Accepted version
Licence: CC-BY-NC-ND
Please cite as:
Giuseppe LAURITI (2023) "Opera mia" European Journal of Pediatric Surgery
DOI: https://doi. 10.1055/s-0043-1772173

European Journal of Pediatric Surgery
Thieme

Neonatal Intestinal Segmental Volvulus: What are the Differences with Midgut Volvulus?

Journal:	*European Journal of Pediatric Surgery*
Manuscript ID	EJPS-2023-05-6616-REV
Manuscript Type:	Review
Date Submitted by the Author:	08-May-2023
Complete List of Authors:	Casalino, Maria; Queen's University, Department of Family Medicine Miscia, Maria Enrica; University Gabriele d'Annunzio of Chieti Pescara Department of Medicine and Aging Science, Pediatric Surgery Unit Lauriti , Giuseppe ; University Gabriele d'Annunzio of Chieti and Pescara Department of Medicine and Aging Science, Department of Pediatric Surgery, Santo Spirito Hospital, Pescara, Italy Gauda, Estelle; The Hospital for Sick Children, Department of Neonatology Zani, Augusto; The Hospital for Sick Children, Division of General and Thoracic Surgery; University of Toronto, Department of Surgery Zani-Ruttenstock, Elke; The Hospital for Sick Children, Division of General and Thoracic Surgery
Keywords:	midgut volvulus, segmental volvulus, neonates, meta-analysis
Abstract:	Aim of the Study Intestinal volvulus in the neonate is a surgical emergency either caused by midgut volvulus (MV) with intestinal malrotation or less commonly, by segmental volvulus (SV) without intestinal malrotation. The aim of our study was to investigate if MV and SV can be differentiated by clinical course, intra-operative findings, and postoperative outcomes. Methods Using a defined search strategy, two investigators independently identified all studies comparing MV and SV in neonates. PRISMA guidelines were followed, and meta-analysis was performed using RevMan5.3. Results Of 1,026 abstracts screened, 104 full-text articles were analyzed, and 3 comparative studies were selected (112 patients). There were no differences in gestational age (37 vs. 36 weeks), birth weight (2,989 vs.2,712 grams), age at presentation (6.9 vs.3.8 days). SV was more commonly associated with abnormal findings on fetal US (65% vs 11.6%; p<0.00001). Preoperatively, SV was more commonly associated with abdominal distension (32% vs.77%; p<0.05), whereas MV with a whirlpool sign on ultrasound (57% vs.3%; p<0.01). Bilious vomiting had similar incidence in both (88±4% vs.50±5%). Intraoperatively, SV had a higher incidence of intestinal atresia (2% vs. 19%; p<0.05) and need for bowel resection (13% vs. 91%; p<0.00001, Figure). There were no differences in postoperative complications (13% MV vs. 14% SV), short

	bowel syndrome (15% MV vs. 0% SV; data available only from 1 study), and mortality (12% MV vs. 2% SV). Conclusion Our study highlights the paucity of studies on SV in neonates. Nonetheless, our meta-analysis clearly indicates that SV is an entity on its own with distinct clinical features and intra-operative findings that are different from MV. SV should be considered as one of the differential diagnoses in all term and preterm babies with bilious vomiting after MV was ruled out – especially if abnormal fetal US and abdominal distension is present.

SCHOLARONE™
Manuscripts

MANUSCRIPT

1. INTRODUCTION

Intestinal volvulus in the neonate is a surgical emergency either caused by midgut volvulus (MV) due to intestinal malrotation or, less commonly, by segmental volvulus (SV) without intestinal malrotation. MV is defined by the twisting of the entire small intestine and parts of the large intestine around the superior mesenteric artery (SMA), superior mesenteric vein (SMV), and its abnormally narrow mesentery [1]. It is an extremely time sensitive entity as, depending on the degree of bowel ischemia, it can result in the loss of most of the intestine and in some cases even death. MV with malrotation is caused by an intestinal rotation anomaly that occurs during the 10th week of gestation, leading to incomplete rotation and abnormal fixation of the intestine [2,3]. There is uniform consensus that MV is a surgical emergency requiring prompt diagnosis and treatment. The classical presentation in neonates is bilious vomiting. The gold standard for investigation is an upper gastrointestinal (GI) contrast study, although some centers also perform abdominal ultrasonography [4]. A normal upper GI contrast study and/or an abdominal ultrasound scan (US) with a normal SMA/SMV relationship in the absence of a “whirlpool sign” typically rule out MV.

From the few case reports and case series, we know that SV occurs when there is twisting of a segment of bowel in the absence of an underlying rotational anomaly [5]. SV can occur pre- or postnatally and can be associated with intestinal pathologies such as intestinal atresia, meconium ileus, congenital bands, or a duplication cyst [5,6]. Contrary to MV, SV may manifest with a vague clinical presentation and with nonspecific radiological findings, thus making it

For Peer Review

Georg Thieme Verlag KG, P.O. Box 30 11 20, 70451 Stuttgart, Germany

challenging to diagnose before the onset of significant bowel ischemia [5]. In most cases, SV diagnosis is only made intraoperatively, and surgical management often entails resection of ischemic bowel. To gain more insights into the specific differences of the two conditions and better understand their different nature, we aimed to compare the clinical course, intraoperative findings, and postoperative outcomes between MV and SV.

2. MATERIALS AND METHODS

2.1 Data sources and study selection

This study was registered on the international prospective register of systematic reviews PROSPERO (registration #CRD42022382088) (National institute for Health Research) [6]. The systematic review was drafted according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement [7]. A systematic review of the English literature was made using a defined search strategy (**Table 1**). Two investigators (MC, MEM) independently searched scientific databases (PubMed, Scopus, Cochrane Collaboration, and Web of Science) looking for studies reporting on malrotation, volvulus or segmental volvulus in newborns published up to March 2023. MeSH headings and terms used were “segmental volvulus”, “malrotation”, “neonates”, and “newborn” (**Supplementary file 1**). Reference lists were searched to identify relevant cross-references. Case reports, opinion articles, experimental studies, and case series with less than 10 patients were excluded. All grey literature publications (i.e. reports, theses, conference proceedings, bibliographies, commercial documentations, and

For Peer Review

Georg Thieme Verlag KG, P.O. Box 30 11 20, 70451 Stuttgart, Germany

official documents not published commercially) were excluded. Full text articles of potentially eligible

studies were retrieved and independently assessed for suitability by two investigators (MC, MEM). We included only studies (trials, cohort, and case-control) that compared the management of MV with SV in newborns. If two or more studies had overlapping patient cohorts, for each outcome measure we included only the article with the largest number of patients. Any disagreement over the eligibility of a specific study was resolved through the discussion with a third author (GL). Outcome measures included patient demographics, clinical features, diagnostic and therapeutic management, and postoperative outcome.

2.2 Statistical analysis

Categorical variable frequencies were compared using Pearson's chi-square test or the two-tailed Fisher exact probability test, as appropriate. When median and range were reported, mean±SD were estimated, as previously reported [8]. Meta-analysis of comparative studies was conducted with RevMan 5.4 [9]. Data are presented as risk ratio (RR) for categorical variables, and mean differences (MD) for continuous variables, along with 95% confidence intervals (CI) using the random-effects model, with p values shown for Z test for overall significance and I2 statistic for

heterogeneity. A p-value <0.05 was considered statistically significant.

2.3 Quality assessment

For Peer Review

Georg Thieme Verlag KG, P.O. Box 30 11 20, 70451 Stuttgart, Germany

Risk of bias for individual studies was assessed in duplicate (MEM and GL) using the methodological index for non-randomized studies (MINORS) [10]. Differences between the two reviewers (MEM and GL) were resolved through consensus and discussion with a third author (EZR). The total score for this 12-item instrument ranges 0–24 points with a validated "gold standard" cut-off of 19.8. We assessed the methodological quality for each outcome by grading the quality of evidence using the Grading of Recommendations Assessment, Development and Evaluation (GRADE) methodology [11]. Quality of evidence was rated as high, moderate, low, and very low for each outcome. Observational studies start with a low quality of evidence. The quality of evidence was rated down in the presence of risk of bias, inconsistency, indirectness, imprecision, and publication bias. For assessment of risk of bias in observational studies, we used the MINORS instrument. Inconsistency was determined according to heterogeneity. We produced I^2 values to assess heterogeneity. I^2 value of 0–40, 30–60, 50–90, and 75–100% were considered as low, moderate, substantial, and considerable heterogeneity, respectively. Imprecision was assessed using optimal information size (OIS), which was based on 25% relative risk reduction, 0.05 of α error and 0.20 of β error [12].

For Peer Review

RESULTS

Of the 1,026 abstracts that were screened, 104 full-text articles were analyzed, and 3 comparative articles were included (4, 13, 14), for a total of 112 patients (69 MV and 43 SV; **Figure 1**). There were no differences in patient demographics between MV and SV with regards to gestational age (36.5±0.8 *versus* 35.7±0.9 weeks; 95% confidence intervals (CI) 0.76 [-0.18, 1.70], I^2=20%; p=ns; **Figure 2a)** and birth weight (2,989±271 *versus* SV 2,712±226 grams; 95% CI

Georg Thieme Verlag KG, P.O. Box 30 11 20, 70451 Stuttgart, Germany

274.48 [-144.53, 693.49], I^2=78%; p=ns; **Figure 2b**). Moreover, the age at presentation was similar between MV (6.9±4.9 days) and SV (3.8±3.8 days; 95% CI 3.65 [-8.51, 15.80], I^2=97%; p=ns; **Figure 2c**). Clinically, bilious vomiting as presenting symptom was similar between MV (44/50 pts, 88±4.3%) and SV (13/26 pts, 50±54.1%; 95% CI 1.82 [0.13, 25.23], I^2=97%; p=ns; **Figure 3a**), whereas, abdominal distension was significantly more often reported in babies with SV (33/43 pts, 76.7±17.9%) compared to MV (22/69 pts, 31.9±25.0%; 95% CI 0.36 [0.14, 0.96], I^2=81%; p<0.05; **Figure 3b**).

At prenatal ultrasonography, the incidence of abnormal findings such as polyhydramnios, bowel dilatation, and presence of an abdominal mass, was less common in MV (8/69 pts, 11.6±1.3%) compared to SV (28/43 pts, 65.1±29.4%; 95% CI 0.19 [0.10, 0.40], I2=0%; p<0.00001; **Figure 4a**). On postnatal abdominal ultrasonography, a classical whirlpool sign was more frequently seen in MV (26/46 pts, 56.5±1.6%) than SV (1/34 pts, 2.9±4.2%; 95% CI 12.22 [2.51, 59.47], I2=0%; p<0.01; **Figure 4b**), although an inversion of SMA/SMV relationship was similarly reported in both conditions (6/46 pts, 13.0±5.2% in MV versus 0/34 pts, 0% in SV; 95% CI 4.59 [0.56, 37.57], I2=0%; p=ns; **Figure 4c**).

Intra-operatively, the incidence of ileal atresia was less commonly detected in MV (1/50 pts, 2±3.0%) compared to SV (5/26 pts, 19.2±15.2%; 95% CI 0.13 [0.02, 0.73], I2=0%; p<0.05; **Figure 5a**). Moreover, resection of bowel was less often needed in MV (9/69 pts, 13.0±0.8%) compared to SV (39/43 pts, 90.7±3.7%; 95% CI 0.17 [0.09, 0.35], I2=20%; p<0.00001; **Figure 5b**).

No differences were found between MV and SV with regards to overall post-operative complications (6/46 pts, 13.0±3.0% versus 7/34, 20.6±4.2%, respectively; 95% CI 0.62 [0.23,

For Peer Review

1.67], I2=0%; p=ns; **Figure 6a**) and post-operative obstructive ileus (3/46 pts, 5.5±1.5% versus 4/34 pts, 11.8±0%, respectively; 95% CI 0.55 [0.13, 2.35], I2=0%; p=ns; **Figure 6b**). The mortality rate was similar between the two groups (MV 8/69 pts, 11.6±11.6% versus SV 1/43 pts, 2.3±7.8%; 95% CI 2.37 [0.55, 10.29], I2=0%; p=ns; **Figure 6c**). The incidence of short bowel syndrome was reported only in one paper (Chung et al 2020), with no significant differences between MV (4/27 pts, 15%) and SV (none; p=ns, **Figure 6d**).

4. DISCUSSION

The present study shows that SV and MV have multiple differences that make these two entities distinguishable from each other. To the best of our knowledge, this is the first meta-analysis that comparatively analyzes MV and SV in terms of demographics, clinical course, intra-operative findings, and outcomes. In the studies analyzed, more than one third of the patients presented with SV. Compared to MV, SV was more commonly associated with the presence of abnormal fetal US, abdominal distension, absence of a whirlpool sign on doppler US, intestinal pathologies such as intestinal atresia and a higher requirement for bowel resection.

With regards to demographics including GA and BW, all three comparative studies noted no differences between SV and MV [4,13,14]. The median gestational age was approximately 36 weeks for both groups, indicating that both pathologies commonly occur in late premature babies. This finding is supported by Kargl et al., that reported on a series of 15 premature patients with SV, suggesting that this commonly prenatally occurring intestinal event would often lead to premature delivery [15]. Furthermore, the median age at presentation was in the immediate

For Peer Review

Georg Thieme Verlag KG, P.O. Box 30 11 20, 70451 Stuttgart, Germany

postnatal period for both entities. Conversely, Maya-Enero et al. reported that surgery was performed at a significantly lower age in SV compared to MV [14]. Overall, this heterogenicity in findings highlights that both entities need to be considered when evaluating preterm as well as term neonates for volvulus especially in their first months of life.

Bilious vomiting was the most common presenting symptom in both MV and SV [4,14], whereas abdominal distension was more commonly associated with SV than MV. The latter was reported in all comparative studies herein analyzed [4, 13, 14], as well as in case series previously reported [5,16]. The higher incidence of abdominal distension in SV could be explained by the difference in pathophysiology compared to MV. SV acts like a distal mechanical obstruction mainly occurring in the ileum, thus more likely causing distension of the jejunal and proximal ileal loops (4). On the other hand, MV is caused by a proximal obstruction that involved all loops of bowel, including the most proximal [17]. With this concept in mind, it is important to still consider the differential diagnosis of SV in a neonate that gets worked up for bilious vomiting after MV has successfully been ruled out, especially when the patient demonstrates abdominal distension.

When investigating a neonate for intestinal volvulus, an upper GI contrast series remains the gold standard [13]. However, abdominal US has gained more popularity in the recent years due to its increased availability and absence of radiation, making it a useful additional imaging modality [2]. The most common US finding in the workup of MV is the inversion of SMA/SMV that is illustrated by the classic “whirlpool sign” on Doppler US [18]. In our meta-analysis, two studies assessed the presence of a whirlpool sign and found that it was significantly more common in

For Peer Review

Georg Thieme Verlag KG, P.O. Box 30 11 20, 70451 Stuttgart, Germany

patients with MV [4,13]. The authors explain this difference as being the consequence of the underlying pathophysiology that profoundly sets the two entities apart. This is also reflected by the fact that malrotation was reported in 68 out of a total of 69 patients with MV (99%), whereas a mesenteric malposition was found only in 4 out of 43 patients with SV (9%) [4,13,14].

MV occurs due to abnormal embryonic gut development, where the normal bowel rotation is either halted or diverted at different stages [2]. Conversely, SV occurs without the underlying presence of malrotation but can be associated with congenital abnormalities, including congenital bands, duplication cysts, intestinal herniation, meconium ileus, and intestinal atresia [13,19]. This is also reflected in our results, whereby intestinal atresia was significantly more common in patients with SV. However, associated intestinal anomalies were found in patients with SV only 35% of the time. This may help explain the typical nonspecific radiologic findings in patients with SV, again making it challenging to diagnose preoperatively and before the onset of significant bowel ischemia.

The mainstay of treatment in both these pathologies is either open or less commonly laparoscopic surgery with removal of necrotic bowel segments [20]. Comparatively, SV had a higher incidence of bowel resection compared to MV [4, 14]. This can be explained by the fact that some neonates with MV require derotation alone without resection, as the bowel has maintained sufficient perfusion. Nonetheless, babies with SV overall have better outcomes, likely due to the fact that the ischemic area is limited to a bowel segment only, compared to patients with MV that may face additional hemodynamic instability[4]. This and the absence of intestinal malrotation in SV may support the argument in favor of laparoscopy for diagnosis and treatment of these neonates.

For Peer Review

Georg Thieme Verlag KG, P.O. Box 30 11 20, 70451 Stuttgart, Germany

In terms of post-operative complications, we did not find differences between the patients with SV and MV. This is in contrary to Khen-Dunlop et al., that reported on a higher incidence of post-operative morbidity in SV patients [13]. Their study included three patients with SV, who required reoperation for secondary intestinal obstruction and abdominal wall hernia[13]. On the other hand, Chung et al. reported no differences in immediate post-operative complications between the two groups [4]. In their study, however, all but one patient with MV that required bowel resection suffered from short bowel syndrome, leading to two cases of mortality [4]. Furthermore, the incidence of mortality was similar between patients with SV and MV. However, these findings are limited by the fact that only one study out of the three provided data on short bowel syndrome.

4.1 Limitation of the study

We are aware of the limitations of our meta-analysis, which relies on the quality of the studies and data available in the literature. All the 3 studies included were retrospective observational studies [4,13,14]. As expected, a blinded evaluation of objective endpoints was not possible. Moreover, none of the study have reported with regards to the loss to follow-up and there were a broad lack of data regards the length of follow-up. Therefore, in our meta-analysis, none of the studies reached the gold standard cut-off on MINORS of 19.8 out of 24 (**Table 2**).

According to the GRADE methodology, the quality of evidence of the meta-analysis was low with regards some pre-operative data (i.e. gestational age and prenatal ultrasonography), the whirlpool sign at abdominal ultrasound scans among the preoperative imaging studies, and the incidence of resection of the bowel among the two groups (**Table 3**). Since the data were

For Peer Review

Georg Thieme Verlag KG, P.O. Box 30 11 20, 70451 Stuttgart, Germany

obtained from a small number of studies, their considerable heterogeneity could generate possible bias. Nonetheless, when independently assessed by two authors (GL and MEM) using A Measurement Tool to Assess Systematic Reviews (AMSTAR) [21], the present systematic review and meta-analysis received a sufficient score (**Supplementary file 2**) and the PRISMA checklist was completed (**Supplementary file 3**).

5. Conclusions

Although SV and MV showed no differences in some demographic and clinical features, there are several aspects of clinical presentation and course that clearly differentiate SV and MV from each other. SV is frequently associated with abnormal fetal US, postnatal abdominal distension, intestinal pathologies such as intestinal atresia, as well as a higher need for bowel resection and should be therefore considered in neonates with bilious vomiting after successful exclusion of MV. This is especially the case when abnormal antenatal US scans and abdominal distension are present.

The literature on SV is currently limited but, with increasing awareness, SV will make its way in the list of differential diagnoses of neonatal bowel obstruction and will have a chance to result in early surgical intervention to prevent morbidity and mortality.

6. Declaration of interest

The authors have no conflicts of interest to declare.

7. Author Contributions Statement

For Peer Review

MC, GL, EZR: Conception/design, analysis and interpretation, participated in drafting.

MC, MEM: Data acquisition.

GL, MEM: Quality assessment.

GL, AZ, EG, EZR: Participated in revision, gave final approval.

8. Submission declaration

This work has not been published previously and is not under consideration for publication elsewhere.

9. Role of the funding source

No funding was secured for this article

10. REFERENCES

1. Le CK, Nahirniak P, Anand S, et al. Volvulus. [Updated 2022 Sep 12]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK441836/

2. Langer JC. Intestinal Rotation Abnormalities and Midgut Volvulus. Surg Clin North Am. 2017 Feb;97(1):147-159.

3. D'Agostino J. Chapter 83 - Malrotation and Midgut Volvulus. In: Baren JM, Rothrock SG, Brennan JA, Brown L, editors. Pediatric Emergency Medicine. Philadelphia: W.B. Saunders; 2008. p. 630-3.

4. Chung JH, Kim W, Kim SY. Comparison of total midgut volvulus and segmental volvulus in neonates. J Pediatr Surg. 2021 Aug;56(8):1375-1377.

5. Kim SH, Cho YH, Kim HY. Primary Segmental Volvulus of Small Intestine: Surgical Perspectives According to Age at Diagnosis. Front Pediatr. 2019 Apr 17;7:146.

For Peer Review

6. PROSPERO international prospective register of systematic reviews. Available at: https://www.crd.york.ac.uk/prospero [on Mar 25th 2023]

7. Moher D, Liberati A, Tetzlaff J, et al. Preferred reporting items for systematic reviews and meta-analyses: the PRISMA statement. PLoS Med. (2009) 21;6(7):e1000097.

8. Hozo SP, Djulbegovic B, Hozo I. Estimating the mean and variance from the median, range, and the size of a sample. BMC Med Res Methodol. (2005) 20:5:13.

9. Review Manager (RevMan). The Nordic Cochrane Centre. The Cochrane Collaboration, Copenhagen. (2014)

10. Slim K, Nini E, Forestier D, et al. Methodological index for non-randomized studies (MINORS): development and validation of a new instrument. ANZ J Surg. (2003) 73:712-6.

11. Guyatt GH, Oxman AD, Vist GE, et al. GRADE: an emerging consensus on rating quality of evidence and strength of recommendations. BMJ. (2008) 336:924-6.

12. Dupont WD, Plummer WD. Power and sample size calculations: a review and computer program. Control Clin Trials. (1990) 11:116-28.

13. Khen-Dunlop N, Beaudoin S, Marion B, et al. Segmental volvulus in the neonate: A particular clinical entity. J Pediatr Surg. 2017 Mar;52(3):454-457.

14. Maya-Enero S, Prat-Ortells J, Martín-Solé O, et al. Distinguishing outcomes of neonatal intestinal volvulus: Review of our experience over the last 20 years. Acta Paediatr. 2022 Feb;111(2):284-290.

15. Kargl S, Wagner O, Pumberger W. Volvulus without malposition--a single-center experience. J Surg Res. 2015 Jan;193(1):295-9.

16. Drewett M, Burge DM: Late-onset volvulus without malrotation in preterm infants. J Pediatr Surg. 2009, 44: 358-361.

17. Turnage RH, Heldmann M. Chapter 119 - Intestinal Obstruction. In: Feldman M, Friedman LS, Brandt LJ, editors. Sleisenger and Fordtran's Gastrointestinal and Liver Disease (Ninth Edition). Philadelphia: W.B. Saunders; 2010. p. 2105-20.e2.

18. Pracros JP, Sann L, Genin G, et al. Ultrasound diagnosis of midgut volvulus: the "whirlpool" sign. Pediatr Radiol. 1992;22(1):18-20.

19. Kitano, Y., Hashizume, K. & Ohkura, M. Segmental small-bowel volvulus not associated with malrotation in childhood. Pediatr Surg Int. 1995; **10**, 335–338.

For Peer Review

20. Hagendoorn J, Vieira-Travassos D, van der Zee D. Laparoscopic treatment of intestinal malrotation in neonates and infants: retrospective study. Surg Endosc. 2011 Jan;25(1):217-20.

21. Shea BJ, Grimshaw JM, Wells GA, et al. Development of AMSTAR: a measurement tool to assess the methodological quality of systematic reviews. BMC Med Res Methodol (2007) 7: 10.

11. TABLE LEGENDS

Table 1: Defined search strategy

Table 2: Risk of bias assessment for individual studies using methodological index for nonrandomized studies (MINORS) [11]

Table 3: GRADE Evidence Profile for the present Meta-Analysis [12]

For Peer Review

13. FIGURE LEGENDS

Figure 1: Diagram of workflow in the meta-analysis

Figure 2: Forest plot comparison of patient's demographics between MV and SV with regards to gestational age (**Figure 2a**), birth weight (**Figure 2 b**), and age at presentation (Figure **2c).**

Figure 3: Forest plot comparison of presenting symptoms between MV and SV with regards to bilious vomiting (**Figure 3 a**) and abdominal distension (**Figure 3b**).

Figure 4: Forest plot comparison of diagnostic imaging between MV and SV with regards to prenatal US (**Figure 4a**), presence of a whirlpool sign on postnatal US (Figure 4b) and SMA/SMV inversion on postnatal US (**Figure 4c**).

Figure 5: Forest plot comparison of the intra-operative findings between MV and SV with regards to the incidence of ileal atresia (**Figure 5a**) and required resection of bowel (**Figure 5b**).

Figure 6: Forest plot comparison of the post-operative outcomes between MV and SV with regards to overall post-operative complications (**Figure 6a**), obstructive ileus (**Figure 6b**), mortality rate (**Figure 6c**), and the incidence of short bowel syndrome (**Figure 6d**).

For Peer Review

For Peer Review

Figure 1

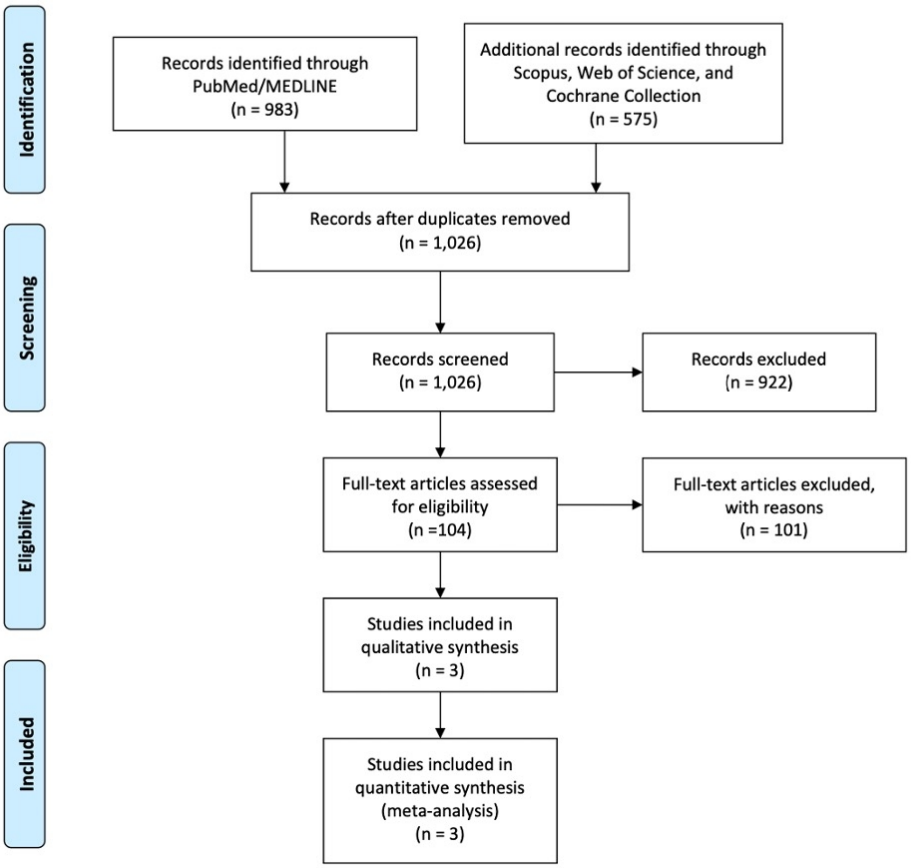
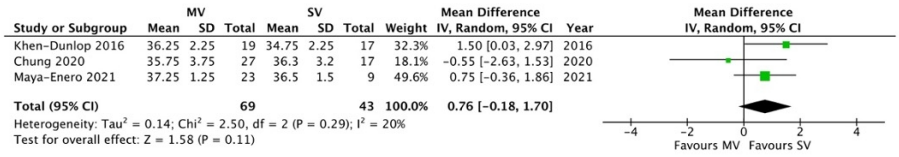


Figure 1

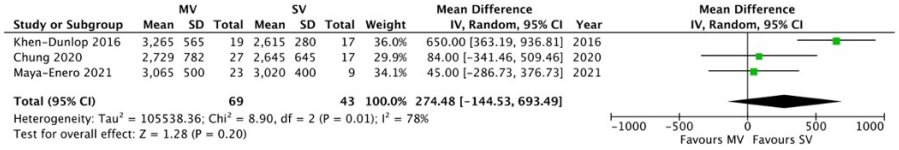
176x190mm (144 x 144 DPI)

Figure 2a-c

2a: Gestational age



2b: Birth weight



2c: Age at presentation

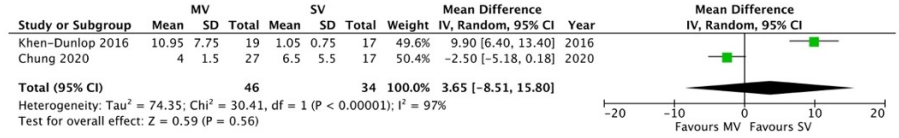
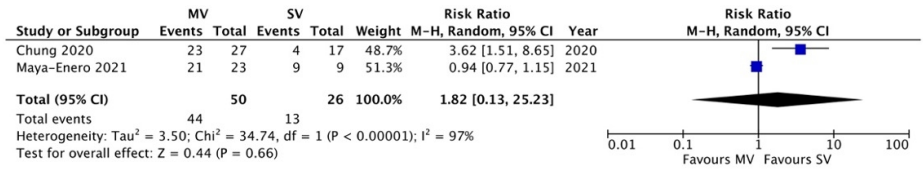


Figure 2

206x190mm (144 x 144 DPI)

Figure 3a-b

3a: Bilious vomiting



3b: Abdominal distension

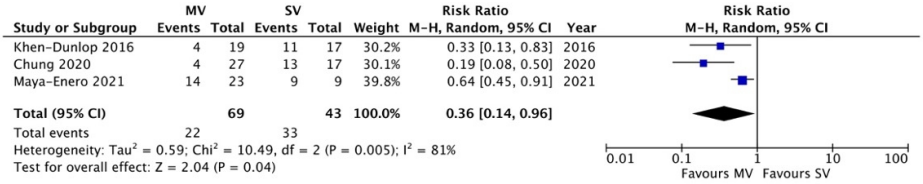
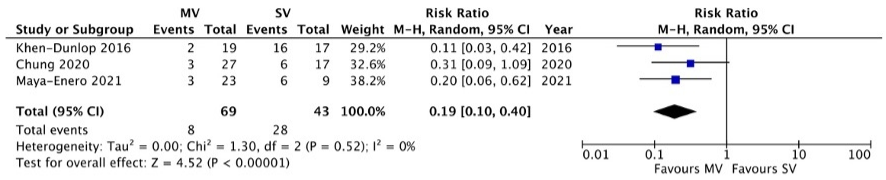


Figure 3

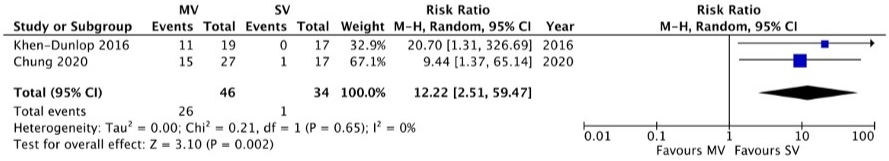
216x167mm (144 x 144 DPI)

Figure 4a-c

4a: Prenatal US



4b: Postnatal US: Whirlpool sign



4c: Postnatal US: SMA/SMV inversion

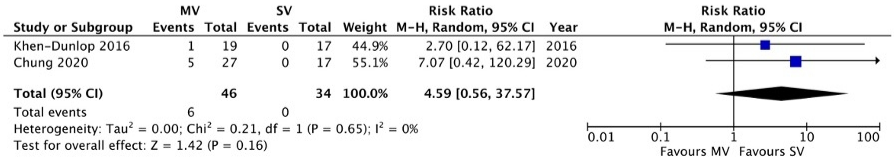
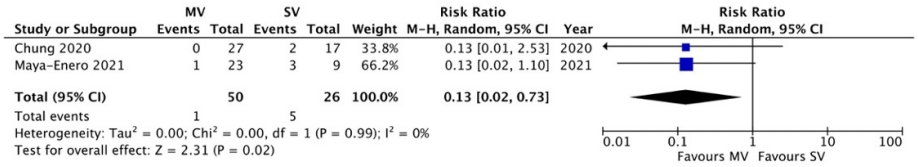


Figure 4

176x190mm (144 x 144 DPI)

Figure 5a-b

5a: Intra-op: Intestinal atresia



5b: Intra-op: Bowel resection

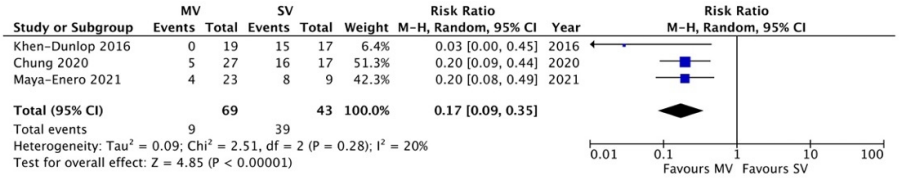


Figure 5

215x170mm (144 x 144 DPI)

Figure 6-d

Figure 6b: Overall complication rate

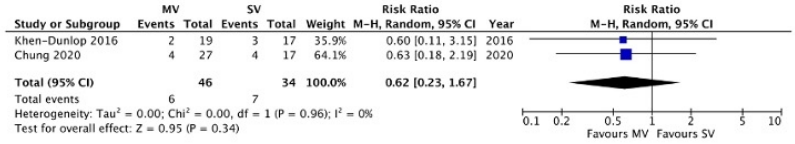


Figure 6b: Obstructive ileus

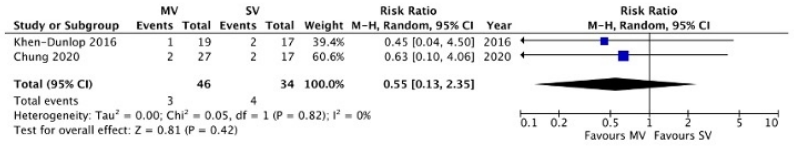


Figure 6c: Mortality rate

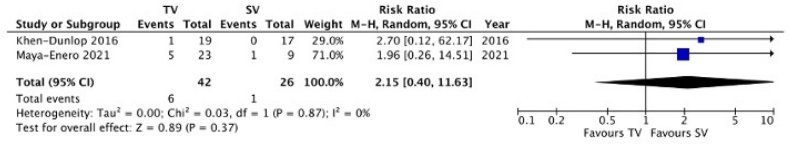


Figure 6d: Short bowel syndrome

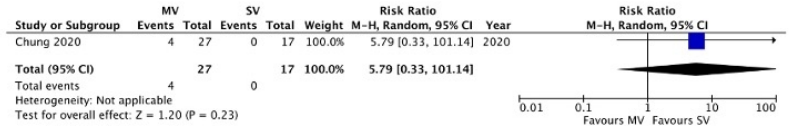


Figure 6

145x190mm (144 x 144 DPI)

Table 1. Inclusion criteria of the Systematic Review

Publication	
Language	English
Time period	January 1950 – July 2022
Subject	Human studies
Study type	Retrospective Prospective Case-control Cohort
Excluded	Case-report Case series (<10 patients) Editorials Letters Grey Literature
Keywords	Segmental volvulus Malrotation Neonates Newborn

Table 1

138x148mm (144 x 144 DPI)

Table 2. Risk of bias assessment for individual studies using methodological index for nonrandomized studies (MINORS) [11]

Item	Chung (4)	Khen-Dunlop (6)	Maya-Enero (14)
1. A clearly stated aim	2	2	2
2. Inclusion of consecutive patients	2	2	2
3. Prospective collection of data	0	0	0
4. Endpoints appropriate to the aim of the study	2	2	2
5. Unbiased assessment of the study endpoint	0	0	0
6. Follow-up period appropriate to the aim of the study	0	0	1
7. Loss to follow-up less than 5%	0	0	0
8. Prospective calculation of the study size	0	0	0
9. An adequate control group	2	2	2
10. Contemporary groups	2	2	2
11. Baseline equivalence of groups	2	2	2
12. Adequate statistical analyses	2	2	2
Total score	**14**	**14**	**15**

0 = not reported; **1** = reported but inadequate; **2** = reported and adequate. Validated "gold standard" cut-off: 19.8.

Table 2

166x179mm (144 x 144 DPI)

Table 3. GRADE Evidence Profile for the present Meta-Analysis [12]

No. of studies	Study design	Risk of bias	Inconsistency	Indirectness	Imprecision	Other Considerations	No. of patients: Cases	No. of patients: Controls	Effect: Relative (95% CI)	Effect: Absolute (95% CI)	Quality
Gestational age in MV *versus* SV							**MV**	**SV**			
3	OS	Moderate[a]	Low	Not serious	Serious[b]	None	69	43	—	MD 0.76 higher (from 0.18 lower to 1.70 higher)	⊗⊗OO LOW
Birth weight in MV *versus* SV							**MV**	**SV**			
3	OS	Moderate[a]	Substantial	Not serious	Serious[b]	None	69	43	—	MD 274.48 higher (from 144.53 lower to 693.49 higher)	⊗OOO VERY LOW
Abnormal prenatal ultrasonography in MV *versus* SV							**MV**	**SV**			
3	OS	Moderate[a]	Low	Not serious	Serious[b]	None	8/69 (11.6%)	28/43 (65.1%)	RR 0.19 (0.10, 0.40)	535 fewer per 1000 (from 594 to 396 fewer)	⊗⊗OO LOW
Age at presentation in MV *versus* SV							**MV**	**SV**			
2	OS	Moderate[a]	Considerable	Not serious	Serious[b]	None	46	34	—	MD 3.65 higher (from 8.51 lower to 15.80 higher)	⊗OOO VERY LOW
Bilious vomiting in MV *versus* SV							**MV**	**SV**			
2	OS	Moderate[a]	Considerable	Not serious	Serious[b]	None	44/50 (88.0%)	28/43 (65.1%)	RR 1.82 (0.13, 25.23)	229 more per 1000 (from 243 fewer to 6766 more)	⊗OOO VERY LOW
Abdominal distension in MV *versus* SV							**MV**	**SV**			
2	OS	Moderate[a]	Substantial	Not serious	Serious[b]	None	22/69 (31.9%)	33/43 (76.7%)	RR 0.36 (0.14, 0.96)	448 fewer per 1000 (from 602 to 28 fewer)	⊗OOO VERY LOW
Whirlpool sign at abdominal USS in MV *versus* SV							**MV**	**SV**			
2	OS	Moderate[a]	Low	Not serious	Serious[b]	None	26/46 (56.5%)	1/34 (2.9%)	RR 12.22 (2.51, 59.47)	536 more per 1000 (from 72 to 2793 more)	⊗⊗OO LOW
SMA inversion at abdominal USS in MV *versus* SV							**MV**	**SV**			
2	OS	Moderate[a]	Low	Not serious	Serious[b]	None	6/46 (13.0%)	0/34 (0%)	RR 4.59 (0.56, 37.57)	130 more per 1000 (from 16 fewer to 1324 more)	⊗OOO VERY LOW
Ileal atresia in MV *versus* SV							**MV**	**SV**			
2	OS	Moderate[a]	Low	Not serious	Serious[b]	None	1/50 (2%)	5/26 (19.2%)	RR 0.13 (0.92, 4.79)	172 fewer per 1000 (from 16 fewer to 749 more)	⊗OOO VERY LOW
Resection of bowel in MV *versus* SV							**MV**	**SV**			
3	OS	Moderate[a]	Low	Not serious	Serious[b]	None	9/69 (13.0%)	39/43 (90.7%)	RR 0.17 (0.09, 0.35)	777 fewer per 1000 (from 852 to 608 fewer)	⊗⊗OO LOW
Post-operative complications in MV *versus* SV							**MV**	**SV**			
2	OS	Moderate[a]	Low	Not serious	Serious[b]	None	6/46 (13.0%)	7/34 (20.6%)	RR 0.62 (0.23, 1.67)	76 fewer per 1000 (from 154 fewer to 134 more)	⊗OOO VERY LOW
Post-operative obstructive ileus in MV *versus* SV							**MV**	**SV**			
2	OS	Moderate[a]	Low	Not serious	Serious[b]	None	3/46 (6.5%)	4/34 (11.8%)	RR 0.55 (0.13, 2.35)	53 fewer per 1000 (from 102 fewer to 159 more)	⊗OOO VERY LOW
Prevalence of deaths in MV versus SV							**MV**	**SV**			
2	OS	Moderate[a]	Low	Not serious	Serious[b]	None	6/42 (14.3%)	1/26 (3.8%)	RR 2.15 (0.40, 11.63)	105 more per 1000 (from 55 fewer to 971 more)	⊗OOO VERY LOW
Short bowel syndrome in MV *versus* SV							**MV**	**SV**			
1	OS	Moderate[a]	—	Not serious	Serious[b]	None	4/27 (14.8%)	0/17 (0%)	RR 5.79 (0.33, 101.14)	148 more per 1000 (from 21 fewer to 3094 more)	⊗OOO VERY LOW
Abnormal UGI contrast study in MV *versus* SV							**MV**	**SV**			
2	OS	Moderate[a]	Low	Not serious	Serious[b]	None	23/23 (100%)	2/5 (40%)	RR 2.09 (0.92, 4.79)	600 more per 1000 (from 44 fewer to 2086 more)	⊗OOO VERY LOW

MV: midgut volvulus; **SV:** segmental volvulus; **USS:** ultrasound scan; **SMA:** superior mesenteric artery; **UGI:** upper gastrointestinal.
[a] Bias due to possible confounding; [b] OIS not met
GRADE Working Group grades of evidence
High quality: Further research is very unlikely to change our confidence in the estimate of effect.
Moderate quality: Further research is likely to have an important impact on our confidence in the estimate of effect and may change the estimate.
Low quality: Further research is very likely to have an important impact on our confidence in the estimate of effect and is likely to change the estimate.
Very low quality: We are very uncertain about the estimate.

Table 3

131x151mm (300 x 300 DPI)

Supplementary file 1: Search strategy.

PubMed/MEDLINE

1. (segmental volvulus OR malrotation).mp.
2. (neonate*adj2 OR newborn*adj2).mp.
3. 1 AND 2

Scopus

TITLE-ABS-KEY ((segmental volvulus) OR (malrotation) AND (neonates) OR (newborn))

Cochrane Collaboration

1. MeSH descriptor: segmental volvulus, malrotation, neonates, newborn
2. Explode all trees

Web of Science

TOPIC ((segmental volvulus) OR (malrotation) AND (neonates OR newborn))

148x178mm (144 x 144 DPI)

Supplementary file 2: AMSTAR criteria [21] for the present systematic reviews and meta-analysis assessed by two authors.

Item	GL	MEM
1. Was an 'a priori' design provided?	1	1
2. Was there duplicate study selection and data extraction?	1	1
3. Was a comprehensive literature search performed?	1	1
4. Was the status of publication (i.e. grey literature) used as an inclusion criterion?	1	1
5. Was a list of studies (included and excluded) provided?	0	0
6. Were the characteristics of the included studies provided?	1	1
7. Was the scientific quality of the included studies assessed and documented?	1	1
8. Was the quality of the included studies used appropriately in formulating conclusions?	0	1
9. Were the methods used to combine the findings of studies appropriate?	1	1
10. Was the likelihood of publication bias assessed?	1	1
11. Was the conflict of interest included?	0	0
Total	8/11	9/11

0 = No, 1 = Yes

216x150mm (144 x 144 DPI)

Supplementary file 3: PRISMA checklist

Section/topic	#	Checklist item	Reported on page #
TITLE			
Title	1	Identify the report as a systematic review, meta-analysis, or both.	1
ABSTRACT			
Structured summary	2	Provide a structured summary including, as applicable: background; objectives; data sources; study eligibility criteria, participants, and interventions; study appraisal and synthesis methods; results; limitations; conclusions and implications of key findings; systematic review registration number.	2, 3
INTRODUCTION			
Rationale	3	Describe the rationale for the review in the context of what is already known.	4, 5
Objectives	4	Provide an explicit statement of questions being addressed with reference to participants, interventions, comparisons, outcomes, and study design (PICOS).	5
METHODS			
Protocol and registration	5	Indicate if a review protocol exists, if and where it can be accessed (e.g., Web address), and, if available, provide registration information including registration number.	5, 6
Eligibility criteria	6	Specify study characteristics (e.g., PICOS, length of follow-up) and report characteristics (e.g., years considered, language, publication status) used as criteria for eligibility, giving rationale.	5, 6
Information sources	7	Describe all information sources (e.g., databases with dates of coverage, contact with study authors to identify additional studies) in the search and date last searched.	5
Search	8	Present full electronic search strategy for at least one database, including any limits used, such that it could be repeated.	5, 6, S1
Study selection	9	State the process for selecting studies (i.e., screening, eligibility, included in systematic review, and, if applicable, included in the meta-analysis).	6
Data collection process	10	Describe method of data extraction from reports (e.g., piloted forms, independently, in duplicate) and any processes for obtaining and confirming data from investigators.	6
Data items	11	List and define all variables for which data were sought (e.g., PICOS, funding sources) and any assumptions and simplifications made.	6
Risk of bias in individual studies	12	Describe methods used for assessing risk of bias of individual studies (including specification of whether this was done at the study or outcome level), and how this information is to be used in any data synthesis.	7, S2
Summary measures	13	State the principal summary measures (e.g., risk ratio, difference in means).	6
Synthesis of results	14	Describe the methods of handling data and combining results of studies, if done, including measures of consistency (e.g., I^2) for each meta-analysis.	6
Risk of bias across studies	15	Specify any assessment of risk of bias that may affect the cumulative evidence (e.g., publication bias, selective reporting within studies).	7
Additional analyses	16	Describe methods of additional analyses (e.g., sensitivity or subgroup analyses, meta-regression), if done, indicating which were pre-specified.	N/A
RESULTS			
Study selection	17	Give numbers of studies screened, assessed for eligibility, and included in the review, with reasons for exclusions at each stage, ideally with a flow diagram.	7, 8
Study characteristics	18	For each study, present characteristics for which data were extracted (e.g., study size, PICOS, follow-up period) and provide the citations.	7, 8
Risk of bias within studies	19	Present data on risk of bias of each study and, if available, any outcome level assessment (see item 12).	S2
Results of individual studies	20	For all outcomes considered (benefits or harms), present, for each study: (a) simple summary data for each intervention group (b) effect estimates and confidence intervals, ideally with a forest plot.	8
Synthesis of results	21	Present results of each meta-analysis done, including confidence intervals and measures of consistency.	8, 9
Risk of bias across studies	22	Present results of any assessment of risk of bias across studies (see Item 15).	15
Additional analysis	23	Give results of additional analyses, if done (e.g., sensitivity or subgroup analyses, meta-regression [see Item 16]).	N/A
DISCUSSION			
Summary of evidence	24	Summarize the main findings including the strength of evidence for each main outcome; consider their relevance to key groups (e.g., healthcare providers, users, and policy makers).	10, 11, 12, 13, 14
Limitations	25	Discuss limitations at study and outcome level (e.g., risk of bias), and at review-level (e.g., incomplete retrieval of identified research, reporting bias).	15
Conclusions	26	Provide a general interpretation of the results in the context of other evidence, and implications for future research.	16
FUNDING			
Funding	27	Describe sources of funding for the systematic review and other support (e.g., supply of data); role of funders for the systematic review.	16

N/A: not available. **S:** Supplementary file.

127x173mm (300 x 300 DPI)