

Is there a role for selective anterior instrumentation in neuromuscular scoliosis?

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Abstract Our objective was to report on the clinical and radiological outcome from a cohort of patients with neuromuscular scoliosis who underwent selective anterior single rod instrumentation for correction of thoraco-lumbar and lumbar scoliosis. Traditionally combined anterior release with long posterior instrumentation has been advocated for the treatment of neuromuscular scoliosis. Neuromuscular curves tend to be long and may have significant pelvic obliquity. However, certain neuromuscular curves with minimal pelvic obliquity may lend themselves to selective anterior correction thereby saving motion segments and allow continued ambulation for those patients. Nine patients with neuromuscular scoliosis underwent selective anterior instrumentation between 1994 and 2000. The mean follow up was 2 years and 9 months (range 24–55 months). The clinical outcome (including parent and caregiver satisfaction), radiological outcome (Cobb angle, apical vertebral translation, pelvic obliquity, truncal shift, thoracic kyphosis, lumbar lordosis, sagittal vertical axis) and complications are reported. Subjective outcome was excellent in six patients and good in three. All nine patients retained their ability to walk. There were no neurological or vascular complications. Supplementary posterior surgery was required in two patients. The mean pre-operative Cobb angle of 52° (range 44–60) improved to 20° (range 10–28) at 3 months, achieving Cobb angle correction of 61% and was 19° (range 7–28) at final follow-up. The mean pre-operative compensatory

curve of 31° (range 20–42) spontaneously corrected to 18° (range 14–24) at 3 months and was maintained at 18° (range 10–26) at final follow up. The mean pre-operative pelvic obliquity of 7° (range 0–14) corrected to 4° (range 0–8) at 3 months and was 3° (range 0–8) at final follow up. Selective anterior instrumentation and fusion in carefully selected patients with neuromuscular scoliosis (short flexible curves, minimal pelvic obliquity, pre-operative walkers, slow or non-progressive pathology) appears to have satisfactory clinical and radiological outcome at least in the short-term.

Keywords Neuromuscular scoliosis · Anterior spinal instrumentation · Pelvic obliquity

Introduction

Surgical correction of spinal deformity in neuromuscular scoliosis has improved significantly over the past 20 years with major innovations in surgical technique and spinal instrumentation, coupled with advances in anaesthesiology, transfusion medicine, critical care and multidisciplinary preoperative and postoperative care of the patients [2, 11].

Traditionally anterior release followed by posterior instrumented fusion has been used to correct deformity in neuromuscular scoliosis. Neuromuscular curves tend to be long C shaped curves, and significant pelvic obliquity has lead many to perform long posterior fusions to include the sacro-pelvis.

Anterior surgery prevents the crankshaft phenomenon in skeletally immature patients, improves correction, and improves fusion rates especially in patients who have posterior element deficiency.

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Selective fusion via anterior instrumentation alone in patients with neuromuscular scoliosis has been recently reported in the literature [1]. Our study was performed on carefully selected neuromuscular patients who underwent selective anterior instrumentation of their thoraco-lumbar and lumbar curves at our institution between 1994 and 2000.

Materials and methods

A retrospective clinical and radiographic study of all patients undergoing selective anterior instrumentation between 1994 and 2000 for neuromuscular spinal deformity was carried out. During this period 148 neuromuscular patients underwent scoliosis correction; nine patients among these had selective anterior instrumentation for the correction of their scoliosis. The mean age at surgery was 14 years (range 11–24 years). The mean follow up period was 2 years and nine months (range 24–55 months). The group consisted of: one each of myotonic dystrophy, arthrogryposis, prune belly syndrome, muscle eye brain syndrome and five patients of spinal dysraphism (partial agenesis of corpus callosum with benign hydrocephalus, Downs syndrome with hydrocephalus, lipomyelomenigocele, lumbosacral diastematomyelia with sacral dysgenesis and syringomyelia following posterior cranial fossa decompression for Arnold Chiari malformation) (Table 1).

All patients were ambulatory. The aim of surgery was to prevent curve progression and achieve a solid fusion of a well balanced spine. A single anterior rod USS (Universal Spine System, Synthes, Switzerland) was used in all patients. Only the major curve was fused. Six patients had structural grafts inserted to improve and maintain lumbar lordosis [femoral ring allograft in three patients, Syncage (Synthes) in two patients and Moss Miami cage (DePuy Acromed,

Raynham, MA, USA) in one]. None of the patients were braced in the postoperative period. Two surgeons performed the procedures (BJF, JKW).

Clinical outcome was assessed from a patient, parent or caregiver completed questionnaire at each follow-up appointment. The respondent was asked to grade the outcome as: (1) excellent, (2) satisfactory, (3) neither satisfactory or unsatisfactory, (4) unsatisfactory and (5) poor. Clinical photographs of the patient were taken pre-operatively and at latest follow-up.

Radiological outcome (Tables 2, 3) was assessed using pre-operative Cobb angle from standing radiographs, supine bending radiographs, 3 months standing radiographs and final standing radiographs. The apical vertebral translation (AVT), truncal shift (coronal balance), thoracic kyphosis, lumbar lordosis, and sagittal vertical axis (SVA) were all measured pre- and post-operatively. Pelvic obliquity (angulation of the pelvis from the horizontal in the frontal plane) was measured on an antero-posterior plain radiograph as an angle formed between a line joining two iliac crests and a horizontal line. For the two patients that required posterior instrumentation, their measurements were excluded from the detailed radiological outcome. Complications including neurological deficit, loss of correction, pseudarthrosis, implant failure and revision surgery were recorded.

Results

Subjective outcome was scored as excellent in six patients and good in three patients. All patients remained ambulatory. There were no neurological or vascular complications. Supplementary posterior instrumentation was required in two patients (one 5 mm rod breakage at 4.5 months postoperatively and one upper thoracic curve progression at 3 years). None of the patients had infection and one patient as mentioned

Table 1 Patient demographics and pelvic obliquity

Case	Sex	Age	Diagnosis	Age at menarche	Risser grade	Pelvic obliquity degrees
1	M	14	Arthrogryposis	NA	4	10
2 ^a	M	11	Syringomyelia	NA	0	5
3	F	11	Muscle eye brain syndrome	Pre-menarche	0	14
4	F	24	Prune belly syndrome	NR	5	7
5	F	12	hydrocephalus	9	4	8
6	F	15	Lipomyelomenigocele	15	4	11
7	F	14	hydrocephalus	Pre-menarche	5	0
8	F	15	Myotonic dystrophy	15	4	0
9 ^a	M	14	Diastematomyelia, sacral dysgenesis	NA	0	5

NA Not applicable, NR not recorded

^aPatients with supplementary posterior surgery

Table 2 Cobb angles and flexibility of the major curves along with correction achieved, levels fused and levels saved in each patient

Case	Major cobb angle	Bending cobb (Major)	Flexibility (%)	Final major cobb	Levels fused	Levels saved
1	52 (T8–L3)	22	58	24	T10–L2	2
2	70 (T9–L3)	24	66	sps	T10–L3	sps
3	55 (T9–L4)	18	67	16	T10–L4	1
4	45 (T11–L3)	20	56	7	T11–L3	1
5	52 (T11–L3)	22	58	18	T11–L3	1
6	53 (T11–L4)	26	51	23	T11–L4	1
7	60 (T11–L4)	32	47	28	T11–L4	1
8	44 (T11–L4)	13	70	19	T11–L3	2
9	54 (T11–L3)	17	69	sps	T11–L3	sps

Levels saved compared to posterior fixation based on Harrington stable zone

sps Supplementary posterior surgery

Table 3 Table illustrating preoperative and final sagittal and coronal balance along with Cobb angles of the minor curves in each patient

Case	Minor cobb angle	Bending cobb (minor)	Final minor cobb	Pre operative coronal balance (cm)	Final coronal balance (cm)	Pre operative sagittal vertical axis (SVA) (cm)	Final SVA (cm)
1	30 (T1–T8)	14	26	4 (L)	2 (L)	+5.6	+1.4
2	50 (T2–T8)	35	sps	0	sps	–2.8	sps
3	32 (T1–T8)	18	20	1.1 (L)	2.5 (R)	+2.7	+1.5
4	20 (T7–T11)	3	10	3.2 (L)	1.3 (L)	+4	–2.0
5	29 (T6–T10)	10	16	4.7 (L)	0.5 (L)	+1.8	0
6	42 (T2–T11)	0	18	0.5 (R)	0.4 (R)	–3.5	–2.2
7	35 (T6–T10)	20	22	5.5 (R)	1.1 (R)	–3.5	–2.2
8	28 (T3–T10)	12	18	3 (L)	2 (L)	+0.7	–1.6
9	52 (T3–T10)	22	sps	0.5 (L)	sps	+6.5	sps

sps Supplementary posterior surgery, L Left, R right

above developed rod breakage due to pseudarthrosis. (We feel this happened too early in the postoperative period to be due to pseudarthrosis alone and it was due to combination of small size rod and non compliance with recommendation of sporting activities.)

The mean preoperative Cobb angle of the major curve was 52° (range 44–60) correcting to 20° (range 10–28) at 3 months postoperatively (61% correction) and was maintained at 19° at final follow up (range 7–28).

The mean preoperative compensatory upper thoracic curve was 31° (range 20–42) which spontaneously corrected to 18° (range 14–24) at 3 months postoperatively and was maintained at 18° (range 10–26°) at final follow up (Cobb angle correction of 42%).

The mean pre-operative apical vertebral translation (AVT) was 5.7 cm (range 5–7.5 cm) correcting to 2.7 cm (range 2–3.7 cm) at 3 months postoperatively (54% correction) and was 2.1 cm (range 1–3.1 cm) at final follow up.

The mean preoperative pelvic obliquity was 7° (range 0–14) which spontaneously corrected to 4° (range 0–8) at 3 months postoperatively, and was 3° (range 0–8) at final follow up.

Preoperatively five patients were off balance in the coronal plane by more than 2 cm (range 3.1–5.5 cm). One patient was off balance in the coronal plane (2.5 cm) at 3 months postoperatively and at final follow up.

The mean preoperative thoracic kyphosis was 29° (range 15–40) increasing to 37° (range 26–45) 3 months postoperatively and was 35° (range 24–45) at final follow up.

The mean preoperative lumbar lordosis was 44° (range 34–60). It was 43° (range 36–55) 3 months postoperatively and 44° (range 32–55) at final follow up.

Preoperatively five patients had a SVA of more than 2 cm (range – 3.5 to + 5.6 cm). Two patients had SVA of more than 2 cm at 3 months postoperatively (– 3.5 cm, + 4 cm) and at final follow up (–2.2, –2.2 cm).

The two cases that subsequently required posterior instrumentation are discussed below:

Case 1: 10-year-old boy with neuromuscular scoliosis due to syringomyelia. Posterior cranial fossa decompression was performed when he was 9 years old with regression of syrinx noted on follow up MRI scan. The thoraco-lumbar curve measured 70° (T9–L3) bending

down to 24° and the thoracic curve measured 50° (T2–T8) bending down to 35°. He underwent anterior correction and instrumented fusion from T10 to L3 using a 5 mm paediatric USS rod. At 4.5 months the patient had been playing football and presented with pain. Plain radiographs revealed a broken rod at the L1/L2 level. He underwent successful posterior instrumentation from T4 to L4 with the left thoracic curve correcting to 28° and the right thoraco-lumbar curve correcting to 12° at final follow-up.

Case 2: 14-year-old boy with sacral dysgenesis, lumbosacral myelomeningocele and mildly decreased lower limb function. He was a national level wheel chair basketball player. His lumbar curve measured 54° (T11–L3) bending down to 17°, thoracic curve of 52° (T3–T10) bending down to 22°. Due to his national level sporting activities, he underwent T11–L3 anterior instrumentation in order to save distal lumbar motion segments. At 3 years following surgery the thoracic curve progressed above the level of instrumentation requiring supplemental posterior instrumentation (T2–L1).

Discussion

Neuromuscular scoliosis is a coronal plane deformity of the spine in patients with abnormalities of the myoneural pathways. This diverse group of diseases are characterised by a breakdown in the normal neural integrated pathway that includes the brain, spinal cord, peripheral nerves, and/or muscle [9]. The well recognised general diagnostic and treatment principles for these group of patients include: (1) prompt identification of the underlying neuromuscular condition, (2) early surgical correction of the primary condition if possible, (3) orthotic bracing to improve functional capacity and postpone surgical intervention in small curves, and (4) operative spinal fixation for large, progressive or functionally debilitating curves [2, 9, 11].

Due to the numerous causes of neuromuscular scoliosis, the pattern and incidence of scoliosis varies with each patient. Although most present with progressive long C-shaped curves often with significant pelvic obliquity, some neuromuscular conditions produce curve patterns similar to those seen in adolescent idiopathic scoliosis [9]. In younger adolescents with limited growth remaining, it has been recognised that posterior-alone surgery may be adequate for arthrodesis and maintenance of correction [2, 4, 12]. In selected patients with major curves that do not include the pelvis and pelvic obliquity of less than 15°, arthrodesis terminating in the lumbar spine may avoid complications associated with fusion to the sacro-pelvis

and may permit improved mobility [13, 14]. Broom et al. [4] considered it mandatory to extend fusion to the pelvis when (1) truncal decompensation occurs such that the vertical plumb line falls *lateral* to the sacroiliac joint, (2) fixed pelvic obliquity of more than 15°, and (3) when the sacrum appears to be part of the curve.

Though the posterior approach is considered less difficult and is the most familiar route for most surgeons, anterior procedures for the spine are being performed more frequently. The main advantage of this approach is that, selective fusion of the major curve alone helps to preserve distal lumbar motion segments, which is important to ambulatory patients. Cochran et al. [5] suggested that when a Harrington rod was used to correct scoliosis, there was increased incidence of disc degeneration and back pain when the instrumentation was distal compared to proximal fixation. Long term follow up with larger number of patients is necessary to study the effects of anterior instrumented correction and fusion on the free disc below the level of fusion. Broom et al. [4] suggested that when the posterior approach is chosen, proximal spinal fixation should extend to at least T3 due to the relative frequency of proximal junctional kyphosis when the construct ended at T4 or below.

Due to the three dimensional nature of a spinal deformity, anterior release can lead to improved curve correction and provide greater surface area between vertebral end plates thereby promoting increased fusion rates. The additional area of fusion is especially important in myelodysplastic patients without posterior elements and neuromuscular patients with severe osteopenia. Anterior release destroys the growth plates of the vertebral bodies and therefore prevents crankshaft phenomenon in skeletally immature patients requiring correction of their scoliosis [6].

The main disadvantage with anterior surgery is that it is kyphogenic, and structural grafts may be required to maintain the lordosis. Rigid packing of the disc space with morcelized grafts and contouring of the rods may be sufficient to prevent kyphosis. Implant failure has been associated with anterior instrumentation alone, particularly with smaller diameter rods; indeed one of our cases involved breakage of a 5 mm rod at 4.5 months postoperatively. Improvement in implant design with stronger rods and larger diameter vertebral body cancellous screws with superior pull-out strength has led to a reduction in failure of the stand-alone anterior instrumentation. Meticulous surgical technique with achievement of bone on bone apposition after anterior release and usage of rib auto graft/structural grafts also helps to decrease the incidence of

pseudarthrosis and metalwork failure. Our study, admittedly with relatively short-term follow up identified only one case of pseudarthrosis. Longer term follow up with larger number of patients is required to study true incidence of pseudarthrosis.

Despite the increased use of anterior instrumentation for adolescent idiopathic scoliosis, it has been used very rarely in patients with neuromuscular scoliosis [1, 7, 10]. Recently Basobas et al. [1] reported on such a series. In this study 29% of patients were non-ambulatory and some patients had pelvic obliquity of greater than 15°. The weaknesses of this study are: (1) combined data that included a mixture of thoracic, thoracolumbar and lumbar curves, (2) not all patient were ambulatory, (3) some patients had pelvic obliquities of greater 15°, and (4) use of four different types of instrumentation systems. In our study, we report the results of surgery on thoracolumbar and lumbar curves, all of our patients were ambulatory and had pelvic obliquity of less than 15°, and only one instrumentation system was used.

Sarwahi et al. [10] also reported that 30% of their patients underwent anterior instrumentation alone for neuromuscular scoliosis whilst others underwent either combined or staged procedures for correction of their scoliosis. They also report that 37 patients had previous spine surgery; it is not clear from their paper whether these were the patients who had anterior instrumentation alone. Hopf et al. [7] reported 33 neuromuscular patients treated by anterior instrumentation. Seventeen patients underwent secondary posterior instrumentation. The ambulatory status, degree of pelvic obliquity, coronal and sagittal balance were not dis-

cussed in this study. Sixteen of these patients had myelomeningocele which suggests that the selection of the anterior approach may have been due to the lack of posterior elements, rather than saving the motion segments and preserving the walking abilities of the patient.

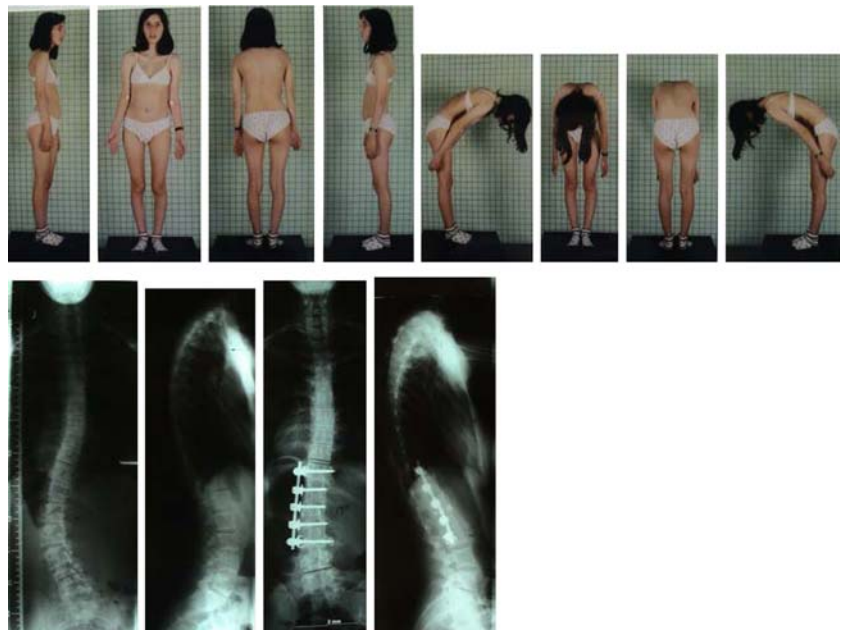
Our study showed that patients with non-progressive pathology with curve patterns similar to idiopathic scoliosis (Group 1, Lonstein and Akbarnia) [8] may be treated with anterior instrumentation.

Conclusion

Anterior instrumentation alone in carefully selected patients with neuromuscular scoliosis results in acceptable clinical and radiological results at least in the short-term. Patients ideally should have flexible thoraco-lumbar or lumbar curves with minimal pelvic obliquity and non-progressive pathology. Anterior surgery achieves better de-rotation and curve correction whilst preserving distal lumbar motion segments [3] important in ambulatory patients. Our study suggests that there is a role for anterior instrumentation in carefully selected patients with neuromuscular scoliosis.

Illustrative case example

Fifteen-year-old female with myotonic dystrophy and a double curve (menarche 15 years). Magnetic resonance imaging showed no evidence of intraspinal



pathology. Plain radiographs revealed Risser grade 4, lumbar curve 44° (T11–L4) bending down to 13°, thoracic curve 28° (T3–T10). The patient underwent anterior instrumentation from T11 to L3 achieving a Cobb angle of 19° which was maintained at the final follow-up (4.5 years). Thoracic curve spontaneously corrected to 18° at final follow up.

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