Laparoscopic Surgery in a Patient with Fontan Physiology
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Introduction: Laparoscopic surgery represents a significant advance in surgical technique; proposed benefits include decreased pain and postoperative complications, and shorter time to discharge. The number of different types of laparoscopic procedures and potential candidates has expanded dramatically. However, there are several potential complications, including carbon dioxide embolus and significant hypercarbia; the importance of these issues could be potentially greater in patients with various forms of congenital heart disease. We present a case of a 19-year-old female with a history of palliated congenital cardiac disease who successfully underwent laparoscopic cauterization of endometriosis and lysis of adhesions.

Case Report: The patient is a 19-year-old female with chronic pelvic pain who elected to undergo laparoscopic evaluation and treatment of presumed endometriosis. She had a history of being a separated conjoined twin, heterotaxy syndrome, dextrocardia, pulmonary atresia and mitral atresia. She had undergone staged single ventricle procedures and at the time of this operation her cardiac anatomy was that of a total cavopulmonary Fontan; a fenestration had previously been device-occluded. In addition, she had a history of sinus node dysfunction necessitating atrial pacemaker placement. She had no clinical signs of reduced contractile function and no evidence of hypoxia or venous congestion. Preoperative echocardiogram revealed good systemic ventricular function, patent Fontan pathways, no intratrial shunting, and no significant atrioventricular or systemic valvar regurgitation. Prior to induction of anesthesia, her pacemaker mode was switched to asynchronous atrial pacing at a modestly increased rate above baseline. An intravenous line was started, midazolam given for sedation, and albumin given for volume expansion. Anesthesia was induced using etomidate, remifentanil and vecuronium without incident. A radial arterial catheter was placed and she was positioned and prepped. Anesthetic maintenance consisted of sevoflurane in air/oxygen, remifentanil and vecuronium. She tolerated incision and CO2 insufflation without evident problems; intraabdominal pressures were kept <10 cm H2O, which did not appear to affect oxygenation, ventilation, or cardiac output (e.g. pulse oximetry, end-tidal CO2, arterial blood pressure). The surgery concluded uneventfully, and after reversal of neuromuscular blockade, she was extubated and taken to the recovery room. She continued to do well and was discharged home the following day.

Discussion: Advances in the medical and surgical management of children with complex congenital heart disease have decreased the early and late mortality from these lesions. As a result, more patients are surviving into adulthood and are presenting more frequently for non-cardiac surgery. In this patient, her complex congenital disease and anatomy ultimately led to Fontan physiology. Originally described for tricuspid atresia in 1971, some version of the Fontan operation is now the ultimate goal for a variety of forms of congenital heart disease that can only be approached as “single ventricle” lesions. In Fontan physiology, systemic venous blood flows essentially passively into the pulmonary circulation where it becomes oxygenated. The blood then drains into what is often a common atrium and into the single ventricle that actively pumps oxygenated blood to the systemic circulation. Determinants of the efficacy of the Fontan circulation include systemic venous pressure and volume, pulmonary vascular anatomy, pulmonary vascular resistance, AV valve function, cardiac rhythm, and the function of the systemic ventricle. The most important consequence of any of these factors, alone or in combination, in Fontan patients is their ability to compromise systemic cardiac output. The physiologic changes produced by CO2 insufflation and positioning for laparoscopy are at odds with Fontan physiology. Increased intraabdominal pressure could lead to a significant decrease in systemic venous return. Hypercarbia (causing increased PVR) could result from CO2 absorption as well as impaired ventilation (due to abdominal distention); increased amounts of positive pressure could increase PVR and impair venous return by increasing intrathoracic pressure. Also, there are numerous reports of CO2 emboli resulting from insufflation. The consequences of CO2 embolus and resultant obstruction to pulmonary blood flow could theoretically be more severe in these patients whose pulmonary blood flow (and systemic preload) is passive. The presence of a patent fenestration would add the risk of paradoxical CO2 embolism to the coronary and/or cerebral circulations. Our patient did not exhibit adverse sequelae from this procedure. Based on her preoperative history, exam, and echocardiogram, she was a good candidate in terms of ventricular function and the status of her Fontan pathways. This case report suggests that laparoscopic abdominal surgery is possible in selected patients with well-compensated Fontan physiology. Minimizing insufflation duration and pressure in this group of patients would appear to be especially important, as would the ability and willingness to convert to an open procedure at the first indication of impaired ventilation or cardiac output. The possibility of pulmonary and paradoxical CO2 embolism, and their potential severity in this patient population, should also be considered.

References: