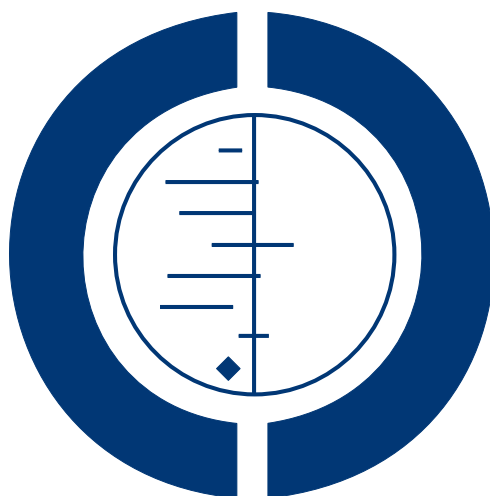


Surgery for scoliosis in Duchenne muscular dystrophy (Review)

Cheuk DKL, Wong V, Wraige E, Baxter P, Cole A



**THE COCHRANE
COLLABORATION®**

This is a reprint of a Cochrane review, prepared and maintained by The Cochrane Collaboration and published in *The Cochrane Library* 2013, Issue 2

<http://www.thecochranelibrary.com>

WILEY

TABLE OF CONTENTS

HEADER	1
ABSTRACT	1
PLAIN LANGUAGE SUMMARY	2
BACKGROUND	2
OBJECTIVES	3
METHODS	3
RESULTS	6
Figure 1.	7
DISCUSSION	8
AUTHORS' CONCLUSIONS	10
ACKNOWLEDGEMENTS	10
REFERENCES	10
CHARACTERISTICS OF STUDIES	13
DATA AND ANALYSES	17
ADDITIONAL TABLES	17
APPENDICES	34
WHAT'S NEW	36
HISTORY	36
CONTRIBUTIONS OF AUTHORS	37
DECLARATIONS OF INTEREST	37
SOURCES OF SUPPORT	37
DIFFERENCES BETWEEN PROTOCOL AND REVIEW	38
INDEX TERMS	38

[Intervention Review]

Surgery for scoliosis in Duchenne muscular dystrophy

Daniel KL Cheuk¹, Virginia Wong¹, Elizabeth Wraige², Peter Baxter³, Ashley Cole⁴

¹Department of Pediatrics and Adolescent Medicine, The University of Hong Kong, Queen Mary Hospital, Hong Kong, China.

²Department of Paediatric Neurology, Evelina Children's Hospital, St Thomas' Hospital, London, UK. ³Ryegate Children's Centre, Sheffield Children's Hospital, Sheffield, UK. ⁴Orthopaedics Department, Sheffield Children's Hospital, Sheffield, UK

Contact address: Daniel KL Cheuk, Department of Pediatrics and Adolescent Medicine, The University of Hong Kong, Queen Mary Hospital, Pokfulam Road, Hong Kong, China. cheukklid@hkucc.hku.hk.

Editorial group: Cochrane Neuromuscular Disease Group.

Publication status and date: New search for studies and content updated (no change to conclusions), published in Issue 2, 2013.

Review content assessed as up-to-date: 31 July 2012.

Citation: Cheuk DKL, Wong V, Wraige E, Baxter P, Cole A. Surgery for scoliosis in Duchenne muscular dystrophy. *Cochrane Database of Systematic Reviews* 2013, Issue 2. Art. No.: CD005375. DOI: 10.1002/14651858.CD005375.pub3.

Copyright © 2013 The Cochrane Collaboration. Published by John Wiley & Sons, Ltd.

ABSTRACT

Background

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.

Objectives

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.

Search 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.

Selection criteria

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.

Data collection and analysis

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.

Main 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.

Surgery for scoliosis in Duchenne muscular dystrophy (Review)

Copyright © 2013 The Cochrane Collaboration. Published by John Wiley & Sons, Ltd.

Authors' conclusions

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.

PLAIN LANGUAGE SUMMARY

Scoliosis surgery for people with Duchenne muscular dystrophy

Scoliosis, curvature of the spine, is common in people with Duchenne muscular dystrophy. It is usually progressive and surgery is often performed aiming to halt its progression, improve cosmetic appearance, facilitate care, preserve upper limb and respiratory function, and hopefully increase life expectancy. However, there were no randomized controlled clinical trials available to evaluate the effectiveness of scoliosis surgery. Randomized controlled clinical trials are needed in this group of patients to evaluate the benefits and risks of different surgical treatments. This is an updated review and an updated search was undertaken in which no new studies were found.

BACKGROUND

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 years (Van Essen 1997). 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 years (Van Essen 1997). 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 uncertain (Manzur 2008).

Spinal deformity, especially scoliosis, is progressive in the majority of people with DMD (Galasko 1995; Miller 1985). From the onset of spinal deformity, progression can be extremely rapid and impair unsupported sitting ability and further compromise the respiratory and cardiac function (Hsu 1983). Kurz observed a 4% decrease in vital capacity for every 10% progression of the spinal curve in people with DMD (Kurz 1983). Galasko found that on average, vital capacity decreases by 8% per year in patients with scoliosis secondary to DMD (Galasko 1992). Long-term corticosteroid treatment may slow the progress of scoliosis in people with DMD and may reduce the need for surgery (Dooley 2010), but adverse effects are frequent (Alman 2004). 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 disease (Cambridge 1987; Colbert 1987). 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 (Forst 1997; Heller 1997; McCarthy 1999).

Spinal fusion surgery with instrumentation remains the mainstay of treatment for people with DMD with scoliosis. Commonly used techniques are either based on sublaminar 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 (Bridwell 1999; Cambridge 1987; Marchesi 1997; Matsumura 1997; Miller 1991; Miller 1992; Rice 1998; Rideau 1984; Shapiro 1992), cosmetic improvement (Bellen 1993; Bridwell 1999), no need for orthopaedic braces (Bellen 1993; Colbert 1987; Miller 1985; Noble Jamieson 1986), easier nursing care by parents (Bellen 1993) and pain relief (Bellen 1993; Galasko 1977; Miller 1991).

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 DMD (Kinali 2006; Miller 1988; Miller 1992; Shapiro 1992), at short-term and five-year follow-up (Miller 1991). In contrast, several studies (Galasko 1992; Galasko 1995; Rideau 1984; Velasco 2007) 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 treated (Galasko 1992; Galasko 1995). However, other studies reported that spinal surgery did not improve life expectancy (Chataigner 1998; Gayet 1999; Kennedy 1995; Kinali 2006; Miller 1988). 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 hand 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 (Manzur 1992). 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.

OBJECTIVES

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.

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 I^2 (Higgins 2002), a quantity which describes approximately the proportion of variation in point estimates that is due to heterogeneity rather than sampling error. In addition, we would use a Chi^2 test for homogeneity to determine the strength of evidence that heterogeneity was genuine.

Assessment of reporting biases

We would have drawn funnel plots (estimated differences in treatment effects against their standard error) if sufficient studies were found. Asymmetry could be due to publication bias, but could also be due to a relationship between trial size and effect size. In the event that a relationship was found, we would examine clinical diversity of the studies (Egger 1997).

Data synthesis

Where the interventions were the same or similar enough, we planned to synthesize results in a meta-analysis if there was no important clinical heterogeneity. If no significant statistical heterogeneity was present, we planned to synthesize the data using a fixed-effect model. Otherwise we would use a random-effects model for the meta-analysis.

Adverse events

Since adverse events were rarely adequately dealt with by randomized studies alone because the numbers were small and follow-up too short, we planned to discuss adverse events taking into account the non-randomized literature.

Cost-benefit analyses

We planned to consider cost-effectiveness of interventions where relevant data were available.

Subgroup analysis and investigation of heterogeneity

If data permitted, we planned to conduct sub-group analyses for:

1. different age groups (younger than 12 years, 12 to 18 years, older than 18 years);

2. different degrees of pre-existing respiratory impairment (mild, severe);
3. different severity of scoliosis (moderate, severe);
4. 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.

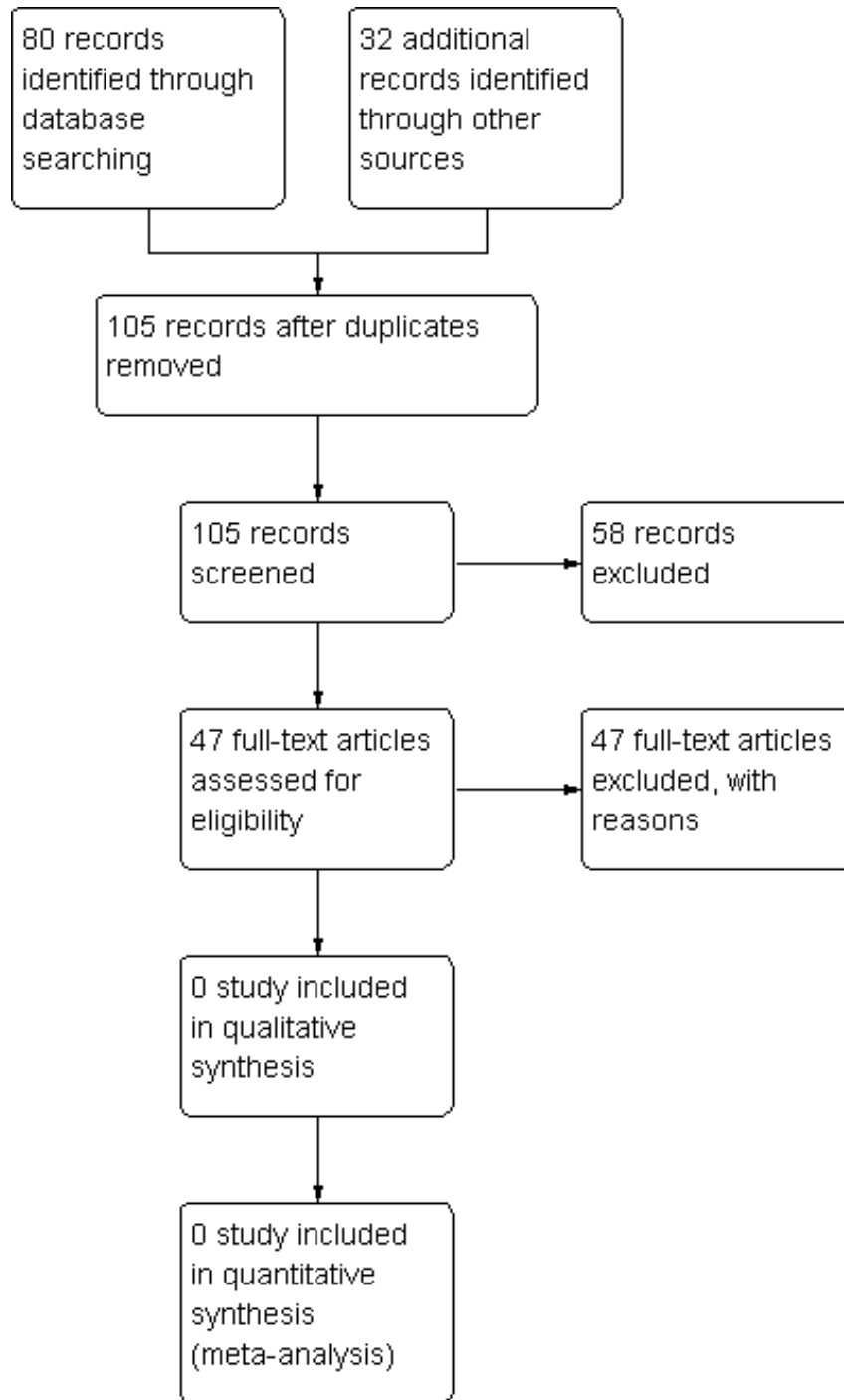
RESULTS

Description of studies

See: [Characteristics of excluded 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](#).

Figure 1. Study flow diagram.



Risk of bias in included studies

Not applicable.

Effects of interventions

No controlled trials met the inclusion criteria of the review for further analyses.

DISCUSSION

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 (Eagle 2007; Galasko 1992; Galasko 1995; Kennedy 1995; Kinali 2006; Miller 1988; Miller 1991; Miller 1992; Sakai 1977).

Outcome measures and comparisons

The studies had different objectives and focused on different outcomes. 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 (Harper 2004; Rideau 1984; Sengupta 2002; Sussman 1984), peri-operative mortality (Alman 1999; Bentley 2001; Brook 1996; Cambridge 1987; Cervellati 2004; Chataigner 1998; Dubousset 1983; Eagle 2007; Gaine 2004; Galasko 1992; Galasko 1995; Gayet 1999; Granata 1996; Hahn 2008; Harper 2004; Heller 2001; Hopf 1994; Kennedy 1995; LaPrade 1992; Marchesi 1997; Marsh 2003; Matsumura 1997; Modi 2009; Rideau 1984; Sakai 1977; Sengupta 2002; Shapiro 1992; Thacker 2002; Weimann

1983) and length of survival (Eagle 2007; Kinali 2006; Miller 1992) in people who had undergone scoliosis surgery. Many studies reported the change in respiratory function after operation (Brook 1996; Cervellati 2004; Chataigner 1998; Dubousset 1983; Eagle 2007; Galasko 1992; Galasko 1995; Gayet 1999; Granata 1996; Kennedy 1995; Kinali 2006; Matsumura 1997; Mehdian 1989; Miller 1988; Miller 1991; Miller 1992; Rideau 1984; Shapiro 1992; Thacker 2002; Velasco 2007). The parameters used included vital capacity, peak expiratory flow rate and forced vital capacity in one second. A few studies also reported patient oriented subjective outcomes such as quality of life, self-image, cosmetic appearance, pain and patient satisfaction (Bentley 2001; Bridwell 1999; Granata 1996; Matsumura 1997; Miller 1991; Miller 1992; Rideau 1984). 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 (Gaine 2004), sublaminar wiring versus intraspinous segmental wiring (LaPrade 1992), Luge instrumentation versus distal instrumentation with Galveston construct and rigid cross-linking (Brook 1996), Harrington-Luge instrumentation versus modified Luque instrumentation (Bentley 2001), Harrington instrumentation versus Luque instrumentation versus segmental spinal instrumentation with fusion (Sussman 1984), sublaminar instrumentation versus pedicle screw versus a hybrid system (Arun 2010), or autogenous versus allogeneous bone graft (Nakazawa 2010). Some studies also compared the outcomes of spinal fusion to different extents (Alman 1999; Bridwell 1999; Gaine 2004; Mubarak 1993; Sengupta 2002; Modi 2010), such as fusion to L5 versus fusion to sacrum. Some studies compared surgical outcomes in patients with different pre-operative respiratory function (Harper 2004; Marsh 2003; Matsumura 1997; Sussman 1984).

Outcomes on survival

Most studies did not demonstrate obvious benefits of scoliosis surgery in terms of prolonging survival (Brook 1996; Cervellati 2004; Chataigner 1998; Gayet 1999; Granata 1996; Hahn 2008; Kennedy 1995; Kinali 2006; Mehdian 1989; Miller 1988; Miller 1991; Miller 1992; Shapiro 1992; Thacker 2002). 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) (Eagle 2007). There was another study showing that survival rate was higher at five years after surgery (61%) compared to those who refused surgery (23%) (Galasko 1995). 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 (Alman 1999; Bellen 1993; Bentley 2001; Bridwell 1999; Brook 1996; Cambridge 1987; Chataigner 1998; Dubousset 1983; Eagle 2007; Galasko 1992; Galasko 1995; Gayet 1999; Hopf 1994; Kennedy 1995; Kinali 2006; LaPrade 1992; Marchesi 1997; Marsh 2003; Matsumura 1997; Mehdian 1989; Miller 1992; Mubarak 1993; Nakazawa 2010; Rice 1998; Rideau 1984; Sakai 1977; Sengupta 2002; Stricker 1996; Sussman 1984; Takaso 2010; Thacker 2002; Weimann 1983), while some studies reported peri-operative mortality ranging from 1.4% to 5% (Modi 2009; Gaine 2004; Cervellati 2004; Granata 1996; Hahn 2008; Harper 2004; Heller 2001; Shapiro 1992).

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 (Galasko 1992; Galasko 1995). Rideau also found that vital capacity could be maintained static for two years (Rideau 1984); and three participants in Matsumura's study had increased forced vital capacity after operation (Matsumura 1997). Velasco found that the average rate of decline of FVC reduced from 4% per year to 1.75% per year after surgery (Velasco 2007). However, most studies did not demonstrate obvious benefits of scoliosis surgery in terms of respiratory function (Brook 1996; Chataigner 1998; Cervellati 2004; Eagle 2007; Gayet 1999; Granata 1996; Hahn 2008; Kennedy 1995; Kinali 2006; Mehdian 1989; Miller 1988; Miller 1991; Miller 1992; Shapiro 1992; Thacker 2002). While some studies found that patients with poor pre-operative respiratory function fared similarly to those with better pre-operative respiratory function (Marsh 2003; Harper 2004), other studies suggested that the prognosis was worse in patients with poorer pre-operative respiratory function (Matsumura 1997; Sussman 1984).

Functional outcome and quality of life

In general, previous descriptive studies suggested that surgical correction of scoliosis resulted in better sitting position, quality of life and patient satisfaction (Bentley 2001; Bridwell 1999; Cambridge 1987; Granata 1996; Marchesi 1997; Matsumura 1997; Miller 1991; Miller 1992; Rice 1998; Rideau 1984; Sakai 1977; Shapiro 1992).

Complications of spinal surgery

Severe complications after spinal surgery are not infrequent and occur in up to 68% of patients (Modi 2009). These include cardiac arrest (Bentley 2001), cardiac arrhythmia (Harper 2004), heart block (Galasko 1992), respiratory failure requiring tracheostomy (Chataigner 1998; Galasko 1992; Galasko 1995;

Harper 2004; Heller 2001; Marsh 2003) or mechanical ventilation post-operatively (Bentley 2001; Brook 1996; Heller 2001; Modi 2009), massive bleeding (Heller 2001; Modi 2008a), pneumonia (Bentley 2001; Galasko 1992; Harper 2004; Heller 2001; Modi 2009; Rideau 1984), pleural effusion (Harper 2004; Modi 2009), hemothorax or pneumothorax (Bentley 2001; Heller 2001; Modi 2009), spinal cord injury (Modi 2009), colonic perforation (Bentley 2001), bladder dysfunction (Bentley 2001; Hopf 1994), urinary tract infection (Modi 2009), deep wound infection (Arun 2010; Modi 2008a; Modi 2009; Sengupta 2002), infection necessitating removal or revision of surgical implants (Eagle 2007; Heller 2001), failure of implants (Arun 2010; Bentley 2001; Gaine 2004; Stricker 1996), dislodgement or dislocation of implants (Heller 2001; LaPrade 1992; Matsumura 1997), loosening of implants (Arun 2010; Modi 2009; Sengupta 2002), mechanical problems requiring revision surgery (Bentley 2001; Gaine 2004; Gayet 1999; Granata 1996; Sengupta 2002), pseudarthrosis (Gaine 2004; Thacker 2002), bone fracture (Alman 1999), pressure sores (Granata 1996; Modi 2009; Modi 2010), dural leak (LaPrade 1992) and deep vein thrombosis (Heller 2001). Several studies reported that postoperative complications were more frequent in patients with greater severity of scoliosis (Bentley 2001; Sakai 1977; Sussman 1984).

Comparisons of different operative methods

In general, fusion to sacrum does not offer benefits over fusion to a more proximal level (Gaine 2004; Mubarak 1993; Rice 1998; Sengupta 2002), unless scoliosis is severe and pelvic obliquity is significant (Alman 1999; LaPrade 1992; Modi 2010). Although none of the surgical methods was uniformly better than others, Isola system (Gaine 2004) or segmental spinal fusion (Miller 1991; Miller 1992) might achieve better correction of deformity, and intraspinal wiring might result in shorter operative time and less blood loss compared to sublaminae wiring (LaPrade 1992). Pedicle screw system might also result in shorter operative time and less blood loss compared to sublaminae instrumentation system (Arun 2010).

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 patient 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 expert 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.

AUTHORS' CONCLUSIONS

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 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.

ACKNOWLEDGEMENTS

We wish to thank Sarah Massey and the Illingworth Library at the Sheffield Children's Hospital for their help and support in locating and retrieving studies. We also thank Angela Gunn for updating search strategies and searching various electronic databases.

Editorial support from the Cochrane Neuromuscular Disease Group for an earlier update was funded by the TREAT NMD Network European Union Grant 036825.

The editorial base of the Cochrane Neuromuscular Disease Group is supported by the MRC Centre for Neuromuscular Diseases and the Muscular Dystrophy Campaign.

REFERENCES

References to studies excluded from this review

Alman 1999 *{published data only}*

Alman BA, Kim HK. Pelvic obliquity after fusion of the spine in Duchenne muscular dystrophy. *Journal of Bone and Joint Surgery. British Volume* 1999;**81**(5):821–4.

Arun 2010 *{published data only}*

Arun R, Srinivas S, Mehdi SMH. Scoliosis in Duchennes muscular dystrophy: A changing trend in surgical management: A historical outcome study comparing sublaminar, hybrid and pedicle screw instrumentation systems. *European Spine Journal* 2010;**19**(3):376–83.

Bellen 1993 *{published data only}*

Bellen P, Hody JL, Clairbois J, Denis N, Soudon P. The surgical treatment of spinal deformities in Duchenne muscular dystrophy. *Journal of Orthopaedic Surgery (Hong Kong)* 1993;**7**:48–57.

Bentley 2001 *{published data only}*

Bentley G, Haddad F, Bull TM, Seingry D. The treatment of scoliosis in muscular dystrophy using modified Luque and Harrington-Luque instrumentation. *Journal of Bone and Joint Surgery - Series B* 2001;**83**(1):22–8.

Bridwell 1999 *{published data only}*

Bridwell KH, Baldus C, Iffrig TM, Lenke LG, Blanke K. Process measures and patient/parent evaluation of surgical management of spinal deformities in patients with progressive flaccid neuromuscular scoliosis (Duchenne's muscular dystrophy and spinal muscular atrophy). *Spine* 1999;**24**(13):1300–9.

Brook 1996 *{published data only}*

Brook PD, Kennedy JD, Stern LM, Sutherland AD, Foster BK. Spinal fusion in Duchenne's muscular dystrophy. *Journal of Pediatric Orthopaedics* 1996;**16**(3):324–31.

Cambridge 1987 *{published data only}*

Cambridge W, Drennan JC. Scoliosis associated with Duchenne muscular dystrophy. *Journal of Pediatric Orthopaedics* 1987;**7**(4):436–40.

Cervellati 2004 *{published data only}*

Cervellati S, Bettini N, Moscato M, Gusella A, Dema E, Maresi R. Surgical treatment of spinal deformities in Duchenne muscular dystrophy: a long term follow-up study. *European Spine Journal* 2004;**13**(5):441–8.

Chataigner 1998 *{published data only}*

Chataigner H, Grelet V, Onimus M. Surgery of the spine in Duchenne's muscular dystrophy. *Revue de Chirurgie*

- Orthopedique et Reparatrice de l'Appareil Moteur* 1998;**84**(3): 224–30.
- Dubousset 1983** *{published data only}*
Dubousset J, Queneau P. Role and indications for surgery in Duchenne de Boulogne muscular dystrophy with rapid development. *Revue de Chirurgie Orthopedique et Reparatrice de l'Appareil Moteur* 1983;**69**(3):207–20.
- Eagle 2007** *{published data only}*
Eagle M, Bourke J, Bullock R, Gibson M, Mehta J, Giddings D, et al. Managing Duchenne muscular dystrophy—the additive effect of spinal surgery and home nocturnal ventilation in improving survival. *Neuromuscular Disorders*. 2007;**17**(6):470–5.
- Gainé 2004** *{published data only}*
Gainé WJ, Lim J, Stephenson W, Galasko CS. Progression of scoliosis after spinal fusion in Duchenne's muscular dystrophy. *Journal of Bone and Joint Surgery. British Volume* 2004;**86**(4):550–5.
- Galasko 1992** *{published data only}*
Galasko CSB, Delaney C, Morris P. Spinal stabilisation in Duchenne muscular dystrophy. *Journal of Bone and Joint Surgery. British Volume* 1992;**74**(2):210–4.
- Galasko 1995** *{published data only}*
Galasko CSB, Williamson JB, Dalaney CM. Lung function in Duchenne muscular dystrophy. *European Spine Journal* 1995;**4**:263–7.
- Gayet 1999** *{published data only}*
Gayet LE. Surgical treatment of scoliosis due to Duchenne muscular dystrophy. *Chirurgie* 1999;**124**(4):423–31.
- Granata 1996** *{published data only}*
Granata C, Merlini L, Cervellati S, Ballestrazzi A, Giannini S, Corbascio M, et al. Long-term results of spine surgery in Duchenne muscular dystrophy. *Neuromuscular Disorders* 1996;**6**(1):61–8.
- Hahn 2008** *{published data only}*
Hahn F, Hauser D, Espinosa N, Blumenthal S, Min K. Scoliosis correction with pedicle screws in Duchenne muscular dystrophy. *European Spine Journal* 2008;**17**(2): 255–261.
- Harper 2004** *{published data only}*
Harper CM, Ambler G, Edge G. The prognostic value of pre-operative predicted forced vital capacity in corrective spinal surgery for Duchenne's muscular dystrophy. *Anaesthesia* 2004;**59**(12):1160–2.
- Heller 2001** *{published data only}*
Heller KD, Wirtz DC, Siebert CH, Forst R. Spinal stabilization in Duchenne muscular dystrophy: principles of treatment and record of 31 operative treated cases. *Journal of Pediatric Orthopedics. Part B* 2001;**10**(1):18–24.
- Hopf 1994** *{published data only}*
Hopf C, Forst R, Forst J, Eysel P, Reitter B. Multi-segmental fusion of scoliosis in Duchenne's muscular dystrophy. *Zeitschrift für Orthopädie und Ihre Grenzgebiete* 1994;**132** (5):377–82.
- Kennedy 1995** *{published data only}*
Kennedy JD, Staples AJ, Brook PD, Parsons DW, Sutherland AD, Martin AJ, et al. Effect of spinal surgery on lung function in Duchenne muscular dystrophy. *Thorax* 1995;**50**(11):1173–8.
- Kinali 2006** *{published data only}*
Kinali M, Messina S, Mercuri E, Lehovsky J, Edge G, Manzur AY, et al. Management of scoliosis in Duchenne muscular dystrophy: a large 10-year retrospective study. *Developmental Medicine & Child Neurology* 2006;**48**(6): 513–8.
- LaPrade 1992** *{published data only}*
LaPrade RF, Rowe DE. The operative treatment of scoliosis in Duchenne muscular dystrophy. *Orthopaedic Review* 1992;**21**(1):39–45.
- Marchesi 1997** *{published data only}*
Marchesi D, Arlet V, Stricker U, Aebi M. Modification of the original Luque technique in the treatment of Duchenne's neuromuscular scoliosis. *Journal of Pediatric Orthopedics* 1997;**17**(6):743–9.
- Marsh 2003** *{published data only}*
Marsh A, Edge G, Lehovsky J. Spinal fusion in patients with Duchenne's muscular dystrophy and a low forced vital capacity. *European Spine Journal* 2003;**12**(5):507–12.
- Matsumura 1997** *{published data only}*
Matsumura T, Kang J, Nozaki S, Takahashi MP. The effect of spinal fusion on respiratory function and quality of life in Duchenne muscular dystrophy. *Rinsho Shinkeigaku* 1997; **37**(2):87–92.
- Mehdian 1989** *{published data only}*
Mehdian H, Shimizu N, Draycott V, Evans G, Eisenstein S. Spinal stabilisation for scoliosis in Duchenne Muscular Dystrophy. Experience with various sublaminar instrumentation systems. *Neuro-orthopedics* 1989;**7**(2): 74–82.
- Miller 1988** *{published data only}*
Miller F, Moseley CF, Koreska J, Levison H. Pulmonary function and scoliosis in Duchenne dystrophy. *Journal of Pediatric Orthopedics* 1988;**8**(2):133–7.
- Miller 1991** *{published data only}*
Miller RG, Chalmers AC, Dao H, Filler-Katz A, Holman B, Bost F. The effect of spine fusion on respiratory function in Duchenne muscular dystrophy. *Neurology* 1991;**41**(1): 38–40.
- Miller 1992** *{published data only}*
Miller F, Moseley CF, Koreska J. Spinal fusion in Duchenne muscular dystrophy. *Developmental Medicine and Child Neurology* 1992;**34**(9):775–86.
- Modi 2008a** *{published data only}*
Modi HN, Suh SW, Song HR, Fernandez HM, Yang JH. Treatment of neuromuscular scoliosis with posterior-only pedicle screw fixation. *Journal of Orthopedic Surgery* 2008;**3**: 23.
- Modi 2008b** *{published data only}*
Modi HN, Suh SW, Song HR, Lee SH, Yang JH. Correction of apical axial rotation with pedicular screws

- in neuromuscular scoliosis. *Journal of Spinal Disorders & Techniques* 2008;**21**(8):606–13.
- Modi 2009** *{published data only}*
Modi HN, Suh SW, Yang JH, Cho JW, Hong JY, Singh SU, et al. Surgical complications in neuromuscular scoliosis operated with posterior-only approach using pedicle screw fixation. *Scoliosis* 2009;**4**:11.
- Modi 2010** *{published data only}*
Modi HN, Woo Suh S, Song HR, Hyuk Yang J, Jajodia N. Evaluation of pelvic fixation in neuromuscular scoliosis: a retrospective study in 55 patients. *International Orthopaedics* 2010;**34**(1):89–96.
- Mubarak 1993** *{published data only}*
Mubarak SJ, Morin WD, Leach J. Spinal fusion in Duchenne muscular dystrophy—fixation and fusion to the sacropelvis. *Journal of Pediatric Orthopaedics* 1993;**13**(6):752–7.
- Nakazawa 2010** *{published data only}*
Nakazawa T, Takaso M, Imura T, Adachi K, Fukushima K, Saito W, et al. Autogenous iliac crest bone graft versus banked allograft bone in scoliosis surgery in patients with Duchenne muscular dystrophy. *International Orthopaedics* 2010;**34**(6):855–61.
- Rice 1998** *{published data only}*
Rice JJ, Jeffers BL, Devitt AT, McManus F. Management of the collapsing spine for patients with Duchenne muscular dystrophy. *Irish Journal of Medical Science* 1998;**167**(4):242–5.
- Rideau 1984** *{published data only}*
Rideau Y, Glorion B, Delaubier A, Tarle O, Bach J. The treatment of scoliosis in Duchenne muscular dystrophy. *Muscle & Nerve* 1984;**7**(4):281–6.
- Sakai 1977** *{published data only}*
Sakai DN, Hsu JD, Bonnett CA, Brown JC. Stabilization of the collapsing spine in duchenne muscular dystrophy. *Clinical Orthopaedics and Related Research* 1977;**128**:256–60.
- Sengupta 2002** *{published data only}*
Sengupta DK, Mehdiian SH, McConnell JR, Eisenstein SM, Webb JK. Pelvic or lumbar fixation for the surgical management of scoliosis in duchenne muscular dystrophy. *Spine* 2002;**27**(18):2072–9.
- Shapiro 1992** *{published data only}*
Shapiro F, Sethna N, Colan S, Wohl ME, Specht L. Spinal fusion in Duchenne muscular dystrophy: a multidisciplinary approach. *Muscle & Nerve* 1992;**15**(5):604–14.
- Stricker 1996** *{published data only}*
Stricker U, Moser H, Aebi M. Predominantly posterior instrumentation and fusion in neuromuscular and neurogenic scoliosis in children and adolescents. *European Spine Journal* 1996;**5**(2):101–6.
- Sussman 1984** *{published data only}*
Sussman MD. Advantage of early spinal stabilization and fusion in patients with Duchenne muscular dystrophy. *Journal of Pediatric Orthopaedics* 1984;**4**(5):532–7.
- Takaso 2010** *{published data only}*
Takaso M, Nakazawa T, Imura T, Okada T, Ueno M, Fukushima K, et al. Segmental pedicle screws instrumentation and fusion to L5 for spinal deformity secondary to Duchenne muscular dystrophy: results with a minimum of 2 years follow-up. *European Journal of Orthopaedic Surgery & Traumatology* 2010;**20**(6):453–61.
- Thacker 2002** *{published data only}*
Thacker M, Hui JH, Wong HK, Chatterjee A, Lee EH. Spinal fusion and instrumentation for paediatric neuromuscular scoliosis: retrospective review. *Journal of Orthopaedic Surgery (Hong Kong)* 2002;**10**(2):144–51.
- Velasco 2007** *{published data only}*
Velasco MV, Colin AA, Zurakowski D, Darras BT, Shapiro F. Posterior spinal fusion for scoliosis in duchenne muscular dystrophy diminishes the rate of respiratory decline. *Spine* 2007;**32**(4):459–465.
- Weimann 1983** *{published data only}*
Weimann RL, Gibson DA, Moseley CF, Jones DC. Surgical stabilization of the spine in Duchenne muscular dystrophy. *Spine* 1983;**8**(7):776–80.

Additional references

- Alman 2004**
Alman BA, Raza SN, Biggar WD. Steroid treatment and the development of scoliosis in males with duchenne muscular dystrophy. *Journal of Bone and Joint Surgery (American Volume)* 2004;**86-A**(3):519–24.
- Colbert 1987**
Colbert AP, Craig C. Scoliosis management in Duchenne muscular dystrophy: prospective study of modified Jewett hyperextension brace. *Archives of Physical Medicine and Rehabilitation* 1987;**68**(5 Pt 1):302–4.
- Dooley 2010**
Dooley JM, Gordon KE, MacSween JM. Impact of steroids on surgical experience of patients with Duchenne muscular dystrophy. *Pediatric Neurology* 2010;**43**(3):173–6.
- Egger 1997**
Egger M, Davey-Smith G, Schneider M, Minder C. Bias in meta-analysis detected by a simple, graphic test. *British Medical Journal* 1997;**315**(7):629–34.
- Forst 1997**
Forst R, Forst J, Heller KD, Hengstler K. Characteristics in the treatment of scoliosis in muscular diseases. *Zeitschrift für Orthopädie und Ihre Grenzgebiete* 1997;**135**(2):95–105.
- Galasko 1977**
Galasko CSB. Incidence of orthopaedic problems in children with muscle disease. *Israel Journal of Medical Science* 1977;**13**(2):165–76.

Heller 1997

Heller KD, Forst R, Forst J, Hengstler K. Scoliosis in Duchenne muscular dystrophy: aspects of orthotic treatment. *Prosthetics and Orthotics International* 1997;**21**(3):202–9.

Higgins 2002

Higgins JP, Thompson SG. Quantifying heterogeneity in a meta-analysis. *Statistics in Medicine* 2002;**21**(11):1539–58.

Higgins 2011

Higgins JPT, Green S (editors). *Cochrane Handbook for Systematic Reviews of Interventions* Version 5.1.0 [updated March 2011]. The Cochrane Collaboration, 2011. Available from www.cochrane-handbook.org.

Hsu 1983

Hsu JD. The natural history of spine curvature progression in the non-ambulatory Duchenne muscular patient. *Spine* 1983;**8**(7):771–5.

Kurz 1983

Kurz LT, Mubarak SJ, Schultz P, Park SM, Leach J. Correlation of scoliosis and pulmonary function in Duchenne muscular dystrophy. *Journal of Pediatric Orthopedics* 1983;**3**(3):347–53.

Manzur 1992

Manzur AY, Hyde SA, Rodillo E, Heckmatt JZ, Bentley G, Dubowitz V. A randomized controlled trial of early surgery in Duchenne muscular dystrophy. *Neuromuscular Disorders* 1992;**2**(5-6):379–87.

Manzur 2008

Manzur AY, Kuntzer T, Pike M, Swan A. Glucocorticoid corticosteroids for Duchenne muscular dystrophy. *Cochrane*

Database of Systematic Reviews 2008, Issue 1. [DOI: 10.1002/14651858.CD003725]

McCarthy 1999

McCarthy RE. Management of neuromuscular scoliosis. *Orthopedic Clinics of North America* 1999;**30**(3):435–49.

Miller 1985

Miller G, O'Connor J. Spinal bracing and respiratory function in Duchenne muscular dystrophy. *Clinical Pediatrics* 1985;**24**(2):94.

Noble Jamieson 1986

Noble Jamieson CM, Heckmatt JZ, Dubowitz V, Silverman M. Effects of posture and spinal bracing on respiratory function in neuromuscular disease. *Archives of Disease in Childhood* 1986;**61**(2):178–81.

Van Essen 1997

Van Essen AJ, Verheij JBG, Reefhuis J, Fidler V, Begeer JH, De Visser M, et al. The natural history of Duchenne muscular dystrophy. Analysis of data from Dutch survey and review of age related events. Thesis 1997. [ISBN 90 367 0711 0]

References to other published versions of this review**Cheuk 2010**

Cheuk DKL, Wong V, Wraige E, Baxter P, Cole A, N'Diaye T, et al. Surgery for scoliosis in Duchenne muscular dystrophy. *Cochrane Database of Systematic Reviews* 2007, Issue 1. [DOI: 10.1002/14651858.CD005375.pub2]

* Indicates the major publication for the study

CHARACTERISTICS OF STUDIES

Characteristics of excluded studies *[ordered by study ID]*

Study	Reason for exclusion
Alman 1999	Retrospective case series, not clinical trial.
Arun 2010	Retrospective case series, not clinical trial.
Bellen 1993	Retrospective case series, not clinical trial.
Bentley 2001	Retrospective case series, not clinical trial.
Bridwell 1999	Retrospective case series, not clinical trial.
Brook 1996	Retrospective case series, not clinical trial.
Cambridge 1987	Retrospective case series, not clinical trial.
Cervellati 2004	Retrospective case series, not clinical trial.
Chataigner 1998	Retrospective case series, not clinical trial.
Dubousset 1983	Retrospective case series, not clinical trial.
Eagle 2007	Retrospective case series, not clinical trial.
Gaine 2004	Retrospective case series, not clinical trial.
Galasko 1992	Retrospective case series, not clinical trial.
Galasko 1995	Retrospective case series, not clinical trial.
Gayet 1999	Retrospective case series, not clinical trial.
Granata 1996	Retrospective case series, not clinical trial.
Hahn 2008	Retrospective case series, not clinical trial.
Harper 2004	Prospective case series, not clinical trial.
Heller 2001	Prospective case series, not clinical trial.
Hopf 1994	Retrospective case series, not clinical trial.
Kennedy 1995	Retrospective case series, not clinical trial.

(Continued)

Kinali 2006	Retrospective case series, not clinical trial.
LaPrade 1992	Retrospective case series, not clinical trial.
Marchesi 1997	Retrospective case series, not clinical trial.
Marsh 2003	Retrospective case series, not clinical trial.
Matsumura 1997	Retrospective case series, not clinical trial.
Mehdian 1989	Retrospective case series, not clinical trial.
Miller 1988	Retrospective case series, not clinical trial.
Miller 1991	Retrospective case series, not clinical trial.
Miller 1992	Retrospective case series, not clinical trial.
Modi 2008a	Retrospective case series, not clinical trial.
Modi 2008b	Retrospective case series, not clinical trial.
Modi 2009	Retrospective case series, not clinical trial.
Modi 2010	Retrospective case series, not clinical trial.
Mubarak 1993	Retrospective case series, not clinical trial.
Nakazawa 2010	Prospective case series, not clinical trial.
Rice 1998	Retrospective case series, not clinical trial.
Rideau 1984	Retrospective case series, not clinical trial.
Sakai 1977	Retrospective case series, not clinical trial.
Sengupta 2002	Retrospective case series, not clinical trial.
Shapiro 1992	Retrospective case series, not clinical trial.
Stricker 1996	Retrospective case series, not clinical trial.
Sussman 1984	Retrospective case series, not clinical trial.
Takaso 2010	Prospective case series, not clinical trial.

(Continued)

Thacker 2002	Retrospective case series, not clinical trial.
Velasco 2007	Retrospective case series, not clinical trial.
Weimann 1983	Retrospective case series, not clinical trial.

DATA AND ANALYSES

This review has no analyses.

ADDITIONAL TABLES

Table 1. Characteristics of excluded studies

Study reference	Number of patients	Treatments	Outcome measures	Findings	Remarks
Arun 2010	43	Sublaminar instrumentation (19) or hybrid sublaminar and pedicle screw (13) or pedicle screw (11)	Cobb angle, flexibility index, blood loss, operating time, complications	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)	Concluded that pedicle screw system might be favored because of the lesser blood loss and surgical time
Alman 1999	48	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	Cobb angle, torso decompensation, sitting obliquity, spinal obliquity, need for revision surgery, mortality	Sitting obliquity and spinal obliquity increased in patients fused to L5. 2 patients had fracture of L5 lamina. 2 patients required revision surgery	

Table 1. Characteristics of excluded studies (Continued)

		the caudal end, or 2 Luque rods			
Bellen 1993	47	Segmental spinal instrumentation according to Luque's technique	Mortality, complications.	Many patients have general and pulmonary and mechanical complications	Concluded that a total spinal arthrodesis could probably be avoided in these patients, which often demonstrate a satisfying spontaneous fusion after instrumentation
Bentley 2001	101 (included 33 patients with SMA and 4 patients with congenital muscular dystrophy)	Modified Luque (87), Harrington-Luque (14)	Cobb angle, pelvic obliquity, mortality, complications, patient satisfaction	Cobb angle decreased from 70 to 37°, pelvic obliquity decreased from 20 to 13°. Early severe complications in 10 patients, late complications in 24 patients. No peri-operative mortality. Excellent satisfaction in 89.6% of patients	Incidence of minor or temporary complications was high, but chiefly occurred in patients with very severe curves and considerable pre-existing immobility
Bridwell 1999	33 (included 21 patients with SMA)	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 (< 40°) 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-	Questionnaires to evaluate function, self-image, cosmesis, pain, pulmonary status, patient care, quality of life, satisfaction, radiographic data.	All patients seemed to have benefited from the surgery. Cosmesis, quality of life, and overall satisfaction rated the highest	

Table 1. Characteristics of excluded studies (Continued)

		like fixation			
Brook 1996	17	L-rod instrumentation (10), distal instrumentation with Galveston construct and rigid cross-linking (7)	Cobb angle and pelvic obliquity, %FVC, mortality, complications	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 < 25%, 2 required ventilation postoperatively. No other respiratory complications. No peri-operative mortality	The effect of surgery on respiratory function remains uncertain
Cambridge 1987	14	Segmental spinal instrumentation (13), Harrington distraction rods (1)	Mortality, complications, sitting tolerance.	No peri-operative mortality, 1 required repeated re-intubation. All achieved excellent long-term sitting tolerance	Recommended posterior spinal fusion with segmental instrumentation when scoliosis > 30°. Spinal fusion did not increase life expectancy or pulmonary function
Cervellati 2004	20	Modified Luque technique (19) or Cotrel-Dubousset instrumentation (1)	Cobb angle, vital capacity, mortality.	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	
Chataigner 1998	27	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)	Cobb angle, pelvic obliquity, coronal imbalance, sagittal imbalance, vital capacity, mortality, complications	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 pa-	Concluded that surgery did not result in respiratory improvement nor in life duration lengthening

Table 1. Characteristics of excluded studies (Continued)

				<p>tients 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</p>	
Dubousset 1983	37	Luque rods, Harrington rods, segmental instrumentation.	Cobb angle, vital capacity, mortality.	Scoliosis reduced from 80 to 24°. No effect on decline of vital capacity. No clear benefit in length of survival	
Eagle 2007	75	Surgery and nocturnal ventilation (27), nocturnal ventilation only (13), no surgery or ventilation (35)	Survival, complications, FVC	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	Spinal surgery does not improve FVC. Combined surgery and nocturnal ventilation improves survival

Table 1. Characteristics of excluded studies (Continued)

				or no intervention (84% versus 34.6% versus 10.7%)	
Gaine 2004	74	Luque rod (55), Isola pedicle screw (19).	Cobb angle, pelvic obliquity, mortality, complications	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), pseudarthrosis (2), metal implant prominence requiring removal (1)	
Galasko 1992	55	Surgery (32), refused surgery (23).	Mortality, complications, FVC, PEFR, Cobb angle.	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° at 5 years. Slightly improved survival with surgery. Complications: respiratory failure requiring tracheostomy (1), pneumonia (1), heart block (1), superficial wound infection (1)	
Galasko 1995	76	Surgery (48), refused surgery (28)	Mortality, complications, FVC, PEFR, Cobb angle.	No pseudarthrosis or post-operative failures. Annual decrease of FVC lower in surgery group (0.07 ver-	Patients with surgery have better lung function and improved survival

Table 1. Characteristics of excluded studies (Continued)

				<p>sus. 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)</p>	
Gayet 1999	37	<p>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</p>	<p>Vital capacity, mortality, complications, Cobb angle, pelvic obliquity</p>	<p>Cobb angle decreased from 19 to 5.2°, 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)</p>	<p>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</p>
Granata 1996	30	<p>Segmental spinal instrumentation and fusion.</p>	<p>Cobb angle, mortality, complications, vital capacity, quality of life, sitting position, aesthetic improvement</p>	<p>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 af-</p>	

Table 1. Characteristics of excluded studies (Continued)

				ter surgery. The sitting position, aesthetic improvement and quality of life were positively evaluated by majority of the patients and their parents	
Hahn 2008	20	Spinal fixation with pedicle-screw-alone constructs	%FVC Cobb angle, degree of pelvic tilt, lumbar lordosis and thoracic kyphosis, mortality, complications	Cobb angle improved from 44 to 10°, pelvic tilt improved from 14 to 3°. Lumbar lordosis improved from 20 to 49°, 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	The rigid primary stability with pedicle screws allowed early mobilisation of the patients, which helped to avoid pulmonary complications
Harper 2004	45	AO Universal Spinal System inserted through a posterior approach	Mortality, complications, hospital stay.	No significant difference in operative and postoperative outcomes between patients with pre-operative forced vital capacity > 30% and ≤ 30%. Complications in 9 patients: pneumonia, respiratory failure requiring tracheostomy, ARDS, pleural effusion, cardiac arrhythmia	Concluded that routine postoperative use of mask ventilation to facilitate early tracheal extubation was vital

Table 1. Characteristics of excluded studies (Continued)

Heller 2001	31	Isola system.	Cobb angle, pelvic obliquity, mortality, complications.	Cobb angle decreased from 48.6 to 12.5°, pelvic obliquity decreased from 18.2 to 3.8°. 1 post-operative death due to cardiac failure. 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)	
Hopf 1994	20	Multi-segmental instrumentation.	Mortality, complications, Cobb angle.	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 peri-operative mortality. Complication: bladder dysfunction in 1 patient	Recommended using multi-segmental instrumentation methods to enable rapid mobilization and a postoperative care without brace or cast
Kennedy 1995	38	Surgery (17), no surgery (21).	Cobb angle, forced vital capacity (FVC), mortality.	Mean Cobb angle of the surgical group at 14.9 years was 57 +/- 16.4°, and of the non-surgical group at 15 years was 45 +/- 9.9°. No difference in the rate of deterioration of % FVC which was 3 to 5% per year. No difference in survival in either group	Spinal stabilization in DMD did not alter the decline in pulmonary function, nor did it improve survival

Table 1. Characteristics of excluded studies (Continued)

Kinali 2006	123	Surgery (43), no surgery (80)	Survival, (FVC, sitting comfort)	No difference in survival, respiratory impairment, or sitting comfort among patients managed conservatively or with surgery	
Laprade 1992	9	Sublaminar wiring (4), intraspinal segmental wiring (5).	Mortality, complications, operative time, blood loss, Cobb angle	Operative time and blood loss lower in sublaminar compared to intraspinal 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)	Recommended segmental fusion and allogeneic bone grafts.
Marchesi 1997	25	Modified Luque: sacral screws in each S-1 pedicle and a device for transverse traction between the caudal right-angle bends of the L-rods	Cobb angle, pelvic obliquity, mortality, instrumental failure, sitting balance	Cobb angle decreased from 68 to 18° and pelvic obliquity decreased from 21 to <15° with mean correction of 75%. No instrumentation failure or loss of correction >3°. In every patient, a good sitting balance could be restored. No perioperative mortality	
Marsh 2003	30	Posterior spinal fusion.	Cobb angle, mortality, complications, hospital stay.	Mean correction of Cobb angle 36°. Two subgroups of patients were compared: those with	Concluded that spinal fusion could be offered to patients with DMD even in the presence of a low

Table 1. Characteristics of excluded studies (Continued)

				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 >30% group, 20 days in the <30% group) and the complication rate was comparable with other published series. No peri-operative mortality	FVC
Matsumura 1997	8	Luque rod (2), Cotrel-Dubousset rod (6).	Cobb angle, FVC, quality of life, mortality, complications, sitting balance	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	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, favorable effect on respiratory function and quality of life could be expected for carefully selected patients with DMD

Table 1. Characteristics of excluded studies (Continued)

Mehdian 1989	17	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)	Cobb angle, respiratory function.	Significant loss of correction in Luque rods secured by sublaminar nylon straps and Hartshill system. Strong correlation between advance of scoliosis and respiratory function	
Miller 1988	67	Surgery (21), no surgery (46).	FVC.	No difference was found in the rate of deterioration of the percentage of normal FVC	
Miller 1991	39	Surgery (17), no surgery (22).	Respiratory function, sitting comfort, sitting appearance.	No significant differences in terms of declining respiratory function. All operated patients reported either improved sitting comfort, appearance, or both	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
Miller 1992	183	Surgery (68), no surgery (115).	Survival, patient comfort, ease of care, respiratory function, quality of life	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, fu-	

Table 1. Characteristics of excluded studies (Continued)

					tion from T2 to the pelvis, correcting or balancing scoliosis, creating normal sagittal plane alignment and correcting pelvic obliquity	
Modi 2008a	26 (including 7 cerebral palsy, 5 SMA, 4 others)	posterior pelvic screw fixation	Cobb angle, pelvic obliquity, complications	Mean Cobb angle: 78. 53° (before surgery), 30.7° (after surgery), 33.06° (final follow-up). There was no difference in the percentage correction between the groups with >90° or <90°. Complications: 1 transient loss of lower limb power, 1 deep wound infection		
Modi 2008b	24 patients (including 6 cerebral palsy, 5 SMA, 4 others) and 12 controls (adolescent idiopathic scoliosis)	Posterior pedicle screw	Cobb angle, pelvic obliquity, apical rotation	Mean Cobb angle decreased from 74 to 32°. Mean pelvic obliquity decreased from 14 to 6°. Mean apical rotation decreased from 42 to 33°. There was no significant difference between different patient groups or between patients and controls		
Modi 2009	50 (including 18 patients with cerebral palsy, 8 patients with SMA and 6 others)	Posterior spinal fusion with segmental spinal instrumentation using pedicle screw fixation	Mortality, complications, Cobb angle, pelvic obliquity	Cobb angle decreased from 79.3+/-30.3° to 31.3+/-21.6°. Pelvic obliquity decreased from 14.6+/-9.4° to 6.8+/-6.3°. 2 deaths (1 due to cardiac arrest, 1 due to hypovolemic shock). 34 patients had at least 1 periop-	DMD patients had higher risk of post-operative coccygodynia.	

Table 1. Characteristics of excluded studies (Continued)

				erative complication (16 pulmonary, 14 abdominal, 3 wound related, 2 neurological, 1 cardiovascular) Post-operative complications: 7 coccygodynia, 3 screw head prominence, 2 bed sore, 1 implant loosening	
Modi 2010	55 (including 28 patients with cerebral palsy and 10 patients with SMA)	Spinal fixation from T2/T3/T4 to L4/L5 with or without pelvic fixation. Group 1: pelvic obliquity >15° with pelvic fixation; group 2: pelvic obliquity >15° without pelvic fixation; group 3: pelvic obliquity <15° without pelvic fixation	Cobb angle, pelvic obliquity, complications	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. Complications: 3 patients had sacral sores in group 1	Patients who have pelvic obliquity >15 degrees require pelvic fixation to maintain correction
Mubarak 1993	22	Luque segmental instrumentation and fusion instrumented to the sacropelvis (12), instrumented to L5 (10)	Cobb angle, pelvic obliquity.	Outcomes similar between the 2 groups.	Concluded that if treatment is initiated early, Luque instrumentation and fusion from high thoracic (T2 or T3) to the fifth lumbar vertebra should be suf-

Table 1. Characteristics of excluded studies (Continued)

					ficent
Nakazawa 2010	36	Autogenous bone graft (20), allogeneic bone graft (16)	Cobb angle, operating time, blood loss	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)	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
Rice 1998	19	Long spinal fusion to L5 and ongoing wheelchair seating attention	Sitting position.	At long-term follow-up 15 patients continued to sit in a well-balanced position	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
Rideau 1984	5	Luque segmental spinal stabilization without bone fusion.	Cobb angle, vital capacity, mortality, complications, hospital stay, pelvic obliquity, patient comfort	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	Concluded that surgical intervention should be prophylactically undertaken when there is high risk of a rapidly evolving curve with a severe restrictive lung syndrome
Sakai 1977	41	Surgery (10), no surgery (31).	Sitting stability, mortality, complications.	Pulmonary complications were minimized by performing preoperative tracheostomy on all patients who had vital capacities less than 40% and or nonfunc-	

Table 1. Characteristics of excluded studies (Continued)

				<p>tional coughs. No peri-operative mortality. Spinal fusion permitted long-term sitting stability despite the progression of the disease</p>	
Sengupta 2002	50	Galveston technique (9), L-rod (22), pedicle screw + sublaminar wires (19)	Cobb angle, pelvic obliquity, mortality, complications, hospital stay	<p>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 main-</p>	

Table 1. Characteristics of excluded studies (Continued)

					tained 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	
Shapiro 1992	27	Harrington rod (2), Harrington rod with sublaminar wires (7), Harrington rod, Luque rod and 2 double sublaminar wires at each level (17)	Cobb angle, FVC, mortality, complications.	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	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	
Stricker 1996	46 (included other neuromuscular diseases)	Modified Luque technique.	Cobb angle, complications.	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 allogeneous bone	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	

Table 1. Characteristics of excluded studies (Continued)

Sussman 1984	11	Harrington instrumentation (group I) (3), Luque instrumentation (group II) (3), segmental spinal instrumentation with fusion (group III) (5)	Complications, Cobb angle, hospital stay.	Mean Cobb angle correction: 40% (I), 35% (II), 60% (III). When surgery to stabilize spinal deformity is done in younger patients in whom pulmonary function is better and curves are milder, complication rate and length of hospital stay are diminished, correction and balance are improved, and patients rapidly return to their normal lifestyle	Concluded that segmental spinal instrumentation had advantage of allowing rapid mobilization without need of a cast or body jacket. Recommended stabilization of the collapsing spine surgically with segmental instrumentation and fusion when scoliosis reached 30 to 40°
Takaso 2010	20	Segmental pedicle screws instrumentation and fusion to L5.	Cobb angle, pelvic obliquity, operating time, blood loss, complications	Mean Cobb angle decreased from 70° to 15°. Mean pelvic obliquity decreased from 13° to 6°. The mean intraoperative blood loss was 890 ml (range: 660 to 1260 ml). The mean total blood loss was 2100 ml (range: 1250 to 2880 ml). There was no major complication	
Thacker 2002	5	Not detailed in DMD patients.	FEV1, FVC, mortality, complications.	FVC and FEV1 maintained, pseudarthrosis in 1 patient, no peri-operative mortality	Included 7 SMA, 6 spastic cerebral palsy, 3 congenital myopathy, 2 spina bifida, 1 paraspinal neuroblastoma in the series
Velasco 2007	56	Posterior spinal fusion	Percent normal FVC	The rates of FVC decline were 4% per year presurgery, which decreased to 1.75% per year post-	

Table 1. Characteristics of excluded studies (Continued)

				surgery	
Weimann 1983	24	Long Harrington instrumentations and spinal fusions from S1 up to the upper thoracic spine (T4, 5, or 6)	Mortality, complications.	One patient died 2 years after his operation from dystrophic cardiomyopathy	Concluded that prophylactic spinal fusion deserved consideration in the care planned for these patients

ARDS: adult respiratory distress syndrome; DMD: Duchenne muscular dystrophy; FVC: forced vital capacity; FEV1: forced expiratory volume in 1 second; PEF: peak expiratory flow rate; SMA: spinal muscular atrophy

APPENDICES

Appendix I. MEDLINE strategy

Database: Ovid MEDLINE(R) <1946 to July Week 3 2012>

Search Strategy:

-
- 1 randomized controlled trial.pt. (332315)
 - 2 controlled clinical trial.pt. (84684)
 - 3 randomized.ab. (235702)
 - 4 placebo.ab. (133040)
 - 5 drug therapy.fs. (1552464)
 - 6 randomly.ab. (169810)
 - 7 trial.ab. (244167)
 - 8 groups.ab. (1114025)
 - 9 or/1-8 (2885687)
 - 10 exp animals/ not humans.sh. (3757814)
 - 11 9 not 10 (2450652)
 - 12 surg\$.mp. or surgery/ (1335297)
 - 13 spine\$.mp. (82686)
 - 14 spinal.mp. (269459)
 - 15 vertebra\$.mp. (166510)
 - 16 or/13-15 (412217)
 - 17 12 and 16 (56524)
 - 18 spinal fusion/ or spinal fusion.mp. (15911)
 - 19 17 or 18 (62847)
 - 20 scolio\$.mp. or Scoliosis/ (15574)
 - 21 duchenne.mp. or Muscular Dystrophy, Duchenne/ (7902)
 - 22 11 and 19 and 20 and 21 (18)
 - 23 remove duplicates from 22 (17)

Appendix 2. EMBASE search strategy

Database: Embase <1980 to 2012 Week 30>

Search Strategy:

1 crossover-procedure.sh. (34521)
2 double-blind procedure.sh. (109963)
3 single-blind procedure.sh. (16165)
4 randomized controlled trial.sh. (326003)
5 (random\$ or crossover\$ or cross over\$ or placebo\$ or (doubl\$ adj blind\$) or allocat\$).tw,ot. (885002)
6 trial.ti. (133129)
7 clinical trial/ (869205)
8 or/1-7 (1482353)
9 (animal/ or nonhuman/ or animal experiment/) and human/ (1194751)
10 animal/ or nonanimal/ or animal experiment/ (3291877)
11 10 not 9 (2727149)
12 8 not 11 (1395248)
13 limit 12 to embase (1081020)
14 Surgery/ or surg\$.mp. (1965351)
15 (spine or spinal or vertebra\$.mp. (474719)
16 14 and 15 (86443)
17 exp Spine Fusion/ (16226)
18 (spinal fusion or spine fusion).mp. (16755)
19 16 or 17 or 18 (92142)
20 exp Scoliosis/ or scoliosis.mp. (20307)
21 Duchenne Muscular Dystrophy/ or duchenne.mp. (11023)
22 13 and 19 and 20 and 21 (11)

Appendix 3. CENTRAL search strategy

#1 MeSH descriptor General Surgery explode all trees
#2 surgery
#3 (#1 OR #2)
#4 (spine or spinal or vertebra*)
#5 (#3 AND #4)
#6 MeSH descriptor Spinal Fusion, this term only
#7 spinal fusion or spine fusion
#8 ((#5 AND #6) OR #7)
#9 scoliosis
#10 duchenne
#11(#8 AND #9 AND #10)

Appendix 4. CINAHL search strategy

Tuesday, July 31, 2012 11:29:22 AM

S29 S18 and S28 13
S28 S25 and S26 and S27 35
S27 ("scoliosis") or (MH "Scoliosis") 3652
S26 ("duchenne") or (MH "Duchenne Muscular Dystrophy") 793
S25 S22 or S24 13207
S24 S23 or spinal fusion or spine fusion 3727
S23 (MH "Spinal Fusion") 3397
S22 S20 and S21 12713

S21 spine or spinal or vertebra* 53209
 S20 S19 or surgery 216179
 S19 (MH "Surgery, Operative") 12808
 S18 S1 or S2 or S3 or S4 or S5 or S6 or S7 or S8 or S9 or S10 or S11 or S12 or S13 or S14 or S15 or S16 or S17 550602
 S17 ABAB design* 77
 S16 TI random* or AB random* 111997
 S15 (TI (cross?over or placebo* or control* or factorial or sham? or dummy)) or (AB (cross?over or placebo* or control* or factorial or sham? or dummy)) 231348
 S14 (TI (clin* or intervention* or compar* or experiment* or preventive or therapeutic) or AB (clin* or intervention* or compar* or experiment* or preventive or therapeutic)) and (TI (trial*) or AB (trial*)) 78188
 S13 (TI (meta?analys* or systematic review*)) or (AB (meta?analys* or systematic review*)) 22863
 S12 (TI (single* or doubl* or tripl* or trebl*) or AB (single* or doubl* or tripl* or trebl*)) and (TI (blind* or mask*) or AB (blind* or mask*)) 18252
 S11 PT ("clinical trial" or "systematic review") 103252
 S10 (MH "Factorial Design") 835
 S9 (MH "Concurrent Prospective Studies") or (MH "Prospective Studies") 182671
 S8 (MH "Meta Analysis") 14368
 S7 (MH "Solomon Four-Group Design") or (MH "Static Group Comparison") 30
 S6 (MH "Quasi-Experimental Studies") 5485
 S5 (MH "Placebos") 7634
 S4 (MH "Double-Blind Studies") or (MH "Triple-Blind Studies") 24614
 S3 (MH "Clinical Trials+") 144869
 S2 (MH "Crossover Design") 9471
 S1 (MH "Random Assignment") or (MH "Random Sample") or (MH "Simple Random Sample") or (MH "Stratified Random Sample") or (MH "Systematic Random Sample") 57405

Appendix 5. Proquest Dissertation & Thesis Database search strategy

Duchenne and surgery and scoliosis

Appendix 6. NIH Clinical Trials Database

Duchenne and surgery and scoliosis

WHAT'S NEW

Last assessed as up-to-date: 31 July 2012.

Date	Event	Description
4 January 2013	New citation required but conclusions have not changed	Review updated with search update to July 31 2012 but no new studies found. Two of the original authors withdrawn
7 November 2012	New search has been performed	Two studies added to excluded studies tables. Minor editorial revisions

HISTORY

Protocol first published: Issue 3, 2005

Review first published: Issue 1, 2007

Date	Event	Description
22 August 2010	New search has been performed	Review updated with search update but no new studies found
13 May 2009	Amended	Acknowledgement added.
2 October 2008	New search has been performed	updated review
23 October 2006	New citation required and conclusions have changed	Substantive amendment

CONTRIBUTIONS OF AUTHORS

Cheuk DKL: protocol development, searching for trials, quality assessment of trials, data extraction, data input, data analyses, development of final review, corresponding author.

Wong V: protocol development, searching for trials, quality assessment of trials, data extraction, data analyses, development of final review.

Wraige E: protocol development, searching for trials, quality assessment of trials, data extraction, data analyses, development of final review.

Baxter P: protocol development, searching for trials, quality assessment of trials, data extraction, data analyses, development of final review.

Cole A: protocol development, searching for trials, quality assessment of trials, data extraction, data analyses, development of final review.

DECLARATIONS OF INTEREST

No potential conflict of interest is known.

SOURCES OF SUPPORT

Internal sources

- None, Not specified.

External sources

- None, Not specified.

DIFFERENCES BETWEEN PROTOCOL AND REVIEW

Risk of bias methodology updated in accordance with the *Cochrane Handbook for Systematic Reviews of Interventions* ([Higgins 2011](#)).

Change in authorship: we were unable to contact original authors N'Diaye T and Mayowe V for this update.

INDEX TERMS

Medical Subject Headings (MeSH)

Muscular Dystrophy, Duchenne [*complications]; Scoliosis [complications; *surgery]; Spine [surgery]

MeSH check words

Humans