

A new technique in the treatment of intestinal malrotation

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ABSTRACT

Introduction. Intestinal malrotation is a congenital pathology with potentially catastrophic complications, such as volvulus, whose treatment has barely not changed in nearly 100 years (Ladd's procedure). Dr. Abu-Elmagd recently described a new technique that was applied in one of our patients.

Clinical case. 12-year-old boy who had undergone Ladd's procedure as a result of intestinal volvulus secondary to malrotation when he was 2 days old. He had subocclusion and eventually obstruction, with intestinal volvulus compatible imaging. Intraoperative findings: duodenal subocclusion, volvulus and lymphangiectasias. Kareem's procedure: bowel positioning in normal rotation, duodenopexy (duodenal C posterior to the mesenteric vessels), formation of neo-Treitz, and fixation of the cecum, the ascending colon, and the mesenteric root. The patient was discharged on postoperative day 6 and remains asymptomatic after 1 year of follow-up.

Discussion. Kareem's procedure is a safe and effective malrotation repair technique. It can replace Ladd's procedure as it reduces the risk of re-volvulation and improves digestive symptoms.

KEY WORDS: Intestinal malrotation; Intestinal obstruction; Intestinal volvulus; Surgical procedures.

UNA NUEVA TÉCNICA EN EL TRATAMIENTO DE LA MALROTACIÓN INTESTINAL

RESUMEN

Introducción. La malrotación intestinal es una patología congénita con complicaciones potencialmente catastróficas, destacando el vólvulo, cuyo tratamiento no ha variado significativamente en casi 100 años (procedimiento de Ladd). Recientemente el Dr Abu-Elmagd describió una técnica que hemos aplicado en un paciente.

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Caso clínico. Varón de 12 años, intervenido con 2 días de vida por vólvulo intestinal secundario a malrotación realizándose procedimiento de Ladd. Presenta cuadros suboclusivos y finalmente obstructivo con imágenes compatibles con vólvulo intestinal. Hallazgos intraoperatorios: suboclusión duodenal, vólvulo y linfangiectasias. Procedimiento de Kareem: posicionando intestino en normorotación, duodenopexia (C duodenal posterior a vasos mesentéricos), formación de neoTreitz, fijación de ciego, colon ascendente y raíz mesentérica. Alta hospitalaria al 6º día postoperatorio, asintomático en 1 año de seguimiento.

Comentarios. El procedimiento de Kareem es una técnica segura y efectiva que corrige la malrotación, pudiendo reemplazar al procedimiento de Ladd al disminuir el riesgo de revolvulación y mejorar síntomas digestivos.

PALABRAS CLAVE: Malrotación intestinal; Obstrucción intestinal; Vólvulo intestinal; Tratamiento quirúrgico.

INTRODUCTION

Intestinal malrotation (IM) is a congenital pathology with potentially catastrophic complications, such as intestinal volvulus, with massive bowel losses that may even require bowel transplantation. IM treatment, which was first described by Dr. Ladd in 1930s, has barely not changed in nearly 100 years. However, in late 2021, a new technique –Kareem's procedure or malrotation repair technique– was described. We report our experience with this technique in one of our patients.

CLINICAL CASE

12-year-old boy who had undergone Ladd's procedure as a result of intestinal volvulus secondary to IM when he was 2 days old. Since he was 5 years old, he had had self-limited subocclusion episodes and constipation. He had been suffering from abdominal pain and intermittent bile vomit for the last month, with a 5 kg weight loss. At

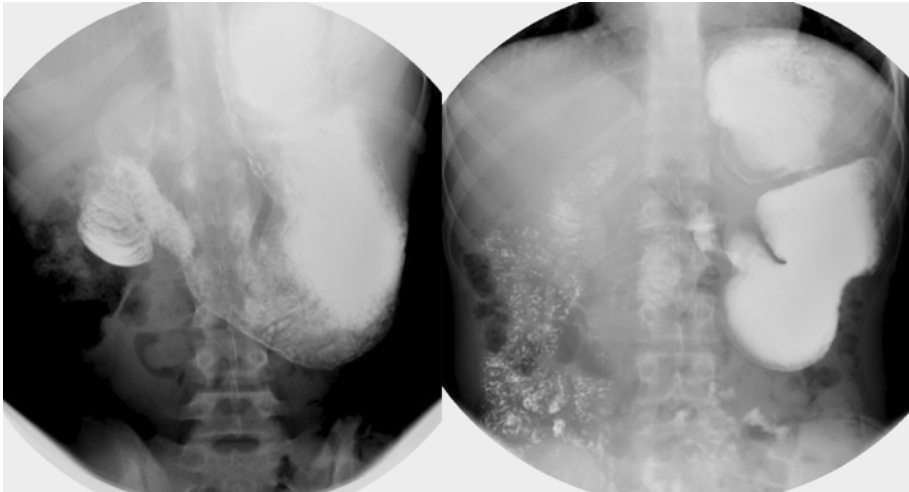


Figure 1. Gastrointestinal transit test revealing gastric dilatation with subocclusion –filiform passage of contrast– in the duodenum.

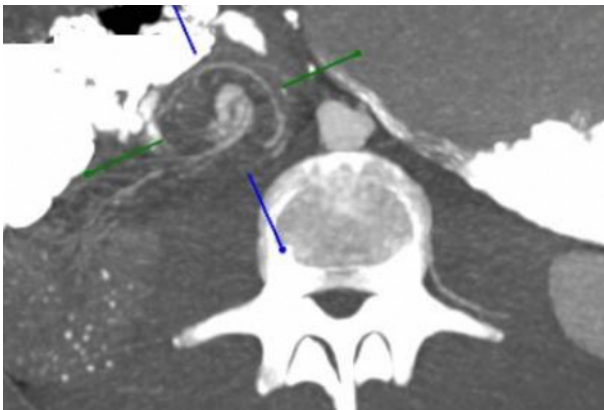


Figure 2. CT-scan axial section demonstrating a “whirl sign,” suggestive of intestinal volvulus.

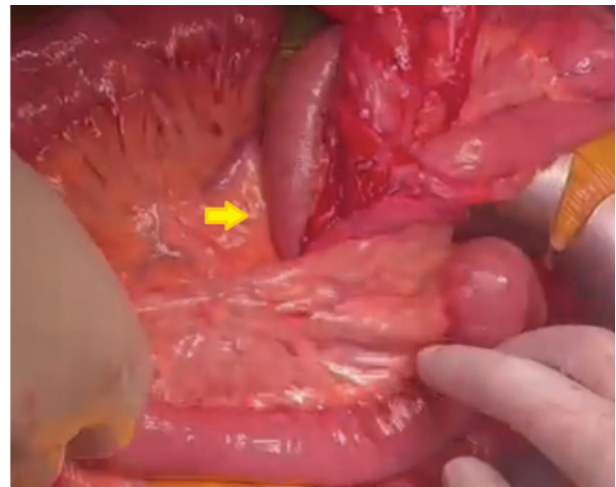


Figure 3. Intraoperative picture of the intestinal volvulus with lymphangiectasias.

exploration, the patient had pain and distension at the upper hemiabdomen. An abdominal X-ray was carried out, with presence of hydroaerial levels, no air within the colon, and no rectal ampulla, compatible with bowel obstruction. Gastrografin® treatment through a nasogastric probe was decided upon. Serial X-rays revealed passage of the contrast to the rectal ampulla. A gastrointestinal transit test was conducted, showing significant gastric and duodenal dilatation, gastroesophageal reflux, and filiform passage of the contrast to the duodenum (Fig. 1). The study was completed with an abdominal CT-scan, which demonstrated a “whirl” sign, compatible with intestinal volvulus (Fig. 2). Therefore, surgical exploration was decided upon.

An iterative middle laparotomy was conducted. It revealed the presence of duodenal subocclusion caused by fibrous bands, along with a partial clockwise volvulus at the duodenojejunal transition and lymphangiectasias (Fig. 3). Following adhesion freeing and de-volvulation,

intestinal malrotation repair surgery –Kareem’s procedure– was carried out. The bowel loops were placed in a normal rotation position, with the duodenum crossing posterior to the mesenteric vessels. A duodenopexy with formation of duodenal C was performed, the ascending colon and the cecum were fixated, and neoformation of Treitz’s angle and the mesenteric root was conducted. The patient had good postoperative progression, with tolerance being initiated on postoperative day 2. He was discharged on postoperative day 6. A gastrointestinal transit test was carried out one month following surgery, with a good passage of contrast throughout the whole bowel, with a visible duodenal C, and the colon in a normal rotation position (Fig. 4). After 12 months of follow-up, the patient reported that symptoms as well as constipation had disappeared, while gaining the weight he had previously lost.

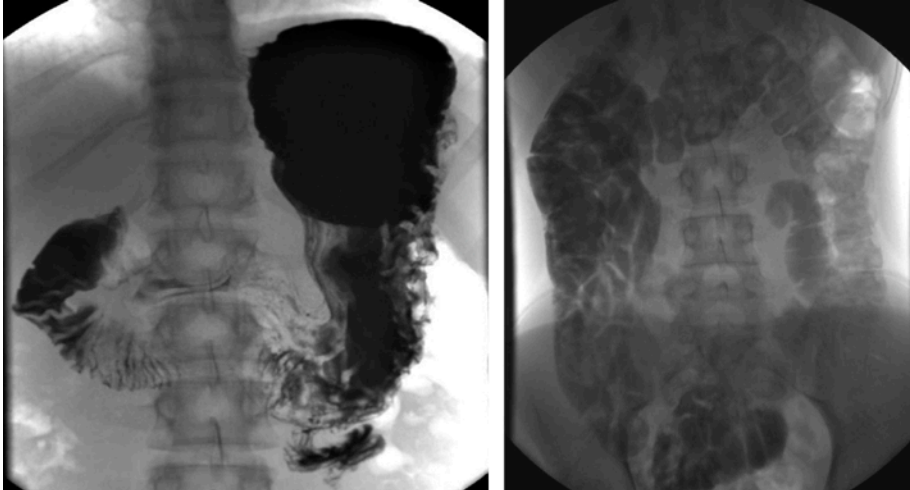


Figure 4. Gastrointestinal transit test after Kareem's procedure showing duodenal malrotation repair, with duodenal C and the cecum in the right iliac fossa.

DISCUSSION

IM is a congenital abnormality which involves an alteration of normal intestinal rotation in the fetal period⁽¹⁾. It occurs in approximately 1 out of 6,000 live newborns, but the absence of symptoms is indicative of a higher incidence in many cases, up to 0.5-1% in the study of autopsies^(1,2). It has traditionally been regarded as a mostly pediatric pathology, with 75% of the cases being diagnosed by the age of 5 years old. However, recent studies have published series where 50% of the patients had been diagnosed in adulthood⁽³⁾.

Normal intestinal rotation represents a 270-degree, anti-clockwise turn. Intestinal rotation disorders primarily consist of lack of rotation and IM/incomplete rotation, with the latter having the highest risk of complications. Even though many patients are asymptomatic, the main clinical manifestations include vomit, abdominal distension, abdominal hypersensitivity/hyperalgesia, hemochezia, peritonitis, and even shock^(1,4). In patients under 1 year of age, it usually starts with vomit, whereas in older patients, it involves more latent clinical signs, such as abdominal pain and intermittent vomit⁽³⁾. In the case reported, the patient had both –sudden bile vomit with abdominal distension in the neonatal period, and intermittent vomit and abdominal pain during adolescence.

IM diagnosis is completed with imaging tests. The main test is the gastrointestinal transit test, with a sensitivity close to 100%^(1,4,6), but ultrasonography is progressively gaining traction. Other tests, such as barium enema, CT-scan or MRI, may prove useful, but they are not routinely used.

The main complications of IM include obstruction –usually duodenal obstruction at Ladd's bands– and volvulus⁽¹⁾. The latter may occur both in the neonatal period and at older ages –as in this case–, and it can be very severe, in some cases with massive small bowel losses

that may require bowel transplantation, and therefore, emergency surgery^(1,4). The risk of intestinal volvulus remains throughout life, with a reported incidence of 22% in pediatric patients, and 12% in adult patients at diagnosis⁽³⁾. In some cases, as in this one, volvulus can occur in a self-limited but recurrent fashion. This is known as chronic intestinal volvulus, with venous and lymphatic obstruction⁽⁴⁾.

The treatment of intestinal malrotation and its complications is surgical. The surgery of choice is Ladd's procedure, which was first described in the 1930s. This technique has undergone minor changes only, which means it virtually remains the same⁽⁷⁾. It involves untwisting the small bowel in the presence of volvulus, separating the adhesions –Ladd's bands– compressing the duodenum, enlarging the mesentery to prevent future volvuli, and placing the bowel in a non-rotation position, with the small bowel on the right side of the abdomen, and the colon on the left one^(1,4,7). Modifications of this technique include laparoscopy, which is limited to selected patients⁽⁸⁾. However, as in the case presented, the risk of volvulus does not disappear, with a re-volvulation rate of 1%-51%^(9,10) –nevertheless, it should be noted that larger series usually belong to reference institutions, which describe more complex cases.

In 2021, Dr. Kareem Abu-Elmagd published a new technique for the treatment of malrotation, known as malrotation repair surgery or Kareem's procedure, which was applied in our case⁽¹⁰⁾. The particularity of this technique lies in the fact that, instead of positioning the bowel in a non-rotation position, malrotation is repaired by placing the bowel in normal rotation⁽¹⁰⁾. These are the main steps of the procedure: 1) the duodenum is freed, along with Ladd's bands, if present; 2) the colon is dissected, with colectomy being required in some cases; 3) the duodenum is rotated 180 degrees anti-clockwise –physiological rotation– until it has been placed in a normal position, that is, posterior to the superior mesenteric artery; 4) the colon

is rotated 180 degrees until the cecum has been placed in the right iliac fossa, which is fixated, along with the ascending colon, to the right parietal peritoneum and the retroperitoneum –this allows the vascular inversion of the superior mesenteric artery and vein to be repaired; 5) a duodenopexy is conducted by fixating the second duodenal portion to the right parietal peritoneum, and a neo-ligament of Treitz is created to fixate the duodenojejunal junction; 6) the mesentery is fixated to the retroperitoneum with a suture, along a diagonal axis between the cecum –which has been previously placed in the right iliac fossa– and the neo-ligament of Treitz⁽¹⁰⁾. The result is a normal rotation bowel position.

This technique was carried out by Dr. Abu-Elmagd in 80 patients, 22 of whom were under 18 years old, and 6 of whom were pediatric patients⁽¹⁰⁾. Mean operating time was 6.5 hours, but in patients without previous surgeries or need for other procedures –mostly colectomy–, mean operating time was 4 hours⁽¹⁰⁾. In the case reported, operating time was 3 hours. Regarding surgery-related postoperative complications, 4 patients had surgical wound infection, 1 patient had intra-abdominal collection requiring percutaneous drainage, 1 patient had intestinal obstruction resolved in a conservative manner, and 11 patients had refractory constipation, with no need for colectomy⁽¹⁰⁾. Our patient had none of these complications.

One of the main advantages of this technique vs. Ladd's procedure is the theoretical absence of volvulus risk when repairing malrotation, which translates into a 0% volvulus rate in 3 years of follow-up⁽¹⁰⁾. A significant improvement of gastrointestinal symptoms –as assessed by the NIH-PROMIS scale– has also been reported, both before and after this surgery, as well as in the 34 patients who initially underwent Ladd's procedure followed by Kareem's procedure⁽¹⁰⁾. As previously stated, our patient has remained asymptomatic following surgery, with normal bowel rhythm.

For the time being, the highly promising data from Dr. Abu-Elmagd cannot be compared, since there are no other series or clinical cases published.

In conclusion, Kareem's procedure is a safe, reproducible technique allowing for definitive repair of intestinal malrotation by normalizing the entero-mesenteric structure and vascular anatomy, with an improvement in gastrointestinal symptoms and no risk of intestinal volvulus when establishing bowel fixations. We believe this technique could replace Ladd's procedure as the technique of choice in the treatment of intestinal malrotation.

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