Anesthesia Management of Anomalous Aortic Origin of a Coronary Artery (AAOCA) Repair Revision

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Abstract

Anomalous Aortic Origin of a Coronary Artery (AAOCA) is a rare congenital cardiac condition with potentially lethal consequences. Variants of this condition accounts for the second leading cause of sudden cardiac death in children and adolescents. The incidence of detection of these anomalies is rising due to the improvement in imaging techniques. From our literature search, there are a few cases reported in the surgical journals, but none is about the anesthesia management. We report a case of AAOCA repair of a previously attempted and failed repair. In summary, anesthesia management of AAOCA depends on the type, anatomy, and pathophysiology of the anomaly. This patient’s previous repair with graft resulted in a competing flow between her native RCA and the right internal mammary artery graft. Understanding and close monitoring of this flow dynamic is the key to manage such patients.

Keywords: Anomalous coronary artery; Transesophageal echocardiography; Un-roof

Introduction

Anomalous Aortic Origin of a Coronary Artery (AAOCA) is a rare congenital cardiac condition with potentially lethal consequences. Variants of this condition accounts for the second leading cause of sudden cardiac death in children and adolescents. The incidence of detection of these anomalies is rising due to the improvement in imaging techniques. From our literature search, there are a few cases reported in the surgical journals, but none is about the anesthesia management. We report a case of AAOCA repair of a previously attempted and failed repair. The patient has provided written consent for this report.

Case Report

A 44-year-old woman (162 cm, 85 kg) with no significant coronary artery disease presented with chest and upper back pain. The intensity and frequency of symptom increased over the past four months. She had a known congenital anomalous right coronary artery origin from the sinus of the left coronary cusp. Due to the intermittent low blood flow in the RCA, Right Internal Mammary Artery (RIMA) to Right Coronary Artery (RCA) bypass was performed two years ago to relieve the symptoms. Sublingual aspirin relieved her symptoms. The patient followed up with her cardiologist where CT angiography demonstrated intertemp flow from the RIMA to RCA graft and a slit-like opening of the RCA ostia at the level of the sinus arising from the left coronary cusp. The patient was then referred to our facility for prompt surgical evaluation. The original surgery of RIMA-RCA bypass was not successful. This was due to the variable flow of native proximal RCA and RIMA-RCA anastomosis. When flow was greater in the native proximal RCA, the flow in RIMA-RCA graft was reduced or completely stopped. The intermittent flow in RIMA-RCA graft eventually caused the graft stenosis and the recurrence of symptoms in the patient.

A pre-induction left brachial arterial line was placed and anesthesia was induced with midazolam (2mg), fentanyl (0.5mg), propofol (100mg) and rocuronium (50mg). After intubation, anesthesia was maintained with isoflurane (1-1.5%) and sufentanil (0.1 mcg/kg/hr). A right internal jugular 9 French central catheter was placed for central venous access. Pre-incision transesophageal (TEE) examination showed normal biventricular systolic function and no regional wall motion abnormality. The altered ostium of the RCA, arising from the sinus of the left coronary cusp, was demonstrated in the mid-esophageal aortic valve short-axis (ME
AV SAX) view. Trace aortic insufficiency was also noted (Figure 1). The patient was closely monitored with TEE for RCA flow, regional wall motion abnormality and EKG for ST segment changes during the pre-bypass period due to the interventricular flow changes of RCA, and a right-dominant PDA. The patient’s aorta and RCA shared a segmental of wall. The ascending aortic flow impacted on the size of the RCA lumen and therefore the RCA flow. The patient’s hemodynamic was managed according to the relative flow between aorta and RCA. The surgeon decided to perform an unroofing of the right coronary ostium instead of re-implantation due to the risk of kinking the artery and impeding flow to the conus branch. The un-roofing procedure included excising the segment of common wall between the right coronary ostium and the aorta. The patient was separated from cardiopulmonary bypass, after 63 minutes clamp time, without difficulty. Color-flow Doppler post-repair demonstrated improved flow through the RCA ostium (Figure 2). Both Bi-ventricular systolic function and pre-existing aortic insufficiency remained unchanged. The patient was extubated in the operating room and taken to the cardiac surgery ICU. She was discharged from the ICU on postoperative day 2, and from the hospital on day 3.

**Discussion**

The incidence of altered coronary artery origins is very low in the general population, 6.4 per 1000 births [1]. The incidence includes separate aortic origins for the conus (RCA) or circumflex arteries, as well as main coronary artery abnormalities. The incidence of right or left main arteries arising from improper positioning or Anomalous Aortic Origin of a Coronary Artery (AAOCA) is even lower (around 1-3 per 1000 births). Interestingly, within that subset RCA arising from a left sinus position is around 3 times more common than the opposite situation [2]. Clinical presentation is extremely variable depending on the nature of vessel involvement, degree of resulting ischemia, patient’s physical activity level, and extent of ischemia-related cardiac changes (i.e. ischemia valvular disease, congestive heart failure, etc.). AAOCA is one of the most common autopsy findings in cases of sudden cardiac death [3].

The two main options for correction of these conditions are re-implantation of the artery elsewhere in the aorta and un-roofing of the aortic intramural section of the affected artery. The decision to undergo correction as well as the timing is dependent on the patient’s symptoms and associated factors such as exertional level and co-existing medical conditions. It is highly recommended that lesions involving the left coronary artery be corrected for the risk of fatal arrhythmias, while right-sided lesions may not warrant such an urgent repair [4]. Timing of repair is often during adolescence or even adulthood, as fatal ischemia during childhood is not common. The decision on appropriate repair often depends on the location of the coronary artery in relation to the structures of the aortic root. Re-implantation may be more prudent in cases where commissural integrity would be compromised by un-roofing of intramural vessels. However, re-implantation may not be suitable for short segments of exposed coronary artery, or the case where altering the course would compromise early takeoff branches. The use of bypass grafting is discouraged because of low long-term success rates secondary to competing flow with the native vessel and thrombosis/sclerosis of the arterial graft [3]. Use of internal mammary artery conduits for grafting also limits options for future grafting if the patient need additional surgery for coronary artery disease.

The anesthetic considerations for these cases revolve around optimizing the myocardial oxygen supply-demand relationship, and the utilization of intraoperative monitors to detect early ischemia. Avoidance of ischemia during the preoperative and induction period is crucial, because ischemia leading to arrhythmias may be fatal. Caution should also be given to the volume status, afterload, and contractility of the heart, as some of these lesions may exhibit shunt physiology with variable flows in different hemodynamic states. Given the tenuous nature of these lesions, pre-induction arterial lines should be placed in all patients presenting for surgery regardless of physical status or age. Central venous access is useful in providing vascular access for resuscitation and administration of inotropes, but the central venous pressure data may have limited use for optimizing these patients. Early post-induction placement of the TEE probe will allow the anesthesiologist to both assess volume status and make note of any pre-repair regional wall motion abnormalities (RWMAs). The aortic root can be accurately imaged from the mid-esophageal long-axis AV (ME AV LAX) view as well as the mid-esophageal short-axis AV (ME AV SAX) view. Valvular dysfunction, annular dilation, and abnormalities of the sinuses can be seen in the long-axis images. The coronary cusps and sinuses are easily visualized in the short-axis images. The ostia can often be seen and doppler assessment of flow through the first part of the coronaries can usually be performed. Post-repair TEE will be able to evaluate new or worsening RWMAs, as well as new or worsening valvular dysfunction. In patients with isolated anomalous

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**FIGURES**

Figure 1: Pre-repair RCA. Figure 2: Post-repair RCA.
coronary origin and only mild preoperative cardiac depression/dysfunction, these patients may be hyperdynamic after separation from cardiopulmonary bypass and may require vasodilators and beta-blockers to control hemodynamics.

In summary, anesthesia management of AAOCA depends on the type, anatomy, and pathophysiology of the anomaly. This patient’s previous repair with graft resulted in a competing flow between her native RCA and the right internal mammary artery graft. Understanding and close monitoring of this flow dynamic is the key to manage such patients.

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None.

Conflicts of Interesting

The authors deny any potential conflicts of interest including commercial relationships such as consultation and equity interests.

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