Operative Techniques in Thoracic and Cardiovascular Surgery

Repair of Anomalous Coronary Artery From the Pulmonary Artery by Aortic Implantation

Anthony Azakie, MD

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), a rare congenital cardiac lesion, is an important cause of myocardial ischemia and infarction in children, and carries a high mortality in the first year of life. In the current era, repair of ALCAPA is most commonly performed by aortic implantation. Coronary elongation techniques are available for cases where the ectopic coronary artery originates at point in the pulmonary artery that is distant from the aortic root. In cases where clinically significant mitral regurgitation (MR) is present mitral valve repair is usually not performed. The early outcomes for aortic implantation of ALCAPA are excellent in the current era, with survival rates exceeding 90%. A small number of children, those who present with cardiogenic shock and very poor ventricular function, may require post-repair mechanical circulatory support using ECMO or LVAD and still have high survival rates and expect good long-term recovery. Following repair of ALCAPA by aortic implantation, ventricular functional parameters such as depressed ejection fraction, ventricular dilation and MR should recover within 8 months of repair.
Operative Techniques in Thoracic and Cardiovasculary Surgery 20:135-147

KEYWORDS coronary anomaly, ALCAPA, aortic implantation, mitral regurgitation

Introduction

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac anomaly that usually presents in infancy.[1] A minority of children who escape clinical detection and survive infancy can present the same in adulthood. The lesion is associated with coronary steal, a left-to-right shunt, and myocardial ischemia that worsens as the pulmonary vascular resistance drops in the first few weeks of life. Myocardial ischemia and infarction ensue. Mitral regurgitation (MR) can develop because of papillary muscle ischemia or infarction, or because of left ventricular (LV) dilation and mitral annular enlargement. If left unrepaired, most patients with this lesion die in infancy.

A number of surgical strategies have been used to address ALCAPA, including anomalous coronary ligation, aortic implantation, Takeuchi tunneling, coronary artery bypass grafting, or anastomosis to the subclavian artery.[2-8] In the current era, immediate repair is indicated and it involves establishment of a dual coronary system. Our preferred approach is aortic implantation even in cases where the anomalous coronary artery originates leftward and laterally from the main pulmonary artery, where tunneling may be a desirable option.[3-6] The Takeuchi tunnel is associated with a number of complications such as tunnel leak or obstruction of the main pulmonary artery.[7] A number of coronary artery elongation techniques are available to allow for successful, tension-free anastomosis to the aortic root.[3-6] In cases where there is clinically significant MR, the mitral valve should be left alone, especially in infants, as recovery of function is expected.[8] In the adult or adolescent with ALCAPA and moderate-to-severe MR, mitral valvuloplasty should be considered.

Although aortic implantation is the technique of choice for repair of ALCAPA, there are very rare circumstances, such as ALCAPA from the right pulmonary artery (PA), where the anomalous vessel courses through the aortic wall and where alternate strategies may need to be developed to avoid coronary artery injury. Most cases of ALCAPA from the right PA, even with an intramural segment in the aortic wall, can be repaired by implantation.[4] In the rare case where mobilization of the vessel appears hazardous, other techniques such as creation of an in situ coronary pouch, subclavian artery anastomosis, or tunneling should be considered.

Postoperatively, mechanical circulatory support may be required in children who fail to wean from cardiopulmonary bypass, an event that is likely in the child who presents cardiogenic shock.[8] Either extracorporeal membrane oxygenation or left ventricular assist device support can be used to allow for LV recovery with good outcomes. We prefer the use of extracorporeal membrane oxygenation to provide right ventricular support as the right ventricle may have been subjected to coronary steal, ischemia, and elevated PA pressures in the setting of MR (Figs. 1-5).

Pediatric Specialists of Texas, Methodist Children's Hospital, San Antonio, TX

Address reprint requests to Anthony Azakie, MD, Pediatric Specialists of Texas, Methodist Children's Hospital, 4410 Medical Dr 540, San Antonio, TX 78229. E-mail: tony.azakie@icloud.com

1522-2942/$-see front matter
http://dx.doi.org/10.1053/j.optechstcvs.2015.09.001

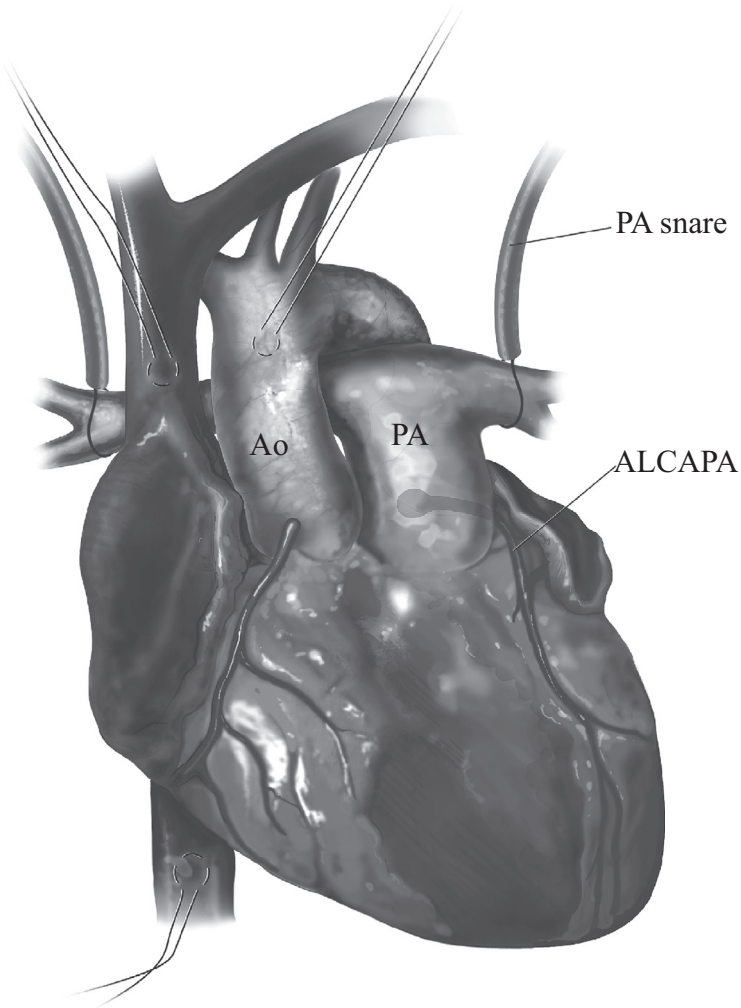

Figure 1 After induction of general anesthesia, central venous and radial arterial monitoring lines are placed. Reduction in pulmonary vascular resistance—as seen with hyperventilation, alkalosis, or administration of high FiO_2—is avoided. A median sternotomy is performed and the thymus gland is removed. The pericardium is opened and a patch is harvested for later use. The child is systemically heparinized and purse strings are placed in the distal ascending aorta, right superior vena cava, and inferior vena cava. The pulmonary arteries are dissected and encircled with heavy silk snares. The ALCAPA may be visualized at this point and fine polypropylene suture can be used to mark the recipient site on the ascending aorta. After ascending aortic and bicaval cannulation, a cardioplegia needle is inserted into the ascending aorta. High-flow CPB is instituted to support the child with mild-moderate hypothermia. CPB = cardiopulmonary bypass.