Paediatric disorders

Hypertrophic pyloric stenosis of infancy

Aetiology

The incidence of this condition is approximately three cases per 1000 births. It is four times as common in males as in females and the aetiology is unknown. It may be that the condition is analogous to achalasia of the oesophagus in which there is a failure of the pylorus to relax, leading to the muscular hypertrophy. In some cases there seems to be a familial association. In such families the mother has suffered from the condition in 50 per cent of cases, and 10 per cent of male siblings and 2 per cent of female siblings are affected.

Pathology

The classical feature is that the musculature of the pylorus and adjacent antrum is grossly hypertrophied, the hypertrophy being maximum in the pylorus itself. The mucosa is compressed such that only a probe can be inserted.

Clinical features

Characteristically it is a first-born male child that is most commonly affected. The condition is most commonly seen at 4 weeks after birth ranging from the third week to, on rare occasions, the seventh. Vomiting is the presenting symptom that after 2—3 days becomes forcible and projectile. The child vomits milk and no bile is present. Immediately after vomiting the baby is usually hungry. Weight loss is a striking feature and rapidly the infant becomes emaciated and dehydrated. Diagnosis can usually be made with a test feed. This may produce characteristic peristaltic waves that can be seen to pass across the upper abdomen. At the same time, using a warm hand, the abdomen is palpated to detect the lump.

Imaging

Ultrasonography is the investigation of choice as it can, without difficulty, detect the classical features in the pyloric canal. Contrast radiology is not now necessary.

Differential diagnosis

The common conditions from which pyloric stenosis must be differentiated are gastro-oesophageal reflux, feeding problems, urinary tract infection and raised

intracranial pressure. The condition cannot normally be confused with duodenal atresia or intestinal obstruction because of the absence of bile in the milk vomit.

Treatment

Following diagnosis the first concern is to correct the metabolic abnormalities. Essentially this is the same situation that pertains in adults with the patient being dehydrated, with low sodium, chloride and potassium, and a metabolic alkalosis. The child should be rehydrated with dextrose—saline and potassium (2.5 per cent dextrose plus 0.45 per cent sodium chloride plus 1 g of potassium chloride per 500 ml of fluid). This will restore the infant's clinical condition and electrolytes to normal. Following this operation is required. Conservative treatment has little place in the management of this condition as with appropriate surgical treatment recovery is virtually 100 per cent.

Ramstedt's operation للأطلاع

In preparing the child for operation it is important that the stomach is emptied and washed out with saline, and that hypothermia is avoided. To achieve this the patient is encased in cotton wool allowing exposure of the upper abdomen. Operation is performed under general anaesthesia, although it is possible to perform the procedure under a local anesthetic. The skin is opened through a transverse incision placed in the upper abdomen over the right rectus sheath, which is opened in the same line. The rectus muscle is then split along the line of its fibres and the posterior rectus sheath opened in the line of the skin incision. The hypertrophied pylorus is delivered and rotated so that its superior surface comes into view. Thus, the least vascular portion can be selected for incision. To ascertain the distal limit of the hypertrophy the surgeon invaginates the duodenum with the index finger. The incision is made through the serosa only and from this point along the whole length of the pylorus and, importantly, the distal antrum. The hypertrophied pylorus has the consistency of an unripe pear, hence splitting the muscle coats can be accomplished by blunt dissection. On separating the edges with artery forceps the pyloric mucosa bulges into the cleft which has been made in the muscle. Great care is taken not to penetrate the mucosa. When this injury occurs it is almost always in dividing the most distal part of the constricting fibres which are in the vicinity of the duodenal fornix. To be sure that there is no perforation some air is squeezed from the stomach into the duodenum. If a perforation has occurred it is closed and a piece of omentum placed over the closure. Haemostasis should be meticulous.

After operation the nasogastric tube can be removed and feeding commenced on the morning after operation. If the infant manages to feed without difficulty

it can be discharged early from hospital. If the mucosa is inadvertently opened it is wise to delay feeding for 48 hours and to retain the child in hospital longer.

Complications of operation

Postoperative pyrexia is common and usually treated with paracetomol elixir. ***Wound disruption*** is rare and is more liable to occur in emaciated subjects. The incidence of ***wound infection*** is around 5per cent, and 1 per cent will suffer wound disruption.

Duodenal atresia

This occurs at the point of fusion between the foregut and midgut, and therefore lies in the neighborhood of the ampulla of Vater. There is a diaphragm, which is usually complete, across the duodenum at this point and the condition is frequently accompanied by other defects. The diagnosis is now made antenatally in most cases through the use of ultrasound. This shows the characteristic appearance of a dilated stomach and first part of the duodenum (double bubble). The child vomits from birth and the vomitus is bile stained. The differential diagnosis includes high intestinal obstruction. Occasionally, however, the diaphragm may be proximal to the ampulla and in these circumstances the condition can be confused with pyloric stenosis, although in pyloric stenosis vomiting does not start from birth. Treatment is by the operation of the duodenoduodenostomy in which the dilated proximal duodenum is anastomosed to the atrophic distal duodenum.

mugdad fuad