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Original Article

The role of cardiac catheterisation in patients with tetralogy of Fallot*

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Abstract Cardiac catheterisation plays an important role in the management of patients with tetralogy of Fallot. There are a number of palliative transcatheter interventions that can be performed in the neonatal period to allow for improved oxygen saturations and interval growth of the pulmonary arteries until corrective surgery is performed. Most patients develop branch pulmonary artery stenosis, right ventricular outflow tract obstruction, pulmonary insufficiency, or significant residual left-to-right shunts during long-term follow-up after corrective surgery. Transcatheter interventions can be performed to treat many of these issues, often eliminating or delaying the need for subsequent surgery. The indications for cardiac catheterisation and the specifics for various interventional procedures for patients with tetralogy of Fallot are reviewed in this manuscript.

Keywords: Cardiac catheterisation; tetralogy of Fallot

Cardiac catheterisation continues to play an important role in the management of patients with tetralogy of Fallot. Diagnostic catheterisation is rarely indicated in the neonatal period; however, there are a number of palliative transcatheter interventions that can be performed to improve oxygen saturations, and allow for interval growth of the pulmonary arteries before proceeding with a definitive surgical repair. During long-term follow-up of patients with tetralogy of Fallot, cardiac catheterisation with transcatheter intervention can be performed to diagnose and treat a number of problems including branch pulmonary artery stenosis, residual right ventricular outflow tract obstruction, pulmonary insufficiency, and residual left-to-right shunts. The current role of diagnostic and interventional catheterisation procedures in patients with tetralogy of Fallot is reviewed here.

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Catheterisation of the neonate with tetralogy of Fallot

Diagnostic catheterisation is rarely indicated for neonates with tetralogy of Fallot. Transthoracic echocardiography accurately delineates the intracardiac and great vessel anatomy in the majority of cases. In patients with more complex anatomy of the pulmonary artery branches, computed tomography angiography or magnetic resonance imaging can be utilised to better image these structures before surgical or catheter-based intervention. Magnetic resonance imaging has been shown to accurately demonstrate branch pulmonary artery anatomy and identify the presence of aorta to pulmonary collateral vessels; in addition, angiography is performed in the cardiac catheterisation laboratory.[1] In the rare patient with late presentation, a diagnostic catheterisation can be performed to delineate the pulmonary arterial blood supply and determine the pulmonary vascular resistance.[2]

There are a number of palliative transcatheter interventions that can be performed in the neonatal period to improve pulmonary blood flow, thereby increasing the saturations and allowing for interval growth of the branch pulmonary arteries. This can be an attractive alternative in patients who would be

at higher risk for complete surgical repair or surgical aorta to pulmonary shunt implantation such as premature or low birth weight infants, patients with genetic syndromes, or those with severely hypoplastic branch pulmonary arteries. Implantation of a stent in the arterial duct can be an attractive alternative to placement of a surgical shunt in patients who have favourable anatomy and orientation of the arterial duct (Fig 1). This is typically done through a femoral arterial approach using a 4 Fr sheath, but has also been described using surgical cutdown access to the carotid artery, and hybrid surgical approach with insertion of the delivery catheter into the main pulmonary artery.[3] This can also be attempted from the femoral venous approach; however, this is often technically more difficult. The typical stent diameter used in most patients is 3–4 mm. In the current era, the risk of complications from the procedure is relatively low, with zero percent procedure-related mortality in recent series.[4] Stenting of the arterial duct has been shown to result in good interval growth of the branch pulmonary arteries and may allow for more uniform growth, and blood flow may be more evenly distributed compared with a surgical Blalock–Taussig shunt.[5] The recovery time following the procedure is often much shorter than following a surgical shunt, with an average reported post-procedure hospital time of 5 days.[4] Care must be taken to cover the entire arterial duct to prevent ductal constriction and obstruction, particularly at the pulmonary artery end. Patency of the stented arterial duct can usually be maintained for at least 4–6 months, allowing for corrective surgery to be performed when the patient weighs 5 kg or more.[6] The limiting factors in maintaining stent patency are neo-intimal proliferation and thrombus. The stented arterial duct is amenable to repeat balloon angioplasty or additional stent implantation to maintain patency until corrective surgery can be performed if needed. The stents can be easily removed or trimmed at the time of corrective surgery.[7]

In patients who have undergone a surgical aorta-to-pulmonary shunt, stent implantation can be performed in the shunt in patients who develop acute cyanosis because of stenosis of the shunt in the presence of an occlusive thrombus. Implantation of a stent in a residual patent arterial duct has also been described as an urgent palliation in patients with acute surgical shunt occlusion.[8]

Stent implantation in the right ventricular outflow tract can be an effective way to augment pulmonary blood flow, particularly in patients with significant hypoplasia of the infundibulum and pulmonary annulus who will require significant surgical reconstruction at the time of complete surgical repair. This can also be a way to palliate patients with hypoplastic branch pulmonary arteries where surgical shunt placement can be challenging. This can be performed percutaneously, or using a hybrid surgical technique where the delivery catheter is inserted into the right ventricular

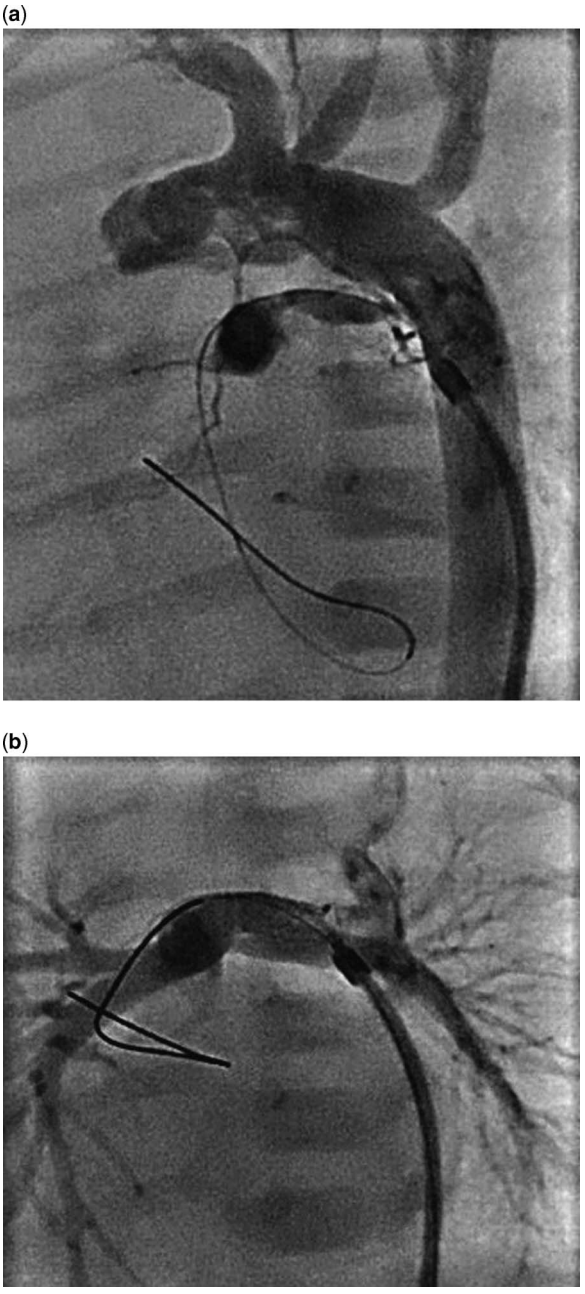

Figure 1.
*(**a**) Angiogram of the descending aorta in a neonate with tetralogy of Fallot and increased cyanosis demonstrating an arterial duct with stenosis of both the aortic and pulmonary ends of the duct. (**b**) Repeat angiogram post stent implantation of the arterial duct demonstrating uniform patency of the duct supplying confluent branch pulmonary arteries.*

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outflow tract through a periventricular approach. The latter technique has been performed in low birth weight infants with eventual successful complete surgical repair, with one reported in a premature neonate who weighed 840 g at the time of stent implantation.[9] Improved pulmonary artery diameter z-score has been reported following stent implantation in the right ventricular outflow tract.[10]

In patients who have right ventricular outflow tract obstruction secondary to valvar stenosis or hypoplasia of the pulmonary annulus, pulmonary balloon valvuloplasty can relieve stenosis and improve oxygen saturations. This can be performed from a percutaneous approach; this results in an increase in the pulmonary annulus size z-score, increased pulmonary artery diameter, and may decrease the need for a transannular patch at the time of corrective surgery.[11] This has also been performed intra-operatively at the time of a valve sparing type of surgical repair. This has shown to give comparable relief of obstruction, interval growth of the pulmonary annulus, and degree of pulmonary insufficiency when compared with surgical valvotomy or rigid valve dilation, even in patients with a hypoplastic pulmonary valve annulus.[12] However, the patients in the pulmonary balloon valvuloplasty group did have a shorter freedom from re-intervention or surgery than the other groups in this series.

In the occasional patient who had extensive aorta to pulmonary collateral vessels with dual supply to the native pulmonary arterial branches, pre-surgical transcatheter occlusion of these vessels with magnetic resonance imaging-compatible coils or other vascular occlusion devices can minimise blood return to the heart during cardiopulmonary bypass.

Post-operative interventions for tetralogy of Fallot

After complete surgical repair of tetralogy of Fallot, most patients require further intervention during long-term follow-up. Although repeat cardiac surgery is needed in many patients, there is an increased risk of morbidity and mortality associated with a repeat open sternotomy. Therefore, non-surgical interventions play an important role in the long-term management of patients with tetralogy of Fallot. In many cases, transcatheter interventions can obviate or significantly delay the need for further surgical intervention. Some patients require placement of a conduit from the right ventricle to the pulmonary artery. When conduit stenosis develops in patients with a conduit that is not large enough for placement of a transcatheter valve, stent implantation can give effective relief of stenosis and delay the need for eventual surgical replacement of the conduit. In larger conduits that develop progressive insufficiency, stenosis, or both, transcatheter valve implantation can also be performed. This is discussed in detail by Dr Jeremy Ringewald elsewhere in this edition.

In patients who have significant residual ventricular septal defects after surgical closure or additional defects in the muscular septum, transcatheter closure with various transcatheter occlusion devices can be performed. This can be done through a percutaneous approach if clinically indicated during long-term follow-up or through a periventricular approach at the time of surgical repair to close a significant additional defect in the muscular septum that may be challenging to close from a surgical approach. Transcatheter occlusion of residual aorta to pulmonary collateral vessels can also be performed in the later post-operative period if they are felt to cause significant additional pulmonary venous return to the heart, leading to symptomatic congestive heart failure.[13]

Branch pulmonary artery stenosis is common in patients with tetralogy of Fallot. This can be associated with decreased exercise intolerance in post-surgical tetralogy of Fallot patients. It may also exacerbate pulmonary insufficiency, although the exact relationship between branch pulmonary artery stenosis and pulmonary insufficiency is not well understood. Percutaneous balloon angioplasty can be performed to relieve stenosis in the branch pulmonary arteries; however, adequate relief of stenosis is not always achieved with balloon angioplasty alone. Stent implantation can provide long-term relief of stenosis the branch pulmonary arteries and subsequently improve blood flow distribution to the lungs, and reduce the right ventricular systolic pressure. Relief of unilateral branch pulmonary artery stenosis has been shown to decrease the degree of pulmonary insufficiency in an animal model.[14] Stents can be placed in the proximal or distal branch pulmonary arteries. In cases where there are stenosis in adjacent or bifurcating vessels, bifurcating or "kissing" stents can be implanted simultaneously to relieve these areas of stenosis. In addition to performing stent implantation through a percutaneous approach, intra-operative stent implantation has become an alternative approach as this gives more direct access to the branch pulmonary arteries for stent delivery and can be performed simultaneously with other planned surgical intervention (Fig 2).

The need for re-intervention is common to continue to enlarge the stented pulmonary artery because of patient growth. Long-term follow-up data have demonstrated that the final pulmonary artery

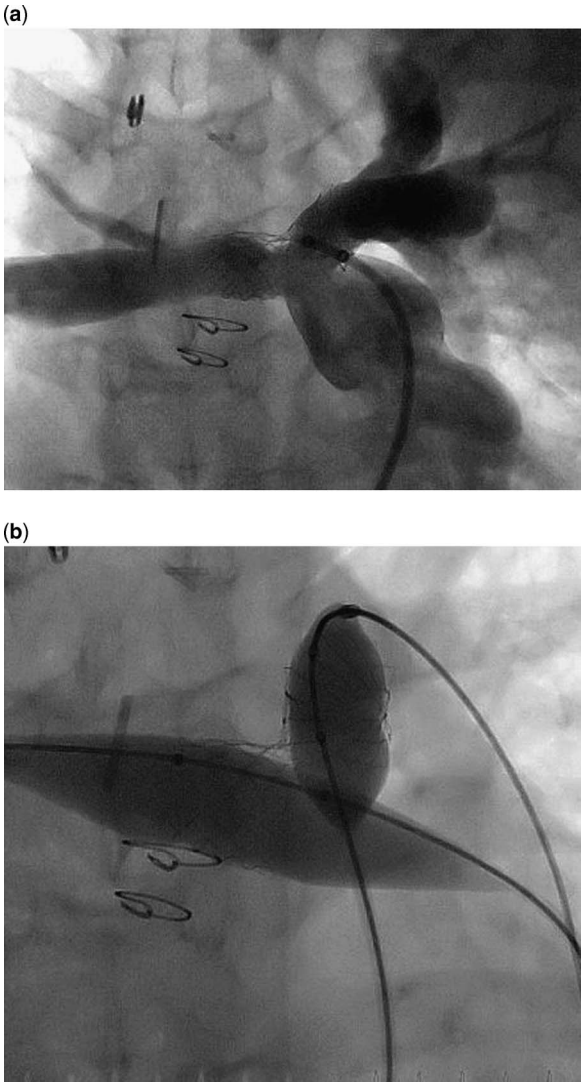

Figure 2.
*A 16-year-old patient who underwent complete surgical repair of tetralogy of Fallot as an infant, and developed bilateral proximal branch pulmonary artery stenosis. He had bilateral pulmonary artery stents placed at 2 years of age, and underwent repeat balloon angioplasty of the stents at 6 years of age. (**a**) Main pulmonary artery angiogram showing good flow into both branch pulmonary arteries. There is some intimal build-up noted in the proximal right pulmonary artery. (**b**) Simultaneous balloon angioplasty of the stents using 14-mm-diameter balloons. Post angioplasty, the right ventricular systolic pressure was 35 mmHg with simultaneous arterial blood pressure of 104 mmHg.*

diameter, pressure gradient across the stenosis, and right ventricular systolic pressure remain significantly improved compared with pre-stent values at the time of repeat catheterisation.[15] Most patients underwent further balloon angioplasty or additional stent implantation during long-term follow-up. Stent fractures do not appear to be common during long-term follow-up and can be treated with second

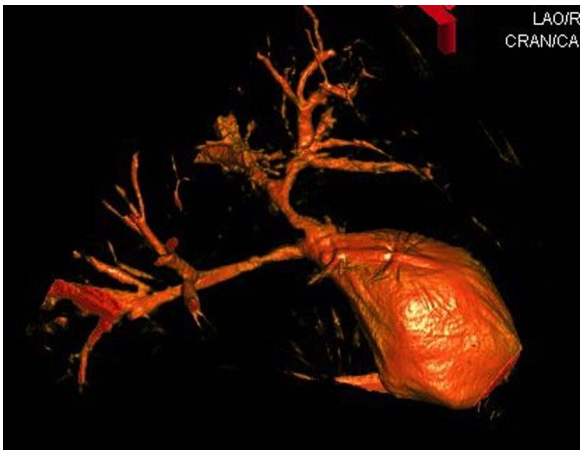

Figure 3.
A three-dimensional computed tomography image obtained from rotational angiography in a 2-year-old patient with Alagille syndrome and tetralogy of Fallot who underwent repair with a transannular patch. There is a large aneurysm of the right ventricular outflow tract and main pulmonary artery. There is severe bilateral branch pulmonary artery stenosis.

stent implantation if necessary. Stents implanted in the branch pulmonary arteries intra-operatively also have a high rate of re-intervention with over half requiring further balloon angioplasty within 7.6 years of implantation.[16] Stents placed in patients <2 years of age or implanted at a diameter <10 mm have an increased rate of re-intervention. Pulmonary artery stents can also be manipulated surgically, including removal of the entire stent, trimming of the proximal ends to allow for conduit placement, or longitudinal transection and patching to increase the diameter of the stented vessel.[17]

More recently, angiographic computed tomography using rotational angiography has been used during cardiac catheterisation of patients with congenital heart disease, including tetralogy of Fallot. It allows for the precise delineation of the three-dimensional anatomy of vascular structures, including the branch pulmonary arteries without a significant increase in contrast or radiation exposure.[18] It can also be useful to guide complex interventions such as pulmonary artery balloon angioplasty, stent implantation, and transcatheter valve deployment (Fig 3).

Summary

Transcatheter interventions play an important role in the long-term management of patients with tetralogy of Fallot. There are a number of palliative interventions that can be performed in the neonatal period to augment pulmonary blood flow, improve oxygen saturations, allow for interval growth of the branch pulmonary arteries, and allow for growth

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and stabilisation of the patient before performing corrective surgery. During long-term post-operative care, most patients with tetralogy of Fallot require further intervention because of branch pulmonary artery stenosis, residual right ventricular outflow tract obstruction, pulmonary insufficiency, or significant residual left-to-right shunts. Transcatheter interventions can improve patient condition and quality of life, and can often delay or may eliminate the need for repeat surgical intervention.

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