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Original Article

Echocardiographic presentation of anomalous origin of the left coronary artery from the pulmonary artery*

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Abstract In the 1970s, diagnosing anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) was often uncertain using imaging alone; however, with the advances in high-frequency transducers, advanced image processing, and other ultrasound modalities such as Doppler colour flow imaging, tissue Doppler imaging, and speckle tracking to asses regional wall motion abnormalities, modern echocardiography now permits accurate diagnosis of ALCAPA with greater certainty. Although many consider ultrasound to be the only imaging test necessary if there is a question as to the diagnosis, other imaging modalities such as MRI, CT, and cardiac catheterisation with angiography remain valuable complementary tests, especially in older patients.

Keywords: Ultrasound diagnosis; ischaemic heart disease in children; Bland, White, and Garland syndrome

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ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY from the pulmonary artery (ALCAPA) is a rare but important and life-threatening abnormality. When it presents in infants and children, it is associated with substantial morbidity and mortality if not treated promptly. In 1886, for the first time, Brooks[1] described two cases of an anomalous coronary artery. Abbot[2] later described the clinical findings of this disorder; her article was published by William Osler in 1908, and was subsequently revised in Osler's Modern Medicine in 1927. She published a major series in her Atlas of Congenital Cardiac Disease, noting eight cases with this anomaly.[3]

Incidence

In 1959, Keith[4] estimated that the incidence of the anomaly was in 1:300,000 live births. Alexander and Griffith[5] estimated that 1.6% of older children had the anomaly; their estimation was based on a catheterisation study of >38,000 patients. In 1962, Fontana and Edwards[6] described the condition pathologically from 58 post-mortem specimens. Neufeld and Schneeweiss[7] described the condition clinically in their book on congenital coronary anomalies. From a cohort of 2388 children and adolescents, Davis et al found a small number of anomalous origins of the coronary arteries in relatively asymptomatic children. When angiograms, exercise perfusion studies, and/or stress tests were performed, four children (0.17%) were identified with anomalous origin of their coronary arteries.[8]

It is important to note that the spectrum of the anomaly includes its discovery in older patients who remain relatively asymptomatic until they present with coronary disease, and clinical evolution of the disorder varies according to the degree of collateral coronary circulation that develops and several other factors. Mortality is high in patients with ALCAPA without treatment during the 1st year of life, and only 10–15% of the individuals with this defect reach adulthood.[9]

Symptoms, signs, and pathophysiology

As patients with this condition frequently arrive with a differential diagnosis of cardiomyopathy,

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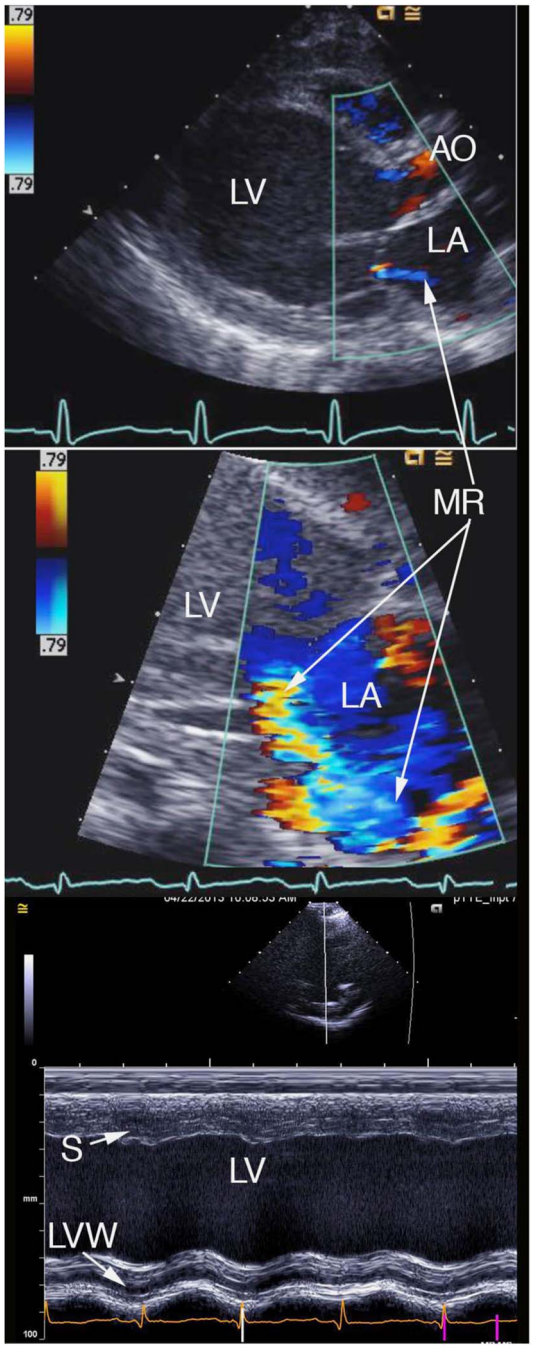

Figure 1.
This composite image shows the heart in the parasternal long-axis view in infants with anomalous origin of the left coronary artery from the pulmonary artery. The top frame *shows the entire image with a markedly enlarged left ventricle (LV). The aorta (AO) and the left atrium (LA) are labelled. Only mild mitral regurgitation (MR) is noted. The LV is dilated. In the* middle frame, *magnified to focus on the left side of the heart, there is substantial MR. The* bottom frame *is an M-mode image taken from a parasternal short-axis reference image. The LV end-diastolic dimension in this neonate is 54 mm, and the systolic dimension is 48 mm, with 11.1% fractional shortening index, all of which are very abnormal. Although the left ventricular posterior wall (LVW) moves appropriately, the ventricular septum (S) moves only minimally and shows inward motion in diastole, reflecting the ischaemic changes in the distribution of the left coronary artery blood supply.*

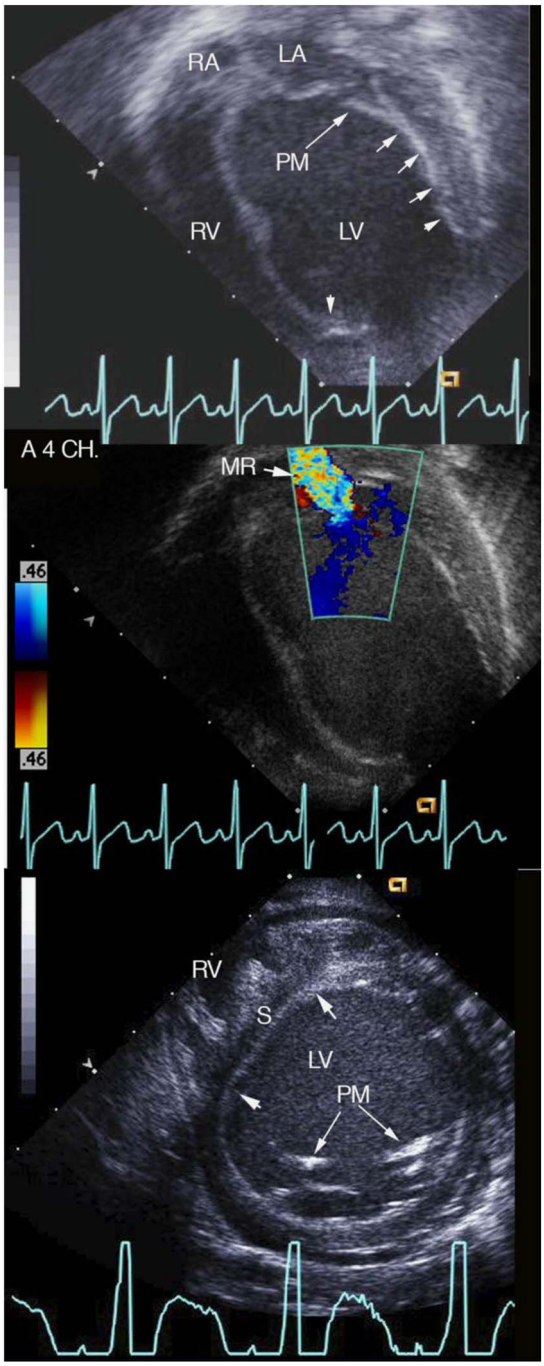

Figure 2.
This figure demonstrates a number of features commonly found in anomalous origin of the left coronary artery from the pulmonary artery. The upper and middle frames *taken from the same patient demonstrate an apical four-chamber view (A 4 CH). The left ventricle (LV) is dilated and the papillary muscles (PM) are bright due to fibrosis. The endocardium (small arrows) is thickened representing endocardial fibrosis. The* bottom frame *shows the cross-section of the LV from the parasternal short-axis view. The ventricle is dilated and circular except in the upper quadrant of the ventricular septum (S) between the arrows. In real time, this segment moved paradoxically outward during systole. LA = left atrium; MR = mitral regurgitation; RA = right atrium; RV = right ventricle.*

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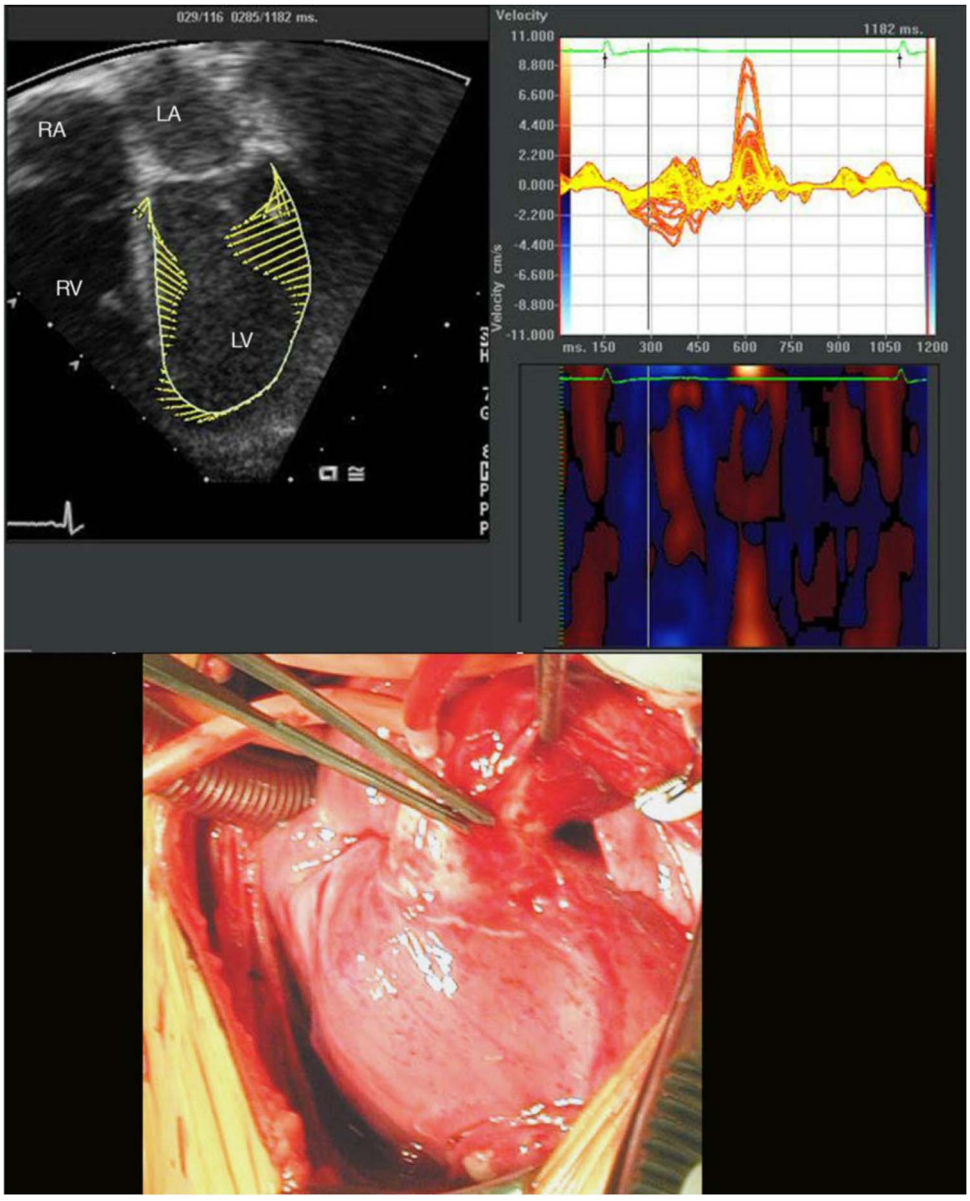

Figure 3.
The top panel on the left *is a tissue Doppler study in a patient with anomalous origin of the left coronary artery from the pulmonary artery, taken during systole using the technique of velocity vector imaging in the apical four-chamber view. The arrows indicate the direction of contraction and its magnitude. This systolic frame shows the best inward contraction of the posterolateral part of the left ventricle (LV) followed by its septal counterpart. The left ventricular apex shows no contraction and, indeed, the septal part of the apex bulges outward. The* right-hand panels *show the individual vectors over the cardiac cycle. There is a very poor peak velocity of excursion at −4 cm/second. The colour map at the bottom part of this panel shows a distorted pattern. In the normal heart, these bars of colour alternate red and blue bands in systole and diastole, but in this panel there is marked fragmentation of the colour, corresponding to that seen above in the upper right panel. The* bottom image *of the heart is that of an infant during repair before commencement of cardiopulmonary bypass. The paler area of the surface of the LV corresponds to the area of infarction in this patient. LA = left atrium; RA = right atrium; RV = right ventricle.*

echocardiographers must use all the clinical tools at their disposal to make the diagnosis, including taking an accurate history as well as electrocardiography. As a rule of thumb, I assume that every infant under 2 years of age referred with a diagnosis of dilated cardiomyopathy should have the diagnosis of ALCAPA excluded as a possible cause of the symptoms. The echocardiographer should have access to a good history as well as a review of ancillary tests. The patient's history often contains episodes of gastrointestinal or pulmonary disease, and clinical examination will show mitral regurgitant murmur and a galloping rhythm. The electrocardiogram is of value and typical for this condition

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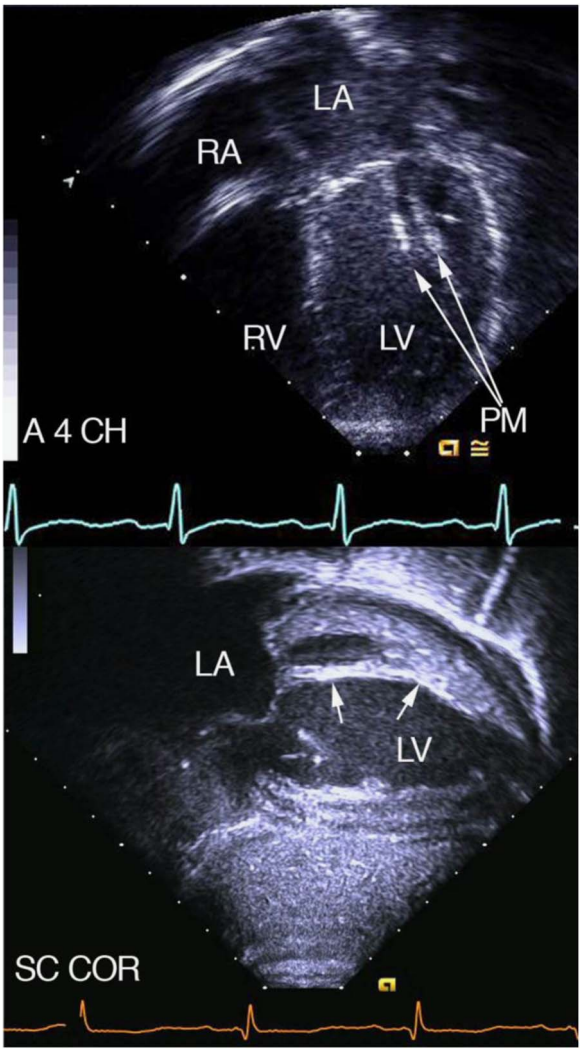

Figure 4.
These figures demonstrate attenuation of the bulk of the papillary muscles (PM) as well as the increase in fibrosis related to the ischaemia. The top frame *is an apical four-chamber view (A 4 CH). The bright PM are well seen, with attenuated thickness (bulk) of the PM mass. The* bottom frame*, taken in the subcostal coronal cut (SC COR), shows the thinned and attenuated PM with brightness extending into the endocardium of the left ventricle (LV) that represents fibrosis. LA = left atrium; RA = right atrium; RV = right ventricle.*

but may overlap with the findings of dilated cardiomyopathy.[10] Chest X-ray shows cardiomegaly and may show pulmonary oedema.

There is no finer description of the symptoms of ALCAPA in the infant than that of Bland, White, and Garland, for whom this disease carries the eponymous name.[11]

> Nothing remarkable was noted about the patient until the tenth week; while nursing from a bottle the onset of an unusual group of symptoms occurred which consisted of paroxysmal attacks of acute discomfort precipitated by the exertion of nursing. The infant appeared to be at first in obvious distress, as indicated by short expiratory grunts, followed immediately by marked pallor and short grunts followed by marked pallor and cold sweats with the general appearance of severe shock. Occasionally with unusually severe attacks there appeared to be a transient loss of consciousness. The eructation of gas at times seemed to relieve the discomfort and shorten the duration of the attack which usually lasted from 5 to 10 minutes, and following which the infant might proceed to nurse without difficulty and remain free of symptoms for several days. … It seems probable that in this infant the curious attacks of paroxysmal discomfort were … those of angina pectoris.

It should be recognised that the diagnosis of an anomalous coronary artery is determined by the nature of its physiology. Essentially, the physiology is that of a coronary-to-pulmonary arterial fistula. There is usually extensive communication between the collateral flows of the coronary arteries. Although pressure in the pulmonary arterial system is elevated before the pulmonary resistance diminishes, the right coronary flow perfuses the myocardium through the collaterals, and it is only when the resistance declines to its normal nadir during the 6th–10th week of life that the coronary flow is "stolen" away from the myocardium.

Infants experience symptoms similar to those experienced by adults with angina pectoris and myocardial infarction, although they cannot describe these symptoms as crushing chest pain. In infants, periods of ischaemia are temporally related to an increase in activity, leading to a concomitant increase in myocardial oxygen demand. These periods include eating, crying, and evacuating, as described originally by Bland, White, and Garland. With feeding, an increased demand for gastrointestinal blood flow steals the flow away from the myocardium as well. Infants who can only express these ischaemic symptoms through crying, yelling, or refusal to feed are often misinterpreted as being "just irritable or colicky". By the time the symptoms lead to a cardiac ultrasound consultation, damage of a greater or lesser degree would have already occured.

Owing to the high risk of sudden cardiac death and myocardial damage, aggressive surgical management and close follow-up are vitally necessary.

Echocardiography

All modalities of ultrasound, including two-dimensional imaging – particularly Doppler colour

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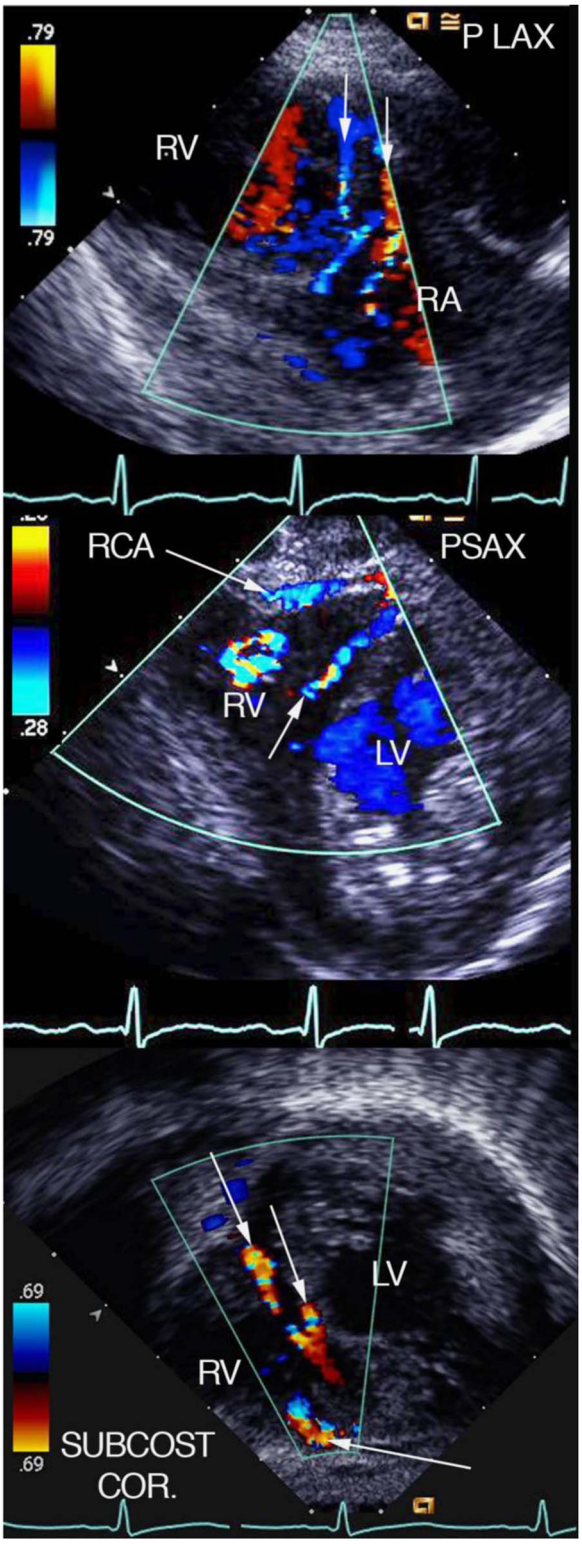
flow – and other Doppler and speckle tracking techniques, are part of a preliminary approach to specific imaging of the coronary artery anomaly itself. Historically, when only cross-sectional imaging was available, the diagnosis was often missed or misinterpreted as being a normal connection between the coronary artery and the aorta.[12] This occurred because the lateral drop-out of the aortic and arterial wall was superimposed by the transverse pericardial sinus, leading to apparent connection between the aorta and the coronary artery.[13] Diagnosis has been substantially improved by the use of modern ultrasonic machines that allow for Doppler colour flow imaging.[13–15] Once ultrasound has been completed, the diagnosis can be clarified by looking at the general features associated with cardiac ischaemia.



General echocardiographic features

Echocardiographic diagnosis must include attention to a number of details obtained from the general ultrasound examination. These features, individually and as a cluster, should lead the echocardiographer to suspect the diagnosis, examine the coronary anatomy very carefully, and look in greater detail at the specific findings.

The echocardiographic approach to appropriate diagnosis begins with taking a history and carrying out a physical examination using all modalities of cardiac ultrasound. The classical history described by Bland, White, and Garland should alert the examiner to the possibility of diagnosing anomalous origin. Unfortunately, many infants with a history of commonly occurring colic have similar symptoms, and patients with true pulmonary problems and cardiac ischaemia may be misinterpreted as having colic.

Associated findings

Decrease in cardiac function with specific attention to regional wall abnormalities. Infants usually present with dilated poorly contracting left ventricles

The left ventricle appears dilated and contracts poorly (Fig 1). In addition, there is usually some degree of

Figure 5.
This series of images demonstrates the increased collateral flow seen with anomalous origin of the left coronary artery from the pulmonary artery. The Doppler colour flow signals through the collateral vessels running within the myocardium itself or on the surface of the heart are shown by arrows. Note the Nyquist scale is as low as 28 cm/second and not higher than 79 cm/second. The top frame *is taken in the parasternal long-axis view (P LAX) with medial rotation towards the right, and shows the right atrium entering the right ventricle through the tricuspid valve. The Doppler signal (arrows) shows two collateral vessels running through the septum. The* middle frame *taken through the parasternal short-axis cut (PSAX) shows a right coronary branch vessel on the surface of the X subcostal coronal cut (SUBCOST COR.); a similar continuity between a right coronary artery (RCA) and septal coronary perforators is shown. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.*

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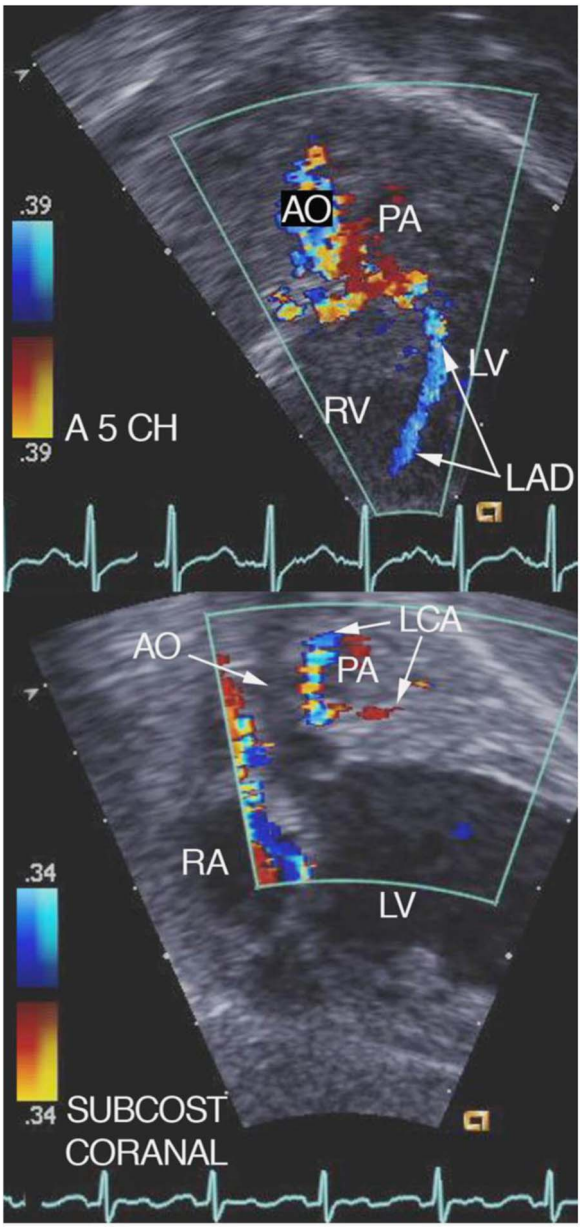

Figure 6.
Top frame*: this anteriorly directed apical five-chamber view (A 5 CH) shows Doppler colour flow signal in the left anterior descending coronary artery (LAD) in the surface of the myocardium paradoxically flowing away from the transducer in a retrograde direction from the apex and heading towards the pulmonary artery (PA). The Doppler signal in the aorta (AO) is also noted.* Bottom frame*: this is a subcostal coronal (SUBCOST CORANAL) frame using Doppler colour flow to show the left main coronary artery (LCA) flowing retrograde up into the PA on its medial side adjacent to the AO. LV = left ventricle; RA = right atrium; RV = right ventricle.*

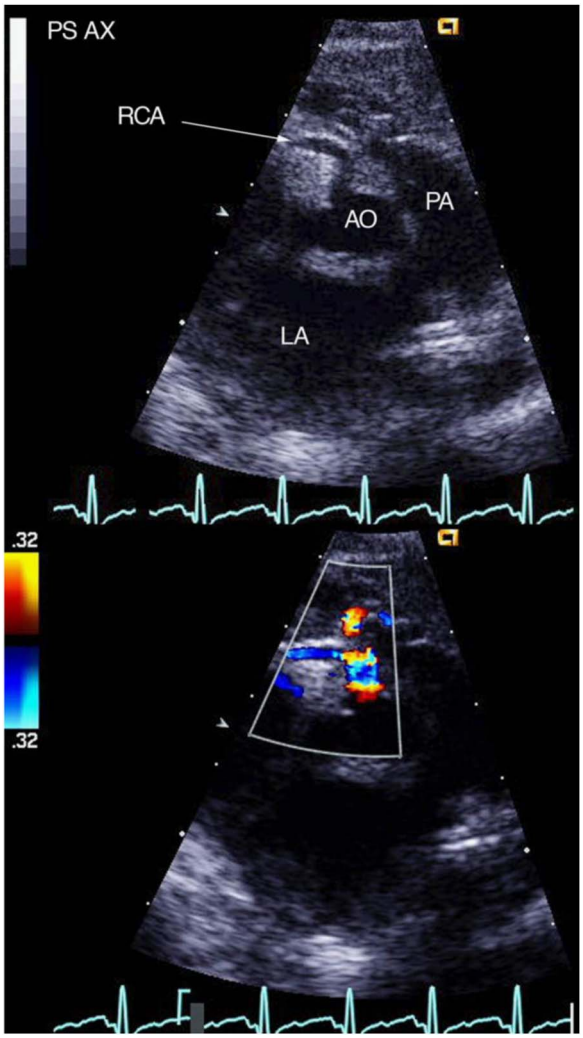

Figure 7.
This parasternal short-axis (PS AX) image is highly indicative of an anomalous origin of the left coronary artery from the pulmonary artery. The top frame *is a cross-sectional image showing a large right coronary artery (RCA) arising from the aorta (AO). The pulmonary artery (PA) lies adjacent to the AO and the left atrium (LA) lies behind the AO. The* bottom frame *is from the same patient with Doppler colour flow highlighting the flow arising from the proximal exit from the AO and entering the proximal RCA.*

mitral regurgitation. The papillary muscles appear very bright due to fibrosis, which can also be demonstrated in the parasternal or apical views (Fig 2). The dyskinesis is largely in the distribution of the left coronary artery, particularly the ventricular septal area and anterior left ventricle. The size of this infarct can be appreciated when the infant is placed on cardiopulmonary bypass (Fig 3). New technology using speckle tracking allows the dyskinesia of the ventricle to become obvious (Fig 3). Such dyskinesis is indicative more of ALCAPA than of a cardiomyopathy.

Papillary muscle fibrosis and endocardial fibrosis

Papillary muscle fibrosis occurs as a consequence of chronic ischaemia. This causes the mitral valve to sit

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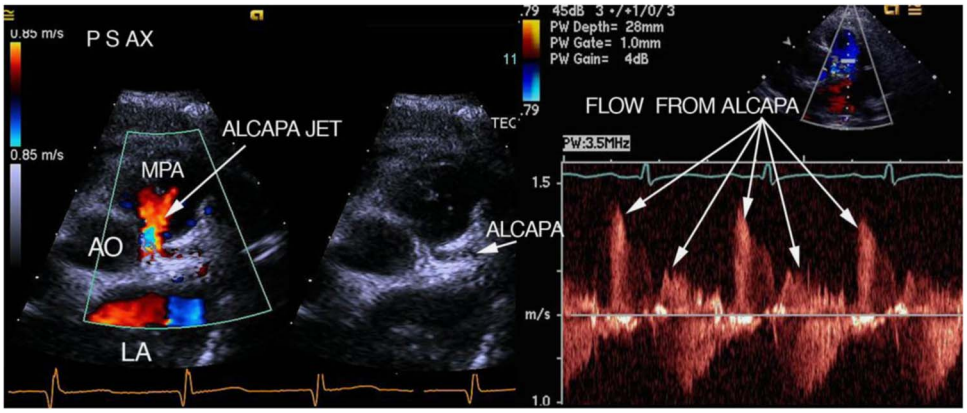

Figure 8.
This series of images displays the flow into the pulmonary artery from the anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). The left-hand image *of this simultaneously recorded image shows the colour flow image modality with the flame-shaped jet entering the pulmonary artery (ALCAPA JET). The aorta (AO), the pulmonary artery, and the left atrium (LA) are demonstrated. The accompanying* middle-panel image *taken in cross-section shows the ALCAPA entering the pulmonary artery. In the* right-hand panel*, the pulsed wave Doppler has been placed in the jet and has captured the higher velocity flow in diastole and the lower flow in systole. MPA = main pulmonary artery; P S AX = parasternal short-axis.*

poorly and causes mitral regurgitation. The intense brightness of the papillary muscle as well as the endocardial area representing endocardial fibrosis is seen due to the same reason and can be demonstrated in many views (Figs 2 and 4). In addition, the papillary muscles are fibrosed, as demonstrated by their dense reflective nature, and are diminished in bulk. This is often accompanied by endocardial fibrosis (Figs 1 and 4).

Mitral regurgitation

Mitral regurgitation is almost invariably present, and its grade can vary from mild to severe (Figs 1 and 2). It is best imaged from views axial to the mitral leaflet plane, such as the apical or some subcostal planes, but can also be observed from the long-axis parasternal view. It is not an observation specific to anomalous coronary artery, being often present in cardiomyopathy as well. Although there are cases of cardiomyopathy that do not exhibit mitral regurgitation, it is almost the rule in patients with ALCAPA.

Increase of coronary flow pattern within the myocardium

As substantial collateral flow is well noted on angiography, it is possible, using the Doppler colour flow modality, to see this flow within the coronary system from a variety of views. In order to observe the low-velocity coronary flow within the myocardium, it is necessary to lower the colour Doppler (Nyquist) scale, because in this condition low velocity flow can be missed if the Nyquist scale is set too high. This is true not only for myocardial flow but also for the low velocity flow with the jet that is seen from the coronary circulation entering the pulmonary artery. This caveat is important, as many of the most modern ultrasound systems seem to default to the highest possible scale setting. Doppler colour signals within the coronary system can be traced from the right coronary artery into the myocardium, across the myocardium, and then retrograde into the left coronary system (Figs 5 and 6). These signals can also be traced back to the proximal left coronary artery with the reversed flow direction that one expects in the left coronary system.

Disparity in size of the coronary arteries: right larger than left

In infants, the left and right coronary arteries are best imaged in the parasternal short-axis view at the base (Fig 7). The right coronary artery – as in any coronary fistula that takes its origin from the aorta – has increased flow and is prominent when viewed by imaging or by the Doppler colour flow modality. This is all the more remarkable, as there is no visible left coronary artery arising from the aorta in its usual position, thus bringing the diagnosis of ALCAPA into strong contention.

Doppler colour signals in the coronary artery

These signals are best obtained by placing the pulsed wave cursor from Doppler colour flow images in the parasternal short-axis plane, directing into the jet that arises from the pulmonary trunk flow disturbance, but signals may be obtained from other

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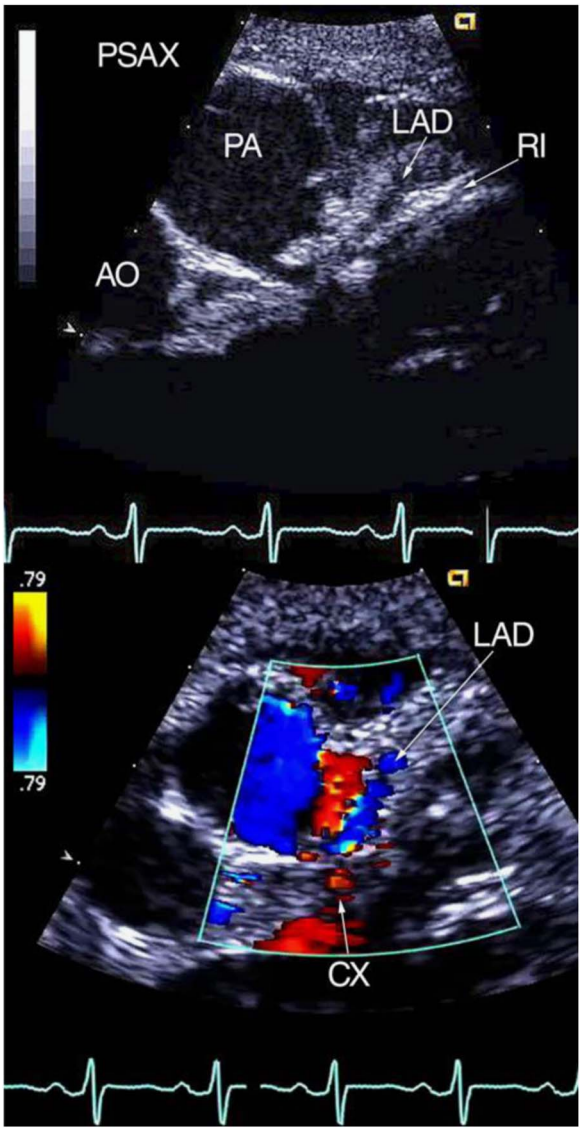

Figure 9.
In these two panels, taken temporally close together, the anomalous entry of the left coronary artery into the pulmonary artery (PA) is seen. In the top frame*, both the left anterior descending coronary artery (LAD) and the ramus intermedius (RI) join as the left main coronary artery, which enters the PA. The circular aorta (AO) is also noted. The* bottom frame *shows the Doppler flow mapping of the paradoxical flow in the LAD as well as flow from the left circumflex artery (CX), directing itself towards the PA. PSAX = parasternal short-axis.*

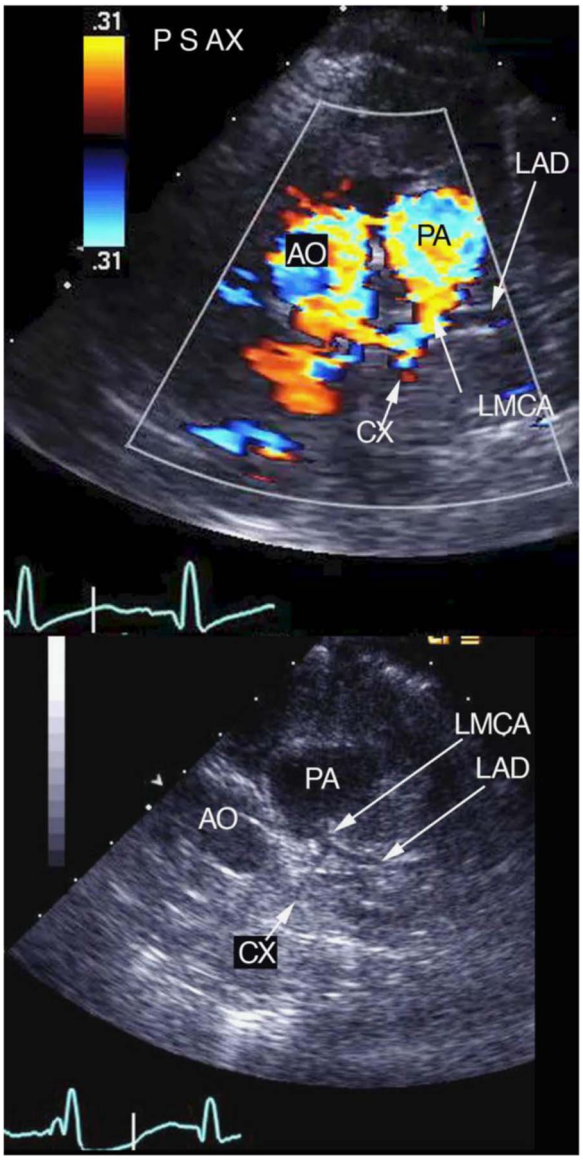

Figure 10.
The coronary connection into the pulmonary artery (PA) is identified through the process of first using Doppler colour flow imaging in the top frame *in the parasternal short-axis images (P S AX). The aorta (AO) and PA are identified. The circumflex coronary artery (CX) and left main coronary artery (LMCA) are identified. The left anterior descending coronary artery (LAD) does not show flow because the artery is running parallel to the flow vector, but can still be identified in this image. In the* bottom frame*, the colour superimposition has been removed and a clear image of the LMCA entering the PA is identified, with the LMCA demonstrated to connect with the anterior descending and CX.*

axial views such as the subcostal views (Fig 8), usually leading to direct imaging of the coronary artery from the pulmonary artery.

Direct imaging of the anomalous left coronary artery

In infants, the parasternal planes are quite easy to achieve. Once the Doppler colour flow images have been obtained, the examiner can make fine adjustments in the transducer position to image the arterial connection to the pulmonary artery.

It is often possible to image the left anterior descending coronary artery filling retrograde and

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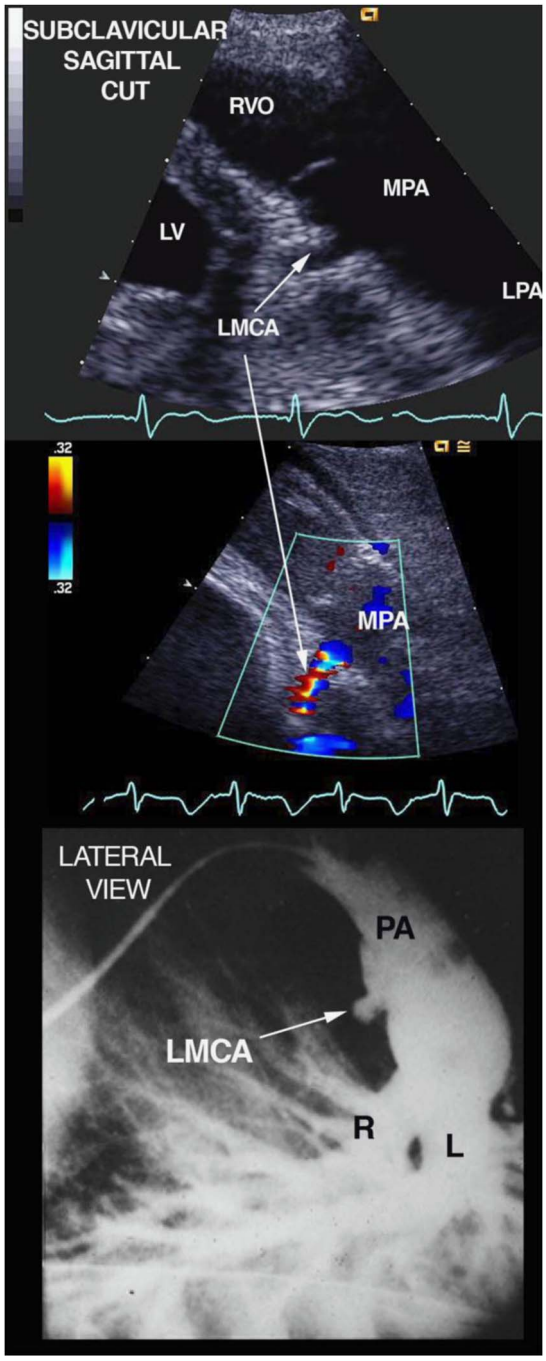

Figure 11.
This series of echocardiograms is taken at 90° to the previous figure. The view is termed the subclavicular sagittal view. The top and middle frames *are taken from two different patients but oriented similarly. The right ventricular outflow tract (RVO) can be seen to be continuous with the main pulmonary artery (MPA) and the left pulmonary artery (LPA). The circular left ventricle (LV) is seen below the right heart structures. The left main coronary artery (LMCA) is seen to drain directly into the MPA. For comparison, in the* bottom panel, *the pulmonary arterial angiogram, rotated to correspond to the echocardiographic plane, is shown in a patient with the same condition. In this frame, there is little reflux into the left main coronary corresponding to its position and alignment with the echocardiograms. L = left pulmonary arteries; R = right pulmonary arteries.*

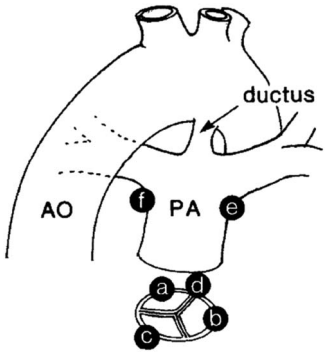

Figure 12.
*Possible sites of origin of the left coronary artery from the pulmonary artery (PA). The frequency of sites of origin and positions are noted: (**a**) right sinus of Valsalva in two, (**b**) left sinus of Valsalva in three, (**c**) anterior in one (malrotation of left anterior descending coronary artery), (**d**) posterior in two, (**e**) left main PA high in one, and (**f**) right main PA at the bifurcation of the right PA in two. AO = aorta (Published from[15] with permission of the Annals of Thoracic Surgery).*

continuing into the main left coronary artery with the ramus intermedius and/or the circumflex coronary artery connection also visible (Fig 9).

Doppler flow imaging, in addition to defining the origin of the vessel, helps define the branches as well. The Doppler colour flow signal can be subtracted from the image uncovering the direct image (Fig 10).

Merely by rotating the transducer 90° into an almost sagittal plane similar to the subclavicular ductus cut, one can image the left main coronary artery; here, the left main coronary artery is seen to connect to the main pulmonary artery directly rather than running below it (Fig 11). In this view, the area of display has to include the surface of the left ventricle and the long axis of the pulmonary artery. In the normal situation, the left coronary artery is circular and not attached to the pulmonary artery, but here the artery can be seen to make contact with either the right or the left posteriorly positioned sinus of Valsalva.

Although for the most part the artery originates from the facing sinus of Valsalva of the pulmonary artery, this is not always so, and one has to guard against presuming that the coronary artery always arises adjacent to the facing sinus of the aorta. The data from Turley et al show that the anomalous artery may arise from different sites within the pulmonary system (Fig 12).[16–19] When the anomalous left coronary artery does not arise in its usual position, it is important to use echocardiography to locate its origin. Particularly important is the use of the Doppler colour flow technique, always remembering to keep the Nyquist limit set low

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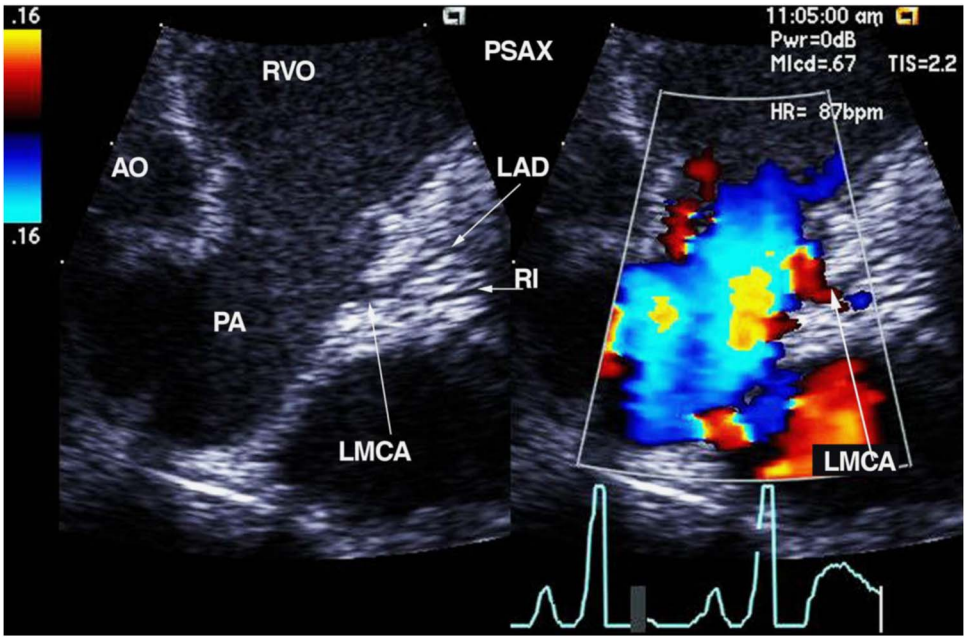

Figure 13.
The figure demonstrates an anomalous origin of the left coronary artery from the pulmonary artery with lateral entry into the pulmonary artery (PA). The entrance is some distance away from the aorta (AO) and will require special surgical attention. The left panel *is a cross-sectional image showing the left arterial origin of the coronary artery. The* right panel *is the Doppler flow information obtained simultaneously. Note the artery is filled with flow signal but there is a blush of colour in the PA, representing the coronary flow. Note the Nyquist limit is set at 16 cm/second. LAD = left anterior descending coronary artery; LMCA = left main coronary artery; PSAX = parasternal short-axis; RI = ramus intermedius; RVO = right ventricular outflow tract.*

because the flow velocity at the entrance into the pulmonary artery is low and easily obfuscated with higher velocity settings. We have encountered many different origins of this vessel (Fig 13).

Later presentation of ALCAPA can occur even during adult life, with presentation only from the time of atherosclerotic association. Exactly why this situation occurs is not clear; perhaps, in some hearts, the right coronary system can maintain excessive flow, and perhaps there is mild stenosis at the entrance of the left coronary artery to prevent the "coronary steal" from occurring.

Associated diseases may precipitate presentation that otherwise would be masked. We have observed this after device closure of a patent ductus arteriosus and with Alagille's syndrome. The mildly elevated central pulmonary pressure from the ductus in the former and the peripheral pulmonary stenosis elevating central pulmonary arterial pressure in the latter prevented the perfusion "steal" from occurring. There are also reports in the adult literature of anomalous coronary artery presenting in adulthood from associated atherosclerotic coronary disease.[20]

Echocardiography remains the primary test for diagnosing ALCAPA. Once it is clear that the patient has presented with ALCAPA, surgery may be undertaken without further imaging or invasive procedures. Many have questioned whether echocardiography alone, without the need for ancillary tests, is sufficient for the diagnosis to be made, and some have insisted on angiography or alternative imaging modalities to substantiate the diagnosis (Fig 14). Although there is no question that cardiac catheterisation and angiography provide exquisite detail of the morphology and physiology of the anomaly, they are invasive procedures and add a substantial amount of time or contrast agent to an already compromised myocardium. A very good estimate of the physiology of this defect can be made using MRI or computerised axial tomography, which both provide even greater spatial detail while being non-invasive (Fig 15).

For sick neonates or infants, CT is perhaps a better option. There is also no doubt that the coronary tree can be demonstrated using a variety of three-dimensional reconstructions. Certainly, if there is doubt about the echocardiographic diagnosis, these options remain available without compromising the patient by assuming a cardiomyopathy or by inappropriate referral to the operating room for repair.

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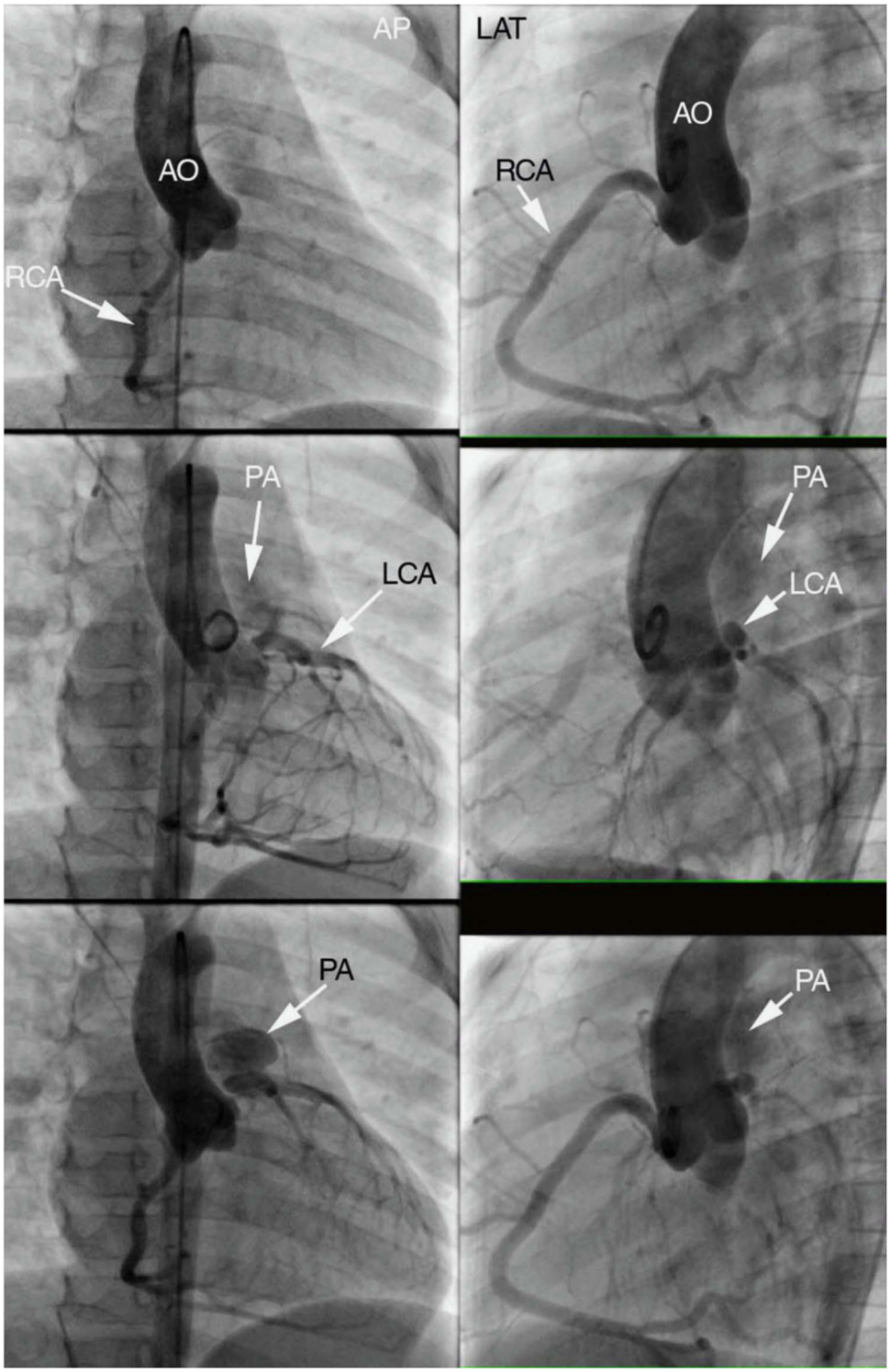

Figure 14.
This series of images was taken from a biplane angiogram of a patent with Alagille's syndrome who also had an anomalous coronary artery. On the left are representative still frames from the posteroanterior (AP) angiogram, and the lateral angiogram (LAT) is on the right. In the top two panels*, the right coronary artery (RCA) fills. No left coronary artery (LCA) is identified. In the* middle two panels*, the contrast dye has passed into the peripheral RCA and then via a myriad of collateral vessels it now fills the left coronary system. A blush of contrast is seen entering the pulmonary artery (PA) from the left coronary system. In the* bottom two panels*, although there is still contrast material in the proximal RCA, the contrast agent in the left system has diminished and the material is now concentrated in the main PA via the left main coronary artery. AO = aorta.*

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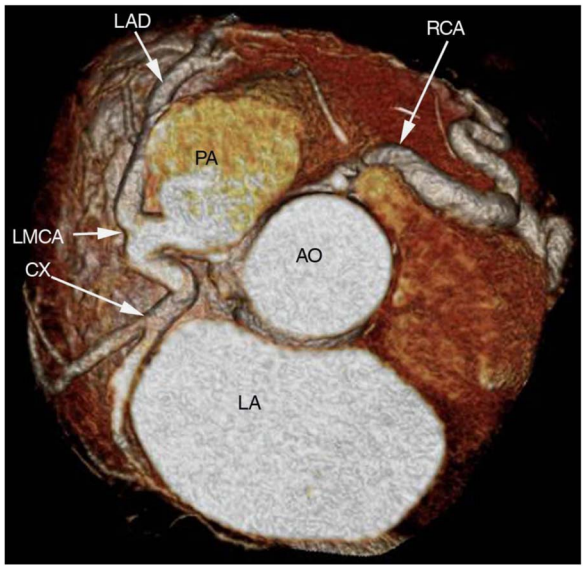

Figure 15.
This is a CT angiogram from an elderly adult with anomalous origin of the left coronary artery from the pulmonary artery. The cross-section is viewed from above. Although the right coronary artery (RCA) arises normally from the aorta, the left main coronary artery (LMCA) and its branches, the left anterior descending (LAD), and circumflex coronary (CX) arteries arise from the pulmonary artery (PA).

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Ethical Standards

The author assert that all procedures contributing to this study comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008.

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