Eur. Radiol. 4, 231–237 (1994)

European
Radiology

Gastrointestinal radiology

Midgut malrotation: radiological features of a twist of nature

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Received 22 July 1993; Revision received 12 November 1993; Accepted 2 February 1994

Abstract. To make an inventory of the radiological findings involved in midgut malrotation 62 consecutive patients with surgically proven isolated malrotation of the midgut were evaluated. Findings at plain abdominal radiography, upper gastrointestinal series, barium enema, ultrasound and/or CT were reviewed, correlated with the findings at surgery and classified into clinical entities related to embryological development. Findings were normal in 21 of 47 plain abdominal radiographs, 3 of 49 upper gastrointestinal series, 10 of 49 barium enemas and 9 of 14 ultrasound and/or CT examinations. All other examinations showed a multitude of abnormalities. Complete radiological classification into the clinical entities was possible in 39 patients (62.9 %). Partial classification could be established in the remaining 23 patients (37.1 %). It is concluded that midgut malrotation is a treacherous disease with a multitude of appearance.

By using the classification described, interpretation of findings becomes easier.

Key words: Abdomen, abnormalities – Children, gastrointestinal tract – Intestines, abnormalities – Midgut, malrotation – Rotational anomalies

Introduction

Malrotation of the midgut is a treacherous disease with a multitude of appearances. In order to make the diagnosis easier, findings of malrotation were reviewed retrospectively in 62 patients with surgically proven malrotation of the midgut. This paper will display the broad variety of findings at plain abdominal radiography, upper gastrointestinal series, barium enema, ultrasound and CT. Embryology, pathogenesis, and classification into clinical entities related to embryological development of the various types of midgut malrotation are presented. Radiographic features, embryological classification and findings at surgery are correlated. The radiological investigation of patients suspected of midgut malrotation is discussed.

Methods

Sixty-two consecutive patients with surgically proven isolated malrotation of the midgut were evaluated retrospectively. Patients with midgut malrotation associated with omphalocoele or diaphragmatic hernia were excluded. The mean patient age at diagnosis was 5.4 years (range 0–70 years). There were 31 female and 31 male patients. Findings in 47 plain abdominal radiographs, 49 upper gastrointestinal series, 49 barium enemas and 14 ultrasound and/or CT examinations were reviewed. The abnormalities were classified into clinical entities related to derangements in one of the three embryological stages (I, II and III) of rotational development. In stage I the midgut lengthens on the superior mesenteric vessels; no rotation has occurred. Stage II consists of withdrawal of the duodenum from the extra-embryonic position followed by its rotation and fixation. Finally, stage III consists of withdrawal of the right colon followed by its rotation and fixation.

Persistence of stage I of fetal development leads to type I(A) malrotation (= non-rotation): no rotation of the bowel has occurred. A narrow mesenteric base exists with a risk of midgut volvulus.

Derangement of the process of rotation in stage II can result in either a type IIA, IIB or IIC malrotation. In type IIA (= malrotation sensu strictiori) non-rotation of the duodenum occurs while the colon rotates normally. When the duodenum comes down to the right of the superior mesenteric vessels, bands cross the duodenum and cause obstruction. Type IIB (= reversed rotation) is the result of reversed rotation of the duodenum and colon. The rotation of the colon beneath the superior mesenteric vessels leads to (partial) obstruction of the mid-transverse colon. Reverse rotation of the duodenum anterior to the superior mesenteric artery (SMA) combined with normal rotation of the colon results in a type IIC rotational anomaly. The small bowel will thus be enfolded by the mesentery of the right colon leading to a paraduodenal hernia.

Rotational anomalies originating in stage III can lead to a type IIIA or IIIB midgut malrotation. In type IIIA partial rotation of the duodenum and non-rotation of the colon occurs leading to a type of midgut malrotation that strongly resembles type I(A). Type IIIB is defined as a situation of normal rotation of the duodenum and partial rotation of the colon with incomplete fixation of the colon at the hepatic flexure and formation of obstructing bands locally. Anomalies of fixation (type IIIC and IIID) that occur at this point in development will not be discussed in this paper.

Results

Of 47 plain abdominal radiographs 21 (21/47 = 44.7 %) showed normal or non-specific findings. Another 8 (8/47 = 17 %) revealed a pattern of high small bowel obstruction, while in 15 patients (15/47 = 31.9 %) the presence of multiple loops of distended bowel was indicative of low small bowel obstruction. In the remaining 3 patients (3/47 = 6.4 %) a gasless abdomen was found.

At upper gastrointestinal series findings were normal in 3 of 49 patients (3/49 = 6.1 %). In 11 patients (11/49 = 22.4 %) partial obstruction in the duodenum was seen with proximal distension. A so-called corkscrew appearance was present in 17 cases (17/49 = 34.7 %). Seven patients (7/49 = 14.3 %) showed an abnormal position of the duodeno-jejunal junction, while in 32 upper gastrointestinal series (32/49 = 65.3 %) the proximal jejunal loops were oriented to the right of the midline.

In the evaluation of 49 barium enemas findings were normal in 10 cases (10/49 = 20.4 %). In 25 patients (25/49 = 51 %) the colon showed different degrees of overlap of the spine (with the presence of the entire colon to the left of the vertebrae in 13/20). An abnormal position of the caecum was noted in 19 cases (19/49 = 38.8 %) while in 2 patients (2/49 = 4.1 %) the terminal ileum entered the caecum from the right aspect of the ascending colon. As can be deduced from these figures some patients presented with more than one feature typical of midgut malrotation on both upper gastroinestinal series and barium enema.

Normal findings at ultrasound or CT were present in 9 patients (9/14 = 64.3 %). In 5 cases (5/14 = 35.7 %) the superior mesenteric vein (SMV) could be found anterior to the SMA.

On the basis of the radiographic features described above a complete radiological classification was possible in 39 patients (39/62 = 62.9 %). Of these, 18 (18/62 = 29 %) showed a type I/IIIA malrotation. Eighteen patients (18/62 = 29 %) presented with features of a

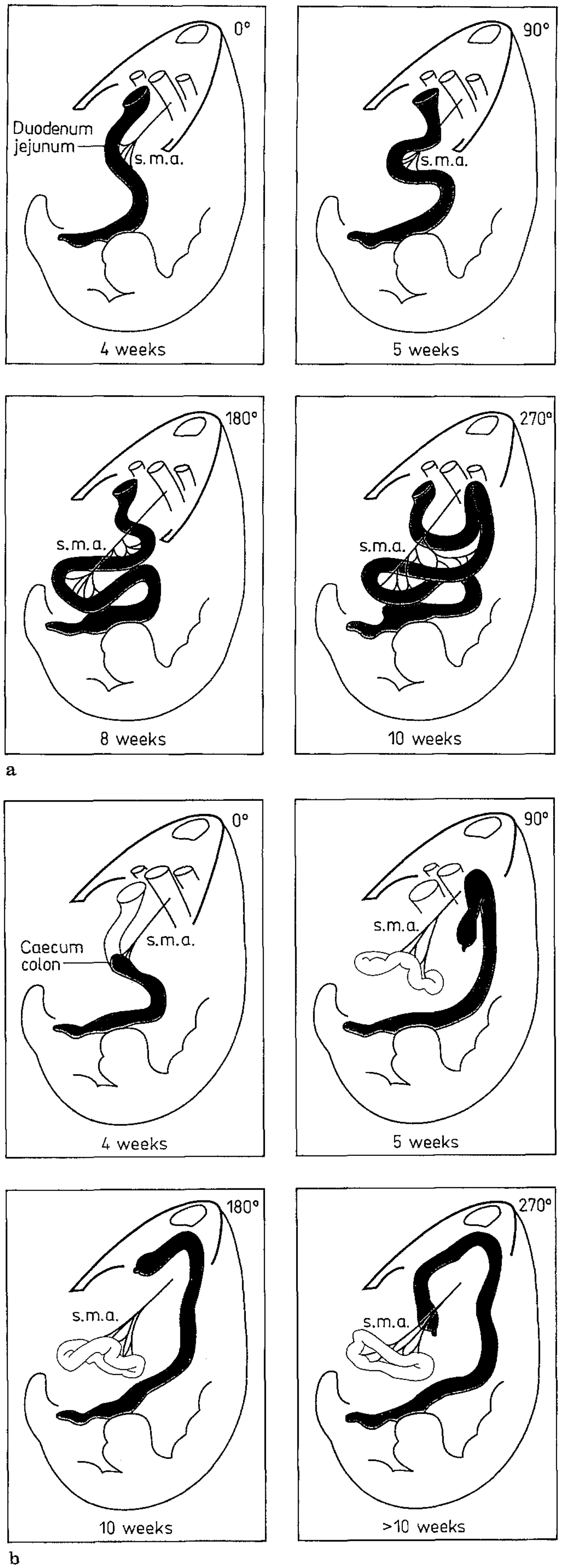


Fig. 1 a, b. The embryological stages of rotation of the duodeno-jejunal **a** and caeco-colic loops **b** For further explanation see text. (Modified from Snyder and Chaffin [23])

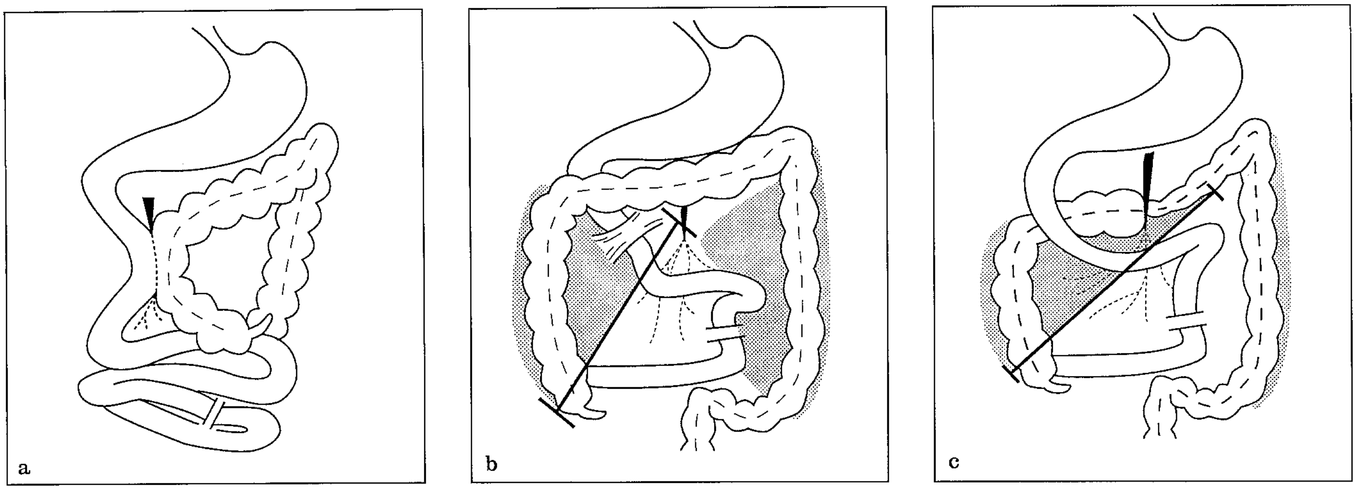


Fig. 2 a–c. The most frequently occurring types of midgut malrotation: non-rotation **a**, malrotation sensu strictiori **b** and reversed rotation **c**

type IIA anomaly, while in 3 patients (3/62 = 4.8 %) a type IIC was diagnosed. All these findings were confirmed at surgery. In the remaining 23 patients (23/62 = 37.1 %) radiological evaluation established the correct diagnosis but allowed only partial classification. Final classification was made according to the findings at surgery. This revealed 22 additional type I/IIIA anomalies and one type IIC malrotation. Thus a total of 40 of 62 type I/IIIA (64.5 %), 18 of 62 type IIA (29 %) and 4 of 62 type IIC (6.5 %) anomalies were revealed. No type IIB or IIIB anomaly was seen.

Discussion

Although approximately 15–50 % of patients with rotational anomalies of the midgut remain asymptomatic throughout their life (which makes estimation of its incidence difficult), midgut malrotation mostly presents in the neonatal period in a dramatic way [1–3]. Three distinct groups of clinical presentation can be discerned [4, 5]. The first, and most common group is related to rotational anomalies presenting during the neonatal period with symptoms of acute intestinal obstruction (e. g. bile-stained vomiting, abdominal distension and secondary dehydration) [6–9]. The second group consists of patients beyond the neonatal period. Complaints include crampy, colic-like abdominal pain, with or without vomiting, often in conjunction with failure to thrive [1, 2, 4, 10–14]. The third type of presentation is that of malabsorption and chronic diarrhoea due to intermittent volvulus, thus mimicking coeliac disease [4, 15]. Symptoms in the latter two groups of patients are frequently misinterpreted and failure to make the proper diagnosis ensues [2, 4, 10].

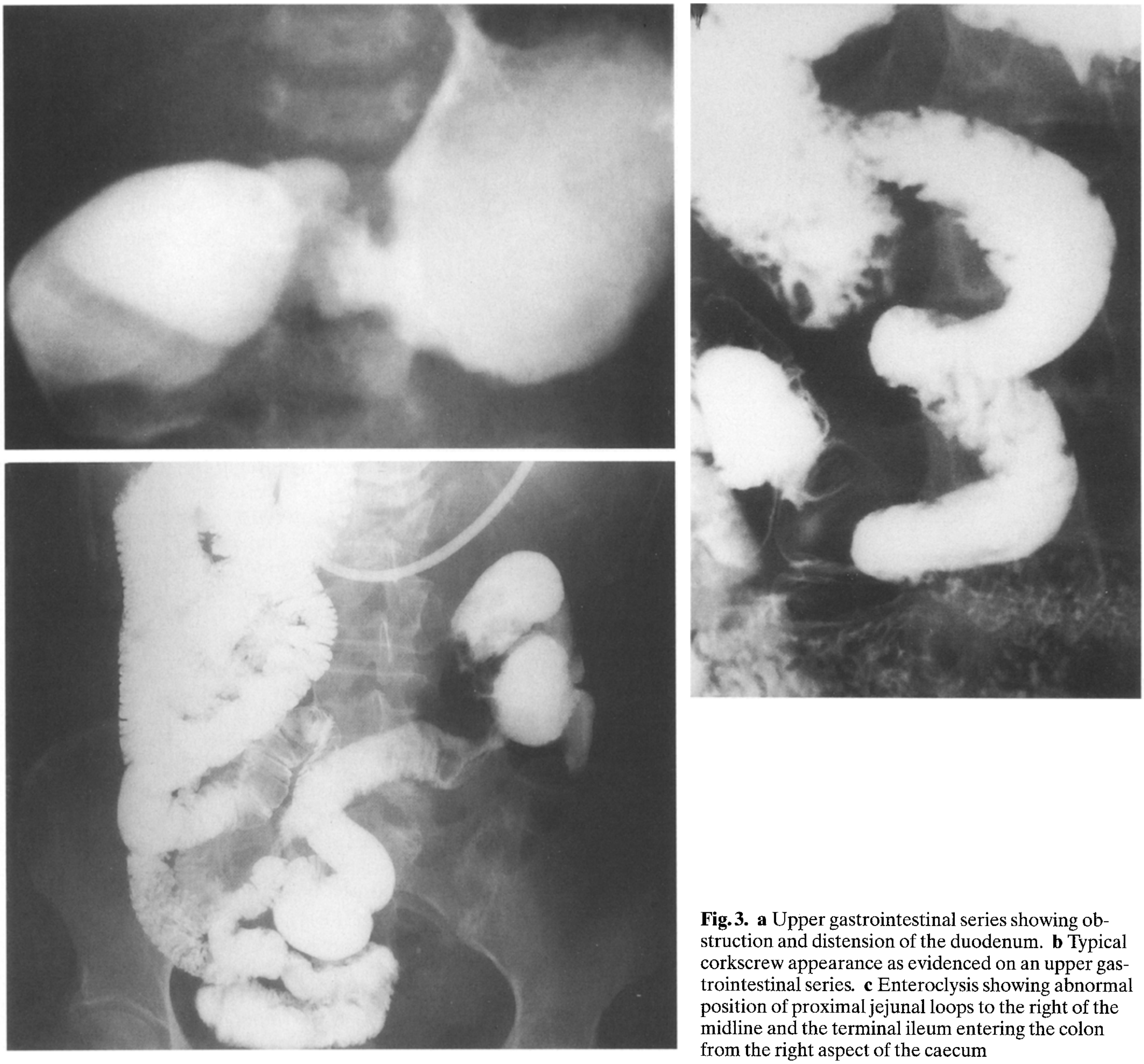
In order to interpret suspected midgut malrotation adequately knowledge of the embryological development of the midgut is necessary. The midgut is defined embryologically as the part of the fetal alimentary tract that is supplied by the SMA [6, 16–18]. Until the fourth week of embryonic life the midgut is a straight tube that can be divided into a pre-arterial and post-arterial segment [17, 19–21]. Rapid longitudinal growth results in the development of two leading loops of bowel: the duodeno-jejunal loop (distal duodenum, jejunum and upper ileum) and the caeco-colic loop (lower ileum, ascending and proximal two-thirds of the transverse colon), both pivoting independently around the SMA in a counter-clockwise fashion [1, 17, 19–23].

At 4 weeks the duodeno-jejunal loop has a midline position. At 5 weeks it adopts a position to the right of the SMA, being pressed inferiorly by the developing liver. At 8 weeks the loop has passed inferior to the artery and acquired an extra-embryonic position (i. e. physiological herniation in the umbilical sac). The final phase of rotation occurs with the return of the intestine into the peritoneal cavity, which brings the first part of the jejunum to the left of the artery, thus completing a total arc of 270 ° (Fig. 1 a) [23].

At 4 weeks the caeco-colic loop has a midline position also. During the fifth week, while the duodeno-jejunal loop is still in the abdominal cavity, the caeco-colic loop has already adopted an extra-coelomic position, lying to the left of the SMA. It is not until the tenth week, after withdrawal into the peritoneal cavity, that the 180 ° arc is completed. Shortly thereafter the final position to the right of the SMA is acquired (Fig. 1 b) [23]. Fixation of the midgut occurs with a broad, diagonally oriented small bowel mesentery.

In order to simplify matters this process of rotation can be divided into three stages [19], as described earlier in this paper. Of these different forms of midgut malrotation type I and IIIA (non-rotation; Fig. 2 a), type IIA (malrotation sensu strictiori; Fig. 2 b) and type IIB (reversed rotation; Fig. 2 c) are considered as occurring most frequently [7, 10, 16, 21]. Non-rotation and malrotation sensu strictiori were most frequently encountered in our series also (in 64.3 % and 30.3 % of cases respectively). Furthermore we found a type IIC in 3 cases (5.3 %) while, in contrast to findings in the literature, no case of reversed rotation was encountered.

In evaluating patients suspected of midgut malrotation plain abdominal radiographs and positive contrast



3a b c

Fig. 3. **a** Upper gastrointestinal series showing obstruction and distension of the duodenum. **b** Typical corkscrew appearance as evidenced on an upper gastrointestinal series. **c** Enteroclysis showing abnormal position of proximal jejunal loops to the right of the midline and the terminal ileum entering the colon from the right aspect of the caecum

examinations of the intestinal tract are used in establishing a radiological classification into the aforementioned clinical entities [7, 8, 10, 23–26]. On plain abdominal radiographs five categories of findings can be discerned [10, 12, 27, 28]. First, there is a pattern of high small bowel obstruction with findings of fluid and air in the stomach and first loop of the small bowel with a distinct paucity of gas in the distal intestinal tract (high small bowel obstruction). This includes the so-called double bubble appearance. Secondly a gasless abdomen can be found. Thirdly the presence of multiple loops of distended bowel displaying air-fluid levels may be encountered, indicative of low small bowel obstruction. Fourthly, nonspecific or normal findings may be seen which might give the sense of false security. Finally the so-called duodenal triangle, consisting of triangular gas shadows in the right upper abdominal quadrant, can be found [28]. In our group of patients normal findings were encountered most frequently (21/47), while low and high small bowel obstruction were less frequent (15/47 and 8/47 respectively). A gasless abdomen was seen by us in only 3 of 47 patients. The 'duodenal triangle' was not noted by us. In some patients the findings on plain abdominal radiography characteristically change with time. In longer-standing volvulus or obstruction signs indicative of bowel gangrene may occur such as intramural bowel gas, gas in the portal venous system, increase in bowel wall thickness and mucosal contour distortion [27].

Positive contrast examinations of the intestines are essential in judging the position of the bowel loops. Malrotation can be evidenced on upper gastrointestinal series by partial obstruction in the second or third part of the duodenum with proximal distension and 'to-and-fro' peristalsis (Fig. 3 a). In these cases the duodenum ends in

Fig. 4. Barium enema demonstrating the presence of the entire colon to the left of the spine

a 'beak of barium' [10, 12, 13, 29–31]. Twisting of the duodenum may occur, giving rise to the 'twisted ribbon' or 'corkscrew' appearance (Fig. 3 b) [3, 7, 8, 12, 13, 29, 31]. Furthermore an abnormal position of the duodeno-jejunal junction may be found – i. e. on the anteroposterior projection overlying the spine or to the right of the midline, on the lateral view lying anterior to the spine (= not retroperitoneal) [8, 9, 10, 12, 13, 14, 20, 29, 31–33] – although it has to be taken into account that in neonates and children under 4 months of age the duodeno-jejunal junction may be very mobile [34]. Finally an abnormal position of the proximal jejunal loops to the right of the midline may occur (Fig. 3 c; differential diagnosis: mesenteric redundancy, acquired right-sided duodenal hernia, previous surgical procedure with adhesions and left-sided abdominal masses) [7, 9, 12, 13, 14, 25, 32]. Of these the twisted ribbon appearance and abnormal position to the right of the midline of the proximal jejunal loops were most frequently seen in our series.

In the radiological evaluation of the distal (caeco-colic) loop several features can be discerned [35–37]. The presence of the entire colon to the left of the spine signifies non-rotation (type 1; Fig. 4), while different degrees of overlap of the spine (and therefore the SMA) are consistent with forms of partial non-rotation (e. g. type IIIA) [8, 10, 20]. This kind of abnormal position was present in 25 cases in our series. An abnormal position of the caecum, i. e. subhepatic or to the left of the spine, was found in 19 patients. It should be noted, however, that in up to 6 % of the normal population the caecum adopts a subhepatic position [10, 12, 13, 29]. Furthermore the terminal ileum can be seen entering the colon from the right aspect of the caecum (Fig. 3 c) [10, 13, 32]. This was noted in 2 of 49 patients. Finally, in type IIB anomaly (not encountered by us), narrowing of the transverse colon due to a retroarterial position can be present [19, 30, 38]. Because of the potential risk of aspiration at upper gastrointestinal series a barium enema is said to be the modality of choice in the neonate [7, 10, 12]. In our series, however, no aspiration was seen, a finding supported by others [8, 14, 29]. Thus one can start the evaluation of suspected midgut malrotation in adults as well as in infants and neonates with an upper gastrointestinal series [6, 13, 29], although in the neonate both may be needed in excluding other pathology (e. g. Hirschsprung's disease or duodenal atresia). Furthermore our findings indicate that an upper gastrointestinal series reveals the diagnosis more frequently because of the lower incidence of normal findings in our series (6.1 % for upper gastrointestinal series vs 20.4 % for barium enemas).

Ultrasound or CT (or MRI) is not the modality of choice in diagnosing midgut malrotation and may be of help in atypical and doubtful cases [9, 18, 39–42], although some authors suggest the use of ultrasound as a primary screening method in the newborn [43, 44]. In the past angiography has been used as well, but this does not seem now to be indicated [3, 21, 26]. The mesenteric vessels should be studied immediately distal to the confluence of the SMV and splenic vein. Normally the SMV lies ventrally and to the right of the SMA. In midgut malrotation this normal relation of the mesenteric vessels can be disturbed. In patients with partial non-rotation the SMV can be found to lie immediately anterior to the SMA (Fig. 5 a). When the vessels are traced caudally the SMV can be found to swing to the left of the SMA. This was noted by us in 5 patients. In cases of non-rotation the SMV lies persistently to the left of the SMA. It should be noted, however, that a normal position of the SMV relative to the SMA does not exclude the presence of malrotation [40, 41], and that an abnormal relationship of the SMA and SMV is not considered pathognomonic of this condition, because anatomical variations exist [42, 44]. In established volvulus ultrasound and CT may either demonstrate a fluid-filled, distended duodenal loop [43] or a mass (consisting of volvulated small bowel loops) and dilated tortuous mesenteric vessels [39, 43, 45–48]. Real-time ultrasound examination of the duodenal loop may reveal hyperperistalsis and to-and-fro motion [43, 48]. Furthermore an abnormal position of the duodeno-jejunal junction, small bowel loops or colon can be seen (Fig. 5 b) [47].

In conclusion, we find that in evaluating patients suspected of midgut malrotation plain abdominal radioa

b

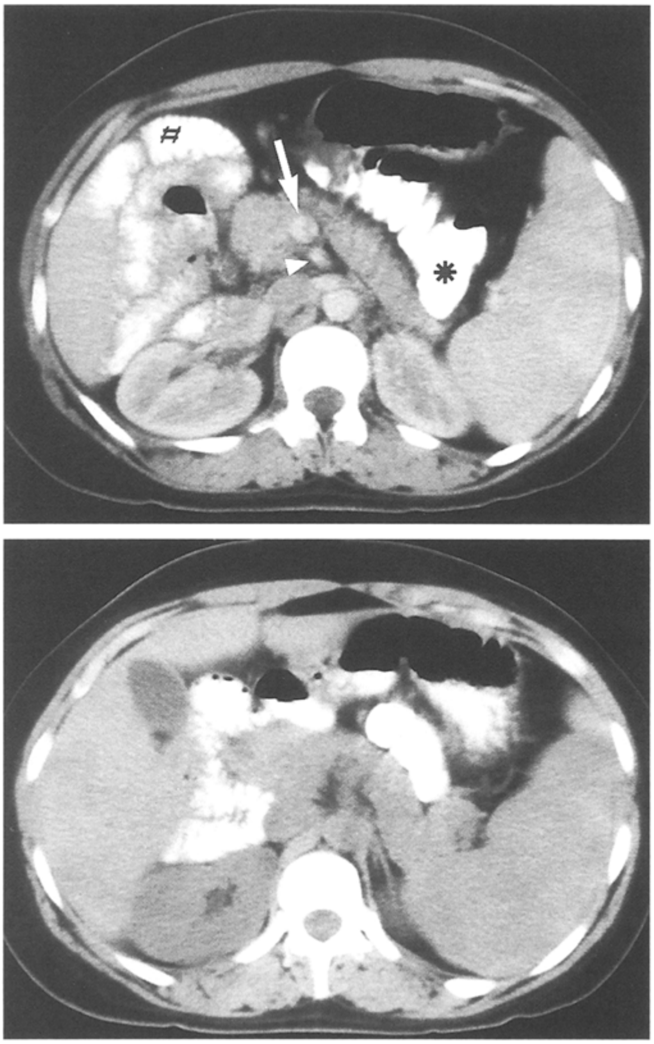


Fig. 5. **a** Contrast-enhanced CT scan showing the presence of the SMV *(arrow)* anterior to the SMA *(arrowhead)*. Small bowel loops are situated to the right (#), and the colon to the left of the midline (*). **b** Unenhanced CT scan showing failure of the third part of the duodenal loop to cross the midline (underneath the SMA), thus causing the small bowel loops to stay in the right half of the abdomen. (Courtesy of L. Meiss, MD, University Hospital Utrecht, The Netherlands)

graphs and positive contrast examinations of the intestinal tract are mandatory. In our opinion plain abdominal radiographs combined with either an upper gastrointestinal series or a barium enema are most useful in the neonatal period, while in older children and adults the combination of plain abdominal radiography and upper gastrointestinal series is preferable. When plain abdominal radiographs and/or positive contrast examinations are not conclusive ultrasound or CT may be performed. It should be noted, however, that radiological investigations should never delay therapeutic intervention [1, 13]. When studying positive contrast examinations in patients with suspected midgut malrotation one should always keep in mind that the midgut does not rotate as a single unit but as two distinct and independent loops. Normal rotation of one loop may be associated with the lack of rotation of the other loop. Normal findings at barium enema alone (or upper gastrointestinal series alone) therefore never exclude the possibility of a midgut malrotation.

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