Cochrane Review (Group): Surgery for scoliosis in Duchenne muscular dystrophy

D KL Cheuk¹, V Wong¹, E Wraige², P Baxter³, A Cole⁴

Abstract

Introduction
Scoliosis in people with Duchenne muscular dystrophy is usually progressive and treated with surgery. However, it is unclear whether the existing evidence is sufficiently scientifically rigorous to support a recommendation for spinal surgery for most people with Duchenne muscular dystrophy and scoliosis. This is an updated review and an updated search was undertaken in which no new studies were found. The aim of this systematic review was to determine the effectiveness and safety of spinal surgery in people with Duchenne muscular dystrophy with scoliosis. We intended to test whether spinal surgery is effective in increasing survival, improving respiratory function, improving quality of life and overall functioning; and whether spinal surgery is associated with severe adverse effects.

Materials and methods
We searched the specialized registers of the Cochrane Neuromuscular Disease Group (31 July 2012), MEDLINE (January 1966 to July 2012), EMBASE (January 1947 to July 2012), CENTRAL (2012, Issue 7 in the Cochrane Library), CINAHL Plus(January 1937 to July 2012), Proquest Dissertation and Thesis Database (January 1980 to July 2012), and the National Institute of Health Clinical Trials Database (July 2012). No language restrictions were imposed. We planned to include controlled clinical trials using random or quasi-random allocation of treatment evaluating all forms of spinal surgery for scoliosis in people with Duchenne muscular dystrophy in the review. The control interventions would have been no treatment, non-operative treatment, or a different form of spinal surgery. Two authors independently examined the search results and evaluated the study characteristics against inclusion criteria to decide which ones would be included in the review.

Results
On searching, 47 studies were relevant but none met the inclusion criteria for the review, because they were not clinical trials but prospective or retrospective reviews of case series.

Conclusion
Since there were no randomized controlled clinical trials available to evaluate the effectiveness of scoliosis surgery in people with Duchenne muscular dystrophy, no evidence-based recommendation can be made for clinical practice. People with scoliosis should be informed about the uncertainty of benefits and potential risks of surgery for scoliosis. Randomized controlled trials are needed to investigate the effectiveness of scoliosis surgery, in terms of quality of life, functional status, respiratory function and life expectancy.

Introduction
Duchenne muscular dystrophy (DMD) is an inherited X-linked muscular dystrophy caused by mutations in the dystrophin gene. It is characterized by progressive dystrophic changes in skeletal and cardiac muscle. Progressive weakness in affected children results in loss of ambulation at a mean age of 9.5 years⁴,⁵. There is progressive cardiomyopathy and respiratory failure occurs secondary to respiratory muscle weakness. The mean survival in the absence of ventilatory support is 19.5 years⁶. In 90% death is the result of respiratory failure and in 10% the result of cardiac involvement. Currently there is no proven effective curative treatment for this debilitating disease. A systematic review has found that glucocorticoid therapy improves muscle strength and function in the short-term. However, adverse effects were common and long-term benefits are uncertain⁷⁸. Spinal deformity, especially scoliosis, is progressive in the majority of people with DMD⁹,⁰,¹¹. From the onset of spinal deformity, progression can be extremely rapid and impair unsupported sitting ability and further compromise the respiratory and cardiac function. Kurz observed a 4% decrease in vital capacity for every 10% progression of the spinal curve in people with DMD¹². Galasko found that on average, vital capacity decreases by 8% per year in patients with scoliosis secondary to DMD¹³. Long-term corticosteroid treatment may slow the progress of scoliosis in people with DMD and may reduce the need for surgery, but adverse effects are frequent⁴⁸. Non-operative treatment such as bracing might not prevent the progression of this kind of spinal deformity because of the progressive nature of the underlying muscle disease⁷,⁹⁹.

Therefore, non-operative treatment is usually considered only in exceptional cases when a person refuses surgery or when a person has a very advanced deformity with poor general health.⁵²,⁵₄,⁶₁. Spinal fusion surgery with instrumentation remains the mainstay of treatment for people with DMD with scoliosis. Commonly used techniques are either based on sublaminar

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segmental wiring, such as Luque instrumentation, or the modern variants based on segmental pedicle screw and hook fixation such as Isola, TSRH or Universal Spine system. Two stainless steel or titanium rods are contoured to the desired spinal shape, and the spine reduced onto the rods, either with the sublaminar wires or segmental screws and hooks. Pelvic fixation is rarely required in DMD scoliosis and the Galveston technique of rod insertion into the ileum, or more modern screw fixation can be used in some circumstances.

Postoperative bracing is not required with modern fixation techniques. The potential advantages of surgery described in the literature include increased comfort and sitting tolerance,3,7,24,26,29,30,36,38,41, cosmetic improvement3,5, no need for orthopaedic braces3,4,9,62,63, easier nursing care by parents3 and pain relief3,29,53. Nevertheless, the effects of spinal surgery on respiratory function and life expectancy are still controversial. Some studies reported that spinal fusion had no effects on the natural deterioration of respiratory function of people with DMD22,28,30,41, at short-term and five-year follow-up.29 In contrast, several studies3,14,38,46 reported stabilization of vital capacity in people surgically treated for two to eight years.

Regarding life expectancy, Galasko observed a lower mortality in people surgically treated13,14. However, other studies reported that spinal surgery did not improve life expectancy9,15,21,22,28. Adverse effects and complications during and after surgery are not uncommon, including ventilator-associated pneumonia (iatrogenic, in the post-operative period), wound dehiscence, surgical wound infection, haemorrhage, loosening of fixation, pseudarthrosis, deteriorated respiratory function and increased difficulty with head to head motions. A randomized trial has demonstrated that although tendon surgery in people with DMD may correct deformities, it might also result in more rapid deterioration of function in some patients and there were no beneficial effects on strength or function.59 With increasing use of non-invasive ventilation (NIV) in DMD patients with respiratory insufficiency which may prolong the life expectancy, it is unclear to what extent increased survival is related to NIV rather than to other interventions, including scoliosis surgery. It remains uncertain whether the existing evidence is sufficiently scientifically rigorous to recommend spinal surgery for most patients with DMD and scoliosis.

In this systematic review, we evaluated the effectiveness of various forms of spinal surgery to prolong life expectancy, retard the natural deterioration of respiratory function, and improve quality of life in people with DMD. We wanted to evaluate whether the benefits outweigh the risks of surgery in general and determine which patient subgroups are most likely to benefit. The review has been updated, most recently in 2012. The objectives of this systematic review were to determine the effectiveness and safety of spinal surgery in people with DMD with scoliosis. We intended to test the following hypotheses: 1.Whether spinal surgery is effective in increasing survival; 2.Whether spinal surgery can improve respiratory function in the short-term and long-term; 3.Whether spinal surgery can improve quality of life and overall functioning; 4.Whether spinal surgery is associated with severe adverse effects.

**Materials and Methods**

**Criteria for considering studies for this review**

**Types of studies**

We planned to include controlled clinical trials using random or quasi-random allocation of treatment in the review.

**Types of participants**

People with Duchenne muscular dystrophy (defined as progressive limb girdle weakness with at least one of: (1) dystrophic changes on muscle biopsy with reduced or absent dystrophin staining; (2) deletion, duplication or point mutation of dystrophin gene) and all degrees of scoliosis documented by appropriate x-rays would be included. It was possible that this definition might have resulted in the inclusion of some individuals with an intermediate or severe Becker phenotype. However, the inclusion of only biopsy proven dystrophin negative cases could potentially result in the loss of some important data.

**Types of interventions**

We planned to include trials evaluating all forms of spinal surgery for scoliosis in the review. The control interventions were to be no treatment, non-operative treatment, or a different form of spinal surgery.

**Types of outcome measures**

**Primary outcomes**

1. Survival: to allow for studies using different follow-up periods, we planned to use hazard ratios from survival data regression analysis.

**Secondary outcomes**

1. Respiratory function, as measured by pulmonary function tests such as forced vital capacity (FVC): medium-term (3 to 12 months), and long-term (more than 12 months). The results from studies with differing lengths of follow-up were to be weighted appropriately to allow for this.

2. Medium and long-term disability as measured by validated scales such as the Barthel index or Functional Independent Measure.

3. Medium and long-term quality of life as measured by validated scales such as the 36-Item Short-Form Health Status Survey (SF-36).

4. Rate of progression of scoliosis, as measured by change of Cobb angle per year.

5. Frequency of severe adverse effects and complications, such as death related to surgery, deep surgical wound infection, wound dehiscence, loosening of fixation, pneumonia, pseudarthrosis, need for revision surgery.
Search methods for identification of studies

We searched the specialized registers of the Cochrane Neuromuscular Disease Group (31 July 2012) using the terms surgery, spine, spinal, vertebra, vertebrae, spinal fusion, scoliosis, Duchenne Muscular Dystrophy and Duchenne. We also searched MEDLINE (January 1966 to July 2012), EMBASE (January 1947 to July 2012), CENTRAL (2012, issue 7 in the Cochrane Library), CINAHL Plus (January 1937 to July 2012), Proquest Dissertation and Thesis Database (January 1980 to July 2012), and the National Institute of Health Clinical Trials Database (July 2012).

Electronic searches

The detailed search strategies in the appendices: MEDLINE (Appendix 1), EMBASE (Appendix 2), CENTRAL (Appendix 3), CINAHL Plus (Appendix 4), Proquest Dissertation and Thesis Database (Appendix 5), and NIH Clinical Trials (Appendix 6). There was no language restriction in the search and inclusion of studies. However, multiple publications reporting the same group of patients or its subsets were excluded.

Searching other resources

The review authors searched the reference lists of all relevant papers for further studies. The process of searching many different sources might have brought to light direct or indirect references to unpublished studies. We planned to seek to obtain copies of such unpublished material. In addition, we contacted colleagues and experts in the field to ascertain any unpublished or ongoing studies.

Data collection and analysis

Selection of studies

Two review authors independently reviewed titles and abstracts of references retrieved from the searches and selected all potentially relevant studies. Copies of these articles were obtained, and reviewed independently by the same authors against the inclusion criteria of the study. Review authors were not blinded to the names of the trial authors, institutions or journal of publication. The authors planned to extract data from included trials and assess trial quality independently. All disagreements would be resolved by consensus.

Data extraction and management

We would have extracted the following data: (1) Study methods (a) Design (e.g. randomized or quasi-randomized). (b) Randomization method (including list generation) (c) Method of allocation concealment (d) Blinding method (e) Stratification factors (2) Participants (a) Inclusion/exclusion criteria (b) Number (total/per group) (c) Age distribution (d) Severity of scoliosis (e) Level of scoliosis (f) Baseline respiratory function (g) Associated morbidities, e.g. cardiomyopathy (h) Previous treatments, including corticosteroids (i) Pre-treatment quality of life and functional status, as measured by validated scales (3) Intervention and control (a) Type of spinal surgery (b) Type of control (d) Details of control treatment including duration of non-operative treatment (e) Details of co-interventions (4) Follow-up data (a) Duration of follow-up (b) Loss to follow-up (5) Outcome data as described above (6) Analysis data (a) Methods of analysis (intention-to-treat/per-protocol analysis) (b) Comparability of groups at baseline (yes/no) (c) Statistical techniques We planned that data would be entered into Review Manager (RevMan) by one review author and then checked by the second author.

Assessment of risk of bias in included studies

We planned to evaluate the validity of the trials by the following criteria: (1) Selection bias (a) Was allocation of participants to treatment and control groups randomized? (b) Was allocation concealed? (2) Performance bias (a) Were participants in the comparison groups treated differently apart from the study treatments? (b) Was there blinding of participants and personnel? (3) Attrition bias (a) Were there systematic differences between the comparison groups in the loss of participants from the study? (b) Were analyses by intention-to-treat? (4) Detection bias (a) Were those assessing outcomes of the intervention blinded to the assigned intervention? (5) Reporting bias (a) Were there systematic differences between reported and unreported findings (incomplete outcome data)? We planned to summarize the quality of a trial into one of the three categories: A. Low risk of bias: all the validity criteria met. B. Moderate risk of bias: one or more validity criteria partly met but none are not met. C. High risk of bias: one or more criteria not met.

Measures of treatment effect

We planned to use risk ratio (RR) estimations with 95% confidence intervals (CI) for binary outcomes. We planned to use mean difference estimations with 95% CI for continuous outcomes. All analyses would include all participants in the treatment groups to which they were allocated.

Dealing with missing data

We planned to contact authors of included studies to supply missing data. We would have assessed missing data and drop-outs/attrition for each included study, and assess and discuss the extent to which the results and conclusions of the review could be altered by the missing data. If less than 70% of patients allocated to the treatments were not reported on at the end of the trial, for a particular outcome, we would not use those data as they would have been considered to be too prone to bias.

Assessment of heterogeneity

We planned to assess clinical heterogeneity by comparing the distribution of important participant factors between trials (age, respiratory function, severity and level of scoliosis, associated diseases), and trial factors (randomization concealment, blinding of outcome assessment, losses to follow-up, treatment type, co-interventions). We would assess statistical heterogeneity by examining...
Previous corticosteroid treatments (yes, no).

**Sensitivity analysis**
We planned to undertake sensitivity analyses to assess the impact of study quality. These would have been undertaken including: 1. All studies; 2. Only those with low risk of selection bias; 3. Only those with low risk of performance bias; 4. Only those with low risk of attrition bias; 5. Only those with low risk of detection bias. Sensitivity analysis would also be performed including and excluding subjects who might have Becker muscular dystrophy or an intermediate phenotype to see whether this would alter any of the results.

**Results**

**Description of studies**
In July 2012, a total of 80 studies were found on electronic search of the databases (Cochrane Neuromuscular Disease Group Registry: 2 studies, MEDLINE: 17 studies, EMBASE: 11 studies, CENTRAL: 1 study, CINAHL Plus: 13 studies, Proquest Dissertation and Thesis Database: 35 studies, and NIH Clinical Trials Database: 1 study). An additional 32 studies were identified on searching the reference lists of relevant studies. After duplicates were removed, a total of 105 studies were screened. Fifty-eight of these studies were excluded as they did not focus on Duchenne muscular dystrophy or scoliosis surgery, or were narrative reviews. We examined the remaining 47 studies in detail but none of these satisfied the inclusion criteria.

All these studies were prospective or retrospective case series and were not clinical trials. Most of these reviews also did not have a control group for comparisons. Where a control group was included, the controls were people who refused surgery or were assigned a different treatment modality by the treating surgeons without randomization or quasi-randomization. We therefore excluded these studies from further analyses because of significant propensity for confounding and bias. The flow of studies is shown in Figure 1.

**Risk of bias in included studies**
Not applicable.

**Effects of interventions**
No controlled trials met the inclusion criteria for further analyses.

**Discussion**
The authors have referenced some of their own studies in this systematic review. These referenced studies have been conducted in accordance with the Declaration of Helsinki (1964) and the protocols of these studies have been approved by the relevant ethics committees related to the institution in which they were performed. All human subjects, in these referenced studies, gave informed consent to participate in these studies. Despite a comprehensive search strategy used for this review, no randomized controlled trial (RCT) of surgery for scoliosis in people with Duchenne muscular dystrophy was identified. Instead we found many retrospective reviews or case series of patients with Duchenne muscular dystrophy and scoliosis treated with surgery. These studies showed varying results and had different conclusions.

Although most agreed that surgery can improve patients’ quality of life and functional status in terms of sitting posture, upper limb function and ease of care, most failed to show a significant improvement in respiratory function or long-term survival, and short-term and long-term postoperative complications occurred not uncommonly. However, a closer look at the relevant studies excluded might be helpful for guiding future clinical trials of scoliosis surgery for patients with DMD (Table 1). These 47 case series included 5 to 70 patients who had undergone scoliosis surgery. Nine of these studies also included a comparison group of 21 to 115 patients without surgery.**

**Outcome measures and comparisons**
The studies had different objectives and focused on different outcomes.
### Table 1 Characteristics of excluded studies

<table>
<thead>
<tr>
<th>Study reference</th>
<th>Number of patients</th>
<th>Treatments</th>
<th>Outcome measures</th>
<th>Findings</th>
<th>Remarks</th>
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<tbody>
<tr>
<td>Arun¹</td>
<td>43</td>
<td>Sublaminar instrumentation (19) or hybrid sublaminar and pedicle screw (13) or pedical screw (11)</td>
<td>Cobb angle, flexibility index, blood loss, operating time, complications</td>
<td>Percentage correction of Cobb angle was 72.5 +/- 14.5% (Group A), 82 +/- 6% (Group B) and 82 +/- 8% (Group C). Flexibility indices were 60 +/- 6.33% (Group A), 70 +/- 4.65% (Group B) and 67 +/- 6.79% (Group C). Mean blood loss was 4.1 L (Group A), 3.2 L (Group B) and 2.5 L (Group C). Mean operating times were 300 min (Group A), 274 min (Group B) and 234 min (Group C). Complications: 3 wound infections and 2 implant failure (Group A), 1 implant failure (Group B), 1 wound infection and 1 partial screw pull out (Group C).</td>
<td>Concluded that pedicle screw system might be favoured because of the lesser blood loss and surgical time.</td>
</tr>
<tr>
<td>Alman¹</td>
<td>48</td>
<td>Spinal fusion to L5 (38) or spinal fusion to sacrum (10) using multiple level sublaminar wires with either a modified unit rod with Galveston extensions to the pelvis cut-off, a modified rod with a cross-link placed at the caudal end, or 2 Luque rods.</td>
<td>Cobb angle, torso decompensation, sitting obliquity, spinal obliquity, need for revision surgery, mortality.</td>
<td>Sitting obliquity and spinal obliquity increased in patients fused to L5. 2 patients had fracture of L5 lamina. 2 patients required revision surgery.</td>
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<tr>
<td>Bellen¹</td>
<td>47</td>
<td>Segmental spinal instrumentation according to Luque’s technique.</td>
<td>Mortality, complications.</td>
<td>Many patients have general and pulmonary and mechanical complications.</td>
<td>Concluded that a total spinal arthrodesis could probably be avoided in these patients, which often demonstrate a satisfying spontaneous fusion after instrumentation.</td>
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<tr>
<td>Bentley¹</td>
<td>101 (included 33 patients with SMA and 4 patients with congenital muscular dystrophy)</td>
<td>Modified Luque (87), Harrington-Luque (14)</td>
<td>Cobb angle, pelvic obliquity, mortality, complications, patient satisfaction</td>
<td>Cobb angle decreased from 70 to 37º, pelvic obliquity decreased from 20 to 13º. Early severe complications in 10 patients, late complications in 24 patients. No peri-operative mortality. Excellent satisfaction in 89.6% of patients.</td>
<td>Incidence of minor or temporary complications was high, but chiefly occurred in patients with very severe curves and considerable pre-existing immobility.</td>
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<tr>
<td>Bridwell⁴</td>
<td>33 (included 21 patients with SMA)</td>
<td>Posterior segmental spinal instrumentation applied from the upper thoracic spine (T2, T3, T4, T5) down to L5 or the sacrum and pelvis. Early in the series, patients with DMD with smaller curves (&lt; 40º) were fixed to L5. All had bilateral segmental fixation with Wisconsin or sublaminar wires at each level and at times with hook supplementation. All patients fused to the sacrum had Galveston or Galveston-like fixation.</td>
<td>Questionnaires to evaluate function, self-image, cosmesis, pain, pulmonary status, patient care, quality of life, satisfaction, radiographic data.</td>
<td>All patients seemed to have benefited from the surgery. Cosmesis, quality of life, and overall satisfaction rated the highest.</td>
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<tr>
<td>Brook et al.</td>
<td>17</td>
<td>L-rod instrumentation (10), distal instrumentation with Galveston construct and rigid cross-linking (7)</td>
<td>Cobb angle and pelvic obliquity, %FVC, mortality, complications</td>
<td>Correction of Cobb angle better in the Galveston group (63% versus 51%). No pseudoarthroses or instrument failures in the Galveston group. Totally 4 patients had FVC &lt; 25%, 2 required ventilation postoperatively. No other respiratory complications. No perioperative mortality.</td>
<td>The effect of surgery on respiratory function remains uncertain.</td>
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<tr>
<td>Cambridge</td>
<td>14</td>
<td>Segmental spinal instrumentation (13), Harrington distraction rods (1)</td>
<td>Mortality, complications, sitting tolerance.</td>
<td>No peri-operative mortality, 1 required repeated re-intubation. All achieved excellent long-term sitting tolerance.</td>
<td>Recommended posterior spinal fusion with segmental instrumentation when scoliosis &gt; 30°. Spinal fusion did not increase life expectancy or pulmonary function.</td>
</tr>
<tr>
<td>Cervellati et al.</td>
<td>20</td>
<td>Modified Luque technique (19) or Cotrel-Dubousset instrumentation (1)</td>
<td>Cobb angle, vital capacity, mortality.</td>
<td>Mean correction at follow-up was 28°. Mean loss of correction was 6°. Vital capacity showed a slow progression, slightly inferior to its natural evolution in untreated patients. Death in 1 patient.</td>
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<td>Chataign et al.</td>
<td>27</td>
<td>Sublaminar wiring with Luque rods (5) or Hartshill rectangle (22) Sacral fixation with ilio-sacral screws linked to the rectangle by Cotrel-Dubousset rods and dominos (15).</td>
<td>Cobb angle, pelvic obliquity, coronal imbalance, sagittal imbalance, vital capacity, mortality, complications.</td>
<td>Scoliosis reduced to 10° after surgery and 13° after 30 months follow-up. Pelvic obliquity was reduced to 4° after surgery and 7° after 30 months. A good spinal balance was present in 20 patients after surgery. A coronal or sagittal imbalance averaging 40 mm was observed in 22 patients at follow-up. Vital capacity had annual decrease of 6.4%. 17 patients were alive with a 50 months follow-up. No operative mortality. 1 patient required tracheostomy post-operatively.</td>
<td>Concluded that surgery did not result in respiratory improvement nor in life duration lengthening.</td>
</tr>
<tr>
<td>Dubousset et al.</td>
<td>37</td>
<td>Luque rods, Harrington rods, segmental instrumentation.</td>
<td>Cobb angle, vital capacity, mortality.</td>
<td>Scoliosis reduced from 80 to 24°. No effect on decline of vital capacity. No clear benefit in length of survival.</td>
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<tr>
<td>Eagle et al.</td>
<td>75</td>
<td>Surgery and nocturnal ventilation only (13), no surgery or ventilation (35)</td>
<td>Survival, complications, FVC</td>
<td>No peri-operative deaths. Complications: GIB (2), postoperative ileus (1), spinal infection requiring removal of surgical rods (1), pressure sores (1), chronic pain due to prominence of metal prosthesis (2). Mean FVC reduced significantly (mean 1.4 L to 1.13 L) after 1 year. Median survival longer in surgery with ventilation group compared to ventilation alone (30 versus 22.2 years). Survival at 24 years higher in surgery with ventilation group compared to ventilation or no intervention (84% versus 34.6% versus 10.7%)</td>
<td>Spinal surgery does not improve FVC. Combined surgery and nocturnal ventilation improves survival</td>
</tr>
<tr>
<td>Gaine et al.</td>
<td>74</td>
<td>Luque rod (55), Isola pedicle screw (19).</td>
<td>Cobb angle, pelvic obliquity, mortality, complications</td>
<td>Fusion to S1 did not offer benefit over fusion to more proximal level. Isola system appears to maintain a slightly better Cobb angle. 1 perioperative mortality due to cardiorespiratory failure. Complications: Failure of implants (3), wound infection (2), pseudoarthrosis (2), metal implant prominence requiring removal (1)</td>
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Table 1 (Continued)
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<td>Galasko(^{12})</td>
<td>55</td>
<td>Surgery (32), refused surgery (23).</td>
<td>Mortality, complications, FVC, PEFR, Cobb angle.</td>
<td>In surgery group, FVC static for 3 years then slightly decreased. Improved PEFR maintained for up to 5 years. Cobb angle improved from 47 to 34(^{\circ}) at 5 years. Slightly improved survival with surgery. Complications: respiratory failure requiring tracheostomy (1), pneumonia (1), heart block (1), superficial wound infection (1)</td>
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<tr>
<td>Galasko(^{14})</td>
<td>76</td>
<td>Surgery (48), refused surgery (28)</td>
<td>Mortality, complications, FVC, PEFR, Cobb angle.</td>
<td>No pseudarthrosis or post-operative failures. Annual decrease of FVC lower in surgery group (0.07 versus 0.15). PEFR increased annually by 7.6 L/min in surgery group but decreased annually by 7.6 L/min in non-surgery group. Cobb angle after 3 years better in surgery group (34 versus 93 degrees). At 5 years, survival higher in surgery group (61% versus 23%). Complications: respiratory failure requiring tracheostomy (1)</td>
<td>Patients with surgery have better lung function and improved survival.</td>
</tr>
<tr>
<td>Gayet(^{15})</td>
<td>37</td>
<td>Pedicular screwing system in the lumbo-sacral area and transversal attachments with steel threads at the thoracic level. A sub-laminar fastening was placed at L1.</td>
<td>Vital capacity, mortality, complications, Cobb angle, pelvic obliquity.</td>
<td>Cobb angle decreased from 19 to 5.2(^{\circ}), and 9.5% at the latest measurement. Pelvic balancing was corrected and results have held over time. Vital capacity was reduced by 3.6% per year. Complications: stem rupture (1), superficial infection (4)</td>
<td>Cardiorespiratory function and life expectancy were not improved, but most patients and families were very satisfied by the comfort brought about by the surgical operation.</td>
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<tr>
<td>Granata(^{16})</td>
<td>30</td>
<td>Segmental spinal instrumentation and fusion.</td>
<td>Cobb angle, mortality, complications, vital capacity, quality of life, sitting position, aesthetic improvement.</td>
<td>29 had a mean 59% correction of scoliosis. Very limited loss of correction over time. One died after cardiac arrest. Complications: pressure sore (1), metal prominence requiring trimming (1). Mean vital capacity decreased from 57 +/- 17% to 34 +/- 13% at 3.9 +/- 2 years after surgery. The sitting position, aesthetic improvement and quality of life were positively evaluated by majority of the patients and their parents.</td>
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<td>Hahn(^{17})</td>
<td>20</td>
<td>Spinal fixation with pedicle-screw-alone constructs</td>
<td>%FVC Cobb angle, degree of pelvic tilt, lumbar lordosis and thoracic kyphosis, mortality, complications</td>
<td>Cobb angle improved from 44 to 10(^{\circ}), pelvic tilt improved from 14 to 3(^{\circ}), lumbar lordosis improved from 20 to 49(^{\circ}); thoracic kyphosis remained unchanged. No problems related to iliac fixation, no pseudarthrosis or implant failures. No pulmonary complications %FVC decreased from 55% preoperatively to 44% at the last follow-up. One patient died intraoperatively due to a sudden cardiac arrest.</td>
<td>The rigid primary stability with pedicle screws allowed early mobilisation of the patients, which helped to avoid pulmonary complications.</td>
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<tr>
<td>Harper(^{18})</td>
<td>45</td>
<td>AO Universal Spinal System inserted through a posterior approach.</td>
<td>Mortality, complications, hospital stay.</td>
<td>No significant difference in operative and postoperative outcomes between patients with pre-operative forced vital capacity &gt; 30% and ≤ 30%. Complications in 9 patients: pneumonia, respiratory failure requiring tracheostomy, ARDS, pleural effusion, cardiac arrhythmia</td>
<td>Concluded that routine postoperative use of mask ventilation to facilitate early tracheal extubation was vital.</td>
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<tr>
<td>Heller(^{19})</td>
<td>31</td>
<td>Isola system.</td>
<td>Cobb angle, pelvic obliquity, mortality, complications.</td>
<td>Cobb angle decreased from 48.6 to 12.5(^{\circ}), pelvic obliquity decreased from 18.2 to 3.8(^{\circ}); 1 post-operative death due to cardiopathy. Complications: pneumonia (1), respiratory arrest (1), pneumothorax (1), respiratory failure requiring tracheostomy (1), dislocation of hook (2), infection requiring revision surgery (5), iliac vein thrombosis (1), massive bleeding (1).</td>
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Competing interests: None declared. Conflict of interests: None declared. All authors abide by the Association for Medical Ethics (AME) ethical rules of disclosure. All authors contributed to conception and design, manuscript preparation, read and approved the final manuscript.


Table 1 (Continued)

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<tr>
<td>Hopp**</td>
<td>20</td>
<td>Multi-segmental instrumentation.</td>
<td>Mortality, complications, Cobb angle.</td>
<td>Mean Cobb angle decreased from 70.6 to 31.2° (mean correction 39.4° or 55.8%). Lordosis of the lumbar spine corrected from 4.1 to 17.8°. No perioperative mortality. Complication: bladder dysfunction in 1 patient.</td>
<td>Recommended using multi-segmental instrumentation methods to enable rapid mobilization and a postoperative care without brace or cast.</td>
</tr>
<tr>
<td>Kennedy**</td>
<td>38</td>
<td>Surgery (17), no surgery (21).</td>
<td>Cobb angle, forced vital capacity (FVC), mortality.</td>
<td>Mean Cobb angle of the surgical group at 14.9 years was 57.4/16.4°, and of the non-surgical group at 15 years was 45.9/5.9°. No difference in the rate of deterioration of % FVC which was 3 to 5% per year. No difference in survival in either group.</td>
<td>Spinal stabilization in DMD did not alter the decline in pulmonary function, nor did it improve survival.</td>
</tr>
<tr>
<td>Kinali**</td>
<td>123</td>
<td>Surgery (43), no surgery (80)</td>
<td>Survival (FVC, sitting comfort)</td>
<td>No difference in survival, respiratory impairment, or sitting comfort among patients managed conservatively or with surgery.</td>
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<tr>
<td>Laprade**</td>
<td>9</td>
<td>Sublaminar wiring (4), intraspinous segmental wiring (5).</td>
<td>Mortality, complications, operative time, blood loss, Cobb angle.</td>
<td>Operative time and blood loss lower in sublaminar compared to intraspinous wiring. Allogeneic bone grafts to supplement the autogenous bone graft allowed for extensive fusion. Cobb angle decreased by a mean of 32°. Complications: dural leak (1), transient numbness of left foot (1), dislodgement of sacral alar hooks (2).</td>
<td>Recommended using multi-segmental fusion and allogeneic bone grafts.</td>
</tr>
<tr>
<td>Marchesi**</td>
<td>25</td>
<td>Modified Luque: sacral screws in each S-1 pedicle and a device for transverse tension between the caudal right-angle bends of the L-rod.</td>
<td>Cobb angle, pelvic obliquity, mortality, instrumental failure, sitting balance.</td>
<td>Cobb angle decreased from 68 to 18° and pelvic obliquity decreased from 21 to &lt;15° with mean correction of 75%. No instrumentation failure or loss of correction &gt;3°. In every patient, a good sitting balance could be restored. No peri-operative mortality.</td>
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</tr>
<tr>
<td>Marsh**</td>
<td>30</td>
<td>Posterior spinal fusion.</td>
<td>Cobb angle, mortality, complications, hospital stay.</td>
<td>Mean correction of Cobb angle 36°. Two subgroups of patients were compared: those with more than 30% pre-operative FVC (17 patients) and those with less than 30% pre-operative FVC (13 patients). One patient in each group required a temporary tracheotomy and there were nine complications in total. The post-operative stay for patients in each group was similar (24 days in the &gt;30% group, 20 days in the &lt;30% group) and the complication rate was comparable with other published series. No peri-operative mortality.</td>
<td>Concluded that spinal fusion could be offered to patients with DMD even in the presence of a low FVC.</td>
</tr>
<tr>
<td>Matsumura**</td>
<td>8</td>
<td>Luque rod (2), Cotrel-Dubousset rod (6).</td>
<td>Cobb angle, FVC, quality of life, mortality, complications, sitting balance.</td>
<td>Cobb angle corrected from 58.8 to 28.6° with the mean corrective rate of 51.3%. FVC increased in 3 patients with moderate scoliosis (Cobb angle: 50 to 80°). Two cases with low % FVC (16.9% and 30.4%, respectively) had poor prognosis in respiratory status. One died of pneumonia at 17 months after the surgery and the other required mechanical ventilation. Sitting balance improved in all patients.</td>
<td>Recommended spinal fusion for patients with Cobb angle more than 30° and with % FVC more than 35%. Although the impact of spinal fusion upon the life expectancy remained unclear, favourable effect on respiratory function and quality of life could be expected for carefully selected patients with DMD.</td>
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<tr>
<td>Mehdian*</td>
<td>17</td>
<td>Luque rods secured by conventional sublaminar wires (9), Luque rods secured by sublaminar nylon straps (4), 2 L-shaped rods connected by H-bars secured by closed wire loops (3), Hartshill rectangle and sublaminar wires (1).</td>
<td>Cobb angle, respiratory function.</td>
<td>Significant loss of correction in Luque rods secured by sublaminar nylon straps and Hartshill system. Strong correlation between advance of scoliosis and respiratory function.</td>
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<tr>
<td>Miller*</td>
<td>67</td>
<td>Surgery (21), no surgery (46).</td>
<td>FVC.</td>
<td>No difference was found in the rate of deterioration of the percentage of normal FVC.</td>
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<tr>
<td>Miller*</td>
<td>39</td>
<td>Surgery (17), no surgery (22).</td>
<td>Respiratory function, sitting comfort, sitting appearance.</td>
<td>No significant differences in terms of declining respiratory function. All operated patients reported either improved sitting comfort, appearance, or both.</td>
<td>Concluded distinct benefits from segmental spine fusion; however, no salutary effect upon respiratory function either in the short term or after up to 5 years follow-up.</td>
</tr>
<tr>
<td>Miller*</td>
<td>183</td>
<td>Surgery (68), no surgery (115).</td>
<td>Survival, patient comfort, ease of care, respiratory function, quality of life.</td>
<td>Patients with surgery were more comfortable in the later years of life and easier to care for, but deteriorating pulmonary function was not affected by spinal fusion. Age at death for the 29 boys who underwent spinal fusion was 18.3 years, similar to that of the 58 boys without surgery. Factors that improved the patients’ quality of life included segmental instrumentation, fusion from T2 to the pelvis, correcting or balancing scoliosis, creating normal sagittal plane alignment and correcting pelvic obliquity.</td>
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<tr>
<td>Modi*</td>
<td>26 (including 7 cerebral palsy, 5 SMA, 4 others)</td>
<td>Posterior pelvic screw fixation</td>
<td>Cobb angle, pelvic obliquity, complications</td>
<td>Mean Cobb angle: 78.53º (before surgery), 30.7º (after surgery), 33.06º (final follow-up). There was no difference in the percentage correction between the groups with &gt;90º or &lt;90º. Complications: 1 transient loss of lower limb power, 1 deep wound infection.</td>
<td></td>
</tr>
<tr>
<td>Modi*</td>
<td>24 patients (including 6 cerebral palsy, 5 SMA, 4 others) and 12 controls (adolescent idiopathic scoliosis)</td>
<td>Posteriod pedicle screw</td>
<td>Cobb angle, pelvic obliquity, apical rotation</td>
<td>Mean Cobb angle decreased from 74 to 32º. Mean pelvic obliquity decreased from 14 to 6º. Mean apical rotation decreased from 42 to 33º. There was no significant difference between different patient groups or between patients and controls.</td>
<td></td>
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<tr>
<td>Modi*</td>
<td>50 (including 18 patients with cerebral palsy, 8 patients with SMA and 6 others)</td>
<td>Posterior spinal fusion with segmental spinal instrumentation using pedicle screw fixation</td>
<td>Mortality, complications, Cobb angle, pelvic obliquity</td>
<td>Cobb angle decreased from 79.3+/−30.3º to 31.3+/−21.6º. Pelvic obliquity decreased from 14.6+/−9.4º to 6.8+/−6.3º. 2 deaths (1 due to cardiac arrest, 1 due to hypovolemic shock. 34 patients had at least 1 perioperative complication (16 pulmonary, 14 abdominal, 3 wound related, 2 neurological, 1 cardiovascular). Post-operative complications: 7 coccycodynia, 3 screw head prominence, 2 bed sore, 1 implant loosening.</td>
<td>DMD patients had higher risk of post-operative coccycodynia.</td>
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<tr>
<td><strong>Moiti</strong></td>
<td>55 (including 28 patients with cerebral palsy and 10 patients with SMA)</td>
<td>Spinal fixation from T2/T3/T4 to L4/L5 with or without pelvic fixation. Group 1: pelvic obliquity &gt;15° with pelvic fixation; group 2: pelvic obliquity &gt;15° without pelvic fixation; group 3: pelvic obliquity &lt;15° without pelvic fixation</td>
<td>Cobb angle, pelvic obliquity, complications</td>
<td>Mean correction of Cobb angle after operation: group 1: 43.8°; group 2: 40°; group 3: 48.7°; Mean loss of correction of Cobb angle at last follow-up: group 1: 0.6°; group 2: 2.3°; group 3: 3°; Mean correction of pelvic obliquity: group 1: 14.4°; group 2: 10.7°; group 3: 5°; Mean loss of correction of pelvic obliquity at last follow-up: group 1: -0.6°; group 2: 6.5°; group 3: 0.8°. Group 2 showed significant loss of pelvic obliquity compared to group 1.</td>
<td>Patients who have pelvic obliquity &gt;15 degrees require pelvic fixation to maintain correction.</td>
</tr>
<tr>
<td><strong>Mubarak</strong></td>
<td>22</td>
<td>Luque segmental instrumentation and fusion instrumented to the sacropelvis (12), instrumented to L5 (10).</td>
<td>Cobb angle, pelvic obliquity</td>
<td>Outcomes similar between the 2 groups.</td>
<td>Concluded that if treatment is initiated early, Luque instrumentation and fusion from high thoracic (T2 or T3) to the fifth lumbar vertebra should be sufficient.</td>
</tr>
<tr>
<td><strong>Nakazawa</strong></td>
<td>36</td>
<td>Autogenous bone graft (20), allogeneic bone graft (16)</td>
<td>Cobb angle, operating time, blood loss</td>
<td>No difference in Cobb angle between the 2 groups. Mean operating time longer in autogenous group (253 min) compared to allogeneous group (233 min). Mean blood loss higher in autogenous group (850 ml) compared to allogeneous group (775 ml).</td>
<td>90% and 50% of patients in autogenous group reported donor site pain after 1 week and 3 months respectively. Concluded against autogenous bone graft for scoliosis surgery in DMD patients.</td>
</tr>
<tr>
<td><strong>Rice</strong></td>
<td>19</td>
<td>Long spinal fusion to L5 and ongoing wheelchair seating attention.</td>
<td>Sitting position.</td>
<td>At long-term follow-up 15 patients continued to sit in a well-balanced position.</td>
<td>Concluded that surgical fusion of the spine to L5 combined with ongoing attention to seating was associated with good long-term functional results in these patients.</td>
</tr>
<tr>
<td><strong>Rideau</strong></td>
<td>5</td>
<td>Luque segmental spinal stabilization without bone fusion.</td>
<td>Cobb angle, vital capacity, mortality, complications, hospital stay, pelvic obliquity, patient comfort.</td>
<td>Cobb angle decreased from 27 to 11°. Pelvic obliquity partially reduced. Static vital capacity after 2 years. No peri-operative mortality, 1 bronchopneumonia. All patients more comfortable during wheelchair activities.</td>
<td>Concluded that surgical intervention should be prophylactically undertaken when there is high risk of a rapidly evolving curve with a severe restrictive lung syndrome.</td>
</tr>
<tr>
<td><strong>Sakai</strong></td>
<td>41</td>
<td>Surgery (10), no surgery (31).</td>
<td>Sitting stability, mortality, complications.</td>
<td>Pulmonary complications were minimized by performing preoperative tracheostomy on all patients who had vital capacities less than 40% and or non-functional coughs. No peri-operative mortality. Spinal fusion permitted long-term sitting stability despite the progression of the disease.</td>
<td>Competing interests: None declared. Conflict of interests: None declared. All authors contributed to conception and design, manuscript preparation, read and approved the final manuscript. All authors abide by the Association for Medical Ethics (AME) ethical rules of disclosure.</td>
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<tr>
<td>Sengupta**</td>
<td>50</td>
<td>Galveston technique (9), L-rod (22), pedicle screw + sublaminar wires (19).</td>
<td>Cobb angle, pelvic obliquity, mortality, complications, hospital stay.</td>
<td>In the pelvic fixation group, the mean Cobb angle and pelvic obliquity were 48° and 19.8° at the time of surgery, 16.7° and 7.2° immediately after surgery, and 22° and 11.6° at the final follow-up (mean 4.6 years). The mean hospital stay was 17 days. 5 major complications: deep wound infection (1), revision of instrumentation prominence at the proximal end (2), loosening of pelvic fixation (2). In the lumbar fixation group, the mean Cobb angle and pelvic obliquity were 19.8° and 9° at the time of surgery, 3.2° and 2.2° immediately after surgery, and 5.2° and 2.9° at the final follow-up (mean 3.5 years). The mean hospital stay (7.7 days) was much less compared with the pelvic fixation group. Pelvic obliquity was corrected and maintained below 10° in all but two cases, who had an initial pelvic obliquity exceeding 20°. 2 complications: instrumentation failure at the proximal end (1), deep wound infection (1). No peri-operative mortality.</td>
<td></td>
</tr>
<tr>
<td>Shapiro**</td>
<td>27</td>
<td>Harrington rod (2), Harrington rod with sublaminar wires (7), Harrington rod, Luque rod and 2 double sublaminar wires at each level (17).</td>
<td>Cobb angle, FVC, mortality, complications.</td>
<td>1 sudden cardiac arrest and died intra-operatively. 3 intra-operative complications reversed without sequelae. Mean post-operative correction 13.1 +/- 11.9°, with mean loss of correction 5.1 +/- 3.1° at 2.4 +/- 1.8 years. Mean FVC preoperatively was 45.3 +/- 15.9° with continuing diminution to 28.7 +/- 14.9% at 3.3 +/- 2.2 years after surgery.</td>
<td>Concluded that the main benefit of surgical stabilization was the relative ease and comfort of wheelchair seating compared with those non-operated patients who develop progressive deformity. No lasting improvement or stabilization in FVC following surgery as decreasing function was related primarily to muscle weakness.</td>
</tr>
<tr>
<td>Stricker**</td>
<td>46 (included other neuromuscular diseases)</td>
<td>Modified Luque technique. Luque</td>
<td>Cobb angle, complications.</td>
<td>Cobb angle decreased from 63 to 24° (correction of about 62%). Failure of implants, pseudarthroses and major losses of correction in purely neuromuscular scolioses could be avoided by using rigid segmental fixation and a dorsolateral fusion with a mixture of autologous and allogenous bone.</td>
<td>Recommended that in DMD the best method of treatment was surgery performed as early as possible, i.e. at the time of loss of walking capacity in the case of a scoliosis exceeding 20° and with two consecutive X-rays proving curve progression.</td>
</tr>
</tbody>
</table>
Most studies aimed to investigate whether spinal surgery improves the degree of scoliosis in the short-term (immediate post-operative period) and in the long-term (years later). Most studies used Cobb angle and degree of pelvic obliquity as outcome measures and described early and late complications of surgery. Some studies also reported duration of hospitalization, peri-operative mortality, operative time, blood loss, complications, mortality, and survival. Many studies reported the change in respiratory function after surgery. The parameters used included vital capacity, peak expiratory flow, and forced vital capacity. A few studies also reported patient-oriented subjective outcomes such as quality of life, self-image, cosmetic appearance, pain, and patient satisfaction. While most studies evaluated the outcomes of spinal surgery in general, some studies tried to compare different surgical techniques, such as Luque instrumentation versus Isola pedicle screw, sublaminar wiring versus intraspinal segmental wiring, and Harrington instrumentation versus distal instrumentation with Galveston construct and rigid cross-linking. Some studies also compared prophylactic spinal fusion with prophylactic spinal fusion versus modified Luque instrumentation, Harrington instrumentation versus Luque instrumentation versus segmental spinal instrumentation with fusion, and length of survival in people who had undergone scoliosis surgery.

Many studies reported the change in respiratory function after surgery. The parameters used included vital capacity, peak expiratory flow, and forced vital capacity. A few studies also reported patient-oriented subjective outcomes such as quality of life, self-image, cosmetic appearance, pain, and patient satisfaction. While most studies evaluated the outcomes of spinal surgery in general, some studies tried to compare different surgical techniques, such as Luque instrumentation versus Isola pedicle screw, sublaminar wiring versus intraspinal segmental wiring, and Harrington instrumentation versus distal instrumentation with Galveston construct and rigid cross-linking. Some studies also compared prophylactic spinal fusion with prophylactic spinal fusion versus modified Luque instrumentation, Harrington instrumentation versus Luque instrumentation versus segmental spinal instrumentation with fusion, and length of survival in people who had undergone scoliosis surgery.
Outcomes on survival

Most studies did not demonstrate obvious benefits of scoliosis surgery in terms of prolonging survival\(^{5,6,9,15,16,17,21,22,27,28,29,30,41,45}\). There was one study showing that when combined with nocturnal ventilation, patients after spinal surgery has longer median survival (30 years) compared with patients on nocturnal ventilation alone (22.2 years)\(^{11}\).

There was another study showing that survival rate was higher at five years after surgery (61%) compared to those who refused surgery (23%)\(^{14}\). In general, the age at death in patients with or without surgery was highly variable in the case series. Although most deaths could be attributed to respiratory infection, respiratory failure, progressive cardiomyopathy and sudden cardiac death, the cause of death could not be ascertained in many cases. However, the age and causes of death did not seem to differ between patients with or without surgery. Peri-operative mortality is generally uncommon. Most studies reported no peri-operative mortality\(^{1,3,4,5,6,7,9,10,11,13,14,15,20,22,26,37,3,8,39,40,42,43,44,45,47}\), while some studies reported peri-operative mortality ranging from 1.4% to 5%\(^{8,12,16,17,18,19,33,41}\).

Outcomes on respiratory function

Galasko found that forced vital capacity could be stabilized for three years and peak expiratory flow rate maintained for up to five years after spinal fusion\(^{13,14}\). Radeau also found that vital capacity could be maintained static for two years\(^{30}\); and three participants in Matsumura’s study had increased forced vital capacity after operation\(^{26}\). Velasco found that the average rate of decline of FVC reduced from 4% per year to 1.75% per year after surgery\(^{46}\). However, most studies did not demonstrate obvious benefits of scoliosis surgery in terms of respiratory function\(^{6,8,9,11,15,16,17,21,22,27,28,29,30,41,45}\). While some studies found that patients with poor pre-operative respiratory function fared similarly to those with better respiratory function\(^{18,25}\), other studies suggested that the prognosis was worse in patients with poorer pre-operative respiratory function\(^{26,47}\).

Complications of spinal surgery

Severe complications after spinal surgery are not infrequent and occur in up to 68% of patients\(^{33}\). These include cardiac arrest\(^4\), cardiac arrhythmia\(^{18}\), heart block\(^{13}\), respiratory failure requiring tracheostomy\(^{9,13,14,18,19,25}\) or mechanical ventilation post-operatively\(^{4,6,19,33}\), massive bleeding\(^{19,31}\), pneumonia\(^{4,13,18,19,33,38}\), pleural effusion\(^{18,33}\), haemothorax or pneumothorax\(^{4,19,33}\), spinal cord injury\(^{33}\), colonic perforation\(^4\), bladder dysfunction\(^{4,20}\), urinary tract infection\(^{33}\), deep wound infection\(^{2,31,33,40}\), infection necessitating removal or revision of surgical implants\(^{11,19}\), failure of implants\(^{2,4,12,42}\), dislodgement or dislocation of implants\(^{19,23,26}\), loosening of implants\(^{2,3,40}\), mechanical problems requiring revision surgery\(^{4,12,15,16,40}\), pseudarthrosis\(^{12,45}\), bone fracture\(^1\), pressure sores\(^{16,33,34}\), dural leak\(^{23}\) and deep vein thrombosis\(^{18}\). Several studies reported that postoperative complications were more frequent in patients with greater severity of scoliosis\(^{4,39,43}\).

Comparisons of different operative methods

In general, fusion to sacrum does not offer benefits over fusion to a more proximal level\(^{12,35,37,40}\), unless scoliosis is severe and pelvic obliquity is significant\(^{2,23,34}\). Although none of the surgical methods was uniformly better than others, Isola system\(^{12}\) or segmental spinal fusion\(^{29,30}\) might achieve better correction of deformity, and intraspinal wiring might result in shorter operative time and less blood loss compared to sublaminar wiring\(^{23}\). Pedicle screw system might also result in shorter operative time and less blood loss compared to sublaminar instrumentation system\(^2\). No meta-analysis of these available data was performed because the retrospective non-randomized, uncontrolled studies were observational in nature and were prone to bias and confounding. There is currently an absence of high level evidence supporting scoliosis surgery in patients with Duchenne muscular dystrophy. There is also a lack of evidence for or against a particular modality of surgical approach. Controlled clinical trials with random allocation into treatment and control groups are needed before firm conclusions on the benefits and risks of scoliosis surgery in patients with DMD can be made. In the absence of evidence it is our view that clinicians might need to consider anecdotal evidence and their personal experience as well as expertise opinions as guidance for their decision on the best care for individual patient. Potential benefits on quality of life and functional status as well as risks of morbidity and mortality should be fully discussed with the patients before embarking on surgery for scoliosis. Patients should also be informed about the uncertainty of benefits on long-term survival and respiratory function after scoliosis surgery.

Conclusion

Implications for practice

Since there were no RCTs available to evaluate the effectiveness of scoliosis surgery in people with Duchenne muscular dystrophy, no recommendation can be made for clinical practice.

Implications for research

RCTs are needed to investigate the effectiveness of scoliosis surgery, in terms of patients’ satisfaction, quality of life, functional status, respiratory function (forced vital capacity, forced

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expiratory volume in one second, peak expiratory flow) and survival. It should be feasible to randomize patients into surgery versus non-surgical management. Although placebo control treatment might not be feasible, random allocation of patients into different treatment groups is essential to avoid selection bias and ensure baseline comparability of different groups. Although blinding of patients and clinicians is almost impossible, blinding of outcome assessors is important and probably feasible. Quality of life and functional status should be assessed by validated questionnaires and instruments. The relative benefits and risks of different surgical treatment modalities and different extents of spinal fusion should also be investigated by RCTs. Stratifications by potentially important prognostic factors such as age, baseline respiratory function and severity of scoliosis should be considered.

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References


