Midgut Malrotation in Children: Our Experience in 33 Cases

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Midgut malrotation (MM) typically presents with bilious vomiting, with or without abdominal distension. However, there is a wide spectrum of associated clinical features that may, on occasion, lead to a delayed or erroneous diagnosis. In this retrospective study, medical records of 33 children with MM were reviewed. Seventeen patients (52%) presented in the neonatal period, seven (21%) in the infancy and nine (27%) after the first year of life. Seventy percent of patients presented with bilious vomiting. Although more common in the neonates, acute volvulus occurred in all age groups. Abdominal distension associated with irritability, bilious vomiting and bleeding per rectum were the most consistent indicators of intestinal strangulation. In the patients presenting beyond the neonatal period, the features of chronic/intermittent volvulus were more evident. These included failure to thrive, chronic non-specific abdominal complaints and malabsorption syndrome. Upper gastrointestinal contrast studies and ultrasound scan were the most commonly used investigations and were almost equally reliable. The mortality associated with acute volulus is well recognised, but the morbidity associated with chronic/intermittent volvulus in infants and older children is not adequately appreciated. Awareness of the unusual presentations in infants and older children and a high index of suspicion is recommended to avoid delayed diagnosis and surgical referral. Because of the unpredictability and high mortality and morbidity associated with acute and chronic volvulus, surgical correction in the form of Ladd's procedure is recommended in every patient with MM, regardless of age.
KEY WORDS: Midgut malrotation, volvulus, children, Ladd's procedure

The normal development of the midgut during early embryonic life involves a 270 counter-clockwise rotation that takes place in three stages[1,2]. Although this concept has been challenged recently [3], it is generally accepted that midgut malrotation (MM) results from an arrest of rotation in the second stage of normal rotation [4,5]. This failure of complete rotation results in a "reversed Comma"-shaped duodenum, as compared to the normal "C"-like arrangement. The duodenum comes to lie behind the superior mesenteric artery, or fails to cross the midline. The caecum also fails to reach its normal position and comes to lie in front of the duodenum. Abnormal peritoneal folds or adhesions run from the caecum across the duodenum to the posterior abdominal wall in the sub-hepatic area in an abortive attempt to fix the malpositioned right colon. These folds, which are often called Ladd's bands may obstruct the duodenum. Lack of rotation also results in failure of development of the root of mesentery, resulting in a narrow base around which the midgut may twist. This volvulus may result in obstruction to the mesenteric venous drainage and arterial inflow, which may lead to intestinal gangrene [5].

A majority of patients present during the first month of life with signs of upper gastrointestinal obstruction like bilious vomiting. However, when presenting beyond the neonatal period, the clinical features are often vague and non-specific, resulting in a delayed or erroneous diagnosis. We reviewed our experience with this condition to observe various modes of clinical presentation in relation to age, validity of different diagnostic investigations and the outcome of the patients.

Patients And Methods

Medical records of consecutive patients with midgut malrotation admitted to The Children's Hospital Sheffield, UK during 1990 and Chelsea & Westminster Hospital London, UK during 1992-94 were reviewed retrospectively. Patients having MM in association with congenital diaphragmatic hernia, exomphalos and gastroschisis were excluded from this study. All patients underwent a classic Ladd's procedure that involved division of Ladd's bands, straightening of the duodenum and widening of the base of the mesentery. An inversion appendicectomy was also performed in all patients.

Table 1: Prevalence of various clinical features, and outcome in 33 patients with midgut malrotation:

Presentation	Age: <1mth Mean: 5.82 days SD: 4.08 n= 17	Age: 1mth-1yr Mean: 5.83 months SD: 3.18 n= 7	Age: >1yr Mean: 8.77 years SD: 4.23 n=9
Sex (M:F)	11:6	3:4	6:3
Bilious vomiting	17	5	1
Non-bilious vomiting	-	2	4
Abdominal distension	8	1	1
Apnoeic episodes	1	2	-
Bleeding P/R	8	2	-
Volvulus with gangrene	2	1	-
Volvulus without gangrene	6	1	1
Irritability	8	1	-
Failure to thrive	-	5	3
Feeding problems	-	1	-
Diarrhoea	5	3	-
Fever	4	1	1
Abdominal pain	-	-	5
Incidental finding	-	1	4
Outcome:	-	-	-
Short-bowel syndrome	2	1	-
Life-long TPN dependent	1	-	-
Mortality	-		-

Three patients with gangrenous bowel underwent resection of variable length of the bowel. The patients were compared regarding age, sex, duration of symptoms, clinical features and outcome (Table 1).

Results

Three clinical entities were recognised: firstly, the neonates presenting acutely; secondly, the infants presenting either acutely or with failure to thrive and thirdly, older children presenting with less dramatic symptoms (Table 1). Overall, males were effected more frequently than females. Seventeen patients presented during the first month of life with a mean age of 5.82 days (range 1-19 days) and a mean duration of illness of 2.11 days (range 1-5days). Bilious vomiting was the main presenting feature. Eight neonates who also had abdominal distension, irritability, circulatory instability and bleeding per rectum were found to have a volvulus at laparotomy. Two of them required resection of variable lengths of intestinal segments that resulted in one being totally dependent on total parenteral nutrition (TPN).

Seven patients presented between the age of 1 month and 1 year with a mean age of 5.83 months (range 2-9 months) and mean duration of symptoms of 19.66 days (range 2-42 days), reflecting a delay in surgical referral. In one infant, MM was an incidental finding during a laparotomy for Hirschsprung's disease. In the rest, intermittent bilious vomiting and failure to thrive were the main clinical features. These patients had been initially investigated by physicians to detect medical illnesses and had been treated by several formula changes and therapies including anti-reflux medications. Two patients from this group developed a volvulus, one requiring resection of a limited segment of the bowel.

Nine children were more than one year old, with a mean age of 8.77 years (range 4-15 years) and mean duration of symptoms of 8.25 months (range 2 months-3 years). The most common presentation was intermittent non-specific abdominal pain and non-bilious vomiting. One patient was a diabetic and his symptoms were attributed to his diabetes, resulting in delay in surgical referral until he developed an acute volvulus. One patient from this group had even been referred to a child psychiatrist, because of his chronic symptoms in the absence of any associated positive physical and laboratory findings. Ultimately, an ultrasound scan of the superior mesenteric vessels revealed the correct diagnosis. In four patients, MM was an incidental finding during laparotomies for Hirschsprung's disease (1) and Nissen fundoplication (3). Older children had a higher incidence of non-bilious vomiting and a longer duration of antecedent symptoms that were mostly non-specific.

Associated anomalies were common. The gastrointestinal system was most commonly affected with gastro-oesophageal reflux and duodenal atresia being the most frequent associations (Table 2). Upper gastrointestinal contrast studies (Fig 1) were performed in 25 patients and ultrasound scans (USS) in 16 patients, to observe the superior mesenteric vascular orientation (Fig 2). Eight patients had both the investigations. USS proved diagnostic in all 16 patients including 2 patients in whom contrast studies had been labelled as normal. The diagnosis was confirmed at laparotomy in all patients.

Table 2: Associated anomalies in patients with midgut malrotation

	No. of cases with associated anomalies
Gastro-oesophageal reflux	6
Duodenal atresia/stenosis	4
Diabetes mellitus	1
Down's syndrome	1
VATER association	1
Cerebral palsy	1
Situs inversus	1
Ventricular septal defect	1
Inguinal hernia	1
Hirschsprung's disease	1
Intussusception	1
Infantile hypertrophic pyloric stenosis	1
Neonatal hepatitis	1
Skeletal deformities	1
Fallot's tetrology	1

The follow-up ranged from 6 months to 4 years. There was no mortality in this series. One patient developed post-operative intestinal obstruction and required a laparotomy for adhesolysis. Out of the 3 patients requiring bowel resection, two tolerated enteral feeding after an initial period of TPN while the remaining patient is surviving on long-term TPN. All the other patients are symptom-free and growing normally.

Discussion

Midgut malrotation classically presents acutely in the neonatal period. Fifty-five percent of patients present within the first week of life and upto 70% in the first month [4,6] Bilious vomiting secondary to duodenal obstruction from Ladd's bands is the most common feature [7,8,9]. Patients with volvulus and resultant bowel strangulation present additionally with persistent irritability, abdominal distension and tenderness, bleeding per rectum and shock [5,10]. The differential diagnosis at this age includes duodenal atresia, gastro-enteritis and necrotising enterocolitis (NEC).

Although, MM commonly presents in the neonatal period, various series have documented that 36% - 47% of children requiring surgery were older than one month of age and 15% - 18% were older than one year [9,11]. When presenting beyond the neonatal period, the diagnosis may be difficult because of multiplicity of clinical symptoms that are often vague and non-specific. Infants may present with intermittent bilious or non-bilious vomiting, feeding problems, malabsorption patterns associated with diarrhoea, protein-calorie malnutrition and failure to thrive [12,13,14]. A majority of these infants are initially referred for a medical opinion and followed by paediatricians. Often,

the symptoms are attributed to non-surgical conditions like milk intolerance, gastrointestinal allergies and celiac disease[14]. This leads to inappropriate extensive investigations and empirical trials of various therapies like anti-reflux medications, jejunal biopsies, gluten-free diet, lactose-free diet and change of formula. This indicates that the morbidity associated with chronic volvulus is generally not well appreciated. In actual fact, these symptom result from chronic/intermittent volvulus and obstruction resulting in blind-loop syndrome, lymphatic obstruction and protein loss in the bowel [11]. The bowel demonstrates a chronic venous and lymphatic congestion of the mesentery that impairs nutrient absorption and transportation [11,12,13]. Various degrees of chronic intestinal ischemia may further damage the absorptive mucosal surface. Infants may also present with intermittent apnoea secondary to gastro-oesophageal reflux precipitated by abnormal gastric motility and delayed gastric emptying [15].

Older children usually present with a longer history of intermittent or cyclical vomiting, intermittent abdominal pain, bloating, nausea, anorexia as a result of pain associated with eating, diarrhoea, constipation and chronic malnutrition[11,16,17]. These children with chronic symptoms may become withdrawn and introverted and face chronic learning and psychomotor disabilities[16]. Many children carry the diagnosis of psychogenic vomiting for months or years before a correct diagnosis of MM is made[9]. The abdominal pain is typically postprandial, colicky, transient, vague and not associated with any physical findings[6]. It must be appreciated that vomiting, bile-stained or not, and colicky abdominal pain in absence of abdominal distension and other physical signs does not exclude MM. These patients must be investigated properly to rule out MM before giving them a diagnostic label of abdominal migraine, cyclical vomiting, worm colic and psychiatric illness. Because of an ill-defined and "benign" past history, these children may present acutely because of sudden development of volvulus that may prove disastrous[10]. Rare presentations of MM include obstructive jaundice, peptic ulcer disease due to gastric and duodenal stasis[18], delayed diagnosis of appendicitis due to abnormal caecal position[19], intussusception due to lack of ileo-caecal fixation[20], chylous ascites, chylous mesenteric cysts, superior mesenteric vein thrombosis and colonic varices[13]. Intussusception associated with MM may present a confusing picture on hydrostatic barium reduction because of the abnormally oriented colon and caecum.

Approximately 0.2% - 0.5% of patients with MM presenting with vague and recurrent abdominal pain may escape the diagnosis until adulthood[19,21]. Cases of bowel gangrene have been documented in adults secondary to volvulus associated with MM[22,23]. MM may exist undetected throughout life (0.2%), or present as an incidental finding during a laparotomy for some unrelated condition[17]. Sudden onset of volvulus has been documented in previously asymptomatic children of different ages[4,14,18,23]. Because of unpredictability and high mortality and morbidity associated with both acute as well as chronic volvulus, surgical correction must be offered to every patient of MM regardless of age, whether symptomatic or asymptomatic, documented or incidental.

Associated anomalies and problems are common, with an incidence of 30% - 62%[4,9]. These include gastrointestinal disorders like duodenal atresia, gastro-oesophageal reflux, jejuno-ileal atresias, Hirschsprung's disease and anorectal anomalies (Table 2). In-utero bowel ischemia caused by Ladd's bands and volvulus has been implicated in the pathogenesis of the common associated anomalies: duodenal atresia and ileo-jejunal atresias respectively [24,25]. The other associated anomalies include congenital diaphragmatic hernia, gastroschisis, exomphalos and congenital heart defects. MM should be considered in the differential diagnosis when evaluating a child with multiple anomalies who presents with recurrent abdominal pain, feeding problems and failure to thrive[16].

The diagnosis of MM is straightforwards when a neonate presents with bilious vomiting or abdominal distension with signs of intestinal strangulation like tenderness, bleeding per rectum and shock. In these situations, urgent surgical intervention is indicated after the baby is intensively resuscitated[26]. Initial workup should always include a plain abdominal radiograph that may demonstrate evidence of duodenal obstruction, distended bowel loops or the ominous sign of bowel infarction: paucity of gas. Plain X-rays, on occasion, may not show any abnormality, leading to a delay in the diagnosis. When the diagnosis is sufficiently in doubt, further confirmatory investigations are indicated. Barium enema has fallen into disrepute because of certain limitations and high incidence of misinterpretation[7,27,28], as the caecum may be high and subhepatic in 5 % of population and mobile in 36% of infants[29,30,31]. Upper gastrointestinal contrast study is the most commonly used investigation. The findings suggestive of MM include partial duodenal obstruction, absence of horizontal part of the duodenum, abnormally positioned duodeno-jejunal flexure and spiral appearance of the duodeno-jejunal loops[26,32] (Fig 1). On occasions, contrast study may be erroneously interpreted as normal, especially in an un-obstructed child[14,29,32]. In addition, the contrast materials may prove hazardous in babies. Recently, interest has been developed in the use of ultrasound scan (USS) for the diagnosis of MM; the classic signs being an abnormally placed superior mesenteric vein in relation to the superior mesenteric artery (Fig 2). We used USS in 16 patients and found it diagnostic in all of them including two patients in whom the upper contrast series had been labelled as normal. We found USS a safe and non-invasive procedure that can be used as an initial investigation as well in those situations where the conventional contrast studies prove impracticable or inconclusive[33]. However, it must be appreciated that USS is very operator dependant and diagnostic only in well trained hands.

The incidence of the most feared complication of MM, midgut volvulus, ranges from 45% - 80% in the neonates and from 14% - 65% in older children [4,6,9,14,34,35]. If not

recognised promptly, it may lead to extensive bowel gangrene, with a disastrous outcome [7,9,36,37]. The mortality can be reduced by virtue of early recognition of the disease process and prompt intervention. Bile-stained vomiting must not be ignored in a neonate. The diagnosis of MM should always be considered during evaluation of infants with failure to thrive, and, older children presenting with recurrent abdominal pain, nausea, anorexia, malabsorption syndrome, even in the absence of physical findings. A high index of suspicion, appropriate investigations and prompt intervention are imperative to avoid the high mortality and morbidity associated with unrecognised acute volvulus.

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