Anesthetic Management of a Patient with Surgically Corrected D-Transposition of the Great Arteries Undergoing Laparoscopic Pyloromyotomy

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Citation

Abstract
We present a case of a newborn with surgically corrected D-transposition of the great arteries (D-TGA) undergoing laparoscopic pyloromyotomy. Anesthetic implications of corrected D-TGA are discussed. As more children with successful repair of D-TGA survive into adulthood, anesthesiologists will increasingly encounter them as patients presenting for noncardiac surgery. It is important to understand the type of repair that was performed as well as the residual comorbidities particular to each procedure. With careful preoperative evaluation and experienced providers, routine anesthetics with noninvasive monitoring can be a safe option.

INTRODUCTION
With the exception of isolated ventricular septal defects, D-transposition of the great arteries (D-TGA) is the most common congenital cardiac defect. As surgical repair has become more successful, long-term survival rates have improved and the need for subsequent noncardiac surgeries for these children has increased in frequency. Few data exist regarding the anesthetic risks faced by patients with surgically corrected D-TGA and only one small case series has attempted to document anesthesia techniques and outcomes in this population. We describe the anesthetic management of an infant with surgically repaired D-TGA undergoing laparoscopic pyloromyotomy.

CASE REPORT
A 7-week-old boy presented for surgical treatment of pyloric stenosis. The patient had presented to the emergency department with a 3-day history of projectile vomiting progressively increasing in frequency. An abdominal ultrasound revealed the diagnosis of pyloric stenosis with a 19 × 6 mm pylorus. The patient had a significant medical history that included an arterial switch operation (ASO) for the treatment of D-transposition of the great arteries (D-TGA), diagnosed prenatally. Shortly after birth prostaglandin therapy was started at a dose of 0.1 μg/kg/min. Soon after initiation of prostaglandin therapy the patient developed apnea requiring intubation and mechanical ventilation. An echocardiogram at that time revealed D-TGA with an intact interventricular septum. The patient also had a restrictive patent foramen ovale and a small patent ductus arteriosus. The patient was therefore taken to the cardiac catheterization laboratory for a balloon atrial septostomy, which was performed without complications. The patient’s oxygen saturation by pulse oximetry (SpO₂) immediately improved from 70% to 95% by the conclusion of the procedure.

On day 5 of life the patient was taken to the operating department for an arterial switch operation. The operation went smoothly and the patient recovered from the procedure and was discharged home on postoperative day 10. The patient’s medical regimen included digoxin and furosemide.

The patient was brought to the operating department for a laparoscopic pyloromyotomy at 7 weeks of age. Preanesthetic evaluation confirmed that the patient had been hydrated and that electrolyte balance had been restored. Intravenous atropine was administered (0.16 mg) and the patient’s stomach contents were evacuated with a 14 French suction catheter placed by the oral route. A modified rapid-sequence induction (positive pressure ventilation during induction with cricoid pressure applied) was performed using propofol 3 mg/kg and rocuronium 1 mg/kg intravenously. Maintenance of anesthesia included the use of desflurane and morphine 0.1 mg/kg. The laparoscopic
pyloromyotomy was performed without complications and insufflation pressure was limited to less than 10 mm Hg. Neuromuscular blockade was reversed with neostigmine and glycopyrolate and the patient was extubated and transferred to the recovery room with 30% inspired oxygen concentration. The patient was monitored overnight in the critical care unit and had an uneventful postoperative course. The patient was discharged on postoperative day 1 after it was confirmed that he was tolerating full feeds by the oral route.

**DISCUSSION**

Patients with D-TGA with intact ventricular septum that has been surgically corrected by ASO have a long-term survival rate of 90% to 92%. Supravalvular pulmonary stenosis and aortic insufficiency are the most common long-term complications and the majority of cases are classified as mild to trivial at 10 years. Asymptomatic atrial and ventricular premature beats can occur but are uncommon. Because the coronary arteries are reimplanted during repair, myocardial ischemia is a potential problem following ASO. Although 90% to 97% of patients have patent coronary arteries by angiography, 89% of coronary events occur in the first 3 months following ASO. Extracardiac defects associated with D-TGA are rare, and patients in whom this defect has been surgically repaired who later undergo noncardiac surgery have been managed as having normal heart structure and function. A recent case series suggests that the majority of these patients can safely undergo routine general anesthesia with no invasive monitoring. 

Consistent with current practice, our patient underwent general anesthesia for laparoscopic pyloromyotomy in the usual fashion using standard monitors and typical rapid-sequence induction technique for this procedure. Although the tachycardia and stress due to pretreatment with atropine and awake orogastric suctioning increase the patient’s myocardial oxygen demand, the risk of aspiration was deemed more significant than the risk of myocardial ischemia. The rapid-sequence induction was modified in order to maintain adequate oxygenation during induction. This technique is commonly used in neonates and was particularly important in this patient, who had a slightly increased risk of stress-induced myocardial ischemia. The standard rapid-sequence induction dose of rocuronium was modestly reduced because the procedure was expected to be relatively short.

The overwhelming majority of pyloromyotomies in infants are done laparoscopically. Laparoscopic surgery can alter cardiopulmonary function due to intra-abdominal pressure, positioning, and absorbed carbon dioxide. Pulmonary effects of decreased functional residual capacity, decreased pulmonary compliance, and ventilation/perfusion mismatch can lead to hypercapnia and hypoxemia. Pneumoperitoneum causes increased pulmonary and systemic afterload and decreases cardiac output. Hypercapnia can initiate a sympathetic response with increases in heart rate, blood pressure, and possibly ventricular arrhythmias. Laparoscopic surgery in children with congenital heart disease (CHD) was originally considered contraindicated due to these physiologic challenges. However, recent literature has suggested that cardiovascular effects are minimal if insufflation pressures are maintained at less than 10 mm Hg, and several case reports have described successful laparoscopic surgery in patients with CHD. Hypertrophic pyloric stenosis (HPS) is a relatively common diagnosis in infancy but the cause is unknown. It has been associated with certain syndromes that can also include CHD, and one small study suggests an association between isolated HPS and CHD. Our patient did not have a congenital syndrome and had no other known gastrointestinal abnormalities.

Gastric outlet obstructive lesions have also been linked to HPS and can mimic the clinical and radiological findings. These include eosinophilic gastroenteritis, transpyloric feeding tubes, and gastric mucosal hypertrophy. Treatment with prostaglandins has been shown to induce proliferation of gastric antral mucosa, occasionally causing gastric outlet obstruction in infants. It has been postulated that secondary HPS can develop in response to the obstruction caused by prostaglandin-induced foveolar hyperplasia. Although our patient did receive a prostaglandin infusion prior to his D-TGA repair, he did not have a history of gastric outlet obstruction during his initial hospitalization.

As more children with successful repair of D-TGA survive into adulthood, anesthesiologists will increasingly encounter them as patients presenting for noncardiac surgery. It is important to understand the type of repair that was performed as well as the residual comorbidities particular to each procedure. With careful preoperative evaluation and experienced providers, routine anesthetics with noninvasive monitoring can be a safe option for this patient population.
References
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