MIDGUT MALROTATION CAUSING INTESTINAL OBSTRUCTION IN ADULT PATIENTS

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Introduction

ALMOST 50 YEARS ago Mr. Zachary Cope delivered an Arris and Gale lecture in this College entitled ' The nerve supply of the parietal peritoneum and subperitoneal tissues with remarks on its clinical application'. This lecture and the subsequent writings of Sir Zachary Cope have greatly influenced our understanding of acute abdominal disease; the concepts of clinical observation enunciated in the lecture of 1922 remain valid to-day, they are part of the heritage of British surgery and form the basis for to-day's lecture on midgut malrotation[1, 2].

Anomalies of development of the intestinal tract usually forcibly reveal themselves during the first days of neonatal life. The anomaly we are concerned with to-day was present from birth but only became apparent when the patients reached adolescence or adult life; it can be difficult to diagnose and this difficulty in diagnosis may lead to delay in treatment. To make any discussion of the syndrome of midgut malrotation meaningful it is essential to have a clear understanding of the basic embryology of the intestinal tract. The embryology will be reviewed first, then the patients and their clinical features will be described, and lastly an attempt be will made to define the pathology and management of midgut malrotation.

Embryology

The midgut becomes a demarcated area of the primitive intestinal tract between the fifth and tenth weeks of intra-uterine life. It is that portion of the gut which is supplied by the superior mesenteric artery and in the adult forms the distal duodenum, the small intestine, the caecum and appendix, and the ascending colon. During the early stages of development of the midgut the true peritoneal cavity is small and the rapidly elongating intestine lies largely in an umbilical hernia, the extra-embryonic coelom. At this time the midgut has a long, attenuated dorsal mesentery containing the superior mesenteric artery and vein (Fig. 1*a*).

As development proceeds the small intestine elongates rapidly and has a tendency to become coiled, whereas the colic midgut elongates much more slowly and retains its straight configuration. While this elongation

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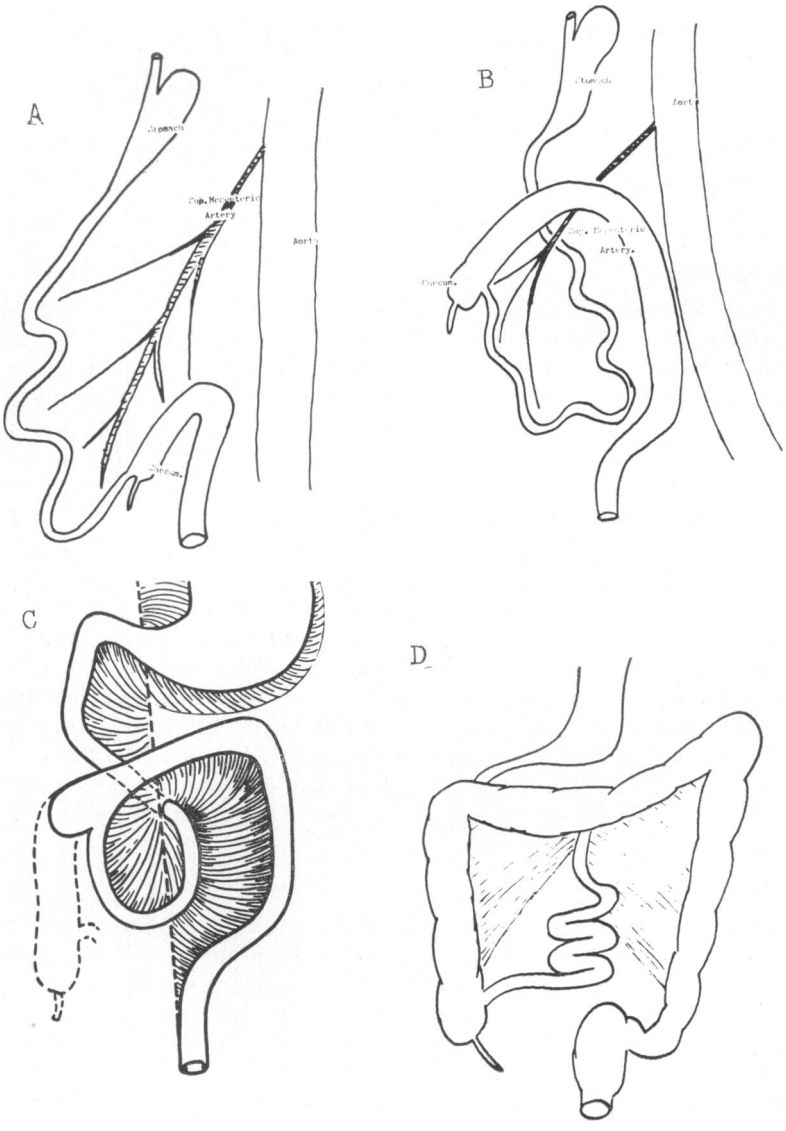
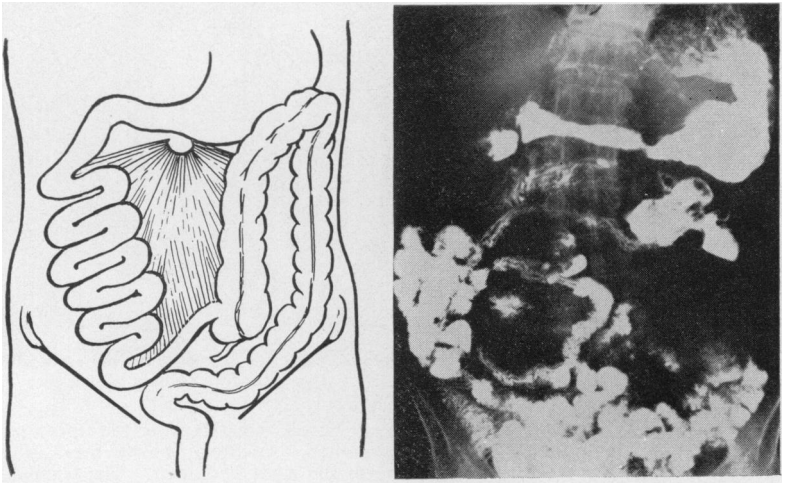


Fig. 1. Sketches to illustrate the embryology. (After Cunningham, *Textbook of Anatomy*.) (*a*) The development of the intestinal canal, and that portion supplied by the superior mesenteric artery (viewed from the left side). (*b*) The rotation of the intestinal loop around the superior mesenteric vessels. (*c*) Rotation of the intestinal loop and the continuous primitive mesentery is shown. (*d*) The final state, the adhesion of portions of the mesocolon to the posterior abdominal wall is shown (shaded). The small intestine mesentery, containing the superior mesenteric vessels, remains free.

is taking place two other events are occurring which dramatically influence the disposition of the adult intestine: these are the progressive reduction of the umbilical hernia (and coincident increase in the size of the true coelom) and the rotation of the midgut. The umbilical hernia is normally completely reduced by the end of the third month and only rarely and under abnormal conditions is it present at birth. As the gut re-enters the abdominal cavity it rotates in an anti-clockwise direction about the axis of the superior mesenteric vessels (Fig. 1*b*). When this rotation is completed the intestine has twisted through an arc of 270 degrees so that the caecum and terminal ileum lie to the right and the colon lies fanned out around the periphery of the abdomen (Fig. 1*c*). At this stage the dorsal mesentery of the duodenum becomes compressed with



(*a*) (*b*)

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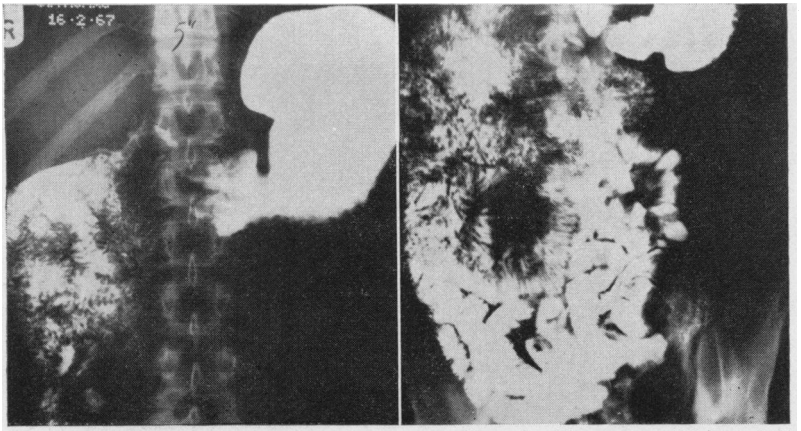
Fig. 2. (*a*) Diagram showing the distribution of the intestines when complete non-rotation is present. The narrow mesenteric origin is indicated. (*b*) Case 3. Small bowel barium meal showing midgut malrotation and the caecum in the left upper abdomen (arrow).

and fused to the parietal peritoneum; the mesentery of the small intestine lies in an oblique plane, still suspended dorsally as a leaf, and the mesocolon becomes adherent to the posterior abdominal wall in the flanks. Thus the intestines assume their adult disposition (Fig. 1*d*).

The clinical syndrome of midgut malrotation

Midgut malrotation may be defined as a partial or complete failure of this complex process of development so that the primitive dorsal mesentery persists to adult life; thus the small intestine will lie in the right side of the abdomen and the large intestine in the left side, the duodenum will be straight and all the colon will have a mesentery. Most importantly the superior mesenteric vessels will form a central fixed point about which a volvulus may occur (Fig. 2*a*).

Although midgut malrotation is a recognized cause of duodenal obstruction and volvulus in neonates[4, 5, 6, 7], this developmental anomaly has received little recognition as a cause of symptoms in adults[3]. During the last four years I have seen six cases of this anomaly which first presented in adults and a further case has recently been reported to me. In all these cases there was an initial confusion of the diagnosis and in some cases this confusion led to delay in instituting surgical management of the patients. It would be best now to describe these patients in order of appearance and then discuss their symptoms and signs and the management of the syndrome.



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(*a*) (*b*)

Fig. 3. Case 2—Barium studies. (*a*) Small bowel meal showing the unrotated duodenum and jejunum in the right abdomen. (*b*) Small bowel meal after 1¼ hours. This shows the small intestine in the right abdomen. The terminal ileum opens into the caecum from the right. The caecum lies in the left abdomen. Subsequent films confirmed the malposition of the caecum and showed the colon in the left abdomen.

Case 1

This patient was a 20-year-old man who was admitted to St. Thomas's Hospital as an emergency in February 1966. He gave a history of severe abdominal colic and vomiting for 72 hours. He had had similar attacks of central abdominal colic frequently since early childhood, each of these attacks lasting about 48 hours and then settling spontaneously. No satisfactory diagnosis had been made, and on one occasion he had been disciplined for malingering when an attack coincided with the end of a period of leave from the Services. A previous barium meal examination was reported as showing 'a normal stomach and duodenum'.

When first seen in St. Thomas's he had generalized abdominal tenderness and an empty rectum, as well as all the signs of dehydration due to his vomiting. An emergency laparotomy showed a midgut volvulus and at its apex a persistent vitello-intestinal duct connected the mass of intestines to the umbilicus. An enteric cyst and a urachal remnant were also present at the umbilicus. After these abnormal ducts and cysts had been divided from the small gut a complete midgut malrotation was found with the small intestine lying in the right abdomen. The midgut arose from a single long narrow mesentery. An area of gangrenous small intestine was resected and the abdomen was then closed. After a difficult postoperative period the patient recovered satisfactorily.

Case 2

A man aged 22 presented in February 1967 with a history of bouts of severe upper abdominal colic and diarrhoea since the age of 3 months. These attacks occurred at intervals of 6 to 8 weeks. Examination in the outpatient department disclosed some tenderness in the left side of the abdomen, and a tentative diagnosis of Crohn's disease was made. A routine barium meal examination was reported as normal, but a formal small-bowel meal showed a complete failure of intestinal rotation, with the small intestines occupying the right side of the abdomen and the colon the left side (Fig. 3*a* and *b*). A diagnosis of recurrent small intestinal volvulus was made.

At laparotomy the whole bowel from the duodenum to the descending colon was found to be suspended from a single narrow dorsal mesentery based on the origin of the superior mesenteric vessels. The duodenum ran caudally in a straight line from its first part onwards. The caecum lay in the left side of the abdomen and the ileum entered it from the right (Fig. 3*b*).

Case 3

A married woman aged 54 was admitted in October 1962 with a history of episodes of central abdominal colic every two to three months since childhood. The attacks of pain were accompanied by vomiting, and then, after two to three days as the pain was settling, she would have a bout of diarrhoea. At the age of 24 she had had an appendicectomy for ' one of these attacks '. Physical examination revealed some tenderness in the right lower abdomen, and a clinical diagnosis of Crohn's disease was made.

A barium small-bowel meal showed an unrotated duodenum with the jejunum lying in the right abdomen. The ileum entered the caecum from the right, and a barium enema confirmed that the colon lay on the left and that there was a complete non-rotation of the midgut. In the small gut there were areas of dilatation and narrowing which suggested Crohn's disease.

At laparotomy the small bowel was found to have a long mesentery which was very narrow at its origin. The duodenum was unrotated and twisted back on itself, and the ileum entered the caecum from the right. No lesions of Crohn's disease were found. In order to prevent further episodes of midgut volvulus the posterior surface of the common mesentery was sutured to the posterior abdominal wall in the sagittal plane as far as the pelvic brim.

After this operation the patient was symptom-free for four years until December 1966, when she had another attack of colic and vomiting. She was seen for follow-up in April 1967, and in view of the recurrent symptoms the possibility of Crohn's disease was raised again. Barium studies at this time confirmed the midgut malrotation but did not entirely exclude Crohn's disease (Fig. 2*b*). For this reason laparotomy was advised.

At operation the bowel was found to be densely adherent to the posterior abdominal wall; there were no lesions of Crohn's disease, and no cause for the obstructive episode in 1966 could be demonstrated. Since operation there has been no return of the symptoms.

Case 4

A man aged 21 first attended in May 1967 with a history of bouts of central abdominal colic, vomiting, and diarrhoea since early childhood. These attacks occurred every three to four months; they began suddenly and ended gradually, the end of the attack being heralded by the onset of diarrhoea, the stools sometimes being bloodstained. At the age of eight he had had bilateral undescended testicles treated surgically. He had also been treated by a psychiatrist for ' reactive depression ' because of his undiagnosable abdominal pain. Physical examination was unrewarding. On the basis of the history we considered midgut malrotation as a possible diagnosis but eventually settled with a tentative diagnosis of Crohn's disease.

Barium studies showed a midgut malrotation, with the small gut in the right abdomen and the large gut on the left (Fig. 4*a* and *b*).

The patient was admitted for laparotomy in September 1967. At operation all the intestine from the duodenum to the descending colon was found to be suspended

on a single dorsal mesentery. This mesentery arose from a narrow base high up in the abdomen and reached down to the pelvic brim. There were numerous areas of fine shiny fibrosis on the small bowel corresponding to sites of previous constriction when a midgut volvulus occurred. This primitive mesentery was sutured to the posterior abdominal wall in the sagittal plane along the axis of the superior mesenteric vessels. He was also found to have a midline gall bladder and biliary apparatus, and the aorta bifurcated high at the level of the renal arteries.

Case 5

A girl aged 18 came to the outpatient department in July 1967 with a history of intermittent attacks of central abdominal colic, sometimes accompanied by vomiting, since the age of 2 years. Each attack lasted up to 24 hours, beginning suddenly and settling gradually. These attacks occurred every few months and on occasions diarrhoea occurred towards the end of the attack.

On examination no abnormality was found in the abdomen. An initial diagnosis of midgut malrotation and intermittent volvulus was made. Barium studies of the

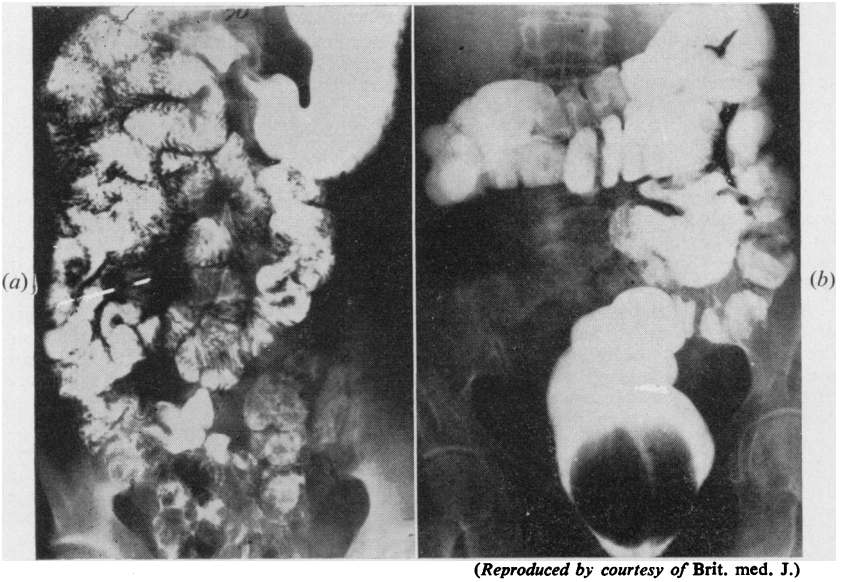

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Fig. 4. Case 4. (*a*) Small bowel meal showing unrotated duodenum. (*b*) Barium enema showing all the colon lying in the left abdomen. No intestinal rotation has taken place.

small bowel confirmed this diagnosis (Fig. 5*a*). In childhood this patient had been seen by a long succession of doctors, who had concluded that her pains were of psychogenic origin and she was given psychiatric treatment. In 1963 a normal appendix had been removed at another hospital after an attack of her usual symptoms. Her general practitioner had at this time diagnosed ' intermittent volvulus ', but this diagnosis was ignored by the surgeon to whom she was referred.

At laparotomy in September 1967 it was found that though the caecum lay to the right of the midline, the small bowel, caecum, and ascending colon arose from a long mesentery with a narrow base. The duodenum was incompletely rotated and it was joined to the caecum by a broad fibrous peritoneal band which effectively divided the abdominal cavity into two compartments, the bulk of the small intestine lying in an upper right-hand compartment (Fig. 5*a*), the colon being on the lower and left-hand side of the abdominal cavity (Fig. 5*a*). The fibrous band between the duodenum and the caecum was divided and excised, then the mesentery and the caecum were carefully sutured to the posterior abdominal wall. Since this operation the patient has been pain-free.

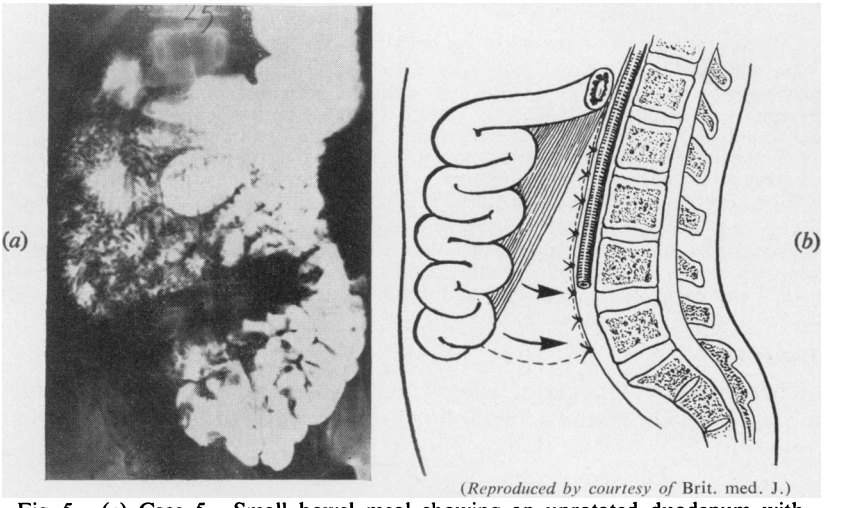

Fig. 5. (*a*) Case 5. Small bowel meal showing an unrotated duodenum with the small intestine apparently in two ' compartments ' (see text). The caecum is marked with an arrow. (*b*) Suggested method of suturing mesentery to posterior abdominal wall to prevent recurrent volvulus.

Case 6

This patient was an elderly lady who was admitted as an emergency to Sutton Hospital. She gave a history of bouts of abdominal colic going back to childhood. These had become less frequent of late and at the time of her admission she had a story of constant abdominal pain and tenderness; a pre-operative diagnosis of appendicitis was made. At operation she was found to have midgut malrotation and appendicitis with localized peritonitis. The appendicitis was caused by a benign adenoma obstructing the appendicular lumen. Subsequent barium studies (Fig. 6*a* and *b*) have confirmed the abnormal configuration of the intestines and no doubt this congenital anomaly was the cause of her frequently recurrent abdominal symptoms.

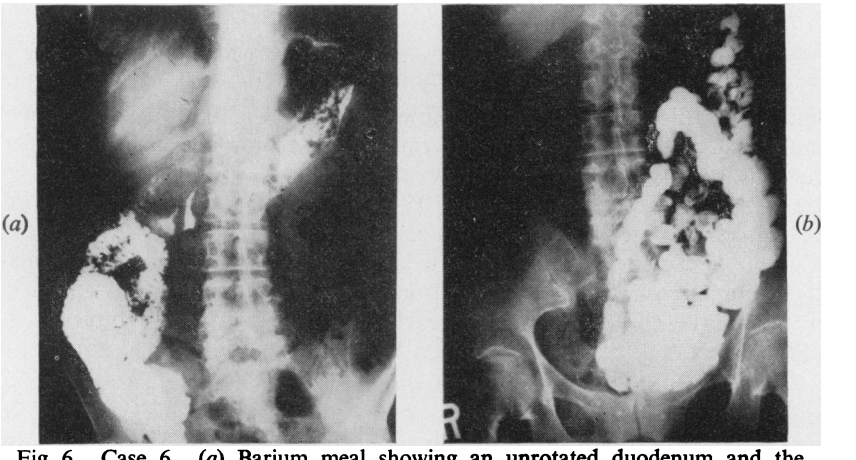

Fig. 6. Case 6. (*a*) Barium meal showing an unrotated duodenum and the small intestine in the right abdomen. (*b*) Barium in the colon which lies in the left abdomen.

Case 7

This case history was reported to me by Mr. B. W. Wells, F.R.C.S.

The patient was a 15-year-old youth who had had symptoms of recurrent high intestinal obstruction for about one year when he presented as an emergency at the Westminster Hospital in 1933. Mr. Tudor Edwards performed a laparotomy then and found the stomach and first part of the duodenum to be distended, the caecum lay in the upper abdomen, the transverse colon was retroperitoneal and there was complete non-rotation of the intestines. The superior mesenteric vessels formed a tight band across the distal duodenum (Fig. 7). A gastro-jejunostomy was performed.

After this the patient had no further symptoms and remained well for 21 years. He served in India throughout the War. Unfortunately the gastro-jejunostomy eventually wrought its havoc, he developed an anastomotic ulcer which perforated in 1954 and eventually a vagotomy and partial gastrectomy were carried out for this ulcer in 1960.

Discussion

There are several papers dealing with malrotation as a cause of obstruction in the newborn and some isolated reports of its occurrence in adults;

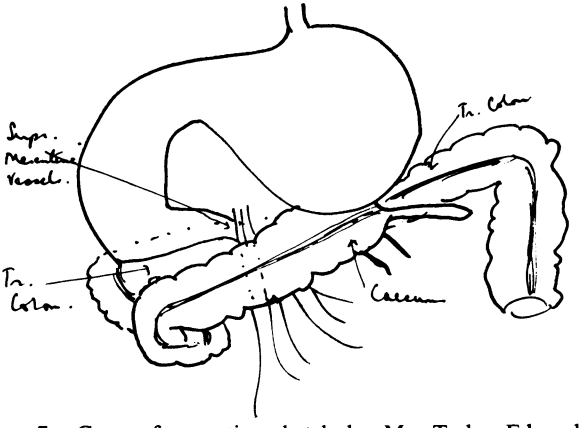

Fig. 7. Case 7. Copy of operative sketch by Mr. Tudor Edwards showing malrotation of the midgut (see text).

definitive clinical papers on this subject are those of Dott[4] and Gardner and Hart[5]. Dott was concerned to classify these anomalies and emphasize them as a cause of neonatal obstruction. Gardner and Hart reviewed the literature and commented on 105 cases. Ladd and Gross[6] described a series of 44 cases, of which 26 were diagnosed in the first three weeks of life. These and other paediatric papers have perhaps given the mistaken impression that this condition forcibly manifests itself in early life, consequently leading one to exclude it as a cause of symptoms in adults. Devlin *et al.*[3] drew attention to the clinical features in adults.

Malrotation of the gut may be discovered accidentally at operation or may present with inflammation of a misplaced organ (Case 6). It may also present with intestinal obstruction (Cases 1, 2, 3, 4, 5, 7). In the newborn this condition is almost always accompanied by high intestinal obstruction due to compression of the duodenum (Case 7). Snyder

and Chaffin[7] reported on 40 cases seen at the Los Angeles Children's Hospital. All 40 had high intestinal obstruction due either to adhesive bands running from the duodenum to the high caecum or to involvement of the duodenum in the base of a midgut volvulus. In the present series midgut volvulus occurred in five cases, one patient (Case 6) had a history of recurrent midgut volvulus but presented with atypical appendicitis, another patient had a combination of volvulus and band obstruction (Case 5).

The cases recorded here have many features in common (Table I). The patients were all apparently healthy adults who since childhood had had intermittent attacks of central abdominal colic unrelated to any

TABLE I

	Sex. Age of onset. Present age.	*Frequency of attacks. Character of pain.*	*Other symptoms and signs during attacks*	*Previous investigations*	*Previous diagnoses*
1.	Male. Childhood. Twenty years.	Every four months. Acute onset of pain. Central abdominal colic. Attacks last about 48 hours.	Vomiting at onset. Diarrhoea at end of attack. Diarrhoea sometimes bloodstained.	Barium meal: 'Stomach and duodenum normal.'	'Malingering.
2.	Male. Three months. Twenty-two years.	Every six to eight weeks. Acute onset of pain. Upper abdominal colic. Attacks last up to 24 hours.	Diarrhoea.	Barium meal: 'Normal.'	? Crohn's Disease.
3.	Female. Early childhood. Fifty-four years.	Every two to three months. Acute onset. Central abdominal colic. Attacks last 48 to 72 hours.	Vomiting at onset. Diarrhoea.	No previous investigations.	'Grumbling Crohn's Disease.'
4.	Male. Early childhood. Twenty-one years.	Every three or four months. Sudden onset. Central abdominal colic gradually settling.	Vomiting. Diarrhoea sometimes bloodstained.	No previous investigations.	'Reactive depression.
5.	Female. Early childhood. Eighteen years.	Every four months. Sudden onset. Central abdominal colic. Attacks last 24 hours.	Vomiting. Diarrhoea at end of attack	No previous investigations.	Depression. 'Chronic appendicitis. 'Intermittent volvulus.'
6.	Female. Childhood. Over sixty.	Frequent attacks of central abdominal colic.	No clear history given.	Unknown.	—
7.	Male. Thirteen years. Fifteen years.	Very frequent attacks of central abdominal colic. Sudden onset.	Vomiting. No definite history of diarrhoea.	None.	High intestinal obstruction.

precipitating cause. The attacks occurred at intervals of a few weeks to some months; sometimes they were accompanied by vomiting, and in all cases they often ended in a bout of diarrhoea, which in two cases was occasionally bloodstained. The pain started acutely but tended to fade out after 24 to 72 hours. Although the patients were obviously obstructed there was little or no abdominal distension even during a severe attack. Clinical examination between attacks was generally normal.

These patients had all suffered with these periodically incapacitating symptoms without any definite diagnosis being made. The probable explanation for this was the absence of physical signs by the time the patient came to be examined by the doctor and the negative results

yielded by routine investigations, such as cholecystography and pyelography. Even a conventional barium-meal examination did not suggest the diagnosis in some cases, presumably because after the first part of the duodenum had been screened further studies were not made of the transit of the contrast medium through the small gut. Overall the most likely reason for not making the diagnosis is simply because the clinical features did not correspond to any condition likely to be considered.

Most hospitals have a residue of patients with bizarre and often paradoxical abdominal disturbances, such as colic without distension, and it is possible that a certain number of these possess anomalies of intestinal rotation which predispose to temporary volvulus that is never severe enough to bring them to emergency laparotomy. Only one of our patients was operated on as an emergency. In this case non-rotation was combined with a persistent vitello-intestinal cord around which the volvulus had become strangulated.

The syndrome of recurrent bouts of upper abdominal or umbilical colic and vomiting with little if any distension, and then bloodstained diarrhoea to complete the episode, is very characteristic of malrotation. Sir Zachary Cope has pointed out[2] that small gut pain is typically in the umbilical region, in the midgut malrotation syndrome there is a small intestine volvulus, hence the umbilical pain, with partial or complete duodenal obstruction being relieved spontaneously. The volvulus occurs around the primitive dorsal mesentery and thus constricts and compresses the superior mesenteric vessels; this process will particularly affect the venous drainage and the involved bowel will become stuffed with blood. The infarcted bowel will bleed into its lumen and if the volvulus is then relieved spontaneously the patient will pass bloodstained diarrhoea signifying the end of the attack.

Investigations are often negative; in particular barium-meal studies without full screening of the small gut will often fail to make the diagnosis, and a misleading report, such as ' stomach and duodenum normal ', may further confuse the situation.

All suspected cases require laparotomy to confirm the diagnosis and to exclude other more dangerous causes of the symptoms. If intestinal malrotation is found, any abnormal bands should be excised and an attempt made to suture the intestines to the posterior abdominal wall to prevent further volvulus (Fig. 5*b*).

When doing this it is important to incise the posterior parietal peritoneum and the posterior mesenteric peritoneal layer, and then to suture these peritoneal layers together. Unless the peritoneum is incised and sutured in this way adequate adherence of the mesentery to the parietes cannot be guaranteed. If the case is operated on urgently and strangulated bowel needs resecting, it is probably wise to defer this mesenteric suturing until a later date.

Summary

Seven adult patients presented with a history of recurrent intestinal obstruction since childhood. Each obstructive episode lasted between 24 and 72 hours and was characterized by colic and vomiting, and diarrhoea towards the culmination of the attack. Physical examination between the attacks showed nothing of note. Barium studies of the whole intestinal tract revealed varying degrees of midgut malrotation, and laparotomy confirmed the diagnosis. At operation the mesentery was sutured to the posterior abdominal wall to prevent further episodes of volvulus. Symptomatic improvement followed this manoeuvre. The pathology, investigation, and treatment of these cases are discussed.

ACKNOWLEDGEMENTS

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GRANT OF DIPLOMAS TO FELLOWS IN DENTAL SURGERY

At the recent Final Examination for the Fellowship in Dental Surgery, 32 candidates out of 62 were successful.

At the meeting of Council held on 11th March, Diplomas of Fellowship in Dental Surgery were granted to the following:

INGLIS, Alexander Thomas (*The London*).
BOULTON, David (*Guy's*).
PELL, Gerard Melvyn (*Royal*).
MATTHEWS, David Owen (*Guy's*).
DAVIES, William Ian Rees (*University College Hospital*).
EYRE, John (*Royal*).
SELWYN-BARNETT, Brian Jack (*The London*).
DWYER, Declan Michael Patrick (*The London*).
RAMSAY, David John (*The London*).
MORGANSTEIN, Stuart Ian (*University College Hospital*).
WILLIAMS, Gerald Howard (*University College Hospital*).
LEWIS, Dwight Anthony (*Leeds*).
REAR, Stephen Bryan (*Guy's*).
HELMY, Mahmoud Helmy Youssef (*Cairo*).
AMARASEKARA, Gunadasa Suriarachchi (*Ceylon*).
PRASAD, Sakhamuri Venkata Leela (*Madras*).
PARRY, James Ebenezer Akyempong (*Liverpool*).
*SMITH, Peta Burton (*The London*).
SOLARIN, Edmund Oluwaseun (*Leeds*).
ROBERTS, George David Dodd (*Liverpool*).
CHAN, Yee Wing (*Singapore*).
KUMMOONA, Raja Kadum (*Baghdad*).
*SAWIRIS, Madiha Maurice (*Cairo*).
COLLIN, Brian David (*Durham*).
KAPLAN, Ross George (*Witwatersrand*).
LUKE, Douglas Alwyn (*Durham*).
BHUMGARA, Marazban Jamshedji (*Bombay*).
SAUNDERS, Ian David Frederick (*Belfast*).
HODGE, Michael George (*Guy's*).
NASH, Eric Stanley (*Bristol*).
WHITE, Peter Roland (*Birmingham*).
WILLIAMSON, David Colin (*Glasgow*).

* Woman.