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ORIGINAL ARTICLE

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Long-term complications following intestinal malrotation and the Ladd's procedure: a 15 year review

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Abstract *Background*: It is accepted that children with volvulus require urgent surgery. However the long term sequelae and late complications of its surgical therapy are less well understood. We hypothesised that the surgical corrected intestinal malrotation have significant long term impact on the patients quality of life. *Methods*: Forty-six children with intestinal malrotation were operated on at a tertiary referral centre over a fifteen year period. Their charts were retrospectively reviewed and the patients were contacted. *Results*: The study revealed two distinct groups, those without complications 25 (54%) and those without 21 (46%). In the acute post operative period four (9%) patients had on going feeding difficulties and one (2%) developed chronic abdominal pain. However 12 (26%) required readmission within the first six months after the initial operation. Eleven (24%) patients were readmitted with acute bowel obstruction. Six (13%) patients required multiple admissions due to small bowel obstruction and six (13%) patients underwent further surgery for adhesion related obstruction. There were four (9%) deaths in the study group, three due to other medical conditions and one following small bowel obstruction. There was zero mortality immediately after the primary operation. There was no significant difference in the initial presentation, age and operative findings in those requiring further surgery and those who did not. *Conclusions*: We demonstrated that there is a significant long-term morbidity associated with intestinal malrotation even after corrective surgery. Detailed education about the potential for small bowel obstruction must be given to the parents of these children.

Keywords Intestinal obstruction · Intestinal volvulus · Intestinal malrotation · Ladd's procedure

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Introduction

Intestinal malrotation is a not uncommon anomaly which frequently occurs during the neonatal period with bilious vomiting. The surgical emphasis is on early diagnosis and treatment in order to minimise the risks of volvulus. The surgical mantra of always ruling out malrotation in bilious vomiting is a safe and proper practice [1, 2]. Yet, although the immediate complications are well known and fears regarding the terrible consequences of short gut syndrome following volvulus are not misplaced, once the procedure is safely performed, the child is frequently discharged with sound health [2]. The aim of the study was to assess the long-term complications of malrotation and the Ladd's procedure.

Methods

All patients aged up to 17 years diagnosed with malrotation in a tertiary referral paediatric surgical centre from January 1986 to June 2000 were included in the study. All clinical records and admission details were reviewed. Data was collected on gender, age, symptoms, duration of symptoms, clinical findings, duration of surgery, operative procedure, period of stay, post-operative complications, further surgery and long-term complications. If possible, the patient or the child's parents were contacted; in case of non-availability of either of them, the patient's general practitioner was contacted. The mean follow-up for the entire group was 10 years ranging from 3 to 18 years.

Results

A total of 57 patients who undeunderwent Ladd's procedure were identified. Of them, 46 were available for assessment and follow-up. The patients presented in three distinct age groups. Twenty-two (48%) presented

Table 1 Predominant symptoms at the age of presentation

	Neonates	1–12 months	> 1 year	Number of patients
Bilious	11	4	4	19
Vomiting	4	3	3	10
Pain	0	0	5	5
Failure to thrive	1	1	2	4
Diarrhoea	1	1	1	3

during the neonatal period with a median age of 7 days ranging from 2 to 28 days. Ten (22%) presented in the first year of life with a median age of 3 months ranging from 5 weeks to 6 months. Fourteen (30%) presented between the ages of 1 and 17 years with a median of 7 years.

Bilious vomiting is classically described as the symptom present in intestinal malrotation and, in our series, this was evident [1] It was present in 50% of neonates and in 33% of those older than 1 month. Of note is the length of the history of bile-stained vomiting with 9 of the 19 patients suffering from bilious vomiting showing the symptoms for more than 24 h. In those presenting acute symptoms ($n = 37$), 78% were operated upon within 12 h of presenting themselves for the first check-up. One neonate had definitive bilious vomiting for 13 days prior to surgical referral while a 4-month-old child revealed symptoms of it since birth (Table 1).

The second commonest complaint was non-bilious vomiting in 10 (22%) patients who had a prolonged history of effortless vomiting. Failure to thrive, pain and diarrhoea were the major presenting symptoms in 4 (9%), 5 (11%) and 3 (7%) patients, respectively. One child presented with a palpable mass and another with chronic pain for 2 years. Two children displayed acute symptoms following surgery. A one-year-old child with Hirschsprung's disease presented with bowel obstruction following closure of a colostomy. On exploration, he was found to have Ladd's bands that were causing a distinct small bowel obstruction. A 7-year-old boy presented symptoms following percutaneous endoscopic gastrostomy formation with acute gastric dilatation. Malrotation was discovered incidentally at the time of surgery in three cases. Six asymptomatic patients with cardiac isomerism who had elective contrast studies to detect malrotation underwent elective surgery, as was the unit policy. Malrotation was associated with other congenital anomalies in 14 (30%) children.

A standard Ladd's procedure was performed in 44 cases while a laparoscopic Ladd's procedure was performed in two. Although acute midgut volvulus was detected on exploration in 30 patients, none required bowel resection. Appendectomy occurred in 37 (80%) patients. The appendix was not detected in one patient. Caecopexy was not performed. The median post-operative stay was 6 days ranging from 3 to 10 days.

Complications occurred both early and late (Table 2). All wound infections occurred within the first week post-operatively. Malabsorption necessitated dietary supplementation in three patients for a mean of 6 months. None developed any of the complications of short gut syndrome. Admission to the intensive care unit was required for one child with post-operative pneumonia. A further patient with chylous ascites was re-admitted for paracentesis. A 4-year-old developed chronic abdominal pain for 2 years following surgery for which all investigations were negative at the time the pain resolved. In total, 12 (26%) patients were re-admitted within the first 6 months after the initial operation.

Eleven (24%) children developed bowel obstruction. Six (12%) patients required multiple admissions due to small bowel obstruction. One child developed six separate episodes of small bowel obstruction. Conservative management was attempted in all 11 patients but six children required surgery. Adhesion-related obstruction was identified in four of the cases and an internal hernia was present in two.

There were four deaths within the group. One died due to post-liver transplant for biliary atresia, another from congenital cyanotic heart disease and a third from sudden infant death syndrome. There was only one death associated with intestinal malrotation and subsequent surgery. The child had cerebral dysgenesis and diabetes inspidius. Malrotation was diagnosed in the child at the age of 7 due to bilious vomiting and a standard Ladd's procedure performed. Six months post-operatively, he developed small bowel obstruction that was successfully managed conservatively. The following year, he was admitted with acute obstruction, sepsis and shock. He underwent an urgent laparotomy revealing an internal hernia and necrotic terminal ileum requiring a right hemi-colectomy and terminal ileostomy. He died 24 h after surgery for systemic sepsis.

Table 2 Incidence of post-operative complications

Complications	Incidence
Wound infection	4 (9%)
Feeding difficulties	4 (9%)
Pneumonia	1 (2%)
Chylous ascites	1 (2%)
Constipation	1 (2%)
Abdominal pain	1 (2%)
Small bowel obstruction	11 (46%)

Discussion

Ladd first described his procedure to treat malrotation and volvulus in 1932 and, since then, it has been the definitive treatment for intestinal malrotation [3, 4]. Ladd's procedure consists of initial untwisting of the volvulus. Secondly, Ladd's bands, which are thick peritoneal bands running from the caecum to the right upper quadrant and to the duodenum, are divided. The

ligament of Treitz is taken down and the duodenum is mobilised to the right and straightened. The narrow base of the mesentery is broadened and adhesions along the small bowel are divided. Finally, the entire bowel is returned to the abdomen in a non-rotated position. The small bowel is on the right and the large bowel is predominantly on the left. Appendectomies can also be performed. The focus in majority of the medical literature is on correct and early identification of the symptoms of malrotation, appropriate investigation and a prompt Ladd's procedure [2, 5]. The pre-operative consent and management of malrotation focuses on the risk of volvulus and its complications such as poor feeding, bowel resection and short gut syndrome. Our data reveals that post-operative complications occur in 46% following an uncomplicated Ladd's procedure.

Malrotation is a recognised risk factor for adhesion related obstruction in neonates [6]. Adhesion formation is in part due to the nature of the operation with the peritoneal mesentery being incised to allow broadening of its base, which in fact exposes more raw area for adhesion formation. The most significant complication within our group was adhesion-related obstruction and mortality associated with it. Although small bowel obstruction has been noted following Ladd's procedure, the incidence has been quite low. In 1980, Stauffer et al. reported 5 (12%) of 41patients required further surgery for bowel obstruction [7]. Mehall et al. [8] in 2002 reported similar results with persistent symptoms following surgery occurring in 11–13% and small bowel obstruction in 11%. Adhesion-related obstruction is most common within the first post-operative year but can occur any time. Late complications have been noted with 8(14%) of 57 requiring further surgery in one study [9]. Despite the fact that no bowel was resected at the initial surgery in our group, eleven (24%) children developed bowel obstruction with six requiring further surgery. Adhesion obstruction occurred in all the age groups and was not related to their initial presentation, their comorbidities, the presence of volvulus at the initial surgery or the length of their post-operative stay.

It is important to remember that intestinal malrotation represents a wide range of anomalies and thus, unsurprisingly, can present with a wide range of symptoms [10]. It is the variation from the classical presentation during the neonatal period hence the term ''volvulus neonatorum'' with bile stained vomitus and a soft non-distended abdomen that accounts for the delays in diagnosis. Studies had reported a low incidence outside the neonatal period [11–13]. However, Malek et al. [14] assessed the frequency of intestinal malrotation in 27 states in US in 2000 and reported that a total of 777 children required a non-elective Ladd's procedure of which 219 (28%) were older than 1 year. Beyond the neonatal period, malrotation may present with bilious vomiting and bowel obstruction; however, more commonly, a more chronic course with abdominal pain, diarrhoea, vomiting, and failure to thrive is present [11–13, 15]. A chronic malabsorptive-like syndrome can occur in patients with chronic midgut volvulus [16]. The vague nature of these symptoms which can persist into adulthood requires a high level of clinical suspicion of intestinal malrotation [17–19]. A small number is diagnosed after an upper intestinal barium meal for investigation of gastro-oesophageal reflux and was discovered incidentally during abdominal surgery. Delays between the initial presentation and the correct diagnosis have been reported up to 5 years [12]. Within our study, symptoms of chronic pain or non-bilious vomiting were confused with gastro-oesophageal reflux in seven patients. The delay in bilious vomiting prior to either upper gastro-intestinal contrast study or surgical referral is a matter of concern. The axiom that bilious vomiting must always be investigated promptly in all children must be reiterated [2].

A plain abdominal X-ray may show the characteristic ''double bubble'' but it is not reliable [17]. Barium enemas have been used to identify the position of the caecum but have been reported normal in 40% of patients with confirmed malrotation [10]. Upper gastrointestinal contrast studies is the investigation of choice in those with bilious vomiting or chronic vomiting with a sensitivity of up to 94% [9]. Abnormal rotation is detected if the duodenal loop does not cross the midline or if there is evidence of an obstruction or the corkscrew pattern. Ultrasonography is sensitive in detecting inversion of the superior mesenteric artery vein but the air within the small bowel or gastric distension can make imaging difficult; this makes upper intestinal barium investigation the investigation of choice [2]. Malrotation is commonly associated with other congenital anomalies with mortality within the group predominant due to these congenital abnormalities [20].

The question of whether asymptomatic patients with a documented malrotation require surgery is still controversial. Many authors recommend elective Ladd's procedure in all patients with intestinal malrotation [1, 21, 22]. In the past, we routinely investigated patients with isomerism who electively went in for treatment of malrotation. Choi et al. [23] reviewed 177 patients with heterotaxia over a 35 year period and found that asymptomatic patients had a low risk of intestinal volvulus and advised that routine investigation, screening and elective surgery was not necessary with close follow-up. However, the concern after diagnosing malrotation on a contrast study and opting for conservative management after volvulus occurs is a terrifying medico-legal nightmare. The potential for volvulus is life long and the complications associated with it do not alter with age [2, 11]. In our institution, we have decided to continue offering elective Ladd's procedures for symptomatic patients but only after detailed parental education and discussion regarding the potential complications of surgery, the feasibility and risks of follow-up in the non-operative group.

Conclusions

Our findings demonstrate that malrotation can present itself in any form and at any age. Bilious vomiting must be promptly investigated whatever one's age. Intermittent vomiting, epigastric pain and failure to thrive do not always imply a diagnosis of gastrointestinal reflux. An upper gastro-intestinal barium meal and follow through should be performed in all cases of bilious vomiting as well as in patients suffering from chronic vomiting in order to rule out malrotation. There is substantial risk of readmission following Ladd's procedure even in uncomplicated cases. Complications can affect up to 46% of patients. Small bowel obstruction occurs in 24% of patients requiring multiple hospitalisation and further surgery. We suggest that long-term follow-up is required for all patients post-Ladd's procedure and that a detailed explanation is given to parents regarding the risk and symptoms of bowel obstruction in order to decrease the morbidity of this condition.

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