Cardiac Magnetic Resonance Imaging of late complications post repair of Tetralogy of Fallot; Pictorial review of sequelae in Grown-Up Congenital Heart disease

Poster No.: C-1167
Congress: ECR 2012
Type: Educational Exhibit
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Keywords: Cardiac, MR, Diagnostic procedure, Congenital
DOI: 10.1594/ecr2012/C-1167

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Learning objectives

In patients with surgically corrected Tetralogy of Fallot, combine the anatomical and functional findings from Cardiac Magnetic Resonance Imaging to facilitate assessment in often asymptomatic patients.

1. Evaluate pulmonary regurgitation including relationship to transannular patch repair.
2. Illustrate right ventricular outflow tract obstruction using cardiac MRI.
3. Display the spectrum of right and left ventricular dysfunction.
4. Outline the manifestations of aortic root dilation and aortic regurgitation.
5. Clarify the imaging parameters which indicate late surgical intervention.

Background

The tetralogy of Fallot is described as

- Right Ventricular outflow tract (RVOT) obstruction
- Ventriculo-Septal Defect (VSD) - right-to-left shunt
- Pulmonary stenosis
- Overriding aorta

Surgery is performed

- To close the VSD
- To relieve the right ventricle outflow tract obstruction

Review of the surgical technique.

The surgical approach is patient-specific and initial assessment of the anatomy is fundamental. The technique is optimised to encompass the spectrum of severity of each component of the tetralogy.

VSD Closure
The VSD in tetralogy of Fallot is usually single, large and perimembranous in approximately 80% of patients. VSD closure is achieved using a synthetic patch and is a very successful technique [1].

A trans-atrial incision is preferred to minimise right ventricular damage. The typical traditional incision for VSD closure is the anterior wall of the right ventricle. As a consequence, it is important to recognise the characteristic post surgical apical thinning of the right ventricle. This is usually best appreciated on the 4 chamber view (Figure 1).

![Fig. 1: 4 chamber HASTE sequence demonstrating apical thinning (red arrow) following a transapical approach for surgical correction.](image)

**References:** K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

**RV outflow tract obstruction**

The spectrum of right ventricular outflow tract obstruction and the age of the child at the time of surgery influence the surgical technique.
The least invasive approach is removal of infundibular muscle bands via a right atrial or pulmonary artery approach. In smaller infants or in cases of severe stenosis a direct incision over the right ventricular outflow tract is required with or without pulmonary valvotomy.

**Transannular Patch**

If the infundibular stenosis is severe, the right ventricular outflow tract is incised anteriorly and a transannular patch extends across the pulmonary valve annulus to increase the lumen of the right ventricular outflow tract.

As previously commonly used pericardium was particularly prone to aneurysm formation, a synthetic material is now applied.

The surgical approach at the level of the RVOT is considered in combination with the degree of pulmonary stenosis distally. If there is distal stenosis, the transannular patch can extend to the level of the main pulmonary artery bifurcation. This wide and long patch is particularly prone to aneurysm formation.

Pulmonary valve implantation, if required, is usually performed with a homograft. The immediate benefit of relieving pulmonary stenosis is balanced with the risks of further staged procedures to modify the conduit as the child grows.

Although there is some variation in opinion, current surgical techniques have advanced such that an infant may undergo correction of the Tetralogy between the neonatal period to 2-6 months. Previously the infant would routinely have undergone palliative systemic to arterial shunts in the neonatal period and a corrective procedure at a later stage.

Adequate relief of right ventricular outflow tract obstruction is critical in the perioperative period of a repaired Tetralogy of Fallot, because persistent RVOT obstruction after successful closure of the VSD can cause significant morbidity.

**Arteriopulmonary Shunts**

In cases of severe pulmonary stenosis that could preclude alleviation of the RVOT obstruction, an initial arteriopulmonary shunt procedure can be performed prior to a definitive repair of the VSD and RVOT obstruction.
The arteriopulmonary shunt affords development of the pulmonary arterial supply. Therefore the pulmonary arteries are larger and more robust prior to the definitive procedure. The increase in calibre is reported to be approximately 20%[2].

New techniques have emerged, such as pulmonary artery annulus angioplasty, which can be performed in early infancy to increase blood flow as an alternative to palliative shunting. The angioplasty can be performed at the same time as corrective surgery.

There are a number of different types of aortopulmonary shunts, the anatomical differences of which are described in the table below.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Arteriopulmonary shunt</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blalock-Taussig (Classical)</td>
<td>Subclavian artery to pulmonary artery (end-to-side)</td>
</tr>
<tr>
<td>Blalock- Taussig (Modified)</td>
<td>Interposition graft between subclavian artery and uni- or bilateral pulmonary artery</td>
</tr>
<tr>
<td>Waterston</td>
<td>Ascending aorta to main or right pulmonary artery (side-by-side)</td>
</tr>
<tr>
<td>Potts</td>
<td>Descending aorta to left pulmonary artery (side by side)</td>
</tr>
</tbody>
</table>

**Major Aorto-Pulmonary Collateral Arteries (MAPCAs)**

In pulmonary atresia the increased resistance causes diversion of blood into the aorta and marked enlargement of the small collaterals that exist between the aorta and pulmonary arteries, called MAPCAs. These can result in complex anastomoses with the atretic native pulmonary arteries and in effect a conduit with the right ventricular outflow tract.

Routine assessment for MAPCAs is important as if they remain patent they may cause a degree of shunting. In the case of solitary or very small numbers of MAPCAs, endovascular coiling can be a less invasive approach for control (Figure 2).
**Fig. 2:** Multi-detector CT Angiography demonstrating a left lower lobe MAPCA. In the left image (yellow asterisk) demonstrates the collateral arteries. In the right image the red asterisk annotates the proximal endovascular coil to occlude the MAPCA.

**References:** K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

Images for this section:
**Fig. 1:** 4 chamber HASTE sequence demonstrating apical thinning (red arrow) following a transapical approach for surgical correction.
Fig. 2: Multi-detector CT Angiography demonstrating a left lower lobe MAPCA. In the left image (yellow asterisk) demonstrates the collateral arteries. In the right image the red asterisk annotates the proximal endovascular coil to occlude the MAPCA.
Imaging findings OR Procedure details

Longitudinal assessment of patients with Grown Up Congenital Heart Disease is routinely performed annually. The aim of this study is to demonstrate the imaging findings of the spectrum of abnormalities within this patient population and describe which require surgical intervention.

Evaluation of Pulmonary regurgitation

Flowing mapping sequences sum the velocities from all the pixels in a given region of interest throughout the cardiac cycle. It is important that the selected slice is perpendicular [3]. This enables accurate quantification of forward and reverse flow and therefore if present, pulmonary regurgitation can be objectively measured. Accurate contour drawing is important to achieve an appropriate area and reliable data using the Argus software(Figure 3).

Fig. 3: Argus analysis of the right pulmonary artery flow mapping, measuring velocity perpendicular to the selected slice of interest.
Velocity v Time graphs can correlate with echocardiography. Although flow mapping is the gold standard the combination of MRI and echocardiography often produce congruent and complementary results (Figure 4). The area under the x-axis is representative of the pulmonary regurgitant fraction.

**Fig. 4:** Left image- Velocity v time plot of flow mapping analysis of the main pulmonary artery. Right image - Echocardiogram of the same patient demonstrates identical velocities on doppler analysis across the pulmonary valve. The regurgitant fraction of 33% is annotated with a red arrow on both images.

*References:* K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

The RVOT views throughout the cardiac cycle demonstrate pulmonary regurgitation (figure 5 and 6).
Fig. 5: SSFP images RVOT view of pulmonary regurgitation as demonstrated by the red arrow. * = right ventricle.

References: K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

Fig. 6

Pulmonary regurgitation is common when the surgical repair has involved a transannular patch repair, when the RVOT is usually dilated (Figure 7). The clinical significance relates to the associated sequelae of the right ventricle and distal RVOT obstruction.
Fig. 7: Right Ventricular Outflow Tract view which is dilated due to previous transannular patch repair. The red arrow indicates the maximal convexity, just caudal to the level of the pulmonary valve.

References: K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

Assessment of Right Ventricular Outflow Tract Obstruction

The cause of Tetralogy is an aberrant embryological development of the infundibulum. The right ventricular outflow tract develops from the infundibular spiral bud and therefore
there can be multiple levels of RVOT obstruction in each patient, with a spectrum of severity.

There can be residual bands of muscular hypertrophy following surgical repair (Figure 8) with a range of haemodynamic effects including significant flow acceleration (Figure 9).
**Fig. 8:** Right Ventricular Outflow tract view which demonstrates the densely thickened infundibular bands (red arrow).

**References:** K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

**Fig. 9:** A series of Right Ventricular Outflow Tract (RVOT) views with dense infundibular bands (red arrow) and turbulent flow acceleration at this level.

**References:** K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

Although less common than pulmonary regurgitation, pulmonary stenosis is seen following surgical repair (Figure 10, Figure 11). In this patient the residual right ventricular outflow tract obstruction can caused some pressure related right ventricular hypertrophy.
Fig. 10: RVOT view with pulmonary stenosis and a hypertrophied right ventricle.

References: K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM
Fig. 11: Transverse axis views of the right ventricular outflow tract with stenosis at the level of the pulmonary annulus (red arrow)

References: K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM
It is important to assess the pulmonary valve at least two planes. The transverse views are particularly useful to assess the relationship of pulmonary stenosis to the main pulmonary artery bifurcation (Figure 12) both in terms of post stenotic dilation or distal stenosis.

Fig. 12: Transverse views of the RVOT. Image A post stenotic dilatation and residual bands at the level of the pulmonary annulus (yellow arrow) and associated turbulent flow (red arrow).

References: K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

In pulmonary artery stenosis, this may be treated with an endovascular stent (Figure 13, 14).
Fig. 13: Maximum Intensity Projection image with red arrow highlighting the severe left main pulmonary artery stenosis.

References: K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM
Fig. 14: Coronal and oblique projection of the contrast enhanced CT (computed Tomography) of the chest with red arrows highlighting the successful result following endovascular stunting of the left main pulmonary artery. Please refer to the pre-procedure images of Figure 13

References: K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

Stenosis of the main pulmonary arteries is often related to palliative procedures[4]. In grown up congenital heart disease there is often a history of multiple operations and therefore the palliative shunt may no longer be visible at the time of imaging but the secondary sequelae are evident. Alternatively if present, the non-functioning arteriopulmonary shunts may be thrombosed, of no clinical significance (Figure 15).
Fig. 15: A Maximum Intensity projection coronal view with post processing. This highlights the relative paucity of pulmonary arteries on the left relative to the right. B The red arrows demonstrate a Modified Blalock-Taussig shunt from the left subclavian artery to the left pulmonary artery. C Contrast enhanced CT demonstrating Left Modified Blalock Taussig conduit thrombosis, highlighted by the red asterix immediately to the left of the conduit.

References: K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

3. Spectrum of Right and Left Ventricular Dysfunction

The right ventricle may be dilated due to severe RVOT obstruction, combined with scar due to a transapical surgical approach. This may cause tricuspid regurgitation and the associated atrial enlargement may predispose to arrhythmias. Often the morphology of right ventricular dysfunction is a combination of volume and pressure overload.

Volume overloaded Right Ventricle (RV)

The features of right ventricular enlargement with trabeculation but without right ventricular hypertrophy are consistent with excessive volume load (Figure 16, 17).
Fig. 16: 2 chamber SSFP image on the left and 4 chamber SSFP image on the right both demonstrate a hypertrabeculated dilated right ventricle in keeping with volume overload.

References: K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

Fig. 17

Pressure Loaded Right Ventricle

Features are of right ventricular hypertrophy, without right ventricular dilation (Figure 18,19).

It is described that restrictive RV physiology does prevent right ventricle dilation and maintains exercise capacity in adults[5].
Fig. 18: SSFP right ventricular outflow tract imaging, demonstrating right ventricular hypertrophy and normal calibre right ventricle, features of pressure overload.

References: K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

Fig. 19

Combined Right ventricular pressure and volume overload

Figure 20
Fig. 20: SSFP cardiac MRI, 4 chamber view (left) and 2 chamber view (right) demonstrating - Right ventricular dilation, (red asterix), - Right ventricular increased trabeculation (red arrow) - Right ventricular hypertrophy (red arrowheads)

References: K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

Left ventricular dysfunction is less prevalent and most commonly related to dilation of the ascending aorta or severe right ventricular dysfunction.

4. Aneurysmal Ascending Aorta and Aortic Regurgitation

Aortic root dilation is evident in 15% of adults after tetralogy repair. It is thought to be related to both cystic medial necrosis and increased flow due to pulmonary atresia. Although aortic regurgitation commonly develops, aortic dissection is very rare (Figures 21-23).
Fig. 21: SSFP Cardiac MRI demonstrating the Left Ventricular Outflow Tract (LVOT) view with dilation of the ascending part and effacement of the sinotubular junction (red arrowhead).

References: K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM
**Fig. 22:** Series of LVOT views A (systole), B mid cycle, C (diastole). The regurgitation jet from the aortic valve is highlighted using the red asterix.

*References:* K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

**Fig. 23**

Other complications of tetralogy repair include VSD patency. In addition presence of delayed myocardial enhancement due to scar tissue with associated poorer prognosis [6].

**Fig. 24:** Delayed Myocardial Enhancement is assessed on these post gadolinium inversion recovery sequences. These modified oblique views demonstrate delayed myocardial enhancement thought to be related to post operative scarring.

*References:* K. Tweed; Papworth Hospital, Cambridge, UNITED KINGDOM

5. Indications for late surgical Intervention
Pulmonary regurgitation is the most frequent condition to prompt consideration for surgery with less than 1 percent mortality.

The decision to proceed is usually guided by a combination of functional assessment and objective measurements, particularly of the degree of PR and the right ventricular dilation.

Longitudinal data are very important to guide intervention, particularly for the functional component. However in terms of objective measures the regurgitant fraction is considered in combination with the right ventricle dilatation. After the end diastolic index exceeds 160 ml/m2 remodeling post repair is doubtful [7].

If planning surgery complete assessment is critical to balance other negating factors, most commonly concurrent pulmonary artery stenosis. This along with distal right or left pulmonary artery stenosis may be amenable to stenting.

Pulmonary valve replacement is usually by a tissue valve with expected lifespan of 10-15 years rather than a mechanical prosthesis. This is in part due to the challenges of maintaining adequate anticoagulation in often relatively young patients.
<table>
<thead>
<tr>
<th>Surgical intervention</th>
<th>Indication</th>
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<tbody>
<tr>
<td>Aortic valve replacement</td>
<td>Severe Aortic Regurgitation</td>
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<tr>
<td></td>
<td>Symptoms of Left Ventricular dysfunction</td>
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<tr>
<td>Pulmonary valve replacement</td>
<td>Symptomatic patients with severe Pulmonary regurgitation</td>
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<td></td>
<td>Symptomatic patients with severe Pulmonary stenosis</td>
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<td></td>
<td>Right ventricular systolic pressure &gt; 60 mmHg</td>
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<tr>
<td>Pulmonary valve replacement</td>
<td>Asymptomatic patients with severe Pulmonary regurgitation and/or Pulmonary</td>
</tr>
<tr>
<td>Consideration</td>
<td>stenosis and at least one of the following</td>
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<tr>
<td></td>
<td>- decrease in objective exercise capacity</td>
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<tr>
<td></td>
<td>- progressive Right Ventricular dilation</td>
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<tr>
<td></td>
<td>- progressive Right Ventricular Systolic Dysfunction</td>
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<td></td>
<td>- progressive Tricuspid Regurgitation (moderate)</td>
</tr>
<tr>
<td></td>
<td>- Right Ventricular Outflow Tract Obstruction,</td>
</tr>
<tr>
<td></td>
<td>systolic pressure &gt; 80 mmHg (TR velocity &gt; 4.3 m/s)</td>
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<tr>
<td></td>
<td>- sustained atrial/ventricular arrhythmias</td>
</tr>
<tr>
<td>VentriculoSeptal Defect (VSD)</td>
<td>Residual VSD and significant Left Ventricular volume overload</td>
</tr>
<tr>
<td>Closure Consideration</td>
<td>or if the patient is undergoing pulmonary valve surgery</td>
</tr>
</tbody>
</table>

**Table 1**: Indications for surgical intervention.


**Images for this section:**
Fig. 6: Right Ventricular Outflow Tract cine imaging (SSFP Cardiac MRI sequence) demonstrating the 33% pulmonary regurgitant jet.
Fig. 7: Right Ventricular Outflow Tract view which is dilated due to previous transannular patch repair. The red arrow indicates the maximal convexity, just caudal to the level of the pulmonary valve.
**Fig. 8:** Right Ventricular Outflow tract view which demonstrates the densely thickened infundibular bands (red arrow).

![Image of Right Ventricular Outflow tract view with a red arrow highlighting the densely thickened infundibular bands.]

**Fig. 13:** Maximum Intensity Projection image with red arrow highlighting the severe left main pulmonary artery stenosis.

![Image of Maximum Intensity Projection image with a red arrow highlighting severe left main pulmonary artery stenosis.]
**Fig. 15:** A Maximum Intensity projection coronal view with post processing. This highlights the relative paucity of pulmonary arteries on the left relative to the right. B The red arrows demonstrate a Modified Blalock-Taussig shunt from the left subclavian artery to the left pulmonary artery. C Contrast enhanced CT demonstrating Left Modified Blalock Taussig conduit thrombosis, highlighted by the red asterix immediately to the left of the conduit.
Fig. 16: 2 chamber SSFP image on the left and 4 chamber SSFP image on the right both demonstrate a hypertrabeculated dilated right ventricle in keeping with volume overload.

Fig. 17: 4 chamber cine SSFP sequence. The right dilated trabeculated right ventricle has typical features of volume overload.
Fig. 19: Cine imaging of the right ventricular outflow tract with pressure overload of the right ventricle.
Fig. 20: SSFP cardiac MRI, 4 chamber view (left) and 2 chamber view (right) demonstrating - Right ventricular dilation, (red asterix), - Right ventricular increased trabeculation (red arrow) - Right ventricular hypertrophy (red arrowheads)
Fig. 23: LVOT cine imaging (SSFP) illustrating aortic regurgitation.
Conclusion

Cardiac magnetic resonance imaging in grown up congenital heart disease is fundamental to evaluate late complications of Tetralogy of Fallot repair. It is particularly useful to assess the right ventricular outflow tract obstruction, in combination with pulmonary valve and right ventricular function.

Early detection of the complex spectrum of sequelae in often asymptomatic patients can guide monitoring, medical therapy and surgical intervention.

Personal Information

References


