PYLORIC STENOSIS

INTRODUCTION

First described in 1788, pyloric stenosis is one of the most common gastrointestinal abnormalities presenting in the first few months of life. Surgical correction was initially described by Ramstedt (1912); at Hasbro Children's Hospital, approximately 50-75 pyloromyotomies are performed yearly. Inheritance is polymodal; occurrence is more frequent in white males, with an overall incidence of approximately 0.3%. Associated anomalies, though unusual, may include cleft palate and esophageal reflux.

Presentation typically occurs between 2-8 weeks of age (range - newborn to 5th month of life); cardinal features include projectile, non-bilious vomiting, visible peristalsis, and a hypochloremic metabolic alkalosis. Jaundice, due to caloric deprivation and hepatic glucuronyltransferase deficiency, occurs in 2-5% of children, and resolves with pyloromyotomy. Although the diagnosis can be made on the basis of history and physical examination alone, most patients will have the diagnosis confirmed by abdominal ultrasonography.

PATHOPHYSIOLOGY

Pathologic findings consist of hypertrophy of the circular and longitudinal fibers of the pylorus. As milk passes through the narrowed pylorus, mucosal swelling occurs, the pyloric channel narrows, and gastric emptying is delayed. Gastric dilatation, regurgitation, and non-bilious vomiting result. An olive-sized mass is often palpable in the upper abdomen; “tumor” size does not tend to correlate, however, with severity of the gastric outlet obstruction.

Persistent vomiting results in a loss of gastric fluid rich in hydrogen and chloride ions, with resultant dehydration, weight loss, anemia, and hypochloremic metabolic alkalosis. Compensatory mechanisms include the (i) respiratory system (hypoventilation - respiratory acidosis) and (ii) renal system. An increased bicarbonate load is presented to the kidney, and alkaline urine ($pH > 8$) is produced, with preferential excretion of HCO_3^- in place of chloride. Sodium ions become depleted along with HCO_3^-, and aldosterone secretion occurs, leading to sodium retention and kaliuresis. Potassium may be further depleted by potassium exchange with hydrogen ions in an effort to normalize serum pH. The situation is exacerbated by further aldosterone secretion in an effort to maintain circulating volume. Finally, paradoxical aciduria (a falling urine pH is generally an ominous sign) occurs as sodium is exchanged for hydrogen ion in the distal tubules.

ANESTHETIC CONSIDERATIONS

Surgical correction of pyloric stenosis is *never* a surgical emergency, and volume and electrolyte deficits of sodium, chloride, and potassium must be repleted to restore acid-base balance prior to surgical intervention. In particular, serum chloride levels provide an index for assessing severity of the metabolic alkalosis. Serum chloride levels of 88 mEq/L, bicarbonate levels less than 30 mmol/L indicate “reasonable” correction prior to surgery.

Dehydration is best assessed by physical and physiologic parameters. Poor skin turgor and dry mucous membranes correlate with 5% dehydration. Sunken fontanels, oliguria and tachycardia suggest closer to 10% dehydration, while hypotension, sunken eyeballs and lethargy are features of 15% dehydration. A reasonable approach to the significantly dehydrated infant with pyloric stenosis would be an initial bolus of 10-20 ml/kg normal saline or lactated Ringer's solution. IV hydration then proceeds, guided by clinical status, urine output, and serum electrolyte determinations. Potassium is usually added to IV solution only after voiding has occurred. Generally, patients are ready for surgery within 12-48 hours. As noted above, key indications of readiness for pyloromyotomy include (i) good hydration status, as indicated by normal skin turgor, urine output, vital signs and activity; (ii) normalization of acid-base status and electrolyte imbalance, as indicated (among other parameters) by serum chloride > 88 and potassium > 3.0.

Even after correction of fluid and electrolyte disturbances, several important considerations bear mention: (i) neontate - pharmacokinetic differences, temperature, labile cardiovascular system, metabolic homeostasis; and (ii) gastrointestinal obstruction - at risk for aspirating gastric contents, which may (in some patients) include barium. Recent data suggests that outcome might be improved in these patients if anesthetized by *pediatric anesthesiologists, as* is our predominant practice at Hasbro Children's Hospital.

Due to young age and full stomach situation, these patients are brought to the operating room without a parent. Routine monitors are established, including precordial stethoscope. The child will almost always have an indwelling intravenous, but it is wise to carefully assess its function before proceeding. Atropine, 0.02 mg/kg IV (minimum 0.1 mg), may be given at this time, and the nasogastric tube suctioned. If no nasogastric tube is present, the infant should be turned left lateral, and orogastric suctioning performed. If awake intubation is planned, I topicalize the throat and epiglottis (by fingertip) with a small amount of 2% lidocaine jelly.

The choices for intubation are awake vs rapid sequence induction. The pros and cons to each technique are cited in "Management of the Routine Pediatric Airway". Most often a rapid-sequence induction is performed, although small tidal volume ventilation is often introduced after induction to prevent oxygen

desaturation. Awake intubation represents a reasonable alternative in infants < 4 weeks of age and 4 kg in weight, especially if airway difficulties are anticipated. An able assistant is required to stabilize the shoulders; an experienced practitioner should be able to intubate expeditiously without undue trauma. Infants > 4 weeks / 4 kg are often too vigorous to intubate via the awake route, and rapid sequence induction is the technique of choice. Succinylcholine (2 mg/kg) is generally the relaxant of choice. Alternatively, rocuronium may be used, although this has been associated with prolonged extubation times in some cases. Preoxygenation is crucial, as rapid desaturation will occur following even a brief period of apnea in neonates. Cook-Sather et al. found in a prospective, non-randomized study that rapid IV (or modified rapid IV) induction was associated with shorter intubation times and no additional complications as compared to awake intubation (*Anesth Analg* 86:945;1998).

The surgical procedure takes 30 minutes or less. Approach is either a small peri-umbilical incision, or via laparoscopy. Anesthesia is maintained with oxygen, nitrous oxide and low-dose isoflurane or sevoflurane. I generally administer an intermediate acting muscle relaxant, such as atracurium or rocuronium, after endotracheal intubation; at completion, the infants are able to be promptly reversed and extubated. Acetaminophen (40 mg/kg) may be administered rectally after induction, and a small amount of bupivacaine 0.25% can be injected into the small (approximately 5 cm) right upper quadrant incision site.

Major postoperative risks are those of virtually any newborn anesthetic, including respiratory insufficiency, hypothermia, and reactive hypoglycemia. Postoperative cardiorespiratory monitoring is indicated, although intensive care observation is generally unnecessary. Feeds are instituted 8 hours after surgery, and advanced as tolerated.

REFERENCES

1) Patel RI. “Pyloric Stenosis” in *Common Problems in Anesthesia*, 2nd edition, Mosby; 1992

2) Caty MG and Azizkhan RG. *Ped Annals* 23:192; 1992

3) Smith RM:*Anesthesia for Infants and Children*, 6th edition, Mosby; 1996