Multimodality Noninvasive Imaging for Assessment of Congenital Heart Disease

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Major advances in the field of pediatric cardiology and cardiac surgery over the last several decades have led to a dramatic improvement in survival rates for most forms of congenital heart disease (CHD). For example, hypoplastic left heart syndrome, a previously lethal defect, now has early survival rates up to 90% at major centers. These improved outcomes have produced a growing population of survivors with complex CHD who are now reaching adulthood (Figure 1). During this period, improvements in surgical and medical treatments have been accompanied by developments in diagnostic modalities. Echocardiography has replaced catheterization as the primary diagnostic modality, and it is now uncommon for newborn infants to undergo catheterization for purely diagnostic purposes. Although echocardiography remains the bedrock of noninvasive cardiac imaging, the array of diagnostic modalities and techniques available continue to grow and this has spawned the specialty of “noninvasive cardiac imaging” and the need for the “cardiac imager” to be adept in all the different modalities.

Modalities

Echocardiography, cardiac magnetic resonance (CMR), and cardiac computed tomography (CCT) are the primary modalities used for noninvasive cardiac imaging in patients with CHD. Nuclear scintigraphy is used in selected circumstances. The Table summarizes the strengths and weaknesses of each modality. Figure 2 shows temporal trends in utilization for the various noninvasive cardiac imaging techniques at our center. It is clear that echocardiography is the most frequently used modality for initial assessment and follow-up of most patients with CHD, with CMR and CCT playing complementary but much smaller roles. Diagnostic cardiac catheterization is used sparingly but continues to be used in selected circumstances where detailed hemodynamic assessment is important. Based on trends at our center, the marked increase in use of noninvasive imaging over the past nearly 3 decades is driven predominantly by a growing population of patients with repaired or palliated heart disease and is not related to increased incidence of CHD or increased frequency of testing in individual patients. Figure 1 shows that a progressively increasing proportion of echocardiograms are now performed on adult survivors of CHD.

Challenges

An ideal modality used for noninvasive imaging of CHD should be able to delineate all aspects of the anatomy, including abnormalities of cardiac structure as well as those involving extracardiac vessels, evaluate physiological consequences of CHD such as measurement of blood flow and pressure gradients across stenotic valves or blood vessels, be cost-effective and portable, not cause excessive discomfort and morbidity, and not expose patients to harmful effects of ionizing radiation. No single modality satisfies all these requirements because noninvasive imaging of CHD poses several unique challenges. Patients range in size from a 200-g fetus to an adult weighing more than 120 kg. Each extreme poses technical difficulties specific to the imaging modality used. For example, in imaging a fetus or newborn infant, a technique with high spatial resolution such as echocardiography performs well, especially because ultrasound windows are usually good. However, the diagnostic yield of echocardiography is often limited in adults, especially after cardiac operations because of diminished ultrasound windows. CMR and CCT are well suited to adults because acoustic windows do not limit image quality but face significant technical challenges in imaging of a fetus or newborn.

Echocardiography

Since the 1980s, echocardiography has been the noninvasive imaging modality of choice for diagnosis and follow-up of children and adults with CHD and has been shown to be reliable in the diagnosis of most forms of CHD, with major diagnostic errors occurring in only 0.2% to 2% infants. New echocardiographic techniques developed in recent years, including real-time 3D echocardiography, strain/strain rate imaging, and speckle tracking, have greatly expanded the capabilities of cardiac ultrasound to evaluate anatomy and physiology of CHD.

CMR

Technical advances during the last decade have brought CMR into the mainstream of noninvasive cardiac imaging. Well-established clinical applications of CMR in patients with CHD include anatomic assessment of cardiovascular anomalies before and after surgery, quantification of biventricular function, magnetic resonance angiography (MRA), measure-
ment of systemic and pulmonary blood flow, quantification of valve regurgitation, identification of myocardial ischemia and fibrosis, and tissue characterization.

**CCT**

The role of CCT in evaluating the thoracic vasculature and the tracheo-bronchial tree is now well established.4,5 Recent advances in multidetector technology have led to improved spatial and temporal resolutions allowing coronary artery imaging and gated cine imaging to evaluate ventricular function. CT scanners are also readily available and easy to use compared with CMR. A major disadvantage of CCT, especially in children, is the exposure to ionizing radiation. This drawback is compounded when multiple follow-up studies are necessary.

**Nuclear Scintigraphy**

These techniques use radioisotope-labeled compounds to quantify ventricular function, blood flow, myocardial perfusion, and metabolism.6 Ventricular volumes, ejection fraction, cardiac output, and regurgitant fraction can be calculated using multiple gated acquisitions obtained after equilibration of 99mTc-labeled red blood cells or albumin. Reversible myocardial ischemia can be identified by assessing myocardial perfusion under stress based on uptake of 99mTc or 201Tl measured using single-photon emission computed tomography (Figure 3). Positron emission tomography can be used to evaluate myocardial viability and metabolism. Lung perfusion can be quantified using 99mTc-labeled macroaggregated albumin. This technique is commonly used to determine differential flow to each lung, which can be helpful in determining the physiological significance of pulmonary artery or vein stenosis. However, nuclear scintigraphy plays a limited role in the pediatric age group due to risks associated with exposure to ionizing radiation and its relatively low spatial resolution.

**Radiation Risks**

Among noninvasive modalities for cardiac imaging, CCT and nuclear scintigraphy involve exposure to ionizing radiation. A growing body of literature has shown that exposure to even low doses of ionizing radiation increases the lifetime risk of developing cancer.7 Importantly, children are at much greater risk than adults from a given dose of radiation both because they are inherently more radiosensitive and because they have more remaining years of life during which a radiation-induced cancer could develop. For example, compared with a 40-year-old adult receiving a similar dose of radiation to the chest, an infant has a 4- to 5-fold higher lifetime risk of developing lung cancer.8 In addition, organs receive higher radiation doses when scanned in a child compared with an adult. Radiation dose associated with gated 16-slice CCT (≈15 mSv) is almost 3-fold larger than that associated with conventional coronary angiography (≈5 mSv).9 Several new techniques to reduce radiation dose of CCT are being developed, but their implementation in clinical practice remains limited at the present time.10 In view of these data, young children undergoing CCT seem especially susceptible to radiation risks. In children with complex CHD, this risk may be compounded by their previous or subsequent exposure to ionizing radiation during cardiac catheterization. Although CCT can provide high-quality imaging in patients with CHD, the often underappreciated risks of exposure to ionizing radiation should be considered carefully when choosing the imaging modality for a particular patient. To reduce these risks to “as low as reasonably attainable” levels,11 it seems sensible that CCT be used only when other imaging modalities such as echocardiography or CMR are inconclusive, unavailable, or contraindicated.

Radiation doses associated with nuclear scintigraphy are also relatively large. The effective doses associated with a myocardial perfusion scan using 99mTc sestamibi at rest and stress (≈11 mSv) are almost twice those associated with coronary angiography.12 Therefore, when myocardial perfusion imaging is necessary in children, alternative modalities such as echocardiography with exercise or pharmacological stress or CMR with pharmacological stress are preferable.

**Age-Specific Considerations**

Because the challenges relevant to cardiac imaging of CHD are different in a fetus, infant, child, adolescent, or adult, age-specific considerations are worth discussing.
Table. Comparison of Imaging Modalities

<table>
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<th></th>
<th>Echocardiography</th>
<th>CMR</th>
<th>CCT</th>
<th>Nuclear Scintigraphy</th>
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<td>22.51‡</td>
<td>14.39§</td>
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<tr>
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<td><strong>Trachea and main stem bronchi</strong></td>
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†Sum of relative value units for CPT codes 93303, 93325, and 93320.
‡CPT code 75562.
§CPT code 71275.
∥CPT code 78465.
¶Temporal resolution for nuclear techniques is variable and depends on the radionuclide and counts.
**When measurement of ventricular stroke volume differential is physiologically appropriate.

**Fetus**

With advances in obstetric ultrasound screening, approximately 30% to 50% of severe CHD is diagnosed before birth. Echocardiography is the primary imaging modality used for fetal cardiac imaging, with excellent diagnostic accuracy for most CHD except for atrial or ventricular septal defects, partially anomalous pulmonary venous connection, anomalies of the coronary arteries, and coarctation of the aorta. Fetal arrhythmias can also be detected and characterized using M-mode, pulsed Doppler, and tissue Doppler imaging. In addition to anatomic evaluation, a wealth of physiological data such as measures of systolic and diastolic ventricular function, cardiac output, and Doppler parameters of fetal well-being can also be obtained. Imaging for suspected CHD is typically performed between 18 and 22 weeks of gestation, although successful imaging during the first trimester (11 to 15 weeks) has been reported using both transabdominal and transvaginal approaches.

Fetal MRI is being used increasingly for the evaluation of noncardiac congenital anomalies such as central nervous system abnormalities and congenital diaphragmatic hernia. CMR for fetal cardiac imaging has also been recently shown to be feasible. Several theoretical advantages make CMR attractive for fetal imaging including the ability to perform tomographic imaging in arbitrary planes independent of acoustic windows. However, significant technical challenges to fetal CMR remain, including limited spatial and temporal resolutions, lack of commercially available methods for fetal cardiac gating, fetal motion during scanning, and paucity of safety data, especially during the first trimester.

**Infants and Children <8 Years**

Echocardiography is the primary cardiac diagnostic modality in this age group, both for establishing initial diagnosis and for follow-up after medical or surgical treatment. Most children in this age group have adequate acoustic windows, and an accurate diagnosis can be established even in patients with the most complex anatomy and in low-birth-weight infants. Sedation may be required in infants and young children who are unable to cooperate with the examination. Rarely, additional imaging modalities are necessary for anatomic assessment of the extracardiac vasculature, further delineation of spatial orientation or relationship of cardiac chambers or vessels, tissue characterization (Figure 4), or to obtain additional physiological data such as quantification of ventricular size and function and quantification of valve regurgitation. In these instances, other diagnostic techniques such as CMR can be used selectively. The accuracy of CMR in this age group in the evaluation of a wide spectrum of congenital heart defects has been established. Questions regarding the extracardiac vasculature can also be answered using CCT. Its use should be balanced against the risks of ionizing radiation exposure.

**Adolescents and Adults**

Typically, as patients grow larger, there is a progressive degradation in the quality of echocardiographic images due to declining acoustic windows, especially in patients who have undergone prior cardiac surgical procedures. Echocardiography continues to remain the primary diagnostic modality due...
to its ready availability and ease of use, but CMR plays an increasing role, especially for the evaluation of the extracardiac thoracic vasculature, ventricular function, and flow measurements (Figure 5). CMR is usually preferred over CCT in this age group because it avoids exposure to ionizing radiation and can provide a wealth of functional information. CCT plays an important role in patients with contraindications to CMR, such as pacemaker dependence, or those in whom concomitant evaluation for coronary artery disease is necessary (Figure 6).

**Imaging Algorithm**

Echocardiography is the initial imaging modality used in the evaluation of patients with suspected CHD and should always be performed before other modalities are used. In most patients, echocardiography alone provides sufficient information to complete the diagnostic evaluation. However, in the small proportion of patients in whom echocardiography is incomplete or inconclusive, other diagnostic techniques should be considered, based on the clinical question. A general diagnostic algorithm is presented in Figure 7 and should be tailored to an individual patient’s specific diagnostic questions. Finally, cardiac catheterization still remains the reference standard for angiography and hemodynamic assessment and should be used when data from noninvasive imaging remain incomplete or inconclusive.

**Figure 2.** Temporal trends in use of diagnostic modalities and surgery at Children’s Hospital Boston from 1983 through 2008. A, Despite a relatively modest growth in surgery and catheterization, use of echocardiography and CMR has increased steadily. B, Decreased utilization of diagnostic catheterization coupled with a steady increase in transcatheter interventions, modest increase in cardiac surgery, and rapid increase in use of CMR. Cath indicates cardiac catheterization; Echo, echocardiography; Surg, cardiac surgery.

**Figure 3.** Multimodality evaluation of cardiac anatomy, function, and myocardial viability in a 4-year-old child with delayed diagnosis of Kawasaki disease and coronary aneurysms. A, Conventional coronary angiography demonstrated multiple aneurysms in the right coronary artery. The circumflex coronary artery was occluded (not shown). B, Myocardial perfusion imaging by single-photon emission computed tomography demonstrated a fixed perfusion defect in the inferior left ventricular wall (arrow). C, Myocardial delayed enhancement CMR imaging demonstrated transmural late gadolinium enhancement in a corresponding area of the left ventricle consistent with myocardial infarction.
Selected Clinical Applications

Evaluation of Ventricular and Myocardial Performance

Systolic ventricular performance is often a determinant of long-term outcomes in patients with CHD. Emerging evidence suggests that diastolic dysfunction may also be an important determinant of outcomes after surgical repair of CHD, and this realization has prompted interest in assessment of diastolic performance using noninvasive techniques.

Ventricular Size and Global Systolic Performance

Echocardiography is the primary modality used in the assessment of left ventricular (LV) size and function. Due to several technical limitations, M-mode–derived measures have been progressively supplanted by measurements made using 2D techniques, which allow more accurate and reproducible measurements of LV volume, mass, and ejection fraction. These methods have been shown to be reasonably accurate in adult patients, but, compared with a reference technique such as CMR, there are several sources of error and considerable

Figure 4. Echocardiographic and CMR evaluation of a large cardiac tumor in a newborn. A, 2D echocardiography demonstrated a single large mushroom-shaped echogenic mass arising from the ventricular septum. Differential diagnosis included fibroma, rhabdomyoma, and hemangioma. B, CMR was performed for tissue characterization and demonstrated the large solitary homogenous septal tumor with a small pericardial effusion. C, Compared with normal left ventricular myocardium, the tumor was isointense on T1-weighted imaging, hyperintense on T2-weighted imaging, showed poor first pass perfusion, and did not show late gadolinium enhancement. These characteristics supported the diagnosis of rhabdomyoma. D, Histological examination was diagnostic for rhabdomyoma with characteristic “spider cells” (arrowhead). T1 indicates T1-weighted imaging; T2, T2-weighted imaging; Perf, first-pass perfusion imaging; MDE, myocardial delayed enhancement imaging.
interstudy and interobserver variability.\textsuperscript{28} The accuracy and reproducibility of 2D echocardiographic techniques for assessing LV volume and function have not been studied systematically in infants and children, especially those with CHD. Despite these limitations, data on the range of normal values for ventricular dimensions, volumes, mass, and ejection fraction derived from echocardiography in the pediatric age group are available and are helpful in evaluating the clinical significance of findings.\textsuperscript{29,30}

The complex geometry of the right ventricle (RV) makes it difficult to accurately quantify its size and systolic performance. Although several 2D echocardiographic methods have been proposed, none has been shown to be reliable and ranges for normal values are not available.\textsuperscript{31} With real-time 3D echocardiography, it is possible to quantify ventricular volume and function without the use of geometric assumptions. These techniques have been shown to be accurate and reproducible compared with CMR for quantifying both left and right ventricular volumes and function.\textsuperscript{32,33} A major limitation of 3D echocardiography is its dependence on adequate acoustic windows, which can be limited after cardiac surgery and in adult patients. Furthermore, unlike 2D echocardiography, ranges for normal values for ventricular volume, mass, and ejection fraction by 3D echocardiography are not available. Expertise in using 3D techniques for routine quantification of LV and RV function is currently limited to a few centers and requires specialized analysis software.

To overcome problems associated with complex ventricular geometry, several nongeometric echocardiographic methods that rely on Doppler echocardiography have also been used to assess global ventricular performance. These include the myocardial performance index, $dP/dt$, systolic annular velocity, and isovolumic acceleration measured using tissue Doppler imaging (TDI).\textsuperscript{34,35} These parameters have the advantage of being relatively independent of ventricular geometry and less influenced by restricted acoustic windows. Disadvantages include a lack of normal values for children with CHD.

CMR is considered the reference technique for quantifying ventricular volume and global systolic function.\textsuperscript{36} The
technique has been extensively validated against direct measurements in explanted hearts and the results in children are highly reproducible for left and right ventricles, including those with abnormal loading conditions. High-quality images can be obtained independent of body size or habitus, which can be especially useful in adolescents and young adults in whom echocardiography is suboptimal. More reproducible measurements are also advantageous in guiding clinical decisions as well as in research trials that rely on longitudinal measures of ventricular size or function. Although CMR has the highest accuracy and reproducibility among all noninvasive techniques, it has important limitations. First, CMR in young children usually requires sedation or general anesthesia. Furthermore, only limited normative data are available for young children. Gated CCT has been shown to be accurate in the quantification of left and right ventricular volumes, mass, and ejection fraction in adults. CCT is generally not the preferred modality for ventricular function assessment because of its use of ionizing radiation and a relatively low temporal resolution compared with CMR and echocardiography. However, it can be useful in patients with contraindications to CMR.

**Regional Systolic Ventricular Function**

Although regional abnormalities are typically associated with ischemic heart disease, they can also occur in patients with CHD. Traditionally, regional function is assessed in a qualitative or semiquantitative fashion, but newer methods for the evaluation of regional abnormalities such as strain and strain rate imaging allow quantitative assessment and have been validated against microsonometry and tagged CMR. Age-related changes in LV strain and strain rate in children have been recently described. In patients after a Senning operation, Pettersen et al. showed that contraction pattern of the systemic RV shifts from longitudinal to circumferential shortening and the ventricle does not exhibit normal torsion. Weidemann et al. used strain rate imaging to demonstrate abnormalities in regional LV and RV function in asymptomatic children after repair of tetralogy of Fallot. Strain imaging has also been shown to be feasible in fetal life. Further investigation is needed to determine the clinical utility of strain imaging in children and adults with CHD and the relationship between measurements and clinical outcomes.

CMR can also evaluate regional ventricular function. In addition to qualitative and semiquantitative assessment using cine steady-state free precession imaging, tissue-tagged CMR has been validated as an accurate method to assess regional myocardial strain. Tagged CMR has been used to show abnormalities in myocardial deformation in children with single ventricle physiology and in patients with a systemic RV. Routine clinical use of tagged CMR is hampered by the need for moderately complex image processing and lack of normative values as well as data relating the results with clinical outcomes. A newly described strain-encoded CMR technique may allow more rapid calculation of myocardial strain and strain rates.
Diastolic Function
Diastolic dysfunction can occur after repair of CHD, and there is some evidence that it may adversely affect outcomes. The reference techniques for assessing diastolic performance involve invasive measurements using micromanometer tipped catheters. Various noninvasive measures of diastolic performance have been proposed, but each has important limitations. Current approach to noninvasive assessment of diastolic function incorporates data from atrioventricular valve inflow parameters, pulmonary vein Doppler, and TDI (Figure 8). Age related maturational changes in TDI velocities have been described.

A few clinical applications of TDI have been reported in the pediatric age group. In children with hypertrophic cardiomyopathy, lower E/E’ ratio has been shown to be associated with an increased risk of adverse clinical outcomes. TDI velocities are often abnormally low after cardiac transplantation and have been shown to be associated with the risk of graft failure.

Several reports have investigated the use of CMR for the evaluation of diastolic function. Phase contrast techniques can be used to evaluate inflow as well as myocardial tissue velocities. In addition, early and late diastolic myocardial strain and strain rate can be calculated using either tissue tagging techniques or the recently reported strain-encoded CMR. Although tissue-tagged CMR has been available for more than 2 decades, clinical applications remain limited. Fogel et al used tagged CMR to study diastolic biomechanics in normal infants and showed that circumferential lengthening strain was highest at the lateral LV wall and that rotational strain was most pronounced at the LV apex.

Although several noninvasive tools for assessing diastolic function are available and some have been found to improve diagnosis and risk stratification in adults with diastolic heart failure, their routine clinical use in patients with CHD remains limited. Most of these indices are sensitive to alterations in preload, afterload, and heart rate, all of which can be abnormal in patients with CHD, making interpretation of findings difficult. Further, the prognostic value of these indices in patients with CHD awaits further studies.

Evaluation of the Functionally Single Ventricle
Anatomic malformations that result in a functionally single ventricle represent some of the most complex forms of
CHD. Surgical treatment involves several stages, culminating in the Fontan operation. Initial diagnosis can be made accurately by echocardiography both in fetal life (Figure 9) and in the newborn period (Figure 10). Rarely, additional imaging using CMR is necessary to clarify details of intracardiac or extracardiac vascular anatomy, quantify ventricular volume for “borderline” sized ventricles, and to evaluate for the presence of endomyocardial fibroelastosis. Diagnostic cardiac catheterization is rarely necessary in the newborn period. Traditionally, cardiac catheterization is performed before the cavopulmonary anastomosis and the Fontan operation. However, a recent prospective randomized trial of CMR versus catheterization showed that CMR is a safe, effective, and less costly alternative to routine catheterization in the evaluation of selected patients before bidirectional Glenn operation. Another retrospective analysis showed that a non-invasive diagnostic algorithm effectively screened for patients who are unsuitable for a Fontan operation and that omission of routine preoperative hemodynamic assessment at catheterization did not impair prediction of adverse postoperative outcomes.

Ventricular function can often be abnormal in patients with a functionally single ventricle. Altmann et al showed that global RV dysfunction identified on echocardiography is a risk factor for intermediate-term mortality after stage I palliation for hypoplastic left heart syndrome. CMR can be useful when abnormal ventricular geometry and/or poor acoustic windows limit quantitative assessment of ventricular function by echocardiography. In a multicenter evaluation of children ages 6 to 18 years after a Fontan operation, systolic dysfunction was identified in 27% subjects and was associated with older age at evaluation and RV morphology. Abnormalities of regional ventricular function and evidence for ventricle-ventricle interactions have been identified after the Fontan operation using inflow Doppler parameters, TDI, Doppler strain imaging, and tagged cine CMR. Rathod et al recently showed that in patients late after the Fontan operation, higher ventricular volumes and abnormalities of global and region ventricular function were associated with myocardial fibrosis identified by myocardial delayed enhancement CMR. There is also evidence for abnormalities of echocardiographic parameters of diastolic ventricular performance in functionally single ventricles. In a multicenter study, echocardiographic diastolic function grade was abnormal in 72% of subjects.

The complex flow dynamics of the Fontan circulation and its relationship with geometric parameters can be evaluated noninvasively using CMR. Fogel et al showed that flow in the Fontan pathway is phasic to both cardiac and respiratory cycles and highest flows occur near end-systole and early diastole, as well as in inspiration, whereas lowest flow occurs
during diastasis. Using multidimensional phase contrast CMR imaging, Be’eri et al demonstrated that blood flow patterns are more organized and uniform in lateral tunnel than in atrioventricular Fontan pathways and are significantly influenced by pathway diameter. CMR has been shown to be more accurate than lung scintigraphy in quantifying blood flow to each lung in the Fontan circulation. Finally, evaluation for abnormalities of the pulmonary vasculature such as pulmonary artery stenosis and aortopulmonary or veno-venous collaterals is challenging using echocardiography but can be performed using CMR.

Vascular Function
Abnormalities in endothelial function have been identified using ultrasound techniques after surgical repair of coarctation of the aorta and the Fontan operation. In addition to endothelial function, other aspects of vascular function such as the elasticity or stiffness of the great vessels may be relevant to patients with CHD. Grotenhuis et al used CMR to demonstrate reduced elasticity of the aortic root after the arterial switch operation. Using Doppler strain echocardiography, Vitarelli et al showed reduced strain and increased stiffness at the site of surgical repair of coarctation of the aorta. Similar abnormalities in aortic stiffness have been demonstrated after aortic arch reconstruction for hypoplastic left heart syndrome.

Hemodynamic Evaluation
Although cardiac catheterization remains the reference standard for assessment of cardiac hemodynamics in patients with CHD, it is not a practical method for longitudinal follow-up. Several noninvasive techniques allow assessment of cardiac physiology.

Pressure gradients across regions of valve or blood vessel stenosis can be estimated using Doppler echocardiography. Pressure gradients and estimated valve areas can also be obtained using phase contrast CMR. Although CMR-derived measurement is more time-consuming to obtain, it can be useful when Doppler evaluation is limited by poor acoustic windows or suboptimal angle of interrogation.
Accurate measurement of the ratio of blood flow in the systemic and pulmonary circulations (Qp/Qs) is important in quantifying the hemodynamic burden of several CHD associated with intracardiac and extracardiac shunts. Doppler techniques for estimation of blood flow are based on measurement of flow velocity using spectral Doppler and vessel diameter on 2D imaging. Because the flow rate and vessel area are not measured directly, the accuracy and reproducibility of this method is limited and hence they are not used routinely in clinical care. CMR overcomes these limitations by measuring flow rate and vessel cross-sectional area independent of vessel location and geometry. In patients with an atrial septal defect, CMR measurement of Qp/Qs agreed well with estimation based on invasive oximetry (mean difference between techniques, 0.06 Qp/Qs units or 2.6%) and had low interobserver variability (<4%).

Quantification of differential blood flow to each lung and individual lung segments is useful in assessing the hemodynamic significance of branch pulmonary artery or pulmonary vein stenosis. These measurements are traditionally performed using nuclear scintigraphy. However, nuclear scintigraphy involves exposure to ionizing radiation, which can be avoided by using CMR. Measurements of differential flow to each lung using phase contrast CMR have shown close correlation and agreement with nuclear scintigraphy. In patients who have undergone a Fontan operation, estimation of differential blood flow by nuclear scintigraphy may be inaccurate due to asymmetrical streaming from the superior and inferior vena cavae. CMR overcomes this limitation and has been shown to provide more accurate estimates. Differential lung perfusion can also be estimated by analyzing signal-time curves during time-resolved contrast enhanced 3D MRA. This technique shows close correlation and agreement with nuclear scintigraphy and also allows assessment of segmental perfusion abnormalities.

Estimation of pulmonary artery pressure and resistance is important in patients with CHD. RV pressure can be estimated by measuring the tricuspid regurgitation jet velocity using Doppler techniques. This technique is accurate in patients with sufficient tricuspid regurgitation and adequate spectral Doppler signal but may underestimate peak pulmonary artery pressure in patients with insufficient tricuspid regurgitation even in the presence of severe pulmonary hypertension. Several investigators have attempted estimation of pulmonary artery pressure or resistance using CMR measures such as flow velocity, pressure wave velocity and acceleration time in the pulmonary artery, and end-systolic curvature of the ventricular septum. However, none of these techniques have been adequately validated and none are currently in routine clinical use. Using specialized imaging suites that combine CMR measurement of blood flow with catheter-based pressure measurement, methods to estimate pulmonary vascular resistance and vascular compliance have also been described. The advantages of this approach include reduced exposure to ionizing radiation and more accurate estimation of pulmonary blood flow, allowing more accurate estimation of pulmonary vascular resistance.

Image-Guided Procedures
Noninvasive imaging modalities are increasingly used during surgical and catheter-based interventions, both to provide guidance during the procedure and to evaluate the results. Echocardiography is usually the modality of choice due to its portability, availability, and ability to provide real-time imaging. Catheter-based closure of atrial septal defects can be successfully performed under transesophageal or intracardiac 2D or real-time 3D echocardiographic guidance. Transesophageal echocardiography can be used to evaluate the results of complex congenital heart surgery in the operating room before the patient is disconnected from cardiopulmonary bypass and has been shown to improve surgical outcomes. Recently, 3D transesophageal echocardiography probes have become available and it is possible to obtain real-time 3D images as well as volumetric 3D data sets that can be reformatted and viewed in arbitrary planes. 3D transesophageal echocardiography can be especially useful in evaluation before and after surgical valve repair to assess the mechanism of valve regurgitation or stenosis.

Although echocardiography is the primary modality used for guidance of surgical or interventional procedures, CMR offers several potential advantages such as the ability to provide high-resolution tomographic images in any arbitrary plane. In preliminary work, the feasibility of placing endovascular stents and devices for closure of atrial septal defects and to balloon dilate aortic coarctation has been demonstrated. Although interventional CMR is the subject of intense investigation, it is not currently in routine clinical use. Impediments to the implementation of interventional CMR include its relatively low temporal resolution, limited availability of MR-compatible catheters and devices and technical difficulties in visualizing and tracking catheter tips and devices during manipulation.

Implications for Training
Because noninvasive imaging is a primary means for diagnosing CHD, competence in performance and interpretation of these modalities is now essential to the practice of pediatric cardiology. Recently published pediatric training guidelines for noninvasive imaging require all pediatric cardiology fellowship trainees to achieve a “core” level of competence in echocardiography and CMR. An “advanced” level of competence is required of those trainees who intend to be dedicated pediatric imagers. Recommended goals for the advanced level of training include competence in all aspects of echocardiography (transthoracic, transesophageal, and fetal) and CMR imaging including independent performance and interpretation of studies, understanding instrumentation and physics, management and quality assurance of the noninvasive laboratories, and basic or clinical research. This level of expertise typically requires an additional year of dedicated training after completing a pediatric cardiology or radiology fellowship program. Several training programs in North America now offer a 4th year fellowship in advanced pediatric cardiac imaging.

Conclusion
Simultaneous advances in several noninvasive imaging techniques have significantly affected the care of children and
adults with CHD. Despite the emergence of other imaging modalities, echocardiography remains the first-line technique in most cases. To choose the best techniques for an individual patient, imagers today must have detailed knowledge of the various modalities available. This has major implications for the training of future noninvasive cardiologists.

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None.

References

71. Fogel MA, Weinberg PM, Hsuyd A, Hubbard A, Rychik J, Jacobs M, Fellows KE, Haselgrove J. The nature of flow in the systemic venous pathway measured by magnetic resonance blood tagging in patients with...


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