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## Abstract

Scoliosis is a common clinical entity ubiquitous to most neurologic disorders encountered by the pediatric orthopaedic surgeon and associated care providers. Until recently, though widely performed, the evidence for scoliosis corrective surgery and its impact on patient-specific outcomes was sparse at best. In addition, the choice of surgical techniques applied during surgery for 'neuromuscular' scoliosis was based mainly on the results of historic case series with a paucity of higher-level evidence available. Over the last few years, however, the number and quality of relevant studies has increased substantially. The aim of this chapter is to provide a review of the best available evidence and provide recommendations as to most appropriate treatment(s) pertaining to the management of neuromuscular scoliosis. Given the results of this evidence-based review, it would seem that neuromuscular scoliosis correction is a worthwhile procedure with expected improvements in function, quality of life, and patient/caregiver satisfaction, albeit with high surgical risks. The ultimate decision as to whether or not scoliosis surgery is performed should be dependent on a disease-specific assessment of the risks and benefits with appropriate communication to the patient and/or caregiver. With respect to surgical fixation, the use of thoracic/lumbar segmental pedicle screws may offer improved outcomes over other methods but the best choice of pelvic fixation is still controversial. Regarding the use of anterior surgery for neuromuscular scoliosis, its role remains unclear except in the case of spina bifida where it is likely to reduce perioperative risks.

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## Keywords

Neuromuscular scoliosis • Cerebral palsy • Spinal bifida • Duchenne muscular dystrophy • Spinal fixation • Pelvic fixation • Anterior surgery • Quality of life

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## Introduction

Spinal deformities associated with neuromuscular diagnoses are common, typically progressive, and often require operative intervention. Despite a wealth of literature in this subject area, most of the published studies are retrospective, overly generalized by diagnosis, focused on radiographic rather than patient-centered outcomes, and are lacking a comparator group. This deficit in high quality studies makes it difficult to draw any hard conclusions regarding the most appropriate choice of operative strategy for scoliosis correction in these disorders. Recently however, there has been a shift towards comparative studies and the use of quality of

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life (QOL) related outcomes that serve to make the picture a little clearer.

The aim of this chapter is to provide a review of the best available evidence and provide recommendations as to most appropriate treatment(s) pertaining to the management of scoliosis associated with neuromuscular diagnoses [1].

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## Etiology and Morphology of Scoliosis in Neuromuscular Disorders

Spinal deformities secondary to neuromuscular disorders are common, with etiologies varied in pathophysiology and anatomic origin. These associated diagnoses can be categorized anatomically, stemming from upper motor neuron causes (e.g. cerebral palsy (CP), Friedrich's ataxia, syringomyelia), lower motor neuron causes (e.g. spinal muscular atrophy (SMA), poliomyelitis), or myopathic causes (e.g. Duchenne muscular dystrophy (DMD), myotonic dystrophy). From a pathophysiological point of view, scoliosis may be secondary to problems with weakness, coordination, and/or hypertonia of the truncal musculature coupled with a lack of effective compensatory mechanisms [1]. Neuromuscular curves typically present at a younger age and are known to exhibit rapid progression during growth that often continues, albeit more slowly, even after skeletal maturity [2].

Irrespective of diagnosis, the curve types associated with neuromuscular etiologies follow typical patterns. The most common curve type is long and C-shaped, often with associated pelvic obliquity, a collapsing kyphosis, and a loss of sitting balance (Fig. 24.1a) [2, 3]. The similarity in curve presentation across diagnoses has led to the convergence of surgical indications and operative strategies for the management of neuromuscular diagnoses, with little difference in approach between disorders except in specific cases (e.g. the exclusion of pelvic fixation in the absence of pelvic obliquity).

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## Goals of Scoliosis Correction: Patient Versus Surgical

The mainstay of treatment for neuromuscular curves involves posterior instrumentation and fusion from the upper thoracic spine (typically T2 or T3) to the pelvis. Many options for spinal fixation have been previously reported including the use of sublaminar wires or bands, segmental pedicle screw fixation, and hybrid methods involving more than one implant type (Fig. 24.1b–d) [4–6].

Indications for surgery have traditionally been met when the major curve reached a Cobb angle of 40–50° and/or there was a significant functional deficit, specifically with respect to sitting tolerance. The goals of the patient and/or caregiver, however, center on the expectation of improvements in activities of daily living (e.g. dressing, independent ambulation, personal hygiene), the absence of pain, ease of care-giving, social interaction, in

addition to comfortable sitting [7]. In other words, the patient's major expectations revolve around issues pertaining to QOL.

Despite these goals, the published literature has, until recently, focused primarily on radiographic outcomes such as Cobb angle correction, implant type and density, pseudoarthrosis rates, and surgical approaches, in addition to reports of peri-operative complications [8–10]. This focus may be at least partially misplaced as several studies have reported a lack of correlation between the extent of Cobb angle correction and QOL improvements in children with neuromuscular disorders [7, 11, 12]. Arguably, the only radiographic goals that might have a substantial overall impact are the achievements of (1) a balanced spine over a level pelvis and (2) a solid spinal fusion.

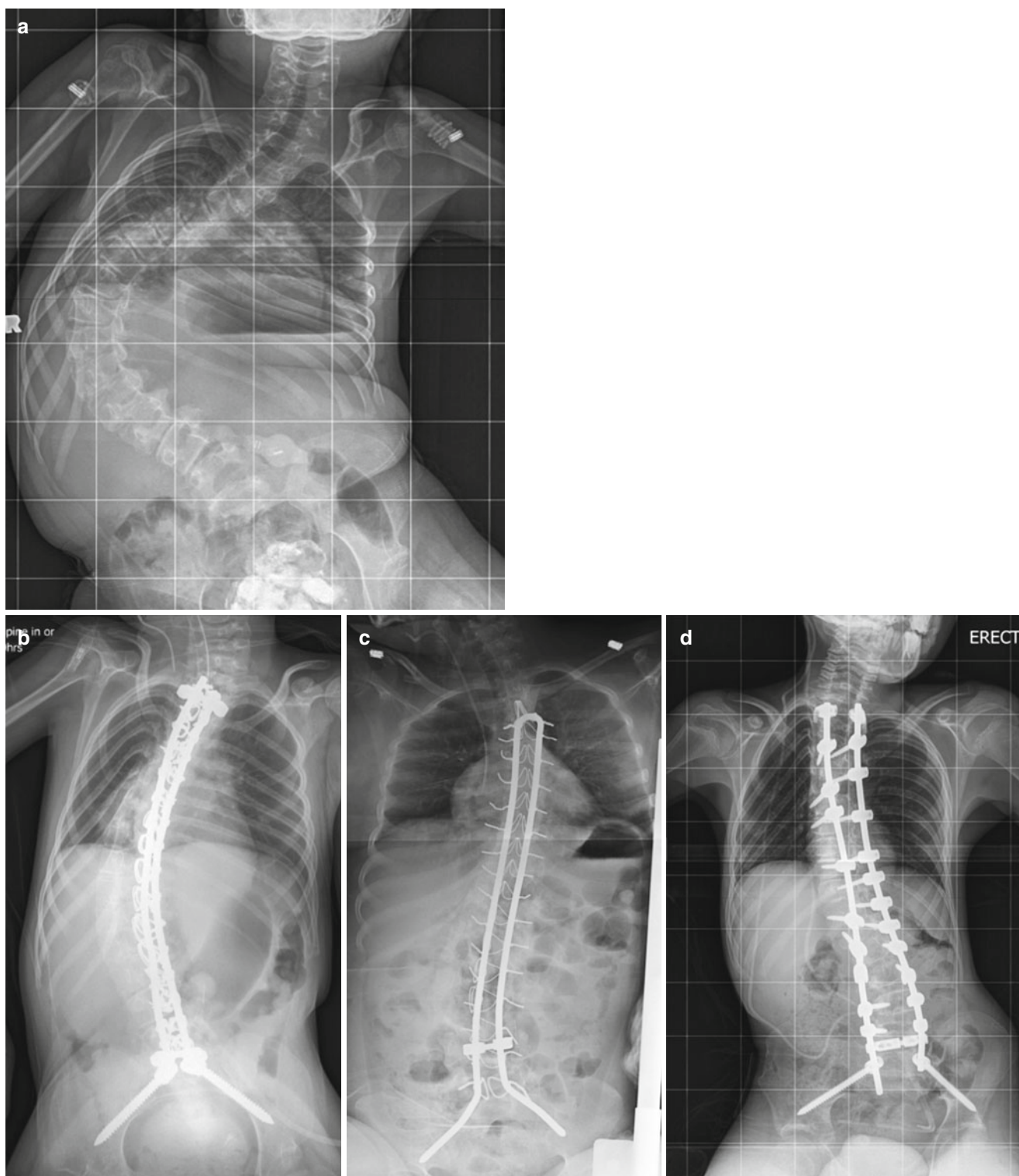
Of course, the patient and/or caregiver have a substantial interest in the surgical risks associated with these procedures but this represents only one side of the risk-to-benefit ratio. A true evidence-based assessment of the available literature must give priority to those studies that report outcomes related to factors that matter to the patient and/or caregiver. At the present time, this may prove to be a difficult task given that, previous to 2011, there were very few published studies that reported QOL measures as outcomes of neuromuscular scoliosis correction [11]. However, in recent years, validated outcome measures have been developed which should prompt an increase in studies that prioritize outcomes on both sides of the risk-benefit ratio [13]. At this time, however, the number of studies that measure patient-specific outcomes remain scant as will be seen throughout the course of this chapter.

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## Is 'Neuromuscular Scoliosis' a Diagnosis to Be Analyzed?

The definition of what is considered a 'neuromuscular' diagnosis is variable amongst clinicians and is a source of some confusion, especially when reviewing the relevant literature on the subject. From the neurologists' view, the term typically refers to neurologic disorders that are progressive with respect to their primary etiology (e.g. DMD) versus those that arise from a static neurologic lesion with associated progressive musculoskeletal manifestations (e.g. CP) [14]. The distinction is important given that these diagnoses have different natural histories, different levels of gross motor function (depending on disease severity), different surgical risk profiles (e.g. ventriculo-peritoneal shunt failure in myelomeningocele), and differing evidence regarding the utility of surgical interventions such as scoliosis correction [3, 7].

Despite these differences, for reasons likely related to improving sample sizes in studies investigating the role of scoliosis surgery for disorders with varying prevalence, disparate diagnoses such as cerebral palsy, spina bifida, Duchenne muscular dystrophy and others are often 'lumped together' and analyzed as if they were equivalent entities [9, 15]. Although the motivations are well understood, the



**Fig. 24.1** Typical neuromuscular curve type and options for spinal fixation. (a) C-shaped thoracolumbar curve with associated pelvic obliquity. (b) Hybrid fixation including Luque sublaminar wires and sacral alar iliac screws. More commonly, lumbar pedicle screws are

present in addition to sacropelvic screw fixation. (c) Unit rod including Luque sublaminar wires and Galveston pelvic fixation. (d) Segmental pedicle screw fixation with sacral alar iliac pelvic fixation

question remains as to the validity of this practice as it undoubtedly skews the interpretation of surgical results in favour of the diagnosis with the largest number of subjects within the analysis, most typically CP [10].

As with other musculoskeletal manifestations of the many diagnoses that fall under the ‘neuromuscular’ moniker, the prevalence of scoliosis in these populations is typically high and most often related to disease severity [16–18]. Like

incidence, surgical success seems to be related to neuromuscular disease severity. In CP, for example, the incidence of hip displacement has been reported to be significantly correlated to functional level (via the Gross Motor Function Classification System (GMFCS)) as have the success rates for hip adductor surgery [19, 20]. Similar to the argument against merging results of different diagnoses, analyzing surgical outcomes without stratifying the analysis by functional level may also skew outcomes in favour of the higher functioning diagnoses and subjects [7].

Given the discussion above, coupled with the reality that most of the published literature regarding the treatment of neuromuscular scoliosis lacks a comparator group and are primarily retrospective in nature, one must be cautious when applying the conclusions of the available literature in clinical practice.

## Search Strategy and Grade Recommendations

For the purposes of this evidence-based chapter, studies for consideration were identified via thorough searches of the PubMed, Cochrane and Web of Science databases, using a combination of keyword and controlled vocabulary searches. Search terms used included but were not limited to: scoliosis, neuromuscular scoliosis, neurogenic scoliosis, cerebral palsy, spina bifida, myelomeningocele and Duchenne muscular dystrophy. When assessing retrieved studies for inclusion, the Journal of Bone and Joint Surgery Level-of-Evidence ratings were utilized [21]. The search focused on systematic

reviews, randomized control trials and studies that contained a comparator group (levels of evidence I, II and III). Lower level studies (level IV) were considered only when there were no higher-level studies available. Searches were limited to include only English-language studies. A sample PubMed search strategy is outlined in Table 24.1.

For each question asked we have provided an overview of the evidence and applied grades of recommendation according to Wright et al. [22]:

GRADE A – good evidence based on level I studies with consistent findings.

GRADE B – fair evidence based on consistent level II or Level III studies.

GRADE C – poor or conflicting evidence based on level IV/V evidence.

GRADE I – insufficient evidence to make a treatment recommendation.

## What Is the Evidence for Scoliosis Correction?

### Scoliosis Correction: Risks Versus Benefits

In the preceding section, it was established that the true benefits of scoliosis correction should be measured in terms of outcomes of interest to the patient and/or caregiver. In a general sense, taking neuromuscular diagnoses as a whole, there are several studies that met the inclusion criteria for this evidence-

**Table 24.1** Sample PubMed search strategy

Search	Query	Items found
#20	Search (((("Infant"[Mesh] OR "Child"[Mesh] OR "Adolescent"[Mesh] OR "Pediatrics"[Mesh] OR "Minors"[Mesh])) OR (infant[Title/Abstract] OR child[Title/Abstract] OR children[Title/Abstract] OR toddler *[Title/Abstract] OR kindergarten *[Title/Abstract] OR adolescent[Title/Abstract] OR adolescence[TIAB] OR minor[Title/Abstract] OR minors[Title/Abstract] OR boy[Title/Abstract] OR boys[Title/Abstract] OR girl[Title/Abstract] OR girls[Title/Abstract] OR pediatr *[Title/Abstract] OR juvenile[Title/Abstract] OR youth[Title/Abstract]))) AND ((((((("Scoliosis"[Mesh] OR "Scoliosis/surgery"[Mesh])) OR (scoliosis OR "neuromuscular scoliosis" OR "neurogenic scoliosis"))) AND (((("Cerebral Palsy/surgery"[Mesh] OR "Muscular Dystrophy, Duchenne/surgery"[Mesh] OR "Meningomyelocele/surgery"[Mesh])) OR ((("cerebral palsy" OR "spina bifida" OR myelomeningocele OR "Duchenne Muscular Dystrophy"))) AND (surgery OR surgical)))))) Filters: English	507
#19	Search (((("Infant"[Mesh] OR "Child"[Mesh] OR "Adolescent"[Mesh] OR "Pediatrics"[Mesh] OR "Minors"[Mesh])) OR (infant[Title/Abstract] OR child[Title/Abstract] OR children[Title/Abstract] OR toddler *[Title/Abstract] OR kindergarten *[Title/Abstract] OR adolescent[Title/Abstract] OR adolescence[TIAB] OR minor[Title/Abstract] OR minors[Title/Abstract] OR boy[Title/Abstract] OR boys[Title/Abstract] OR girl[Title/Abstract] OR girls[Title/Abstract] OR pediatr *[Title/Abstract] OR juvenile[Title/Abstract] OR youth[Title/Abstract]))) AND ((((((("Scoliosis"[Mesh] OR "Scoliosis/surgery"[Mesh])) OR (scoliosis OR "neuromuscular scoliosis" OR "neurogenic scoliosis"))) AND (((("Cerebral Palsy/surgery"[Mesh] OR "Muscular Dystrophy, Duchenne/surgery"[Mesh] OR "Meningomyelocele/surgery"[Mesh])) OR ((("cerebral palsy" OR "spina bifida" OR myelomeningocele OR "Duchenne Muscular Dystrophy"))) AND (surgery OR surgical))))))	569
#15	Search (((("Scoliosis"[Mesh] OR "Scoliosis/surgery"[Mesh])) OR (scoliosis OR "neuromuscular scoliosis" OR "neurogenic scoliosis"))) AND (((("Cerebral Palsy/surgery"[Mesh] OR "Muscular Dystrophy, Duchenne/surgery"[Mesh] OR "Meningomyelocele/surgery"[Mesh])) OR ((("cerebral palsy" OR "spina bifida" OR myelomeningocele OR "Duchenne Muscular Dystrophy"))) AND (surgery OR surgical))))	649

based review and used a QOL measure to assess patient-centered benefits.

In a prospective study utilizing the Swedish spine registry, Ersberg and colleagues used the EQ-5D (a validated instrument with sections that evaluate self-care, mobility, usual activities, pain, and anxiety) and the SRS-22 questionnaire to compare QOL between idiopathic, neuromuscular, and congenital scoliosis groups [15]. From a surgical risk perspective, patients with neuromuscular scoliosis experienced statistically significant increases in intra-operative blood loss, duration of surgery, and length of hospital stay as compared to the idiopathic group. With respect to QOL measures, there were significant increases in the post-operative EQ-5D total score and in particular the reduction of post-operative pain. When assessed via the SRS-22 instrument, neuromuscular patients experienced significantly improved function and better self-image. Interestingly, despite the significant number in complications in the neuromuscular group, when stratified into groups with and without complications, there were no significant differences in QOL scores. The conclusion of this study was that QOL was improved after scoliosis surgery even despite the high risk of complications.

In another prospective Swedish study, the impact of scoliosis surgery in neuromuscular patients was assessed in terms of outcomes pertaining to QOL, using a questionnaire that assessed sitting, care-giving, reaching, pain, rest time, seating supports, and activities of daily living (ADL), Cobb angle correction, and respiratory function as measured by vital capacity (VC) at a mean follow-up of 7 years [7]. This landmark study stratified patients who underwent scoliosis surgery into 4 subgroups including those that: (1) understood verbal instructions, (2) did not understand verbal instructions, (3) had progressive disease (e.g. DMD), and (4) had non-progressive disease (e.g. CP). Overall, surgical patients had significant QOL improvements in sitting balance, weight distribution, ADL, time used for resting, number of seating supports in addition to improvements in Cobb angle (~50 %) and respiratory function. When analyzed by subgroups, both the non-progressive and verbal instructions subgroups maintained improvements in outcomes as per the study results overall, but those that did not understand verbal instructions showed no improvement in ADL, ease of care giving, or respiratory function. Even more telling, scoliosis surgery had no impact on QOL-related outcomes or respiratory function in the progressive disease subgroup in which the only significant improvement was in Cobb angle correction. The sample size in this subgroup however, was quite small (only 14 patients) so this should be interpreted with caution.

In a general sense, the results of these two prospective studies support the surgical treatment of scoliosis in neuromuscular diagnoses, citing significant improvements in QOL-related outcomes, radiographic measures and, in some cases, respiratory function. That said, an objective assessment of the

literature with respect to the risks involved with such procedures is required to balance the risk-benefit equation.

In a large retrospective study utilizing the Kids Inpatient Database (US based database of nationwide hospital discharges), 437 children with progressive neurodegenerative disorders (e.g. SMA, myopathies, etc.) who underwent scoliosis surgery were identified and their results were compared to non-progressive patients. In general, the progressive group had significant increases in length of stay (10.3 versus 7.7 days), pulmonary complications, in-hospital mortality (1.6 % versus 0.6 %), and hospital costs [14].

These results were similar to a study from the Scoliosis Research Society Morbidity and Mortality Committee, which analyzed complication rates from a database of 19,360 patients who underwent pediatric scoliosis surgery; 4657 of which had a neuromuscular diagnosis (including non-progressive and progressive diagnoses) [10]. Within this large sample, as compared to idiopathic scoliosis (IS), neuromuscular patients again had the highest risk of complications at 18 % versus 6 %. As with the previous study, mortality rates were significantly higher in the neuromuscular group (0.3 % versus 0.02 % for IS) with respiratory complications being the leading cause of death. Non-fatal complications such as blood loss (NM: 1.2 % versus IS: 0.2 %) and deep wound infection (NM: 3.8 % versus IS: 0.8 %) were also significantly higher in the neuromuscular group.

Continuing the trend, a systematic review analyzing the rate of complications in scoliosis surgery substantiated these results with significantly increased risks of death, infection, and pseudoarthrosis associated with a neuromuscular diagnosis when compared to IS [9].

The results of these studies confirm that the risks associated with scoliosis correction in neuromuscular diagnoses are substantial but, at the same time, the best evidence available also suggests that QOL is improved for these patients despite the risks. These points must be carefully considered and effectively communicated by the treating surgeon so the patient and/or caregiver can make an informed decision as to whether or not they proceed with surgery. Furthermore, any discussion regarding disease-specific risks and benefits needs to take into consideration the differences in outcomes that are beginning to emerge in the recent literature when analyzed by diagnosis.

## Cerebral Palsy

Though typically associated with high patient/care-giver satisfaction, scoliosis surgery in children with CP is fraught with high complication rates likely related to the increased prevalence of co-morbidities inherent to this patient population including: poor nutritional status, epilepsy, infections of the urinary and respiratory tract, feeding disorders, and relative immunodeficiency [23]. Despite these risks, children with CP

seem to be highly tolerant of spinal surgery with a relatively long predicted life expectancy post-operatively [24]. The evidence regarding the risks and benefits of scoliosis correction in CP, as in other diagnoses, is scant but a few studies did meet our inclusion criteria.

In a recent systematic review with an aim to determine the risks, benefits of scoliosis correction in CP, in addition to the pre-operative factors affecting surgical outcome, only 1 prospective and 3 retrospective cohort studies were identified, with the rest being retrospective case series. Unfortunately, none of these studies analyzed included an observational group and the conclusions of the review were that the “overall strength of the evidence was insufficient” to make any firm recommendations for or against surgical intervention [25]. The authors also revealed that outcomes in these studies were “poorly delineated with limited or no use of validated outcome instruments”. They suggested that future studies needed to employ validated outcomes relating to patient satisfaction and function.

Recently, the development of the Caregiver Priorities & Child Health Index of Life with Disabilities (CP CHILD) questionnaire by Narayanan and colleagues has provided a validated disease-specific outcome measure to apply to patients with CP [13]. In a prospective longitudinal cohort multi-center study investigating the utility of the CP CHILD questionnaire for children with severe CP who underwent scoliosis surgery, the authors found that by 12 months post-operatively, significant improvements in positioning/transfers, health, and overall QOL were achieved. The instrument was found to be sensitive to change and suggested as a meaningful outcome measure for evaluating this patient population [26].

These results were corroborated by a recent retrospective case-control study that also used the CP CHILD as its primary outcome measure [27]. In this study, children with severe CP (GMFCS IV and V) and scoliosis greater than 40 degrees were analyzed to determine the impact of scoliosis correction. The operative group demonstrated significant improvements in overall CP CHILD scores, personal care/activities of daily living, positioning/transferring/mobility, comfort/emotions, and communication/social interactions while the observational group deteriorated. In the surgical group, the complications included wound infections (22 %), pneumonia (17 %), reoperations due to post-surgical collections (12 %), pneumothorax (6 %), and recurrent hip dislocation (6 %).

Two other retrospective case-control studies also concurred with the findings of the previous studies, reporting a high level of patient and/or caregiver satisfaction but with a high rate of associated complications [28, 29].

In conclusion, given the best evidence available, it would seem that scoliosis correction improves quality of life in CP albeit with a high rate of complications.

## Spina Bifida

Spina bifida represents a spectrum of disease severity and patho-anatomical variations (including deficiencies of the bony posterior elements in conjunction with congenital abnormalities of the spinal cord, brain stem, and peripheral nerves) that make it a difficult entity to assess from a disease-specific standpoint. To this point, many previous studies that refer to ‘spina bifida’ as the diagnosis in question typically focus on patients with myelomeningocele rather than typically higher functioning forms of spinal dysraphism including meningocele, lipomeningocele and others. As in other neuromuscular disorders, this distinction is relevant since the incidence of scoliosis and risks associated with curve correction vary according to these differing manifestations of the ‘spina bifida’ diagnosis. For example, disease severity in myelomeningocele is commonly related to neurosegmental level, with the risk of scoliosis progression being more prevalent at higher motor levels (e.g. T12 and above (high risk) versus L5 and below (low risk) [18, 30]. Furthermore, QOL and functional scores have also been linked to neurosegmental level, a potential confounder when assessing the impact of interventions without taking disease severity into consideration [31].

Scoliosis is one of the most common musculoskeletal manifestations in spina bifida with up to 50 % prevalence being reported [3]. As curve magnitude increases, a progressive loss in truncal balance and sitting stability can occur which may impact QOL [32]. The indications for scoliosis correction in spina bifida have mirrored that of other neuromuscular diagnoses including a progressive curve greater than 50° and functional concerns such as sitting balance and wheelchair tolerance. However, the operative risks associated with spinal fusion have been reported to be among the highest for scoliosis of any diagnosis which necessitates a comprehensive assessment of the benefits and risks of surgery, and their effective communication to patient and/or caregiver, before embarking on such a procedure.

Until recently, there were no validated outcome measures to evaluate the impact of scoliosis correction of QOL and patient satisfaction in spina bifida despite complication rates approaching 75 % [33]. To rectify this, Wai and colleagues developed the Spina Bifida Spine Questionnaire (SBSQ), a validated tool that evaluated self-perception and overall physical function for these children with associated spinal deformity [34]. Interestingly, in this cross-sectional study, the authors found no relationship between spinal deformity and self-perception or physical function.

In a follow-up retrospective case-control study involving the same institution, the SBSQ and 36-Item Short Form Health Survey (SF-36) were used to assess the impact of scoliosis correction on QOL in patients with spina bifida at a mean 14-year follow-up [35]. Like the previous study, they

found no difference in SBSQ and SF-36 scores between the operative and non-operative groups and no relation to curve magnitude. In addition, the study showed that, of the patients that could walk pre-operatively, only 50 % remained ambulatory after scoliosis correction. The authors suggest that this finding, coupled with an increased spinal stiffness after fusion and a lack of improvement in sitting balance between the groups, may have contributed to the lack of impact on QOL.

A prospective study from Poland also investigated the relationship between QOL/functional scores and the presence of spinal deformity in spina bifida in a non-operative cohort [32]. They used the Quality of Life in Spina Bifida Questionnaire (QLSBQ) to assess QOL and the SBSQ to assess physical function in addition to a self-perception outcome measure. They found that very large curve magnitudes did have a negative affect on QOL but no impact on physical function or self-perception was identified. Since there was no surgical arm in the study, no inference can be made as to whether surgery would improve these QOL limitations.

A recent evidence-based review on the subject identified 9 level III studies involving the spine but only two that evaluated physical function within both operative and non-operative groups [36]. The authors concluded that surgery had little effect on physical function and cautioned that the risks may outweigh the benefits when considering scoliosis correction for children with spina bifida. If surgery was to be done, a combined anterior and posterior fusion was reported to have a decreased complication rate over other approaches.

As previously mentioned, complication rates associated with scoliosis correction in spina bifida are very high and are an important consideration. In one retrospective comparative study investigating complication rates associated with different surgical approaches, an overall complication rate of 48–60 % was found with infection (19 %), shunt insufficiency (12 %), pseudoarthrosis (22 %), and hardware-related problems (30 %) being the most notable [33]. They also found that the addition of anterior instrumentation and fusion to a posterior fusion provided the lowest rate of hardware-related complications and loss of correction. Furthermore, one death from shunt insufficiency was identified and as such the authors stressed that shunts should be evaluated pre-operatively to help decrease the rates of peri-operative malfunction.

In summary, unlike CP, current evidence suggests that the risk-benefit ratio for scoliosis correction in spina bifida is not favourable given the lack of significant improvements in QOL or physical function coupled with very high complication rates. As such, unless a substantial functional problem (e.g. severe pain from costo-pelvic impingement; functional sitting imbalance) is identified which is unresponsive to conservative management (e.g. wheelchair modifications), scoliosis surgery should not be recommended for children with spina bifida.

## Duchenne Muscular Dystrophy

Duchenne Muscular Dystrophy (DMD) is an inherited X-linked myopathic disorder secondary to mutations in the gene coding for dystrophin, a muscle cell membrane stabilizer. The absence, or reduced action, of dystrophin renders muscle cells susceptible to damage that is thought to result in an inflammatory response with eventual replacement of viable muscle with fibro fatty scar tissue [37]. As a result of this process, unlike CP and other non-progressive disorders, the natural history of DMD is typified by progressive deleterious changes in muscle strength, static contractures, and the development of scoliosis. Concomitant to these changes, progressive decreases in respiratory and cardiac function, along with a significantly reduced life span, increase the risks associated with the correction of scoliosis in this population.

Key questions regarding scoliosis correction in DMD center around whether it: (1) improves long-term survival, (2) improves respiratory function, (3) improves QOL and overall physical function, (4) has a positive benefit-to-risk ratio, and (5) is still required given the potential for scoliosis prevention or delay in onset by the use of newer corticosteroids with fewer side effects.

In an attempt to answer some for these questions, a recent Cochrane review was published which investigated the role of scoliosis correction in DMD [38]. Expectedly, no randomized controlled studies were identified and the authors' conclusions were that the available literature was insufficient to make any direct recommendations regarding the application of scoliosis correction in DMD. However, based on the available literature, some of which was at least level III, a few general statements were offered.

Most of the studies reviewed were in agreement that spinal surgery improved sitting position, patient satisfaction, and overall QOL, in addition to improvements in Cobb angle and pelvic obliquity for children with DMD. However, most failed to show significant improvements in respiratory function or long-term survival despite the many studies investigating this over the past 35 years [39–42]. In fact, one retrospective case-control study cited found no significant differences in respiratory function deterioration between operative and non-operative groups. Furthermore, the addition of surgery did not improve respiratory function despite significant Cobb angle correction (61.7 %) [43].

The published complication rates were significant as in other neuromuscular disorders. In a recent level III study comparing complication rates for scoliosis surgery in DMD and CP, it was suggested that DMD involves even higher perioperative risks with significantly higher complication rates (DMD: 38 % vs CP: 18 %) [44]. A systematic review from Mercado and colleagues supported these statements suggesting that cosmesis, QOL, and overall patient satisfaction are generally improved after scoliosis correction in

**Table 24.2** GRADE recommendations for scoliosis correction

Evidence-based statement	GRADE recommendation
The risks of scoliosis correction for neuromuscular diagnoses are significantly more than for idiopathic diagnoses	B
Scoliosis correction in neuromuscular diagnoses, on the whole, improves QOL, radiographic measures and, in some cases, respiratory function	B
Scoliosis correction improves quality of life in CP	B
Scoliosis correction does not improve quality of life or physical function in spina bifida and has a very high risk profile	B
Scoliosis correction improves quality of life and patient satisfaction in DMD albeit with a high complication rate	B
Scoliosis correction does not improve or preserve respiratory function in DMD	B
The use of deflazacort significantly reduces the need for scoliosis correction in DMD	B

DMD despite the fact that most patients undergo surgery at a relatively early stage (e.g. 25° Cobb angle) [3]. Given the lack of controlled studies however, the authors stated that the relationship between spinal surgery and improvement in respiratory function is still unclear. They also cautioned that a consistent negative aspect was the adverse impact on self-feeding due to the combination of a stiff, straight spine and upper limb weakness post-operatively.

Recently, it has been suggested that perhaps scoliosis surgery in DMD might be completely avoided. In the absence of effective medical management, it has been reported that over 90 % of patients with DMD will develop scoliosis, most of which will require spinal stabilization [45, 46]. Previous studies have shown that corticosteroid administration can slow the decline in muscle strength, prolong walking in children, and delay the onset of scoliosis with DMD but often at the expense of unacceptable side effects [47]. However, several recent studies have demonstrated that deflazacort (an oxazolone derivative of prednisone) can achieve similar improvements in function with substantially better side effect profile.

In a prospective cohort study investigating the role of deflazacort in the development of scoliosis, the Kaplan-Meier survival rate of not developing scoliosis or needing spinal surgery was 78 % in the treatment group and 8.3 % in the non-treatment group at a mean follow-up of 15 years [48]. Scoliosis measuring 20° or more developed in 20 % of patients for the treatment group and in 92 % for the non-treatment group, all of which required surgery. Of further benefit, patients in the treatment group had significantly improved pulmonary function, prolongation of walking, and ability to climb stairs. Side effects were common with 70 % (21 patients) in the treatment group developing cataracts but only two patients required cataract surgery. Other side effects included decreased height (17 cm shorter), weight gain (55 kg vs 51 kg), and no difference in bone fractures (treatment group was given bisphosphonates). The authors concluded that glucocorticoids have a long-

term protective effect against the development of scoliosis in DMD.

Two additional retrospective cohort studies investigating the role of deflazacort in DMD supported the findings above [46, 49]. In each of these studies, the treatment group showed a similar decrease in the need for spinal surgery with an acceptable side effect profile. In one of these cohorts with a mean 8-year follow-up, surgery was completely avoided in the treatment group while 43 % of the non-treatment group underwent surgery [46]. In addition to prolonged walking, this study also reported significant improvements in cardiac function.

Given the studies above, it would seem that the use of deflazacort prevents, or at least delays, the onset of scoliosis in DMD with secondary benefits of preserving respiratory and cardiac function with an acceptable side effect profile. When surgery is required, it can be expected to result in improvements in QOL and patient satisfaction albeit with high perioperative risks. At this time, the available evidence regarding scoliosis correction in DMD does not seem to support improvements in survival and/or respiratory function.

Evidence-based statements and GRADE recommendations for this section are provided in Table 24.2.

## What Is the Best Choice of Spinal Fixation?

In previous sections, it was established that the goals of surgical management for neuromuscular scoliosis typically involved the achievement of a balanced spine over a level pelvis via a spinal fusion extending from the upper thoracic spine to the lower lumbar spine and/or the pelvis. Due to the high rate of vertebral osteopenia in neuromuscular patients, segmental fixation is typically the rule to help prevent implant-related complications. The choice of spinal fixation used to achieve these goals has evolved over the years from Harrington instrumentation to Luque sublaminar wires and, more recently, to segmental pedicle screw fixation [1, 2, 50].



Proponents of pedicle screw fixation cite improvements in Cobb correction, a decreased need for anterior release, and a favourable risk profile over earlier methods despite the associated increase in implant costs [51]. Proponents of sublaminar fixation cite the ability to achieve the desired goals, albeit with a smaller Cobb correction (~50 %), decreased costs, and comparable complication rates [52]. Although most of the literature in this area is comprised of uncontrolled case series, several level III studies were identified and available for review.

### Hybrid Versus Pedicle Screw Fixation

Regarding the assessment of spinal fixation methods in neuromuscular scoliosis, our literature search revealed no prospective studies and a small number of retrospective comparative studies. One such study compared hybrid fixation (sublaminar wires/hooks in the thoracic spine, pedicle screws in the lumbosacral spine) to segmental pedicle screw fixation and found significant improvements in curve correction (75 % vs 59 %), operating time (6.0 vs 7.4 h), blood loss (1785 vs 3760 mL), and the need for anterior surgery (12 % vs 40 %) for the pedicle screw group as compared to the hybrid group [53]. As might be expected, the authors reported no change in QOL as measured by the SRS-24 questionnaire and no significant difference in overall complication rate between the two groups. They stressed that, although Cobb correction was greater in the pedicle screw group, the attainment of spinal balance was most important and was achieved by both groups.

In another retrospective comparative study with 44 of 68 patients having a neuromuscular diagnosis, four different types of apical spinal fixation were compared for large curves greater than 100°: sublaminar wires, hooks, anterior vertebral screws, and all pedicle screw constructs [5]. Like the previous study, all pedicle screw constructs demonstrated significant improvements in Cobb correction, a decreased rate of anterior release, and the lowest complication rate between the groups. The all-pedicle screw group was also better at maintaining curve correction by final follow-up as compared to other methods, which tended to be associated with a significant loss of curve correction over time. That said, there were no significant differences in the ability to achieve coronal/sagittal balance or in the neurologic risk profile between the wire and screw groups.

A third retrospective cohort comparing ‘rigid’ constructs (greater than 50 % pedicle screw fixation) against ‘non-rigid’ constructs (greater than 50 % sublaminar fixation) in neuromuscular scoliosis correction supported the claim that pedicle screw constructs achieved greater Cobb angle correction and had a decreased need for anterior release as compared to

sublaminar wires [54]. The pseudoarthrosis rate was also significantly less in the rigid group (5 % vs 22 %). The authors concluded that, despite a fivefold increase in implant costs, the overall charges associated with the higher rates of pseudoarthrosis in the wire group would more than offset any implant-related differences in cost. They suggested that there was a need for a future study that incorporates a more comprehensive economic analysis to more fully elucidate the true value of using pedicle screws over cheaper implants such as sublaminar wires.

Lending further support to the evidence above, in a retrospective study investigating the use of sublaminar (SL) wires, hybrid (H) constructs, and segmental pedicle screw (PS) fixation in patients with DMD, significant increases in operative time, blood loss, and Cobb correction were again identified in the SL group [55]. The increased Cobb correction was likely related to the increased pre-operative Cobb angle in the wire group as compared to the other groups (50° (SL) vs 18° (H), 26° (PS)) rather than improved mechanical capabilities. The authors suggested that the increased blood loss in the SL group was likely due decreased vasoconstriction in DMD and epidural vessel injury during wire passage. In this study, based on radiographic and intra-operative outcomes, it would seem that pedicle screw constructs performed most favourably as compared to other implants.

### Newer Implants

Regarding the Universal Clamp (Zimmer, Warsaw, USA), a relatively new titanium implant which incorporates a sublaminar polyester tape, only one comparative study was identified which compared its use to the standard sublaminar wire in scoliosis correction [6]. This cohort of 50 patients (25 in each group) showed no differences in Cobb correction and no change in operative time or blood loss between the two groups. The costs, though not stipulated in the study, are known to be quite high for the Universal Clamp as compared to the stainless steel sublaminar wire, leaving the sole reported benefit to be related to the MRI compatibility of the titanium implant. Additional studies comparing the Universal Clamp to other types of spinal fixation will be required to ascertain its place in the surgeon’s armamentarium.

Given the above discussion, it would seem that segmental pedicle screw fixation provides the best Cobb correction, decreased operative time, and a decreased need for anterior release, as compared to hooks or sublaminar wires. Prospective studies that incorporate comprehensive economic analyses would be beneficial to assess the outcomes-to-cost ratio as it pertains to the most appropriate choice of spinal fixation for these children.

Evidence-based statements and GRADE recommendations for this section are provided in Table 24.3.

**Table 24.3** GRADE recommendations for choice of spinal fixation

Evidence-based statement	GRADE recommendation
The use of segmental pedicle screw fixation in neuromuscular scoliosis results in the largest Cobb angle correction as compared to other methods	B
The use of segmental pedicle screw fixation in neuromuscular scoliosis results in a reduced rate of anterior release as compared to other methods	B
The use of segmental pedicle screw fixation in neuromuscular scoliosis results in a reduced operative time over other methods	B
The use of segmental pedicle screw fixation in neuromuscular scoliosis results in a reduced rate of complications over other methods	I
Quality of life improvements are unrelated to the choice of spinal fixation in neuromuscular scoliosis	I

### What Is the Best Choice of Pelvic Fixation?

In neuromuscular scoliosis, reducing pelvic obliquity is one of the major technical goals of spinal correction. Coupled with an unbalanced spine, its presence can adversely affect sitting balance, wheelchair tolerance, weight distribution, and QOL [56]. Pelvic obliquity correction is typically achieved by instrumentation and fusion to the sacropelvis through various implant types and configurations. There has been some controversy as to the indications for fusion to the pelvis but, when required, achieving stable fixation to the sacropelvic unit that resists post-operative loss of correction is essential to surgical success [56–59]. Although a myriad of implant configurations for the treatment of pelvic obliquity have been described, this section will focus on three of the most prevalent types of pelvic fixation: (1) Galveston, (2) iliac screws, and (3) sacral alar iliac (SAI) screws (Fig. 24.2).

### Galveston Versus Iliac Fixation

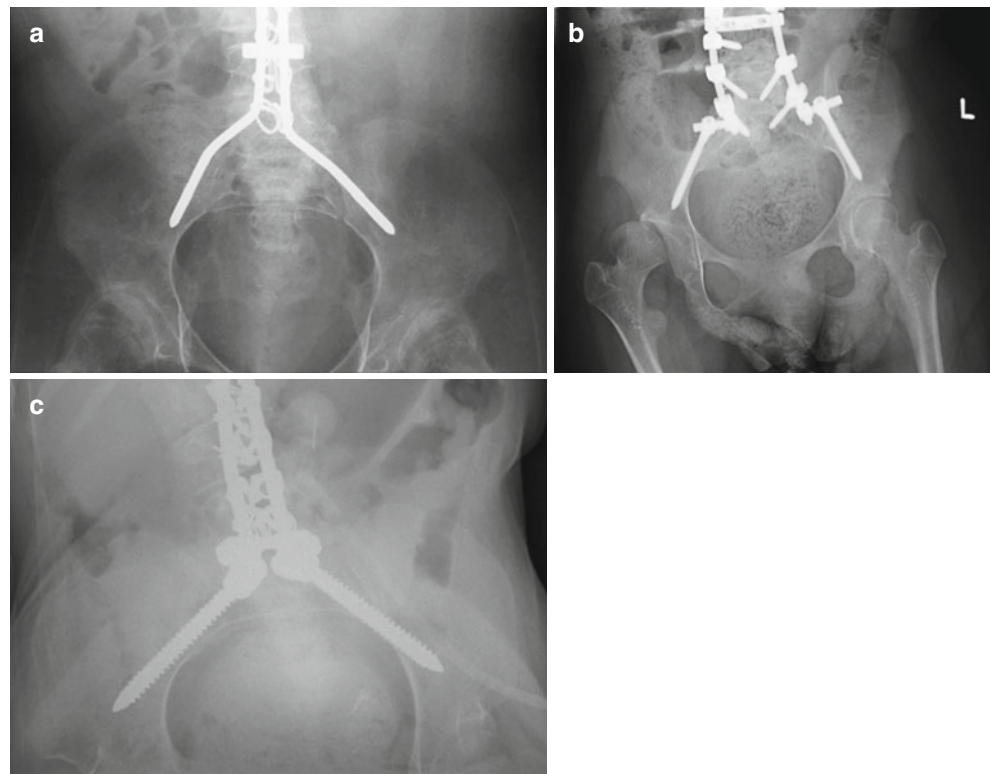
Galveston bilateral iliac fixation is one of the best-known techniques for the treatment of pelvic obliquity in neuromuscular scoliosis although its use has been waning in favour of more modern techniques utilizing screw fixation. Typically, it has been utilized in conjunction with Luque segmental sublaminar wire fixation in the thoracic and lumbar spine and has evolved as a system incorporated into a single, pre-contoured ‘unit rod’ [60, 61]. The technique has been well described elsewhere but in essence, involves the insertion of stainless steel smooth tines into tracts made between the tables of the ilia bilaterally.

Iliac screw fixation takes a similar approach but substitutes long screws for the tines described for the Galveston technique. It has been suggested that the use of screws over smooth tines may diminish implant pullout and loss of pelvic obliquity correction [62]. A frequent need for lateral offset connectors, however, has been reported to increase pullout forces rather than decrease them and serves as an additional potential point of failure [63]. Clearly, comparative studies are needed to ascertain whether or not these ‘more advanced’ systems actually improve outcomes. Despite an exhaustive literature search, very few studies with comparator groups and/or adequate sample size were identified [64].

In a retrospective cohort of forty patients with neuromuscular diagnoses, radiographic outcomes and complication rates between Galveston and iliac screw pelvic fixation were compared to assess superiority of one technique over the other [62]. Each patient underwent the standard posterior instrumentation and fusion from the upper thoracic spine to the pelvis with no differences in curve correction or apical vertebral translation between the groups. In addition, an equal number of patients in each group underwent intra-operative halo femoral traction. The study showed no difference in initial pelvic obliquity correction between the two pelvic fixation groups. At a final follow-up of 3 years however, the percentage of patients with residual pelvic obliquity greater than 10° was significantly increased in the Galveston group; a fact the authors claimed was not clinically significant given that both groups achieved “an excellent overall correction”. In addition, pelvic anchor motion (‘wiper blading’) was significantly increased in the Galveston group but with uncertain clinical significance. The complication rates were similar overall. The authors concluded that iliac screws are equivalent to the Galveston technique but the former allows for sacral and lumbar screw fixation that may offer better construct stability with the downside of having an increased implant profile.

Despite these claims, the Galveston technique may still prove to be more efficacious over more modern implants. In a multicenter retrospective study of 157 children with CP comparing the pre-contoured unit rod (including Galveston pelvic fixation) to custom-bent rods utilizing various methods of pelvic anchorage (including iliac screws or rods, S-rods, and sacral screws), pelvic obliquity was found to be significantly improved in the unit rod group (74 % vs 22 %) with no significant differences in operative time or intraoperative blood loss [65]. The authors offered that the likely reason for the improvement could be explained by the fixed 90° angle between the unit rod’s pelvic limbs and spinal rods. Transfusion requirements, implant prominence, as well as inpatient and intensive care unit length of stays, were significantly increased in the unit rod group. Accordingly, results from the non-validated caregiver-reported outcome measure used in the study suggested a higher satisfaction rate in the custom-bent rod group.

**Fig. 24.2** Types of pelvic fixation. (a) Galveston, (b) Iliac screws with lateral offset connectors, and (c) Sacral alar iliac (SAI) screws



One potential advantage of the use of iliac screw fixation is that it allows for augmentation with additional implants to theoretically improve construct stability. In a retrospective study with 5-year follow-up, two pelvic fixation constructs utilizing 1 or 2 iliac screws were compared with respect to pelvic obliquity correction and associated complications [66]. The single screw group experienced 2.5 times greater rod dislodgement than the double screw group but this did not reach statistical significance. However, a sevenfold increase in proximal (i.e. thoracic and lumbar spine) implant failure was seen in the single screw group. The authors surmised that the extra screw provided a more secure base that dampened proximal motion and thus reduced this risk. The overall pelvic obliquity correction was reported to be 59 % (from 18.8° to 7.6°) with no mention of correction stratified by treatment group. Given the decreased complication rate, the authors recommended the adoption of the double screw technique over the single screw for neuromuscular pelvic fixation.

### Sacral Alar Iliac Fixation

Recently, a new pelvic screw fixation technique was described that utilized a tract traversing the sacral ala, sacroiliac joint, and ilium [67]. The proposed advantages of this sacral alar iliac (SAI) screw fixation over iliac screws were reported to be due to its low profile and decreased need for both lateral offset connectors and the associated subparaspinal muscle dissection required. In a retrospective

comparative study with 2-year follow-up, SAI screws demonstrated significant improvements in pelvic obliquity correction over an iliac screw group (70 % vs 50 %, respectively) with no difference in implant-related complications or pain between the groups. The authors suggested that the improvement in pelvic obliquity correction might have stemmed from a better mechanical advantage in manipulating the pelvis directly rather than working through the lateral offset connectors used in the iliac screw group. Additional studies investigating patient-specific outcomes would be beneficial to further elucidate the clinical superiority of SAI screws in pelvic fixation.

### Choice of Surgical Approach

The choice of surgical approach may have an impact on the success of pelvic fixation. In a retrospective comparative study analyzing 54 patients with muscular dystrophy, one group underwent a posterior-only approach while the second underwent a combined anterior and posterior approach for the correction of pelvic obliquity [68]. The anterior approach chosen involved an extraperitoneal midline lumbosacral release and fusion with mesh cage and allograft. The anterior-posterior group achieved a significant improvement in pelvic obliquity correction over the posterior-only group (21° vs 13°, respectively) but at the expense of significant increases in intraoperative blood loss (2.4 L vs 345 mL, respectively) and operative time (611 vs 440 min, respectively). Since the study only evaluated radiographic parameters, rather than

sitting tolerance or other patient-specific outcomes, the clinical significance of this modest improvement in pelvic obliquity is not known and may not outweigh the additional risks imparted by the anterior approach.

### Fusion to Lumbar Spine Versus Pelvis

The indications for pelvic fixation in neuromuscular scoliosis correction have been controversial with some authors calling for all non-ambulatory patients to be instrumented and others only in the face of substantial pelvic obliquity [2, 69]. Reasons to exclude the pelvis have included: minimal preoperative pelvic obliquity, increased operative time, potential loss of walking ability, and higher complication rates [56, 70].

In support of these reasons, the extent of caudal fixation for patients with and without pelvic obliquity was investigated in a retrospective cohort of 55 patients with neuromuscular scoliosis [69]. The cohort was stratified into 3 groups according to the distal fixation level and the severity of pelvic obliquity including: (1) pelvic obliquity greater than 15° with pelvic fixation, (2) pelvic obliquity greater than 15° without pelvic fixation, (3) pelvic obliquity less than 15 degrees with pelvic fixation. Pelvic obliquity correction was found to be significant for all groups but Group 2 displayed a significant loss in correction at final follow-up. The authors concluded that the presence of pelvic obliquity greater than 15° necessitates the addition of pelvic fixation while lesser amounts remain stable without fixation.

Lending further support to the exclusion of pelvic fixation for patients with minimal pelvic obliquity, a retrospective study of 36 patients with DMD compared two groups according to the magnitude of tilt [70]. In this cohort, pelvic obliquity was found to be (1) more than 15° in 10 patients and (2) less than 15° in 26 patients. In Group 1, pelvic tilt was improved by 62 % using iliac screw fixation. Group 2 also experienced stable pelvic tilt correction by 42 % even without pelvic fixation at a final follow-up of 37 months. As for complications, 24 % in Group 2 had postoperative coccygodynia compared with only 10 % in Group 1. The authors concluded that instrumentation of the pelvis is unwarranted for pelvic obliquity less than 15°.

Given the above discussion with respect to pelvic fixation, the evidence seems to support the use of the Galveston technique, a posterior-only approach, and the exclusion of pelvic fixation for patients with pelvic tilt less than 15°. However, the available evidence is based on a limited number of studies with small sample sizes and, as such, more studies are required to make definitive conclusions regarding pelvic fixation in neuromuscular scoliosis.

Evidence-based statements and GRADE recommendations for this section are provided in Table 24.4.

**Table 24.4** GRADE recommendations for choice of pelvic fixation

Evidence-based statement	GRADE recommendation
For pelvic obliquity correction, iliac screw fixation is not superior to Galveston fixation	B
For pelvic obliquity correction, sacral alar iliac fixation may be superior to iliac screw fixation	I
Pelvic fixation is not be required for pelvic obliquity less than 15 degrees	B

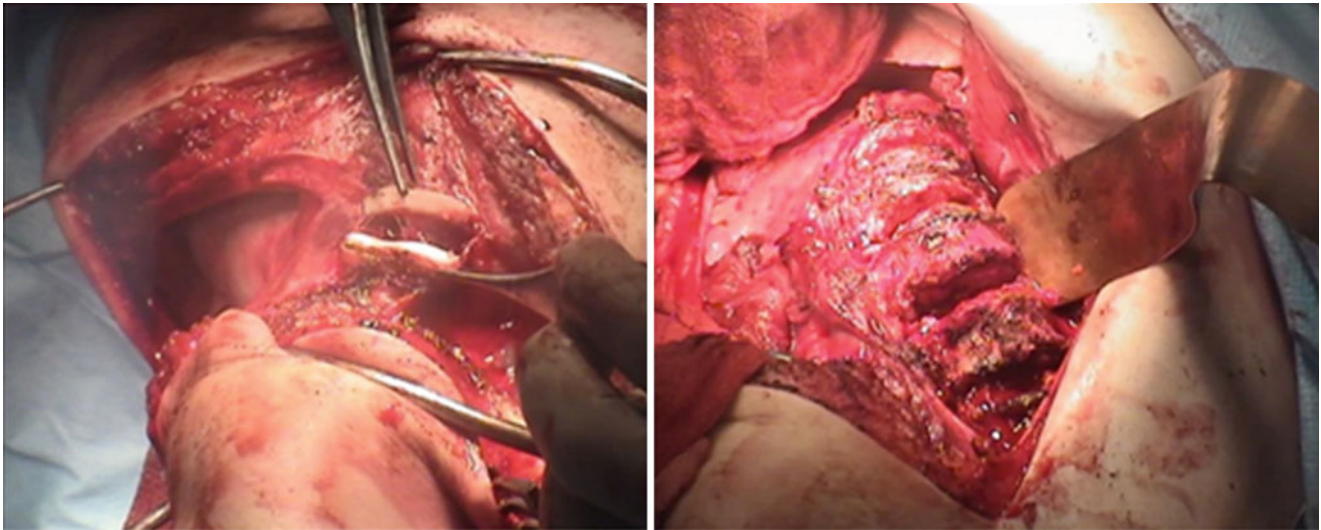
### What Is the Evidence for Anterior Fusion?

The indications for anterior release for neuromuscular scoliosis have traditionally been described for curves with residual magnitude greater than 50–70° on bending or traction radiographs and for the prevention of crankshaft in immature patients [71]. The rationale behind the anterior approach is twofold: (1) to improve the capacity for correction in large, stiff curves by releasing the anterior longitudinal ligament and performing discectomies at multiple levels about the apex of the major curve (Fig. 24.3), (2) to facilitate the development of an anterior fusion to reduce the risk of pseudoarthrosis [24]. It was believed that the addition of an anterior fusion allowed for a better curve correction than posterior fusion alone. With the advent of ‘heavy constructs’ utilizing segmental pedicle screw fixation, in addition to the use of peri-operative adjuncts such as skull-femoral traction, the routine use of the anterior approach has been challenged. Though few in number, several studies that focused in this area and met our inclusion criteria were identified for review.

### Anterior Surgery: Risk to Benefit Balance

Like any surgical intervention, a comprehensive assessment of the risk-benefit balance is paramount, particularly for procedures that violate the thoracic cage in a patient population that is already predisposed to pulmonary complications and have significant co-morbidities. Indeed, the risks of anterior surgery are substantial. In a large retrospective comparative study comparing risks of anterior surgery as a function of diagnosis, children with CP were more likely to have a major complication, most of which were pulmonary, as compared to spina bifida (52 % vs 41 %, respectively). In addition, curve size greater than 100° was found to be a significant risk factor for a major complication [72].

Another retrospective study assessing the risks pertaining to surgical approach for children with neuromuscular scoliosis, reported significantly longer intensive care unit (ICU) stays and increased pulmonary complications with combined



**Fig. 24.3** Multilevel anterior discectomies via thoracoabdominal approach for severe scoliosis and pelvic obliquity in a 15 year-old with spastic quadriplegia

anterior-posterior fusions as compared to posterior instrumentation and fusion (PSF) alone [73].

Despite the findings presented above, there are instances where the benefits of an anterior fusion may very likely outweigh the potential risks. In a systematic review investigating the role of spine surgery in spina bifida, it was found that curve correction and fusion rates were significantly improved when an anterior fusion was added to the standard PSF [36]. Specifically, the risk of pseudoarthrosis was reported to be 46 % for PSF only compared to 14–23 % for combined anterior and posterior fusion. Wright assigned a B GRADE recommendation for the combined approach but stated there was insufficient evidence to recommend that the addition of anterior instrumentation improved outcomes further.

For children with neuromuscular scoliosis who, for whatever reason, require the addition of anterior fusion, the question of whether to do both procedures staged or same day is a topic of interest. In a retrospective comparative study involving children with spastic quadriplegic CP, significant increases in blood loss and operative time for single stage combined anterior-posterior fusion compared to performing the anterior and posterior procedures in two stages were demonstrated [24]. Based on this and other studies, it would seem that, although there are substantial risks associated with the combined approach, staging the procedures might mitigate the risk to some extent. Given the contradictory results of other comparative studies in this area, however, the evidence is likely insufficient to lend definitive guidance [74–76].

As outlined above, one of the most common reasons to perform an anterior release is to improve curve flexibility and accordingly, the potential for increased curve correction

over PSF alone for large, stiff curves. A retrospective case-control study investigated the legitimacy of this practice by comparing matched groups of children with CP and scoliosis that underwent PSF with or without an anterior release [77]. The PSF-only group was augmented with intraoperative skull-femoral traction. There were no significant differences in curve correction, coronal/sagittal balance, or pelvic obliquity correction, between the groups. There were however, significantly increased complication rates demonstrated in the anterior release group including increased operative times, blood loss, postoperative intubation, and pneumonias. There were no traction-related complications reported. The authors concluded that the addition of an anterior release did not improve radiographic outcomes as compared to PSF (with skull-femoral traction) alone and demonstrated a significantly higher complication rate.

### The Role of Intra-operative Skull-Femoral Traction

Although the preceding study suggested that curve correction was equivalent for both combined approaches and PSF alone, the addition of skull-femoral traction in the preceding study represented a significant confounding factor given that the technique has been shown to significantly improve major curve correction and pelvic obliquity when coupled to PSF versus PSF alone [78]. Therefore, one cannot conclude that scoliosis correction via a combined approach is equivalent to PSF alone without traction.

In recent years, intra-operative halo-femoral traction has become more commonplace, particularly for stiff curves, and

**Table 24.5** GRADE recommendations for the role of anterior fusion

Evidence-based statement	GRADE recommendation
Overall, anterior release in addition to posterior fusion does not provide better outcomes over posterior fusion alone	I
The combined use of anterior and posterior fusion results in superior outcomes to posterior fusion alone in spina bifida	B
The risks associated with the combined use of anterior and posterior fusion are higher than for posterior fusion alone	I
Staging anterior and posterior fusion for combined approaches is associated with a decreased complication rate compared to single stage surgery	I
The use of skull-femoral traction as an adjunct to PSF may provide equivalent curve correction to the combined use of anterior and posterior fusion	C
The use of skull-femoral traction as an adjunct to PSF reduces the need for anterior release in neuromuscular scoliosis	I

is often combined with PSF following an anterior release. A recent systematic review on the subject suggested that the use of intraoperative skull-femoral traction may improve radiographic outcomes, decrease complication rates, and reduce the need for anterior procedures in neuromuscular scoliosis surgery [79]. The included studies in this review, however, were mixed in diagnosis and surgical approaches, and were marred by small sample sizes. As such, these recommendations should be interpreted with caution.

Overall, the availability of level III or better evidence studies concerning the role of anterior surgery in neuromuscular scoliosis is scarce and definitive conclusions in this area remain elusive. Given the available evidence, however, it is reasonably well established that the addition of anterior fusion to PSF in spina bifida leads to improved outcomes. Lesser evidence is available regarding the use of skull-femoral traction and its ability to obviate the need for anterior release in stiff curves.

Evidence-based statements and GRADE recommendations for this section are provided in Table 24.5.

### Conclusions

Given the results of this evidence-based review of the best available literature, it would seem that for most cases, neuromuscular scoliosis correction is a worthwhile procedure with expected improvements in function, quality of life, and patient/caregiver satisfaction. These positive benefits likely outweigh the high surgical risks associated with these procedures but the ultimate decision as to whether or not scoliosis surgery is performed should be dependent on a disease-specific assessment of risks and benefits and appropriate communication with the patient and/or caregiver. The use of segmental pedicle screw fixation may offer improved outcomes over other methods of spinal fixation but the best choice of pelvic fixation is still controversial. Regarding the use of anterior surgery for neuromuscular scoliosis, its role is also controversial except in the case of spina bifida where it is likely to reduce perioperative risks.

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