Indian J Pediatr (September 2015) 82(9):852–859
DOI 10.1007/s12098-015-1842-z

REVIEW ARTICLE

Home Mechanical Ventilation in Children

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Received: 14 April 2015 / Accepted: 29 June 2015 / Published online: 31 July 2015

Abstract The number of children dependent on home mechanical ventilation has been reported to be increasing in many countries around the world. Home mechanical ventilation has been well accepted as a standard treatment of children with chronic respiratory failure. Some children may need mechanical ventilation as a lifelong therapy. To send mechanically ventilated children back home may be more difficult than adults. However, relatively better outcomes have been demonstrated in children. Children could be safely ventilated at home if they are selected and managed properly. Conditions requiring home ventilation include increased respiratory load from airway or lung pathologies, ventilatory muscle weakness and failure of neurologic control of ventilation. Home mechanical ventilation should be considered when the patient develops progressive respiratory failure or intractable failure to wean mechanical ventilation. Polysomnography or overnight pulse oximetry plus capnometry are used to detect nocturnal hypoventilation in early stage of respiratory failure. Ventilator strategy including non-invasive and invasive approach should be individualized for each patient. The author strongly believes that parents and family members are able to take care of their child at home if they are trained and educated effectively. A good team work with dedicated members is the key factor of success.

Keywords Children · Home ventilation · Long term ventilation · Chronic respiratory failure · Invasive ventilation · Non-invasive ventilation · Tracheostomy

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Introduction

The number of children dependent on home mechanical ventilation has been reported to be increasing in many countries around the world [1–6]. This growing trend could be explained by a variety of reasons. Firstly, advancement in intensive care dramatically improves prognosis of patients with severe illnesses. Much more patients survive and live longer, although some may remain on high technology support. The progression to detect nocturnal hypoventilation by overnight polysomnography, pulse oximetry and capnometry leads to early initiation of ventilator support in children, which beneficially can prevent complications from overt respiratory failure. In children with static or progressive nervous system disorders, ventilator support has been considered as a standard life-long therapy [7]. In addition, the development of noninvasive ventilation has simplified ventilator strategy and broadened choices of ventilators used for chronic respiratory failure. Innovations in respiratory home care equipment with remote alarm features make patients to live at home with more confidence and comfort. Economically, home-based care is far less expensive than in-hospital care when home nursing service is omitted [8, 9]. Socially, home environment has been found to enhance quality of living, unite children with their families and community [10]. Psychologically, most children prefer living at home to staying in the hospital for prolonged periods of time.

Special Considerations in Children

To initiate home mechanical ventilation in children is much more difficult than in adults. Children have smaller airway diameters. Collateral ventilation such as Pore of Khon or Canal of Lambert has not been fully developed in young children. Therefore, airway clearance procedures are essential. The danger of airway blockage with secretions increases with

a decreasing inner diameter of the tracheostomy cannula [11]. Children are at increased risks of accidental removal of cannula and aspirations. Fewer choices of mechanical ventilators are available for children. Small children with poor effort may not be able to trigger the ventilator. So, home ventilators designed for children must be able to provide more sensitive trigger, lower tidal volume and lower flow rate. Commercial masks have a relatively high amount of dead space and often do not fit children well, especially infants [11]. The risk of developing mid-facial hypoplasia is increased in children when using masks with high contact pressure [12]. Some children are too young to help themselves in emergency situations such as ventilator malfunction, electrical power failure *etc*. [11]. So they require well trained care givers who can observe them closely and help them immediately.

Even though there are a lot of difficulties as described above, many studies have shown that properly selected infants and children could be safely ventilated at home [1–3, 5, 6]. As compared to adults with chronic obstructive pulmonary disease (COPD), better outcomes have been reported in children especially those with reversible underlying diseases such as bronchopulmonary dysplasia and tracheobronchomalacia [2, 13]. More than half of the children suffering from severe bronchopulmonary dysplasia were liberated from positive pressure ventilation and decannulated before their sixth birthday [13]. A good quality of life has been also demonstrated in ventilator-dependent children with congenital central hypoventilation syndrome (CCHS), who were able to attend school and function normally in society [14, 15]. Figure 1 shows one of the author's patients with CCHS who has normal growth and development. She can go to primary school like other normal children in the day and has been on mechanical ventilator at night for 6 y. Some patients with spinal cord injury and neuromuscular conditions, who became ventilator-dependent before 6 y of age, could graduate from universities, earned degree and run their own business [16]. Similarly, Fig. 2 shows author's patient with spinal muscular atrophy who has been on non-invasive ventilator for 17 y. At present she supports herself and raises the family by writing novels.

Fig. 1 A Thai girl with congenital central hypoventilation syndrome who has been on a mechanical ventilator *via* tracheostomy during sleep for 6 y (**a**) She can enjoy outdoor activities as normal children (**b**)

Candidates for Home Mechanical Ventilation

Pediatric diseases and conditions causing respiratory failure may be classified according to physiologic abnormalities into three groups as shown in Table 1 [17, 18]. Neuromuscular disorders have been listed in the majority of reports as the first or second most common diagnostic categories in pediatric population [2, 19–22]. It should be noted that a number of children have had multiple problems and disabilities such as cardiac diseases, seizure disorders, endocrine disorders and so on. These co-morbidities make their care even more complex.

Long-term mechanical ventilation is indicated either when the patients are not be able to wean from mechanical ventilation or demonstrate evidence of progressive respiratory failure with or without pulmonary hypertension. Since minute ventilation is relatively diminished during sleep as compared to awake state, detection of hypoventilation and hypoxemia during sleep, established by either polysomnography or overnight pulse oximetry plus capnometry, is considered as the earliest signs of respiratory failure in these candidates [23]. Nocturnal ventilatory support should be initiated. Waiting until the patients become hypercapnic during the day may result in uncontrolled decompensation [24].

Children who are suitable to be sent home with mechanical ventilation must fulfill the following criteria:

1. Medical stability without frequent monitoring, test or treatment changes [25]. Unchanged ventilator settings.

Fig. 2 One of the patients with spinal muscular atrophy who has been on a non-invasive ventilators for 17 y (**a**) She supports herself and raises the family by writing novels (**b**)

2. Positive trend in weight gain and growth curve [26].
3. Freedom from frequent infection and fever [26].
4. Motivated family with willingness and commitment to provide support and care at home [25].
5. Adequate and adaptable home setting [25].
6. Accessible financial resources and health care support in the community [25].

Table 1 Candidates for pediatric home mechanical ventilation as classified by physiologic abnormalities

Increased respiratory load
Obstructive disorders
Obstructive sleep apnea
Tracheobronchomalacia
Bronchiectasis
Bronchiolitis obliterans
Restrictive disorders
Kyphoscoliosis
Chest wall deformities
Lung hypoplasia
Interstitial lung diseases
Combined obstructive and restrictive disorders
Bronchopulmonary dysplasia
Heart failure
Ventilatory muscle weakness or pumping failure
Spinal muscular atrophy
Myasthenia gravis
Spinal cord injury
Diaphragmatic dysfunction
Motor neuron disease
Congenital muscular dystrophy
Failure of neurologic control of ventilation
Congenital central hypoventilation syndrome (CCHS)
Post encephalitis
Brain stem tumor
Brain infarct, trauma or degeneration

Adapted from references [17, 18]

Ventilators Designed for Home Use

The goal of home mechanical ventilation is to correct respiratory failure and maximize oxygenation and ventilation in order to allow children to reach their developmental potential [27]. Long-term ventilators designed for home use must be small, lightweight, portable and durable. It must have an internal air compressor, which can be run on electricity. It should also be powered by the internal battery in case of inaccessible electricity. Home ventilators must offer adequate audible alarms to alert caregivers in time to check the child and the ventilator.

Home mechanical ventilators are classified into invasive and non-invasive mechanical ventilators. Advantages of non-invasive ventilation relate to the fact that no tracheostomy is required, which reduces complications and discomfort related to tracheostomy. Caregivers do not need to take care of tracheostomy, which is a heavy burden. In general, a requirement for only nocturnal support warrants non-invasive ventilation, while a requirement for 24 h support often warrants invasive ventilation [18]. The decision to choose whether

invasive or non-invasive ventilation for children needing more than nocturnal but less than 24 h support must be individualized [18]. Due to a lot of advantages, non-invasive ventilation should be considered first for children with chronic respiratory insufficiencies [28]. Invasive ventilation is used only when the children show evidence of either failure to adequately ventilate with non-invasive ventilation, failure to tolerate masks, bulbar dysfunction with a high risk of ongoing aspiration and ineffective non-invasive management of secretions or high level of dependence on assisted ventilation (>16 h/d) [27–29].

Modes of Mechanical Ventilation

Non-invasive Mechanical Ventilation

Non-invasive ventilators generate high flow to compensate leak around masks in order to reach the pre-set pressure [30]. There is a single-tube circuit with passive exhalation port incorporated in the circuit near the patient or into the interface [30, 31]. Patient's acceptance of the mask is the most important indicator of success. It should ensure comfort and fit while minimizing leak [31]. A poorly fitting mask interface will decrease the clinical effectiveness of treatment through leak, and may impact on adherence. Excessive leak can impact on sleep quality, patient-ventilator synchrony and the amount of effective ventilation delivered to the patient [32].

Basically, there are two modes; continuous positive airway pressure (CPAP) and bi-level positive pressure (BPAP). CPAP is commonly used in children with obstructive sleep apnea to distend upper airway and relieve airway obstruction. BPAP provides tidal breaths for patients with alveolar hypoventilation. The pressure difference between inspiratory and expiratory positive airway pressure in pressure support mode determines the amount of tidal volume delivered to the patient. BPAP can be set in the spontaneous mode (the patient cycles the device), the spontaneous timed mode (a back-up rate and inspiratory time can be set in case the patient does not trigger the device) and the timed mode (inspiratory time and respiratory rate are fixed) [33].

Modern non-invasive ventilators additionally offer more modalities including average volume-assured pressure support (AVAPS) that automatically adjusts the pressure support level in order to provide a consistent desired tidal volume; auto-adjusting positive airway pressure (APAP) that automatically adjusts the levels of continuous positive airway pressure in order to relieve airway obstruction in obstructive sleep apnea; and C-flex or Bi-flex that soften airflow at only upon exhalation or both exhalation and inhalation respectively to make breathing more comfortable. Each device has data storage in a sim card so that usage information can be downloaded such as hours of usage patterns and leakage which is helpful to assess adherence of the patient and adjust mask fitting.

Invasive Mechanical Ventilation

There have been a number of modern home ventilators available in markets, which offer different modes of setting including volume-targeted, pressure-targeted, in assisted controlled or controlled mode as well as pressure support. The choice of mode should be individualized depending on patient's comfort and underlying conditions. Alarms should be set up appropriately for patients' safety.

The settings of both non-invasive and invasive ventilators are first established by bedside titration and monitoring. Patients are allowed to try to use each ventilator for a short time and select the ventilator according to their preference. It is most helpful to adjust the settings under polysomnography, by which leakage can be detected and inspiratory and expiratory pressure, back-up rate, inspiratory time, trigger sensentivity can be precisely adjusted. The goal is to achieve adequate ventilation and oxygenation, good quality of sleep with minimal arousals [33]. The leakage around masks can be fixed by selecting the right size together with the right type of mask and headgear. If a child mouth-breathes a lot, a chin strap may be needed to keep the mouth closed. Full face masks may be used to minimize leak in some patients [34]. The leakage around the tracheostomy tube can be corrected by using cuffed tracheostomy tube and inflating cuff with appropriate volume.

If the child is optimally ventilated either by a non-invasive or invasive ventilator and normal $PaCO_2$ is achieved, but there is still hypoxemia, then long-term oxygen therapy should be given in addition to ventilatory support. The oxygen requirement should be assessed by pulse oximetry monitoring for at least 6–12 h during all levels of activity, including sleep and feeding [35]. The goal is to maintain oxygen saturation above 92–93 % [35, 36]. When oxygen gas is administered through the ventilator circuit, CO_2 level and respiratory rate should be monitored closely to ensure that any hypercapnia or hypopnea is not exacerbated by the removal of the hypoxic drive [35]. Oxygen alone should not be used to correct hypoxemia caused by alveolar hypoventilation in patients with neuromuscular weakness since it will mask cyanosis, worsen hypercapnia and delay initiation of ventilatory support [29].

Various airway clearance techniques are routinely recommended to improve mucociliary clearance, prevent or treat atelectasis, minimize chances of acquired respiratory infection. For tenacious airway secretions, in addition to increasing fluid intake, author uses heated humidification system, nebulised normal saline or 3 % hypertonic saline [29]. Aerosolized bronchodilators, either by nebulization or metered dose inhaler *via* spacer, are given for those patients who have

bronchial hyperresponsiveness. Standard chest physiotherapy consisting of postural drainage, manual chest percussion and vibrations is carried out to mobilize secretions into the central airways. For a tracheostomized patient who has weak cough, author recommends the caregiver to give a positive pressure ventilation by a resuscitation bag and deep suctioning through a tracheostomy tube. For a patient who uses a non-invasive ventilator, author recommends to support deep breathing by using his or her own non-invasive ventilator intermittently during airway clearance. Several manual augmented coughing options and respiratory muscle training are tailored and advised to each child depending on his or her age, cooperation and underlying diseases. Although sophisticated devices such as mechanical insufflation/exsufflation [37] and high frequency chest wall oscillation [38] have been shown to have a lot of advantages over conventional airway clearance techniques, they rarely are administered in our home setting due to the lack of funding resources.

If the underlying diseases are getting better, there is no need to change the ventilator setting. The patient can be weaned from the ventilator by increasing free time so that they can independently participate in more daily activities. Free time can be increased gradually in daytime first. If the patient improves, the ventilator can be weaned to only nighttime. Overnight monitoring with pulse oximetry and capnometry or polysomnography are performed to confirm whether or not the patient can be taken off nighttime mechanical ventilation.

Discharge Planning

Because of the shortage of home health care nurses as well as complexity of children's conditions and technological supports, it is imperative that the children's family and caregivers must be involved and capable of learning all aspects of home care [39]. Caregivers need to be trained in using, cleaning and caring for the ventilators and accessories; basic trouble shooting skills; emergency problem solving; understanding alarms and corrective measures to take; tracheostomy care as well as basic life support [40]. In addition, they need to be trained to competently manage airway secretions either by suctioning through a tracheostomy, or by using manual and/or mechanical cough assist techniques in the patients with neuromuscular weakness being on noninvasive ventilation. Their skills need to be reassessed periodically [40]. Assuming the responsibility for the care and management of ventilator dependent patients is a life-altering event for most families and caregivers. The parental responsibility was found to be highly stressful and sometimes overwhelming [41]. However, the success in training the parents and family members as caregivers has been reported in many studies [2, 42–44].

In Thailand, where the cost of home care is directly charged to the family, it is impossible to have health care professionals as caregivers at home. The author strongly believes that in order to assist complicated children to live at home, the most suitable persons to be trained as caregivers are the ones who really love them. Among 53 children sent home with invasive home mechanical ventilation, 35 (66 %) of primary caregivers were parents, 9 (17 %) were relatives, 9 (17 %) were nursery maids. All of them were non-medical professionals [2]. One advantage in Thai society is the extended family setting in which two or three generations live together in the same house. Either parents or family members have been selected and trained to be a caregiver [2]. Experience for more than 15 y has taught the author that the level of the caregiver's education is not as important as dedication. Commitment, motivation and preparation from patients' families and caregivers are crucial for a successful transition to home [2].

The author's home care team consists of pediatric pulmonologists, respiratory nurses, home visiting nurses and equipment organizers [2]. As professionals of respiratory therapists do not exist in Thailand, respiratory nurses take on their role to train lay people to be able to take care of their mechanically ventilated children at home. They also work as nurse coordinators, discharge planners, social workers and so on. Teaching and training the care of ventilated children is a gradual process that requires practice and repetition. Initially it is very stressful due to the huge learning curve required to care for the child and to maintain the equipment safely. More time and experience subsequently reduces their stress. Once competence is gained, the parents are able to advocate for their child to get appropriate treatment. They are also trained to know their limits, so that they seek help when necessary. The family can call respiratory nurses for help 24 h a day whenever any problems occur. When the children go home, the home visiting nurses go to see their home at the beginning, may be once a week, to supervise the caregivers and coordinate with the hospital team [2]. The distributors of home ventilators are responsible to visit patients' home and check ventilator function periodically. This model approach may be practical in other countries with low income and limited resources.

Obstacles to Establish Home Mechanical Ventilation

The levels of financial support for at-home care for children on long-term ventilator support are highly different across the world. In high income countries, home health care service are covered by either the government or medical health insurance, while in lower income countries, patients and families are responsible for all expenses that may occur [2]. The cost of equipment, supplies, and caregivers must be met by the family themselves. A number of patients whose conditions are

suitable for home care still have to undergo long-term hospitalization unnecessarily because of the lack of family resources. The feasibility of home care depends very much on the family's resources and their ability to cope with ensuing problems [2]. BPAP and CPAP *via* tracheostomy have been using instead of standard home ventilator to cut down the expense [2]. Fortunately some children received ventilators from donations. In case of mechanical ventilator failure or electrical power failure or outage, patients need to go to nearby hospitals as soon as possible since they are provided only one set of each device.

In order to effectively run home care program in this difficult situation, the team has to face a lot of barriers including designating family members as caregivers, seeking for funding for home care equipment, teaching and training family members who mostly are lay persons for all aspects of sophisticated home care, emotional and social support to the family, preparing safe home environment, coordinating with other specialties involved in the care of the child such as nutritionists, neurologists, speech therapists, occupational therapists, social workers, physical therapists and schools. A checklist of caregiver's evaluation form should be completely filled-in before discharge (Appendix). All tasks require a good team work; dedicated members of the team are the key factor.

Conclusions

Home mechanical ventilation has been well accepted as a standard treatment of children with chronic respiratory failure. Although it is more difficult in children than adults, better outcomes have been reported in children. Children can be safely ventilated at home if they are selected and managed properly. Home mechanical ventilation is classified as non-invasive and invasive ventilation. The ventilator strategy should be individualized. Parents and family members are able to take care of their child at home if they were trained and educated effectively. A good team work with dedicated members is the key factor of success.

Acknowledgments The author wishes to thank Varangkana Lamsam and family for their unconditioned continuous funding support to the underprivileged children and Jeeraporn Pongsasanongkul, Jongrak Utrarachkit, Thitida Chaisupamongkollarp, Anchalee Leejakapai, Dr. Harutai Kamalaporn, Dr. Teeradej Kuptanon, Malinee Nugboon, Porntip Tachanives, Jarinya Prempreungves, Wannapa Yensirikul and Kesanee Chaicoming, whose dedication and extraordinary effort has enabled the complicated patients to live happily at home.

Conflict of Interest None.

Source of Funding None.

Appendix Checklist of caregiver's evaluation before discharge at Ramathibodi hospital

Topics	Yes/No
1. General	
- Understand underlying disease	
- Signs and symptoms must see MD.	
- Nutrition/ Milk / Feedings	
- Aerosol therapy	
- Chest physiotherapy	
- Airway clearance	
- Development/ Immunizations	
- Emergency management & CPR	
- Emergency call number	
- Letter (summary of medical history)	
- Follow-up appointments	
2. Oxygen therapy	
- Understand why it is necessary	
- Type of O_2 source	
- Care/ Maintenance of equipment	
- Flow rate of O_2	
- Warning signs	
- Complications of O_2 therapy	
3. Tracheostomy care	
- Understand why it is necessary	
- Safe activities of daily living	
- Communication/ Speech	
- Cleaning & sterilization of all supplies and equipment	
- Suction techniques: how and when?	
- Self-inflating bag	
- Suction machine & catheters	
- Trach tube & tie changing	
- Accidental decannulation	
- Safety & home environment	
- Transportation	
- Wean off trach: when and how?	
4. Ventilator care	
- How ventilator works? Complications	
- Check ventilator & heated humidifier	
- Mask fitting and interface	
- Problem solving of ventilator malfunction	
- Cleaning & sterilization of all supplies and equipment	
- Safety & home environment	
- Battery / power supply	
- Wean off ventilator	

References

1. Amin R, Sayal P, Syed F, Chaves A, Moraes TJ, MacLusky I. Pediatric long-term home mechanical ventilation: twenty years of

follow-up from one Canadian center. Pediatr Pulmonol. 2014;49: 816–24.
2. Preutthipan A, Nugboon M, Chaisupamongkollarp T, Kuptanon T, Kamalaporn H, Leejakpai A. An economic approach for children with chronic ventilation support. Curr Pediatr Rep. 2014;2:1–8.
3. Edwards EA, Nixon GM; Australasian Paediatric Respiratory Group Working Party on Home Ventilation. Paediatric home ventilatory support: changing milieu, proactive solutions. J Paediatr Child Health. 2013;49:13–8.
4. Com G, Kuo DZ, Bauer ML, et al. Outcomes of children treated with tracheostomy and positive-pressure ventilation at home. Clin Pediatr (Phila). 2013;52:54–61.
5. Paulides FM, Plotz FB, LP V-v d O, van Gestel JP, Kampelmacher MJ. Thirty years of home mechanical ventilation in children: escalating need for pediatric intensive care beds. Intensive Care Med. 2012;38:847–52.
6. Fauroux B, Boffa C, Desguerre I, Estournet B, Trang H. Long-term noninvasive mechanical ventilation for children at home: a national survey. Pediatr Pulmonol. 2003;35:119–25.
7. Weese-Mayer DE, Berry-Kravis EM, Ceccherini I, et al. An official ATS clinical policy statement: congenital central hypoventilation syndrome: genetic basis, diagnosis, and management. Am J Respir Crit Care Med. 2010;181:626–44.
8. Noyes J, Godfrey C, Beecham J. Resource use and service costs for ventilator-dependent children and young people in the UK. Health Soc Care Community. 2006;14:508–22.
9. Bach JR, Intintola P, Alba AS, Holland IE. The ventilator-assisted individual. Cost analysis of institutionalization *vs* rehabilitation and in-home management. Chest. 1992;101:26–30.
10. Boroughs DS, Dougherty J. A multidisciplinary approach to the care of the ventilator-dependent child at home: a case study. Home Healthc Nurse. 2010;28:24–8. quiz 8–30.
11. Windisch W, Walterspacher S, Siemon K, Geiseler J, Sitter H; German Society for Pneuomology. Guidelines for non-invasive and invasive mechanical ventilation for treatment of chronic respiratory failure. Published by the German Society for Pneumology (DGP). Pneumologie. 2010;64:640–52.
12. Li KK, Riley RW, Guilleminault C. An unreported risk in the use of home nasal continuous positive airway pressure and home nasal ventilation in children: mid-face hypoplasia. Chest. 2000;117: 916–8.
13. Cristea AI, Carroll AE, Davis SD, Swigonski NL, Ackerman VL. Outcomes of children with severe bronchopulmonary dysplasia who were ventilator dependent at home. Pediatrics. 2013;132: e727–34.
14. Liess BD, Dost JS, Templer JW, Tobias JD. Congenital central alveolar hypoventilation syndrome (Ondine's curse) with survival into adulthood. Clin Pediatr (Phila). 2008;47:941–6.
15. Healy F, Marcus CL. Congenital central hypoventilation syndrome in children. Paediatr Respir Rev. 2011;12:253–63.
16. Gilgoff RL, Gilgoff IS. Long-term follow-up of home mechanical ventilation in young children with spinal cord injury and neuromuscular conditions. J Pediatr. 2003;142:476–80.
17. Make BJ, Hill NS, Goldberg AI, et al. Mechanical ventilation beyond the intensive care unit. Report of a consensus conference of the American College of Chest Physicians. Chest. 1998;113: 289S–344.
18. King AC. Long-term home mechanical ventilation in the United States. Respir Care. 2012;57:921–30. discussion 30–2.
19. Racca F, Berta G, Sequi M, et al. Long-term home ventilation of children in Italy: a national survey. Pediatr Pulmonol. 2011;46: 566–72.
20. Hsia SH, Lin JJ, Huang IA, Wu CT. Outcome of long-term mechanical ventilation support in children. Pediatr Neonatol. 2012;53: 304–8.
21. McDougall CM, Adderley RJ, Wensley DF, Seear MD. Long-term ventilation in children: longitudinal trends and outcomes. Arch Dis Child. 2013;98:660–5.
22. Goodwin S, Smith H, Langton Hewer S, et al. Increasing prevalence of domiciliary ventilation: changes in service demand and provision in the South West of the UK. Eur J Pediatr. 2011;170: 1187–92.
23. Simonds AK. Recent advances in respiratory care for neuromuscular disease. Chest. 2006;130:1879–86.
24. Simonds AK. Home ventilation. Eur Respir J Suppl. 2003;47: 38s–46.
25. McKim DA, Road J, Avendano M, et al. Home mechanical ventilation: a Canadian Thoracic Society clinical practice guideline. Can Respir J. 2011;18:197–215.
26. Eigen H, Zander J. Home mechanical ventilation of pediatric patients. American Thoracic Society. Am Rev Respir Dis. 1990;141:258–9.
27. Amin RS, Fitton CM. Tracheostomy and home ventilation in children. Semin Neonatol. 2003;8:127–35.
28. Nixon GM, Edwards EA, Cooper D, et al. Ventilatory support at home for children. a consensus statement from the Australasian Paediatric Respiratory Group. February 2008. APRG Working. Available at http://www.thoracicorgau/documents/papers/aprghomeventilationguideline.pdf. Accessed 31 Mar 2015.
29. Hull J, Aniapravan R, Chan E, et al. British Thoracic Society guideline for respiratory management of children with neuromuscular weakness. Thorax. 2012;67:i1–40.
30. Boldrini R, Fasano L, Nava S. Noninvasive mechanical ventilation. Curr Opin Crit Care. 2012;18:48–53.
31. Pavone M, Verrillo E, Caldarelli V, Ullmann N, Cutrera R. Noninvasive positive pressure ventilation in children. Early Hum Dev. 2013;89:S25–31.
32. Teschler H, Stampa J, Ragette R, Konietzko N, Berthon-Jones M. Effect of mouth leak on effectiveness of nasal bilevel ventilatory assistance and sleep architecture. Eur Respir J. 1999;14: 1251–7.
33. Berry RB, Chediak A, Brown LK, et al. Best clinical practices for the sleep center adjustment of noninvasive positive pressure ventilation (NPPV) in stable chronic alveolar hypoventilation syndromes. J Clin Sleep Med. 2010;6:491–509.
34. Willson GN, Piper AJ, Norman M, et al. Nasal versus full face mask for noninvasive ventilation in chronic respiratory failure. Eur Respir J. 2004;23:605–9.
35. Balfour-Lynn IM, Field DJ, Gringras P, et al. BTS guidelines for home oxygen in children. Thorax. 2009;64:ii1–26.
36. Chye JK, Gray PH. Rehospitalization and growth of infants with bronchopulmonary dysplasia: a matched control study. J Paediatr Child Health. 1995;31:105–11.
37. Chatwin M, Bush A, Simonds AK. Outcome of goal-directed noninvasive ventilation and mechanical insufflation/exsufflation in spinal muscular atrophy type I. Arch Dis Child. 2011;96:426–32.
38. Keating JM, Collins N, Bush A, Chatwin M. High-frequency chest-wall oscillation in a noninvasive-ventilation-dependent patient with type 1 spinal muscular atrophy. Respir Care. 2011;56:1840–3.
39. Dumas HM. Rehabilitation considerations for children dependent on long-term mechanical ventilation. ISRN Rehabil. 2012;2012:15.
40. Piper A. Discharge planning and management for patients with chronic respiratory failure using home mechanical ventilation. Breathe. 2010;6:322–33.
41. Kirk S, Glendinning C, Callery P. Parent or nurse? The experience of being the parent of a technology-dependent child. J Adv Nurs. 2005;51:456–64.
42. Tearl DK, Hertzog JH. Home discharge of technology-dependent children: evaluation of a respiratory-therapist driven family education program. Respir Care. 2007;52:171–6.

43. Mah JK, Thannhauser JE, McNeil DA, Dewey D. Being the lifeline: the parent experience of caring for a child with neuromuscular disease on home mechanical ventilation. Neuromuscul Disord. 2008;18:983–8.

44. Mah JK, Thannhauser JE, Kolski H, Dewey D. Parental stress and quality of life in children with neuromuscular disease. Pediatr Neurol. 2008;39:102–7.