Imaging congenital heart disease

By Dr Helen Mathias*, Dr Nathan Manghat* and Dr Mark Turner†
Departments of Radiology* and Cardiology†, Bristol Heart Institute, Bristol Royal Infirmary

Congenital heart disease encompasses a spectrum of anomalies from the simple ASD to the complex multiple anomalies such as tetralogy of Fallot and hypoplastic left heart.

Imaging of these patients can be complex and the diagnosis and follow-up is largely performed in tertiary centres by multi-disciplinary teams including specialist radiologists and cardiologists. In children, echocardiography is the main imaging modality with reference to catheter angiography, MRI and occasionally CT for specific indications. In adults, echocardiography is also widely used, but poor echo windows and difficulty measuring the right ventricle has resulted in increasing reliance on MRI for these patients.

MRI is less limited by body habitus and its lack of ionising radiation makes it ideal for the repeated investigations required in the follow-up of these young patients. CT is used in certain circumstances, eg in a patient with a contraindication for MRI (eg pacemaker), as a problem-solving tool in specific cases and remains the first line cross-sectional imaging modality in the emergency setting. CT is also used to image stents and devices which cause susceptibility artefact limiting MRI assessment.

As treatment of complex cardiac anomalies has evolved (prenatal diagnosis, improving surgical and intensive care etc), an increasing number of children are surviving to adulthood and these patients are encountered in general radiology practice. These encounters are often as part of the on-call radiology service and can represent an acute presentation of a problem related to the patient’s congenital condition or previous surgery, or be unrelated. In either case, a knowledge of the relevant anatomy and surgical procedure along with potential complications is imperative to enable prompt diagnosis. A previously undiagnosed congenital anomaly may also be uncovered when imaging the acutely unwell patient.

Accurate interpretation of imaging of these patients demands an understanding of the anatomy. A detailed description of the relevant nomenclature is beyond the scope of this article but can be found in most paediatric cardiology texts (eg Anderson et al, 2001). The cardiac connections encountered may represent abnormal congenital anatomy or, more likely, a surgical repair (some of which restore the anatomy towards normal and some make it more abnormal, eg the Fontan circulation). Operative notes are invaluable but may not be available outside the tertiary centre, so liaison with the surgical centre is often helpful. As a specialist centre we have round-the-clock consultant cardiologist on-call cover for both adult and paediatric congenital heart disease.

This article cannot provide a comprehensive guide to imaging these patients but merely seeks to raise awareness and highlight a few examples of scenarios that may be encountered by the general radiologist.

A detailed explanation of radiological follow-up can be found in the recently published Guidelines from the European Society of Cardiology.

Patients with a history of aortic surgery presenting with haemoptysis

Aortic coarctation occurs in 1 in 3000-4000 live births and is generally diagnosed in childhood. It comprises a focal narrowing of the aorta in the proximal descending aorta, just distal to the ductus/ligamentum arteriosus. Treatment options include balloon angioplasty or stenting and open surgical repair, usually with resection and end-to-end anastomosis. Subclavian flap repair and patch aortoplasty are rarely used today but patients may be seen for imaging. Follow-up is important to exclude late complications associated with the repair, such as recoarctation or pseudoaneurysm, and is performed with focused CT or MRI.

An uncommon but potentially life-threatening complication is an aortobronchopulmonary fistula. This can present with haemoptysis which may initially appear minor or can be immediately life-threatening. Patients with previous Dacron patch repair are at particular risk of this and despite the technique no longer being widely used, there are many patients with this repair many of whom may have been lost to follow-up and may present to a general medical take. More generally, any patient with a history of thoracic aortic surgery is at risk of aortobronchopulmonary fistula.

Chest X-ray may reveal a thoracic aortic aneurysm or air space opacification within the adjacent lung, but in one series chest X-ray only demonstrated the aneurysm in 16% of cases. Therefore, urgent contrast-enhanced CT of the thoracic aorta should be performed. This should demonstrate the thoracic aortic aneurysm or pseudoaneurysm and may show airspace opacification/consolidation within the adjacent lung. These findings with the appropriate history are diagnostic of an aortobronchopulmonary fistula, although the fistula itself is rarely visualised. If diagnosed in a timely fashion, endovascular or surgical treatment can be successful (figure 1).

Complex congenital including single ventricle CTPA

In complex congenital cardiac conditions, anatomical correction is not possible. The Fontan procedure is performed in a range of congenital conditions where a biventricular repair is not possible due to hypoplasia of a ventricle or inability to make two atrio-ventricular valves. Repair involves creating a total cavopulmonary connection (TCPC) where the SVC and IVC are directed to the pulmonary artery, thus bypassing the heart entirely. Pulmonary blood flow then relies on passive flow of blood driven by respiratory activity and ventricular suction. These patients will be seen in general radiological practice in infection or numbers. Pulmonary embolus can occur in these patients because of thrombosis in the venous pathways and can be catastrophic for the patient. However, due to the abnormal central vascular connections, imaging of the pulmonary arterial tree is challenging and if the anatomy is not understood the potential for erroneous interpretation of such imaging is high.

As the blood from the SVC drains directly into the RPA, an upper limb contrast injection may only opacify the right pulmonary artery if a standard CTPA protocol is employed. This may lead to unopacified blood within the left pulmonary artery, giving the false impression of a large central embolus. These findings with the appropriate history are diagnostic of an aortobronchopulmonary fistula, although the fistula itself is rarely visualised. If diagnosed in a timely fashion, endovascular or surgical treatment can be successful (figure 1).

Various complex patient-specific contrast and radiation strategies are proposed to overcome this problem, eg:

1. The need for a test bolus of contrast to optimise the scan to the patient’s own haemodynamics
2. Use a prolonged contrast bolus or biphasic contrast injection protocol (eg 80mls at 7mls/sec followed by 40mls at 2-3 mls/sec) with a second delayed phase scan allowing for contrast recirculation
3. Simultaneous upper and lower limb contrast injections (this is more difficult to achieve without a second pump injector or appropriate ‘y’ connectors)
A larger bolus of contrast at a significantly higher flow rate may be needed for adequate arterial opacification (with reference to standard contrast haemodynamic flow curves) in the presence of an intra or extracardiac shunt, if there is substantial valvular regurgitation (notably pulmonary and tricuspid) and/or when there is severe chamber dilatation.

A delayed phase scan increases the radiation burden but additional clinical information that can be gained may justify it. Patient-specific radiation reduction strategies are therefore of paramount importance, notably reduction in kilovoltage. Awareness of the potential pitfalls and specialist advice can help to optimise the clinical information and minimise the radiation dose in these young patients.

**Right heart dilatation**

CTPA is a commonly performed investigation in the acute setting in patients with shortness of breath and/or chest pain. Despite these scans being performed using a non-gated technique, a significant amount of diagnostic information regarding the heart can be obtained.

When pulmonary emboli are present it is important to look for signs of right heart strain, including right ventricular dilatation, flattening of the interventricular septum or bowing to the left in severe cases, and reflux of contrast into the IVC, hepatic andazygous veins indicating raised right heart pressures.

In the absence of pulmonary emboli other causes to explain the presenting symptoms should be sought and right heart dilatation may be an important observation. Such findings can be easily overlooked and a mental “checklist” can be helpful.

Left to right shunts at atrial level are an important cause of right heart dilatation and may be identified on CT as a jet of contrast traversing the atrial septum depending on the relative contrast opacification of the cardiac chambers (figure 3).

A commonly overlooked abnormality is the anomalous pulmonary venous drainage in which one or more pulmonary veins drain into the systemic venous circulation, including the SVC, IVC or brachiocephalic vein. Anomalous venous drainage can also be seen as part of sinus venous ASD (where there is an ASD where the SVC or IVC join the heart) (figure 4).

Pulmonary valve abnormalities are another cause of right heart abnormalities and may cause dilatation in the presence of pulmonary regurgitation or right ventricular hypertrophy with pulmonary stenosis. This finding is particularly common in patients following tetralogy of Fallot repair but may also be present with isolated, possibly previously undiagnosed pulmonary valve abnormalities. Enlarged central pulmonary arteries should raise the suspicion of pulmonary valve dysfunction and prompt echocardiographic and clinical assessment.

Primary right ventricular disease, such as right ventricular cardiomyopathy (ARVC), may present with right heart dilatation and requires specialist assessment including MRI.

**Bicuspid aortic valve**

Bicuspid valve is the most common congenital heart abnormality occurring in 1-2% of the population and can present at any age. Associated abnormalities occur in up to 50% of patients, the most commonly encountered being aortic dilatation that occurs as a consequence of structural abnormalities of the aortic wall. Coarctation of the aorta and other left-sided lesions need to be sought using cross-sectional imaging as necessary.

Patients with bicuspid aortic valves may be more prone to aortic dilatation and dissection. A prevalence of dissection of 0.1% per year has been reported. Long-term imaging follow-up is performed to monitor aortic dimensions and identify progressive aortic dilatation before complications develop. Bicuspid aortic valve patients with acute chest pain should be clinically assessed with a low threshold for performing echocardiography and CT. Equally, when a CT reveals thoracic aortic dilatation, the possibility of a bicuspid aortic valve or coarctation should be considered. The Marfan syndrome should be considered in the presence of aortic dilatation, particularly in young people without a bicuspid valve (figure 5).

**Summary**

Congenital heart disease encompasses a complex variety of conditions which require treatment and follow-up in specialist centres. The configuration of such services is currently undergoing immense change with the designation of services and development of clinical networks for congenital heart disease follow-up. While the centralisation of such services may continue, radiologists in non-specialist centres may be asked to undertake imaging, particularly in the emergency setting. For these networks to succeed, improved integration of IT systems will be needed to allow easy access to imaging across centres; this should allow increased support for non-specialist colleagues imaging these patients. It, however, remains important for the general radiologist to have an awareness of the potential presentations of these patients and the specific considerations when imaging them.

We have tried to provide a few examples of clinical scenarios where an understanding of imaging of congenital heart disease may be needed by the general radiologist. However, with such a wide variation of possible anatomy and pathology, patients with congenital heart disease should be assessed on an individual basis and rigid protocols are rarely suitable for all. Specialist colleagues are always available and happy to discuss complex cases.

**References**

FIGURE 1
Aortobronchopulmonary fistula post-coarctation repair. (1a and b) Axial CT images showing a pseudoaneurysm in the distal aortic arch at the site of a previous coarctation repair with surrounding low density haematoma. (1c) Oblique sagittal CT demonstrating the pseudoaneurysm. (1d) Axial CT image on lung settings demonstrating air space opacification within the upper lobe of the left lung adjacent to the descending thoracic aorta. This sign in the presence of a thoracic aortic aneurysm at the site of previous surgery is diagnostic of an aortobronchopulmonary fistula.

FIGURE 2A and B
Multiplanar reformatted CT images in a patient with a Fontan circulation. Images show the connections between the cavae and the right pulmonary artery and a metal stent in the central pulmonary artery. Contrast administered via an upper limb injection results in mixing of opacified blood from the SVC with unopacified blood from the IVC, resulting in partially opacified blood within the central pulmonary artery. This can be misinterpreted as thrombus.

FIGURE 2C
MPR CT image of a patient with a previous arterial switch procedure. There is mixing of opacified blood from the SVC with unopacified blood from the IVC which can cause mixing artefact within the baffle (star) and can give the appearance of thrombus.

FIGURE 2D
MPR of another patient with a previous atrial switch in which a biphasic contrast protocol was used. This approach can overcome the mixing artefact seen in the previous two cases and good, almost equal, contrast opacification is seen in all cardiac chambers.

FIGURE 3
Axial CT image demonstrating a blush of contrast across the inter-atrial septum consistent with an ASD.

FIGURE 4A
Four-chamber axial CT demonstrating a dilated right atrium and ventricle.

FIGURE 4B
CT multiplanar reformats demonstrating a superior sinus venosus defect (star).

FIGURE 4C
CT multiplanar reformat showing anomalous drainage of the right upper pulmonary vein into the superior vena cava (arrow).

FIGURE 4D
3D volume rendered CT of the same patient illustrating the sinus venous atrial septal defect, with the over-riding SVC and the associated anomalous drainage of the right upper pulmonary vein into the SVC.
FIGURE 5A and B
CT multiplanar reformat showing a bicuspid aortic valve with thickened valve leaflets, while closed (a) and open (b).

FIGURE 5C
CT multiplanar reformat of the thickened valve leaflets which are domed on this systolic image.

FIGURE 5D
CT multiplanar reformat of the dilated ascending aorta in the same patient.