Case Report

Anaesthetic management of infants posted for repair of anomalous origin of left coronary artery from pulmonary artery

Address for correspondence:
Dr. Chitralekha Patra, Department of Cardiac Anaesthesiology, Sri Jayadeva Institute of Cardiovascular Sciences and Research, 9th Block, Jayanagar, Bengaluru - 560 069, Karnataka, India.
E-mail: Chitralekhapatra@gmail.com

Access this article online

Website: www.ijaweb.org

DOI: 10.4103/ija.IJA_212_17

Quick response code

Chitralekha Patra, Naveen G Singh, N Manjunatha, Anand Bhatt

Department of Cardiac Anaesthesiology, Sri Jayadeva Institute of Cardiovascular Sciences and Research, Bengaluru, Karnataka, India

ABSTRACT

First described in 1908, anomalous origin of left coronary artery from pulmonary artery is a very rare congenital anomaly. Here, the right coronary artery is usually enlarged and has a normal origin from aorta. Numerous collaterals connect the two coronary arteries over right ventricular outflow tract or interventricular septum. It is one of the most common causes of myocardial ischaemia and infarction in children.

Key words: Anomalous origin of left coronary artery from pulmonary artery, coronary steal, left ventricular assist device, myocardial ischemia

INTRODUCTION

Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) is a rare entity occurring 1 in 3,00,000. It is characterised by anomalous origin of the left main coronary artery (LMCA) from pulmonary artery (PA).[1,2] Blood supply to the myocardium is derived through collaterals arising from the right coronary artery (RCA) and exiting into the PA which is liable for coronary steal. Endocardial fibrosis and scarring may involve the papillary muscle of the mitral valve resulting in valvular insufficiency.[2,3] Anaesthetic management poses a unique challenge as ALCAPA patients have increased chances of perioperative myocardial ischaemia, cardiac arrest and sudden death. Here, we discuss the anaesthetic management and post-operative outcome of a series of three cases over 1 year in our institution.

CASE REPORTS

Case 1

A 6-month-old, 3 kg boy presented with resting tachypnoea and diaphoresis while feeding and failure to thrive. The heart rate (HR) was 160/min and blood pressure (BP) was 90/60 mmHg. No murmur was present. Electrocardiogram (ECG) showed T-wave inversion in lead II, III and AVF and chest radiograph showed significant cardiomegaly.

Transthoracic echocardiography revealed retrograde flow in LMCA draining into the PA. There was dilatation of left atrium and ventricle with Grade II mitral regurgitation (MR). There was severe left ventricular (LV) dysfunction with ejection fraction (EF) of 20%. Since the diagnosis was clear, cardiac catheterisation and angiography were not done. His medications included digoxin, spironolactone and ramipril.

How to cite this article: Patra C, Singh NG, Manjunatha N, Bhatt A. Anaesthetic management of infants posted for repair of anomalous origin of left coronary artery from pulmonary artery. Indian J Anaesth 2017;61:676-8.

The child was taken to operating room, where non -invasive monitors such as 5 lead ECG, Pulse oximetry and non-invasive BP (NIBP) were attached. BP was 90/50 mmHg, HR was 135/min and SpO_2 was 100%. There was no evidence of ischaemia on ECG. General anaesthesia was induced with intravenous (iv) injection ketamine (2 mg/kg), followed by iv injection fentanyl (2 μg/kg) and iv injection atracurium (0.5 mg/kg) and the trachea was intubated with 4.0 mm cuffed endotracheal tube (CETT). The femoral artery was cannulated for invasive arterial BP monitoring, and the right internal jugular vein was cannulated for central venous pressure monitoring. Anaesthesia was maintained with isoflurane and intermittent fentanyl.

The ventilatory settings were aimed to prevent hypocarbia, hyperoxia and alkalosis. An FiO_2 of 0.5 was targeted to maintain normoxia, with a PaO_2 80-100mmHg which was used to maintain pulmonary vascular resistance (PVR). End-tidal carbon dioxide was maintained between 40 and 45 mmHg. Re-implantation of the LMCA was performed under cardiopulmonary bypass (CPB). During weaning from CPB, dopamine (5 μg/kg/min) and milrinone (0.33 μg/kg/min) were started. Patient remained haemodynamically stable throughout the perioperative period. The child was ventilated for 24 h and discharged to ward on 5th post-operative day.

Case 2

An 8-month -old, 5.5 kg baby girl presented with a history of excessive crying and difficulty in breathing since 3 months. HR was 106/min, regular and BP was 80/46 mmHg. Transthoracic echocardiography revealed ALCAPA, reduced LV global function and EF 20%. She was on ramipril (5 mg/day) and furosemide (10 mg/day). The child was taken to OT, where non-invasive monitors such as ECG, NIBP and SpO_2 were attached. Anaesthesia was induced with iv injection etomidate (0.3 mg/kg), iv injection fentanyl (2 μg/kg) and iv injection atracurium (0.5 mg/kg) and intubated with CETT (3.5 mm). Femoral artery and vein were cannulated for invasive monitoring. Anaesthetic goals of maintaining a slightly higher PVR and higher $EtCO_2$ of 40–45 mmHg was done by preventing hypocarbia, hyperoxia and alkalosis. Relocation of LMCA was performed under CPB where bypass time was 106 min and cross-clamp time was 73 min. After rewarming was complete, inotropes such as dopamine 5 μg/kg/min and dobutamine 5 μg/kg/min were started. Sinus rhythm returned, and patient was haemodynamically stable throughout post-operative period. The patient was ventilated for 36 h and shifted to ward on 8th post-operative day. Transoesophageal echocardiogram (TOE) after 1 week revealed LMCA arising from aorta and slightly improved EF of 30%.

Case 3

A 1-month-old, 3.5 kg baby girl presented with breathlessness. Her HR was 150/min and BP was 64/35 mmHg. Auscultation revealed S1, S2 and pansystolic murmur. ECG revealed ST depression and T-wave inversion in leads aVL, I and II. TTE showed ALCAPA, severe LV dysfunction, severe MR and EF of 20%. In the OT, HR was 145/min, BP was 60/38 mmHg and ECG revealed T-wave inversion. Anaesthesia was induced with iv injection ketamine (2 mg/kg), injection fentanyl (2 μg/kg) and injection atracurium (0.5 mg/kg) and intubated with CETT (3.0 mm). Femoral artery and vein were cannulated. The anaesthetic goals of preventing hypocarbia, hyperoxia and alkalosis and maintaining the PVR were achieved by keeping FiO_2 of 0.5 to maintain PaO_2 between 80-100mmHg, and $EtCO_2$ of 40–45 mmHg. Re-implantation of LMCA was performed under CPB. After the patient was rewarmed completely, the heart did not beat for a long time, and pacing of the ventricle was commenced. Inotropic supports i.e. dopamine (5 μg/kg/min) and adrenaline (0.1 μg/kg/min) were started. The heart started to beat in sinus rhythm, however, with very poor contractility. An attempt was made to wean off CPB, but there was severe distension of the ventricles with low systemic pressures of around 45/30 mmHg. CPB was recommenced and later three more attempts were made to wean off CPB again but was unsuccessful due to severe myocardial dysfunction. The total CPB time was 300 min and cross-clamp time was 126 min. Finally, the patient was weaned off CPB with very high inotropic support of Dopamine 10 μg/kg/min, Noradrenaline 0.1 μg/kg/min and Milrinone 0.99 μg/kg/min. Patient was shifted to paediatric intensive care unit (PICU) with a BP of 50/34 mmHg and poor contractility of heart. Patient had unstable haemodynamics, severe metabolic acidosis with low cardiac output inspite of high inotropic support resulting in cardiac arrest after one hour of shifting to PICU.

DISCUSSION

There are unique anaesthetic considerations for patients presenting with ALCAPA repair. Of utmost importance is maintaining adequate coronary perfusion pressure through the single coronary artery and minimising myocardial oxygen consumption. Hence, laryngoscopy

and intubation should be smooth and rapid, and swings in BP and HR should be avoided as tachycardia can alter myocardial oxygen supply and demand and predispose to myocardial infarction (MI).[2,3]

Coronary blood flow in ALCAPA is dependent on pressure gradient between RCA and PA, and low PA pressures can worsen the steal phenomenon. Hence, factors decreasing PVR such as hyperventilation, hypocarbia or hyperoxia must be avoided. Patients are ventilated using oxygen and air mixture, with an aim to maintain normoxia targeting a PaO_2 of 80-100 mmHg. An $etCO_2$ of 40-45 mmHg is achieved to maintain PVR. An increase in afterload is prevented, to optimize the stroke volume.[3] However, aggressive reduction in afterload may be deleterious as it attenuates perfusion of the RCA, thus decreasing blood flow to LMCA. Inotropes such as dopamine, dobutamine and milrinone should be used cautiously as they increase myocardial oxygen consumption and increase the risk of MI.[3,4] Isoflurane is the inhalation agent of choice as it causes less myocardial depression.

In our case series, case 1 and 2 showed remarkable recovery post-bypass. A possible mechanism for the recovery of cardiac function after repair is myocyte hyperplasia in young infants. In addition, there may be compensatory hypertrophy of remaining viable myocytes if muscle necrosis occurs. Alternatively, phenomenon of hibernation may explain the complete myocardial recovery after revascularisation.[5] Hibernating myocardium specifically refers to the occurrence of persistent contractile dysfunction associated with chronic ischaemia but preserved myocardial viability. It has been suggested that the chronic myocardial hypoperfusion of ALCAPA leads to myocyte adaptation rather than diffuse infarction.[5,6] The finding of complete recovery of LV function in these patients without evidence of infarction supports this hypothesis. However, at times, the recovery may be prolonged or may not occur at all as in case 3 which represents a reversal of these adaptive cellular changes. Cardiac dysfunction may be acutely compounded by prolonged CPB time and post-operative myocardial stunning, the transient mechanical dysfunction that persists after reperfusion and restoration of normal coronary blood flow. Myocardial stunning may contribute to some patient dependence on left ventricular assist device (LVAD) in the immediate post-operative period. ALCAPA patients with severe LV dysfunction exacerbated with prolonged CPB time are ideal candidates for successful use of a LVAD when separation from bypass cannot be achieved.[5,6] The advantages of LVAD are its simplicity, better ventricular decompression, absence of oxygenator and low anticoagulation requirements. Alternatively, extracorporeal membrane oxygenator (ECMO) can be considered as a strategy for recovery of poor LV function in ALCAPA patients. Survival has been found to be 33% when ECMO was initiated.[7] However, it is associated with complications such as bleeding and intracerebral haemorrhage. Mild and moderate MR usually improves with post-operative recovery of LV function. However, it remains possible that patients with severe pre-operative MR, as in case 3 would have a better outcome if MV was repaired at the time of coronary revascularisation. Furthermore, better results could be expected with minimal bypass and cross-clamp time. Moreover, mechanical support of LV with a LVAD or ECMO could have helped for better management in the above case.

CONCLUSION

Survivors of perioperative period in ALCAPA repair have excellent prognosis for functional recovery of LV regardless of pre-operative state. Normalisation of LV function occurs once dual coronary circulation is restored, which may take as long as 2 years.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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