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Author/s:

Chawla, J;Edwards, EA;Griffiths, AL;Nixon, GM;Suresh, S;Twiss, J;Vandeleur, M;Waters, KA;Wilson, AC;Wilson, S;Tai, A

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## POSITION STATEMENT

# Ventilatory support at home for children: A joint position paper from the Thoracic Society of Australia and New Zealand/ Australasian Sleep Association

Jasneek Chawla<sup>1,2</sup>  | Elizabeth A. Edwards<sup>3</sup> | Amanda L. Griffiths<sup>4,5,6</sup> | Gillian M. Nixon<sup>7,8</sup> | Sadasivam Suresh<sup>1,2</sup> | Jacob Twiss<sup>3</sup> | Moya Vandeleur<sup>4,5</sup> | Karen A. Waters<sup>9,10</sup> | Andrew C. Wilson<sup>11,12</sup> | Susan Wilson<sup>13</sup> | Andrew Tai<sup>14,15</sup>

<sup>1</sup>Department of Respiratory and Sleep Medicine, Queensland Children's Hospital, Brisbane, Queensland, Australia

<sup>2</sup>School of Clinical Medicine, University of Queensland, Brisbane, Queensland, Australia

<sup>3</sup>New Zealand Respiratory & Sleep Institute, Starship Children's Hospital, Auckland, New Zealand

<sup>4</sup>Respiratory & Sleep Medicine, Royal Children's Hospital, Melbourne, Victoria, Australia

<sup>5</sup>Murdoch Children's Research Institute, Melbourne, Victoria, Australia

<sup>6</sup>University of Melbourne, Melbourne, Victoria, Australia

<sup>7</sup>Melbourne Children's Sleep Centre, Monash Children's Hospital, Melbourne, Victoria, Australia

<sup>8</sup>Department of Paediatrics, Monash University, Melbourne, Victoria, Australia

<sup>9</sup>Sleep Medicine, The Children's Hospital at Westmead, Sydney, New South Wales, Australia

<sup>10</sup>University of Sydney, Sydney, New South Wales, Australia

<sup>11</sup>Respiratory & Sleep Medicine, Princess Margaret Hospital for Children, Perth, Western Australia, Australia

<sup>12</sup>School of Paediatrics and Child Health, University of Western Australia, Perth, Western Australia, Australia

<sup>13</sup>Child Youth Mental Health Services, Queensland Children's Hospital, Brisbane, Queensland, Australia

<sup>14</sup>Respiratory & Sleep Medicine, Women's and Children's Hospital, Adelaide, South Australia, Australia

<sup>15</sup>Robinson Research Institute, University of Adelaide, Adelaide, South Australia, Australia

## Correspondence

Jasneek Chawla, Department of Respiratory and Sleep Medicine, Queensland Children's Hospital, 501 Stanley Street, South Brisbane, QLD 4101, Australia.  
Email: jasneek.chawla@health.qld.gov.au.

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## Abstract

The goal of this position paper on ventilatory support at home for children is to provide expert consensus from Australia and New Zealand on optimal care for children requiring ventilatory support at home, both non-invasive and invasive. It was compiled by members of the Thoracic Society of Australia and New Zealand (TSANZ) and the Australasian Sleep Association (ASA). This document provides recommendations to support the development of improved services for Australian and New Zealand children who require long-term ventilatory support. Issues relevant to providers of equipment and areas of research need are highlighted.

## KEYWORDS

home ventilation, paediatrics, respiratory, sleep, ventilatory support

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This document has been endorsed by the Thoracic Society of Australia and New Zealand and the Australasian Sleep Association. It is due for review in 2027.

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## THE PURPOSE OF THIS DOCUMENT

This position statement informs the principles of care for a child on long-term ventilatory support. Appropriate levels of service provision are highlighted and compared to those currently available in Australia and New Zealand. The aim is to highlight the need for services that are equitable, appropriate and safe to these children across Australia and New Zealand.

Details regarding the provision of ventilatory support at home will vary, depending on the severity of the underlying condition and presence of co-morbidities, the mode of ventilation and the number of hours of ventilation required per day. Changes in these factors may occur over time.

*This position statement represents consensus opinion of paediatric respiratory and sleep specialists on the management of children on ventilatory support at home. It is not a comprehensive 'standards of care' document.*

## DEFINITIONS

Paediatric long-term ventilation (LTV) refers to 'any child who, when medically stable, continues to require a mechanical aid for breathing, after an acknowledged failure to wean, or a slow wean, 3 months after the institution of ventilation'.<sup>1</sup> Children with a wide variety of underlying medical conditions can require long-term ventilatory support. Technical advances have led to an increasing number of children being managed with long-term ventilatory support using a non-invasive interface and therefore it is increasingly important to define the group of children being referred to.

The United Kingdom Long Term Ventilation Working Party define paediatric long-term ventilatory support as any child below the age of 17 yrs who is medically stable and requires a mechanical aid for breathing either invasively by tracheostomy or by non-invasive mask interface for all or part of the 24 h per day.<sup>2</sup> They specifically excluded children who are likely to wean from ventilation in 3 months. The American Thoracic Society Paediatric Chronic Home Ventilation Workgroup refer only to children requiring *invasive* ventilation in the home.<sup>3</sup>

In this document, the following definitions are used:

1. *Ventilatory support*: Assistance to breathing provided by mechanical means. This includes continuous positive airway pressure (CPAP) and mechanical ventilation delivered by any means (non-invasive or invasive) but excludes supplemental oxygen alone.<sup>4</sup> We do not discuss

the care of a child with a tracheostomy alone (without ventilation); for this, the reader is referred to the American Thoracic Society Guidelines.<sup>5</sup>

2. *Long-term ventilation*: Failure to wean from ventilatory support once medically stable, 3 months after the institution of ventilation.
3. *Invasive ventilation*: Ventilatory support delivered via an endotracheal tube or a tracheostomy.
4. *Non-invasive ventilation (NIV)*: Ventilatory support delivered via a nasal mask or similar non-invasive interface. This includes mask bi-level support (bi-level positive airway pressure [BPAP]) and CPAP support.
5. *Other forms of ventilatory support*: This includes negative-pressure ventilation, phrenic nerve pacing and high-flow nasal cannula therapy.
6. *Home*: May be the child's home, a foster home or a group living environment.

Children have been classified into subgroups according to the type of ventilatory support they require and the main goal of ventilation at home (Table 1). This includes:

1. Children dependent on ventilation
  - A. Continuous life support
  - B. Sleep life support
2. Children requiring ventilation for health optimization
  - A. Fragile health support
  - B. Health support
3. Ventilatory support for children with life-limiting conditions

This document is divided into sections covering these subgroups, with a section on issues common to all groups.

## ASSESSMENT OF EVIDENCE

Methods used in development of this position paper are summarized in Figure 1, with further information provided in Appendix S1 (Supporting Information).

The working party reviewed relevant literature to inform this update of a previous document.<sup>6</sup> Literature relevant to each section was identified if published after the original guideline, covering the period 2008–2018. Key search terms included: 'ventilation', 'invasive ventilation', 'non-invasive ventilation', 'bi-level or BiPAP or BPAP' and 'CPAP or PAP' for children. The search yielded small cohort studies or case series, with no randomized controlled studies. Due to the absence of randomized controlled trials, the authors reviewed all available literature with consensus of expert opinion reached to form the following recommendations.

## SUMMARY OF RECOMMENDATIONS

A summary of key recommendations is available in Appendix S2 (Supporting Information).

## 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 ventilatory support for all or part of everyday.<sup>7–9</sup> In the past 20 years, the number and complexity of children receiving home ventilatory support has increased significantly.<sup>2,10–12</sup> In 2019, there were 1359 children across Australia and New Zealand receiving home ventilatory support (Appendix S3 in the Supporting Information).

This growth is driven by a change in societal expectations of adequate support for those with long-term disability in the community, the pressure to reduce the duration of hospital stay and recognition that children with a broad range of medical conditions may benefit from long-term ventilatory support. Technical improvements have also meant that ventilatory support at home is feasible for more children. Care requirements at home vary and an individualized clinically driven approach is required. A principle of care is that long-term hospitalization is inappropriate for a developing child,<sup>13</sup> puts them at risk of hospital acquired infection and is detrimental to family functioning and relationships. Furthermore, it is expensive, an unnecessary use of resources and may block intensive care or medical specialty beds for other children.

The role of non-invasive ventilatory support to facilitate discharge home and improve quality of life in patients with life-limiting conditions has also expanded. Here, the goal is not to prolong life, but to increase the time families can spend away from hospital. A further large group of children have an unrecognized need for ventilatory support due to the lack of diagnostic and treatment facilities.<sup>14</sup> The equipment and expertise required to care for these children does not match the rapidly increasing number of children already on NIV and CPAP.<sup>11</sup> Furthermore, the level of support provided for home care of children on long-term ventilatory support is highly variable across Australia and New Zealand, highlighting the need to improve equity of access.<sup>15</sup>

As well as technological requirements, children on assisted ventilation require standard general paediatric health care. A child's developmental and educational needs should be a high priority, and health care should be delivered in a way that promotes participation in family life and education, maximizing the child or young person's potential. Maintaining their role and relationships within their family is paramount. 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, including siblings.

The need for psychological and emotional support for children receiving home ventilation and their families is increasingly recognized. Survey data from children using home mechanical ventilation and their families have demonstrated lower quality of life scores than both healthy populations and children with other chronic conditions.<sup>16</sup> Many studies have highlighted that family dysfunction can occur in this population.<sup>16–20</sup> Families report that caring for their children has negative implications such as marital conflict, emotional overload or the feeling of

**TABLE 1** Classification of subgroups of children requiring ventilatory support

| Support level                                                                              | Description                                                                                                                                                                                                                                                                                                                                                                 | Example                                                       |
|--------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------|
| 1 Children dependent on ventilation                                                        | <ul style="list-style-type: none"> <li>Those who require a mechanical aid for ventilation to maintain life in the short or long term, with a high risk of death or significant adverse outcome if ventilatory support is discontinued</li> </ul>                                                                                                                            | Two subgroups are recognized: 1A and 1B                       |
| 1A: Continuous life support                                                                | <ul style="list-style-type: none"> <li>The child is dependent on ventilatory support for at least 16 h per day to maintain life awake and asleep</li> <li>Life would be threatened by loss of ventilatory support</li> </ul>                                                                                                                                                | End-stage respiratory failure<br>High cervical cord injury    |
| 1B: Sleep life support                                                                     | <ul style="list-style-type: none"> <li>The child is dependent on ventilatory support to maintain life asleep and life would be threatened by continued sleep without ventilatory support</li> <li>In the event of equipment/power failure, the child would be safe if able to be woken</li> </ul>                                                                           | Congenital central hypoventilation syndrome                   |
| 2 Children requiring ventilation for health optimization                                   | <ul style="list-style-type: none"> <li>Those in whom ventilation is required for optimal health but is not 'life support' as defined above</li> <li>This group can discontinue ventilatory support during sleep for short periods without immediate threat to life but there may be medium- or long-term negative effects without regular utilization of therapy</li> </ul> | Two subgroups are recognized                                  |
| 2A: Fragile health support                                                                 | <ul style="list-style-type: none"> <li>The child is not dependent for awake or asleep on the ventilatory support for continuation of life on a short-term basis but has fragile health status and may be subject to adverse outcomes without daily use or in the event of a failure of the ventilator</li> </ul>                                                            | Severe neuromuscular disease or young infant on mask NIV/CPAP |
| 2B: Health support                                                                         | <ul style="list-style-type: none"> <li>The child is not dependent for awake or asleep on the ventilatory support for continuation of life on a short-term basis</li> <li>Regular use optimizes health, but a major adverse event is unlikely without use in the event of failure of the ventilator</li> </ul>                                                               | Majority of CPAP for OSA                                      |
| 3 Ventilatory support for children with life-limiting conditions receiving palliative care | <ul style="list-style-type: none"> <li>ventilatory support provided with a focus on palliation of symptoms and to facilitate quality of life (e.g., discharge from hospital) in those with a life-limiting condition</li> <li>Prolonging life is not the goal in this situation</li> </ul>                                                                                  | End-stage respiratory failure                                 |

Abbreviations: CPAP, continuous positive airway pressure; NIV, non-invasive ventilation; OSA, obstructive sleep apnoea.

neglecting their other children.<sup>16</sup> Despite this evidence, resources to provide psychological support are still lacking in most centres across Australia and New Zealand.

While home-based care is often feasible<sup>21</sup> and may be less expensive than in-hospital care, increasing demand for care at home has highlighted funding issues for the home care of children with significant medical and technological needs. Reviews demonstrate that once a child requiring LTV is home, non-elective readmission to hospital is unusual.<sup>22–24</sup> Limited information is available regarding long-term outcomes of children cared for at home on ventilation via a tracheostomy, and children using NIV.<sup>9,25–30</sup> Further information on such outcomes for both child and family will inform future funding and care decisions, and the development of standard of care guidelines.

## PART ONE: CHILDREN DEPENDENT ON VENTILATION

### (A) Continuous life support

#### Introduction

Continuous ventilatory support is usually delivered via tracheostomy when there is either a need for prolonged hours on ventilatory support each day or if the child has an abnormal upper airway. The ventilators used in this setting and

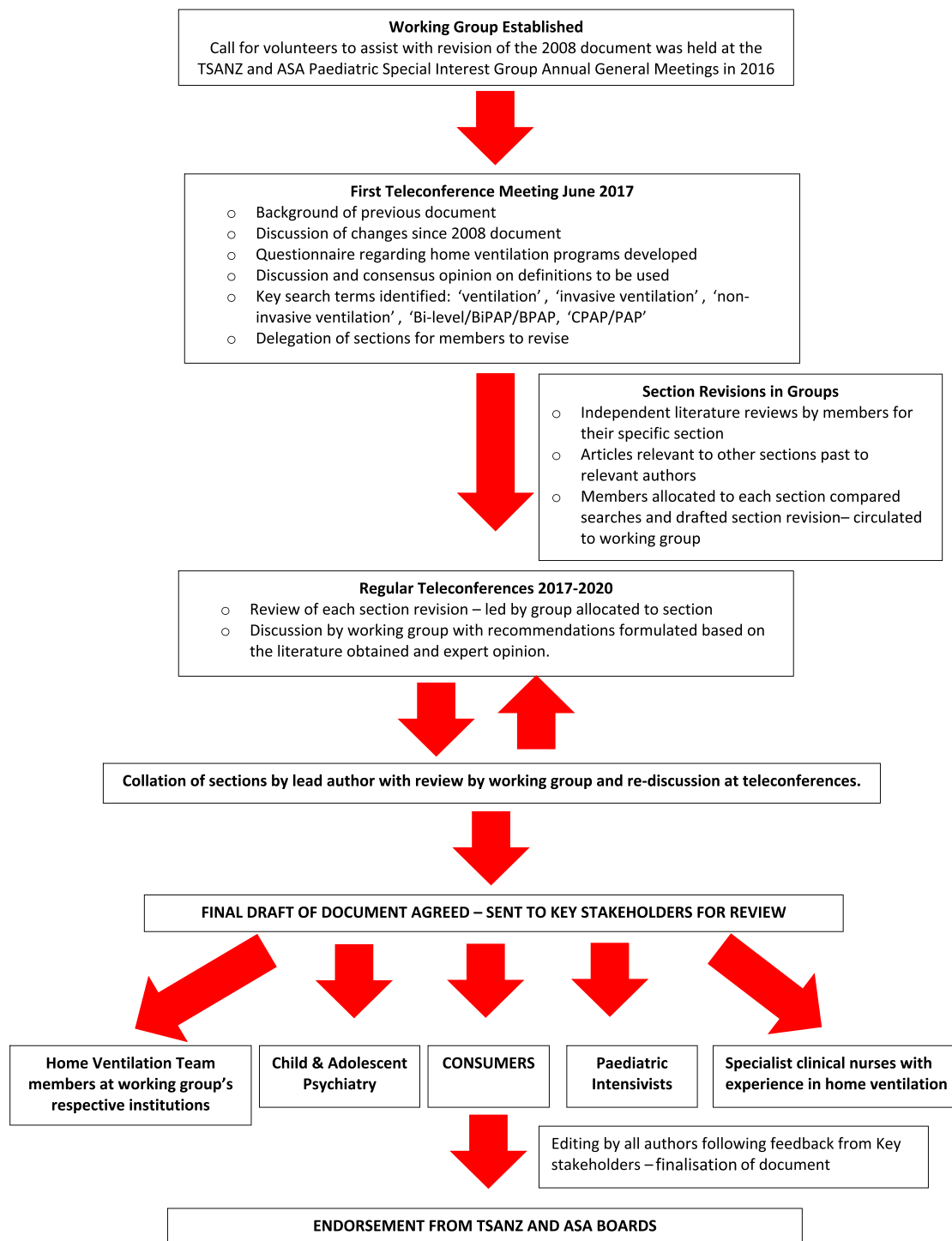
the presence of a tracheostomy add to the care needs of this group of children.

#### Existing guidelines

Worldwide, there are substantial differences in practices, care packages and financial support for children managed by paediatric home-based ventilation programmes.<sup>1,8,31–36</sup> Only the United Kingdom<sup>1</sup> and the United States<sup>37</sup> have published guidelines for home ventilation in children. The American Thoracic Society guideline<sup>37</sup> was published in 1990 with an update for chronic home invasive ventilation in 2016.<sup>3</sup> In 1998, guidelines produced by the UK Working Party on Paediatric Long-Term Ventilation<sup>1</sup> suggested that, with appropriate support and careful planning, LTV for children in the home is feasible. These recommendations have been updated and can be found on the UK website ([www.longtermventilation.nhs.uk](http://www.longtermventilation.nhs.uk)).

A UK survey published in 1999<sup>8</sup> highlighted obstacles to hospital discharge: failure to recruit qualified nursing staff or trained carers, delay in obtaining funding and unsuitable housing. Perspectives on community support, care pathways and discharge planning for home ventilation in the United Kingdom have been published.<sup>38–41</sup>

A review by Edwards et al. in 2004,<sup>26</sup> 5 years following the publication of the UK guidelines,<sup>1</sup> reiterated



**FIGURE 1** Summary of process for developing position statement

that, although transitioning a child on tracheostomy ventilation from hospital to home was feasible, the obstacles to discharge remained frustratingly similar. In Australia and New Zealand, although the patient group is similar,<sup>9</sup> community support services for children requiring LTV differ significantly from those in the United Kingdom, although many of the same obstacles are faced by families.

## Practical issues

### *Considering the options*

The medical and ethical aspects of whether a child should be ventilated long term should be discussed in paediatric multidisciplinary meetings prior to consideration of home therapy.<sup>35</sup> 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.<sup>42</sup> Discussions should include explanation of the expected benefits and risks of treatment and the anticipated burden of treatment on family life. Withdrawal of ventilatory support may be appropriate where the child's underlying condition is progressive or irreversible, with poor quality of life.<sup>43</sup> These issues are outlined further in Part four, but a detailed discussion of the ethics of introducing or withdrawing treatment is outside the scope of this document.<sup>3,44,45</sup>

### *Indications*

A requirement for long-term mechanical ventilation most often starts during an admission to the intensive care unit (ICU), with failure to wean from full-time invasive to intermittent NIV, leading to tracheostomy for ongoing provision of invasive ventilation. Examples of conditions in which this may be required include cervical cord injury, tracheo-bronchomalacia and central alveolar hypoventilation. Eligibility for care at home requires stability in the child's medical condition and ventilatory support requirements. This is usually considered when no changes have occurred for a month.<sup>46</sup>

In contrast, implementation of ongoing NIV is more likely to occur through monitoring of disease with overnight polysomnography (PSG), although individual centres may or may not involve hospital admission in this process. The level of support families receive tends to be limited to equipment and/or financial support (carers' payments), without in-home carers.

The management of discharge to home<sup>47</sup> ideally requires a centre with the capacity to set up, monitor and optimize the ventilation therapy; provide for emergency admission in cases of deterioration; and be a contact point for those managing the patient at home. A consistent process for discharge within an expert team improves the time to discharge.<sup>48</sup> Recommended discharge requirements are summarized in Table 2. Failure to meet discharge criteria can constitute a contraindication to the therapy and needs to be addressed early in the planning. Recently published German national guidelines provide excellent detailing of the requirements for each step of transitioning to home, from the perspective of the managing team.<sup>47,49</sup>

### *Service provision*

Financial and carer support packages for the care of children on LTV vary widely in Australia and New Zealand, between countries, regions and even between centres. Packages need to be flexible in terms of content and time, allowing for adjustments when the support needs of the child change. Early implementation of appropriate funding packages prevents inappropriate long-term hospitalization. Factors known to delay discharge include parent training, obtaining approval from the agency financing home care and staffing for any home carers. Anticipation of these issues and optimizing the processes can minimize delays to discharge.<sup>50</sup>

## **Management recommendations**

The following recommendations acknowledge that for an individual child many aspects of care will be influenced by

co-morbid conditions, which need to be included in the assessment of home care requirements.

Providing ventilation in the home increases quality of life through family caregiving, the ability to participate in social life with psychosocial support and increased patient autonomy. It is also linked with reduced infection rates.<sup>15,51</sup> Key elements in home management for invasively ventilated children include provision of appropriately trained carers and support by multidisciplinary team. Discharge home of a child who is ventilated places considerable demands on the parents and family. Studies of the stresses and mechanisms that staff and family use to cope are mainly from ICUs.<sup>52</sup> The alteration in parental role is a major stressor, as they shift to become an active carer for their child.

### *Carers in the home*

Existing guidelines recommend that an awake, trained caregiver be present at all times for a child who is chronically, invasively ventilated, with at least two family caregivers receiving specific training.<sup>1,3</sup> The presence/absence of associated disabilities will affect the level of care a child requires.<sup>47</sup> Support from professional, appropriately trained in-home caregiver(s) is required by most families, to allow time to sleep, work and maintain a life balance. However, the presence of regular carers in the home may also have a negative impact on families.

Close supervision (24 h per day) is required to ensure patency of the tracheostomy and on-going adequate ventilation for children who cannot address these issues themselves. Lay healthcare 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. This initial and ongoing training should be supervised by specialist paediatric nurses and should never be the responsibility of parents. Trained carers (nurses, family members or healthcare assistants) should be provided every night, with 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 addressed. Organizations that specialize in coordinating complex home care offer a significant advantage. Recommendations for the provision of home ventilation care provided by the disability sector are summarized in Appendix S4 (Supporting Information). The discharging tertiary paediatric service has the overall responsibility for ensuring that the child is discharged to a safe environment.

### *Multidisciplinary care team*

Families should be made aware early on of the large multidisciplinary team that will be involved in caring for their child both in hospital and at home. This will include not only health professionals, but also additional supports from community services (e.g., disability services, housing and education).<sup>53</sup>

Consensus is that expertise can be developed in multidisciplinary teams to coordinate the management of these children. Elements of importance include: a nominated team

**TABLE 2** Discharge requirements for the child dependent on home ventilation and relative contraindications for consideration prior to discharge

| Recommendation                                                                                      | Additional information                                                                                                 |
|-----------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------|
| 1. Stable underlying disease and co-morbidities                                                     | Defined as no recent requirements for treatment changes (as assessed by the child's clinical team)                     |
| 2. A (verified) safe environment                                                                    |                                                                                                                        |
| 3. Stable ventilation status without complications                                                  | Using the same ventilator as intended for home use                                                                     |
| 4. Stable tracheostoma (where present)                                                              |                                                                                                                        |
| 5. Funding in place                                                                                 | Arrangement to take over the costs for nursing care and medical aid supplies                                           |
| 6. Equipment in place                                                                               | Supply with all the necessary devices, consumables and materials and ensure all are available at the time of discharge |
| 7. Follow-up plan in place                                                                          | Appointment for the first follow-up should be scheduled pre-discharge                                                  |
| <b>Relative contraindications (issues that may need to be addressed prior to initial discharge)</b> |                                                                                                                        |
| 1. Unstable medical condition                                                                       | Either respiratory or other organ system                                                                               |
| 2. Household factors                                                                                | Factors that make ventilation at home unduly difficult or dangerous                                                    |
| 3. Care and protection issues                                                                       | Include screening of parental health and provision of psychosocial support as appropriate                              |
| 4. Financial hardship                                                                               |                                                                                                                        |

member to communicate with the family, respiratory/sleep paediatrician, general paediatrician, paediatric intensive care specialist, specialist nurses and/or sleep technologists/physiologists familiar with home ventilation equipment, allied health (child mental health professionals, physiotherapists, occupational and speech language therapists), a social worker and the community agency coordinating home carers. Regular meetings with team members should occur, with the frequency of meetings determined by the child's underlying illness, rate of change, age, medical complexity and the social situation. The general practitioner and community nursing teams should be engaged early and kept up to date with progress, particularly closer to discharge from hospital or after any acute admission.

### Parents and family

The first major change for the family is the shift from a 'parenting' role to that of 'carer' with expertise in their child's condition and liaising with the multiple professionals involved. This most commonly occurs during an intensive care admission, often followed by a prolonged admission, after the child's health has stabilized, due to the time taken for organizational (particularly funding) processes and living arrangements to be finalized.<sup>50,54</sup> Staff communication and

behaviour become more important in determining parental stress as their stay lengthens.<sup>54</sup> Families find strength in their role as advocates for their child.<sup>55</sup> The transition from acute to chronic care also requires the implementation of routines for exercise, mobility and communication devices, as well as attention to indicators of anxiety in the patient and family, sometimes requiring referral to psychological services.<sup>56</sup>

Parents adopt different roles when faced with a stressful admission.<sup>54</sup> Transition to home and then ongoing care in the home demand considerable family and professional caregiver support.<sup>57</sup> Parents are expected to care for their child with disability at home and most are not offered an option or alternative.<sup>58</sup> Undertaking this care at home creates a radically different model of parenting and family life to that which they may have had before.<sup>53</sup> The impact on siblings also needs to be considered and should not be underestimated.<sup>52,59</sup>

For the child to be cared for at home, the parents must take on the challenges of their child's health needs including roles of nurse, manager and technician required to fulfil this task.<sup>58</sup> These sometimes contradictory roles can challenge parents to their physical and psychological limits. Summaries of parents' experiences include recurrent themes of loss of privacy and authority that change the meaning of home, dependence for financial support and decision-making, social isolation for many parents (especially mothers), a requirement to develop coping strategies for dealing with an inherently unstable situation and confronting the moral dilemmas posed by the situation.<sup>17,60</sup>

### Risk minimization

A child who is dependent on assisted ventilation and their family lives with a level of risk that others do not. Discharge from hospital comes with additional risks, and parents need to be clear that the benefits of home care outweigh these risks.

The risk management considerations of the ventilator-dependent patient at home have been highlighted previously and are reproduced in Table 3 (with permission from Simonds).<sup>61,62</sup> Additional considerations when preparing for transition to home are shown in Table 4.

Discharge from hospital in staged fashion allows parents and healthcare teams to troubleshoot any gaps in the care plans, equipment or home set-up. Close liaison with the child's regional medical team is imperative, particularly when families live at a distance from the tertiary hospital. Training of local staff and/or transfer to a regional hospital may be required prior to discharge home.

### Equipment requirements

The requirements listed are in addition to any equipment needed for the care of a tracheostomy itself, for which the reader is referred to the American Thoracic Society Guidelines for the Care of a Child with a Tracheostomy.<sup>5</sup> Prior to discharge, a child ventilated via a tracheostomy should be

stabilized on their home equipment. Table 5 summarizes the recommended minimum equipment requirements for children dependent on ventilation.

## 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 precise location will vary between hospitals but a high dependency area or transitional care unit near to or within the paediatric intensive care unit (PICU) is ideal.

### *Funding*

The working group noted major disparities (and often inadequacies) in the provision and timing of funding provided to children needing LTV, suggesting a need for this to be documented for Australia and New Zealand.

## (B) Sleep life support

This section addresses the needs of infants, children and young people whose high ventilatory needs are supported via a non-invasive interface. Other international guidelines/standards recognize that the 'interface' used does not distinguish those with high needs or risks *per se*, and a proportion of those using NIV require the same levels of training, support and planning as those using invasive ventilation, particularly when ventilation cannot be discontinued for any period during sleep without risk to life. A small but increasing number of children with complex multi-system disorders are dependent on non-invasive ventilatory support and individuals may transition between invasive and non-invasive interfaces over time.

Defining 'dependency' and 'need' for ventilatory support is a continuum. For our purpose, dependency applies to those at risk of serious or irreversible harm if their ventilatory support was discontinued or malfunctioned for a 24-h period. As with the invasively ventilated children described above, this spectrum is best defined through an individualized assessment by the clinical team involved.

## Existing guidelines

No specific standards/guidelines for high dependency non-invasive ventilatory support were identified in our literature search, including grey literature from relevant national and international organizations and societies, apart from one document specific to management of children with central congenital hypoventilation syndrome.<sup>70</sup> However, several standards/guidelines acknowledge that those supported with a non-invasive interface may have needs as high as those with a tracheostomy and combined these groups in their recommendations.<sup>47,71</sup>

## Practical issues

### *Considering the options*

A tracheostomy remains the option of choice for children who require ventilation >16 h/day.<sup>70,72–74</sup> However, a small but increasing number of children who are dependent on ventilatory support are managed with NIV.<sup>11</sup> This option avoids the potential morbidity/mortality of a tracheostomy and offers advantages to the child and family. The choice of invasive or NIV may be made at diagnosis (including for infants), or a child may be transitioned to NIV from tracheostomy ventilation as their condition improves or when age or other factors influence this choice. These decisions should be made in conjunction with the parents and, if appropriate, with the child, recognizing that the optimal interface may change over time.

Decisions around the choice of interface are typically based on multiple factors including the child's:

- Overall therapeutic goals.
- Age and development.
- Ventilatory requirements including aspects such as wake, sleep and hours per day.
- Ability to cope with planned periods off ventilatory support.
- Upper airway function and security.
- Bulbar function.

### *Indications*

NIV is ventilatory support delivered via a nasal mask or similar non-invasive interface. NIV is an option (above invasive support) when the goals of ventilation can be safely and effectively achieved without tracheostomy. This includes symptom reduction, improved quality of life, improved pulmonary development, reduced complications, reduced hospitalization, prolonged life and 'life support'. The potential adverse consequences of a mask interface are less than tracheostomy, and NIV can be undertaken on a trial basis.

Children in whom NIV is indicated may have a congenital or acquired underlying medical condition. They generally fall into three main groups: neuromuscular disease (NMD), central hypoventilation and severe respiratory insufficiency.

### *Contraindications*

NIV is contraindicated or not practical for long-term home use:

- In the absence of a secure airway during wakefulness.
- If NIV fails to normalize ventilation in a child whose underlying condition is considered reversible or non-progressive.
- If ventilation is required more than 16 h per day, tracheostomy should be considered due to the side effects and practicalities of delivering NIV for this time period.<sup>70,72–74</sup>

### *Funding packages*

Funding packages for those with high-dependency non-invasive ventilatory support should be based on those for

**TABLE 3** Risk management considerations (adapted with permission from Simonds et al.<sup>61</sup>)

| Problem                                                     | Consequence                                         | Risk management                                                                                                                                                                                                                                                                                                                                                        |
|-------------------------------------------------------------|-----------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Power failure (e.g., power cut)                             | Ventilator failure                                  | Mains power alarm. Battery backup (internal or external battery in circuit). Ambu bag with connector for ventilator-dependent patient                                                                                                                                                                                                                                  |
| Ventilator malfunction                                      | Ventilator failure                                  | Regular service and planned preventative maintenance. Emergency contact line to report problems. Back-up ventilator in patients with less than one night ventilator-free time                                                                                                                                                                                          |
| Accidental disconnection                                    | Failure of ventilation                              | Low pressure and low minute volume alarms. Secure attachment of ventilator/circuitry connections. Support of ventilator tubing to prevent dragging on tracheostomy or mask                                                                                                                                                                                             |
| Circuit obstruction                                         | Failure or suboptimal ventilation                   | High pressure alarm                                                                                                                                                                                                                                                                                                                                                    |
| Mask fit                                                    | Too tight: pressure sore<br>Too loose: leaks        | Close attention to mask fit, variety of interfaces to alternate. Skin protective dressing. Low pressure, low minute volume alarms                                                                                                                                                                                                                                      |
| Tracheostomy blocked                                        | Failure of ventilation                              | Efficient suction with battery power or manual operation. Carers trained to change tracheostomy. Effective humidification. High pressure alarm                                                                                                                                                                                                                         |
| Tracheostomy falls out or cannot be replaced after changing | Failure of ventilation                              | Improve fixation of tube. Carers trained to change tube, smaller size tracheostomy tube available. Ambu bag and mask are available, which carers can use                                                                                                                                                                                                               |
| Medical problems                                            | Acute deterioration, settings no longer appropriate | Rapid access to advice. Carers and patients trained to recognize early signs of chest infection or ventilatory decompensation. Carers trained in basic life support. Immediate access to hospital care. Battery-powered ventilator for use in transit                                                                                                                  |
| General                                                     |                                                     | Patients and carers competent in ventilator operation, basic maintenance, problem solving and when to seek help. Written plan of action for predictable problems such as power cuts, chest infection and equipment failure. Equipment problems notified to manufacturer and central agency where available (e.g., Medicines and Healthcare Products Regulatory Agency) |

children on tracheostomy ventilation. The working group noted that this group often receive less funding due to not having a tracheostomy, even when their care needs are equivalent.

## Management recommendations

Specific issues that apply to high-dependency NIV include:

### *Interface choice, fit and care*

The non-invasive interface may not be simpler or safer than a tracheostomy. The type and size of mask must be appropriate and requires ongoing assessment/monitoring, particularly with facial growth. Any interface has a limited lifespan and will need regular checks and replacement. Specific risks may have equivalent life-threatening implications to a tracheostomy. These include partial mask displacement occluding an infant or child's airway, impaired clearance of airway secretions and/or vomit, imposition of additional dead space or rebreathing if the mask or ventilator is not operating correctly, risk of skin pressure ulcers and abnormal growth of the mid-face, due to long-term pressure from the mask. Carer training is vital to minimize these risks. Mask pressure effects can be reduced by the use of multiple masks used on a rotating schedule, mouthpiece ventilation for daytime use and so on. The application of adhesive pads and/or cushioning solutions may also be beneficial.

### *Ventilator*

The ventilator must be appropriate for NIV, with appropriate alarms and power failure provision, to accommodate the child's ventilatory needs, age and size.

### *Back up*

As with tracheostomy ventilation, a back-up device, circuit and mask must be available within an appropriate timeframe.

### *Infection control*

Attention to appropriate infection control procedures is also recommended, including mask care. Further infection control measures relevant to all patients on home ventilation are discussed in section on Infection control.

## **PART TWO: VENTILATORY SUPPORT FOR OPTIMIZATION OF A CHILD'S HEALTH: INCLUDING BPAP AND CPAP**

Children for whom ventilatory support has been prescribed to normalize ventilation during sleep 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.<sup>74-77</sup> Demonstrated benefits include decreased incidence of hospital admission,<sup>75</sup> improved quality of sleep, decreased daytime

**TABLE 4** Recommended requirements for discharge home

| Recommended requirement                                            | Details                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                       |
|--------------------------------------------------------------------|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| 1. Medical stability for discharge                                 | <ul style="list-style-type: none"> <li>The child should have been using the ventilator that will be used at home for a sufficient period to establish that the settings are appropriate to achieve ventilatory support goal<sup>3,63</sup></li> </ul>                                                                                                                                                                                                                                                                                                                                                                                                                                                         |
| 2. Home visits by nursing staff and/or other team members          | <ul style="list-style-type: none"> <li>Should occur early in the discharge process to assess the home environment in relation to the new needs of the child—inappropriate home environments are a frequent cause of delayed discharge for children with complex healthcare needs<sup>1,8,26</sup></li> <li>Ventilation equipment requires considerable space and access to power outlets—home modifications to meet equipment specifications and workplace standards may be required. Parents may need support to find alternative housing</li> </ul>                                                                                                                                                         |
| 3. Parent/caregiver education                                      | <ul style="list-style-type: none"> <li>Parent willingness and capacity to undertake complex medical care at home should be assessed early in the admission</li> <li>Parents are required to have completed a standardized education programme and have demonstrated proficiency and confidence in all aspects of their child's care</li> </ul>                                                                                                                                                                                                                                                                                                                                                                |
| 4. Ventilator maintenance plan                                     | <ul style="list-style-type: none"> <li>A programme for regular maintenance and replacement of ventilator equipment is recommended</li> </ul>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                  |
| 5. Daily care plans, action plans and risk minimization strategies | <ul style="list-style-type: none"> <li>It is recommended that these are individually written for each child</li> <li>Action plans for respiratory exacerbations after initial discharge are recommended and should be regularly reviewed (e.g., instructions on how to arrange urgent medical review, changes to ventilatory settings and addition of oxygen)</li> </ul>                                                                                                                                                                                                                                                                                                                                      |
| 6. Emergency plan                                                  | <ul style="list-style-type: none"> <li>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 is recommended</li> <li>An ambulance plan should be organized. 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 ambulance officers. This emergency plan should be discussed and practiced with the family and carers prior to discharge</li> </ul> |
| 7. Power supply plan                                               | <ul style="list-style-type: none"> <li>The relevant power supply company should be contacted and advised of the patient's medical condition, and priority requested for power restoration in the case of power outage and for power not to be removed in the absence of bill payment</li> <li>A back-up plan for power outage should be included in the discharge plan, availability of back-up battery support, emergency contacts and consideration of a generator, depending on distance to closest hospital</li> </ul>                                                                                                                                                                                    |
| 8. Trained carer availability                                      | <ul style="list-style-type: none"> <li>Sufficient carers should be trained in all aspects of the child's care and available at the time of discharge</li> </ul>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                               |

Abbreviation: PICU, paediatric intensive care unit.

sleepiness, increased sense of well-being, improved concentration and reduced incidence of morning headaches.<sup>78–80</sup>

## Existing guidelines

No guidelines were found for the general management of children on NIV, although specific guidelines address certain diagnostic groups, highlighting issues in these populations (e.g., for Duchenne muscular dystrophy [DMD]<sup>81–83</sup> and for the respiratory management of children with neuromuscular weakness).<sup>84</sup> Reports from centres involved in commencing children on NIV<sup>9,28–30,76</sup> highlight the need for paediatric expertise, the use of behavioural modification approaches and allied health support to improve adherence to therapy, as well as parental commitment for successful initiation.

## Practical issues

### Considering the options

Non-invasive support extends survival and improves quality of life in patients with progressive neuromuscular conditions

like DMD.<sup>75,77,78</sup> Children who would potentially benefit from NIV should have access to a paediatric facility with tertiary respiratory and sleep medicine services, enabling informed discussions regarding the advantages and limitations of ventilatory support, including escalation of support during a significant respiratory infection,<sup>42,77</sup> at the time of spinal surgery<sup>85–87</sup> and towards the end of life.<sup>44,88</sup>

## Indications

### *Bi-level positive airway pressure*

Children with hypoventilation during sleep, even if isolated to rapid eye movement sleep, benefit from NIV.<sup>78,80,89–91</sup> The presence and severity of hypoventilation during sleep is best established by PSG. Indications for long-term ventilatory support with NIV include hypoventilation secondary to:

- NMD.
- Chest wall restriction (bony abnormalities, etc.).
- Chronic lung disease.
- Severe upper airway obstruction not adequately treated with CPAP.
- Obesity hypoventilation.

**TABLE 5** Recommended equipment requirements for children dependent on ventilation

| Recommended requirement                                                                                    | Details                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                      |
|------------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| 1. A portable ventilator, suitable for home use in children                                                | <ul style="list-style-type: none"> <li>One or two preferred ventilators are recommended, so that workers are familiar with the capabilities, limitations and operation of those ventilators</li> <li>Summaries of different ventilators are available<sup>64–66</sup></li> <li>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</li> <li>Heated humidification is essential for tracheostomy ventilation<sup>67,68</sup></li> <li>Ventilators should be regularly maintained according to the manufacturer's specifications and documentation made of maintenance and repair</li> </ul> |
| 2. Alarms should be available to alert the carer to disconnection, decannulation or tracheostomy occlusion | <p>The following built-in alarms are recommended:</p> <ul style="list-style-type: none"> <li>Low tidal volume (unless the patient's tidal volume is less than the minimum detectable tidal volume for the ventilator used)</li> <li>Low minute ventilation</li> <li>High pressure</li> <li>Disconnect (low pressure),<sup>69</sup> noting that this alarm may fail to detect decannulation with small tracheostomy size (alarm function should be evaluated with the child's specific circuit and tracheostomy size)</li> <li>Power failure</li> </ul>                                                                                                                                                                                                       |
| 3. A sufficient alternate power source as appropriate to the child's setting                               | <ul style="list-style-type: none"> <li>This may be a battery or generator</li> </ul>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                         |
| 4. Hand ventilation equipment for manual ventilation                                                       | <ul style="list-style-type: none"> <li>Will be needed in the event of mechanical or electrical failure</li> <li>It may also be required for use away from home in children who do not require 24-h ventilation but may need support unexpectedly</li> </ul>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                  |
| 5. An alternative ventilator                                                                               | <ul style="list-style-type: none"> <li>For use in the event of a mechanical failure</li> <li>Where this ventilator is kept will depend on the geographical location of the patient and the accessibility of services to provide an alternative ventilator at short notice</li> <li>In most cases, a second ventilator will need to be kept in the child's home</li> </ul>                                                                                                                                                                                                                                                                                                                                                                                    |
| 6. Oxygen saturation monitor                                                                               | <ul style="list-style-type: none"> <li>Ventilator alarms should not be relied on alone, and continuous oximetry monitoring is recommended to augment carer observation and ventilator alarms, in order to facilitate early recognition of a problem</li> </ul>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                               |
| 7. Portable suction                                                                                        | <ul style="list-style-type: none"> <li>For all tracheostomy ventilated patients</li> </ul>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                   |
| <b>Additional equipment to be considered depending on individual patient needs</b>                         |                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                              |
| 1. Oxygen concentrator or cylinders                                                                        | <ul style="list-style-type: none"> <li>Capable of providing sufficient oxygen flow for emergency use</li> </ul>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                              |
| 2. Mechanical insufflation–exsufflation device                                                             |                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                              |
| 3. Nebulizer equipment                                                                                     |                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                              |

### *Continuous positive airway pressure*

The largest group of children requiring CPAP have obstructive sleep apnoea (OSA), many of whom have significant medical co-morbidities,<sup>9,28–30</sup> which need to be considered when planning support for the home. Indications for long-term ventilatory support with CPAP include:

- OSA not successfully treated with surgical options such as adenotonsillectomy or in children where surgery is contraindicated.
- Craniofacial and airway abnormalities.
- Chronic lung disease.

### Initiation of therapy

Initiation of BPAP/CPAP is usually not urgent and can be instigated through a planned process, including staged approach of mask fitting as an outpatient  $\pm$  a period of

acclimatization to the mask at home, to enhance the child's acceptance of the treatment in the long term.<sup>29</sup> BPAP/CPAP may be initiated in a hospital ward, PICU, sleep laboratory or even at home. Regardless of the location, ventilatory settings and equipment need to be individualized for each child by trained paediatric staff. Compliance with these therapies is enhanced in units with skilled paediatric staff and appropriate behavioural management programmes.<sup>29,30</sup>

### Monitoring progress of therapy

Close follow-up is recommended following the initiation of therapy, particularly over the first month.<sup>92</sup> Telephone contact from experienced staff can support parents with maintaining therapy at home. Outpatient clinic visits should include a review of interface, overall comfort as well as general equipment checks. Objective assessment of adherence is now possible through digital download of data or remote

monitoring and is useful for identifying suboptimal adherence. The frequency of formal monitoring with PSG depends on many factors but is generally recommended at 6–12-month intervals.

## Adherence to therapy

The challenges associated with maintaining NIV therapy in children are increasingly recognized especially for children with developmental delay, anxiety or behavioural problems.<sup>93</sup> What exactly constitutes good or adequate adherence to therapy continues to be debated. Following early adult studies that suggested average CPAP use was 4.7 h/night, CPAP use of 4 h/night of 70% of nights was established as a clinical and empiric benchmark of CPAP adherence.<sup>94,95</sup> However, such levels of therapy may not promote all health and functional outcomes, and the cut off for adequate levels of therapy is still to be determined in paediatrics, given the higher sleep requirements of children.<sup>96</sup> Regardless, studies in children have consistently found low levels of usage. Marcus et al. found that adherence averaged only  $3.8 \pm 3.3$  h per night.<sup>27</sup> O'Donnell et al. and Nixon et al. reported similar suboptimal adherence, with mean usage of 4.7 h per night.<sup>29,92</sup> Identification of specific factors affecting adherence to therapy and the impact of potential behavioural interventions continue to be explored.<sup>93,94,97–100</sup>

## Funding and service provision

Appropriate funding for diagnosis, equipment and long-term management is important for NIV, regardless of the child's age, and should be equitable across regions.

## Management recommendations

### Family support

The care and supervision needs of children on ventilatory support should be eligible for government financial assistance (e.g., Child Disability Allowance). The need for home carers will depend on the co-morbidities/associated disabilities of the child. Respite care should also be considered.

### Home ventilation teams

Staff experienced in using NIV can support successful initiation and ongoing care of these children, providing access to the specialized advice needed to ensure rapid rectification of problems. Teams with processes to ensure adequate follow-up of all children on ventilatory support, both for clinical review and review of ventilation settings (with polysomnography if appropriate), will make sure that changing needs are anticipated and addressed.<sup>1,26,101,102</sup>

## Risk minimization

NIV therapy for health optimization is usually low risk. Parents require education on how to anticipate, avoid 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 prioritize and seek assistance in the event of equipment failure (resulting in timely replacement) and have a documented action plan. Regular equipment servicing according to the manufacturer's recommendations is essential.

## Children receiving CPAP or BPAP via tracheostomy

A small group of children that require special mention are those who may require a tracheostomy for anatomical reasons (upper airway obstruction) but who also receive CPAP or BPAP support for health optimization through this rather than through a non-invasive interface. The presence of a tracheostomy adds complexity to their management and they may require care equivalent to those in the high-dependency groups.

## Equipment requirements

The most problematic issues for NIV in children are achieving low dead space and appropriate triggering sensitivity. Ideally, centres should have one or two preferred machines, so that staff are familiar with the capabilities, limitations and operation of each. 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.<sup>67</sup>

When prescribing NIV for infants and children in high-risk groups (include those under 5 years of age and children with mobility difficulties restricting mask removal), monitoring at home should be done appropriate to the specific child's vulnerabilities, regardless of the model of ventilator used. Monitoring may include oximetry or cardiorespiratory monitoring, disconnect, pressure (low or high) and/or power failure alarms. High-risk infants and children may also warrant use of ventilators with higher specifications to ensure appropriate alarms (e.g., use of a bi-level machine in CPAP mode for additional alarms to be available).

Masks designed specifically for children are ideal, with low dead space and an appropriate size and shape to limit leak and minimize pressure marks on the face. All interfaces require regular review and adjustment as children grow. Full face masks covering the mouth should be avoided in infants and very young children, children with frequent vomiting and children unable to independently remove the mask or seek assistance in the case of overnight vomiting or other emergencies, due to risk of aspiration. If a full-face mask or

total face mask is used (covering the mouth), it must be fitted with an anti-asphyxia valve.

## Key challenges

### Readmission to hospital

Staff caring for children on ventilatory support require specific training in the use of ventilation equipment. Children on NIV should be advised to take their equipment with them for any hospital admission, regardless of the reason for hospital admission.

## PART THREE: VENTILATORY SUPPORT FOR CHILDREN WITH LIFE-LIMITING CONDITIONS RECEIVING PALLIATIVE CARE

### Introduction

NIV is increasingly being used in children with complex life limiting conditions during acute illnesses and these children may demonstrate a need for ongoing ventilatory support. A specific cohort of these children are receiving end-of-life care.

### Clinical context

Advances in medical care have led to a cohort of children with complex medical needs, who may develop the need for ventilatory support during acute illnesses, with some children benefiting from NIV when being discharged for end-of-life care.

Close liaison between the primary team, intensive care, palliative care and respiratory/sleep ventilation teams needs to be established early, to facilitate discussions regarding appropriateness and feasibility of ventilatory support as part of the palliative care plan. The goal is to facilitate discharge of the child from an acute hospital setting to a domiciliary setting.

This cohort of children are an evolving group, where a clear understanding of the goals of therapy is needed to avoid increasing the burden of care by using NIV. A subgroup of children receiving end-of-life care have a life expectancy that extends beyond a few weeks. The nature and burden of home ventilation in this cohort need to be carefully considered, with plans for treatment escalation and/or agreed limits of care. The funding considerations for this group of children include equipment for home ventilation and extra support for the family in relation to the increased burden of care.

## PART FOUR: ISSUES COMMON TO ALL GROUPS

Ventilatory support is a highly specialized treatment that should always be commenced and followed up with involvement of a tertiary specialized paediatric service.

## Medical review

A multidisciplinary clinic can help 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, for example, two to three monthly for children highly dependent on ventilation and three to six monthly for those on NIV for health optimization. It is likely to be more frequent directly after the initiation of ventilatory support and decrease with time. The review should include assessment of adherence (subjective and objective<sup>27</sup>) and complications of therapy. Clinical review should be coordinated with appropriate investigations (e.g., lung function testing) and other specialist assessments if indicated. Vaccination against respiratory pathogens should be offered.<sup>103,104</sup>

Tertiary paediatric centres should have a designated team for children on ventilatory support. Each child should have a case manager responsible for coordinating follow-up within a multidisciplinary framework, to ensure the changing needs of the child and family are anticipated and addressed. Additional to this, an overall care coordinator is recommended for children receiving LTV, to work with and guide the team process and tasks while building collaboration with all parties involved.

Follow-up physiology studies are recommended at regular intervals. The nature of those studies (Level 1–4 sleep studies) will depend on the child's health condition, their ventilatory support and their stability over time, and is best determined by their supervising sleep specialist.<sup>101,102</sup>

Changes to ventilatory settings are usually required with increasing age and growth or due to changes in the underlying condition of the child but vary annually for children stable on CPAP for OSA, to more frequently for those early in the course of treatment or in children with an unstable medical condition.

Checking of ventilatory 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.

## Infection control

It is recommended that infection control procedures at home are aligned with hospital practice, which should be continually reviewed appropriate to local circumstances. Families require education on infection control measures, to ensure a good understanding of the importance of this aspect of their child's care. For all patients, manufacturer guidelines should be followed regarding cleaning/disinfection of equipment. Chronic colonization with highly resistant organisms can occur in patients on LTV, particularly following prolonged hospitalization. Families should be made aware of the additional measures that may be required

in this instance. NIV (CPAP/BPAP) increases the dispersal of aerosolized droplets and therefore may require special considerations to avoid risk of transmission of respiratory viruses. Specific measures should be guided by hospital policy and be practical and easy to undertake at home. Information regarding any changes to local infection control policies needs to be regularly communicated to both families and carers. There are significant risks to ceasing respiratory support without seeking expert advice. Therefore, in the context of infection risk, these decisions should always be made in consultation with the treating respiratory/sleep paediatrician.

## Psychological support

The need for home ventilatory support places a substantial burden on the child or adolescent patient and their family, including parents and siblings.<sup>15–19</sup> The nature of the social, emotional, developmental, educational and practical challenges faced by children and parents vary.

The provision of in-home support workers often adds to the burden of care impacting family privacy and functioning. Psychological preparation of families and ongoing training of support workers can help to reduce stress for all concerned and minimize the risk of breakdown in these critical relationships.

Psychological support may be required at any phase of the illness process and additionally as the child moves through various key developmental stages including commencing school, adolescence and transition to adult care and independence.

Psychological support should be available and offered to children, parents and siblings. Psychology may also have a role in the training of support workers, given the complexity of their in-home role, and in the provision of secondary consultation and support for all members of the home ventilation team.

## Ethics of paediatric home ventilation

Evidence shows that home mechanical ventilation is a cost-effective method to deliver the therapy, optimizes use of healthcare resources, provides mental and emotional benefits for the patients and their families and that patients experience increased independence.<sup>105</sup> Home ventilation is effective, but it imposes significant burden on families, particularly those of children dependent on ventilation for life support. The burden of care is also affected by the duration (per day) of therapy required, other care needs of the patient and the child's tolerance of therapy balanced against the parent's conscientiousness in applying it.

Apart from the burden of care imposed on the families, ethical challenges also surround the decision of who should be offered the therapy while also maintaining respect for the family and patient's wishes, and quality of life.<sup>106</sup> Equity of access to therapy is important and is acknowledged to be an issue worldwide. The level of support provided at home

varies widely, depending largely on the availability of local resources. Ethical challenges can arise at all points of changing therapy, whether commencing more intensive therapy or ceasing support. The cessation of support is an issue covered most extensively in the literature pertaining to the ICU, but it is also likely to be faced by teams who provide ventilatory support at home. Evaluation of the 'best interests of the child/family' or the balance of relative benefits against harm is important, in particular balancing beliefs about duration versus quality of life particularly in children where many have significant co-morbidities that the family may already have dealt with for long periods of time.<sup>59</sup> This dilemma can compound, or be compounded by, the resources required to manage the home ventilatory support, where the decision to provide home therapy may implicitly demand admission to the ICU during any acute illnesses.

## Transition from paediatric to adult care

Long-term ventilatory support is associated with an increase in survival in a range of conditions leading to ventilatory failure in children,<sup>107–109</sup> resulting in increasing numbers of such children surviving to adulthood and transitioning to adult services. Evidence suggests that insufficiently managed, non-standardized transition of children requiring ventilatory support to adult care increases the risk of adverse outcomes.<sup>110–112</sup> Many reviews discuss the medical transition process from paediatric to adult care and the reader is referred to consensus statements on healthcare transition for young adults with complex and chronic healthcare needs.<sup>113–115</sup>

Transition to adult-orientated health services should be a formally planned and coordinated process, which begins early in adolescence. The choice of primary adult physician and ventilatory support service will depend on the underlying diagnosis, the locality of services and the wishes of the patient. During transition, health care for adolescents on ventilatory support may need to address ethical decisions such as need for ongoing or escalating ventilatory support and end-of-life decisions.<sup>116</sup> The timing of transition can be especially difficult in adolescents with a progressive disorder who are nearing the terminal stages of their disease. Ideally, the transfer process should involve close collaboration between treating paediatric and adult teams and should include one of the following care models: paediatric clinicians attending adult clinics, adult clinicians attending paediatric clinics or attendance by both at a planned 'handover' clinic with the young person and her/his family or carers.<sup>117,118</sup>

## Workforce issues/manpower planning

The initial treatment of children requiring home ventilatory support should ideally be within a tertiary paediatric service with respiratory paediatricians and intensivists. Continuing increase in the numbers of children receiving ventilatory support at home will necessitate increase in the capacity of healthcare

systems to deal with the demands for appropriate diagnostic services, medical and nursing support.

Once home, the day-to-day care of these children may rest with a regional general paediatrician and local allied health professionals<sup>1</sup> who may have very little experience or expertise in ventilatory support therapy. Geographical distances from a tertiary centre can be large and educational packages, teleconferencing, telephone help and written action guidelines for families and local teams help to ensure that therapy is not compromised. Including local paediatricians in the planning stage will help them to provide adequate support and education to families.

## Travel and fitness to fly

Children on ventilatory support frequently seek to access air travel. Long flights where the child may need to sleep may necessitate a fitness to fly test.<sup>119–122</sup> Families need to be informed, ideally at the outset, that airline travel can be difficult, especially internationally, with 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. Travel can be facilitated by contacting the medical officer of the airline early for information specific to the airline. The family will require appropriate carry bags to protect equipment and the correct electrical adapters for the countries being visited.

## WHAT IS NEEDED IN THE FUTURE?

- Research should focus on improving the evidence base for current models of care.
- Further development of education packages for parent/family is required.
- Appropriate funding is needed to set up and maintain an Australian and New Zealand registry of patients on home ventilatory support. This is required:
- To facilitate equitable standards of care.
- To inform future recommendations and assist in resource allocation planning.
- To liaise with manufacturers to improve equipment options for children.

## CONCLUSIONS AND KEY MESSAGES

Children with chronic respiratory failure vary in their level of dependence on mechanical ventilation. Policy makers and practitioners at all levels can help to ensure that their rights are respected and their needs met. Coherent, multi-disciplinary and inter-agency planning is required to discharge these children from hospital to home, particularly those dependent on ventilation. The use of ventilatory support adds a significant burden of care to parents and families caring for these children, so that provision of appropriate

carer support and respite care is crucial to the well-being of the child and family.

The levels of support for care at home for children on LTV support are highly variable across Australia and New Zealand in both our previous and current surveys of specialists involved in delivery of home ventilation programmes. The establishment of a multi-centre working group and patient registry will help to advocate for equity of access across Australia and New Zealand.

### SUMMARY STATEMENTS

- The Thoracic Society of Australia and New Zealand (TSANZ) and Australasian Sleep Association (ASA) support the equitable access to diagnostic and therapeutic options for children who require ventilatory support.
- The TSANZ and ASA support the care of technology-dependent children in their own home.
- Implementation of a standard template for surveillance and audit of this increasing/expanding patient cohort of children who require ventilatory support is recommended to facilitate cross-centre collaboration and service enhancement.
- The establishment of a multi-centre working group and a patient registry should be undertaken to inform individual patient care and policy development.

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### CONFLICT OF INTEREST

Andrew Tai (chair), Jasneek Chawla, Amanda L. Griffiths, Sadasivam Suresh, Jacob Twiss, Moya Vandeleur, Karen A. Waters, Andrew C. Wilson and Susan Wilson have no conflicts of interest to declare. Elizabeth A. Edwards' spouse is an employee of Fisher and Paykel Healthcare, and Gillian M. Nixon's spouse is an employee of Bird Healthcare.

### ORCID

Jasneek Chawla  <https://orcid.org/0000-0001-5035-0744>

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## SUPPORTING INFORMATION

Additional supporting information may be found online in the Supporting Information section at the end of this article.

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