CLINICAL VIGNETTE

Challenges in the Perioperative Management of Infantile Hypertrophic Pyloric Stenosis

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Introduction

Infantile hypertrophic pyloric stenosis (IHPS) is a unique and uncommon disorder that presents in older neonates or younger infants. Definitive treatment is often performed by a relatively quick surgical procedure shortly after diagnosis, but patients often bring distinct preoperative, intraoperative, and postoperative challenges.

Case Report

A 7-week-old infant born at 38 weeks gestational age was brought to the ER for vomiting of one week duration and one episode of watery stool that same day. Previously, his birth history was eventful for a cesarean delivery secondary to failure to progress with no other complications. He began to have small amounts of emesis following feedings for the past week that were not bloody nor bilious. He was afebrile, did not appear lethargic to his parents, and had consistently normal numbers of feedings and wet diapers during the week.

His vital signs were HR 145 bpm, BP 80/42 mmHg, RR 37/min, T 36.8°C, and S_pO_2 100%. On physical exam, he appeared interactive with a normal fontanelle, mucous membranes, and capillary refill time. No discrete abdominal masses were palpable. His labs were notable for Na 135 mmol/L, K 5.5 mmol/L, Cl 105 mmol/L, HCO3 23 mmol/L, and Cr 0.3 mg/dL. He underwent an abdominal ultrasound that revealed findings concerning for pyloric stenosis. The consulting surgeon requested to perform an open pyloromyotomy later the same afternoon.

The patient was given a peripheral intravenous line and placed on maintenance fluids of D5 ½ NS prior to arriving to the preoperative area. Prior to anesthetic induction, the patient's stomach was suctioned via temporary placement of an orogastric tube in the operating room. The patient was induced with fentanyl, propofol, and succinylcholine and was intubated immediately. The anesthetic was maintained with sevoflurane, and bupivacaine local anesthetic was infiltrated in the incision for analgesia at conclusion of the pyloromyotomy.

Discussion

The exact etiology of IHPS is unknown and likely both genetics and environmental factors have some influence. The incidence varies somewhat among studies but has been found to be approximately 1 to 3 cases per 1,000.[1-3] The disease has been associated in studies with various environmental factors including exposure to macrolide antibiotics, bottle feeding, and living in a rural area.[3-6] Patients are most often first-born males, and there may be some small association of IHPS with prematurity.[7]

The classical presentation of IHPS is a three-to five-week old infant with non-bilious, projectile vomiting that continuous to have a strong appetite. On physical exam, there can be a palpable pyloric "olive-like" mass in the abdomen with signs of dehydration. Patients can have associated electrolyte abnormalities from the loss of gastric acid and significant hypovolemia, typically resulting in hypochloremic, hypokalemic metabolic alkalosis. Severe hypovolemia may lead to acidosis when perfusion is compromised.

The patient that is presented did not have some of the classic presentations described above, including his older age, lack of forceful vomiting, benign physical exam and vital signs, and hyperkalemia. The child's near normal chloride and bicarbonate levels and reassuring exam indicate that he was not significantly hypovolemic. This is consistent with the possibility that the expected findings in pyloric stenosis have become attenuated compared to past decades due to earlier diagnosis, better nourishment, and wider healthcare availability. Interestingly, similar to the case presentation above, one study reviewed 140 patients with pyloric stenosis and found approximately one-third of them to have increased potassium levels.[8]

Ultrasonography has replaced barium studies as the diagnostic procedure of choice for IHPS. Common criteria measured include the pyloric muscle thickness, length, and diameter. Patients with IHPS have greater than normal measurements for these dimensions. Endoscopy and upper GI contrast studies are other possible diagnostic modalities.

The most common and definitive management of IHPS is pyloromyotomy in which the hypertrophied pylorus is incised and dissected to the submucosa. This can be performed either as an open surgery or laparoscopically. Studies show an overall complication rate to be similar between the two methods, though there is some evidence that laparoscopy may slightly reduce time to full enteral feeding and length of postoperative hospital stay.[9-12] Medical management of pyloric

stenosis is rare. It includes using anticholinergic medications, such as atropine, or duodenal feedings using an enteral tube.

Hypovolemia and electrolyte abnormalities should be corrected with intravenous therapy prior to surgery as pyloromyotomy is generally not an emergency. Ensuring an appropriate NPO interval while receiving fluid therapy may also decrease the risk of aspiration during the anesthetic. It is also possible to attempt to suction any gastric contents with an orogastric or nasogastric tube immediately prior to induction to further minimize the possibility of aspiration. In this instance, the patient received some continuous intravenous fluids but did not require aggressive volume replacement as evidenced by his exam and electrolytes.

Rapid-sequence induction, including a paralytic agent and cricoid pressure, is the most common strategy for airway management in light of the risk of aspiration. Awake intubation allows for maintaining protective reflexes and spontaneous respirations, but appears to confer no added benefit in otherwise normal-appearing infants.[13] Intubating a vigorous infant without anesthesia may prolong intubation time, require more laryngoscopy attempts, and result in episodes of trauma, breath-holding, laryngospasm, bradycardia, and hypertension. Though not widely accepted for instances with a high aspiration risk, inhalation induction has been used for pyloromyotomy as well. A retrospective review at a single institution where gas induction followed by intravenous muscle relaxant is commonly administered for these cases found no aspiration events in 252 inhalation inductions.[14]

Succinylcholine is a common paralytic agent used in rapid sequence induction, though the FDA has given the drug a black box warning secondary to possible hyperkalemic rhabdomyolysis in children with undiagnosed myopathy. Rocuronium is an alternative muscle relaxant, but its longer duration of action may be a cause of delay after a relatively short pyloromyotomy procedure. However, a retrospective study at a single center showed no significant difference in time from surgery stop to the time of patient transport out of the OR when comparing induction with rocuronium to succinylcholine.[15]

Given the relatively young postconceptual age of these infants, patients should be maintained on cardiorespiratory monitoring overnight. There are specific reports of postoperative apneic episodes in formerly full-term children with IHPS following surgery.[16,17] An exact cause for the propensity is unknown, but it is possible that metabolic alkalosis resulting in an increase in CSF pH causes respiratory depression.

Following surgery, this patient was admitted to the pediatric ward for postsurgical monitoring including pulse oximetry. He was able to begin oral feedings overnight and was discharged on the first postoperative day. The infant's return to clinic confirmed that the patient had no complications and no further regurgitation.

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