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

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Abstract

Objective Intestinal volvulus in the neonate is a surgical emergency caused by either 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, intraoperative 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 a meta-analysis was performed using RevMan 5.3.

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 g), and age at presentation (6.9 vs. 3.8 days). SV was more commonly associated with abnormal findings on fetal ultrasound (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 ($88 \pm 4\%$ vs. $50 \pm 5\%$). Intraoperatively, SV had a higher incidence of intestinal atresia (2 vs. 19%; $p < 0.05$) and need for bowel resection (13 vs. 91%; $p < 0.00001$). There were no differences in postoperative complications (13% MV vs. 14% SV), short bowel syndrome (15% MV vs. 0% SV; data available only from one 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 intraoperative 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.

Keywords
- midgut volvulus
- segmental volvulus
- neonates
- meta-analysis

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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 (US) scan 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 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.

Materials and Methods

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 (M.C., M.E.M.) independently searched scientific databases (PubMed, Scopus, Cochrane Collaboration, and Web of Science) looking for studies reporting on malrotation, volvulus, or SV 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 official documents not published commercially) were excluded. Full-text articles of potentially eligible studies were retrieved and independently assessed for suitability by two investigators (M.C., M.E.M.). We included only studies (trials, cohort, and case–control) that compared the management of MV with SV in newborns. MV was defined as complete MV involving the entire small bowel related to intestinal malrotation, while SV was defined as twisting of only a part of the small bowel, irrespective of the mesenteric pattern.

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 (G.L.). Outcome measures included patient demographics, clinical features, diagnostic and therapeutic management, and postoperative outcome.

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 ± standard deviation 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 for continuous variables, along with 95% confidence intervals (CIs) using the random-effects model, with *p*-values shown for Z-test for overall significance and I^2 statistic for heterogeneity. A *p*-value of less than 0.05 was considered statistically significant.

Table 1 Inclusion criteria of the systematic review

Publication	
Language	English
Time period	January 1950–March 2023
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

Quality Assessment

Risk of bias for individual studies was assessed in duplicate (M.E.M. and G.L.) using the methodological index for non-randomized studies (MINORS).[10] Differences between the two reviewers (M.E.M. and G.L.) were resolved through consensus and discussion with a third author (E.Z.-R.). The total score for this 12-item instrument ranges from 0 to 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, which was based on 25% relative risk reduction, 0.05 of α error, and 0.20 of β error.[12]

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; ►Fig. 1). There were no differences in patient demographics between MV and SV with regards to gestational age (GA; 36.5 ± 0.8 vs. 35.7 ± 0.9 weeks; 95% CI: 0.76 [−0.18,

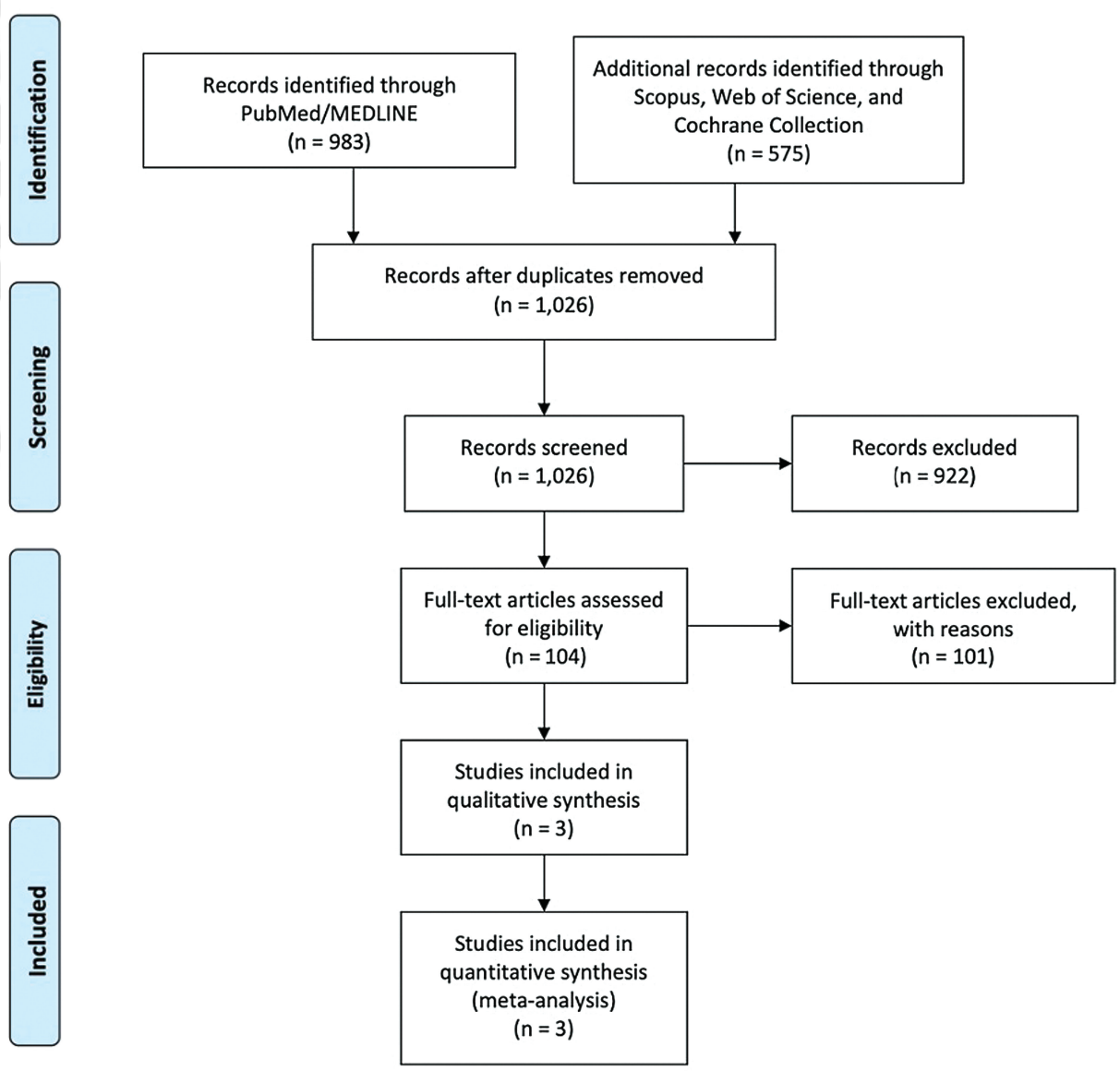

Fig. 1 Diagram of workflow in the meta-analysis.

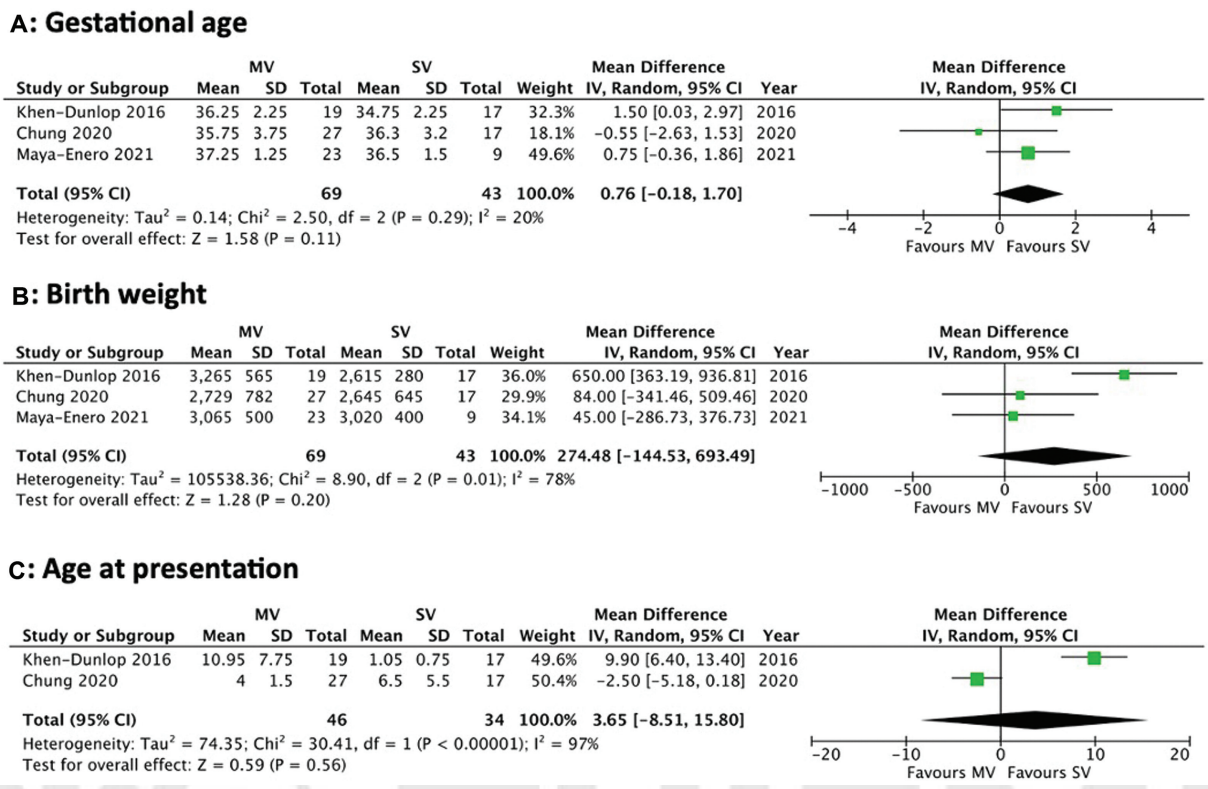

Fig. 2 Forest plot comparison of patient's demographics between MV and SV with regards to gestational age (**A**), birth weight (**B**), and age at presentation (**C**). MV, midgut volvulus; SV, segmental volvulus.

1.70], $I^2 = 20\%$; $p =$ ns; ►**Fig. 2A**) and birth weight (BW: 2,989 ± 271 vs. SV: 2,712 ± 226 g; 95% CI: 274.48 [−144.53, 693.49], $I^2 = 78\%$; $p =$ ns; ►**Fig. 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; ►**Fig. 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; ►**Fig. 3A**), whereas, abdominal distension was significantly more often reported in babies with SV (33/43 pts,

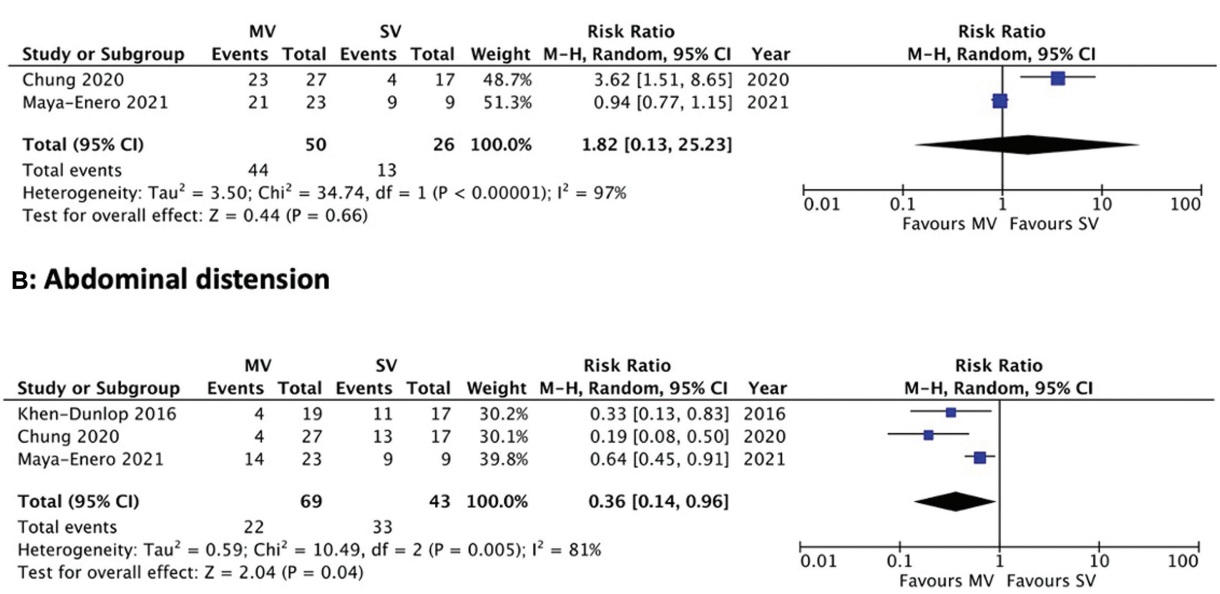

Fig. 3 Forest plot comparison of presenting symptoms between MV and SV with regards to bilious vomiting (**A**) and abdominal distension (**B**). MV, midgut volvulus; SV, segmental volvulus.

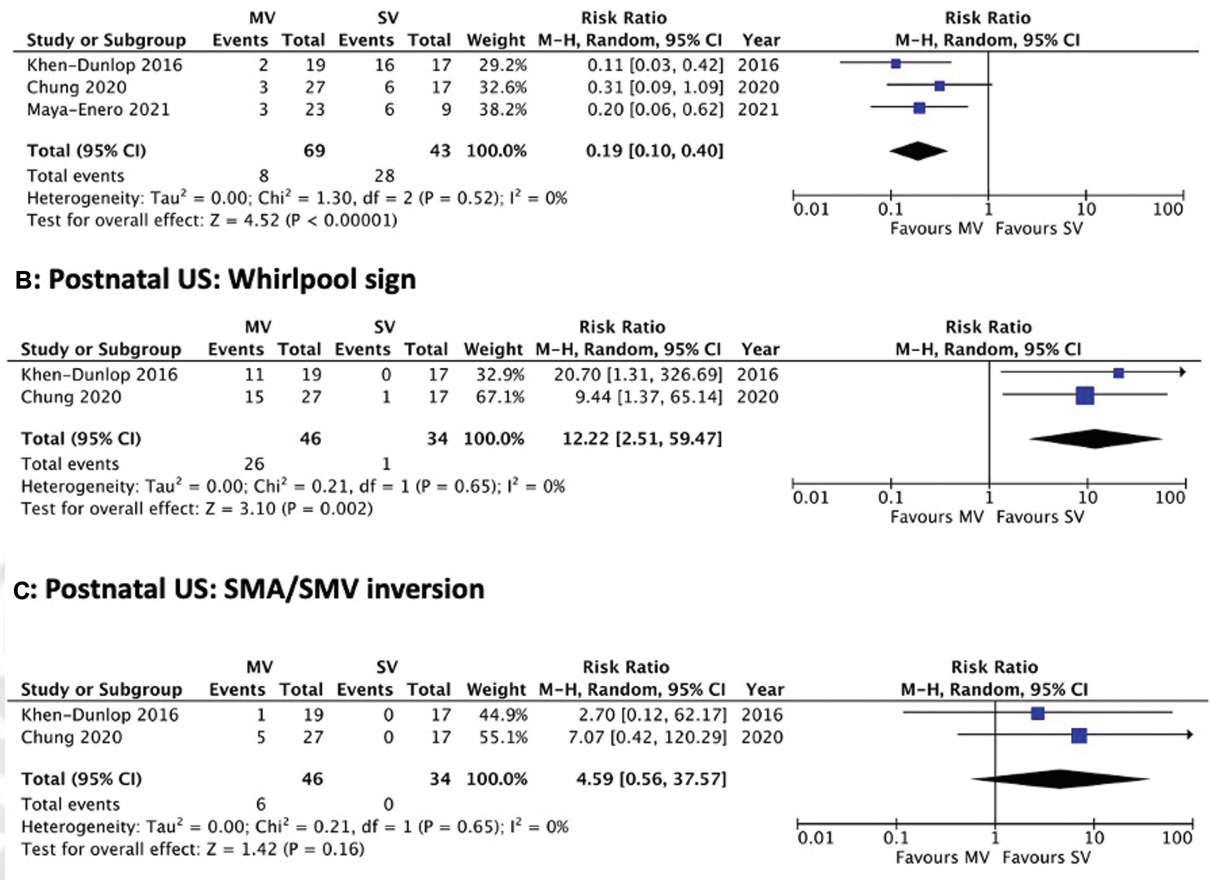

Fig. 4 Forest plot comparison of diagnostic imaging between MV and SV with regards to prenatal US (**A**), presence of a whirlpool sign on postnatal US (**B**), and SMA/SMV inversion on postnatal US (**C**). MV, midgut volvulus; SMA, superior mesenteric artery; SMV, superior mesenteric vein; SV, segmental volvulus; US, ultrasound.

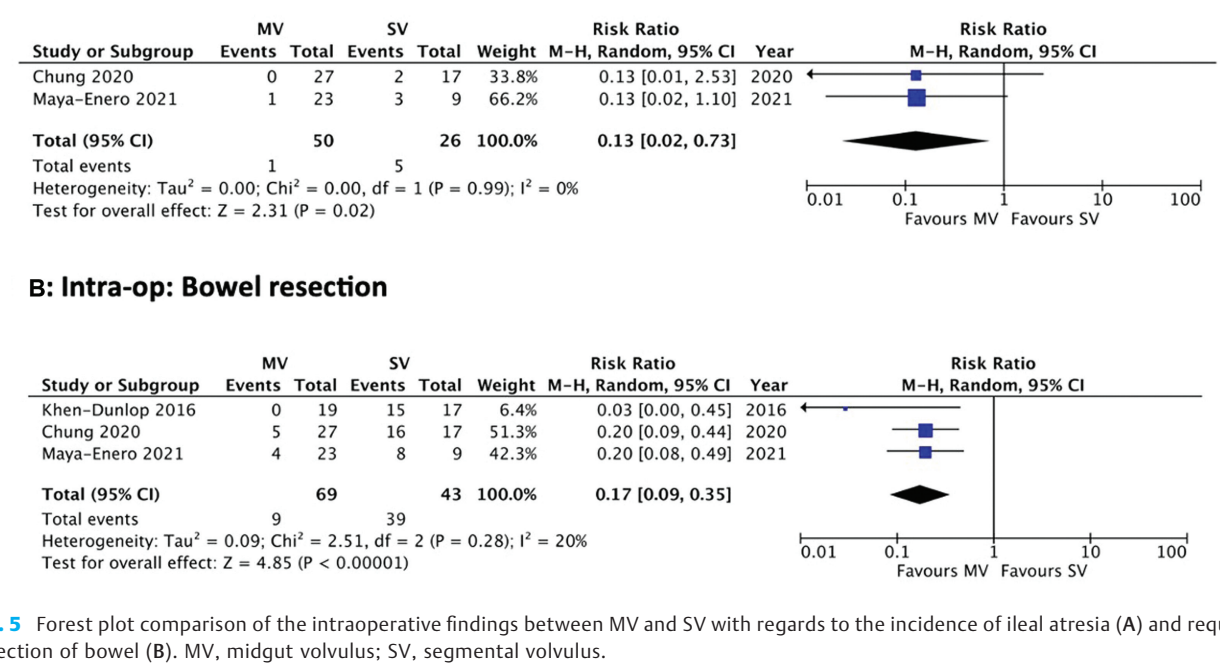

Fig. 5 Forest plot comparison of the intraoperative findings between MV and SV with regards to the incidence of ileal atresia (**A**) and required resection of bowel (**B**). MV, midgut volvulus; SV, segmental volvulus.

76.7 ± 17.9%) compared to MV (22/69 pts, 31.9 ± 25.0%; 95% CI: 0.36 [0.14, 0.96], $I^2 = 81\%$; $p < 0.05$; ►Fig. 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], $I^2 = 0\%$; $p < 0.00001$; ►Fig. 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], $I^2 = 0\%$; $p < 0.01$; ►Fig. 4B), although an inversion of SMA/SMV relationship was similarly reported in both conditions (6/46 pts, 13.0 ± 5.2% in MV vs. 0/34 pts, 0% in SV; 95% CI: 4.59 [0.56, 37.57], $I^2 = 0\%$; $p =$ ns; ►Fig. 4C).

Intraoperatively, 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], $I^2 = 0\%$; $p < 0.05$; ►Fig. 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], $I^2 = 20\%$; $p < 0.00001$; ►Fig. 5B).

No differences were found between MV and SV with regards to overall postoperative complications (6/46 pts, 13.0 ± 3.0% vs. 7/34, 20.6 ± 4.2%, respectively; 95% CI: 0.62 [0.23, 1.67], $I^2 = 0\%$; $p =$ ns; ►Fig. 6A) and postoperative obstructive ileus (3/46 pts, 5.5 ± 1.5% vs. 4/34 pts, 11.8 ± 0%, respectively; 95% CI: 0.55 [0.13, 2.35], $I^2 = 0\%$; $p =$ ns; ►Fig. 6B). The mortality rate was similar between the two groups (MV: 8/69 pts, 11.6 ± 11.6% vs. SV: 1/43 pts, 2.3 ± 7.8%; 95% CI: 2.37 [0.55, 10.29], $I^2 = 0\%$; $p =$ ns; ►Fig. 6C). The incidence of short bowel syndrome was reported only in one paper (Chung et al[4]), with no

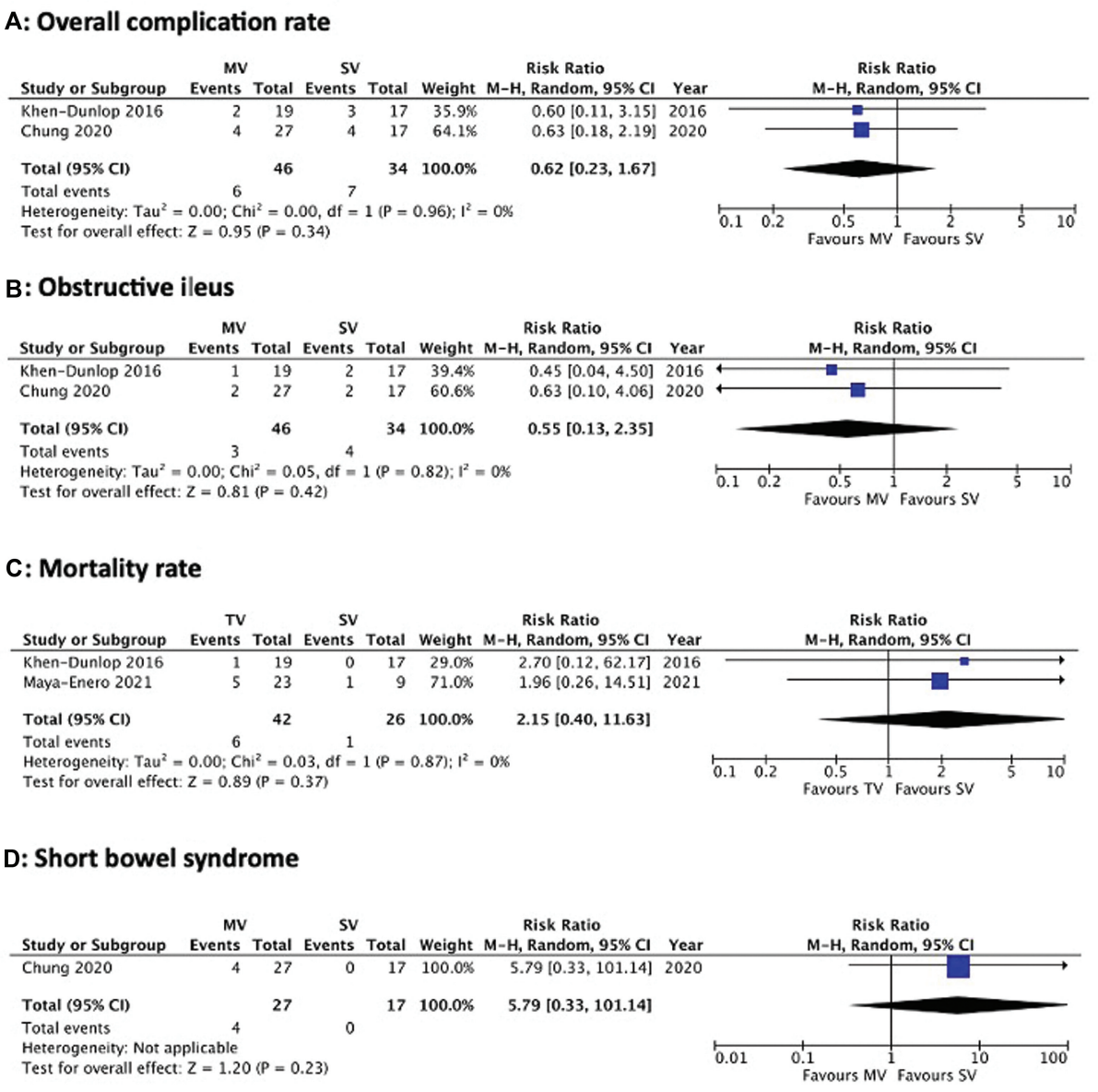

Fig. 6 Forest plot comparison of the postoperative outcomes between MV and SV with regards to overall postoperative complications (**A**), obstructive ileus (**B**), mortality rate (**C**), and the incidence of short bowel syndrome (**D**). MV, midgut volvulus; SV, segmental volvulus.

Table 2 Risk of bias assessment for individual studies using methodological index for nonrandomized studies (MINORS)[10]

Item	Chung et al[4]	Khen-Dunlop et al[13]	Maya-Enero et al[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**

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

significant differences between MV (4/27 pts, 15%) and SV (none; p = ns, ►**Fig. 6D**).

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, intraoperative 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 GA 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, who 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 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 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

Table 3 GRADE evidence profile for the present meta-analysis[11]

							No. of patients		Effect		Quality
No. of studies	Study design	Risk of bias	Inconsistency	Indirectness	Imprecision	Other considerations	Cases	Controls	Relative (95% CI)	Absolute (95% CI)	
Gestational age in MV vs. 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 vs. 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 vs. 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 1,000 (from 594 to 396 fewer)	⊗⊗OO LOW
Age at presentation in MV vs. 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 vs. 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 1,000 (from 243 fewer to 6,766 more)	⊗OOO VERY LOW
Abdominal distension in MV vs. 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 1,000 (from 602 to 28 fewer)	⊗OOO VERY LOW
Whirlpool sign at abdominal USS in MV vs. 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 1,000 (from 72 to 2793 more)	⊗⊗OO LOW
SMA inversion at abdominal USS in MV vs. 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 1,000 (from 16 fewer to 1324 more)	⊗OOO VERY LOW
Ileal atresia in MV vs. 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 1,000 (from 16 fewer to 749 more)	⊗OOO VERY LOW

Table 3 (*Continued*)

							No. of patients		Effect		Quality
No. of studies	Study design	Risk of bias	Inconsistency	Indirectness	Imprecision	Other considerations	Cases	Controls	Relative (95% CI)	Absolute (95% CI)	
Resection of bowel in MV vs. 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 1,000 (from 852 to 608 fewer)	⊗⊗OO LOW
Post-operative complications in MV vs. 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 1,000 (from 154 fewer to 134 more)	⊗OOO VERY LOW
Post-operative obstructive ileus in MV vs. 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 1,000 (from 102 fewer to 159 more)	⊗OOO VERY LOW
Prevalence of deaths in MV vs. 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 1,000 (from 55 fewer to 971 more)	⊗OOO VERY LOW
Short bowel syndrome in MV vs. 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 1,000 (from 21 fewer to 3094 more)	⊗OOO VERY LOW
Abnormal UGI contrast study in MV vs. 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 1,000 (from 44 fewer to 2086 more)	⊗OOO VERY LOW

Abbreviations: MD, mean difference; MV, midgut volvulus; RR, risk ratio; SMA, superior mesenteric artery; SV, segmental volvulus; UGI, upper gastrointestinal; USS, ultrasound scan.
Note: 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.
[a]Bias due to possible confounding.
[b]Optimal information size not met.

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 de-rotation 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.

In terms of postoperative complications, we did not find differences between the patients with SV and MV. This is in contrary to Khen-Dunlop et al, who reported on a higher incidence of postoperative 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 postoperative 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.

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 three studies included were retrospective observational studies.[4,13,14] As expected, a blinded evaluation of objective endpoints was not possible. Moreover, none of the studies have reported with regards to the loss to follow-up and there was a broad lack of data with regards to 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 to some preoperative data (i.e., GA and prenatal ultrasonography), the whirlpool sign at abdominal US scans among the preoperative imaging studies, and the incidence of resection of the bowel among the two groups (►**Table 3**). Since the data were obtained from a small number of studies, their considerable heterogeneity could generate possible bias. Nonetheless, when independently assessed by two authors (G.L. and M.E.M.) 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**).

Conclusion

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.

Author Contributions Statement

M.C., G.L., E.Z.-R. contributed to conception/design, analysis and interpretation, participated in drafting. M.C., M.E.M. contributed to data acquisition. G.L., M.E.M. contributed to quality assessment. G.L., A.Z., E.G., E.Z.-R. participated in revision, and gave final approval.

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Conflict of Interest

None declared.

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