INTESTINAL MALROTATION IN THE OLDER CHILD: COMMON DIAGNOSTIC PITFALLS.

By

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ABSTRACT

8 children aged 4 to 14yrs (mean age 9yrs) with a clinical diagnosis of intestinal malrotation were studied prospectively over a 4yr period at the University of Nigeria Teaching Hospital Enugu. Despite, the reported normal barium studies of the upper gastrointestinal tract in the children, surgery were done based on strong clinical suspicion, chronic morbidity, and our interpretation of the barium studies. Operative findings were consistent with intestinal malrotation. 7 patients were symptom-free after 3months to 3yrs follow-up. One patient died of gastroenteritis 2 weeks post-operatively. A high index of suspicion and a close co-operation between Radiologists and Paediatric Surgeons are required for prompt and accurate diagnosis of intestinal malrotation in the older child.

ABSTRAIT

8 enfants âgés 4 à 14yrs (âge moyen 9yrs) avec un diagnostic clinique de malrotation intestinal ont été étudiés pour l'avenir sur une période 4yr à l'université de l'hôpital d'enseignement Enugu du Nigéria. Outrage, les études normales rapportées de baryum de l'appareil gastro-intestinal supérieur dans les enfants, la chirurgie ont été faites a basé sur le soupçon clinique fort, morbidité chronique, et notre interprétation des études de baryum. Les résultats effectifs étaient conformés au malrotation intestinal. 7 patients étaient sans symptômes après 3months au suivi 3yrs. Un patient est mort de la gastroentérite pendant 2 semaines postopératoirement. Un index élevé de soupçon et une collaboration étroite entre les radiologistes et les chirurgiens pédiatriques sont exigés pour le diagnostic prompt et précis du malrotation intestinal dans l'enfant plus âgé.

INTRODUCTION

Intestinal malrotation is a spectrum of clinical conditions resulting from anomalies of migration, rotation, and fixation of the midgut during fetal development[1,2]. The clinical manifestation can be acute and severe in neonates and infants as a result of midgut volvulus or duodenal obstruction. In contrast to neonates and infants, the clinical manifestations of intestinal malrotation in older children are chronic, non-specific, and often obscure, leading to delay in diagnosis and treatment. Intermittent, and partial intestinal obstruction give rise to prolonged, atypical and obscure symptoms.[3,4] Upper GIT contrast studies regarded as the gold standard for diagnosis of this condition [1,2,5,6] are often atypical in these older patients and may be wrongly interpreted. This paper describes the clinical features,

radiological features, operative findings, treatment and outcome of these children and calls for the attention of both clinicians and radiologists to these diagnostic pitfalls that lead to delay in treatment.

PATIENTS AND METHOD

We prospectively documented the clinical data of 8 patients aged between 4yrs and 14years seen from January 2000 to December 2003 with intestinal malrotation and whose upper GI barium studies were reported as normal. Collected data included age, sex, clinical features and findings on barium studies (both radiologist report and surgeon's impression). Also included are operative findings, operative treatment and outcome.

RESULTS

Table I shows the clinical data of the 8 children with malrotation. None of the radiologist's reports of the barium studies indicated malrotation. Our clinical diagnosis of intestinal malrotation was based partly on the chronicity of the symptoms (Table 1), our interpretation of the barium films, and partly on experience. This was confirmed by the operative findings which were consistent with malrotation - multiple congenital bands, defects in the mesentery, and chronic volvulus (Table 1). One patient (14 year old) who had had 4 previous admissions came in with intestinal obstruction, and malrotation was diagnosed at laparotomy (Fig 1V). Another patient (6 year old) who was severely malnourished and who had chronic volvulus at operation (Fig 11) died two weeks post-operatively from acute gastroenteritis (14.3% mortality).

DISCUSSION

Rotational anomalies occur when the proximal (duodeno-jejunal) and distal (coeco-colic) loops of the embryonic midgut fail to complete their required 270^0 counter clockwise rotation[1,7,8]. This rotations when completed ensures that the base of the small bowel is spread along a diagonal from the ligament of Trietz to the ileocaecal attachments, thereby stabilizing the midgut, and protecting it from volvulus and strangulation[1,8,9]. Malrotation results when the duodeno-jejunal loop remains to the right of the spine, but the caeco-colic loop rotates 180^0, passing in front of the superior mesenteric artery (SMA), and fails to descend to the right lower quadrant. This leaves the two points of fixation of the small bowel mesentery almost superimposed upon each other at the base of the SMA[1,2,8]. The resultant mesenteric attachment is narrow and predisposes to volvulus[10]. The presentation may be acute and persistent or chronic and intermittent. The latter picture is seen in the older children and is illustrated by the 8 cases reported in this study. Normal rotation and fixation of the midgut also results in the fixation to the posterior abdominal wall of the right and left colon and the duodenuim.[1,8]. Potentially, obstructive aberrant peritoneal bands (Ladd's bands) as well as mesocolic envelops form when the mesentery of these parts fail to become fixed retroperitonealy[1,7]. These may restrict small bowel loops to one or other side of the abdomen. Wrong interpretation of barium studies in this condition has been documented[5]. Similar reports in the literature show that the diagnosis of malrotation was not clear until exploration was performed[6,11]. Our findings are similar to other reports[1,12,13]

CONCLUSION

The common diagnostic pitfalls of intestinal malrotation syndrome in the older child arise from the fact that symptoms are usually chronic, non-specific, and obscure. Physical signs are often absent or unremarkable, and upper GI barium studies are often reported as normal. We therefore recommend that

the older child with recurrent attacks of abdominal colic should have a comprehensive evaluation. Their upper GI contrast studies should, preferably, be jointly reviewed by an experienced Paediatric Radiologist and the Paediatric Surgeon.

Table I: Clinical Data, Upper GI barium & Operative findings, in 8 children with intestinal malrotation syndrome

S/N	Age	SEX	Clinical Features	Upper GI Barium Studies (Surgeon's impression)	Operative findings
1.	4yrs	M	Intermittent abdominal pain & vomiting post-prandial. Weight loss.	Small bowel loops crowded to left side of abdomen	Multiple congenital bands. Sub-hepatic, mobile caecum.
2.	5yrs	M	Recurrent attacks of abdominal pain & vomiting. 3 previous admissions.	Small bowel loops crowded to the right side of the abdomen.	Multiple congenital bands around the D-J flexure. Mobile high riding caecum
3.	5 ½ yrs	M	Recurrent attacks of vomiting & abdominal pain. 2 previous admissions	Small bowel loops are crowded to left upper abdomen	Multiple congenital bands around the D.J junction which is to the right of midline. (Fig 1)
4.	6yrs	F	Intermittent vomiting & abdominal colic. 3 previous admissions	Small bowel loops restricted to the left side of the abdomen	Multiple congenital bands around D-J junction. Chronic volvulus. (Fig 11)
5.	8yrs	M	Recurrent episodes of abdominal pain & vomiting. Poor weight gain	Bowel loops confined to left side of abdomen	Multiple congenital bands. D-J junction to the right of midline. Mobile right colon located on left side.
6.	8yrs	M	Recurrent episodes of abdominal pain & vomiting. Poor weight gain	Bowel loops confined to left side of abdomen	Multiple congenital bands. Mobile right colon located on left side. Small bowel loops to right side. D-J junction on the right of midline.
7.	9yrs	F M	Recurrent attacks of abdominal pain & vomiting. Failure to thrive 4 previous admissions	Dilated stomach (Fig 111)	Multiple congenital bands. Large bowel all located on the left side & small bowel on right.
8	14yrs		4 previous admissions with diagnosis of peptic ulcer, typhoid ileitis, and adhesive intestinal obstruction.	Intestinal obstruction	Chronic volvulus, congenital defect in the mesentery, and perforated proximal bowel. (Fig 1V)

Fig 1: Operative finding showing D-J flexure (arrow) to the right of the midline

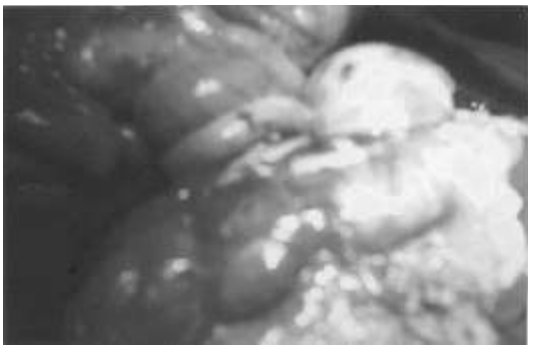

Fig 2: Operative finding showing showing volvulus of the midgut

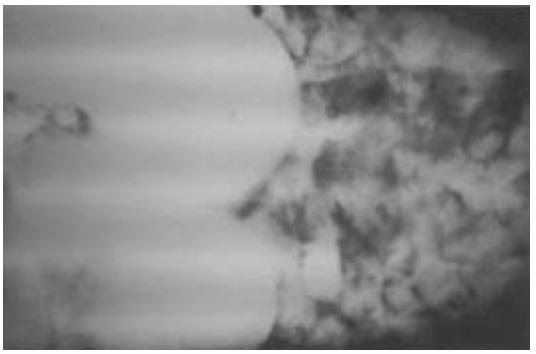

Fig 3: Barium meal and follow through X-ray showing hugely dilated stomach

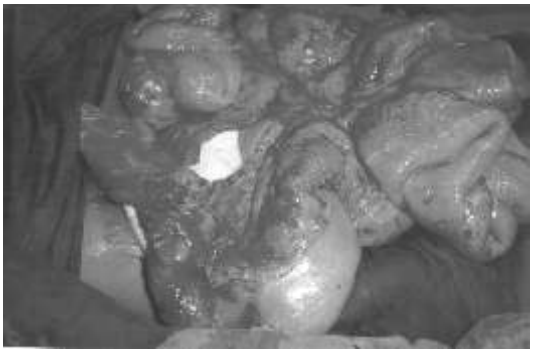

Fig 4: Operative finding showing the defect in the mesentery. The perforated volvulus has been resected and the ileo-colonic anastomosis is shown.

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