

Video Article

Direct Re-implantation of Left Coronary Artery into the Aorta in Adults with Anomalous Origin of Left Coronary Artery from the Pulmonary Artery (ALCAPA)

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Abstract

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly which is one of leading causes of myocardial ischemia and infarction in children. If left untreated, it results in a 90% mortality rate in the first year of life. In patients who survive to the adulthood, the coronary steal phenomenon and retrograde left-sided coronary flow provide a substrate for chronic subendocardial ischemia, which may lead to left ventricular dysfunction, ischemic mitral regurgitation, malignant ventricular arrhythmias, and sudden cardiac death. The average age of life-threatening presentation is 33 years and of sudden cardiac death 31 years. Therefore, surgical correction is highly recommended as soon as the diagnosis is made, regardless of age. In adult-type ALCAPA originating from the right-facing sinus of the pulmonary artery, direct re-implantation of the ALCAPA into the aorta is the more physiologically sound repair technique to re-establish the dual-coronary perfusion system and is recommended. This protocol describes the technique of direct re-implantation of adult-type ALCAPA into the aorta.

Video Link

The video component of this article can be found at <https://www.jove.com/video/55590/>

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly usually seen as an isolated lesion¹. The incidence of ALCAPA is estimated at 1 in 300,000 live births, comprising between 0.24% and 0.46% of congenital cardiac diseases^{2,3}. It is one of the most common causes of myocardial ischemia and infarction in children and, if left untreated, results in a 90% mortality rate in the first year of life⁴. Only 10 - 15% of infants survive to adulthood due to the rapid development of a large dominant right coronary artery (RCA) with extensive intercoronary collaterals⁴. During the neonatal period, high pulmonary vascular resistance and the resultant pulmonary artery (PA) pressures ensure that antegrade flow is maintained from the PA into the anomalous left coronary artery. As the pulmonary vascular resistance gradually decreases, antegrade flow to the left coronary artery reduces. This eventually leads to reversal of flow, and left-to-right shunting into the PA, thus resulting in a "coronary steal"⁵. Thus, left ventricular (LV) myocardial perfusion depends on intercoronary collaterals from an RCA^{5,6}.

The coronary steal phenomenon and retrograde left-sided coronary flow provide a substrate for chronic subendocardial ischemia, which may lead to left ventricular dysfunction, ischemic mitral regurgitation, and malignant ventricular arrhythmias precipitated by acute myocardial ischemia⁷. In a subset of adult patients, the mean age at presentation is 41 years, with a shift in sex distribution toward female patients (Female-to-male ratio: 2:1)⁸. In this patient population, 14% are asymptomatic; 66% present with symptoms of angina, dyspnea, palpitations, or fatigue; and 17% present with life-threatening symptoms, including ventricular arrhythmias, syncope, and sudden cardiac death⁸. The average age of life-threatening presentation is 33 years and of sudden cardiac death 31 years⁸. Therefore, surgical correction is highly recommended as soon as the diagnosis is made, regardless of age or the degree of intercoronary collateralization^{1,9}.

Depending on the origin of the anomalous left coronary artery, direct re-implantation of the ALCAPA into the aorta is the more physiologically sound repair technique to reestablish the dual-coronary perfusion system. Most commonly, ALCAPA takes off from the right-hand pulmonary sinus (sinus 1 of the PA), which faces the aortic sinus where the main left coronary artery usually originates (sinus 2 of the aorta)¹⁰. This coronary anatomy is most suited to the direct re-implantation technique. The aim of this report is to describe, in detail, the technique for the direct re-implantation of the left coronary artery in ALCAPA in adult patients. The rationale behind direct re-implantation is the advantage-the physiological

reestablishment of dual-coronary perfusion-it offers over the ligation of the anomalous left coronary combined with coronary artery bypass grafting^{11,12,13}.

Protocol

The protocol follows the institutional guidelines of the human research ethics committee of the University of Zurich.

1. Preparation for Surgery

1. Clean and prepare the surgical suite in a typical manner. To facilitate communication between the surgeon and the perfusionist, place the heart-lung machine to the left of the patient, opposite to the surgeon.
2. Pre-medicate the patient by the oral administration of 5 mg of midazolam, 30 - 60 min prior to the induction of anesthesia.
3. Let the patient be monitored according to standard guidelines, with direct arterial and central venous pressure access by the anesthesiologists. Induce anesthesia using an initial intravenous injection of 0.5 - 1.5 mg/kg propofol, 1 - 2 µg/kg fentanyl, and 0.6 mg/kg rocuronium.
4. Perform tracheal intubation and maintain the anesthesia through the intravenous infusion of 100 - 150 µg/kg/min propofol, 0.015 - 0.03 µg/kg/min fentanyl, and 0.6 - 1.2 µg/kg/min rocuronium as needed.
5. Install the patient in a supine position and drape her/him in a sterile fashion, leaving the chest, the abdomen, and the groin free in the operative field.

2. Surgery

1. To minimize crowding of the operative field by cannulas, establish the cardio-pulmonary bypass access outside the thorax.
 1. Perform a 6 cm-long incision 2 cm below and parallel to the right clavicle. Prepare the right subclavian artery, freeing it from the surrounding tissue. Give 300 I.U. of heparin/kg through the IV line.
 2. Side-clamp the right subclavian artery with a Cooley Derra clamp. Perform a 10 mm-long arteriotomy of the vessel parallel to its axis.
 3. Sew an 8-mm Dacron graft end-to-end to the arteriotomy using a running 6/0 polypropylene monofilament suture (e.g., Prolene). Cannulate the graft with a 24 Fr elongated one-piece arterial cannula for arterial return to the patient.
 4. Under transesophageal echocardiographic guidance, cannulate the right femoral vein percutaneously over a guidewire for venous drainage.
2. Access the heart through a median sternotomy.
 1. Incise the skin longitudinally over 15 cm, starting 1 cm below the suprasternal notch. Take care to stay in the middle of the sternal width.
 2. Saw the sternum with an oscillatory saw. Take care to stay in the middle of the sternal width.
 3. Incise the pericardium over the aorta to uncover the massively dilated and tortuous right coronary artery.
 1. Grab the pericardium with Carpentier dissection forceps. Cut the pericardium with Metzenbaum scissors. Continue cutting the pericardium up to its reflection line over the ascending aorta using electrocautery.
 4. Start the extra-corporeal circulation and dissect the aorta circumferentially, freeing it from the surrounding tissue, including the pulmonary artery, taking care not to injure the anomalous left coronary artery.
 1. Gently push the ascending aorta to the left using a lung retractor. Grab the soft tissue behind the ascending aorta with Carpentier dissection forceps.
 2. Separate the posterior wall of the ascending aorta from the surrounding tissue using electrocautery. Continue caudally behind the ascending aorta. Make sure not to injure the right pulmonary artery behind the ascending aorta.
 3. Grab the main pulmonary artery with Carpentier dissection forceps and push it gently towards the left side. Separate the left lateral wall of the ascending aorta from the main pulmonary artery using electrocautery.
 5. Dissect the main pulmonary artery free from the surrounding tissue.
 1. Pull the ascending aorta towards the right side using a peanut gauze mounted on an Allis clamp.
 2. Bluntly dissect the posterior wall of the main pulmonary artery with a second peanut gauze mounted on an Allis clamp. Make sure not to injure the main left coronary artery behind the main pulmonary artery.
 3. Pass a dissector under the main pulmonary artery. Gently open the dissector to prepare a tunnel for the passage of the vessel loop.
 4. Bring down a vessel loop mounted on Carpentier dissection forceps and place it between the open jaws of the dissector. Close the jaws of the dissector to grab the vessel loop.
 5. Grab the adventitia of the ascending aorta and pull the vessel loop held by the dissector to encircle the main pulmonary artery.
 6. Place a left ventricular vent.
 1. Place a purse-string on the right upper pulmonary vein with a 4/0 polypropylene monofilament suture.
 2. Gently push the superior vena cava towards the left side.
 3. Make a small cut in the middle of the purse-string on the right upper pulmonary vein with an 18-blade knife (see the Materials List).
 4. Gently dilate the opening with a Leriche hemostatic clamp.
 5. Insert a vent through the right upper pulmonary vein and the mitral valve into the left ventricle to unload the left heart. Secure it with a braided polyester 2/0 ligature with polybutylene coating.

7. Place a retrograde cardioplegic cannula through the coronary sinus. Secure it with a braided polyester 2/0 ligature with a polybutylene coating.
8. Install the antegrade cardioplegic root.
 1. Place a purse-string on the main pulmonary artery with a 4/0 polypropylene monofilament suture.
 2. Make a small cut in the middle of the purse-string on the main pulmonary artery with an 18-blade knife (see the Materials List).
 3. Gently dilate the opening with a Leriche hemostatic clamp. Insert an antegrade cardioplegic cannula in the main pulmonary artery. Secure it with a braided polyester 2/0 ligature with polybutylene coating.
 4. Place a purse-string on the ascending aorta with a 4/0 polypropylene monofilament suture.
 5. Make a small cut in the middle of the purse-string on the ascending aorta with an 18-blade knife.
 6. Gently dilate the opening with a Leriche hemostatic clamp.
 7. Insert an antegrade cardioplegic cannula into the ascending aorta. Secure it with a braided polyester 2/0 ligature with polybutylene coating. Cut the ligature with Metzenbaum scissors. Verify the correct placement of the antegrade cardioplegic cannulas in the main pulmonary artery and ascending aorta.
3. Prepare to open the aorta.
 1. Cross-clamp the aorta as distally as possible. Deliver the antegrade cold blood cardioplegia simultaneously through the aorta and the pulmonary artery.
 2. Remove the cannulas from the aorta and the pulmonary artery and repeat the cold blood cardioplegia retrogradely every 20 min.
 3. Transect the aorta.
 1. Grab the ascending aorta with Carpentier dissection forceps on each side of the opening left following the removal of the cardioplegic cannula. Enlarge the opening with an 18-blade knife.
 2. Finish the transection of the aorta with Metzenbaum scissors. Verify the absence of the left coronary ostium inside the aorta in the left sinus of Valsalva.
 4. Transect the main pulmonary artery.
 1. Grab the main pulmonary artery with Carpentier dissection forceps on each side of the opening left following the removal of the cardioplegic cannula. Enlarge the opening with an 18-blade knife.
 2. Finish the transection of the main pulmonary artery with Metzenbaum scissors. Confirm the presence of the left coronary ostium, originating from the right-facing sinus 1¹⁰ of the pulmonary artery.
 5. Detach the left coronary ostium from the right-facing sinus 1 of the pulmonary artery.
 1. Grab the proximal main pulmonary artery with Carpentier dissection forceps. Using Metzenbaum scissors, separate the left coronary ostium from the right-facing sinus 1 of the pulmonary artery with a generous surrounding patch of the pulmonary root wall, taking care not to injure the pulmonary valve.
 2. Mobilize the main left coronary artery up to its bifurcation.
4. Prepare for the re-implantation of the left coronary ostium.
 1. Grab the proximal ascending aorta with Carpentier dissection forceps.
 2. Use a straight blade to create a neo-ostium in the left sinus 2 of the aorta. Leave a 10-mm margin of aortic root wall around the neo-ostium towards the commissure between the left sinus 2 and the right coronary sinus of the aorta, as well as towards the aortic annulus.
 3. Connect the main left coronary ostium end-to-end to the neo-ostium in the left sinus 2 of the aorta using a running 6/0 polypropylene monofilament suture. Start the anastomosis at the deepest point of the main left coronary ostium and allow it to come up on the right-hand side of the anastomosis. Complete the anastomosis by running the left-hand side of the suture to meet the other end.
 4. To relieve tension on the tissue when pulling the sutures, bring the aorta and the main left coronary ostium together each time.
5. Repair the defect in the pulmonary artery root.
 1. Use a non-treated autologous pericardial patch.
 2. Connect the autologous pericardial patch at the deepest point of the right-facing sinus 1 of the pulmonary artery using a running 6/0 polypropylene monofilament suture.
 3. First run up the left and then the right end of the suture to mid-height of the defect in the right-facing sinus 1 of the pulmonary artery. Leave the two ends of the suture under tension.
6. Reestablish the continuity of the great vessels.
 1. Reconnect the proximal part of the aorta to its distal part with an end-to-end anastomosis using a running 5/0 polypropylene monofilament suture. Start the anastomosis at the deepest point, allowing it to come up on the right-hand side of the anastomosis. Complete the anastomosis by running the left-hand side of the suture to meet the other end.
 2. De-air the aorta. Remove the aortic cross-clamp. Start rewarming the patient.
 3. To shorten the ischemic time, perform the reconnection of the proximal and distal parts of the pulmonary artery on the beating heart. Put the pump sucker into the distal pulmonary artery to improve the sight of the operative field.
 4. Continue in a running fashion the left end of the 6/0 polypropylene suture that had been stopped at mid-height of the defect in the right-facing sinus 1 of the pulmonary artery to complete the posterior and left aspects of the anastomose.
 5. Continue in a running fashion the right end of the 6/0 polypropylene suture that had been stopped at mid-height of the defect in the right-facing sinus 1 of the pulmonary artery to complete the posterior and right aspects of the anastomose. Tie the two ends of the suture to finish the connection.

Representative Results

Presentation

The patient was a 48-year-old woman presenting with the recent onset of angina Canadian Cardiovascular Society (CCS) grad III and occasional palpitations. She reported three uneventful pregnancies. Moderate smoking was the main cardiovascular risk factor. Trans-thoracic echocardiography showed a moderately impaired (45%) left ventricular ejection fraction and no mitral regurgitation. A coronary angiography was then performed. It demonstrated the absence of the left main coronary artery arising from the aorta. The right coronary artery was considerably enlarged and perfused the main left coronary artery via intraseptal collaterals (**Figure 1**). Thus, the diagnosis of ALCAPA was made. The anatomical type of ALCAPA was further defined by bi-plan (**Figure 2**) and three-dimensional (**Figure 3**) CT scans.

Post-operative Course

The patient was separated from the cardiopulmonary bypass at a core temperature of 37 °C. No post-operative myocardial ischemia occurred. Bleeding from the chest tube drain was less than 30 mL/h. The patient was weaned from the ventilator and extubated 6 h post-operatively. She was discharged from the intensive care unit to the normal ward on the first post-operative day. Her course on the ward remained uneventful; she was discharged to a cardiac rehabilitation program on post-operative day 9.

One-year Follow-up

The patient was seen at the outpatient clinic one year after surgery. She was working full-time and had no angina or dyspnea. Her exercise test was negative. **Figure 4** shows her CT scan one year after surgery. The re-implanted ALCAPA is widely patent at the site of anastomosis to the aorta. The pulmonary artery does not present any narrowing at the site of reconstruction with the autologous pericardial patch used to repair the defect in the right-facing sinus of the PA.

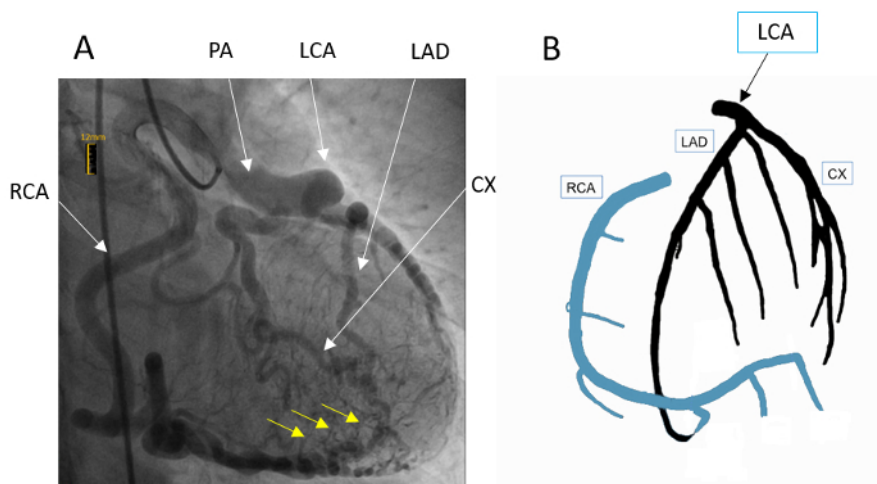


Figure 1: Pre-operative Coronary Angiography. **A)** The coronary angiography of the patient is remarkable for the absence of the ostium of the main left coronary artery (LCA) in the aorta. In a right anterior oblique view, the opacified right coronary artery (RCA) is considerably enlarged, measuring 12 mm. It follows a tortuous path across the right heart while supplying large collaterals to the left-sided circulation via intraseptal collateral vessels (yellow arrows). These collaterals feed the left anterior descending (LAD) and left circumflex (CX) arteries, which join the main left coronary artery and drain in a retrograde fashion into the main pulmonary artery (PA). **B)** This diagram shows a schematic representation of the normal distribution of the coronary arteries. (1 mm on figure scale = 2 mm) [Please click here to view a larger version of this figure.](#)

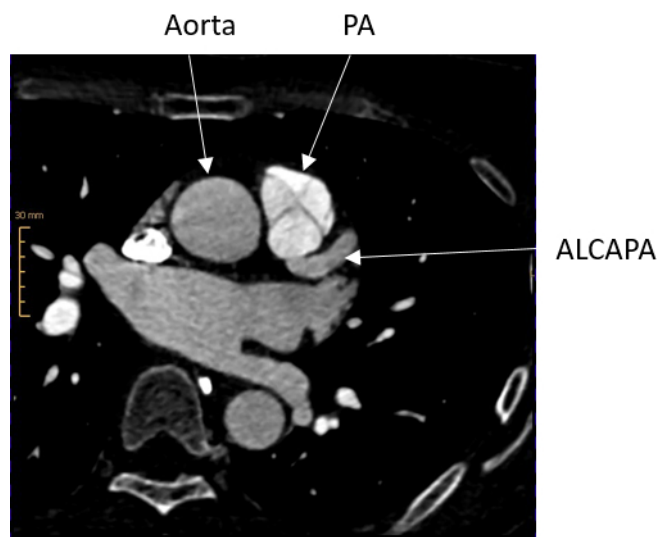


Figure 2: Pre-operative Computed Tomography. A cardiac computed tomography scan shows the take-off of the ALCAPA from the right-facing sinus of the PA according to the Dodge-Khatami classification¹⁰. (1 mm on figure scale = 2.14 mm)

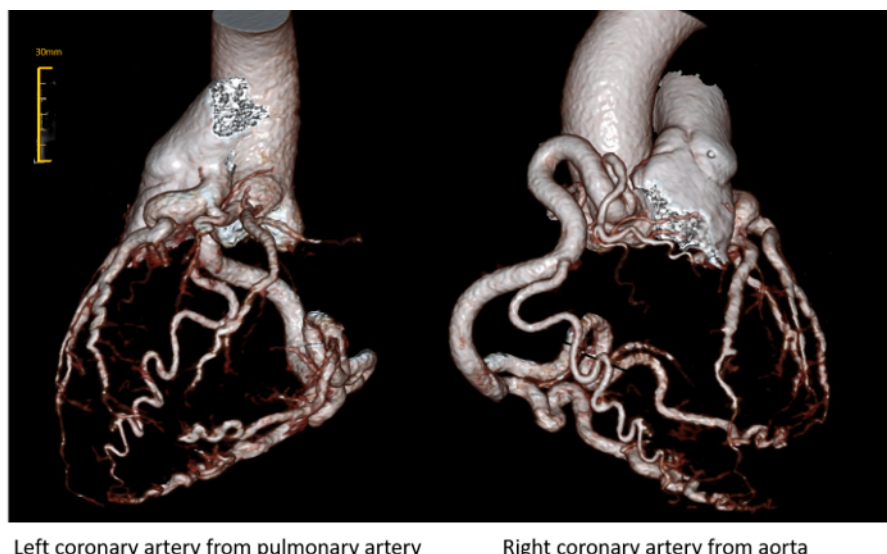


Figure 3: Pre-operative Three-dimensional Computed Tomography. A three-dimensional reconstruction of the computed tomography of the great vessels confirms the topography of the ALCAPA originating from the right-facing sinus 1 of the PA and the RCA from the facing sinus 1 of the aorta¹⁰. From this anatomy, a direct translocation of the ALCAPA to the aorta seems feasible. (1 mm on figure scale = 1.66 mm) [Please click here to view a larger version of this figure.](#)

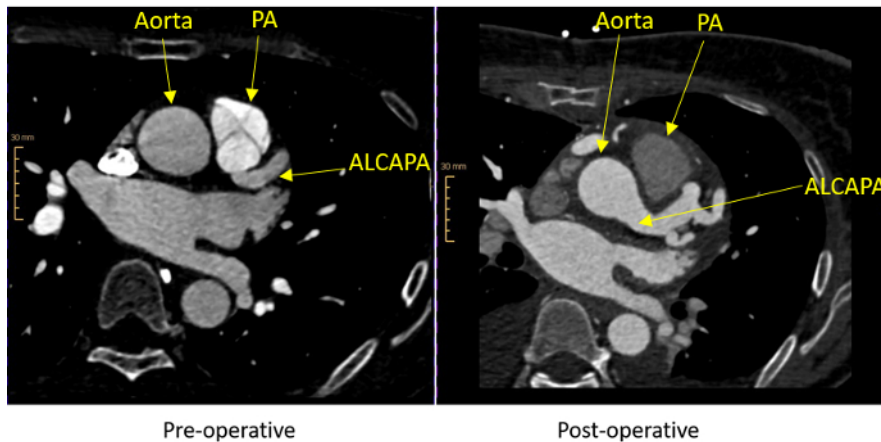


Figure 4: Follow-up Post-operative Computed Tomography. In the right panel, the computed tomography scan of the patient one year after surgery shows a widely patent main left coronary artery (ALCAPA) connected to the aorta. The reconstructed pulmonary artery (PA) does not present any narrowing. In the left panel, the pre-operative computed tomography depicts the ALCAPA originating from the right-facing sinus of the PA according to the Dodge-Khatami classification¹⁰. (1 mm on figure scale = 2.14 mm in the left panel, 1 mm on figure scale = 2.5 mm in the right panel) [Please click here to view a larger version of this figure.](#)

Discussion

This protocol describes a detailed technique for the direct re-implantation of the ALCAPA into the aorta in an adult patient with the origin of main left coronary artery from the right-facing sinus of the pulmonary artery according to the Dodge-Khatami classification¹⁰. The myocardial protection strategy and the reconstruction of the pulmonary artery are clearly demonstrated. The major critical step of this technique is represented by the generous mobilization of the left coronary artery to achieve a tension-free anastomosis.

Prolonged desaturated coronary perfusion is tolerated into adulthood in a small number of individuals. ALCAPA patients who survive to adulthood present with a spectrum of clinical manifestations, ranging from the absence of symptoms to acute myocardial infarction and/or chronic myocardial ischemia⁸. The latter would ultimately lead to left ventricular dysfunction, ischemic mitral regurgitation, malignant ventricular arrhythmias, and sudden cardiac death⁷. Therefore, surgical correction is highly recommended as soon as the diagnosis is made, regardless of age or the degree of intercoronary collateralization^{1,9}.

In both adults and infants, the direct re-implantation of the ALCAPA represents the physiological repair method and is the preferred technique when the anatomy is suitable¹. However, in adults, the direct re-implantation of the ALCAPA might be more challenging because of a very short main left coronary artery, increased coronary artery friability, diminished vessel elasticity for mobilization, and the potential for tearing and the resultant uncontrollable bleeding^{14,15}. In these situations, coronary artery bypass grafting associated with the ligation of the ALCAPA may be more suitable^{11,12,13}. In a series of 30 patients with ALCAPA, 3 of which were adults, Neumann *et al.* performed 19 direct re-implantations, 9 Takeuchi repairs, and 2 ligations¹⁶. The early and late 24-year survival rates for direct re-implantation were both 100%, and the 10-year rate of freedom from reoperation was 94.1%¹⁶. At the last follow-up, 95.5% of the patients were in New York Heart Association functional class I¹⁶.

Other options for surgical repair aim to prevent the coronary steal phenomenon and, ideally, to restore dual-coronary circulation. The simplest corrective procedure involves ligating the LMCA, which does prevent steal, but does not allow for antegrade flow into the left-sided circulation. Therefore, this is most often combined with bypass grafting^{11,12}. This approach, less physiological than the previous, is generally the most straightforward in adults, but there remains a risk of late graft stenosis, especially with saphenous vein grafts¹³. In adults, creating an intrapulmonary baffle (Takeuchi procedure) is a more complex option that does restore the dual-coronary supply but that may be complicated by supravulvar pulmonary stenosis and baffle obstruction or leakage^{13,17}.

The treatment of associated ischemic mitral valve regurgitation remains controversial and depends on the degree, as well as the functional versus structural type, of regurgitation. The age of the patient, the personal experience of the surgeon, and the ability of the center must also be considered. In infants, most authors recommend an expectative approach, unless the mitral regurgitation is severe^{1,16}. In adults, the decision on the concomitant surgical correction of functional ischemic mitral regurgitation during the operation for ALCAPA should be made with regard to the severity of the mitral regurgitation and the possibility for it to worsen after intra-operative norepinephrine tests. An aggravation of mitral regurgitation following norepinephrine challenge, evidenced by intra-operative transesophageal echocardiography, would support the correction of the mitral regurgitation. For structural mitral regurgitation, correction would be recommended for more than mild regurgitation, unless the prolongation of the cross-clamp time is deemed undesirable for the patient.

In conclusion, in adult-type ALCAPA originating from the right-facing sinus of the pulmonary artery, direct re-implantation into the aorta is recommended, provided that the tissue is of adequate elastic quality. Thickening and calcification of the main left coronary could be detected in the pre-operative computed tomography. However, the final evaluation of the tissue elasticity is performed by intra-operative visual inspection and gentle palpation of the vessel wall for the absence of calcified plaques and by the tactile perception of tissue resistance during mobilization.

Disclosures

The authors have nothing to disclose.

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