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ORIGINAL ARTICLE

Malrotation of midgut in adults, an unsuspected and neglected condition—An analysis of 64 consensus confirmed cases

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Abstract
Introduction Malrotation of midgut is considered to be a condition of childhood. This study evaluated malrotation in adults with recurrent abdominal pain (RAP).
Methods Sixty-four consensus-confirmed cases of intestinal malrotation were reviewed. The diagnosis was based on radiological criteria, and the consensus was arrived at by at least three of the five authors in any individual case.
Results Abnormal duodenojejunal junction (DJJ) was a consensus finding in 64 cases referred for RAP. Most were in their fourth decade of life, and 12 were beyond 60 years. Besides RAP, intolerance to food was the next common symptom. Acute intestinal obstruction was seen in 16. Forty-two of 64 patients consented for surgery. Ladd's procedure was the commonest. All patients who underwent surgery were symptom free except for two, of which, one had liver cyst and the other had hernia. Of those who refused surgery (22), all had continued symptoms and 10 patients took alternative therapies. On follow up of initially unwilling patients (for surgery) with abnormal DJJ, only eight consented for surgery; three underwent open Ladd's procedure, and one had laparoscopic Ladd's done.
Conclusion Malrotation is not uncommon as a cause of RAP in adults.

Keywords Duodenojejunal junction · Intestinal obstruction · Ladd's procedure · Malrotation of midgut · Recurrent abdominal pain · Volvulus

Introduction

Malrotation of midgut was and is conventionally taught to be a condition of infancy [1–4]. Although most of the cases are described in first month to first year of life, malrotation can present even in adults. This is obvious in most reports being case reports. Malrotation of midgut has been mistakenly considered to be always associated with volvulus, a myth that continues, a myth that does not get erased off the mindset of clinicians, and a myth that makes adult clinicians not to consider the entity in differential diagnosis of recurrent abdominal pain (RAP). RAP in adults is a heterogeneous group of conditions, ranging from adhesions, tuberculosis, chronic cholecystitis, acid peptic disease, irritable bowel syndrome (pain) to psychological overlay. Malrotation of midgut can be a cause of RAP, and the commonest presenting symptom of malrotation in adults is RAP.

The site of duodenojejunal junction (DJJ) is the most diagnostic sign on imaging of upper gastrointestinal tract. Here, we analyze 64 consensus confirmed cases of malrotation in cases of RAP in adults.

Methods

This is an analysis of a contiguous observational cohort of "consensus confirmed" malrotation in adults. From 1995 till 2012, 64 consensus confirmed cases were referred to the Department of Surgery and a Pediatric Surgery. Consensus was defined when the radiologist, physician, gastroenterologist,

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and surgeon jointly evaluated the imaging studies and accepted to the site of DJJ. At least three members were required at the time of the consensus meeting. The abnormalities noted were as follows: (1) exact site of DJJ by drawing a line at pyloroduodenal junction at L1, (2) exact DJJ in a given case either below L1 or on the spine or right of the spine, (3) superior mesenteric vein (SMV)/superior mesenteric artery (SMA) relationship, exactly noted if SMV is below or right side or above SMA, (4) straightened second (D2) and third (D3) parts of duodenum, (5) jejunal loops on right side, (6) hypertrophied jejunal loops, (7) clustering of jejunal loops, (8) splayed out mesentery, (10) whirlpool sign, and (11) other obvious signs of volvulus if any.

Results

Only consensus confirmed malpositioned DJJs were accepted for treatment. Other cases of RAP wherein any other lesion was seen were excluded. During the same time, 116 cases of RAP were referred and were treated for adhesions, bands, hiatus hernia, chronic calculus cholecystitis, and duodenal ulcer. Tuberculosis mesenteric lymphadenitis was also excluded when their tuberculosis was diagnosed by ESR, chest radiograph, history of exposure, Mantoux test, and, in some cases, TB PCR. In addition, there were four cases referred at first instance as malrotation by radiologist and were also excluded.

All children treated for malrotation were excluded. During the period, the malrotation treated by senior pediatric surgeon was as follows: a total of 124 cases with 71 babies less than 1 month, 42 children less than 1 year, and 11 more than 1 year to 5 years of age. There was a diagnostic difficulty to get a consensus on malrotation of midgut in RAP in children beyond 5 years of age. The trends were similar to the situation of RAP in adults. Despite being considered to be a disease of childhood, 30 out of 71 neonates, 11 out of 42 in infants, and 2 out of 11 pre-school children had volvulus at presentation and had hypertrophied mesentery and duodenum, chylous ascites, and hypertrophied proximal jejunal loops. Even bilious vomiting was often taken lightly by pediatricians and referred late.

Records of these 64 cases were evaluated using the following parameters: (1) age of referral, (2) symptoms at presentation, (3) number of visits to hospitals and physicians before referring to surgery, (4) various investigations done during those visits, (5) number of surgically treated patients, (6) relief of symptoms after surgery, and (7) symptoms of unoperated cases. These were the parameters chosen based on questions during the first conference presentation. Most cases of RAP with consensus confirmed malrotation of midgut were referred between 41 and 50 years of age. There were 12 patients beyond 60 years of age (Table 1). All patients had RAP, 41 had recently intolerance to food, and one fourth presented as acute abdomen (Table 2). Twenty-four out of 64 had at least ten visits, and 30 of them changed and visited more physicians (Table 3). All patients had upper gastrointestinal (GI) endoscopy at least once. Thirty-five patients had more than one upper GI endoscopy (Table 4). The commonest report was chronic duodenitis. Normal study was the next common report (Table 5). Upper GI contrast (barium meal follow through) was done at least once in all sometime during their evaluation for RAP, and 40 had upper GI series twice as there was no consensus. Ten patients had upper GI series thrice. The commonest diagnosis was normal study, normal C loop, and unreported DJJ till forced for a consensus. Consensus meetings were often inconclusive; the fact supported by only 64 cases had consensus confirmed abnormal DJJ. Many radiologists never thought of reporting the site of DJJ in normal study report. Consensus sometimes required fluoroscopic reconfirmation of site of DJJ with the consensus team. The observation during discussions was that the exact site of DJJ was the point of consensus. The commonest reason was overfilling proximal jejunal loops with contrast. Another reason was radiologists insisting the use of right anterior oblique view. Position of DJJ is best seen in true frontal projections, when spinal pedicles will be symmetrical and the place of DJJ can be related to the spine. The other radiological features looked for are cited earlier in text (Table 4).

Contrast-enhanced computed tomography (CECT) was done once in 51 patients, 38 patients had twice repeated, and 13 patients had three times CECT repeated. The commonest report was normal study and no mention of exact site of DJJ. The observation was that the more educated the patients, the more number of times the CECT. The radiologists often never considered malrotation when referred for CECT for RAP. Consensus team looked for relation to SMA to SMV, and whirlpool sign a sign of volvulus. Point of conflict was describing DJJ on CECT. Often, it was followed by fluoroscopic

Table 1 Agewise distribution of recurrent abdominal pain with abnormal duodenojejunal junction

Age (years)	Number	%
25–40	14	21.8
>41–50	22	34.3
>51–60	16	25
>61–70	9	14.0
>71	3	4.6

Table 2 Symptoms at presentation

Symptom	Number	%
Recurrent abdominal pain	64	100
Vomiting	18	28.1
Intolerance to food	41	64.0
Diarrhea	7	10.9
Acute intestinal obstruction	16	25

Table 3 Presurgical referrals

No. of visits	Number of patients	%
<10	24	37.5
11–20	30	46.8
>21	10	15.6

evaluation with the team. Radiologists were not comfortable at all to be precise on DJJ on CECT. Most of CECT reports ranged from normal study to multiple small volume mesenteric nodes and a small liver cyst in one. Obviously, radiologists were not comfortable and confident in reporting DJJ in CECT. Six patients had a report on a repeat CECT in coronal reconstruction malpositioned DJJ but confirmed after fluoroscopy together with the team. Routine reconstructions were not done by radiologists unless specifically asked and felt they take a lot of time and patients do not wait for report.

Color Doppler was done in 15 patients. The commonest report was a normal study. When probed by the team member, SMA relationship with SMV was reported. The commonest report was a normal study with normal portosystemic confluence. One patient had a report of SMV below SMA but did not conclude malrotation of midgut.

Twenty-seven patients had diagnostic laparoscopy, and operative findings reported only "no adhesions" followed by chronic appendicitis and multiple small lymph nodes in mesentery. No attempt was made to evaluate DJJ, and when the team retrospectively probed, the surgeon that felt that locating the DJJ was a major laparoscopic intervention and did not feel it to be necessary, as the rest of the abdomen was normal. Obvious observation was that malrotation was not in the surgeon's mind at the time of laparoscopy. Laparoscopic appendectomy was performed. These laparoscopies were performed for RAP when all reports were reported as normal and

Table 4 Investigations and the number of repetitions before referral to surgery

Investigation	Number of times investigation was done	Patients (n=64)	%
Upper GI endoscopy	Once	64	100
	>Once	35	54.6
Barium meal	Once	14	21.8
	Twice	40	62.5
	>3 times	10	15.6
CT scan	Once	51	79.6
	Twice	38	59.3
	>Twice	13	20.3
Color Doppler	Once	15	23.4
Laparoscopy	Once	27	42.1

Table 5 Details of reports of tests performed before surgery

Name of test	Reports
Upper GI endoscopy, n=64	a. Normal 34/64 b. Chronic duodenitis 23/64 c. Mild antral inflammation 7/64
Barium meal, n=64	a. Normal study 51/64 before consensus b. Jejunal mucosal fold thickening 2/64 c. Visualized appendix and normal 13/64 Consensus malposition of DJJ by team
CECT, n=51	a. Normal 61/64 b. High cecum 4/64 c. Small liver cyst 1/64 d. After consensus abnormal DJJ 6/64, but confirmed only after repeat fluoroscopic team observed study
Color Doppler, n=15	a. Normal study 11/15 b. SMV below SMA 1/64 c. Unremarkable 3/15
Laparoscopy, n=21	a. Normal 14/21 b. Chronic appendicitis 3/21 c. Multiple mesenteric lymph nodes 4/64

DJJ duodenojejunal junction, *CECT* contrast-enhanced computed tomography, *SMV* superior mesenteric vein, *SMA* superior mesenteric artery

multiple courses of H_2 blockers, and anti-spasmodic and even empirical anti-tuberculous treatment.

Table 6 shows presurgical referral medical treatment till consensus was achieved. All patients had multiple courses on H_2 blockers and antacids. Nearly half of the index series had empirical anti-tuberculous treatment before referring for surgery. Forty-two out of 64 were referred to psychiatrist; the commonest diagnosis was depression. Three patients had CT scan of the brain too; the observation made was when cornered by frustrated patients with RAP, and more courses of H_2 blockers were prescribed.

Table 7 shows the type of surgery performed. Only 42 out of 64 patients with consensus confirmed abnormal DJJ were accepted for surgery. Ladd's procedure of non-rotation and widening duodenocolic isthmus was done. Ladd's procedure and intestinal resection were done in 12, as they had evidence of ischemia presenting as acute abdomen. These patients with acute abdomen in retrospect gave history of RAP and visited a number of clinicians before landing in emergency. All patients' site of DJJ was marked in relation to the spine and

Table 6 Type of surgery performed in willing patients

Treatment	Number of patients n=64	%
Willing for surgery	42/64	65.6
Surgery (Ladd's)	30/42	71.4
Ladd's+resection	12/42	28.5
Refused to surgery	22/64	34.3

Table 7 Follow up of operated patients with malrotation

Years	Number of patients	%
<1	11	26.1
>1–3	2	4.7
>3–5	11	26.1
>5	8	19.0
Not followed up	10	23.8

pyloroduodenal junction. The images performed in the past for RAP of these 12 cases were reviewed by the consensus team and confirmed as malrotation. No other cause was found in these patients. Those who had resection also showed signs of chronic and recurrent volvulus with hypertrophy of mesentery and proximal jejunal loops. All operative findings were recorded and shown by the consensus team.

Twenty-two out of 64 refused surgery despite a consensus confirmation of abnormal DJJ and absence of any other cause. The observation made was that other physicians expressed doubt while they did not have any diagnosis to offer, and this led to reluctance to consent for surgery.

Table 8 shows the symptomatic relief of operated cases of RAP with consensus confirmed malrotation. Thirty-two out of 42 followed up for 1 to 8 years. In two patients having symptoms of persisting pain, one had small liver cyst and the other had case of internal hernia. Internal hernia was through a mesenteric defect and the patient was relieved of symptoms. The live cyst was reevaluated and found to have been mentioned in CECT but was not consistent with GI symptoms.

Table 9 shows the follow up of non-operated consensus confirmed abnormal DJJ with RAP. The follow up was done for 1 to 8 years through mails, telephone calls, and contacting the referring physicians. All of them continued to have symptoms despite physician and medication change. Ten out of 22 opted for alternative treatment like Unani, Ayurveda, and Homeopathy. At the end of 8 years, 8 out of 22 was accepted for surgery. Four of these eight undergone open Ladd's procedure, and one had laparoscopic Ladd's operation. Three are still awaiting for surgery.

Table 8 Symptomatic relief following surgery

Symptom	Number of patients	%
Symptom free	40	95.2
Vague symptoms	2	4.7
Concomitant lesions		
Liver cyst	2	4.7
Internal hernia	1	2.3

Table 9 Follow up of non-operated patients with malrotation

Symptoms	Non-operated patients $n=22$	%
Symptoms continued	22/22	100
Symptoms+alternate treatment (yoga, Unani, Ayurveda, Homeopathy	10/22	45.4
Willing for surgery at the end of 8 years	8/22	36.3
Open Ladd's procedure	4/8	50.0
Laparoscopic Ladd's procedure	1/8	12.5
Still waiting	3/8	13.6

Discussion

Malrotation of midgut in cases of RAP was unconsidered and missed in most patients leading to repeated investigations and empirical medication. Symptomatic relief in 40 out of 42 supports that malpositioned DJJ was responsible for symptoms. Even those who presented with acute abdomen in retrospect revealed multiple doctor visits for RAP and signify missing malrotation till they landed up in emergency and malrotation with intestinal obstruction. Then, Ladd's widening of duodenocolic isthmus relieved the symptoms also, which indicates malrotation as the cause of RAP in these patients.

Malrotation is often mistakenly considered to be "always" associated with volvulus and that is the cause of symptoms. RAP in adults is caused by myriad of conditions like adhesions, inflammatory bowel disease, cholelithiasis, internal hernias, complicated acid peptic disease, tuberculosis (visceral and or nodal or peritoneal), and irritable bowel syndrome (pain). Many standard books do not even mention malrotation in the list of differential diagnosis of RAP in adults. Pickhardt and Bhalla [5] and Berg [6] presented radiological features and presentation of malrotation in adults. Even these reports are based in case series. Gong et al. [7] and Dietz et al. [8] reported malrotation in adults presenting with duodenal obstruction. Gilbert et al. [9] and, Seymour and Andersen ([10] presented three cases of malrotation with RAP in adults. Emanuwa et al. [11] and Singh et al. [12] reported a case of malrotation presenting as acute intestinal obstruction. The index series also had patients presenting with acute intestinal obstruction. The very fact search reveals only case reports, which is a reflection not considering malrotation of midgut as one of the causes of RAP, and this observational cohort stresses a fact that malrotation of midgut does present in adults, and if searched for with focus, malpositioned DJJ can be picked up. Wanjari et al. [13] reported a case of malrotation treated by laparoscopic Ladd's procedure in a 17-year-old patient. Index series also show that Ladd's operation is the procedure to address malrotation. Fu et al. [14] reported 12 cases of malrotation in adults, confirmed by upper GI series. The authors also feel that radiologists are not comfortable in reporting site of

DJJ on CECT and relied more on barium upper GI series. Members of consensus confirmed malrotation team also mostly reported on upper GI contrast series. Fu et al. had some patients with laparoscopically confirmed malrotation.

We feel that DJJ position is underreported. Tacket et al. [15] have also suggested that focused and concerted efforts are required to detect and diagnose malrotation in adults with RAP.

Yousefzedeh [16] has questioned the very existence of DJJ but concluded that abnormal cecum left is more for a non-rotation. Retromesenteric D3 malrotation excludes volvulus, and pushing of D3 medially is more reliable. An ultrasound compression technique to see D3 may aid the diagnosis [16].

Abnormally located cecum and high positioned cecum are not valid to diagnose malrotation, as cecum may be normal in malrotation. But, they might be an indicator to search more pointedly the malpositioned DJJ. Normal position of DJJ is on left side of the spine at the level of pyloroduodenal junction. The relationship of SMA to SMV may be altered when volvulus complicates malrotation.

We conclude that malrotation of midgut is a rare cause of RAP in adults and should be looked for in cases where a specific cause of RAP in adults is not found.

Compliance with ethical standards

Conflict of interest GRP, JVSR., HF, MMS, AS, and GS declare that they have no conflict of interest.

Ethics statement The study was performed in a manner to confirm with the Helsinki Declaration of 1975, as revised in 2000 and 2008 concerning human and animal rights, and the authors followed policy concerning informed consent as shown in Springer.com

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