RadioGraphics

ALCAPA Syndrome: Not Just a Pediatric Disease[1]

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TEACHING POINTS
See last page

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome is a rare congenital coronary artery anomaly. There are two types of ALCAPA syndrome: the infant type and the adult type, each of which has different manifestations and outcomes. Infants experience myocardial infarction and congestive heart failure, and approximately 90% die within the 1st year of life. Rarely, ALCAPA syndrome manifests in adults; it may be an important cause of sudden cardiac death. Historically, ALCAPA syndrome was diagnosed at conventional angiography. However, the development of electrogardiographically gated multidetector computed tomographic (CT) angiography and magnetic resonance (MR) imaging enables accurate noninvasive imaging. At MR imaging and multidetector CT angiography, findings include direct visualization of the left coronary artery arising from the main pulmonary artery. Reversed flow from the left coronary artery into the main pulmonary artery may be seen at steady-state free-precession cine and fast cine phase-contrast MR imaging. Because of its ability to assess myocardial viability, which can be used as a prognostic factor to direct the need for surgical repair, MR imaging plays an important role in patient treatment. Restoration of a dual-coronary-artery system is the ideal surgical treatment for ALCAPA syndrome.

Abbreviations: ALCAPA = anomalous origin of the left coronary artery from the pulmonary artery, CABG = coronary artery bypass graft, ECG = electrocardiographically, LAD = left anterior descending, LCA = left coronary artery, RCA = right coronary artery, SSFP = steady-state free precession

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Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome, also known as Bland-White-Garland syndrome, is a rare congenital abnormality that affects 1 of every 300,000 live births (1) and accounts for 0.25%–0.5% of all congenital heart defects (2). It usually manifests as an isolated defect, but in 5% of cases it may be associated with other cardiac anomalies such as atrial septal defect, ventricular septal defect, and aortic coarctation (3).

ALCAPA syndrome results in the "coronary steal" phenomenon, in which a left-to-right shunt leads to abnormal left ventricular perfusion. ALCAPA syndrome is one of the most common causes of myocardial ischemia and infarction in children. If left untreated, up to 90% of patients with ALCAPA syndrome die within the 1st year of life (4). In patients who live to adulthood, ALCAPA syndrome may cause myocardial infarction, left ventricular dysfunction and mitral regurgitation, or silent myocardial ischemia, which can lead to sudden cardiac death. Early diagnosis and prompt surgical intervention with the aim of restoring a two-coronary-artery circulatory system have excellent results and lead to gradual myocardial recovery.

In this article, we review the pathophysiology of the different types of ALCAPA syndrome, clinical manifestations of the disease, and imaging features at cardiac catheterization, magnetic resonance (MR) imaging, and electrocardiographically (ECG) gated multidetector computed tomographic (CT) angiography. We also describe different surgical approaches and the appearance of postoperative complications on CT images.

Pathophysiology

In fetal and early neonatal life, the origin of the left coronary artery (LCA) from the pulmonary artery is well tolerated because pulmonary arterial pressure equals systemic pressure, which leads to antegrade flow in both the anomalous LCA and the normal right coronary artery (RCA) (5). Soon after birth, when pulmonary arterial pressure decreases, flow in the LCA decreases and then reverses, which leads to myocardial ischemia and infarction. The extent of acquired collateral circulation between the RCA and LCA during the critical period, when pulmonary arterial pressure gradually decreases, determines the extent of myocardial ischemia (Fig 1). Patients with well-established collateral vessels have the adult type of the disease, and those without collateral vessels have the infant type. Both types of the disease have different manifestations and outcomes (Table 1).

Teaching Point

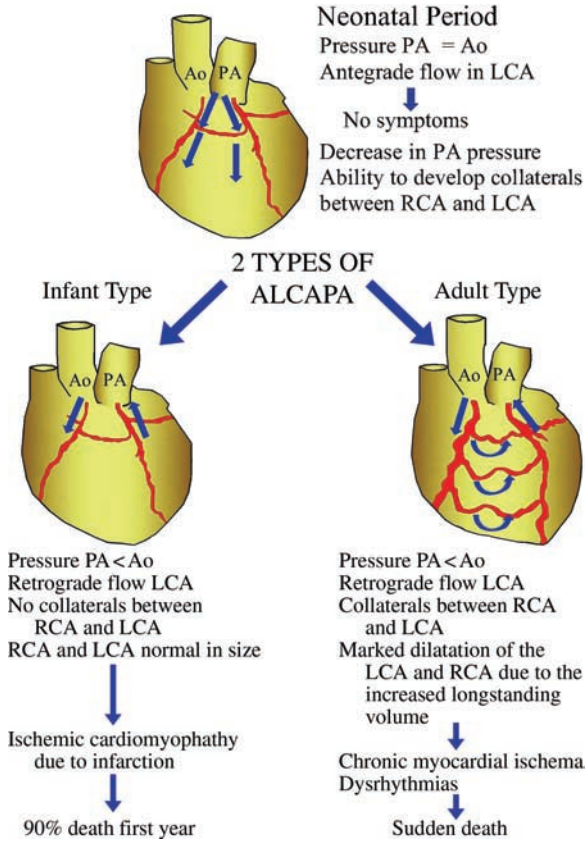


Figure 1. Diagram shows the spectrum of pathophysiologic changes that take place after birth in patients with ALCAPA syndrome. The differences between the infant and adult types of ALCAPA syndrome, the importance of developing intercoronary collateral vessels, and the different clinical manifestations are summarized.

Infant Type

The onset of symptoms usually occurs about 8 weeks after birth. There is little or no coronary collateral development. When the reversal of flow in the LCA is established, a limited blood supply to the left ventricular myocardium leads to congestive heart failure and mitral insufficiency secondary to myocardial infarction (4,6). The most important differential diagnosis in this age group is dilated cardiomyopathy (1).

Infants typically present with a failure to thrive, profuse sweating, dyspnea, pallor, and atypical chest pain while eating or crying. Without surgical repair, rapid death ensues in up to 90% of patients within weeks or months of birth (4).

Table 1
Differences between the Infant and Adult Types of ALCAPA Syndrome

Factor	Infant Type	Adult Type
Clinical manifestations	Symptomatic (infarction and heart failure)	Asymptomatic (subclinical ischemia and sudden death)
ECG findings	Ischemic changes	Left ventricular hypertrophy or ischemic changes
Right coronary artery	Mildly dilated	Markedly dilated
Septal collateral vessels	Insufficient	Abundant
Wall motion abnormalities	Anterior and lateral wall hypokinesis	None or global hypokinesis
Left ventricle	Dilated	Normal or mildly dilated, hypertrophy
Outcome	Myocardial infarction and death in the first year of life	Sudden death due to ischemic ventricular dysrhythmias

Table 2
Factors in Infants that Enable Survival to Adulthood

Factor	Description
Abundant interarterial collateral vessels between the RCA and the LCA	Retrograde left ventricular perfusion from the RCA
RCA dominance	Smaller myocardial area supplied by the LCA leads to less extensive myocardial ischemia
Minimal coronary steal from the pulmonary artery	Ostial stenosis of the LCA or a restrictive opening into the pulmonary artery limits the left-to-right shunt
Development of systemic blood supply to the LCA	Bronchial artery collateral vessels increase oxygenated blood flow and perfusion pressure to the ischemic myocardium

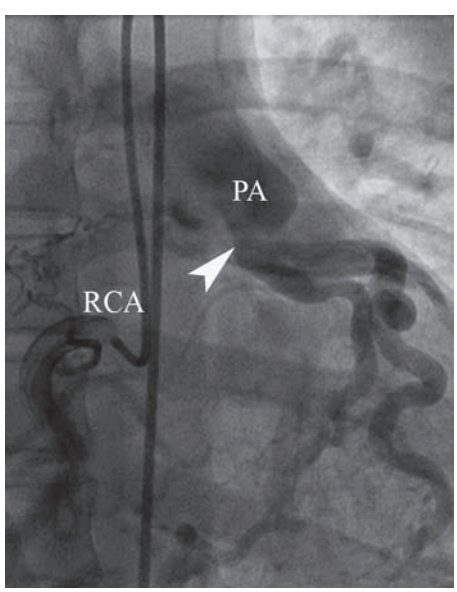


Figure 2. ALCAPA syndrome in a 35-year-old woman. Coronary angiogram obtained with a single RCA injection shows a tortuous and dilated RCA as well as an equally tortuous and dilated LCA. The LCA opacifies the main pulmonary artery *(PA)*. The connection of the LCA with the main pulmonary artery (arrowhead) and the resultant steal phenomenon are the main diagnostic features of ALCAPA syndrome.

Adult Type

Teaching Point

As pressure decreases in the pulmonary circulation and as flow reverses in the LCA, the LCA fails to supply the myocardium and "drains" fully oxygenated blood into the main pulmonary artery. Thus, there is preferential blood flow into the low-pressure pulmonary circulation rather than into the high-resistance myocardial circulation. This left-to-right shunt is known as the steal phenomenon. To survive beyond infancy, patients with ALCAPA syndrome develop significant collateral circulation from the RCA to the LCA (Table 2). However, often it is not sufficient to supply the left ventricle, especially in the subendocardial region; chronic left ventricular subendocardial ischemia ensues. As a result, patients may develop malignant ventricular dysrhythmias. These patients are at risk for sudden cardiac death, which occurs in 80%–90% of cases (7–11). Patients may be asymptomatic, or they may present with mitral insufficiency, ischemic cardiomyopathy, or malignant dysrhythmias, which lead to sudden death (4).

Imaging Findings

Conventional angiography can depict the course of the anomalous coronary artery (Fig 2).

Table 3
Comparison of Multidetector CT Angiography and MR Imaging in the Diagnosis of ALCAPA Syndrome

Modality	Spatial Resolution	Temporal Resolution	Additional Factors	When to Use
Multidetector CT angiography	Excellent	Low	Fast, widely available, uses ionizing radiation	Initial diagnostic work-up, postoperative follow-up
MR imaging	Low	Excellent	Cine phase-contrast imaging (to quantify the steal phenomenon), delayed enhancement (to assess left ventricular viability), SSFP cine imaging (to assess wall motion, the jet from the LCA into the main pulmonary artery, and valvular abnormalities)	Preoperatively to assess the extent of left ventricular myocardial infarction, wall motion abnormalities, mitral valve impairment, and the degree of left-to-right shunt

*SSFP = steady-state free precession.

Table 4
Features of ALCAPA Syndrome on Multidetector CT Angiographic and MR Images

Findings	Pathophysiologic Significance
Primary findings	
Direct visualization of the LCA arising from the main pulmonary artery	Diagnostic hallmark
Retrograde flow from the LCA into the main pulmonary artery	Steal phenomenon
Secondary findings (adult type)	
RCA dilated and tortuous	Chronic left-to-right shunt
Dilated intercoronary collateral vessels	Collateral pathways between the RCA and the LCA
Left ventricular hypertrophy and dilatation	Chronic myocardial ischemia
Mitral insufficiency and prolapse	Myxomatous degeneration and myocardial ischemia
Left ventricular wall motion abnormalities	Global hypokinesis
Dilated bronchial arteries	Systemic supply to the LCA territory and increased perfusion pressures
Delayed subendocardial enhancement	Subendocardial infarction

However, angiography is invasive and associated with a low but well-known risk of complications (12,13). ECG-gated multidetector CT angiography and MR imaging are valuable noninvasive modalities that can be used to help diagnose ALCAPA; radiologists should be aware of the advantages and disadvantages of each (Table 3).

Multidetector CT angiography is fast and widely available. ECG-gated multidetector CT angiography offers excellent spatial resolution, which is required to assess small vessels such as the coronary arteries. The main disadvantages of multidetector CT angiography are its relatively high radiation dose and its inability to assess flow.

MR imaging does not use ionizing radiation. SSFP cine imaging can demonstrate communication and flow from the LCA into the pulmonary artery, and it is useful in assessing mitral valvular function. In addition, fast cine phase-contrast imaging can help assess qualitative and quantitative

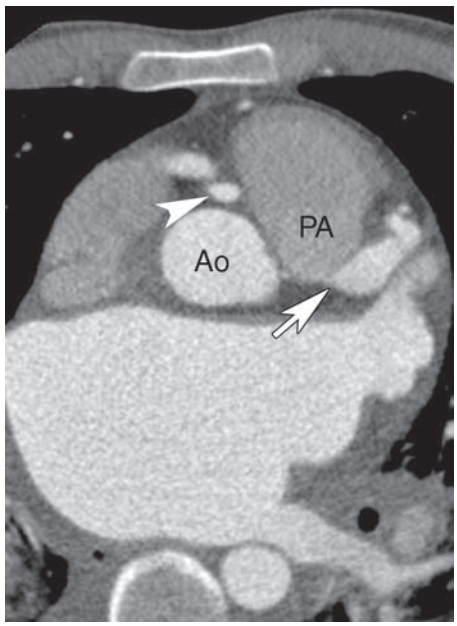


Figure 3. ALCAPA syndrome in a 30-year-old woman. Axial multidetector CT angiogram shows the origin of the LCA (arrow) from the main pulmonary artery *(PA)* and the dilated RCA (arrowhead). *Ao* = aorta.

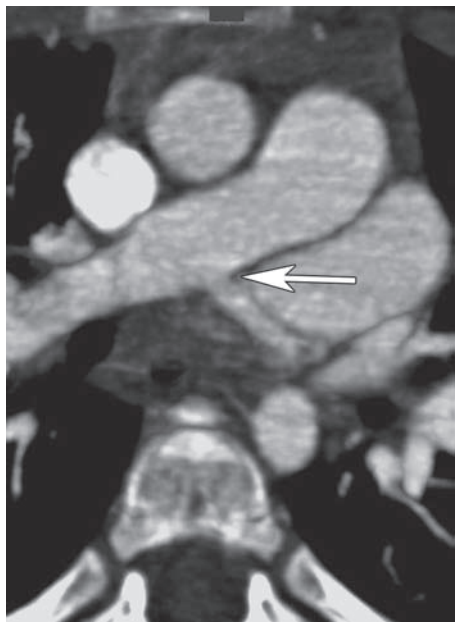


Figure 4. ALCAPA syndrome variant in a 3-year-old boy. CT angiogram shows the left circumflex artery (arrow) arising from the posterior wall of the right pulmonary artery and coursing above the left atrial appendage to reach the atrioventricular groove.

flow. Delayed gadolinium enhancement may help determine myocardial viability. The main disadvantages of MR imaging in comparison with CT are its relatively long examination times and its low spatial resolution.

ECG-gated multidetector CT angiography plays an important role as a first-line modality in assessment of ALCAPA syndrome, and it may be a valuable postoperative follow-up tool for adult patients. MR imaging is useful for assessment of left ventricular myocardial viability to determine if asymptomatic adult patients may benefit from surgical correction. Imaging findings of ALCAPA syndrome are classified as either primary or secondary (Table 4).

Primary Imaging Findings

Teaching Point

Direct visualization of the LCA arising from the main pulmonary artery is the diagnostic hallmark of ALCAPA syndrome (Fig 3). In ALCAPA syndrome, the LCA typically arises from the left inferolateral aspect of the main pulmonary artery just beyond the pulmonary valve. It then courses toward the interventricular groove and branches into the left anterior descending (LAD) and circumflex arteries. An isolated anomalous origin of the RCA, circumflex artery, and LAD artery from the pulmonary artery also has been described (Fig 4). However, in more than 90% of cases, the LCA originates anomalously from the pulmonary artery (10).

The retrograde flow from the LCA to the main pulmonary artery is well depicted on MR images, a characteristic finding of the steal phenomenon (5). The reversal of flow at the origin of the LCA into the main pulmonary artery appears as a jet entering the main pulmonary artery on SSFP cine images, a finding caused by turbulent flow that leads to spin dephasing in the main pulmonary artery at the origin of the anomalous LCA. The direction and volume of flow from the LCA to the pulmonary circulation may be accurately quantified on fast cine phase-contrast images.

Secondary Imaging Findings

The RCA and LCA appear markedly dilated and tortuous. In neonates, these vessels are normal

in size. But if the abnormality is detected only in adulthood, marked dilatation develops over time as blood is rapidly shunted from the RCA into the LCA and then into the main pulmonary artery.

Dilated intercoronary collateral arteries are seen along the epicardial surface of the heart or within the interventricular septum. They represent the collateral pathways between the RCA and the LCA (Fig 5).

Left ventricular hypertrophy and dilatation result from chronic myocardial ischemia (Fig 6) and may be seen at both SSFP cine MR imaging and ECG-gated multidetector CT angiography, both of which also may be used to assess left ventricular function.

Myxomatous degeneration and ischemic dysfunction of the papillary muscles and adjacent myocardium may cause mitral insufficiency and prolapse, findings best seen at SSFP cine MR imaging, although mitral valve prolapse may be seen at ECG-gated multidetector CT angiography. Left ventricular wall motion abnormalities, most commonly global hypokinesis, may be seen at SSFP cine MR imaging and ECG-gated multidetector CT angiography.

Dilated bronchial arteries, which act as systemic collateral vessels to the LCA, are best depicted at ECG-gated multidetector CT angiography because of their small size (Fig 7) (14–16).

Delayed subendocardial enhancement may be seen on MR images, a finding caused by subendocardial infarction that results from chronic subendocardial ischemia. This finding is important, especially in asymptomatic adults and those in whom ALCAPA is incidentally discovered, because it may be predictive of the onset of malignant dysrhythmias, which lead to sudden death. If subendocardial enhancement is seen, surgical repair should be strongly considered (Fig 8).

Surgical Correction

Teaching Point

Surgical correction performed on making a diagnosis of ALCAPA syndrome is considered to be the standard treatment. The aim of surgery is to restore a two-coronary-artery circulation system. In neonates, early correction is essential because in this age group repair is associated with improvement in ventricular function. If ALCAPA syndrome is left untreated, mortality in the 1st year of life approaches 90% (17).

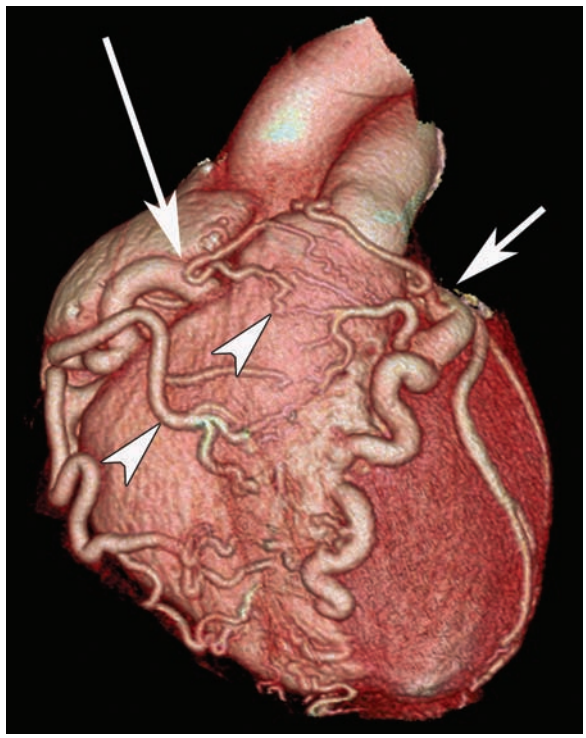
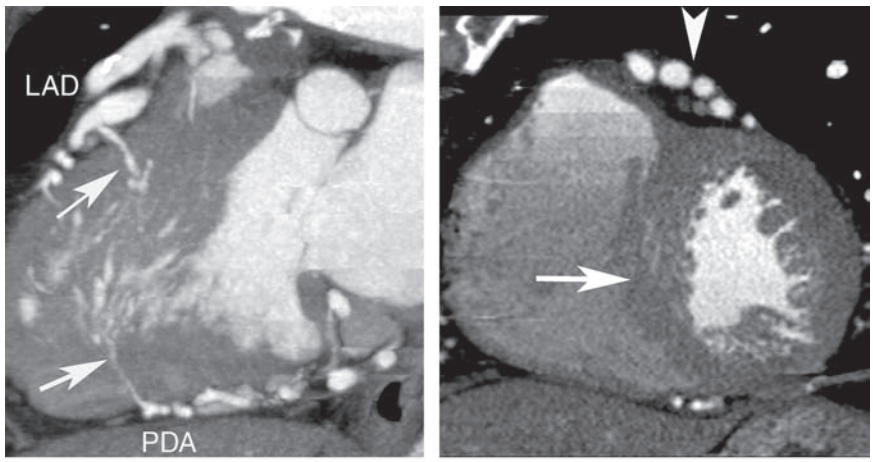


Figure 5. Three-dimensional volume-rendered multidetector CT angiogram shows dilated intercoronary collateral arteries (arrowheads), which connect the tortuous RCA (long arrow) to the LCA (short arrow).

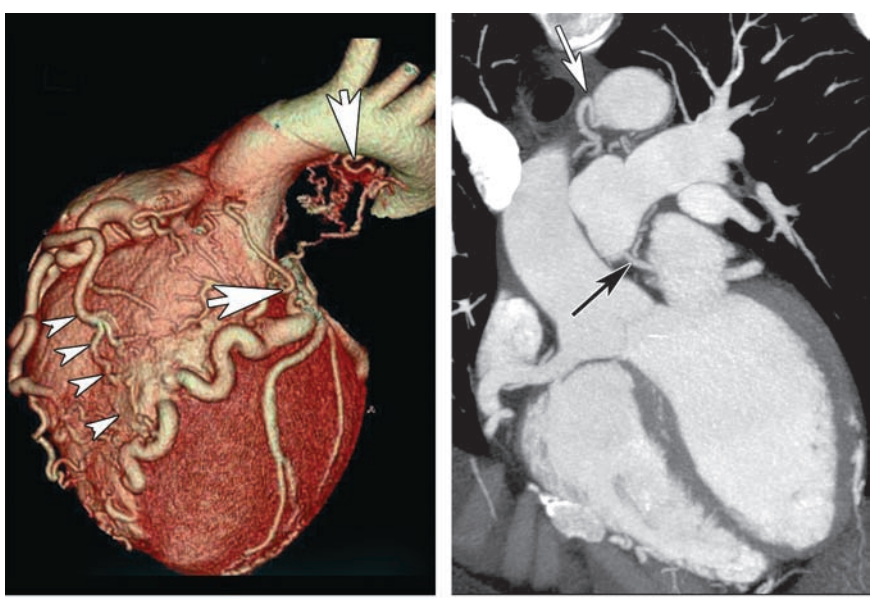
In asymptomatic adults, recent literature suggests that if only moderate chronic ischemia and limited necrosis are present, survival without surgical correction is possible. Because of its ability to provide information on myocardial function and viability, MR imaging is an excellent tool in the therapeutic decision algorithm in asymptomatic adults (18,19). When extensive delayed subendocardial enhancement caused by infarction is seen on MR images, surgery should be performed (1,19,20). Surgery enables correction of chronic subendocardial ischemia and recovery of ventricular function, and it reduces the risk for malignant dysrhythmias and sudden death. However, cases of uncorrected ALCAPA syndrome have been described in asymptomatic adults, even in older patients, when the risk-benefit ratio of surgery is less clear (21).

There are several surgical approaches, which are classified into two groups: one-coronary-system and two-coronary-system repairs (20). Single-coronary-system repair includes ligation of the anomalous LCA at its pulmonary origin, a procedure that is currently avoided due to the high rate of complications such as recanalization of the ALCAPA, a greater risk of atherosclerosis, severe mitral regurgitation resulting from ischemic



a. b.

Figure 6. ALCAPA syndrome in a 51-year-old woman. **(a)** Multiplanar reformatted CT image shows thickened septal myocardial wall and multiple septal collateral vessels (arrows) between the posterior *(PDA)* and anterior *(LAD)* descending arteries. **(b)** Oblique short-axis multiplanar reformatted CT image obtained at the base of the left ventricle shows left ventricular hypertrophy (arrow). Note the intercoronary collateral vessels along the epicardial surface (arrowhead).



a. b.

Figure 7. ALCAPA syndrome in a 35-year-old woman. Volume-rendered **(a)** and maximum intensity projection **(b)** CT images show multiple dilated bronchial arteries arising from the descending aorta and anastomosing to the anomalous LCA (arrows). Note the multiple collateral vessels between both coronary arteries (arrowheads in **a**), which extend across the surface of the pulmonary trunk and right ventricle.

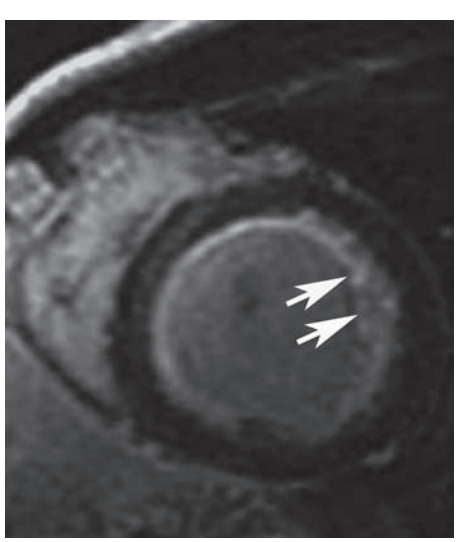


Figure 8. ALCAPA syndrome in a 38-year-old man. Oblique short-axis MR image acquired 10 minutes after gadolinium-based contrast material was administered shows subendocardial enhancement (arrows) at the base of the left ventricle, a finding consistent with an infarct. The patient underwent surgical repair.

Table 5
Types of Surgical Repair for ALCAPA Syndrome

Type of Repair	Description	Indications
One-coronary system	Ligation of the anomalous LCA at the pulmonary origin	No longer used
Two-coronary system		
Coronary button transfer	Direct reimplantation of the anomalous artery into the aorta by transferring it with a button of the main pulmonary artery	Used in infants; the most anatomic correction and yields excellent long-term results
Takeuchi procedure (transpulmonary baffling)	A baffle made from the pulmonary artery wall is used to tunnel the coronary artery through the main pulmonary artery into the left coronary ostium	Used in infants when coronary button transfer is not feasible due to unfavorable coronary anatomy
Placement of a CABG combined with ligation of the anomalous LCA	A venous or arterial bypass graft is placed from the aorta to the proximal LAD artery, and the anomalous LCA is ligated at its pulmonary origin	Preferred technique in adults

Table 6
Complications of Surgical Correction

Surgical Correction	Complications
One-coronary system	
Ligation of the ALCAPA	Recanalization of the ALCAPA, atherosclerosis, severe mitral regurgitation, and persistent silent ischemia (causes sudden death)
Two-coronary system	
Coronary button transfer	In adults, coronary friability with potential for tearing, bleeding, and kinking of the LCA
Takeuchi procedure (transpulmonary baffle)	Supravalvular pulmonary stenosis, aortic valve insufficiency, baffle obstruction, and leaks
CABG placement with ligation of the ALCAPA	Saphenous vein and arterial graft stenosis and occlusion with a high percentage of re-do procedures, especially in vein grafts

cardiomyopathy (22,23), and a persistent risk of sudden death due to silent ischemia.

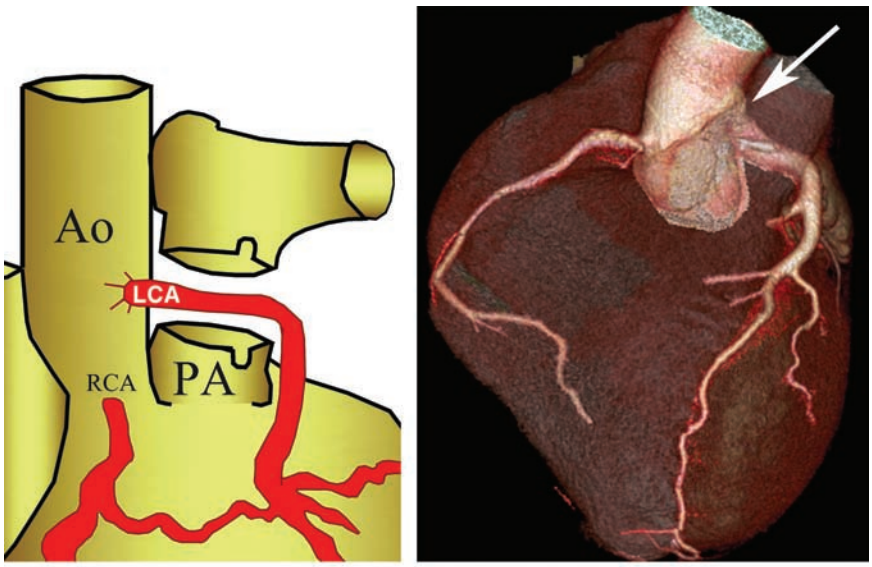
Two-coronary-system repairs are preferred and include coronary button transfer, (Figs 9, 10) (24), the Takeuchi procedure (Figs 11, 12) (25), and placement of a coronary artery bypass graft (CABG) combined with ligation of the origin of the LCA (Fig 13) (Table 5). Of these options, coronary button transfer is considered to be the most anatomic correction (22,23,26,27), and it has excellent long-term results. It is the preferred method of treatment in infants (20). In adults, the preferred method is ligation of the LCA at its origin from the main pulmonary artery to stop competitive flow combined with CABG placement by using the internal mammary artery or a saphenous vein (7,8). Other surgical repairs include cardiac transplantation in cases of significant left ventricular dysfunction (28) and percutaneous transcatheter closure of the ALCAPA (29).

Teaching Point

Postoperative Complications

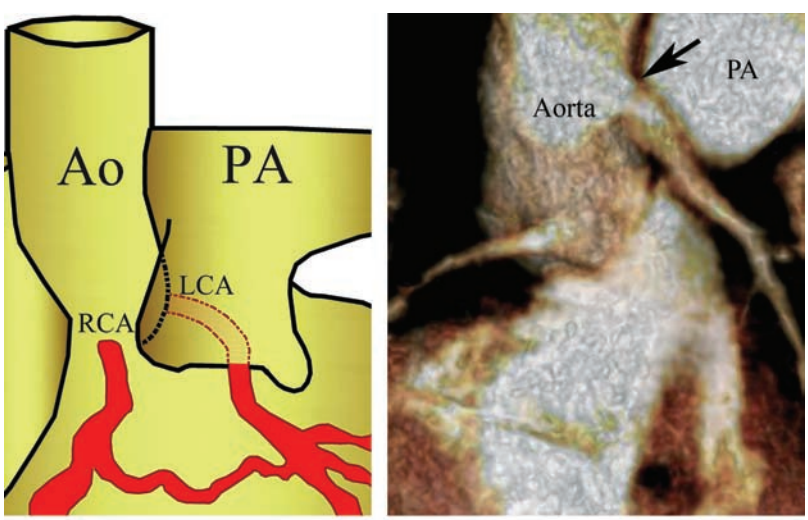
Although early diagnosis and prompt surgical intervention lead to excellent results, the possibility of postoperative complications necessitates long-term follow-up imaging. Because of its excellent spatial resolution, ECG-gated multidetector CT angiography is a valuable follow-up tool in adults after surgery.

Each type of surgical repair is associated with specific complications (Table 6). CABG placement combined with ligation of the anomalous



9. 10.

Figures 9, 10. **(9)** Diagram shows coronary button transfer. In this procedure, the LCA is reimplanted into the aorta *(Ao)* with a button from the pulmonary artery *(PA)* wall. Coronary button transfer is the most commonly used procedure in newborns and is the most anatomic correction. **(10)** Corrected ALCAPA syndrome in a 16-year-old boy. As a neonate, the patient underwent coronary button transfer with end-to-side anastomosis of the LCA to the aorta. Volume-rendered CT image shows the reimplanted LCA into the aorta (arrow). The normal size of the RCA is due to this surgical correction.



11. 12.

Figures 11, 12. **(11)** Diagram shows the Takeuchi procedure. In this procedure, a transpulmonary baffle between the coronary ostium in the pulmonary artery *(PA)* and the aorta *(Ao)* is created. Because the pulmonary artery wall is used to create the baffle, supravalvular pulmonary stenosis is a common complication. **(12)** Corrected ALCAPA syndrome in a 12-year-old girl who underwent the Takeuchi procedure. Multidetector CT angiography was performed to assess the postoperative patency of the LCA. Volume-rendered CT image shows the LCA reimplanted above the sinotubular junction (arrow) and the patent transpulmonary baffle. *PA* = pulmonary artery.

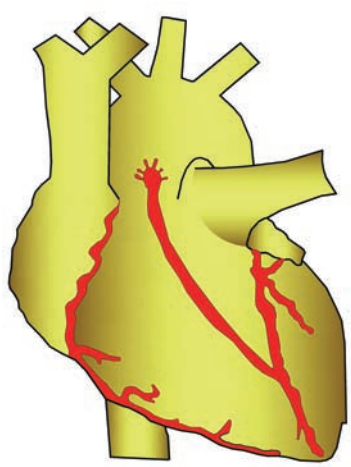


Figure 13. Diagram shows placement of a CABG with ligation of the anomalous LCA. This procedure is the preferred method of surgical correction for ALCAPA syndrome in adults.

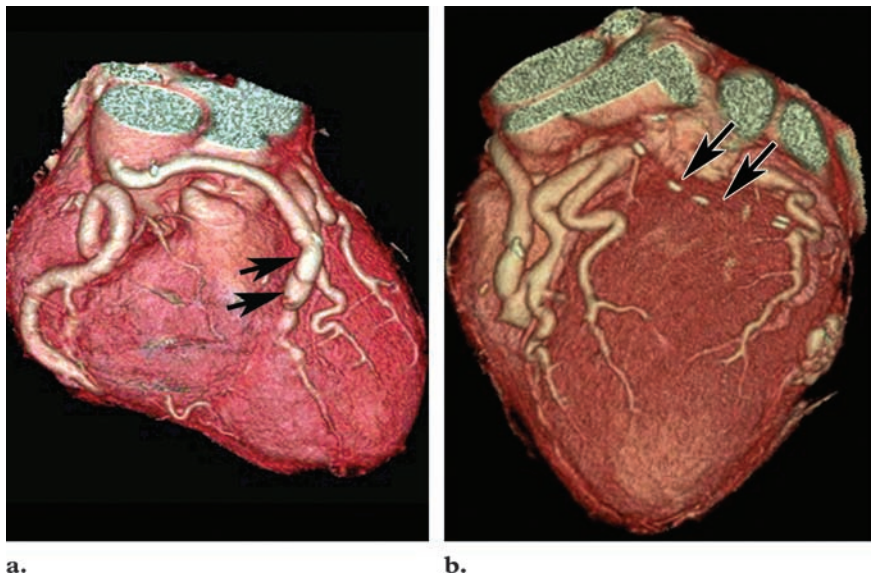


Figure 14. CABG placement with ligation in a 50-year-old man with ALCAPA syndrome. **(a)** Volume-rendered CT image shows a venous graft extending from the aorta to the mid LAD artery with focal atherosclerotic dilatation but no narrowing (arrows). **(b)** Volume-rendered CT image shows a second graft extending from the aorta to the first marginal artery; this graft is occluded (arrows).

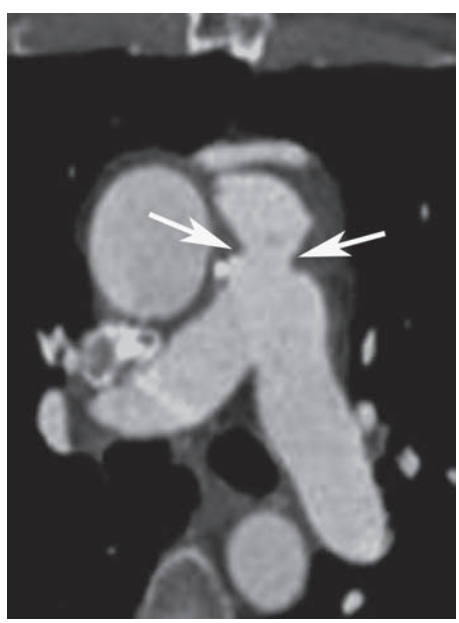
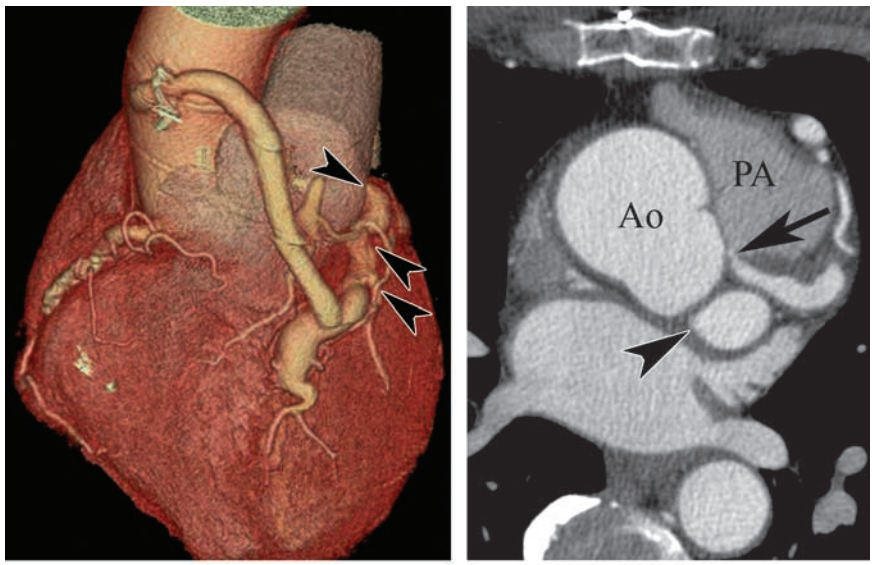


Figure 15. Unsuccessful coronary button transfer in a 52-year-old woman with ALCAPA syndrome. The friable LCA was torn intraoperatively, and the coronary button transfer procedure was halted. Two venous CABGs were then constructed to the LAD artery and the left circumflex artery. Multidetector CT angiogram acquired 2 years later shows postoperative supravalvular pulmonary stenosis (arrows) at the site of the coronary button transfer.

LCA is associated with potential graft occlusion and stenosis (Fig 14). In adults, coronary button transfer carries the risk for tearing of the anomalous LCA and massive bleeding due to increased friability and diminished elasticity when the anomalous LCA is mobilized for repair (Fig 15). In the Takeuchi procedure, a transpulmonary baffle made from the pulmonary arterial wall is used to create a coronary tunnel to connect the anomalous LCA to the aorta. Complications such as supravalvular pulmonary stenosis (26,30) and baffle obstruction or leakage have been reported. Simple ligation of the anomalous LCA is no longer performed because of the high prevalence of persistent shunting and the risk of sudden death from persistent chronic ischemia (Figure 16) (1).

Differential Diagnosis

The differential diagnosis for dilatation of the coronary arteries includes Kawasaki disease, coronary artery–coronary sinus fistula, atherosclerosis-related coronary artery ectasia, vasculitis (polyarteritis nodosa or Takayasu arteritis), scleroderma, Ehlers-Danlos syndrome, hereditary hemorrhagic telangiectasia, trauma, and hyperlipidemia (Table 7) (30–32).



a. b.

Figure 16. ALCAPA syndrome in a 64-year-old man who underwent ligation of an LAD artery arising from the pulmonary artery and placement of a venous bypass graft from the aorta to the proximal LAD artery distal to the ligation. **(a)** Volume-rendered CT image shows the venous graft and a patent proximal LAD artery (arrowheads). **(b)** Axial CT image shows a faint jet of contrast material in the main pulmonary artery *(PA)* that emanates from the previously ligated LAD artery (arrow), a finding indicative of recanalization of the ALCAPA. Note the dilated left main coronary artery (arrowhead). *Ao* = aorta.

Table 7
Key Differentiating Features of Dilated Coronary Arteries

Disease Entity	Imaging Findings	Differentiating Features
ALCAPA syndrome (adult type)	Diffuse dilatation of the anomalous LCA and the RCA with dilated intercoronary collateral vessels	LCA arises from the main pulmonary artery
Coronary artery dilatation related to atherosclerosis	Diffuse coronary artery dilatation	Atherosclerotic plaque in affected coronary arteries
Kawasaki disease	Multiple focal coronary artery aneurysms	Seen in young patients with a history of viral infection
Coronary artery–coronary sinus fistula	Tortuous coronary artery associated with dilated epicardial veins and coronary sinus	Arteriovenous communication; only the artery leading to the fistula is dilated
Takayasu arteritis	Coronary artery aneurysms and stenoses	Involvement of the aorta and great vessels

Summary

ALCAPA syndrome is a rare congenital anomaly in which the LCA originates from the main pulmonary artery. There are two types: the infant type and the adult type, each of which has different clinical manifestations and carries a different prognosis. On ECG-gated multidetector CT angiographic and MR images, direct visualization of the LCA originating from the main pulmonary artery is a primary imaging feature of ALCAPA syndrome. Another important imaging feature is retrograde flow from the LCA into the main pulmonary artery on cine MR images. Because of its ability to demonstrate

subendocardial infarction, delayed enhancement MR imaging plays an important role in clinical management in adult patients who present with this anomaly.

The treatment of choice for ALCAPA syndrome is surgical repair. The preferred surgical option is to restore a two-coronary-artery system to correct the coronary steal phenomenon. There are characteristic postoperative complications associated with each type of surgical repair that may be seen at ECG-gated multidetector CT angiography. Although ALCAPA syndrome is rare, it is important to be aware of this potentially lethal condition. The diagnosis may be made with ECG-gated multidetector CT angiography and MR imaging. Prompt treatment may prevent life-threatening complications, such as myocardial infarction or sudden death.

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ALCAPA Syndrome: Not Just a Pediatric Disease

Elena Peña, MD, et al

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Page 554
The extent of acquired collateral circulation between the RCA and LCA during the critical period, when pulmonary arterial pressure gradually decreases, determines the extent of myocardial ischemia. Patients with well-established collateral vessels have the adult type of the disease, and those without collateral vessels have the infant type. Both types of the disease have different manifestations and outcomes.

Page 555
As pressure decreases in the pulmonary circulation and as flow reverses in the LCA, the LCA fails to supply the myocardium and "drains" fully oxygenated blood into the main pulmonary artery. Thus, there is preferential blood flow into the low-pressure pulmonary circulation rather than into the high-resistance myocardial circulation. This left-to-right shunt is known as the steal phenomenon. To survive beyond infancy, patients with ALCAPA syndrome develop significant collateral circulation from the RCA to the LCA. However, often it is not sufficient to supply the left ventricle, especially in the subendocardial region; chronic left ventricular subendocardial ischemia ensues. As a result, patients may develop malignant ventricular dysrhythmias. These patients are at risk for sudden cardiac death, which occurs in 80%-90% of cases.

Page 557
Direct visualization of the LCA arising from the main pulmonary artery is the diagnostic hallmark of ALCAPA syndrome.

Page 558
Surgical correction performed on making a diagnosis of ALCAPA syndrome is considered to be the standard treatment. The aim of surgery is to restore a two-coronary-artery circulation system.

Page 560
Of these options, coronary button transfer is considered to be the most anatomic correction, and it has excellent long-term results. It is the preferred method of treatment in infants (20). In adults, the preferred method is ligation of the LCA at its origin from the main pulmonary artery to stop competitive flow combined with CABG placement by using the internal mammary artery or a saphenous vein.

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