Chapter 21

# Pyloric Stenosis

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A six-week-old male arrives to the emergency room with sudden onset of projectile vomiting, which occurs shortly after feeding. He was born via spontaneous vaginal delivery at 38 weeks. There is no additional medical or surgical history.

His current vital signs are: blood pressure 60/37 mmHg; heart rate 146/min; respiratory rate 32/min; temperature 37.1°C; $SpO_2$ 100% on room air. Weight is 3.8 kg.

## DIAGNOSIS

### What Are the Important Diagnoses to Consider?

Severe reflux, esophageal atresia, and pyloric stenosis should be considered in a patient with this presentation. Intussusception may also be part of the differential but usually results in intermittent pain and vomiting.

Severe reflux is usually a medical diagnosis and common at this age. Treatment includes oral medication for gastric acid secretion such as H2 receptor blockers and/or proton pump inhibitors. Esophagogastroduodenoscopy (EGD) may be considered as well as placement of a pH probe.

Pyloric stenosis (PS) is a potentially life-threating condition in which the pyloric musculature becomes hypertrophied leading to gastric outlet obstruction. The muscle is usually normal at birth and hypertrophies in the first few weeks to months of life. Children with PS present with feeding difficulties, weight loss, significant dehydration, and electrolyte disturbances. The vomiting is usually non-bilious as the thick pylorus prevents bile regurgitation. Interestingly, the pyloric hypertrophy generally resolves if left untreated. Historically, the treatment was frequent but small feedings until spontaneous resolution and thus why PS remains a medical and not surgical emergency. With the advances in surgical treatment and technique, the preferred method after medical stabilization is via a small incision and pyloromyotomy (incising of the pyloric muscle), immediately relieving the gastric outflow obstruction.

### Briefly Review the Historical Overview of Pyloric Stenosis

Reports of PS exist as early as the 15th century, however, the modern description is owed to Hirschsprung who reported clinical and autopsy findings of pyloric hypertrophy in 1887. Initially, PS was uniformly fatal in the first weeks to months, with most infants succumbing to malnutrition and dehydration. Early surgical intervention was via full thickness incision of the pyloric musculature and transverse closure. This was successful but mortality rates were still as high as 50%. In the early 20th century, Ramstedt introduced the suture-less technique of longitudinal division of the pyloric muscle while leaving the mucosa intact. This modification still serves as the basis for current operative management. Presently, advances in minimally invasive surgery make the laparoscopic approach the preferred method owing to decreased wound complications and a shorter length of stay.

### What Is the Incidence of Pyloric Stenosis?

The incidence of PS is 1.8 per 1,000 births. There is a significant male predominance of 5:1 for development of pyloric stenosis.

### What Is the Most Likely Age for Presentation?

Pyloric stenosis can present from infancy to around four months of age. Most patients typically present between 3 and 10 weeks.



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## Is There a Genetic Component to Pyloric Stenosis?

To date, five genetic loci have been identified for idiopathic infantile pyloric stenosis. Pyloric stenosis is also associated with the following congenital syndromes: trisomy 21, trisomy 18, Cornelia de Lange, Apert, Opitz FG, Marden-Walker, Smith-Lemli-Opitz, Zellweger, duplication 1q, duplication 9q, deletion 11q, ring 12, Denys–Drash, and paramyotonia congenita.

## What Are the Risk Factors for Development of Pyloric Stenosis?

Risk for development of PS can be divided into maternal and infant factors.

Maternal factors include: smoking of >10 cigarettes/day in early pregnancy, erythromycin use, age <25 years, and being overweight (but not obese).

Infant factors include: Very preterm infants, delivery by cesarean section and a birth order <2.

It has been suggested that infant exposure to prostaglandins (usually for congenital heart disease) may relay an increased risk of development of PS.

Preterm infants are generally diagnosed at a later post gestational age.

# PREOPERATIVE EVALUATION

## The Surgeon Requests to Perform the Pyloromyotomy after His Next Case. Is This Appropriate?

The next step is a thorough examination of the infant followed by IV placement in which labs are obtained. Dehydration and electrolyte status dictates when the patient is ready for operative intervention.

## How Is the Diagnosis of Pyloric Stenosis Made?

Typically, the history and physical examination are suggestive. A small (olive size) mass may be palpated in the upper abdomen and carries a 99% positive predictive value but cannot always be felt.

In the past, upper GI series was the imaging method used to diagnose PS (Figure 21.1). Widespread use and availability of ultrasound has made sonography the imaging test of choice in these patients. While the diagnosis of PS can be made clinically based on the history and physical examination, most patients will undergo ultrasound evaluation.

## What Is the Appropriate Workup for Patients with Pyloric Stenosis?

Evaluation should begin with a history of presenting symptoms to assess the degree of dehydration. Hydration status at this age should include observation for skin turgor, sunken eyes, mucous membranes, fontanelle, weight change, and number of dry diapers per day (Table 21.1).

Unlike adults, children have the ability to maintain their hemodynamics until they are profoundly dehydrated. Estimation of the degree of dehydration in children can be found in Table 21.1.

Laboratory evaluation should focus on the electrolytes to assess the degree of derangement. Intravenous access should be established.

Fluid administration in the form of 0.9% saline or D5NS should be given until the electrolytes, and

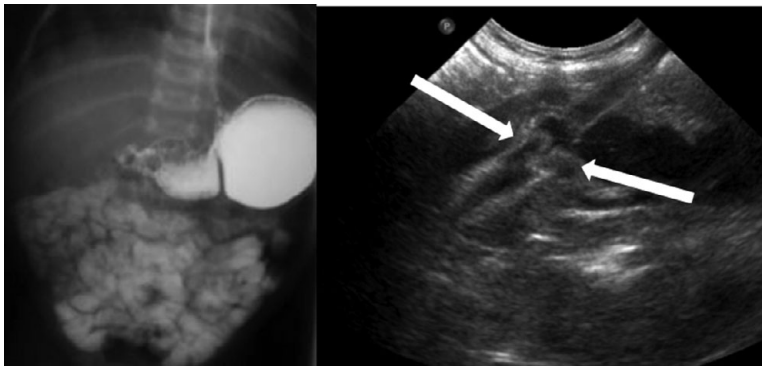

**Figure 21.1** **Left**: Upper GI demonstrating slow emptying of barium past the pylorus. **Right**: Abdominal ultrasound



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**Table 21.1** Assessment of dehydration severity in children

| | Mild | Moderate | Severe |
|---|---|---|---|
| Percentage of fluid loss | 5 | 10 | 15–20 |
| Fontanelle and eyes | Normal | Sunken fontanelle | Sunken eyes |
| Mucous membranes | Dry | Dry | Parched |
| Skin turgor | Normal / poor | Poor | Parched |
| Blood pressure | Normal | Mildly hypotensive | Severe hypotension / shock |
| Heart rate | Normal | Mild tachycardia / weak pulse | Significant tachycardia |
| Respiratory rate | Normal | Deep | Deep and rapid |
| Urine output | Normal / mild oliguria | Oliguria | Oliguria / anuria |

especially the chloride, return to normal. Potassium is often added to fluids to avoid hypokalemia that may occur with absorption of sodium.

## What Is the Expected Initial Metabolic Profile in Pyloric Stenosis?

Due to chronic emesis, sodium, chloride, and hydrochloric acid become depleted. This leads to a hyponatremic, hypokalemic, hypochloremic metabolic alkalosis.

## What Is the Endpoint of Resuscitation?

The goal of fluid resuscitation is aimed at normalizing intravascular volume and repletion of electrolytes. Fluid administration should ensue with the initial goal of approximately 2 cc/kg/h of urine production.

Electrolyte correction should continue with a goal of reaching chloride values of >100 mEq/L and a bicarbonate of <28 mEq/L.

## Why Is Bicarbonate Correction Important in the Preoperative Period?

Correction of the metabolic alkalosis is important in prevention of alkalosis induced postoperative apnea. Persistent alkalosis can depress the respiratory drive and preclude safe extubation.

## Is the Preferred Method of Induction an Awake Intubation?

Formerly, awake intubation was considered the method of choice for securing the airway in patients with PS. Cook-Sather et al. revealed that awake suctioning of the stomach decreases the gastric volume to a safe level, allowing for induction of anesthesia and asleep intubation.

Generally, the stomach is suctioned with an OGT multiple times in the awake infant, rotating the child until minimal to no gastric contents return. This should be followed by a rapid sequence intubation to secure the airway.

# POSTOPERATIVE CONSIDERATIONS

## What Is the Optimal Time for Postoperative Discharge?

Patients should be able to tolerate PO feeding prior to discharge. Young infants <50 weeks post conceptual age are at risk for postoperative apnea and should be monitored. Risk of postoperative apnea is increased with opioid administration.

## When Should Patients Resume a Regular Diet Postoperatively?

Numerous studies have attempted to determine the optimum time for resumption of feeding post PS repair, as there is concern for post-op vomiting. While there is no consensus as to the timeframe for feeding, a small period of NPO time appears to decrease the frequency of post-op emesis and decreases hospital length of stay.


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## Suggested Reading

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