Eur Respir J 2000; 16: 476–481
Printed in UK – all rights reserved

European Respiratory Journal
ISSN 0903-1936

Outcome of paediatric domiciliary mask ventilation in neuromuscular and skeletal disease

A.K. Simonds*, S. Ward*, S. Heather*, A. Bush*, F. Muntoni**

Outcome of paediatric domiciliary mask ventilation in neuromuscular and skeletal disease. A.K. Simonds, S. Ward, S. Heather, A. Bush, F. Muntoni. ©ERS Journals Ltd 2000.

ABSTRACT: Noninvasive positive pressure ventilation delivered by nasal mask or facemask has been used widely in the last decade to manage chronic ventilatory failure in adults with neuromuscular and chest wall disease. However, it has been thought that paediatric patients would not be able to tolerate masks, and previous anecdotal reports on the paediatric application of mask ventilation have not assessed the effects on nocturnal and arterial blood gas control.

Domiciliary mask ventilation has been used in 40 children with ventilatory insufficiency due to congenital neuromuscular and skeletal disease aged 9 months–16 yrs. Eighteen patients had symptomatic nocturnal hypoventilation, 17 had diurnal ventilatory failure, three were referred for weaning and two had frequent chest infections associated with sleep-disordered breathing.

Thirty eight of the 40 patients tolerated mask ventilatory support long-term. Diurnal mean±SD oxygen tension in arterial blood (P_{a,O_2}) increased from 8.5±1.8–10.9±1.7 kPa ($p<0.001$) and meansd carbon dioxide tension in arterial blood (P_{a,CO_2}) fell from 7.0±1.6–5.9±0.8 kPa (p=0.01) following initiation of ventilatory support. Mean and minimum nocturnal P_{a,O_2} and peak transcutaneous carbon dioxide tension (P_{tc,CO_2}) (n=21) improved significantly.

Mask ventilation can be used successfully in young children and reverses ventilatory insufficiency due to congenital neuromuscular and skeletal disease.

Eur Respir J 2000; 16: 476–481.

*Royal Brompton and Harefield NHS Trust, and **Hammersmith Hospital, London, UK.

Correspondence: A.K. Simonds, Respiratory Support Service, Sleep & Ventilation Unit, Royal Brompton and Harefield NHS Trust, Sydney Street, London SW3 6NP, UK. Fax: 44 2073518911.

Keywords: Mechanical ventilation
neuromuscular disease
NIPPV

Received: March 19 1998
Accepted after revision November 7 1998

Noninvasive ventilation (NIV) using a nasal mask or facemask is an effective form of ventilatory support in adults with chronic ventilatory failure arising from stable neuromuscular disease and chest wall disorders [1, 2], and in selected cases of obstructive lung disease [3]. NIV has also been extended to adolescents and adults with hypercapnic respiratory failure arising from progressive neuromuscular disorders such as Duchenne muscular dystrophy (DMD) [4, 5]. Encouraging results have been obtained with nasal ventilation improving survival, arterial blood gas tensions, and health-related quality of life [6].

Current guidelines [7] suggest that ventilatory support should be considered in DMD when daytime carbon dioxide tension in arterial blood (P_{a,CO_2}) exceeds 6.0 kPa, but there is no consensus on the role of NIV in other congenital neuromuscular disorders. There are only a few studies of the outcome of long-term nasal ventilation [8, 9] and continuous positive airway pressure (CPAP) [10] in paediatric patients, despite an early case report [11], and it has been suggested that these techniques might not be feasible in young children because of difficulty coping with the mask interface [11, 12].

FAUROUX *et al.* [13] have described the long-term use of home ventilation in France. The majority of children had restrictive disorders, but a small number with cystic fibrosis were included. However, the proportion of children using noninvasive as opposed to invasive ventilation in each diagnostic group is unclear, and the average age on initiation of ventilatory support is not given. In addition, previous reports have not provided data on the effects of NIV on nocturnal and diurnal hypoventilation. Ethical concerns have also been raised regarding the appropriateness of ventilatory support in progressive neuromuscular disease after a study of the prophylactic use of nasal ventilation in normocapnic boys with DMD showed that mask ventilation offered no benefit compared with a control group who did not receive ventilatory assistance [14].

In this series 40 children with diurnal and/or nocturnal ventilatory insufficiency associated with congenital neuromuscular, neurological or skeletal disease have been treated with domiciliary mask ventilation, with the aim of assessing the tolerance of the technique and its impact on nocturnal hypoventilation and diurnal arterial blood gas tensions.

Patients

Forty children presented consecutively with ventilatory problems, aged 9 months–16 yrs, were included. Diagnostic categories were intermediate spinal muscular atrophy (SMA) n=8; congenital muscular dystrophy (CMD) n=8 (five merosin positive, three merosin negative); congenital myopathy (CM) n=6 (two centronuclear, two nemaline, two minimal change); DMD n=7; and a miscellaneous

group n=11 (three limb girdle muscular dystrophy, two congenital central hypoventilation syndrome, and one each of: facioscapulo-humeral muscular dystrophy, Prader Willi syndrome, Leigh's syndrome, osteogenesis imperfecta, congenital idiopathic thoracic scoliosis and hereditary sensory motor neuropathy).

Methods

Indications for mask ventilation

Seventeen patients presented with diurnal hypercapnic respiratory failure, three were referred from intensive care units elsewhere for weaning from conventional mechanical ventilation, 18 had symptomatic nocturnal hypoventilation and two had frequent chest infections associated with sleep-disordered breathing.

Investigations

Baseline diurnal blood gas measurement was carried out using earlobe arterialized samples, and overnight monitoring of respiration (sleep study) was performed in individuals with suspected nocturnal hypoventilation. Variables monitored during the sleep study included arterial oxygen saturation (S_{a,O_2}) and transcutaneous CO_2 tension P_{tc,CO_2} in all cases. In some patients multichannel polysomnography (S_{a,O_2}, P_{tc,CO_2}, oronasal airflow, chest and abdominal movement, chest wall paradox, body position and snoring detection) was performed using the Densa 600 Pneumograph [15]. Initial sleep studies were not performed in the three subjects referred for weaning and in 16 subjects who presented with uncontrolled diurnal hypercapnia.

Ventilatory equipment

Ventilatory support was initiated in the hospital in all cases. Thirty-six patients with nocturnal and/or diurnal hypercapnia were treated with nasal ventilation, four with frequent serious chest infections (more than three infective episodes requiring hospital admission in the previous year) and nocturnal arterial oxygen desaturation in the presence of normocapnia, were treated with nasal continuous positive airway pressure (CPAP) therapy at night. Two of these were subsequently transferred to nasal ventilation after several months because of the development of hypercapnia (P_{a,CO_2} >7.5 kPa) while using CPAP.

Nineteen children used the bilevel positive airway pressure (BiPAP) ventilator (Respironics Inc., Murrysville, USA) in spontaneous/timed (S/T) mode, 12 used the pressure preset Nippy ventilator (B & D Electromedical, Warwickshire, UK), six the bilevel pressure support DP90 machine (Taema, Cedex, France), and one used the Breas PV401 (Breas, UK). Those receiving CPAP used the Sullivan Elite CPAP generator (ResMed, Abingdon, UK). The total number of ventilators exceeded 40 as two children had back-up ventilators to use in the event of equipment malfunction. Masks were selected taking into account the preference of the child, comfort, and the adequacy of ventilation. Twenty children used facemasks, 18 nasal masks and two the Adams circuit (Puritan Bennett, UK). Medium size adult nasal masks were used as facemasks in some cases. Despite the increased deadspace of the facemask, control of CO_2 was achieved by careful monitoring of P_{tc,CO_2} and titration of the ventilatory settings. Customized masks were required for two children and constructed by S. Ward and S. Heather. Most children required help in securing the interface, but all could summon help when the mask was in place by calling or using a buzzer alarm system. Ventilator settings were determined in each child by overnight monitoring of S_{a,O_2} and P_{tc,CO_2}, with the aim of maintaining P_{tc,CO_2} within the normal range. Supplemental oxygen was added only if S_{a,O_2} could not be maintained >90% overnight in the presence of optimal CO_2 control. Patients were advised to use nasal ventilation overnight. A few children used their ventilator for short periods during the day, *e.g.* after returning from school. One child with severe limb girdle muscular dystrophy used nasal ventilation for 22 $h \cdot day^{-1}$. Twelve patients used machines with low pressure, high pressure and power failure alarms. All ventilators had a backup respiratory rate set just below the patients resting respiratory rate.

Parents and carers were taught to perform daily chest physiotherapy for the children during a session of ventilatory support. A modified active cycle of breathing was used to achieve effective coughing. The inspiratory positive pressure (IPAP) level was temporarily increased by 2–4 cmH_2O, and manual chest capping and shaking combined with assisted cough/huffing to facilitate clearance of sputum.

Follow-up and maintenance of equipment

By the time of discharge each family completed competency training in equipment use, and was provided with a care plan giving advice regarding readmission, outpatient follow-up, and simple equipment maintenance. All patients and families had access to a 24 h telephone hotline service to contact if medical or technical problems arose. Triage was carried out by either a respiratory nurse or respiratory support coordinator who could arrange physician review or technical support, depending on the nature of the problem. Children were routinely reviewed every 3–6 months in the ventilatory support outpatient clinic. Respiratory support technicians provided a daily walk-in clinic for ventilator and mask-related problems. Ventilators and CPAP generators were repaired when necessary and serviced in the patient's home, in accordance with the manufacturers' instructions. Patients who were unable to breathe spontaneously for more than 12 h without a ventilator were provided with a backup machine and battery pack to power the ventilator in the event of power cuts, for use during trips outside the home, and to facilitate transfer to hospital.

Analysis of data

Diurnal arterial blood gas tensions, mean and minimum nocturnal S_{a,O_2}, and peak nocturnal P_{tc,CO_2} were compared before and after NIV using a paired t-test. Diurnal blood gas tensions were measured with the patient breathing spontaneously on air at the same time of the day in each subject (apart from two subjects who had initial

measurements made while receiving oxygen therapy). Nocturnal values were obtained in the baseline study with the patient breathing spontaneously and in the follow-up study during NIV support.

Results

The mean±SD age at the start of nasal ventilation in each diagnostic category is given in table 1. Diurnal arterial blood gas results before NIV and after application of NIV while breathing spontaneously, and before discharge home are shown by one diagnostic group in table 1. Individual results for each child are given in fig. 1. For the group as a whole (n=40), mean±SD P_{a,O_2} increased from 8.5±1.8–10.9±1.7 kPa ($p<0.001$) and mean±SD P_{a,CO_2} fell from 7.0±1.6–5.9±0.8 kPa (p=0.01). It should be noted that 18 subjects were treated for symptomatic nocturnal hypoventilation, and in these children baseline diurnal P_{a,CO_2} was not elevated. Before and after NIV paired sleep studies were available in 21 patients. Overnight S_{a,O_2} and P_{tc,CO_2} improved significantly on nasal ventilation for the group as a whole (fig. 2). Nocturnal mean±SD minimum S_{a,O_2}, mean S_{a,O_2}, and maximum P_{tc,CO_2} were 65±19.2%, 85.2±7.2% and 8.8±2.15 kPa, respectively during the baseline sleep study and improved to 84±8.6%, 92.4±2.7% and 7.6±1.4 kPa (p=0.03) while receiving noninvasive positive pressure ventilation (NIPPV). After the sleep study on NIV, modifications to the initial ventilator settings were made with the aim of improving nocturnal CO_2 control further. Subgroup analysis of changes in diurnal and nocturnal blood gas tensions within diagnostic groups showed trends in improvement, which did not reach significance in all categories, probably because of the small numbers in each subgroup.

However, there was a significant increase in mean±SD diurnal P_{a,O_2} in the DMD and CM groups before and after NIV (DMD 9.3±2.3 *versus* 12.17±1.46 kPa, p=0.025; and CM 8.2±0.6 *versus* 11.3±1.15 kPa, p= 0.02). Both minimum and mean nocturnal S_{a,O_2} were improved in the DMD and CMD categories (DMD min S_{a,O_2} 55.4±15 *versus* 88.6±3.0%, p=0.015, mean S_{a,O_2} 81.6±9.1 *versus* 93.6±1.5%, p=0.043; CMD min S_{a,O_2} 72.5±17 *versus* 90.2±3.1%, p=0.05, mean S_{a,O_2} 88.8±5 *versus* 94.2±2.2%, p=0.05). Five children (two with CMD, two with CM and one scoliosis) required supplemental oxygen therapy at night. Two were able to discontinue oxygen therapy after 6 months of NIV because of improvement in P_{a,O_2}. Average ventilator settings were inspiratory positive pressure 14 cmH_2O, expiratory positive pressure level (where available) 4 cmH_2O. CPAP was used in four children with nocturnal S_{a,O_2} in the absence of a significant rise in P_{a,CO_2}. However, two developed hypercapnia on CPAP and were transferred to nasal ventilation. Mean duration of use of mask ventilation for the group so far is 30 months (range 1–105 months). One child with CMD (aged 11 yrs) could not tolerate nasal ventilation long-term and did not return for follow-up. She died of respiratory failure after 21

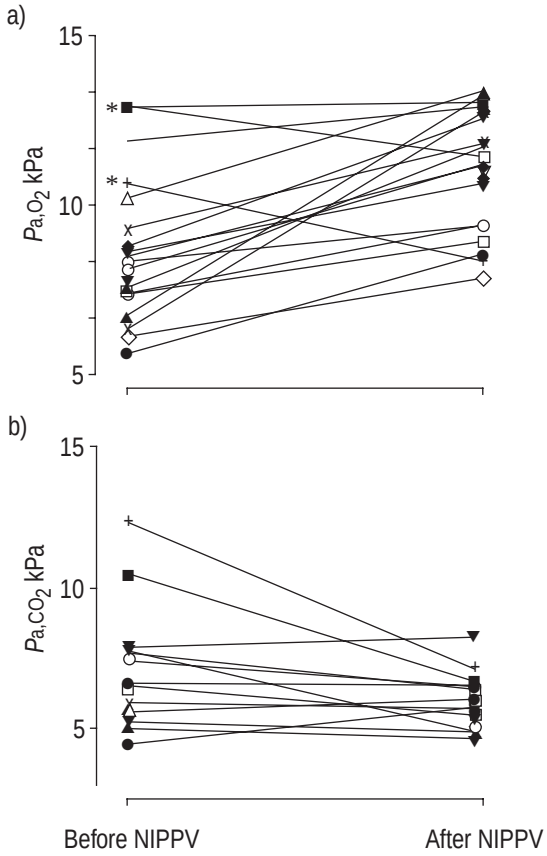

Fig. 1. – a) Arterial oxygen tension (P_{a,O_2}) and b) arterial carbon dioxide tension (P_{a,CO_2}) breathing spontaneously during the day before and after initiation of nocturnal noninvasive positive pressure ventilation (NIPPV). *: measurements obtained with patients using oxygen therapy 2 $L \cdot min^{-1}$. Symbols represent the different children.

Table 1. – Diagnostic categories, mean age at starting noninvasive positive pressure ventilation (NIPPV) and diurnal arterial oxygen (P_{a,O_2}) and carbon dioxide (P_{a,CO_2}) tensions before and after NIPPV by diagnostic categories

Diagnosis	Age yrs	Before NIPPV		After NIPPV	
		P_{a,O_2} kPa	P_{a,CO_2} kPa	P_{a,O_2} kPa	P_{a,CO_2} kPa
CMD	11.6±3.7	8.2±1.2	6.6±1.28	10.65±2.2	5.97±0.9
SMA	5.7±4.2	7.9±1.5	6.1±0.85	11.9±1.1	5.4±1.2
CM	7.3±3.3	8.2±0.6	7.73±1.25	11.3±1.15	6.00±0.9
DMD	13.9±1.5	9.3±2.3	6.33±1.13	12.17±1.46	5.75±0.41
Misc	9.4±4.8	9.4±2.3	7.6±1.37	9.33±1.87	6.33±1.1
Overall		8.5±1.8	7.0±1.6	10.9±1.7	5.9±0.8

Data are presented as mean±SD. CMD: congenital muscular dystrophy; SMA: spinal muscular atrophy; CM: congenital myopathy; DMD: Duchenne muscular dystophy; Misc: miscellaneous; P_{a,O_2}: oxygen tension in arterial blood; P_{a,CO_2}: carbon dioxide tension in arterial blood; NIPPV: noninvasive positive pressure ventilation.

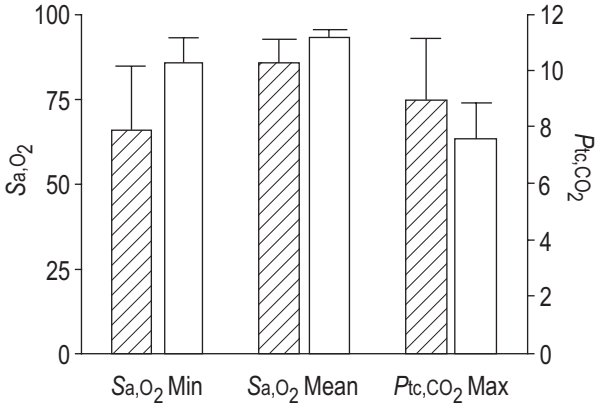

Fig. 2. – Nocturnal arterial blood gas values before (▨) and during (□) noninvasive positive pressure ventilation (NIPPV). MinS_{a,O_2}: minimum arterial oxygen saturation; mean S_{a,O_2}: mean arterial oxygen saturation; max P_{tc,CO_2}: maximum transcutaneous carbon dioxide tension.

months of intermittent ventilator use. Two other children with SMA died at 7 and 26 months respectively, following chest infections. A decision not to institute invasive ventilation was agreed with the family in both cases, as quality of life had deteriorated. Another child with SMA aged 13 yrs used mask ventilation intermittently for 7 months and then transferred to nocturnal oxygen therapy, which provoked worsening nocturnal hypercapnia; the child has recently restarted NIPPV.

Seven children developed nasal bridge sores which were resolved with simple measures. In two patients customized masks were required. For the others, the substitution of the bubblemask (ResMed, Abingdon, UK) or Adams circuit nasal pillows (Puritan Bennett, UK) and/or nasal bridge skin dressings helped the lesion heal. No cases of aspiration were seen in children using facemasks. Four children developed mild-mid facial hypoplasia, which was managed by rotation of the masks on a weekly basis to reduce pressure over the maxillary region. Three of these children had a congenital myopathy associated with facial muscle weakness, and all began NIPPV at ≤8 yrs of age.

Discussion

Ventilatory failure is the most common terminal complication of neuromuscular and chest wall diseases, which affect the respiratory muscles. In many cases a reduction in inspiratory muscle strength is complicated by the development of a thoracic scoliosis, which further reduces respiratory muscle efficiency and increases the work of breathing [16]. Some myopathies and muscular dystrophies are associated with a rigid spine, which reduces chest expansion and can lead to ventilatory insufficiency [17]. Expiratory muscle weakness impairs cough ability, which, when combined with an underlying tendency to atelectasis, predisposes the individual to frequent respiratory tract infections. These, in turn, may precipitate hypercapnic decompensation. Over the last decade it has become evident that diurnal ventilatory failure is preceded by sleep-related abnormalities in arterial blood gas tensions, usually in the form of nocturnal hypoventilation. This first occurs in rapid eye movement (REM) sleep due to the reduction in intercostal muscle tone and central drive, which is most profound during this sleep stage, but later extends into non REM (NREM) sleep and ultimately diurnal respiratory failure results. This progressive sequence provides the rationale for monitoring children at risk of decompensation during sleep and providing ventilatory support at night. REM related S_{a,O_2} desaturation and hypercapnia are most pronounced in children with conditions that are associated with early diaphragm involvement, such as the congenital myopathies, as the reduction in intercostal muscle activity in REM sleep causes maximum compromise of the inspiratory muscles [17]. However, the effect is not as pronounced in children with SMA, in whom diaphragm strength is generally preserved, enabling them to continue to utilize this principal muscle of inspiration during REM sleep.

Symptoms and signs of sleep disordered breathing in children with neuromusculo-skeletal disease can be vague but include failure to thrive, anorexia at breakfast time, daytime fatigue, morning headaches, and cyanosis during meals and on transfer from a wheelchair. Some children develop erratic noisy breathing during sleep. Obstructive apnoeas and hypopnoeas may precede or accompany nocturnal hypoventilation particularly in children with DMD or bulbar weakness [18]. Upper airway obstruction may also be seen in Prader-Willi syndrome, spina bifida and other primary neurological diseases. CPAP may be used initially in these situations when P_{a,CO_2} is normal, but careful observation is required as it may subsequently rise necessitating the use of mask ventilation. Bilevel ventilatory support, which incorporates positive pressure during expiration (EPAP) may be particularly helpful in stabilizing the upper airway in these situations. Cough assist devices (*e.g.* Emerson Insufflator/Exsufflator) may also be helpful, but were not assessed in this series of investigation as they are not currently available in hospitals in the UK.

The average age of the DMD patients in this series is 13.9 yrs (range 12.7–16 yrs). This is younger than the average age for developing ventilatory failure in DMD reported previously, which is ~20 yrs [5]. Five out of seven patients had a diurnal P_{a,CO_2} >6kPa, and only two had symptomatic nocturnal hypoventilation alone. The vital capacity range was 0–700 mL (mean 290 mL). It is likely that these children represent the severe end of the DMD spectrum.

In the past it has been thought that children <10 yrs old might be incapable of using nasal or facemasks successfully, as in one report four out of five younger children could not cope with the equipment [11]. The study presented confirms that mask ventilation can be used in young children and the more favourable results may be attributable to, the extensive range of masks now available, use of pressure preset ventilators, and greater experience with the technique. Side effects such as nasal bridge sores can be minimized with good mask fit and skin dressings. The association of facial mask use and mid facial hypoplasia is the subject of further investigation. Neonatal masks can now be obtained, but careful monitoring is mandatory to ensure that adequate ventilation is achieved. It should be noted that most domiciliary ventilators in current use were not designed for the paediatric age range. The patients in the present study, mainly used pressure preset ventilators, but in other studies volume preset machines have predominated [19], suggesting that both have a role.

The authors believe that overnight P_{a,CO_2} monitoring during NIV is essential to determine ventilator settings. Titration using S_{a,O_2} alone is inadequate and may lead to significant over- or under-ventilation. In addition, if S_{a,O_2} levels are low, it is impossible without CO_2 data to determine whether desaturation is due to underventilation requiring an increase in inspiratory pressure, or that supplemental oxygen is required. Ventilator settings require regular assessment to ensure optimal CO_2 control. Masks should be fitted carefully to minimize deadspace. Using the BiPAP machine an EPAP level of ~4 cmH_2O is necessary to flush deadspace and reduce rebreathing. A plateau expiratory valve (Respironics Inc.) substituted for the standard whisper swivel valve may improve P_{a,CO_2} control. For very young children the PLV 100 (Lifecare, Medic Aid, West Sussex, UK) ventilator was employed as the trigger facility offered favourable characteristics. In general, however, pressure preset ventilators have been used for the children in this series as the machines are smaller and more portable, and the addition of EPAP is often helpful.

Two older children were unable to cope with mask ventilation long-term and this was discontinued. One died after several months without ventilatory support and the other was transferred to nocturnal oxygen therapy, but NIV has recently been recommenced. Both attended hospital for only a short period of acclimatization to mask ventilation (24–48 h) and did not return for follow-up as they attended a local hospital. It is possible that a longer initial period of familiarization with the equipment for the child and family/carers may have reduced this failure rate. Thirty-six children continue to use nasal ventilation and none have developed obtrusive bulbar symptoms. Four children have a feeding gastrostomy. In three cases this was placed to supplement oral intake, and in one child gastrostomy feeding is used exclusively, as swallowing incoordination precludes oral feeding. All school-age children attend mainstream school.

It is important to assess the impact of domiciliary ventilatory support on the family as well as the child as it has been suggested that while the quality of life of children discharged on ventilatory support may improve, the quality of life of their family may deteriorate [20, 21]. This area is being monitored, but as of yet no family has requested discontinuation of NIV.

Mouthpiece NIV has been employed in some paediatric series. Intermittent use for short periods during the day has been advocated to reduce atelectasis and improve chest wall and lung growth [22], but simple mouthpieces cannot be used to deliver overnight ventilation. Ventilatory support during sleep using a mouthpiece retention device has been well-described in adults with neuromuscular disease [23], but these oral interfaces may cause dental maldevelopment in young children. A noninvasive alternative to positive pressure ventilation is negative pressure ventilation using either a tank ventilator, jacket device or cuirass. These devices have been employed successfully in neuromuscular and chest wall disease for many years [24], but are less portable than mask ventilators and may need to be constructed individually, especially in patients with chest wall deformity. Negative pressure devices also tend to be less efficient than positive pressure ventilators, as they may provoke upper airway collapse; and experience in applying negative pressure ventilation is limited to a relatively small number of centres in Europe. The Hayek oscillator (Medicom, London, UK) combines negative pressure with high frequency oscillation and has been used to treat acute ventilatory failure, but is not used widely in the home. In the past, children receiving home ventilation have been treated with tracheostomy intermittent positive pressure ventilation (T-IPPV). Mask ventilation is likely to supersede T-IPPV in those with preserved bulbar function and some degree of spontaneous ventilatory capacity. Not only does NIV facilitate speech development and circumvent tracheostomy-related complications, it can also be readily adapted to the needs of a growing child. Carers and patients report that noninvasive methods are preferable where these are feasible [25]. For practical purposes, therefore, mask ventilation is the noninvasive method of choice and is likely to be applied more widely. However, negative pressure ventilation is still useful in some cases and T-IPPV will remain essential for patients with bulbar insufficiency and extreme ventilator-dependence.

In summary, it is possible to provide mask ventilatory support for children with a variety of neuromuscular and skeletal disorders from <1 yr of age and this intervention reverses ventilatory failure and is likely to extend life expectancy. The optimum time to introduce noninvasive ventilation is not yet clear, but these results would suggest that children most likely to benefit have diurnal hypercapnia or sleep disordered breathing and/or frequent recurrent chest infections. The effects of nasal ventilation on the evolution of muscle weakness, cardiac function, and quality of life need further evaluation.

References

1. Leger P, Bedicam JM, Cornette A, *et al*. Nasal intermittent positive pressure ventilation. Long term follow-up in patients with severe chronic respiratory insufficiency. *Chest* 1994; 105: 100–105.
2. Simonds AK, Elliott MW. Outcome of domiciliary nasal intermittent positive pressure ventilation in restrictive and obstructive disorders. *Thorax* 1995; 50: 604–609.
3. Meecham Jones DJ, Paul EA, Jones PW, Wedzicha JA. Nasal pressure support ventilation plus oxygen compared to oxygen therapy alone in hypercapnic COPD. *Am J Respir Crit Care Med* 1995; 152: 538–544.
4. Bach JR, Ishikawa Y, Kim H. Prevention of pulmonary morbidity for patients with Duchenne muscular dystrophy. *Chest* 1998; 112: 1024–1028.
5. Simonds AK, Ward S, Heather S, Bush A, Rosenthal M, Muntoni F. Outcome of domiciliary nocturnal non-invasive mask ventilation in paediatric neuromuscul-skeletal disease. *Thorax* 1998; 53: A10.
6. Vianello A, Bevilacqua M, Salvador V, Cardaioli C, Vincenti E. Long-term nasal intermittent positive pressure ventilation in advanced Duchenne′s Muscular Dystrophy. *Chest* 1994; 105: 445–448.
7. Rutgers M, Lucassen H, Kesteren RV, Leger P. Respiratory insufficiency and ventilatory support. 39th European Neuromuscular Centre International workshop. *Neuromusc Disord* 1996; 6: 431–435.
8. Barois A, Estournet-Mathiaud B. Ventilatory support at home in children with spinal muscular atrophies (SMA). *Eur Respir Rev* 1992; 10: 319–322.
9. Teague WG. Pediatric application of noninvasive ventilation. *Respir Care* 1997; 42: 414–423.

10. Waters KA, Everett FM, Bruderer JW, Sullivan CE. Obstructive sleep apnea: the use of nasal CPAP in 80 children. *Am J Respir Crit Care Med* 1995; 152: 780–785.
11. Heckmatt JZ, Loh L, Dubowitz V. Night-time nasal ventilation in neuromuscular disease. *Lancet* 1990; 335: 579–582.
12. Kinnear WJM. Assisted ventilation of children at home. *In*: Kinnear WJM, ed. Assisted ventilation at home. A Practical guide, 1994; Oxford, Oxford University Press, pp. 70–75.
13. Fauroux B, Sardet A, Foret D. Home treatment for chronic respiratory failure in children: a prospective study. *Eur Respir J* 1995; 8: 2062–2066.
14. Raphael J-C, Chevret S, Chastang C, Bouvet F. Randomised trial of preventive nasal ventilation in Duchenne muscular dystrophy. *Lancet* 1994; 343: 1600–1604.
15. Simonds AK. Paediatric non-invasive ventilation. *In*: Simonds AK, ed. Non-invasive Respiratory Support, London, Chapman & Hall Medical, 1996; 102–115.
16. Bergofsky EH. Thoracic Deformities. *In*: Charis Roussos, ed. The Thorax Part C: Disease, Second ed. New York, Marcel Dekker, Inc, 1995; pp. 1915–1949.
17. Beckerman RC, Hunt CE. Neuromuscular diseases. *In*: Beckerman RC, Brouillette RT, Hunt CE, eds. Respiratory control disorders in infants and children, Baltimore, Williams & Wilkins, 1992; pp. 251–270.
18. Khan Y, Heckmatt JZ. Obstructive apnoeas in Duchenne muscular dystrophy. *Thorax* 1994; 49: 157–161.
19. Barois A, Estournet-Mathiaud B. Nasal ventilation in congenital myopathies and spinal muscular atrophies. *Eur Respir Rev* 1993; 3: 275–278.
20. Aday LH, Wegener DH, Anderson RM, Aitken MJ. Home care for ventilator-assisted children. *Health Affairs* 1989; 8(2): 137–147.
21. Robins Milller J, Colbert AP, Schock NC. Ventilator use in progressive neuromuscular disease: impact on patients and their families. *Dev Med Child Neurol* 1988; 20: 200–207.
22. Barois A, Estournet-Mathiaud B. Spinal muscular atrophy: respiratory management as a function of clinical aspects. *In*: Robert D, Make BJ, Leger P, Goldberg AL, Paulus J, Willig TN. eds. Home mechanical ventilation, 1 ed. Paris, Arnette Blackwell, 1995; pp. 261–275.
23. Bach JR, Alba AS, Bohatiuk G, Saporito L, Lee M. Mouth intermittent positive pressure ventilation in the management of postpolio respiratory insufficiency. *Chest* 1987; 91: 859–864.
24. Thomson A. The role of negative pressure ventilation. *Arch Dis Child* 1997; 77: 454–458.
25. Bach J. A comparison of long-term ventilatory support alternatives from the perspective of the patient and caregiver. *Chest* 1993; 104: 1702–1706.