Hypertrophic Pyloric Stenosis

22

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Infantile hypertrophic pyloric stenosis (IHPS) is a common surgical condition encountered in early infancy, occurring in 2–4 per 1000 live births. It is characterized by hypertrophy of the circular muscle, causing pyloric narrowing and elongation. The incidence of disease varies widely with geographic location, season, and ethnic origin. Boys are affected four times more often than girls.

There is evidence of a genetic predisposition to the development of this condition. Siblings of patients with IHPS are 15 times more likely to suffer the condition than children who have no family history of IHPS. The cause of hypertrophic circular muscle of the pylorus is still obscure. Various hypotheses have been advocated, including abnormal peptidergic innervation, abnormality of nitrergic innervation, abnormalities of extracellular matrix proteins, abnormalities of smooth-muscle cells, and abnormalities of intestinal hormones.

22.1 Presentation

The typical clinical presentation of infants with IHPS is nonbilious vomiting, usually occurring at 2–8 weeks of age in full-term infants. The development of IHPS in preterm infants has been described previously as a rare entity, but a recent large population study in the United States reported a significant increase in IHPS rates among premature infants, and the preterm infants typically presented at a later chronological age than full-term infants. Initially there is only regurgitation of feeds, but over several days, vomiting progresses to be characteristically projectile. It occasionally contains altered blood in emesis (appearing as brownish discolouration or coffee grounds) as a result of gastritis and/or oesophagitis.

22.2 Diagnosis

The diagnosis is usually based on the clinical history and physical examination of a "palpable pyloric tumour". Ultrasonographic scanning of the abdomen reveals a typical hypoechoic ring with an echogenic centre of increased muscle thickness. In difficult and/or complicated presentations, a contrast meal may be required, which shows a characteristic narrowed, elongated pyloric canal.

22.3 Treatment

Persistent nonbilious vomiting in these patients results in chloride depletion, metabolic alkalosis, and dehydration. Haematological and biochemical analysis should be undertaken. Any fluid, electrolyte, and acid-base imbalance should be corrected prior to surgery. Oral feeding should be discontinued and a nasogastric tube should be inserted prior to surgery to keep the stomach empty. The operation for pyloric stenosis is not an emergency and should never be undertaken until serum electrolytes have returned to normal.

Ramstedt's pyloromyotomy is the universally accepted operation for pyloric stenosis. A 3-cm transverse right upper quadrant, muscle-splitting incision provides excellent exposure and direct access to the pylorus with minimal retraction. Another incision that is commonly used is a supra-umbilical fold incision. Although a supra-umbilical skin-fold incision has a better cosmetic result, it has been argued that delivery of the pyloric tumour can be difficult and time-consuming and may damage the serosa of the stomach or duodenum by tearing. Some surgeons have used transumbilical intracavitary pyloromyotomy without delivering the pyloric tumor outside. In recent years, more and more centres are employing laparoscopic pyloromyotomy as the surgical approach. The main advantage of the laparoscopic pyloromyotomy is the superior cosmetic result.

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A nasogastric tube must be placed before the induction of anaesthesia, if the tube was not placed preoperatively. If a barium meal study has been carried out prior to surgery, it may be necessary to remove the residual barium meal by gastric aspiration and irrigation.

22.3.1 Operative Procedure: Ramstedt's Pyloromyotomy

Figures 22.1, 22.2, 22.3, 22.4 and 22.5 illustrate an open procedure.

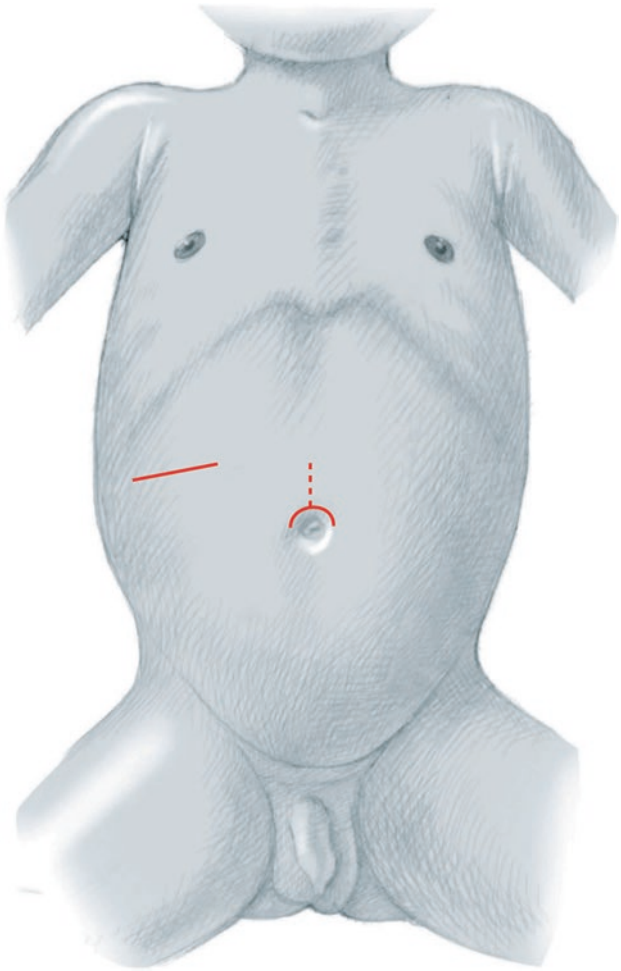

Fig. 22.1 The patient is placed in the supine position. After the induction of anaesthesia and endotracheal intubation, careful abdominal palpation will usually identify the site of the pyloric tumour. A transverse incision 2.5–3 cm in length is made lateral to the lateral border of the rectus muscle. The incision is deepened through the subcutaneous tissue; the underlying external oblique, internal oblique, and transverse muscles are split. The peritoneum is opened transversely in the line of the incision.When a supraumbilical skin fold incision is employed, a circumumbilical incision is made through about two thirds of the circumference of the umbilicus. The skin is undermined in a cephalad direction above the umbilical ring and the linea alba is exposed. The linea alba is divided longitudinally in the midline from the umbilical ring to as far cephalad as necessary to allow easy delivery of the pyloric tumour

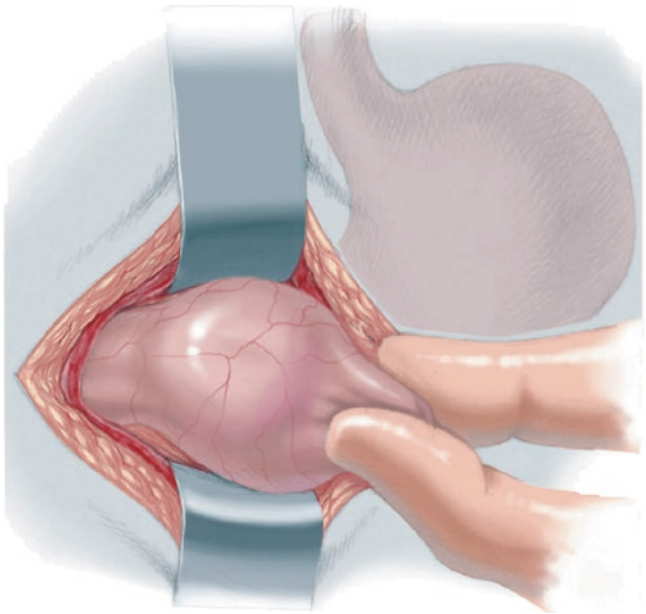

Fig. 22.2 The stomach is identified, is grasped proximal to the pylorus with a noncrushing clamp, and is brought through the wound. The greater curvature of the stomach then can be held in a moist gauze swab, and the pylorus can be delivered through the wound with traction inferiorly and laterally. Grasping the duodenum or pyloric tumour directly by forceps should be avoided, as it often results in serosal laceration, bleeding, or perforation

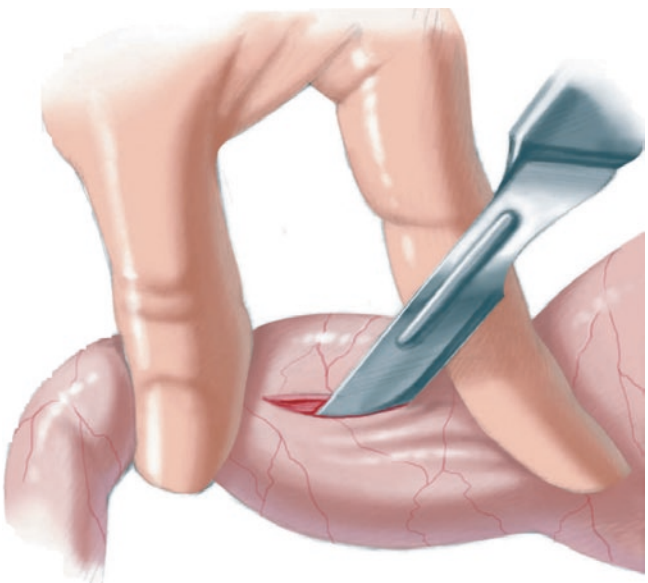

Fig. 22.3 The pylorus is held with surgeon's thumb and forefinger to stabilize and assess the extent of hypertrophied muscle. A seromuscular incision is made over the avascular area of the pylorus with a scalpel, commencing 1 or 2 mm proximal to the prepyloric vein along the gastric antrum. The incision should go far enough onto the gastric antrum (at least 0.5–1.0 cm from the antropyloric junction, where the muscle is thin)

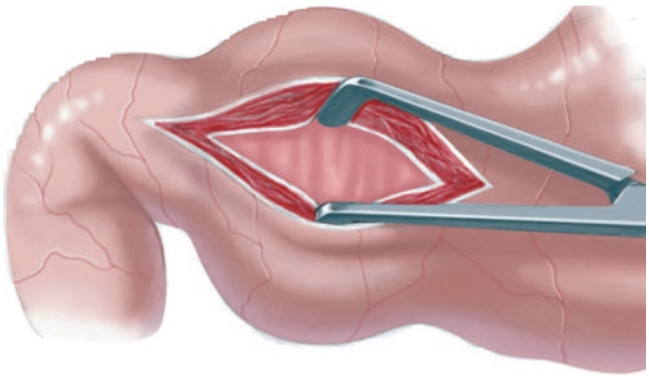

Fig. 22.4 The scalpel handle is used to further split the hypertrophied muscle down to the submucosal layer. Then the pyloric muscle is spread widely. The spreader is placed at the midpoint of the incision line and the muscle is spread perpendicularly; spreading must be continued proximally and distally. Gentle spreading is required to obtain a complete myotomy. Mucosal tears are most common at the pyloroduodenal junction because of the attempt to split all remaining muscle fibres. To reduce the risk of mucosal tears, care should be taken when spreading the pyloric muscle fibres at the duodenal end

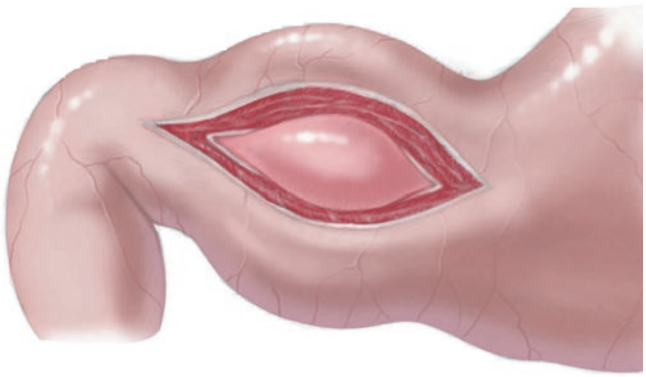

Fig. 22.5 Loose prolapsing of intact mucosa is evidence of a satisfactory myotomy. To test the mucosal injury, the stomach is inflated through the nasogastric tube, and passage of air through the pylorus to the duodenum is confirmed. Then the pylorus is dropped back into the abdomen. Bleeding from the myotomy edge or submucosal surface is frequently seen, but it is generally venous and always stops after returning the pylorus to the abdominal cavity. The posterior rectus fascia and peritoneum are approximated with a running 4/0 absorbable suture material, and the anterior fascia is closed with 5/0 absorbable suture material

22.3.2 Operative Procedure: Laparoscopic Pyloromyotomy

Figures 22.6, 22.7, and 22.8 show the procedure for laparoscopic pyloromyotomy (LP).

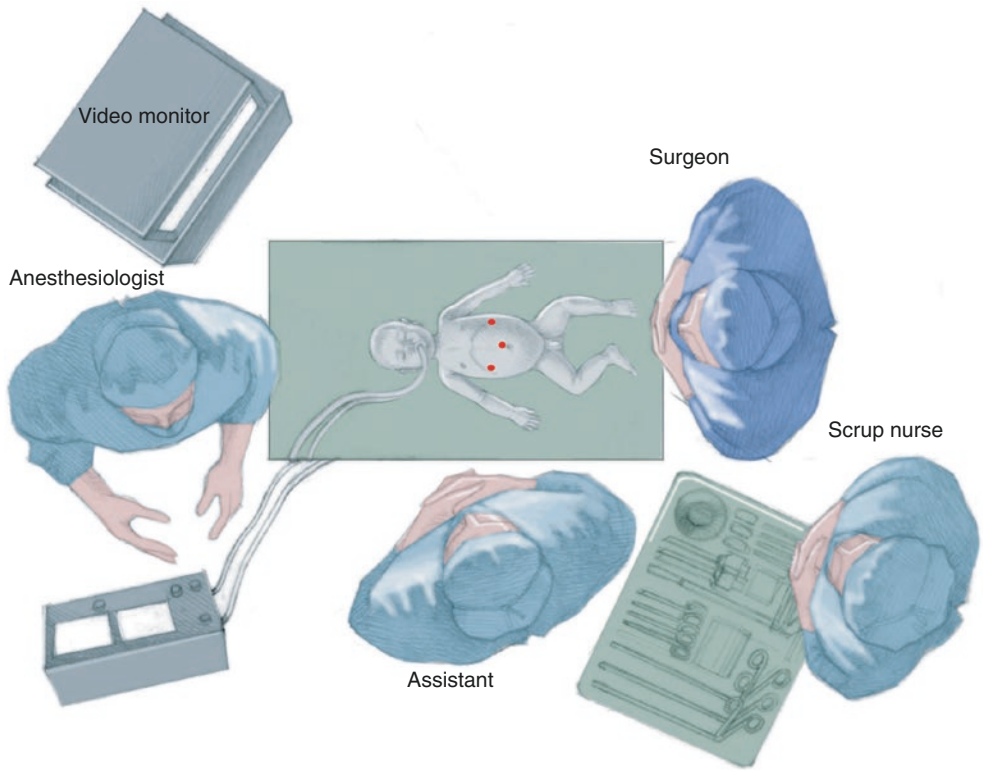

Fig. 22.6 For the laparoscopic procedure, the patient is placed in the supine position at the end of the operating table (or 90° to the anaesthesiologist). The video monitor is placed at the head of the table, and the surgeon stands at the end of the table with the assistant to the patient's right. The abdomen is scrubbed and draped in a sterile fashion. Attention must be paid to ensure the appropriate preparation of the umbilicus. The access sites are injected with local anaesthetic (0.25% bupivacaine) with epinephrine, which is used to reduce the postoperative pain and reduce the risk of bleeding from the stab wound. The author prefers an open procedure for insertion of the primary port. A curvilinear supraumbilical incision (4.0–5.0 mm) is made and carried down to the peritoneal cavity. At the level of the umbilical fascia, 4/0 absorbable suture material is placed circumferentially to anchor the port; it will also be used for closure of the peritoneal cavity after the laparoscopic pyloromyotomy is completed. Intra-abdominal pressure is maintained at 8 mm Hg, and the insufflation rate is set at 0.5 L/min. In the right midclavicular line just below the costal margin (just above the liver edge), a no. 11 scalpel blade is used to make a 2- to 3-mm stab incision under direct vision. Also using the no. 11 scalpel blade, a second stab incision is made under direct vision, just below the costal margin in the left midclavicular line. An atraumatic grasper is placed directly through the right upper quadrant stab wound and is used to retract the inferior border of the liver superiorly and expose the hypertrophic pylorus. A retractable myotomy knife (retractable arthrotomy knife or Endotome) is inserted directly through the left stab wound. Working ports are usually not necessary and instruments are directly introduced through these stab wounds

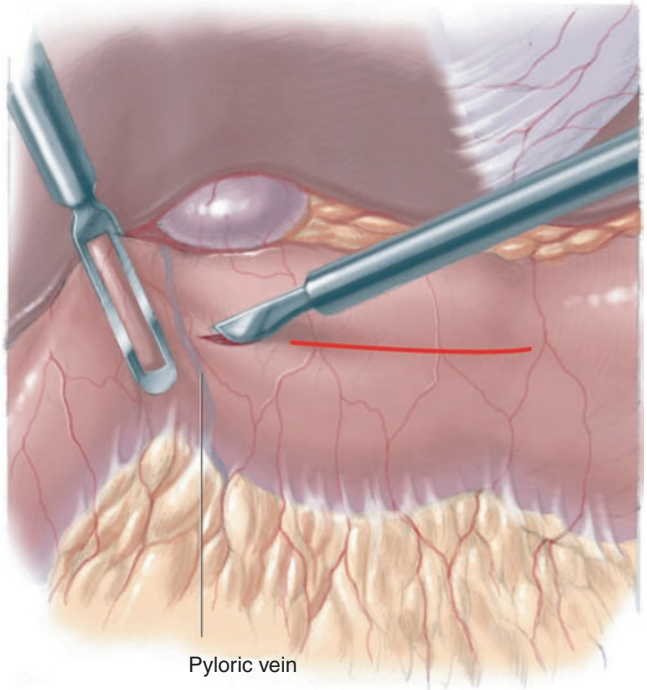

Fig. 22.7 The working instruments (the retractable myotomy knife and atraumatic laparoscopic grasper) are used to assess the extent of the hypertrophied pylorus by palpating the margins of the pylorus as one would do with thumb and forefinger in the open procedure. The duodenum is then grasped just distal to the pyloric vein (pyloroduodenal junction) and retracted using the atraumatic grasper to expose the avascular surface of the hypertrophic pylorus. In positioning the pylorus for myotomy, lateral and slightly anterocephalad retraction of the distal pylorus achieves excellent exposure of the avascular surface of the hypertrophic pylorus. This manoeuvre also exposes the proximal margin of hypertrophied muscle that is seen as a deep fold in the wall of the stomach. A seromuscular incision is made over the hypertrophic pylorus with the retractable myotomy knife, commencing 1 to 2 mm proximal to the pyloroduodenal junction and extending to the gastric antrum. The incision should go far enough onto the antrum (at least 0.5–1.0 cm proximal to antropyloric junction). Care must be taken at this stage that this incision is deep enough to allow the insertion of the pyloric spreader blades; it must penetrate the pyloric muscle somewhat deeper than is usual with the conventional open procedure

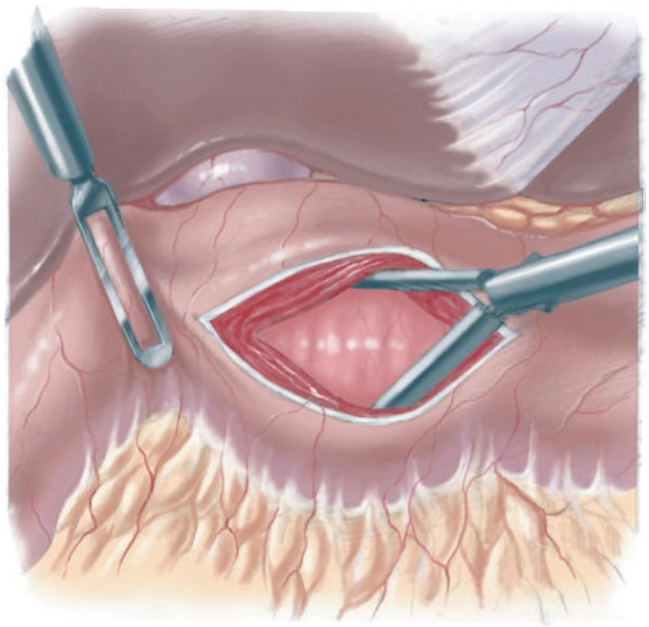

Fig. 22.8 After the muscle is incised, the blade is then retracted and the sheath of the knife is used to further split the hypertrophied muscle fibres, as the scalpel handle is used in the open procedure, until mucosa is visualized. The retractable myotomy knife is removed and a laparoscopic pyloromyotomy spreader is introduced into the abdominal cavity directly through the left stab wound to complete the pyloromyotomy. The spreader is placed in the midpoint of the seromuscular incision line and the muscle is spread perpendicularly. Once the initial spread reaches the mucosa, spreading must be continued proximally and distally. Pushing the spreader towards the mucosa or rapid spreading can result in mucosal tear. To avoid mucosal tears, the spreader should not be placed at the proximal and distal edges of the incisional (myotomy) line. To test for mucosal injury, the stomach is inflated through the nasogastric tube (160–180 mL), as is usually done in open techniques. Bulging of the mucosal layer with no evidence of defect should be confirmed. Greenish or yellowish fluid at the myotomy area is a sign of mucosal tear. After the successful myotomy, the instruments are withdrawn under direct vision and the pneumoperitoneum is evacuated. The nasogastric tube is also removed after completing the surgery. The umbilical fascia is reapproximated with the 4/0 absorbable suture material, which is already in place, and the skin of all the wounds is reapproximated with skin adhesive tapes

22.4 Complications

Mortality associated with pyloromyotomy is rare today. Early diagnosis and proper perioperative management reduces complications. In spite of these advances, there remains about an 8–10% incidence of associated perioperative morbidity such as perforation, wound infection, and wound dehiscence.

In an open procedure, essentially right upper quadrant incision and circumumbilical incision, manipulation of and tension on the pylorus to deliver it through the wound can induce oedema in the muscle layer, mucosal swelling, and occasionally serosal laceration. A laparoscopic pyloromyotomy (LP) is a less traumatic operation. The tolerance of an early feeding regimen after LP confirms the lack of trauma to the pylorus during the procedure, which we feel is the most important benefit of LP. The use of 3.0-mm instruments improves cosmesis.

Suggested Reading

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