

SPINE

The treatment of neuromuscular scoliosis

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Abstract Neuromuscular scolioses are difficult deformities to treat. A careful assessment and an understanding of the primary disease and its prognosis are essential for planning treatment which is aimed at maximising function. Conservative treatment of these deformities is difficult. Braces may be of short-term value. Surgery is the mainstay of treatment for appropriately selected patients. The goals of surgery are to correct the deformity producing a balanced spine with a level pelvis and a solid spinal fusion to prevent or delay secondary respiratory complications. Improvements in anaesthetic techniques and recognition of the importance of the timing of surgery have reduced the overall complication rate. As deformities involve most of the thoracic and lumbar spine posterior instrumentation is the standard surgical approach. Anterior release is necessary only in severe deformities. The use of pedicle screws improves fixation in the lumbar spine and has reduced the need for instrumentation to the sacrum. Early referral of patients who are candidates for surgery simplifies treatment and may improve its outcome. This can only be achieved by better education of referring physicians on the importance of timing of surgery. © 2003 Elsevier Ltd. All rights reserved.

INTRODUCTION

Scoliosis frequently develops in patients with neurological or muscle diseases. The aetiology of the spinal deformity is a muscle imbalance which can be caused by diseases of the brain, spinal cord, peripheral nerves, neuromuscular junctions or the muscles. Although the underlying neuromuscular conditions from which the patients suffer vary, the scoliosis that they develop shows certain common features:

1. The deformities are progressive and may continue to progress after skeletal maturity.
2. The curves involve most of the thoracic and lumbar spine.
3. The commonest pattern in the non-ambulant patient is the collapsing 'C' shaped curve with associated obliquity of the pelvis.
4. Inadequate compensatory curves in these patients increase the risk of loss of spinal balance.
5. The deformities whilst initially mobile rapidly become stiff, making satisfactory correction difficult if the treatment is delayed.

The fundamental principle in the treatment of orthopaedic deformities secondary to neuromuscular diseases is to maximise function. This is particularly true for patients with neuromuscular scoliosis.

Neuromuscular scolioses can be among the most challenging deformities to treat satisfactorily. There is no ideal method of treatment. Conservative treatment is usually unsuccessful as these deformities are difficult to control with orthoses. Operative treatment involves major surgery on frail patients with a higher complication rate than is seen when operating on most other types of scoliosis. In early reports of the surgical correction of neuromuscular scoliosis the complication rate was alarmingly high up to 81% in some series.¹ Despite its limitations, surgery is the most effective form of treatment in suitably selected patients.²

There are a number of factors which make surgery technically demanding and increase the risk of complications in these patients:

- The patients have a poor bone quality. This is most marked in the non-ambulant patients with disuse osteoporosis.
- Soft tissues are uncompliant and oedematous particularly in the myopathic patients. This is a

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problem even in early deformities. As the curves progress severe soft tissue contractures may develop.

- The patients often have associated medical problems such as respiratory dysfunction or cardiomyopathy.
- Nutritional problems are common in these patients and their nutritional status needs to be carefully assessed.
- The patients may be uncooperative, suffer from epilepsy or have involuntary movements, all of which considerably increase the risk of the loss of fixation following surgery.
- Mobilising the patients after surgery is difficult and it is particularly important to obtain rigid fixation to permit early mobilisation without the need for external support.

CLASSIFICATION (SCOLIOSIS RESEARCH SOCIETY)

- I. Neuropathic
 - A. Upper motor neurone
 - i. Cerebral palsy
 - ii. Spinocerebellar degeneration
 - a. Friedreich's ataxia
 - b. Charcot Marie Tooth disease
 - c. Roussy-Levy syndrome
 - iii. Syringomyelia
 - iv. Spinal cord tumour
 - v. Spinal cord trauma
 - B. Lower motor neurone
 - i. Poliomyelitis
 - ii. Other viral myelitides
 - iii. Traumatic
 - iv. Spinal muscular atrophy
 - v. Werdnig-Hoffmann disease
 - a. Kugelberg-Welander disease
 - v. Dysautonomic disorders (Riley-Day syndrome)
- II. Myopathic
 - A. Arthrogryposis
 - B. Muscular dystrophy
 - i. Duchenne muscular dystrophy
 - ii. Limb-girdle dystrophy
 - iii. Fascio-scapulo-humeral dystrophy
 - C. Fibre-type disproportion
 - D. Congenital hypotonia
 - E. Myotonia dystrophica

In this review particular consideration will be given to the two groups of patients in whom treatment is most frequently required for neuromuscular scoliosis. These are Duchenne muscular dystrophy (DMD) and cerebral palsy (CP). There are some interesting contrasts between the management problems posed by these two groups of patients.

TREATMENT OPTIONS IN NEUROMUSCULAR SCOLIOSIS

The objective of treatment in neuromuscular scoliosis is to maximise patient's function whilst preventing progression of the scoliosis. The treatment options available in neuromuscular scoliosis are limited. The decision on the choice of treatment involves an understanding of the nature of the underlying neurological disease and the level of function at the time of presentation. If it is a progressive disease with a very limited life expectancy. It would be inappropriate to put the child and their family through the trauma of major spinal surgery and bracing may be the best treatment option. If the patient has a non-progressive neurological condition and is ambulant with a progressive deformity surgical treatment is the best option. Similarly, patients who are confined to a wheelchair with a scoliosis which is interfering with their sitting balance may well be candidates for surgical correction of their deformity.

Choosing the correct option is much easier for the surgeon, patient and their families if the child has normal or near normal intellectual function and can be involved in the decision. It is therefore easier for the family of a boy with Duchenne muscular scoliosis to accept the choice for surgery if their son is involved in the decision. The family of the child with severe cerebral palsy on the other hand may have to choose for them and will be left with a greater sense that they are subjecting an unwilling child to an unpleasant operation. This is one of the factors which may explain the tendency for late presentation in patients with cerebral palsy.

Non-operative treatment and bracing

The role of bracing in patients with neuromuscular scoliosis is contentious. A review of the literature shows that there is no convincing evidence to suggest that any form of brace prevents progression of deformity in neuromuscular scoliosis. The types of brace which are available for use in patients with scoliosis include the dynamic, Milwaukee and underarm braces. Dynamic bracing is illogical in patients with neuromuscular scoliosis as these braces require normal muscle function to be effective. The role of the Milwaukee brace is very limited in this group of patients, and is contraindicated in wheelchair-bound patients. The most commonly used orthosis therefore, is an underarm brace. These can either be made of rigid plastic or softer materials. Softer braces are better tolerated, and in the short term seem to be effective for flexible deformities. These braces can hold the patient in a better position thereby improving sitting balance and maintaining their level of function in the short term. While there is little evidence that bracing alters the outcome, it may delay progression. Bracing may therefore be used to hold the curve and allow

growth in younger patients before definitive surgery is undertaken.

Another option available for the disabled and non-ambulant patient is to modify the wheelchair with supports to improve the child's sitting position. One such modification is a moulded half shell in which the patient lies and can be tilted up to 60°. The advantage of this is that it is better tolerated by the patient, is simpler for the carers and does not restrict chest movements unlike braces. This is important in these children who are prone to frequent chest infections. The abdomen is also freely accessible for the ease of managing a feeding gastrostomy.

Physiotherapy with regular stretching is an important part of the general care of these children, but it will not prevent progression of an established scoliosis.

Surgery

Surgery is the only form of treatment that will improve an established deformity and prevent its progression. With surgery, however there is a significant risk of complications. The complication rate in the past was very high, but a number of factors have reduced the complication rate. These include improvement in anaesthetic techniques, operating earlier in the course of the disease before the deformities become rigid, and improvement in surgical techniques.

Despite these advances the complication rate remains higher when operating on neuromuscular deformities than most other spinal deformities. Some reports have shown that the infection rate in neuromuscular scoliosis maybe as high as 10%. Should this occur, surgery to remove the metal-ware may be necessary. It must be borne in mind that wherever possible the initial surgery needs to be definitive. Multiple operations are highly undesirable for these patients.

Assessment

Prior to deciding on the treatment plan for these patients they need to be carefully assessed.³ This should include assessment of the spinal deformity and investigation to identify and correct any associated medical problems such as gastro-intestinal reflux. The specific risks need to be evaluated prior to surgical intervention to minimise post-operative complications. A complete assessment should include the following.

Clinical assessment

- **Function:** the status of ambulation, independent sitting ability, head control and the level of intellectual function,

- Examination of the spine in the sitting and prone position to determine the presence of pelvic obliquity and to assess the curve flexibility,
- Examination of hips to look for fixed deformities and the possibility of hip dislocation,
- Neurological assessment and careful charting of neurological status.

Lung function

A history of frequent chest infections should be sought. The presence of an adequate cough is a good clinical indicator of a satisfactory forced expiratory volume. They are difficult to perform in non-compliant patients.

The assessment should include:

- Examination of the chest,
- Chest X-ray,
- Lung function tests. The simplest test is still spirometry although this is only of value in compliant patients. Sniff nasal inspiratory pressures (SNIP) are currently under evaluation as a further method of assessing global respiratory function.
- Assessment of overnight oxygen saturation may be of value.

Cardiac status

There is a potential for considerable blood loss during surgery. Anti-epileptic medications such as Valproate can impair platelet function. A full evaluation, therefore, is necessary in children in the myopathic group with cardiac problems since cardiomyopathy is commonly encountered.

Assessment should include:

- Chest X-ray,
- ECG,
- Echocardiogram.

Nutritional status

In this group of patients nutritional status is frequently suboptimal. Poor nutritional status increases the post-operative complication rate. The nutritional evaluation should include serum levels of:

- Total protein,
- Albumin (≥ 3.5 g/l)
- Lymphocyte count (≥ 1.5 g/l).

Imaging of the spine

- Erect radiographs (standing/sitting) in the antero-posterior and lateral planes.
- Flexibility of the curve is assessed by traction views or prone push-pull views.

- Additional imaging of the spine with MRI scanning is not widely used in this group of patients.

Surgical planning and selection of levels for surgery

The surgical objective in these patients is to produce a balanced spine in the frontal and sagittal plane, a level pelvis and a solid fusion. The general principle in neuromuscular scoliosis is to operate long. Short segment corrections are illogical since these patients have an underlying neuromuscular disease and hence lack the ability to compensate. If fusion is too short junctional deformities develop with progression above or below the instrumented levels. Posterior instrumentation and fusion therefore remains the mainstay of surgical correction in neuromuscular scoliosis.^{2, 4-8} Anterior release of the apex of the scoliosis is reserved for severe deformities where a satisfactory correction cannot be obtained by posterior surgery alone.^{2,9}

Anterior surgery is sometimes used in this group of patients to prevent 'crankshaft' when operating on young patients (less than age 10 years). The Crankshaft phenomenon was first described by Dubousset and occurs when anterior growth continues with a posterior tethering fusion, resulting in a gross rotational deformity. Anterior and posterior surgery also improves the fusion rate in patients who are felt to be at particularly high risk of pseudarthrosis.

The most contentious question in planning correction is whether the pelvis should be included in the instrumentation and fusion.^{4, 6-8, 10} This partly depends on the level of function. It is generally accepted that fixation to the pelvis is undesirable in patients who are ambulant. In the non-ambulant patient if the deformity can be corrected and a level pelvis is achieved, instrumentation to L5 may be adequate. Patients with a residual pelvic obliquity should be fused to the pelvis.

Surgical technique

Since Paul Harrington first developed his instrumentation system of distraction rods in the treatment of scoliosis in patients with poliomyelitis during the epidemics of the 1950s, there have been considerable advances in fixation techniques. The greatest advance has been the development of the segmental instrumentation technique described by Eduardo Luque (Fig. 1). This provides a greater rigidity of fixation, which avoids the need for post-operative bracing and produces a better control of the sagittal contour. This technique involves the use of sublaminar wires, at each of the levels to be fused, attached to two rods. The fixation of the posterior elements at each level spreads the forces over a large area.²

The gold standard operative technique for the treatment of neuromuscular scoliosis in the non-ambulant pa-

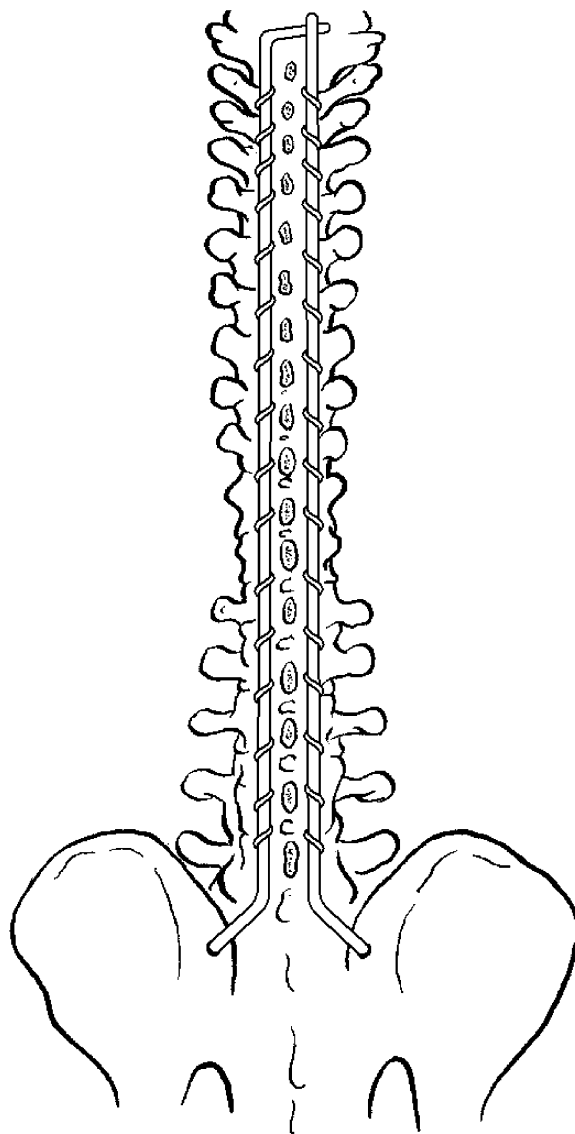


Figure 1. Luque-Galveston technique in treating neuromuscular scoliosis with pelvic obliquity.

tient has been the use of Luque Galveston fixation to the pelvis and segmental sublaminar wiring at all the levels up to T2 or T3. This technique is difficult and time-consuming and is associated with a considerable blood loss.

Modern scoliosis instrumentation systems can be used to provide a rigid fixation. The fixation is obtained with pedicle screws between T8 and L5. Higher in the thoracic spine the fixation is usually obtained with hooks (pedicle or laminar). Pedicle screws may sometimes be used in the upper thoracic spine (Fig. 2).

Although fixation techniques are important it must be remembered that the objective of surgery is to obtain a spinal fusion. Autografts are frequently inadequate in these patients who often have a poor bone quality, particularly as in these patients the posterior pelvis may be required to anchor the distal end of the fixation. Milled



Figure 2. Radiographs: A: preoperative and B: post-operative. 15-year-old girl with a multiple Acyl CoA dehydrogenase deficiency presented with myopathy, respiratory insufficiency and secondary neuromuscular scoliosis. She underwent a single stage posterior instrumentation and fusion from T3 to L5. A good correction is maintained at the 3-year follow-up. She has a well-balanced trunk, level pelvis, ambulates independently and is on regular nocturnal ventilatory support.

allograft femoral head bone has proven to be very useful in this situation and is probably the best type of bone graft for these patients. The main concern about using allograft bone is that it may increase the risk of wound infection.

Blood loss is the most important per-operative complication in this group of patients.^{2,8,9} Cross-matching of blood is therefore essential for these patients. The amount of blood required may be reduced by the use of the cell-saver intra-operatively. Predonation of blood which is a standard technique in patients with idiopathic scoliosis is of less value in this group of patients but may be appropriate in selected patients. It has also been suggested that per-operative blood loss may be reduced by the use of intravenous Trasylo1 during surgery.

The incidence of neurological injury during operations for neuromuscular scoliosis is higher than for idiopathic scoliosis. Intraoperative monitoring of spinal cord function is recommended. Spinal somato-sensory evoked potentials (SSEP) can be monitored in most of these patients despite their underlying neurological condition.¹¹

DUCHENNE MUSCULAR DYSTROPHY

Duchenne muscular dystrophy is the commonest cause of myopathic neuromuscular scoliosis. It is an X-linked condition affecting 1 in 3500 male live births. Shortly

after these boys go off their feet and into a wheelchair they begin to develop a spinal deformity. Progressive scoliosis develops in 95% of boys with Duchenne muscular dystrophy while 5% develop a hyperextended posture which poses almost as much of a functional disadvantage as a scoliosis.¹² The scoliosis once present follows a predictable course. A deformity with a Cobb angle over 20° will inevitably progress.¹³ The options of treatment for these boys are either bracing or surgery. There is no evidence in the literature to support bracing for long-term management of the deformity. Braces are not tolerated well by most of the boys with DMD.

The mainstay of treatment of scoliosis in Duchenne muscular dystrophy is surgery.⁴⁻¹⁰ The main area of contention in surgical treatment is whether the fusion should end to L5 or extend to the pelvis. There are two schools of thought. One states that provided the spine is corrected and the pelvis is level after instrumentation, fixation can end at L5,^{6,7} the other states that problems with late pelvic obliquity can develop affecting sitting balance and hence the fusion should extend to the pelvis.^{4,10} Sussman⁷ reviewed 10 patients with muscular dystrophy treated with Luque segmental wiring from the upper thoracic spine to L5 and reported an excellent clinical outcome, concluding that fusion to L5 is adequate. Brooks⁴ compared 10 patients fused to L5 with seven patients having been fused to the pelvis using the Galveston construct. In the group fused to L5, four patients had problems with sitting balance and six patients lost correction, with no complications being reported in the Galveston group. They suggest that pelvic obliquity was better corrected and maintained by fixation to the pelvis. Fixation to the pelvis makes the operation longer, technically more difficult, and increases the potential for per-operative complications, especially blood loss. Surgery should be kept as simple as possible and any technique that makes the operation longer or more difficult must be of proven benefit to be justifiable. At the present therefore our practice is to fuse to L5, in the ma-

jority of these patients. The possibilities for basing fixation in the lumbar spine have been considerably increased by the use of pedicle screws, which provide a firm distal anchor for the fixation.

One of the most important factors in the reduction of the complication rate and improvement in the outcome of surgery in these patients has been the change in timing of surgery. Operating on the patients early before a severe fixed deformity is present makes the surgery easier, with a lower blood loss and produces a better correction so that it is rarely necessary to extend the fusion to the pelvis. A recent report by Sengupta et al. confirms that fixation to L5 is adequate if the surgery is performed early.⁸

Cardio-respiratory function in Duchennes muscular dystrophy

Cardiomyopathy is present to varying degrees in all boys with DMD. There is however, a small subgroup (approximately 10%) of patients who develop severe cardiomyopathy which results in early death.¹⁵

Respiratory function steadily deteriorates with time in children with DMD. The progressive myopathy causes an average reduction in vital capacity of 4% each year. A further 4% reduction in vital capacity is noted for every 10° progression of the Cobb angle of the scoliosis.¹³ A patient with a curve over 35° is usually associated with a vital capacity < 40% of the predicted normal¹⁴ (Fig. 3).

Correction of the spinal deformity does not in itself improve respiratory function.^{16, 17} However, Galasko et al. have reported an improvement in the peak expiratory flow rate after spinal stabilisation.⁵ Most other authors suggest that respiratory function initially deteriorates post-operatively before returning to the preoperative base-line level. Correcting the deformity and fusing the spine may, at best, delay the subsequent deterioration in respiratory function.

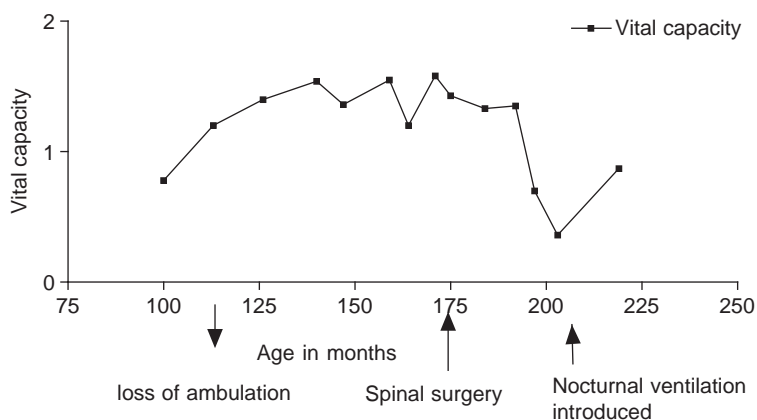


Figure 3. A typical curve demonstrating longitudinal changes in vital capacity in DMD.

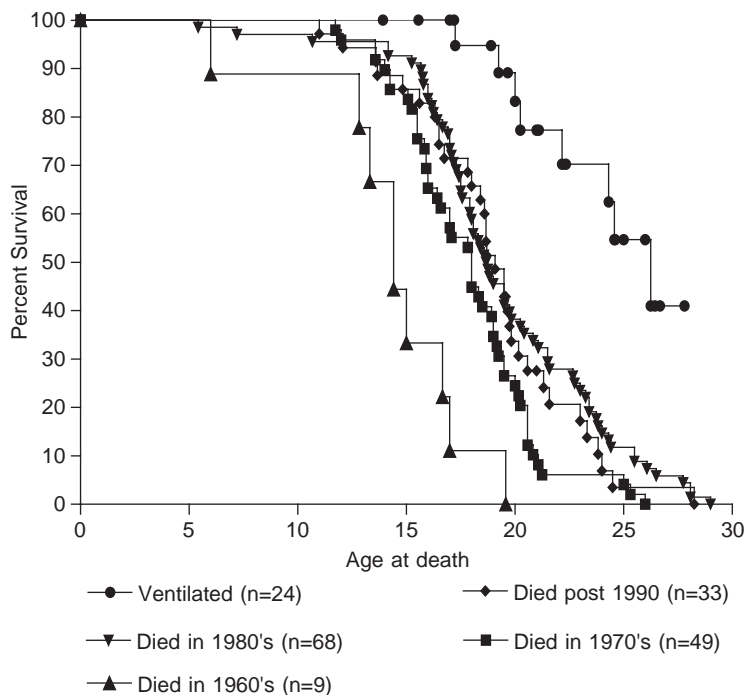


Figure 4. Impact of nocturnal ventilation on survival in DMD.

The early use of nocturnal home ventilation has proved valuable for these children after the onset of respiratory complications. A recent retrospective review from Newcastle documents the impact of nocturnal ventilation on the survival of such patients.¹⁵ Overnight oxymetry was commenced after the vital capacity dropped below 1 l/m. Children with respiratory symptoms and/or an abnormal oxymetry in association with deterioration in vital capacity were provided with nocturnal home ventilation. The mean survival in the non-ventilation group was 19.3 years, while that in the ventilation group was 25.3 years. The group of patients with early onset severe cardiomyopathy died younger with the mean survival in this group of only 16.2 years (Fig. 4).

We are currently evaluating the combined effects produced by nocturnal home ventilation and spinal surgery on a group of patients treated in our unit. Figure 3 shows the typical changes in vital capacity in relation to predictable milestones such as loss of ambulation and onset of the spinal deformity.

In some of the other less common forms of myopathic scoliosis, respiratory failure occurs relatively early in the course of the disease. These patients may require nocturnal ventilatory support at an early stage when they still have a reasonable life expectancy and sometimes when they are still ambulant. Anecdotal experience suggests that in this small group of patients correction of the scoliosis may produce an improvement in respiratory function in the short term.

CEREBRAL PALSY

Spinal deformity develops in 25% of patients with cerebral palsy. Spinal deformities develop most commonly in the patients who are the most severely disabled. Samilson reported the incidence of scoliosis in patients with cerebral palsy as being 7% in the ambulant patients, 39% in the bed care patients and 83.2% in patients with spastic quadriplegia.¹⁸

Understandably there is a significant resistance amongst the carers and health-care professionals, who look after these severely disabled children, to subject them to major spinal surgery. These patients therefore frequently present late to the spinal surgeon. By the time of presentation there is often a gross rigid scoliosis, usually associated with marked fixed pelvic obliquity. From the surgeons' perspective it is difficult to decide which of these severely disabled children should be offered surgery. The decision can only be made after the child is carefully assessed and a detailed discussion is undertaken with the family on the benefits and risks of surgery.

It is difficult to measure outcome in this group of patients and there are no good outcome studies looking at patients with CP following scoliosis correction. The decision will always be in part philosophical with some surgeons operating on most of the children because of the severity of their deformities, while others operate on very few because of the high complication

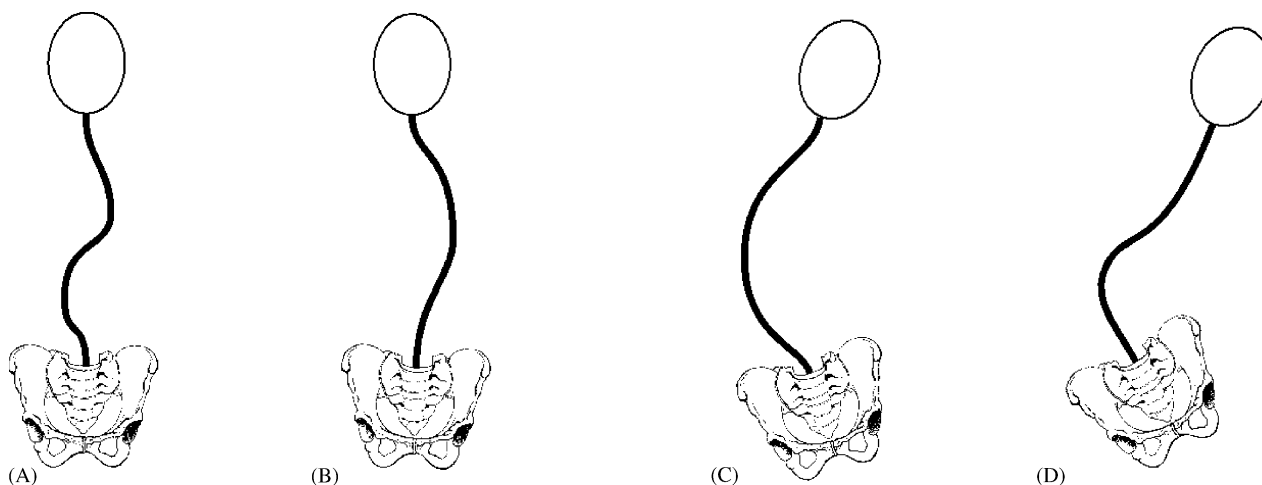


Figure 5. Curve types in cerebral palsy associated neuromuscular scoliosis. (A, B) represent Group 1. The spine is well balanced with little or no pelvic obliquity. (C, D) represent Group 2. The large lumbar or thoracolumbar curve extends into the sacrum with a significant fixed pelvic obliquity.

rate. Our approach is based on the basic premise that surgery is to maximise function. If the patient is so disabled that they can do very little, have no sitting balance, no head control and are unaware of the surroundings major spinal surgery is difficult to justify. When deciding not to operate it must be accepted that the patients may develop gross deformities, which can be distressing for both parents and carers. Furthermore, curve progression can compromise respiratory function and thereby shorten the patient's life expectancy.

In this group of patients fixed pelvic obliquity and hip dislocations frequently co-exist.¹⁹ It is often difficult to establish which of these occurred first. Hence it is also difficult to establish the order in which to address the problems surgically. Our approach is to operate on the hips first if they are symptomatic. If the hip dislocation is asymptomatic we correct the scoliosis first.

A variety of curve patterns are seen in patients with cerebral palsy and these have been classified by Lonstein and Akbarnia¹ (Fig. 5). Group 1 comprises the patients with double curves, that are either well balanced or have a small fractional lumbar curve with little or no pelvic obliquity. These are usually seen in ambulant patients (A, B). Group 2 curves comprise a large lumbar or thoracolumbar curve, extending into the sacrum with significant fixed pelvic obliquity seen most commonly in non-ambulant patients (C, D).

Treatment in group 1

These patients are ambulant, have a good trunk control and sitting balance. Here the treatment of choice is surgery. Indeed in these patients there is evidence to suggest the outcome is better if the surgery is performed before

the Cobb angle of the curve has reached 60°. When surgery is required it is usually possible to correct the scoliosis without extending the fixation to the pelvis.

Treatment in group 2

The treatment in this group is much more difficult since these are the most disabled patients. They present late with severe stiff curves and gross fixed pelvic obliquity. Surgical correction almost always involves fixation from T2 or T3 to the pelvis. If a reasonable correction is to be obtained it may also be necessary to perform an anterior release first.⁹ Combined anterior and posterior surgery also improves the fusion rate which is an advantage because pseudarthrosis is a significant problem when treating these severe deformities. This is, however, a major undertaking for these severely disabled children.

PRACTICE POINTS

- The goal of the treatment is to maximise function
- Bracing is of limited value
- The aim of surgery is to produce a balanced spine in the coronal and sagittal planes with a level pelvis and a solid fusion
- Early surgery reduces the complication rate, and may improve the outcome

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