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Review

New Directives in Cardiac Imaging: Imaging the Adult With Congenital Heart Disease

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ABSTRACT

Advances in pediatric surgical and interventional techniques and medical care over the past 50 years have revolutionized the care of children with congenital heart disease. Survival to adulthood is now expected and, as such, there is a growing population of adults which is exceeding the pediatric population with congenital heart disease. Noninvasive cardiac imaging with modalities such as echocardiography, computed tomography, and cardiac magnetic resonance imaging are integral to the care of adults with congenital heart disease. These modalities are used for diagnosis, surveillance for complications late after surgery and catheter-based interventions, and in decision-making for medical, interventional, and surgical therapies. In this review we will discuss noninvasive imaging modalities used to assess congenital cardiac lesions, imaging strategies for select congenital lesions, and comment on the future of cardiac imaging in congenital heart disease.

RÉSUMÉ

Les progrès en matière de chirurgie pédiatrique, de techniques interventionnelles et de soins médicaux au cours des 50 dernières années ont révolutionné les soins aux enfants ayant une cardiopathie congénitale. On s'attend maintenant à une survie jusqu'à l'âge adulte et, à ce titre, la population adulte est supérieure à la population pédiatrique ayant une cardiopathie congénitale. L'imagerie cardiaque non effractive qui utilise des modalités comme l'échocardiographie, la tomodensitométrie et l'imagerie cardiaque par résonance magnétique fait partie intégrante des soins aux adultes ayant une cardiopathie congénitale. Ces modalités sont utilisées pour le diagnostic, la surveillance des complications tardives de la chirurgie et les interventions par cathéter, ainsi que pour la prise de décision quant aux traitements médicaux, interventionnels et chirurgicaux. Dans cette revue, nous discuterons des modalités d'imagerie non effractive utilisées pour évaluer les lésions cardiaques congénitales et des stratégies d'imagerie de certaines lésions congénitales, et émettrons des commentaires sur l'avenir de l'imagerie cardiaque dans l'évaluation de la cardiopathie congénitale.

Advances in pediatric surgical and interventional techniques and medical care have improved survival for children born with congenital heart disease. As a consequence, there are a growing number of adults with congenital heart disease. Cardiac imaging plays an integral role in the management of adults with congenital heart disease aiding with diagnoses, surveillance for complications late after surgeries and catheter-based interventions, and in the decision-making for interventional, surgical, and medical therapies.

Knowledge and understanding of congenital cardiac anatomy, terminology, pathophysiology, and surgical and/or interventional procedures are fundamental in adult congenital heart disease (ACHD) imaging. When performing and interpreting complex congenital imaging studies, a segmental approach to the cardiac anatomy is used to ensure that important pathology is not missed.[1] Standard views might be inadequate and atypical views might be required. Complementary multimodality imaging is often needed for comprehensive assessment of these patients because of the complex nature of the cardiac anatomy and pathophysiology. Longitudinal imaging data are also very important when following adults with congenital cardiac lesions.

Cardiac imaging should be performed at centres with technicians and readers with expertise in ACHD.[2-5] Even adults with simple congenital cardiac lesions might benefit from an initial assessment at a specialized centre because simple lesions have associated defects that can be overlooked.

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Information pertaining to ACHD centres in Canada can be found at www.cachnet.org. General cardiac centres are encouraged to collaborate with specialized ACHD centres when imaging adults with complex congenital cardiac lesions.

In this review we will discuss the noninvasive imaging modalities commonly used to assess congenital cardiac lesions, discuss imaging strategies for select congenital lesions, and comment on the future of imaging in ACHD.

Imaging Modalities

Echocardiography

Echocardiography is the primary modality used for cardiac imaging in ACHD. It is safe, portable, readily available, and is easy to perform. The main limitation in the adult patient population, specifically in those with previous cardiac surgery, relates to poor acoustic windows.

Understanding cardiac structure is fundamental when imaging adults with congenital cardiac lesions and cardiac anatomy is often well visualized with echocardiography. Colour Doppler can detect small intracardiac shunts which might not be detected using other imaging modalities. Saline contrast can be used to help identify small intracardiac shunts, particularly at the atrial level. Doppler echocardiography assesses diastolic function of the ventricles and pressure gradients between the chambers, between the chambers and the vessels, and within the pulmonary arteries.

Determination of left ventricular dimensions and global systolic function are readily performed with transthoracic echocardiography (TTE). However, echocardiographic assessment of right ventricle (RV) systolic size and function is more difficult, in part because of the shape of the RV. Indirect measures of right ventricular function such as fractional area change,[6] tricuspid annulus planar excursion,[7] velocity of tricuspid annular systolic motion,[8] myocardial performance index,[9,10] and isovolumic acceleration[11,12] can be helpful. Echocardiographic methods to assess RV dimensions and estimates of RV volumes have been proposed.[13] Three-dimensional (3D) echocardiography aids the assessment of RV size and function as it is not limited by geometric assumptions or by foreshortened views. It remains limited, however, by its poorer spatial and temporal resolution in comparison with 2-dimensional (2D) echocardiography and is not yet routinely used in clinical practice. Systems have been developed that use 2D images to create a 3D model of the RV.[14] At present, cardiac magnetic resonance (CMR) imaging is considered the most accurate method to measure RV size and systolic function. RV volumes and ejection fraction tend to be underestimated in comparison with CMR imaging.[15] Guidelines for the echocardiographic assessment of the RV in adults are published elsewhere.[16]

New quantitative measures of ventricular function using tissue Doppler and speckle tracking are helping to improve our understanding of ventricular mechanics. For example, in repaired tetralogy of Fallot (TOF), strain imaging has demonstrated abnormalities in regional systolic function, diastolic dysfunction, and the presence of dyssynchrony.[17-20] In congenitally corrected transposition of the great arteries, abnormalities in strain patterns are observed in the subaortic RV.[21,22] Though these techniques have been used for research, they are not currently use in routine clinical practice.

Transesophageal echocardiography (TEE) is most commonly used for diagnosis of atrial septal defects (ASDs), for assessment of the ASD anatomy, and to provide device sizing and placement guidance during percutaneous ASD closure. Intracardiac echocardiography can be used as an alternative during device placement.[23] TEE is also used to assess valve structure and function, patency of baffles, or baffle leaks in patients who have had atrial switch surgery, and to examine for intracardiac thrombus in adults at risk such as those with Fontan circulation.

Use of stress echocardiography in the assessment of adults with congenital heart disease is limited. It can be useful assessing adults who have had surgeries that manipulate the coronary arteries, such as individuals who have undergone an arterial switch repair.

CMR imaging

There is a growing role for CMR imaging in the field of ACHD. CMR, like echocardiography, is relatively safe and does not expose the patient to nonionizing radiation. It can be a difficult imaging modality for patients with claustrophobia. The contrast agent used, gadolinium, is avoided in patients with a glomerular filtration rate less than 50 mL/min/kg because of the risk of nephrogenic systemic fibrosis. CMR is contraindicated in patients with standard pacemakers or internal cardiac defibrillators. Magnetic resonance imaging-compatible devices are being increasingly used and though some centres perform cardiac imaging in individuals with these new devices, at present these devices are not licensed for use in cardiac imaging.[24]

CMR imaging is a useful tool to define anatomy in adults with complex congenital structural lesions. The heart can be imaged in any plane allowing for thorough assessment of anatomy of the heart, great vessels, and surrounding thoracic structures. This is especially useful in the assessment of baffles, conduits, and Fontan connections which might be poorly visualized with echocardiography.[25,26] CMR is the reference standard method of assessing ventricular size and systolic function; particularly for the RV.[27] There are a number of newer CMR techniques available to assess strain or strain rate without the limitations of acoustic windows often encountered in echocardiography. Myocardial tagging can quantify myocardial strain and has the potential to assess regional contractile function in a variety of heart lesions.

Phase contrast imaging allows for accurate assessment of regurgitant fractions, regurgitant volumes, and shunt calculations. Fibrosis and scar can be visualized with the use of the contrast agent gadolinium. Stress CMR might be useful in the assessment of coronary artery disease, but more frequently in the assessment of myocardial ischemia secondary to primary or coronary anomalies, ostial stenosis after congenital heart disease surgery, or right coronary artery caused by a conduit.

Cardiac computed tomography

Advances in technology now allow for accurate assessment of cardiac structure and function using computed tomography (CT). Contrast allergy can limit use, but allergies are much less common with the use of newer contrast agents. Though current CT scans have lower temporal resolution than CMR, they have superior 3D spatial resolution and the advantage of much shorter acquisition times than CMR.[28] This allows for imaging

of the epicardial coronary arteries and the assessment of their relation to other structures such as conduits.[29] Assessment of ventricular size and function can be performed. Ventricular volumes might be overestimated but overall have a good correlation with CMR.[30] Radiation limits the repeated use of CT in the young ACHD population, however, improved technology and protocols can substantially limit the amount of radiation.[31] In addition, there is a risk of contrast nephropathy especially in individuals with pre-existing renal insufficiency.

Specific Congenital Cardiac Lesions

ASDs

There are several types of ASD. The most common type is the secundum ASD located in the region of the fossa ovalis. Less common are the primum ASD, the unroofed coronary sinus, and the sinus venosus defect. An intracardiac shunt at the atrial level results in volume overload of the RV. Many patients only develop symptoms in adulthood including functional decline, atrial arrhythmias, stroke from paradoxical emboli, right heart failure, and less commonly, pulmonary arterial hypertension. When indicated, ASDs can be closed percutaneously or surgically.[3,4,32]

Echocardiography is an excellent tool to assess an ASD.[33] TTE can detect most secundum ASDs with the exception of very small ASDs.[34] Saline contrast can help to identify small intracardiac shunts. TEE is useful for measuring the maximal diameter of the defect and sizing of percutaneous devices. TEE allows for visualization of the rims of the ASD and their relation to important structures (tricuspid valve, ostia of the coronary sinus, vena cava). TEE also assesses the connections of the pulmonary veins to the left atrium and aids in the detection of anomalous pulmonary venous connection which is associated with ASDs. With 3D echocardiography advances, the shape of the ASD and the change in the size of the ASD during the cardiac cycle can be visualized. This information might be of value when sizing percutaneous closure devices (Fig. 1).[35] Although initial closures of secundum ASDs in the cardiac catheterization laboratory were aided by TEE guidance, some centres now use intracardiac echocardiography.[36]

CMR and CT can be useful in assessing sinus venosus defects. These defects are associated with anomalous pulmonary venous return and this is best visualized with CMR or CT. CMR and CT can also be used to assess the RV size and function and the pulmonary veins when echocardiographic images are suboptimal. CMR can quantify shunt flow by directly measuring the pulmonary and systemic blood flow,[37] although typically the hemodynamic significance of a shunt is characterized by its effect on the RV. Hemodynamically significant shunts are associated with right ventricular enlargement.

Ventricular septal defects

The anatomic classification of ventricular septal defects (VSDs) is determined by their location within the septum (membranous, muscular, inlet, or outlet). A small restrictive VSD is associated with a significant systolic gradient across the VSD and do not typically result in increased pulmonary blood flow or left ventricular volume overload. Medium or larger sized VSDs can result in left ventricular volume overload, left ventricular dysfunction, and increase pulmonary artery systolic pressures. Unrepaired large VSDs are occasionally seen in adults and can be associated with pulmonary hypertension and shunt reversal (Eisenmenger syndrome). Cardiac complications that can occur in adulthood include heart failure, arrhythmias, infective endocarditis, and, in the case of a perimembranous or outlet VSD, aortic regurgitation because of prolapse of the right and/or noncoronary cusp.[38] When indicated, VSDs can be closed surgically and in some instances, percutaneously.[3,4,32]

TTE with colour Doppler remains the most sensitive means of detecting small or multiple defects.[39] Transseptal peak instantaneous systolic gradient can be assessed with continuous wave Doppler[40] (Fig. 2). There might be no increased shunt flow velocities with large nonrestrictive VSDs. The hemodynamic significance of the VSD shunt is often determined by its effect on the left ventricular size and function.

Pulmonary artery systolic pressures are easily assessed using echocardiography. Aortic valve prolapse and associated regurgitation, right ventricular outflow tract (RVOT) obstruction secondary to infundibular hypertrophy or muscle bundles, or partial closure of VSDs by tricuspid valve tissue are best detected echocardiographically. CMR has a limited role to play in individuals with a VSD because TTE can provide the relevant information in most cases. It can define

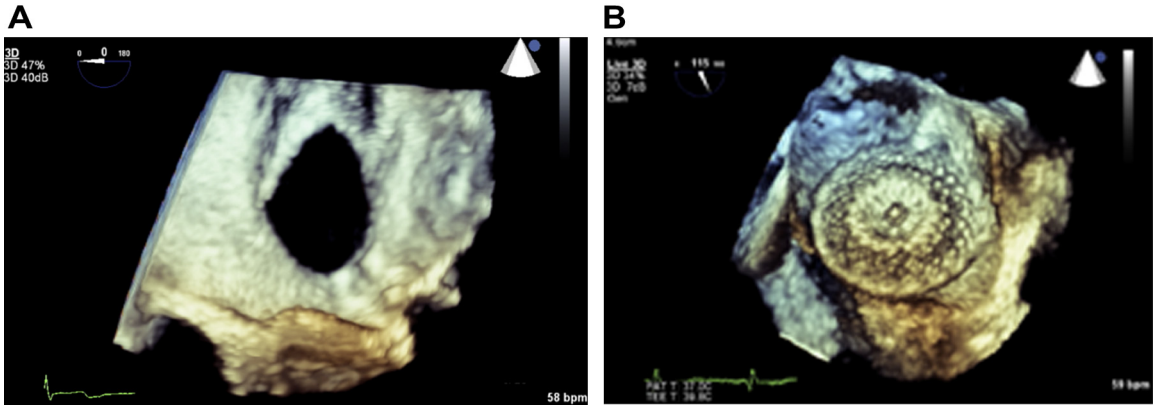

Figure 1. Transesophageal echocardiogram 3-D images of a secundum atrial septal defect. (**A**) A large secundum atrial septal defect. (**B**) An Amplatzer closure device.

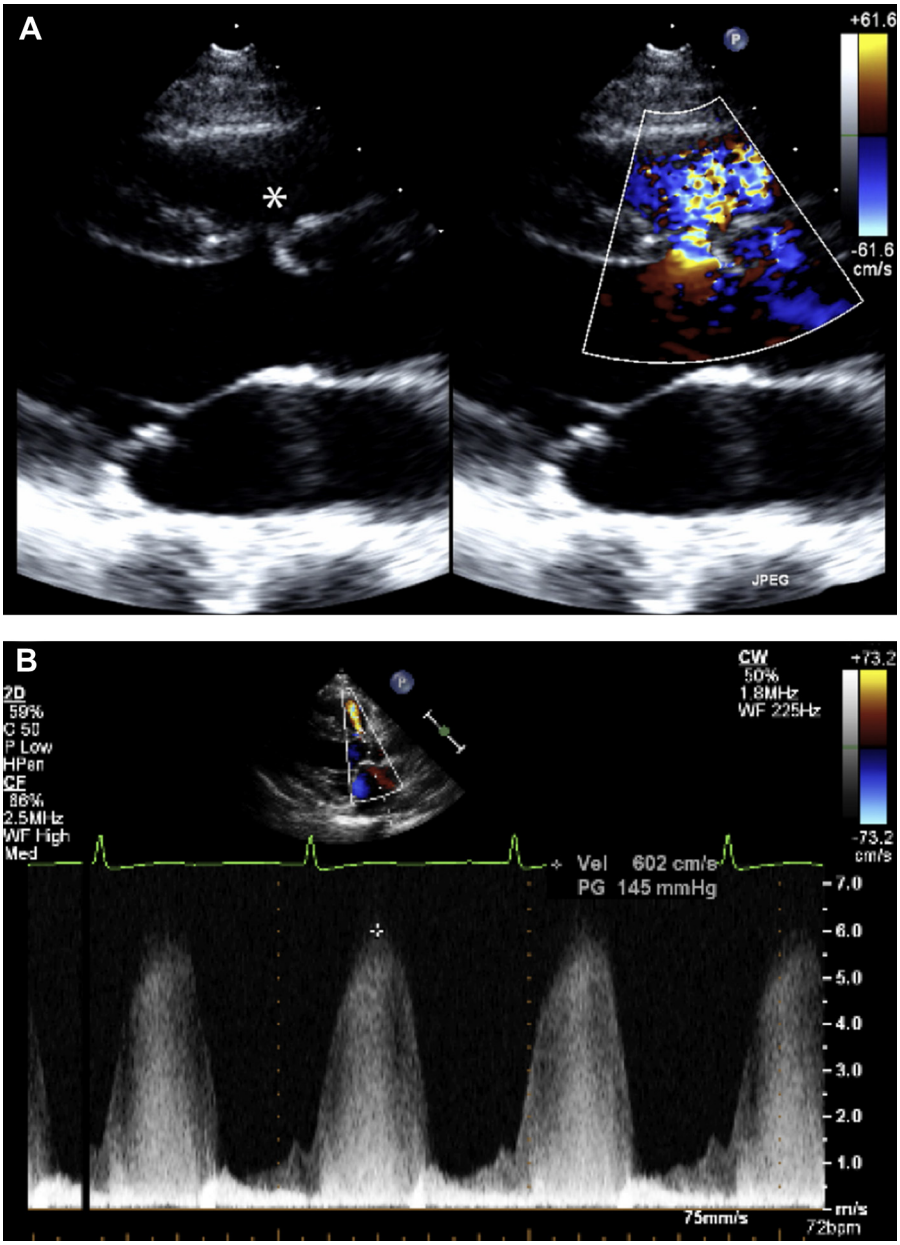

Figure 2. Transthoracic parasternal of a perimembranous ventricular septal defect. (**A**) Parasternal long axis view (end-systole) of a perimembranous ventricular septal defect (**asterisk**) with colour Doppler flow across the ventricular septal defect. (**B**) A peak systolic pressure gradient of 145 mm Hg across a restrictive perimembranous ventricular septal defect.

the site and size of a VSD, but is more commonly used to assess the hemodynamic consequences of the lesion, the size, and systolic function of the left ventricle. The pulmonary to systemic flow ratio can be calculated using CMR.

TOF

TOF is the most common cyanotic heart lesion. Older adults often have had palliative shunts (Blalock-Taussig shunts, Potts shunts, Waterston shunts) before their intracardiac repair. Most adults will have undergone an intracardiac repair in childhood. The repair comprises a VSD patch closure, infundibular resection, and in some cases, transannular patching or pulmonary arterioplasties. The types of RVOT repairs have been modified over the years. Transannular patches were originally used; however, they resulted in severe pulmonary regurgitation and have been replaced, when possible, by valve sparing operations. Adults with TOF are at risk for heart failure, atrial and ventricular arrhythmias, and sudden cardiac death.

The degree of pulmonary regurgitation and its effect on the RV are important determinants of outcome in this population. Echocardiography can provide a qualitative assessment of pulmonary regurgitation, whereas CMR, using either phase contrast measurements or stroke volume differential, can provide estimates of regurgitant fraction and regurgitant volume.[41] Use of CMR allows visualization of the pulmonary arteries and determination as to whether there is differential

regurgitation from the right and left pulmonary arteries.[42] Accurate assessment of the RV is important for the assessment of adults with TOF. There are a number of methods of assessing RV size and function in adults with TOF. Though echocardiographic techniques are useful for serial follow-up, CMR allows for the most accurate assessment of the RV size and function[43] (Fig. 3). For adults with severe pulmonary regurgitation, RV size is an important variable when deciding on timing of pulmonary valve replacement. When the RV end diastolic volume of 150-170 mL/m^2 is reached, the RV does not remodel to normal size after pulmonary valve implant.[44] The unique remodelling of the volume-overloaded RV can be seen with 3D CMR modelling. Using tagging techniques, areas of the RV that do not contract normally, such as an aneurysmal RVOT patch, can be identified.[45] The burden of fibrosis can be determined using CMR and might have prognostic importance. Three-dimensional echocardiography can also be used to assess the size of the RV though it is dependent on adequate acoustic windows. CT is an alternative in patients with a pacemaker or AICD. A restrictive right ventricular filling pattern with antegrade blood flow in the pulmonary artery is sometimes noted in this population and is prognostically important.[46]

An additional strength of CMR and CT is the ability to accurately assess the size of the RVOT and the main and branch pulmonary arteries. Imaging of the right ventricular outflow track is particularly important when implanting a percutaneous pulmonary valve. Using a combination of cine imaging and contrast angiography, the dimensions of the outflow tract, the main pulmonary artery, and the course of the proximal coronary arteries in relation to the RVOT can also be determined before valve placement.[5]

Coarctation of the aorta

Coarctation of the aorta is a narrowing of the aortic isthmus with or without hypoplasia of the aortic arch. It can be associated with other congenital lesions, the most common of which is the bicuspid aortic valve. Most adults with significant coarctation of the aorta will have undergone interventions as children. A variety of repairs have been performed over the years and include left subclavian flap repair, patch aortoplasty, end-to-end anastamosis, and jump interposition grafts. Some individuals might have undergone angioplasty or stent placement. Imaging surveillance is required to assess for long-term complications such as hypertension and associated left ventricular hypertrophy, recoarctation or aneurysms at the site of repair, aortic valve lesions, and proximal thoracic aortic dilation in patients with bicuspid aortic valves and premature coronary artery disease.

Although use of echocardiography might allow visualization of the coarcation site, CMR and CT are better imaging options because they allow for visualization of the entire thoracic aorta and help provide information on the geometry of the arch, which has been shown to influence vascular remodelling after aortic arch repair.[47] Doppler is used to assess pressure gradients in the descending thoracic aorta. The Doppler velocity trace shows a typical high systolic peak velocity with accompanying diastolic antegrade flow (Fig. 4A). The diastolic antegrade flow velocities correlate with the severity of coarctation.[48] However, the severity of the coarctation might be underestimated if there is significant collateral flow. When estimating the peak systolic gradient at the site of coarctation and/or recoarctation, it is important to consider the velocity proximal to the coarctation site and to remember that a long segment of stenosis will make the Bernoulli equation invalid.[49]

CMR and CT are the preferred means of assessing for coarctation and aneurysm formation because they allow for complete visualization of the aorta (Fig. 4B).[50,51] When a stent has been implanted, CT is often superior to CMR which is limited by artifact from the stent. New imaging techniques will allow assessment of flow within the vessels and the wall stresses. The CMR technique, 4-dimensional velocity mapping, is a phase contrast imaging method which provides information on complex flow patterns. Distorted flow patterns in the descending aorta with marked helical and vortical flow are seen in regions of poststenotic dilatation. In individuals with bicuspid aortic valves, abnormal flow patterns in the

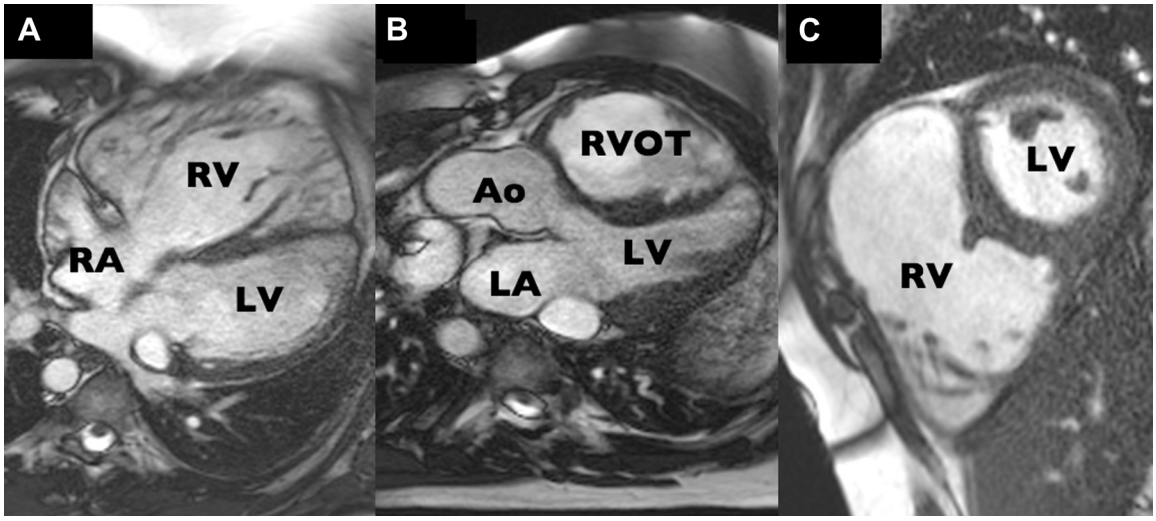

Figure 3. Cardiac magnetic resonance images in an adult with repaired tetralogy of Fallot. (**A**) The right ventricle is dilated secondary to significant pulmonic regurgitation. (**B**) A 3-chamber view showing an aneurysmal right ventricular outflow tract. (**C**) A short axis view of the heart showing the dilated right ventricle. Ao, aorta; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; RVOT, right ventricular outflow tract.

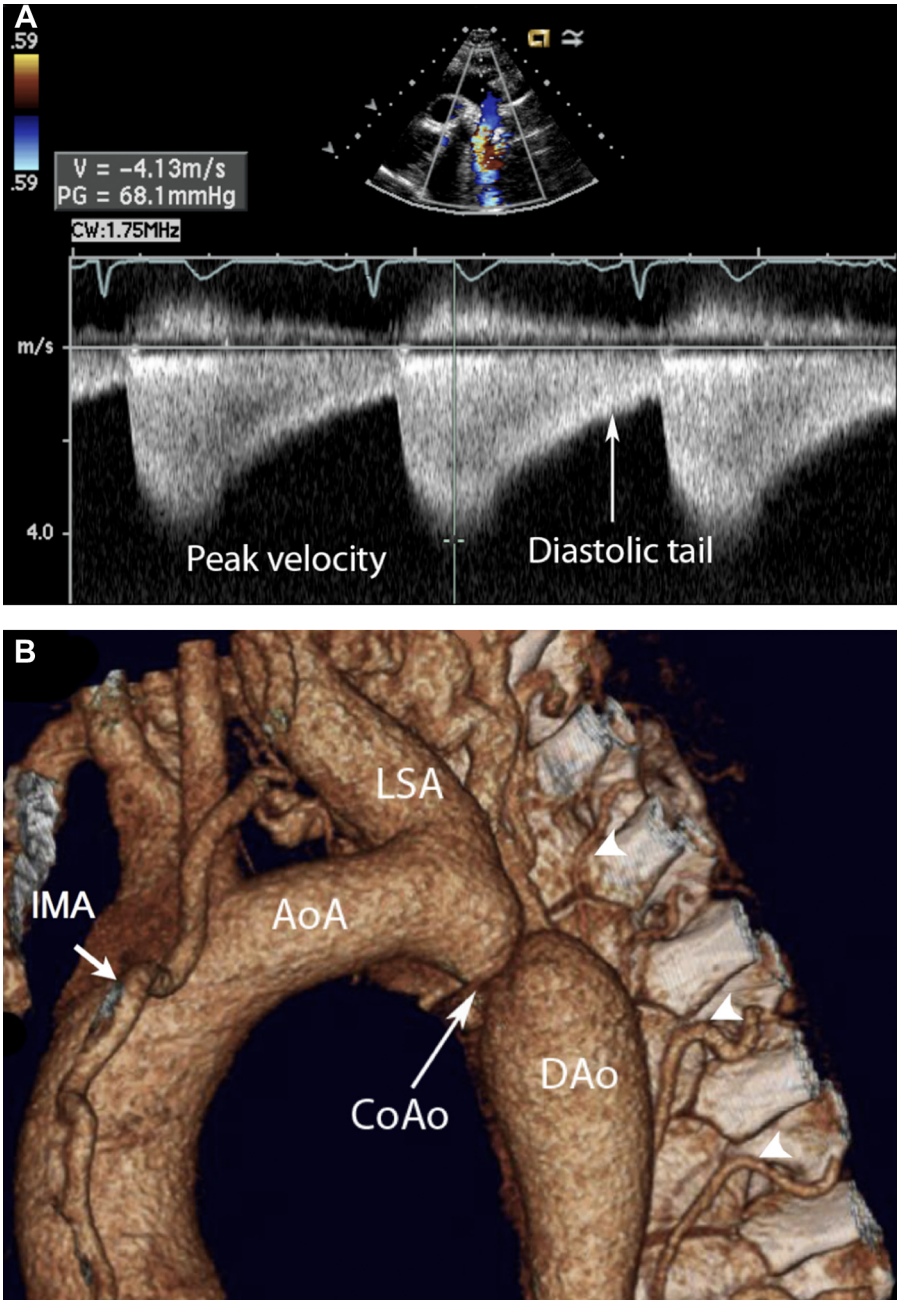

Figure 4. Cardiac computed tomography and transthoracic echocardiography in an adult with coarctation of the aorta. (**A**) A Doppler velocity tracing obtained from the suprasternal view. The tracing shows an increased peak velocity with diastolic antegrade flow. (**B**) A multiplanar cardiac CT image of the aortic arch demonstrating a discrete narrowing distal to the left subclavian artery (**arrow**). There is an aneurysmal segment of aorta distal to the site of coarctation. Prominent internal mammary artery and collaterals (**arrow heads**) are also seen. AoA, aortic arch; CoAo, coarctation of aorta; DAo, descending aorta; IMA, internal mammary artery; LSA, left subclavian artery.

ascending aorta with eccentric jets and abnormal helical systolic flow patterns are seen.[52] In the future, such techniques might be of clinical use in identifying individuals who are at risk of developing aneurysms.

Ebstein anomaly

Ebstein anomaly is a rare congenital anomaly of the tricuspid valve and RV. It results from failure of delamination of the tricuspid valve leaflets from the endocardium. Echocardiography allows assessment of the morphology of the tricuspid valve which is characterized by apical displacement of the septal leaflet, dysplasia of the septal and/or the posterior leaflet, elongation of the anterior leaflet, a dilated right atrium, and an abnormal "functional" RV (Fig. 5A). A wide variation in severity of Ebstein anomaly is seen from quite mild cases that are noted incidentally on echocardiography to severe cases that require the Fontan palliation. Tricuspid regurgitation is

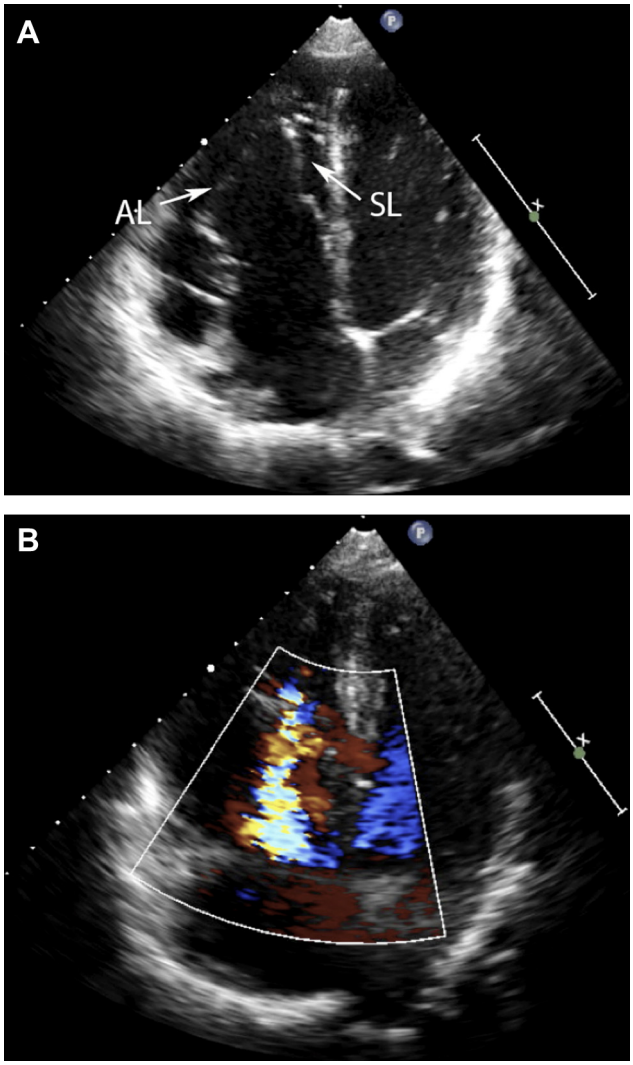

Figure 5. A transthoracic echocardiogram 4-chamber view showing Ebstein anomaly. (**A**) Apical displacement of the tricuspid valve, a large anterior leaflet with tethering of the leaflet, and a dilated right atrium that includes the atrialized portion of the right ventricle. (**B**) Colour Doppler of tricuspid regurgitation. The origin of the tricuspid regurgitation jet is near the right ventricular apex. AL, anterior leaflet; SL, septal leaflet.

common (Fig. 5B). Potential complications in adults with Ebstein anomaly include atrial arrhythmias, tricuspid regurgitation with RV volume overload, right heart failure, RVOT obstruction, and paradoxical embolism when a patent foramen ovale or ASD is present. Tricuspid valve repair or replacement might be necessary.

TTE is important in establishing a diagnosis allowing for assessment of the tricuspid valve anatomy and function. Evaluation of the tricuspid valve includes measurement of the apical displacement of the septal or posterior leaflets (> 8 mm/m^2 in Ebstein anomaly) and mobility and size of the anterior leaflet. The degree to which the anterior leaflet is tethered to the RV free wall is important when attempting tricuspid valve repair. The size and systolic function of the "functional" RV is an important prognostic marker. Echocardiographic methods to evaluate the "functional" RV have been proposed.[53] The right atrium is typically enlarged and includes the atrialized portion of the RV. Colour Doppler can be used to investigate the associated resulting regurgitation, or rarely, stenosis. Patent forman ovale and ASD are commonly observed with shunt reversal (right-to-left shunting) being noted in more severe cases. TEE can aid in the assessment of the valve when images are suboptimal with TTE but is particularly useful intraoperatively when performing valve repairs. Recent advances in 3D technology allow for better assessment of the valves and might ultimately improve our ability for repair these complex valve lesions.[54] CMR has an important role to play in accurately assessing right heart volumes and function, especially when surgery is being considered.[55]

Transposition of the great arteries: atrial switch operation (Mustard and Senning operation)

Older adults with transposition of the great arteries might have an atrial switch procedure. Using surgically created baffles, the Mustard and the Senning operations redirect blood flow at the atrial level. This results in a morphologic subaortic RV and a tricuspid valve in the subaortic position. The most severe common complication observed in adults with atrial switch repairs is failure of the subaortic RV, the prevalence of which increases with time. Tricuspid regurgitation develops and progresses, often as subaortic RV function declines. Intra-atrial baffles can become obstructed or can leak. Arrhythmias are common with interatrial reentrant tachycardia being the most common, but ventricular arrhthymias can occur. Sinus node dysfunction is also common.

Echocardiographic assessment of the subaortic RV can be difficult because of the shape of the ventricle. Echocardiographic methods that have been proposed to assess the subaortic RV include tricuspid annulus planar excursion, fractional area change, myocardial perfusion index, and the

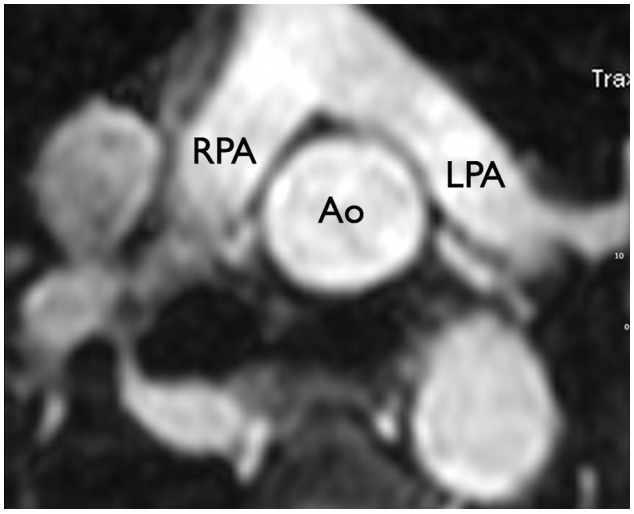

Figure 6. An axial cardiac magnetic resonance image of a patient with an arterial switch operation (Jatene operation). Characteristic of this repair, there is draping of the left and right pulmonary arteries over the ascending aorta. Ao, aorta; LPA, left pulmonary artery; RPA, right pulmonary artery.

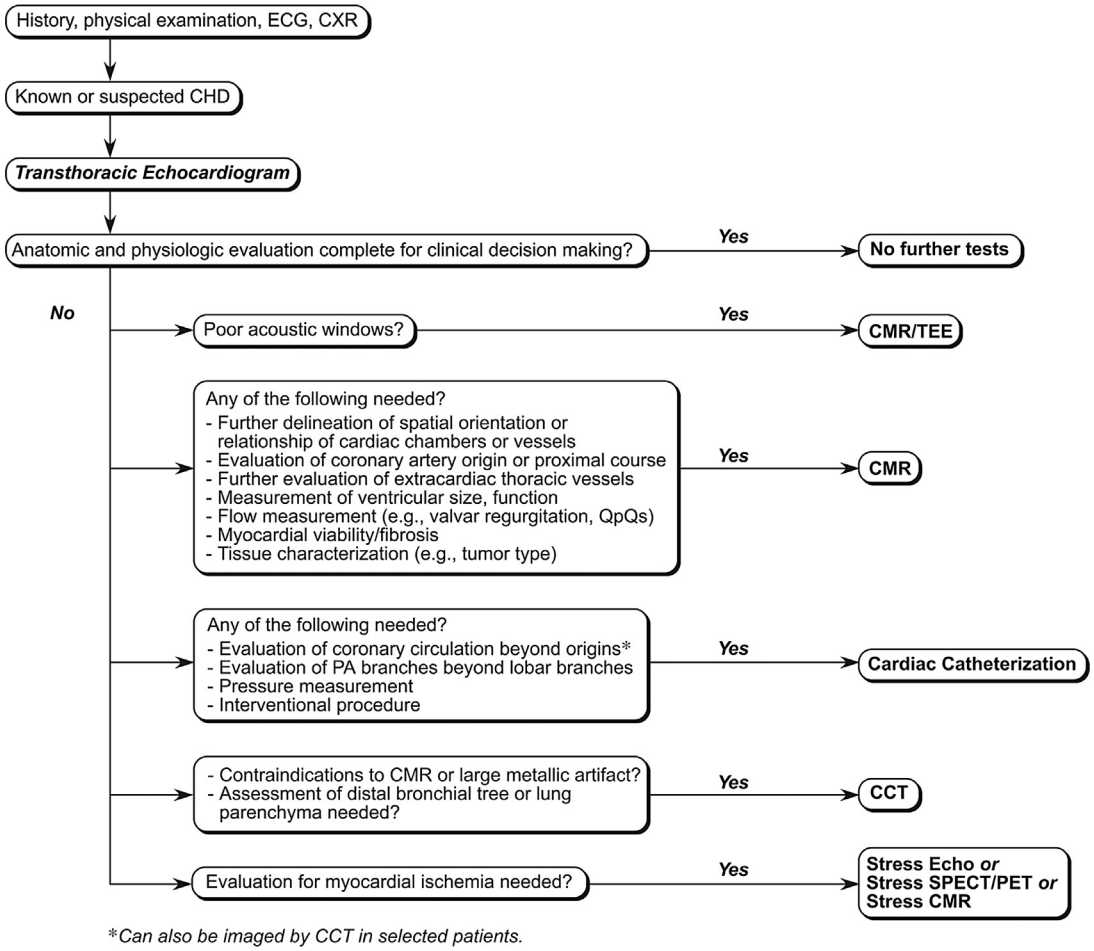

Figure 7. Algorithm for the evaluation of patients with known or suspected congenital heart disease. CCT, cardiac computed tomography; CHD, congenital heart disease; CMR, cardiac magnetic resonance; CXR, chest x-ray; ECG, electrocardiogram; Echo, echocardiography; PA, pulmonary artery; PET, positron emission tomography; QpQs, pulmonary-systemic flow ratio; SPECT, single-photon emission computed tomography; TEE, transesophageal echocardiography. Reproduced from Prakash et al.[63] with permission from Wolters Kluwer Health.

rate of pressure rise at the onset of ventricular contraction (Dp/dt) (if there is tricuspid regurgitation).[10,22] There has been recent interest in the use of tissue Doppler and strain rate imaging in this group of patients.[56,57] The subaortic RV contraction pattern is noted to adapt and to shift from its normal longitudinal shortening to a circumferential shortening pattern, but with an abnormal torsion.[58] CMR and CT are usually superior in determining subaortic RV size and systolic function. It is important to note that the subpulmonic left ventricle has a characteristic D shape, a normal shaped left ventricle might be seen with a baffle leak, pulmonary hypertension, or obstruction within the RVOT.

In some cases, 3D TTE might have an additive benefit to normal 2D TTE in the assessment of the tricuspid valve and the atrial baffles.[59] Echocardiography with colour Doppler can help to identify baffle leaks; this can be further confirmed or excluded by the injection of agitated saline contrast. Doppler assessment can be performed to determine the flow velocity in the baffles, but assessment for baffle stenosis is often inadequate. CMR and CT are better modalities to assess the patency of the intracardiac baffles and ensure that there is no significant baffle stenosis.[60]

Transposition of the great arteries: arterial switch operation (Jatene operation)

In 1975, Jatene first described the successful anatomic correction of the great arteries. This procedure is now the standard method of repair for children with transposition of the great arteries. The aorta and pulmonary artery are transected above the sinuses, the coronary arteries are detached from the aorta with a surrounding button of aortic wall, and then sutured into the neoaorta. The pulmonary trunk is moved forward anterior to the aorta whereupon the switched great arteries are sutured into place. Long-term complications seen in adults who have undergone an arterial switch operation include dilatation of the neoaortic root, neoaortic valve regurgitation, coronary ostial stenosis, supravalvar pulmonary stenosis at the suture sites, and stenosis of the pulmonary arteries.

Echocardiography in the adult can assess the neoaortic root but is limited in assessing the pulmonary arteries because of their anterior position. Doppler gradients across the pulmonary artery will identify stenosis at suture sites. Colour Doppler can be used to identify any significant valve regurgitation. CMR and

CT, however, are ideal imaging modalities to visualize the pulmonary arteries because there are no restrictions to field of view (Fig. 6). CMR and CT can allow assessment of the proximal coronary arteries[61,62]; CT has the advantage of higher spatial resolution to examine the coronary arteries. Stress CMR and stress echocardiography allow assessment of myocardial perfusion without radiation exposure.

Future Directions in Cardiac Imaging

Cardiac imaging is very important in ACHD. Noninvasive cardiac imaging helps to identify important residuae and sequelae, determine when surgery or therapy are indicated, and determine long-term prognosis. Physicians have a number of complementary imaging modalities available and typically a number of modalities are required for complete assessment of the patient (Fig. 7). TTE remains the most common imaging modality. For adults with lesions such as TOF, periodic CMR is recommended to accurately monitor right ventricular size. CMR and cardiac CT are also used to address specific questions. It is important for the technicians performing the studies and the physicians interpreting the studies to be aware of the strengths and limitations of the various imaging modalities. Inappropriate imaging studies occur when physicians are not familiar with congenital heart disease and/or imaging modalities.

There will likely be a growing role for 3D imaging in the future. For instance, 3D echocardiography might allow for better understanding of underlying ASDs or cardiac valves. CMR and CT have been fundamental in advancing our understanding of complex congenital cardiac lesions including the anatomy of the pulmonary vascular bed. CMR provides additional functional information about cardiac hemodynamic characteristics and blood flow. With CMR-derived 4-dimensional flow analysis, blood flow patterns can be assessed within the heart and vessels. Because ventricular structure is often abnormal in the setting of complex congenital heart disease, new echocardiographic techniques, such as myocardial acceleration during isovolumic contraction, might be helpful in assessing ventricular systolic function. The importance of regional ventricular function is being increasingly recognized in ACHD. New imaging techniques such as echocardiographic strain imaging and CMR tagging are providing insights into regional ventricular function. CMR imaging with gadolinium provides information about the burden of fibrosis which might be prognostically important. Stress imaging using echocardiography or CMR is being increasingly used for specific cases such as after surgeries that manipulate the coronary arteries. Such new imaging technologies have the potential to change clinical practice by improving our understanding of the pathophysiology of the lesions and identifying individuals at increased risk. Future developments in cardiac imaging in ACHD will hopefully lead to improvements in the care of ACHD patients in the long-term.

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Disclosures

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