Infantile pyloric stenosis is one of the most common gastrointestinal abnormalities presenting in the first 6 months of life. Although the definitive treatment for this condition is surgical pyloromyotomy, it is important to realize that pyloric stenosis is a medical and not a surgical emergency. Effective pre-operative rehydration and correction of acid/base abnormalities is the primary treatment goal and is the main factor contributing to the present-day low operative mortality. Recent studies quote a mortality of less than 0.3% compared with that of 10% in the early years of the 20th century.

**Incidence and aetiology**

The condition has a polygenic mode of inheritance. The incidence is higher in the offspring of affected parents and more common in autumn and spring. White infants are more likely than black or Asian infants to develop pyloric stenosis and there is a male predominance of approximately 4:1. World-wide incidence may approach 3 per 1000 live births with large regional variations.

Various theories have been proposed for the aetiology of infantile pyloric stenosis. Hypoganglionosis or immaturity of ganglion cells within the pyloric muscle, infection (*Helicobacter pylori*), and hypergastrinaemia with pylorospasm have been proposed. The definitive aetiology remains unknown.

**Table 1** Clinical signs of dehydration in infants

<table>
<thead>
<tr>
<th>Severity</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fluid loss (% body weight)</td>
<td>5%</td>
<td>10%</td>
<td>15%</td>
</tr>
<tr>
<td>Anterior fontanelle</td>
<td>Normal</td>
<td>Sunken</td>
<td>Markedly depressed</td>
</tr>
<tr>
<td>Skin turgor</td>
<td>Normal</td>
<td>Decreased</td>
<td>Greatly decreased</td>
</tr>
<tr>
<td>Mucous membrane</td>
<td>Moist</td>
<td>Dry</td>
<td>Very dry</td>
</tr>
<tr>
<td>Eyes</td>
<td>Normal</td>
<td>Sunken</td>
<td>Markedly sunken</td>
</tr>
<tr>
<td>Pulse</td>
<td>Normal</td>
<td>Increased</td>
<td>Greatly increased and feeble</td>
</tr>
<tr>
<td>Respiration*</td>
<td>Normal</td>
<td>Tachypnoea</td>
<td>Rapid and deep</td>
</tr>
<tr>
<td>Urine output (ml kg⁻¹ h⁻¹)</td>
<td>&lt; 2</td>
<td>&lt; 1</td>
<td>&lt; 0.5</td>
</tr>
</tbody>
</table>

*Respiratory signs may be altered in the presence of metabolic alkalosis.

**Key points:**

- Pyloric stenosis is a medical and not a surgical emergency
- Hypovolaemia and biochemical abnormalities should be corrected before anaesthesia
- The metabolic picture is a chloride-responsive hypochloraemic, hypokalaemic, hyponatraemic metabolic alkalosis
- Nasogastric tube aspiration before induction of anaesthesia is essential to minimize the risk of regurgitation and aspiration
- Postoperative pain is usually well controlled with rectal paracetamol and wound infiltration with bupivacaine; avoid intraoperative opioids

**Signs and symptoms**

Clinical manifestations of pyloric stenosis usually present between the 3rd and 5th weeks of life, but earlier or later presentation is also encountered. It is usually a disease of full-term infants and only a small percentage of cases are found in premature infants. The infant presents with a history of progressive non-bilious vomiting which may become projectile and blood-tinted. Concurrent constipation is common. The infant may be mildly jaundiced and this is attributed to glucuronyl transferase deficiency which develops as a consequence of starvation.

Associated abnormalities are found in 6–20% of the infants and comprise oesophageal atresia, congenital cardiac anomalies, Hirschprung’s disease, intestinal malrotation, anorectal anomalies, minor renal anomalies and inguinal hernias.

There is loss of weight or failure to gain weight and the infant is ravenously hungry and attempts to feed avidly. On examination, varying degrees of dehydration, loss of skin turgor and apathy may be present (Table 1). Capillary refill time may be prolonged (>2 s) when dehydration is severe, with decreased circulating volume and compensated or impending circulatory failure. The limb is examined by raising it slightly above the heart level (to assess arteriolar rather than venous capillary return) and compressing the skin for 5 s. Return of blood is indicated...
Infantile pyloric stenosis

by disappearance of blanching normally in less than 2 s. Heart rates in infants are variable but should not normally exceed 160 min\(^{-1}\). Assessment of fontanelle tension, ocular tension and skin turgor are subjective. It should be unusual in UK practice to see such severe signs of dehydration as a result of pyloric stenosis.

The upper abdomen is distended and gastric peristaltic waves may be seen during feeding with an appearance resembling a golf ball moving under the skin from left to right. An olive-sized mass may be felt to the right of the umbilicus more readily after the infant has vomited. Blood tests reveal hypochloaemic, hypokalaemic, hyponatraemic metabolic alkalosis.

**Surgical management**

The diagnosis usually can be made on clinical presentation alone, but ultrasound scan, endoscopy and barium meal are sometimes used. The definitive treatment of infantile pyloric stenosis remains surgical pyloromyotomy. Described by Ramstedt in 1912, this involves division of the pyloric muscle leaving the mucosa intact. A subcostal, upper midline or, more recently, a peri-umbilical incision is used. The pyloric tumour is delivered by gentle traction on the stomach and an anteroposterior incision is made along the axis of the pylorus. The pyloric muscle is then widely split down to the submucosa. The end result should be an intact bulging submucosa from the duodenal fornix to the gastric antrum. Complications of surgery are few, but mucosal perforation may occur. Pyloromyotomy is now performed as a laparoscopic procedure in some centres.

**Pathophysiology**

Gastric outlet obstruction initially produces regurgitation and eventually vomiting. As a result of vomiting, the infant loses gastric acid, water and a variable amount of Na\(^+\) and K\(^+\). Normally, as the gastric acid passes into the duodenum, it is neutralised by pancreatic HCO\(_3\)\(^-\). The acid (H\(^+\)) and the HCO\(_3\)\(^-\) ions are generated by the activity of the gastric parietal cells. H\(^+\) ions are secreted into the gastric lumen while HCO\(_3\)\(^-\) reaches the extracellular fluid (ECF) and is regenerated in the duodenal secretions. If vomiting occurs with an intact communication between stomach and duodenum, both H\(^+\) and HCO\(_3\)\(^-\) are lost with a neutral effect on acid/base balance. However, in pyloric stenosis there is loss of H\(^+\) and a consequent net increase in plasma HCO\(_3\)\(^-\) concentration. This increased HCO\(_3\)\(^-\) presented in the glomerular filtrate in the early stages of the disease overwhelms the resorptive capacity of the proximal convoluted tubule (PCT). The resulting loss of HCO\(_3\)\(^-\) in the urine makes it alkaline with a pH >7.

The kidney then attempts to conserve Na\(^+\) (in the face of ECF volume depletion) by stimulating aldosterone secretion. Hence, Na\(^+\) ions are retained at the cost of significant kaliuresis. H\(^+\) in the glomerular filtrate is also exchanged for K\(^+\) in an unsuccessful effort to maintain normal serum pH. These factors, along with the loss of K\(^+\) during vomiting and the extracellular to intracellular shift of K\(^+\) because of plasma alkalosis, result in significant hypokalaemia.

Loss of Cl\(^-\) during vomiting results in hypochloaemia. The kidney tries to conserve Cl\(^-\) by maximising Cl\(^-\) absorption, but there is insufficient Cl\(^-\) in the glomerular filtrate to absorb along with Na\(^+\) and in exchange for HCO\(_3\)\(^-\). Sodium re-absorption takes place in exchange for K\(^+\) across the distal renal tubules until hypokalaemia forces Na\(^+\) to exchange preferentially for H\(^+\) resulting in the paradox of acidic urine during alkalosis in pyloric stenosis. Only when Cl\(^-\) status is restored, is there sufficient Cl\(^-\) concentration in the glomerular filtrate to permit tubular exchange with HCO\(_3\)\(^-\) and excretion of bicarbonate in the urine, which restores pH towards normal.

Measurement of low or absent urinary Na\(^+\) concentration is normally diagnostic of hypovolaemia. An exception occurs when Na\(^+\) is excreted with another anion. In pyloric stenosis, Na\(^+\) loss in urine occurs as Na\(^+\) is delivered along with HCO\(_3\)\(^-\) to the distal tubule and collecting ducts, so urinary sodium concentration is unreliable in this condition.

However, since all the Cl\(^-\) is re-absorbed in exchange for HCO\(_3\)\(^-\), urine Cl\(^-\) results are more relevant than serum electrolytes when assessing volume status in infants awaiting pyloromyotomy. Urine Cl\(^-\) concentration >20 mmol l\(^{-1}\) suggests that ECF volume status has been corrected. Plasma potassium is not a reliable indicator of total body potassium. Correction of potassium depletion is essential to achieve complete correction of alkalosis. Plasma Cl\(^-\) concentrations of >106 mmol l\(^{-1}\) are necessary to ensure that alkalosis has been corrected in the infant.

In summary, the metabolic picture is a chloride-responsive hypochloaemic, hypokalaemic, hyponatraemic metabolic alkalosis. The urine is alkaline at first with little or no chloride. In later stages, the urine may become acidic (paradoxical aciduria) because of dehydration, more efficient HCO\(_3\)\(^-\) absorption by the PCT, re-absorption of Na\(^+\) in exchange for H\(^+\), and also because lactic acidosis and starvation ketosis become significant factors. Respiratory compensation for alkalosis is ineffective.
Correction of acid/base and fluid abnormalities

Infants with pyloric stenosis should not be considered for surgery unless the metabolic abnormalities have been corrected and the ECF volume restored. This may take 24–48 h or more. An intravenous line is first secured and blood samples are analysed for measurement of baseline electrolytes and haemoglobin concentration. A nasogastric tube (size 8 or 10 Fr) is inserted, and gastric washouts are performed at least 4 hourly using saline until the aspirate is clear. Two hourly aspirations are carried out thereafter. A capillary blood gas sample is taken for measurement of $\text{HCO}_3^-$. The degree of dehydration is then established.

For severe dehydration (>15% loss of body weight) with severe alkalaemia and impending circulatory failure, a bolus of 20 ml kg$^{-1}$ of normal saline (0.9%) or a colloid (Haemaccel® or Gelofusine®) should be given initially to correct intravascular fluid deficits. For mild-to-moderate dehydration and moderate alkalaemia ($\text{HCO}_3^- 32–42$ mmol l$^{-1}$), fluid replacement is achieved with glucose saline (5% glucose, 0.45% saline with 10 mmol of potassium chloride per 500 ml bag) given at the rate of 6–8 ml kg$^{-1}$ h$^{-1}$. Nasogastric losses are replaced with normal saline, ml for ml. Repeated capillary and venous blood samples are used to guide therapy pre-operatively.

Once targets are approached or achieved (Table 2), glucose saline (4% glucose, 0.18% saline) can be given as maintenance intravenous fluid at the rate of 4 ml kg$^{-1}$ h$^{-1}$ along with potassium supplementation (10 mmol of potassium chloride per 500 ml bag). The importance of up-to-date electrolyte measurements and capillary $\text{HCO}_3^-$ is stressed. Infants scheduled for surgery should have had these measurements performed within the previous 6 h. Failure to correct hypokalaemia can lead to intra-operative cardiac arrhythmias and persistent dehydration will result in intra-operative hypotension. Similarly, failure to correct alkalaemia can result in postoperative apnoeas, particularly if combined with intra-operative hyperventilation and opioid administration.

Anaesthetic management

Once the infant arrives in the operating room, a pulse oximeter, ECG and blood pressure cuff are placed and monitoring started. The infant is adequately wrapped and kept warm. Regular nasogastric tube suction pre-operatively should ensure a milk-free stomach. However, some gastric secretions may persist. Accordingly, nasogastric tube aspiration with the patient in the right and left lateral and head down positions is imperative before induction. Saline infiltration and gastric lavage may also be performed at this time, in an attempt to minimise the risk of regurgitation and tracheal aspiration during induction. Both intravenous and inhalational induction are safe techniques following this manoeuvre (although the latter should rarely be necessary as most patients will arrive at the operating room with intravenous access already established). A combined oesophageal stethoscope and temperature probe is a useful monitor and can be placed after tracheal intubation.

The patency of the intravenous line is checked with a saline flush and atropine 20 µg kg$^{-1}$ is given to obtund vagal reflexes during intubation or gastric manipulation. Pre-oxygenation is then followed by inhalational induction with halothane or sevoflurane. Alternatively, intravenous induction of anaesthesia is performed with thiopental (5–7 mg kg$^{-1}$) and succinylcholine (2 mg kg$^{-1}$). If gastric emptying has been assured, atracurium or another non-depolarising neuromuscular blocker may be employed in preference in experienced hands. The trachea can usually be intubated with a 3.5 mm uncuffed tracheal tube, but a smaller sized tube must be available if required. The endotracheal tube is secured and anaesthesia is maintained with pulmonary ventilation of oxygen, nitrous oxide and halothane, sevoflurane or isoflurane. Awake tracheal intubation is not superior to anaesthetised, paralysed intubation in maintaining adequate oxygenation and heart rate or in reducing complications. This practice should be abandoned.

The use of a muscle relaxant improves surgical conditions and also permits the inspired concentration of inhalation agents to be reduced. There is also a decrease in the risk of perforation of the pyloric mucosa during surgery. Atracurium (0.3–0.5 mg kg$^{-1}$) is appropriate and IPPV is achieved with manual or mechanical ventilation, ensuring the avoidance of high minute volumes and hyperventilation. Glucose saline (4% glucose, 0.18% saline) is continued at the rate of 4 ml kg$^{-1}$ h$^{-1}$ as maintenance fluid. Particular attention is required to keep the infant warm. A heat and moisture exchanger, a warm under-mattress and the occasional use of warm air device should ensure that the infant does

### Table 2 Targets for resuscitation

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Target</th>
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<tbody>
<tr>
<td>Serum $\text{Cl}^-$</td>
<td>$\geq 106$ mmol l$^{-1}$</td>
</tr>
<tr>
<td>Serum $\text{Na}^+$</td>
<td>$&gt; 135$ mmol l$^{-1}$</td>
</tr>
<tr>
<td>Serum $\text{HCO}_3^-$</td>
<td>$\leq 26$ mmol l$^{-1}$</td>
</tr>
<tr>
<td>Urine $\text{Cl}^-$</td>
<td>$&gt; 20$ mmol l$^{-1}$</td>
</tr>
<tr>
<td>Urine output</td>
<td>$&gt; 1$ ml kg$^{-1}$ h$^{-1}$</td>
</tr>
</tbody>
</table>
not become hypothermic despite the short duration of surgery. Antibiotic prophylaxis should be given to infants with a cardiac lesion.

Analgesia comprises a combination of systemic non-opioid analgesia and local anaesthesia. Opioids are rarely given and may be contra-indicated. The combination of corrected peripheral venous alkalosis, persistent and slower correction of CSF alkalosis, intra-operative hyperventilation and opioids can lead to postoperative apnoea.

Paracetamol is used as the main systemic analgesic and postoperative pain can be well controlled with rectal paracetamol given in a loading dose of 30–40 mg kg\(^{-1}\) followed postoperatively by 15–20 mg kg\(^{-1}\) rectally to ensure a maximum daily dose of approximately 90 mg kg\(^{-1}\). Wound infiltration with 0.25% bupivacaine (2 mg kg\(^{-1}\)) delays the time to administration of first postoperative analgesia. Injection of 0.25% bupivacaine with epinephrine (adrenaline) 1:200,000 under the anterior rectus sheath at the wound edge has been shown to reduce heart rate and respiratory rate postoperatively.

It may be necessary to inject air through the nasogastric tube after the pyloromyotomy to help the surgeon test the integrity of the pyloric mucosa. Care is taken to ensure that the air is injected correctly into the nasogastric tube. After completion of the procedure and wound infiltration with bupivacaine, muscle relaxation is reversed with a combination of neostigmine (50 µg kg\(^{-1}\)) and glycopyrronium (10 µg kg\(^{-1}\)) or atropine (20 µg kg\(^{-1}\)). The trachea is extubated when the infant is fully awake, breathing spontaneously and when upper airway reflexes have returned fully.

Postoperatively, supplemental oxygen is administered and the infant is placed on an apnoea monitor for 6–12 h. Feeding is usually commenced 12 h postoperatively. At first, 20 ml of a clear fluid such as dioralyte is given 2 hourly. If tolerated well, 40 ml of half-strength milk is given 3 hourly before returning to full feeds. Earlier return to feeding is associated with a high incidence of vomiting. It is important at this time to administer maintenance intravenous fluids in the form of glucose-saline (4% glucose, 0.18% saline) and watch out for hypoglycaemia as a result of hepatic glycogen depletion. The signs of hypoglycaemia are lethargy and irritability progressing to convulsions and cardiac arrest. Hypoglycaemia is treated promptly with intravenous 10% glucose (2 ml kg\(^{-1}\) over 3 min as a bolus followed by an infusion of 6 ml kg\(^{-1}\) h\(^{-1}\)). The infant is usually discharged from the hospital 36–48 h postoperatively.

**Key references**

Anderson BJ. What we don’t know about paracetamol in children. *Paediatr Anaesth* 1998; 8: 451–60


See multiple choice questions 52–54.