



Review Article

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INTRODUCTION

The incidence of scoliosis is higher in neuromuscular diseases than in other etiologies. The paraspinal muscles can be flaccid, spastic, or dyskinetic, according to the etiology. The abnormal muscle architecture, muscle imbalance, and related abnormal musculoskeletal growth are the causes of neuromuscular scoliosis. Neuromuscular scoliosis has the greatest risk of progression and tends to progress even after skeletal maturity compared to idiopathic scoliosis. The treatment for neuromuscular scoliosis is based on the natural history of the underlying neu-

Clinical Issues in Indication, Correction, and Outcomes of the Surgery for Neuromuscular Scoliosis: Narrative Review in Pedicle Screw Era

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Surgical treatment for neuromuscular scoliosis has evolved with pedicle screw instrumentation and the improvement of perioperative management. We aimed to review recent papers related to posterior surgical correction with a pedicle screw system for neuromuscular scoliosis, particularly cerebral palsy and muscular dystrophy, in terms of indication, correction method, and outcomes. The most remarkable change was the posterior-only operation with all-pedicle screw fixations. With this change, operation time, blood loss, and postoperative complications decreased. Furthermore, spinal osteotomy could be performed with the pedicle screw system for severe scoliosis. S2-alar-iliac screws demonstrated favorable outcomes in terms of stability and complication, but a question remains about saving the mobile segment for sitting balance, interaction between the hip and spine, effect to the sagittal balance, and proximal junctional problem. The quality of life improvement was more definite in patients with cerebral palsy. The improvement of respiratory function in Duchenne muscular dystrophy was not certain, although an increase of spinal height, thoracic cavity size, and absolute forced vital capacity have been reported. Further prospective studies or randomized clinical trials are needed to evaluate the long-term outcomes of lumbosacral fixation, preservation of the lumbosacral joint, or functional improvement considering the different etiology.

Keywords: Scoliosis, Cerebral palsy, Muscular dystrophy, Pedicle screws

romuscular disorder, patient's needs, and quality of life (QoL).

Many patients with neuromuscular scoliosis undergo surgical treatment. Surgical treatment for neuromuscular scoliosis has evolved with recent advances in instrument and technique. One of the significant changes is a shifting trend from Luque instrumentation with and without the Galveston technique to a pedicle screw construct.¹ Pedicle screws can achieve a 3-column fixation with a better vertebral grip and longer moment arm due to the anatomical location of the pedicle screw. This mechanical advantage improves the correction rate and decreases the operation time. Recent research using an inpatient da-

tabase in the United States revealed an increasing adoption of the all-pedicle screw fixation technique.² A 30-year surgical correction in patients with Duchenne muscular dystrophy (DMD) at a single institute showed less postoperative complications with pedicle screw fixation.³

The improved outcome of posterior correction using the pedicle screw system and the advancement of perioperative management in patients with neuromuscular scoliosis makes it possible to expect favorable results even in patients with more severe scoliosis or poor general condition. Because posterior-only correction with 3-column fixation by pedicle screw system may be comparable to the traditional posterior and anterior combined operation, and the spinal osteotomy in osteoporotic bone could be performed with the improved stability and rigidity of the pedicle screw system. The previous surgical guideline using Luque instrumentation with or without the Galveston technique should be modified based on the pedicle screw system. Furthermore, there has been tremendous advancement in understanding the natural course of neuromuscular scoliosis, the interaction between spine and hip, and growth-friendly surgery. We aimed to review the recent literature regarding posterior correction and fusion by mainly using a pedicle screw system for patients with neuromuscular scoliosis. In this review, we discussed the surgical indication, correction technique, outcome, and complications in patients with neuromuscular diseases, particularly cerebral palsy (CP) and muscular dystrophy, the most common neurologic and muscular disorder.

METHODS

We searched the PubMed database using the following terms: scoliosis, neuromuscular, cerebral palsy, Duchenne muscular dystrophy, and pedicle screw system. We reviewed only articles published in English from January 1, 2015 to October 1, 2021; 491 papers were found. All the articles were reviewed, and their references were also searched for any further, additional pertinent articles. The first author (HSK) and corresponding author (KBP) identified articles that had important new findings.

INDICATION OF SCOLIOSIS CORRECTION

Patients with neuromuscular scoliosis who cannot walk independently may have pain due to rib-pelvis impingement and trouble sitting due to trunk imbalance. Few nonoperative modalities have been effective in patients with neuromuscular sco-

liosis. Furthermore, due to the rapid progression of neuromuscular scoliosis compared to idiopathic scoliosis and the anticipated deterioration of respiratory and cardiac functions, early surgery has been recommended.

For patients with CP, early teenagers with a Cobb angle of $>40^\circ$ are considered for spinal fusion because the curves progress even after skeletal maturity and have a poor prognosis. In a retrospective study by Hollenbeck et al.,⁴ delaying surgery in those with a curve $>90^\circ$ increased the risk of infection, blood loss, and the need for anterior/posterior procedures. They recommended surgical correction for curves $<90^\circ$.

The use of steroid therapy has been associated with a reduction in the incidence of scoliosis and decreased progression of scoliosis in DMD, although adverse effects, such as the development of cataracts, risk of spine and long bone fractures, and reduced bone density, should be considered.^{1,5} However, early surgery is essential in patients who have a rapid progression of scoliosis, or whose respiratory and cardiac functions are anticipated not to be viable for later surgery.⁵

Hip dislocation and scoliosis are common in children with CP or other neuromuscular diseases. There was a concern that the pelvis can become a stiff “end vertebra” that prevents compensatory mechanisms from adjusting to hip deformities. Crawford et al.⁶ reported that 45% (21 of 47) of neuromuscular patients who underwent posterior spinal fusion required a hip procedure, either before or after spinal surgery with an average follow-up of 3.5 years. In patients who had containment surgery of the hip before posterior spinal fusion, the hips maintained the reduction after spinal fixation, and new hip subluxation/dislocations occurred in 8 cases (17%) after correction of pelvic obliquity. However, the development of subluxation and dislocation of the hip is multifactorial. Current expert opinion suggests that when hip dislocation and scoliosis are present at the same time, scoliosis-associated pelvic obliquity should be corrected before hip reconstruction; however, whether adequate treatment of the hip prevents scoliosis or vice versa in patients with CP remains unclear.⁷ The close observation of the progression of hip dislocation or scoliosis after surgical treatment of one of these problems is important in patients with neuromuscular disease (Fig. 1).

SURGICAL TECHNIQUE

1. Correction Method

Changes in the surgical fixation system from wire, hook, or hybrid instrumentation to pedicle screw-only instrumentation

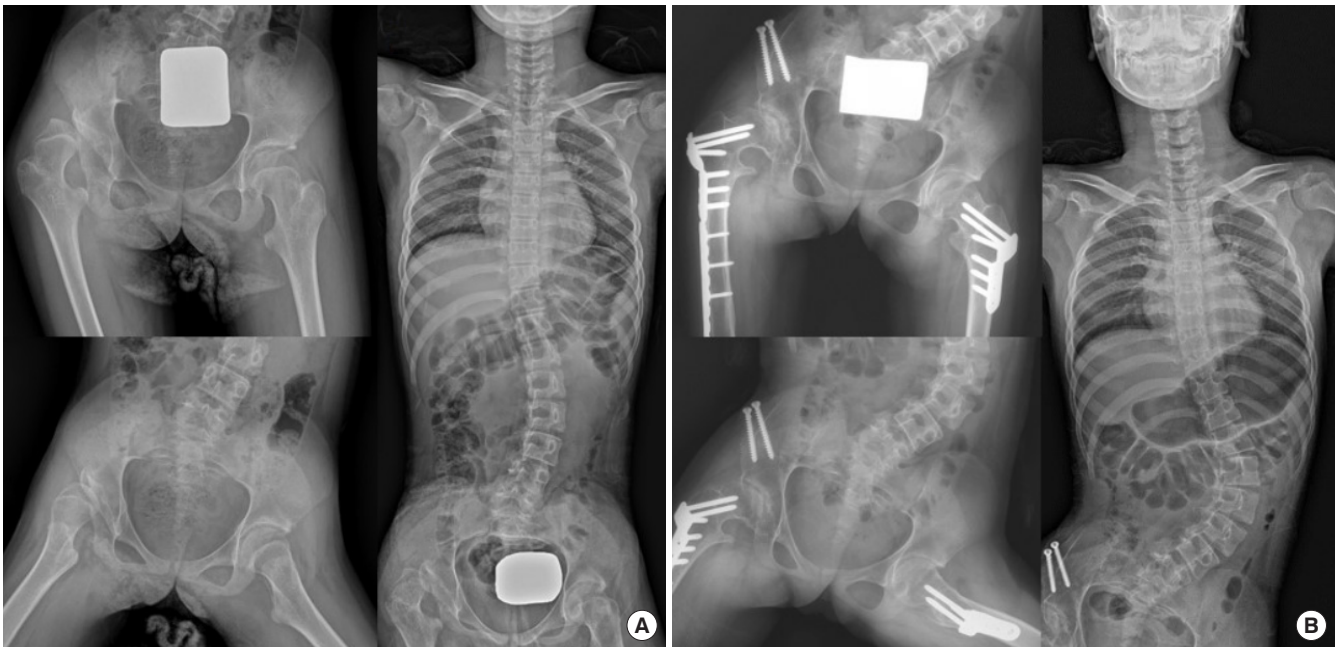


Fig. 1. (A) An 11-year-old girl with spastic quadriplegic cerebral palsy had a right hip dislocation and 37° of scoliosis from T11 to L4 in supine radiography. (B) At the postoperative 2-year follow-up after reconstructive surgery for hip dislocation, the reduction of the hip was maintained, but the scoliosis had progressed to 82° from T11 to L4 in supine radiography.

also made the posterior correction of curve more effective. Posterior-only surgery appears to lead to comparable radiological results (overall Cobb correction of 54% in the posterior-only group and 50% in the anterior-posterior group). This included shorter operation times and shorter intensive-care unit and hospital stays than the combined anterior-posterior instrumentation in patients with severe CP.⁸ Shao et al.⁹ performed a meta-analysis and described that the posterior-only approach is similar to the anterior-posterior approach for the correction of scoliosis in coronal and sagittal planes. However, the anterior-posterior approach has advantages of correcting pelvic obliquity and decreasing the loss of angle between postoperation and follow-up in the main scoliosis. In contrast, the posterior-only approach has the advantage of decreased operation time, blood loss, duration of hospital stay, and complications. The traditional anterior approach needs a wide incision or longer operation time. However, in our opinion, advanced anterior operation, such as oblique lateral interbody fusion, may be an alternative surgical option in selected cases (Fig. 2).

Spinal osteotomies have been performed for the correction of stiff curvatures with the advanced pedicle screw system and spinal cord monitoring. Bekmez et al.¹⁰ established a protocol based on the correction of the curve with traction films under general anesthesia. If the pelvis was leveled, and the correction of the

deformity was >50%, partial facetectomy was performed. However, if the residual pelvic obliquity was >15°, then an apical pedicle subtraction osteotomy was a better choice because residual pelvic obliquity over 10° was detected despite multiple posterior column osteotomy. Alternatively, preoperative or intraoperative traction is helpful for the correction of a stiff curve. In a retrospective study of 23 patients with CP scoliosis of more than 100°, intraoperative traction was a useful curve correction maneuver with shorter operation time than anterior-posterior surgery.¹¹ Furthermore, intraoperative traction showed a better correction in pelvic obliquity (33.5° vs. 14° of correction).

Besides coronal plane deformity, the angle and balance on the sagittal plane change after scoliosis correction. The lumbar lordosis or pelvic parameter is related to thoracic sagittal balance. In progressive muscular dystrophy scoliosis patients with thoracic lordosis, thoracic lordosis correction surgery could be an effective treatment method to improve the sacral slope in the standing position and the anterior pelvic tilt during gait if the power of the back and hip extensor muscles are healthy.¹² Cervical deformities associated with neuromuscular disorders also should be considered.¹³ The effects of sagittal alignment after scoliosis operation on the cervical alignment in patients who could not stand independently are unknown. Ilharreborde et al.¹⁴ classified 23 nonambulating patients with CP scoliosis into

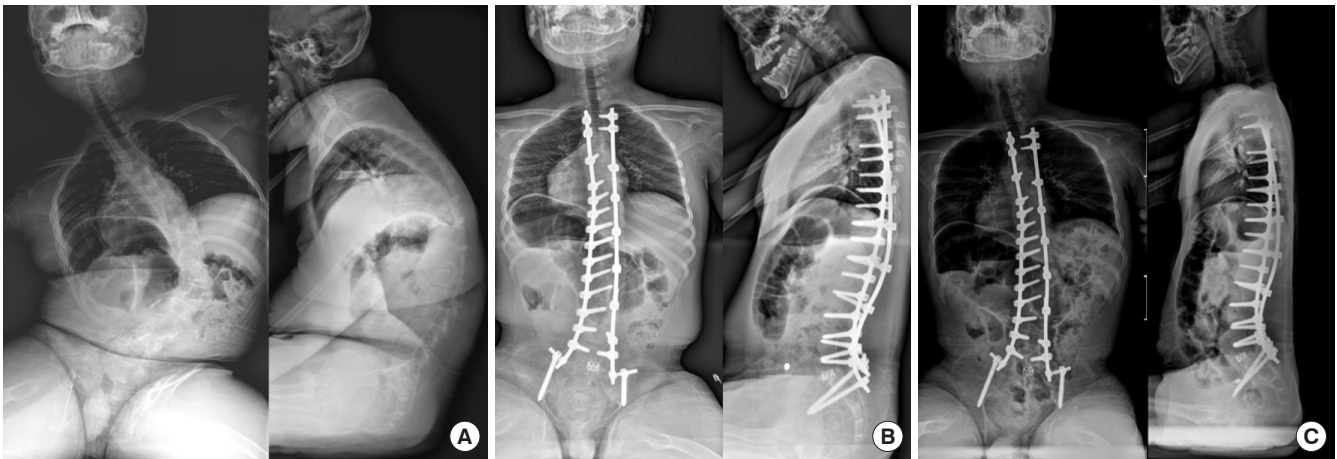


Fig. 2. (A) A 15-year-old girl with muscular dystrophy had 56° (T6–T12) of neuromuscular scoliosis in the posteroanterior whole spine radiography in the sitting position. (B) She underwent L5/S1 oblique lateral interbody fusion and posterior instrumentation from T1 to the pelvis. (C) Pelvic obliquity was successfully corrected and maintained until the postoperative 6-year follow-up.

2 groups: 61% of the patients with less trunk control had lumbar lordosis $< 50^\circ$, retroverted and vertical pelvis, and anterior imbalance, and 39% of the patients with better trunk control had lumbar lordosis $> 60^\circ$, anteverted and horizontal pelvis, and posterior imbalance. The clinical significance of this classification should be further evaluated. Additionally, factors that could influence the head and trunk posture should be systematically considered.

A growing rod that can adjust the changes of scoliosis according to the growth of spine is also used in neuromuscular scoliosis. Miladi et al.¹⁵ used 4 hook claws at the proximal end with an iliosacral screw in the pelvis in 100 patients and did not perform a final fusion. During the follow-up period (3⁺⁹ years; range, 2 years to 6⁺³ years), 12 mechanical complications and 16 wound infections were noted. Successful application of fusionless surgery in patients with spinal muscular atrophy was also reported.¹⁶ The use of a growing rod may be an alternative technique for the treatment of neuromuscular scoliosis; however, a long-term follow-up study is essential to assess the outcome of the fusionless technique. Magnetically controlled growing rods also show favorable outcomes in neuromuscular scoliosis.¹⁷ However, the growing rate of spine in neuromuscular scoliosis is not certain compared to the typically growing children. Furthermore, the limited use of magnetic resonance imaging to evaluate the central nervous system after the operation with a magnetically controlled growing rod should be considered.

2. Pelvic Fixation

In posterior correction and fusion for neuromuscular scoliosis, fusion to the pelvis is recommended to correct pelvic obliquity and prevent loss of correction.

A rigid construct for spinopelvic fixation with pedicle screws showed improved deformity correction and a lower rate of pseudarthrosis (22% vs. 5%) compared with sublaminar wire fixation with Galveston or iliac screw pelvic fixation in a retrospective comparative study of 80 patients with neuromuscular scoliosis.¹⁸ Sacral-alar-iliac fixation, intraoperative computed tomography-guided navigation, screw augmentation using bisphenol-a-glycidyl dimethacrylate composite resin, or transiliac bar have been performed with favorable outcomes in neuromuscular scoliosis.^{19–23}

Concerning the biomechanical aspect, pelvic fixation requires more stiff implants in order to be resistant to forces generated during motion and rotation at the lumbosacral junction. Schoenleber et al.²⁴ compared both iliac screws alone and iliac and S1 screws. No differences in complications were observed, but the correction of pelvic obliquity was superior in the iliac-S1 group in a 2-year follow-up. Myung et al.²⁵ reported early failures of pelvic fixation in 12 of 41 patients (29%). The failure rate with < 6 screws in the L5, S1, and pelvis was higher at 35%, and no failures occurred if at least 6 screws were used in the L5, S1, and pelvis. To achieve a 6-screw fixation, they recommended S2-alar-iliac screws instead of traditional iliac screws due to the in-line nature of the screws' heads in a construct with bilateral L5, S1, and S2-iliac screws. S2-alar-iliac screws also do not need a screw-to-rod connector, resulting in decreased modes of failure and less muscle dissection.²⁵ Abousamra et al.²⁶ also reported inferior outcomes of iliac screws compared with unit rod and S2-alar-iliac screws in terms of complication and curve correction. In a retrospective study of 50 patients with neuromuscular scoliosis (28 standard iliac screws, 22 S2-alar-iliac screws), no difference

was found between the groups for frequency or severity of postoperative complications, including wound infections. However, a statistically significant difference in radiographic implant failure rates was noted between the standard iliac screw and S2-alar-iliac screw groups (57% vs. 27%).²⁷ Shabtai et al.²⁸ reported a lower implant failure rate (7%, 3 of 46) in the S2-alar-iliac screw group than in the iliac screw group (24%, 13 of 55) in a review of 101 patients. In a meta-analysis with 6 clinical studies comparing outcomes of S2-alar-iliac screws versus iliac screws, a superior complication profile in terms of screw head prominence, loosening, and revision rates was noted in S2-alar-iliac screws;²⁹ however, the traditional iliac screw technique was an early technique compared with the S2-alar-iliac screw technique and loosening of S2-alar-iliac screw was reported in older patients.³⁰ Although the S2-alar-iliac screw technique needs technically advanced skills, it would be most appropriate for the pelvic fixation in terms of stability and low profile.

However, pelvic or sacral fixation has several disadvantages, including increased blood loss, longer operation time, and technical difficulty. Takaso et al.³¹ performed segmental pedicle screw instrumentation and fusion only up to L5 in patients with 20 DMD and 7 spinal muscular atrophy (SMA). Preoperatively, the apex of the curve was L2 or higher, and the L5 tilt was $<15^\circ$. In a minimum 2-year follow-up, pelvic obliquity improved from 15° preoperatively to 5° postoperatively, and 6° at the last follow-up. The L5 tilt improved from 9° preoperatively

to 2° postoperatively, and 2° at the last follow-up. No significant loss of correction was found in the coronal curve and pelvic obliquity. They described that pelvic obliquity could be effectively corrected and maintained in select cases after proper correction of the thoracolumbar scoliotic curve because lumbosacral articulation is stable and pelvic obliquity is due to the suprapelvic effects of scoliosis. The advantage of the mobile L5–S1 disc space includes the absorption of the angular and rotational movements of the trunk.

On the contrary, Tøndevold et al.³² recommended pelvic fixation in neuromuscular scoliosis because correction of the main curve and pelvic obliquity was superior in patients with pelvic fixation in their retrospective study on 91 nonambulatory neuromuscular scoliosis patients. Loss of pelvic obliquity correction was also more frequently seen in the L5 group (25% vs. 0%). However, no significant difference was found between the L5 group and the pelvic group in terms of perioperative blood loss and postoperative complications. They described that patients with pelvic fixation maintained their coronal balance better, and this was most likely due to better stability at the lumbosacral junction.

The simple comparison between these 2 studies is inappropriate because Takaso et al.³¹ included only flaccid neuromuscular scoliosis. In contrast, the patients in the study by Tøndevold et al.³² had a different etiology (52% of CP, 12% of central motor neuron involvement, 10% of SMA, and others). The ad-

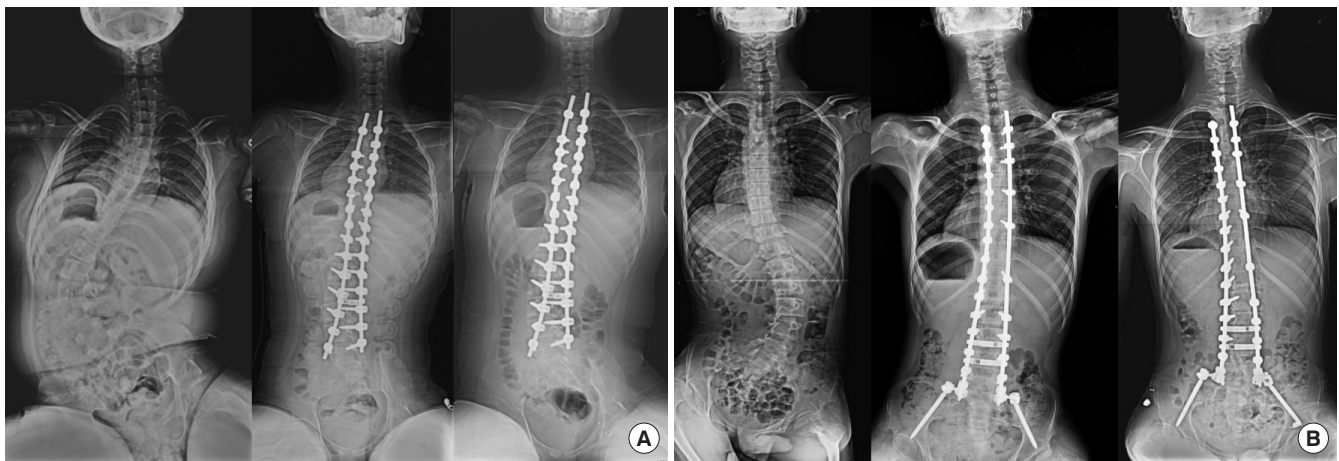


Fig. 3. (A) A 12-year-old girl with spastic quadriplegic cerebral palsy underwent posterior instrumentation from T2 to L5. She underwent hip reconstructive surgery 3 years ago. Preoperatively, hip joint was stable and L5 tilt on pelvis was not prominent, although pelvic obliquity on sitting position was severe. She was very thin (38 kg), and had a gastrostomy tube and sore on coccyx. Because of the possibility of postoperative infection, fusion was stopped at L5. Pelvic obliquity improved after operation, but the remaining pelvic obliquity was slightly progressed at the 12-month follow-up. However, the patient and parents did not complain of any discomfort during sitting. (B) A 17-year-old boy with spastic quadriplegic cerebral palsy underwent posterior instrumentation from T3 to the pelvis. Pelvic obliquity was improved and maintained during the 5-year follow-up.

vantage of the mobile L5–S1 disc space may be the absorption of stress during wheelchair activities, sitting on the floor, or crawling. Although the fixation to L5 in patients without severe pelvic obliquity has not achieved wide acceptance, it certainly merits strong consideration, particularly with early diagnosis and adequate counseling of the family before the curves become severe and rigid (Fig. 3).³³ In a recent review article, fusion short of the pelvis was considered in patients with adequate head control without the presence of hip subluxation or dislocation and when pelvic obliquity is $< 15^\circ$.³⁴ In our opinion, if pelvic obliquity is associated with coronal imbalance, especially C-type neuromuscular scoliosis, pelvic fixation should be performed considering the correction of the curve and postoperative coronal balance.

3. Intraoperative Monitoring

Intraoperative monitoring of transcranial electric motor-evoked potentials (tce-MEPs) and somatosensory-evoked potentials (SSEPs) is safe and effective for spinal cord monitoring during scoliosis surgery. The use of intraoperative monitoring has increased during neuromuscular scoliosis operation.^{2,35} However, the reliability of spinal cord monitoring in patients with neuromuscular scoliosis is conflicting, because the preoperative baseline reduced due to immaturity of motor pathways in younger children or atrophy of cortical dendrites. A relatively high incidence of false-positive cases (4 of 29, 13.8%) was noted in patients with neuromuscular scoliosis related to the central nervous system, such as CP.²⁷ However, there was no false-positive in peripheral nervous system, such as DMD, spinal muscular atrophy, or myopathy. Another question may be which monitoring is more reliable between tce-MEPs and SSEPs. Thirumala et al.³⁶ confirm the high sensitivity and specificity of SSEPs monitoring alone in idiopathic scoliosis. In neuromuscular scoliosis, Pastorelli et al.²⁷ described that monolateral or bilateral reduction of peripheral and cortical SSEPs with or without significant changes in tce-MEPs occurring at any time during surgery suggests peripheral nerve lesion due to malposition related to muscular atrophy, tendon retractions, and skeletal malformations. Application of both tce-MEPs and SSEPs may be more reliable in patients with neuromuscular scoliosis.

OUTCOMES AND COMPLICATIONS AFTER SURGERY

A high rate of complications occurs after the surgical correction of neuromuscular scoliosis. Sullivan et al.³⁷ reviewed data

on deep surgical site infections after spinal arthrodesis in 1,353 patients with idiopathic, neuromuscular, or genetic and syndromic scoliosis. Deep surgical site infections were more common in neuromuscular scoliosis (10.0%) than in adolescent idiopathic scoliosis, kyphosis (2.7%), or genetic and syndromic scoliosis (4.2%). However, consideration of nutrition status decreases the postoperative complication and the incidence of surgical site infection in neuromuscular scoliosis decreased significantly (16.1% vs. 4.4%) after the implementation of the strategies mentioned in the 2013 Best Practice Guideline.³⁸⁻⁴⁰ Postoperative fever is a frequent complication after surgery. Yousef et al.⁴¹ reviewed 76 patients who had undergone scoliosis surgery and 49 patients (64.5%) who developed postoperative fever with a temperature of $> 38^\circ\text{C}$. This temperature reading was a sign of infection in 32.7% of patients, and urinary tract infection was the most frequent finding.

Another concern is a risk factor about the postoperative complication.⁴² Obese neuromuscular patients undergoing posterior fusion have higher odds of experiencing adverse outcomes, particularly surgical site infections, urinary tract infections, and readmissions.^{43,44} Toll et al.⁴⁵ reported a perioperative complication rate of 27% in 102 cases of neuromuscular scoliosis at a single center. They suggested a preexisting pulmonary compromise and greater intraoperative blood loss as the greatest risk factors for a major perioperative complication. Luhmann and Furdock⁴⁶ also reported a postoperative respiratory complication rate of 15.3% (17 of 111), and a history of pneumonia and gastrostomy tube as risk factors. Gross Motor Function Classification System (GMFCS) level 5, based on the mobility of patients, was subcategorized according to preoperative central neuromotor impairments by Jain et al.⁴⁷ The rate of major complications increased significantly with more involvement of a gastrostomy tube, tracheostomy, history of seizures, and nonverbal status.

Because of the high incidence of complications, surgical correction for neuromuscular scoliosis must provide meaningful benefits in the improvement of QoL in proportion to the risks. In a prospective multicenter analysis performed by Miyanji et al.⁴⁸ using the Child Health Index of Life with Disabilities (CPCHILD) questionnaire for 5 years, significant improvements in CPCHILD personal care, positioning, and comfort domains were noted. The overall complication rate was 46.4% at 1 year, with an additional 5.7% up to 5 years. No correlation was found between complications and CPCHILD scores postoperatively at 1, 2, and 5 years. They concluded that the substantial complication rate does not correlate with health-related QoL changes postoperatively, suggesting that the benefits of surgery outweigh

the risks in this fragile population. In another aspect, improvement in surgical skill and evolution of instrumentation decreases the rate of complication. The Scoliosis Research Society Morbidity and Mortality database shows a significant decrease in wound infections (superficial and deep), respiratory complications, and implant-associated complications; and the overall complication rate decreased by approximately 10% from 2004–2007 to 2012–2015.⁴⁹

Jain et al.⁵⁰ compared treated and untreated CP scoliosis patients' qualitative and quantitative health-related QoL assessments. Caregivers reported overall improvement of patients' lives after spinal fusion, and ranked scoliosis surgery as the most beneficial intervention in the children's lives, secondary to gastrostomy tube insertion. Disappointingly, DiFazio et al.⁵¹ demonstrated that the health-related QoL improved 1 year following spine fusion but regressed to baseline after 2 years in children with CP.⁵¹ However, Sewell et al.⁵² demonstrated the improvement in caregiver-assessed QoL and pain in the spinal fusion group in a retrospective study of 33 children with GMFCS IV/V CP scoliosis.⁵² In patients with CP, caregiver-assessed QoL is important, because most of the patients are quadriplegic CP, and they are not ambulatory and have low level of cognition. We also recommend operative correction for patients with CP scoliosis because pelvic obliquity related to scoliosis may affect hip stability and sitting balance, which is also related to the QoL of patients.

Suk et al.⁵³ reviewed the postoperative QoL in patients with DMD (27 patients), SMA (15 patients), and progressive muscular dystrophy (16 patients). All patients were nonambulatory, and pelvic fixation was performed in all patients. The Muscular Dystrophy Spine Questionnaire results indicated that the patients had significantly improved sitting balance-related outcomes, whereas the 36-item Short Form Health Survey indicated improvements only in bodily pain and social functioning scales. The collapse of the rib cage ("parasol rib deformity") that was found in patients with hypotonic neuromuscular diseases, such as SMA and muscular dystrophies, may be associated with poor pulmonary function. Unfortunately, parasol rib deformity did not improve after treatment with growing rods or a vertical expandable prosthetic titanium rib.⁵⁴ However, after the insertion and lengthening of the posterior-based growing rods in the SMA population, the clinical respiratory support requirements appeared to stabilize. Increased spinal height and thoracic cavity size were noted with an increasing absolute forced vital capacity (FVC), although the percent predicted FVC diminished.⁵⁵

To decrease postoperative pulmonary complications, noninvasive positive-pressure ventilation (NIPPV) and pulmonary

rehabilitation were attempted. Chong et al.⁵⁶ compared 45 patients with no mechanical ventilation and 28 patients who received NIPPV for respiratory support. Although statistically not significant, patients in the non-NIPPV group had a higher incidence of pulmonary complications (38% vs. 21%). In another study, 24 patients with an FVC of < 30% and neuromuscular scoliosis had undergone staged anterior and posterior scoliosis correction safely through pulmonary rehabilitation.⁵⁷ Pneumonia and upper respiratory infection were noted in only 3 cases. However, Farber et al.⁵⁸ reported 6 patients with SMA, and most cases of 7 patients with DMD lost vital capacity after posterior fusion.

Many patients undergoing spinal fusion for neuromuscular scoliosis have preexisting neurosurgical implants, including ventricular shunts for hydrocephalus or baclofen pumps for spasticity control. A recent study suggested that spinal fusion for neuromuscular scoliosis correction does not increase the rates of complications involving previously placed neurosurgical implants.⁵⁹

PROBLEMS IN PEDICLE SCREW ERA

The pedicle screw system improved the surgical outcome of neuromuscular scoliosis and decreased postoperative complications; however, there are still unsolved problems. The most commonly encountered problem is mechanical issues related to the small bone or osteopenia in nonambulatory patients with neuromuscular scoliosis. The small distorted pedicle makes screw insertion difficult, and the weak pull-out strength due to osteopenia may induce implant failure or proximal junctional problem.

Due to hypoplastic pedicles, osteoporosis, and severe rotation of the vertebral body in neuromuscular scoliosis, pedicle screw insertion is difficult particularly at the apex of the curve. Hybrid fixation using sublaminar wire or hook had been tried, but the sublaminar wire is associated with a high rate of neurologic complications, and the hook is inferior in terms of fixation strength.⁶⁰ Several studies reported favorable outcomes using sublaminar polyester bands.^{61–63} Sublaminar polyester bands have advantages of achieving bidirectional fixation, in contrast to the 1-way fixation achieved by pedicle screws and increased contact surface compared to wire, thereby decreasing the stress per unit area. The bicortical fixation through the lamina is especially important in patients with neuromuscular scoliosis due to osteopenia. In our opinion, additional sublaminar polyester bands would provide better pull-out strength of pedicle screw in the correc-

tion of neuromuscular scoliosis.

S2-alar-iliac screw is an excellent solution for stable spinopelvic fixation. However, mechanical failure was still reported, and Hyun et al. suggested a larger-diameter screw (at least 8.0 mm in diameter).^{30,64,65} Patients with neuromuscular scoliosis are generally small due to developmental delay or nutritional problems. If a larger-diameter screw is unable due to small bone, the dual screw may be another option.⁶⁶ Although we can do the stable lumbosacral fixation, the selection between the mobility for the advantages in performing transfers, weight shifts, or rotational mobility, and the fusion for the correction of pelvic obliquity for sitting balance is questionable. Douleh et al.³⁴ suggested adequate head control as a guide for L5–S1 preservation. This indication looks ideal considering proximal junctional problem.

Compared to the adult population, proximal junctional kyphosis or proximal junctional failure following long fusions to the sacrum and pelvis in children with neuromuscular scoliosis is much less known.⁶⁷ Menger et al.⁶⁸ reported 12% proximal junctional failure in 25 ambulatory pediatric patients with non-idiopathic spinal deformity. They recommended considering sagittal plane dynamics when performing fixation to the pelvis. Toll et al.⁶⁹ reported 27% proximal junctional kyphosis with more than 10° increase and 7% proximal junctional failure in 60 neuromuscular scoliosis. They suggested preoperative halo-gravity traction or greater postoperative C2 sagittal translation, loss of primary curve correction, and smaller preoperative proximal kyphosis as risk factors of proximal junctional kyphosis. Because the cause of the proximal junctional disease is multifactorial, the prevention method could not be suggested. Hook or ligament augmentation to lessen instrumentation rigidity and suddenness of transition at the upper instrumented vertebra could be utilized selectively in neuromuscular scoliosis considering proper sagittal alignment and osteopenia.⁷⁰ Most of all, surgeons should examine the functional status of patients with neuromuscular scoliosis and vertebral body deformity before selecting fusion level and fixation method.

Our study has limitations to suggest specific guidelines because this study is not a systematic review. However, this study will provide the current concept about surgical correction for neuromuscular scoliosis using a pedicle screw system and reawake what we should consider in the pedicle screw era.

CONCLUSIONS

The posterior correction for neuromuscular scoliosis could be effectively achieved with a 3-column fixation using a pedicle

screw system because of the immediate stability in all directions, accompanied by spinal osteotomies or intraoperative traction. The improved correction of scoliotic curvature raises the question of whether to sacrifice the lumbosacral joint. Many studies suggest pelvis fixation with a low profile instrument, such as S2-alar-iliac screws; however, the surgeon should consider not only pelvic obliquity but also sagittal balance and patient's gross motor function. The improvement of QoL was more definite in patients with CP. However, the differences in the level of cognition and activity depending on the neurologic and muscular disorder would have to be considered. Most of the included studies in this review were retrospective in nature. Prospective and randomized clinical trials are warranted, especially to prove the long-term outcomes of lumbosacral fixation, preservation of the lumbosacral joint, and growth-friendly surgery.

NOTES

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