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Ciclo XXVI

CONGENITAL AND ACQUIRED CORONARY ARTERIES
ANOMALIES IN INFANT CHILDREN AND
ADOLESCENTS:
COMBINATION OF TRANSTHORACIC
ECHOCARDIOGRAPHY AND CARDIAC MAGNETIC
RESONANCE IMAGING

Coordinatore:
Chiar.mo Prof. Aldo Agnetti

Tutor:
Chiar.mo Prof. Aldo Agnetti

Dottorando: Dott. Bertrand Tchana.

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INTRODUCTION

Coronary arteries, which seat on the heart as a crown, are the main subject of concern in adult cardiology due to the prevalence of acquired coronary arteries diseases, particularly related to atherosclerosis. In the pediatric population, because of the involvement of coronary circulation in an increasing number of acquired systemic diseases and psychosocial impact, surgical issues in the surgery of structural congenital heart diseases, coronary arteries are becoming an important topic in pediatric cardiology.

In the pediatric patient, the main concern about coronary arteries regards congenital malformations, as far as some of the arrangements implicated in sudden death can remain asymptomatic throughout the lifetime of the individual, even if some important systemic diseases can lead to significant coronary arteries anomalies.

Coronary arteries can have multiple anomalous origins, and epicardial courses, in a otherwise normally structured heart. The anomalous patterns are relatively frequent; using coronary arteriography as the diagnostic modality, a prevalence of abnormal arrangement was shown in around 1% of a large cohort of more than 100,000 patients; on all ages autopsies there were found in 0.2-2.2%, 0.5% on pediatric autopsies (1,2,3,4,5). In the setting of congenital heart disease coronary artery abnormalities are common. Hence, delineation of the origin, course and accurate visualization of the coronary artery anatomy is critical for surgical planning and assessment of results in congenital heart disease. Some systemic diseases, particularly connectivitis which involve vascular wall, can lead to coronary arteries persistent deformation with flow alteration, thrombosis and subsequent myocardial ischemia and infarction. In this setting Kawasaki Disease features the leading cause of acquired coronary arteries anomalies.

Coronary arteries imaging has two technical requirements: high temporal resolution because of the near constant motion during respiratory and cardiac cycle and high spatial resolution for accurate imaging of small vessels. Conventional angiography and Computed Tomography (CT scan), which fulfilled these requirements, are currently the modalities of choice to assess and delineate coronary arteries anatomy in therapeutic decision - making. However both techniques carry risks associated with the exposure to ionizing radiation and iodinated contrast agents, moreover conventional angiography presents the disadvantage of its invasive nature, the lacks of three-dimensional information and ability to image soft tissue,

making it difficult to demonstrate the spatial relationship between the coronary arteries course, myocardium, and great vessels. Continuous progress and improvement and the extraordinary sophistication of non – invasive imaging technique such as echocardiography and magnetic resonance imaging (MRI), alone or better in combination, can now provide a complete non-invasive alternative evaluation of the anatomic origins and courses of the coronary arteries with exquisite accuracy. MRI can provide high resolution, high contrast, anatomical images in freely-selectable 2D planes or true 3D volume scans. Anatomical images, combined with the functional information provided by cine, velocity mapping, and dynamic contrast angiography techniques make MRI ideally suited to the evaluation of the coronary anomalies. The use of these techniques in combination, therefore, will increase the ability to diagnose an increased number of abnormal arrangements.

CORONARY ARTERIES

1. ANATOMY AND DISTRIBUTION

There are normally two major coronary arteries. Usually they come from the middle of the sinuses, but they may arise from the supra-ventricular ridge or even above it (fig. 1).

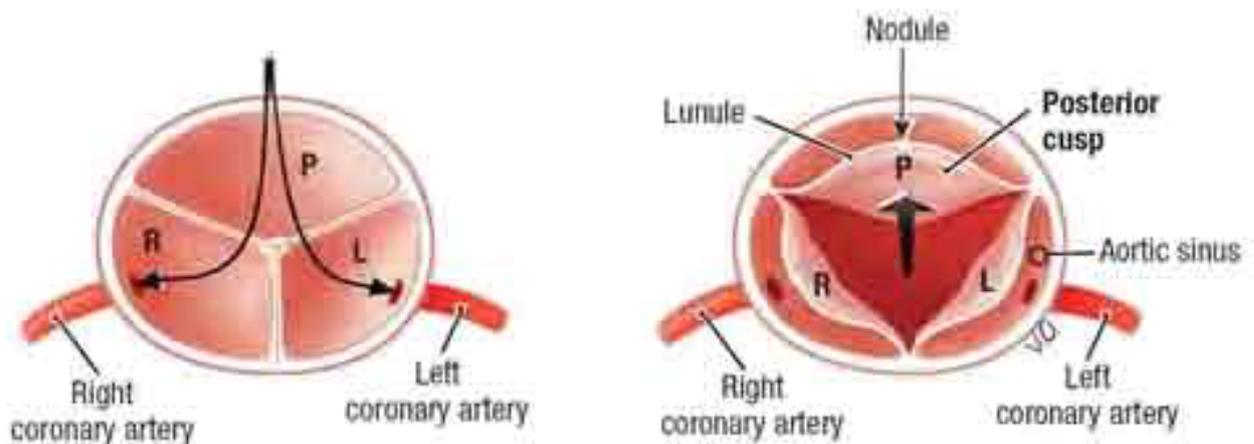


Figure 1.: a superior view of aortic with the origin of coronary arteries (6)

The position of the ostium does not appear to affect the flow through it. The ostia may be round, oval, or elliptical. The arteries are perpendicular to the aortic wall; that is, they are radially arranged relative to the center of the aorta (Fig. 1.). The left main coronary artery originates from the left aortic sinuses and gives rise to the circumflex branch, which courses posteriorly, then continues as an anterior descending branch (Fig. 2.). The right coronary artery which originates from the right aortic sinuses and gives rise to a small conal branch and then courses posteriorly along the atrioventricular groove (Fig. 2.). There is no separate septal branch, the septum being supplied by perforating branches that enter the septum from the anterior and posterior descending coronary arteries (Fig. 3.).

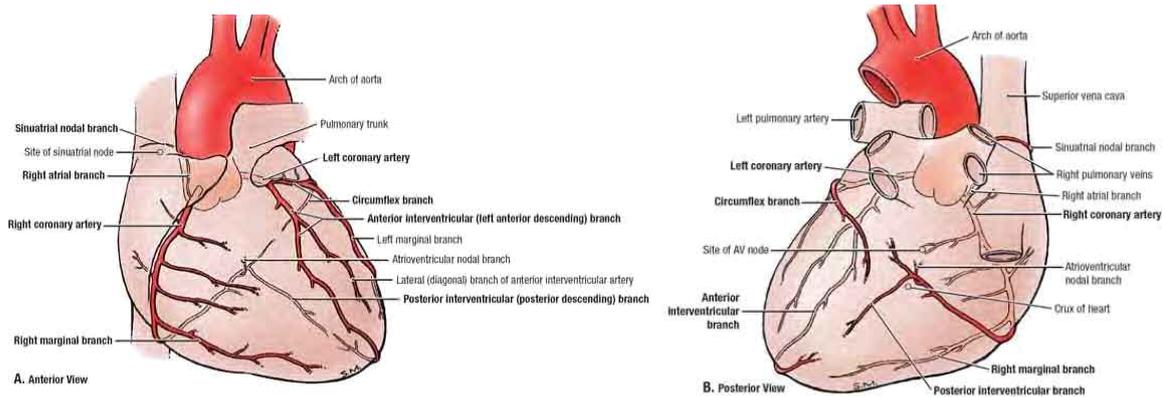


Figure 2. A and B: anterior and posterior view of the heart figuring the epicardial course of the coronary arteries (6)

In 90% of the population the right coronary artery is described as dominant, giving rise to the posterior descending coronary artery, although it is certainly not posterior when viewed in attitudinally appropriate orientation, and may even ascend as it course towards the cardiac apex, and supplies the posterior part of the ventricular septum, the inferior wall of the left ventricle, and the atrioventricular node (Fig. 2). The left coronary artery supplies only the free wall of the left ventricle. Separate origin of the conus branch of the right coronary artery occurs commonly (Fig. 2.). The corresponding anomaly on the left side—separate origins of the left anterior descending and left circumflex coronary arteries—occurs in about 1% of people and is more frequent with bicuspid aortic valves. Interestingly, a significant number of patients with bicuspid aortic valves or aortic stenosis have left dominant systems and a short left main coronary artery.

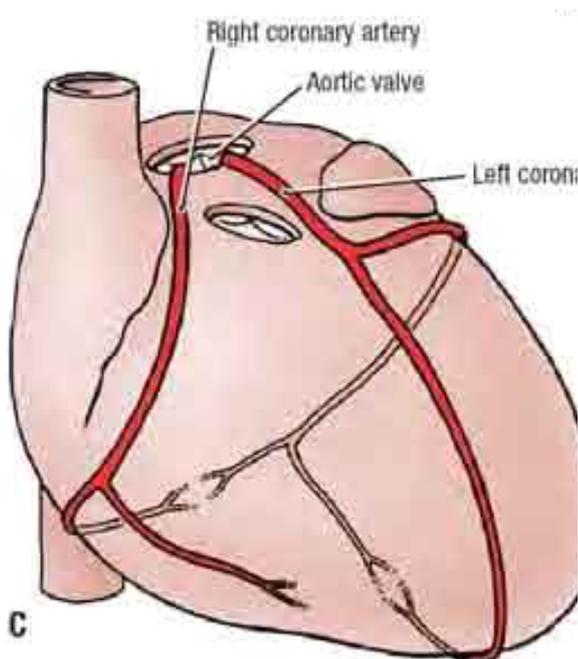


Figure 2. C: course of the main coronary arteries (6)

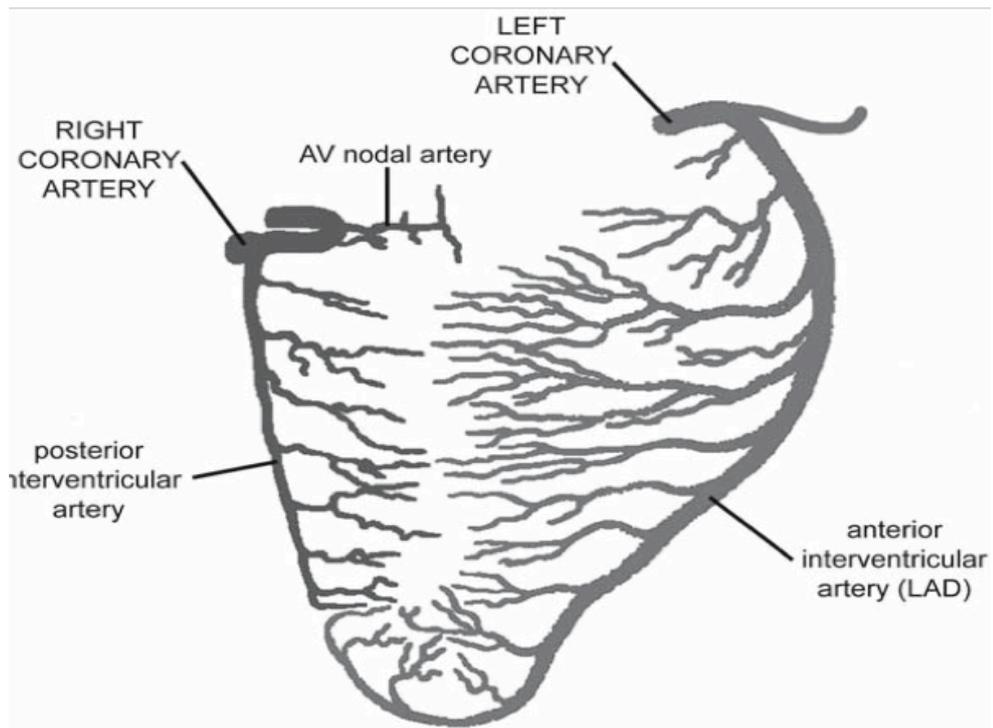


Figure 3.: Arterial supply to the interventricular septum. The right coronary artery supplies the posterior one-third of the interventricular septum, and the left coronary supplies the anterior two-thirds. The artery to the atrioventricular node commonly branches off the posterior interven- tricular artery.

2. CONGENITAL CORONARY ARTERIES ANOMALIES CLASSIFICATION

Anomalies of the coronary arteries can be considered the result of a rudimentary persistent of an embryologic coronary arterial structure, a failure of normal coronary development, a failure of the normal atrophic process of development, or the misplacement of a connection of an otherwise normal coronary artery. At present, there is no consensus as how best to classify the multiple variations.

A. CONGENITAL CORONARY ARTERY ANOMALIES IN NORMALLY STRUCTURED HEART

In the past, the lesions were considered to be of major or minor significance. Many of the allegedly major variations fail to produce symptoms during life, so such a system is less than ideal. Thus, one can classified anomalies of the coronary arteries as:

- > Abnormal origin
 - o Anomalous aortic sinusal origin
 - Right coronary artery arising from left coronary sinus (fig. 1)
 - Main stem of left coronary artery arising from right coronary sinus (fig. 2)
 - Circumflex artery arising from right coronary sinus (fig. 3)
 - Anterior interventricular artery arising from right coronary sinus
 - o Ectopic origin of coronary artery
 - From pulmonary trunk (fig. 4)
 - From right pulmonary artery
 - From left pulmonary artery
 - From brachiocephalic artery
- > Abnormal course
 - o Anomalous epicardial course
 - Retroaortic
 - Interarterial
 - Prepulmonary
 - Intramural (fig. 5)
- > Abnormal number of coronary arteries

- o Solitary right coronary artery (fig. 6)
- o Solitary left coronary artery (fig. 7)
- o Duplication of coronary arteries
- > Abnormal connections of the arteries.
 - o Connections with a cardiac cavity
 - o Coronary arteriovenous fistulas
 - o Coronary to extracardiac arterial or venous connections
- > Lack of patency of the orifice of a coronary artery
- > Myocardial bridging

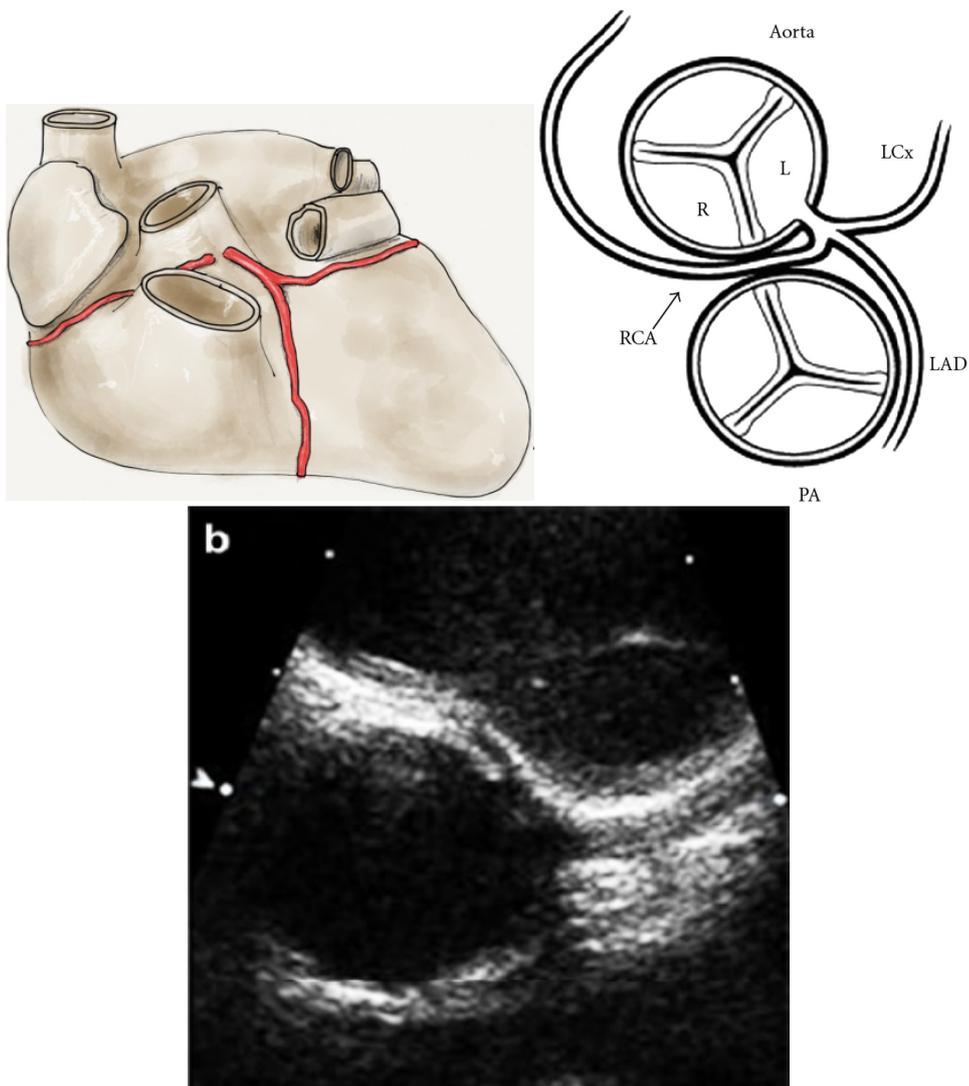


Figure 1.: Right coronary artery arising from left coronary sinus (5, 7)

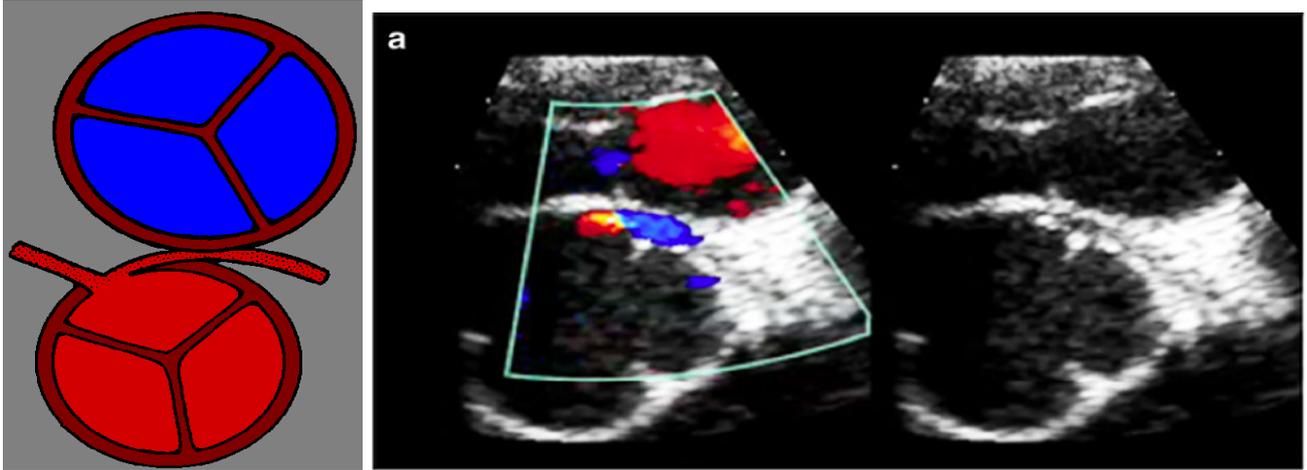


Figure 2.: Main stem of left coronary artery arising from right coronary sinus (5; courtesy of Dr. S, Sanders, Boston Children Hospital)

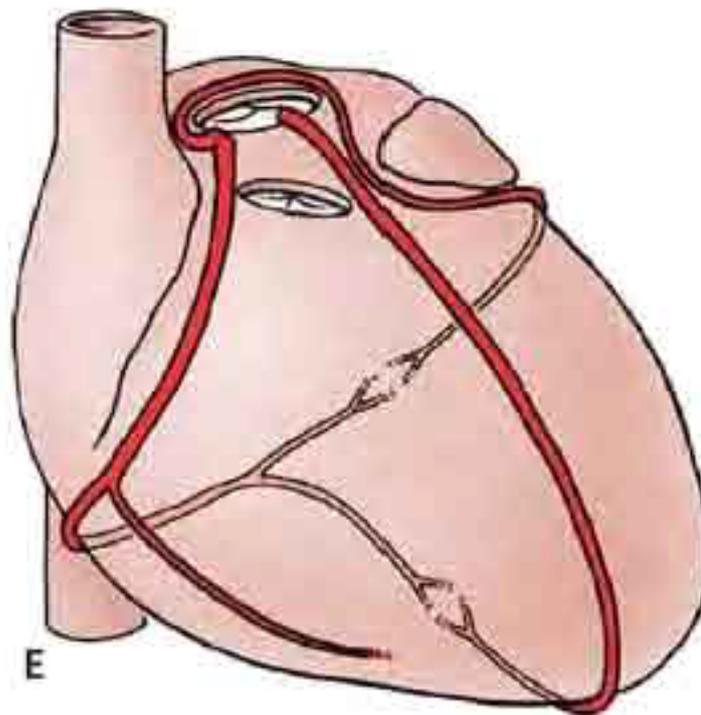


Figure 3: Circumflex artery arising from right coronary sinus (6)

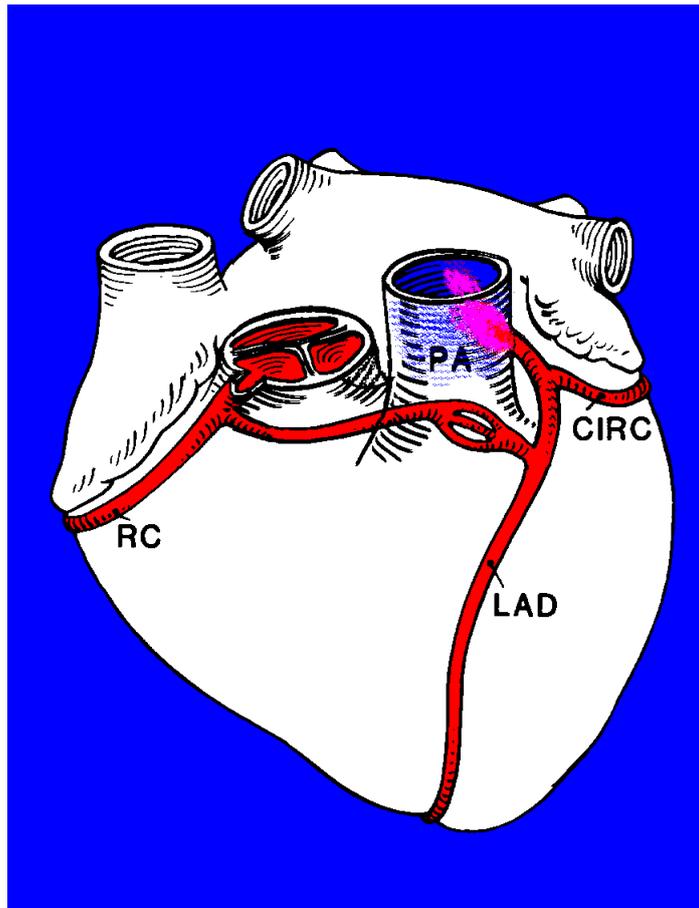


Figure 4.: Anomalous origin of the left coronary artery from pulmonary trunk (courtesy of Dr. S. Sanders, Boston Children Hospital)

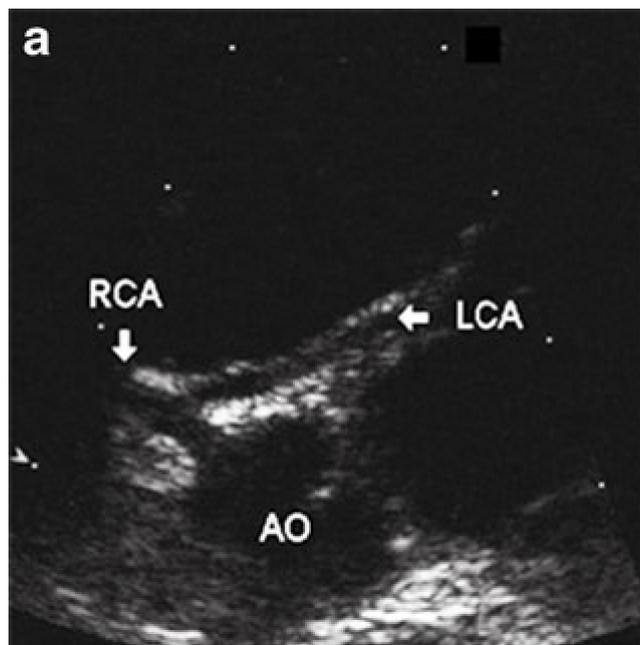


Figure 5.: intramural course in the conal septum of the left coronary artery arising from the right coronary artery (5).

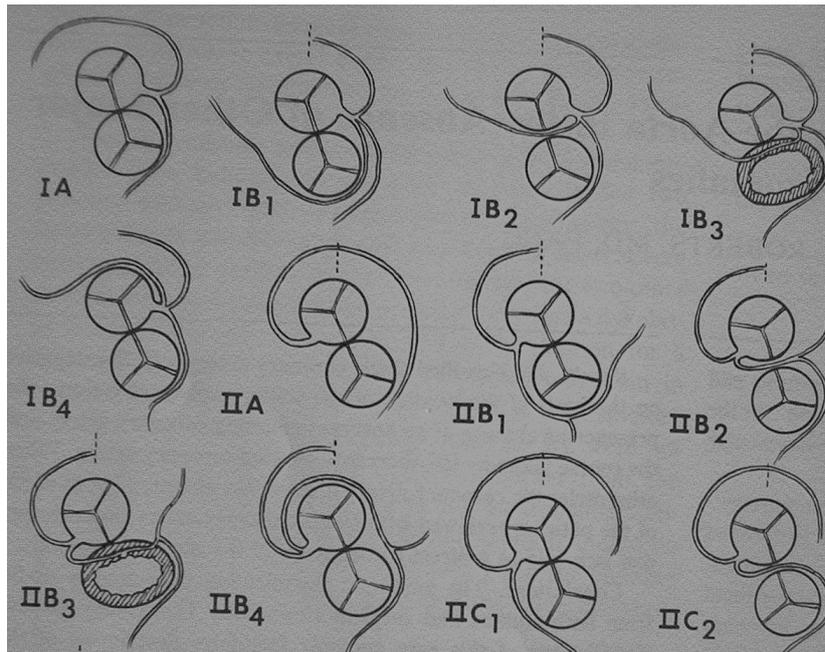


Figure 6.: Single right coronary artery and other examples of single coronary artery (9)

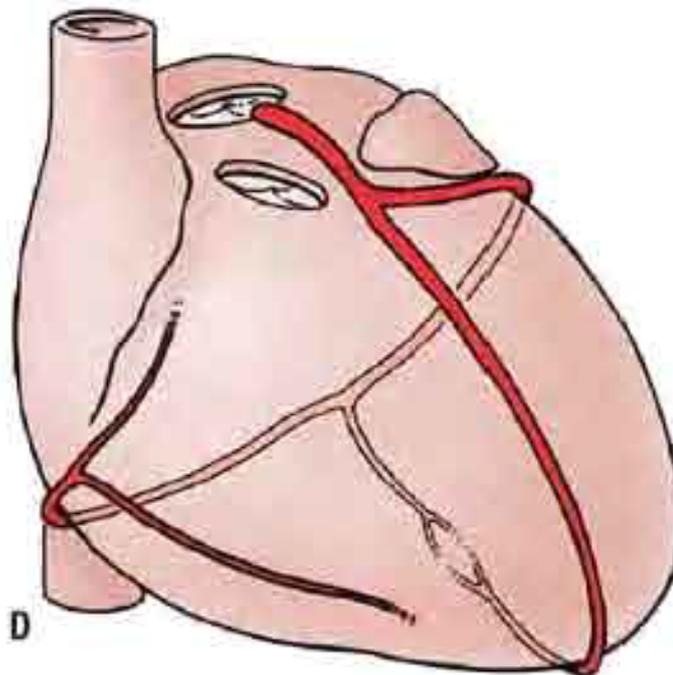


Figure 7.: Single left coronary artery (6)

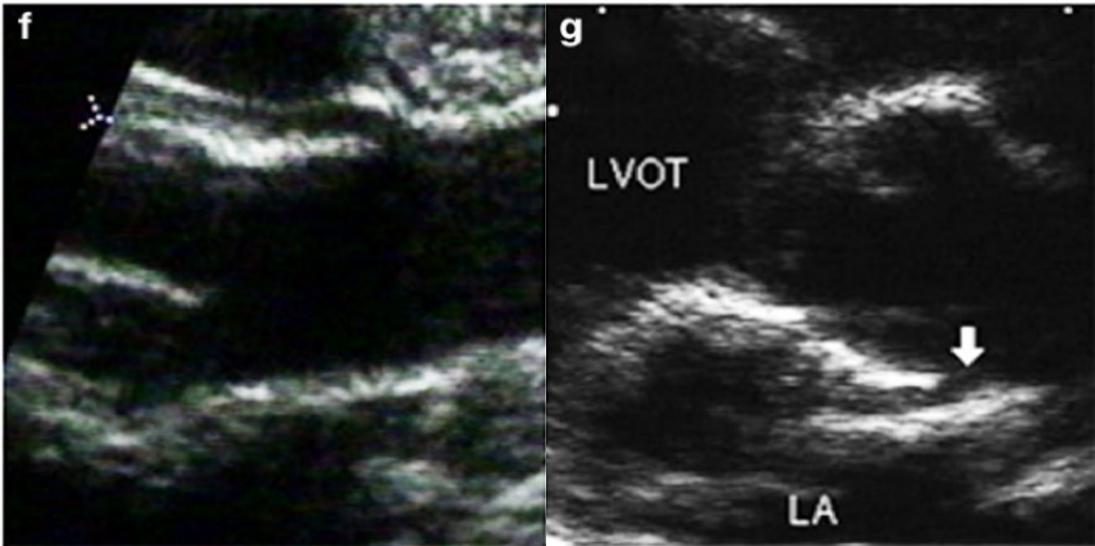


Figure 8.: parasternal long-axis view with coronary arteries arising above the sinotubular junction (5)

B. CONGENITAL CORONARY ARTERY ANOMALIES AND STRUCTURAL CONGENITAL HEART DISEASES

As one might expect with the heterogeneous nature of congenital heart disease, many CHD are associated with aberrant coronary artery systems:

- > Double outlet right ventricle, the coronary artery distribution may follow patterns similar to those seen in tetralogy of Fallot or in transposition of the great arteries:
 - o DORV TOF type: LAD arising from the right coronary artery
 - o DORV TGA type: coronary anomalies similar to those seen in TGA
- > Tetralogy of Fallot: incidence reported ranging from 18% to 31%, with the two anomalies with major clinical relevance being:
 - o A coronary artery crossing the right ventricle outflow tract (reported incidence of 5%)
 - o A coronary artery supplying part of the pulmonary blood flow

Others abnormalities are:

- a large conal branch arising from the right coronary artery (fig. 9)
 - left anterior descending arising from the right coronary artery
 - dual or paired anterior descending coronary arteries, one arising from the right sinus of Valsalva (fig. 9)
 - both coronary arteries arising from a single left ostium
- > Double-inlet left ventricle,
 - > D-Transposition of the great arteries, nine major anatomical patterns accounting for more than 95% of the coronary variability can be defined:
 - o Morphologically left and right coronary arteries (named by their distal course and area of distribution) arising from the posterior-leftward and posterior-rightward facing sinuses respectively (about 67%, fig. 10)
 - o Circumflex arising from the right coronary artery (about 16%) and traveling behind the arterial roots (it is important not to confuse a diagonal branch or great cardiac vein with the origin of the left circumflex) (fig. 10)
 - o Take-off at or above sinotubular junction

- o Paracommissural take-off
- > L-transposition of great arteries:
 - o Single coronary artery
 - o Coronary arteries crossing the outflow tract
- > Truncus arteriosus:
 - o High take-off of one or both coronary arteries
 - o Paracommissural take-off
 - o Ostia stenosis
- > Pulmonary atresia with intact ventricular septum:
 - o Absence of proximal aorto – coronary connection
 - o Coronary arterial stenosis or interruption
 - o Major coronary to right ventricle fistula

Not surprisingly, an especially high incidence of coronary anomalies is found in lesions involving malformation of the outlet portion (conotruncus) of the developing heart.

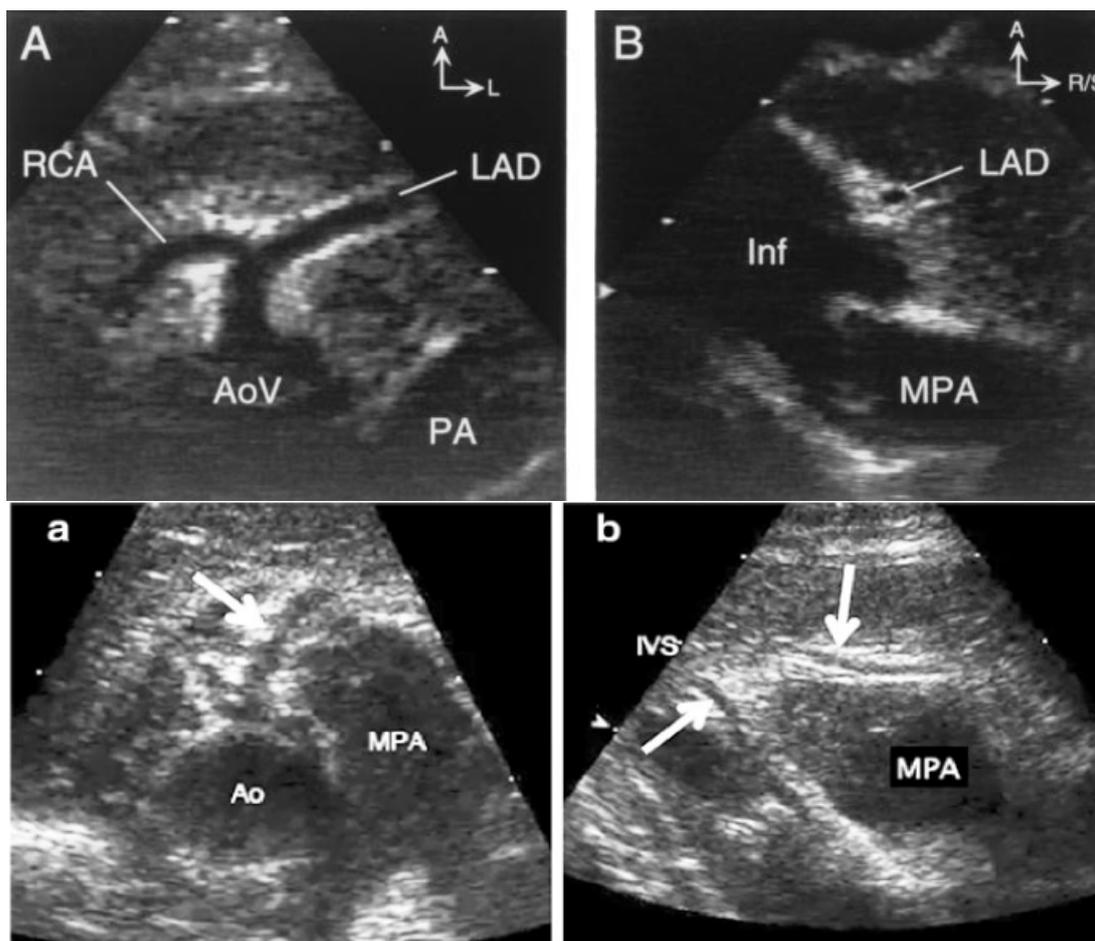


Figure 9.: echocardiography images of a crossing coronary artery (left) and dual anterior descending coronary arteries (5).

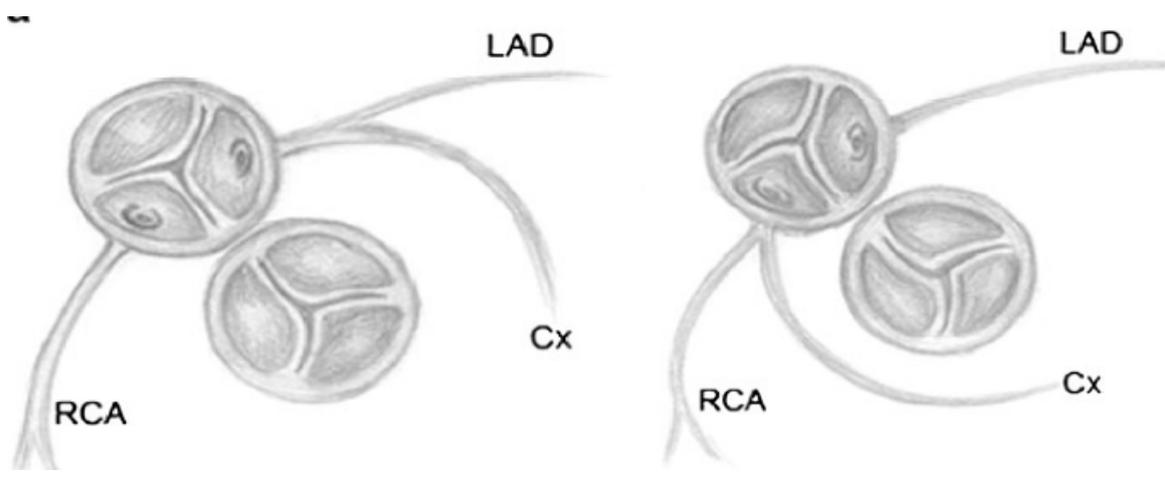


Figure 10.: coronary artery anatomy in D- transposition of great arteries (5)

3. ACQUIRED CORONARY ARTERY ANOMALIES: KAWASAKI DISEASE

Kawasaki disease (KD), is an acute systemic vasculitis of unknown etiology, with a worldwide distribution, that predominantly occurs in young children, with 80% of cases reported in children under 5 years of age and 90% occurring in children under 8 years of age. The disease occur with highest frequency in children of Asian descent, follow by African-Americans, and Caucasian. Male to female ratio is 1.5/1. The acute febrile phase is characterized by diffuse microvascular angiitis with endarteritis and perivascular inflammation of coronary arteries. The subacute phase features a persistent panvasculitis of the coronary arteries which may be associated with ectasia, aneurysm and/or thrombosis. The convalescent phase presents replacement of microvascuar angiitis by intimal thickening of the coronary arteries eventually with scarring, stenosis and calcifications. It is associated with the development of coronary artery aneurysms (CAAs) in 15% to 25% of untreated cases and in 10% of cases treated, and represents the most important cause of acquired coronary artery disease in childhood. In approximately one half of patients, CAAs resolve within 1 to 2 years, whereas in the other patients, the aneurysms persist and may lead to thrombosis and stenotic lesions that can cause myocardial ischemia and infarction. As a consequence, serial coronary artery surveillance is necessary in patients with a history of KD.

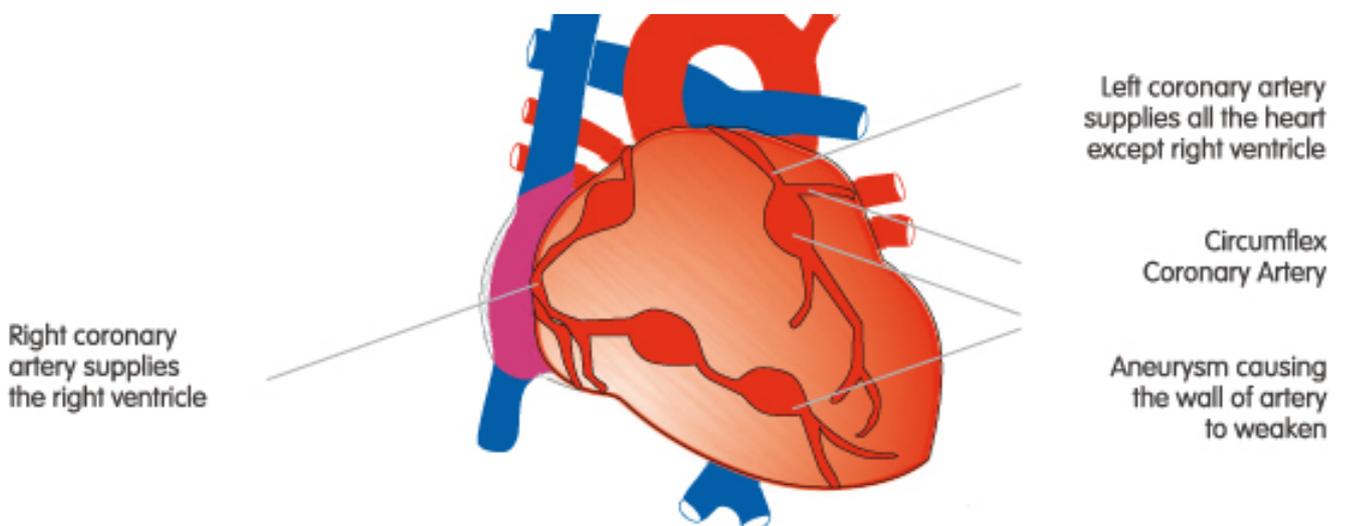


Figure 7: coronary aneurysms in

Kawasaki disease (www.corience.org)

NON-INVASIVE CORONARY ARTERY IMAGING

Historically, coronary arterial anomalies have been difficult to diagnose by noninvasive methods. With improvement in the acquisition of images, and the technology used to display them, the ability to identify the origins and courses of the coronaries has markedly improved over the last years. Routine imaging of the coronary arteries is important because anomalies may be asymptomatic and prospectively identifying them may save lives.

CORONARY ARTERIES TRANSTHORACIC ECHOCARDIOGRAPHY

1. 2D AND DOPPLER COLOR CORONARY ARTERIES VIEWS

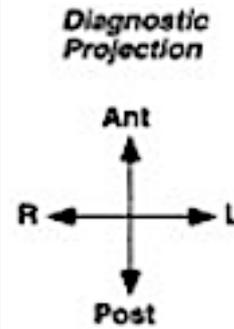
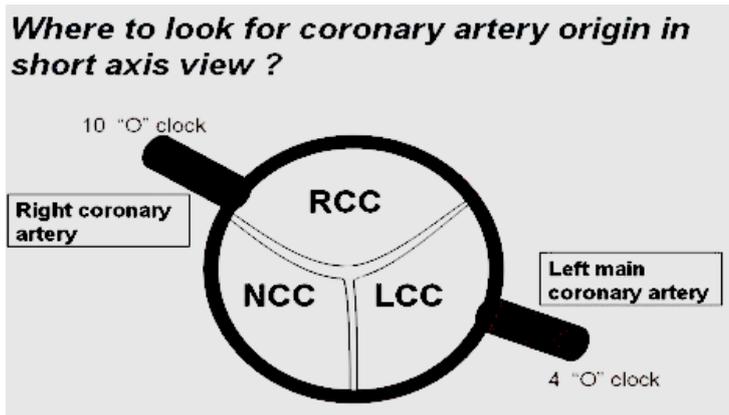
Identification of coronary arterial origins is now a routine component of the standard pediatric echocardiogram. Two – dimensional echocardiography with Doppler is the most common practical tool to investigate infant children with suspected anomalies of the coronary arteries.

The goals of the examination are (5, 8):

- identification of the origin of each major coronary artery
- identification of the proximal course
- demonstration of flow direction by color Doppler

The highest frequency probe that allows adequate penetration may offer a distinct identification of the coronary arteries as they are often in the near-field of the sector (5,8).

The proximal coronary arteries are best imaged in the parasternal (long and short axis) and subcostal views. In the parasternal short axis view, at the base of the heart, both main coronary arteries origins may be seen simultaneously from their respective sinuses of valsalva (Fig. 1A e B). Clockwise or counterclockwise rotation, with very subtle movement, are often necessary for optimal imaging of the vessels.



RCC: Right Coronary Cusp

LCC: Left Coronary Cusp

NCC: Non Coronary Cusp

N.B.: Aortic trileaflet attachment is highly variable, so as the origin of coronary arteries, the right may arise anywhere between 9 and 11 O'clock, the left anywhere between 3 and 5.

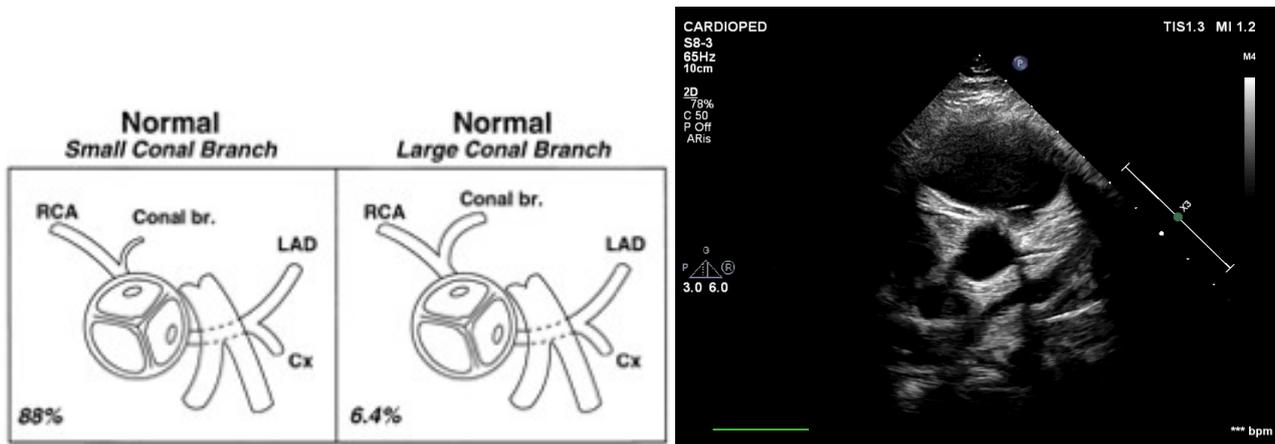


Figure 1B: Parasternal short axis view where both coronary arteries can be seen (8)

Parasternal long axis view with leftward angulation of the probe allows visualization of the left main coronary artery bifurcation as well as a long length of the left anterior descending artery down in the ventricular septum (Fig. 2). An inferior angulation allows to visualize the posterior descending coronary artery.

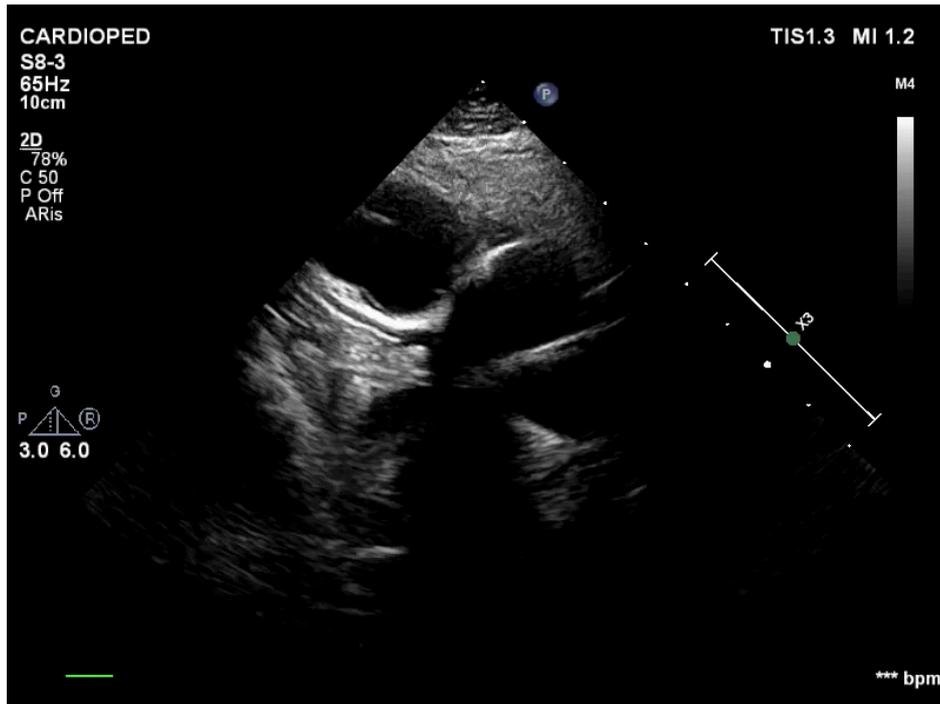


Figure 2.: Parasternal long axis view with leftward angulation of the probe visualization of the left main coronary artery bifurcation as well as a long length of the left anterior descending artery down in the ventricular septum

Subcostal long axis (coronal) view at the level of the aortic root allows to visualize the proximal course of the coronary arteries.

Apical 4-chamber view is the best to image the distal right coronary artery in the posterior right atrio-ventricular groove (fig. 3.).

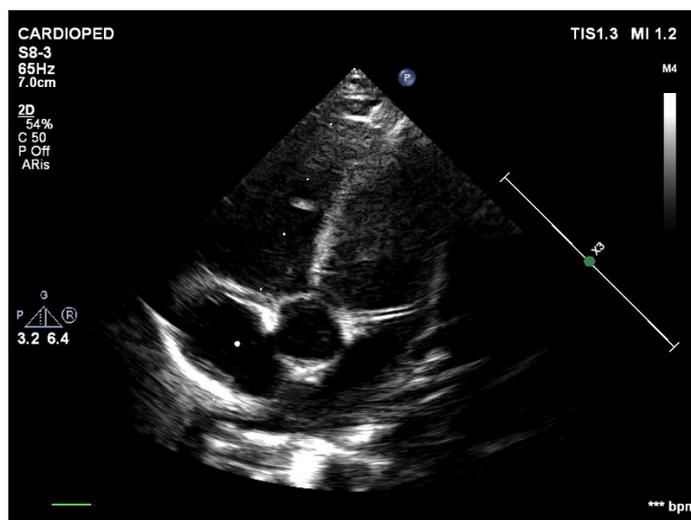


Figure 3.: modified 4 chamber view with right coronary artery and left circumflex artery

Left Anterior Descending coronary artery

The proximal LAD is visualized in a modified parasternal short-axis view just above the level of the aortic valve (fig. 4).

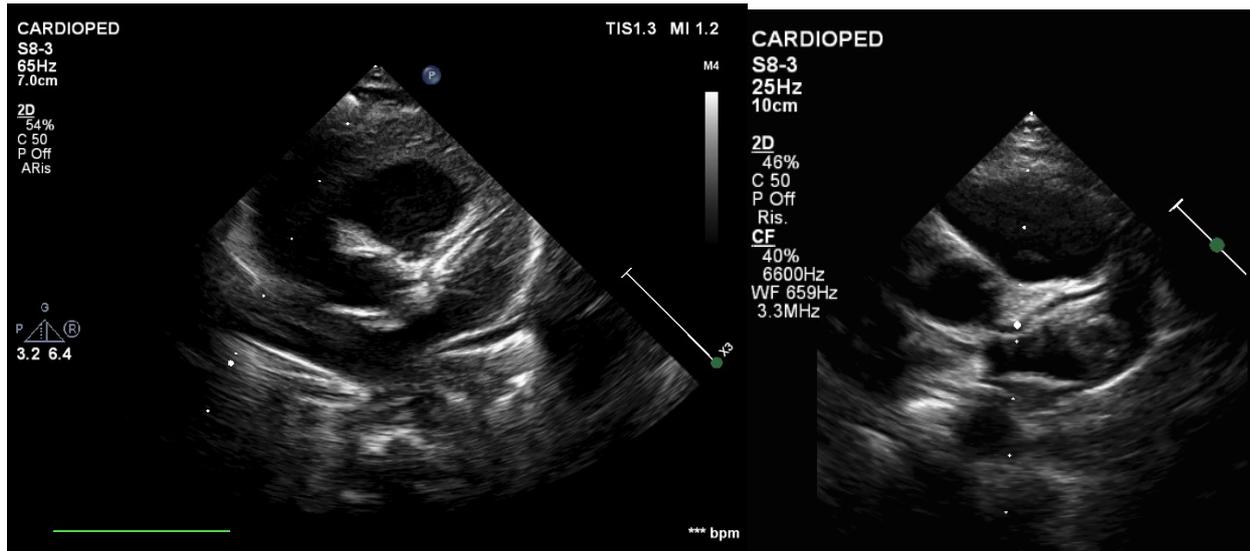


Figure 4.: modified parasternal short axis view showing left main coronary artery, left anterior descending branch and the origin of circumflex

A septal branch of LAD may also be visualized in the parasternal short-axis view at the mid-papillary muscle level. To visualize the distal LAD, the optimal acoustic window is found close to the mid-clavicular line in the fourth or fifth intercostal space with the patient positioned in the left lateral decubitus position. The distal LAD can best be identified in the long-axis view with the ultrasound beam inclined laterally (fig. 2).

In addition to the anatomic information on the coronary arteries, echocardiography with Doppler color flow can demonstrate the direction of flow that may provide important information regarding anomalies of origin (5,10,11,12), function and regional wall motion assessment eventually pointing to specific areas of perfusion (5,10,11,12). Coronary blood flow is identified using Doppler color flow mapping with the Nyquist limit set at 10 to 20 cm/s (10,11,12). Because the coronary artery is not in the same plane throughout the entire cardiac cycle, because of translational motion, flow sampling should be optimized during diastole when the position of the coronary artery is most stable (10,11,12). The probe angle

and sample volumes should be adjusted to orient the Doppler ultrasound beam parallel to coronary flow (10,11,12). In normal coronary arteries, the color and spectral Doppler signals of distal LAD show diastolic dominant flow with a smaller systolic component (5,10,11,12). The flow patterns are very similar to that in the proximal LAD measured using transesophageal Doppler echocardiography, and the flow patterns obtained invasively using Doppler guidewires or Doppler catheters. Extravascular compression and lower coronary perfusion pressure during systole is likely to be responsible for the lower systolic component.

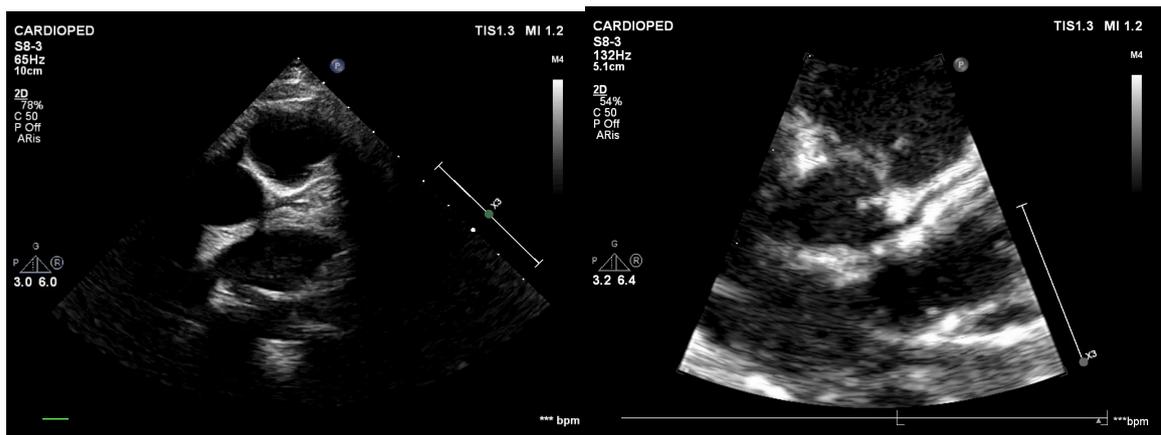


Figure 5: main left coronary artery and left anterior descending in modified parasternal short axis view

Right Coronary Artery

For imaging peripheral RCA flow, a lower-frequency transducer is required because of the distance between the transducer and the basal inferior cardiac wall (fig. 6). The posterior descending branch of distal RCA may be visualized in the 2-chamber view adjacent to the ostium of the coronary sinus (5,10,11,12). The patient should be positioned in the left lateral decubitus position. The ultrasound beam should be inclined laterally or rotated to visualize the coronary blood flow close to the epicardial layer of the proximal portion or midportion of the posterior interventricular sulcus under color flow mapping guidance (5,10,11,12). A recent study shows coronary flow velocities measured with pulsed wave Doppler using minimum angle correction (0-15 degrees) (10,11,12).

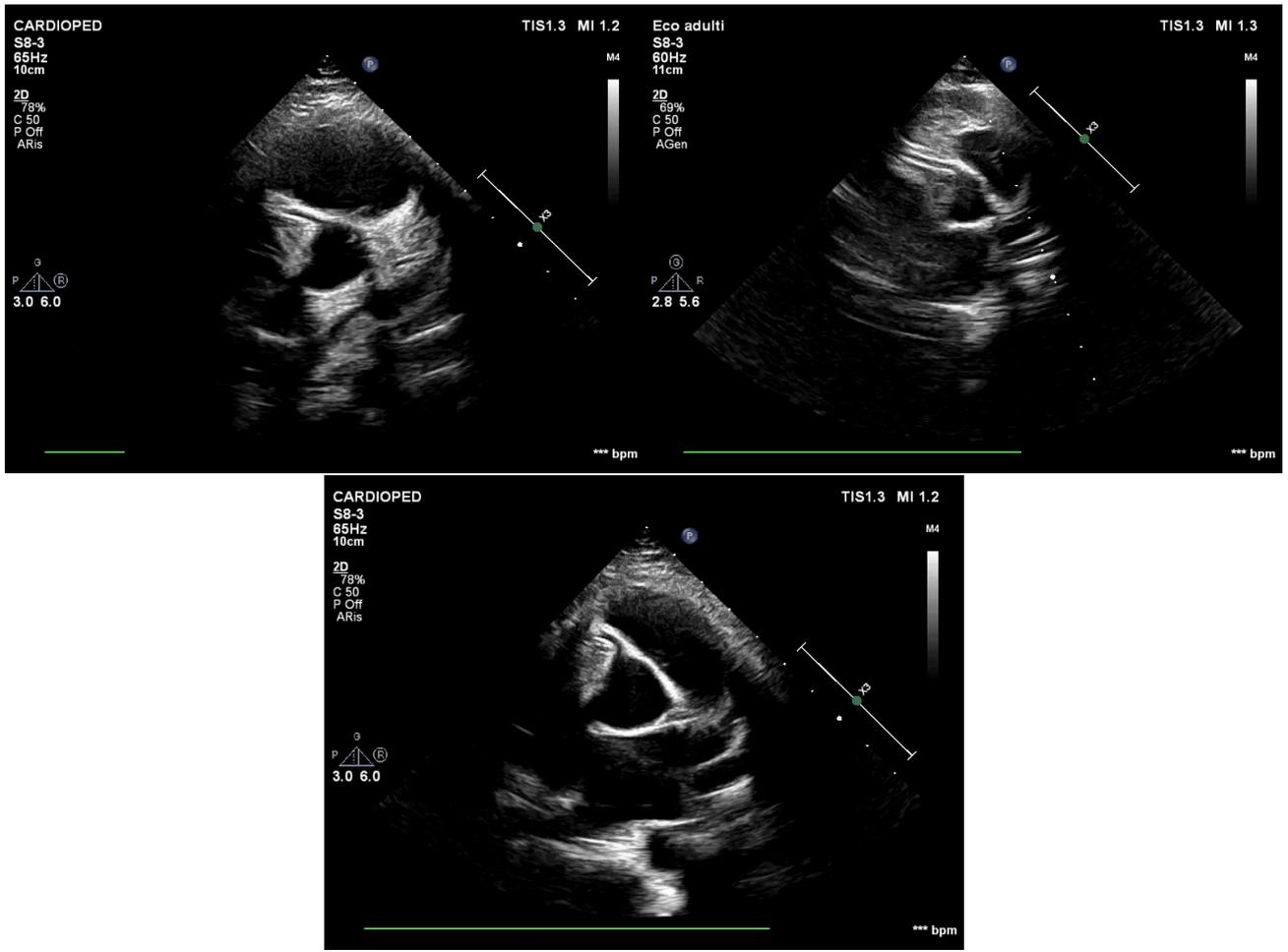


Figure 6.: parasternal short axis views of the proximal right coronary artery

2. CONGENITAL CORONARY ARTERIES ANOMALIES

1. Anomalous coronary artery from the pulmonary arteries

The most significant ectopic origin is from the pulmonary arteries, particularly the trunk of the pulmonary artery. Either the left or right coronary artery can arise from the pulmonary trunk. The coronary arteries may also arise from the right or left pulmonary artery.

a. Anomalous origin of the left coronary artery from the pulmonary trunk

Also known as the Bland–White–Garland syndrome, its one of the more common coronary arterial abnormalities encountered in children. Referred to as anomalous left coronary artery from the pulmonary artery, (ALPACA), it occurs from 1 in 250 to 1 in 400 of all congenitally malformed hearts, with an overall incidence of approximately 1 in 300,000 children, with male/female ratio of 2/1. The anomalous artery, almost without exception, arises from the sinus of the pulmonary trunk adjacent to the left coronary sinus of the aortic root, although it may also arise from the right or the left pulmonary artery. Typically, the right coronary artery is enlarged at its aortic origin, whereas the anomalous left artery tends to be small, and relatively thin walled.

The diagnosis can be confirmed by using transthoracic echocardiography.

The parasternal short-axis plane is one of the best views to visualize the origins of the coronary arteries. In ideal circumstances, interrogation reveals the anomalous origin of the left coronary artery from the pulmonary trunk. This is more feasible when the arterial origin is located anteriorly from the pulmonary trunk. Typically, however, the origin is posterior or leftwards, just adjacent to the normal origin from the aorta. Color flow mapping is quite helpful, but. Lowering the Nyquist limit permits greater sensitivity for imaging the coronary arteries. When the color scale is lowered to 18–50 cm/s, it is possible to assess the direction of coronary arterial flow. With pulmonary arterial origin, it is often possible to identify reversal of flow from the myocardium towards the pulmonary trunk, at least during part of the cardiac cycle. This is one of the most important findings suggestive of the diagnosis. In some cases, off-axis parasternal short-axis views help identify the anomalous origin, and color flow mapping reveals a jet emptying into the pulmonary trunk.

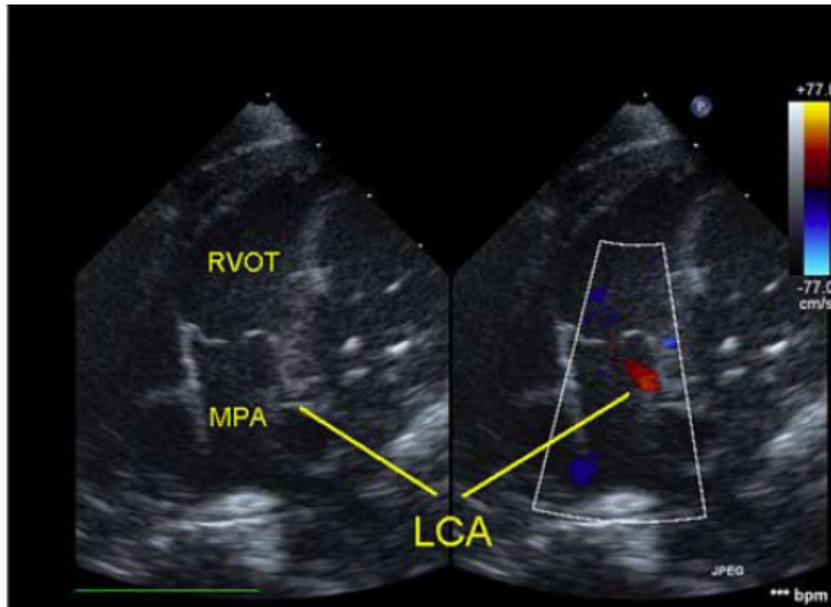


Figure 1,; High parasternal short-axis view in color-compare mode of the right ventricular outflow tract and pulmonary trunk demonstrating the leftward and anterior origin of the left coronary artery from the pulmonary trunk. Color flow demonstrates flow from the artery filling the pulmonary trunk in retrograde fashion; RVOT: right ventricular outflow tract; MPA: pulmonary trunk; LCA: left coronary artery (13)

In contrast to those presenting as infants, late presentation is not usually associated with significant left ventricular dysfunction. It is perhaps because of a better-developed collateral arterial system, but also because of some narrowing of the left coronary arterial orifice as it enters the pulmonary root. In addition, any elevation of pulmonary arterial pressure allows better perfusion of the myocardium, such as is the case with patency of the arterial duct. The most striking echocardiographic features in such patients are the dilation of the right coronary artery and the collateral arterial flow seen in the ventricular septum.



Figure 2.: Parasternal short-axis view demonstrating a dilated right coronary artery in a patient with the anomalous origin of the left coronary artery from the pulmonary trunk (12)

The collateral flow is often misdiagnosed as multiple muscular ventricular septal defects.

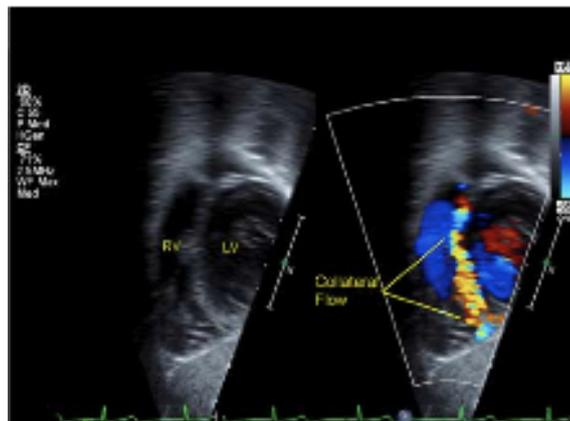


Figure 3.: Subxiphoid sagittal view in colour-compare mode demonstrating significant coronary collateral flow in the ventricular septum in a child with a late presentation of the anomalous origin of the left coronary artery from the pulmonary trunk; (12)

The right coronary artery has connections to the anomalously arising left coronary artery similar to fistulous connections, and therefore is often tortuous in its appearance. In those presenting at this age, the lesion may easily be misinterpreted as a coronary

arterial fistula because, in the era of color flow mapping, it is an easy matter to visualize the collateral arteries. In addition, the enlarged right descending coronary artery may have the appearance of a fistula.

This diagnosis can also easily be missed, even with a thorough and detailed examination. Should it arise from the pulmonary trunk, the left coronary artery then follows its normal path, often coursing quite close to the aorta, and sometimes even taking an intramural course through the aortic wall. Thus, it often appears to have a normal aortic origin.

b. Anomalous origin of the right coronary artery from the pulmonary trunk

Anomalous pulmonary origin of the right coronary artery (ARCAPA) is much less common, with approximately 61 cases in the literature, only half of which are patients of pediatric age and typically has a more benign course.



Figure 4.A and B: Parasternal short-axis view in colour-compare mode demonstrating the anomalous origin of the right coronary artery from the pulmonary trunk. The right coronary artery is seen entering the pulmonary trunk just above the pulmonary valve. Colour flow mapping demonstrates that the flow is from the artery into the pulmonary trunk, which is dilated as a result of this flow; Parasternal short-axis view in the same patient with the anomalous origin of the right coronary artery from the pulmonary trunk demonstrating a dilated left coronary artery that provides collateral flow to the right coronary artery (12)

ARCAPA differs from ALCAPA in being commonly diagnosed in association with other congenital heart defects (55% of pediatric cases). Aortopulmonary window and Tetralogy of Fallot TOF account for the majority of these associated lesions. It is often diagnosed later, typically in childhood or early adulthood after a cardiac murmur has been recognized. Patients are usually asymptomatic, or have other associated congenital diseases requiring surgical treatment. It is easier to recognize the pulmonary truncal origin of the right coronary artery, since retrograde flow into the pulmonary trunk is typically seen. The left coronary artery is usually enlarged and tortuous because of the collateralization to the right coronary artery, and collateral arteries are seen in the ventricular septum as with anomalous origin of the left coronary artery. Left ventricular performance is usually normal, and right ventricular performance may be diminished or normal. Significant tricuspid regurgitation is not a common feature.

2. Anomalous aortic origin of the coronary arteries

When the heart is otherwise normally structured, abnormal coronary arteries most frequently continue to have their origin from the aorta. It could be argued that the high take-off relative to the sinutubular junction, if excessive, should be considered an anomaly.

The difficulty comes in determining the height of take-off judged to represent the abnormality. The difficulty comes in determining the height of take-off judged to represent the abnormality. As origins minimally above the sinutubular junction are common,⁷ the arteries probably need to arise at least 1 centimeter above the junction in adults to be considered anomalous, with appropriate consideration given for growth when assessing the origins in infants and children.

Abnormal sinusal origin is of far more significance, particularly if associated with a so-called intramural course. Almost without exception, even if they have an anomalous origin, the coronary arteries will continue to arise from one or the other of the two sinuses adjacent to the pulmonary trunk. These can be distinguished as being to the right hand or left hand of the observer, with convention dictating that the right-handed sinus is considered to be no. 1, and the left-handed sinus no. 2. (4, Fig 5.)

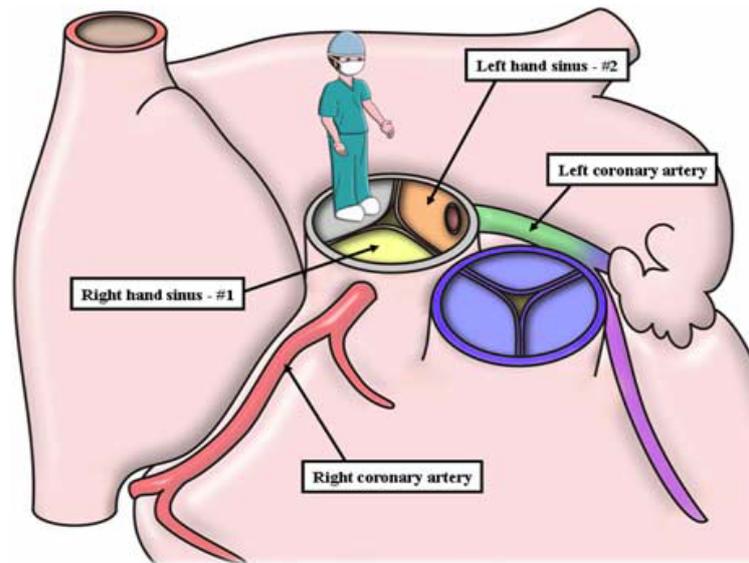


Figure 5.: The cartoon shows how, figuratively speaking, and when the coronary arteries arise from the aortic valvar sinuses adjacent to the pulmonary root one of the sinuses will always be to the right hand of the observer standing in the non-adjacent sinus. This sinus is now defined as being n°1. The other sinus, defined as n°2, is to the observer's left hand. In the normal heart, it is sinus #1 that gives rise to the right coronary artery, and sinus #2 that supports the left coronary artery. When defined in this manner, it is always possible to distinguish the left-handed and right-handed sinuses, irrespective of the interrelationships of the arterial trunks. (4)

This convention has proved its value for surgeons transposing the coronary arteries as part of the so-called arterial switch procedure (4). It is of equal relevance for those seeking a suitable means of describing the abnormal aortic sinusal origin in the otherwise normally structured heart. The convention also allows description of the anomalous sinusal origin of a major coronary artery from the non-adjacent sinus, albeit that this sinus would then no longer be a non-coronary aortic sinus. It would remain non-adjacent when considered relative to the pulmonary trunk.

1. Anomalous origin of the left coronary artery from the right coronary sinus

The anomalous origin of the main stem of the left coronary artery from the right coronary sinus is rare, but of great clinical importance, because it has been associated with sudden cardiac death in children and adolescent. After its anomalous origin, the left main coronary artery may take one of the four different routes. It can run anterior to the pulmonary trunk, posterior to the aorta, through the tissue plane between the arterial trunks, or through the crest of the muscular ventricular septum. It is those in whom the artery takes a course between the arterial trunks who are at greatest risk for sudden death.

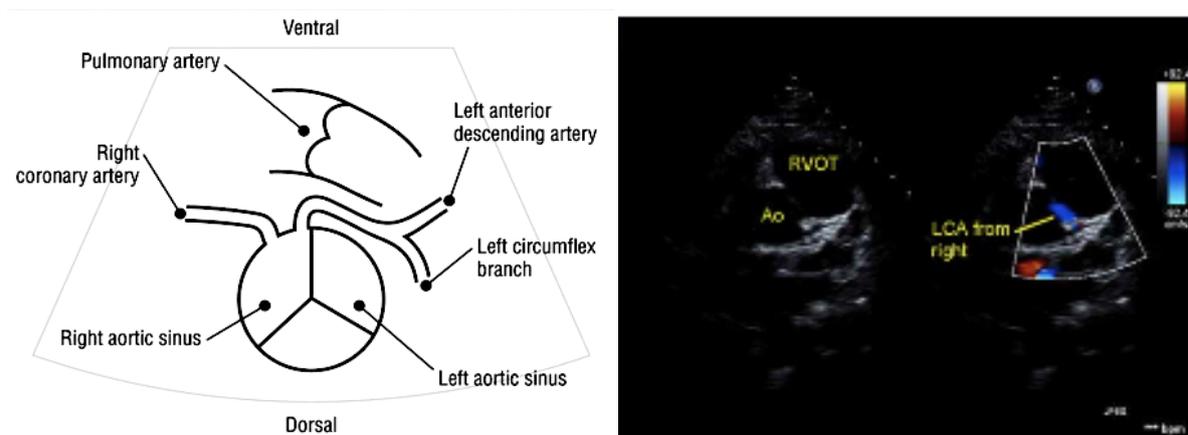


Figure 6.: Parasternal short-axis view in colour-compare mode in a patient with an anomalous aortic origin of the left coronary artery from the right coronary aortic sinus. Although the origin is not well visualised by cross-sectional imaging in this still frame, colour flow mapping suggests that an artery courses from the right, between the aorta and the pulmonary trunk to the left (12)

The mechanism of such sudden death is acute myocardial ischemia, produced by reduced flow in the anomalously arising artery. This occurs most often during exercise, when myocardial demand for oxygen is at its highest, and the increased stroke volume causes outward expansion of the roots of both arterial trunks. The mechanism of obstruction to flow seems to be a combination of compression of the intramural portion of the coronary artery itself, compression of the artery between the aorta and the pulmonary trunk, and worsening of the intrinsic narrowing or kinking at the anomalous ostium.

The definition of the origins and proximal courses of the anomalous coronary artery may often be achieved with echocardiography. The parasternal short-axis view at the base of the heart is the best view to appreciate both the relationship of the origin of the left coronary artery to the zone of apposition between the aortic valvar leaflets guarding the coronary aortic sinuses, its often intramural course, and its relationship to the right ventricular outflow tract. On account of the extreme translation of the coronary arterial origins during the cardiac cycle, the excellent temporal resolution of echocardiography, compared with that of other non-invasive techniques, makes this the diagnostic technique of choice in most cases. However, this diagnosis is often difficult to make in real time. Thus, obtaining a loop that is one to several heartbeats in duration, and then playing this loop back slowly, is often of great help in visualizing the anomalous coronary artery, as well as discerning its relationship to the zones of apposition between the aortic valvar leaflets. Color Doppler should be used to identify any intramural segment of the main stem. In fact, an abnormal jet seen during diastole over the space between the aorta and the pulmonary trunk is often the first indication of an abnormal coronary arterial course. The inter-arterial course can often be seen to advantage in the leftward angled parasternal long-axis view.

2. Anomalous origin of the right coronary artery from the left coronary sinus

It is reported less often than anomalous origin of the main stem of the left coronary artery from the right coronary sinus, but its true incidence is probably greater. It is common for the anomalous right coronary artery to pass between the arterial trunks, although other courses are possible. Such an inter-arterial course provides the anatomic substrate for myocardial ischemia in an identical manner to that with the anomalous origin of the left coronary artery from the right coronary aortic sinus. The anomaly is most often diagnosed by echocardiography, either as an unexpected finding or as part of an evaluation for a patient with chest pain or exercise-induced syncope. It is found using the same principles as for the anomalous origin of the left coronary artery from the right coronary aortic sinus, in that the parasternal short-axis view allows determination of the anomalous origin and proximal course of the right coronary artery. Without careful delineation, the right coronary artery can appear to take its normal origin from the right coronary aortic sinus.

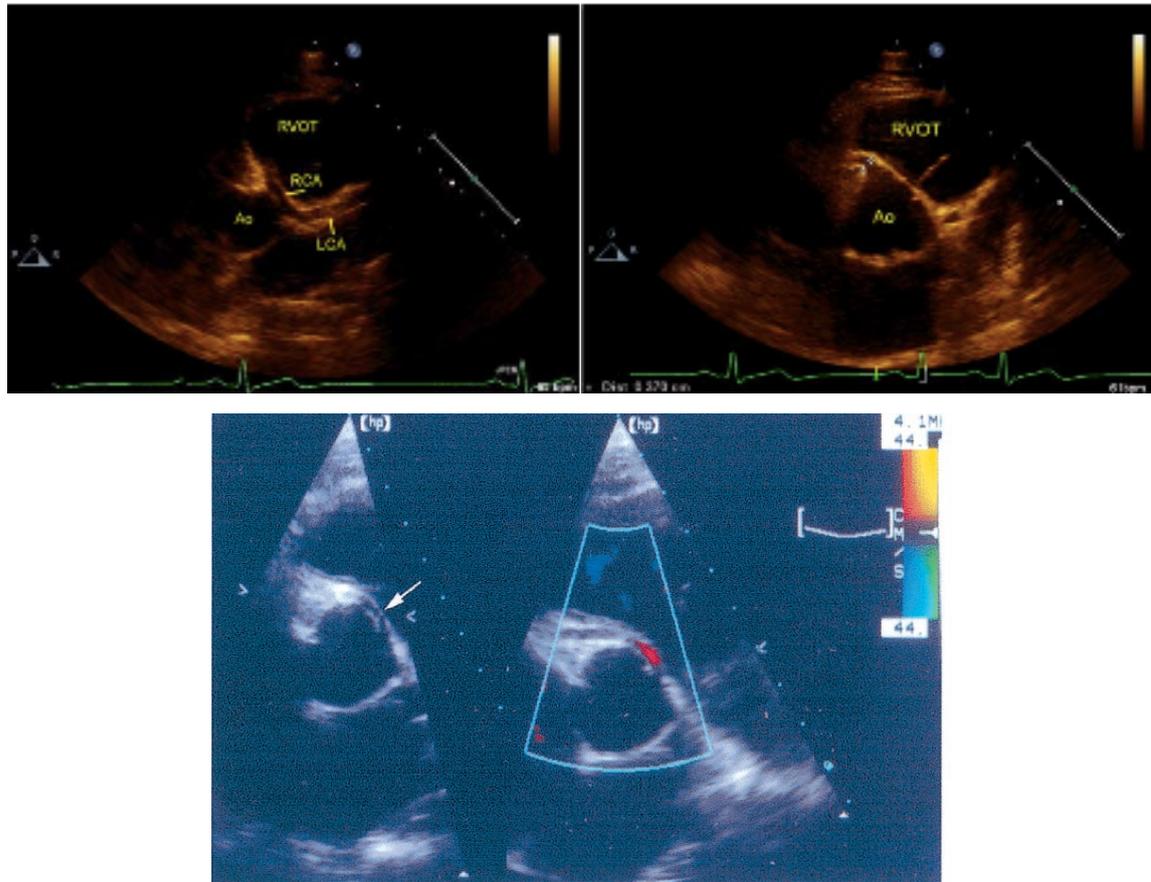


Figure 7.: Parasternal short-axis view demonstrating anomalous aortic origin of the right coronary artery from the left coronary aortic sinus. The right coronary artery can be seen coursing between the arterial trunks; Parasternal short-axis view in the same patient with anomalous aortic origin of the right coronary artery from the left coronary aortic sinus demonstrating that the right coronary artery can appear to have a normal origin (12); origin of the RCA from the left sinus of Valsalva. Abnormal direction of blood flow is shown by color Doppler (red) (13)

When the right coronary artery arises from the left coronary aortic sinus, it can also take an unusually high origin at or slightly above the sino - tubular junction. With such a high origin, take - down of the inter-coronary commissure during an unroofing. Identifying the origin in relation to the sino-tubular junction is best accomplished in the parasternal long-axis view or in a long-axis transoesophageal echocardiographic view. Transoesophageal echocardiography is the imaging modality of choice if such an anomalous aortic origin is suspected but not definitively diagnosed by transthoracic imaging.

3. Solitary coronary artery

This anomaly is characterized by a single coronary artery arising from a single ostium in one of the aortic sinuses of Valsalva and follows the peripheral course and distribution of one or both right and left coronary arteries or an abnormal distribution, in a patient with an otherwise structurally normal heart.

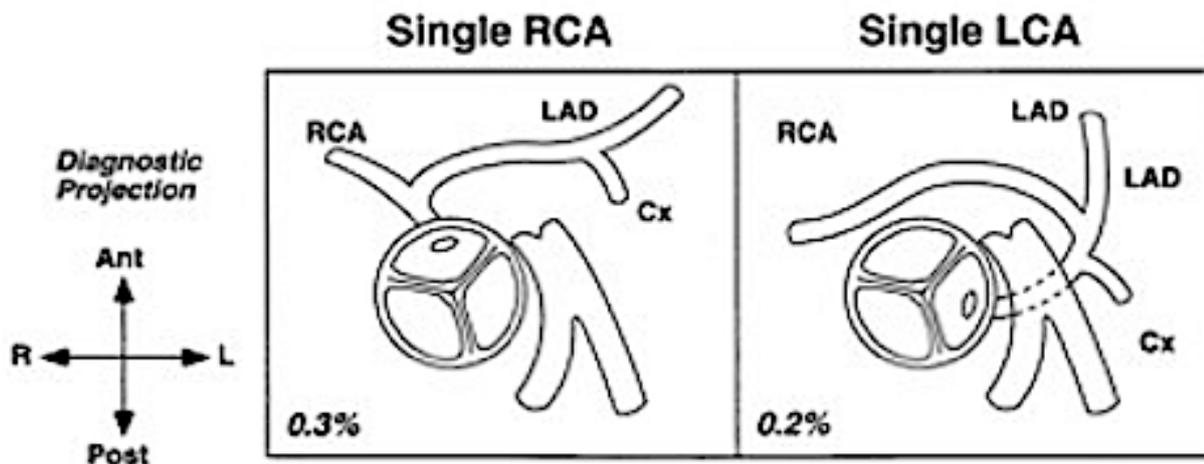


Figure 8.: single coronary artery pattern (8)

The majority of these patients are asymptomatic, thus the anomaly is compatible with a normal life, even if the patients are also known to be at risk of severe ischemic cardiac disease, including infarction. The anomaly is almost always an incidental finding, and when suspected the origin and proximal course of each of the three major coronary arteries should be confirmed in multiple views.

4. Congenital coronary artery termination anomalies

a. *Congenital coronary arterial atresia*

In this anomaly, the left coronary arterial system has a normal epicardial course, but ends blindly, usually in close proximity to the aorta. The left coronary arterial bed is supplied by the right coronary artery. This disorder, ironically coined CALM syndrome for Congenital

Absence of the Left Main, must be distinguished from a single right coronary artery, the latter arrangement carrying a more benign course in most cases. Almost uniformly, patients with atresia of the arterial orifice develop cardiac symptoms similar to the infantile presentation of the Bland–White–Garland syndrome. Associated lesions are unusual, but may include supralvalvar aortic stenosis and ventricular septal defect.

The echocardiographic findings are very similar to those of anomalous origin of the left coronary artery from the pulmonary trunk, except that the left coronary artery does not arise from the pulmonary trunk. Again, it must be emphasized that the artery may appear to arise from the aorta, especially when its atretic end is in close approximation to the left coronary aortic sinus. Demonstration of flow in the coronary artery is extremely helpful, when retrograde flow in at least part of the cardiac cycle should arouse suspicion. The myocardium may also have the abnormal appearance of non-compaction.

b. Coronary artery fistula

Coronary artery fistula is a rare, usually solitary, anomaly accounting for approximately 0.4% of congenital heart defects. The great majority of cases in the pediatric population are congenital in nature, possibly arising from persistence of sinusoidal_ coronary arterial connections. Congenital fistulas most frequently arise from the right coronary artery system and the great majority (90%) exit into right heart structures, including the vena cava, coronary sinus or pulmonary arteries. The site of termination of the fistula may be a single entry site, multiple entry sites, a plexiform communication or a side-to-side communication between the coronary artery and a cardiac chamber. The fistula has the potential to produce a coronary artery steal, and may cause myocardial ischemia or rarely infarction. Children are usually asymptomatic at the time of diagnosis, which can often be confirmed with identification of CAF origin and exit sites by echocardiography. Two – dimensional echocardiography may show dilation of the proximal portion of the coronary artery feeding the fistula and chamber dilation consistent with the fistula physiology. Fistula detection is greatly helped by color Doppler imaging. Small fistula are usually unexpected findings, first detected by abnormal diastolic flow into right heart chamber or into pulmonary arteries, without dilation of any cardiac chamber or of the proximal coronary artery.

3. ACQUIRED CORONARY ANOMALIES: KAWASAKI DISEASE

Echocardiography is highly sensitive, specific and non invasive means of assessing the proximal coronary arterial system and thus occupies a central role in the diagnosis workup (5,11,13,14).

The goal of the examination is:

- identification of coronary artery anomalies
 - o perivascular inflammation
 - o ectasia
 - o localization of anomalies
 - o classification of severity
 - o identification of thrombus
 - o identification of stenosis
- identification of valvar involvement
- identification of myocardial involvement
- identification of pericardial involvement.

Coronary artery imaging should be performed at the highest feasible transducer frequency. Reducing two-dimensional gain and dynamic range will usually improve demonstration of the endovascular lumen (5,11,14). Imaging at a greater depth will also enhance anatomic visualization (5,11,14).

Coronary aneurysm should be characterized both qualitatively and quantitatively.

Coronary artery ectasia is defined as coronary enlargement without aneurysm

Coronary aneurysm are described (5,14):

- as saccular when the axial and longitudinal dimensions are nearly equal,
- as fusiform when the longitudinal dimension is longer with the aneurysm tapering at its ends

Coronary artery dimensions should be assessed relatively to published normal values (z-scores), understanding that they exist for RCA, LMCA, and LAD. Internal vessel diameters should be measured from inner edge to inner edge. Aneurysm have been classified as (11,14):

- small – medium (>3 mm but<6 mm, coronary z-score of +3 to +7)
- large (>6 mm)
- giant (> 8 mm).

Japanese Ministry of Health has defined coronary artery dilation (11,14):

- internal diameter >3 mm in children aged under 5 years
- internal diameter > 4 mm in children aged 5 years or above
- internal diameter 1.5 times the diameter of an adjacent coronary segment.

It is important to make every effort to image each segment of the coronary artery system to determine the sites involved when coronary artery aneurysm occur. The most common sites for coronary artery aneurysm are (11,14):

- left anterior descending artery
- proximal right coronary artery
- left main coronary artery
- left circumflex artery
- distal right coronary artery
- proximal posterior descending artery.
- Usually multiple lesions involving right and left coronary systems are seen.

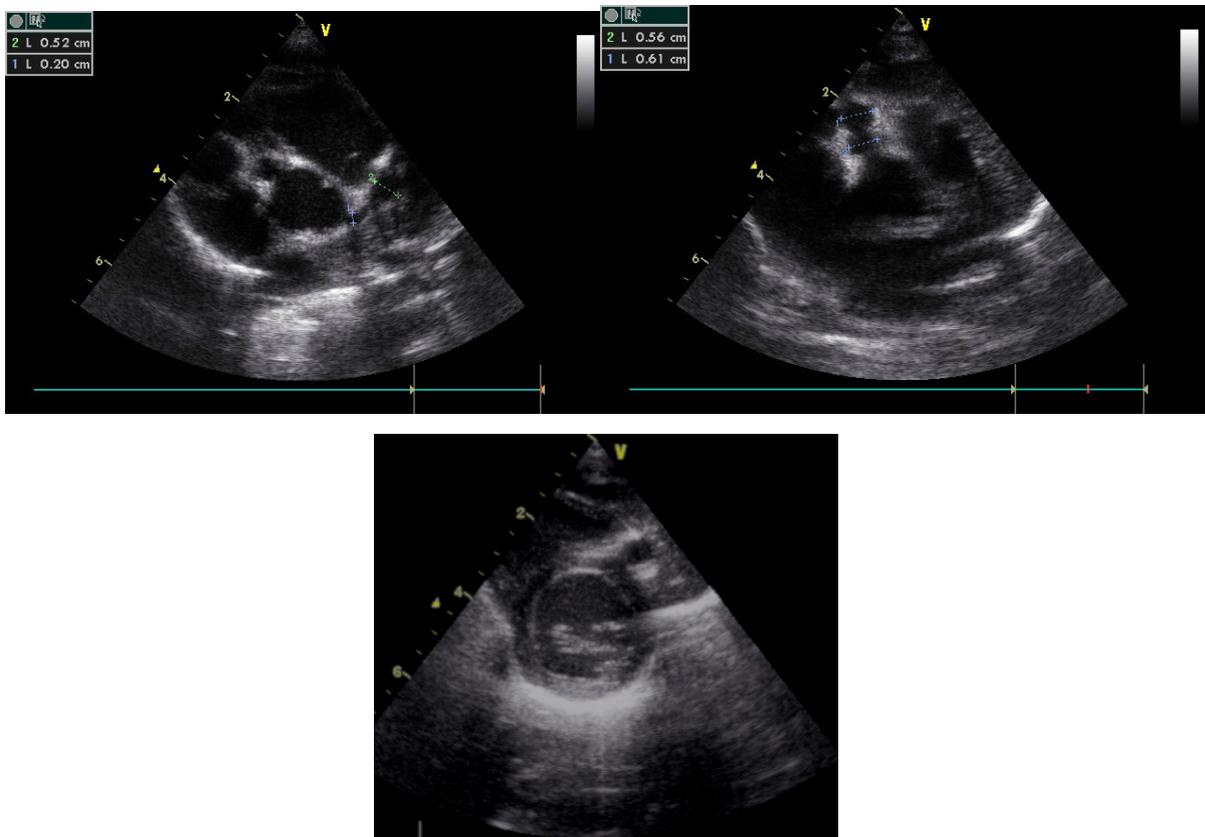


Figure 9.: coronary artery aneurysms in a 2 months old boy

One commonly performed practice that should be stringently avoided when imaging coronary arteries is that of obtaining still frames of the arterial origins, and making diagnostic decisions based upon these still images. With the anomalous aortic origin of a coronary artery from the pulmonary trunk or the aorta, the proximal coronary artery often approaches the appropriate sinus of Valsala very closely, and resolving the thin tissue between the coronary arterial and the aortic lumens can be difficult or impossible in a still frame image. It is easy, therefore, to be falsely assured of a normal origin of the coronary artery when, in fact, the coronary artery arises anomalously from another location. This is particularly true of the right coronary artery

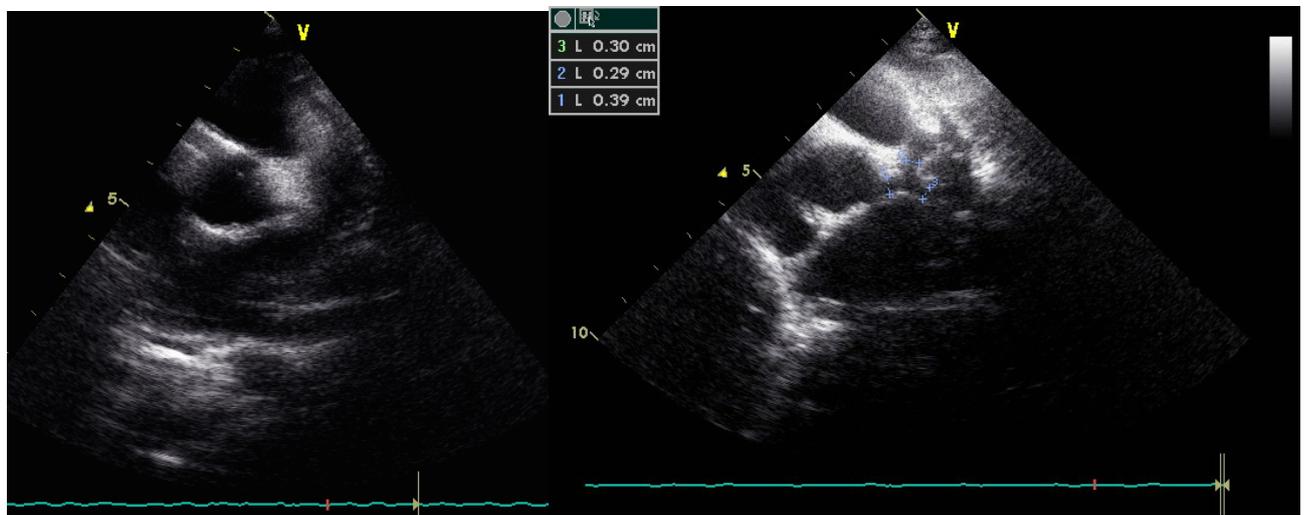


Figure 10.: coronary artery dilation in a 4 y.o. young boy.

CARDIAC MAGNETIC RESONANCE IMAGING IN PEDIATRIC AGE

1. CORONARY MRI ANGIOGRAPHY (MRA): TECHNICAL CONSIDERATIONS

Coronary artery assessment by cardiac MRI has the advantage of (5, 15):

- 3 D image acquisition;
- Post processing analysis in any plane
- Better signal-to-noise ratio between coronary blood pool and surrounding tissues.

The small caliber of the coronary vessels, as well as the elevated anatomical variability, cardiac and respiratory motion pose major challenges to coronary MRA and require dedicated techniques for image quality optimization.

a. Sequences and vessel wall visualization

The first approaches to coronary artery angiography were attempted by 2-dimensional (2D) gradient-echo techniques (16,17). One slice was acquired in 16 heartbeats during a single breath-hold. Patients could breathe between acquisitions. Two-dimensional coronary MRA using breath hold is subject to several limitations such as partial volume effects, poor overall signal-to-noise, misregistration between adjacent slices due to inconsistent breath holding, and a long overall acquisition time.

Improved hardware and sophisticated pulse sequences have allowed three-dimensional coronary MRA that acquires a complete volume during a single breath hold (18). Three-dimensional data acquisition allows for extended coverage of anatomical structures, improved signal-to-noise ratio, sophisticated k-space sampling schemes, and isotropic spatial resolution. 3D techniques adopting a whole heart or target volume approach became feasible after the introduction of navigator techniques (15, 19, 20, 21). These can visualize the entire heart, and be performed in less than 30 minutes. Three-dimensional data acquisition diminishes the operator dependency and allows for a variety of post processing techniques such as maximum intensity projection (MIP), curved multiplanar reformatting (curved-MPR), and volume rendering techniques (15, 20, 21). The main disadvantage is the reduced contrast between blood and the myocardium due to the reduction of inflow effects. All state-of-the-art techniques use bright blood imaging, such as gradient echo, or more commonly, steady state free precession (SSFP), known under the acronyms FISP, True-FISP (Fast Imaging with Steady-state free Precession, balanced FFE (balanced Fast Field Echo), and FIESTA (Fast Imaging Employing STEady-state Acquisition) (15, 20, 21). SSFP is characterized by an alternating phase of the excitation pulse combined with the application of time-balanced gradients for all gradient directions: slice selection, frequency, and phase encoding (15, 20, 21). The practical application of SSFP is enabled by modern MR systems that allow short repetition times and short echo times. SSFP provides high signal intensity for tissues with a high T2/T1 ratio (e.g., blood), with a relative independency of the repetition time and flow artifacts. The SSFP sequence however, is sensitive to field inhomogeneities, which makes field shimming an important issue. In two studies, SSFP was compared to gradient-echo imaging, and improved endocardial border delineation was reported for the SSFP images, which facilitated automated edge detection during cardiac function analysis. The potential use of SSFP for coronary MRA was shown in a study comparing conventional FLASH (fast low-angle shot) to three-dimensional true-FISP. The signal-to-noise and contrast-to-noise were improved with 55% and 178% respectively for the SSFP acquisitions.

Conventional bright-blood coronary MRA is usually based on gradient-echo

sequences with or without exogenous contrast enhancement. Contrast in bright-blood techniques can be improved with the application of various prepulses such as an inversion prepulse, spectral fat-suppression, and T2-preparation. Black-blood spin-echo coronary MRA offers particular advantages as compared with bright-blood coronary MRA, such as a high contrast-to-noise ratio, reduced susceptibility to turbulent flow allowing improved coronary artery visualization. In black-blood imaging, the blood signal is selectively suppressed in favor of the other tissues by using a double-inversion recovery spin-echo sequence. This resulted in the visualization of the coronary artery tree with 390 to 700mm in-plane resolution, within a reasonable 10 minutes acquisition time. Three-dimensional black-blood imaging allows high contrast between the blood pool, the vascular wall, and the perivascular tissues such as fat and myocardium, which can be traded for improved spatial resolution. The black-blood imaging sequence reduces blood flow artifacts and motion-related artifacts that are characteristic of nonblack-blood techniques, which can be particularly useful in regions of focal stenosis.

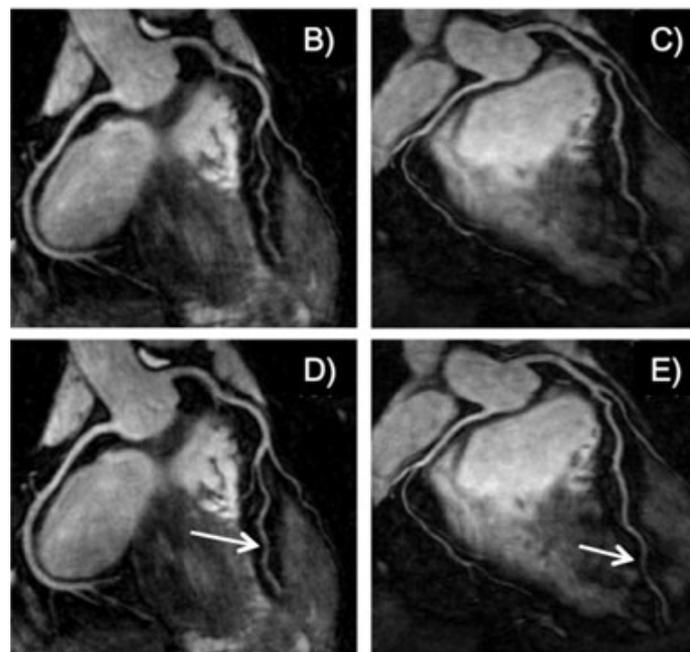


Figure 1.: Examples of right and left coronary artery visualized with the 2Dnav sequence (15)

b. Compensation for cardiac and respiratory motion

Cardiac motion occurs in both systole and diastole, but is said to be minimal in mid-diastole (at diastasis). Cardiac motion correction is therefore usually achieved by timing the acquisition to the mid-diastolic phase of the cardiac cycle. Cardiac magnetic resonance techniques used to visualize the coronary arteries generally use a “free breathing navigator” approach (15, 20, 21), where the motion of the diaphragm is monitored, and imaging data is obtained only at certain points in the respiratory cycle. In addition, acquisition of imaging data is timed to the quiescent period of the cardiac cycle during diastole. Both these methods are used to image the coronary arteries when there is little motion and, in the case of imaging in diastole, where the artery contains the most blood (15, 21). The contraction of the heart is not symmetrically distributed over the various areas of the cardiac muscle, which causes the coronary artery segments to be subject to different rates of motion, depending on their anatomical location. Coronary artery motion has been measured using dynamic x-ray angiography (22). Authors concluded that there is considerable variation of motion patterns, motion ranges and motion velocities among individual patients (22). The rest period was strongly heart-rate dependent (22). On average, the right coronary artery had greater movement and greater velocity as compared with the left coronary artery, up to a factor of two for the proximal segments. An important finding of this study was that the coronary arteries returned to the same location from heartbeat to heartbeat during the rest period, which is an absolute requirement to perform coronary MRA (15, 21, 22). The optimal time delay between the electrocardiographic trigger point (the R-wave of the QRS complex) and the acquisition window can be determined in various ways. Several studies have empirically determined the mid-diastolic period in the cardiac cycle using data from heart sound recording, carotid arterial pulse tracing, electrocardiography, and multiphase gradient-echo breath

hold MRI, based on the assumption that the systolic part of the cardiac cycle has a relatively constant duration.

In a later study some authors recommended to perform a pre-scan prior to the actual coronary MRA acquisition for the estimation of the electrocardiographic trigger delay of each patient. The efficacy of ECG sensing during MR acquisitions depends on both the patient and the operator. Imprecise ECG triggering may result from traditional noise sources such as poor electrode contact, lead wire noise, patient movement, muscle contraction, ECG baseline drift, or amplitude modulation due to respiration. The main interference is caused by a false triggering on the ST segment instead of the R-wave due to the magnetohydrodynamic effect that results from the magnetic field and induces voltages that superimpose the ST-segment voltage. The rise of the ST segment eventually results in false triggering of the T-wave in favor of the R-wave. In contrast to conventional scalar ECG, a vector cardiogram system uses multiple ECG channels simultaneously to reconstruct a vector cardiogram, in which the QRS loop is spatially separated from the MR related artifacts (23). In addition to the cardiac motion, coronary MRA suffers from artifacts due to respiratory motion. As the heart lies on the diaphragm, it translates during the respiratory cycle in a supero-inferior direction. Artifacts caused by respiration movements can be minimized with the aid of several respiratory motion correction approaches.

Currently, two major approaches are distinguished: breath holding and free-breathing navigator gating. During breath holding, the position of the diaphragm is immobilized and thus the translational shift of the heart. The main limitation of breath holding is the acquisition time, which is restricted particularly in critically ill patients (18,22). The VCATS technique relies on a three-dimensional localizer scan that covers the entire heart in a single breath-hold. The three-dimensional information thus obtained is used to determine the optimal planes for high - resolution three - dimensional scans that specifically target the coronary segments during breath-hold acquisitions.

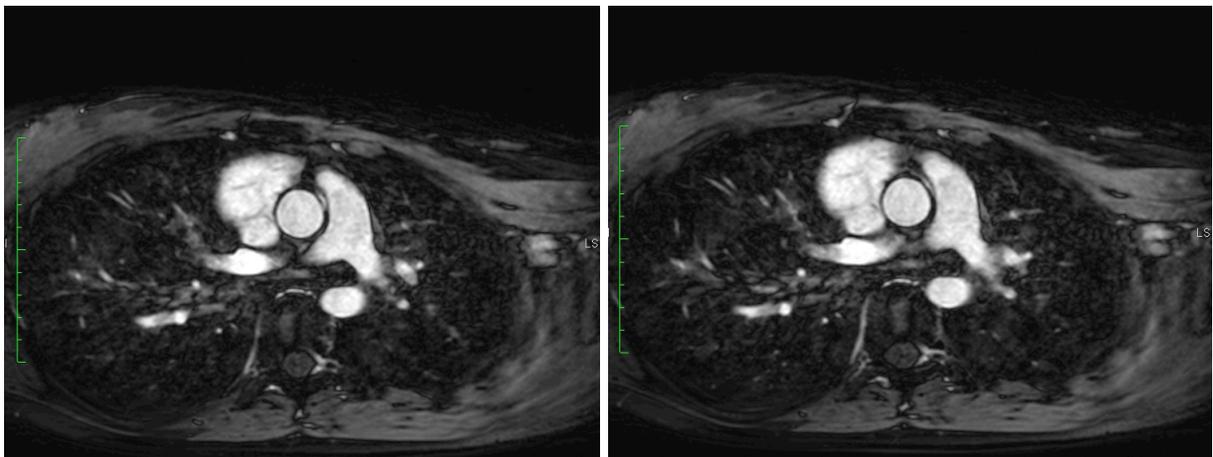
The free-breathing navigator-gating approach is another way to correct for respiratory motion. During navigator gating, the position of the right hemi-diaphragm is deduced in real-time from a navigator pencil beam acquisition (24). A gating acceptance window is predefined. Image data that are acquired while diaphragm position is within the acceptance window are accepted for filling of the k-space,

all other data are discarded. The gating window is chosen at the end - expiratory motion range (24),

as this is assumed to be the period of the least diaphragmatic movement. The navigator beam is usually placed on the dome of the right hemi-diaphragm. Navigator gating implies that only a small fraction of the total imaging time is used for actual data acquisition. This causes the overall imaging time to be prolonged, depending on the navigator efficiency: an acquisition that theoretically takes 4 minutes, will take the twice the amount of time with a navigator acquisition efficiency of 50%. The navigator efficiency depends on several factors, including the anatomical location of the navigator beam and the regularity and nature of the patient's breathing pattern. A clear patient instruction and the patient feeling comfortable therefore play an important role in improving the navigator efficiency. In practice, an average navigator efficiency of 40% – 60% can easily be obtained, which however may drop down to 20%–30% in patients that have trouble with regular breathing. The efficiency of navigator gating can be extended by the application motion adapted gating (MAG). The clinical feasibility of MAG has recently been illustrated in a study comparing three-dimensional gradient-echo MRA combined with motion-adapted gating to conventional x-ray angiography. A way to optimize the image quality of navigator - gated acquisitions is the use of slice tracking. This method moves the position of the image slice according to the observed motion of the diaphragm within the constraints of the acceptance window. It has been shown that the supero - inferior motion of the coronary arteries is approximately 60% of the superoinferior displacement of the diaphragm. During the breath hold, the most important central portion of the k-space is sampled; the remaining k-space data are acquired using a conventional free-breathing navigator method. This combined approach yielded images with increased signal-to-noise ratio and may be a valuable technique for contrast-enhanced (first-pass) coronary MRA.

2. CORONARY MRI ANGIOGRAPHY (MRA) IN CONGENITAL AND ACQUIRED CORONARY ANOMALIES IN THE PEDIATRIC PATIENT

Cardiac magnetic resonance (CMR) of the pediatric patient involves a unique set of technical challenges above and beyond those encountered in adult imaging. Anatomical structures are smaller, demanding greater spatial resolution; heart rates are high, demanding higher temporal resolution; and patients may be sedated or uncooperative, rendering breath-hold imaging strategies useless. Despite these difficulties, CMR offers several advantages over other imaging modalities, including soft tissue contrast, lack of ionizing radiation, a capacity for true three-dimensional imaging, accurate flow quantification, and freely selectable imaging planes. These advantages and continued advances in MR hardware, software, and imaging techniques are bringing CMR into more widespread use in pediatric cardiology



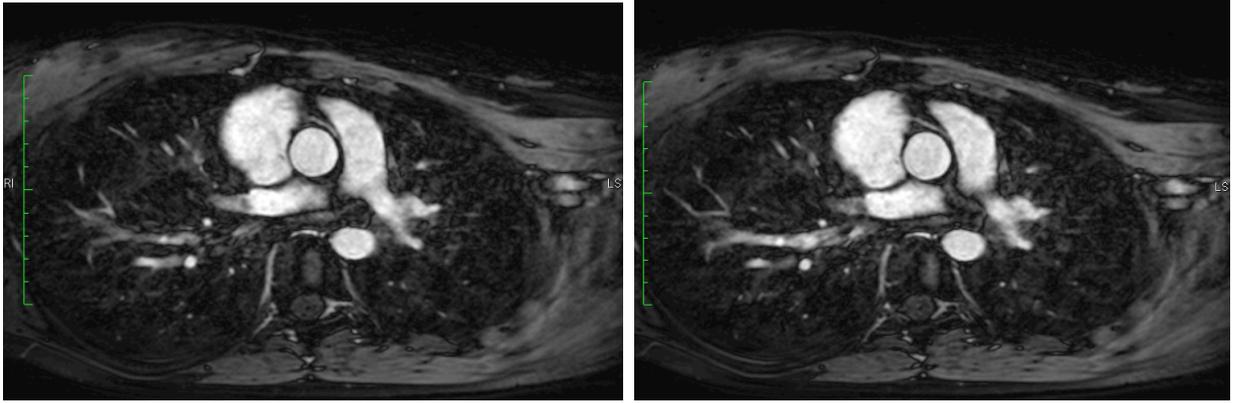


Figure 2.: anomalous origin of the right coronary artery, small in caliber, which passes between ascending aorta and right ventricular outflow tract.

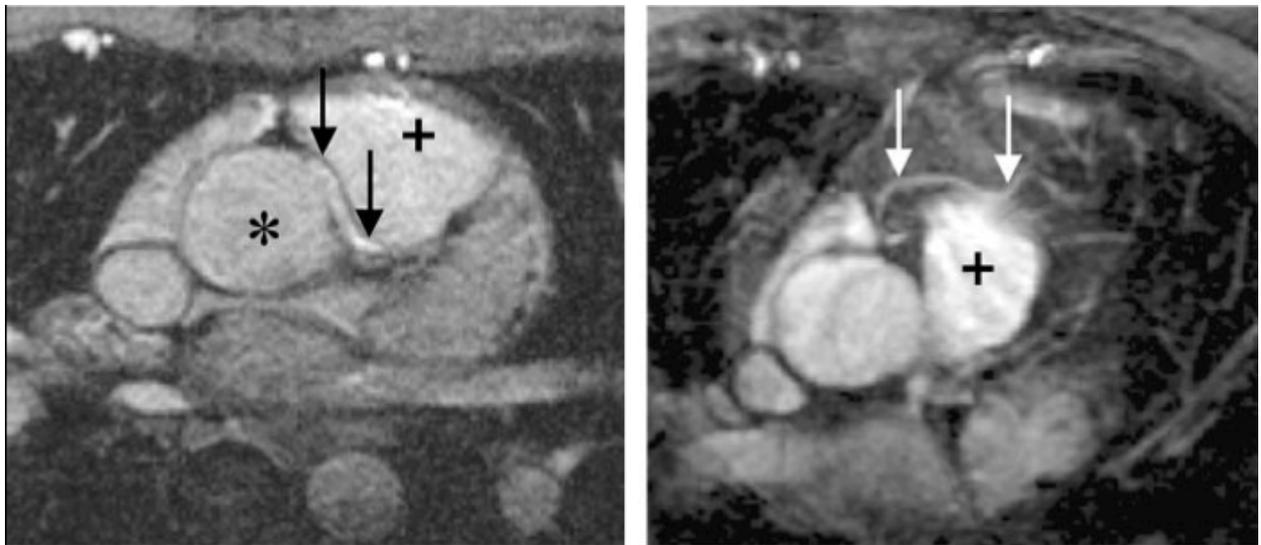


Figure 3.: Image shows an anomalous left coronary artery (black arrows) that arises from the right sinus of Valsalva and passes between the aortic root and the RVOT in an intraarterial course. The left anterior descending artery (white arrows) courses anteriorly to the RVOT (25)

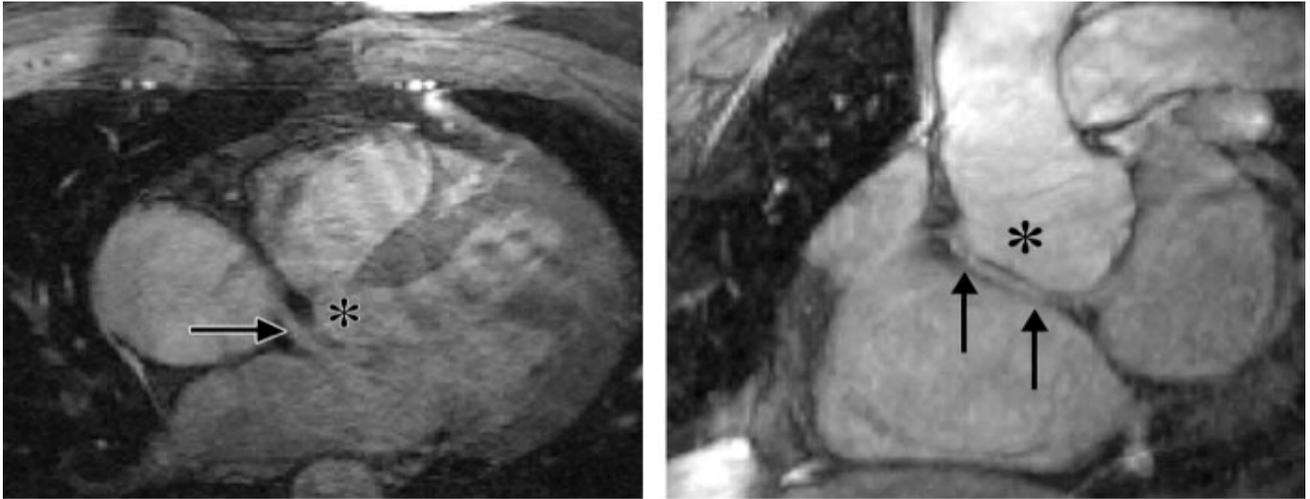


Figure 4.: Anomalous circumflex artery (arrow) courses posterior to the aortic root. Oblique sagittal slab positioned in plane with the anomalous vessel. The anomalous circumflex artery (arrows) courses an inferior course as a branch from the right coronary artery. It passes inferior and posterior to the aortic root (25).

One of the first major studies of modern cardiac magnetic resonance used to image coronary appeared in 1995, when McConnell et al (26) identified 14 of 15 anomalous coronary arterial lesions correctly as validated against angiography. This was then followed rapidly by numerous other reports, one of them (25) where cardiac magnetic resonance identified the anomalous course of the coronary arteries in 26 patients, with 15 confirmed by angiography, and 11 where angiography could not define the anatomy with certainty, 8 of these running an intramural course. Other authors had examined a larger series of 65 children, providing unambiguous diagnosis of coronary arterial anomalies in 62, with the other 3 having significant arrhythmias, which precluded imaging. There are also multiple reports of cardiac magnetic resonance performed for lesions other than anomalous left coronary arteries in children, such as Kawasaki's disease (26 - 32), and in the setting of other congenital cardiac malformations (33 -34).



Figure 5.: the right coronary artery (*RCA*) in a 4-month-old patient who had undergone Norwood I operation (33)

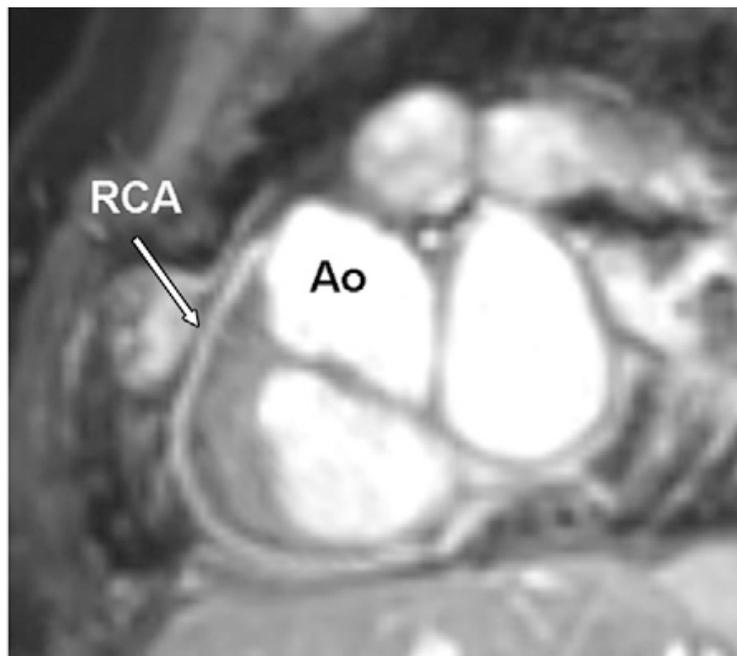


Figure 6.: 1-year-old patient who had undergone repair of tetralogy of Fallot (33)

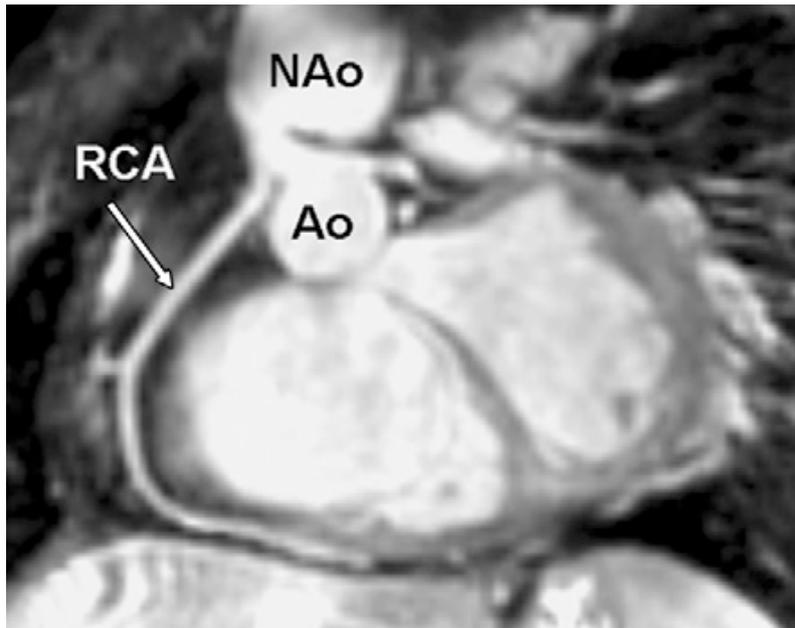


Figure 7.: 10-year-old patient who had undergone Fontan procedure (33)

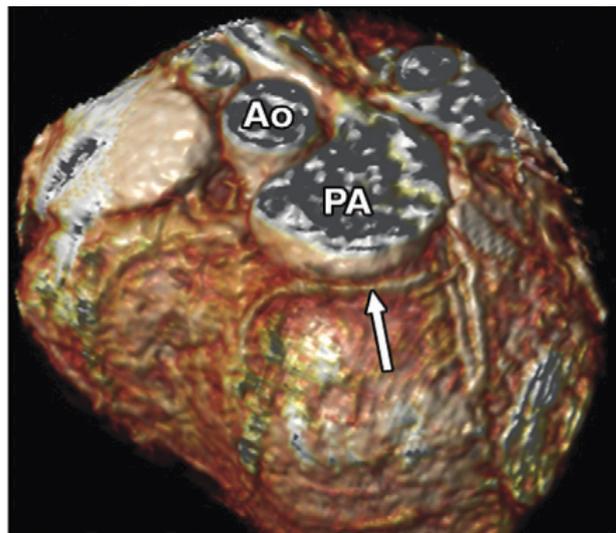


Figure 8.: Volume-rendered whole-heart image in a 3.5-year-old patient, who had undergone repair of tetralogy of Fallot. An accessory branch (arrow) from the left anterior descending coronary artery runs across the right ventricular outflow tract (33)

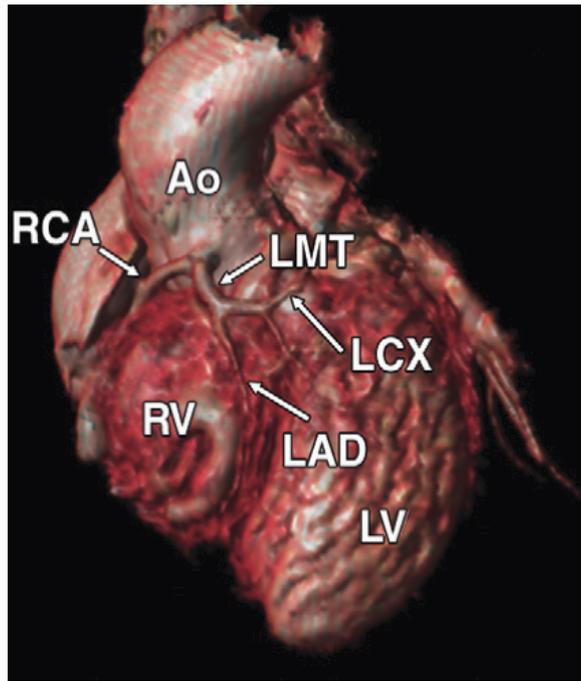


Figure 9.: Volume-rendered whole-heart image in a 3-year-old patient with a hypoplastic right ventricle who had undergone Glenn and Fontan procedures shows a single coronary artery. The left main trunk (LMT) arises from the right coronary artery (RCA). (33)

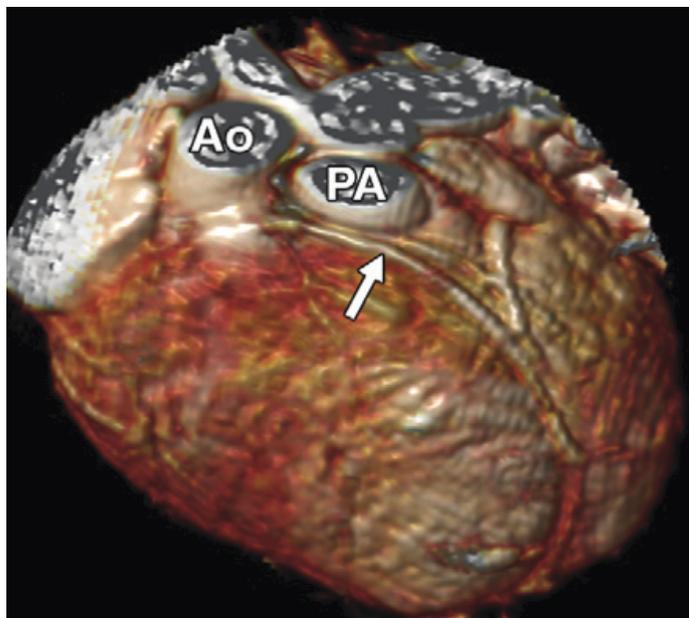


Figure 10.: Volume-rendered whole-heart image in a 3.7-year-old boy with double-outlet right ventricle, ventricular septum defect, and subvalvular pulmonary stenosis. The boy had undergone creation of a right Blalock-Taussig shunt. The left anterior

descending coronary artery (arrow) arises from the aorta (Ao) anterior to the pulmonary artery (PA). (33)

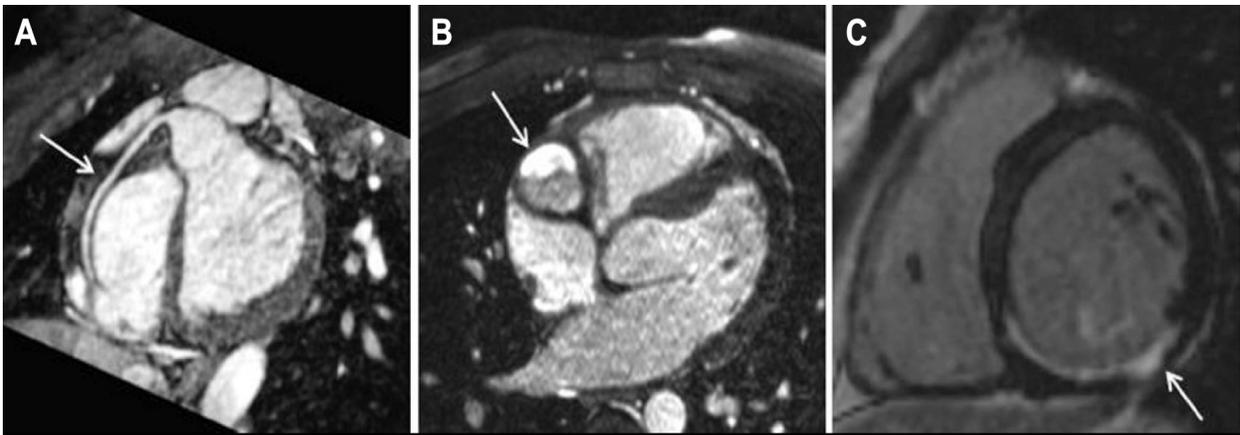


Figure 11: Examples of cardiac MRI images from patients with Kawasaki disease, showing a normal right coronary artery (A), a giant aneurysm of the right coronary artery with thrombosis (B), and a basal inferoseptal-inferior myocardial infarction (C). (34)

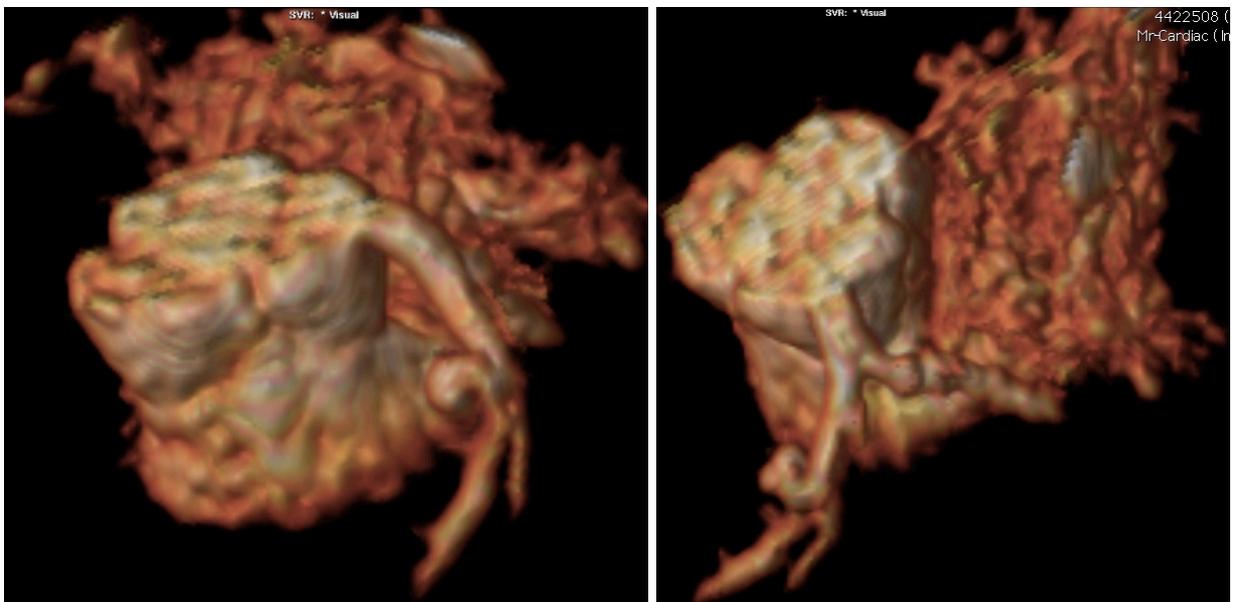


Figure 12.: volume rendering of left coronary artery arteries in a 7.5 y.o girl with previous Kawasaki disease, and with LAD saccular aneurysm that evolved in a smaller aneurysm separated from the lumen.

The appeal of cardiac magnetic coronary arterial system in children

resonance goes beyond visualizing the with congenitally malformed hearts, as

it also makes it possible to assess the effect of coronary arterial flow of blood. Cardiac magnetic resonance can evaluate the myocardium for scar tissue that may have resulted from coronary ischemia utilizing delayed enhancement after injection with gadolinium (5,11), a magnetic contrast agent. Indeed, the technique has been used for that purpose in Kawasaki's disease (26 – 32)), and in patients subsequent to repair of transposition using the arterial switch (34).

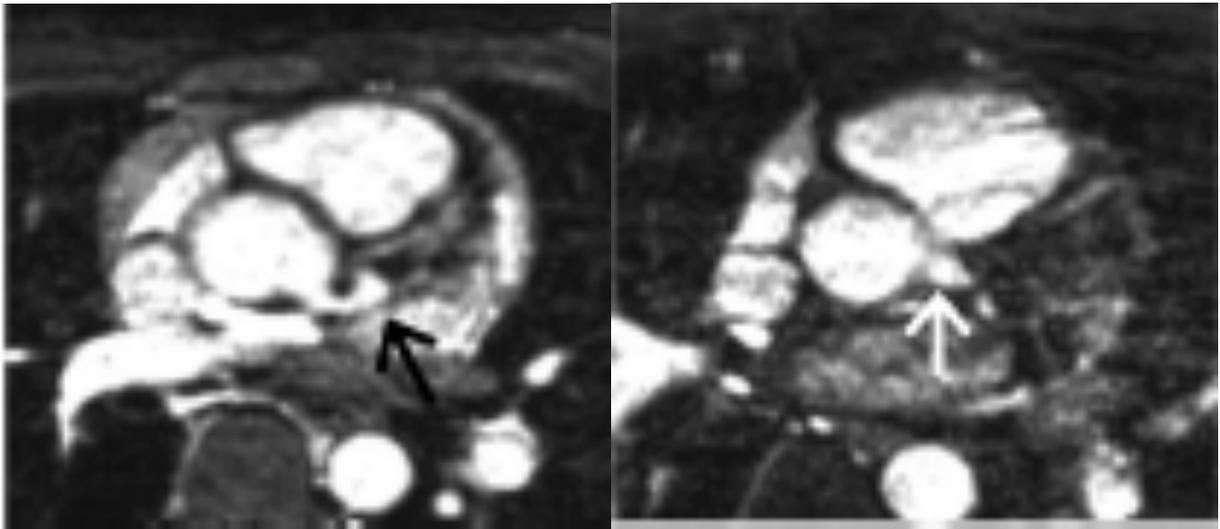


Figure 13.: left coronary artery bifurcation aneurysm (5.1 mm) and left main coronary artery aneurysm (5.8 mm); (31)

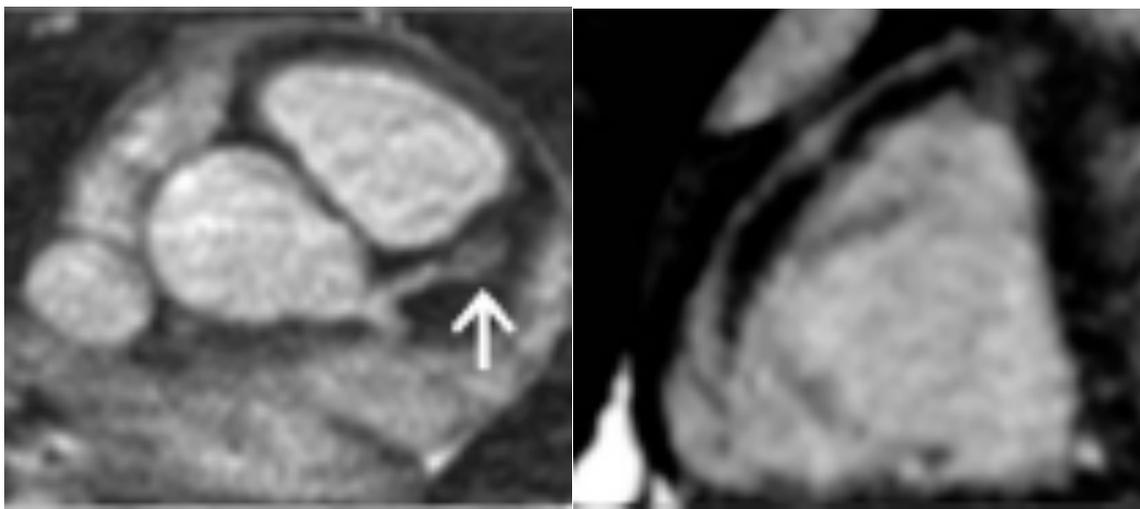


Figure 14.: left anterior descending coronary artery aneurysm (4.9 mm); right coronary artery aneurysm (5.1 mm) (31)

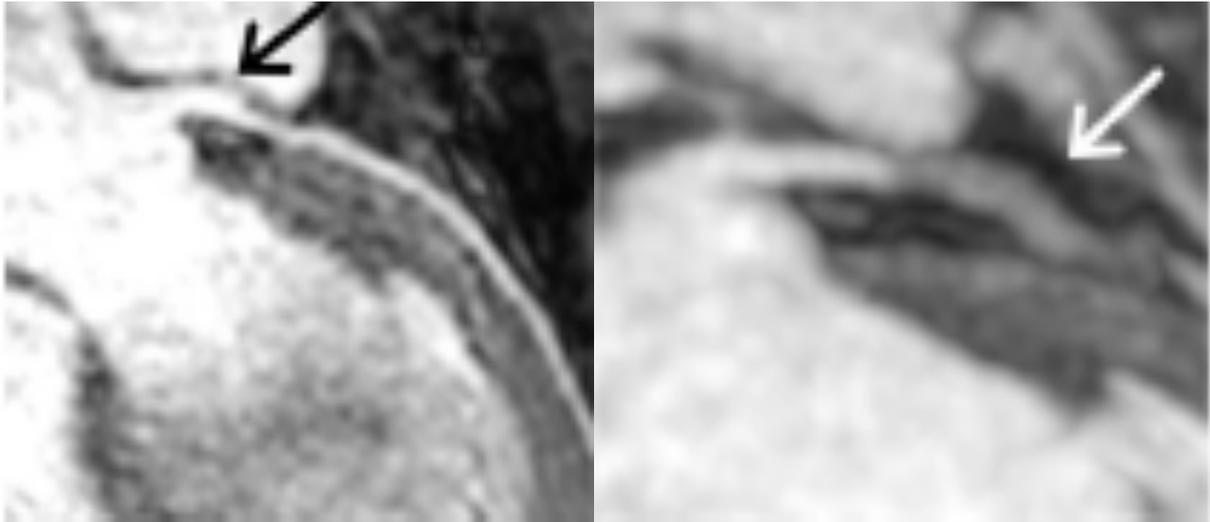


Figure 15.: proximal left coronary artery aneurysm (5.8 mm); left anterior descending coronary artery aneurysm (4.8 mm). (31)

Adenosine stress perfusion cardiac magnetic resonance is complementary to delayed enhancement, and is used to detect regional defects in myocardial perfusion (31). Another complementary technique is measuring the coronary flow reserve by estimating flow in the coronary sinus with phase-encoded velocity mapping (5, 11). Further, the use of cine cardiac magnetic resonance and myocardial tagging makes it possible to identify regional abnormalities of wall motion, which may be related to the affected coronary artery (5, 11).

The primary advantages of coronary MRA over X-ray catheter angiography are that it is non-invasive and does not expose the patient to ionizing radiation. These features are particularly important in pediatric patients, motivating the continued development of the technology. Non-invasive coronary angiography by X-ray computed tomography (CTA) is gaining in clinical importance but also carries the risks of X-ray exposure. Coronary MRA has proven to be technically challenging due to the small size of the vessels and the effects of cardiac and respiratory motion. The most common imaging strategy used currently is a segmented, ECG-triggered 3D SSFP sequence with data acquisition timed to the relatively quiescent phase of mid-diastole. Navigator-echo respiratory gating is used both to avoid respiratory motion, as well as to prospectively adjust slice position based on diaphragm position.

Coronary MRA is still in an active state of technical development, but it has found important pediatric applications in the assessment of anomalous coronary artery origins (33, 34), and in the serial evaluation of coronary artery aneurysms in Kawasaki disease patients (26 - 32).

CONCLUSIONS

Imaging is fundamental for the diagnosis of coronary arteries anomalies. Imaging outlines anatomy and physiology, evaluates the consequences of interventions and helps guide prognosis. However, no single available imaging modality fulfils these roles for all patients and anomalies. Therefore, assessment for coronary arteries must involve a variety of modalities that can be used in a complementary fashion, and that together are sensitive, accurate, reproducible, and cost effective, whilst minimizing harm. Coronary artery anomalies pose an array of diagnostic challenges. Currently, echocardiography is the first-line imaging method for delineating the origin and course of the proximal portion of the coronary arteries, particularly in neonates and infants. However patient size, body habitus, and acoustic windows, along with inability to visualize distal coronary arteries are all limiting factors when the children grow. Conventional angiography, or CT scan angiography, are currently the modality of choice when echocardiography becomes progressively more difficult, when acoustic windows are poor, the course of the distal portion of the coronary arteries is needed. Due mainly to the risks associated with ionizing radiation iodinated contrast they carry, they have some limitation in the pediatric population, particularly in the setting of anomalies requiring a long-life follow-up with serial imaging exams. With the evolution of Cardiac MRI, it is now possible to definitively evaluate the proximal and distal coronary arteries with volume-scanning MRI technique. using real-time, free-breathing navigator technique both the proximal and distal coronary arteries. CMR provides a powerful tool, giving anatomical and physiological data, assessment of coronary arteries, cardiac function, viability, perfusion and cardiac anatomy, with a high spatial resolution and three-dimensional dataset, allowing reconstruction of data in any imaging plane information that echocardiography and catheterization alone cannot provide. Echocardiography combined with coronary MR angiography offers a valid non-invasive radiation-free alternative.

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