Laparoscopic Resection of a Pheochromocytoma in the Prone Position in a Patient with Fontan Physiology

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Introduction: As patients with congenital heart disease are living longer into adulthood, it becomes increasingly more common for them to present for non-cardiac surgery. Fontan physiology is especially challenging perioperatively, due to the nature of its non-pulsatile, low flow system through the pulmonary vasculature. We present a case of an adult with Fontan physiology undergoing a laparoscopic resection of a pheochromocytoma in the prone position.

Case Presentation: A 19 year old woman presented for left total adrenalectomy for an extra-adrenal paraganglioma resulting in a functional pheochromocytoma that was diagnosed incidentally on CT scan of the abdomen. Past medical history was complex and included a history of congenital pulmonary atresia with Ebstein's malformation of the tricuspid valve. Multiple shunts were performed before a Fontan circulation with fenestration was finally established. She was admitted a month prior to surgery for initiation of phenoxybenzamine and was subsequently discharged. During this admission, communication and planning was established with surgery, cardiology, and anesthesia.

In the operating room, an awake radial arterial line was inserted. Stable induction and intubation were achieved with midazolam, remifentanil, propofol, and rocuronium. Additional monitoring included CVP and TEE. She was placed in the prone position. Magnesium was loaded and then infused throughout the case for the prevention of arrhythmias. Insufflating pressures were maintained at 12–15 cm H2O. Dobutamine was started to maintain hemodynamic stability after the TEE exam (performed in the prone position) demonstrated LV dysfunction. Phentolamine was available but never administered. She was extubated in the OR and transferred directly to the cardiac ICU. Despite our attempts at fluid restriction, she required diuresis with Lasix postoperatively due to fluid overload, but was able to be discharged home after 1 week.

Discussion: Fontan physiology can be especially challenging in the perioperative setting. As has previously been described, laparoscopy, prone positioning, and the catecholamine surge from pheochromocytomas all individually pose unique and potentially detrimental physiologic sequelae in these patients. There has yet to be a case reported involving all of these variables. In the management of our patient we collaborated with the cardiologist and surgeon pre-operatively, used magnesium for the prevention of arrhythmias, placed a TEE probe intraoperatively, and kept insufflation pressures less than 15 mm Hg. With these measures, our patient remained hemodynamically stable throughout the procedure, and was discharged home after one week with the post-operative challenge being fluid overload. This case demonstrates that with proper planning and collaboration, more complex surgical procedures can be safely performed on the ever growing number of adults with Fontan physiology.

References: