Post-discharge follow-up of congenital duodenal obstruction patients: A systematic review

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Abstract

PURPOSE

Long-term follow-up of congenital duodenal obstruction patients often falls on care providers with little experience of this condition. We performed a systematic review of the long-term outcomes of duodenal obstruction and provide a summary of sequelae care providers should anticipate.

METHODS

In 2022, after registering with PROSPERA, Medline (Ovid), EMBASE, PSYCHINFO, CNAHL and SCOPUS databases were searched using the title keyword 'intestinal atresia'. Abstracts were filtered for inclusion if they included the duodenum. Papers of filtered abstracts were included if they reported post-discharge outcomes. Methodological Index for Non-Randomized Studies was used to grade the papers.

RESULTS

Of the 1068 abstracts were screened, 32 papers were reviewed. Eleven studies were included. Thirty additional papers were included after reviewing references, for a total of 41 papers. The average MINORS was 7/16.

CONCLUSIONS

There is good evidence that children with congenital duodenal obstruction do well in terms of survival, growth and general well-being. Associated cardiac, musculoskeletal and renal anomalies should be ruled-out. Care providers should be aware of anastomotic dysfunction, blind loop syndrome, bowel obstruction and reflux. Reflux may be asymptomatic. Laparoscopic repair does not change long-term outcomes, and associated Trisomy 21 worsens neurodevelopmental outcomes.

Introduction

Congenital duodenal obstruction is a complete or partial embryologic occlusion of the duodenum. Congenital duodenal obstruction may be due to intrinsic or extrinsic causes. Intrinsic obstructions result from mucosal webs or atresia, and may be the result of the failure of the fetal intestine to recannulate [1]. Duodenal atresia is the most common of the intestinal atresias, occurring once in 6000 to 10,000 live births [2]. Extrinsic obstructions are the result of extra-intestinal anomalies such as annular pancreas, Ladd's band or preduodenal celiac vessels. Intrinsic and extrinsic obstructions may occur simultaneously. Most patients present shortly after birth, but patients with stenosis may present later in life [1]. Management requires excision of intrinsic obstructions or by-passing the obstruction to establish intestinal continuity.

After repair of duodenal obstruction, patients may be followed by their surgeons, but commonly, long-term care is managed by primary care providers. Because congenital intestinal obstruction is uncommon, few clinicians know what outcomes to screen for during the long-term follow-up. Routine follow-up of patients with surgical congenital anomalies has been endorsed and recommendations for domains to be assessed at follow-up have been made [3]. And, although studies have reported specific outcomes for children after duodenal obstruction repair, a systematic

review of the long-term outcomes has not been published. The purpose of this review was to summarize the literature about the outcomes of duodenal obstruction patients after discharge, and to suggest specific complications and associations for which screening should be considered as supported by the literature.

Methods

We registered our systematic review with PROSPERO (2020 CRD4204018344).

Identification of studies

We performed a literature search of the databases Medline (Ovid), EMBASE, PSYCHINFO, CINAHL and SCOPUS using the keywords 'intestinal atresia', 'apple peel', 'jejunal atresia' and 'intestinal atresia'. Including 'ileal', 'duodenal' and 'colonic' did not change the results. We initially included studies for any site of intestinal atresia then divided the papers by the site of intestinal atresia: duodenal, jejuno-ileal and colonic. This review includes our summary of the duodenal literature. Jejuno-ileal and colonic reviews will be addressed in separate manuscripts. The original search was performed in June 2020 and a second search was performed in June 2022. No limitations were placed on publication date or language. Abstracts were uploaded to Rayyan, an online systematic review website [4]. Within Rayyan, abstracts were independently filtered for possible inclusion by two authors (MI, SLM). The same two authors then agreed on which complete studies to review based on the filtered abstracts. Complete papers were independently assessed for possible inclusion by two authors (MI, SLM). If a conflict arose regarding inclusion or exclusion, papers were assessed by a third reviewer (AS). References of included papers were manually searched to identify papers that met the inclusion criteria but were not identified through the electronic database searches.

Inclusion and exclusion criteria

Included papers needed to have reported post-discharge outcomes for patients who had surgical repair of intrinsic or extrinsic duodenal obstruction. When full papers were not available, the abstracts were included if sufficient data was reported in the abstract [5]. Google Translate® was used to interpret papers in languages other than English [6]. Papers were excluded if results did not include long-term outcomes. Long-term was defined as 'after discharge' for the admission to correct the duodenal obstruction or more than 30 days post-surgery. Papers describing patients with duodenal obstruction with or without associated anomalies, such as tracheoesophageal fistula, were included. When cases were included in subsequent case series by the same authors, the earlier studies were excluded. Reviews, case reports, surveys, protocols and studies with five or fewer duodenal obstruction patients were excluded. Studies were excluded if they included multiple congenital surgical anomalies but the duodenal cohort outcomes could not be isolated.

Included papers were critically appraised to identify limitations that may have affected the results. Methodological Index for Non-Randomized Studies (MINORS) scores were independently assigned by two reviewers (MI, SLM) to quantify the quality of each paper [7].

Extraction of outcomes

Data abstracted from each paper were: description of patients, type of surgery, age of follow-up, outcomes measured and results. Data was independently abstracted from each manuscript by two authors (MI, SLM) who then conferred to reach a consensus about the details abstracted from each paper. After all of the data was

abstracted, the outcomes were classified as relating to: late mortality, gastrointestinal function, anthropometric and musculoskeletal outcomes, neurologic or neurodevelopmental outcomes and quality-of-life or general well-being.

Results

Included and excluded studies

Figure 1 illustrates how papers were selected for inclusion. A total of 1068 abstracts were screened in Rayyan. Thirty-two complete papers that included duodenal obstruction were reviewed. Eleven of these studies were included. An additional 30 papers were included after reviewing the references of the manuscripts selected. A total of 41 papers met the inclusion criteria.

Table 1 lists the studies included in this review. Table 2 shows the breakdown of the MINORS score for each included paper. Two papers were not accessible but the abstracts provided sufficient data for inclusion [8, 9]. Google Translate→ was used to interpret one manuscript from Russian which was included [10].

Table 3 lists the studies excluded and the reasons for exclusion. Eight studies were excluded because they did not contain long-term outcomes [11–18]. Twelve studies did not differentiate duodenal obstruction cases from other pathology [19–29]. Two studies for which the complete manuscripts were unavailable were excluded because the abstracts did not contain enough detail [30, 31]. Five manuscripts were excluded because each included 5 or fewer duodenal patients [32–36]. Grosfeld *et al.*, Della Vecchia *et al.* and three papers by Grosfeld and Rescorla were excluded because the patients in these manuscripts were subsequently included in an expanded case series published by Escobar *et al.* [37–41]. One survey [42], two reviews [43, 44], one case report [45] and one protocol [46] were excluded.

Data extracted

Of the 41 included manuscripts, 33 papers were retrospective cohort studies, and 8 were retrospective case control studies [2, 10, 47–52]. Six papers limited their population to cases without associated anomalies; 35 papers included cases with duodenal obstruction with or without associated anomalies. The total number of patients included was 2702; 2297 patients survived to discharge or for more than 30 days of life. Duodenoduodenostomy was the most commonly performed surgery (approximately 50%) followed by duodenojejunostomy, web excision and gastrojejunostomy, in the that order. Endoscopic dilatation of duodenal stenosis was reported in one study of six cases [53]. Less than 10% of surgeries were performed laparoscopically, but laparoscopic surgery was increasingly common in the more recent papers. Trisomy 21 was reported in 650 cases but five papers did not report this association; after considering only papers that included cases with and without Trisomy 21 the prevalence of Trisomy was 24%. The longest follow-up was 35 years [54].

The average MINOR score was 7/16 (IQR 5, 9). MINOR score breakdown for included manuscripts is shown in Table 2.

Late mortality

Sixty-four (2.79%) late mortalities were described among the 2297 children who survived to discharge, as shown in Table 4. The most common causes of late mortality were congenital heart disease and complications of cardiac surgery. Other common causes of death were associated congenital anomalies and respiratory infections. The oldest reported death occurred at 14 years of age [39].

Gastrointestinal outcomes

Common gastrointestinal complications that occurred after discharge were: anastomotic dysfunction, blind loop syndrome, adhesive bowel obstruction, reflux and ulcers. Anastomotic leak and wound dehiscence were unlikely to have evaded detection for the long-term and are not summarized herein [39, 55–57]. Eight occurrences of biliary atresia and three choledochal cysts were reported, but the long-term outcomes of these uncommon cases is reported elsewhere [39, 55, 58–60].

The terms 'anastomotic dysfunction', 'anastomotic stricture', 'blind pouch', 'duodenal diverticuli' and 'megaduodenum' were assumed to refer to the same morphology and were grouped together. Although the proximal duodenum can be dilated prior to surgical correction of the congenital obstruction, 'megaduodenum' was considered an 'anastomotic dysfunction' when it persisted after surgery. In this review 'blind loop syndrome' was limited to symptoms resulting from dysfunction of a gastrojejunostomy or duodenojejunostomy, as shown in Figs. 2 and 3. Occurrences of 'blind loop syndrome' in the absence of an anatomic loop without ingress were censored. For example, Kimura *et al*. demonstrated the utility of the diamond-shaped duodenoduodenostomy anastomosis; they noted the absence of blind loops on follow-up contrast studies but none should have been expected [61]. We used 'adhesive bowel obstruction' to refer to obstructions distal to the duodenal anastomosis or bypass, and we addressed them as outcomes separate from anastomotic dysfunction.

Twenty-three manuscripts addressed anastomotic anomalies or anastomotic dysfunction [39, 47, 53–56, 59, 61–68]. Four of these studies used screening contrast studies to detect anastomotic anomalies [53, 54, 61, 64]. A total of 82 contrast studies were done from 6 months to 35 years of age. Findings consistent with anastomotic dysfunction were reported in 35 patients. Megaduodenum, duodenal diverticuli or pouches and strictures were the most common irregularities; windsocks and bezoars were also noted [54, 64]. Despite the radiographic aberrations, only two patients in these four studies required surgical revision [64]. Symptomatic anastomotic dysfunction was described in 51 (4.70%) patients in twenty papers that clearly looked for anastomotic dysfunction in a total of 1084 cases. The presenting complaints from the patients with anastomotic dysfunction included: early satiety, vomiting, bilious emesis, abdominal pain with eating and halitosis [35, 69]. The oldest case presented at 18 years of age [39]. Balloon dilation, with or without cautery ablation, was used to manage anastomotic strictures, but more commonly, surgical revision was required.

Early satiety, emesis, halitosis or an abdominal mass may be due to blind loop syndrome rather than anastomotic dysfunction [35, 69]. In our review, only two cases were consistent with blind loop syndrome [39, 64]. A 16-year-old patient in Escobar *et al.*'s study required conversion from a duodenojejunostomy to a duodenoduodenostomy; the presenting symptoms were not described [39]. The second case occurred in a child initially treated with an ileo-transverse colostomy, an unusual procedure likely employed because of an associated intestinal atresia [64]. Spigland and Yazbeck performed seven duodenojejunostomies and five required reoperations for 'blind loop syndrome' [59]. However, careful review of the operative findings (a missed membrane, bowel necrosis, adhesive bowel obstruction, volvulus and H-type tracheoesophageal fistula) suggested that none of the revisions were for blind loop syndrome. Zani *et al.* found no blind loop syndrome in their 45 duodenojejunostomy cases, the largest series reported to date [67]. Admittedly, their mean follow-up was only 6.5 months.

Adhesive bowel obstructions in 60 (3.87%) of 1549 cases from 16 studies that clearly looked for obstructions [10, 39, 49, 54–57, 59, 60, 63, 66–68, 70–72]. The oldest child was 10 years of age at presentation [60]. Surgical

intervention was required in 37 of the described cases. Oral Gastrografin→ was effectively used to resolve an obstruction in one patient [49].

Malrotation was identified in 492 (35.94%) cases in 17 papers that reported this associated anomaly in a total of 1369 survivors [10, 39, 49–52, 56, 57, 59, 64–66, 70, 72–75]. Only one case of volvulus was reported [59]. Most, but not all, cases of malrotation identified at surgery underwent a Ladd's procedure [51].

Reflux was inconsistently described in the reviewed papers. Some researchers reported gastroesophageal reflux only in patients that required fundoplication [39, 55, 70]. Other researchers reported surgically and medically managed patients [59, 72]. Nineteen cases of gastroesophageal reflux were managed with surgery, and an additional 36 were managed medically. Therefore, 6.29% of the 715 survivors in papers that reported this outcome were believed to have gastroesophageal reflux. Fragoso *et al.* included only patients with duodenal atresia and esophageal atresia [76]. Sixty percent of Fragoso *et al.*'s 10 cases had gastroesophageal reflux, and 4 required surgery [76].

Duodenogastric and/or biliary reflux was observed on upper gastrointestinal study and isotope biliography in 12 of 28 asymptomatic patients [54]. In the same series, endoscopic biopsies from 20 cases located found reflux gastritis in 7 and duodenal irritation and retention in 3 [54]. Four cases of gastric ulcers were reported [39, 51].

Ventral wall hernias occurred in 13 (2.62%) of 497 open repairs [10, 39, 47, 51, 58, 66, 73, 74]. No trocar site hernias were reported.

Eleven cases of associated congenital intestinal obstruction that eluded detection at the initial surgery were described [39, 47, 50, 55–57, 59, 74]. Most cases presented shortly after the duodenal surgery, but two children presented 17 and 18 years after duodenojejunostomy with missed membranes [39, 59]. One case presented with bleeding from a peptic ulcer in the retained membrane, and the other presented with symptoms consistent with anastomotic dysfunction leading to megaduodenum.

Other uncommon gastrointestinal long-term complications described in the literature included: abdominal pain, scar pain, constipation and diarrhea [54, 57, 60, 70].

Anthropometric and musculoskeletal

Growth, reported for a total of 45 patients in four papers, was normal in all but five children followed from 3 months to 15 years [49, 53, 61, 77]. Only Parmentier *et al.* reported weights below the 10th percentile in five cases at a median of 150 days [49]. Atwell and Klidjian observed vertebral anomalies in 37% of their duodenal atresia cases [78]. None of the observed anomalies were clinically relevant. Two children from the same series had other musculoskeletal anomalies: one had bilateral absence of their thumbs and the other had bilateral talipes equinovarus [78]. Kullendorf reported one child with syndactyly and another with talipes equinovarus [74]. Singh *et al.* reported occurrences of sacral deficiency, vertebral anomalies, talipes, dislocated knees, absent digits and fused ribs [50].

Neurologic outcomes

Specific neurologic deficits were reported in two studies. At 4.5 years of age, 11 of 86 children with Trisomy 21, and 4 of 141 without Trisomy 21 had hearing loss [72]. Hearing loss was reported in one child from another study [54]. Niramis *et al.* also reported 'lower limb paralysis' in five children without Trisomy 21; details regarding the cause and sequelae were not provided [72].

Neurodevelopmental outcomes

Two studies assessed neurodevelopment in duodenal atresia survivors. Batta *et al.* reviewed the Griffiths Mental Development Scales II assessment of 15 children at 1 year of age; the median score was normal [79]. Children born earlier than 34 weeks' gestation and children with chromosomal anomalies were excluded [79]. Niramis *et al.* reported severe neurodevelopmental delay in 24 (27.9%) of 86 duodenal atresia children with Trisomy 21, but only 5 (3.55%) of 141 cases without Trisomy 21 [72]. The means by which Niramis *et al.* defined neurodevelopment delay was not clear.

Quality of life/general well-being

The validated Pediatric Quality of Life Inventory™ was used to compare patients with duodenal atresia to population norms at a median of 6.7 years [80]. Children with isolated duodenal atresia had quality of life scores similar to age-matched norms, but children with duodenal atresia and Trisomy 21 had significantly lower social scores compared to norms [80]. Type of duodenal atresia did not affect the quality of life score [80]. Six other studies commented on general well-being, reporting the survivors as 'perfectly well' or 'without significant sequalae' [8, 9, 60, 68, 73, 81, 82].

Discussion

Our objective was to review the literature on the long-term, post-discharge outcomes of children after congenital duodenal obstruction repair. To this end, we determined that there are many studies to inform evidence-based, long-term follow-up, but most of the evidence comes from retrospective case series. We have, herein, compiled the findings from these studies to assist care providers as they follow these children to maturity.

The available evidence suggested that after discharge, children with repaired duodenal obstruction do well. Late mortality, neurologic, neurodevelopmental, growth deficits are rare. Quality of life can be expected to be normal. However, gastrointestinal complications may persist into or present in adulthood with symptoms that may be over-looked by clinicians not forewarned of their risk.

Late mortality after duodenal atresia repair is more common than other types of intestinal atresia, and is due to the higher rate of association with other congenital anomalies or syndromes [1]. The available literature suggested that late mortality occurred in approximately 2.79% of duodenal atresia survivors and was most often due to cardiac anomalies.

Long-term gastrointestinal function was good. Patients' self-reported perception of their gastrointestinal quality of life, quantified by Vinycomb *et al.* using the gastrointestinal module of the Pediatric Quality of Life Inventory™, found no difference between duodenal atresia patients and controls [80].

Anastomotic dysfunction presented as early satiety, pain with eating, vomiting, failure to thrive, halitosis and an abdominal mass. Stricture at the anastomosis may precipitate intestinal dysmotility, megaduodenum or duodenal diverticulum. A contrast study may be helpful in the presence of symptoms. However, routine screening with contrast studies in the absence of symptoms may not be worthwhile. Huang *et al.* and Kimura *et al.* found no anastomotic dysfunction on contrast studies performed in asymptomatic patients, and Kokkonen *et al.* found no correlation between symptoms and radiographic anomalies [53, 54, 61]. Despite failing to correlate investigations

to symptoms, Kokkonen *et al.* endorsed both contrast studies and endoscopy in asymptomatic patients. Endoscopy may be helpful, however, to simultaneously rule-out asymptomatic reflux.

Blind loop syndrome was a rare complication in the papers included in this review. This may reflect our limiting 'blind loop syndrome' to symptoms resulting from dysfunction of gastrojejunostomy and duodenojejunostomy. Gastrojejunostomy and, more commonly, duodenojejunostomy were routinely used to bypass duodenal obstructions in the remote past. These reconstructions have largely been replaced by the duodenoduodenostomy which eliminates the blind loop. Symptoms consistent with blind loop syndrome included: early satiety, pain with eating, vomiting old and/or undigested food and/or bile, failure to thrive, halitosis and abdominal mass [35, 69]. Although it is uncommon, care providers should consider blind loop syndrome in patients presenting with these symptoms and a history of gastrojejunostomy or duodenojejunostomy.

Adhesive bowel obstructions occurred in 3.87% of survivors. This rate is similar to 4.6%, the incidence of adhesive small bowel obstructions after any laparotomy [83]. No unique symptoms were described in duodenal obstruction cases who presented with adhesive bowel obstruction. Therefore, care providers can expect the typical complaints of acute abdominal pain, vomiting, abdominal distention and obstipation in duodenal atresia cases with an adhesive bowel obstruction.

Despite the common association of malrotation with duodenal atresia, 36% in this review, volvulus was rare after long-term follow-up. This may be due to the routine performance of a Ladd's procedures at correction of the duodenal obstruction. Note, however, that confirmation and correction of malrotation were not always performed. Therefore, malrotation should be documented if not corrected to prevent intestinal loss.

The incidence of gastroesophageal and duodenogastric reflux is difficult to estimate. The evidence suggests that gastroesophageal reflux is uncommon, but duodenogastric reflex is more common and may be asymptomatic. Many asymptomatic patients had histologic evidence of reflux. Therefore, endoscopy in asymptomatic patients may prove helpful to rule-out complications of reflux. An endorsement for routine screening will require more research.

Associated intestinal atresia occurs in approximately 7% of duodenal atresia [84]. These would hopefully have been identified at the time of duodenal atresia repair but they have been missed and are more likely to be missed with laparoscopic duodenal atresia repair [84].

Altered neurologic or developmental outcomes are unlikely to be due to duodenal atresia directly, but may be an uncommon association or result from the treatment of duodenal atresia. Hearing loss occurred in 15 of 206 (7.28%) survivors for whom hearing was assessed [72]. The type and degree of loss was not described, but it occurred in survivors with and without a history of congenital heart disease and/or Trisomy 21 [72]. The unexpected incidence of lower limb paralysis described in one study requires further investigation [72].

Neurodevelopmental delay in patients with both duodenal atresia and Trisomy 21 were common and not unexpected. In the absence of Trisomy 21, neurodevelopment was normal. Hamrick *et al.* published the only paper to describe neurodevelopment in children with intestinal atresia using an objective measurement: use of special education services [23]. Hamrick *et al.* observed a modest, but not significant, increase in the use of special education services in children with isolated intestinal atresia compared to the general population. When bowel atresia occurred in association with other congenital anomalies, educational support was significantly more common, occurring in 25.9% of cases versus 8% of controls. Batta *et al.* showed that low birth weight and extended

length of hospital admission significantly decreased neurodevelopment at 1 year of age. Lund *et al.* found the strongest indicator of poor intellectual development in children who had neonatal surgery at 6 months was the length of perinatal admission, and at the age of 3 years was the number of procedures under general anesthesia [85, 86]. After an uncomplicated perinatal repair of isolated duodenal atresia, a child can expect to suffer no neurodevelopmental sequelae.

Associated musculoskeletal anomalies have not been well studied in duodenal atresia. Only three manuscripts that met our inclusion criteria included musculoskeletal assessments. Vertebral anomalies, sacral deficiency, talipes equinovarus, syndactyly, absent digits, dislocated knees and fused ribs were reported in association with duodenal atresia. Schierz *et al.* was not included in this review because they did not distinguish duodenal atresia from other atresias. They recommended routine pelvic radiographs to rule out anomalies of the coccyx, ilium, ischium, pubis and vertebrae after finding 34.8% of children with congenital malformations of the digestive system had congenital pelvic skeletal anomalies [27].

Renal anomalies reported in association with duodenal atresia include: vesicoureteral reflux, hydronephrosis, hypotonic bladder, renal dysplasia or agenesis, vesicoureteral junction obstruction, multicystic kidneys, ambiguous genitalia and hypospadias [71]. Screening for these anomalies should be completed in the perinatal period or during follow-up and documented.

Laparoscopic repair of duodenal obstruction was first reported in 2001, and reports comparing long-term outcomes have started to appear in the literature [2, 10, 47, 49, 51, 52]. These studies showed that perioperative and mid-term outcomes are similar for open and laparoscopic repair. The results from recent reports of exclusively laparoscopic case series suggest that laparoscopic repair will replace open repair [75, 82]. For care providers following children after laparoscopic repair, however, the same long-term complications should be considered.

Approximately one quarter of duodenal atresia patients have Trisomy 21. The effect of Trisomy 21 on specific long-term outcomes has yet to be determined. Niramis *et al.* demonstrated more long-term complications in Trisomy 21 patients with duodenal atresia but the specific complications contributing to the disparity were not reported [72]. Singh *et al.* found no difference in gastrointestinal outcomes between Trisomy 21 cases and cases without Trisomy 21 [50]. But, Stauffer and Irving described increased re-operation in Trisomy 21 cases [60]. Because children with Trisomy 21 constitute a large portion of the duodenal atresia population, further investigation into combined effect of these anomalies is required.

We acknowledge several limitations of this review. One, most of the evidence came from small, retrospective case series studies without controls or standardized follow-up protocols. Two, we were forced to exclude many excellent studies that did not clearly distinguish duodenal atresia from other intestinal atresia and case series that included too few duodenal obstruction cases. However, the knowledge imparted from these excluded studies informed our discussion. Third, terms used to define outcomes varied from researcher to researcher. For example, one researcher may have described megaduodenum as an outcome while another researcher may have reported anastomotic dysfunction. To mitigate this variability, we reported outcomes in groups that included similar pathophysiology and manifestations. Our fourth limitation pertained to the original electronic search method. We did not include ‘obstruction’ in the search. This may explain why so many manuscripts were found only after reviewing the included papers. Finally, not all outcomes were reported by all researchers. As a consequence, our estimations for the incidence of each outcome are limited by our assumption that if a manuscript did not report an outcome, then it did not occur; it is more likely that researchers did not look for certain outcomes.

Conclusion

There is good evidence in the literature to suggest that children with repaired congenital duodenal obstruction do well in terms of survival, growth and general well-being. If not previously screened, cardiac, musculoskeletal and renal anomalies should be ruled-out. Care providers should be aware of the potential for long-term gastrointestinal complications, even in the absence of symptoms. Routine assessment should include inquiry regarding abdominal pain, halitosis, vomiting and early satiety. These symptoms may herald anastomotic dysfunction or blind loop syndrome. Acute abdominal pain with distension should alert one to bowel obstruction. Reflux may be asymptomatic. Further investigation into the frequency and sequelae of duodenogastric and gastroesophageal reflux is warranted. Until then, a low threshold for investigation would be wise. One might expect that after laparoscopic repair children will experience outcomes similar, if not better, than after laparotomy. The effect of Trisomy 21 on neurodevelopmental outcomes is clear but its effect on other, particularly gastrointestinal, long-term outcomes requires further study.

Declarations

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Conflict of interest statement

On behalf of all authors, I hereby declare that we have no competing interests that might be perceived to influence the results and/or discussion reported in this paper.

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Tables

Table 1

	Citation	Cases	Males	Cases with T21	Cases survived to discharge or for > 30 days	Included cases with associated anomalies?	Intrinsic, extrinsic or both	Duration or age of follow-up
1	Atwell & Klidjian 1982	35	NA	13	32	Yes	Both	Not reported
2	Bailey *et al.* 1993	138	65	15	128	Yes	Both	Not reported
3	Bairdain *et al.* 2014	87	32	33	84	Yes	Intrinsic	20 months (5 to 48 months)
4	Batta *et al.* 2020	19	NA	0	19	Yes	Intrinsic	1 year of age
5	Burjonrappa *et al.* 2021	59	29	18	59	Yes	Intrinsic	12.5 months (1 to 8 years)
6	Chen *et al.* 2014	287	193	9	270	No	Both	6 months to 5 years
7	Eek, S. 1955	29	21	4	20	Yes	Both	4 months to 23 years
8	Escobar *et al.* 2004	169	80	46	164	Yes	Intrinsic	Not reported
9	Feggetter, S. 1969	20	10	0	22	No	Intrinsic	6 to 28 years of age
10	Fragoso *et al.* 2015	20	NA	2	10	Yes	Intrinsic	Median 9 years
11	Gfroerer *et al.* 2018	47	22	14	46	Yes	Both	44.9 months
12	Helbig & Sallandt 1980*	80	NA	NA	101	Yes	Intrinsic	Not reported
13	Hill et al. 2011	58	30	26	58	Yes	Both	Not reported
14	Huang *et al.* 2015	6	2	1	6	No	Intrinsic	3 to 24 months
15	Jensen *et al.* 2013	64	NA	NA	64	No	Both	Not reported
16	Jerry *et al.* 2022	10	9	5	10	Yes	Intrinsic	4 years (0.9 to 6.5 years)
17	Kimura *et al.* 1990	44	21	NA	39	Yes	Intrinsic	6 months to 15 years
18	Kokkonen *et al.*1988	180	NA	5	107	No	Intrinsic	15 to 35 years of age
19	Kozlov *et al.*	211	NA	53	205	Yes	Both	Assessed at

	2021							1, 3, 6 & 12 months
20	Kraeger *et al.* 1973	19	10	3	12	Yes	Intrinsic	Not reported
21	Kullendorff, C.M. 1983	21	10	11	20	Yes	Intrinsic	Not reported
22	Li et al. 2015	11	7	3	11	Yes	Extrinsic	Mean 15.2 months (4 to 39 months)
23	Louw J.H. 1952	31	NA	9	6	Yes	Both	Not reported
24	Maassel *et al.* 2021	209	108	209	209	Yes	Intrinsic	1 year of age
25	Miller R.C. 1979	16	NA	2	11	Yes	Intrinsic	Not reported
26	Mooney *et al.* 1987	20	6	2	19	Yes	Intrinsic	Not reported
27	Niramis *et al.* 2010	227	62	86	206	Yes	Intrinsic	1 to 7 years
28	Oh *et al.* 2017	22	11	6	22	Yes	Intrinsic	Median 3.5 months (1 to 21 months)
29	Parmentier *et al.* 2015	29	NA	0	29	Yes	Intrinsic	149.5 days (28 to 388 days)
30	Piper *et al.* 2008	63	29	22	58	Yes	Intrinsic	2 years of life
31	Rousková *et al.* 2008*	77	30	NA	70	Yes	Both	5 months to 16 years
32	Salonen & Makinen 1976	27	NA	NA	27	No	Both	Mean 10 years 2 months (3 to 21 years)
33	Samuel *et al.* 1997	36	NA	6	36	Yes	Intrinsic	2 years (4 months to 14 years)
34	Singh *et al.* 2004	79	46	28	76	Yes	Intrinsic	Not reported
35	Smith & Landman 2019	43	22	19	43	Yes	Intrinsic	Not reported
36	Spigland & Yazbeck 1990	33	13	7	31	Yes	Both	Mean 2 years (1 month to 8 years)

37	Spilde et al. 2008	29	13	12	29	Yes	Intrinsic	Not reported
38	Stauffer & Irving 1977	85	NA	32	53	Yes	Intrinsic	10 to 23 years
39	van der Zee D.C. 2011	28	17	11	28	Yes	Intrinsic	6 months to 2.5 years
40	Vinycomb *et al.* 2020	110	54	21	98	Yes	Intrinsic	Median 6.7 years age (2.7 to 17.3 years)
41	Zani *et al.* 2017	92	61	30	90	No	Intrinsic	Mean 6.5 months (2 to 26 months)

T21 Trisomy 21 *****Abstract only available **NA** Not available

Table 2

	Clear aim	Consecutive	Prospective	Endpoint	Unbiased	Follow-up	Loss <5%	Prospective
Bailey *et al.* 1993	2	2	0	1	0	2	0	0
Bairdain *et al.* 2014	2	0	0	2	2	2	0	0
Batta *et al.* 2020	2	2	2	2	1	2	0	0
Burjonrappa *et al.* 2021	2	0	0	1	1	0	0	0
Chen *et al.* 2014	2	0	0	2	1	2	0	0
Eek, S. 1955	0	0	0	0	0	2	0	0
Escobar *et al.* 2004	1	0	0	1	1	2	0	0
Feggetter, S. 1969	2	2	1	2	0	2	0	0
Fragoso *et al.* 2015	2	1	0	2	2	2	0	0
Gfroerer *et al.* 2018	2	2	0	2	2	1	0	0
Helbig & Sallandt 1980*	0	0	0	0	0	2	0	0
Hill et al. 2011	2	2	0	1	1	1	0	0
Huang *et al.* 2015	2	0	0	2	1	2	0	0
Jensen *et al.* 2013	1	2	0	0	2	1	0	0
Jerry *et al.* 2022	2	0	0	1	0	2	0	0
Kimura *et al.* 1990	2	2	1	1	1	2	0	0
Kokkonen *et al.*1988	2	0	2	2	0	2	0	0
Kozlov *et al.* 2021	2	0	0	2	2	1	0	0
Kraeger *et al.* 1973	0	0	0	0	1	0	0	0
Kullendorff, C.M. 1983	1	0	0	0	1	0	0	0
Li et al.	2	0	0	2	2	2	0	0

2015								
Louw J.H. 1952	2	0	0	1	1	0	0	0
Maassel *et al.* 2021	2	2	0	2	2	2	2	0
Miller R.C. 1979	2	2	0	0	1	0	0	0
Mooney *et al.* 1987	2	0	0	1	0	0	0	0
Niramis *et al.* 2010	2	1	1	1	1	1	1	0
Oh *et al.* 2017	2	0	0	1	0	0	0	0
Parmentier *et al.* 2015	2	2	0	2	2	2	2	0
Piper *et al.* 2008	2	2	0	2	2	1	0	0
Rouskovà *et al.* 2008*	1	0	0	1	1	2	0	0
Salonen & Makinen 1976	2	0	1	0	0	2	2	0
Samuel *et al.* 1997	2	0	0	0	0	2	2	0
Singh *et al.* 2004	2	2	0	2	2	0	0	0
Smith & Landman 2019	2	0	0	2	2	2	0	0
Spigland & Yazbeck 1990	2	0	0	2	2	2	0	0
Spilde et al. 2008	2	0	0	2	2	2	0	0
Stauffer & Irving 1977	2	0	0	2	1	2	0	0
van der Zee D.C. 2011	2	2	0	2	2	2	2	0
Vinycomb *et al.* 2020	2	2	2	2	0	2	0	0
Zani *et al.* 2017	2	2	0	2	2	0	0	0

*only abstract available

Table 3

	Citation	Reason for exclusion from systematic review
1	Adzick *et al.* 1986	No long-term outcomes
2	Affourtit *et al.* 1989	Did not separate types of intestinal atresia
3	Amoury *et al.* 1977	Less than 5 cases of duodenal atresia
4	Bax *et al.* 2001	Case report
5	Bethell *et al.* 2020	No long-term outcomes
6	Cavusoglu *et al.* 2012	Did not separate types of intestinal atresia
7	Charlorin *et al.* 2021	No long-term outcomes
8	Chiesa *et al.* 2000	Abstract only; insufficient detail
9	Chung *et al.* 2017	Review
10	Crowle *et al.* 2018	Did not separate types of intestinal atresia
11	Della Vecchia *et al.* 1998	Cases reported in subsequent case series (Escobar *et al.* 2004)
12	Dickson 1970	Less than 5 cases of duodenal atresia
13	Eeftinck Schattenkerk *et al.* 2021	Did not separate types of intestinal atresia
14	Ein & Shandlin 1986	Less than 5 cases of duodenal atresia
15	Ein *et al.* 2000	Less than 5 cases of duodenal atresia
16	Fonkalsrud *et al.* 1969	Survey
17	Ghafouri-Taleghani *et al.* 2015	Did not separate types of intestinal atresia
18	Grosfeld *et al.* 1979	Cases reported in subsequent case series (Escobar *et al.* 2004)
19	Grosfeld & Rescorla 1993	Cases reported in subsequent case series (Escobar *et al.* 2004)
20	Hamrick *et al.* 2010	Did not separate types of intestinal atresia
21	Li *et al.* 2013	Did not separate types of intestinal atresia
22	Li *et al.* 2014	Did not separate types of intestinal atresia
23	Mazer *et al.* 2010	Did not separate types of intestinal atresia
24	Murshed *et al.* 1999	No long-term outcomes
25	Nixon & Tawes 1971	No long-term outcomes
26	Rescorla & Grosfeld 1985	Cases reported in subsequent case series (Escobar *et al.* 2004)
27	Rescorla & Grosfeld 1988	Cases reported in subsequent case series (Escobar *et al.* 2004)
28	Roorda *et al.* 2021	Review
29	Sarin *et al.* 2012	No long-term outcomes
30	Schierz *et al.* 2020	Did not separate types of intestinal atresia
31	Solanki *et al.* 2022	Did not differentiate pyloric webs from duodenal webs

32	Strobel *et al.* 2020	Did not separate types of intestinal atresia
33	Tchirkow et al. 1980	Less than 5 cases of duodenal atresia
34	Toyama *et al.* 2021	No long-term outcomes
35	Wang & Chen 2011	No long-term outcomes
36	Wright *et al.* 2019	Protocol
37	Zhang *et al.* 2018	Abstract only; insufficient detail

Table 4

Author	Number of deaths	Age of deaths	Causes of death
Bairdain *et al.* 1993	1	1 year of age	Cardiac surgery complication
Eek, S. 1955	1	1 year of age	Not reported
Escobar *et al.* 2004	10	3 months to 14 years after surgery	5 congenital heart disease 1 neurosurgery complication 1 TEF complication 1 hepatoportoentrostomy complication 1 respiratory failure after small bowel resection 1 anastomotic leak
Feggetter, S. 1969	2	10 weeks of age 4 1/2 years of age	Hepatitis Splenectomy complication
Hill *et al.* 2011	1	5 months	Sepsis
Kimura *et al.* 1990	9	Not reported	All due to associated anomalies
Kozlov *et al.* 2021	4	Not reported	Not reported
Kraeger *et al.* 1973	1	1 month after discharge	Cardiac surgery complication
Niramis *et al.* 2010	22	Not reported	Congenital heart disease Recurrent pneumonia
Parmentier *et al.* 2015	1	Not reported	Bacterial translocation
Stauffer & Irving 1977	12	6 weeks to 14 months of age	Gastroenteritis Gangrenous volvulus (adhesions) Congenital heart disease Aspiration/pneumonia Sepsis Bronchiolitis
Total	64		

Figures

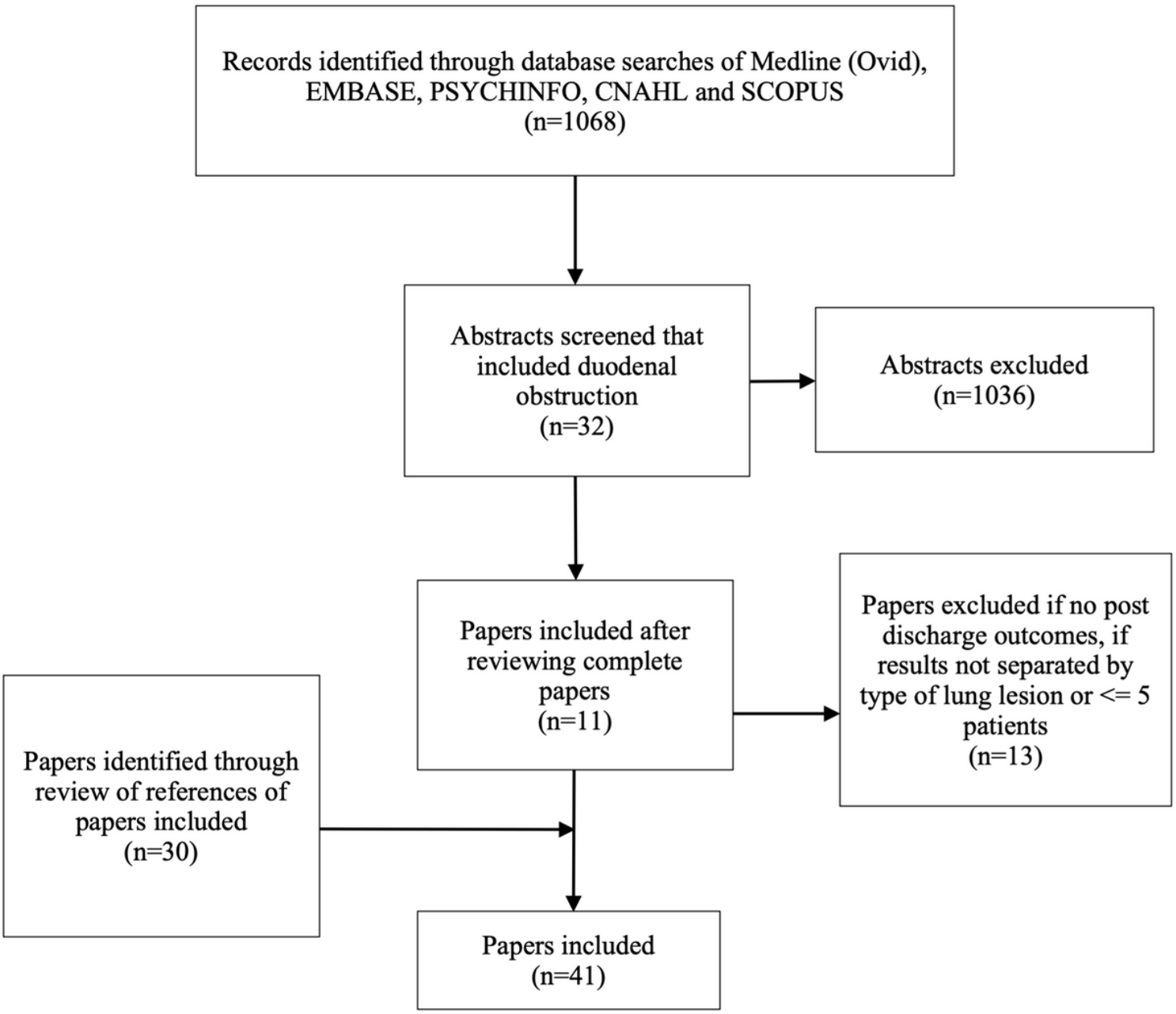

Figure 1

Flow diagram of inclusion and exclusion of papers retrieved through database search of ‘intestinal atresia’ and subsequent review of references

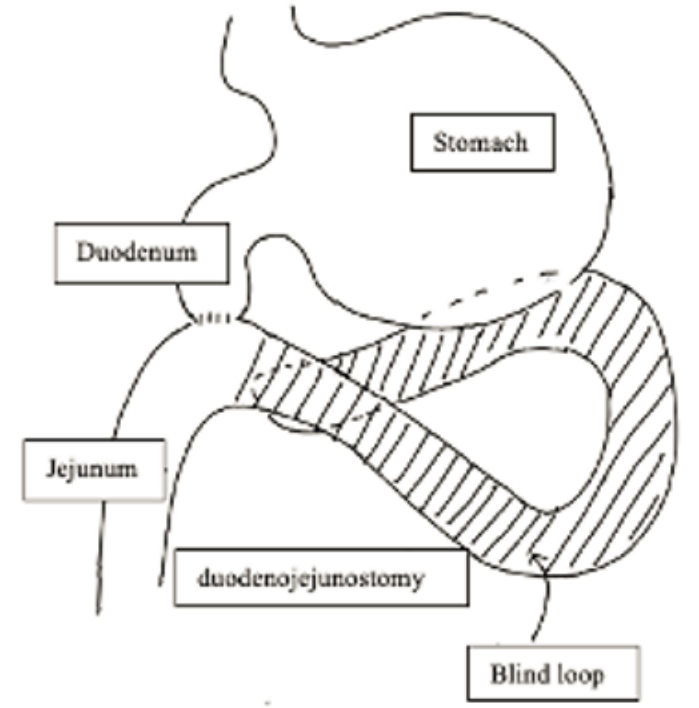

Figure 2

Blind loop secondary to duodenojejunostomy

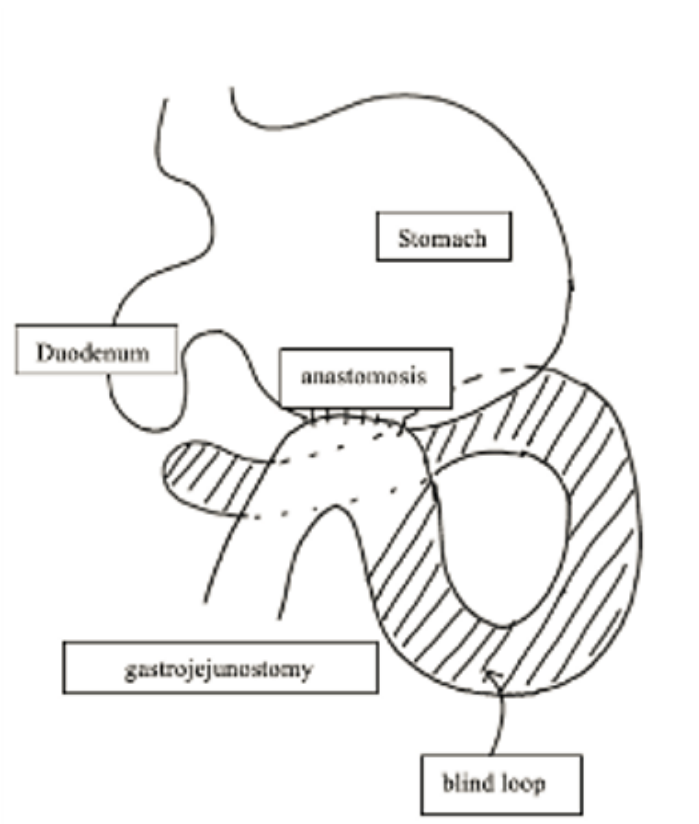

Figure 3

Blind loop secondary to gastrojejunostomy