Laparoscopic Robotic Radical Hysterectomy in an Adult with Congenital Heart Disease: A Case Report and Review of the Literature

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Abstract: *Purpose*: Non-cardiac surgeries in adults with congenital heart disease are increasing due to improved survival of children with congenital heart disease. There has also been an increase in laparoscopic procedures due to benefits including decreased length of stay and improved quality of life. There have been concerns regarding laparoscopic surgery in patients with congenital heart disease especially with pneumoperitoneum and positioning related to hemodynamic effects. We present a case of laparoscopic surgery in an adult with congenital heart disease who underwent robotic radical hysterectomy and provide a review of the literature.

Methods/Results: The case of a 39 year old female with palliated congenital heart disease who underwent laparoscopic robotic radical hysterectomy for cervical cancer was summarized. A review of the literature was performed. Ten surgeries in 9 adult patients were reported with good outcomes. Insufflation pressures were reported in 8 of 10 surgeries and kept at or below 15 mmHg.

Conclusions: When performed in centers with experience in CHD, non-cardiac laparoscopic surgery has been shown to be safe and effective. CO_2 abdominal insufflation pressures should be reduced as much as possible without compromising visualization, in order to decrease potential negative effects on hemodynamics.

Keywords: Congenital heart defect, adult congenital heart disease, laparoscopy, gynecologic oncology, radical robotic hysterectomy.

INTRODUCTION

Improvements in medical and surgical management of patients born with congenital heart disease (CHD) have resulted in increased survival into adulthood. Adults with CHD may require non-cardiac surgery as longevity improves. Patients with adult congenital heart disease (ACHD) are at increased risk during surgery [1]. Laparoscopic surgeries have become more common due to decreased length of stay and improved post-operative pain management. In gynecologic oncology, the laparoscopic approach has been shown to result in improved quality of life and decreased length of stay [2]. Laparoscopic and robotic radical hysterectomies (LRH and RRH) have been shown to decrease blood loss, morbidity from infection, and length of stay as compared to traditional abdominal radical hysterectomy (ARH), and is considered standard of care in patients without CHD [3].

Historically, there have been concerns about laparoscopic surgery in patients with congenital heart disease (CHD), especially patients with cyanotic lesions and single ventricle physiology. A major concern involves the effects of pneumoperitoneum and body positioning on hemodynamics [4-6]. We present a case of an adult with complex CHD who had robotic radical hysterectomy for cervical adenocarcinoma and review of the literature on this topic.

CASE SUMMARY

A 39 year old female with a history of pulmonary valve atresia and ventricular septal defect presented for a robotic radical hysterectomy, bilateral salpingectomy, and sentinel lymph node biopsy due to Stage IB1 adenocarcinoma of the cervix. At a young age, she had bilateral Blalock-Taussig (BT) shunts placed. At age 6, her right BT shunt was taken down and a Waterston shunt was placed from the ascending aorta to the pulmonary artery. She also had revision of her left BT shunt at age 10. At age 28, she had a laparoscopic cholecystectomy performed for cholecystitis with cholelithiasis, which she tolerated well. Her post-operative course was complicated by hypoxia and fluid overload which were managed with oxygen and diuretics. She had one previous episode of atrial flutter at age 37 which was treated with successful cardioversion.

During pre-operative cardiology evaluation an electrocardiogram (ECG) showed 2:1 atrial flutter. An

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echocardiogram demonstrated an ejection fraction (EF) of 53%. She was admitted two days prior to the procedure for management of her atrial flutter. A diltiazem drip was started and her metoprolol was increased for rate control. She received a heparin infusion for anticoagulation.

General anesthesia was administered with propofol, fentanyl, lidocaine, and vecuronium in preparation for surgery, a transesophageal echocardiogram (TEE) was performed which demonstrated an EF of 55 to 60% and atrial flutter rhythm with no intracardiac thrombus. The patient was placed in a steep Trendelenburg position for surgery. An insufflation pressure of 12 mmHg was used due to her underlying congenital heart disease to try to avoid hemodynamic complications. A robotic radical hysterectomy and salpingectomy was performed. The ovaries were spared and secured to the pelvic side walls. A sentinel node biopsy was performed bilaterally using indocyanine green. Phenylephrine 50mcg was used at 50 minutes into the procedure due to hypotension (85/39 mmHg) which resolved afterwards. The patient tolerated the procedure. Her post-operative stay was prolonged by several days due to complications from bleeding due to the anticoagulation post-cardioversion. Heparin was discontinued and bleeding resolved.

DISCUSSION

Patients with CHD can present many anesthetic management challenges depending on the complexity of their cardiac lesion. Dysrhythmias are problematic, especially in patients with poor ventricular function or a history of ventriculotomy or atrial surgery [7, 8]. Increased risk of bleeding or thrombosis in cyanotic patients are secondary to abnormalities in the coagulation cascade and erythrocytosis with microcytosis, respectively [8]. Balancing pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR) is extremely important and can be difficult, especially in patients with intracardiac or systemic-to-pulmonary artery shunts [8, 9].

Laparoscopic surgery has been an area for particular concern in patients with CHD. Studies have shown, in adults without cardiac disease, pneumoperitoneum with CO_2 insufflation can cause increased mean arterial pressure (MAP) and systemic vascular resistance (SVR) as well as decreased cardiac index (CI) [10-13]. End-diastolic area (EDA) and pulmonary capillary wedge pressure (PCWP) also increase with pneumoperitoneum [14]. Trendelenburg positioning may contribute to some of these effects including increased mean pulmonary arterial pressure (MPAP) and central venous pressure (CVP) [11, 13-15]. Both pneumoperitoneum and positioning result in increased levels of aldosterone and arginine [16]. Increased intra-abdominal pressure secondary to pneumoperitoneum can also decrease venous return. Patients with complex CHD may be more sensitive to changes in PVR and SVR and have historically created concern for laparoscopic approach.

There have been multiple reports of laparoscopic surgeries in patients with CHD. In children, laparoscopic surgeries have been shown to be safe [17-20]. The most common laparoscopic surgeries are gastrostomy tube placement, Nissen fundoplication, and pyloromyotomy. The CHD lesions reported in these patients varied from simple patent ductus arteriosus (PDA) to complex single ventricle physiology such as hypoplastic left heart syndrome (HLHS). Cribbs *et al*. reported 104 laparoscopic fundoplications with one death [21]. Thirty-seven of these patients had single ventricle physiology.

There are fewer reports of cases of laparoscopic surgery in adults with CHD. Laparoscopic cholecystectomy has been well reported. Andrews reported successful laparoscopic cholecystectomy in a patient with unrepaired tetralogy of Fallot (TOF) [22]. Hemodynamics remained relatively unchanged after insufflation and the patient recovered well. Another cholecystectomy in a patient with Fontan physiology was managed with low ventilator pressures and continuous infusion of hetastarch for mild hypertension [23]. McClain *et al*. reported both a laparoscopic cholecystectomy and a laparoscopic cauterization of endometriosis and lysis of adhesions three years later in a patient with an unfenestrated Fontan physiology with dextrocardia, heterotaxy, pulmonary valve, and mitral valve atresia [24]. The patient tolerated both procedures very well. Intra-abdominal pressures were kept below 10 cm H_2O. A successful laparoscopic cholecystectomy in a patient with hypoplastic right ventricle and fenestrated Fontan was reported [25]. Zach *et al*. described a colectomy in a patient with ulcerative colitis and single ventricle physiology with pulmonary and tricuspid valve atresia and only a BT shunt [26]. Hypotension did develop after pneumoperitoneum, but it was managed successfully with vasopressin infusion.

Laparoscopic surgery in patients with Eisenmenger syndrome has also been described. It is important to

maintain systemic vascular resistance (SVR) as decreases in this can cause right-to-left shunting due to increased right sided pressures resulting in desaturation and cyanosis. Kopka *et al*. described two cases of women, ages 42 and 39 years, who had laparoscopic cholecystectomy [27]. Both had a VSD with bidirectional shunting and elevated pulmonary artery blood pressures. The first patient tolerated anesthesia and pneumoperitoneum well. Intra-abdominal pressure was kept below 9 mmHg and head was kept at 20 degree head-up tilt. The second patient had decreased blood pressure after insufflation which was controlled successfully with metariminole. Her intra-abdominal pressure was kept below 12 mmHg and she also was tilted at 20 degrees head-up. Successful laparoscopic cholecystectomy was reported in a 46 year old female with Eisenmenger complex. SVR was maintained with a norepinephrine infusion [28]. Removal of a pheochromocytoma has also been reported [29].

Laparoscopic assisted hysterectomy in a patient with complex CHD has not been previously reported to our knowledge. Hirvonen reported hemodynamic changes in women who underwent laparoscopic hysterectomy [4]. CO_2 insufflation was shown to result in a significant increase in systemic MAP, MPAP, CVP, and PCWP at the beginning of the procedure. Over time, all of these parameters decreased. Trendelenburg positioning resulted in increased MPAP, CVP, and PCWP.

According to ACC/AHA 2008 guidelines for managing adult congenital heart disease (ACHD), it is recommended that patients with ACHD undergo perioperative evaluation at a regional center that specializes in congenital cardiology and has experienced surgeons and cardiac anesthesiologists [30]. This is in agreement with the more recent 2014 ESC/ESA guidelines on non-cardiac surgery [31]. Neither guideline statement has recommendations specific to laparoscopic procedures, although the ESC/ESA guidelines do outline negative effects of pneumoperitoneum and Trendelenburg positioning in laparoscopic surgery and increased cardiac risk in patients with heart failure [31]. Continuous ECG monitoring is recommended in patients with CHD undergoing non-cardiac surgery, and perioperative use of TEE should be considered in patients who are deemed high risk for ischemia or who develop ST-segment changes [31].

PLAN FOR MANAGEMENT

Adults with CHD undergoing non-cardiac laparoscopic surgery should have a preoperative evaluation performed at a center that is experienced with congenital heart disease involving cardiologists, surgeons, and cardiac anesthesia. Continuous monitoring of hemodynamics during the operation is critical, especially with abdominal insufflation and changes in positioning. It is important to maintain a balance of SVR and PVR to maintain adequate hemoglobin oxygenation and systemic perfusion. Insufflation pressures should be decreased while still maintaining adequate visualization to decrease negative hemodynamic effects. Trendelenburg positioning greater than 20 degrees should also be avoided if feasible. Continuous ECG is recommended, and it may be helpful to perform TEE if ST-segment changes are elevated or the patient is deemed high risk for ischemia.

CONCLUSION

Adults with CHD are at increased risk during non-cardiac surgery. There are multiple case reports of laparoscopic surgeries performed in patients with complex anatomy and physiology with good outcomes. Large studies in children have shown very low mortality as a result of laparoscopic surgeries. Data from large groups of adults with CHD undergoing laparoscopic surgery are lacking. When performed in centers with experience in CHD, non-cardiac laparoscopic surgery has been shown to be safe and effective. CO_2 abdominal insufflation pressures should be reduced as much as possible without compromising visualization, in order to decrease potential negative effects on hemodynamics.

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