Predominantly posterior instrumentation and fusion in neuromuscular and neurogenic scoliosis in children and adolescents

**Abstract**

We studied the results in 46 patients with neuromuscular and neurogenic scoliosis (average age 13.5 years, range 6–19 years) who had had posterior fusion with a modified Luque technique between May 1985 and June 1992. The main criteria to recommend surgery were curve progression, loss of balance when sitting, control of the head and difficulties in wearing an external orthotic support. The mean preoperative curve was 63°, the postoperative value was 24°, representing a correction of about 62%. The average number of stabilized segments was 13. In 39 out of 46 patients, lumbo-sacral fixation was included in the construct. Failure of implants, pseudarthroses and major losses of correction in purely neuromuscular scolioses could be avoided by using rigid segmental fixation and a dorso-lateral fusion with a mixture of autologous and allogeneous bone. The scoliosis most difficult to influence was found to be Friedreich’s ataxia. In Duchenne muscular dystrophy 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. Analysis of our series does not confirm the morbidity and complication rates of previous studies.

**Key words** Scoliosis · Neuromuscular · Posterior fusion · Adolescents

**Introduction**

Until the end of the 1970s operative treatment of neuromuscular scoliosis was performed only exceptionally and in few centres. In the last few years there have been several studies reporting encouraging results of operative treatment [1, 7, 8, 13, 17, 20]. This was made possible due to improvement in surgical techniques, using new implants, as well as to more successful peri- and postoperative management of patients.

The term “neuromuscular disease” is not used uniformly in the literature, however. In traditional terminology, neurogenic deformities of the spine are divided into upper motor neuron (encephalopathy, Friedreich’s ataxia, etc.) and lower motor neuron (polio, spinal muscular atrophy, etc.) according to the location of affection, and include cerebral palsy and poliomyelitis. Later classifications treat the two latter clinical pictures as independent entities [10]. In the comprehensive study by Daher [5] “neuromuscular scoliosis” even includes scoliosis in myelomeningocele (MMC). Since problems and operative techniques differ considerably, we shall not cover our MMC cases in this study. In children suffering from neuromuscular disease, secondary scolioses are frequent, with some authors reporting a prevalence of up to 60% [3, 13], the figure for spinal muscular atrophy sufferers could be as high as 85% [4]. In these patients, the gains to be made from surgical reduction of the scoliotic curve are very significant, especially where the capacity to walk has...
been lost, as for example in patients with Duchenne (MDD) muscular dystrophy, in whom scoliosis progresses rapidly, particularly following the prepubertal growth spurt. Such patients require a comprehensive orthosis in order to be able to sit, but even with this, there is loss of balance while sitting in the wheelchair, and problems arise from pressure points and intolerance of the corset [20].

Crippled posture causes psychological problems in the mostly intelligent children, which could be largely transformed into a very positive experience by surgical correction of the scoliosis. Another problem was the increasing restriction in vital capacity [16] particularly in cases of MDD, which further deteriorates due to the patient’s posture and as a result of forcing the thorax and abdomen into a corset. Operative treatment of these types of scoliosis offers patients the possibility of freeing themselves from external restrictions while improving their posture and quality of life.

Scoliosis develops somewhat differently in children with cerebral palsy; these patients, as well as their caregivers and parents had already had to deal with many different orthopaedic measures of either surgical or conservative nature for several years. The asymmetrically spastic muscles can cause contractual deformities in the hips, knees and feet even before scoliosis requires treatment. In this paper we shall focus on the problems of the spine only, since co-existing problems of the hip or lower limb were managed by our colleagues in paediatric orthopaedics. No operation was carried out simultaneously with spinal surgery in any patient.

All orthopaedic measures focused on improving the patient’s capacity to sit, reducing care measures, relieving pain and increasing the patient’s self-esteem.

Materials and methods

The cases of 46 patients with neuromuscular and neurogenic scoliosis (31 male, 15 female) were reviewed after surgery performed between May 1985 and June 1992. The distribution of the type of neuromuscular disease is shown in Table 1. The curve was thoracolumbar in 35 cases, thoracic in 9 and purely lumbar in 2 cases; 28 scolioses were converted to the right, 18 to the left.

Preoperative planning of each case was based on a total of nine radiographs, i.e. AP and lateral views in supine and sitting positions, side bending, maximal flexion and extension, as well as an AP view under maximal axial traction. In 11 patients, preoperative radiographs could only be taken in a supine position. The preoperative Cobb angle measured on average $63^\circ$ (range 28–140). In all 19 patients suffering from MDD, an echocardiograph and lung function test were carried out, showing a mean vital capacity of 54% (range 32–86%) of the norm (Table 2).

All operations were performed by the same surgeon (M.A.). Mean patient age at the time of operation was 13.5 years (range 6–19.5 years). Progression of the curve, impaired balance while sitting up, reasons of nursing, and anatomical limits to corset treatment were the predominant criteria for surgical indication. All patients had delayed extubation within the first 36 h after surgery to provide continuous adaptation to spontaneous respiration.

### Table 1 Different etiology of the treated scolioses ($n = 46$)

<table>
<thead>
<tr>
<th>Disease</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duchenne dystrophy</td>
<td>19</td>
</tr>
<tr>
<td>Cerebral palsy</td>
<td>8</td>
</tr>
<tr>
<td>Poliomyelitis</td>
<td>5</td>
</tr>
<tr>
<td>Friedreich’s ataxia</td>
<td>5</td>
</tr>
<tr>
<td>Spinal muscular atrophy</td>
<td>5</td>
</tr>
<tr>
<td>Myopathy (unclassified)</td>
<td>3</td>
</tr>
<tr>
<td>Syringomyelia</td>
<td>1</td>
</tr>
</tbody>
</table>

During an average postoperative period of supervision of 30 months (range 18–74 months) the following objective parameters were evaluated:

1. Loss of correction judged on the basis of the Cobb angle
2. Loosening of implants
3. Success of fusion radiologically
4. Sitting balance, i.e. the ability to sit upright without support from the hands, and
5. Bearing of the head, defined as the ability to hold the head upright over the trunk without any external support

Subjective assessment of the surgical result was carried out by the patient (except for these with cerebral palsy), the parents and the carers according to four categories based on an arbitrary scale (Table 3). This was compared with the objective assessment, which was scaled independently of the surgeon.

### Operative technique

The procedure most frequently used ($n = 39$) was a dorsal segmental instrumentation according to Luque with our own modified lumbosacral fixation (Table 4, Fig. 1).

In the remaining seven patients, dorsal stabilization and fusion using CD instrumentation was carried out in four cases and a two-step procedure in two cases, which consisted of a ventral derotation spondylodesis according to Zielke and subsequent dorsal instrumentation according to Harrington and Luque. In a 12-year-old girl suffering from syringomyelia and severe thoracic kyphosis, a ventral release, including interposition of autologous ribs, was performed first, followed by dorsal stabilization according to Harrington and Luque.

In all cases, extensive dorsolateral spondylodesis with a mixture of autogenous and allogeneic bone graft was performed. The source of the allogeneic bone graft was exclusively deep-frozen femoral heads, which were cut in to pieces and then passed through a bone mill during surgery. The serum of all donors of femoral heads had been previously screened for HIV, hepatitis and syphilis. The average number of segments stabilized was 13 (range 8–17).

When using the Luque sublaminar wiring technique, threads were first inserted with a Dechamps passing instrument; these threads then served to insert the Luque wires. In several hundred sublaminar wire insertions, this technique has proved to be harmless with no complications related to injuries of the dura or the neural structures. The two L-shaped Luque rods were anchored in the sacrum with CD sacral screws in the S1 pedicles and were inserted into the OS ileum through a horizontal hole in order to fix the spine to the pelvis (Fig. 1). Whenever possible, the S1 lamina was additionally used for sublaminar wiring, resulting in a three-point fixation of the distal rod with the sacro-pelvic complex. This allowed for manipulation and correction of pelvic obliquity as well as the lumbosacral hyperlordosis by prebending the rod. Once this three-point fixation had been achieved the rods were levered...