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

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 $\text{cm H}_2\text{O}$) to be set by a healthcare professional. When patients inhale through Threshold IMT, a spring-loaded valve provides a resistance that exercises respiratory muscles

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 $\text{cm H}_2\text{O}$.

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.

Fig. 2 a,b A 12-year-old boy with Duchenne muscular dystrophy (DMD) being trained to use the Threshold IMT in a sitting position

Results

Details of all the patients are shown in Table 2. Changes in FVC are shown in Fig. 3. The mean FVC at the time of admission was 21.6% (range, 16–27%). There were no ventilator dependent patients preoperatively. FVC increased in all patients at three weeks before operation to 24.1% (range, 19%–28%). FVC increased to 26.2% (range, 22–31%) the day before operation. The mean preoperative scoliotic curvature was 98° (range, 81°–130°). All patients had posterior spine fusion and all-screw construction from the upper thoracic spine (T3 or T4) to the lower lumbar spine (L5) for scoliosis. No anterior surgery was performed

in any of this consecutive series. All patients were extubated on the day of operation in spite of poor preoperative pulmonary function. Frequent aspiration of debris was performed by nurses in the immediate postoperative period. No patients showed abnormal pulse oximetry monitoring values in the immediate postoperative period. No patients developed any respiratory complications such as postoperative pneumonia or required reintubation or tracheotomy. The postoperative scoliotic curvature was 34° (range, 20°–40°) (65%). FVC increased very slightly or remained stable in all patients at six weeks after operation. Although there was very slight increase in FVC of six patients after operation, there was no significant difference in

Table 1 Six-week preoperative IMT exercise training protocol

Subject's baseline MIP pressure: <50cm H ₂ O				
Borg RPE	<13	13–15	>15	>17
Pressure resistance (cm H ₂ O)	Increased by 2	Increased by 1	Maintained at same level	Reduced by 2
Subject's baseline MIP pressure: <50 cm H ₂ O				
Borg RPE	<13	13–15	>15	>17
Pressure resistance (cm H ₂ O)	Increased by 4	Increased by 2	Maintained at same level	Reduced by 2
If subject developed symptoms (i.e. dizziness, lightheadedness, or shortness of breath) while performing IMT exercise, the resistance was adjusted as follows until no symptoms persisted				
Symptoms	Two or more symptomatic episodes in a row per week		1–2 isolated symptomatic episodes per week	
Pressure resistance (cm H ₂ O)	Decreased by 2. Subjects were called back 3 days later to monitor subject's response		Held constant, subjects were called back three days later to monitor subject's response	

IMT inspiratory muscle strength training, *MIP* maximal inspiratory pressure, *RPE* rating of perceived exertion

IMT exercises were performed daily for ten weeks with three sets of 15 repetitions. If a subject achieved the maximum IMT trainer pressure resistance of 41 cm H₂O and resistance could no longer be increased, a fourth set of exercises was added with an increased number of repetitions up to a maximum of 15 repetitions. Initial resistance (H₂O cm) of the IMT was set at 30% of the subjects pretest MIP. Subjects were called once per week by one of the investigators to assist with IMT pressure resistance training progression. IMT pressure resistance was increased weekly according to the subject's symptoms

Table 2 Details of the patients and operative details

Patient no.	Age (y)	Six weeks before operation	The day before operation		Immediately after operation		Two years after operation		Operative time (min)	Intraoperative blood loss (ml)	Complications
		FVC(%)	Scoliotic curvature (°)	FVC(%)	Scoliotic curvature (°)	FVC(%)	Scoliotic curvature (°)	FVC(%)			
1	11	23	100	28	38	29	40	19	249	889	Tachycardia
2	12	22	93	27	35	28	35	20	267	867	-
3	12	27	91	31	32	31	33	25	271	921	-
4	14	18	81	23	27	24	29	19	264	964	-
5	13	26	93	28	20	28	23	22	268	868	-
6	14	16	130	20	38	21	39	18	327	933	Tachycardia
7	11	26	101	29	35	29	34	21	268	968	Tachycardia
8	17	21	101	28	40	28	42	21	256	856	-
9	11	19	81	25	32	25	32	16	267	767	-
10	13	22	98	28	35	29	35	21	266	866	Paralytic ileus
11	13	19	128	22	28	22	28	16	277	820	Paralytic ileus
12	13	19	100	24	35	25	34	19	301	744	-
13	12	25	85	29	30	29	32	20	266	806	-
14	11	20	95	26	35	26	37	20	270	760	-
Mean	13	21.6	98	26.2	34 (65%)	26.7	35 (64%)	19.8	273	859	-

FVC before and after operation (26.2% vs 26.7%). Curve correction was excellent but the operation did not increase FVC. FVC continued to decrease after operation and it was 19.8% (range, 16–25%) at two years after operation. The scoliotic curvature at two years after operation was 35° (range, 23°–42°) (64%) and 35° (range, 23°–42°) (64%) at the last follow-up. Maintenance of correction was excellent. Figure 4 is a radiographic example.

Fig. 3 Changes in FVC are demonstrated. After preoperative respiratory muscle training for six weeks, FVC increased in all patients. Although FVC of 13 patients did not exceed 30% at the time of operation, 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 and two years after operation

Five minor complications occurred immediately after operation. Three of five were transient tachycardia for about seven days due possibly to the presence of profound cardiomyopathy but they were treated successfully with medications and resolved uneventfully. Two had paralytic ileus, both of which were treated only by observation without oral intake and cleared in 48 hours. No respiratory complications such as postoperative pneumonia or prolonged respiratory dependence were found. There were no neurological complications, infection or instrumentation failure during the study period. Up to the present, all patients are still alive at 3.5 years (range, 2.0–5.5 years) after operation.

Discussion

Posterior spinal fusion for scoliosis in DMD should be done early enough in the course of curve progression, when pulmonary and cardiac function are sufficient, so the patients can be anaesthetised and operated upon relatively safely and in order to reduce the likelihood of major complications [3, 5, 7–9, 11, 18–22]. Some authors recommended early operation on these patients in order to avoid anaesthetic, peri/postoperative complications [3, 5, 7–9, 18–22]. Correction or prevention of scoliosis by posterior spinal fusion has been highly effective in stabilising the spine and maintaining seating balance and comfort, and it is accepted as the optimal procedure [3, 8, 18–22]. However, this major procedure is accompanied by many significant complications [3, 9, 16–18]. These complications include superficial and deep wound infection, pseudoarthrosis of the fusion, which may result in pain, progression of the deformity, and implant failure. Furthermore, due to the cardio-

Fig. 4 Preoperative sitting radiographs of a 12-year-old boy with Duchenne muscular dystrophy (DMD). Anteroposterior radiograph shows a severe scoliosis of 128° . FVC at six weeks before operation was 19% and FVC increased to 22% the day before operation. **C** and **D** Surgical correction of scoliosis was performed successfully. Successful extubation was performed on the operative day. Postoperative sitting views show significant coronal correction of 28° (78%). At six weeks after operation, FVC remained stable at 22%

pulmonary compromise of these patients, they are more susceptible to postoperative pneumonia, prolonged respirator dependence, and death. Significant complication rates from 44% to 62% have been reported in some studies [3]. To the best of our knowledge, there has been little information about clinical outcomes and peri/postoperative complications in DMD patients with scoliosis and poor pulmonary function undergoing spine surgery.

Surgeons are often reluctant to perform elective surgery on patients who are respirator dependent or whose forced vital capacities or vital capacities do not exceed 40% of predicted normal [2]. Usually, the limits for the operation are not the extent of the curvature but a minimum forced vital capacity of about 30% of normal [9, 14, 18, 19]. Several previous

studies have attempted to quantify peri/postoperative complications in patients with various diagnoses and poor pulmonary function undergoing surgery for scoliosis. In 1977, Sakai et al. [17] recommended preoperative tracheotomy if the FVC was less than 40% of predicted normal in patients with DMD undergoing scoliosis surgery. In 2005, Bach et al. [2] referred to the role of noninvasive ventilation and noted that all patients with neuromuscular scoliosis were extubated by the third postoperative day and maintained by noninvasive intermittent positive-pressure ventilation (IPPV). Despite continuous ventilator dependence, no patient developed any postoperative respiratory complications or required a tracheotomy. In our method, all patients had preoperative respiratory muscle training using pulmonary trainers for six weeks after admission and the FVC increased. Then, surgical correction of scoliosis was undertaken and extubation on the operative day was also undertaken. In our series, FVC in all patients increased. However, 13 of 14 patients had FVC of less than 30% at the time of operation. Then, surgical correction of scoliosis was performed successfully, and extubation was performed on the operative day for all patients without any respiratory complications. However, if respiratory complications occur, noninvasive intermittent positive-pressure ventilation (IPPV) or other kinds of assistance of breathing including reintubation and tracheotomy should be considered.

We used Threshold IMT for preoperative respiratory muscle training. Current consensus statements confine recommendation of IMT to patients with inspiratory muscle weakness [15]. As well as in patients with obstructive lung disease, there is a theoretical rationale for strengthening the inspiratory muscles of any patient who experiences inappropriate breathlessness, abnormal respiratory mechanics and/or inspiratory muscle weakness/fatigue. Furthermore, there are a small number of randomised, controlled trials in which IMT has elicited a positive effect upon neuromuscular diseases [23, 24] as well as less rigorously designed and executed studies in conditions such as kyphoscoliosis [10]. Although the number of our patients is small, the efficacy of preoperative respiratory training using Threshold IMT is shown clinically in this study.

Prevention of pulmonary function deterioration has been one of the important goals of scoliosis surgery in DMD patients. Because of the lack of controlled studies, it is uncertain as to whether posterior spinal fusion improves pulmonary function. There are few reports indicating that the rate of pulmonary function deterioration is reduced by surgical correction of scoliosis [6–8]. Kurz et al. [14] suggested negative mechanical effects of the deformed thorax associated with scoliosis on the underlying lung. However, many studies have reported that the surgical correction of scoliosis does not increase FVC and suggested that longevity is not increased by posterior spinal fusion

[4], and prevention or correction of scoliosis in DMD does not alter the decline in pulmonary function, nor does it improve survival [1, 12, 14, 20, 21].

Thus, although there have been controversies regarding the effect of posterior spinal fusion on pulmonary function, there is overwhelming support in the literature regarding the beneficial effects of well-timed posterior spinal fusion on the general well-being of DMD patients [5, 7]. Many studies noted that most patients and their families believed that posterior spinal fusion improved their function, cosmesis, seating balance and quality of life [1, 3, 5, 7–9, 16].

While our patients presented late at the time of operation, surgical correction was performed successfully with no respiratory complications. Also, despite preoperative severe scoliosis in all patients, curve correction and maintenance of correction was excellent. Although longer-term follow-up must be performed to determine the final fate of this series, this study shows that our patients with high-risk pulmonary dysfunction and severe scoliosis in DMD could undergo surgical correction of scoliosis with general anaesthesia after respiratory muscle training. Severe scoliosis with poor pulmonary function in DMD patients has been considered a reason to avoid surgery because of the fear of peri/postoperative respiratory complications. However, we felt such patients with FVC considered too low to permit reasonable surgical risk could undergo surgery and could benefit from posterior spinal fusion for scoliosis and pelvic obliquity.

Conclusions

FVC in DMD patients increased after respiratory muscle training using Threshold IMT. DMD patients with severe scoliosis and FVC considered too low to permit reasonable surgical risk could undergo surgery with general anaesthesia without respiratory complications and could benefit from posterior spinal fusion for scoliosis and pelvic obliquity.

Competing interests The authors declare that they have no competing interests. The funding source had no involvement in the decision to submit the paper for publication.

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