Medical Xpress

Study describes new procedure that successfully treated patients with congenital intestinal malrotation

September 13 2021

Credit: Pixabay/CC0 Public Domain

A Cleveland Clinic study has introduced a new surgical procedure to treat both children and adults with congenital intestinal malrotation, an inherited disorder that can cause the intestines to twist.

The study, published in the *Annals of Surgery,* also defines the disease presentation in both children and adults, identifies the patients at risk of intestinal loss, and assesses the long-term outcomes after different surgical interventions.

Congenital intestinal malrotation, also known as gut malrotation, can cause the small and large intestines to twist, which potentially can lead to the development of life-threatening complications. The condition, often overlooked or misdiagnosed, can affect people at any age.

Malrotation occurs in about one out of every 500 births in the United States. During pregnancy, a baby's bowel normally rotates 270-degree counterclockwise and then becomes fixed in position. With gut malrotation, however, this embryonic process is defective and sometimes associated with mal-development of the gut, abdominal wall and the nerve supply of the intestine. As a result, babies, children and adults with the disorder can suffer from several gastrointestinal symptoms—from abdominal pain, nausea and bloating to food intolerance and altered bowel habits. Most seriously, twisted intestines—known as volvulus—often result in loss of most of the small bowel and half of the colon with the need for long-term intravenous nutrition. Some patients may ultimately need life-saving intestinal or multivisceral transplantation.

Drawing on 30 years of experience in digestive surgery and transplantation, Kareem Abu-Elmagd, M.D., Ph.D. – director of Cleveland Clinic's Center for Gut Rehabilitation and Transplantation and lead author of the study—developed a new gut malrotation correction surgery to alleviate the gastrointestinal symptoms as well as prevent the need for gut transplantation. "During the procedure, the digestive organs are rearranged and fixed in their proper anatomic locations, which prevents the intestines from twisting and resolves the digestive symptoms with improved quality of life," said Dr. Abu-Elmagd.

The operation that has been used traditionally to address intestinal malrotation is called the Ladd's procedure, which untwists the intestines and alleviates the bowel obstruction by dividing adhesive bands between the duodenum (upper part of the small intestine) and colon.

However, despite the relief of the obstruction with some temporary improvement in patient's symptoms, the Ladd's procedure does not prevent the intestine from recurrent future twisting, which can cut off the blood supply and be life-threatening at any age.

This study is the largest series worldwide with the longest follow-up in the literature.

Of the 500 study patients, 41% were referred after losing their intestine due to volvulus and the remaining 59% had intact gut with a wide range of digestive symptoms. Gut transplantation was required for patients with massive intestinal loss. The new operation ("Kareem's procedure") was performed in patients with intact gut and disabling symptoms.

The study further revealed:

- The patients most at risk of losing their intestines had a history of congenital anomalies, including the gastrointestinal tract and

abdominal wall, and were male—traits that had not been reported previously in medical literature.
- 21% of the patients who had a previous Ladd's procedure developed another volvulus.
- Gut transplantation is effective in saving the lives of the pediatric patients who suffered intestinal loss. The survival of these children reached 63% at 10 years and 54% at 20 years. Infants had the best survival with up to 64% at 20 years. Most survivors had a better quality of life. (Most of the patients who received intestinal and multivisceral transplantation as part of the study were children, including infants.)

"This study shows the favorable outcomes of gut transplantation in saving the lives of babies with intestinal loss," said Dr. Abu-Elmagd. "These encouraging results must be shared with parents to guide their decision-making process along with their clinical care team."

This study also shows that more awareness about intestinal malrotation is needed to proactively treat the disorder and prevent its potentially life-threatening complications.

"Overlooked symptoms or misdiagnosis can result in a delay to receive life-changing care," said Dr. Abu-Elmagd. "Therefore, it is important to diagnose the congenital disorder early in life—possibly with increased efforts to establish perinatal screening programs—so patients can receive an effective surgical treatment in a timely manner to prevent volvulus and save their gut."

In this study, the Kareem's procedure was performed on 80 patients with intact gut and disabling symptoms—92% were adults and 8% were children. Soon after surgery, the digestive symptoms improved significantly. With up to 10 years of follow-up, most of those patients are able to eat normally and have a better quality of life. None of the

patients developed volvulus following the Kareem's procedure.

"The new operation is safe and effective, and can be performed in patients of all ages with intestinal malrotation, especially in the presence of disabling symptoms and those who had volvulus after Ladd's," said Dr. Abu-Elmagd. "It is my hope that this procedure can become part of the surgical training for both pediatric and adult surgeons."

A feasibility study evaluating a laparoscopic approach to safely perform the procedure is currently underway at Cleveland Clinic.

More information: Kareem Abu-Elmagd et al, Five Hundred Patients with Gut Malrotation, *Annals of Surgery* (2021). DOI: 10.1097/SLA.0000000000005072

Provided by Cleveland Clinic

Citation: Study describes new procedure that successfully treated patients with congenital intestinal malrotation (2021, September 13) retrieved 16 April 2024 from https://medicalxpress.com/news/2021-09-procedure-successfully-patients-congenital-intestinal.html

This document is subject to copyright. Apart from any fair dealing for the purpose of private study or research, no part may be reproduced without the written permission. The content is provided for information purposes only.