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Correction of Neuromuscular Scoliosis in Patients With Preexisting Respiratory Failure

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Study Design. A prospective observational study in scoliosis patients who were on noninvasive night ventilation for respiratory failure.

Objective. To report the results of spinal deformity correction in a group of patients with progressive scoliosis and rare forms of muscular dystrophy/myopathy with respiratory failure who were on nocturnal ventilatory support at the time of surgery.

Summary of Background Data. This is the first study on the results of deformity correction in a series of patients on ventilatory support.

Materials and Methods. Eight patients (6 males, 2 females) presented with progressive scoliosis and respiratory failure. The mean age at surgery was 12 years (range, 8–15 years). The mean follow-up was 48 months (range, 12–80 months). Outcome measures include lung function (spirometry), overnight pulse oximetry, Cobb angles, duration of stay in Intensive care (ICU), and the total hospital stay.

Results. The mean stay in the ICU was 2.7 days (range, 2–5 days). The mean hospital stay was 14.2 days (range, 10–21 days). The mean preoperative Cobb angle was 70.2° (55°–85°). This changed to 32° (16°–65°) after surgery ($P = 0.0002$). The mean vital capacity at the time of surgery was 20% (range, 13%–28%). The mean vital capacity of patients at last follow-up was 18% (range, 10%–31%). The desaturation noted on the preventilation overnight oximetry was reversed by nocturnal ventilation. All patients recovered well following surgery with no major cardiac or pulmonary complications.

Conclusion. Patients with preexisting respiratory failure on nocturnal noninvasive ventilation can be safely operated for deformity correction. This can help to significantly improve their quality of life.

Key words: muscular dystrophy, neuromuscular scoliosis, respiratory failure, nocturnal ventilation. **Spine 2006; 31:2478–2483**

Surgical correction of the neuromuscular scoliosis is aimed at maximizing function and improving the quality of life. Irreversible changes in cardiac and respiratory function pose serious risks to anesthesia and hence are relative contraindications to surgery. Lung function can deteriorate due to progression of the spinal deformity; hence, achieving spinal fusion is one of the foremost goals of surgical treatment. However, whether scoliosis correction improves respiratory function is contentious. Most research is in patients with Duchenne muscular dystrophy or spinal muscular atrophy.[1–6] Most of this literature suggests that respiratory function following surgery initially deteriorates before returning to preoperative levels, and then it gradually deteriorates with progression of the underlying neuromuscular disease.

In this study, we report on a group of patients with rare forms of muscular dystrophy/myopathies that develop respiratory failure early in the course of their disease. This complicates the management of their spinal deformity. These patients with progressive scoliosis and respiratory failure represent an anesthetic challenge.

We present the results of scoliosis surgery in this series of patients with respiratory failure and on nocturnal ventilatory support.

Materials and Methods

Eight patients were referred to the spinal unit by the Muscle Clinic for management of spinal deformity. All the patients had underlying myopathies. There were 4 patients with multicore myopathy, 2 with merocin-negative congenital muscular dystrophy, 1 with Ullrich's muscular dystrophy, and 1 patient with congenital acyl-CoA dehydrogenase deficiency.

There were 6 males and 2 females. The mean age at surgery was 12 years (range, 8–15 years).

All the patients were commenced on nocturnal ventilation by the home ventilation team when symptomatic nocturnal hypoventilation was diagnosed secondary to Type I respiratory failure.

Type I respiratory failure was diagnosed when patients had low oxygen saturation on overnight pulse oximetry.

All patients were on nocturnal home ventilation using noninvasive ventilation, at the time of surgery. The mean forced vital capacity (FVC) at the time of surgery was 20% (range, 13%–28%). The cardiac function of the patients was normal in 6 patients, 1 patient with multicore myopathy had global impairment of ventricular function, and another had mitral valve prolapse.

Nocturnal Ventilation

Indications. One patient with multicore myopathy was admitted in severe respiratory failure. The other patients were ventilated following symptoms of hypoventilation and episodes of desaturation during overnight pulse oximetry study and the documentation of a FVC $\leq$ 1 L.

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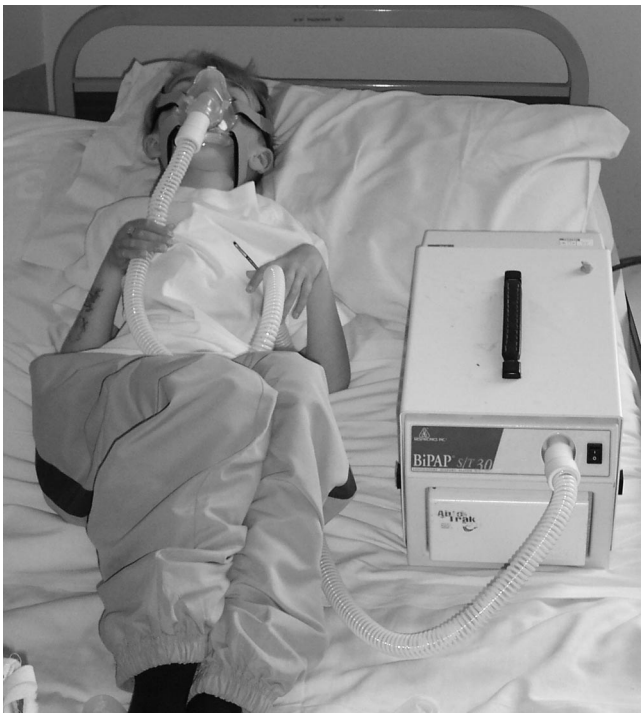

Figure 1. Clinical picture of a patient using ventilator *via* nasal mask.

Symptoms of hypoventilation. Frequent waking at night, unrefreshing sleep, and daytime somnolence. General malaise and weight loss are also reported. Morning headaches are a cardinal sign of hypercapnea.

Technique. Ventilators are used overnight while the patient is asleep (Figure 1). The interface can be either a mask or nasal pillows; some people alternate between them if they have problems with pressure sores or discomfort. In the initial stages, blood gases are done to ensure correct levels. Once a routine is established successfully, then they are monitored 3 to 6 months at the muscle clinic where lung function tests and a clinical assessment of symptoms and well-being are made. Yearly overnight oximetry at home is conducted to check oxygen saturations. The ventilators are serviced regularly. Sometimes, especially in later stages of the disease, the patients have brief top up sessions during the day if they have had a particularly tiring or energetic time. There is a 24-hour system in case of ventilator failure for replacements. The home ventilation services are available anytime for replacement parts, masks, troubleshooting, etc.

Results. Repeated episodes of desaturation during sleep at night were reversed using biphasic positive airway pressure (BIPAP) ventilation with mask/nasal pillows (Figure 2). Clinically, the symptoms regressed within a few months of commencing the ventilation. The patients were now more active in the daytime and could use this time productively. There were no problems reported with the patient compliance with this technique.

Surgical Procedure. All the patients had standard posterior fusion performed by the same surgeon. 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. All curves were fused to L5 distally. Patients were placed prone on a Relton-Hall frame. The posterior elements of the spine were exposed from the upper thoracic spine to the sacrum by stripping the muscles subperiosteally. Spinal cord function was monitored throughout the procedure. The spine was instrumented with the Universal Spinal System (Stratec). Distal fixation comprised of 6-mm pedicle screws in the lumbar spine. Apical and proximal fixation was with a combination of pedicle screws and pedicle hooks with a claw construct at the proximal end of the convex side for proximal fixation. The rods were contoured to recreate the sagittal profile and coronal balance was achieved by sequential reduction of the segments toward the rods. The posterior elements were decorticated and bone grafts placed on the decorticated bed along the length of the instrumentation from the upper thoracic to L5. The pelvis was not included in the instrumentation unless the pelvic obliquity was over 15°. None of the patients in our series required extension of the fusion to the pelvis. Bone grafts for the fusion comprised of a combination of autografts, from the decorticated posterior elements, and milled femoral head allografts.

After surgery, patients were transferred to an ICU where they were gradually weaned off the ventilator and extubated. Postoperative hemoglobin was maintained over 10 g/dL. Sitting up in bed was encouraged the day after surgery, and they subsequently sat in their wheelchairs for increasing times over the next few days. The ambulatory patients were encouraged to return to their preoperative level of ambulation. Clinical and radiologic outpatient reviews were carried out at 6 weeks, 3 and 6 months, and then yearly thereafter. They were discharged from orthopedic follow-up if symptom free after 5 years post surgery. However, they continued to be regularly reviewed at the Newcastle Muscle Clinic.

■ Results

All patients recovered well following surgery with no major cardiac or pulmonary complications. There was no postoperative mortality. The mean stay in the intensive care (ICU) was 2.7 days (range, 2–5 days). The mean hospital stay was 14.2 days (range, 10–21 days). The mean intraoperative blood loss was 1450 mL (500–2600 mL) (Table 1).

The mean vital capacity of patients at last follow-up was 18% (range, 10%–31%). There was no significant difference between the preoperative and postoperative values. However, there was an appreciable increase in the vital capacity in the patient with multiple acyl-CoA dehydrogenase deficiency.

The mean preoperative deformity in the coronal plane as measured by Cobb angle was 70.2° (range, 55°–85°). The mean postoperative deformity was 32° (range, 12°–47°). The average change in the Cobb angle after surgery was 38° (range, 17°–65°). The difference in the preoperative and postoperative Cobb angles was significant ($P = 0.0002$). The follow-up was from 12 to 80 months with a mean of 48 months. The correction of scoliosis was maintained at the last follow-up with no loss of correction or progression of the deformity. There were no implant failures, pseudarthrosis, or major infection.

There were no chest infections or spontaneous pneumothorax after surgery. No patient needed a tracheos-

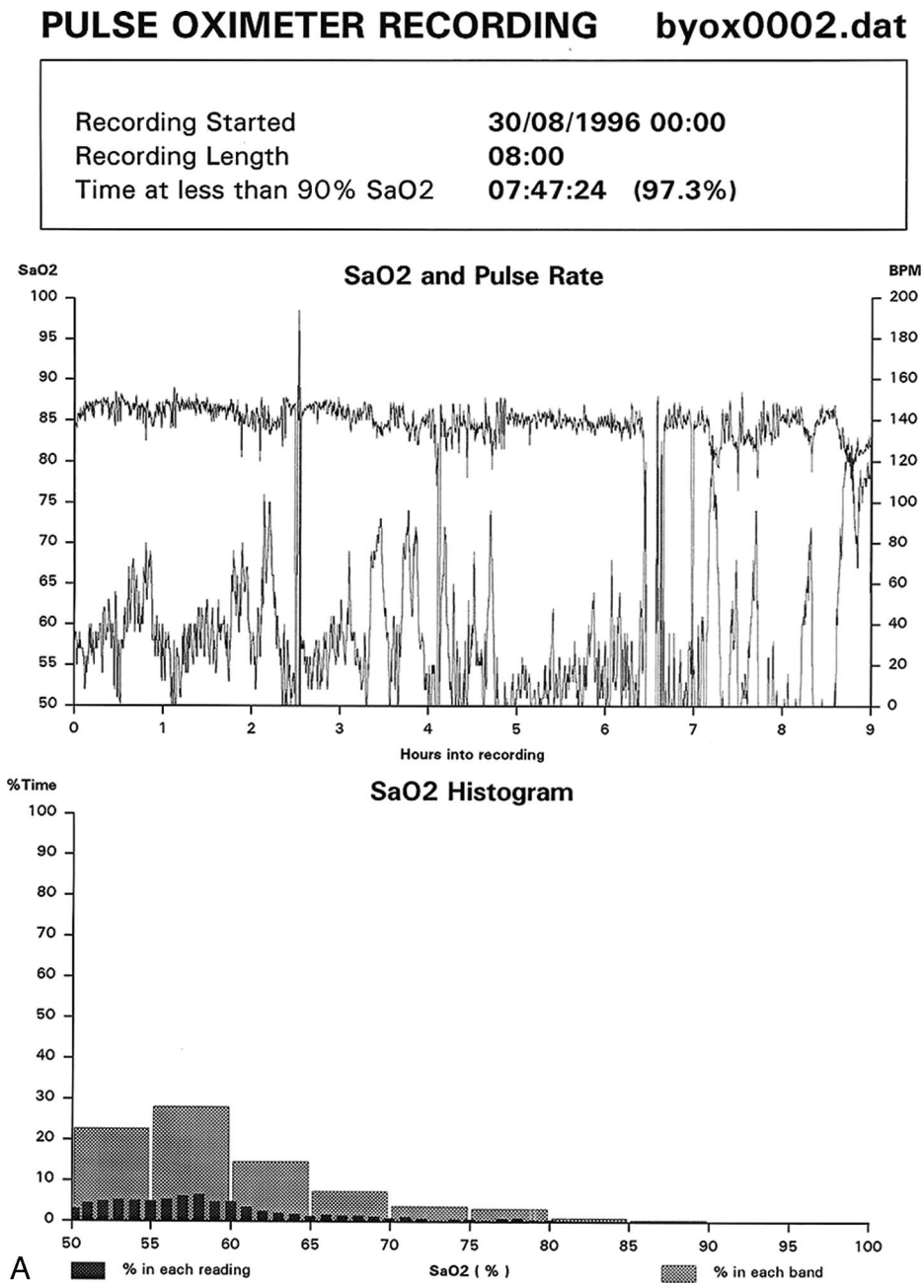

Figure 2. **A**, Overnight pulse oxymetry before institution of nocturnal ventilation showing poor oxygen saturation. **B**, Reversal of hypoventilation after institution of nocturnal ventilation showing dramatic improvement in oxygen saturations.

tomy. All patients were gradually downgraded from CMV-BIPAP to genuine BIPAP over 48 hours after surgery while monitoring the arterial blood gases and pulse oximetry. They were transferred out of ICU when they were stable on genuine BIPAP. They returned to their preoperative ventilation status over the next few days. Thus, there was a smooth transition from no spontaneous breathing (CMV-BIPAP) to spontaneous breathing at both CPAP levels (genuine BIPAP) in these patients reducing the ICU stay and avoiding any problems of weaning off the ventilator. No regional pain control measures were used in any patient.

All the patients maintained their functional status after surgery. Three patients (Cases 3, 6, and 8) were ambulatory at the time of surgery and returned to their preoperative level of function. Thus, none of the ambulatory patients lost this ability as a result of the operation.

■ Discussion

Artificial ventilation technology has made significant advances in the past few decades. In the past, preoperative tracheostomy has been reported in patients with severe restrictive disease.[7] Improved techniques have led to the use to lesser invasive options. Pressure and volume cycled ventilator systems are particularly useful in patients with neuromuscular diseases. Noninvasive home ventilation using bilevel positive airway pressure (BiPAP) has been shown to be safe and effective in pediatric patients.[8]

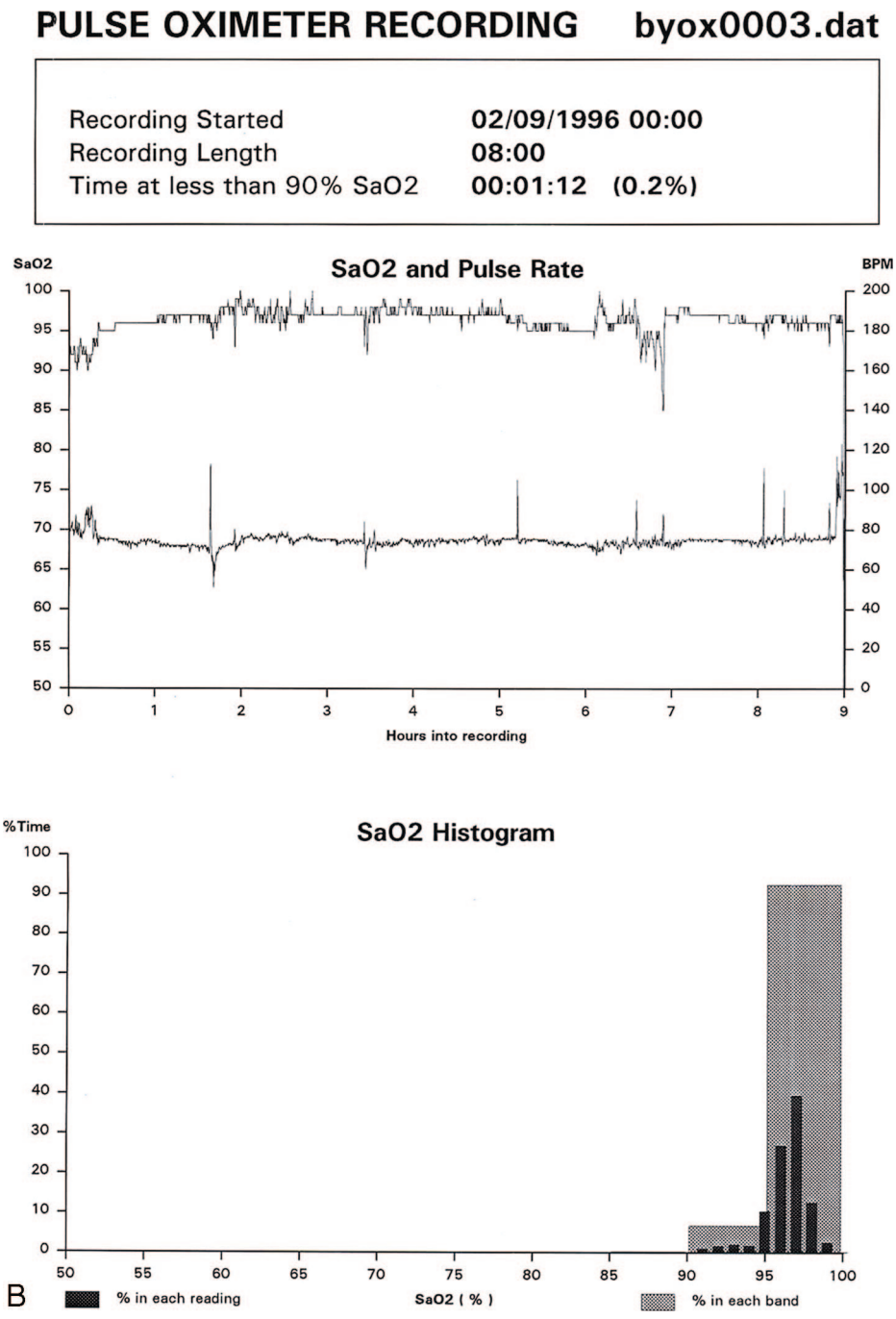

Figure 2. (Continued).

Depending on spontaneous breathing activity, BiPAP can be subdivided into 4 categories[9]:

No spontaneous breathing: CMV-BIPAP

Spontaneous breathing at lower pressure level: IMV-BIPAP

Spontaneous breathing at higher pressure level: APRV-BIPAP

Spontaneous breathing at both CPAP levels: genuine BIPAP

A smooth transition can be achieved from CMV-BIPAP to genuine BIPAP in the postoperative period. Hence, these methods are particularly useful when patients on ventilation need to be given a general anesthesia. This was our observation since all the patients had a smooth transition from assisted ventilation to spontaneous breathing.

Patients with neuromuscular disorders are at high risk for development of sleep-related respiratory disorders and respiratory failure. Nocturnal ventilation is an accepted method to improve life quality and longevity in patients with DMD.[10,11] In patients with multicore myopathy also, it has been shown to reverse the symptoms of nocturnal hypoventilation.[12] Chest wall weakness along with restrictive lung disease caused by chest wall deformity and kyphoscoliosis contribute to this hypoventilation.[13] Polysomnographic evaluation in sleep

Table 1. Patient's Details

No.	Age (yr)	Sex	Diagnosis	FVC Preoperative (%)	FVC Postoperative (%)	Cobb Angle Preoperative (°)	Cobb Angle Postoperative (°)	Follow-up (mo)
1	12	F	Ullrich's congenital muscular dystrophy	24	10	55	30	48
2	13	M	Multicore myopathy	26	17	65	40	80
3	15	F	Multiple acyl-COA dehydrogenase deficiency	13	31	60	12	60
4	14	M	Multicore myopathy	28	25	60	43	80
5	8	M	Merocin-positive congenital muscular dystrophy	16	12	85	20	24
6	14	M	Multicore myopathy	23	19	80	35	18
7	9	M	Merocin-negative congenital muscular dystrophy	14	16	83	47	12
8	11	F	Multicore myopathy	23	17	74	32	60

laboratory or continuous pulse oximetry is required to diagnose nocturnal sleep-related ventilatory alterations. Institution of noninvasive ventilation using BIPAP reverses this ventilatory alterations.[13] The institution of home ventilation also probably helps to optimize these patients for surgery.

There is the first study on the results of deformity correction surgery in a series of patients on ventilatory support for respiratory failure. A strong association between restrictive lung disease and increased pulmonary complications has been reported.[14,15] Padman *et al* reported a positive correlation between decreased vital capacity and increased postoperative complications in children with neuromuscular scoliosis that underwent posterior spinal fusion.[16] Rawlins *et al* found 19% pulmonary complications in patients undergoing reconstructive spinal surgery when the vital capacity was <40% of predicted.[17] None of these patients was on ventilatory support. However, there were no postoperative pulmonary complications in our group of patients, although our patients had a mean vital capacity of 20%. There was no significant difference in the FVC before and after the operation. However, looking closely, there was an appreciable increase in the postoperative vital capacity of 1 patient with multiple acyl-CoA dehydrogenase deficiency. In other patients with congenital muscular dystrophies and multicore myopathies, the postoperative vital capacity was lower than preoperative levels as expected with the progression of the disease.

■ Conclusion

Spinal deformity correction in this group of patients with muscular dystrophy/myopathy on ventilatory support for respiratory failure presented no increased risk of complications. Nocturnal ventilation allowed these patients to tolerate major surgery to prevent progression of scoliosis. They returned to their preoperative level of function and had smooth postoperative recovery. Ventilatory support thus helps these patients to safely undergo major surgery, which can improve their quality of life.

■ Key Points

- Patients with rare forms of muscular dystrophy/myopathy develop respiratory failure early in the course of disease.
- Noninvasive nocturnal ventilation reverses the episodes of hypoventilation and improves well-being.
- Deformity correction in this group of patients represents an anesthetic challenge.
- Nocturnal ventilation helps these patients to undergo major scoliosis correction surgery with no increased complications.

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