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RESEARCH REPORT

Gastroenterology

Long-term gastrointestinal outcomes in pediatric intestinal malrotation patients following operative treatment

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1 | INTRODUCTION

Intestinal malrotation (IM) is a rare congenital birth defect that occurs in utero due to failure of intestinal rotation and fixation into the normal anatomic position.[1] Generally, IM is diagnosed once a patient experiences a life-threatening complication (e.g., midgut volvulus) or as part of the workup for gastrointestinal symptoms. For those with symptomatic IM and for some who are asymptomatic, operative treatment, typically the Ladd procedure, is often performed to reduce the likelihood of a future volvulus.[2] Previous studies investigating outcomes after the Ladd procedure typically have focused only on short-term operative outcomes[3–6] with only a few identifying ongoing gastrointestinal symptoms.[4,7–9]

In this study, we used TriNetX, a large multi-institutional aggregated database of electronic medical records (EMRs), to estimate the rates of diagnosed gastrointestinal symptoms several years postoperative treatment for pediatric IM patients compared to a general pediatric cohort.

2 | METHODS

2.1 | Ethics statement

West Virginia University's Institutional Review Board determined that this study does not meet the definition of human subjects research.

2.2 | Study population

Data were extracted on August 1st, 2024 from the TriNetX Research Network, which provides access to

For a complete list of the Intestinal Malrotation Clinician Group, see the Acknowledgments section.

aggregated deidentified EMR for over 130 million patients from 92 healthcare organizations (HCOs) across the world. We conducted a retrospective matched cohort study of postoperative gastrointestinal outcomes in pediatric IM patients across 32 HCOs. The control group for comparison also came from TriNetX data and consisted of a general pediatric cohort from 92 HCOs, including the HCOs in the pediatric IM cohort. TriNetX maps International Classification of Diseases (ICD), Ninth Revision, Clinical Modification (CM) codes (ICD-9-CM) to ICD, Tenth Revision, CM codes (ICD-10-CM) and ICD-9-CM procedure codes (PCS) to ICD-10-PCS codes using General Equivalence Mappings.[10]

TriNetX presents medical outcomes for periods of time after a defined index event, such as a diagnosis or procedure. Table 1 provides the inclusion and exclusion criteria for determining the cohorts. Cohorts were matched using propensity score matching (1:1) based on sex, current age, race, and ethnicity.

To measure gastrointestinal symptoms, ICD-10-CM codes for constipation, abdominal pain, nausea and vomiting, diarrhea, and gastro-esophageal reflux disease (GERD) were queried (see Table S1C for ICD-10 codes).

2.3 | Statistical analyses

We present descriptive statistics of demographic data before and after matching. Cells with fewer than 10 cases but more than 0 cases are automatically rounded up to 10. Mean is displayed for the current age. Percentages are displayed for categorical variables.

Data analysis was performed at two time intervals after the index event to evaluate overall (from Years 1 to 5) and longer-term outcomes (from Years 3 to 5). Follow-up began at 1 year after operation to exclude symptoms that could be related to operative intervention and recovery. The cohorts are the same for both follow-up periods. The former identifies whether the patient had one or more of the gastrointestinal outcomes in the 1–5 years after the index event, whereas the latter captures whether the patient had one or more of the gastrointestinal outcomes 3–5 years after the index event. Rates of the gastrointestinal outcomes for those follow-up periods were compared between cohorts. Risk ratios (RRs) with 95% confidence intervals were calculated.

3 | RESULTS

Three hundred fifty-four pediatric patients with IM met the inclusion criteria; these patients were matched 1:1 to 354 general pediatric patients. Table S2 provides cohort demographic data before and after matching. Before matching, the cohorts differed in current age, race, and sex. After matching, there were no significant differences in socio-demographic variables between the cohorts.

TABLE 1 Overall and cohort-specific inclusion and exclusion criteria.

Overall inclusion criteria	Overall exclusion criteria
1. ≤17 years old at time of data extraction	1. Index event >20 years ago[a]
2. Had at least one medical visit ≥5 years after the index event[b]	2. Had any of the following diagnoses: short bowel syndrome, postoperative malabsorption, omphalocele, gastroschisis, diaphragmatic hernia, situs inversus, levocardia, prune belly syndrome, congenital hernia of the bladder, intestinal atresia or stenosis, and hiatal hernia (see Table S1A for ICD-10 codes)

Cohort-specific inclusion criteria		Cohort-specific exclusion criteria	
Pediatric IM cohort	**Pediatric general cohort**	**Pediatric IM cohort**	**Pediatric general cohort**
3. Had a diagnosis of IM (ICD-10 code Q43.3) within the past 20 years (first instance was used)[c]	3. Had at least one medical visit for any reason[c]		3. Had a diagnosis of IM (ICD-10 code Q43.3)
4. Underwent a Ladd procedure or abdominal operative treatment on the same day or after the first IM diagnosis within the past 20 years (CPT code 44055 or at least one ICD-10 procedure code, see Table S1B), and it was the first instance of such an operative procedure[c]			4. Had a Ladd procedure or abdominal operative procedure (CPT code 44055 or an ICD-10 procedure code, see Table S1B)

Abbreviations: CPT, Current Procedural Terminology; ICD-10, International Classification of Diseases, Tenth Revision; IM, intestinal malrotation.

[a]TriNetX excludes patients in whom the index event occurred over 20 years ago.

[b]To follow patients for 5 years after their index event, we needed to ensure that the cohorts only included patients with medical records spanning that duration. Because of this, we only included patients that had at least one medical visit for any reason ≥5 years after the index event.

[c]Index event.

TABLE 2 Gastrointestinal outcomes for pediatric IM cohort postoperative treatment compared to a pediatric general cohort.

	Years 1–5 post-index event				Years 3–5 post-index event			
	IM cohort *n* (%)	General cohort *n* (%)	Risk ratio (95% confidence interval)	*p*	IM cohort *n* (%)	General cohort *n* (%)	Risk ratio (95% confidence interval)	*p*
Constipation	147 (41.5%)	26 (7.3%)	5.65 (3.83–8.35)	<0.001	104 (29.4%)	14 (4.0%)	7.43 (4.34–12.72)	<0.001
Abdominal pain	96 (27.1%)	24 (6.8%)	4.00 (2.62–6.10)	<0.001	58 (16.4%)	15 (4.2%)	3.87 (2.23–6.69)	<0.001
Nausea and vomiting	145 (41.0%)	41 (11.6%)	3.54 (2.58–4.84)	<0.001	75 (21.2%)	24 (6.8%)	3.12 (2.02–4.83)	<0.001
Diarrhea	80 (22.6%)	17 (4.8%)	4.71 (2.85–7.78)	<0.001	34 (9.6%)	10 (2.8%)	3.40 (1.71–6.77)	<0.001
GERD	123 (34.7%)	10 (2.8%)	12.30 (6.57–23.04)	<0.001	79 (22.3%)	10 (2.8%)	7.98 (4.16–15.00)	<0.001

Abbreviations: GERD, gastro-esophageal reflux disease; IM, intestinal malrotation.

Table 2 compares the cohorts in terms of the gastrointestinal outcomes for different follow-up periods. Compared to the pediatric general cohort, the pediatric IM cohort was at a higher risk of all gastrointestinal outcomes for both follow-up periods. Between 1 and 5 years post-index event, 147 (41.5%) patients in the pediatric IM cohort had a diagnosis of constipation compared to 26 (7.3%) patients in the pediatric general cohort (RR, 95% confidence interval: 5.65, 3.83–8.35), 96 (27.1%) IM patients had a diagnosis of abdominal pain compared to 24 (6.8%) in the general cohort (4.00, 2.62–6.10), and 145 (41.0%) IM patients had a diagnosis of nausea and vomiting compared to 41 (11.6%) patients in the general cohort (3.54, 2.58–4.84). Between 1 and 5 years post-index event, 80 (22.6%) IM patients had a diagnosis of diarrhea compared to 17 (4.8%) patients in the general cohort (4.71, 2.85–7.78) and 123 (34.7%) IM patients had a diagnosis of GERD compared to 10 (2.8%) patients in the general cohort (12.3, 6.57–23.04).

Between 3 and 5 years post-index event, 104 (29.4%) IM patients had a diagnosis of constipation compared to 14 (4.0%) patients in the general cohort (7.43, 4.34–12.72), 58 (16.4%) IM patients had a diagnosis of abdominal pain compared to 15 (4.2%) patients in the general cohort (3.87, 2.23–6.69), and 75 (21.2%) IM patients had a diagnosis of nausea and vomiting compared to 25 (6.8%) patients in the general cohort (3.12, 2.02–4.83). Between 3 and 5 years post-index event, 34 (9.6%) IM patients had a diagnosis of diarrhea compared to 10 (2.8%) patients in the general cohort (3.4, 1.71–6.77) and 79 (22.3%) IM patients had a diagnosis of GERD compared to 10 (2.8%) patients in the general cohort (7.98, 4.16–15.00).

4 | DISCUSSION

Few prior studies have examined gastrointestinal symptoms after operative treatment, although some smaller-scale studies have been conducted.[4,7–9] Two studies found that a large majority of patients were symptom-free after having a Ladd procedure.[2,3] However, the symptoms and follow-up considered in these studies were ambiguous. Recently, one multicenter study and two larger-scale studies identified persistent gastrointestinal symptoms following operative treatment.[1,11,12] However, prior research did not compare pediatric IM patients to a general pediatric cohort to determine if their rates of gastrointestinal symptoms are higher than what one would expect if they did not have IM and operative intervention.

In the current work, we utilized an EMR database and presented the first large-scale study comparing the long-term gastrointestinal outcomes of pediatric patients with IM following operative intervention to a general pediatric population. The findings demonstrate the presence of higher rates of gastrointestinal symptoms post-index event for pediatric IM patients compared to a matched general pediatric cohort. Consistent with other studies that call for long-term follow-up for IM patients after operative treatment, an automatic gastroenterology referral or a coordinated care model may be important in helping to manage and treat these ongoing symptoms in pediatric IM patients.[1,4,9] As such, the current data can inform the management strategy of physicians taking care of pediatric IM patients who have had operative treatment.

This study has several limitations. The data only include diagnosed symptoms, and we cannot identify if they were chronic symptoms. Individuals may have such symptoms and not seek medical care for them, which may be the case for those who have recurring symptoms for extended periods of time. The results may also potentially overrepresent gastrointestinal issues, as only patients who had at least one medical visit at least 5 years after the index event were included. Since the data comes from aggregated medical records, there is no way to assess if a patient has no medical records 5 years later due to having no medical visits or due to having dropped out of the system. It seems unlikely for it to be the former as one

might expect the majority of patients within the health system to have at least one medical visit of any kind ≥5 years after the index event. Gastrointestinal symptoms may also be overrepresented if diagnosed symptoms are carried over into future medical records through duplication despite resolution of those symptoms. Additionally, the general pediatric cohort may reflect a sicker population than a true baseline for pediatric patients, since they are drawn from a sample of those who had at least one medical visit followed by one other medical visit at least 5 years later. Because we examined a long-term follow-up period (5 years), the IM cohort's sample size is not large enough to stratify by age at surgery, volvulus, or new versus persistent GI symptoms postoperatively. Another limitation is that TriNetX's aggregate-level data cannot identify individual-level factors associated with an increased risk of experiencing postoperative gastrointestinal symptoms. To protect patient privacy, TriNetX rounds up to 10 any cell with between 1 and 9 patients. The general cohort gastrointestinal rates with a frequency of 10 may thus overestimate the rates and underestimate the differences between cohorts.

5 | CONCLUSION

Our findings demonstrate that a large group of pediatric IM patients experience gastrointestinal symptoms several years after operative treatment for IM, and that these rates are significantly higher than those of a propensity-matched general pediatric cohort. Given the substantial risk of gastrointestinal symptoms, all pediatric IM patients who undergo operative treatment should have routine gastroenterology follow-up in addition to the already recommended postoperative follow-up.[1,4,9] These results also suggest that IM should be considered a chronic disease.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest.

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

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