ORIGINAL ARTICLE

Adult intestinal malrotation; a significant surgical entity

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Objective: The aim of this study was to review diagnosis and surgical management of adult patients with intestinal Malrotation which presented to our institute.

Patients and methods: A retrospective review of the surgical outcome of adults with intestinal malrotation was performed. Twenty patients were observed and treated between November 1984 and January 2014 (8 women and 12 men; the mean age of the patients was 40 years) at Aga Khan University Hospital, Karachi. Patient demographics, surgical outcomes, including perioperative morbidity and mortality were measured.

Results: Most of the patients were symptomatic at the time of presentation. Abdominal pain was the most frequently occurring symptom. CT scan abdomen was done in 14 patients. In 5 of these patients diagnosis was missed. 8 patients had upper GI contrast study. 16 patients underwent laparotomy and four were managed conservatively. Follow-up ranged from 2 to 52 months. Two patients had post-operative complications: both had adhesive obstruction and were managed conservatively in both the cases. No deaths occurred as a result of surgical intervention in the first 30 day period.

Conclusions: Intestinal malrotation is a rare but important cause of abdominal pain in adults. Patients may present with variety of nonspecific clinical symptoms and signs. Successful management includes early diagnosis and appropriate management.

Key words: adult intestinal malrotation, midgut volvulas, whirlpool sign, Ladd's procedure.

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Introduction:

Malrotation of the intestine results due to incomplete or failure of rotation of the gut during fetal development, the axis of this is based on the superior mesenteric artery.[1] It mostly manifests at an earlier age but may rarely present in the later life. It is hard to estimate the exact incidence of the late presenting adult variant of malrotation because majority of these patients present with acute signs within the first month of life.[2] As a consequence this may result in a twist compromising the blood supply of the bowel and eventually result in intestinal ischemia. The disease classically presents with an acute onset of mid gut obstruction resulting in vomiting with duodenal, midgut or cecal volvulus. In adult malrotation it becomes trickier to detect these cases due to lack of discrete symptoms.[3] The most consistent finding in adults is the presence of diffuse mild abdominal pain or discomfort which is attributed to the peritoneal bands and this may mimic several other conditions like irritable bowel syndrome, peptic ulcer disease, bilio-pancreatic disease, and psychiatric disorders.[4] Occasionally these cases may be incidentally discovered at laparotomy. In these circumstances it is imperative that adult general surgeons be aware of the embryologic origin and variants of the disorder to manage it effectively.[5] Ladd procedure is usually performed to correct this congenital defect which can benefit at any age, it is crucial that all surgeons operating on adult patients have an understanding of not only adult malrotation and its various manifesta-

tions, but also have a firm grasp of intestinal embryology and its anatomic variations[6].We aimed at reviewing our experience with these patients and to briefly reflect on this condition.

Methodology:

A retrospective review of records was performed for all adult patients over the age of 16 years who had presented to Aga Khan University Hospital, Karachi, between November 1984 and January 2014 and had been diagnosed to have intestinal malrotation. Data recorded included information about patient demographics, presentation, diagnostic investigations, Management, postoperative morbidity and mortality, length of hospital stay and followup. Means and proportions were calculated using SPSS 19.

Results:

Total of 12 patients were diagnosed to have intestinal malrotation. Male to female ratio was 2:1. Mean age at presentation was 40 years, ranging from 23 – 64 years of age. Most of our patients were symptomatic (n=14). Those who were asymptomatic (n=6) were diagnosed to have intestinal malrotation while being investigated or treated for some other cause. The most frequently occurring symptom was abdominal pain (n=12), followed by vomiting (n=6), nausea (n=4), constipation (n=3), early satiety (n=2) and weight loss (n=2). Duration of symptoms ranged from 1 to 120 months. Almost all the patients who were symptomatic in our series had a delay in diagnosis most likely due to their non specific symptoms and lack of suspicion because of low incidence while making judgment of clinical diagnosis.

CT scan was done in 14 patients. Diagnosis was missed in 5 patients, which was confirmed on subsequent small bowel follow through in 1 patient and on surgical exploration in 2 patients. Small bowel follow through was diagnostic in 8 patients. Barium enema was done in 2 patients, along with small bowel follow through.

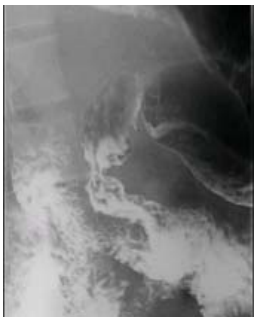
One patient did not undergo any investigation and was diagnosed on surgical exploration because of gunshot abdomen.

Out of 20, sixteen patients underwent surgical intervention. 14 patients underwent standard Ladd's procedure. Appendectomy, division of ladd's bands & widening of small bowel mesentery was done in all 14 ladd's procedures. Cecopexy was done only in initial two procedures. 1 patient who underwent Whipple procedure because of periampullary CA also had appendectomy and widening of small bowel mesentery. 1 patient who underwent exploration because of gunshot abdomen had repair of jejunal perforations and appendectomy as well. Patients who were managed conservatively had advanced malignancy.

4 patients had postoperative adhesive bowel obstruction, both were managed conservatively. No mortalities were observed in our patient group.

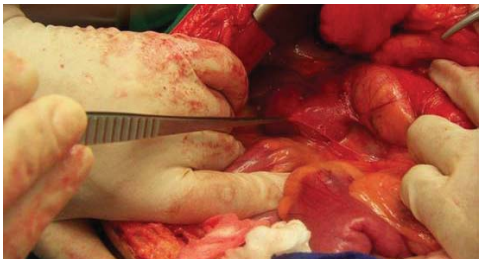
Mean length of stay was 10 days (range 1 – 30 days). Follow up was available from 2 – 52 months.

case 1, Fig.1: Upper GI Contrast: DJ and small intestine lying in right hemi-abdomen



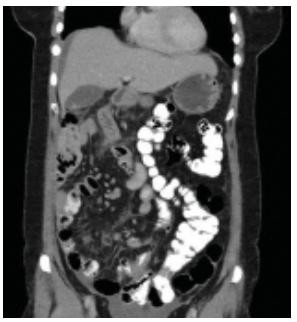
case 1, Fig. 2: Upper GI contrast: partial obstruction at D2(because of external compression by Ladd's band

Case 1, Fig.3 operative picture: small intestine on right side of field and large bowel on left side

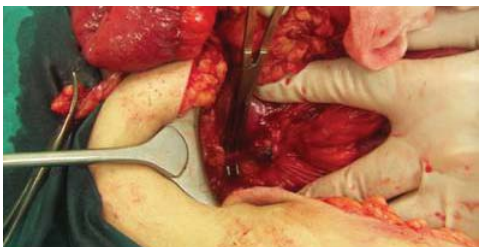


case 1, Fig.4:
Ladd's band (over duodenum) being picked up by forceps

case 1, Fig.5
Ladd's band (over duodenum) being divided



case 2 Fig. 1:
CT scan abdomen (coronal reconstruction): DJ and small intestine lying in right hemi-abdomen and large bowel occupying left hemi-abdomen



Case 2 Fig.2:
operative picture Ladd's band being divided

Discussion:

Intestinal malrotation is not just a disease of children as the stigma goes. Adult general surgeons need to be familiar with the condition itself, signs and symptoms, management, outcome and prognosis. Our results highlight the need for awareness regarding the intestinal embryology and Ladd's procedure although it is not a condition encountered frequently by adult general surgeons.

During embryonic development, normal intestinal rotation starts around 4th week and continues till birth; abnormalities occur during this period when normal intestinal fixation and rotation fail to occur normally.[7-8] Malrotations can present in various forms including, complete, incomplete, reversed or nonrotation depending on the axis of vessels and the intestinal rotation.

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Symptoms of intestinal malrotation are linked to the anatomical anomalies which occur during embryonic life.9 For instance the cecum is anchored to the retroperitoneum by fibrous bands known as the Ladd's bands which cross anterior to the duodenum and can cause partial obstruction hence resulting in recurrent abdominal pain, nausea and vomiting. Also the non rotated midgut has a small bowel mesentery thus increasing the risk of clockwise rotation resulting in midgut volvulus. Appendix in nonrotated or partially rotated mid gut can be present anywhere in the abdomen and appendicitis can be easily missed if the surgeon is unaware of malrotation. [10]

Nausea, vomiting, abdominal pain, bloating and weight loss are the vague symptoms such patients present with and these symptoms become so chronic over period of time that these patients are labelled as psychosomatics and the underlying condition is often overlooked until they present in the emergency room.[11-12] CT scan abdomen can prove to be diagnostic if certain confirmatory signs such as small bowel in right hemi abdomen, abnormal relationship between SMA and SMV or whirlpool sign in which mid gut is rotated around axis of SMA.[13] The gold standard for the diagnosis remains upper GI contrast study along with barium enema.[14] DJ flexure appears to lie on the right side of midline and follow up films show an abnormally placed small bowel. Barium enema confirms the abnormally placed cecum which could be anywhere except he right iliac fossa. But both these investigations are in lieu with one another, if done in isolation neither can rule out intestinal obstruction.[15]

The essential component in management of intestinal obstruction is awareness regarding the Ladd's procedure. Every adult general surgeon needs to be aware of the procedure, its technique and indications. The operative technique consists of division of Ladd's bands, mobilization of duodenum, derotation of midgut volvulus, mesenteric widening and appendectomy.[16] Initially cecopexy was a component of the procedure but over the years after much research it was found to have limited role in outcome so it is not performed anymore. The dilemma which arises now is whether to perform Ladd's procedure in every patient of intestinal malrotation whether asymptomatic or symptomatic. A lot of times malrotation is discovered while the patient is being operated upon for any other reason and here the question arises whether Ladd's procedure should be performed without informed consent since it's a completely different procedure.[17]

There are different schools of thoughts regarding this dilemma. Choi et al after reviewing 177 patients with midgut volvulus who were asymptomatic recommended conservative management with vigilant follow up since even after performing Ladd's procedure electively, the possibility of a future volvulus cannot be ruled out. [18] But on the other hand several authors after conducting studies on abnormal positioning and rotation of intestine recommend an elective Ladd's procedure to prevent an abdominal catastrophe.[19] But the mystery still needs to resolved and further studies need to be conducted to find a definite solution to the query along with awareness of midgut volvulus and Ladd's procedure in adult general surgeons.

Conclusion:

As a general rule intestinal malrotation presents in childhood. Nevertheless a significant number of people may carry this condition to adult life, being asymptomatic for many decades, but at a higher risk of developing an abdominal catastrophe (midgut volvulus). A general surgeon may encounter such case while practicing, and lack of clinical suspicion might result in to misdiagnosis and severe consequences for the patient. Successful management of such patients is dependent on a well-informed surgeon, accurate diagnosis, and prompt and appropriate management. In 1932, William Ladd1 cautioned in his landmark article, "the condition is rare enough so that it is likely to escape the mind, and it is common enough to be important".

Conflict of Interest: None

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