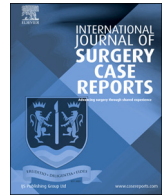


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# Intestinal malrotation in an adult patient with other congenital malformations: A case report

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## ABSTRACT

**INTRODUCTION:** Intestinal malrotation refers to a variety of abnormalities which occur between weeks 5–12 of embryological development. Most presentations occur before the first year of life. However, patients persisting beyond this period report chronic abdominal symptoms making it difficult to diagnose. Although uncommon, it is important that emerging surgeons and radiologists are made aware of the diagnosis and management of adult intestinal malrotation cases.

**PRESENTATION OF CASE:** We present the case of a 40-year old patient admitted with subacute abdominal pain on a background of chronic abdominal pain, alternating constipation and diarrhoea requiring several previous hospitalisations and other congenital malformations. Outpatient computed tomography (CT) of her abdomen demonstrated intestinal malrotation and emergency laparotomy revealed Ladd's bands compressing the duodenum. Ladd's procedure was performed and she had an uncomplicated recovery in hospital.

**DISCUSSION:** Intestinal malrotation can present acutely as volvulus mimicking an obstruction or more commonly, as chronic symptoms such as intermittent cramping, alternating constipation and malnourishment. Gold standard diagnosis in adults is by computed tomography imaging with oral contrast demonstrating inappropriate bowel position and/or inversion of superior mesenteric vessels. It is accepted that the definitive management is via the Ladd's procedure although there is controversy regarding when laparoscopy or laparotomy should be considered.

**CONCLUSION:** Intestinal malrotation is uncommon amongst adults but its complications can be devastating if not recognised early. Ladd's procedure either laparoscopically or via laparotomy can provide good resolution of symptoms if performed astutely.

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## 1. Introduction

Cases of adult intestinal malrotation are rare but can have devastating consequences if not recognised early. Usually, adults with intestinal malrotation report chronic abdominal symptoms such as intermittent abdominal pain and vomiting, bloating, malabsorption and alternating constipation and diarrhoea [1–3]. Gold standard diagnosis in adults is usually with cross-sectional imaging with oral and intravenous contrast (computed tomography (CT)), although it is often encountered intra-operatively as well [4]. There is still no means of predicting which patients will proceed to midgut volvulus or bowel ischaemia [5]. Due to the low incidence and nonspecific presenting symptoms, it is important that emerging general

surgeons and radiologists are made aware of this diagnosis and subsequent management.

The standard for definitive management is with Ladd's procedure which can be performed via laparotomy or laparoscopically. Adult patients undergoing either surgical approach have good resolution of both acute and chronic symptoms.

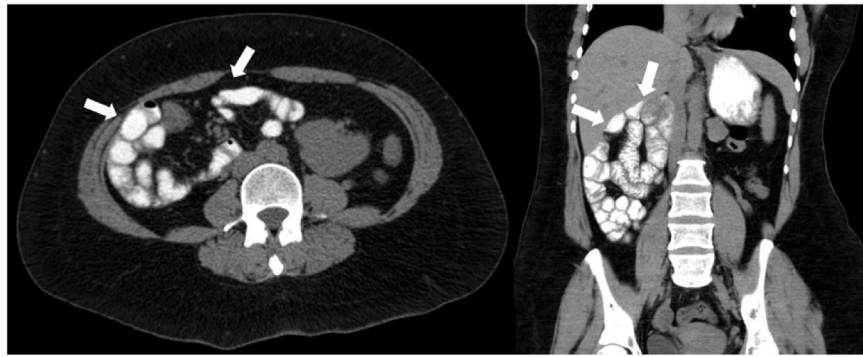
We present a unique case of a 40-year old lady with a background of congenital abnormalities including Goldenhar and Klippel-Feil syndrome and contrast-media allergy who presented with acute on chronic abdominal pain. An approach for diagnosis, surgical management and when to consider laparotomy or laparoscopic Ladd's procedure for the modern-era general surgeon is elucidated. This case has been reported in line with the SCARE criteria [6].

## 2. Presentation of case

A 40-year old lady presented to our emergency department with generalised abdominal pain. She was noted to be opening her bow-

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**Fig. 1.** Axial and coronal CT images with barium oral contrast, of our patient with arrows demonstrating all small bowel positioned on the right side; this is diagnostic of malrotation.

els and was not vomiting on initial presentation. Five days previous to this, she had presented and was admitted to another peripheral hospital with similar symptoms. During the admission, she was noted to have a normal abdominal ultrasound, her pain improved and she was discharged home.

On presentation to our hospital, her respiratory rate was 18 breaths per minute, oxygen saturation was 98% on room air, blood pressure was 121/75 mmHg, heart rate was 55 beats per minute and she was afebrile at 36.5 °C. Physical examination revealed a tender abdomen especially in the epigastric region with voluntary guarding.

She was known to have other congenital abnormalities including Goldenhar Syndrome and Klippel-Feil Syndrome. She described a background of chronic abdominal pain with alternating diarrhoea and constipation requiring many previous hospital admissions. However, she explained that her abdominal pain on this presentation was different (sharper and cramping). She had multiple previous gastroscopies and colonoscopies which had revealed no obvious abnormalities, a cholecystectomy and an open duplex appendectomy over 25 years ago in a peripheral hospital. The patient also described that she had diagnostic laparoscopies performed by a gynaecologist for her chronic ‘abdomino-pelvic pain’ for possible endometriosis; she was never formally diagnosed with any gynaecological abnormality.

Despite several years of chronic abdominal pain, the patient did not have any contrast studies or CT of her abdomen because she had a severe allergy to iodine contrast media. However, she presented on this occasion with an outpatient CT (with oral barium contrast, no intravenous contrast) which demonstrated all of the small bowel on the right side of her abdomen (Fig. 1).

The general surgeon to whom she was known to was on call for acute surgery the day she had presented and immediately reviewed the patient. She was kept nil by mouth, commenced on IV antibiotics and booked for emergency surgery.

A midline laparotomy was performed under general anaesthesia by the consultant general surgeon. Intra-operatively, the patient was noted to have a mobile caecum and the small bowel was noted to all be on the right side of the abdomen (Fig. 2). Ladd’s bands from the caecum to the right abdominal wall compressing the duodenum were divided and the Ladd’s procedure was completed by fixing the caecum to the left upper quadrant (Figs. 3 & 4). None of the small bowel was noted to be ischaemic or necrotic and bowel resection was not necessary. Due to previous appendectomy, this part of the Ladd’s procedure was not necessary.

She had an uneventful post-operative recovery and was discharged eight days after laparotomy. She was followed up by the surgeon at three and seven weeks post-operatively and there were no major post-operative complications. The chronic abdomi-

nal pain she had pre-operatively had completely resolved and the patient was content with the outcome.

### 3. Discussion

Midgut malrotation is a congenital anomaly of intestinal rotation. It is estimated that more than 90% of patients with intestinal malrotation will present in the first 12 months of life [7,8]. However, some children may escape this period if they were asymptomatic or only had vague abdominal symptoms which were misinterpreted for another cause [5]. Diagnosis is rare and unexpected in adults due to presentation with non-specific symptoms such as colicky abdominal pain, bloating, chronic vomiting, malabsorption/inability to gain weight and alternating diarrhoea and constipation [1–3].

Midgut malrotation occurs due to the failure of the normal 270° anti-clockwise rotation of the midgut along its vascular pedicle as



**Fig. 2.** Small bowel noted completely on right side of abdomen.

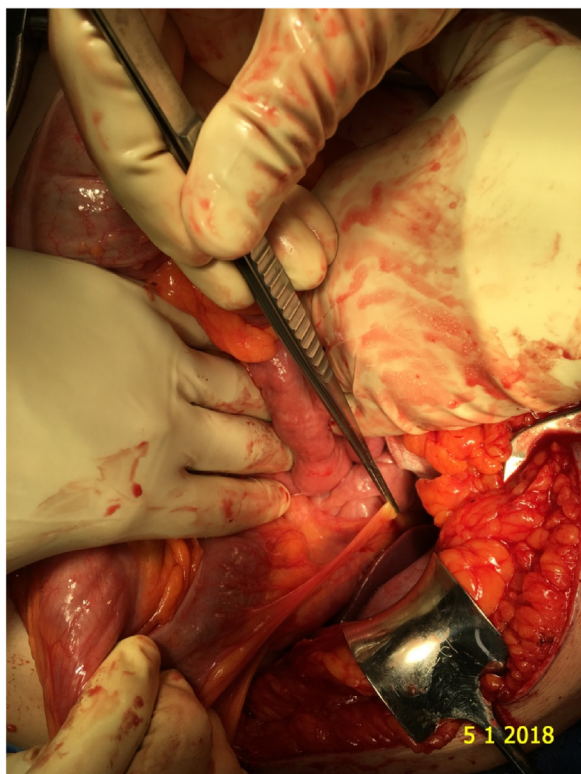


Fig. 3. Ladd's band extending from right abdominal wall to caecum.

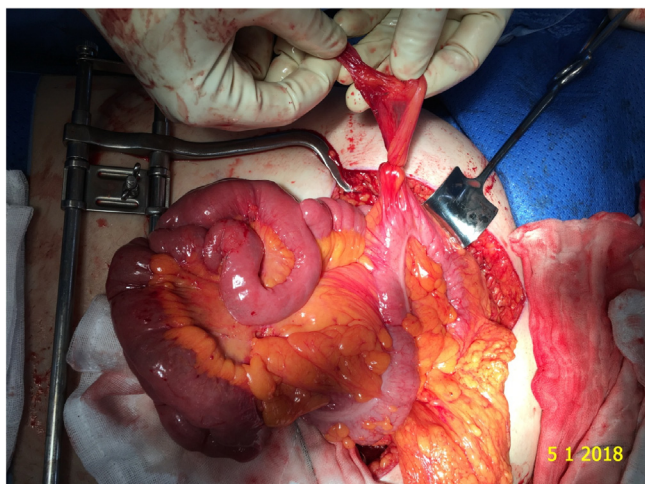


Fig. 4. Ladd's band after division.

it returns from herniation from the umbilical cord in the 5th to 12th week of embryological development [7,9,10].

The clinical importance of malrotation abnormalities lies within two important features: an abnormally positioned duodenojejunal loop compressed by peritoneal Ladd's bands causing obstruction of the duodenum and the narrow fixation of the base of the mesentery placing the SMA at risk of volvulus [9].

Adult presentation of midgut malrotation may present in two distinct patterns: acute and chronic [12]. Vague chronic symptoms such as abdominal pain, bloating, intermittent constipation and diarrhoea and malabsorption are more common in adults [12]. As highlighted in our case report, our patient had a background of years of abdominal pain and varying bowel habit but presented with subacute abdominal pain during this admission. Acute presentation (sudden onset abdominal pain, vomiting and obstipation) is

due to midgut volvulus, where the small bowel twists in clockwise manner due to the narrow base of the mesentery [13].

It is suggested that the gold standard for diagnosis for intestinal malrotation is either an upper gastrointestinal (UGI) contrast series or CT imaging with oral and intravenous contrast [2,4]. While the use of UGI series is common in children, CT has been more favoured in adults. CT can demonstrate inversion of the superior mesenteric artery and vein, bowel position and viability and volvulus (whirlpool sign) if present [4,14].

It is unusual that despite previous abdominal operations, that malrotation remained undiagnosed in our patient. Intestinal malrotation is not likely to be observed through an open appendicectomy incision or during a laparoscopic cholecystectomy, as bowel is not routinely examined. Although no gynaecological abnormality was found during diagnostic laparoscopies performed by gynaecology, it is unlikely they sought intestinal malrotation as the cause of her chronic symptoms. Furthermore, it is possible that in the pursuit of common adult pathology that the diagnosis of intestinal malrotation was overlooked. The striking aspect in the case of our patient was her severe allergy to contrast media and therefore the lack of previous CT imaging despite several presentations to hospital with abdominal pain. She presented soon after having an outpatient CT with premedication, which was instantly diagnostic of intestinal malrotation.

There are controversies regarding management of patients found to have incidental intestinal malrotation intra-operatively or incidentally on CT. Often, further questioning of these patients yields a history of nonspecific abdominal pain and symptoms. There is no conclusive imaging modality that can exclude the catastrophic outcomes of intestinal malrotation (bowel ischaemia and necrosis) [2,15]. It would be reasonable to suggest that any symptomatic (even if nonspecific) undergo elective surgical management, if they are appropriate surgical candidates, as there is no way to predict which of these patients will proceed to have catastrophic sequelae from their intestinal malrotation [5,13,16,17].

Surgical management of intestinal malrotation at any age is by the Ladd's procedure [3]. This procedure was first described by William Ladd in 1936 and consists of the following steps [2,12,18,19]:

- 1) Division of Ladd's bands (fibrous bands) lying over the duodenum to the caecum
- 2) Widening of the narrowed root of the mesentery
- 3) Counterclockwise detorsioning of the midgut volvulus if present and inspecting the bowel to observe if bowel resection is required
- 4) Appendicectomy if required
- 5) Placing the small bowel to the right and fixing the colon to the left

Most adult patients have resolution of their symptoms of intestinal malrotation after the Ladd's procedure [16].

Ladd's procedure can be performed through midline laparotomy or laparoscopically. Usually if an adult patient presents acutely, as in our case, it is reasonable to proceed to midline laparotomy [3]. Laparoscopy appears to be safe and effective when performed by experienced laparoscopic surgeons and usually in the absence of volvulus [12]. This suggests that laparoscopy is perhaps an option when intestinal malrotation is found incidentally in adult patients and an elective setting is chosen. Other factors to consider include the skill of the primary surgeon, if the diagnosis is unclear or if diagnostic laparoscopy is being used prior to definitive management.

The Ladd's procedure yields excellent results in all ages. Most adult patients, as in the case of our patient, report complete resolution of their symptoms.

An important feature noted in our case is our patient's previous congenital abnormalities. Although not intellectually disabled, our patient had been previously diagnosed with Klippel-Feil and Goldenhar syndromes and duplex appendix. All rare congenital malformations not associated with intestinal malrotation in previous articles. However, a study by Husberg et al. describes that concomitant congenital malformations were observed in 38% of adult patients with intestinal malrotation [17]. This suggests that intestinal malrotation and volvulus may be worth suspecting in adult patients with known congenital disorders.

#### 4. Conclusion

Intestinal malrotation is an aberration of midgut embryological development. Although a common diagnosis amongst neonates, it is uncommon and unsuspected in the adult patient. Although it can present as an acute abdomen, it tends to present in adults as it has done so in our patient; as subacute or chronic symptoms. Because of its low incidence and its nonspecific presentation it is neither suspected nor detected by surgeons or radiologists until possibly, when severe consequences such as ischaemia or necrosis have occurred. When surgeons encounter this diagnosis in symptomatic adult patients who are deemed appropriate surgical candidates, arrangements should be made to proceed to surgical management by the Ladd's procedure either laparoscopically or by laparotomy. The two main factors to consider when deciding the surgical approach is the acuity of the presentation and the skill of the primary surgeon. Furthermore, acute on chronic abdominal pain in adult patients with other congenital malformations should raise suspicion of intestinal malrotation.

#### Conflicts of interest

There are no conflicts of interest to declare.

#### Funding source

We have not received any funding for this research.

#### Ethical approval

Ethical approval was not obtained from the institution as the article is a complete de-identified case report and nil experimentation was performed. However, clear written consent was obtained from the patient regarding photographs and information that was going to be published (see below).

#### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Author contribution

Dr Pratima Herle – Data collection and analysis, Reviewing recent literature, writing the paper, editing the paper.

Dr Tushar Halder – concept planning, writing the paper, editing the paper.

#### Registration of research studies

N/A not first in-man case report.

#### Guarantor

Dr Pratima Herle & Dr Tushar Halder.

#### Provenance and peer review

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