The management of a coronary cameral fistula: lessons learnt

Mokgwathi GT, MBChB, Registrar
Department of Anaesthesiology, University of Limpopo (Medunsa Campus)

Ogunbanjo GA, MBBS, FCFP(SA), MFamMed, FACRRM, FACTM, FAFP(SA), FWACP(Fam Med)
Department of Family Medicine and Primary Health Care, University of Limpopo (Medunsa Campus)

Correspondence to: Gaorutwe Mokgwathi, e-mail: gaorutwe@hotmail.com

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Abstract
Coronary arteriovenous fistula (CAVF) is a rare congenital anomaly. Literature on CAVF perioperative anaesthetic management is limited, but the consequences of mismanaging symptomatic CAVF are dire, with the potential risk of perioperative myocardial infarction, and even death. We present a 48-year-old male with symptomatic congenital coronary cameral fistulae, who underwent a successful surgical repair of the fistulae. This case study describes this very rare phenomenon, and its perioperative anaesthetic management, including the lessons learnt.

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Introduction
A coronary arteriovenous fistula (CAVF) is an abnormal communication between one of the coronary arteries and a cardiac chamber (coronary cameral fistula) or vessels, including the coronary sinus, pulmonary artery, or superior vena cava (coronary arteriovenous malformation).1-3 CAVF is seen in 0.002% of the general population, and visualised in nearly 0.25% of patients undergoing cardiac catheterisation.1-3 It may be congenital or acquired.1-3 The origin of CAVF can be any of the major coronary arteries. The majority arise from the right coronary artery and left anterior descending artery, while the circumflex artery is rarely involved.3-4 Over 90% of these fistulae drain into the venous structures of circulation (the right-sided chambers, pulmonary artery, coronary sinus and superior vena cava), but drainage into the left-sided chambers is less frequent.4 This case study describes this very rare phenomena, and its anaesthetic management.

Case report
A 48-year-old man with a body mass index of 31 kg/m² was admitted to Dr George Mukhari Hospital (Ga-Rankuwa Hospital), with ischaemic heart disease, with poor left ventricular function, type II diabetes mellitus, dyslipidaemia, and an aortocoronary fistula, detected by coronary angiography. He had a five-year history of chest pains with progressive, intermittent dyspnoea. The chest pains were described as retrosternal, sharp, and radiating to the left arm, neck and back. Reportedly, they increased with frequency over the years, despite medical therapy. The 12-lead electrocardiograph showed a sinus rhythm (70 beats/minute) with evidence of left ventricular hypertrophy, and T-wave inversion from V4-V6, but no S-T changes.

The cardiothoracic ratio measured on the posterior-anterior (PA) view of his chest X-ray was 65%, with radiological evidence of left ventricular hypertrophy. A repeat angiogram and transthoracic echocardiography at the same hospital confirmed the diagnosis of large proximal and distal coronary artery fistulae from the circumflex artery draining into the right ventricle, left anterior descending artery (LAD) steal syndrome, and reduced left ventricular function, with an ejection faction of 55%. Initially, the patient was managed in the cardiology ward on medical therapy, and later followed up at the medical outpatient clinic. Surgery for closure of the fistulae was planned months later when the patient was symptom-free. Preoperatively, on admission to the cardiothoracic ward, his blood pressure was 138/89 mmHg and the pulse was 61 beats/minute and regular. The first and second heart sounds were normal, and there were no murmurs.
In addition, examination of his respiratory system revealed a respiratory rate of 20 breaths/minute. The rest of his physical examination was unremarkable. He had angina-like chest pains with strenuous rapid or prolonged exertion, and he was classified as Canadian Cardiovascular Society functional classification of angina, Class I. Ordinary physical activity such as walking and climbing stairs did not cause angina. The premedications that were administered were an anxiolytic, hydroxyzine tablet 75 mg, continuation of anti-anginal therapy (aspirin 150 mg, atenolol 50 mg, perindopril 2 mg, and statin 20 mg tablets) for myocardial protection, while oral hypoglycaemic agents (metformin and gliclazide tablets) were stopped a day before surgery.

Prior to radial artery cannulation, the patient was premedicated with a 2 mg intravenous bolus of midazolam, and then pre-oxygenated with 100% oxygen. He was slowly induced over 10 minutes with fentanyl (5 µg/kg), etomidate (0.3 mg/kg) and vecuronium (0.02 mg/kg). Endotracheal intubation was performed with a 8-mm endotracheal tube. Anaesthesia was maintained using a continuous fentanyl infusion, at a rate of 0.1 µg/kg/hour, supplemented with midazolam 0.2 mg/kg, vecuronium 20 µg/kg, sevoflurane 2%, air and oxygen at the fraction of inspired oxygen 2 of 0.6. Intraoperative monitoring included electrocardiography, pulse oximetry, end-tidal carbon dioxide, a rectal temperature probe, a urinary catheter for fluid balance, intra-arterial blood pressure monitoring and central venous pressure. Pre-bypass systolic blood pressure (SBP) was kept between 100-130 mmHg, mean arterial pressures above 70 mmHg, and heat rate between 50-60 beats/minute by deepening the anaesthesia with sevoflurane and fentanyl boluses.

The surgical repair of the fistulae was carried out under cardiopulmonary bypass (CPB). CPB time (3 hours and 52 minutes) was prolonged because of the complicated nature of the fistulae with its multiple branching. Aortic cross-clamp time was 2 hours and 44 minutes. Duration of anaesthesia and surgery was 7 hours and 35 minutes in total. Perfusion flow rates ranged between 3.5-6 l/min/minute. Rectal temperature was 32.5-35.8°C.

Intraoperatively, three units of packed red blood cells were transfused. As a rescue plan, the following drugs were prepared: anti-ischaemic, nitroglycerin, beta blockers (labetalol), inotropes (dobutamine and adrenaline) and vasoconstrictors (phenylephrine). Inotropic support with dobutamine and adrenaline infusions running at 5 and 0.2 µg/kg/minute, respectively, were administered after weaning from CPB and continued postoperatively in the intensive care unit. The patient’s trachea was extubated after six hours in the intensive care unit after which he received oxygen by face mask and anti-anginal therapy (aspirin 150 mg, atenolol 50 mg, perindopril 2 mg and statin 20 mg). Postoperative recovery was uneventful and the patient was subsequently discharged after nine days.

**Discussion**

CAVF is a very rare anomaly which was first described by Krause in 1865 and the first surgical treatment was carried out by Bjork and Crafoord in 1947. The pathophysiological mechanisms of fistulae depend principally on the resistance of fistulous connection and on the site of fistula termination. Blood follows the lower resistance pathway though the fistula rather than traversing the smaller arterioles and capillaries of the myocardium. With larger fistulae, a “diastolic run-off” may occur, drawing blood away from the normal coronary pathway, with widened pulse pressure and coronary “steal” phenomena. The myocardial stealing or reduction in coronary blood flow distal to the site of connection causes angina. It has also been proposed that a congenital coronary artery fistula proximal to a segment of acquired atherosclerotic stenosis aggravates the distal perfusion deficit by acting as a low resistance alternative to the zone of coronary artery stenosis. The patient was at risk of perioperative myocardial ischaemia because he had underlying atherosclerosis (diabetes mellitus was confirmed both clinically and serologically, and dyslipidaemia was confirmed serologically), and angiographic LAD steal syndrome which explained the recurrent angina. Surprisingly, this patient did not have a continuous murmur in the praecordium, as often described in the literature. Untreated CAVF can result in several complications, namely congestive cardiac failure, infective endocarditis, pulmonary hypertension, aneuysmal dilatation and death. Management of CAVF is controversial and often depends on the experience of a medical centre. Valid options described in the literature include primary surgical treatment and trans-catheter closure with coil embolisation. The former is advocated in the case of asymptomatic (angina and infarction) or haemodynamically significant CAVF. The median sternotomy approach is the preferred surgical technique in cases of multiple branching and laterally localised fistulae. The decision to carry out a surgical procedure by bypass was based on the significant history of angina and atherosclerosis, multiple branching and laterally localised fistulae.

The anaesthetic management of these patients focuses on the prevention of coronary steal and perioperative myocardial ischaemia. Keeping in mind the determinants of myocardial oxygen balance, it was important to attenuate both the sympathetic and neuroendocrine responses to...
surgical stress that could increase the myocardial demand, while simultaneously optimising the myocardial oxygen supply. The patient was adequately pre-medicated with hydroxyzine and midazolam, and subsequently given oxygen therapy prior to induction to avoid hypoxaemia and respiratory depression that are associated with benzodiazepines. During induction, the goals were to avoid hypertension, tachycardia, hypotension and excessive myocardial depression, and to provide smooth intubation. Hence, the rescue plan was to include drugs that would manipulate the haemodynamics in the above manner. Clinical evidence has shown that inhaled anaesthetics, such as sevoflurane, have favourable properties of myocardial protection and preconditioning. Therefore, they should be included in an anaesthetic technique.6 Intraoperative monitoring was conducted according to standard American Society of Anesthesiologists monitoring guidelines, especially the pulse oximetry, intra-arterial blood pressure and electrocardiogram (lead II and V5), which is 80% sensitive in detecting myocardial ischaemia.6 The trans-oesophageal echocardiogram (TEE) would have been ideal in this patient, but was not available in our institution. TEE is able to detect the earliest signs of myocardial ischaemia in a few seconds.6 Postoperative continuation of the intraoperative management is crucial to maintain a balance between myocardial oxygen supply and demand.

Conclusion

Symptomatic CAVF is a rare phenomenon that can present with angina due to coronary steal phenomena. Therefore, perioperative anaesthetic management focuses on avoiding myocardial ischaemia. If this rare condition is not managed properly, perioperative myocardial infarction, and even death, can occur.

Conflict of interest

We declare that we have no financial or personal relationships that may have inappropriately influenced the writing of this paper.

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