Case Report

Laparoscopic colectomy in an adult with single ventricle physiology: Anesthetic implications and management

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ABSTRACT

Increasing numbers of adult patients with complex congenital heart conditions are presenting for noncardiac surgery later in life. These disorders can present challenges for surgical and anesthesia providers. Specifically, single ventricle lesions offer anatomic and physiologic concerns during the perioperative period. Single ventricle physiology represents a delicate balance between systemic and pulmonary blood flow. Any alterations in blood flow through these systems can produce undesirable hemodynamic changes, especially during the perioperative period. We present a case of an adult patient with a single left ventricle who presented for laparoscopic total colectomy due to inflammatory bowel disease. His abnormal anatomy coupled with the hemodynamic disruptions caused by laparoscopy presented significant anesthetic challenges. We highlight the anesthetic concerns of single ventricle physiology, specifically pertaining to laparoscopic surgery. We provide recommendations for safely managing these patients perioperatively. With detailed preoperative evaluation and close hemodynamic monitoring during the perioperative period, these patients can experience successful surgical and anesthetic outcomes.

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INTRODUCTION

Advances in the medical and surgical management of patients with adult congenital heart disease (CHD) have improved survival and longevity in this patient population. In 2010, the percentage of adult patients with CHD surpassed that of the pediatric patient population with CHD.[1] Expertise in the surgical correction of single ventricle lesions has led to a growing number of adults with these defects. For example, 70% of infants born with hypoplastic left heart syndrome are now expected to survive to adulthood.[2] As a survival improves, more of these patients are presenting for noncardiac surgeries later in life. The complex anatomy and physiology of these lesions presents challenges for both the surgical team and the anesthesiologist. A clear understanding of each patient's anatomy is imperative for quality perioperative care.

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Single ventricle physiology occurs due to a variety of congenital conditions that result in a hypoplastic or absent right or left ventricle (LV). This results in inadequate perfusion as the single functioning ventricle pumps blood to both the pulmonary and systemic vascular beds. These disorders are treated with either heart transplantation or staged palliative reconstruction.[3] The goal of the staged procedures is to convert this parallel circulation to serial circulation whereby the single ventricle directs blood systemically, and pulmonary flow relies on passive venous return.[4,5] The initial stage is performed in the neonatal period to connect the aorta to the single ventricle in

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the Norwood procedure. Additionally, unobstructed venous return from the systemic and pulmonary systems to the ventricle is fashioned by an inter-atrial communication. Pulmonary blood flow is facilitated by the creation of a systemic to pulmonary shunt. This is usually created between the subclavian and pulmonary arteries (PAs), (Blalock-Taussig [BT] shunt). The second stage, termed the Glenn procedure, involves the anastomosis of the superior vena cava (SVC) to the PA. This stage results in the SVC as the sole provider of pulmonary blood flow, allowing the ventricle to remodel and function at lower end-diastolic pressures in anticipation for the final stage, the Fontan procedure. This procedure entails the redirection of blood flow from the inferior vena cava into the pulmonary circulation. This operation finalizes the anatomic goals in which systemic flow is diverted directly into the pulmonary circulation, bypassing the nonfunctioning right or left heart.[4-6] The resulting Fontan circulation relies on passive, nonpulsatile flow through the lungs. The single ventricle has increased work to generate enough force to pump blood through the body but also overcome resistance across the pulmonary vascular bed.[7] Balancing pulmonary/systemic blood flow (Qp/Qs) is the primary goal in the management of single ventricle physiology. This ensures the optimization of systemic oxygen delivery.[8] Alterations in pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR) can lead to hypotension or hypoxia, hemodynamic derangements especially common during the perioperative period.[9,10] While low and intermediate risk noncardiac surgery is considered safe in patients with a single ventricle, these lesions add a significant risk of mortality.[11,12]

Despite all the well documented benefits (earlier return of gastrointestinal function, shorter hospital stay, less postoperative pain and earlier ambulation),[13] laparoscopic surgery can be especially concerning for patients with a single ventricle. By increasing intra-abdominal pressure, the pneumoperitoneum produces significant cardiovascular and respiratory alterations. Declines in cardiac output have been observed following abdominal insufflation as decreased venous return lessens cardiac preload, leading to hypotension. Additionally, the increased abdominal pressure elevates cardiac afterload and SVR, further compromising cardiac output. Peritoneal traction can lead to vagal stimulation and bradyarrhythmias.[14,15] Respiratory function can also be affected by elevated intra-abdominal pressures. Elevations in PVR due to hypercarbia can be caused by either direct carbon dioxide absorption or hypoventilation. Extreme patient positioning can exacerbate these hemodynamic changes. The "head-up" position often required for appropriate laparoscopic technique can decrease preload and lower cardiac output.[16,17] As mentioned, single ventricle physiology necessitates balance between systemic and pulmonary blood flow. The pneumoperitoneum and positional changes disrupt this balance and can lead to significant hypoxemia, hypotension and hemodynamic instability.[18,19] On the other hand, laparoscopic surgery also offers several aforementioned advantages. Therefore, the risks and benefits of laparoscopy for patients with single ventricle physiology must be carefully considered.

We present the unique case of an adult patient with a single ventricle heart defect presenting for a major laparoscopic intra-abdominal procedure. The case outlines the anesthetic management for patients with single ventricle physiology. Specifically, we highlight the concerns of laparoscopy and the challenges that this surgical technique presents in patients with complex congenital cardiac defects.

CASE REPORT

A 32-year-old male with a history of a cyanotic CHD and ulcerative colitis presented for a laparoscopic total colectomy. He was born with tricuspid and pulmonary atresia and underwent a right BT shunt at 3 days of age. At 3 months, he had an atrial septectomy performed. Persistently elevated PA pressures precluded him from undergoing a Fontan procedure. He therefore functioned with a single LV filled by both right and left atria. His pulmonary flow remained via the BT shunt with baseline oxygen saturations around 80%. Additional medical issues included a 10-year history of recurrent atrial fibrillation requiring multiple cardioversions. He was placed on amiodarone and systemic anticoagulation. He also had pulmonary hypertension for which he was medically managed with an endothelin receptor antagonist, ambrisentan. Preoperatively, cardiology was consulted and performed a transthoracic echocardiogram. This revealed normal left ventricular function with an ejection fraction of 62%. The single ventricle produced a high output state with a cardiac output near 17 L/min, as measured by quantitative Doppler stroke volume calculation. The echo also demonstrated mild LV hypertrophy without wall motion abnormalities. The remainder of the exam was consistent with tricuspid atresia, pulmonary atresia, an atrial septectomy, and a right BT shunt. Cardiology requested a specialist cardiothoracic anesthesiologist for this patient given his high-risk situation [Figure 1].

A laparoscopic approach was initially planned. His preoperative oxygen saturation was 78% with central and peripheral cyanosis. His baseline blood pressure was approximately 110/60 mmHg. A preoperative arterial catheter was placed for blood pressure monitoring. Anesthesia was induced with etomidate, fentanyl, and succinylcholine. Volume control ventilation was utilized with 600 ml tidal volumes resulting in peak airway pressures around 25 cm H_2O. Following induction, a central venous catheter was placed without difficulty and a transesophageal echo probe inserted. He was maintained on sevoflurane at an end tidal concentration of 2.0% along with 50% oxygen. Target oxygen saturations were between 75% and 85%. Intermittent boluses of vecuronium and hydromorphone were titrated during the case. In addition, two units of packed red blood were infused at the onset of the case due to anemia from persistent hematochezia. His preoperative hemoglobin was 12.9 g/dl, decreased from a baseline of 17 g/dl. To maintain optimal oxygen delivery during the perioperative period, a hemoglobin >16 g/dl was targeted. Following induction, positional maneuvers were tested to assess hemodynamic stability. Both Trendelenburg (head-down) and reverse Trendelenburg (head-up) positions produced little change in hemodynamics. A pneumoperitoneum was created, and the patient was placed in a head-up position for surgical access. These maneuvers resulted in hypotension with systolic blood pressures declining to 70 mm Hg. Multiple doses of phenylephrine were administered to temporarily resolve the hypotension. However, a vasopressin infusion was subsequently initiated due to persistently low blood pressures. He required increasing doses of vasopressin to a maximum of 2.4 units/h. Despite the abdominal distension created by the pneumoperitoneum, his blood pressures were adequately maintained with the vasopressin infusion. The surgical team proceeded with extensive exploration by a laparoscopic approach. This revealed dense adhesions and a significantly inflamed distal colon. Due to the difficult mobilization created by the adhesions, the decision was made to convert to an open surgical approach. The pneumoperitoneum was released, and the patient returned to the supine position. Within minutes, his blood pressure improved and the vasopressin was titrated off. The operation was resumed and completed without complications. The patient's neuromuscular blockade was reversed and he was extubated without difficulty. He was taken to the intensive care unit where he was monitored for hemodynamic changes, bleeding complications, arrhythmias, and other postoperative complications seen in patients with single ventricle conditions. He was eventually discharged home on postoperative day 10 [Figure 2].

DISCUSSION

Complicated congenital heart defects are increasingly prevalent in adults due to advances in the surgical repair of these lesions. As more of these patients survive into adulthood, higher numbers are presenting for noncardiac surgery. Overall, utilization of hospital resources is increased for this patient population.[20] Furthermore, adults with CHD presenting for noncardiac surgery have increased perioperative morbidity and mortality compared with a matched control cohort.[12] Risk factors for perioperative complications include baseline cyanosis, treatment for congestive heart failure, and poor general

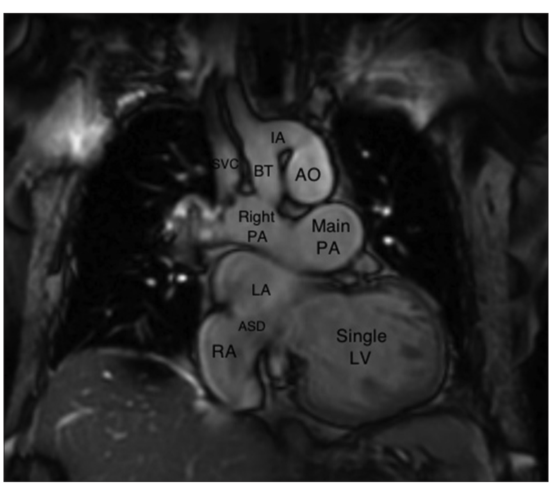

Figure 1: Cardiac magnetic resonance imaging showing the patient's Blalock-Taussig shunt connecting the innominate artery to the right pulmonary artery

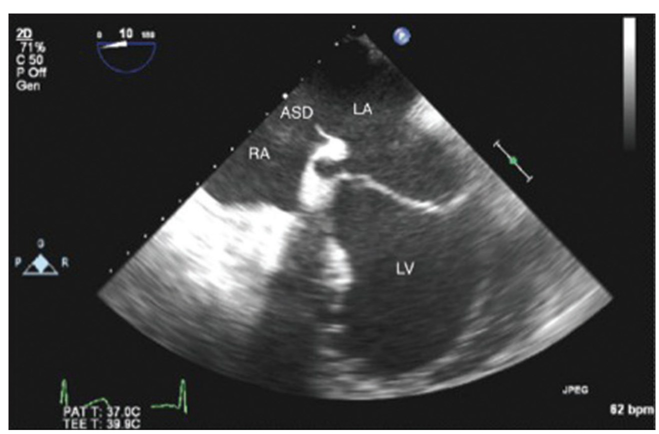

Figure 2: Intra-operative two-dimensional-echo demonstrating the surgically created atrial septal defect between the right atrium and left atrium as well as the patient's single left ventricle

health.[21] Conditions with the greatest complexity include single ventricle abnormalities, pulmonary and tricuspid atresia, transposition variants, pulmonary vascular disease, and Tetralogy of Fallot. These more complex patients are susceptible to comorbidities including cardiac, renal, and pulmonary disease. In turn, they are at significant risk of premature death, re-operation, and other complications.[22] Therefore, these patients require detailed preoperative workups and an understanding of their complex physiology for successful perioperative care. In addition, they necessitate a multidisciplinary care team. The consensus statement from the ACC/AHA 2008 Guidelines for Management of Adults with CHD recommends that patients with complex CHD undergoing surgery should be performed in a regional adult CHD center. Cardiology consultation should be completed prior to procedures in moderate to high-risk patients and an anesthesiologist familiar with CHD should care for these adults during the perioperative period.[23] Furthermore, these procedures are not suited for ambulatory surgery centers given the high-risk nature of these patients.

We outline a case of a patient with a single LV who presented for a laparoscopic colectomy for ulcerative colitis. The operation required a pneumoperitoneum and extreme patient positioning, leading to significant hypotension. Hemodynamic stability was maintained with vasopressor therapy; however poor surgical conditions necessitated a conversion to a laparotomy. Following release of the pneumoperitoneum and return to a supine position, the patient's blood pressure improved and the vasopressor support was titrated off.

This case highlights the anesthetic concerns for patients with complex congenital heart defects, specifically single ventricle conditions. As mentioned, single ventricle physiology leads to a delicate balance between systemic and pulmonary blood flow. Having not undergone a Fontan operation due to persistently elevated pulmonary pressures, this patient relied on a BT shunt to maintain blood flow from the systemic system (subclavian artery) to the pulmonary system (PA). As a result of the mixing between the systemic and pulmonary circulations, his baseline oxygen saturation and target for the perioperative period was 80%. Higher saturations might indicate excessive pulmonary flow, risking pulmonary edema and respiratory decline. Furthermore, patients with chronically low saturations undergo compensatory erythropoiesis resulting in higher than normal hematocrits. While no optimal hematocrit has been identified, it is important to maintain hematocrit levels near baseline to ensure optimal systemic oxygen delivery.[7] In patients with systemic to pulmonary shunts, pulmonary blood flow will vary with changes in the pressure gradient across the pulmonary vascular bed. Elevations in PVR due to hypercarbia or hypoxemia should be avoided to maintain adequate pulmonary flow and prevent arterial desaturation. Ventilatory management should promote maximal pulmonary flow by minimizing peak airway pressures. At the same time, significant increases in pulmonary flow can worsen pulmonary hypertension and decrease lung compliance. Inadequate anesthesia or suboptimal pain control can increase SVR, exacerbating left to right shunting as blood is directed away from the systemic circulation. As a result, cardiac output would be compromised. Similarly, decreased cardiac output can occur following declines in venous return from systemic hypotension or elevated intrathoracic pressures.[8-10] These are some examples of the complex physiology that must be considered in the setting of patients with single ventricles presenting for surgery.

To assist anesthetic management, invasive monitoring should be considered. However, placement can be technically difficult on account of anatomical abnormalities. In turn, the provider must have a precise understanding of a patient's anatomic variations and surgical repairs prior to placing central venous or PA catheters. As was the case for this patient, systemic to pulmonary shunts require that arterial line and pulse oximetry monitors be placed on the contralateral arm. Transesophageal echocardiography can be helpful to follow cardiac performance and fluid status. Intravenous line filters should be applied to prevent paradoxical air embolism. Postoperatively, these patients should be managed in an intensive care setting with continued meticulous attention to hemodynamics.

CONCLUSION

This was a challenging case of an adult patient with a complex congenital heart condition who underwent an extensive open abdominal operation due to failed laparoscopy. A growing number of adults with CHD are presenting for similar interventions. With detailed preoperative workups and close hemodynamic monitoring during the perioperative period, these patients can experience successful surgical outcomes.

REFERENCES

1. Williams RG, Pearson GD, Barst RJ, Child JS, del Nido P, Gersony WM, *et al.* Report of the National Heart, Lung,

and Blood Institute Working Group on research in adult congenital heart disease. J Am Coll Cardiol 2006;47:701-7.
2. Feinstein JA, Benson DW, Dubin AM, Cohen MS, Maxey DM, Mahle WT, *et al*. Hypoplastic left heart syndrome: Current considerations and expectations. J Am Coll Cardiol 2012;59 1 Suppl: S1-42.
3. Gulack BC, Adibe OO. Laparoscopic antireflux surgery in infants with single ventricle physiology: A review. J Laparoendosc Adv Surg Tech A 2013;23:733-7.
4. Delmo Walter EM, Hübler M, Alexi-Meskishvili V, Miera O, Weng Y, Loforte A, *et al.* Staged surgical palliation in hypoplastic left heart syndrome and its variants. J Card Surg 2009;24:383-91.
5. Petit CJ. Staged single-ventricle palliation in 2011: Outcomes and expectations. Congenit Heart Dis 2011;6:406-16.
6. Jaquiss R, Imamura M. Single ventricle physiology: Surgical options, indications and outcomes. Curr Opin Cardiol 2009;24:113-8.
7. McRae ME. Long-term issues after the Fontan procedure. AACN Adv Crit Care 2013;24:264-82.
8. Yuki K, Casta A, Uezono S. Anesthetic management of noncardiac surgery for patients with single ventricle physiology. J Anesth 2011;25:247-56.
9. Leyvi G, Wasnick JD. Single-ventricle patient: Pathophysiology and anesthetic management. J Cardiothorac Vasc Anesth 2010;24:121-30.
10. Cannesson M, Earing MG, Collange V, Kersten JR. Anesthesia for noncardiac surgery in adults with congenital heart disease. Anesthesiology 2009;111:432-40.
11. Torres A Jr, DiLiberti J, Pearl RH, Wohrley J, Raff GW, Bysani GK, *et al*. Noncardiac surgery in children with hypoplastic left heart syndrome. J Pediatr Surg 2002;37:1399-403.
12. Maxwell BG, Wong JK, Kin C, Lobato RL. Perioperative outcomes of major noncardiac surgery in adults with congenital heart disease. Anesthesiology 2013;119:762-9.
13. Harji DP, Griffiths B, Burke D, Sagar PM. Systematic review of emergency laparoscopic colorectal resection. Br J Surg 2014;101:e126-33.
14. Gerges FJ, Kanazi GE, Jabbour-Khoury SI. Anesthesia for laparoscopy: A review. J Clin Anesth 2006;18:67-78.
15. Struthers AD, Cuschieri A. Cardiovascular consequences of laparoscopic surgery. Lancet 1998;352:568-70.
16. Joris JL, Noirot DP, Legrand MJ, Jacquet NJ, Lamy ML. Hemodynamic changes during laparoscopic cholecystectomy. Anesth Analg 1993;76:1067-71.
17. McLaughlin JG, Scheeres DE, Dean RJ, Bonnell BW. The adverse hemodynamic effects of laparoscopic cholecystectomy. Surg Endosc 1995;9:121-4.
18. Gueugniaud PY, Abisseror M, Moussa M, Godard J, Foussat C, Petit P, *et al.* The hemodynamic effects of pneumoperitoneum during laparoscopic surgery in healthy infants: Assessment by continuous esophageal aortic blood flow echo-Doppler. Anesth Analg 1998;86:290-3.
19. McClain CD, McGowan FX, Kovatsis PG. Laparoscopic surgery in a patient with Fontan physiology. Anesth Analg 2006;103:856-8.
20. Mackie AS, Pilote L, Ionescu-Ittu R, Rahme E, Marelli AJ. Health care resource utilization in adults with congenital heart disease. Am J Cardiol 2007;99:839-43.
21. Warnes CA, Liberthson R, Danielson GK, Dore A, Harris L, Hoffman JI, *et al.* Task force 1: The changing profile of congenital heart disease in adult life. J Am Coll Cardiol 2001;37:1170-5.
22. Foster E, Graham TP Jr, Driscoll DJ, Reid GJ, Reiss JG, Russell IA, *et al.* Task force 2: Special health care needs of adults with congenital heart disease. J Am Coll Cardiol 2001;37:1176-83.
23. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, *et al*. ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: Executive Summary: A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines for the management of adults with congenital heart disease). Circulation 2008;118:2395-451.

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