

SUMMARY

Congenital hypertrophic pyloric stenosis (CHPS) is an important and interesting disease : important, because it is common with an incidence of three per 1000 live births, it affects healthy babies and an effective surgical cure is available; interesting, because the cause is still mystery despite much speculation and research over a period of nearly 100 years since the condition became widely recognized.

The average age of onset of symptoms is 3.5 weeks and most cases occur around this age though more rarely symptoms may start at birth or be delayed until 3 months of age. The natural history is remission over a period of 2.5 months and the disease is not a significant problem after the first few months of life.

CHPS is three to four times more common in boys than in girls. There is no doubt that there is a strong hereditary factor in the aetiology of CHPS. There are many instances of high familial incidence.

The cause of CHPS is still unknown. Many theories of pathogenesis have been proposed over the years.

From the pathological point of view, the pylorus is increased in length and diameter to an average size of 3 cm X 1.5 cm, the stomach is dilated and the antrum hypertrophic. The circular muscle layer is up to four times thicker than normal, the longitudinal muscle layer on the other hand is rather attenuated. There is a fibrous block or septum between the circular muscle of the pylorus and that of the duodenum, whereas the longitudinal muscle forms a continuous layer outside this. Leucocyte infiltration is prominent and there is an increase in connective tissue. Ganglion cells are more prominent in Auerbach's than in Meissner's plexus.

The biochemical disorder is so characteristic as to be almost diagnostic of CHPS. As distinct from other causes of vomiting, only gastric secretions are lost. The result is large losses of H^+ and Cl^- and smaller losses of Na^+ and K^+ , reflected mainly as hypochloraemia and alkalaemia.

The cardinal symptom is vomiting which usually starts between 3 and 4 weeks of age. The vomiting is forceful and projectile. It usually occurs at the end

of the feed but may happen during or sometime afterwards. On examination, visible gastric peristalsis may be seen.

The cardinal physical sign is palpation of the pyloric tumour during a test feed and it should be the clinician's aim to confirm the diagnosis by this means if possible.

If the clinical features are characteristic of CHPS, and a pyloric mass is palpable, no further investigations are necessary. Plain abdominal radiographs are indicated if intestinal obstruction is suspected. Ultrasound may be helpful in confirming the diagnosis. Contrast radiology is essential if gastro-oesophageal reflux, or malrotation are being considered in the differential diagnosis.

Nowadays there are few advocates of medical treatment. Its success depends upon meticulous nursing care and refeeding after each vomit, aided by the anticholinergic drugs methylscopolamine nitrate (Skopyl) or atropine methylnitrate 1 : 10,000 of water (Eumydrin). Surgical treatment in the form of

Ramstedt's pyloromyotomy is considered to be the treatment of choice, except in those cases in which the length of history and good condition of the child indicate that the disease is spontaneously resolving.

Recently, extramucosal pylorotomies can be successfully performed by laparoscopy since May 1990 .