

A Practical Guide to Pediatric Coronary Artery Imaging with Echocardiography

Lynne M. Brown, BA, RDCS (AE) (PE), FASE, C. Elise Duffy, MBBS, FRACP,
Carol Mitchell, PhD, RDMS, RDCS, RVT, RT(R), FASE, FSDMS, and
Luciana Young, MD, FACC, FASE, *Chicago, Illinois; and Madison, Wisconsin*

Few resources exist to educate cardiac sonographers regarding proper techniques and specific issues to consider when performing pediatric coronary artery imaging. The main objective of this report is to review the echocardiographic techniques used to image the coronary arteries of children when screening for anomalous origin of the coronary arteries, as well as coronary involvement in the setting of Kawasaki disease. The authors discuss the physics and instrumentation for developing optimal coronary artery imaging system settings and present the commonly used anatomic and echocardiographic views. Use of the correct settings and a thorough understanding of the associated ultrasound physics are crucial for obtaining quality images. With this report, the authors provide guidance to sonographers and a resource for pediatric echocardiography laboratories to help ensure high-quality echocardiographic imaging of the coronary arteries. (*J Am Soc Echocardiogr* 2015;28:379-91.)

Keywords: Coronary artery, Ultrasound imaging, Coronary artery aneurysm, Kawasaki disease, Pediatric echocardiography

Current pediatric echocardiographic guidelines published by the American Society of Echocardiography list imaging of the coronary arteries as a standard component of a pediatric echocardiogram.¹ Numerous reports have described the assessment of abnormal origins of coronary arteries using echocardiography and the association between abnormal coronary artery origins and an increased risk for sudden cardiac death.²⁻¹¹ Pediatric patients presenting with syncope, chest pain, exercise-induced arrhythmias, and Kawasaki disease undergo detailed imaging of the coronary artery origins. Additionally, patients with Kawasaki disease undergo serial echocardiographic examinations to assess the progression of disease as it relates not only to the coronary artery origins but also to the more distal segments. Although studies have proved that coronary artery imaging in children by echocardiography is feasible and reproducible,^{2,3,6,9} it poses a particular set of challenges for sonographers.

There are few resources that comprehensively describe the techniques and views needed to obtain high-quality diagnostic images of

the coronary arteries in children.^{4,12} An important place to begin with any echocardiographic examination is the use of proper system settings. In this review, our objective is to first discuss normal coronary artery anatomy, followed by the physics and instrumentation for developing optimal coronary artery imaging settings, and to then describe the diagnostic echocardiographic views of the coronary arteries specifically as they relate to abnormal origins of the coronary arteries and Kawasaki disease. Just as pediatric echocardiography is a subspecialty of echocardiography, pediatric coronary artery imaging can be considered a subspecialty of pediatric echocardiography. Our overall goal is to provide sonographers and pediatric echocardiography laboratories that perform coronary artery imaging with a resource that will help ensure the successful imaging of these small yet significant structures in children.

UNDERSTANDING CORONARY ARTERY ANATOMY

Accurate imaging and diagnosis rely on a familiarity with cardiac anatomy, specifically the normal arrangement of the coronary arteries. Random searching is not usually a successful strategy when imaging the coronary arteries, and in the interest of timeliness, which is especially important in the pediatric setting, knowing where normal coronary arteries should be will reduce the time needed to complete an examination.

The standard echocardiographic views will not provide all of the information required to rule out potential abnormalities, and nonstandard echocardiographic views are routinely used to identify coronary origins and to follow the course of the vessels around the heart. Sonographers should thus have a solid understanding of normal coronary artery branching and course over the surface of the heart. Heart models, pathologic specimens, anatomy textbooks, cardiac angiograms, or cardiac computed tomographic three-dimensional reconstructions are helpful resources that can give context to what a sonographer is trying to capture with two-dimensional (2D) echocardiography. Once

From the Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, Illinois (L.M.B., C.E.D., L.Y.); and the Section of Cardiovascular Medicine, University of Wisconsin Atherosclerosis Imaging Research Program, Madison, Wisconsin (C.M.).

Reprint requests: Lynne M. Brown, BA, RDCS (AE) (PE), FASE, Ann & Robert H. Lurie Children's Hospital of Chicago, 225 E Chicago Avenue, Box 287, Chicago, IL 60611 (E-mail: imbrown@luriechildrens.org).

Attention ASE Members:

ASE has gone green! Visit www.aseuniversity.org to earn free CME through an online activity related to this article. Certificates are available for immediate access upon successful completion of the activity. Non-members will need to join ASE to access this great member benefit!

0894-7317/\$36.00

Copyright 2015 by the American Society of Echocardiography.

<http://dx.doi.org/10.1016/j.echo.2015.01.008>

Abbreviations

ALCAPA = Anomalous origin of the left coronary artery from the pulmonary artery
AV = Atrioventricular
LAD = Left anterior descending coronary artery
LCX = Left circumflex coronary artery
LMCA = Left main coronary artery
PDA = Posterior descending coronary artery
PLAX = Parasternal long-axis
PSAX = Parasternal short-axis
RCA = Right coronary artery
2D = Two-dimensional

normal anatomy is mastered, additional study of abnormal arrangements will prepare the sonographer for atypical presentations. Taking the time to understand this anatomy in advance will reduce the amount of time spent and level of frustration when performing the echocardiographic examination.

Coronary arteries have corresponding coronary veins that, because of their often larger diameters, tend to be more prominent (especially in the posterior interventricular sulcus). The coronary veins may also be affected in Kawasaki disease, which can make them even more conspicuous. Assessment of vessel flow is helpful in differentiating the arteries from the veins. The direction of coronary artery flow is

away from the aorta and base of the heart (predominantly during diastole), whereas coronary vein flow is toward the coronary sinus and base of the heart (predominantly during systole). A quick check of vessel flow using color Doppler is prudent before acquiring images that may result in inaccurate diagnoses (Figure 1).⁵ Coronary vein walls are thinner than coronary artery walls, which can also be helpful in differentiating the two structures.⁴

The following sections provide strategies to obtain quality diagnostic echocardiographic images and a brief overview of the potential anomalous and aberrant coronary artery findings during a pediatric echocardiographic study. It is important to remember that coronary artery imaging takes time and skill, and not every study will produce all of the images presented in this report.

OPTIMIZING SYSTEM SETTINGS FOR PEDIATRIC CORONARY ARTERY IMAGING

When imaging the coronary arteries in children, it is important that sonographers apply their knowledge regarding ultrasound physics and system optimization. In 2D imaging, the best resolution is obtained when the coronary vessel is perpendicular to the beam (axial resolution),⁴ while color and spectral Doppler quantification of flow velocities are most accurate when the beam is parallel to flow.¹³ Sonographers should continually strive to optimize images by referencing these basic principles.

Imaging of the coronary arteries should be performed with the highest frequencies possible relative to patient size. The use of higher frequencies (i.e., 8, 10, or 12 MHz) provides superior grayscale resolution and should be readily accessible during coronary imaging.^{13,14} Depending on the imaging system, sonographers may need to change transducers multiple times during a study to obtain the best images. High-frequency transducers should not be reserved for neonates and small children, as they are also useful in larger patients when imaging in the near field.

Frame rate is an important factor in all ultrasound imaging, especially for imaging coronary arteries. Because the coronary vessels are small, superficial structures, and heart rates in children are typically higher

than in adults, temporal resolution is a priority.^{4,15} Reducing depth and sector size has the greatest impact on improving frame rate. To obtain detailed information, sonographers should also zoom in on the structure or increase the magnification. However, enlarging images that have not been properly optimized does not improve their resolution and should be avoided.¹⁵ The focal zone length should be adjusted and placed appropriately. Beam-width artifacts can be minimized by proper placement of the focal zone at the site of interest.¹⁶

Harmonics and compression should be used sensibly, and both of these imaging adjustments are particularly important in the setting of Kawasaki disease. Sonographers should exercise sound judgment on the basis of an understanding of the benefits and pitfalls of harmonics and increased compression with regard to imaging and resolution. Harmonics degrade axial resolution, particularly in the near field, and may give 2D structures a “thickened” appearance.¹³ Consequently, the coronary artery wall may appear falsely abnormal. Bright, echogenic walls may be seen in the acute phase of Kawasaki disease, but this finding can be exaggerated with the use of harmonics. On the other hand, harmonics may be helpful in clarifying coronary artery origins in larger patients or those with technically challenging acoustic windows. Excessive compression may actually filter out low-level sound waves that might otherwise identify a subtle abnormality such as a thrombus within the vessel lumen.¹⁵ Alternately, when measuring the internal lumen of coronary arteries in Kawasaki disease, gain can be judiciously lowered and compression increased to achieve a more black-and-white image to help define the intima-lumen interface in better detail.⁴

Color Doppler settings should also be carefully optimized, as the flow in coronary arteries is subtle and often difficult to capture. A Nyquist limit of 30 cm/sec may be used as the starting point for color Doppler interrogation; however, some situations may require a lower scale. The actual value of the Nyquist limit is not as important as the quality of the Doppler signal. A systematic approach of gradually lowering the limit until flow is detected while avoiding indistinguishable artifacts can help achieve diagnostic accuracy. Sonographers should use their judgment, remembering that a lower scale with a suboptimal angle of insonation may increase the potential for artifact. Noise in the color flow electronics used to produce the color Doppler signals can appear as artificial “flow” in hypoechoic areas, and clutter from tissue motion can obscure the true underlying blood flow.¹⁵ These artifacts are often mistaken for coronary artery blood flow, offering yet another reason why the angle of the ultrasound beam should be as parallel to flow as possible (Figure 1C). This may require acquisition of a 2D image in one view and a color Doppler image in an alternative view. Higher frequency transducers may not be sensitive enough to demonstrate color Doppler flow in coronary imaging, and attempts should be made to use lower frequency transducers if this information is the focus of the examination. Vascular studies use transducers with slightly lower frequencies, as echoes returning from blood flow are weaker than those from anatomic imaging.¹⁵ The same principles apply to echocardiography, and further support the use of multiple transducers to achieve the highest quality images. Small adjustments to persistence may also be helpful if color flow imaging is difficult to obtain.⁶ Persistence averages individual frames, which reduces noise and results in a smoother image.¹⁵ Flow detection in the right coronary artery (RCA) can be challenging to demonstrate using the standard parasternal short-axis (PSAX) view in healthy patients due to the perpendicular angle of insonation. Flow may appear more prominent if there is pathophysiology present that increases flow velocity.⁶

Image acquisition should include moving clips followed by still images. Still images alone do not provide adequate diagnostic

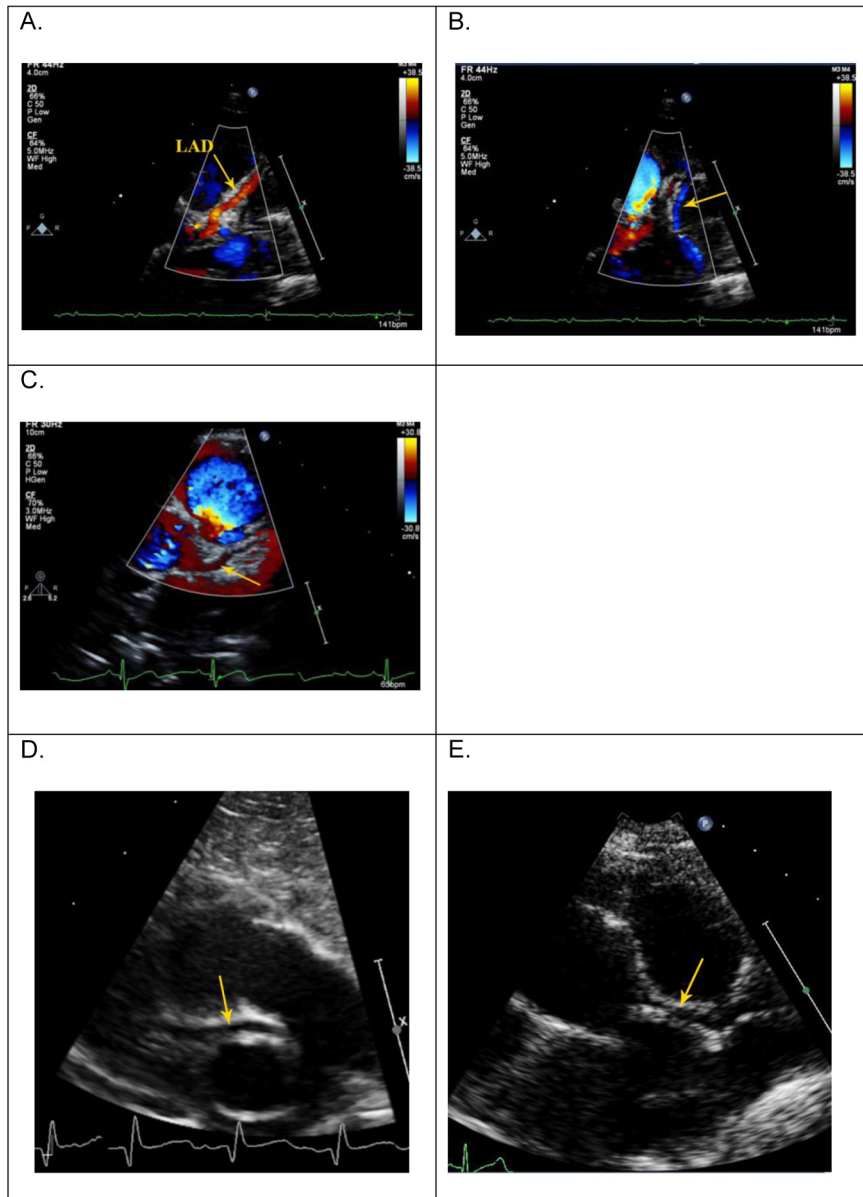


Figure 1 Example of a normal color Doppler signal in the left anterior descending coronary artery (LAD) and left circumflex coronary artery during diastole (**A**) and the great cardiac vein during systole (**B**). Left coronary artery flow during systole (**C**) along with color artifact in the surrounding tissue provides images that are not diagnostically convincing. Small amounts of fluid in the pericardial sinuses near the RCA (**D**) and left (**E**) coronary arteries can be mistaken for coronary vessels. See [Videos 1A, 1B, and 1C](#) (available at www.onlinejase.com).

information and may bias the interpreting physician into making a false diagnosis.¹⁷ Moving images provide a contextual reference in relation to other cardiac structures and capture additional information that might be overlooked when concentrating on specific anatomy. The reading physician does not have the benefit of hands-on scanning and thus relies on the sonographer to provide as much information as possible so that an accurate diagnosis can be made.

An electrocardiographic tracing is necessary to correctly assess coronary artery blood flow. Left coronary artery flow occurs predominantly in diastole,¹² while RCA flow occurs in both diastole and systole.^{18,19} Comparing the color Doppler flow to the electrocardiographic tracing will confirm timing of flow in the coronary arteries. Coronary artery flow reversal in any part of the

cardiac cycle is abnormal and should increase suspicion of a potentially serious anomaly, necessitating a thorough investigation of the anatomy.

Coronary arteries are very small superficial structures that may be difficult to image in any patient. The technical issues are further confounded by the fact that children, especially those in the painful, acute phase of Kawasaki disease, may not tolerate lying still for extended periods of time. If concerns exist regarding the coronary artery anatomy for any reason, sedation may be required to obtain the best diagnostic images. If the coronary arteries are not adequately visualized with echocardiography, other imaging modalities (i.e., computed tomography, magnetic resonance imaging, or cardiac catheterization) may need to be considered.

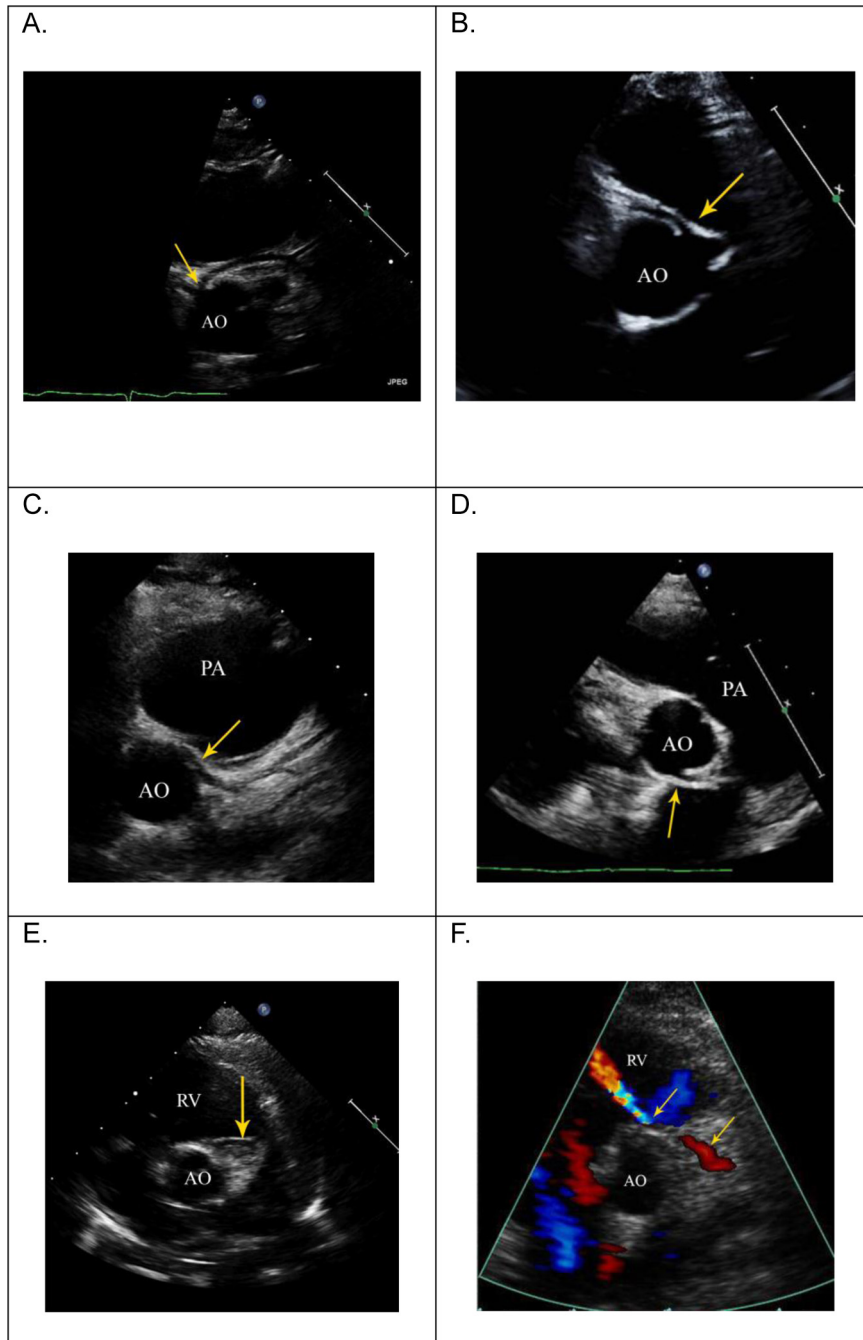


Figure 2 Example of a single coronary origin from the right sinus of Valsalva (**A**), right coronary artery from the left sinus of Valsalva (**B**), and left main coronary artery (LMCA) from the right sinus of Valsalva with acute angle of the arterial course (**C**). Note that the origin is not perpendicular to the left sinus of Valsalva. (**D**) A rare case of LMCA from the “noncoronary” sinus of Valsalva and (**E**) an intramyocardial course of the left coronary artery (image courtesy of Alaina Kipps). (**F**) A coronary artery fistula from the left anterior descending artery to the right ventricle (RV). AO, Aorta; PA, pulmonary artery. See [Videos 2A, 2B, and 2C](#) (available at www.onlinejase.com).

ANOMALOUS OR ABERRANT CORONARY ARTERIES

Although the incidence of anomalous or aberrant¹⁰ coronary arteries is rare, 0.6% to 1.3% as reported in several series,²⁰ the risks associated with this diagnosis can be serious. Congenital coronary anomalies are the second most common underlying cause of sudden cardiac death or arrest in athletes (15%–25% of cases),

with hypertrophic cardiomyopathy being the most common.²¹ Exertional chest pain, syncope, and exercise-induced arrhythmias in children may be caused by coronary artery abnormalities.^{3,5,17,20,22} Echocardiography is a low-risk procedure that has been proven successful in diagnosing these anomalies.²⁻¹⁰ The coronary abnormalities that are most often seen are anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA),

aberrant origin of the coronary artery from the incorrect sinus of Valsalva, single coronary artery, and coronary artery fistula (Figure 2). Aberrant coronary arteries that have interarterial, intramural courses within the wall of the aorta are of great concern.²¹ Intramural segments can be difficult to visualize and easy to miss without a careful assessment of coronary origins, and confirmation of an intramural segment may be possible only by direct inspection. The course of the proximal coronary segment is usually perpendicular to the aortic sinus, and unusual orientations should be highly suspect. Acute takeoffs of either coronary artery from the opposite coronary sinus of Valsalva with an intramural course and ostial valve-like ridge have also been associated with sudden death (Figure 2C).^{11,21-23} Although the significance of an intramyocardial course of the left coronary artery is somewhat controversial, the risk for sudden death with this anomaly is considered low (Figure 2E).^{20,21}

Superior origin of the RCA from the ascending aorta is considered a normal variant; however, this finding is important to note before surgery to avoid accidental damage or clamping of the vessel.²⁰ Incidentally, a superior origin of the RCA may be associated with abnormal origin and should be carefully inspected.¹⁷ The origin of the RCA may even “straddle” the imaginary line of the intercoronary commissure, and therefore careful sweeps should be performed. A left circumflex coronary artery (LCX) that arises from the RCA is also considered a normal variant, as are separate origins of the LCX and left anterior descending coronary artery (LAD) from the left sinus of Valsalva and separate origins of the RCA and right conal branch from the right sinus of Valsalva. An aberrant origin of either the left or both coronary arteries from the noncoronary sinus (posterior sinus) is extremely rare but has also been reported in association with sudden cardiac death (Figure 2D).^{11,24,25}

ALCAPA does not present with a predictable pattern imaged with specific echocardiographic views. Absence of a left coronary artery ostium or presence of flow reversal are both possible findings. In older patients, the RCA may be notably dilated. Infants who present with severe left ventricular dysfunction and mitral regurgitation of unknown etiology should be carefully screened for ALCAPA.¹⁷ In the presence of pulmonary hypertension and ALCAPA, color flow in the left coronary artery may appear to be antegrade. The use of transient supplemental oxygen to lower pulmonary vascular resistance during echocardiography has been reported to enhance flow into the main pulmonary artery and reversal of flow in the left coronary system.²⁶ The rarer variant of anomalous origin of the RCA from the pulmonary artery is usually diagnosed later in life, and patients are typically asymptomatic.¹⁷ With both anomalies, collateral arteries are usually seen within the ventricular septum but prominence may vary depending on the age at presentation.¹⁷ An LCX from the pulmonary artery is very rare but should also be assessed in a symptomatic patient.²⁷ If echocardiography cannot delineate the coronary origins in these patients, other imaging modalities should be considered in case urgent repair is needed. A rapid diagnosis can have a tremendous impact on improving the outcomes of these patients.

Coronary artery fistulas are rare and usually asymptomatic in childhood; however, presentation with heart failure is possible. Treatment is controversial for small silent shunts, but continued follow-up is recommended (Figure 2F).²⁸ Because of their unpredictable course, echocardiographic assessment of fistulas is similar to the techniques used for Kawasaki disease. Nonstandard views should be incorporated into the examination to follow the course of the fistula and to assess for vessel dilatation in other coronary segments.

ECHOCARDIOGRAPHIC VIEWS USED TO ASSESS CORONARY ARTERY ORIGINS

Unique imaging techniques and nonstandard views are needed to visualize the various coronary segments and origins. The coronary arteries are visualized primarily from the PSAX view, just superior to the aortic valve,¹⁷ and from the parasternal long-axis (PLAX) view (Table 1). The origins can also be imaged from the apical²⁹ and subcostal views if the parasternal views are not adequate. Additional views are used to map more distal segments of each vessel, and further details will be discussed later in relation to coronary artery assessment in Kawasaki disease.

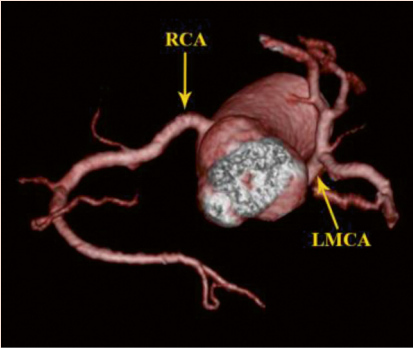
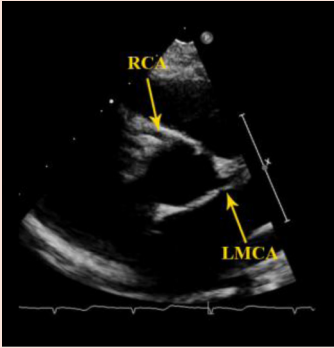
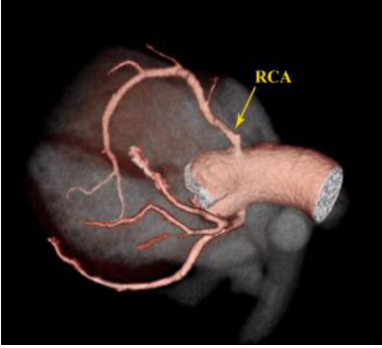
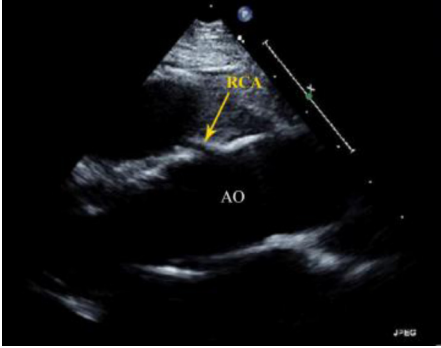
The PSAX view at the level of the aortic valve is standard for identification of the coronary artery origins and is an important component of any pediatric protocol (Table 1, row A). Diagnosing abnormal origins of the coronary arteries is possible in this view because of the clear delineation of right versus left (which is not possible in the PLAX view). Coronary arteries originate above the level of the aortic valve and therefore will not be visualized in the same plane as the aortic leaflets. Imaging sweeps may be necessary to prove which aortic sinus is related to each coronary artery origin. Slight rotation (clockwise for left and counterclockwise for right) and/or angulation to the left or right is frequently required to image the coronary artery origins³⁰ (Table 1, row A). Sonographers should ignore intracardiac anatomy and focus on the coronary vessels when obtaining these images. These are superficial anterior views and will not be in the standard planes of imaging. Scanning through the interarterial space with a superior-to-inferior angulation is fraught with pitfalls due to small amounts of fluid in the transverse sinus of the pericardial sac (Figure 1D). The echo-free space between the pericardial tissue and the left atrial appendage can be mistaken for the left coronary artery (Figure 1E).³¹ Although both origins may sometimes be seen in the same plane, sonographers should interrogate each origin individually, as subtle defects are easy to miss. When abnormal origins are detected, orthogonal imaging should also be performed to better demonstrate the slitlike opening, which might not be appreciated otherwise.

In the standard PLAX view, the RCA is usually visualized; however, this view is not sufficient to diagnose abnormal coronary arteries from the opposing sinus, as the orientation is superior/inferior rather than left/right (Table 1, row B). The benefit of this view is that it can identify a superior origin of the RCA from the ascending aorta, above the sinotubular junction. This angle is also ideal for color Doppler interrogation of the RCA, as it is the view most parallel to flow. If a coronary vessel is seen in this view as a discrete circle in cross-section, an abnormality should be suspected until ruled out.³²

KAWASAKI DISEASE

The views discussed thus far are just the beginning of a complete assessment of the coronary artery system—patients with Kawasaki disease require a more comprehensive inspection to rule out potentially serious sequelae. Kawasaki disease is a childhood vasculitis of unknown etiology, which may be associated with coronary artery ectasia and aneurysms.¹⁴ Ectasia refers to the pathologic dilatation of the coronary vessels and does not imply irregularity in the vessel lumen.¹⁴ Aneurysms can be saccular when axial and lateral diameters are nearly equal, or fusiform if the dilatation is symmetric with gradual tapering on the proximal and distal ends of the affected segment.¹⁴

Table 1 Routine views to obtain in the standard pediatric echocardiographic examination to assess coronary artery origins

A.	Coronary CT image 	Coronary echocardiogram 
<p>PSAX view, just above the level of the aortic valve leaflets. The origins of the RCA and left coronary artery are often not seen in the same plane and will need to be interrogated independently. Rotating clockwise for the LMCA and counterclockwise for the RCA is often helpful.</p>		
B.	Coronary CT image 	Coronary echocardiogram 
<p>PLAX view, right coronary origin. This view is helpful to define a superior origin of the RCA from the ascending aorta but will not accurately diagnose other abnormal origins, because of the lack of a right and left reference.</p>		

CT, Computed tomographic.

Corresponding CT images are included for anatomic correlation.

Affected coronary vessels can develop proliferation of the intimal wall (Figure 3A) that may lead to stenosis, ischemia, and myocardial infarction. Additionally, in larger aneurysms that develop quickly, there is high risk for thrombus formation that can also interrupt blood flow, leading to the same outcome (Figure 3C).¹⁴ Serial echocardiography and measurements are required to follow the progression of the disease as it relates to the coronary arteries. The presence of echo-bright coronary arteries and perivascular tissue has also been reported, although detection by echocardiography is not well validated.^{14,33}

In Kawasaki disease, the most common sites of coronary artery aneurysms, in decreasing frequency, are the proximal LAD, proximal RCA, left main coronary artery (LMCA), LCX, junction between the RCA and posterior descending coronary artery (PDA), and junction of any marginal branch (Figure 4).^{4,14} Careful measurement of the coronary artery lumen should be made from inner edge to inner edge (Figure 5) while avoiding measurements at branch points, as these areas may be slightly larger, even in normal patients. To assist in diagnosing coronary artery ectasia and aneurysm, body surface

area-adjusted Z scores are available for the proximal RCA, LMCA, and proximal LAD.³⁴

Patients with Kawasaki disease may lack the tapering seen in normal coronary arteries.¹⁴ Dilated coronary arteries are more conspicuous and, as a result, usually easier to image. If coronary arteries are difficult to image when the acoustic windows are satisfactory, suspicion of coronary artery dilatation is low. When the coronary arteries are dilated, careful assessment of the internal lumen for thrombus formation is required. Thrombi are subtle to detect, and the use of too much compression, or harmonics in the near field, may effectively erase the low-level sound waves reflecting from these masses. Color Doppler may be helpful to delineate flow around a thrombus, as well as to rule out stenotic segments resulting from myointimal proliferation that may be missed by 2D echocardiographic imaging.⁶ There should be a suspicion for coronary artery stenosis if flow disturbances are noted with color Doppler (Figure 3B).

Variation exists in labeling coronary segments in relation to Kawasaki disease.^{4,14,35,36} We describe the following seven segments (Table 2): LMCA, LAD, LCX, RCA proximal segment,

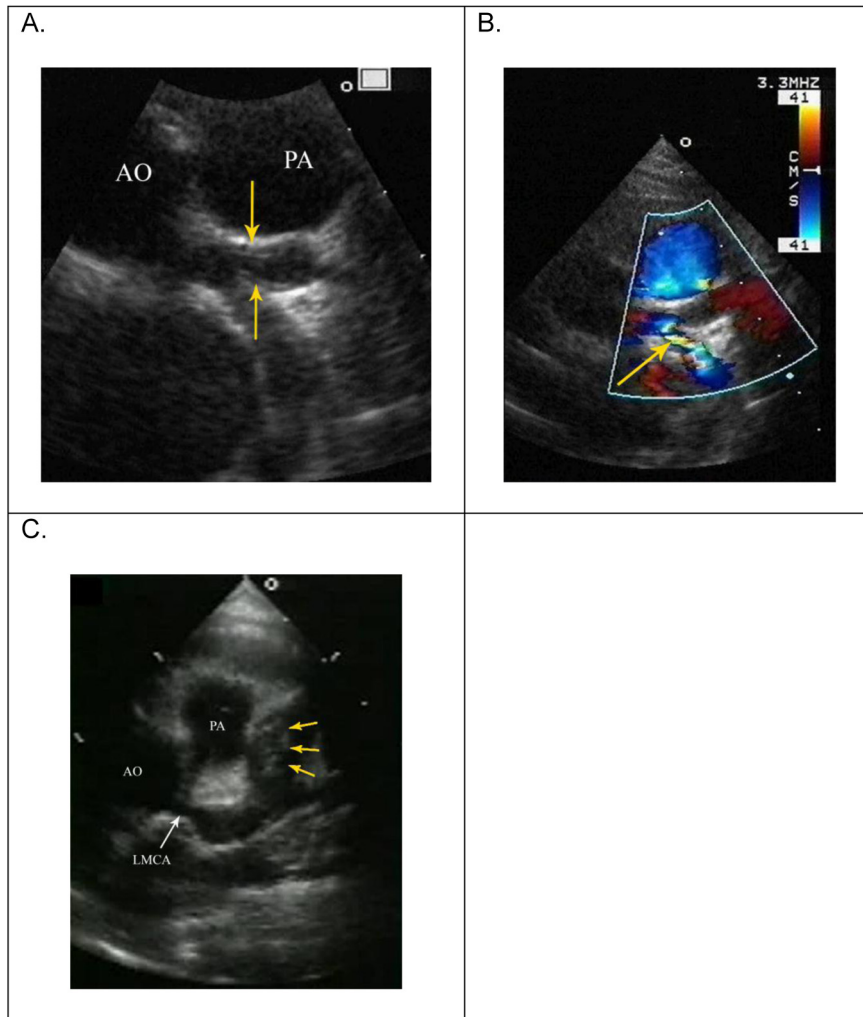


Figure 3 Thickening of the coronary artery luminal wall within an aneurysmal coronary artery in a patient with Kawasaki disease, presumably due to myointimal cellular proliferation (**A**). Turbulent color Doppler signal in the left main circumflex coronary artery, even with color Nyquist limit of 41 cm/sec, suggests stenosis (**B**). A severely dilated left anterior descending coronary artery with thrombus formation in the midsegment (**C**). AO, Aorta.

RCA middle segment, RCA distal segment, and PDA. Detailed interrogation of all coronary artery segments should be attempted on each echocardiographic examination.¹⁴ Sedation may be necessary, particularly in pediatric patients, who may find it difficult to remain still for prolonged periods, so that a comprehensive evaluation can be performed. Serial imaging of all segments of the coronary system requires skill and critical thinking on the part of the sonographer. Complete coronary artery assessment can be challenging, and having a checklist of views and segments to include in the examination can be helpful.

ECHOCARDIOGRAPHIC SCANNING PROTOCOL AND CRITICAL THINKING STRATEGY FOR IMAGING THE CORONARY ARTERIES IN PATIENTS WITH KAWASAKI DISEASE

In addition to the proximal coronary segments, which are the most common location for aneurysms, a survey of as much of the coronary system as possible should be attempted in each Kawasaki study. This

should be performed on the initial echocardiographic examination to provide a baseline and on each follow-up study to assess ongoing changes in the vasculature. The views that follow will provide opportunities to uncover any concealed pathology related to the coronary arteries.

After locating the coronary origins in the PSAX view, slight rotational modifications will provide more distal imaging of the RCA and left coronary artery (Table 2, rows A and B). Starting at the standard PLAX view and pointing the transducer anterolaterally and toward the left axilla (also referred to as the superior tangential view)¹⁴ will provide a window perpendicular to the course of the LMCA and LAD (Table 2, row C). This view focuses on the area between the aorta and pulmonary artery. Because of the anterior-to-posterior direction of the sound beam, the proximal LCX is often best seen in this view and provides the most parallel orientation for color Doppler interrogation. The sonographer should avoid rotating counterclockwise in this view, as this will result in a mirror image of the PSAX view instead of a unique long-axis image of the left coronary artery. This angulation is not intended to capture the LMCA origin but is ideal for imaging the more distal portions of the LAD as the sonographer attempts to follow

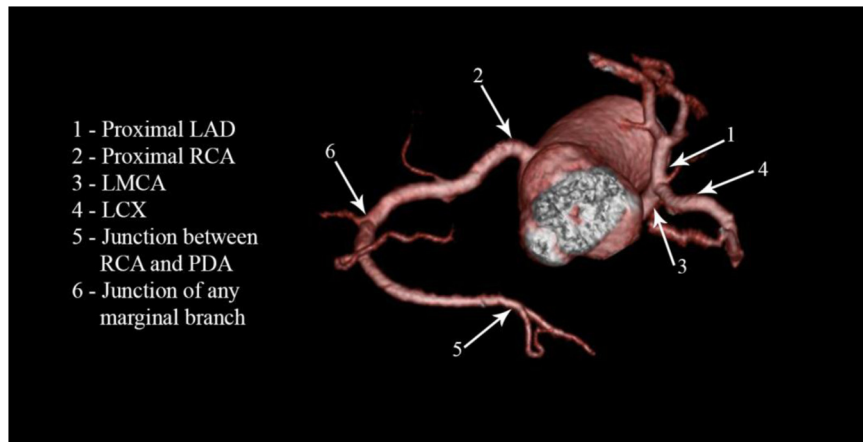


Figure 4 Common locations of aneurysms in patients with Kawasaki disease in decreasing order of frequency. Segments 1 to 5¹⁴ and segment 6.⁴



Figure 5 Proper technique for measuring coronary artery diameter. For best axial resolution, a plane as perpendicular as possible to the vessel should be attempted. Measurements are performed from inner edge to inner edge.

its course toward the apex of the heart and that of the LCX into the lateral atrioventricular (AV) groove. Image acquisition will be in segments, proximal to distal, as it is usually not possible to capture long uninterrupted portions of the coronary arteries in the same plane because of the curvature of the surface of the heart.

By angling the transducer toward the right ventricular inflow tract view (inferiorly toward the patient's right hip), the distal RCA and PDA can be viewed (also referred to as the inferior tangential view; Table 2, row D).¹⁴ Remembering that the distal portion of the RCA resides in the posterior AV groove of the right heart, the transducer beam will capture the RCA between the PLAX and right ventricular inflow tract views. Some rotation of the transducer will be necessary, and this may not be possible on every patient. By angling the transducer even more inferiorly, the sound beam will pass through the inferior surface of the interventricular septum/sulcus, and the PDA with its associated vein can be imaged (Table 2, row D).

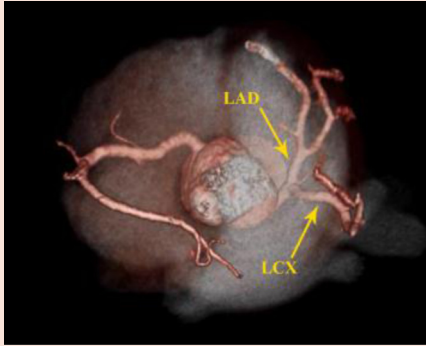
The apical four-chamber view is helpful in assessing both the right and left coronary systems (Table 2, row E). Exaggerated anterior angu-

lation of the transducer, focusing on the apex, may allow imaging of the most distal segments of the LAD. Posterior angulation of the transducer, while sliding medially, will provide an excellent view of both the distal RCA and the PDA (Table 2, row F). The middle cardiac vein in this view is usually the more prominent vessel, and careful attention to direction of blood flow is helpful in distinguishing between vein and artery. The coronary artery will have blood flow coursing toward the apex, which is flowing toward the transducer in this view. Rotating the transducer toward the apical long axis from the standard four-chamber view (clockwise in the pediatric apex-down orientation) and angling anteriorly will usually show a segment of the LCX (Table 2, row G). The retroaortic course of the LCX from the right aortic sinus can be readily visualized from the apical four-chamber view behind the aortic root, and the apical five-chamber view (left ventricular outflow tract) can also be used to identify coronary origins. This view is helpful in patients who do not have adequate parasternal windows. Left- versus right-sided coronary artery dominance will dictate the coronary patterns on the posterior aspect of the heart. The most common coronary artery arrangement is right dominant, with the RCA supplying circulation to the inferior portion of the interventricular septum via the PDA. Patients with left-dominant coronary systems will have lateral and posterior extension of the LCX around the left AV groove, with the artery extending to the right AV groove and then coursing inferiorly to become the PDA. Posterior angulation, focusing on the left AV groove, can provide a glimpse of the distal LCX (Table 2, row H). For this view, using the coronary sinus as a landmark may help in finding the distal LCX; however, in right-dominant systems, the LCX usually begins to dive apically near the obtuse margin of the heart, which complicates the ability to trace the vessel. Even when a posterior vessel is found, differentiating the artery from the vein may be challenging. This imaging plane is most helpful for ruling out ectasia and aneurysms rather than identifying specific coronary segments. A nonstandard right parasternal border view with the patient in a steep right decubitus position can also allow further imaging of the proximal RCA (Table 2, row I).

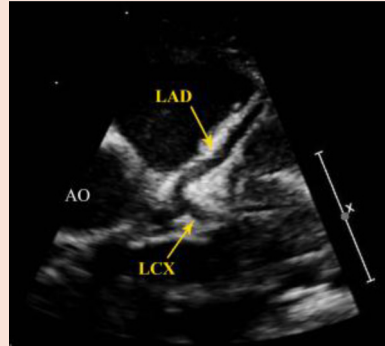
Subcostal imaging can be very helpful for assessing coronary artery origins in challenging patients and is especially useful with neonates; however, distinguishing coronary artery origins from a pericardial reflection is important with the latter. Similar to the PSAX view, careful assessment to differentiate small amounts of fluid in the pericardial

Table 2 Routine views to obtain when imaging a patient with Kawasaki disease to assess for aneurysms and ectasia

A. Coronary CT image

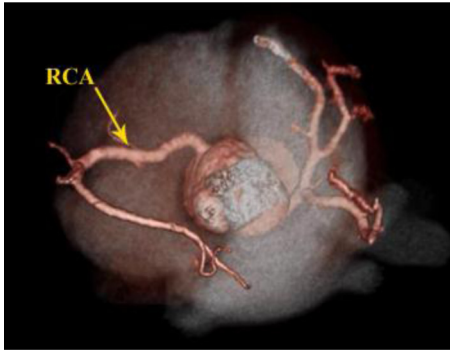


Coronary echocardiogram

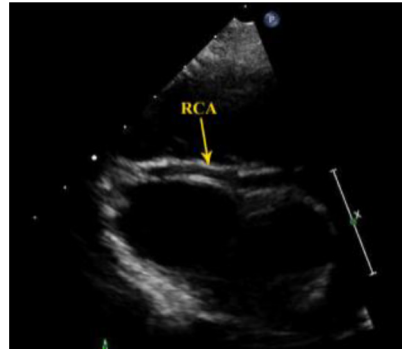


PSAX view rotating clockwise with a slight leftward and anterior angulation following the LAD toward the apex. A portion of the proximal LCX will also be seen.

B. Coronary CT image

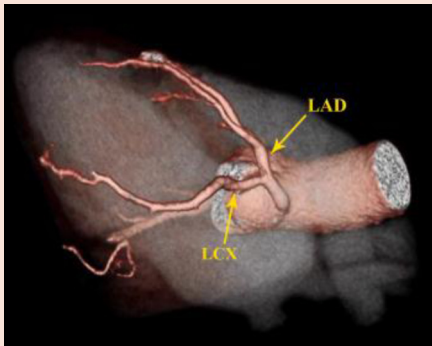


Coronary echocardiogram

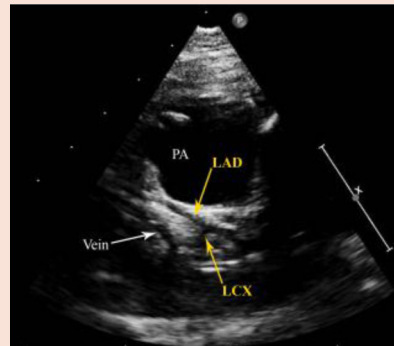


Modified PSAX view following the course of the RCA around the tricuspid annulus. Proximal RCA with a portion of the mid-RCA. Slight counterclockwise rotation of transducer with anterior angulation.

C. Coronary CT image



Coronary echocardiogram

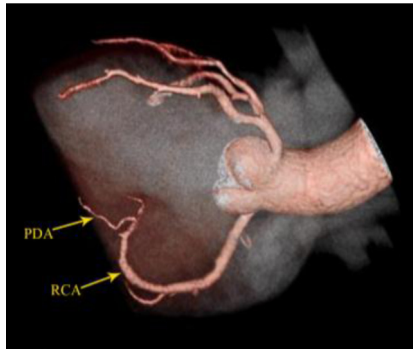


PLAX view angling anterolaterally toward the RVOT and lateral left ventricular wall (superior tangential view). This is an imaging plane between the aorta and pulmonary artery that helps define the more distal LAD and LCX.

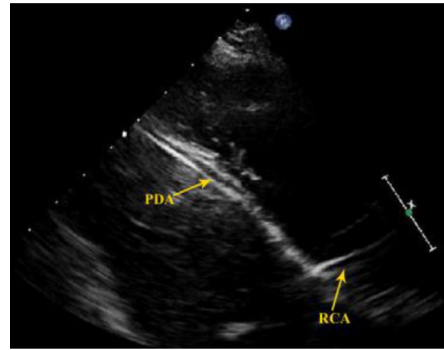
(Continued)

Table 2 (Continued)

D. Coronary CT image

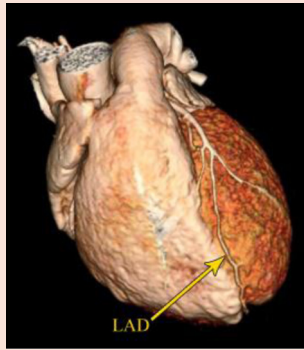


Coronary echocardiogram

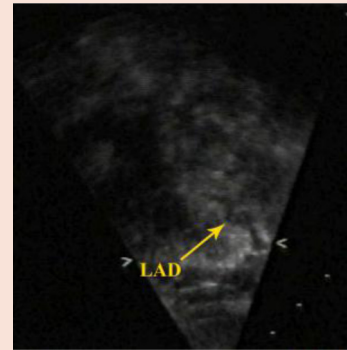


RVIT view (inferior tangential). Distal RCA in posterior right AV groove and PDA in ventricular sulcus. It is difficult to differentiate the middle cardiac vein from the PDA without the use of color Doppler. Coronary veins are usually more prominent because of their larger size.

E. Coronary CT image

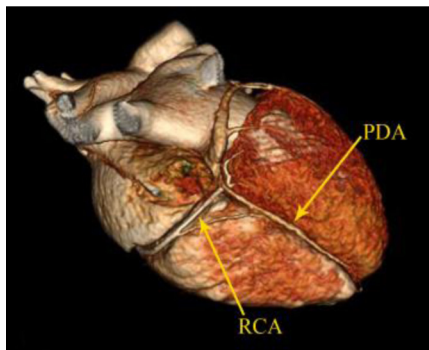


Coronary echocardiogram

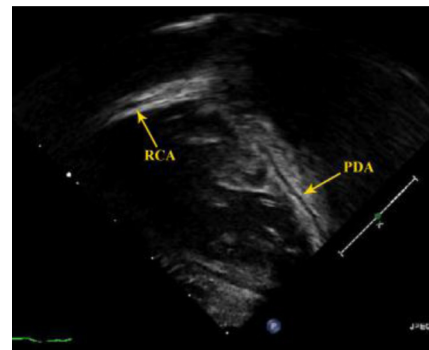


Anterior apical four-chamber view with focus on the apex. Distal segments of the LAD can be imaged in this view. Echocardiographic image shows dilated distal LAD in the setting of Kawasaki disease.

F. Coronary CT image



Coronary echocardiogram



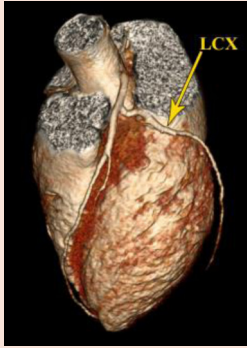
Modified apical four-chamber view with transducer placed more medially and aimed posteriorly. Distal RCA is seen in the AV groove, and a prominent vessel is present in the posterior ventricular sulcus. Again, it is difficult to differentiate the artery from the middle cardiac vein without the use of color Doppler. (CT image reversed to correlate with echocardiogram.)

(Continued)

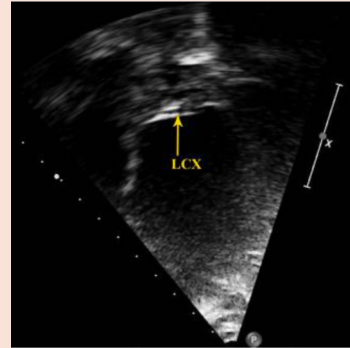
Table 2 (Continued)

G.

Coronary CT image



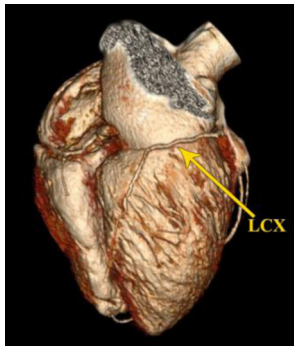
Coronary echocardiogram



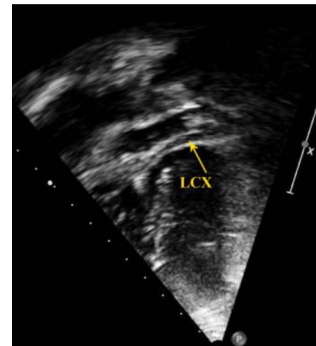
Anterior angulation of the apical long-axis/three-chamber view. A portion of the left LCX can be followed in the left AV groove.

H.

Coronary CT image



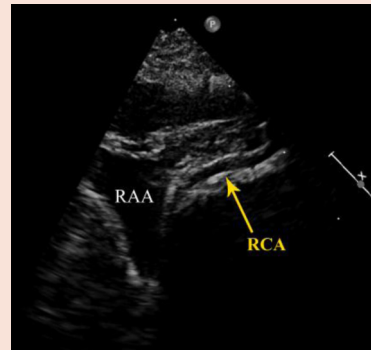
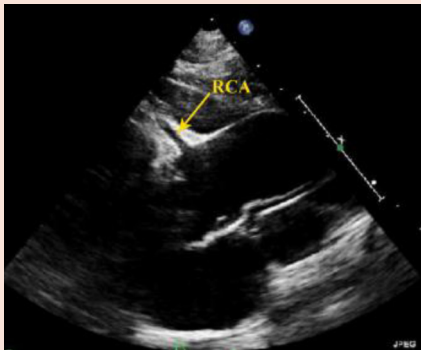
Coronary echocardiogram



Apical four-chamber view angled posteriorly. The left LCX follows the left AV groove around the annulus of the mitral valve. This vessel may travel apically at any point of this course. If the LCX continues to follow the annulus posteriorly, it may be possible to image in this view. The coronary sinus is a good reference point for this vessel. (CT image reversed to correlate with echocardiogram.)

I.

Coronary echocardiogram

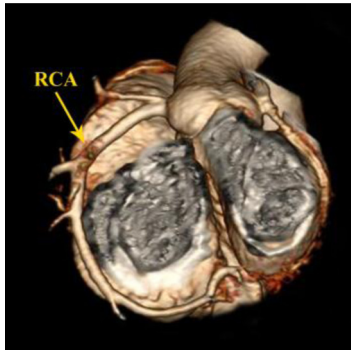


Right parasternal border with patient in right decubitus position. Transducer marker position near 12 o'clock but may require modification. This view, along with the PLAX and/or PSAX view, will often provide a better view of the proximal and mid-RCA. RAA, Right atrial appendage.

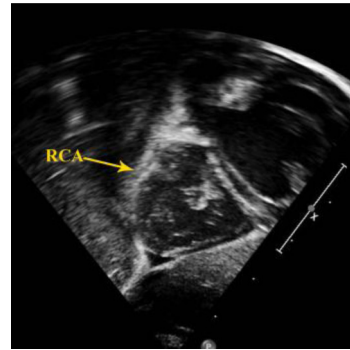
(Continued)

Table 2 (Continued)

J. Coronary CT image

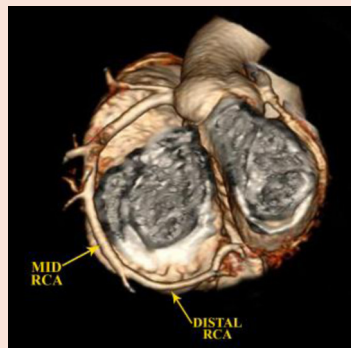


Coronary echocardiogram

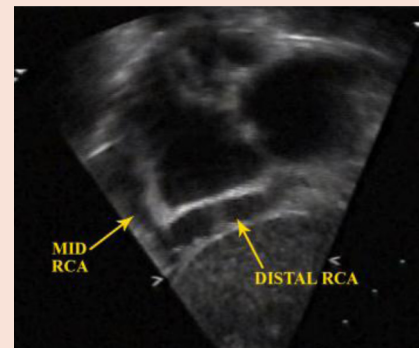


Subcostal view between the long and short axis just posterior to the pulmonary outflow tract. Transducer at a 4:30 rotation. The imaging plane should focus on the tricuspid valve annulus. The proximal segment of the RCA is usually seen in this view.

K. Coronary CT image



Coronary echocardiogram



Subcostal 4:30 view showing the mid and distal segments of the RCA. Although difficult to image in normal patients, this dilated RCA in Kawasaki disease is obvious.

CT, Computed tomographic; RAA, right atrial appendage; RVIT, right ventricular inflow tract; RVOT, right ventricular outflow tract. Corresponding CT images are included for anatomic correlation.

spaces from coronary vessels is important. A nonstandard subcostal view between the long and short axes is obtained by rotating the transducer to approximately a 4:30 marker position. The proximal segment of the RCA is usually easily visualized at the level of the right ventricular outflow (Table 2, row J). The most difficult segment of the coronary anatomy to image is the midportion of the RCA. By focusing on the tricuspid annulus, this view is usually the most successful to use for finding this segment. In normal patients, visualization may be impossible because of the small size of the vessel and the motion of the heart. In patients with grossly dilated coronary arteries, however, clear delineation of the coronary course around the right AV groove is often the case (Table 2, row K).

critical thinking skills and the ability to tailor an examination to answer the clinical question require preparation, study, and skill. A regular practice of obtaining these views in compliant patients with normal cardiac anatomy will increase the chances of success when imaging in more difficult situations. When performed correctly, these studies will assist physicians in making accurate diagnoses and treatment plans and, in most cases, will save patients from undergoing more invasive diagnostic testing.

ACKNOWLEDGMENTS

Special thanks to Brian Reilly, RT(R), for providing three-dimensional computed tomographic renderings.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.echo.2015.01.008>.

CONCLUSIONS

It is important for sonographers to be aware of the optimal system settings, anatomy, and echocardiographic views to provide high-quality diagnostic images of the coronary arteries in children. Application of

REFERENCES

- Lai WW, Geva T, Shirali GS, Frommelt PC, Humes RA, Brook MM, et al. Guidelines and standards for performance of a pediatric echocardiogram: a report from the Task Force of the Pediatric Council of the American Society of Echocardiography. *J Am Soc Echocardiogr* 2006;19:1413-30.
- Frommelt PC, Frommelt MA, Tweddell JS, Jaquiss RD. Prospective echocardiographic diagnosis and surgical repair of anomalous origin of a coronary artery from the opposite sinus with an interarterial course. *J Am Coll Cardiol* 2003;42:148-54.
- Frommelt PC, Berger S, Pelech AN, Bergstrom S, Williamson JG. Prospective identification of anomalous origin of left coronary artery from the right sinus of Valsalva using transthoracic echocardiography: importance of color Doppler flow mapping. *Pediatr Cardiol* 2001;22:327-32.
- Fuse S, Kobayashi T, Arakaki Y, Ogawa S, Katoh H, Sakamoto N, et al. Standard method for ultrasound imaging of coronary artery in children. *Pediatr Int* 2010;52:876-82.
- Frommelt PC, Friedberg DZ, Frommelt MA, Williamson JG. Anomalous origin of the right coronary artery from the left sinus of Valsalva: transthoracic echocardiographic diagnosis. *J Am Soc Echocardiogr* 1999;12:221-4.
- Jureidini SB, Marino CJ, Singh GK, Fiore A, Balfour IC. Main coronary artery and coronary ostial stenosis in children: detection by transthoracic color flow and pulsed Doppler echocardiography. *J Am Soc Echocardiogr* 2000;13:255-63.
- Agarwala B, Ghosh A. 2-Dimensional and color-flow Doppler imaging of coronary anomalies. *Tex Heart Inst J* 2010;37:250-1.
- Thankavel PP, Brown PS, Carron HD, Ramaciotti C. Imaging the coronary artery: is it really normal? How to avoid common echocardiographic pitfalls. *Circ Cardiovasc Imag* 2012;5:415-8.
- Clouse M, Cailles C, Devine J, Jordan M, Lester J, Lo L, et al. What is the feasibility of imaging coronary arteries during routine echocardiograms in children? *J Am Soc Echocardiogr* 2002;15(10 Pt 2):1127-31.
- Jureidini SB, Marino CJ, Singh GK, Balfour IC, Chen SC. Assessment of the coronary arteries in children: an integral part of each transthoracic echocardiographic study. *J Am Soc Echocardiogr* 2003;16:899-900; author reply 900.
- Catanzaro JN, Makaryus AN, Catanese C. Sudden cardiac death associated with an extremely rare coronary anomaly of the left and right coronary arteries arising exclusively from the posterior (noncoronary) sinus of Valsalva. *Clin Cardiol* 2005;28:542-4.
- Jureidini SB, Marino CJ, Waterman B, Syamasundar Rao P, Balfour IC, Chen SC, et al. Transthoracic Doppler echocardiography of normally originating coronary arteries in children. *J Am Soc Echocardiogr* 1998;11:409-20.
- Miele FR. *Essentials of Ultrasound Physics: The Board Review Book*. Forney, TX: Pegasus Lectures; 2008.
- Newburger JW, Takahashi M, Gerber MA, Gewitz MH, Tani LY, Burns JC, et al. Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. *Pediatrics* 2004;114:1708-33.
- Kremkau FW. *Diagnostic Ultrasound: Principles and Instruments*; 6th ed. Philadelphia, PA: W.B. Saunders Company; 2002.
- Scanlan KA. Sonographic artifacts and their origins. *AJR Am J Roentgenol* 1991;156:1267-72.
- Cohen MS, Herlong RJ, Silverman NH. Echocardiographic imaging of anomalous origin of the coronary arteries. *Cardiol Young* 2010;20(Suppl 3):26-34.
- Akasaka T, Yoshikawa J, Yoshida K, Hozumi T, Takagi T, Okura H. Comparison of relation of systolic flow of the right coronary artery to pulmonary artery pressure in patients with and without pulmonary hypertension. *Am J Cardiol* 1996;78:240-4.
- Ofili EO, Labovitz AJ, Kern MJ. Coronary flow velocity dynamics in normal and diseased arteries. *Am J Cardiol* 1993;71:3D-9.
- Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn* 1990;21:28-40.
- Camarda J, Berger S. Coronary artery abnormalities and sudden cardiac death. *Pediatr Cardiol* 2012;33:434-8.
- Maron BJ, Shirani J, Poliac LC, Mathenge R, Roberts WC, Mueller FO. Sudden death in young competitive athletes: clinical, demographic, and pathological profiles. *JAMA* 1996;276:199-204.
- Virmani R, Chun PK, Goldstein RE, Robinowitz M, McAllister HA. Acute takeoffs of the coronary arteries along the aortic wall and congenital coronary ostial valve-like ridges: association with sudden death. *J Am Coll Cardiol* 1984;3:766-71.
- Nishiyama M, Doi S, Matsumoto A, Nishioka M, Hosokawa S, Sasaki A, et al. Exercise-induced myocardial ischemia in a case of anomalous origin of the left main coronary artery from the noncoronary sinus of Valsalva. *Pediatr Cardiol* 2011;32:1028-31.
- Lo PH, Chang KC, Hung JS, Chen HL, Fang CY, Fu M, et al. Anomalous origin of left main coronary artery from the noncoronary sinus: an intravascular ultrasound observation. *Cathet Cardiovasc Diagn* 1997;42:430-3.
- Blanco VM, Blalock SE, Ramaciotti C, Lemler M, Heistein L, Moore J, et al. Oxygen supplementation is helpful for the echocardiographic detection of anomalous left coronary artery from the pulmonary artery. *J Am Soc Echocardiogr* 2010;23:1099-102.
- Page HL, Engel HJ, Campbell WB, Thomas CS. Anomalous origin of the left circumflex coronary artery: recognition, angiographic demonstration and clinical significance. *Circulation* 1974;50:768-73.
- Hsieh KS, Huang TC, Lee CL. Coronary artery fistulas in neonates, infants, and children: clinical findings and outcome. *Pediatr Cardiol* 2002;23:415-9.
- Ross JJ Jr, Mintz GS, Chandrasekaran K. Transthoracic two-dimensional high frequency (7.5 MHz) ultrasonic visualization of the distal left anterior descending coronary artery. *J Am Coll Cardiol* 1990;15:373-7.
- Caiati C, Montaldo C, Zedda N, Bina A, Iliceto S. New noninvasive method for coronary flow reserve assessment: contrast-enhanced transthoracic second harmonic echo Doppler. *Circulation* 1999;99:771-8.
- Attili A, Hensley AK, Jones FD, Grabham J, DiSessa TG. Echocardiography and coronary CT angiography imaging of variations in coronary anatomy and coronary abnormalities in athletic children: detection of coronary abnormalities that create a risk for sudden death. *Echocardiography* 2013;30:225-33.
- Eidem BW, Cetta F, O'Leary PW, editors. *Echocardiography in Pediatric and Adult Congenital Heart Disease*. Philadelphia, PA: Lippincott Williams & Wilkins, a Wolter Kluwer Business; 2010.
- McCulloch MA, Gutgesell HP, Saulsbury FT, Hellems M, Hammill WW, Lim DS. Limitations of echocardiographic periaortic brightness in the diagnosis of Kawasaki disease. *J Am Soc Echocardiogr* 2005;18:768-70.
- Manlhiot C, Millar K, Golding F, McCrindle BW. Improved classification of coronary artery abnormalities based only on coronary artery z-scores after Kawasaki disease. *Pediatr Cardiol* 2010;31:242-9.
- Margossian R, Lu M, Minich LL, Bradley TJ, Cohen MS, Li JS, et al. Predictors of coronary artery visualization in Kawasaki disease. *J Am Soc Echocardiogr* 2011;24:53-9.
- Capannari TE, Daniels SR, Meyer RA, Schwartz DC, Kaplan S. Sensitivity, specificity and predictive value of two-dimensional echocardiography in detecting coronary artery aneurysms in patients with Kawasaki disease. *J Am Coll Cardiol* 1986;7:355-60.