Imaging congenital heart disease in the adult

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Introduction
Congenital heart disease (CHD) has a worldwide incidence of approximately 1% and is the most common inborn defect. Advances in diagnosis and treatment have led to increased patient survival with an estimated 90% of affected children reaching adulthood. Soon, it is estimated, there will be more adults than children with CHD.

The increased prevalence in the general population of adults with CHD necessitates follow-up surveillance and frequent medical or surgical intervention.

Imaging techniques
Imaging outlines anatomy and physiology, helps to refine management decisions, evaluates the result of interventions and guides prognosis. However, no single available modality fulfils these roles for all cardiovascular structures and physiological states. As serial imaging is a key factor in the management of the adult patient, the complementary modalities used must be sensitive, reproducible and do least harm.

The most commonly used modality for CHD diagnosis and follow-up is echocardiography. This combines the benefits of being able to accurately assess anatomy and function with portability and ease of use. However, echocardiography is limited by acoustic windows, provides poor images of distal vasculature and is highly operator dependent. This is especially so in adults and patients who have undergone multiple procedures.

For more detailed anatomical and functional information, cardiac catheterisation is still considered an important factor. Although, due to the invasive nature, use of ionising radiation and limited three-dimensional (3D) capabilities, its role is limited (mainly used for patients for whom haemodynamic data is essential). Historically, diagnosis was achieved in the catheter lab with the operating theatre the site of treatment. Now, the diagnostic site is the imaging lab with invasive treatment performed in the catheter lab.

The newest of the imaging modalities is cardiovascular magnetic resonance. CMR imaging plays an essential role in the evaluation and follow-up of adult congenital heart disease, providing safe, high-resolution imaging of some of the most complex anatomies encountered. This tool can be used to acquire accurate functional assessment with high reproducibility, e.g. measurement of blood flow for quantification of left-to-right shunts, regurgitant fractions and pressure gradients. Unimpaired by acoustic windows and capable of tissue characterisation, CMR is devoid of ionising radiation and provides superior 3D spatial resolution to transthoracic echocardiography and superior temporal resolution to computed tomography, making it the ‘gold-standard’ for various cardiac and great vessel imaging indications in CHD.

Cardiovascular CT is equally good for visualising anatomy and has slightly better spatial resolution than CMR. However, due to potentially large doses of ionising radiation (particularly ECG gated studies) and inability to easily quantify cardiac function, the role of cardiovascular CT is limited. Its main uses are imaging vascular rings (providing airway anatomy), for patients who are unable to cooperate with a CMR examination (or CMR contra-indicated) and for imaging small structures such as pulmonary veins and coronary arteries (the latter becoming important, as the patient population ages, their prevalence to develop ischaemia increases).

Strategies for imaging some specific common adult CHD
Approximately 60% of all CHD is diagnosed in babies less than one year old, 30% in children and 10% in adults. Many patients who have undergone total corrective surgery will have few if any haemodynamic residua (e.g. septal defects), thus requiring infrequent evaluation and treatment. However, patients with more complex lesions require more frequent evaluation, medical treatment and consideration for further surgical or catheter-based intervention.

For these adults, late-onset complications need to be identified and managed. Regarding these more complex CHDs, there follows a brief imaging overview for some of the most common (see reference 12 for an overview of CMR findings in the majority of CHDs).

Tetralogy of Fallot (figure 1)
Tetralogy of Fallot is the most common form of cyanotic CHD with an incidence of approximately 420 per million live births. It has four characteristic features, a ventricular septal defect (VSD), an overriding aorta, right ventricular outflow tract (RVOT) infundibular obstruction and right ventricular hypertrophy.

FIGURE 1
Schematic diagrams of unrepaired (A) and completely repaired (B) tetralogy of Fallot. (C) 3D MR angiogram, viewed from anterior, showing dilated right ventricle (RV), RVOT and pulmonary arteries. (D) Four-chamber view, axial, with dilated hypertrophied RV.

The main post-repair complications include pulmonary regurgitation, right ventricular dilatation, residual RVOT obstruction and residual VSD. Doppler echocardiography should be used to assess post-op residual RVOT stenosis when conduit position allows; functional data is often difficult to collect due to the irregularity in ventricular chamber geometry. CMR plays a major role in the post-op imaging assessment, especially with respect to pulmonary regurgitation. Accurate quantification of any regurgitation and good definition of RVOT anatomy is required for the surgical/transcatheter approach to patient management. Phase contrast velocity mapping provides the quantified regurgitant infor-
Coarctation of the aorta (figure 2)

Coarctation of the aorta accounts for 7% of live births with CHD. It comprises a stenosis in the thoracic portion of the aortic arch in the region of the insertion of the arterial duct. In adults, there is often a more definite obstruction with narrowing distal to the left subclavian artery.

The main post-repair complications include repair site aneurysm, aortic valve disease and re-coarctation, which can occur in 3–35% of surgical patients. Although echocardiography can be used to assess aortic arch stenosis severity and size of the transverse arch, the distal segment of the ascending aorta is poorly visualised. Also, aneurism formation and re-stenosis are often difficult to demonstrate. CMR provides information on the exact location of the stenosis, collateral pathways and quantifies flow acceleration. 3D data sets also afford accurate measurements of vessel dimensions. Due to increased incidence of aortic valve disease, its morphology and competency can be evaluated and left ventricular function, size and mass may be assessed to provide significance of any residual obstruction. Importantly, imaging can help decide if percutaneous stenting or surgery are required for treatment. When stenting has been performed, CT imaging is used to assess metallic stent integrity and the potential complication of proximal and distal pseudo-aneurysms.

Previously, cardiac catheterisation had been considered as the imaging modality of choice; now though, with its increased access, CMR is becoming a more common method to assess aortic coarctation.

Transposition of the great arteries

Transposition of the great arteries (TGA) is the second most common cyanotic CHD in the first year of life, with an incidence of 315 per million live births. It is defined as atrioventricular concordance with ventriculoarterial discordance. Thus, the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle.

The surgical repair for TGA will either involve the atrial switch procedure (figure 3), in which the patient still has a systemic right ventricle (subject to the associated pressures), or, preferably, the more modern arterial switch, which leaves the patient with a systemic left ventricle (figure 4).

The main post-repair complications include:
- For the atrial switch procedure, tricuspid valve regurgitation, right ventricular dysfunction, superior/inferior vena cava pathway obstruction and atrial baffle leak.
- For the arterial switch procedure, RVOT/branch pulmonary artery obstruction, neo-aortic valve regurgitation and coronary artery narrowings.

Echocardiography, with the aid of Doppler, can be used in the assessment of most of the post-op complications after the atrial and arterial switch procedures. The exceptions are when imaging the superior/inferior vena cava pathways in atrial switch patients and RVOT/branch pulmonary arteries after the arterial switch procedure, due to their position just behind the sternum. CMR is not limited by the position of these post-sternal vessels and is therefore perfect for imaging this area. Although the atrial switch has been superseded by the arterial switch, there are still significant numbers of atrial switch patients requiring follow-up imaging. Such is the complexity of the vasculature in these adults, 3D imaging is essential, which CMR excels at. These 3D images can then be used to plan orientations for the more conventional imaging techniques. When it is not possible to undertake a CMR examination, CT provides excellent 3D anatomical data and is the modality of choice when imaging metallic implants. Coronary artery obstruction due to re-implantation during the arterial switch procedure is a less common post-op complication; in such cases, cardiac catheterisation is the preferred imaging modality.

Conclusion

Echocardiography still remains the imaging modality of choice for serial follow-up of adult congenital heart disease due mainly to its availability and inexpense. As previously stated, the vast majority of CHDs will be repaired in childhood; as the child grows to adulthood, cardiovascular MR (which easily overcomes the limitations of echocardiography) is playing an increasing role in assessing changes during this maturation period in the young adult, with the majority of patients undergoing CMR several time during their adolescent and early adult years.
CMR has changed our approach to the adult CHD patient, shifting indications for cardiac catheterisation from diagnostic to therapeutic. The need for angiography to evaluate ventricular function, aortic arch stenosis or aneurysm or branch pulmonary artery stenosis is rarely necessary as CMR provides the diagnostic basis for catheter-based interventions. Despite major advances in multi-detector technology, the role of CT remains limited.

Although the prognosis for patients with congenital heart disease is improving, significant cardiovascular problems still develop. Recurrent vascular lesions, residual shunts, ventricular dysfunction, heart failure, pulmonary hypertension, cyanosis, valvular disease and sudden cardiac death are all realistic problems. The main imaging question now is how often a CMR examination should be performed to complement echocardiography.

References