually due to a partial VSD patch dehiscence. Large leaks are rare but are important as they may lead to volume loading of the left ventricle and increase the risk of heart failure. Small leaks have no haemodynamic consequences but do present a risk of VSD patch endocarditis, and prophylaxis for subacute bacterial endocarditis remains recommended in such a case. Although both CT and CMR can identify patch leaks, echo is generally the modality of choice (Fig. 12) where this complication is
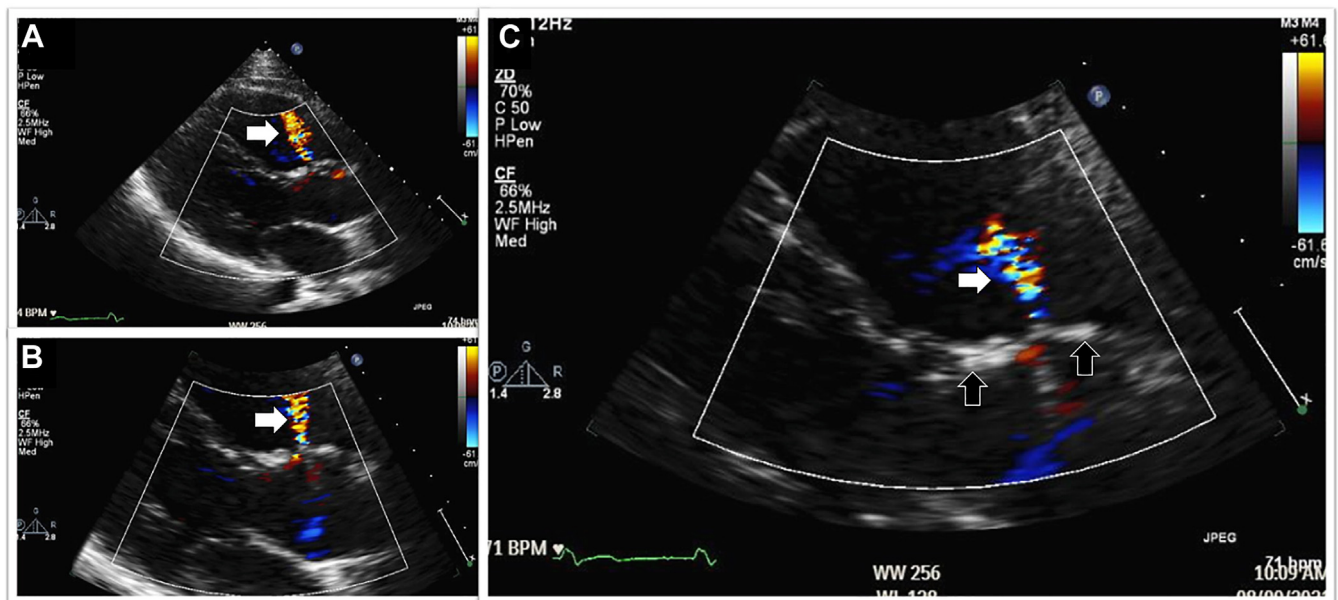

Figure 12. When CMR fails. This patient with tetralogy was noticed to have a loud systolic murmur that was out of keeping with expected physical findings. Pulmonary or RVOT stenosis was suspected clinically, and the patient underwent CMR, which failed to identify the cause. (**A-C**) Subsequent echo colour Doppler clearly demonstrated a small residual restrictive VSD with a systolic jet (**arrow**) through a tiny hole in the artificial (and hyperechogenic) VSD patch (**black arrows**). CMR, cardiac magnetic resonance; RVOT, right ventricular outflow tract; VSD, ventricular septal defect.
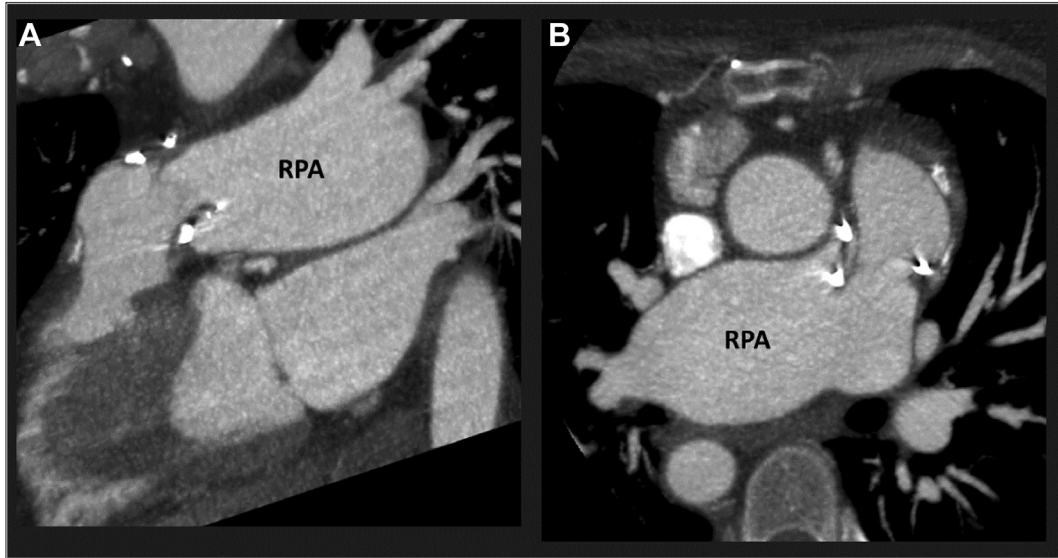
Figure 13. Example of a patient with tetralogy with absent pulmonary valve (after pulmonary valve replacement). (**A**) Sagittal oblique and (**B**) axial gated computed tomography images demonstrating a hugely enlarged right pulmonary artery (RPA). In this example, there was no major compression of any adjacent structures, but in some cases, there may be important dynamic compression of either the coronary arteries or adjacent airways, and evidence for this should be carefully sought.

suspected, especially where endocarditis is also present. Cross-sectional imaging may still be indicated, of course, for identifying embolic infection.

Special Cases of Tetralogy

Tetralogy with absent pulmonary valve

This is a relatively uncommon form of tetralogy that is seen in only 3%-6% of patients with ToF. The majority of cases

will need repair in infancy or childhood due to breathing difficulties, heart failure, or failure to thrive.[26] However, a small number may progress unrepaired to adulthood. These patients generally have very enlarged and highly pulsatile central PAs (Fig. 13) that may compress surrounding structures including airways and coronary arteries.[27] Compression of airways is particularly problematic in infants who have relative tracheomalacia compared with adults.

The author has personally encountered one middle-aged adult who developed angina of effort as a result of
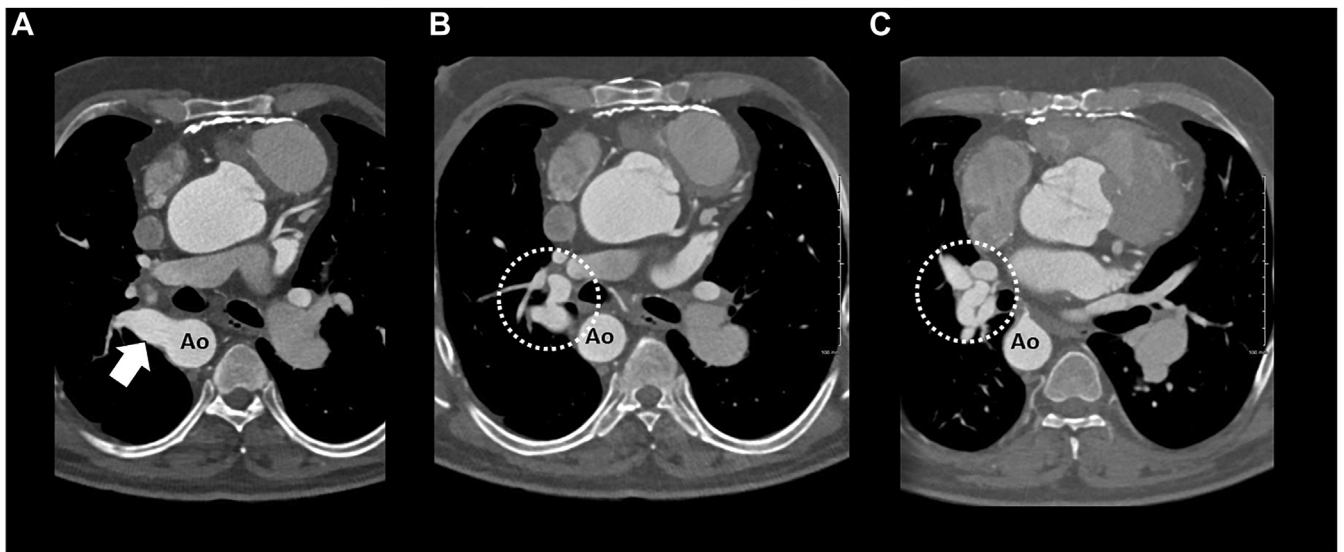

Figure 14. Major aortopulmonary collaterals in tetralogy with pulmonary atresia. Sequential axial slices from gated cardiac computed tomography. (**A**) A large aortopulmonary collateral (**arrow**) is seen to emerge from the proximal descending thoracic aorta (Ao). (**B** and **C**) The vessel takes a tortuous path (**dotted circles**) as it heads towards the mediastinum, from where it will travel out to supply a lung segment (not shown).

compression of the left main coronary artery, and symptoms could only be relieved by pulmonary artery plication. Sometimes the imaging specialist is required to think "out of the box" with unusual patients. In this case, we used full cardiac cycle coronary computed tomography angiography to confirm our suspicion of compression of the left main coronary by the enlarged pulmonary artery.

Tetralogy with pulmonary atresia

This condition resembles standard tetralogy except for the fact that there is complete obstruction between the RV and the pulmonary trunk.[28] The incidence is roughly 13-100 per million live births. In this condition the pulmonary trunk is atretic or hypoplastic; only very rarely is it normal. The central PAs are confluent in up to 86% of cases but are often underdeveloped with origin PA stenoses. Peripheral pulmonary arterial stenosis may also be seen in 1 or more areas.[25] Collateral pulmonary blood flow is the rule and may include flow from the subclavian, intercostal, and even the coronary arteries. However, the commonest source by far is the descending thoracic aorta, which may give rise to numerous and often quite large APCs (Fig. 14). On occasion these vessels are stenotic in places, which helps to prevent over circulation, but in many cases, pulmonary vascular disease and heart failure are common in later adult life.

If there are confluent central PAs, then an attempt may have been made to use these. One option is to encourage arterial growth by the use of a central shunt, and there will be occasional adult patients in whom these have been created and then left as a purely palliative procedure. However, the majority of patients will end up with a conduit placed between the RV outflow tract and the central PA confluence. Like any conduit these suffer age-related degeneration and calcification, and the need for eventual revision is very common.

The complexity of the initial surgical approach to tetralogy with pulmonary atresia also depends on the presence and size of major APC vessels. Where possible, these may be unifocalized to an adequate PA branch segment. If this is not technically achievable, they may be ligated—although if there is no dual pulmonary arterial supply, this is unsafe as it will likely lead to lung infarction. Another role for imaging is to identify the presence of stenoses within APCs, once they have been unifocalized. Although these are protective when the APCs are attached to the aorta, they diminish pulmonary flow once attached to that circulation. Both CT and magnetic resonance angiography are able to demonstrate such stenoses clearly, although the author favours CT if intervention is being considered, due to the intrinsically higher spatial resolution achievable.

Tetralogy with atrioventricular canal defect

This is a particular form of tetralogy seen most commonly in patients with trisomy 21. Surgical repair for these children is more complex than for standard tetralogy. In cases of staged repair, the PR expected after the repair of the RV outflow tract may lead to worsening right atrioventricular (AV) valve regurgitation. Furthermore, pre-existing left AV regurgitation may raise pulmonary pressures and worsen postoperative PR. Therefore, palliation with later repair (age 4-6 years) runs the risk of complications. Others argue for early complete repair as

a single-stage intervention, and this is now the choice of most centres. The problems faced in adult life are similar to standard tetralogy, and the same imaging protocols may be used. Small residual leaks at the atrial or ventricular level are usually best characterized by echo, even if they can be detected on cross-sectional imaging. Note that these AV valves are morphologically quite different from mitral and tricuspid valves and are composed of tissue known as "bridging leaflets." As a result, the correct terminology is "left and right AV valves," and the terms mitral and tricuspid are discouraged. A full description of this entity is beyond the scope of this article, but Umapathi et al.[29] provide a useful summary.

Concluding Thoughts

Hopefully, this review will have demonstrated that it is not difficult to image ToF competently and completely. However, it does require basic knowledge of the underlying lesions and an appreciation of the corrective surgeries that may have been applied at different points in the patient's life. Furthermore, the imaging clinician must be responsive to not only the questions of the referring physician but also the ability of the patient to tolerate the examination. Our job is to consider the question being asked and also think beyond the confines of CHD itself. If the patient with tetralogy is breathless but appears to have normal ventricular volumes and function with no intracardiac obstruction, we must ask ourselves whether there is another issue at play—whether that be weight, lung disease, or pulmonary vascular disease. After all, the patient with tetralogy may become obese, or get pneumonia, interstitial lung disease, emphysema, and pulmonary emboli just like every other adult. There has been a deliberate focus on the bread and butter imaging of tetralogy in this article. For those interested, more advanced imaging techniques—including 4D flow and tissue mapping—are covered in a separate review in this journal issue.

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