A Review Paper

The Operative Treatment of Scoliosis in Duchenne Muscular Dystrophy

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ABSTRACT

The results of segmental spinal stabilization and fusion in nine patients with Duchenne muscular dystrophy are reviewed. The average follow-up time was 3.2 years. Vital capacities averaged 46% (range, 20% to 70%), and there were minimal pulmonary complications. Operative time and blood loss decreased when the spinal fixation method was changed from sublaminar to intraspinous wiring. Segmental wiring anchored through the spinous processes also maintained reduction and distraction until fusion occurred; we recommend this technique. The use of allogenic bone grafts to supplement the autogenous bone graft allowed for extensive fusion; we recommend this technique as well. Furthermore, fusion to the sacrum to prevent further pelvic obliquity should be indicated in all patients who develop scoliosis.

Duchenne muscular dystrophy (DMD), the most common and severe form of muscular dystrophy, is a progressive, degenerative childhood disorder. In the past 20 years, children with DMD have experienced an improved quality of life largely due to orthopaedic procedures that prolong their ambulation by 3 to 5 years and effectively treat their scoliosis.[1-3]

Not long ago, DMD children with scoliosis were not treated surgically because of poor results and a frequent postsurgical demise due to their weakened condition.[4,5] However, recent studies have demonstrated that mechanical ventilation can markedly extend survival and productivity in patients with late-stage DMD.[6-7] In addition, spinal stabilization and fusion procedures have given these patients the stability needed to remain comfortable when seated.[1,3,8]

The incidence of scoliosis, a major problem in DMD management, has reportedly ranged from 60% to 95%.[1-3,9-11] Scoliosis develops in DMD patients in the later stages of ambulation[12,13] or when patients become wheelchair-bound.[14-16] Unless the spinal curvature is treated, it invariably progresses;[11,17] this process may lead to incapacitating deformities (Figure 1).

This study provides a retrospective review of the results of segmental spinal stabilization and fusion in patients with DMD. Our results and modifications are compared with previously published reports to identify factors influencing the probability of improving mobilization in these patients while decreasing morbidity.

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Accepted for publication June 21, 1991.

This paper will be judged for the Resident Writers' Award.

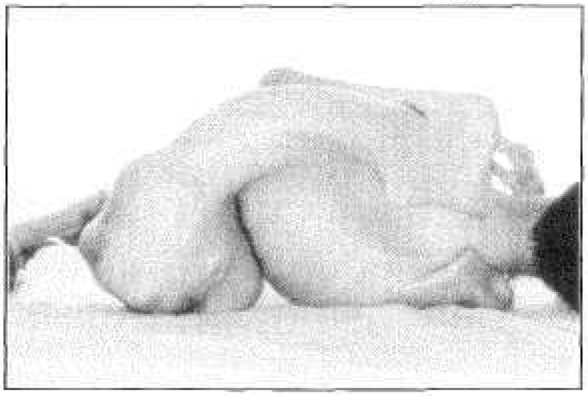

Figure 1. *A 17-year-old male with severe deformities secondary to scoliosis. Reprinted from reference 22, p 29. (Permission obtained).*

MATERIALS AND METHODS

The study group included nine DMD patients who received segmental spinal stabilization and fusion for progressive scoliosis from 1981 to 1987 at the Borgess Medical Center Muscular Dystrophy Clinic. Two other DMD patients were also offered this surgical option. Although both patients initially refused surgery, they later requested the spinal stabilization procedures; however, surgery was withheld because of their poor pulmonary reserve.

Curve progression (all patients) and seating difficulties (three patients) were the most common indications for surgery. Although occasional attempts were made to schedule spinal surgery once the curve approached 30°,[3,18] missed clinic visits or delays in surgery contributed to patients having much larger curves than 30° by the time surgery was performed.

OPERATIVE TECHNIQUE

The patients underwent surgery using segmental spinal stabilization and fusion comprising Harrington rods with segmental wiring to hold the spine after distraction. Fixation was performed from T-3 or T-4 to the sacrum in seven patients and to L-5 in two patients. Fusion was obtained by using autogenous iliac crest bone grafts supplemented with allogenic femoral heads from the hospital bone bank.

Segmental wires were placed sublaminarly in four patients. The process was monitored using intraoperative, cortical-evoked, spinal cord monitoring. In the remaining five patients, wires were placed through the spinous processes with the aid of an awl.

RESULTS

Thoracolumbosacral orthoses (TLSO) were used in four patients preoperatively to attempt to either halt or retard advancement of a lateral scoliotic curve. All cases were considered failures (Table I).

Preoperative vital capacity averaged 46% of normal (Table II). One patient (Case 8) was noted to have a vital capacity of 32% on surgical evaluation; however, his surgery was delayed for several months, and a repeated pulmonary function test revealed a vital capacity of 20%.

Table I. Preoperative Curve Progression in DMD Patients Using TLSO Orthoses to Control Scoliosis

Case	Total Time Used (Years)	Initial Curve* (Degrees)	Final Curve (Degrees)	Progression of Curve (Degrees)
		15° T	23° T	7° T
1	1.5	18° L	30° L	12° L
		18° T	26° T	8° T
2	0.9	19° L	28° L	9° L
6	2	15°	64°	49°
8	3.2	10°	100°	90°

*For double major curves: L = lumbar, T = thoracic.

Table II. Perioperative Results for DMD Patients Having Segmental Spinal Stabilization and Fusion for Scoliosis

Case	Age at Surgery (Years)	Vital Capacity (%)	Total Anesthesia Time (Hours)	Fusion Levels	No. of Segmental Wires	Estimated Blood Loss (mL)	Length of Hospital Stay (Days)
1	12.9	38	7.25	T3–S1	14	6,300	11
2	16.1	70	6.16	T3-S1	13	2,000	9
3	14.3	51	5.67	T3–S1	11	3,500	9
4	11.8	70	6.25	T4-L5	13	2,400	9
5	16.7	35	5.50	T4-S1	11	3,000	8
6	14.3	31	5.53	T4-S1	12	1,500	7
7	13.9	51	4	T4-L5	9	2,400	8
8	13.7	20	5.45	T4-S1	7	2,600	10
9	11.3	45	4.30	T4–S1	6	2,000	10
Average	13.9	46	5.6	NA	10.7	2,856	9

Surgery was performed in this patient because of the severe curve and seating difficulties. He did well postoperatively, and was extubated at 26 hours and discharged on day 10.

Operative results for this series are summarized in Table II. Overall anesthesia time averaged 5.6 hours. Anesthesia time required for sublaminar wiring was 6.3 hours; 4.9 hours was the average time for the intraspinous wiring techniques. The average number of segmental wires used for all patients was 10.7. Sublaminar wiring held an average of 12.8 wires. The total number of intraspinous wires was less (average, 9.1), because some spinous processes were unable to support tightening of the wires. No breakage or pullout of wires was noted on roentgenograms on completion of this review. Estimated blood loss was 2,856 mL overall, with an average of 3,550 mL for patients with sublaminar wiring and 2,300 mL for patients with intraspinous wiring. The duration of hospitalization averaged 9 days.

The average preoperative curve was 58° in six patients with one major curve. Three patients had double major curves, with an average thoracic scoliosis of 31° and lumbar scoliosis of 26°. The average postoperative curve in patients with a single major curve was 18°, with a final follow-up curve of 25°. Two patients with a double major curve had correction to a single major curve, averaging 16° postoperatively and 25° at final follow-up. The one patient with a remaining double major curve had correction to 8° and 20°, respectively, on his last clinic visit. The average follow-up for all patients was 3.2 years (range, 1.6 to 6.1) (Table III).

All patients had solid fusion as indicated by trabeculated bone, fused facets, and lack of roentgenographic pseudarthrotic signs. When solid fusion was obtained, no curve progression was noted in the fused regions. Two patients were fused to L-5 rather than the sacrum to gain additional mobility.[3] One patient, who died 20 months postoperatively of pneumonia, had no further pelvic obliquity. By 8-month follow-up, the other patient who was fused to L-5 developed a 30° pelvic obliquity inferior to the fusion level (Figure 2); however, despite this pelvic obliquity and its attendant discomfort, he was more comfortable in his wheelchair after spinal stabilization.

All patients had improved or maintained seating balance postoperatively. Surgery resulted in minimal interruption of the patients' lifestyles.

Table III. Postoperative Results for DMD Patients After Segmental Spinal Stabilization and Fusion for Scoliosis

Case	Length of Follow-up (Years)	Preoperative Curve* (Degrees)	Postoperative Curve (Degrees)	Final Curve (Degrees)	Complications
1	6.1	23° T 30° L	15°	18°	Dural sac leak
2	5.5	26° T 28° L	16°	32°	Sacral alar hook dislodged
3	5.4	41°	8°	8°	Sacral alar hook dislodged, transient left foot numbness
4	1.6	43° T 20° L	12° T 24° L	8° T 20° L	Died 20 months postoperatively
5	2.4	39°	11°	11°	
6	2	75°	20°	35°	
7	2.4	25°	14°	30°	Pelvic obliquity
8	2	100°	32°	40°	
9	2	70°	26°	27°	

*For double-major curves: L == lumbar, T = thoracic.

One patient who was virtually bedridden was able to ride in a wheelchair again. Another patient was attending college 5 years postsurgery.

COMPLICATIONS

One patient had a dural leak intraoperatively during the exposure for sublaminar wiring. The leak was repaired, and no further leaks or complications were noted. Another patient who had undergone sublaminar wiring complained of postoperative numbness of his medial left foot, probably due to irritation from the sublaminar wiring; the discomfort resolved by the second postoperative month.

Two patients had dislodgment of sacral alar hooks. One patient continued to have excellent alignment and spinal fusion after dislodgment. The other patient had a 14° progression of his scoliosis at 1-month surgical follow-up; over 5 years, he developed an excellent fusion mass with no further curve progression.

DISCUSSION

Over the past 20 years, many advances have been made in recognizing and treating scoliosis in DMD patients. Benign neglect and use of spinal orthoses to control scoliosis[5,19,22] have been replaced with a more aggressive surgical regimen[1,3,8] that, when combined with newer respiratory care technology,[6,7] has extended the useful life spans of these patients into a third decade.

The two patients who initially declined spinal stabilization procedures demonstrated the potential clinical course of untreated disease. The first patient, 13 years' of age and of average intelligence, refused surgery when his thoracolumbar curve had progressed to 70°. Two

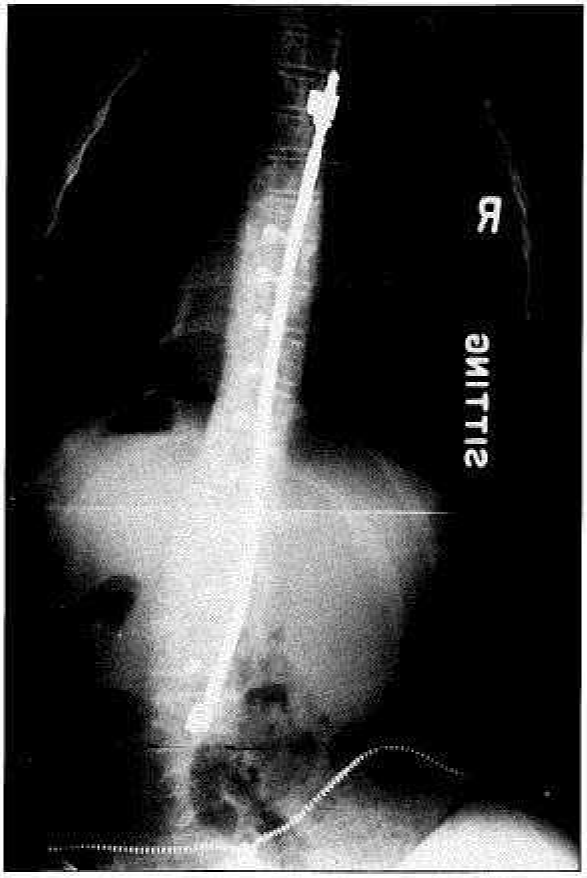

Figure 2. *A 16-year-old male who underwent spinal fusion from T-4 to L-5. Although the patient was asymptomatic, a 30° pelvic obliquity developed postoperatively.*

years later, he wanted to proceed with surgery after meeting a patient who had undergone the spinal stabilization procedure. His vital capacity was 12%, and surgery was withheld because of poor pulmonary reserve. He died 8 months later of pneumonia.

The course of the second patient who declined surgery was similar. His thoracolumbar curve was 40° and his vital capacity was 38%. Twenty-seven months later, he could no longer endure his seating discomfort and mobilization problems. However, his curve was now 95°, and his vital capacity was 7%; therefore, surgery was canceled. He succumbed to respiratory complications 15 months later.

Specialized preoperative screening and postoperative management for DMD patients has decreased postoperative complications significantly.[23] In our series, preoperative pulmonary consultations were obtained to assist in postoperative pulmonary care. Early postoperative mobilization was also emphasized. After remaining supine for the first day to achieve further wound hemostasis through compression, all patients were sitting in a chair within 2 or 3 days. As a result, these patients experienced a significant decrease in pulmonary complications and permanent loss of motor strength that had been observed in DMD patients treated with bed rest for long periods of time.[21,24] In addition, no postoperative cast or brace immobilization was required; therefore, the added expenses of braces and patient monitoring for poor compliance were eliminated.[11] All patients were discharged from the hospital within 7 to 11 days with no significant postoperative cardiopulmonary complications.

Spinal stabilization surgery is now offered to all patients whose Cobb angle approaches 30°, since subsequent progression to a much larger curve is usually inevitable.[17] We observed this finding in two patients who had rapid curve progression of 49° and 52° over a 12-month routine follow-up.

In addition to curve progression, deterioration of the patients' pulmonary function with time is inevitable. In this series, the two patients who initially refused surgery and later requested spinal stabilization demonstrated the dramatic decline of vital capacity that others have reported.[11,25] Therefore, spinal stabilization surgery is better performed in younger patients who have a higher baseline pulmonary status. Younger patients also have fewer muscular contractions and fibrosis, which makes spinal distraction easier and decreases intraoperative bleeding.

During this series, we switched from a sublaminar wiring technique to intraspinous wiring because of complications associated with the former technique. Intraspinous wiring was preferred to avoid compromising the dura and impinging on nerve roots. Anesthesia time was decreased from an average of 6.3 hours in the first four patients undergoing sublaminar wiring to 4.9 hours in the succeeding five patients undergoing intraspinous wiring; much of this saved time was attributed to the change in technique. Furthermore, intraoperative blood loss was decreased from 3,550 mL to 2,300 mL in the patients who underwent sublaminar wiring.

Although some intraspinous wires did pull out on distraction, the number retained was adequate to maintain reduction in these patients until fusion occurred (Figure 3). We now anchor segmental wires through the spinous processes rather than sublaminarly in all patients because of the decreased anesthesia time, decreased blood loss, avoidance of the dura and nerve roots, and apparent adequate strength of the spinous processes to maintain distraction until spinal fusion occurs.

A minimum vital capacity for this procedure is considered to be 30%, since poor results have been obtained in DMD patients who have undergone spinal surgery with lower vital capacities.[9,15,25] In this series, however, one patient with a vital capacity of 20% had spinal stabilization surgery with minimal complications. Virtually bedridden before surgery, this patient could satisfactorily maneuver his wheelchair postoperatively. We thus recommend that selected patients with a vital capacity of less than 30% be evaluated individually for spinal surgery.

Fusion to the L-5 level instead of the sacrum was initially advocated by Sussman.[3] Fusion to L-5 only was performed in two patients in this series. One patient developed no change in pelvic obliquity before his death 20 months postoperatively; the other patient developed 30° of pelvic obliquity by 8-months follow-up, which subsequently stabilized. No additional benefit was found in either patient in terms of increased mobility because of a nonfused lumbosacral junction.

All patients who underwent allogenic bone grafting tolerated and incorporated these grafts well. Allogenic bone supplementation in these osteoporotic patients with limited iliac crest autogenous bone allowed performance of extensive fusion. Previous reports have indicated that the fusion mass heals slowly in these patients because of poor quality and insufficient amounts of autogenic bone graft.[24] We now use allogenic bone grafts for fusion in all DMD patients who undergo segmental spinal stabilization.

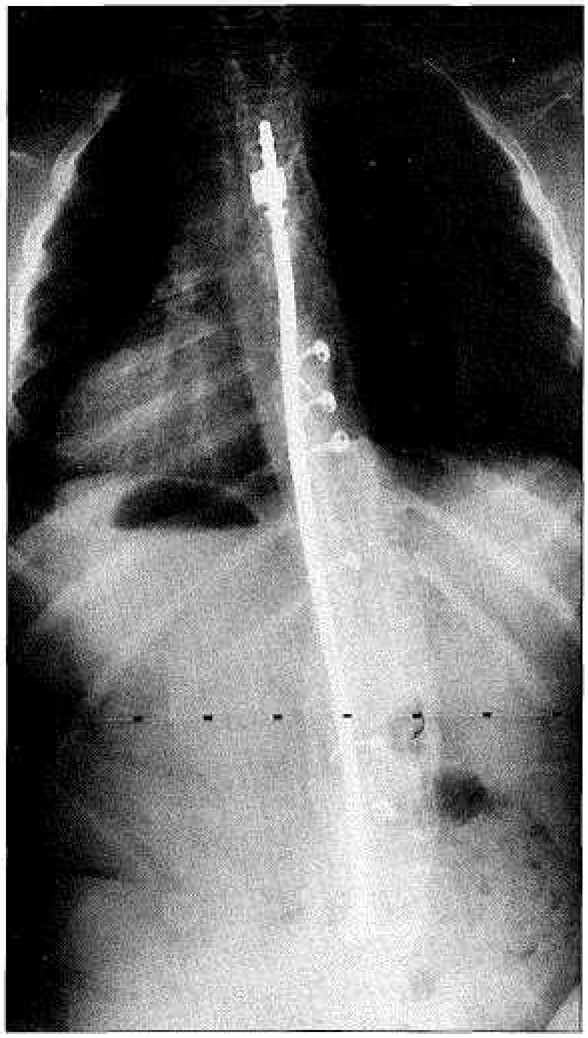

Figure 3. *A 13-year-old male who underwent fusion from T-4 to the sacrum. Although only six intraspinous wires held, sufficient distraction was maintained until fusion occurred.*

Advances in treatment and care of DMD patients has progressed rapidly over the past two decades. Improved respiratory care technology and rapid mobilization make segmental spinal stabilization a viable alternative for patients who, until recently, were unsuccessfully treated with spinal orthoses. We should abandon previous attitudes of avoiding surgery altogether or until absolutely necessary in these patients because of potential complications. Although spinal stabilization with fusion is an extensive procedure, these patients tolerate surgery well. We believe that spinal stabilization and fusion are possible in most DMD patients with the use of perioperative pulmonary consultation and screening, segmental wires anchored through the spinous processes, autogenous bone supplementation with allogenic bone graft, and fusion from the upper thoracic level to possibly the sacrum. Our results support recent recommendations[8,11] that all nonambulatory DMD patients developing scoliosis receive routine spinal stabilization and fusion for a stabilized, relatively straight spine.

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