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LITERATURE REVIEW

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Scoliosis in myelomeningocele: epidemiology, management, and functional outcome

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OBJECTIVE The authors aimed to provide an updated and consolidated report on the epidemiology, management, and functional outcome of cases of myelomeningocele (MMC) in patients with scoliosis.

METHODS A comprehensive literature search was performed using MEDLINE, Embase, Google Scholar, and the Cochrane Database of Systematic Reviews on cases of MMC in patients with scoliosis between 1980 and 2016. The initial search yielded 670 reports. After removing duplicates and applying inclusion criteria, we included 32 full-text original articles in this study.

RESULTS Pooled statistical analysis of the included articles revealed the prevalence of scoliosis in MMC patients to be 53% (95% CI 0.42–0.64). Slightly more females (56%) are affected with both MMC and scoliosis than males. Motor level appears to be a significant predictor of prevalence, but not severity, of scoliosis in MMC patients. Treatment options for these patients include tethered cord release (TCR) and fusion surgeries. Curvature improvement and stabilization after TCR may be limited to patients with milder (< 50°) curves. Meanwhile, more aggressive fusion procedures such as a combined anterior-posterior approach may result in more favorable long-term scoliosis correction, albeit with greater complication rates. Quality of life metrics including ambulatory status and sitting stability are influenced by motor level of the lesion as well as the degree of the scoliosis curvature.

CONCLUSIONS Scoliosis is among the most common and challenging comorbidities from which patients with MMC suffer. Although important epidemiological and management trends are evident, larger, prospective studies are needed to discover ways to more accurately counsel and more optimally treat these patients.

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KEY WORDS epidemiology; myelomeningocele; neural tube defect; scoliosis; spina bifida; treatment; spine

Myelomeningocele (MMC), characterized as a protruding sac of both the meninges and spinal cord, is the most frequent manifestation of neural tube defects, affecting nearly 1500 newborns in the United States annually and 0.2–6.5 newborns from every 1000 births globally.[4,13,27,30] One of the most common and most severe skeletal complications of MMC is scoliosis,[45] which can result in respiratory compromise, decreased mobility, skin breakdown, sitting and ambulation issues, and worsening of neurological symptoms.[11,42] Thus, an understanding of the effects of scoliosis on patients with MMC is particularly important.

A thorough review on the intersection between MMC and scoliosis has not been recently conducted. Therefore, we aimed to provide an updated and consolidated report on the epidemiological observations and clinical outcomes on this topic. In this study, we aggregated data addressing the following questions, which were formulated both from clinical curiosity and literature shortcomings: 1) What is the prevalence of scoliosis in MMC patients? 2) Does a sex predilection exist for scoliosis in MMC? 3) How does motor level correlate with presence and degree of scoliosis? 4) What scoliosis treatment options are most commonly employed for patients with MMC, and what outcomes can be expected? 5) What is the quality of life in MMC patients with scoliosis?

Methods

A comprehensive literature search was performed using MEDLINE, Embase, Google Scholar, and the Cochrane Database of Systematic Reviews in May of 2016.

ABBREVIATIONS MeSH = Medical Subject Headings; MMC = myelomeningocele; PRISMA = Preferred Reporting Items for Systematic Reviews and Meta-Analyses; TCR = tethered cord release.

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To achieve an encompassing and focused literature search, relevant Medical Subject Headings (MeSH) terms were divided into 2 groups: Group 1 (terms associated with MMC) and Group 2 (terms associated with scoliosis). Group 1 consisted of the following terms: "myelomeningocele," "meningomyelocele," "spina bifida," "spina bifida cystica," "tethered spinal cord," and "neural tube defect." Group 2 consisted of the following: "scoliosis" and "neuromuscular scoliosis." Relevant combinations of the MeSH terms in the groups were searched. The articles from these initial searches were screened so that we included only original English-language full-length articles published between 1980 and 2016. Review papers and any original publications with a sample of fewer than 25 patients were excluded. The articles were further screened to ensure that they provided data relevant to one or more of the aforementioned questions. Reports were excluded if they did not provide data relevant to the questions or if there was no clear delineation of patients with both MMC and scoliosis. The remaining articles were included in this systematic review. Within these included articles, patients were excluded if they had a form of spina bifida other than MMC. Furthermore, patients were excluded if they did not have scoliosis, even if they had other deformity (e.g. kyphosis). No other exclusion criteria were applied. The literature search was conducted according to Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines (Fig. 1).[22]

Two authors (N.M. and M.R.M.) independently reviewed the articles, extracted data, and discussed any disagreements before including or excluding any studies. Disputes were resolved by an arbiter (M.C.D.) before data were included in this manuscript. Level of evidence for each article was assigned according to Oxford Centre for Evidence-Based Medicine 2011 guidelines (http://www.cebm.net/ocebm-levels-of-evidence/). Significant findings from the review are shown in Figs. 2 and 3 and Tables 1–3.

For data analysis, no strict definition of scoliosis was applied while evaluating the patients in the articles owing to a lack of information about each patient's degree of curvature. As a result, we accepted the articles' definition of scoliosis. Many of the articles determined scoliosis based on the Cobb angle identified on radiographs. However, some of the reports did not clearly define their methods of determining scoliosis. There were no instances of reverse lordosis being considered as scoliosis.

Data analysis was performed using *Meta-Essentials* (http://www.erim.eur.nl/research-support/meta-essentials/). Standard error of prevalence was calculated using the following formula: $(p \times (1 - p)/n) ^ 0.5$, where p is the prevalence and n is the study sample size. A random effects model with 95% CIs was used to generate the forest plots.

Results

Article Selection and Characteristics

The initial literature search with different combinations of MeSH terms, as described in the *Methods* section, yielded a total of 670 reports. Once duplicate articles were removed and exclusion criteria applied, 32 reports

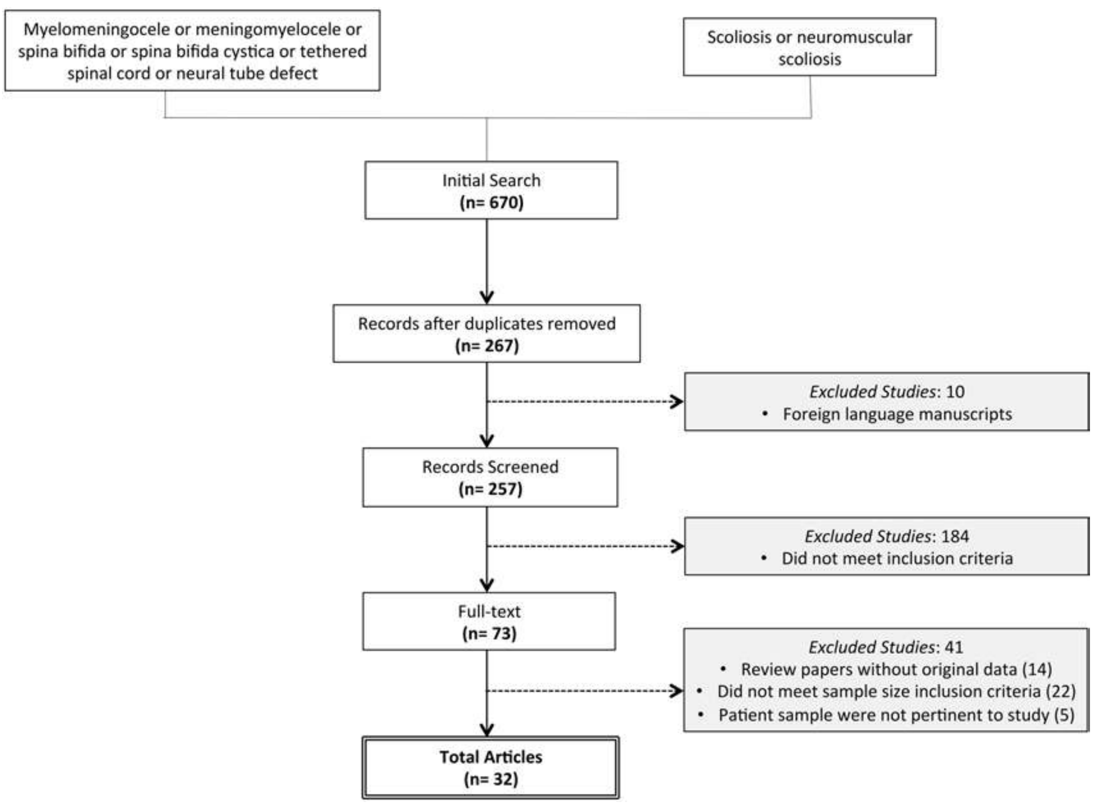

FIG. 1. PRISMA flow diagram for this study.

remained and were included in this study (Fig. 1). Of these 32 articles, 7 were published in the 1980s, 11 in the 1990s, 5 in the 2000s, and 9 from 2010 until the present day. All but 3 of these articles were retrospective in design. The sample size of patients with both MMC and scoliosis in source articles ranged from 29 to 343 patients.

Prevalence and Demographics of Scoliosis in MMC Patients

A total of 15 articles provided data addressing the prevalence of scoliosis in patients with MMC (Fig. 2). Based on these studies, the percentage of MMC patients with scoliosis ranged widely from 23% to 88%.[9,21,34,38] Pooled statistical analysis of the included studies revealed a prevalence rate of 52.8% (95% CI 42%–64%). Díaz Llopis et al. and Reigel et al. conducted the 2 largest studies in terms of sample size with 1500 and 262 patients, respectively. Díaz Llopis et al. reported a prevalence rate of 23% in their population while Reigel et al. found a prevalence rate of 25%.[9,31]

Details about the sex ratio of patients with MMC and scoliosis were discussed in 17 studies (Fig. 3). In all but 3 studies, there were more females than males with MMC-associated scoliosis. Two studies found that males and females were affected equally, and only 1 article described males more commonly affected with both MMC and scoliosis at a ratio of 1.2:1.[33,24,41] The largest sex disparity ratio of 2.2 females to 1 male was described in a 54-patient study by Banta in 1990.[2] Pooled statistical analysis of the included studies revealed that 56% (95% CI 52%–59%) of the MMC patients with scoliosis were females.

Effects of Motor Level on Scoliosis in MMC Patients

One or more aspects of this topic were discussed in reports of 9 studies (Table 1). Five of these studies examined the percentage of MMC patients with scoliosis based on motor level, the lowest level of functionality in the spinal cord.[5,23,31,35,42] Trivedi et al. found that 93% of MMC patients with thoracic lesions had scoliosis, whereas only 72% of patients with upper lumbar lesions (L1–3), 43% of patients with lower lumbar lesions (L4–5), and 8% of patients with sacral lesions did.[42] Similarly, the remaining 4 studies also reported that lesions at the thoracic level resulted in the highest percentage of MMC patients with scoliosis. Most of these studies also found that sacral level lesions resulted in the least prevalence of scoliosis in MMC patients. However, a 71-patient prospective study by Bowman et al. reported in 2001 that upper lumbar lesions resulted in the lowest prevalence of scoliosis.[5]

One article published findings about the effects of motor level on the degree of scoliosis.[1] This study found no association between motor level/MMC lesion level and the scoliosis curve. In addition, the authors reported the average preoperative scoliosis curves for thoracic, upper lumbar, and lower lumbar lesions to be 74°, 75°, and 74°, respectively. After surgery, these curves were reduced to 41°, 36°, and 40°, respectively.[1] One study in 2009 by Bowman et al. found that a majority (54%) of the patients who experienced curve progression even after tethered cord release (TCR) had a thoracic motor level.[6]

In 1981, Kahanovitz and Duncan reported data regarding the effects of motor level on sitting stability and ambulation. They found that 55% of MMC patients with scoliosis who were unbalanced sitters had a motor level of T-12. Furthermore, 100% of MMC and scoliosis patients with L-5 or S-1 motor levels were ambulatory. However, only 35% of the patients with a motor level above L-5 were ambulatory.[14] Future research looking at MMC populations without scoliosis needs to be performed to establish an accurate comparison of motor level and ambulatory status. In addition, Samuelsson and Skoog found that an increasing scoliosis curve resulted in worse ambulatory outcomes.[35]

Management of MMC Patients With Scoliosis

Eighteen articles discussed one or more aspects of this topic, most of which addressed various surgical treatment

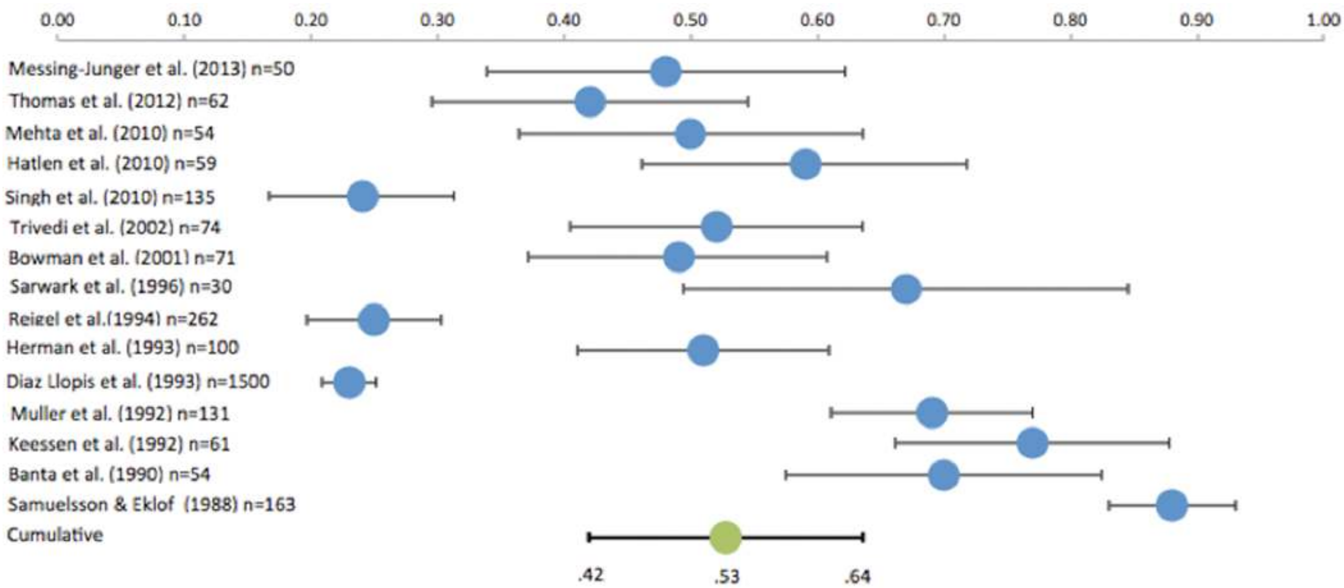

FIG. 2. Forest plot showing that 53% of patients with MMC also suffer from scoliosis. Figure is available in color online only.

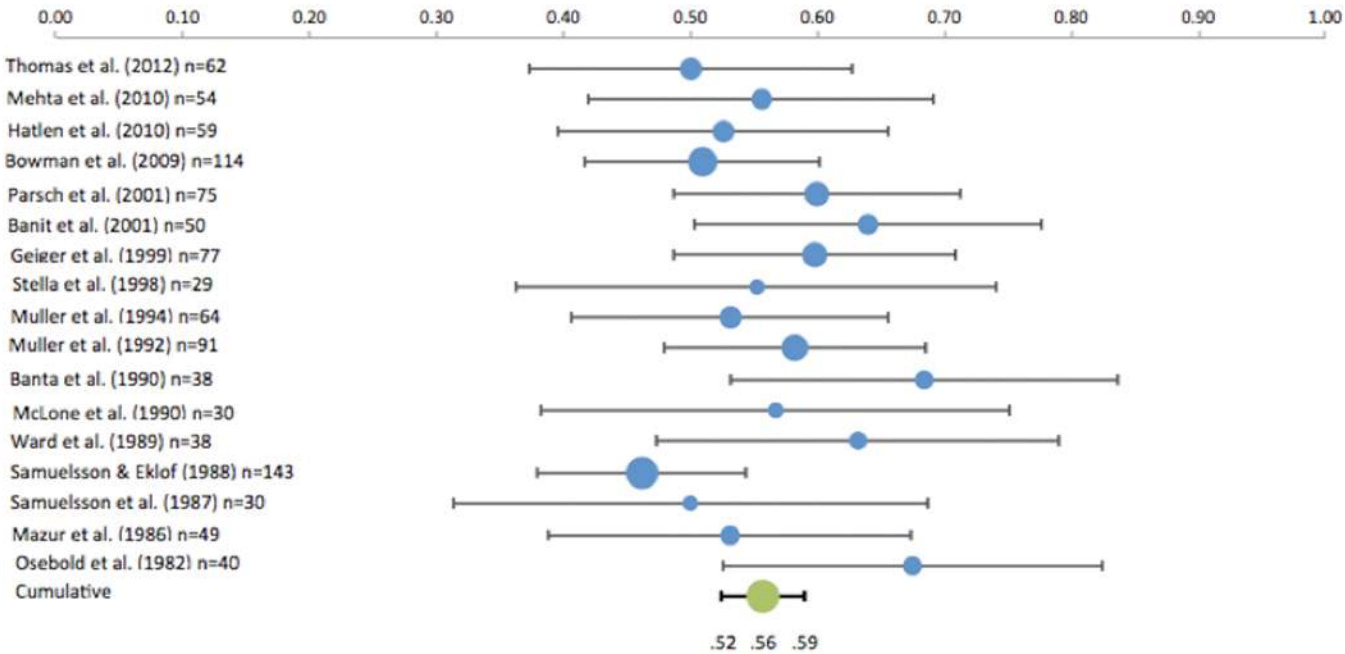

FIG. 3. Forest plot showing percentage of MMC patients with scoliosis who were female. More female than male patients tend to have both MMC and scoliosis. Figure is available in color online only.

options for these patients. Generally, the surgical approaches can be divided into one of two entities: TCR and instrumentation/fusion for scoliosis correction.

Seven studies examined the effects of TCR on MMC patients with scoliosis. Several reports found modest to moderate improvement in various symptoms after TCR.[5,6,36] Sarwark and colleagues noted that 15% of their patients had improvement in scoliosis, while 60% had stable curvature and 25% had curve progression during the 1-year follow-up after TCR.[36] In a retrospective study

TABLE 1. Effects of motor level on scoliosis in MMC patients

Authors & Year	Study Design	No. of Patients	Findings	Level of Evidence
Bowman et al., 2009	Prospect	114	54% of the patients w/ progressive scoliosis after TCR had function at the thoracic level.	3b
Trivedi et al., 2002	Retro	74	93% of patients w/ thoracic lesions had scoliosis. 72% w/ lumbar lesions had scoliosis. 43% of patients w/ lower lumbar lesions had scoliosis. 8% of patients w/ sacral lesions had scoliosis.	4
Banit et al., 2001	Retro	50	Mean scoliotic curve for thoracic, upper lumbar (L1–3), & lower lumbar (L4–5) lesions were 74°, 75°, & 74°, respectively, preop & 41°, 36°, & 40° postop.	4
Bowman et al., 2001	Prospect	71	51% of scoliosis patients had a thoracic motor level, 6% an upper lumbar level, 29% a lower lumbar level, & 11% a sacral level.	3b
Müller et al., 1994	Retro	64	Level of lesion was not significantly associated w/ change in scoliosis angle.	4
Reigel et al., 1994	Retro	262	77% of patients w/ thoracic lesions had scoliosis. 40% w/ lumbar lesions had scoliosis. 13% w/ lower lumbar lesions had scoliosis. 3% w/ sacral lesions had scoliosis.	4
Müller et al., 1992	Retro	131	94% of patients w/ thoracic lesions had scoliosis. 75% w/ lumbar lesions had scoliosis. 50% w/ lower lumbar lesions had scoliosis. 20% w/ sacral lesions had scoliosis. Significant correlation existed btwn functional level & scoliosis prevalence.	4
Samuelsson & Skoog, 1988	Retro	163	100% of patients w/ thoracic lesions had scoliosis. 95% w/ L1–2 lesions had scoliosis. 90% w/ L-3 lesions had scoliosis. 85% w/ L-4 lesion had scoliosis. 88% w/ L-5 lesion had scoliosis. 72% w/ sacral lesions had scoliosis. Increasing scoliosis curve resulted in worse ambulatory outcomes.	4
Kahanovitz et al., 1981	Retro	39	55% of the patients who were unbalanced sitters had a T-12 motor level. 100% of L-5 & S-1 patients were ambulators. 35% of patients w/ neurological levels above L-5 were ambulators.	4

Prospect = prospective; retro = retrospective.

TABLE 2. Surgical treatment outcomes and complications in MMC patients with scoliosis

Authors & Year	Study Design	No. of Patients	Treatment	Findings	Level of Evidence
Mehta et al., 2010	Retro	54	Surgery (TCR)	Retethering occurred in 41% of scoliosis patients but only 22% of patients w/o scoliosis (p = 0.149); furthermore, retethering occurred earlier than in patients w/o scoliosis.	4
Hatlen et al., 2010	Retro	59	Surgery (spinal fusion)	Complications, arising in more than half the cases, included infection & respiratory failure. 12 patients required a 2nd op due to infection. No correlation btwn MMC lesion & complication rate.	4
Cahill et al., 2010	Retro	84	Surgery	18% experienced postop infection.	4
Bowman et al., 2009	Prospect	114	Surgery (TCR)	Scoliosis progressed after TCR in 52%. 70% of patients showed improved LE muscle strength. Gait was also improved. Spasticity improved in 67%. 64% showed improvement in bladder function. Complications of the op included CSF leak (6%), infection (10%), & neurological deterioration (4%).	3b
Parsch et al., 2001	Retro	54	Surgery (Group I: PF/instrumentation; Group II: AF/no instrumentation combined w/ PF/instrumentation; Group III: CF/instrumentation)	Cobb angle for Group I changed from a mean of 79° preop to 35° postop to 45° at 3.5-yr follow-up. Cobb angle for Group II changed from mean of 97° preop to 52° postop to 61° at 3.4-yr follow-up. Cobb angle for Group III changed from mean of 92° preoperative to 38° postop to 40° at 2.5-yr follow-up. Overall hardware complication rate was 30%; these rates were not significantly different btwn groups. Patients w/ thoracic lesion had a greater loss of correction than patients w/ lumbar lesion (p < 0.06).	4
Banit et al., 2001	Retro	50	Surgery (PF)	Cobb angle changed from a mean of 75° preop to 39° postop to 45° at follow-up. 48% had complications (infection, pseudomeningocele, & UTI).	4
Bowman et al., 2001	Prospect	71	Surgery (TCR)	10 patients developed symptomatic tethering involving scoliosis; 50% of these patients had improvement in symptoms & 50% became stable. 43% of these patients eventually required spinal fusion ops, the majority of which were at the thoracic level.	3b
Geiger et al., 1999	Prospect	77	Surgery (Group I: anterior correction & Harrington rods; Group II: posterior instrumentation; Group III: anterior release & posterior instrumentation; Group IV: anterior & posterior instrumentation)	Cobb angle: Group I: preop 84.5°, postop 48°; Group II: preop 79.5°, postop 38°; Group III: preop 103.2°, postop 49.4°; Group IV: preop 94.8°, postop 38.4°. 52.9% of patients had complications including hardware problems, shunt failure, infection, & anesthetic concerns. Hardware problems occurred most often w/ Harrington rods rather than CD instrumentation. Hardware problems were associated w/ loss of correction (loss of 21.7° vs loss of 6.5°). No difference in complication rate among groups. Group IV had the lowest loss in correction (p < 0.05).	3b
Stella et al., 1998	Retro	29	Surgery (7 PF, 3 AF, 19 CF)	Posterior fusion resulted in scoliosis curve correction of 22% in thoracic & thoracolumbar curves & 32% in lumbar curves. Anterior fusion resulted in scoliosis curve correction of 43% in lumbar curves. Combined fusion resulted in scoliosis curve correction of 55% in thoracic & thoracolumbar curves & 61% in lumbar curves. Overall, ops resulted in 47% mean correction in thoracic & thoracolumbar curves & 50% mean correction in lumbar curves. Complications of ops included infection, bed sores, & pneumothorax.	4
Sarwark et al., 1996	Retro	30	Surgery (TCR)	First-yr follow-up results showed 15% of patients w/ MMC & scoliosis improved, 60% were stable, & 25% had curve progression.	4
Reigel et al., 1994	Retro	262	Surgery (TCR)	25% of patients developed significant scoliosis (>30°) after TCR. Highest prevalence (77%) of scoliosis occurred in thoracic lesions. Only for thoracic group did progression of scoliosis continue after TCR.	4
Herman et al., 1993	Retro	100	Surgery (TCR)	Correction in the scoliosis curve by at least 7° occurred in 51% of the patients. 10% had progression of the curve at 1-yr follow-up. 63% improved or were stable in the follow-up exams after 1 year. CSF leak & infection were the most common complications.	4

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TABLE 2. Surgical treatment outcomes and complications in MMC patients with scoliosis

Authors & Year	Study Design	No. of Patients	Treatment	Findings	Level of Evidence
Keessen et al., 1992	Retro	48	Surgery (PF n = 15, AF n = 8, circumferential fusion n = 25)	AF: preop 88.8°, postop 37.1; PF: preop 71.2°, postop 45.7°; circumferential: preop 84.6°, postop 31.1°. Correction was 45% for thoracolumbar, 20% for thoracic, & 30% for lumbar. Complications included infection, instrumentation related, skin issues, & neurological.	4
Banta, 1990	Retro	54	Surgery (CF)	Pre-op: 73 Post-op: 34 4 patients needed revision surgery. 47% of patients had complications w/ the procedures. The 2 most common were infection & pseudarthrosis.	4
McLone et al., 1990	Retro	30	Surgery (TCR)	At 1-yr follow-up, 83% of patients w/ curves >50° progressed & required fusion. 96% w/ curves <50° improver or were stable. However, at last follow-up, 37% progressed, 21% improved, & 42% remained the same.	4
Ward et al., 1989	Retro	38	Surgery (Group A: 1-stage AF or PF w/ or w/o instrumentation; Group B: anterior interbody fusion w/ posterior fusion using Harrington rod; Group C: anterior interbody w/ Dwyer & posterior fusion using Harrington; Group D: anterior interbody fusion w/ posterior fusion using Luque; Group E: anterior interbody w/ Dwyer & posterior fusion w/ Luque)	Initial fusion success rates for the 5 groups were 50%, 100%, 92%, 83%, & 100%, respectively. 16% of patients needed to have a 2nd procedure. Group A: preop 69, postop 64; Group B: preop 81, postop 35; Group C: preop 77, postop 43; Group D: preop 57, postop 28; Group E: preop 62, postop 18. Group B & E improvements were significantly better than A. 58% of patients had complications.	4
Mazur et al., 1986	Retro	49	Surgery (Group A: PF; Group B: AF; Group C: CF)	Cobb angle for Group A changed from a mean of 78° preop to 41° postop to 53° at follow-up. Cobb angle for Group B changed from a mean of 61° preop to 27° postop to 32° at follow-up. Cobb angle for Group C changed from a mean of 78° preop to 33° postop to 45° at follow-up. No significant difference among groups in terms of postop function. Infections only occurred during posterior ops (Group A 7%; Group C 11%). Pseudarthrosis rate was 33%, 29%, & 11%, respectively.	4
Osebold et al., 1982	Retro	40	Surgery (Group A: PF; Group B: PF & instrumentation; Group C: PF & instrumentation along w/ AF; Group D: PF & instrumentation along w/ AF & instrumentation)	Average correction at follow-up for scoliosis was 6°, 12°, 52°, & 45°, respectively, in Groups A–D. Pseudarthrosis occurred in 67%, 46%, 100%, & 23%, & infection in 23%, 33%, 29%, & 8%.	4

AF = anterior fusion; CD = Cotrel-Dubousset; CF = combined fusion; LE = lower extremity; PF = posterior fusion; UTI = urinary tract infection.

in 1993, Herman et al. found that there was a correction of the scoliosis curve by at least 7° in 51% of the patients postoperatively after TCR. This study also reported that 63% of the patients with at least a 1-year follow-up improved or demonstrated curve stability.[12] McLone and colleagues examined curve reduction after surgery, based on the degree of scoliosis. Almost all patients (96%) with a curve less than 50° had curve improvement during the 12-month post-TCR follow-up period. However, they did note that curves in only 63% of the patients with initial curves less than 50° were considered to be improved or stable at the last follow-up visit several years after the procedure. Meanwhile curves in 83% of the patients with a curve greater than 50° progressed and required spinal fusion surgery.[17] Retethering of the spinal cord occurred in 41% of patients with scoliosis as opposed to only 22% of

TABLE 3. Effects of scoliosis on quality of life for myelomeningocele patients

Authors & Year	Study Design	No. of Patients	Findings	Level of Evidence
Thomas et al., 2012	Retro	62	Mean Hoffer score was 2.6. 16.4% scored 1, 8.2% scored 2, & 75.4% scored 3.	4
Patel et al., 2011	Retro	32	80% of patients used wheelchairs for all mobility. 20% were household ambulators & used wheelchairs in the community. As magnitude of curve increased, there was an increase in the percentage of seat w/ a pressure btwn 38–70 mm Hg.	4
Bowman et al., 2009	Prospect	114	78% showed improvement in gait, 19% remained stable, & 3% got worse. All showed postop improvement in pain.	3b
Trivedi et al., 2002	Retro	74	Significant association btwn scoliosis & nonambulatory status.	4
Stella et al., 1998	Prospect	29	4 patients w/o orthoses or crutches; 2 w/ orthoses & w/o crutches; 9 w/ both. 14 dependent on wheelchairs.	3b
Sarwark et al., 1996	Retro	30	Sitting balance was improved after surgery. All patients w/ preop back pain had pain resolution.	4
Mazur et al., 1986	Retro	49	Ability to ambulate deteriorated in 27% of Group A, 57% of Group B, & 67% of Group C. 73% of Group A, 43% of Group C, & 33% of Group C remained the same. 67% of Group A & 70% of Group C showed improved sitting ability. Group C showed most improvement w/ sitting. In Groups A & C, the majority of patients' back pain was unchanged. In Group B, 43% improved, 43% remained the same.	4
Osebold et al., 1982	Retro	40	57% of interviewed patients said major benefit was improved sitting stability.	4
Kahanovitz & Duncan, 1981	Retro	39	At final follow-up, 20 patients were nonambulators, 9 were balanced sitters, & 11 were unbalanced sitters. The remaining 19 patients were ambulatory (5 household, 14 community).	4

patients without scoliosis in a study performed by Mehta et al. However, this difference was not significant ($p = 0.149$) In addition, the retethering occurred significantly earlier in patients with scoliosis than it did in patients without scoliosis ($p = 0.042$).[19]

Spinal fusion for MMC patients with scoliosis was discussed in 11 studies. Several of these reports included comparisons of different spinal surgical approaches with one another.[10,15,28,38] Parsch et al. examined the Cobb angle change in patients who underwent posterior instrumentation/fusion (Group 1), anterior fusion with posterior instrumentation/fusion (Group 2), and combined instrumentation/fusion (Group 3). The Cobb angle for patients in Group 1 changed from an average of 79° preoperatively to 35° postoperatively to 45° at follow-up. In Group 2, the Cobb angle changed from 97° preoperatively to 52° postoperatively to 63° at follow-up. In Group, 3 the Cobb angle changed from 92° preoperatively to 38° postoperatively to 40° at last follow-up.[28] Similar to these findings, Mazur et al. and Geiger et al. also published series in which they found a greater correction of the scoliosis curve when combined fusion was used as the surgical intervention.[10,16]

In terms of complication rate after posterior fusion surgery, one study found that 48% of patients had complications while another reported that 53% of their patients suffered complications such as hardware problems, shunt failures, infections, and pseudomeningoceles.[1,10] In a study by Banit et al. with a 48% complication rate, no patient's treatment resulted in cardiopulmonary complications or death. However, 63% of the patients with complications required an additional surgery.[1] Geiger and colleagues reported a complication rate of 53%, but they found that there was no difference in the complication rates among the different surgical techniques. The 4 main groups of complications included hardware problems, shunt failures, infection, and anesthetic complications.[10] Cahill and colleagues found that an alarming 18% of their population had infection after surgery.[7] In addition, one study found no correlation between MMC level and complication rate, whereas another published data suggesting that postoperative infection only occurred in patients who had undergone posterior surgical approaches.[11,16]

Quality of Life in MMC Patients With Scoliosis

Quality of life in terms of ambulatory status and sitting stability was discussed in 9 articles (Table 3). Thomas et al. reported a mean Hoffer score of 2.6 in a sample of 62 MMC and scoliosis patients, with 16.4% of the patients scoring a 1 (ambulatory with prosthetic device), 8.2% scoring a 2 (partially ambulatory with prosthetic device), and 75.4% scoring a 3 (wheelchair bound). The median motor level was L-1, with a range from T-4 to S-1.[41] In the cohort of 32 patients that Patel et al. studied, 80% of the patients used wheelchairs for all mobility, while 20% of the patients were ambulatory in the household but used wheelchairs in the community.[29] As would be expected, one study found that nonambulatory patients were more likely to develop scoliosis ($p < 0.001$).[42] This suggests that nonambulatory status plays a causative role in the development of scoliosis, not vice versa. However, Kahanovitz and Duncan reported a much more balanced ratio of patients who were nonambulatory (51%) and patients who were ambulatory (49%).[14]

Sitting stability is a valid quality of life metric in children with neuromuscular pathology as abnormal balance can cause discomfort and skin breakdown.[3] In patients with MMC-related scoliosis, Patel et al. noted that an increase in the magnitude of the scoliosis curve resulted in an increased percentage of the contact area between the buttocks and the wheelchair with a pressure between 38 and 70 mm Hg.[29] Three studies reported improvement in sitting stability for their patients who underwent a surgical procedure.[16,26,36]

Discussion

In the present study, we screened a total of 670 reports and ultimately included 32 full-length articles that discuss scoliosis in MMC patients. Although the prevalence of MMC in the population has declined since the addition of folate into a regular diet, a significant population still suffers from MMC and its severe bony complication, scoliosis.[13] A thorough understanding of this topic, therefore, is essential when counseling and managing these patients. To our knowledge this study is the most comprehensive and up-to-date systematic review examining the most important clinical questions surrounding MMC patients suffering from scoliosis. We have sought to address the epidemiology and management outcomes in these patients in a concise and straightforward report.

From our literature search and analysis, several significant patterns were identified and we elaborate on them below. More female MMC patients had scoliosis than male MMC patients.[1,43] Whether this increased female predilection is due to an underlying pathophysiological process or whether it is simply due to higher rates of females suffering from MMC remains uncertain. Sawin and colleagues published a report highlighting the demographics of the patients included in the National Spina Bifida Patient Registry, a multi-institution project with over 1500 patients with MMC. They reported that more females (51.6%) suffered from MMC than males.[37] Given that the higher percentage of MMC patients was female (51.6%) and the higher percentage of patients having both MMC and scoliosis was also female (56%), it seems likely that the increased prevalence of females suffering from MMC and scoliosis is a result of more females suffering from MMC than males. However, there is a slight difference in the percentages of the 2 groups. Whether this difference is significant or due to study limitation remains to be determined.

This bias toward females being more likely to have scoliosis with MMC is particularly important given that some of these females could get pregnant during their lives. A recent review on the effects of scoliosis on pregnancy in females suffering from adolescent idiopathic scoliosis found that these females tended to have higher rates of nulliparity and increased back pain prepartum (M. Dewan et al., unpublished data). Future studies evaluating the effects of scoliosis on pregnancy in MMC patients may provide useful answers in an otherwise unstudied area of the literature.

Several of the articles we reviewed indicated that the MMC lesion level/motor level was highly correlated with the prevalence of scoliosis in those patients but not with the severity of the curve.[1,5,24,24,31] Basically, the higher the lesion level, the more likely the patient is to get scoliosis. This finding could potentially be an important diagnostic tool to determine whether early treatment, or at minimum screening, for scoliosis is warranted. While this topic is still hotly debated, early detection of scoliosis has been associated with better results for the use of noninvasive brace therapy.[8,18,40] Regardless of treatment approach, MMC patients might benefit from earlier treatment of their scoliosis or more frequent monitoring based on lesion level; clinicians should be cognizant of this observation.

The data presented here show that TCR for MMC patients with scoliosis has mixed results. Several studies reported that a significant number of patients had improvement in their scoliosis after TCR, whereas a sizeable subset had scoliosis progression even after TCR.[6,12,36] However, several of the articles did present data that suggested certain populations of MMC patients with scoliosis could benefit more from TCR than others. McLone et al. found that 96% of their MMC patients with a scoliosis curve of less than 50° either improved or were stable during the 1-year follow-up period.[17] Whether 50° of scoliosis curvature is a strong predictor of TCR surgery success needs to be validated with larger studies. However, based on the available data, this factor could serve as a tool for clinicians to counsel patients. In addition, it must be noted that because these results were based on a short follow-up interval of 1 year, the true efficacy of this surgery can only be gleaned from an analysis of results over a longer follow-up period. In addition, Reigel and colleagues reported that progression of the scoliotic curve after TCR occurred solely in patients with thoracic lesions.[31] These collective findings suggest that TCR is potentially beneficial to some but not all MMC patients with scoliosis, specifically patients with smaller scoliosis and curves and lower-level lesions.

Whether untethering of the spinal cord should be performed before fusion surgery, at the same time of fusion surgery, or not at all still remains to be determined. Mehta and colleagues found that patients who were treated with concomitant TCR and scoliosis corrective surgery had less retethering, lower prevalence of wound infection, shorter operative time, and fewer hospital days than patients who had a 2-staged surgery for untethering at one stage and then scoliosis correction at another.[20] However, this study included patients with pathologies other than MMCs, including thickened filum terminale and lipomyelomeningocele. In addition, Mehta and colleagues looked primarily at patients with large curves (> 40°) and patients who had curves that were progressing. Another study showed that an additional neurosurgical procedure at the time of scoliosis correction surgery did not increase the complication rate compared with that in patients who only underwent scoliosis correction surgery.[25] These reports suggest that performing TCR and scoliosis correction simultaneously may carry benefits for patients without increasing the complication rate. However, selecting the correct patient for this combined approach is absolutely necessary to ensure the best care. Samdani et al. found that untethering prior to fusion may actually be unnecessary in patients with MMC who do not have clinical symptoms of tethered cord, even if tethering is radiologically demonstrated.[32] As a result, being asymptomatic may be a contraindication to combined TCR and scoliosis correction surgery. Since all of these studies were retrospective in nature with no controls, it is difficult to generate a concrete set of guidelines on this topic. Indeed, larger studies with a prospective design and objective end points are needed to provide a conclusive answer.

In terms of treating MMC patients with scoliosis by implanting spinal instrumentation, we found that indications for spinal fusion were only defined in some of the articles we reviewed. Specific indications were rarely defined, as the most common indication for surgery was listed as paralytic scoliosis.[1,16,25,28] In addition, Keessen and colleagues used progressive sitting imbalance, pain, and pressure sores as indications for surgery.[15] Again, larger studies that track patients for long periods both before and after treatment will help clinicians determine the optimal surgical candidates in the future. Furthermore, none of the reviewed articles reported any concrete numerical cutoffs in terms of goals for scoliosis correction. The authors of the reviewed articles were attempting to reduce the scoliosis curve as much as possible for the longest period of time.

Multiple studies have indicated that combined anterior-or-posterior fusion results in greater correction than an anterior or posterior approach alone.[10,16,26,28] Despite the increase in the correction, the combined instrumentation/fusion surgeries are usually more invasive, which seems to result in more complications. Although 2 studies indicated no significant difference in complication rates between the combined fusion group and other groups, these studies did not analyze important perioperative considerations such as length of hospitalization, blood loss, and recovery time.[10,28] Furthermore, one study reported no significant difference in function between the different surgical groups.[16] The heterogeneous nature of scoliosis demands customized treatment modality to address the level of the lesion, degree of the curve, and functional capacity of each individual patient. Realistically, answers are likely to emerge not from randomized controlled trials but rather from larger cohort studies evaluating long-term outcomes and patient-reported metrics.

Complication rates for spinal surgery in MMC patients with scoliosis were reported to be 48% by Banit et al. and 53% by Geiger et al.[1,10] These rates are higher than the average complication rate of 22% for spinal surgeries in patients with idiopathic scoliosis published by Weiss and Goodall.[44] However, variations in complication rates for idiopathic scoliosis patients ranged from 0% to 73%. The higher complication rate seen in MMC patients could be due to a number of factors including larger scoliosis curves, more comorbidities, and a lower capacity for rehabilitation after surgery.[1,10,44]

Study Limitations

It is important for readers to be cautious while interpreting conclusions from this review because of several limitations. First, the majority of the articles reviewed in this study were retrospective in nature. Thus, the level of evidence was relatively low. Furthermore, each of the articles had unique methodologies and criteria for inclusion, which could result in similarities and/or differences that might not be completely accurate. For example, the definition of scoliosis was not standardized across the articles reviewed, mainly due to missing information about the topic in the original reports. Nonstandardized disease definition may result in variations more reflective of literature disparities rather than true biological or clinical variation. As a result, prospective, controlled cohort studies with strict inclusion criteria are necessary to garner more conclusive data.

Conclusions

Scoliosis is among the most common skeletal abnormalities from which MMC patients suffer, potentially leading to respiratory comprise and significant mobility limitations. Pooled statistical analysis demonstrated a prevalence of scoliosis in MMC patients of 53%, with females being slightly more affected (56%) than males. Motor level is a significant predictor of prevalence of scoliosis in MMC patients but not of severity. Treatment options for these patients include TCR and fusion surgeries; however, not all patients benefit from these procedures. One useful marker to predict the success of TCR release might be the degree of scoliosis curvature. Furthermore, combined anterior-posterior fusion surgeries may result in better long-term outcomes despite greater perioperative complications. In the future, larger prospective studies will be important to gain conclusive evidence regarding the optimal surgical treatment of patients with MMC and scoliosis.

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Disclosures

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Conception and design: Dewan, Naftel, Wellons, Bonfield. Acquisition of data: Mummareddy, Mercier. Analysis and interpretation of data: Mummareddy, Dewan, Mercier, Bonfield. Drafting the article: Mummareddy, Dewan. Critically revising the article: Mummareddy, Dewan, Naftel, Wellons, Bonfield. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Mummareddy. Statistical analysis: Mummareddy.

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