Congenital hypertrophic pyloric stenosis

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SUMMARYCongenital hypertrophic pyloric stenosis (CHPS) isan important and interesting disease important,because it is common with an incidence of three per1000 live births, it affects healthy babies and aneffective surgical cure is available; interesting,because the cause is still mystery despite muchspeculation and research over a period of nearly 100years since the condition became'widely recognized.The average age of onset of symptoms is 3.5 weeksand most cases occur around this age though more rarelysymptoms may start at birth or be delayed until 3months of age. The natural history is remission over aperiod of 2.5 months and the disease is not ·asignificant problem after the first few months of life.CHPS is three to four times more common in boysthan in girls. There is no doubt that there is a strongheredítary factor in the aetiology of CHPS. There aremany instances of high familial incidence.The cause of CHPS is still unknown. Many theoriesof pathogenesis have been proposed over the years.149from the pathological point of view, the pylorusis increased in length and diameter to an average sizeof 3 cm X 1.5 cm, the stomach is dilated and the antrumhypertrophic. The circular muscle layer is up to fourtimes thicker than normal, the lonqitudinal musclelayer on the other hand is rather attenuated. There isa fibrous block or septum between the circular muscleof the pylorus and that of the duodenum, whereas thelongitudinal muscle forms a continuous layer outsidethis. Leucocyte infiltration is prominant and there isan increase in connective tissue. Ganglion cells aremore prominant in Auerbach's than in Meissner'splexus.The biochemical disorder is so characteristic asto be almost diagnostic of CHPS. As distinct from othercauses of vomiting, only gastric secretions are lost.The result is large losses of H+ and cl and smallerlosses of Na+ and K+, reflected mainly ashypochloraemia and alkalaemia.The cardinal symptom is vomiting which usuallystarts between 3 and 4 weeks of age. The vomiting isforceful and projectile. It usually occurs at the end150of the feed but may happen during or sometimeafterwards. On examination, visible gastric peristalsismay be seen.The cardinal physical sign is palpation of thepyloric tumour during a test feed and it should be theclinician's aim to confirm the diagnosis by this meansif possible.If the clinical features are characteristic ofinvestigations are necessary. Plainno furtherabdominalCHPS, and a pyloric mass is palpable,radiographs are indicated if intestinal obstruction issuspected. Ultrasound may be helpful in confirming thediagnosis. Contrast radiology is essential if gastrooesophagealreflux, or malrotation are being consider~din the differential diagnosis.Nowadays there are few advocates of medicaltreatment. Its success depends upon meticulous nursingcare and refeeding after each vomit, aided by

theanticholinergic drugs methylscopolamine nitrate(Skopyl) or atropine methylnitrate 1 : 10,000 of water(Eurnyd r Ln l . Surgical treatment in the form of151Ramstedt’s pyloromyotomy is considered to be thetreatment of choice, except in those cases in which thelength of history and good condition of the childindicate that the disease is spontaneously resolving.Recently, extramucosal pylorotomies can besuccessfully performed by laparoscopy since May 1990.