J Minim Invasive Surg Sci. 2014 February; 3(1):e7834.

Published online 2013 December 30. **Case Report**

Perioperative Management of Bariatric Surgery in a Patient with Transposition of the Great Arteries Following an Atrial Level Switch

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Received: August 20, 2012; **Revised:** October 17, 2012; **Accepted:** November 13, 2012

Introduction: Patients with palliated congenital heart disease may present later in life for major, non-cardiac surgical procedures. Although alternative surgical techniques are now available for the treatment of the transposition of the great arteries (TGA), an atrial level baffle (Mustard or Senning procedure) was previously performed. Long-term adverse effects of the atrial level switch include myocardial dysfunction, rhythm disturbances requiring pacemaker placement, pulmonary hypertension and baffle obstruction.
Case report: We present a 23-year-old woman, who had undergone an atrial level switch during infancy for palliation of TGA and now presents for laparoscopic vertical sleeve gastrectomy for the treatment of obesity.
Conclusions: The long term end-organ effects of an atrial level switch for TGA are reviewed and the perioperative of these patients is discussed.

Keywords: Transposition of Great Arteries, Congenital Heart Disease, Bariatric Surgery

1. Introduction

With improvements in surgical techniques, cardiopulmonary bypass (CPB), and perioperative care, survival into the teenage years is common for patients with congenital heart disease (CHD). We present a patient with transposition of the great arteries (D-TGA), who had undergone an atrial level switch or Mustard procedure during infancy. She subsequently presented for laparoscopic vertical sleeve gastrectomy for the treatment of obesity. The long term end-organ effects of an atrial level switch for TGA is presented and the perioperative care of these patients is discussed.

2. Case Report

Institutional Review Board approval for single case reports is not required at Nationwide Children's Hospital (Columbus, Ohio). The patient was a 23-year-old, 113 kilogram (body mass index = 48 kg/M^2) woman presenting for a laparoscopic vertical sleeve gastrectomy for the treatment of severe obesity. Her past medical history was significant for D-TGA with intact ventricular septum. She had undergone a balloon atrial septostomy and Mustard procedure during the first year of her life. Additional co-morbid diseases included atrial fibrillation, 3rd degree AV block requiring pacemaker placement, transient ischemic attacks requiring chronic coumadin therapy, hypertriglyceridemia, and superior vena caval stenosis with baffle leak, which had required placement of a covered vena caval stent. Her current medications included sotalol 80 mg twice daily, sertraline 100 mg daily, fenofibrate 158 mg daily, and warfarin 6 mg daily. Vital signs revealed a regular paced rhythm with a heart rate of 70 beats/minute, blood pressure (BP) 108/40 mmHg, respiratory rate of 16 breaths/minute, and room air oxygen saturation of 100%. Cardiac examination was unremarkable with the exception of an II/VI systolic ejection murmur. Transthoracic echocardiography revealed D-TGA following Mustard repair. The right ventricle was mildly dilated and hypertrophied with an estimated ejection fraction of 39%. There was mild pulmonary and tricuspid regurgitation with trace mitral regurgitation. The superior vena caval and inferior vena caval limbs were widely patent. A covered stent was in place from the superior vena cava toward the mitral valve and no significant gradient was noted in the pulmonary venous baffle. A pacemaker catheter was seen in the superior limb with the ventricular lead crossing the mitral valve into the left ventricle. Preoperative electrocardiography demonstrated the presence of atrial pacemaker spikes with intact atrio-

Implication for health policy/practice/research/medical education:
Perioperative concerns for patients with palliated transposition of the great arteries may be related to the long term consequences of the atrial level switch, the end-organ effects of insufflation during a laparoscopic procedure, as well as other co-morbidities of obesity.

ventricular (AV) conduction and right ventricular hypertrophy. Recent pacemaker evaluation indicated that the patient was paced approximately 99% of the time with her baseline rhythm being atrial pacing with intact AV conduction. The baseline pacemaker settings were AAIR mode with a lower rate limit of 60 beats/minute and an upper rate limit of 150 beats/minute. Prior to her arrival in the operating room, the patient's pacemaker settings were changed to the AOO mode with a pacing rate of 70 beats/min. The patient was admitted to the hospital the day before surgery, a cardiology consult was obtained at the time of admission and she was started on a heparin infusion. Warfarin therapy had been discontinued five days prior to surgery while the heparin infusion was discontinued 6 hours prior to surgery. On the morning of surgery, with the exception of sotalol 80 mg, all of the patient's routine medications were held and the patient was kept nil per os for 8 hours. Preoperative laboratory evaluation revealed a negative urine HCG, hemoglobin of 10.9 gm/dL, hematocrit of 32.6%, and platelet count of 182,000/mm3. The prothrombin time was 18.5 seconds, the INR was 1.55, and the partial thromboplastin time was 28 seconds. Premedication included midazolam 2 mg intravenously via a pre-existing 20 gauge intravenous cannula in the right forearm. The patient was then transported to the operating room where standard American Society of Anesthesiologists' monitors were placed. Additionally, cerebral oxygenation was measured using near infrared spectroscopy. Preoxygenation with 100% oxygen was provided via a tight-fitting anesthesia face mask. Intravenous induction included lidocaine 60 mg, fentanyl 100 μg and propofol 200 mg followed by the application of cricoid pressure. Tracheal intubation was facilitated by the administration of rocuronium (100 mg). Following cessation of spontaneous respirations, mask ventilation with sevoflurane mixed with 100% oxygen was provided while cricoid pressure was held. Following uncomplicated intubation of the trachea, a left radial arterial line, and a second peripheral intravenous cannula were placed. Using ultrasound guidance, a central venous catheter was placed via the right internal jugular vein. Maintenance anesthesia included desflurane (expired concentration 5.1-6.1%) and fentanyl (250 μg). An additional intravenous dose of rocuronium (20 mg) was administered for maintenance of neuromuscular blockade. Clindamycin (600 mg) was administered for antibiotic prophylaxis. The patient was placed supine with bilateral upper extremities padded and abducted less than 90°. Throughout the case, appropriate access to the patient's pacemaker module was ensured, and a magnet was available for use. Insufflation pressures were maintained at 10-12 mmHg. The duration of the case was 2.5 hours. Mean arterial pressure (MAP) was maintained between 65-100 mmHg with a baseline of 65 mmHg, pulse oximetry was maintained between 98-100% with an inspired oxygen concentration of 50%, and cerebral oximetry was maintained at greater than 75% throughout the case. Intraoperative blood loss was approximately 25 mL. A total of 1960 mL of lactated Ringers solution was administered. Urine output during the case was 450 mL. Dexamethasone (10 mg) and ondansetron (4 mg) were administered for postoperative nausea and vomiting prophylaxis. Morphine sulfate (2 mg) and hydromorphone (1 mg) were administered to treat surgical pain. Upon completion of the surgical procedure, desflurane was discontinued, the presence of twitches in response to train-of-four stimulation was confirmed and residual neuromuscular blockade reversed with glycopyrrolate (0.8 mg) and neostigmine (5 mg). After the resumption of spontaneous ventilation, demonstration of adequate strength and spontaneous eye opening, the patient's trachea was extubated. She was then transferred to the post-anesthesia care unit (PACU) with supplemental oxygen via nasal cannula at 3 liters per minute. Postoperative analgesia was provided by patient-controlled analgesia with hydromorphone. Following discharge from the PACU, the patient was transferred to the cardiothoracic intensive care unit (CTICU) for further monitoring. The patient's pacemaker settings were returned to baseline with a set heart rate of 60 beats/minute after the arrival to the CTICU. The heparin drip was resumed on postoperative day 0 and her routine dose of sotalol was continued. Warfarin was restarted on postoperative day 2. She was transitioned to oral analgesics (oxycodone) on postoperative day 4 and discharged home on postoperative day 7 after warfarin therapy was therapeutic with an INR greater than 1.8. The remainder of her postoperative course was unremarkable.

3. Discussion

In transposition of the great arteries, the aorta originates from the right ventricle and the pulmonary trunk arises from the left ventricle (1). Anatomically, D-TGA consists of a morphological right atrium that is connected to a morphological right ventricle emptying into the aorta. The morphological left atrium connects to the left ventricle that empties into the pulmonary artery. This anatomy results in the presence of two parallel circulations as compared to the physiologically normal circulation that exists in series. Following birth, survival is dependent on pathways that allow for the intracardiac mixing of blood including an atrial septal defect (ASD), a patent foramen ovale (PFO), or a ventricular septal defect (VSD). Other than atrial septostomy, the first palliative approach for D-TGA was the Senning operation in 1959. In 1964, Mustard proposed a new procedure that excised the atrial septum and used synthetic material to create the atrial baffle (2) with improved long-term survival of these patients, sequelae of atrial-level repairs including atrial arrhythmias, baffle leaks and obstructions, and right ventricular dysfunction became increasingly evident. Concerns re-

garding the perioperative care of complex surgical procedures in patients with co-morbid conditions may arise related to the patient's co-morbid CHD, other co-morbid conditions unrelated to the CHD, the impact of the surgical procedure itself on physiologic function or more commonly, the interaction of these factors. In our patient, there were concerns related both to the long term consequences of the atrial level switch, the end-organ effects of insufflation during a laparoscopic procedure (see below), as well as her other co-morbid condition (obesity) (3, 4). The long term cardiovascular complications and sequelae following an atrial level switch (Mustard or Senning procedure) are listed in Table 1. Fewer than half of patients remain in sinus rhythm 15-20 years post-surgery due to sinus node dysfunction and there is a high percentage of late deaths related to the abrupt onset of polymorphic ventricular tachycardia/fibrillation (5). The management of sinus node dysfunction and arrhythmias frequently requires radiofrequency ablation, pacemaker placement, or insertion of an implantable cardioverter-defibrillotor (ICD) (6). Right ventricular dysfunction (systemic ventricle) and the development of pulmonary hypertension are two other late complications following an atrial level repair that can significantly impact perioperative care. Clinical signs related to pulmonary hypertension may not appear until adult years, as the LV can effectively function against a high-resistance system. Given the significant perioperative risk imposed by pulmonary hypertension, its identification is key to successful perioperative management (7). Additional late sequelae may be related to the baffle including leaks or stenosis. Up to 15% of patients will develop stenosis with obstruction of the systemic venous baffle. The superior vena cava (SVC) is more frequently affected by obstruction than the inferior vena cava (IVC). Obstruction of the SVC generally develops gradually, allowing for the development of venous collaterals. IVC baffle obstruction results in the acute onset of symptoms from elevated venous pressures with hepatic congestion, hepatomegaly and ascites. A small intra-atrial baffle leak is more common than obstruction and although asymptomatic, it functions as a left-to-right shunt with the risk for paradoxical embolus. The latter may be of concern during insufflation for a laparoscopic procedure as was performed in our patient (see below). Additional perioperative management and evaluation is required in patients with a pacemaker or ICD to address and prevent potential device-related adverse events (8, 9). With these devices, electromagnetic interference (EMI) is the most commonly encountered problem intraoperatively. Current perioperative guidelines on cardiac rhythm management devices have been extensively reviewed elsewhere and should be reviewed prior to the perioperative care of these patients (8 , 9). In our patient, after a discussion with the pediatric cardiology service, the pacemaker was left in the VVI mode during the case. Since the patient was pacemaker dependent less than 5% of the time, there was little concern that EMI interference would result in interruption of pace maker function leading to bradycardia and decreased cardiac output. After patient positioning, we ensured easy access to the pacemaker and had a magnet in the room in the event that its use became necessary. Perhaps the most important aspect following an atrial level repair for D-TGA is an assessment of the function of the systemic ventricle (RV). As the RV serves as the pumping chamber for the systemic circulation, progressive RV dysfunction is commonly seen during the 2nd and 3rd decades of life. This concern was magnified in our patient as the mere presence of obesity, even in the absence of CHD, may compromise myocardial function (3, 10). Transthoracic or when necessary transesophageal echocardiography plays a primary role in the assessment of the right ventricular structure and function. Echocardiography should be used to evaluate myocardial contractility, tricuspid valve competency, the presence of an atrial baffle leak or obstruction, as well as to estimate pulmonary artery pressure (11). Further evaluation of RV function may require the use of cardiopulmonary exercise testing, dobutamine stress echocardiography, or cardiac MRI and adenosine stress imaging. Depressed myocardial function may be further compromised by the increase in systemic vascular resistance that is induced by abdominal insufflation (12). Given these concerns, in our patient, insufflation pressure was closely monitored and limited while near infrared spectroscopy was used to monitor cerebral oxygenation as a measure of end-organ oxygen delivery. The presence of pulmonary hypertension is associated with a significant risk of perioperative cardiac complications including cardiac arrest, pulmonary hypertensive crisis and death. Intraoperatively, hypercarbia, alveolar hypoxia, systemic hypoxemia, metabolic acidosis, and noxious stimuli can trigger a pulmonary hypertensive (PH) crisis. Even in the absence of these physiologic and biochemical changes, an increase in pulmonary artery pressure may occur following abdominal insufflation (12) Ventilation strategies to minimize pulmonary hypertensive triggers and pulmonary artery pressure include the use of high FiO_2 , low tidal volume ventilation (6 mL/kg predicted body weight), mild hypocarbia by increasing the respiratory rate, and an optimal level of positive end-expiratory pressure (5-10 cm H_2O) to maintain normal lung volume, reduce the degree of atelectasis and maintain functional residual capacity. Moderate hyperventilation with 100% oxygen, volume resuscitation as needed, and the initiation of inhaled nitric oxide (iNO) should be considered as the first tier of therapy for an intraoperative pulmonary hypertension crisis and RV failure (13, 14). In the presence of preoperative ventricular dysfunction or PH, the preoperative administration of milrinone should be considered to optimize ventricular performance and control

PVR. The volatile anesthetic agents and β-adrenergic antagonists may have significant negative inotropic effects especially in patients with pre-existing myocardial dysfunction. Perioperative central venous pressure (CVP) monitoring or TEE may be useful to guide volume management in patients with compromised myocardial contractility. Additional concerns in patients with palliated CHD include their chronic medication regimen. Intraoperative hemodynamic management may be affected by medications that act on the renin-angiotensin-aldosterone system (angiotensin-converting enzyme inhibitors and angiotensin-receptor blockers) and chronic anticoagulation therapy. The agents may lead to clinically significant hypotension that is recalcitrant to usual therapies including adrenergic agonists such as phenylephrine, requiring the administration of vasopressin (15) given these concerns; these agents should be held on the morning of surgery. Additionally, the perioperative conduct of anticoagulation must be considered in patients such as ours who are receiving warfarin therapy (16). In our patient, perioperative anticoagulation therapy was bridged using intravenous heparin. In summary, we present the perioperative considerations of a patient with D-TGA following an atrial level (Mustard) switch during infancy. Perioperative concerns include those related to the primary cardiac lesion (D-TGA), those related to co-morbid conditions (obesity), and those related to the end-organ physiologic effects of the surgical procedure (laparoscopy). Cardiac and hemodynamic concerns include the presence of RV (systemic ventricle) dysfunction, baffle leak or obstruction, pulmonary hypertension, arrhythmias, and sinus node dysfunction. Preoperative evaluation generally includes echocardiography and consultation with cardiology to evaluate pacemaker and ICD function. The impact of anesthetic agents on myocardial function, sinus node function, and PVR should be considered. Additional concerns include those related to the cardiovascular and respiratory physiologic changes induced by laparoscopy and insufflation. Consideration of these factors should help in the safe perioperative care of such patients.

Table 1. Long Term Cardiovascular Sequelae of an Atrial Level Switch

1	**Sinus node dysfunction**
1.1	Heart block
1.2	Atrial and ventricular arrhythmias
1.3	Sudden death
2	Right ventricular (systemic ventricle) dysfunction and failure
3	Atrioventricular (systemic) valve regurgitation
4	Pulmonary hypertension
5	Atrial baffle leak or obstruction

Acknowledgements

None declared.

Authors' Contribution

All of the authors participated in the preparation of the manuscript.

Financial Disclosure

There are no financial disclosures.

Funding Support

There was no external funding or support for the preparation of this manuscript.

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