

A NEW NASAL MASK FOR NOCTURNAL HOME VENTILATION IN CHRONIC NEUROMUSCULAR DISORDERS

B. Klefbeck,¹ L. Remmer,² J. Weinberg¹ and J. Borg¹

Departments of ¹Neurology and ²Oral Surgery, Södersjukhuset, Stockholm, Sweden

ABSTRACT. A new nasal mask for nocturnal, intermittent positive pressure ventilation was used by patients with symptomatic sleep hypoventilation due to chronic neuromuscular disorders. The custom-fabricated mask is cast in visible light curing acrylic from a plaster model. Air is delivered through plastic tubes cast in the mask. The treatment was introduced through close cooperation between a physiotherapist and a technician. Evaluation was performed after 4-34 months' regular use. The mask was well tolerated. All patients reported comfortable sleep and symptom relief. The effectiveness was confirmed by whole-night measurements of oxygen saturation and end-tidal carbon dioxide.

Key words: home ventilation, nasal mask, neuromuscular disorders.

INTRODUCTION

Respiratory muscle involvement might cause chronic alveolar hypoventilation and respiratory failure in several chronic neuromuscular disorders including muscle dystrophies, motor neuron diseases and residuals after prior polio (4, 6, 12, 13, 14, 16). Chronic hypoventilation might cause an insidious development of incapacitating symptoms including severe reduction of physical and mental capacity due to nocturnal blood gas and sleep disturbances (4, 12, 13, 15). The treatment possibilities have increased by the introduction of improved techniques for non-invasive, assisted ventilation (1, 3, 5, 8, 10). Assisted ventilation with intermittent positive pressure by home ventilators connected with a nasal mask is more safe and efficient than negative pressure methods and is a less complicated alternative to permanent tracheostomy (3, 10). A comfortable mask, permitting minimal air leakage without causing tissue damage is crucial for treatment success. This is not always achieved by commercially available nasal masks (2, 9, 10). We have developed a new, custom-fabricated mask aimed at minimizing

skin pressure and discomfort and achieving high effectiveness. The mask, training procedure and long-term evaluation are reported.

PATIENTS AND METHODS

Patient data are summarised in Table I. Eleven patients with respiratory failure due to muscular dystrophy (3), chronic neuropathy (2), slowly progressive motoneuron disease mainly affecting the lower moto neurons (1) or prior polio (5) were treated and used the individually fitted nasal mask during 4-34 months (Median 21).

Nine patients were able to handle the equipment without assistance, 2 patients needed assistance due to reduced function of the upper extremities. Six patients used wheel chairs, 3 patients used crutches, 2 patients used no aids for locomotion.

The mean age of the patients was 53.7 years (range 39-64). In the polio group the mean time elapsed since the acute infection was 45 years (range 36-53 years). The mean vital capacity was 1.5 liter (range 0.7-2.9) corresponding to 37% (range 21-60) of the predicted value.

In 3 patients, assisted ventilation was started during intensive care due to severe carbon dioxide retention. In 8 patients, assisted ventilation was started due to a history of increasing dyspnoea, sleep disturbances, morning headache

Table I.

Pat. no	Age (years)	Diagnosis	Vital capacity		Ass.v. months
			% predicted	liters	
1	56	MD	22	0.7	33
2	58	MD	60	2.9	14
3	39	MD	21	1.2	22
4	53	MND	43	1.8	4
5	61	NP	31	1.1	21
6	58	NP	53	1.3	10
7	46	PP	32	1.6	34
8	43	PP	29	1.2	25
9	64	PP	30	1.3	14
10	59	PP	38	0.8	19
11	54	PP	49	2.4	30

MD = Muscle Dystrophy

MND = Motorneuron Disease

NP = Chronic Neuropathy

PP = Post polio

Ass.v. = assisted ventilation with a nasal mask

and daytime hypersomnia and nocturnal blood gas disturbances (cf. Results).

Treatment evaluation included whole-night recordings of the oxygen saturation (Radiometer Oximeterd connected by an ear probe) and carbon dioxide tension in end-tidal expired air (Engström Eliza CO₂ analyzer). A separate chart recorder was used (paper speed 10 mm/min).

The ventilator

All patients used PLV-100 (Life Care Products) which is a volume-cycled, portable home-ventilator.

The nasal mask

To provide a mask model an alginate impression of the nose is used to make a plaster. The plaster model is modified to obtain maximal tightness of seal. Air is delivered through plastic tubes with an inner diameter of 6.8 mm. The acrylic gel is formed on the plaster model and preshaped plastic tubes are connected. It is important to consider the patients nose/ear angle to get an optimal position of the tubes permitting the patient to rest on one side without the mask being displaced and without discomfort. When the plastic tubes have been attached to the plaster model the mask is cast in visible blue light (400 nm) curing acrylic, which is a hard acrylic material. The mask is then tried in connection with the ventilator and minor adjustments are made if required. The tubes are then attached to the individually adjusted headgear and care is taken to obtain correct angles at the traction lines to ensure tightness of seal. If necessary, a dental splint is used to fixate the mask (Figs. 1 and 2).

RESULTS

Training procedure

Initially a Respironics CPAP mask was used to get the patient accustomed to the ventilator and nasal ventila-

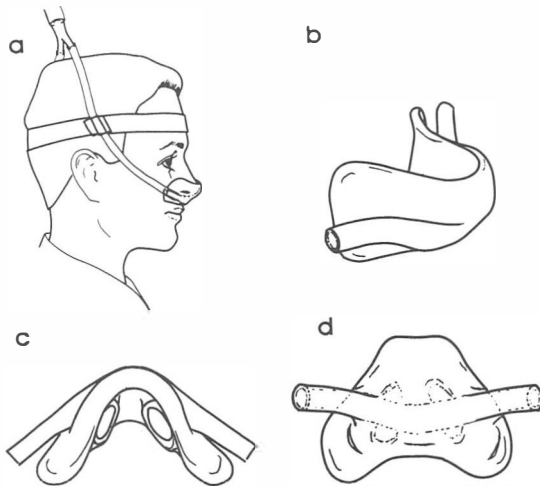


Fig. 1. Nasal mask for nocturnal ventilation: (a) connected, (b) lateral view, (c) dorsal/inside view, (d) frontal view.

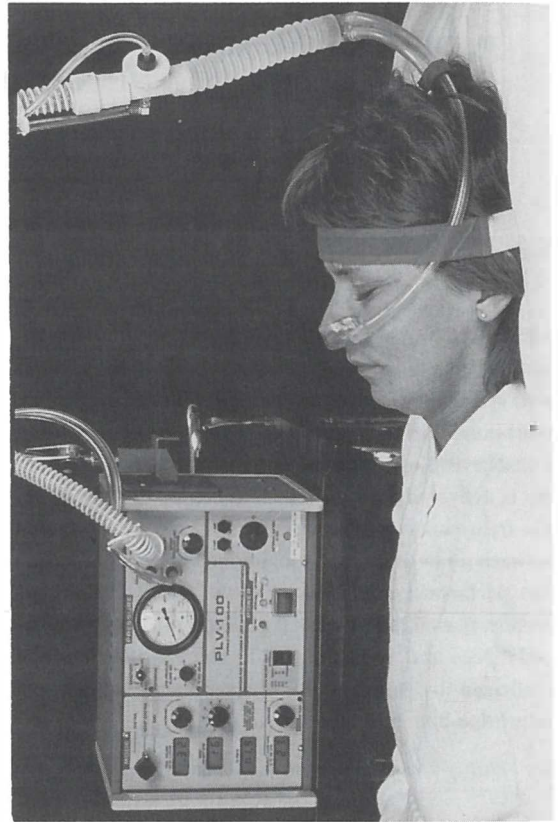


Fig. 2. The patient connected to a PLV 100.

tion. The individually fitted nasal mask was then introduced and ventilator settings were adjusted. The patients were taught how to use the mask and the headgear. Only a few hours of daytime training was necessary before nocturnal use. After two to ten nights all patients reported a comfortable sleep with the equipment. In patients with a tendency to air leakage through the mouth, straps around the chin were used.

Clinical evaluation

All patients reported improved sleep and improved physical and mental capacity as compared to before the treatment. No one complained of morning headache or significant day-time fatigue. At the long-term follow up, the nasal mask was well tolerated by all patients. There were few complaints of air leakage and none of irritated eyes. Impressions on the skin were minor and no patient exhibited pressure sores. The head-gear requires low pressure of the straps due to the

smaller surface area of the mask. The mask allows the use of spectacles (at reading before going to sleep) and permits variation of sleeping position without increased air leakage. The material or the shape of the mask did not change during the follow-up period and no further adjustments were needed after the initial.

Nocturnal blood gas evaluation

Nocturnal blood gas monitoring showed significant improvement. Before treatment daytime $p\text{CO}_2$ was > 7.0 kPa in 5 patients. In all, except the patient with motoneuron disease, nocturnal CO_2 -retention exceeded 13%, which was considered significant (9), with maximal $p\text{CO}_2$ -levels ranging 6.4–14.0 kPa. The patient with motoneuron disease had repeated oxygen desaturations within the 90–96% range and according to a sleep study never got into stage 2 sleep.

At the follow up daytime $p\text{CO}_2$ was < 6.4 kPa in all patients. Nocturnal $p\text{CO}_2$ was < 7.0 kPa in all patients except for a few episodic values above this level in two patients at most to 8.2 kPa. None exhibited significant, nocturnal CO_2 -retention.

Before treatment all patients except the motoneuron disease patient (cf. above) exhibited severe oxygen desaturations with frequent episodes within the 50–80% range. At follow up the nocturnal oxygen saturation was continuously above 90%, usually about 95%, except for a few episodic values below this level in 3 patients, at most to 82%.

DISCUSSION

Nocturnal home ventilation with this custom-fabricated nasal mask was efficient with regard to blood gas restoration and symptom relief. The new mask caused no tissue damage and was considered comfortable. This mask is an alternative to commercially available masks when there are problems with skin pressure or discomfort.

The treatment procedure was appropriate with regard to easy introduction, also in an intensive-care unit, and with regard to the long-term results.

A prerequisite of the mask is the new visible light curing material used. Another is a close cooperation between the technician and the physical therapist both initially and during long-term follow-up.

ACKNOWLEDGEMENT

This study was supported by grants from the Swedish association for Traffic and Polio Disabled and Eugeniastiftelsen.

REFERENCES

1. Borg, J., Weinberg, J. & Klefbeck, B.: When is assisted ventilation indicated in neuromuscular diseases? Abstract of the VII International Congress on Neuromuscular Diseases, J Neurol Sciences 98: 342, 1990.
2. Bach, J. R. & Alba, A.: Management of chronic alveolar hypoventilation by nasal ventilation. Chest 97: 52–57, 1990.
3. Branthwaite, M.: Mechanical ventilation at home. Br Med J 298: 1409, 1989.
4. Bye, P., Ellis, E., Faiq, I., Donnelly, P. & Sullivan, C.: Respiratory failure and sleep in neuromuscular disease. Thorax 45: 241–247, 1990.
5. Carroll, N., Branthwaite, M.: Intermittent positive pressure ventilation by nasal mask: technique and applications. Intens Care Med 14: 115–117, 1988.
6. Ciriginotta, F., Mondini, S., Zucconi, M., Barrot-Cortes, E., Sturani, C., Schiavina, M., Coccagna, G. & Lugaresi, E.: Sleep-related breathing impairment in myotonic dystrophy. J Neurol 235: 80–85, 1987.
7. Corbascio, M., Granata, C., Capelli, T., Sabattini, L., Bartone, M. T. & Martini, L.: Mechanical ventilation in Duchenne muscular dystrophy to counter vital capacity loss. Abstract of the VII International Congress on Neuromuscular Diseases. J Neurol Sci 98: 340, 1990.
8. Ellis, E., Bye, P., Bruderer, J. & Sullivan, C.: Treatment of respiratory failure during sleep in patients with neuromuscular disease. Am Rev Resp Dis 135: 148–152, 1987.
9. Kerby, G. R., Mayer, L. S. & Pingleton, S. K.: Nocturnal positive pressure ventilation via nasal mask. Am Rev Resp Dis 135: 738–740, 1987.
10. McDermott, I., Bach, J., Parker, C. & Sortor, S.: Custom-fabricated interfaces for intermittent positive pressure ventilation. Int J Prosthodont 2: 224–233, 1989.
11. Midgren, B.: Continuous non-invasive assessment of blood gases during sleep. Thesis, Univ. Hosp. Lund, 1987.
12. Newsom-Davis, J., Goldman, L. & Casson, M.: Diaphragm function and alveolar hypoventilation. Q J Med 177: 87–100, 1976.
13. Newsom-Davis, J.: The respiratory system in muscular dystrophy. BR Med Bull 36: 135–138, 1980.
14. Smith, P., Calverley, P. & Edwards, R.: Hypoxemia during sleep in Duchenne muscle dystrophy. Am Rev Resp Dis 37: 884–888, 1988.
15. Steljes, D., Kryger, M., Kirk, B. & Millar, T.: Sleep in postpolio syndrome. Chest 98: 133–140, 1990.
16. Svanborg, E., Borg, J., Weinberg, J.: Sleep and respiration in chronic neuromuscular disorders. Abstract of the VII International Congress on Neuromuscular Diseases. J Neurol Sci 98: 342, 1990.

Address for offprints:

Brita Klefbeck
Department of Neurology
Södersjukhuset
S-118 83 Sweden
Tel 08/6162191
Fax 08/6162917