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The presentation of malrotation of the intestine in adults

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Three cases of intestinal malrotation presenting in adults are reported. Each illustrates various aspects of symptomatology, investigation and treatment which are discussed.

Although malrotation has long been recognised as a cause of intestinal obstruction in neonates *(1,2)*, its clinical importance in adults is less well documented. In adults, if symptoms arise they are either due to the abnormality itself or to the presence of coincidental intestinal disease. The three cases described below illustrate these alternative modes of presentation.

Case reports

Case 1

A 37-year-old woman presented as an emergency with a 12 h history of upper abdominal colicky pain, abdominal distension and diarrhoea. She had suffered many previous episodes of similar pain of lesser severity. Physical examination demonstrated tenderness without guarding in the right upper quadrant of the abdomen with normal bowel sounds. The white cell count was 7.4×10^9/litre and the serum amylase 240 IU/litre. Plain radiographs showed gross faecal loading. A provisional diagnosis of chronic constipation was made. Over the following 48 h the symptoms subsided and she was allowed home with arrangements made for bowel transit time studies. She re-presented 1 week later with similar symptoms, but examination on this occasion revealed generalised abdominal tenderness with guarding. Radiography suggested large bowel obstruction. Following resuscitation the patient underwent laparotomy during which an ileocaecal volvulus was discovered lying in the right side of the abdomen. Having reduced the volvulus the entire large bowel was seen to occupy the left half of the abdomen with the ileum joining the midline caecum from the right. The duodenum, which was free on its own mesentery, passed straight down to join the jejunum to the right of the midline. The small intestine was therefore all held in the right hemi-abdomen. These appearances are typical of non-rotation. An ileocaecal resection was performed without intestinal fixation. The patient made a good recovery and was symptom free 12 months after her operation.

Case 2

A 57-year-old woman presented electively with a history of bouts of central abdominal colicky pain, each lasting for a few hours. She had suffered many similar episodes over the preceding 5 years preventing her from continuing in employment. Ten years before presentation she had undergone cholecystectomy for gallstones, when the midgut was found to be malrotated. Prior to investigation, she was admitted to hospital as an emergency, complaining of a 2-day history of central abdominal pain which settled in the left hypochondrium. Examination demonstrated localised peritonitis in the left upper quadrant of the abdomen.

The white cell count was 13.5×10^9/litre and the serum amylase was normal. Plain abdominal radiography showed a linear array of opacities in the left hypochondrium consistent with the appearance of the calcified

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contents of the appendix (Fig. 1). Acute appendicitis was confirmed at laparotomy and the patient was found to have non-rotation of the intestine similar to that described in the first case. No fixation procedure was performed after the appendix was removed. The patient remains well and pain free after 8 months.

Case 3

A 55-year-old man presented as an emergency with a 1-week history of lower abdominal colicky pain, distension and vomiting. He had previously been entirely well but had experienced decreasing frequency of bowel action and had occasionally noticed blood in the motions. Examination supported the overall clinical picture of large bowel obstruction, which was confirmed by radiography. Laparotomy revealed a stenosing carcinoma at the splenic flexure with gross dilatation of all the proximal intestine. Malrotation of the same pattern as described above was discovered but, in this case, the duodenum was in the normal retroperitoneal position. The caecum lying in the midline was in imminent danger of perforation. After decompressing the bowel and repairing the caecum, a defunctioning 'transverse' colostomy was formed to the left of the umbilicus. The patient made a good recovery. Postoperative barium enema examination, via both the anus and the colostomy, delineated the abnormal colon and confirmed the intraoperative findings (Fig. 2). He underwent an uneventful colectomy with reversal of the defunctioning colostomy 2 months later. No hepatic abnormality was palpable at this time. Histological examination of the resected specimen confirmed it to be an adenocarcinoma of the colon (Dukes' stage C). The patient later succumbed to the effects of multiple hepatic matastases.

Discussion

By the end of the 4th week of intrauterine life, the developing intestinal tract is supported by a short mesentery. The midgut, already identifiable as that part supplied by the superior mesenteric vessels, elongates faster than the rest of the gut and herniates out of the intraembrionic coelom, which is too small to hold it. The complicated return to the developing abdominal cavity was described and divided into three stages by Frazer and Robbins *(3)*. On this basis, Dott *(4)* was able to classify the various forms of intestinal malrotation. This

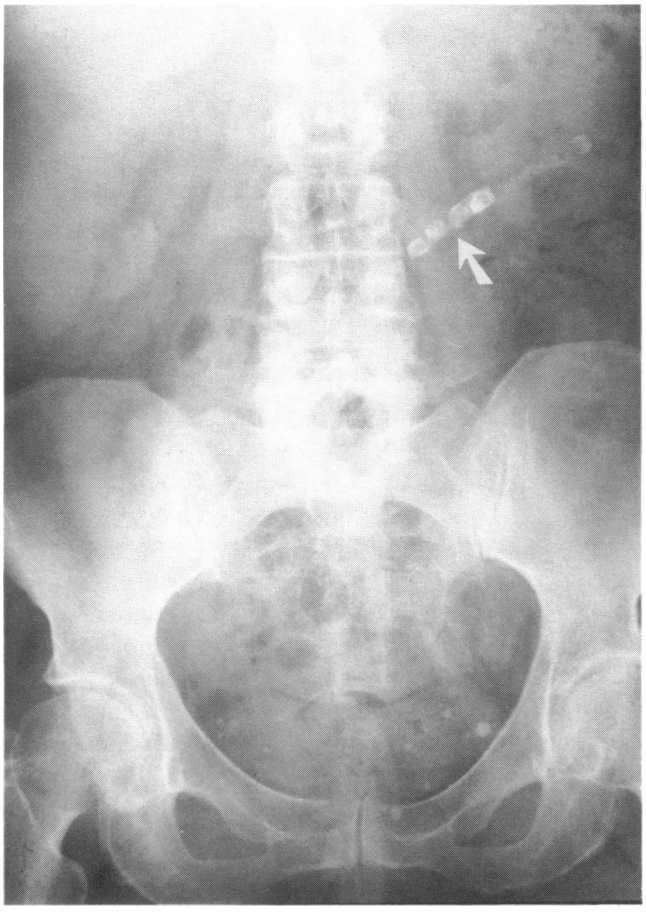

Figure 1. Plain abdominal radiograph (Case 2); faecoliths arrowed.

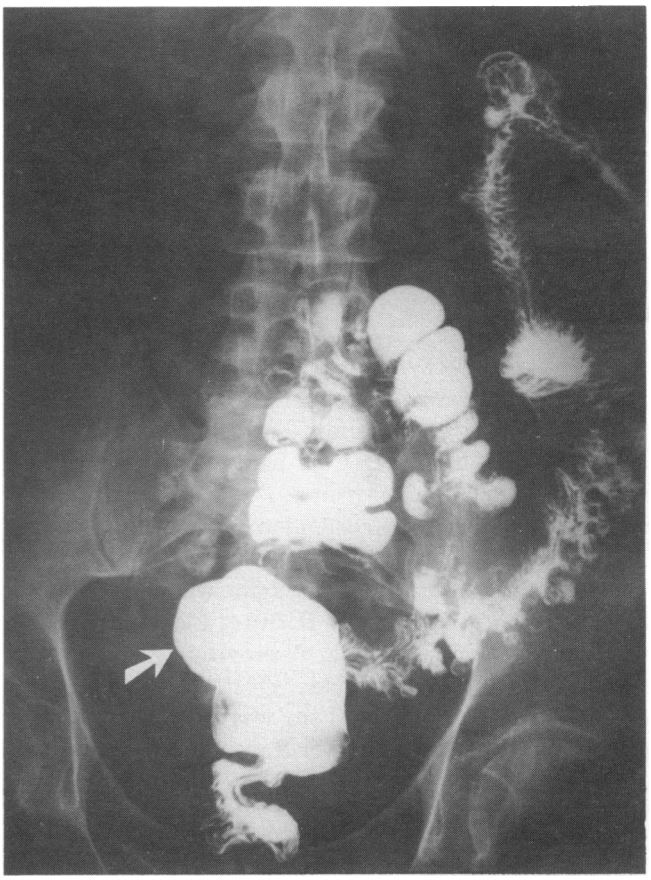

Figure 2. Barium contrast enema (Case 3); the entire colon lies within the left hemi-abdomen with the caecum (arrowed) overlying the rectum.

classification falsely divides a continuous process but facilitates the understanding of its complexities:

Stage 1: Exomphalos; failure of the intestine to return to the abdominal cavity presents at birth.

Stage 2: Non-rotation and partial rotation; the failure of the midgut to undergo rotation. If the duodenum is seen to descend in a straight line to the right of the superior mesenteric vessels to join the small bowel all of which lies in the right hemi-abdomen with the colon lying all to the left then no rotation has occurred. Partial rotation is the arrest of the intestinal migration at any stage between the position of non-rotation and the normal adult position. Since the original descriptions, it has been realised that rotation of the proximal and distal loops occurs independently so that failure of the duodenojejunal flexure to achieve the normal position is not necessarily associated with caecocolic malrotation and vice versa *(5)*. Nevertheless, both segments must be thoroughly examined if one is found to be abnormal. Reverse rotation, in which the caecum passes dorsal to the mesenteric vessels with the duodenum in front of both, is very rare. Paraduodenal hernias are attributed to abnormal intestinal rotation *(6)*.

Stage 3: Failure of retroperitonealisation (deficient fixation): Parts of the midgut are supported by persistent mesenteries. The caecum and duodenum are most commonly affected but the mesentery may also fail to extend its insertion and the small bowel is therefore, as in cases of non-rotation, suspended on a long, narrow mesentery.

Although 85% of cases of malrotation and non-rotation present in the first 2 weeks of life *(2)*, the cases described above demonstrate that these congenital anomalies can persist into adulthood. Many cases will remain asymptomatic, and in the absence of radiological or autopsy studies the true incidence is difficult to determine. In the largest reported series of these anomalies (50 cases) deficient fixation was the most frequently encountered abnormality; partial rotation accounted for 13 cases whereas non-rotation was found only three times *(7)*.

Symptoms can arise from acute or chronic intestinal obstruction which may either be due to the presence of abnormal peritoneal bands (eg Ladd's Bands) or a volvulus. In order for the bowel to twist fixation must be deficient. Ileocaecal volvulus, which is more common in adults *(4)*, occurs when the primitive caecal mesentery persists; the relatively broader insertion of the mesentery in such cases may confer a degree of protection against volvulus and hence delay presentation. In contrast, the limited insertion of the small bowel mesentery in cases of, for example, non-rotation explains the ease of formation of volvulus and consequently its earlier presentation.

As described in the first case, the pattern of recurrent bouts of central abdominal pain lasting for up to 72 h, followed by short-lived, sometimes blood stained, diarrhoea is typical in cases in malrotation. It is, however, so non-specific that when other more common pathologies, such as Crohn's disease or adhesive obstruction, have been excluded the patient may be labelled either as hysterical or a malingerer *(8)*.

The difficulty lies in both the abscence of specific physical signs and a straightforward method of investigation with which to confirm the diagnosis. The patient usually presents between episodes of pain so that plain radiographs are unhelpful. Contrast radiology will confirm the diagnosis *(9)* but is not usually contemplated in the symptomatic group into which these patients fall. Barium enema is more reliable than barium meal; the latter will demonstrate duodenal abnormalities but can fail to demonstrate small bowel malrotation unless delayed films are taken. Ultrasonography can define the characteristic abnormal vascular anatomy of malrotation and has been used successfully in diagnosis *(10)*. This technique may be able to identify the subgroup of patients, from amongst those with undiagnosed abdominal pain, who should proceed to have contrast studies in order to confirm malrotation.

Malrotation presents some peculiar problems to the surgeon. Two of the cases described above show how the clinical and radiological signs of some very familiar conditions can be confused by the abnormal anatomy. Many cases of malrotation will only be discovered when laparotomy is performed for coincidental disease. No operative intervention can be justified if the patient is free of symptoms directly attributable to the congenital abnormality. The first case illustrates that malrotation can be the cause of both acute and chronic symptoms which in either case requires treatment. The aim of any operative intervention is to alleviate and prevent recurrence of intestinal obstruction. At laparotomy, the abnormal anatomy must be deciphered which, even in experienced hands and especially at the time of emergency laparotomy, can be difficult. It should be remembered that malrotation is an anomaly which affects the whole midgut and, therefore, this should be examined in its entirety. Any abnormal adhesions are divided and the bowel is arranged in the configuration least likely to lead to further obstruction. This may require complete evisceration and the fixation of the midgut in the position of complete non-rotation rather than the normal position. In order to produce adherence, incision of the posterior peritoneal leaves before stitching has been recommended *(8)*. This is ineffective in the case of ileocaecal volvulus when, in a fit patient, right hemicolectomy, rather than caecopexy, is the best method of preventing recurrence *(11)*.

Conclusion

Malrotation of the midgut does not only present in childhood. In adult life it may cause chronic but nebulous symptoms which defy diagnosis. The presence of malrotation does not necessarily account for symptoms and careful consideration of other possible pathologies must be made. There is no reliable method of linking such symptoms with the abnormality other than by the meticulous interpretation of the patient's history.

Malrotation is usually found coincidentally at surgery, when it is important to identify the exact abnormality and plan the anatomical dissection appropriately.

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Notes on books

Renal Transplantation edited by Edgar L Milford. 313 pages, illustrated. Churchill Livingstone, New York. 1989. £50.00. ISBN 0 443 08627 3

This book is Volume 19 in the series *Contemporary Issues in Nephrology*. It contains eleven chapters written by North American experts on the immunologic, surgical, infectious, neoplastic, pathologic and pharmacologic aspects of renal transplantation.

Passing the FRCS by Ivor A Sewell. 331 pages, paperback. Butterworths, London. 1989. £10.95. ISBN 0 407 00725 3

The author of this book is a well-known surgical tutor in the North of England and he has now produced a study guide which will certainly be of value to many prospective surgeons. The first part outlines the implications of becoming a Fellow of a Royal College and gives practical advice on methods of studying and revising. The second part analyses the format of the various examinations in considerable detail, while the third part covers the post-examination period. Chapter 15 is entitled 'What if you fail?' and Chapter 16 'Careers after becoming a Fellow'. There are useful appendices on courses, books and journals.

Local Invasion and Spread of Cancer edited by Kenneth W Brunsen. 238 pages, illustrated. Kluwer Academic Publishers, Dordrecht. 1989. £59.95. ISBN 0 89838 996 8

A highly specialised volume of limited interest to clinical surgeons but of great interest to research workers studying cancer growth and progression. Twenty chapters covering general aspects of cancer spread and also dealing with specific tumours in detail.

Anesthesia and the Brain by John D Michenfelder. 215 pages, illustrated. Churchill Livingstone, New York. 1988. £27.50. ISBN 0 443 08628 1

The intention of the author is to bring together all the clinically relevant information available regarding the effects of currently used anaesthetics on the brain. Functional, metabolic and vascular effects are all considered in detail.

Endoscopic Biliary and Pancreatic Drainage by K Huibregtse. 144 pages, illustrated, paperback. Georg Thieme Verlag, Stuttgart. 1988. DM64. ISBN 3 13 731301 5

This slim, readable and well-illustrated monograph will interest all those who carry out ERCP or who manage patients with biliary obstruction. The author has very extensive experience of endoscopic biliary drainage and his authority is reflected throughout the volume. It has a strong 'How I do it' character which adds to the interest.

Surgery of the Peripheral Nerve by Susan E Mackinnon and A Lee Dellon. 638 pages, illustrated. Georg Thieme Verlag, New York. 1988. DM298. ISBN 3 13 728701 4

This beautifully produced and illustrated book will interest many readers of this notice—orthopaedic surgeons, neurosurgeons and general surgeons all operate from time to time on peripheral nerves. Beginning with anatomy and physiology of nerves and nerve injuries, the authors go on to discuss nerve repair and nerve grafting including results. The various entrapment syndromes are covered in depth and then extensive sections follow on specific nerve lesions including injuries and tumours. Each section begins with a historical review and is followed by the surgical anatomy before the clinical aspects are covered. The volume concludes with a bibliography of over one thousand references. An important and authoritative book.