The Thoracic Society of Australia and New Zealand

Ventilatory Support at Home for Children. A Consensus Statement from the Australasian Paediatric Respiratory Group

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APRG Working Party on Home Ventilation

GM Nixon [Chair]	Melbourne Children's Sleep Unit, Monash Medical Centre, Melbourne, Australia
EA Edwards	Starship Children's Hospital, Auckland, New Zealand
DM Cooper	Mater Children's Hospital, Brisbane, Australia
DA Fitzgerald	The Children's Hospital at Westmead, Sydney, Australia
M Harris	Mater Children's Hospital, Brisbane, Australia
J Martin	Women's and Children's Hospital, Adelaide, Australia
RJ Massie	Royal Children's Hospital, Melbourne, Australia
S Suresh	Mater Children's Hospital, Brisbane, Australia
KA Waters	The Children's Hospital at Westmead, Sydney, Australia
A Wilson	Princess Margaret Hospital for Children, Perth, Australia

Corresponding author:
Dr Gillian M. Nixon
Department of Respiratory and Sleep Medicine
Monash Medical Centre
246 Clayton Road
Clayton Victoria 3168 AUSTRALIA
Phone: +61 3 95942900
Fax: +61 3 95946270
Email: gillian.nixon@southernhealth.org.au

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GOALS OF DOCUMENT

- To outline an Australian and New Zealand consensus opinion regarding optimal care of children on respiratory support at home, both non-invasive and invasive.
- To make recommendations that support the development of improved services for Australian and New Zealand children who require long term respiratory support.
- To develop standards of care for respiratory support at home within paediatric respiratory and intensive care communities in Australia and New Zealand.
- To highlight issues relevant to providers of equipment related to respiratory support at home for children.
- To highlight areas of research need.

This document represents consensus opinion of paediatric respiratory specialists on aspects of the management of children on ventilatory support at home. It is not intended as a comprehensive 'standards of care' document.

DEFINITIONS

- **Ventilatory support** - assistance to respiration provided by mechanical means. For the purposes of this document, this includes continuous positive airway pressure (CPAP) and artificial ventilation delivered by any means, but excludes supplemental oxygen alone[1]. This guideline does not discuss the care of a child with a tracheostomy alone; for this, the reader is referred to the American Thoracic Society Guidelines[2].
- **Ventilator dependency** - requirement of a mechanical aid for ventilation for any period of the day or night in order to maintain optimal health.
- **Long-term ventilation** - failure to wean from ventilatory support once medically stable, three months after the institution of ventilation[3].
- **Invasive ventilation** - ventilatory support delivered via an endotracheal tube or a tracheostomy.
- **Non-invasive ventilation (NIV)** - ventilatory support delivered via a nasal mask or similar non-invasive interface.
- **Home** - may be the child’s home, a foster home or a group living environment.

LIST OF ABBREVIATIONS

BPAP	bi-level positive airway pressure
CPAP	continuous positive airway pressure
NIV	non-invasive ventilation
VS	Ventilatory Support
DMD	Duchenne Muscular Dystrophy
LTV	Long term ventilation
OSA	Obstructive sleep apnoea
APRG	Australasian Paediatric Respiratory Group
TSANZ	Thoracic Society of Australia and New Zealand

SUMMARY OF RECOMMENDATIONS

The number of children who receive long term ventilatory support is increasing. Long term hospitalisation is inappropriate for the developing child. Care of children requiring long term ventilatory support is feasible and should occur out of hospital ("home", see definitions). Dedicated government funding is needed for children who require long term respiratory support in the home. The APRG and TSANZ support the equitable access to diagnostic and therapeutic options for children who require ventilatory support. The APRG and TSANZ support the care of technology-dependent children in their own home.

Children dependent on ventilation

Eligibility for home ventilation via tracheostomy depends on stable medical condition and ventilatory requirements for four weeks. Hospitalisation is frequently delayed in children on home ventilation via tracheostomy, and funding channels for home care should be clear, so that care packages can be readily put together to facilitate optimal timing of discharge. Funding for home ventilation should be equitable, optimal and accessible across Australia and New Zealand. Discharge should be a staged process, ensuring high quality of care and parental and carer confidence. Discharge planning must provide families with the education, skills and resources to care for their child. A case manager should be identified for each child. Each centre should have one or two preferred ventilator models, so that workers are familiar with the capabilities, limitations, and operation of those ventilators. Minimum standards for ventilatory equipment should be universally applied across Australia and New Zealand. Children on tracheostomy ventilation require awake overnight carers who are trained and credentialed in all aspects of the child's care. A formal accreditation of achievement of a satisfactory standard of training should be part of the contractual agreement between the hospital and the home care organisation. A risk minimisation strategy and action plan should be formally documented for each child.

Non-invasive ventilation is an alternative to tracheostomy ventilation in children who are predominantly dependent on night-time support. Levels of funding, supervision, training and equipment are comparable to those required for children dependent on tracheostomy ventilation.

Ventilatory support for optimisation of a child's health

Access to paediatric diagnostic services should be available to all children who would potentially benefit from non-invasive ventilation (NIV) or continuous positive airway pressure (CPAP). NIV and CPAP should be available and accessible to all children whose health could be optimised with such therapy. NIV and CPAP should be instituted and monitored by multidisciplinary paediatric teams experienced in such treatment for children, and ventilatory settings and equipment should be individualised for each child. Equipment for CPAP and non-invasive ventilation should be funded by designated government authorities and equitable across regions, with allowance within the funding packages for changing needs over time.

Issues common to all children on ventilatory support

The frequency of medical review will be determined by the individual clinical situation, but should be at least every three months for children on invasive ventilation and three to six monthly for those on NIV or CPAP. Regardless of the child's residence appropriate funding is required for review to occur at a tertiary centre with access to lung function testing facilities and paediatric sleep medicine services including experience in NIV and CPAP. Follow-up sleep studies are recommended for all children on respiratory support. Tertiary paediatric centres require a coordinator for home ventilatory support patients. Home ventilation equipment should be checked/serviced as recommended by the manufacturer. Transition to a local adult service should be arranged for adolescents, having ascertained that the service is appropriate for adolescents with complex health needs. Funding for home ventilation should be maintained through the transition process.

Increasing demand for home ventilatory support should be matched with appropriate funding, so that specialised diagnostic and treatment services can be maintained appropriate to the needs of this patient group. The authors recommend the establishment of a multi-centre working group and a patient registry to inform individual patient care and policy development.

INTRODUCTION

Advances in neonatal and paediatric care have reduced mortality, but have introduced a new morbidity: a growing number of children who are medically stable but require respiratory support for all or part of every day[4-6]. This growth has been perpetuated by a change in society's expectations of long term disability, the pressure to reduce duration of hospital stay, and recognition that children with a broad range of medical conditions may benefit from long-term respiratory support. Improvements in equipment design and technical support have also broadened the group of children for whom respiratory support at home is a possibility. Increasingly, children who would previously have died or been managed in hospital with respiratory support via a tracheostomy are being managed with less invasive forms of respiratory support (non-invasive ventilation [NIV] or continuous positive airway pressure [CPAP] via a mask). Requirements for transfer to care at home differ for children on these various types of support. A key principle of care is that long-term hospitalisation is inappropriate for a developing child[7], puts them at risk of hospital acquired infection, is expensive, is detrimental to family functioning and relationships, is an unnecessary use of resources and may block intensive care or medical specialty beds for other ill children.

In the past ten years there have been exponential increases in the numbers of children on NIV and CPAP [5, 6], whereas the numbers of children supported with ventilation via tracheostomy has remained relatively static. There is a further large group of children whose need for ventilatory support is unrecognised and untreated due to the lack of diagnostic and treatment facilities (European data suggests 6.6 per 100,000 require ventilatory support [VS])[8]. The provision of equipment and expertise to care for these children is outstripped by the rapidly increasing number of children on NIV and CPAP.

In addition to the myriad of technological requirements for children on assisted ventilation, the care of such children must meet all the general standards of paediatric health care. A child's developmental and education needs should have a high priority, and health care should be delivered in a way which promotes participation in family life and education, thereby maximising the child or young person's potential. Maintaining their role and relationships within their family is paramount, and can only be achieved if the child is cared for at home. Children with high technological dependency and complex medical needs place a considerable additional burden on any parent or caregiver. Planning for care at home must address appropriate respite and psychological support for carers and other family members, e.g. siblings.

While home-based care is often feasible[9] and may be less expensive than in-hospital care, the increasing demand for care at home has highlighted funding issues for the care of children with significant medical and technological needs in their own home. Once a child requiring long-term ventilation is established at home, reviews have demonstrated that non-elective readmission to hospital is unusual[10-12]. There is emerging but limited information regarding the long-term outcomes of children who are maintained at home on ventilation via a tracheostomy, and children using nocturnal non-invasive ventilation or CPAP[6, 13-18]. Further information on outcomes for both the child and family is required, both to inform funding and care decisions, and also for the development of standard of care guidelines.

Key points

1. The number of children who receive long term ventilatory support is increasing.
2. Long term hospitalisation is inappropriate for the developing child.
3. Home care of children requiring long term ventilatory support is feasible and should be a priority of the health care team.
4. Dedicated government funding is needed for children who require long term respiratory support in the home.

The purpose of this document

The purpose of this document is to discuss the principles of care of a child on long term ventilatory support. We aim to outline existing services for such children in Australia and New Zealand and to highlight appropriate levels of service provision in comparison to what is currently provided.

This document is divided into sections covering the different types of ventilatory support, with a section on common issues. Readers should bear in mind that practical details regarding the provision of ventilatory support at home depends on the severity of the underlying condition and presence of co-morbidities, the route of ventilation, and the number of hours of ventilation required per day, and that these factors may change with time.

The structure of this document

Part 1:

Part 1 relates to children who are dependent on ventilation for life support. Ventilatory support may either be via:

A]: Tracheostomy: the child who requires ventilation via tracheostomy is an obligatory ventilator dependent patient. The care pathways and risk management are unique to this group.

B]: Non-invasive ventilation: this is defined for the purposes of this document as those in whom ventilation cannot be discontinued for any period during sleep without risk to the child's life.

Part 2:

Part 2 discusses ventilatory support for children in whom CPAP and NIV are appropriate health strategies for health maintenance and illness prevention, including reductions in mortality, morbidity, frequency, duration and cost of hospital admissions, and improvements in quality of life[19-27].

Part 3:

Part 3 covers issues common to all patients receiving mechanical support to respiration at home. The reader should bear in mind that the other medical conditions of the child (e.g. co-morbid developmental delay or feeding issues) will also affect the support needs of their family at home and should be considered in the overall care package for the child.

ESTABLISHMENT OF CONSENSUS

In response to an initiative of several members of the Australasian Paediatric Respiratory Group (APRG), a core working group was formed at the meeting of the APRG in August 2005. Members of that group represented paediatric respiratory physicians with experience in home ventilation from around Australasia. The initial draft of this document was written and circulated by email to all members of the APRG prior to the next meeting in August 2006. A workshop was then held at that meeting, when all APRG members had the opportunity to discuss the draft. A subset of the core working group also met at the Australasian Sleep Association meeting in October 2006. Further adaptations to the manuscript were made by the core working group (named as authors of this document) following these meetings and a teleconference. The working group met at the APRG meeting in August 2007, and key areas of the paper were also presented to the entire APRG group for discussion at that time. The final version of the document was submitted to the Thoracic Society of Australia and New Zealand for review and was endorsed by its Executive Committee in February 2008.

Assessment of evidence

A critical review of the relevant literature was performed for the formation of this document and its recommendations. The literature in this area is of small cohort studies or case series. Very few randomised controlled studies exist. This statement uses expert consensus in the majority of its recommendations, as randomised controlled trials are largely not ethical or clinically feasible within this patient population.

PART 1: CHILDREN DEPENDENT ON VENTILATION

A] VIA TRACHEOSTOMY

Introduction

Ventilatory support is usually delivered via tracheostomy when there is either a need for prolonged hours on ventilatory support each day or if a child has an abnormal upper airway. Usually, different ventilators are used in this setting compared to non-invasive ventilation, and the presence of a tracheostomy adds another dimension to the care of this group of children.

Existing guidelines

Authors from several countries have described their experience with paediatric home-based ventilation programmes for children (United Kingdom[3, 5], Canada[28], America[29], France[30, 31], Switzerland[32], and Japan[33]). All but one of these papers were published in the last five years. They highlight substantial differences in practices, care packages and financial support. Only the United Kingdom[3] and the USA[34] have published guidelines for home ventilation in children. The American Thoracic Society guideline[34] was published in 1990 and has not been revised. In 1998, guidelines produced by the UK Working Party on Paediatric Long Term Ventilation (LTV)[3] suggested that, with appropriate support and careful planning, long-term ventilation for children in the home was feasible. These recommendations have been updated and can be found on the UK website (www.longtermventilation.nhs.uk).

The UK survey[5] highlighted specific obstacles to discharge: failure to recruit qualified nursing staff or trained carers; delay in obtaining funding; and unsuitable housing. Various aspects of home ventilation in the UK have also been published[35, 36, 37, 38]. A review by Edwards five years following implementation of these guidelines reiterated that, although transitioning a child on tracheostomy ventilation from hospital to home was feasible, the obstacles to discharge remained frustratingly similar[14]. In Australia and New Zealand, although the patient group is similar[6], community support services for children requiring long term ventilation differ significantly from those in the UK. There is an urgent need for guidelines and recommendations for Australia and New Zealand.

PRACTICAL ISSUES

Considering the options

Discussions regarding the medical and ethical aspects of whether a child should be ventilated long-term should take place in paediatric multidisciplinary meetings, and a consensus reached prior to consideration of home therapy[32]. Any form of assisted ventilation has enormous implications for the quality of life of the child and their family, and tracheostomy ventilation has the most impact[39]. Discussions should include explanation of the expected benefits and risks of treatment and the anticipated burden of treatment on family life. Withdrawal of respiratory support may be appropriate where the child's underlying condition is progressive or irreversible, with poor quality of life[40]. Detailed discussion of the ethics of introducing or withdrawing treatment is outside the scope of this document and is covered in recent reviews[41, 42].

Indications

The main indication for long-term ventilation via tracheostomy is the inability to wean from invasive ventilatory support to intermittent non-invasive ventilation. Examples of conditions in which this treatment may be applicable include cervical cord injury, tracheobronchomalacia, and central alveolar hypoventilation. In order to be eligible for care at home, the child's medical condition and ventilatory support requirements should be stable. "Stable ventilatory support requirements" is regarded as there being no requirement for changes for a month[43].

Contraindications

- Unstable medical condition (either respiratory or other organ systems)
- Household factors making ventilation unduly difficult or dangerous (relative)
- Care and protection issues (relative)
- Financial (relative)[43]

Service provision

At present government funding for the care of children on long term ventilation varies between countries, regions and even between centres within one region. Appropriate funding packages for home care should be able to be put in place immediately when a child meets criteria of stability for discharge, to prevent inappropriate long-term hospitalisation. Clear pathways should be accessible for channelling funding applications. Funding packages should be flexible in terms of content, and time, allowing for increases and decreases in support as needed.

Recommendations

- Eligibility for home ventilation via tracheostomy depends on stable medical condition and ventilatory requirements for four weeks.
- Funding channels for home care should be clear, so that care packages can be readily put together to facilitate optimal timing of discharge.

MANAGEMENT GUIDELINES

The following recommendations have been made acknowledging that for an individual child many aspects of care will be influenced by co-morbid conditions and these factors should be included in assessment of home care requirements.

Carers in the home

Children requiring ventilation via a tracheostomy require close supervision 24 hours a day, to ensure patency of the tracheostomy and ongoing adequate ventilation. Families are unable to sustain this level of care without assistance. On discharge, trained carers (nurses, family members or health care assistants) should be provided every night, plus extended periods in the daytime to support the child at home or in school. Carers should be awake during the night so that problems can be immediately attended to. Older children without conditions affecting their ability to arouse to respiratory stimuli may be able to manage with carers who sleep during the night, but this should not be considered the standard of care for initial discharge. The amount of support provided during the day will depend on the child's co-morbidities, mobility and communication skills, the need for daytime ventilation and family factors.

Lay health care assistants and/or family members can be comprehensively trained in all aspects of the child's care to provide appropriate care in the home environment. Home care and ongoing training should be supervised by specialist paediatric nurses. The availability of an organisation that specialises in co-ordinating complex home care is a significant advantage. Training of carers may be provided by specialist nurses and medical staff in the hospital setting or in the community, depending on local resources and regulations. The discharging tertiary paediatric service has the overall responsibility for ensuring that the child is discharged to a safe environment. Training of the carers should never be the responsibility of the parents. If a community agency is utilised to supply home carers, the care agency contract should have an audit feature so that training credentials of carers can be assessed in an ongoing way.

Multidisciplinary care team

The care of a child dependent on long term ventilation is best achieved with a multi-disciplinary team, with a key person nominated to lead the group. The team should include a respiratory paediatrician, a general paediatrician, +/- a paediatric intensive care specialist (or other paediatrician with experience in ventilation), specialist nurses and/or sleep technologists familiar with home ventilation equipment, allied health (child mental health professionals, physiotherapist, occupational and speech language therapists), a social worker familiar with community support available to parents of children with complex health needs, and the community agency co-ordinating the home carers. Other health care workers may be called upon in specific instances. Regular meetings involving all team members should occur, with the frequency of meetings determined by the child's underlying illness, rate of change, age, medical stability and the social situation.

Parents & family

Most parents will not have a realistic idea of what life with a ventilated child will mean to them and their families, to their other children or to the child in question. While parents can become experts in their child's specific health care needs, it should not be assumed that parents are able or willing to take on the additional roles required for the care of a ventilated child. From the beginning, parents must be involved in the decision-making process regarding discharge and home care. In some cases, foster care or shared care may be appropriate. Discharge planning must provide families with the education and skills to safely care for their child[44-46]. Parents and extended family members require training similar to the professional carers in all aspects of the child's needs.

Risk minimisation

A child who is dependent on assisted ventilation and their family live with a level of risk that other people do not. Discharge from hospital care comes with additional risks, and parents need to be clear that the benefits of non-institutional care outweigh these risks. The risk management considerations of the ventilator- dependent patient at home have been highlighted recently[47] and a multicentre quality control study has examined the performance of mechanical ventilators at home[48]. Major risks include accidental disconnection of tubing from the tracheostomy and equipment failure. It is impossible to determine the absolute frequency of disconnections, but an American study published in the early 90's found that every family experienced some kind of difficulty with the ventilation equipment (up to 20 times per year for two thirds of the families in the study)[49]. Equipment for home mechanical ventilation can be expected to fail once every 1.25 years[50]. Overall, the morbidity for home care of ventilator dependent children is low, with the greatest risk being in the initial transition period from hospital to home[51].

Daily care plans, action plans and risk minimisation strategies need to be individually written for each child. Action plans for respiratory exacerbations need to be in place at discharge and reviewed regularly, including how to arrange urgent medical review, changes to ventilatory settings, addition of oxygen etc. Planning for emergencies should include provision of a list of contacts (e.g. PICU, respiratory paediatricians or paediatrician on call in regional area) whom the parents and carers can call on at any hour for advice or support. The action plan should include calling for an ambulance if more urgent support is required. At the time of discharge, ambulance services should be aware of the child's condition and ventilatory support needs, so that appropriate teams can be sent in an emergency. A clear written emergency plan with medical contact details included should be available to be given to ambulance officers when they arrive. This emergency plan should be discussed and practiced with the family and carers prior to discharge.

Transition to home

Home visits by nursing staff and/or other team members should occur early in the discharge process to assess the home environment in relation to the new needs of the child. Several reviews have highlighted that inappropriate home settings are a frequent cause of delayed discharge for children with complex health care needs[3, 5, 14]. Ventilation equipment requires considerable space and access to power outlets, and modifications to meet equipment specifications and work place standards may be required. Parents may need support to find alternative housing.

Discharge from hospital in a supportive and staged fashion allows parents and health care teams to trouble-shoot any gaps in the care plan, equipment or home set-up. Discharge may be preceded by a period spent in a care-by-parent unit or transitional care unit within the hospital or its grounds, where parents and carers look after the child with easy access to help if required. Close liaison with the child's regional medical team is imperative, particularly if the family lives a significant distance from the tertiary hospital; this may involve training of local staff and/or transfer to a regional hospital prior to discharge home.

Recommendations

- Children on tracheostomy ventilation require awake overnight carers.
- Carers should be trained and credentialed in all aspects of the child's care.
- A formal accreditation of achievement of a satisfactory standard of training should be part of the contractual agreement between the hospital and the home care organisation.
- If a community agency is utilised to supply home carers, the care agency contract should have an audit feature so that training credentials of carers can be assessed in an ongoing way.
- A case manager should be identified for each child.
- Discharge should be a staged process, ensuring high quality of care and parental and carer confidence.
- Discharge planning must provide families with the education, skills and resources to care for their child.
- A risk minimisation strategy and action plan should be formally documented for each child.

EQUIPMENT REQUIREMENTS

The requirements listed below are in addition to any equipment needed for the care of a tracheostomy itself. The reader is referred to the American Thoracic Society Guidelines for the Care of a Child with a Tracheostomy for more detail in this regard[2]. Prior to discharge, a child ventilated via a tracheostomy should be stabilised on the equipment they will use at home.

Minimum requirements

1. A portable ventilator, suitable for home use in children. It is recommended that each centre have one or two preferred ventilators, so that workers are familiar with the capabilities, limitations, and operation of these ventilators. Excellent summaries of different ventilators are available[52, 53]. The ventilator model chosen will depend on local availability and individual requirements. The ventilator should be set up for that individual child and then locked so that settings cannot be adjusted accidentally. Humidification is essential for tracheostomy ventilation[54, 55].
2. The following built-in alarms are mandatory regardless of the model of ventilator used:
    - Low tidal volume
    - High pressure
    - Disconnect (low pressure)
    - Power failure
3. An alternative power source - this may be a battery or a generator.
4. Hand ventilation equipment for manual ventilation will be needed in the event of mechanical or electrical failure. It may also be required for use away from home in children who do not require 24-hour ventilation but may need support unexpectedly.
5. An alternative ventilator for use in the event of a mechanical failure. Where this ventilator is kept will depend on the geographical location of the patient and the accessibility of services able to provide an alternative ventilator at short notice. In the majority of cases, a second ventilator will need to be kept in the child's home.

Other potential requirements

1. Oxygen saturation monitor: Most children who are dependent on tracheostomy ventilation should have an appropriate oximeter available in the home. This may not be required continuously.
2. Oxygen concentrator or cylinders capable of providing sufficient oxygen flow.

Recommendations

- Each centre should have one or two preferred ventilator models, so that workers are familiar with the capabilities, limitations, and operation of those ventilators.
- Minimum standards for ventilatory equipment should be universally applied across Australia and New Zealand.

KEY CHALLENGES

Readmission to hospital

When readmitted, a child on tracheostomy ventilation will require care by nursing staff skilled in tracheostomy care and the use of ventilators. The location of care will therefore vary between hospitals. The ideal would be a high dependency area or transitional care unit near to, or within the paediatric intensive care unit.

Funding

The experience of the group is that there are currently major disparities and inadequacies in funding for children needing long term ventilation. This is an area that needs to be formally documented for Australia and New Zealand. When a child is identified as suitable for home ventilation, funds for these children should be available within an appropriate time frame from governments and health authorities.

Recommendations

- Funding for home ventilation should be equitable, optimal and accessible across Australia and New Zealand.

B]: CHILDREN DEPENDENT ON NIGHTLY NON-INVASIVE VENTILATION

This section addresses the needs of children on non-invasive ventilation, where care requirements and risks are of similar intensity to those on tracheostomy ventilation, as ventilation cannot be discontinued for any period during sleep without risk to the child's life. A small but increasing number of children with complex multi-system disorders are dependent on this mode of ventilatory support.

Existing guidelines

No formal guidelines exist for the management of children dependent on non-invasive ventilation, apart from a document specific to central congenital hypoventilation syndrome [56].

PRACTICAL ISSUES

Considering the options

Children that are dependent on ventilatory support have traditionally been managed with a tracheostomy, and this remains the option of choice for ventilation >16 hours/day[57, 58, 59]. However, a small but increasing number of children who are dependent on supported ventilation are managed with non-invasive ventilation. This option avoids the potential morbidity/mortality of a tracheostomy and offers advantages to the child and family. The choice of invasive or non-invasive ventilation may be made at diagnosis (including for infants), or a child may be transitioned to non-invasive ventilation from tracheostomy ventilation as their condition improves or when age or other factors influence this choice. These decisions should be made by a home ventilation team in conjunction with the parents and, if appropriate, with the child.

Indications

Long term ventilation is defined as failure to wean from ventilatory support once a child is medically stable, three months after the institution of ventilation[3]. Non-invasive ventilation is ventilatory support delivered via a nasal mask or similar non-invasive interface. The underlying paediatric medical conditions fall into three main groups: neuromuscular disease, central hypoventilation, and severe respiratory insufficiency. Any of these conditions may be congenital or acquired.

Contra-indications

Non-invasive ventilation is contraindicated or not practical for long-term home use:

- in the absence of a secure airway during wakefulness
- if NIV fails to normalise ventilation, in a child whose underlying condition is considered reversible or non-progressive.

If ventilation is required more than 16 hours per day tracheostomy should be considered, due to the side effects and practicalities of delivery[57, 58, 59].

Funding Packages

Funding packages should be based on those for children on tracheostomy ventilation, with flexibility for change over time following review. At present, this group can be discriminated against in the provision of funding (as they do not have a tracheostomy), despite the fact that their care needs are the same.

MANAGEMENT GUIDELINES

The requirement for carers, training of parents and carers, transition from hospital to home and equipment requirements are the same as for children dependent on tracheostomy ventilation, and are discussed in Part 1.

Recommendations

- Non-invasive ventilation is an alternative to tracheostomy ventilation in children who are dependent on respiratory support during sleep.
- Comparable levels of funding, supervision, training and equipment apply as for children dependent on tracheostomy ventilation.

PART 2: VENTILATORY SUPPORT FOR OPTIMISATION OF A CHILD'S HEALTH: INCLUDING CPAP AND NIV

This section addresses those children for whom ventilatory support has been prescribed to normalise ventilation during sleep. These children are able to discontinue ventilatory support during sleep for short periods without immediate threat to their life. However, failure to adequately treat their respiratory insufficiency in the medium to long term will lead to serious morbidity and premature mortality[19, 20, 60, 61].

Existing guidelines

No guidelines currently exist for the management of children on non-invasive ventilation.

Several centres have published their experience with commencing children on NIV [6, 16-18, 62]. These papers highlight the need for paediatric experience, a behavioural modification approach, and parental commitment for successful initiation of non-invasive respiratory support in children. In 2004 the American Thoracic Society published a consensus statement regarding the respiratory care of the patient with Duchenne Muscular Dystrophy (DMD)[63]. This document highlights issues relating to provision of respiratory support for this patient group.

PRACTICAL ISSUES

Considering the options

There is now clear evidence that non-invasive support extends survival and improves quality of life in patients with progressive neuromuscular conditions like DMD[19, 20, 60]. Children who would potentially benefit from NIV or CPAP (see below) should have access to a paediatric facility with tertiary respiratory and sleep medicine services, so that informed discussions can occur regarding the advantages and limitations of ventilatory support. Discussions should include escalation of support during a significant respiratory infection[39, 60], at the time of spinal surgery[64-66] and towards the end of life[41, 67, 68].

Indications

Non-invasive ventilation

Children with hypoventilation during sleep, even if isolated to rapid eye movement sleep, benefit from non-invasive ventilation[20, 26, 69-71]. The presence and severity of hypoventilation during sleep is best established by polysomnography. Indications for long-term respiratory support with NIV include hypoventilation secondary to:

- neuromuscular disease
- chest wall restriction (bony abnormalities etc)
- chronic lung disease
- severe upper airway obstruction not adequately treated with CPAP
- obesity hypoventilation.

CPAP

The largest group of children requiring CPAP have obstructive sleep apnoea (OSA). A large proportion of these children have significant medical co-morbidities[6, 16-18] which need to be taken into account when planning support for the home. Indications for long-term respiratory support with CPAP include:

- Obstructive sleep apnoea not successfully treated with surgical options such as adenotonsillectomy, or in children where surgery is contra-indicated;
- Craniofacial and airway abnormalities;
- Chronic lung disease.

Initiation of therapy

Initiation of CPAP or NIV is not urgent in most cases and can usually be instigated through a planned process. A staged approach including mask fitting as an outpatient +/- a period of acclimatisation to the mask at home may enhance the child's acceptance of the treatment in the long term[17]. CPAP or NIV may be initiated in a hospital ward, PICU or sleep laboratory (as an outpatient). In all cases, ventilatory settings and equipment need to be individualised for each child by trained paediatric staff. Compliance with therapy is enhanced by commencement of therapy in a unit with skilled paediatric staff and an appropriate behavioural management programme[17, 72].

Funding and service provision

Appropriate funding for diagnosis, equipment and long term management is important for NIV and CPAP. This should be funded by designated government authorities regardless of the child's age, and be equitable across regions, including equipment funding packages.

Recommendations

- Access to paediatric diagnostic services should be available to all children who would potentially benefit from NIV or CPAP.
- Ventilatory settings and equipment should be individualised for each child by trained paediatric staff.
- Equipment for CPAP and non-invasive ventilation should be funded by designated government authorities and equitable across regions.

MANAGEMENT GUIDELINES

Family support

Children at home on respiratory support have care and supervision needs over and above those of another child of the same age, and should be eligible for government financial assistance (e.g. Child Disability Allowance). Carers are not usually needed in the home, but this will depend on the co-morbidities / associated disabilities of the child. Respite care may be needed and should be incorporated into funding packages where required.

Home ventilation teams

It is important that staff be experienced in using CPAP and NIV for the successful initiation and ongoing care of children on respiratory support. All families should have access to specialised advice at all times during the course of treatment, to ensure assistance with trouble shooting and rapid rectification of problems. The team needs a process to ensure adequate follow-up of all children on respiratory support, both for clinical review and review of ventilation settings (with polysomnography if appropriate), ensuring changing needs are anticipated and addressed[3, 14, 73, 74].

Risk minimisation

NIV and CPAP are usually low risk therapies. Parents should receive education on how to avoid, anticipate and treat complications that arise, such as skin breakdown or mask leak. Higher risks may apply to infants and children who are unable to remove their mask or signal for assistance. All families should know how to prioritise and seek assistance in the event of equipment failure (replacement within 24 hours or longer) and have a documented action plan. Regular equipment servicing according to the manufacturer's recommendations is essential.

Recommendations

- NIV and CPAP should be available and accessible to all children whose health could be optimised with such therapy.
- NIV and CPAP should be instituted and monitored by multidisciplinary paediatric teams experienced in such treatment for children.
- Funding packages should allow for changing needs over time.

EQUIPMENT REQUIREMENTS

The most problematic issues for NIV in children are achieving low dead space and appropriate triggering sensitivity. It is recommended that each centre have one or two preferred machines, so that staff are familiar with the capabilities, limitations, and operation of each. The machine settings should be lockable so that settings cannot be adjusted accidentally. Although humidification is optional, it is increasingly being recommended and may improve compliance[54].

For NIV and ideally for CPAP for infants and children in high risk groups (includes those under five years of age and children with mobility difficulties restricting mask removal), the following alarms are advised, regardless of the model of ventilator used:

- Mask alarm (to detect accidental removal of mask during sleep)
- High pressure alarm
- Disconnect / low pressure alarm
- Power failure alarm

Masks designed specifically for children are ideal, with low dead space and appropriate size and shape to limit leak and minimise pressure marks on the face. These interfaces should be reviewed and adjusted regularly as the child grows.

KEY CHALLENGES

Readmission to hospital

Children on respiratory support require staff specifically trained in the care of ventilation equipment and their underlying disease. This will vary from a ward with trained staff to a high dependency unit to PICU depending on the local situation. Children on NIV or CPAP should be advised to take their equipment with them for any hospital admission, regardless of the reason for hospital admission.

PART 3: ISSUES COMMON TO ALL GROUPS

Ventilatory support is a highly specialised treatment that should always be commenced and followed up under the auspices of a tertiary specialised paediatric service.

Medical review

A multi-disciplinary clinic would be ideal to avoid frequent outpatient visits. The timing of clinical review by a paediatrician experienced in home ventilation will vary depending on the child's needs. It is likely to be more frequent in the period after initiation of respiratory support and decrease with time, e.g. two to three monthly for children on invasive ventilation and three to six monthly for those on NIV or CPAP. The review will include assessment of compliance (subjective & objective[15]) and complications of therapy. Clinical review should be coordinated with appropriate investigations (e.g. lung function testing) and other specialist assessment if indicated. Vaccination against respiratory pathogens should be considered[75, 76].

Tertiary paediatric centres should have a designated team for children on ventilatory support, with each child having a case manager. The case manager should have responsibility for ensuring follow-up within a multidisciplinary framework to ensure the changing needs of the child and family are anticipated and addressed.

Follow-up sleep studies are recommended at regular intervals for all children on respiratory support. Full polysomnography is ideal to ensure that sleep architecture as well as gas exchange is normalised[73, 74]. Changes to settings may be required with increasing age and growth or due to changes in the underlying condition of the child. These will vary from three to six monthly early in the course of treatment or in children with an unstable medical condition to 18 – 24 monthly for children stable on CPAP for OSA.

Checking of ventilator / CPAP settings, alarm functioning, battery power level and machine servicing should occur annually or as recommended by the manufacturer to ensure appropriate functioning. This may be performed by the hospital biomedical engineering department, the manufacturer or by a specific service provider.

Recommendations

- Medical review will be determined by the individual clinical situation, but should be at least every three months for children on invasive ventilation and three to six monthly for those on NIV or CPAP.
- Funding should be available for review to occur at a tertiary centre with access to lung function testing, paediatric sleep medicine services and experience in NIV and CPAP.
- Follow-up sleep studies are recommended for all children on respiratory support.
- Tertiary paediatric centres require a coordinator for home ventilatory support patients.
- Home ventilation equipment should be checked /serviced as recommended by the manufacturer.

Transition from paediatric to adult care

There are many reviews on the medical transition process from paediatric to adult care. We refer the reader to consensus statements on health care transition for young adults with special health care needs[77,78]. The choice of primary adult physician and respiratory support service for adolescents and young adults on respiratory support will depend on the underlying diagnosis of the young person, the locality of services and the wishes of the patient. The timing of transition can be especially difficult in adolescents with a progressive disorder who are nearing the terminal stages of their disease. The timing of transition should be discussed at least 24 months prior to transfer and hand over of care should be a staged process.

Recommendations

- Appropriate adult services should be identified locally that are appropriate for adolescents with complex health needs.
- Funding should be maintained through the transition process.

Workforce issues / manpower planning

Due to the nature of the conditions of children requiring home ventilatory support, the initial treatment should ideally be within a tertiary paediatric service with respiratory paediatricians and intensivists. Due to the continuing increase in the numbers of children receiving ventilatory support at home, there will need to be an ongoing increase in the capacity of health care systems to deal with the needs of these children. In addition to equipment requirements, the provision of appropriate diagnostic and nursing services is essential.

Once a child is home, the day to day care may rest with a regional general paediatrician and local allied health professionals[3] who may have very little experience or expertise in ventilatory support therapy. In Australia and New Zealand, children may live many kilometers from a tertiary centre, and educational packages, teleconferencing, telephone help and written action guidelines for families and local teams are important to ensure therapy is not compromised. It is important to include local paediatricians in the planning stage, and provide adequate support and education for those in regional centres.

Recommendation

- Increasing demand for home ventilatory support should be matched with appropriate funding, so that specialised diagnostic and treatment services can be maintained appropriate to the needs of this patient group.

Travel and fitness to fly

Airline travel for children on ventilatory support is an issue that is increasingly encountered. Long flights where the child may need to sleep are the most problematic and may necessitate a fitness to fly test[79-82]. We recommend that families are informed at the outset about the difficulties of airline travel, especially internationally. There will be restrictions in the choice of airline due to carry-on luggage rules, which machines can be used in flight (electrical isolation) and difficulties procuring travel insurance. The medical officer of the airline in question may provide information specific to the airline, and should be consulted well in advance of travel. It is also important to ensure that the family has appropriate carry bags to protect equipment and the correct electrical adapters for the countries they are visiting.

Recommendation

- The need for ventilatory support does not usually preclude air travel, but appropriate investigations and arrangements should be made with the airline in advance of travel.

WHAT IS NEEDED IN THE FUTURE

- Future research should focus on improving the evidence base for current medical models of care.
- An Australian and New Zealand registry of patients on home ventilatory support is required to inform future recommendations and assist in resource allocation planning. This process requires appropriate funding for set-up and maintenance.
- Development of education packages for parent/family is required.
- Establish a funded multi-centre committee to facilitate equitable standards of care.
- The committee would also work with manufacturers to improve equipment options for children.

CONCLUSIONS AND KEY MESSAGES

Although medically stable, children with chronic respiratory failure are dependent to varying degrees on mechanical ventilation in order to survive. Attention is required by policy makers and practitioners at all levels to ensure that their rights are respected and their needs met. Without coherent multi-disciplinary and inter-agency planning, some children, particularly those requiring invasive ventilation, endure unnecessarily long periods in hospital. Children discharged on respiratory support add a significant burden to parents and families. Provision of appropriate carer support and respite care is crucial to the well-being of the child and family.

Summary statements

- The APRG and TSANZ support the equitable access to diagnostic and therapeutic options for children who require respiratory support.
- The APRG and TSANZ support the care of technology-dependent children in their own home.
- The establishment of a multi-centre working group and a patient registry would inform individual patient care and policy development.

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