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Malrotation of the intestine in childhood

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Sixty patients were operated on at Children's Hospital between 1985-1995 for abnormalities of intestinal rotation with or without volvulus. Thirty five were male and twenty five were female. All had clinical manifestations of high gut obstruction. The most common symptom of malrotation in neonates and infants was bilious vomiting while the older children had a long course of vague abdominal pain with or without bilious vomiting. Forty one patients (68.3 per cent) developed clinical presentations in the first month of life. Upper gastrointestinal series were performed for diagnosis in 50 patients, with no false-negative studies. Ten in 50 cases (20 per cent) were found to have the corkscrew sign of volvulus. Two patients required a barium enema in order to confirm the diagnosis because of suspicion of a volvulus with gangrenous bowels. An abnormal location of the cecum was found in both of them.

Sixty patients underwent a Ladd's procedure,including appendectomy. Twenty four (40 per cent) had malrotation alone while 36 (60 per cent) had malrotation with midgut volvulus. Eight of the 36 patients had bowel gangrene. A gastric perforation was noted in two patients. Eight patients were found to have both malrotation and intrinsic duodenal obstruction.

Eight patients died due to bowel gangrene, sepsis, pneumonia and serious congenital anomalies. The total mortality rate was 13.3 per cent. The mortality rate in the patients with malrotation alone was 8.3 per cent while the mortality rate in the patients with malrotation and volvulus increased to 16.7 per cent. This study emphasizes the need for consideration of Ladd's procedure for children of all ages when a malrotation of the intestine is discovered. If the operation is delayed, midgut volvulus may occur at any time.

Index : Malrotation, Non-rotation

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Malrotation of the intestine is the most common cause of extrinsic duodenal obstruction in infancy and childhood. It can produce acute episodes of duodenal obstruction by Ladd's band compression and midgut volevulus. Ladd1 described this anomaly and the operative treatment in 1932, and fibrous band anomalies and the operative procedure came to bear his name. Subsequent to Ladd's report, Gross2 revealed his experience with abnormalities of intestinal rotation in 1953. His paper presented a further analysis of the problems of malrotation, the improvement of diagnostic methods and the operative results. Nowadays, this anomaly is not well known in the general surgeon's concept and this results in misdiagnosis and improper management. Unfortunately, some patients have died because the correct diagnosis was delayed until they had developed bowel necrosis and septicemia. The purpose of this study was to review our experience with malrotation at Children's Hospital in the past ten years.

Materials and Methods

The records of total of 174 patients who were treated surgically for duodenal obstructions at Children's Hospital from January 1985 to December 1995 were reviewed. For the study, selected only 60 of the patients with malrotation. Children having congenital anomalies with associated malrotation (ie, diaphragmatic hernia, gastroschisis and omphalocele) were excluded from this study. In reviewing the medical records, special attention was paid to the initial presentation, operative findings and eventual outcome.

Table 1 Clinical presentation in 60 patients

Symptom/Sign	No. of patients	Per cent
Bilious vomiting	46	76.6
Non-bilious vomiting	12	20
Jaubdice	15	25
Abdominal distension	13	21.6
Lethargy	12	20
Abdominal pain	11	18.3
Constipaton	10	16.7
Bloody stool	3	5

Table 2 Associated anomalies

Anomaly	No. of patients
Down's syndrome	4
Meckl's diverticum	4
Congenital heart disease	3
Situs inversus	2
Jejunal atresia	2
KUB anomalies	2
Imperforate anus	1
Esophageal atresia	1
Duodenal duplication	1
Vertebral anomalies	1

Table 3 Analysis of the radiological findings

Radiological finding	No. of patients	Per cent
Plain abdominal films	60	
1. Incomplete small bowel obstruction	47	78.3
-gastric dilatation+ decreased distal bowel gas	(25)	(41.7)
-gastric and duodenal bulb dilatation+decreased distal bowel gas	(22)	(36.6)
2. Complete small bowel obstruction	10	16.7
3. Normal findings	3	5
UGI series	50	
1. Abdominal position of DJ juandice	50	100
2. Duodenal obstruction	50	100
3. Corkscrew sign	10	20
BE study	2	
Abnormal position of cecum	2	100

Results

Of the sixty patients with malrotation of the intestine (34.5% of all duodenal obstructions) that we studied, 35 were male and 25 female. The patient who had the lowest birth weight weighed 1,240 gm. The youngest was one day old and the oldest was 6.7 years of age. Eight patients (13.3%) had both malrotation and or intrinsic duodenal obstruction.

Bilious vomiting was the most common symptom and was found in 46 cases (76.6%). Twelve patients (20%) had a history of non-bilious vomitus (Table 1). We found 13 patients with abdominal distension in this study. Only three patients had passed bloody stool while there were eight patients of malrotation with volvulus and bowel gangrene. Eleven patients (18.3%) who were more than one year old had long courses of intermittent abdominal pain.

Forty one patients (68.3%) developed the clinical presentations of high gut obstruction within one month of age. Only 11 children (18.3%) had the onset of gut obstruction when over one year old. Associated congenital anomalies were common in patients who had malrotation, including intrinsic duodenal obstructions. Down's syndrome, Meckel's diverticulum and cardiac anomalies were frequent (Table 2). Anomalies were found in a total of 14 patients.

Barium enema (BE) studies were performed in two patients who diclosed the serious conditions. The BE finding was abnormal location of the cecum in the mid-upper abdomen. We used the upper gastrointestinal (UGI) series to confirm the diagnosis in 50 patients. The results of the UGI series revealed incomplete "C" loop formation, complete and incomplete obstruction of the duodenum in the third or fourth part, and abnormal location of the duodeno-jejunal (DJ) junction. Ten patients demonstrated the

corkscrew sign of volvulus form the UGI series (Fig. 3).

Sixty patients underwent Ladd's procedure, including appendectomy. At the time of laparotomy, midgut volvulus was found in 36 patients (60% of malrotation). Twenty six of the 36 (72.2%) in neonates and 10 (27.8%) in infants and children. Eight of these 36 patients (22.2%) required segmental resection for bowel necrosis. Two neonates also had gastric perforations. Three of these eight patients survived and two developed the clinical features of short bowel syndrome. Both of them had small bowel remainders of about 25 and 60 cm. They received parenteral hyperalimentation in the hospital for nearly one year. Six of the 36 patients with malrotation and volvulus died because of bowel necrosis and septicemia. The mortality rate in this group was 16.7%

In the 24 patients (without midgut volvulus) who underwent Ladd's procedure, eight patients were found with both malrotation and intrinsic duodenal obstructions. These lesions included duodenal web, annular pancreas and duodenal stenosis in 3, 3, and 2 cases respectively. Two of the 24 patients died because of severe associated anomalies and pneumonia. The mortality rate of malrotation without volvulus was 8.3%

Among the 60 patients, there were a total of eight deaths. Therefore, the total mortality rate was 13.3%. In comparision, the mortality rate of malrotation with midgut volvulus was higher than the other group with statistical difference ($p< 0.05$). The age incidence and the prognosis of the patients are shown in Table 4.

Follow-up for more than six months was available in 52 patients. Three of the survivors developed intestinal obstructions from adhesions and required exploration and enterolysis. Two patients with short bowel syndrome are still alive. They were 7 and 9 years of age on the last follow up in 1996. Both have had normal growth and development. There were no episodes of recurrent volvulus following Ladd's procedure in all of the surviving patients.

Discussion

Malrotation is a condition in which the midgut fails to complete its 270° counterclockwise rotation during return of the midgut from its ventral extracelomic herniation. This process occurs between 10-12 weeks of gestation[3-6]. In normal children, the duodenum is formed to be a "C" loop. The DJ junction is located in the left- upper quadrant and or the left side of the vertebrae. If the midgut fails to complete the rotation, the duodenum is not well formed in its "C" loop and leaves the DJ junction to the right side of the spine (Fig 4). The cecum is closed to the left side of the DJ junction. There is a fibrous band (Ladd's band) which originates from the cecum and attaches to the posterior aspect of the peritoneum. This band crosses over the duodenum and may be the cause of the extrinsic duodenal obstruction. Depending or the shortening of the mesenteric pedicles, midgut volvulus may easily occure.[1,2,5] Although more accurate terms for this

Table 4 Correlation between age incidence and prognosis

Age	Malrotation		Malrotation with volvulus	
	Total	Died	Total	Died
0-1 month	15	1	26	4
>1 month-1 yr	5	1	3	2
>1 yr	4	0	7	0
Total	24	2	36	6

diagnosis would be "nonrotation", "incomplete rotation" or "fixation" malrotation remains in current usuage.

Malrotation may present as acute duodenal obstruction alone or as duodenal obstruction accompanied by signs of mesenteric vascular compromise. Volvulus due to malrotation may occur in utero, producing variable degrees of ischemic necrosis and resulting intestinal atresia[7]. At birth, there will besigns of upper gastrointestinal obstruction.

In general, the clinical manifestations of patients with malrotation,with or without volvulus, vary somewhat with theirage. More than half are seen in the first month of life [5,8-10]. However, midgut volvulus may occur at any time. The most common symptom of malrotation in neonates and infants is bilious vomiting, while the older children have had a long course of vague abdominal pain with or without bilious vomiting[9-14]. In many infants, an abrupt change from a normal feeding pattern to pernicious vomiting and irritability signifies the development of midgut volvulus requiring immediate surgical intervention.

Abdominal distension may be present or not, depending upon the site of obstruction. If extrinsic duodenal obstruction is present, the infants may have gastric decompression caused by frequent vomiting and the distension may be conspicously absent or confined to the upper abdomen. If midgut volvulus has occurred, abdominal distension is likely to be more marked.

Bloody stool following the acute onset of gut obstruction from malrotation is a dire sign indicating compromise of intestinal circulation. This sign indicates that midgut volvulus has occurred. In our study, there were eight volvulus patients with extensive small bowel necrosis, but only, three patients, passed bloody stool. This indicates that the patients may have infarcted bowel without bloody stool.[9,10]

The diagnosis of malrotation is made by a combination of plain abdominal films followed by BE examination and a UGI series [15-21]. The radiological findings in the plain abdominal films may reveal both obvious signs of intestinal obstruction and partial duodenal obstrucion. If there are both gastric and duodenal dilatations (Fig 2.), the patients may have the combination of malrotation and intrinsic duodenal

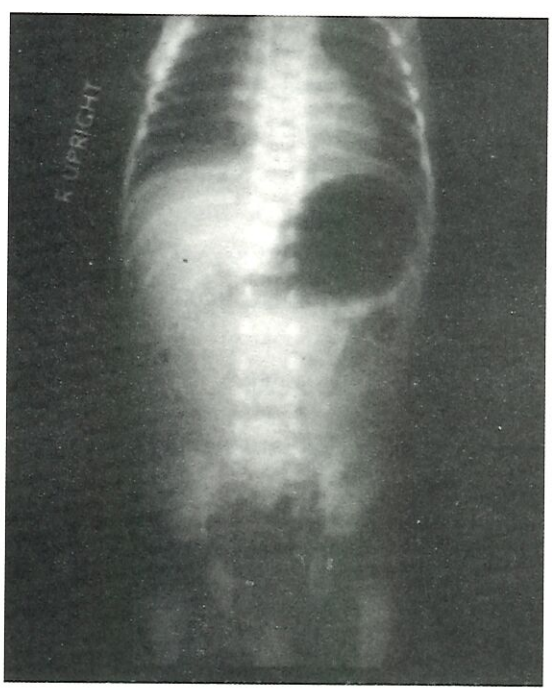

Fig1. Infant with malrotation. plain abdominal examination shows incomplete duodenal obstruction with only gastric dilatation and decreased distal bowel

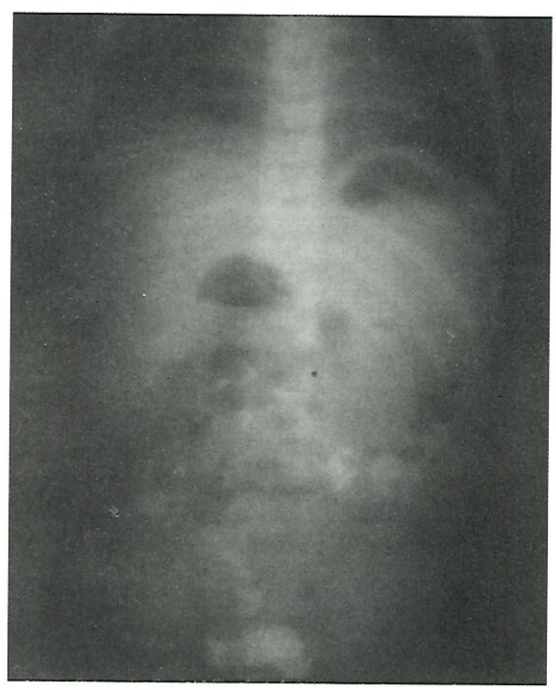

Fig2. Malrotation in infants. incomplete duodenal obstruction is showed in a plain film of abdomen. Gastric and proximal duodenal dilatation including minimal distal bowel gas are noted.

Fig3. This film is one of UGI study in a neonate with malrotation and midgut volvulus. Clockwise rotations of small bowel are present as a corkscrew sign of volvulus.

Fig4. And UGI study of a child who has abnormalities of midgut rotations shows nonformed "C" loop of duodenum, abnormal location of small bowel in the left and in the right side of abdomen.

obstruction[15]. Three patients in our serieshad normal roentgenological findings in the plain abdominal films [7]. A barium enema (BE) study is recommended as the initial examination to confirm the diagnosis[17]. It demonstrates malposition of the incompletely rotated cecum and colon in the right upper quadrant. Sometimes, the BE study is inconclusive ; therefore, the UGI series is made to confirm the diagnosis. However, the advantages and safety of the UGI series in malrotation are now well recognized[17-20]. A UGI study will demonstrate duodenal obstruction which is usually seen in the third part rather than in the second portion of the duodenum in the patients with intrinsic duodenal obstruction. The contrast study will also show the abnormal position of the proximal jejunal loop to the right of the midline. Corkscrewing of the intestinal loops may be seen in malrotation with volvulus[10,16]. Ten of the 36 volvulus patients in our study displayed the cockscrew sign during the UGI series.

For correction, we prefer to perform a celiotomy through a midline incison. The entire bowel is extracted out of the abdomen by gentle pressure on the abdominal cavity. Failure to extract the entire bowel outside the abdomen can lead to misdiagnosis or incomplete dissections of the bands[10]. If the volvulus is present, it is reduced by counterclockwise rotation. The peritoneal bands (Ladd's bands) are freed by sharp and blunt dissection from the duodenum to the mesenteric pedicles. An appendectomy is done in order to avoid the future likelihood of a diagnosis of appendicitis presenting in the left side of abdomen.

In cases of midgut volvulus and bowel necrosis, we use warm saline and gauze coverage on the devitalized bowel. If extensive bowel necrosis persists, the abdomen is closed and the patient is returned to the ward. A second look operation is performed after 24-48 hours from the initial procedure. Occasionally, the ischemic bowel is more vitalized in the second operation. Bowel necrosis is the most common cause of death among patients with midgut volvulus. Extensive midgut resection is a risk for short bowel syndrome. In our study, twopatients with only 25 and 60 cm. of the small bowel remainingwere survived by for a long period of time by use of

parenteral hyperalimentation.

When suspecting that there is also intrinsic duodenal obstruction, a gastrotomy or proximal duodenotomy is performed. A Foley catheter is passed through the stomach or duodenum to the upper jejunum. The balloon is inflated and the catheter is pulled proximally into the stomach or duodenum to rule out associated intrinsic lesions in the duodenum and upper jejunum. Eight patients of our study (13.3%) had intrinsic lesions including duodenal web or diaphragm, duodenal stenosis and annular pancreas. Other reports have shownor incidence of malrotation with intrinsic obstruction in the range of 5.6-10 per cent[9,10].

After completion of the Ladd's procedure, the entire bowel lies free of its peritoneal attachments. The colon is returned to the left side and the small bowel to the right side of the abdomen. It is not necessory to fix the intestines with the lateral peritoneal side wall andwe have not experienced recurrent volvulus in our patients. Many other authors we reviewed had also not found any recurrent volvulus.[9,10,21]

According to the study of Messineo et al,[22] the mortality risk of malrotation is associated with the increased percentage of neorotic bowel, the presence of serious associated anomalies and younger age of the patients at the time of operation. Nowadays, the mortality rate is reduced because of the improvement in diagnosis, surgical therapy and postoperative care. Early diagnosis followed by immediate surgical intervention and careful exploration at laparotomy for associated anomalies is mandatory. The improvement in postoperative neonatal care, including total parenteral nutrition, has alsoimproved the postoperative course. However, we recommend prompt elective Ladd's procedure in all patients who are incidentally found to have abnormalitis of intestinal rotation or fixation, even if they are asymptomatic children.

Conclusion

Our experience documents that the clinical onset of malrotation frequently occurs in the neonatal period. Most patients developed midgut volvulus in the period of presenting symptoms. The factors which influence the mortality rate are age of the patients and delayed diagnosis. Our study helps confirms the association between high mortality and younger age, as demonstration in Table 4. A delay in diagnosis, leading to a delay in surgery, is also associated with an increased risk of mortality. We reccomend Ladds procedure in children of all ages with malrotation because midgut volvulus may occur at any time if the operation is delayed.

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