# 30 Anomalous Left Coronary Artery from the Pulmonary Artery

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Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a congenital coronary abnormality associated with high infant mortality and adult sudden cardiac death. In ALCAPA, the left coronary artery arises from variable locations in the PA system.

## Pathophysiology and Clinical Presentation

In ALCAPA, as the PVR decreases after birth, the resistance in the coronary arterial system becomes higher than in the pulmonary circulation and the blood flow in the anomalous coronary artery reverses, causing primarily a left-to-right shunt. This coronary steal in the absence of adequate collateralization results in severe myocardial ischemia and dysfunction, classically within a few weeks to months of life.

ALCAPA is characterized by chronically ischemic hypocontractile, yet potentially salvageable myocardium. The variable equilibrium between timing of closure of the ductus arteriosus, pulmonary hypertension, and the speed of development of preexisting collateral circulation between the right and left coronary arteries define the extent of myocardial necrosis and scarring of the LV. If left uncorrected, the mortality is very high. Extensive collateral arteries may enable some patients to survive beyond infancy. However, chronic hypoperfusion causes subendocardial ischemia and later fibrosis, increasing the risk of sudden death secondary to ventricular arrhythmias. There are some rare situations of advanced-age patients presenting with ALCAPA.

When presenting in infancy (most common), the history will be significant for crying during feeds (angina on exertion), diaphoresis, and tachypnea with intermittent grunting – this is a distinctive constellation of symptomatology known as Bland-White-Garland syndrome. As ischemia progresses, feeding sessions will be more brief (infant becomes a "snacker") and pallor, fatigue, and grunting become prominent. When there is good collateralization, symptoms might be more subtle with failure to thrive being a typical clinical presentation.

On physical exam, the child is typically tachycardic and tachypneic. Grunting may be present with angina or when there is established pulmonary edema from poor ventricular function and high LAP +/- MR. The precordium is hypoactive. The second heart sound may be narrowly split from elevated PA pressures and sometimes there could be a single $S_2$. There is usually a gallop ($S_3$) and at times an $S_4$ (atrial kick on a poorly compliant LV).

## Diagnosis

- **CXR (Figure 30-1).** Cardiomegaly with pulmonary edema and pulmonary hyperinflation are typical findings.
- **ECG (Figure 30-2).** Atrial enlargement (LA or biatrial). Abnormal (deep and wide) Q waves in leads I and aVL tend to be a classic finding. Nonspecific ST-T wave changes.

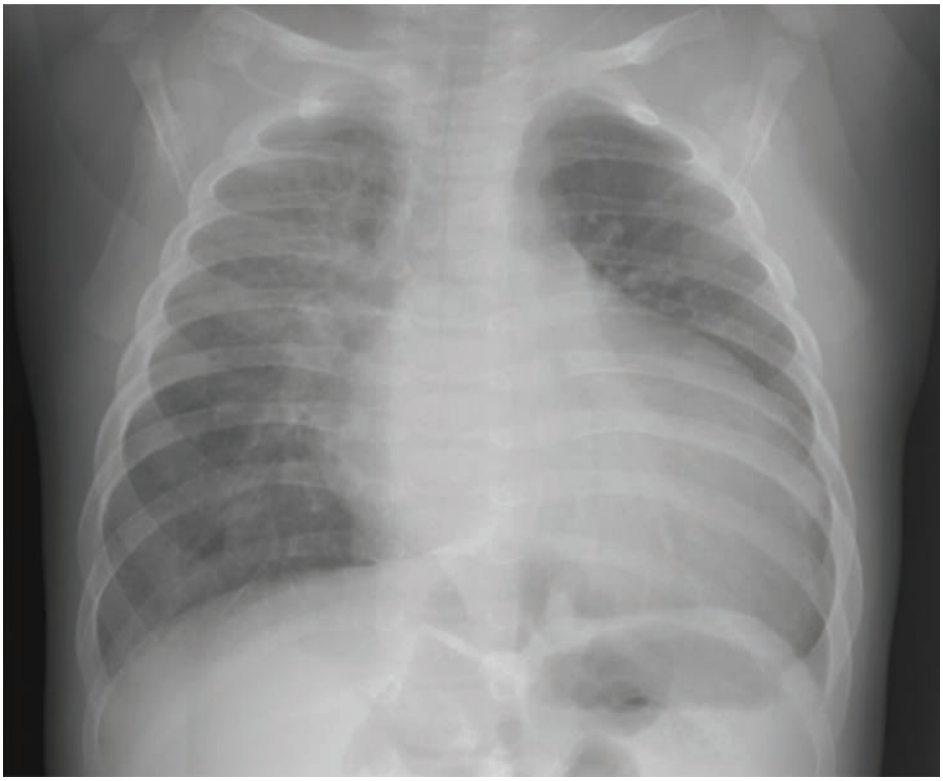

**Figure 30-1.** Anteroposterior CXR of a 5-month-old patient with ALCAPA showing cardiomegaly with LV dilation and bilateral pulmonary edema.

- **Echocardiogram (Figure 30-3).** Surface echocardiography is the primary diagnostic mode. The most important feature includes a severely depressed global LV function with MR. The LV endocardium will often appear echo bright with profound depression of contractility (ejection fraction <10%). The left coronary artery is found to arise from the main PA trunk in most cases and the ostial location is highly variable (sometimes at higher/more distal positions). Systolic and diastolic flow from the suspected anomalous coronary into the PA is pathognomonic. It is important to note that the 2D appearance of the left main coronary artery in relationship to the leftward facing aortic sinus can be very misleading and *appear* to connect to the aorta. We have seen cases in which experienced echocardiographers believe the left main coronary originates from the aorta, *but they are unable to demonstrate prograde flow by color Doppler in the coronary.* This is a situation that mandates another diagnostic study.
- **Cardiac CTA.** A properly conducted contrast CTA of the coronaries is an important diagnostic adjunct in suspected cases where the echocardiogram is insufficient or incomplete at showing the defect and there is high clinical suspicion.
- **Cardiac catheterization.** Catheterization is rarely necessary for diagnosis. This

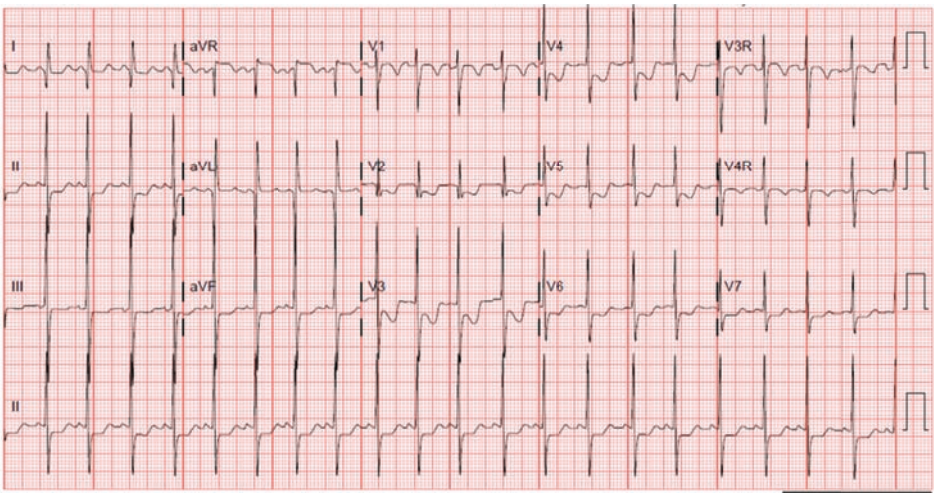


**Figure 30-2.** ECG of a patient with ALCAPA showing deep and broad Q waves in I and aVL (pathognomonic of ALCAPA). In addition, there is significant ST-segment depression in precordial leads.

may be helpful in patients in which a CTA cannot be obtained or the diagnosis remains equivocal and there is high clinical suspicion. Catheterization may be very dangerous in patients with profound ventricular dysfunction and a highly irritable myocardium. Coronary angiography is the only goal of catheterization in these cases.

## Indications / Timing for Intervention

The diagnosis of ALCAPA is an indication for surgical intervention. Surgical intervention should be performed once the diagnosis is obtained.

## Preoperative Management

Preoperatively, these infants may present in a warm/cold + wet state. Not all patients will need inotropes, but likely all will need diuresis to decrease the pulmonary symptoms from pulmonary edema and congestion. One should stay away from pure vasoconstricting agents, as they are likely to increase afterload and deteriorate function further. Noninvasive ventilation with HFNC or CPAP should be considered.

## Anesthetic Considerations

The main goals of the pre-CPB period are to maintain myocardial perfusion without increasing oxygen consumption or drastically reducing BP. If the patient is not intubated, the process of intubation may represent a high risk for cardiac arrest. One should be prepared for emergent initiation of CPB, with a primed and ready CPB circuit and the surgeon present for induction. Induction is usually incremental, with titrated doses of

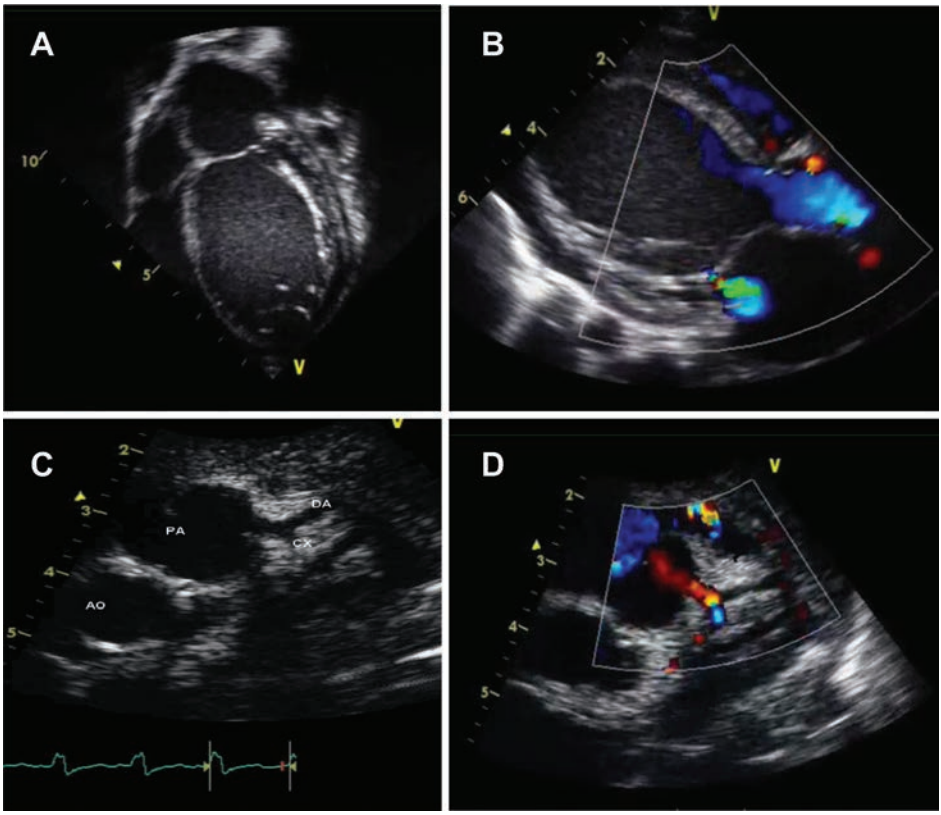


**Figure 30-3.** Echocardiograms of patients with ALCAPA. The apical 4-chamber (A) and parasternal long-axis (B) views show a severely dilated and globular LV with secondary MR. Of note, this can be easily misdiagnosed as dilated cardiomyopathy. A 2D parasternal short-axis view (C) demonstrates the takeoff of the anomalous left coronary from the PA. A color Doppler image (D) shows reverse flow from the left coronary artery into the PA. Ao: Aorta, CX: Circumflex coronary, DA: Anterior descending coronary, PA: Pulmonary artery. Images courtesy of Dr. Josh Kailin, www.pedecho.org.

anxiolytic and narcotic with or without small doses of anesthetic vapor. Arterial access should be obtained as quickly as feasible to facilitate close hemodynamic monitoring. Remember that while it is important to maintain normal oxygen saturation, maneuvers which decrease PVR should be avoided in order to keep (left) coronary perfusion pressure as high as possible.

Post-CPB, even with the "revascularization" of the left system, myocardial function does not usually immediately improve. Inotropic agents (milrinone/epinephrine) at moderate doses are often needed to maintain good cardiac output. While afterload reduction might be desirable, it is not uncommon to need vasoconstriction to maintain an adequate BP. Discuss with the surgeon the need or desire for an agent like nitroglycerin, if tolerated, to promote dilation of the reimplanted coronary.

## Surgical Repair

Direct aortic reimplantation (Figure 30-4) or creation of an intrapulmonary baffle (Figure 30-5) when coronary translocation is not feasible (rarely) have been the primary modes of repair in the recent era. Primary ligation has been virtually abandoned as it is well established that even in the setting of profound LV dysfunction, establishing a two-coronary system by whatever means confers a survival advantage over simple ligation. Preservation of the two-coronary system leads to recovery of LV function and long-term survival rates greater than 80% or better in patients with profound heart failure at presentation. Historical operations including left subclavian artery "turn-down" and coronary artery bypass grafting are now almost never indicated. MR from LV dilation, mitral annular dilatation, or papillary muscle ischemia and secondary dysfunction may also occur in ALCAPA patients and may require the consideration of concomitant surgical treatment.

Intraoperative management of these patients must be a highly choreographed scenario. The anesthesiologist will be alerted to the very precarious nature of the patient's condition and as such, will avoid techniques that increase myocardial demand or lower systemic BP precipitously. *All members of the surgical team must be in the OR during the induction of anesthesia and early preparation of the patient.* This includes the perfusion

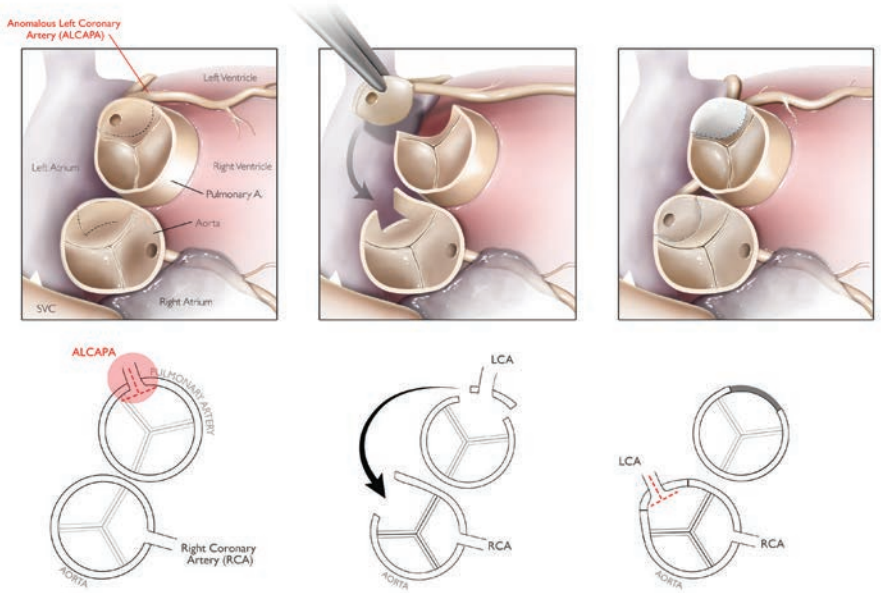


**Figure 30-4.** Translocation of a left coronary artery arising from the left posterior sinus of the PA. A generous button is harvested around the coronary artery, the coronary artery is widely mobilized, a trapdoor incision is created in the optimal location of the aortic root, and the button is reimplanted avoiding any torsion of the vessel. The PA defect is reconstructed with an autologous pericardial patch.

team who should be prepared for the urgent institution of CPB in critical situations. During sternal entry, the surgeon and assistants must be extremely careful in avoiding unnecessary manipulation of the heart. These hearts may be extremely irritable and even a minimal brush of the heart can induced intractable dysrhythmias or even ventricular fibrillation, a scenario that may require urgent initiation of CPB.

**TCH experience with ALCAPA repair (1996-2011)** (Cabrera et al. 2015)
Number of patients: 34
Median age at surgery: 5 months (3 days – 39 years)
Median ICU length of stay: 7 days (1-26 days)
Median hospital length of stay: 16 days (3-540 days)
Perioperative mortality: 0
Postoperative mechanical circulatory support: 0

We have favored separate vena caval cannulation and moderate hypothermia (nasopharyngeal temperature of ~30-32 °C). After initiating CPB, the pulmonary vasculature is completely decompressed leading to further coronary steal. It is critical for the surgeon to gain circumferential control of either the main or branch PAs *distal* to the anomalous coronary ostium such that the PA may be occluded after initiating bypass. This also facilitates the effectiveness of antegrade, aortic root cardioplegia. After the heart is arrested, the main PA is strategically opened anteriorly at or just proximal to the bifurcation. The location of the anomalous ostium is then visualized. One should note that the ostium may originate from anywhere on the main or proximal branch PAs. Given the extensive and very successful experience congenital heart surgeons have with the arterial switch operation for transposition of the great arteries, coronary artery translocation to the ascending aorta is performed now in almost all newborn and infant cases. As patients get older, and particularly in rare adult cases, the coronary ostium is not as elastic and translocation may be inappropriate. This decision has to be made on a case-by-case basis. Our experience has been that in small children, all ostial locations, including an anterior and leftward location of the coronary, are amenable to translocation to the aorta (Figure 30-4). The surgeon should mobilize the ostium as a very liberal "button" of PA wall. In posteriorly located ostia, this may include taking down the posterior pulmonary valve commissure. The coronary is then mobilized to optimize the translocation with minimal traction and avoidance of axial torsion. The location for translocation to the aorta is selected on the basis of the best available geometric location; this is not necessarily the true leftward-facing aortic sinus in all cases. We have used an appropriately oriented "trapdoor" flap incision in the aorta to facilitate the coronary anastomosis, which is accomplished with a running, non-absorbable suture. In some cases, surgeons have used a small, anterior patch (typically autologous pericardium) to augment the reconstructed neo-ostium, although we have not found this to be necessary. It is very important to reconstruct the PA sinus defect with a generous pericardial patch, again much as one would do with an arterial switch operation, prior to removing the aortic cross-clamp.

In cases where coronary ostial translocation is not believed to be feasible, an intrapulmonary tunnel (as originally described by Takeuchi and colleagues) may be constructed across the back wall of the PA (Figure 30-5). Options for tunnel construction include a native anterior flap of PA wall or some form of prosthetic material. A neo-ostium is

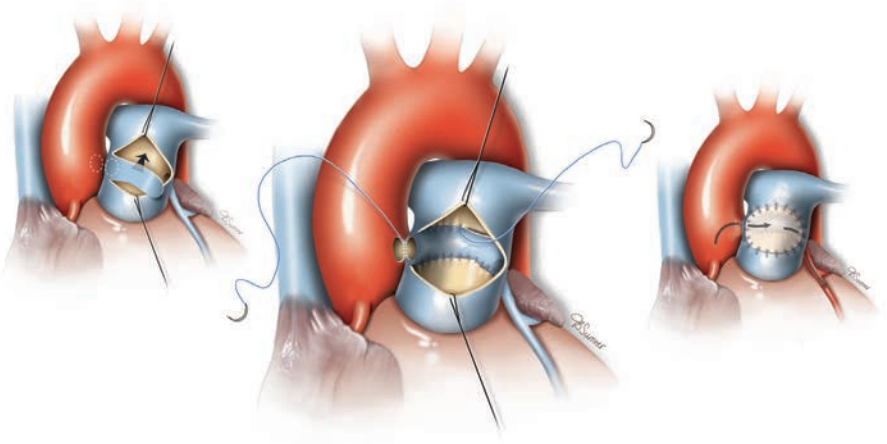

**Figure 30-5.** Takeuchi repair for ALCAPA. A baffle is created inside of the PA with either an anterior flap of PA tissue or other type of material, and an aortopulmonary window is created to redirect the flow from the aorta, through the intrapulmonary baffle, and into the left coronary artery. The anterior PA segment is reconstructed with a generous patch.

constructed which essentially amounts to the creation of an aortopulmonary window. It is critical to liberally augment the anterior PA wall deficit with pericardium or some other patch. Failure to do so has been associated with a high incidence of supravalvar PS in patients undergoing this operation.

Deciding whether or not to intervene on important MR at the index coronary artery operation can be very challenging. In patients with profound ventricular dysfunction, the additional obligate myocardial ischemic time required for mitral valve repair/annuloplasty may be critical. As such, our approach has been that in patients with massively dilated LV with MR in the setting of profound LV dysfunction, we have not proceeded with mitral repair. Our belief has been, and this has been born out in our complete avoidance of postoperative mechanical circulatory support, is that with improved myocardial performance, the MR may improve. Alternatively, persistent important MR may be dealt with at a subsequent operation, at a time when LV function is improved.

Despite being compromised, these patients are for the most part managing an adequate cardiac output when they come into the OR. It is our belief that we should, through diligent attention to myocardial protection, be able to get the patient through the operation without needing a VAD or ECMO. In the setting of refractory heart failure or inability to wean from CPB, we would likely favor a temporary LVAD (left atrium to ascending aorta) over ECMO.

Although rare, some patients will present with ALCAPA late in life. We have seen several adult patients with ALCAPA and massive right-to-left coronary collateralization who have normal LV function. In general, we have still favored operating to create a two-coronary system in such individuals, although this may be a difficult and controversial decision.

## Postoperative Management

The postoperative management is mainly directed to address LV systolic dysfunction, early identification of arrhythmias, and transitioning from IV vasoactive medications to oral decongestive therapies (i.e., angiotensin converting enzyme-inhibitors, beta-blockers, diuretics).

### General Management

- **Fluids.** 25% or less maintenance with D5%/0.45% NS is standard. Careful attention should be paid to manage the patient with the minimal necessary preload. Unnecessary preload may worsen myocardial wall stress and lead to further ventricular dysfunction and hypotension. An LA line will facilitate adjudication of intracardiac filling.
- **Analgesia and sedation.** Analgesics and sedatives are adjusted for patient's comfort. For analgesia, a fentanyl infusion is used with scheduled acetaminophen (enteral, rectal, or IV) as an adjuvant. Sedation is achieved with a combination of dexmedetomidine (both intubated and extubated patients) and/or benzodiazepines. Midazolam as a drip is preferred, as significant shifts in afterload or BP may produce instability.
- **Vasoactive drugs.** Most patients will arrive from the OR on a milrinone infusion 0.25-0.75 mcg/kg/min and an epinephrine infusion 0.02-0.05 mcg/kg/min. Hypotension should be primarily managed with inotropes when LV filling pressure (LAP) is >5-10 mmHg.
- **Mechanical ventilation.** Patients are usually ventilated on SIMV-VC with pressure support, Vt 8-10 mL/kg and PEEP 5-7 $cmH_2O$, aiming for a pH of 7.35-7.45 and $SaO_2$ >95%. Postoperative ALCAPA patients are expected to be extubated when their pulmonary edema has improved. Ventricular dysfunction takes weeks to months to show measurable echocardiographic improvement. Extubation should not be delayed as long as the patients are managing an adequate systemic cardiac output. Active preload reduction with diuretics before extubation tends to attenuate the effects of significant MR.

### What to Expect in the First 24 Hours Postoperatively

- **Vasoactive drugs.** Probably reasonable to manage milrinone and low-dose epinephrine (<0.03 mcg/kg/min) through extubation to support the LV, as extubation will lead to an increase in transmural pressure and consequently higher afterload.
- **Ventilation.** Transitioning from the OR, the lungs will be significantly improved from the preoperative period secondary to continuous ultrafiltration. There may still be some lung injury from pulmonary edema as the LV end-diastolic pressure is unlikely to change significantly in the immediate postoperative period.
- **Fluids.** Even to slightly negative. If a peritoneal dialysis catheter is present, it should be used from the day of surgery.
- **Nutrition.** If considering to extubate within 24 hrs, it is not necessary to write for TPN. If longer periods of mechanical ventilation are anticipated, full TPN should be ordered. Oral feeds should be reinstated once successfully extubated.

## Complications

The most common postoperative complications after ALCAPA repair are:

- **LCOS.** Primarily treated with inotropes. A combination of low-dose epinephrine and standard-dose milrinone. High inotrope doses increase the likelihood of arrhythmias.
- **Arrhythmias.** The incidence of arryhthmias after ALCAPA repair at TCH is 9%. Any arrhythmia (atrial tachycardia or ventricular tachycardia), should be treated promptly and should raise the clinical suspicion of deterioration in ventricular function or worsening MR.
- **Mechanical circulatory support.** Although other institutions have used pre- and/or postoperative mechanical circulatory support, at TCH we have been able to recover all of these patients without mechanical assistance.

## Long-Term Follow-Up

Despite excellent LV recovery and long-term survival rates after ALCAPA repair, follow-up complications such as persistent MR, late-onset CHF, and coronary arterial stenosis may necessitate reinterventions, including heart transplantation. As such, long-term follow-up with appropriate diagnostic testing (including provocative testing for ischemia later in life) is paramount.

Outpatient management of patients after ALCAPA repair requires ongoing assessment of systolic and diastolic ventricular function. Conventional methods have largely relied on echocardiographic parameters such as shortening fraction or ejection fraction. Myocardial strain has been a useful tool in detecting myocardial dysfunction to identify subclinical dysfunction before the echocardiogram can detect meaningful differences in ejection or shortening fraction.

### Suggested Reading

Cabrera AG, Chen DW, Pignatelli RH, et al. Outcomes of anomalous left coronary artery from pulmonary artery repair: beyond normal function. Ann Thorac Surg 2015;99:1342-1347.