DOLICHOCOLON AND HIRSCHSPRUNG'S DISEASE

BY

ALAN MONCRIEFF, M.D., F.R.C.P., and T. V. CRICHLOW, B.Sc., D.M.R.E.

(From the Children's Department and Radiological Department, Middlesex Hospital)

Dolichocolon is the term given to a condition of a long and redundant sigmoid colon. As it occurs in childhood it is associated with obstinate constipation, occasionally a sort of 'overflow diarrhoea', and not infrequently acute obstruction from impaction or even volvulus. In general the prognosis is held to be good, for as the child grows the colon becomes relatively the correct length. No case has been found recorded in which the 'long colon' has developed into the 'megacolon' as described by Hirschsprung. This latter is usually congenital, with constipation and increasing abdominal distension. It is thought to be due to neuromuscular incoordination, especially in relation to the pelvic-rectal junction. The case here recorded is believed to show characteristics of both these abnormalities of the colon.

Clinical details

A. W., a male infant, was first admitted to Tindal House Emergency Hospital (E.M.S.), Aylesbury, on January 5, 1942, at the age of ten months. He was the first child of healthy parents who were not related. Birth was normal and he was breast-fed for four-and-a-half months, followed by a milk mixture with mixed feeding after six months. He cut his first tooth at eight months and at this age was taken to a medical practitioner because he had been constipated for six days, without pain, but associated with abdominal distension. There had been no disturbance of the bowels before this time. The practitioner ordered an enema twice a week, liquid paraffin and cascara. However, the abdominal distension continued and the child was admitted to hospital.

Examination (first admission) showed a moderate degree of abdominal distension in an otherwise healthy child. Some vague visible peristalsis was seen. Nothing abnormal was felt on abdominal palpation. Rectal examination was normal. X-ray examination (fig. 1) by means of a barium enema showed an extra loop of sigmoid colon which passed over to the right side of the abdomen. The colon was of normal calibre. The enema was completely evacuated in twenty-four hours. A diagnosis of dolichocolon was made and treatment instituted. An enema was given daily for two weeks, then every few days and then omitted. A mixture containing 25 minims of liquid extract of cascara daily was given. After six weeks in hospital he was discharged gaining weight with regular daily motions.

Progress. He returned home and continued to have regular bowel actions with the help of cascara for the next four months. Then increasing constipation necessitated the use of enemas every few days for two weeks. He then remained well again for another four months. He was now nearly twenty months of age. Increasing constipation again developed and occasional enemas kept it under control until May, 1943. Then an enema became necessary every few days until September, 1943. There then occurred a period of eight days during which he had no action of the bowels. At the end of this period he was admitted to hospital, now aged 2½ years. (His residence in the country had precluded supervision during the period between the two admissions.)

Examination (second admission September 14, 1943) showed a small child with enormous abdominal distension with a girth of 28 inches. The abdomen was tympanitic with visible coils of gut but without visible peristalsis. There were dilated veins over the abdominal wall and the child frequently cried with pain. X-ray examination showed (fig. 2) enormously distended large gut with fluid levels. He was treated by olive oil enemas to break up impacted faeces and daily soap enemas. Pain was relieved and the abdominal girth went down to 23 inches. X-ray examination after a barium enema showed the rectum and recto-sigmoidal junction to be of normal size. There was no obvious constriction at the pelvi-rectal junction. Above this point, the sigmoid colon and the whole
or the rest of the colon was enormously dilated, the sigmoid colon being most enlarged and extending well to the right and upwards into the epigastrium. Peristalsis appeared to be vigorous but spontaneous evacuations became infrequent. Within a fortnight it became necessary to give an enema every few days and peristalsis was only seen occasionally. An attempt was made to get bowel action by means of increasing doses of liquid extract of cascara in a rhubarb and soda mixture by mouth. But this produced pain and a very poor result as regards spontaneous evacuations. A second spinal anaesthetic was given on November 30, 1943, with 3.5 c.c.m. of hypobaric nupercaine. Peristalsis began at once but ceased after a few minutes. There was spontaneous evacuation of the bowels. Some intercurrent bronchitis now occurred and sulphadiazine was used in treatment. Pain and visible peristalsis occurred intermittently but no spontaneous evacuations occurred except on one occasion on the day before death. Cascara was omitted and enemas were given as required.

Terminal episode. For about a week the child refused food and became increasingly toxic. There was spontaneous evacuation of the bowels. Some intercurrent bronchitis now occurred and sulphadiazine was used in treatment. Pain and visible peristalsis occurred intermittently but no spontaneous evacuations occurred except on one occasion on the day before death. Cascara was omitted and enemas were given as required.

FIG. 2. (14.9.43.) Erect view of abdomen showing multiple fluid levels indicating some obstruction.

Fig. 2—Erect view of abdomen showing multiple fluid levels indicating some obstruction.

FIG. 3.—This shows a typical Hirschsprung with much dilation of the colon, but with normal sized rectum and pelvirectal junction.

Fig. 3—This shows a typical Hirschsprung with much dilation of the colon, but with normal sized rectum and pelvirectal junction.

FIG. 4.—After anaesthesia showing the enormous sigmoid loop and a peristaltic wave.

Fig. 4—After anaesthesia showing the enormous sigmoid loop and a peristaltic wave.

FIG. 5.—Post-mortem photograph. (Kindly made by Dr. P. Apperly).

Fig. 5—Post-mortem photograph. (Kindly made by Dr. P. Apperly).
was no vomiting. Abdominal distension increased and enemas produced a poor result. Death occurred suddenly one night.

Post-mortem examination was made by Dr. John Murray. There was enormous distension of the whole of the colon with a large amount of semi-fluid faeces (fig. 5). Barium was still present. There were formed faeces in the rectum. There was no obvious organic stenosis in the large gut or rectum. There was marked kinking of the ascending colon and at the recto-sigmoid junction. No histological examination was made.

Summary
The case here described began as what appeared to be an ordinary dolichocolon: a long, redundant colon with no obvious enlargement of the bowel. It developed into a typical megacolon of the Hirschsprung type, in which atony and constipation were temporarily relieved by spinal anaesthesia. It throws doubt upon the usually accepted good prognosis of dolichocolon and on the usually accepted etiology of Hirschsprung’s disease as a congenital, idiopathic, dilatation of the colon.