Short reports

Neonatal small left colon syndrome

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SUMMARY Five patients with neonatal small left colon syndrome are described together with some early investigative studies on rectal biopsy material. Current reports on this condition are reviewed. The precise aetiology remains unknown, but vigilance and early referral are recommended in view of the several reports of colonic perforation.

Delay in diagnosis and treatment can result in serious complications in the otherwise benign neonatal small left colon syndrome. This condition was first distinguished from other forms of micro-colon by Davis *et al.* (1974), and subsequently reports have suggested a surgical orientation. The condition presents as a low obstruction of the large bowel, similar to Hirschsprung's disease in the newborn. There is failure to pass meconium, with tympanitic distention of the abdomen and bilious vomiting.

Contrast radiography, particularly with Gastrografin, shows a smooth, narrow sigmoid and descending colon, with proximal dilatation of the large bowel. However, the enema also promotes both the rapid evacuation of meconium and the early establishment of normal defecation. Many of the mothers of such babies have diabetes mellitus.

A group of 5 babies illustrates the practical management of the condition; 2 of them are described in detail.

Case reports

Case 1. A baby boy, weighing 2.65 kg, delivered normally at 37 weeks. There was no history of maternal diabetes. He failed to pass meconium and was transferred at 30 hours with tympanitic abdominal distention and bile-stained vomiting.

Plain x-rays of the abdomen showed dilated loops of small and large bowel, with multiple fluid levels consistent with a low obstruction of the large bowel (Fig. 1). A Gastrografin enema was performed to assist radiological diagnosis. This showed a smooth,

Fig. 1 Erect abdominal x-ray showing multiple fluid levels in dilated small and large bowel.

narrow descending and sigmoid colon with a marked 'transition zone' to distention at the splenic flexure (Fig. 2). The colon proximal to the splenic flexure was overfilled with air and meconium. These appearances are typical of the neonatal small left colon syndrome but simulate long-segment Hirschsprung's disease. The latter diagnosis was made in error, and a left transverse colostomy established. Subsequent investigations showed no evidence of Hirschsprung's disease and at 11 months the colostomy was closed. The infant has remained well since.

Case 2. A baby girl weighing 3.4 kg of a diabetic mother, delivered by forceps at 36 weeks' gestation. She was referred at 24 hours with failure to pass

Fig. 2 Gastrografin enema showing narrowed descending and sigmoid colon and transition zone at splenic flexure.

meconium, tympanitic abdominal distention, and bile-stained vomiting. Plain x-rays indicated a low colonic obstruction and a Gastrografin enema showed the typical appearances of the small left colon syndrome.

Evacuation of meconium followed rapidly and subsequent bowel function was normal. No further investigations were made and the baby has remained well.

Three other infants were seen in whom the history and progress were similar to Case 2, although none was known to have a diabetic mother. Further description of the clinical details is redundant but the results of investigations undertaken to exclude Hirschsprung's disease are of interest.

In Case 1 quantitative estimations of total cholinesterase activity were made in rectal biopsy specimens, using the method described by Boston *et al.* (1975). At 72 hours, the value was in the high normal range (21.8 rate units) and barely excluded Hirschsprung's disease but, when it was repeated at 2 weeks, the result was unequivocally normal (10.0 rate units). In 2 of the other patients, estimations were done for total cholinesterase and acetyl cholinesterase, using the method described by Dale *et al.* (1977). In one, the results were normal at 4 days, but in the other patient, in whom the quantity

of enzyme present was also normal, the ratio of acetyl cholinesterase to total cholinesterase was 82% which is consistent with Hirschsprung's disease. This estimation was performed at 3 days, but repetition was not justified in view of the patient's good progress.

Discussion

Sluggish or delayed passage of meconium in the newborn is well recognised, and there are various reasons for this condition. Two of the causes to be distinguished from the small left colon syndrome are the meconium plug syndrome (Clatworthy *et al.*, 1956), and long segment Hirschsprung's disease.

The small left colon syndrome is distinct from either as there is no meconium plug and the intrinsic neurohistology of the affected bowel is apparently normal. In the first description of the syndrome (Davis *et al.*, 1974) 20 infants were reported, 2 of whom were operated on as suspected of having Hirschsprung's disease. One other patient died soon after birth of hyaline membrane disease. After administration of a diagnostic barium enema, all the remaining 17 patients began and continued to evacuate their bowels normally. Eight of them had diabetic mothers and this high correlation has subsequently been confirmed (Davis and Campbell, 1975; Philippart *et al.*, 1975; Stewart *et al.*, 1977). Later reports have not agreed with the initial opinion that complications are rare. Philippart *et al.* (1975) and Stewart *et al.* (1977) reported that in 4 babies out of 12 there were perforations of the bowel. These perforations are often in the caecum and, in one baby, this was 6 days after a barium enema. For this reason vigilance is recommended before x-ray examination with contrast enemas is undertaken, and early referral of these patients to paediatric surgical centres for diagnosis and treatment is to be preferred.

The aetiology of the small left colon syndrome is obscure. No abnormality of meconium has so far been shown and it is unusual for a meconium plug to be passed (Clatworthy *et al.*, 1956; Ellis and Clatworthy, 1966).

Davis *et al.* (1974) reported that in biopsy material from 4 babies increased numbers of immature small cells were noted in the myenteric plexus, and these were present in both the narrow and dilated areas of colon. These findings were confirmed by Stewart *et al.* (1977) but not by Philippart *et al.* (1975), and none of the biopsy material now reported has shown any gross abnormality in the intramural ganglion cells and nerves, but studies of the cholinesterase suggest a temporary ganglion cell dysfunction.

Philippart *et al.* (1975) confined their attention to

the children of diabetic mothers and suggested that the problem in the infant results from impaired intestinal motility, possibly promoted by glucagon, rather than from an intraluminal plug. In several of their patients an element of obstruction persisted after complete evacuation of meconium and it has been shown that the small calibre of the left colon can persist for variable periods despite normal bowel activity (Davis *et al.*, 1974). Their hypothesis is that an increase in glucagon excretion occurs in babies in the perinatal period either secondary to hypoglycaemia associated with maternal diabetes in a complex way, or with stress. Glucagon is known to decrease bowel motility in the jejunum and sigmoid colon. Conversely hypoglycaemia can also increase bowel motility by vagal and sympathetic stimulation, and consequently Stewart *et al.* (1977) have proposed that the immature small intramural nerve cells are unable to respond to increased sympathetic stimulation. The results of further studies are awaited.

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Acute necrotising fasciitis due to streptococcal infection in a newborn infant

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SUMMARY A 3-day-old baby girl developed septicaemia, meningitis, and necrotising fasciitis due to group A β -haemolytic streptococcus, type M52, which was also cultured from the mother's cervix. Necrotising fasciitis is a severe infection of the skin and subcutaneous tissues with infarction, necrosis, and sloughing of the affected areas. Early recognition of this condition is essential so that appropriate treatment can be given.

Acute necrotising fasciitis is a rare but severe bacterial infection of the subcutaneous tissue and fascial planes, producing extensive destruction of tissues (Hammar and Wagner, 1977). It is often associated with blood stream invasion by the

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causative pathogen, and the mortality is high. The condition is familiar to the surgeon, but it appears to be rare in children (Wilson and Haltalin, 1973), and only 3 cases have been reported in newborn infants (Weinberger *et al.*, 1972; Ramamurthy *et al.*, 1977).

We report a newborn infant who developed septicaemia, meningitis, and necrotising fasciitis caused by group A β -haemolytic streptococcus.

Case report

A term baby girl of 2.7 kg was born to healthy non-related parents after a pregnancy complicated only by a heavy leucorrhoea during the 3rd trimester. The membranes ruptured spontaneously 14 hours before a normal delivery. The Apgar score at one minute