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ORIGINAL ARTICLE

Can the caudal extent of fusion in the surgical treatment of scoliosis in Duchenne muscular dystrophy be stopped at lumbar 5?

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Abstract Instrumentation and fusion to the sacrum/pelvis has been a mainstay in the surgical treatment of scoliosis in Duchenne muscular dystrophy (DMD) and is recommended to correct pelvic obliquity. The caudal extent of instrumentation and fusion in the surgical treatment of scoliosis in DMD has remained a matter of considerable debate, and there have been few studies on the use of segmental pedicle screw instrumentation for this pathology. From 2004 to 2007, a total of 28 patients with DMD underwent segmental pedicle screw instrumentation and fusion only to L5. Assessment was performed clinically and with radiologic measurements. All patients had a curve with the apex at L2 or higher preoperatively. Preoperative coronal curve averaged 74°, with a postoperative mean of 14°, and 17° at the last follow-up. The pelvic obliquity improved from 17° preoperatively to 6° postoperatively, and 6° at the last follow-up. Good sagittal plane alignment was recreated after surgery and maintained long term. In 23 patients with a preoperative L5 tilt of less than 15°, the pelvic obliquity was effectively corrected to less than 10° and maintained by adequately addressing spinal deformity, while five patients with a preoperative L5 tilt of more than 15° had a postoperative pelvic obliquity of more than 15°. Segmental pedicle screw instrumentation and fusion to L5 was effective and safe in patients with DMD scoliosis with a minimal L5 tilt (<15°) and a curve with the apex at L2 or higher, both initially and long term, obviating the need for fixation to the sacrum/pelvis. Segmental pedicle screw instrumentation and fusion to L5 was safe and effective in patients with DMD scoliosis with stable L5/S1 articulation as evidenced by a minimal L5 tilt of less than 15°, even though pelvic obliquity was significant. There was no major complication. With rigid segmental pedicle screw instrumentation, the caudal extent of fusion in the treatment of DMD scoliosis should be determined by the degree of L5 tilt. This method in appropriate patients can be a viable alternative to instrumentation and fusion to the sacrum/pelvis in the surgical treatment of DMD scoliosis.

Keywords Duchenne muscular dystrophy · Scoliosis · Pelvic obliquity · Lumbar 5 tilt · Pedicle screw instrumentation

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Introduction

Duchenne muscular dystrophy (DMD) is the most common and severe form of muscular dystrophy and is a flaccid neuromuscular disorder in which gradual deterioration is the hallmark [7, 11, 19, 22, 27, 29, 33]. Spinal deformity such as scoliosis is almost universal in patients with DMD [7, 11, 19, 22, 27].

Non-operated scoliotic curves in patients with DMD progress relentlessly to levels as great as 80° or more [31, 33]. Natural history studies on neuromuscular scoliosis demonstrated an almost invariable progression [2, 18, 31, 33]. The progression of untreated spinal deformity in this population causes increasing pain [14, 35], difficulties in sitting [2, 9, 22, 27], decreased pulmonary function [4, 27] and an increase in mortality [28]. Surgical management is the most reliable treatment for this deformity. Posterior spinal fusion in patients with DMD has been accepted as an optimal procedure to stabilize the spine and maintain an upright and comfortable sitting balance [5, 9, 13].

Major changes affected scoliosis surgery: earlier spinal stabilization and improvement in implants and instrumentation technique. However, most reports on scoliosis surgery in patients with DMD have dealt with spinal instrumentation using hooks, wires and hybrid constructs with lumbar pedicle screws [4, 12, 17, 22, 23, 30], yet it is difficult to provide strong and stable fixation with hook and wire anchors in patients with DMD [23, 38].

The development of the intrailiac post by Allen and Ferguson [1] has provided a reliable means to achieve pelvic fixation and address pelvic obliquity. Several studies have reported that instrumentation and fusion to the sacrum/pelvis should be reserved for patients with large coronal curves (>40°) and severe pelvic obliquity (>10°) [27, 35, 37]. However, the indications for extending the instrumentation and fusion to the pelvis/sacrum have remained controversial [23, 30]. Furthermore, pelvic or sacral fixation has several disadvantages [25, 27].

The purpose of this study was to determine the efficacy and safety of stopping segmental pedicle screw instrumentation constructs at L5 in the treatment of scoliosis in DMD.

Patients and methods

Patients

From December 2004 to October 2007, a total of 28 consecutive patients with DMD underwent posterior spinal fusion and segmental pedicle screw instrumentation for scoliosis at our institute. All patients were non-ambulatory. All the operations were performed by the same surgeon (M.T.).

Surgical techniques

The primary aim of the surgery was to obtain a solid fusion, level pelvis, and a balanced spine in the coronal and sagittal planes in these patients. The incision was midline and extended over. The posterior elements of the spine were exposed from the upper thoracic spine to the sacrum by stripping the muscles subperiosteally. The spinous process, the lamina, and the base of the transverse process of the vertebrae were stripped of periosteum. After removal of all soft tissue, local autograft bone was obtained from the spinous process, laminae, and transverse process of all the vertebrae, which did not support instrumentation, as a bone graft source. All articular facets were removed carefully. Spinal cord function was monitored throughout the procedure. Autotransfusion via preoperative storage and intraoperative collection was used. Correction of the curves was maintained by segmental pedicle screw and rod instrumentation. Instrumentation was Expedium instrumentation with a 5.5 mm titanium rod (Expedium, DePuy-Acromed, Raynham, MA, USA). Pedicle screws with a diameter of 5 mm were used in the thoracic and lumbar spine. Cross-links were employed as needed. The pelvis was not included in the instrumentation. All curves were instrumented and fused from T3 or T4 to L5 regardless of the severity of spinal deformity and pelvic obliquity. For the segmental pedicle screw instrumentation, every level was instrumented on at least one side. No image-guided spinal navigation system was employed. Screws were placed with free-hand technique. Fluoroscopy was employed to confirm acceptable screw position. The segmental pedicle screw correction was performed with rod insertion, rod rotation, translation of the rod, appropriate distraction and compression to level the proximal and distal end vertebra. Extensive posterior elements decortication for local bone graft was performed using a motorized gouge. The local bone graft was packed onto the prepared surfaces and placed carefully in each facet. The wound was sutured in three layers with two drainage tubes.

Radiographic assessment

Preoperative and postoperative radiographic evaluation included sitting posterior–anterior (PA) and lateral radiographs and best-effort spine side-bending radiographs. The sitting PA radiograph and side-bending films were examined and Cobb angles of the curve were needed to determine flexibility and correction. Sitting PA and lateral radiographs were taken the day before surgery, in a week after surgery and thereafter at a 3-month interval after surgery. On the coronal plane, the Cobb angles of the curves were measured. Lumbar 5 (L5) tilt (the angle between a line across the top of L5 and the intercristal line) and spinal pelvic obliquity (SPO) (the angle between the perpendicular of the spine line from T1 to S1 and a line across the top of the pelvis) were measured (Fig. 1). There have been some definitions of pelvic obliquity. It is important to identify a method to measure pelvic obliquity. SPO is the most commonly used method to calculate pelvic obliquity, while the line from T1 to S1 is not necessarily the same as the line that represents the obliquity of the trunk and the chest cage. However, several studies have concluded that obliquity measured by SPO is the most reliable and the standard method by which pelvic obliquity can be defined [15, 23]. Therefore, the current authors used SPO to measure the pelvic obliquity in this study. On the sagittal plane, thoracic kyphosis between T3 and T12, and lumbar lordosis between L1 and L5 were measured. Fusion was defined as: first, stable coronal and sagittal alignment over the follow-up period; second, no clinical complaints; third, no evidence of nonunion radiographically; and fourth, stable instrumentation. All four criteria must be present for the definition of fusion.

Results

Twenty-eight patients who were followed up for a minimum of 2 years were enrolled into this study. No patients were lost to follow-up. Demographic details and surgical parameters for the study group patients are demonstrated in Table 1. Radiographic measurements in the coronal and sagittal plane alignment are demonstrated in Table 2. Changes in the pelvic obliquity and L5 tilt are demonstrated in Fig. 2. The mean age at surgery was 13 years and 6 months (range: 11 years 8 months–14 years 10 months). All scoliosis curves were single curves (23 right thoracolumbar, three left thoracolumbar and two right thoracic curves). The mean operating time was 282 min (range: 232–308 min). The mean intraoperative blood loss was 950 ml (range: 660–1,260 ml). The mean total blood loss was 2,150 ml (range: 1,250–2,880 ml). The mean follow-up period was 40 months (range: 24–57 months).

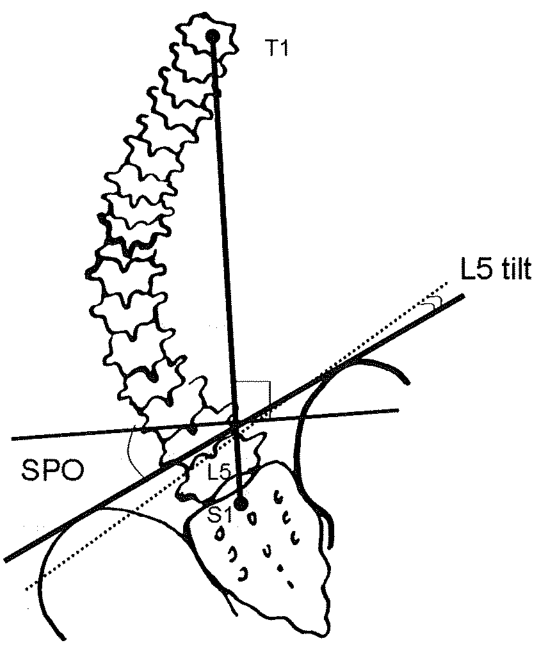

Fig. 1 The technique of determining the L5 tilt and spinal pelvic obliquity (SPO). L5 tilt (the angle between a line across the top of L5 and the intercristal line) and spinal pelvic obliquity (the angle between the perpendicular of the spine line from T1 to S1 and a line across the top of the pelvis) were measured

During hospitalization, most patients reported back pain in the region of surgery. At 3 months after surgery, no patients reported back pain and clinical complaints in the region of surgery. On the coronal plane, the mean preoperative coronal curve measured 74° (range: 51–88°). The mean preoperative coronal curve on best-effort spine side-bending radiograph was 16° (range: 10–27°) and the preoperative flexibility of the curve averaged 78%. The mean immediate postoperative coronal curve measured 14° (range: 8–25°) (81% correction). At the last follow-up, the mean curve measured 17° (range: 9–27°) and we noticed less than 3° in the loss of correction in all patients at the last follow-up. On the sagittal plane, the mean preoperative sagittal thoracic curve (T3–T12) measured 10° (range: −8° to 23°). The mean immediate postoperative sagittal thoracic curve measured 20° (range: 12–28°). At the last follow-up, this curve measured 21° (range: 13–28°). The mean preoperative sagittal lumbar curve (L1–L5) measured 15° (range: −18° to 58°), including many kyphotic patients and the mean immediate postoperative curve measured 34° (range: 8–45°). This curve measured 35° (range: 7–44°) at the last follow-up. Therefore, no significant loss of correction in the coronal and sagittal plane was noted between postoperative time and the last follow-up. The L5 tilt improved from a preoperative mean of 9° (range: 5–20°) to a postoperative mean of 3° (range: 0–18°) and 3° (range: 0–20°) at the last follow-up. The pelvic obliquity improved from a preoperative mean of 17° (range: 10–38°) to a postoperative mean of 6° (range: 2–22°) and 6° (range: 2–25°) at the last follow-up. We noticed no significant loss of correction of pelvic obliquity and L5 tilt.

Of the 28 patients, 23 had an SPO of less than 10° postoperatively. A good pelvic balance was achieved and maintained despite the fusion only to L5 in these patients. These 23 patients had a preoperative L5 tilt of less than 15° and the SPO was effectively corrected to less than 10° and maintained by adequately addressing spinal deformity (Table 2, Fig. 2). Figure 3 shows a radiographic example. Of the 28 patients, five had an SPO of more than 15° postoperatively. In these five patients, scoliotic curvature

Table 1 Details of the patients and operative parameters in the study group

Patient no.	Age	Follow-up (month)	Operative time (min)	Intraoperative blood loss (ml)	Total blood loss (ml)	Complications
1	13	57	278	980	2,050	(−)
2	13	56	285	990	2,200	(−)
3	12	56	284	1,000	3,200	(−)
4	11	55	248	880	2,110	(−)
5	13	55	260	870	1,880	Paralytic ileus
6	13	54	270	900	1,920	Paralytic ileus
7	13	50	266	780	2,230	(−)
8	13	48	277	900	1,670	(−)
9	12	44	308	890	1,890	(−)
10	13	44	245	890	2,000	(−)
11	14	42	267	970	2,100	(−)
12	13	40	278	900	2,200	(−)
13	11	39	299	1,260	2,780	(−)
14	13	39	232	660	1,250	(−)
15	11	36	278	780	1,880	Paralytic ileus
16	13	35	269	890	2,000	(−)
17	13	32	243	820	2,200	(−)
18	13	32	301	1,120	2,880	(−)
19	12	31	278	820	1,970	Paralytic ileus
20	11	31	279	980	2,230	Paralytic ileus
21	12	30	255	740	1,990	(−)
22	13	29	272	990	2,620	(−)
23	13	29	289	880	2,340	(−)
24	14	28	288	1,030	2,200	(−)
25	14	28	290	1,040	2,360	(−)
26	11	26	260	980	2,030	(−)
27	12	26	255	960	1,870	(−)
28	13	24	250	860	1,950	(−)
Mean	13	40	282	950	2,150	

was well corrected and maintained but a residual SPO of more than 15° was observed (Table 2, Fig. 2). These five patients had an L5 tilt of more than 15° preoperatively. Figure 4 shows a radiographic example.

The mean number of levels fused was 14.5 (range: 14–15). In the 28 patients, a total of 616 screws was placed safely with no reoperations for screw malposition. The mean number of screws per patient was 22. Clinically, there were no neurologic deficits or radicular symptoms.

There were five postoperative complications. All complications were paralytic ileus, occurring in five patients, which resolved with observation without oral intake in at least 48 h. There were no neurologic complications, instrumentation failure, pull out of fixation or infections. There were no reoperations for any reason, nor any second hospitalizations related to scoliosis surgery.

Discussion

Spinal deformity is almost universal and progressive in patients with DMD [7, 11, 19, 29, 33]. The posterior spinal fusion for neuromuscular scoliosis has been highly effective in stabilizing the spine and maintaining an upright and comfortable sitting balance [5, 9, 13].

The most recent instrumentation innovation for the treatment of scoliosis is the pedicle screw. To our knowledge, this study is the only reported series of consecutive cases of scoliosis in DMD treated with segmental pedicle screw instrumentation and fusion to L5. Excellent minimum 2-year results are shown in this study, with no reoperations for nonunion, infection or instrumentation failure. Radiographically, the coronal curves were flexible (78% compensation on preoperative best-effort spine side-bending radiographs) preoperatively and there was 81%

Table 2 Radiographic measurements in the study group

Patient no.	Curve apex	Scoliotic curvature (°)					Thoracic kyphosis (°)				Lumbar lordosis (°)			
		Preop	Side bend	Immediate postop	2-year postop	The last	Preop	Immediate postop	2-year postop	The last	Preop	Immediate postop	2-year postop	The last
1	L1	86	17	15	15	17	20	22	23	22	8	35	36	35
2	T12	95	21	20	19	20	8	28	29	28	0	28	30	30
3	L1	86	16	14	16	16	10	24	24	23	8	40	41	38
4	L1	80	10	8	8	9	23	20	21	21	25	30	28	28
5	L1	60	13	10	10	11	5	18	20	20	10	35	38	38
6	L1	72	10	9	9	10	10	28	26	26	8	30	32	32
7	T12	65	14	10	11	11	5	20	22	22	5	38	40	40
8	T11	55	16	13	13	15	5	18	19	19	39	32	30	30
9	T12	70	19	15	17	16	−7	22	24	24	47	37	38	38
10	L1	51	14	12	13	15	5	12	13	13	0	45	42	42
11	T11	81	19	17	18	20	6	16	18	18	6	36	35	35
12	L2	72	19	15	16	18	−8	20	22	21	58	28	29	29
13	T10	71	20	18	18	19	10	15	18	18	10	33	35	38
14	L1	62	14	10	11	11	22	20	22	20	−18	8	8	7
15	T12	81	13	12	14	13	5	17	20	20	55	42	45	44
16	L1	78	27	25	28	27	5	15	17	17	5	35	38	38
17	LI	58	19	18	22	22	13	18	20	20	36	43	40	41
18	L1	70	16	15	16	18	3	20	24	24	13	37	39	39
19	T9	85	16	15	16	17	−8	20	22	22	58	28	28	29
20	L1	75	18	15	17	17	11	14	16	16	−6	31	33	34
21	L2	68	20	18	20	22	5	20	17	17	11	45	45	43
22	T12	77	20	18	18	19	20	25	19	19	10	30	32	32
23	T10	71	23	20	21	22	5	28	27	27	6	35	38	38
24	T11	72	17	15	16	16	20	22	22	24	0	36	37	38
25	T12	66	10	8	8	10	18	25	26	27	8	38	37	38
26	L2	86	12	10	12	12	22	26	27	25	12	38	38	35
27	L1	55	15	12	12	13	8	22	25	27	−7	35	36	35
28	T12	88	17	16	15	16	6	26	28	28	−8	33	36	34
Mean		74	16 (78%)	14 (81%)	15	17	10	20	21	21	15	34	35	35

coronal curve correction and normalization of sagittal plane postoperatively. Loss of correction was minimal at the last follow-up. Good pelvic balance was achieved and maintained in all but five patients.

In the surgical treatment of spinal deformity in DMD, strong anchors are needed because of increased loss of muscular stability in neuromuscular disorders that causes asymmetrical muscle balance leading to progression of spinal deformity [8, 38] and because of osteopenia in this population [6, 24, 38]. In addition, it is difficult to provide strong and stable fixation with hook or wire anchors [38]. Pedicle screws offer a better vertebral grip with three-column purchase and a longer moment arm due to the anatomic location of the pedicle screw as compared to other forms of spinal bone–implant interfaces such as hook placement on the laminar or sublaminar wires. The screws are immediately stable in all directions after insertion. The superior biomechanical advantages of pedicle screws over other forms of spinal bone–implant interfaces allow the correction technique to generate powerful corrective forces in all planes. Segmental pedicle screw instrumentation has been reported to offer a significant coronal curve correction and maintenance of correction in patients with adolescent idiopathic scoliosis [20, 36]. In addition, significant correction and maintenance of correction in sagittal plane alignment has been reported with the use of segmental pedicle screw instrumentation as compared to those with other forms of instrumentation in patients with adolescent idiopathic scoliosis [20, 36]. Although pedicle screws have been thought to carry a higher complication risk

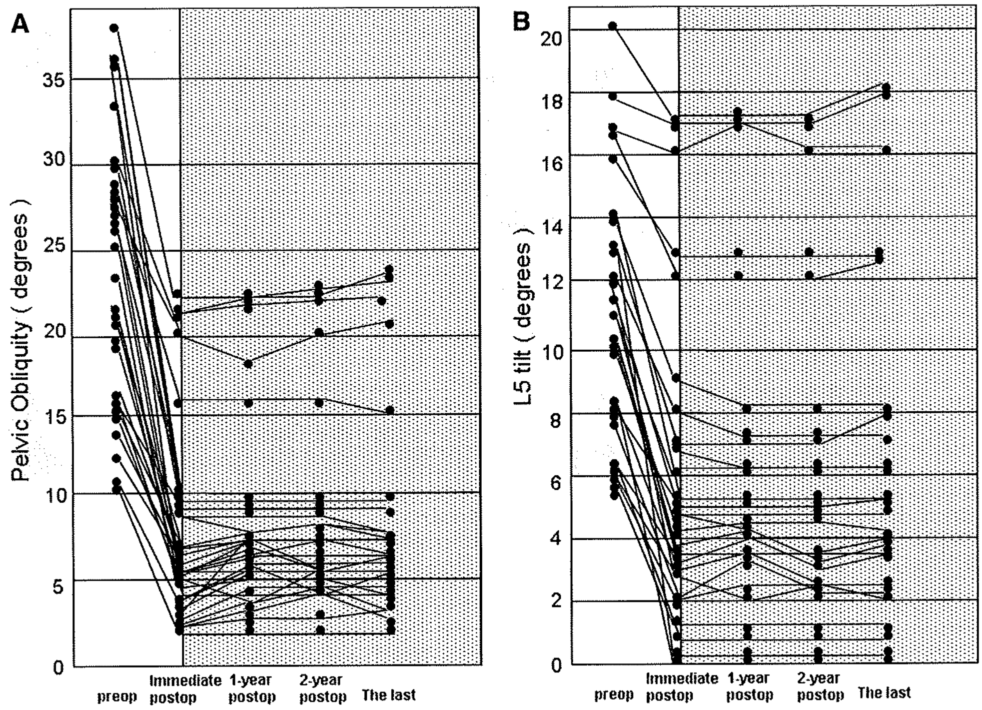

Fig. 2 Changes in pelvic obliquity (**a**) and L5 (**b**). Twenty-one of 25 patients had pelvic obliquity of less than 10° postoperatively. All these patients had L5 tilt of more than 15° preoperatively. Four patients had pelvic obliquity of more than 10° postoperatively and all these patients had L5 tilt of more than 15° preoperatively

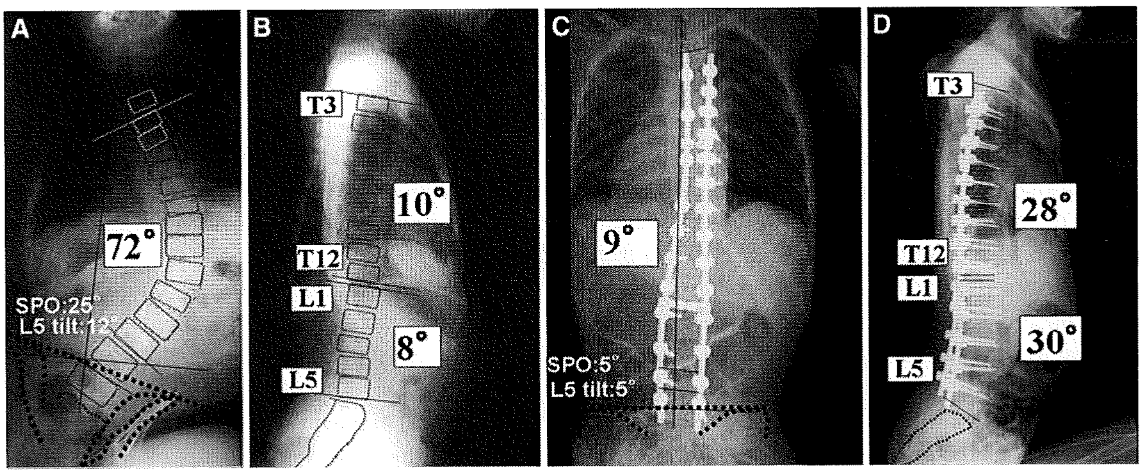

Fig. 3 A 13-year 2-month-old boy (patient 6). **a** and **b** Preoperative sitting anteroposterior radiograph shows significant thoracolumbar curve of 72° and SPO of 25°. L5 tilt is 12°. Sagittal thoracic kyphosis is 10° and lumbar lordosis is 8°. **c** and **d** Postoperative sitting views show significant correction of scoliosis of 9° and SPO of 5°. L5 tilt is 5°. Good reconstruction of thoracic kyphosis and lumbar lordosis is achieved postoperatively

(particularly, potential for neurologic complications), many studies have reported no increased neurologic risk with pedicle screw fixation and confirmed that pedicle screw placement in the thoracic and lumbar spine can be performed with both accuracy and safety [20, 36, 38].

Most studies on scoliosis surgery in patients with DMD have dealt with spinal instrumentation using hooks, wires or hybrid constructs with pedicle screws in the lumbar spine. Sussman [37] reported 25 patients with neuromuscular scoliosis who were treated with Luque rod instrumentation and fusion to L5, with a 48% postoperative correction and 35% correction at the last follow-up. The preoperative curve size in that study was 67°. Whitacker et al. [38] reported 23 patients with neuromuscular scoliosis, who were treated with hybrid constructs using pedicle screws ending in the distal lumbar spine. The preoperative curve size in that study was 71°, with a 64% initial correction and 55% correction at the last follow-up. McCall and Hayes [23] reported 75% initial correction of scoliotic curvature and 69% correction at the last follow-up using the U-rod and fusion to L5. The curve size was 65°. They reported 76% initial correction of pelvic obliquity and 68% final correction. They also reported 80%

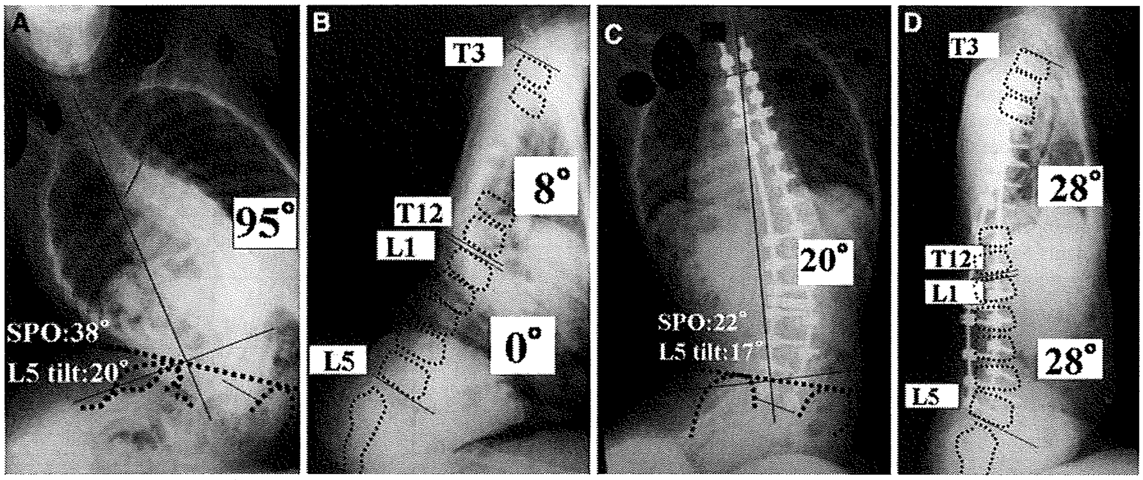

Fig. 4 A 13-year 4-month-old boy (patient 2). **a** and **b** Preoperative sitting anteroposterior radiograph shows significant thoracolumbar curve of 95° and SPO of 38°. L5 tilt is 20°. Sagittal thoracic kyphosis is 8° and lumbar lordosis is only 0°. **c** and **d** Postoperative sitting views show significant correction of scoliosis, yet show an appreciable residual SPO of 22°. L5 tilt is 17°. Sagittal plane alignment is well created

initial correction of scoliotic curvature and 70% correction at the last follow-up using the unit rod and fusion to the pelvis. The curve size was 70°. They reported 80% initial correction of pelvic obliquity and 75% correction at the last follow-up. Our results compared favorably with those published data. There are few studies on the use of pedicle-screws-alone fixation for scoliosis in DMD. Hahn et al. [16] reported excellent results for the coronal deformity with 77% correction, both initially and long-term, using pedicle-screw-alone fixation to the pelvis (not segmental pedicle screw instrumentation). The mean preoperative curve size in that study was 44°. They reported 78% initial correction of pelvic obliquity and 80% correction at the last follow-up. Modi et al. [26] found that in patients with neuromuscular scoliosis, acceptable amounts of curve correction (a mean of 61% initial correction and 58% correction at the last follow-up) can be achieved and maintained with posterior-only pedicle screw instrumentation to the pelvis without anterior release procedure. The mean preoperative curve size in that study was 79°. They reported 45% initial correction of pelvic obliquity and 47% correction at the last follow-up. We have challenged the long-term belief that fusion to the pelvis can be avoided even in non-ambulatory patients with DMD. In the current study, the scoliotic curvature improved 80% from a preoperative mean of 73° to an immediate postoperative mean of 14° and a mean of 17° at the last follow-up. The pelvic obliquity improved 65% from a preoperative mean of 17° to a postoperative mean of 6°, and 6° at the last follow-up. However, four patients had pelvic obliquity of more than 15°, postoperatively.

Pelvic obliquity associated with scoliosis in DMD continues to pose one of the most challenging instrumentation problems there is [3, 10, 27]. Instrumentation and fusion to the sacrum/pelvis has been a mainstay in the treatment of spinal deformity in DMD since the development of intra-iliac post and recommended to prevent and/or correct pelvic obliquity [1, 9]. However, controversy remains concerning the necessity of extending the implant constructs and fusion to the sacrum/pelvis [23, 30, 38]. In addition, pelvic fixation has certain disadvantages, including increased blood loss, longer operative time and technical difficulty [5, 10, 25, 36]. In addition, the sacrum and the pelvis in patients with DMD are often small and osteoporotic [4, 16].

With severe scoliotic curve and higher pelvic obliquity, suprapelvic fusion has shown higher loss of long-term correction, especially, of pelvic obliquity. However, there is growing evidence that adequate correction can be obtained and maintained by fusion to the distal lumbar spine or even short segment anterior instrumentation [12, 26, 38].

The L5/S1 articulation is normally a very stable joint secondary to traction exerted by the iliolumbar ligament and stability imparted by the disc complex (annulus fibrosis and anterior longitudinal ligament) [21, 34]. Therefore, with stable L5/S1 articulation, pedicle screw fixation into L5 should allow correction of pelvic obliquity [23]. Pelvic obliquity is due to the suprapelvic effects of scoliosis [38]. Therefore, addressing the spinal deformity must effectively correct pelvic obliquity. Frischut et al. [10] found pelvic obliquity was effectively corrected by correcting the spinal deformity. Wild et al. [39] noted a spontaneous correction of pelvic obliquity after the spinal deformity was adequately addressed. Thus, we have challenged the long-term belief that instrumentation and fusion to the sacrum/pelvis can be avoided even in non-ambulatory DMD patients. In all our patients, there was a significant improvement in the Cobb angle. Of the 28 patients, 23 had pelvic obliquity of less than 10° postoperatively and a good pelvic balance was achieved and maintained in these patients. However, five patients had pelvic obliquity of 15° postoperatively. These five patients had preoperative Cobb angles of 86°, 95°, 86°, 86° and 88°, respectively, and an SPO of 36°, 38°, 33°, 36° and 30°, respectively; in addition, they had a preoperative L5 tilt of 18°, 20°, 17°, 17° and 16°, respectively. They should have had fusion to the sacrum/pelvis.

The basis of successful treatment with instrumentation and fusion only to L5 appears to be a stable L5/S1 articulation. Some authors have noted that an L5 tilt of more than 15° represented an unstable L5/S1 joint complex caused by attenuation of the iliolumbar ligaments or the disc complex, or both, and consequently, would not support L5-based constructs [21, 34]. McCall and Hayes [23] documented that possible contraindications to instrumentation (using the U-rod) and fusion only to L5 included indication of L5/S1 instability as evidenced by an excessive L5 tilt of more than 15° and a lumbar curve with the apex at L3 or lower as a result of the inability to achieve an adequate number of distal fixation points. In the current study, all the patients had a curve with the apex at L2 or higher. Of the 28 patients, 23 with a preoperative L5 tilt of less than 15° had a postoperative pelvic obliquity of 10°, even if they had high pelvic obliquity preoperatively. However, five patients with pelvic obliquity of more than 15° postoperatively had an L5 tilt of 15° preoperatively.

Thus, with rigid segmental pedicle screw instrumentation and fusion only to L5, the pelvic obliquity, even if it was high, was effectively corrected and maintained within 10° by adequately addressing spinal deformity in patients with stable L5/S1 articulation as evidenced by a preoperative L5 tilt of less than 15°. Instrumentation and fusion to the sacrum/pelvis should be reserved for the patients with unstable L5/S1 articulation as evidenced by an excessive L5 tilt of more than 15° preoperatively.

The sagittal plane alignment was well recreated in our series. Correction of thoracic hypokyphosis was found, with a preoperative mean of 10° to a postoperative mean of 20°. With a postoperative mean of 35°, an excellent reconstruction of lumbar lordosis was achieved in all our patients. The change in the sagittal plane alignment reflected the intention to re-create a good sagittal profile. Adequate lumbar lordosis is important for good and balanced sitting in patients with DMD in whom flexion contractures of the hips and knees are often present.

There was no significant loss of correction or progression of scoliosis at the last follow-up. Some authors described a loss of correction with the Luque or hook instrumentation [8, 32], which was not the case when the U-rod, the unit rod or pedicle-screw-alone fixation systems were used according to other studies [3, 4, 23]. We suggest a diligent procedure during the exposure of the spinal posterior element to enhance an excellent arthrodesis. The lack of loss of correction in our series is felt to be due to the diligent procedure, which exposes a large surface area of contact within facets possibly permitting early facet arthrodesis. This is coupled with the use of anchors at every level providing true segmental fixation.

Instrumentation and fusion to L5 has been reported to decrease operating time and complexity, decrease blood and avoid injury to the sacrum/pelvis. In our series, this procedure could be accomplished with less exposure and less complexity, translating into decreased blood loss and decreased operating time. In our series of patients treated with fusion to L5, the L5/S1 articulation is preserved. The advantage of the mobile L5/S1 disc space includes absorption of much of the angular and rotational movement of the trunk during wheelchair activities. The presence of mobility at L5/S1 may assist in sitting and transfer activities. However, longer-term follow-up should be performed to determine whether segmental pedicle screw instrumentation and fusion only to L5 is adequate to maintain spinal alignment and pelvic balance and prevent junctional problems at the L5/S1 motion segment.

In our series of segmental pedicle screw instrumentation, every level was instrumented on at least one side and several vertebrae received two pedicle screws. Controversy may arise concerning the necessity of placing two pedicle screws at every level, especially for mild or moderate curves. Also, this method increases operating time and blood loss and represents a significant cost. It is important to provide strong and stable fixation in the lumbar spine. Strong anchors in the lumbar spine are needed. We believe pedicle screws should be placed bilaterally at every level in the lumbar spine to give a solid foundation, on which the rest of the spine may be held upright, well balanced, and without rotation, thus this helps to correct and maintain pelvic obliquity. However, we also think screw placement at every two or three levels in the thoracic spine may be sufficient in this patient population.

Conclusions

Segmental pedicle screw instrumentation and fusion to L5 was safe and effective in patients with DMD scoliosis with stable L5/S1 articulation as evidenced by a minimal L5 tilt of less than 15°, even though pelvic obliquity was relatively large. This method in appropriate patients can be a viable alternative to instrumentation and fusion to the sacrum/pelvis in the surgical treatment of DMD scoliosis.

Study limitations

Although the minimum 2-year results are encouraging, and there has been no significant deterioration in curve magnitude, pelvic obliquity or development of lumbosacral junctional instability and pain, it remains to be elucidated whether segmental pedicle screw instrumentation and fusion to L5 is adequate to maintain spinal and pelvic alignment and sitting balance and prevent junctional problems at the L5/S1 motion segment with a longer-term follow-up. Furthermore, the number of the patients, who had a preoperative L5 tilt of more than 15° and a postoperative pelvic obliquity of more than 10°, was too small. Therefore, it may be impossible to draw significant conclusions from the measurement of the L5 tilt in a small group of only five patients. However, this study shows the introduction of segmental pedicle screw instrumentation and fusion to increase the potential amount of curve correction and correction of pelvic obliquity in patients with DMD scoliosis.

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Conflict of interest statement None.

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ORIGINAL PAPER

Surgical management of severe scoliosis with high risk pulmonary dysfunction in Duchenne muscular dystrophy: patient function, quality of life and satisfaction

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Abstract In a previous study, the authors reported the clinical and radiological results of Duchenne muscular dystrophy (DMD) scoliosis surgery in 14 patients with a low FVC of <30%. The purpose of this study was to determine if surgery improved function and QOL in these patients. Furthermore, the authors assessed the patients' and parents' satisfaction. %FVC increased in all patients after preoperative inspiratory muscle training. Scoliosis surgery in this group of patients presented no increased risk of major complications. All-screw constructions and fusion offered the ability to correct spinal deformity in the coronal and pelvic obliquity initially, intermediate and long-term. All patients were encouraged to continue inspiratory muscle training after surgery. The mean rate of %FVC decline after surgery was 3.6% per year. Most patients and parents believed scoliosis surgery improved their function, sitting balance and quality of life even though patients were at high risk for major complications. Their satisfaction was also high.

Introduction

Posterior spinal fusion and instrumentation for scoliosis in Duchenne muscular dystrophy (DMD) has been aimed at maximising function and improving patients' quality of life [1–5, 7, 10, 13, 15–18, 20, 22]. This surgery has been widely accepted as an optimal procedure to prevent

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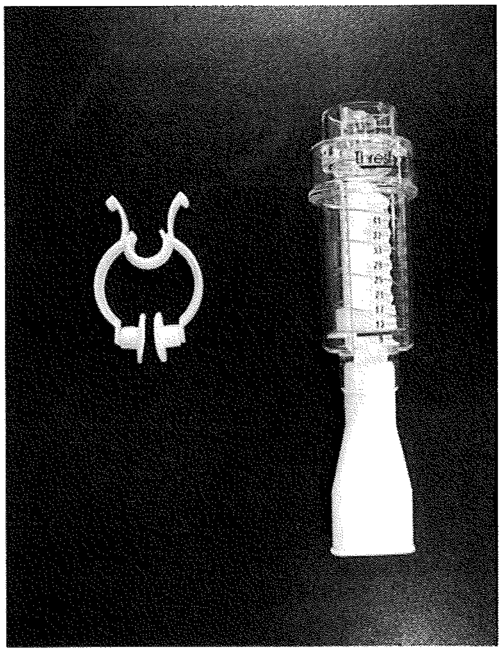

Fig. 1 Pulmonary trainer (Threshold IMT, Philips Respironics, Inc., Murrysville, PA, USA) provides consistent and specific pressure for inspiratory muscle strength and endurance training, regardless of how quickly or slowly patients breathe. This device incorporates a flow-independent one-way valve to ensure consistent resistance and features an adjustable specific pressure setting (in cm H_20) to be set by a healthcare professional. When patients inhale through Threshold IMT, a spring-loaded valve provides a resistance that exercises respiratory muscles through conditioning

progression of scoliosis and maintain upright and comfortable sitting balance [1–5, 7, 10, 13, 15–18, 20, 22]. Although many studies have reported the results of scoliosis surgery in patients with DMD, most studies have lacked a postoperative evaluation of the quality of life (QOL) of the patients and the overall effects on the parents. The majority of studies have reported only data on changes in scoliotic curvature and complications. Irreversible changes in cardiac and pulmonary function pose serious risks to anaesthesia and have been relative contraindications to surgery [7, 17, 18, 20, 22, 23].

In a previous study, the authors reported the clinical and radiological results of DMD scoliosis surgery in patients with a low forced vital capacity (FVC) of <30%, conventionally considered too low to permit reasonable surgical risk [23]. Scoliosis surgery for such patients must provide meaningful benefit in their function, QOL and satisfaction in proportion to risks involved for patients and parents.

The purpose of this study was to determine if scoliosis surgery improved function and QOL in patients with severe DMD scoliosis and high risk pulmonary dysfunction. Furthermore, the authors assessed the patient's and parent's satisfaction after scoliosis surgery.

Materials and methods

This study was approved by the Institutional Review Board of Kitasato University and informed consent was obtained from all patients prior to participation in this study.

Between 2005 and 2007, a total of 14 patients with scoliosis secondary to DMD and a FVC of <30% of predicted value at the time of admission underwent scoliosis surgery. Before surgery, all patients were non-ambulatory. All patients were able to ventilate autonomously and were not respirator dependent. They were trained with preoperative inspiratory muscle training by a pulmonary trainer (Threshold IMT) (Figs.1 and 2) for six weeks prior to surgery [23]. All patients recovered well following the surgery with no major complications [23]. They were encouraged to continue inspiratory muscle training after surgery and even at home after discharge.

Pulmonary function tests [23]

Pulmonary function tests were performed with a computerised pulmonary function system (FUDAC-50, Fukuda Denshi, Tokyo). Pulmonary function values reported in this study are forced vital capacity (FVC) expressed as percent of predicted value (%FVC) on the published normative data. Pulmonary function tests were evaluated at six weeks and three weeks before surgery, the day before surgery, and then at six weeks, one year and two years after surgery as well as at the last follow-up. %FVC was measured three times and evaluated by one independent observer. The best trial for the test was used for further analysis.

Fig. 2 An 11-year-old boy being trained with inspiratory muscle training by a pulmonary trainer (Threshold IMT)

Inspiratory muscle training [9, 11, 23]

After surgery and even in the home after discharge, patients had to perform both resistive breathing manoeuvres and maximal static inspiratory efforts. They were trained with inspiratory muscle training by a pulmonary trainer.

The pulmonary trainer resistive exercise intervention was based on previously published protocols. Inspiratory muscle training was performed daily. Three sets of 15 repetitions were performed.

Radiographic assessments

Radiographic assessments were performed on anteroposterior spinal radiographs covering the whole spinal column with the patient in sitting position. These assessments were made the day before surgery, in the postoperative week, at one and two years after surgery and at the last follow-up. The Cobb angles of the curves and the pelvic obliquity (spinal pelvic obliquity [SPO]) on the coronal plane were measured.

Surgical procedure [23]

The primary aim of this surgery was to obtain a solid fusion, level pelvis and a balanced spine. All patients had posterior spine fusion and all-screw construction from the upper thoracic spine (T3 or T4) to the lower lumber spine (L5) for scoliosis, performed by the same surgeon (M.T.) as described in the previous study (Fig. 3). No anterior surgery was performed in any of these consecutive series. All operations were performed under general anaesthesia.

Patient/parent evaluation of surgery

At the last follow-up, patients and parents were interviewed and asked to complete an outcome questionnaire by one independent observer. The authors modified the previous questionnaire described by Bridwell et al. [5] and made our own simple outcome questionnaire. Six of the questions addressed function (questions 1–6), two addressed quality of life (questions 7 and 8) and three targeted satisfaction (questions 9–11).

Results

A total of 14 consecutive patients were enrolled in this study. No patient was lost to follow-up. The mean follow-up period was four years one month (range, 2 years 6 months to 5 years 11 months). Details of radiographic parameters in the study group are shown in Table 1. Changes in %FVC are shown in Fig. 4. The mean %FVC at the time of admission was 21.6% (range, 16–27%). There were no ventilator dependent patients preoperatively. %FVC increased to 26.2% (range, 22–31%) the day before surgery after six-week inspiratory muscle training. %FVC remained stable or increased slightly in all patients at six weeks after surgery. %FVC continued to decrease to 23.2% (range, 19%–27%) at one year after surgery, 20.1% (range, 16%–25%) at two years after surgery and 12.0% (range, 9%–22%) at the last follow-up (four years one month after surgery). The mean rate of %FVC decline after surgery was 3.6% per year in our patients.

The mean preoperative scoliotic curvature was 98° (range, 81°–130°).The mean immediate postoperative scoliotic curvature was 34° (range, 20°–40°). The mean scoliotic curvature was 35° (range, 23°–42°) at the last follow-up. The pelvic obliquity improved from a preoperative mean of 22° (range, 15°–34°) to an immediate postoperative mean of 8° (range, 4°–17°) and a mean of 8° (range, 5°–17°) at the last follow-up. No significant loss of correction of scoliotic curvature and pelvic obliquity was found at the last follow-up.

Balanced sitting posture was achieved and maintained in all but three patients who had postoperative pelvic obliquity of >15°. These three patients had preoperative Cobb angles of 101°, 128° and 130° and pelvic obliquities of 29°, 30° and 34°, respectively.

Table 2 shows the questionnaire with answers and the tally for each of the questions broken down into the total answers. All patients reported difficulty in sitting. After surgery, all patients reported major or moderate improvement (question 1). All patients reported difficulty using their arms and hands before surgery due to poor sitting balance. After surgery, 14 patients reported improvement and only one reported no change (question 2).

Before surgery, all patients were able to feed themselves. At the last follow-up, ten patients were able to feed themselves but four patients were unable to feed themselves because their arms were getting weaker (question 3). Two of the 14 patients/parents reported problems with digestive systems before surgery. Both of them reported major or moderate improvement (question 4). Nine patients reported respiratory problems before surgery. At the last follow-up, six reported moderate to major improvement and three reported minimal improvement. In the assessment of the child's overall activity level, 14 patients reported improvement (question 6). Only two patients had pressure sores before surgery. Both of them reported a major improvement after surgery (question 7). Regarding the overall quality of life, all 14 children reported improvement (question 8). All patients considered the surgery successful and satisfactory (question 9). Twelve of the 14 patients definitely would have the surgery again, and only two probably would have (question 10). Twelve of the 14 patients definitely would

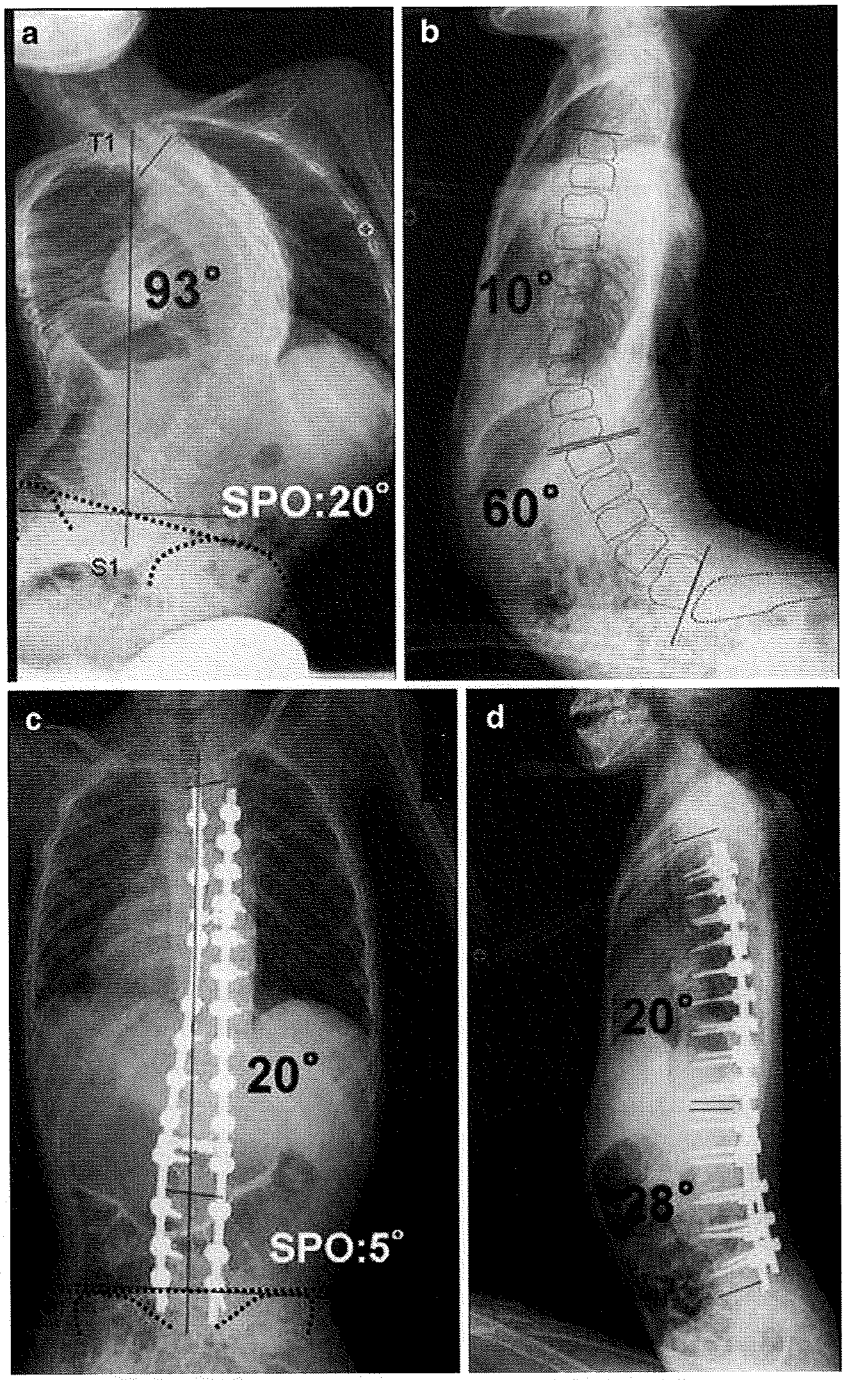

Fig. 3 **a**, **b** A 13-year-old boy with DMD. Anteroposterior radiograph shows severe scoliosis of 93° with significant pelvic obliquity of 20°. Thoracic hypokyphosis and lumbar hyperlordosis were present. **c**, **d** All-screw constructs and fusion to L5 were performed. Postoperative sitting views show significant coronal curve correction of 20° with normalization of sagittal plane. Pelvic obliquity improved to 5°

recommend the surgery and two probably would recommend it (question 11).

Discussion

Motor strength and pulmonary function are the key issues of function and QOL in patients with DMD, while scoliosis surgery has been highly effective in stabilising the spine and maintaining comfort and sitting balance [1–5, 7, 10, 13, 15–18, 20, 22]. Scoliosis surgery should be early enough in the course of the underlying disease, when cardiac and pulmonary function is sufficient, so that patients can be anaesthetised and operated relatively safely in order to reduce the likelihood of major complications [1, 3, 22].

Due to the profound cardiopulmonary compromise of the patients, they are more susceptible to postoperative pneumonia, prolonged ventilator dependence, cardiac failure and even death [1, 5, 16, 20]. We routinely provide the preoperative information and guidance (informed consent)

Table 1 Details of radiographic parameters in the study group patients

Patient number	Age (years)	Scoliotic curvature (°)			Pelvic obliquity (°)		
		The day before operation	Immediately after operation	Last follow-up	The day before operation	Immediately after operation	Last follow-up
1	11	100	38	40	20	8	9
2	12	93	35	35	20	4	5
3	12	91	32	33	19	5	7
4	14	81	27	29	15	8	8
5	13	93	20	23	20	5	5
6	14	130	38	40	34	17	17
7	11	101	35	35	20	7	7
8	17	101	40	42	29	16	16
9	11	81	32	33	18	4	4
10	13	98	35	35	21	5	5
11	13	128	28	29	30	15	15
12	13	100	35	36	25	8	8
13	12	85	30	32	18	6	6
14	11	95	35	37	23	7	7
Mean	13	98	34 (65%)	35 (64%)	22	8 (64%)	8 (64%)

concerning this high pulmonary risk scoliosis surgery for the patients/parents at the time of admission.

Although potential risks and peri/postoperative complications should be considered, there has been also overwhelming support for the beneficial effect of scoliosis surgery on general well-being of the patient. Despite the magnitude of this surgery, the successful outcome of an operation for DMD scoliosis has been considered beneficial by most patients and their principal care providers [1, 5, 7]. The patient's function will be worse because of the progressively deteriorating nature of the disease as well as further progressive scoliosis if surgery is not performed. Therefore, we have recommended this surgery even if the patient presents late and is at high risk. However, surgery should be discussed thoroughly and we are of the opinion that the decision whether the patient will undergo surgery

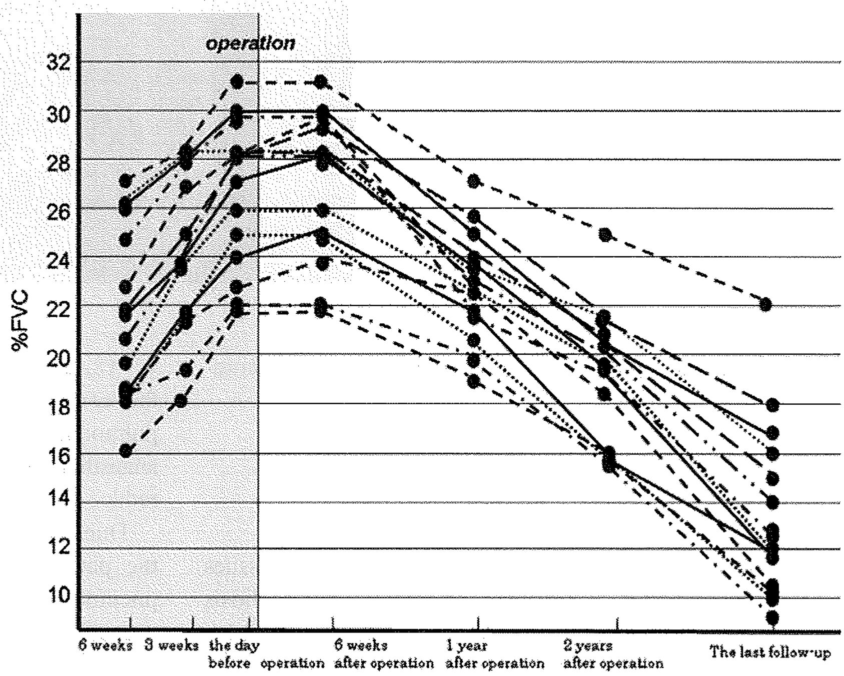

Fig. 4 Changes in %FVC are demonstrated. After preoperative respiratory muscle training for six weeks, %FVC increased in all patients. All 14 patients underwent surgical correction of scoliosis successfully without any complications. At six weeks after operation, %FVC remained stable or increased slightly. %FVC decreased in all patients at one year, two years after operation and at the last follow-up. The average rate of decline of %FVC was 3.6% per year at the last follow-up

Table 2 Questionnaire

Question	Patient/parent answers
1. How was your child's posture/sitting prior to surgery?	
Very crooked; pelvis very tilted; sat with rib cage on hip	4
Very crooked with head and shoulders very forward	6
Moderately crooked	4
Minimally crooked; required help to sit up	0
No problem with sitting balance	0
1a. How is your child's posture/sitting balance at the follow-up compared to before surgery?	
Major improvement	2
Moderate improvement	8
Minimal improvement	0
Unchanged	0
2. Did your child have difficulty using his arms and hands before surgery due to poor sitting balance?	
Yes	14
No	0
2a. If yes, has this been changed by the surgery?	
Major improvement	2
Moderate improvement	6
Minimal improvement	6
Unchanged	0
3. Was your child able to feed himself before surgery?	
Independent in feeding	5
Able to feed himself only with assistive devices	9
Only able to feed himself finger foods	0
Unable to feed himself	0
3a. Compared to before surgery, is your child:	
Independent in feeding	5
Able to feed himself only with assistive devices	5
Only able to feed himself finger foods	0
Unable to feed himself	4
4. Did your child have a problem with eating/digestion before surgery?	
Yes	2
No	12
4a. If yes, has this changed by the surgery?	
Major improvement	1
Moderate improvement	1
Minimal improvement	0
No change	0
Worse	0
5. Did your child have respiratory problems before surgery (i.e., frequent colds, pneumonia)?	
Yes	9
No	5
5a. If yes, has this been changed by the surgery?	
Major improvement	3
Moderate improvement	3
Minimal improvement	3
Unchanged	0
Worse	0
6. Compared to before surgery, is there any change in your child's overall activity level during the day (e.g., going to school, reading, playing, etc.)	
Major improvement	8
Moderate improvement	3
Minimal improvement	0
Unchanged	0
Worse	0
7. Did your child have a problem with pressure sores before surgery?	
Yes	2
No	12
7a. If yes, has this been changed by the surgery?	
Major improvement	2
Moderate improvement	0
Minimal improvement	0
Unchanged	0
Worse	0
8. Please rate the change in your child's and your own quality of life since the surgery:	
For child	
Major improvement	6
Moderate improvement	8
Minimal improvement	0
Unchanged	0
Worse	0
For Parent	
Major improvement	8
Moderate improvement	6
Minimal improvement	0
Unchanged	0
Worse	0
9. Overall, how would you rate the outcome of surgery?	
Very satisfactory	11
Satisfactory	3
Neither satisfactory nor unsatisfactory	0
Unsatisfactory	0
Very unsatisfactory	0
10. If given the opportunity, would you want your child to have his scoliosis surgery over again?	
Yes, definitely	12
Probably	2
Unsure	0
No	0
11. Would you recommend the surgery for others with DMD scoliosis?	
Yes, definitely	12
Probably	2
Unsure	0
No	0

or not must be made by the patients/parents after sufficient discussion.

Pulmonary complications in the peri/postoperative period in DMD patients have been reported to be frequent [1, 16, 18, 21]. Higher pulmonary complications have been reported when the FVC is <30% [7, 8, 10, 19].

Pneumonia, respiratory arrest, pneumothorax as well as difficulties in weaning from the respirator have been reported [1, 8, 16, 19, 21]. Thus, many surgeons have recommended scoliosis surgery only for patients who have a FVC of at least 35–40% [7, 18, 21]. To prevent pulmonary complications and to increase pulmonary function, we recommend preoperative respiratory muscle training using threshold IMT when the patient decides to undergo surgery.

Thus, scoliosis surgery in patients with DMD is challenging, and yet the authors' contention has been that the rate of complications with scoliosis surgery in patients with DMD and poor pulmonary function is low and that the surgery is substantial. However, there has been concern that patients/parents may not believe the scoliosis surgery to be of significant benefit, considering the magnitude of the surgery, high rates of complication and mortality and the generally downhill course of the underlying neuromuscular disorders. This study shows that our patients have been managed successfully with no major complication. Although many studies have reported that the role of scoliosis surgery in patients with DMD is to prevent or retard deterioration of function and sitting balance rather than to improve it because DMD is a progressively deteriorating myopathic disorder [1, 15–22], this study also shows most parents/patients believed that high risk scoliosis surgery improved their function, sitting balance and QOL. The authors were surprised with the strongly positive perceptions of most of the patients/parents with DMD scoliosis and high risk pulmonary dysfunction.

There have been few QOL measures appropriate for DMD scoliosis. In 1999, Bridwell et al. [5] devised a questionnaire that assessed patient function, pain, cosmesis, self-image, and the QOL for flaccid neuromuscular (Duchenne muscular dystrophy and spinal muscular atrophy) scoliosis patients. In their work, they reported that most patients/parents believed scoliosis surgery improved patient function, cosmesis, sitting balance and QOL. The authors modified their questionnaire and devised our simple questionnaire that assessed patient function, the QOL and satisfaction.

With the results of this study, most patients/parents believed scoliosis surgery improved function, sitting balance and QOL with a long-term follow-up. The favourable results related to QOL and overall satisfaction. However, DMD is a flaccid neuromuscular disorder in which gradual deterioration is the hallmark, and thus the less favourable evaluations appeared to concern function.

It may be that it reflects the progressively deteriorating nature of the disorder. Balanced sitting posture was achieved in all patients except for three who had postoperative pelvic obliquity of >15°. These three patients probably should have had pelvic fixation. However, they reported improvement in sitting balance and did not complain of difficulties in sitting. The coronal balance has been reported to be the most important parameter affecting sitting [4]. With a mean of 65% correction, our study shows good results for coronal deformity despite preoperative severe scoliotic curvature. Thus, the sitting balance appeared to be improved and maintained in our patients.

It is interesting to note that all patients reported improvement of pulmonary function, although there was a significant decrease in %FVC in all patients at the last follow-up. While pulmonary function deterioration is primarily due to respiratory muscle weakness, the negative mechanical effects of the deformed thorax on the underlying lung has been suggested. The authors believe that chest wall deformity and respiratory tract deformity caused by spinal deformity are effectively treated by correcting spinal deformity and thereby patients felt improvement of pulmonary function.

Because of the absence of control groups, the authors can not contribute to the controversy whether scoliosis surgery affects the rate of decline of pulmonary function in DMD patients. Most studies could not document positive effects of scoliosis surgery on pulmonary function [12, 14, 19]. Beneficial effects of scoliosis surgery on pulmonary function have been reported in only a few studies [6, 24]. Recently, Velasco et al. [24] showed positive effects of posterior spinal stabilisation on pulmonary function by showing slowed rate of %FVC decline from 8.0% per year before surgery to 3.9% per year after surgery with a long-term follow-up. Koessler et al. [9] have shown that inspiratory muscle training in patients with DMD improves inspiratory muscle function and that the training leads to a stabilisation of %FVC even in the long-term of two years. Although our patients were encouraged to continue inspiratory muscle training to stabilise or retard %FVC decline even after surgery, there was a continuous %FVC decline in all patients and the rate of %FVC decline was 3.6% per year with a mean follow-up of three years five months. This value compares favourably with Velasco's result even though the number of our cases is too small.

In conclusion, %FVC in patients with severe scoliosis secondary to Duchenne muscular dystrophy increased after preoperative inspiratory muscle training prior to scoliosis surgery. Scoliosis surgery in this group of patients presented no increased risk of major complications. There was no mortality. In all cases, patients were extubated immediately after surgery in the operating room and no respiratory support was required in the peri/postoperative period. All-screw constructions and fusion to L5 in DMD patients with severe spinal deformity and high pelvic obliquity offered the ability to correct scoliotic curvature and pelvic obliquity initially, intermediate and long-term. All patients were encouraged to continue inspiratory muscle training after surgery. The mean rate of %FVC decline after surgery was 3.6% per year. Most patients/parents believed scoliosis surgery improved their function, sitting balance and quality of life even though patients were at high risk for major complications. Patient satisfaction was also high. They benefitted from scoliosis surgery.

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ORIGINAL PAPER

Surgical management of severe scoliosis with high-risk pulmonary dysfunction in Duchenne muscular dystrophy

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Abstract Between 2005 and 2007, 14 patients who had severe scoliosis in Duchenne muscular dystrophy (DMD) and a poor forced vital capacity (FVC) of <30% at admission underwent scoliosis surgery. FVC on admission was 21.6% (range, 16–27%). The patients were given respiratory muscle training using a pulmonary trainer (Threshold IMT, Philips Respironics, Inc.) for six weeks before operation. FVC increased to 26.2% (range, 22–31%) the day before operation. The mean preoperative scoliosis was 98° (range, 81°–130°). All patients underwent posterior fusion and all-screw construction and were extubated on the operative day. No patients developed any respiratory complications. The postoperative scoliosis was 34° (range, 20°–40°) (65%). FVC remained stable at six weeks after operation. FVC decreased to 19.8% (range, 16–25%) and the mean scoliosis was 35° (range, 23°–40°)(64%) at two years after operation. DMD patients with severe scoliosis and FVC considered too low to permit reasonable surgical risk could undergo surgery and could benefit from surgery.

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Introduction

Duchenne muscular dystrophy (DMD) is the most common form of muscular dystrophy and it is often accompanied by deterioration of cardiac and pulmonary function and spinal deformity. The natural history of scoliosis as well as the rapid deterioration of pulmonary function in DMD patients has been well established [11, 19, 25]. Usually, progressive curves require surgical correction and stabilisation. In DMD scoliosis, progression occurs much more frequently and more quickly than in idiopathic scoliosis; therefore, early surgical correction of scoliosis is recommended [1, 5, 7, 8]. Despite the magnitude of this surgery, the successful outcome of an operation for scoliosis in DMD is considered beneficial by most patients and their principal care providers [1–3, 5, 7–9].

However, surgeons are often reluctant to perform elective surgery on such patients who have a history of respiratory failure, are respirator dependent, or whose forced vital capacities (FVC) are very poor.

Generally, the limit for the operation is minimum FVC of about 30% of predicted normal [9, 14, 18, 19]. Most surgeons are inclined to avoid surgery for those patients because of the fear of respiratory complications, and therefore many such patients with severe respiratory muscle weakness associated with DMD who could benefit from spinal fusion with instrumentation for scoliosis and pelvic obliquity are not being offered surgical correction.

As our high pulmonary risk patients (FVC <30%) with scoliosis in DMD have been successfully managed without any pulmonary complications, the authors are of the opinion that patients with FVC conventionally considered too low to permit reasonable surgical risk can undergo scoliosis surgery with general anaesthesia.

The purpose of this study was to report on a prospectively collected series of patients with DMD and very high-risk pulmonary dysfunction (FVC <30%) who underwent surgical correction of scoliosis after preoperative respiratory muscle training.

Materials and methods

A total of 14 consecutive patients with DMD (all non-ambulatory boys) undergoing scoliosis surgery between 2005 and 2007 were prospectively studied. Inclusion criteria required that each patient had difficulties in seating balance due to scoliosis and pelvic obliquity, FVC <30%, and no history of continuous dependence on respirator.

Pulmonary function tests

Pulmonary function tests were performed with a computerised pulmonary function system (FUDAC-50, Fukuda Denshi, Tokyo). The various measurements of pulmonary function were assessed. The tests were performed on each occasion with the patients seated in a wheel chair. Arm-span measurements were selected for calculations for the predicted heights. Pulmonary function value reported in this study is forced vital capacity (FVC) expressed as percent of predicted value (%FVC) on the published normative data. Pulmonary function tests were evaluated in all patients included in this study at six weeks and three weeks before operation, the day before operation, as well as at six weeks, one year and two years after operation.

Respiratory muscle training

All 14 patients were admitted at our hospital six weeks before operation. After admission, they were given preoperative respiratory muscle training using a pulmonary trainer (Threshold IMT, Philips Respironics, Inc., Murrysville, PA, USA) (Figs. 1 and 2) for six weeks prior to operation. The pulmonary trainer resistive exercise intervention, presented in Table 1, was based on previously published protocols [13]. Respiratory muscle training was performed daily for six weeks. Three sets of 15 repetitions were performed. Initial resistance (H_2O cm) was set at 30% of the patients preset maximal inspiratory pressure (MIP). Threshold IMT pressure resistance was increased weekly according to the patient's baseline MIP and Borg Rating of Perceived Exertion (RPE) [12] as well as the patient's symptoms. Patients ranked perception of fatigue during training using a Borg RPE Scale. Patients completed a log documentation for exercise adherence, Borg RPE, and symptoms following the daily exercise sessions. All the patients tolerated the exercise training program except for one subject who complained of lightheadedness during the initial training session. When the light-headedness occurred, pressure resistance was reduced by 2 cm H_2O.

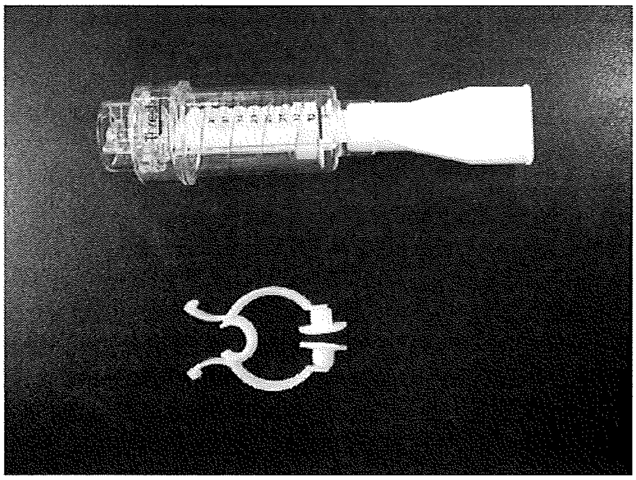

Fig. 1 The pulmonary trainer (Threshold IMT, Philips Respironics, Inc., Murrysville, PA, USA) provides consistent and specific pressure for inspiratory muscle strength and endurance training, regardless of how quickly or slowly patients breathe. This device incorporates a flow-independent one-way valve to ensure consistent resistance and features an adjustable specific pressure setting (in cm H_20) to be set by a healthcare professional. When patients inhale through Threshold IMT, a spring-loaded valve provides a resistance that exercises respiratory muscles

Radiographic assessments

Sitting anteroposterior and lateral spinal radiographs covering the whole spinal column were available for all patients the day before operation, at six weeks after operation, at two years after operation and the last follow-up. The Cobb angles of the curves on the coronal plane were measured.

All operations were performed by the same surgeon (T. M.) using all-screw construction. Autotransfusion via both preoperative storage and intraoperative collection was used. During operation, monitoring of spinal cord function was conducted by somato-sensory/motor-evoked potentials. Extubation was undertaken when the patients could ventilate their lungs spontaneously immediately after operation. Overnight pulse oximetry monitoring was used to evaluate pulmonary function for all patients in the immediate postoperative period. The patients were encouraged to sit on the wheelchair on the first postoperative day and were discharged at two weeks. The patients were encouraged to continue respiratory muscle training using pulmonary trainers even after operation and discharge.