Cardiac Magnetic Resonance Imaging and Computed Tomography in Newborns With Congenital Heart Disease

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Author Disclosure
Drs Gupta and Chandran have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

Educational Gaps

1. Neonatologists may be unfamiliar with history and evolution of cardiac imaging.
2. Neonatologists may be unfamiliar with imaging principles behind the use of computed tomography angiography (CTA) and cardiac magnetic resonance imaging (CMRI).
3. Neonatologists may be unfamiliar with indications and utility of CTA and CMRI in congenital heart disease.

Abstract
Congenital heart disease continues to be a leading cause of neonatal morbidity; however, advancements in diagnostic technology and surgical techniques have markedly improved patient survival. In particular, in-depth noninvasive imaging with computed tomography angiography (CTA) and cardiac magnetic resonance imaging (CMRI) has had a tremendous effect on the amount of information available to the surgeon or interventionalist before any procedure. CTA and CMRI have their own advantages and disadvantages, and this review provides an overview of these technologies, highlighting clinical indications. This article also provides images and videotapes to better highlight our understanding of various complex congenital heart lesions.

Objectives
After completing this article, readers should be able to:

1. Understand the basic imaging principles behind computed tomography angiography (CTA) and cardiac magnetic resonance imaging (CMRI).
2. Understand basic indications for CTA and CMRI in the diagnosis of congenital heart disease (CHD).
3. Understand the advantages and disadvantages of CTA vs CMRI.
4. Become familiar with key CTAs and CMRIs of common CHD lesions.

Introduction
The incidence of congenital heart disease (CHD) in the United States is approximately 1%. (1)(2) CHD is the leading cause of neonatal mortality attributable to birth defects, accounting for 27.5% of affected neonates, with chromosomal anomalies being the second highest at 22.4%. (3) Echocardiography continues to be the primary initial modality used to define these congenital heart lesions, but there are limitations to its use, especially as it applies to extracardiac arterial or venous and thoracic delineation. Often, further diagnostic workup is required before any form of intervention. With recent advancements in technology in computed tomography angiography (CTA) and cardiac magnetic resonance imaging (CMRI), pediatric cardiologists have been able to furnish these answers, thus obviating the need for more traditional invasive procedures, such as diagnostic cardiac catheterization. In our institution, we primarily use CTA for extracardiac great vessel imaging, including aortic arch, pulmonary artery, and venous anomalies, as well as defining adjoining thoracic structures, including the airway. In addition, CMRI can be used for these purposes with the added benefit of intracardiac anatomical and functional imaging.

Cardiac CTA
Computed tomography (CT) was introduced in the 1970s by Sir Godfrey Hounsfield, and since then, there has been tremendous development in CT technology. (4)(5) CT uses
x-rays or ionizing radiation, which is emitted from the source and detected by special detectors opposite to the source. After data acquisition, special algorithms are used to create the final image. Initially, single-slice scanners were used. Today, CT contains 320-slice scanners called multidetector-row CT (MDCT). The temporal and spatial resolution of CT with the advent of MDCT scanners has paved the way for improved imaging of complex CHD, thus bypassing the restrictions otherwise posed by faster neonatal heart rates and smaller neonatal structures.

**Indications for Neonatal Cardiac CTA**

The indications for neonatal cardiac CTA include the following: (1) aortic arch anomalies (coarctation of the aorta, interrupted aortic arch, and vascular ring or sling), (2) pulmonary artery anomalies (pulmonary artery stenosis and aortopulmonary collaterals), (3) anomalies of pulmonary venous return (anomalous pulmonary venous return and pulmonary vein stenosis), (4) coronary artery anomalies (anomalous coronary artery origins or course), (5) extracardiac thoracic anatomy (extracardiac masses, airway compression, and lung disease), and (6) postoperative assessment after CHD surgery.

**Sample CTA Protocol**

At our institution, we use a 320-slice Aquilion ONE scanner (Toshiba Medical Systems Corp, Tustin, CA). Neonatal images are typically obtained without any sedation or breath-holding and usually without administration of β-blockers even with heart rates greater than 80 beats per minute. Low-osmolar iodine contrast medium (Visipaque) is injected intravenously at a dose of 2 mL/kg via a 24-gauge or larger peripheral catheter or a peripherally inserted central catheter not exceeding the rate of 0.5 mL/s manually. The scan is initiated after the injection of contrast without using bolus tracking. The scan volume typically extends superiorly from the thoracic inlet to just below the diaphragm inferiorly. With suspected heterotaxy, interruption of the inferior vena cava, and infracardiac anomalous pulmonary venous return, the scan volume is extended to the infrarenal region. All scans are performed in a cranial-caudal direction with CT parameters adapted to the patient’s weight. Postprocessing of the MDCT scans is performed on a dedicated Vital Images workstation using Vitrea 2 software, version 4.0 (Vital Images Inc, Minnetonka, MN).
Cardiac Morphologic Findings in Complex CHD

AORTIC ARCH ANOMALIES. Coarctation of the aorta (CoA) is associated with clinical symptoms in the newborn period after the patent ductus arteriosus (PDA) closes and can manifest variably as an asymptomatic neonate with a murmur to outright left ventricular dysfunction as a result of increased afterload. Cardiac CTA is extremely useful in providing detailed anatomy of the arch and the pattern and caliber of the branches. CoA can present as a focal discrete juxtaductal lesion with poststenotic dilatation (Fig 1 and Video 1), transverse arch hypoplasia (Fig 2 and Video 2), or interrupted aortic arch with or without the presence of aberrant head and neck vessel branching and other associated anomalies. Aortic arch anomalies can also be well delineated using cardiac CTA (Fig 3 and Video 3), and CTA has the added benefit of clarifying thoracic abnormalities (eg, tracheal compression in the double aortic arch; Video 4). These images negate the need for other procedures, such as bronchoscopy or barium swallow. (9) CTA has enabled us to diagnose these lesions before neonates become markedly symptomatic.

PULMONARY ARTERY ANATOMY. Tetralogy of Fallot (TOF) is the most common cyanotic CHD, and its morphologic spectrum ranges from TOF with pulmonary stenosis to pulmonary atresia. In severe cases when the pulmonary blood flow is duct dependent, the traditional approach to delineate the source of the patient’s pulmonary blood flow was cardiac catheterization, which was invariably associated with increased cost, need for vascular access, and sometimes hemodynamic instability. Today, cardiac CTA can be used to image branch pulmonary arteries, the PDA (Figs 4 and 5 and Videos 5 and 6), and the number or size and distribution of major aortopulmonary collaterals (MAPCAS). (10) It can also be used postoperatively to assess the adequacy of pulmonary artery repair (Fig 6). This approach has also been applied to other lesions, such as truncus arteriosus and isolated pulmonary artery stenosis. (9)

ANOMALIES OF PULMONARY VENOUS RETURN. Pulmonary venous anomalies can be categorized into total anomalous pulmonary venous return (TAPVR) and partial anomalous pulmonary venous return (PAPVR) based...
on the number of veins that drain abnormally. Infants with TAPVR present in the newborn period with significant hypoxia; if there is obstruction to venous return (typically with the infradiaphragmatic type), infants will also have hypotension and require urgent surgical correction. Preoperative diagnosis of TAPVR can be challenging because the imaging by echocardiography is extremely difficult, even in the hands of an experienced operator. Cardiac catheterization has been used in the past, but this is a high-risk procedure, particularly because of the potential for pulmonary hypertensive crises and further hemodynamic instability. We have used cardiac CTA (Fig 7) as an expeditious method to diagnose this lesion, thus providing the surgeon with a clear diagnosis without any procedural complications. We have also applied CTA to delineate the pulmonary veins in PAPVR, which can be especially useful if CMRI is contraindicated.

CORONARY ARTERY ANOMALIES. Congenital coronary artery anomalies can be categorized as primary or related to anomalies of the origin of the coronary arteries (anomalous left coronary artery from the pulmonary artery) (Fig 8) or coronary anomalies associated with other CHD (left anterior descending artery from the right coronary artery in TOF). Infants with anomalous left coronary artery from the pulmonary artery can present acutely in shock; accurate diagnosis requires echocardiography in the appropriate clinical milieu and possibly cardiac catheterization. In
unclear situations, we have attempted to use CTA to delineate the coronary artery origins, but spatial resolution is limited by the rapid heart rates in newborns. In contrast to adult coronary studies, we do not recommend administering β-blockers to neonates to obtain an optimal heart rate because there is a risk of symptomatic bradycardia or depressed cardiac function in this otherwise vulnerable population.

Figure 4. Sagittal image in a newborn with tetralogy of Fallot with pulmonary atresia showing a tortuous patent ductus arteriosus (PDA) arising from undersurface of the arch.

Figure 5. Modified axial thick-section image of the newborn with tetralogy of Fallot with pulmonary showing a patent ductus arteriosus (PDA) arising from undersurface of the arch of aorta supplying confluent branch pulmonary arteries. Bibasilar atelectasis is present. LPA = left pulmonary artery; RPA = right pulmonary artery.

Figure 6. Axial section in an infant with right pulmonary artery (RPA) coming off the aorta with a band placed on the RPA to prevent pulmonary overcirculation. PA = pulmonary artery.

Video 5. Modified thick-section image in a newborn with tetralogy of Fallot with pulmonary atresia showing a patent ductus arteriosus arising from undersurface of the arch of aorta supplying the confluent branch pulmonary arteries. Click here to play the video.
Extracardiac Thoracic Anatomy and Postoperative Assessment

CTA is extremely useful in providing additional information about extracardiac structures, including the lung parenchyma, airways (caliber and compression by surrounding structures), and thoracic wall structures. Technology has made it possible to perform dynamic studies in which the airway caliber can be evaluated in both phases of the breathing cycle, thus obviating the need for bronchoscopy for diagnosing tracheal compression (Video 4) in either preoperative or postoperative cases. CTA can be helpful for evaluating the lung parenchyma, particularly for bronchoalveolar changes (atelectasis or pneumonia). In Fig 5, the infant’s atelectasis in dependent areas can be clearly seen and appears more pronounced in the lung window views. The resolution of CTA is definitely superior to CMRI for these extracardiac structures.

Radiation Exposure

Exposure to ionizing radiation continues to be the biggest risk factor for cardiac CTA, especially in infants with complex CHD, because multiple studies may be obtained in a lifetime. The detrimental effect is the risk of genetic mutations and malignant neoplasms, with a median lifetime cumulative dose of 7.7 mSv per patient. (13)(14) (15)(16) We have previously published our experience with minimizing radiation exposure using the 320-detector scanner. In our nongated protocol, the mean (SD) dose administered is 1.8 (0.7) mSv, and by using a gating protocol,

Video 6. Three-dimensional reconstruction in a newborn with tetralogy of Fallot with pulmonary atresia showing a patent ductus arteriosus arising from the undersurface of the arch of the aorta supplying the confluent branch pulmonary arteries. Click here to play the video. LPA = left pulmonary artery; RPA = right pulmonary artery.

Figure 7. Modified axial thick-section image showing supracardiac total anomalous pulmonary venous return with common pulmonary vein (CPV) draining through left vertical vein to the proximal left innominate vein. SVC = superior vena cava.

Figure 8. Coronal section showing an anomalous left coronary artery from the pulmonary artery (ALCAPA). LCA = left coronary artery; MPA = main pulmonary artery.
the dose was further reduced to 0.8 (0.39) mSv. (8) To put this into perspective, the approximate effective radiation dose of chest radiography for an average-sized adult is 0.1 mSv, and a similar scan for pediatric patients exposes them to a much higher radiation dose. In the United States, the mean exposure to radiation is 3 mSv per year. The best way to decrease this risk is by applying the principle of ALARA (as low as reasonably achievable) and reducing the dose of ionizing radiation using various techniques, such as decreasing the scan time and/or modifying the kilovoltage, current, and scan pitch. (4)(7)

CMRI

Historical Perspective
Initial use of CMRI dates back to the 1970s when it was used to measure in vitro myocardial metabolism in perfused rat hearts. In the past 3 decades, its use has evolved to assist with cardiac anatomy, ventricular function, extracardiac vasculature, and viability and metabolism of the myocardium. Because CMRI can provide such an extensive amount of information, it has been described as a one-stop-shop for cardiac imaging. (17)

MRI Physics
Felix Bloch and Edwin Purcell independently described the phenomenon of nuclear magnetic resonance in...
1946 and were awarded the Nobel Prize in Physics in 1952. Since then, many other principles have been described that have enabled technology to improve the resolution and information obtained by CMRI. Hydrogen nuclei (protons) are present in large numbers in the body and have an intrinsic alignment, which is essentially random without any net magnetization. On application of a high-strength magnetic field, the spins of these protons align with the direction of the magnetic field. For the purposes of CMRI, the magnetic field strength typically used is 1.5 T, although other centers have started using 3-T scanners. Radiofrequency energy applied during the alignment excites the spins and changes their alignment relative to the direction of energy application. In the process of returning to their baseline state (relaxation), the released magnetic energy is then converted to a resonant signal in a receiver signal placed perpendicular to the transverse plane (based on the principle of the electromagnetic effect). During the last step, the computer converts these electrical signals to an image by the complex process of Fourier transformation. (4)(17)(18)

**Indications of Neonatal CMRI**

Indications of neonatal CMRI include the following: (1) cardiac chamber volume, mass, and functional analysis; (2) cardiac morphologic findings in complex congenital heart disease (including aortic arch anomalies [coarctation of the aorta, interrupted aortic arch, and vascular...
ring or sling], pulmonary artery anomalies [pulmonary artery stenosis and aortopulmonary collaterals], anomalies of pulmonary venous return [total or partial anomalous pulmonary venous return and pulmonary vein stenosis], and complex CHD); (3) tissue characterization and viability imaging (including evaluation of cardiac masses and tumors, evaluation of cardiomyopathies and myocarditis, and evaluation of ischemic tissue or fibrosis using delayed enhancement sequences); (4) hemodynamic data (including quantification of pulmonary to systemic blood flow ratio when assessing shunt lesions, right and left ventricular ejection fraction, and assessment of valvular regurgitation and stenosis); (5) extracardiac thoracic anatomy (including complex vascular anatomy, extracardiac masses, airway compression, and lung disease); and (6) postoperative assessment after CHD surgery. (19)

Myocardial Ventricular Volume, Mass, and Function

CMRI remains the gold standard for quantification of ventricular volumetrics and function, predominantly because of its ability to measure acquired images without the need for any geometric assumptions. (18) This is especially useful in newborns with complex CHD, such as hypoplastic right or left heart syndrome or an unbalanced atrioventricular septal defect with borderline chamber sizes. The actual chamber volume thresholds determine adequacy for a single-ventricle vs a biventricular or 1½-ventricle repair. (20)(21)(22)

The general approach involves acquiring multiple cine sections in the short-axis plane of the ventricles and tracing both the epicardial and endocardial margins in systole and diastole. The volumes and ejection fractions are then calculated using the Simpson method. This method allows for accurate and reproducible calculations of the ejection fraction, and the cine loops provide an extremely useful visual impression of overall cardiac function and wall motion (Video 7). The applications of CMRI are multifold, and it remains the standard for diagnosing cardiomyopathies (Videos 8–10 and Figs 9 and 10), especially in older children because the neonatal presentations of such diseases are rare. (23)(24)(25)
Cardiac Morphologic Findings in Complex CHD
In complex CHD, even though the cardiac anatomy is usually easily delineated using echocardiography, there are multiple situations wherein CMRI is required for further enhancement of the diagnosis. In our center, where CTA is used to delineate extracardiac anomalies, we tend to use CMRI in the neonatal period for evaluating a combination of both complex intracardiac and extracardiac lesions. CMRI is extremely helpful in these situations, and it also provides the additional benefit of obtaining information about abdominal and thoracic viscera and other associated anomalies, as well as functional information.

AORTIC ARCH ANOMALIES. In the newborn period, echocardiography is usually a very good tool for assessing for aortic arch anomalies. Major anomalies in neonates consist of a CoA, vascular rings or slings, or a double aortic arch. Suspicion based on initial evaluation needs confirmation with advanced imaging. In this case, CMRI can be beneficial with the major limitations of longer scan time and need for sedation to diminish breathing or motion artifact, adding potential risks. At our center, we prefer to use CTA for the assessment of a neonate with a diagnosis of neonatal arch anomalies. CMRI, however, remains a mainstay of assessing aortic arch defects in older children (Fig 11). Videos 11 and 12 show a child with a significant recoarctation with collateral formation after a previous end-to-end CoA repair.

RIGHT VENTRICULAR OUTFLOW TRACT AND PULMONARY ARTERY ANOMALIES. In cases with duct-dependent pulmonary blood flow (especially TOF and pulmonary atresia), it is essential to assess the source of pulmonary blood flow. The presence and size of MAPCAS (Fig 12) and the size and confluence of the pulmonary arteries have an effect on the surgical repair and prognosis. In our center, we have selectively used CMRI with magnetic resonance angiography to delineate the right ventricular outflow tract, branch pulmonary arteries (Fig 13), and MAPCAS, while dynamically assessing biventricular function and relative branch pulmonary artery flow using differential flow quantification.

ANOMALIES OF PULMONARY VENOUS RETURN. As described previously, infants with infradiaphragmatic TAPVR usually require emergency surgery. In this scenario, longer scan times or additional procedures that involve sedation and intubation can be detrimental. In our center, CTA is often used to establish this diagnosis. Patients with
PAPVR may be asymptomatic and may not have their conditions diagnosed until adolescence or adulthood after presenting with unexplained right heart enlargement. CMRI is extremely useful for such cases and accurately reveals the anomalous drainage of one or more pulmonary veins into another systemic venous structure while providing additional information about right heart dimensions and shunt fraction (Videos 13 and 14).

INTRACARDIAC DEFECTS. CMRI is extremely effective in clarifying the presence and severity of different forms of intracardiac defects, ranging from simple shunts, such as ventricular septal defects (Fig 14 and Video 15), to more complex forms, including midline liver in a newborn with heterotaxy (Fig 15).

Video 13. Steady-state free-precession cine image showing a supracardiac type of total anomalous pulmonary venous return. Click here to play the video.

Video 14. Steady-state free-precession cine image showing an enlarged right ventricle in a patient with anomalous pulmonary venous return. Click here to play the video.

Figure 13. Steady-state free-precession cine image showing a supracardiac type of total anomalous pulmonary venous return. Click here to play the video.

Figure 14. Steady-state free-precession cine of the basal short axis plane showing a perimembranous ventricular septal defect (VSD) flow jet from the left ventricle to the right ventricle.

Figure 15. Cardiac magnetic resonance image showing a midline liver in a newborn with heterotaxy.
complex conditions, such as heterotaxy syndrome in which the CMRI can provide additional information about the abdominal viscera and systemic and pulmonary venous drainage abnormalities. In the first scenario (Figs 15–17 and Videos 16–18), we describe an infant who was born with heterotaxy (right atrial isomerism); Fig 15 shows a midline liver with asplenia, and Figs 16 and 17 reveal an infant with an interrupted inferior vena cava with azygous continuation. Cine loops from the same patient show the complex intracardiac anatomy, consisting of bilateral right atrial appendages that are broad and triangular in contrast to the left atrial appendages that are finger-like (Video 16). Video 17 shows a cine en-face view of a common atrioventricular valve, and Video 18 shows a bilateral morphologic right atria with a common atrioventricular valve, large inlet ventricular septal defect, and transposition of great vessels with anomalous venous return to the right-sided atrium. In the second scenario (Videos 19–21), Video 19 shows a newborn with tricuspid atresia and a dilated mitral valve, which opens into a morphologic left ventricle, whereas the right ventricle is hypoplastic. Video 20 is on the other end of this spectrum because it shows a newborn with mitral atresia and a hypoplastic left ventricle. A case of severe Ebstein anomaly (Video 21) involves a newborn with severe enlargement of the right atrium and an atrialized right ventricle, leaving a small functional part of the right ventricle. (29)

Figure 16. Coronal cardiac magnetic resonance image in same patient in Fig 15 showing an interrupted inferior vena cava and azygous continuation.

Figure 17. Coronal cardiac magnetic resonance image in same patient in Fig 15 showing an azygous vein draining into the superior vena cava (SVC).

Tissue Characterization and Viability Imaging

CMRI has the capability of distinguishing between tissues by exploiting the variability in returned magnetic signal, depending on the water and fat content of the different tissues. Its applicability in the neonatal period is small, but it is used extensively in older individuals. Its use has extended to patients with primary or secondary intracardiac tumors, such as rhabdomyoma, fibroma, leiomyoma, and teratoma. (30–35) Figure 18 shows a black blood image of a mass (later confirmed to be a papillary
fibroelastoma) on the left coronary cusp of the aortic valve, and the corresponding Video 22 shows a dense mass at the same position. Delayed enhancement after use of gadolinium is suggestive of injury that results in myocardial fibrosis, especially in this patient who has a history of myocarditis (Fig 10).

Technical Considerations
The CMRI and CTA have their own individualized features that make them suitable for different situations. The Table summarizes the key differences between the CTA and CMRI.

CMRI. The advantages of CMRI include better temporal resolution, quantitative volumetric and functional

Video 15. Steady-state free-precession cine image of the same patient in Fig 14 showing the ventricular septal defect flow. Click here to play the video.

Video 16. In the infant born with heterotaxy, cine loops show the complex intracardiac anatomy. Click here to play the video.

Video 17. In the same infant, a cine en-face view of a common atrioventricular valve is shown. Click here to play the video.

Video 18. In the same infant, bilateral morphologic right atria are shown. Click here to play the video.

information, lack of radiation exposure, and tissue characterization for special purposes, such as fibrosis or tumor differentiation. The disadvantages are need for sedation or general anesthesia, intubation, and possibly complete muscle relaxation to achieve breath-holding and avoid artifact; longer study duration; risk of hypothermia; higher cost; need for experienced personnel; and inability to scan
patients with certain stents, pacemakers, and other implantable devices.

**CARDIAC CTA.** The advantages of cardiac CTA include better spatial resolution; excellent extracardiac vascular anatomical delineation; short study duration (12–15 seconds), obviating the need for sedation; better cost profile; excellent coronary artery evaluation; and less prone to artifact, such as motion or breathing artifact. A disadvantage is exposure to ionizing radiation.

**Conclusion**

Advanced diagnostic modalities, such as CMRI and CTA, are used extensively in patients with CHD for both
preoperative and postoperative evaluation. It is important to understand the advantages and limitations of this technology and choose accordingly for optimum results. Currently, there are center-specific variations in the use of this technology in neonates. In our experience, we use CTA in the neonatal period predominantly for extracardiac anatomical delineation, whereas we reserve CMRI for older children. In neonates, the use of CMRI is limited to complex lesions that involve deciphering both intracardiac and extracardiac anomalies in the setting of other associated thoracic or abdominal malformations. We have found this approach to be an accurate, reproducible, and cost-effective practice with few adverse effects to the neonates involved.

Table. Key Differences Between the CTA and CMRI

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<th>Variable</th>
<th>CTA</th>
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<td>Study duration</td>
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<td>Functional information</td>
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<tr>
<td>Patient with stent or pacer</td>
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CTA=computed tomography angiography; CMRI=cardiac magnetic resonance imaging; ECG=electrocardiography; MRI=magnetic resonance imaging.

American Board of Pediatrics Neonatal-Perinatal Content Specifications

- Know the appropriate techniques to assess cardiovascular function in the fetus and newborn infant.
- Recognize the laboratory, imaging, and other diagnostic features of a cyanotic neonate.
- Recognize the laboratory, imaging, and other diagnostic features of a neonate with a left-sided cardiac obstructive lesion.
- Recognize the laboratory, imaging, and other diagnostic features of a neonate with a right-sided cardiac lesion.

References


Parent Resources from the AAP at HealthyChildren.org
• http://www.healthychildren.org/English/health-issues/conditions/heart/Pages/Common-Heart-Defects.aspx (English only)

**Answer Key for June 2015 NeoReviews:**
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