Vol. 3(4), 155-165, 2025

PEDIATR RESPIR J

REVIEW

Long-term ventilation in children: state of the art and future perspectives

Mihail **Basa** [1], Aleksandar **Sovtic** [1, 2, *]

[*] **Correspondence to:**
adsovtic@gmail.com

Doi
10.56164/PediatrRespirJ.2025.82

[1] Mother and Child Health Institute of Serbia, Belgrade, Serbia
[2] Faculty of Medicine, University of Belgrade, Serbia

ABSTRACT

Home mechanical ventilation (HMV) has transformed the care of children with chronic respiratory failure, improving survival, reducing hospital dependence, and enabling better quality of life. The choice between invasive and noninvasive modalities must be individualized, but successful outcomes universally depend on structured initiation, careful parameter adjustment, and systematic follow-up. Local practices, health system organization, reimbursement policy, and home care provider availability have significant influence on HMV successes. Future advances will rely on technology, multidisciplinary expertise, and broader access to specialized home-care services.

IMPACT STATEMENT

This review provides a comprehensive and up-to-date overview of long-term home mechanical ventilation in children, integrating current evidence with practical clinical considerations. It aims to support clinicians in decision-making, optimize patient selection and ventilatory strategies, and guide future development of pediatric home ventilation programs.

KEY WORDS
NIV; IMV; HMV in children; chronic respiratory failure; home care.

INTRODUCTION

Chronic respiratory failure (CRF) is a major cause of morbidity and mortality in both adult and pediatric populations. Advances in diagnostic and therapeutic care have enabled survival in many patients with conditions once deemed untreatable or incompatible with long-term life (1, 2). Consequently, the number of children living with CRF and requiring long-term assisted ventilation has steadily increased. The introduction of home mechanical ventilation (HMV) has opened new possibilities for comprehensive care, while simultaneously reducing the burden on healthcare systems and facilitating social integration for some of the most vulnerable children. Although HMV has been available for decades, continuous technological innovation in ventilator design and functionality, along with the advent of disease-modifying therapies, has made this field highly dynamic and constantly evolving (1, 2).

HMV is typically delivered through two modalities: invasive mechanical ventilation (IMV) via tracheostomy and non-invasive ventilation (NIV) using an appropriate interface such as a nasal or oronasal mask. The choice between these approaches largely depends on the underlying disease, the degree of respiratory dysfunction, and the individual patient's characteristics (1, 2).

DIAGNOSIS OF SLEEP-DISORDERED BREATHING (SDB)

Diagnostic procedures for early detection of sleep-disordered breathing (SDB) are recommended in all children with chronic, stable medical conditions that increase the risk of SDB, regardless of the presence of symptoms (3-5). In otherwise healthy children, diagnostic evaluation is warranted when clinical signs suggest SDB (3-5). The gold standard for early detection is video-polysomnography (PSG) combined with continuous non-invasive transcutaneous capnometry during sleep. In resource-limited settings, respiratory polygraphy may serve as an acceptable alternative (6).

When advanced diagnostics are unavailable, morning arterial blood gas analysis (ABG) and overnight oximetry trends can provide useful additional information (7, 8). However, elevated $PaCO_2$ (>50 mmHg) in morning samples usually indicates a late manifestation of chronic respiratory failure. A difference in $PaCO_2$ >10 mmol/L favoring the morning sample strongly suggests alveolar hypoventilation. Elevated serum bicarbonate (HCO_3^- >28 mmol/L) reflects metabolic compensation but is not specific for respiratory acidosis; hence, these findings must always be interpreted in clinical context (7, 8).

Children at increased risk—including those with neuromuscular disorders, chronic primary lung diseases, severe obesity, craniofacial malformations, or impaired respiratory control—should undergo systematic evaluation as part of a standardized diagnostic protocol (2, 5, 6).

Long-term noninvasive ventilation (NIV) in children generally encompasses modes that provide ventilatory assistance, most notably bilevel positive airway pressure (BiPAP). Continuous positive airway pressure (CPAP), although frequently classified under the broader NIV umbrella due to its noninvasive interface, is not strictly considered a *ventilatory* modality, as it delivers a constant distending pressure without augmenting tidal volume or providing true ventilatory support. Accordingly, both BiPAP and CPAP may be indicated in children with chronic, stable conditions characterized by severe SDB and impaired gas exchange (**Table 1**), although their mechanisms of action and therapeutic objectives differ (5).

In pediatrics, severe SDB is typically defined by an apnea–hypopnea index (AHI) greater than 10 events per hour (6). Reference values differ significantly between adults and children, with pediatric thresholds being much stricter (**Table 2**).

Table 1. *Key differences between CPAP and BiPAP in pediatric home ventilation.*

Feature	CPAP	BiPAP
Pressure pattern	Constant, fixed pressure throughout the entire respiratory cycle	Variable pressures: higher inspiratory pressure (IPAP) and lower expiratory pressure (EPAP)
Indications	Primarily obstructive disorders (*e.g.*, OSAS)	Central, restrictive, and obstructive disorders
Mode of breathing	Spontaneous breathing only	Can support spontaneous breathing and provide backup ventilation if needed
Flexibility	Limited – single continuous pressure	Flexible – can adapt to more complex ventilatory needs
Transition	May be escalated to BiPAP if CPAP is insufficient	Typically used when CPAP fails or in more severe disorders

Table 2. *AHI thresholds for sleep-disordered breathing (SDB) in pediatric and adult populations.*

Severity of SDB	Pediatric population (AHI, events/hour)	Adult population (AHI, events/hour)
Normal finding	<1.5/h	<5/h
Mild SDB	1.5–5/h	5–15/h
Moderate SDB	5–10/h	15–30/h
Severe SDB	>10/h	>30/h

Criteria for nocturnal alveolar hypoventilation in children vary slightly depending on the source (5, 6). The European Respiratory Society (ERS) defines hypoventilation when gas exchange impairment is documented by any of the following:

- morning arterial $PaCO_2$ ≥50 mmHg on ABG, or
- transcutaneous CO_2 ($TcCO_2$) >50 mmHg for ≥2% of total sleep time, or
- oxygen saturation (SpO_2) <90% for >2% of total sleep time (5).

However, the American Academy of Sleep Medicine (AASM) applies a different threshold for scoring hypoventilation during sleep in children (6). According to pediatric AASM criteria, hypoventilation is scored when **>25% of total sleep time is spent with PCO_2 >50 mmHg**, measured either by arterial sampling or a validated surrogate. This definition is therefore more stringent in terms of required duration of hypercapnia compared with the ERS threshold of ≥2% TST with CO_2 >50 mmHg. In adults, AASM criteria differ further and define hypoventilation when either:

- PCO_2 exceeds 55 mmHg for ≥10 minutes, or
- PCO_2 increases by ≥10 mmHg from the awake supine value to a level >50 mmHg for ≥10 minutes.

These differing reference standards are clinically relevant, as certain patient populations—such as children with neuromuscular disorders—may require adapted $TcCO_2$ thresholds or additional indicators of respiratory compromise. A recent international expert panel (RIND study) proposed criteria for nocturnal hypoventilation in these patients, including $TcCO_2$ >45 mmHg for >25% of total sleep time, or $TcCO_2$ >50 mmHg for >2% of total sleep time or at least 5 continuous minutes (9). Importantly, initiation of nocturnal ventilation is not based solely on $TcCO_2$, but also considers functional parameters such as reduced forced vital capacity (FVC <50% predicted), weakened maximum inspiratory pressure (<60 cm H_2O), or daytime SpO_2 <95% (9). In Duchenne Muscular Dystrophy (DMD), these adapted criteria are particularly critical due to progressive weakness of the diaphragm and accessory respiratory muscles, which often leads to alveolar hypoventilation first occurring during sleep. Routine screening for nocturnal hypoventilation is recommended when FVC falls below 50% predicted and mandatory at ≤40%, with NIV initiated when these functional and $TcCO_2$ criteria are met or when clinical symptoms such as morning headaches or daytime fatigue appear (5, 9).

Although the proportion of children requiring invasive mechanical ventilation (IMV) has declined markedly in recent decades, it remains the therapy of choice for a subset of patients with the most severe phenotypes (7, 10). Typical candidates include children with global chronic respiratory insufficiency requiring ≥16 hours of assisted ventilation, as well as those with bulbar dysfunction in whom noninvasive modalities are insufficient or not feasible (7, 10).

Importantly, the decision between noninvasive and invasive home mechanical ventilation does not need to be final or immutable over time. For example, certain conditions—such as congenital central hypoventilation syndrome (CCHS) or severe forms of bronchopulmonary dysplasia (BPD)—may necessitate IMV during infancy or early childhood, yet decannulation and transition to NIV can be achieved later in selected patients (11, 12). Conversely, in progressive disorders where NIV initially provides satisfactory support, the clinical course may eventually require tracheostomy and initiation of IMV. The best way to make these complex decisions is within a multidisciplinary framework, always in close consultation with the family and caregivers.

INPATIENT INITIATION OF LONG-TERM HMV

Research indicates that the initiation of home mechanical ventilation (HMV) in children can be effectively managed in either inpatient or outpatient settings (13, 14). Traditionally, initiation of long-term NIV was undertaken in the hospital setting, where the process requires close collaboration between healthcare professionals, social services, parents, and the child. Traditionally, several days of inpatient training for both the patient and caregivers have been recommended. Although some healthcare systems with well-developed home-care infrastructure have reported successful home-based initiation, safety and efficacy in children remain insufficiently validated (13, 14). For this reason, inpatient initiation continues to represent the standard of care, particularly in resource-limited settings or in children with complex medical needs (15, 16).

In contrast, IMV is almost invariably initiated in the hospital, as it is most often required in children with complex underlying conditions (17). Hospitalization is typi-

cally longer than for NIV, reflecting both the severity of the primary disorder and the necessity of establishing a stable tracheostomy, which is essential for IMV. Preparation for discharge is also more demanding, as it involves not only caregiver training but also securing a wide range of supportive equipment, such as oxygen concentrators, suction devices, and other home-care aids. Collectively, these factors make the initiation of IMV considerably more resource-intensive compared with NIV (17, 18).

PATIENT SELECTION AND CLINICAL APPROACH TO LONG-TERM HMV

In children with obstructive sleep apnea syndrome type 1 (OSAS type 1), long-term nocturnal CPAP is indicated when symptoms and abnormal polygraphic findings persist after adenotonsillectomy. If residual symptoms remain following surgery, repeat polygraphy after 4–6 weeks is recommended, and CPAP should be initiated when the apnea–hypopnea index (AHI) exceeds 10 events per hour (19). The prevalence of obesity-related obstructive SDB (OSAS type 2) is steadily rising. In some of these children, dietary interventions and innovative pharmacological approaches may reduce body weight and lead to symptom resolution. In cases of severe SDB without alveolar hypoventilation, long-term CPAP therapy is indicated, accompanied by active nutritional management (19). The rarest yet most therapeutically challenging group includes patients with genetically determined craniofacial anomalies and upper airway malformations (OSAS type 3). These involve abnormalities of the maxilla and mandible, palate, tongue, or pharyngeal and laryngeal structures. In such cases, CPAP is indicated when a fixed level of positive pressure is sufficient to normalize breathing patterns and restore gas exchange (20-22).
Long-term BiPAP therapy at home is indicated during sleep and, when necessary, during the daytime in children with disorders characterized by alveolar hypoventilation (5). BiPAP with a backup rate is the first-line therapeutic option for alveolar hypoventilation due to:

- neuromuscular disorders,
- pediatric obesity hypoventilation syndrome (OHS),
- disorders of respiratory rhythm control in selected cases,
- advanced primary pulmonary diseases (*e.g.*, cystic fibrosis, interstitial lung disease, bronchopulmonary dysplasia).

In addition, BiPAP is recommended in two specific contexts: (i) in children with obstructive SDB who fail CPAP or cannot tolerate the high pressures required to maintain airway patency, and (ii) in those with OSAS type 2 (obesity-related) or OSAS type 3 (craniofacial anomaly-related) when alveolar hypoventilation is documented (5, 9, 19, 22).
By contrast, IMV is reserved for children in whom noninvasive modalities cannot ensure adequate ventilation or are not feasible. Certain conditions require IMV from the outset—most notably disorders of respiratory rhythm control, whether primary (*e.g.*, CCHS) or secondary (*e.g.*, sequelae of severe perinatal asphyxia, severe metabolic diseases) (11, 23, 24). Children with restrictive or mixed obstructive–restrictive ventilatory patterns, as seen in neuromuscular disorders, may also ultimately require IMV during acute decompensation of previously stable respiratory insufficiency, particularly when endotracheal intubation is necessary and extubation fails (25). In addition, neonates with generalized muscle weakness who cannot be weaned from the ventilator represent another group requiring early IMV (26).

VENTILATOR SETUP: DEVICE, CIRCUIT, AND INTERFACE

Devices for mechanical ventilation can generally be divided into two categories: intensive care unit (ICU) ventilators and those specifically designed for home mechanical ventilation (HMV). Home ventilators are typically smaller, more portable, and optimized for ease of use in a non-hospital environment (27).
The choice of ventilator type is guided primarily by the underlying indication. While many different models from various manufacturers are currently available, most share a core set of technical features and clinical functionalities. These common elements provide a framework for classifying pediatric HMV devices, as outlined in **Table 3** (28, 29).
The ventilator circuit is a critical, though sometimes underappreciated, component of the setup. Two main types are commonly employed: single-limb and dual-limb circuits (**Figure 1**) (30, 31).
In a **single-limb circuit**, a single tube serves both inspiratory and expiratory flow. Depending on how exhaled gas is eliminated to prevent rebreathing, two main configurations exist:

Table 3. Ventilators for pediatric home mechanical ventilation: technical features and clinical use.

Device type	Key features	Limitations	Typical use
Level 1 – Standard CPAP devices	Easy to handle, integrated humidifier	No battery or alarm; limited flow detection (13–30 kg); auto-CPAP usable >30 kg	OSAS without hypoventilation
Level 2 – Intermediate devices (BiPAP)	Support <16 h/day; integrated humidifier; medium size; basic alarms; battery (2–6 h); flow detection >2.5–5 kg	Limited backup capacity, not robust for continuous support	Children with SDB and hypoventilation, neuromuscular diseases, obesity-hypoventilation
Level 3 – Life-support ventilators	Full support (24 h/day); integrated alarms; longer battery (~8 h); precise flow detection (>2.5–5 kg)	More expensive; larger; external humidifier required	Children needing continuous ventilation, invasive ventilation via tracheostomy, medically fragile patients

- **Vented circuits**, in which the mask itself incorporates exhalation ports (intentional leak openings). In this design, a minimum PEEP of approximately 4 cm H_2O is required to ensure effective clearance of exhaled CO_2 and avoid rebreathing.
- Circuits with an active expiratory valve, in which the valve is positioned close to the patient. The valve opens during exhalation, actively directing expired gas out of the circuit and thereby preventing rebreathing.

Single-limb circuits are most often used for NIV, as they are simple, portable, and cost-effective. Nevertheless, they present limitations, including reduced accuracy in monitoring exhaled volumes, reliance on intentional leaks, and greater susceptibility to unintentional air leaks. By contrast, a **dual-limb circuit** employs separate inspiratory and expiratory tubes connected via a Y-piece. This arrangement allows more precise regulation of tidal volume and gas exchange, reduces the risk of rebreath-

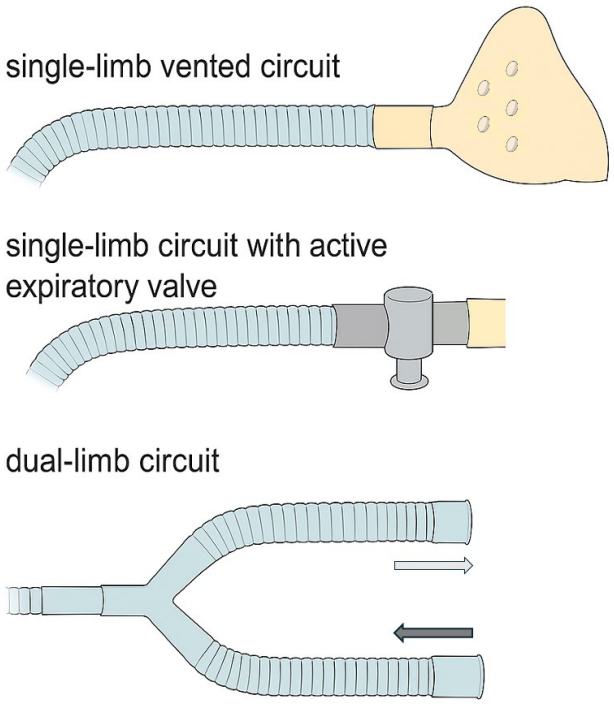

Figure 1. Types of ventilator circuits used in pediatric home mechanical ventilation.

ing, and facilitates accurate monitoring of exhaled volumes. Dual-limb systems are the standard for IMV via tracheostomy, but they may also be applied in NIV when precise volume monitoring is required. Their disadvantages include greater technical complexity, larger size, and reduced portability compared with single-limb systems (30, 31).

The choice of mask that best matches the child's long-term ventilatory requirements and facial anatomy at different ages is a critical determinant of successful NIV (31, 32). Current evidence does not demonstrate the superiority of any specific mask type with respect to ventilation efficacy. Nevertheless, the **nasal mask** is generally the preferred interface in children, given its wide availability in different sizes, the possibility of oral feeding and speech, and its relative ease of application. However, it may be associated with complications such as nasal bridge skin breakdown, xerophthalmia, or midface flattening, and its effectiveness can be reduced by mouth air leaks. In some cases, chin straps may help mitigate this problem (31, 32).

Alternative interfaces include **oronasal and full-face masks**, although these may be difficult to use in younger children because of claustrophobic sensations and the limited availability of appropriately sized models. Furthermore, they can increase the risk of aspiration in children with gastroesophageal reflux (31, 32). **Mouthpiece ventilation** can be useful in selected patients with stable chronic respiratory insufficiency, particularly during daytime use, and is often combined with another interface at night (**Table 4**) (33).

ADJUSTMENT OF LONG-TERM NIV PARAMETERS

For **CPAP**, treatment should be initiated at 4 cm H_2O and titrated upward until both adequate oxygenation and relief of obstruction are achieved, while maintaining patient tolerance (34, 35). PSG-guided titration remains the gold standard; however, in settings without PSG availability, oximetry trends and subsequent ventilator software analysis provide reliable alternatives (36). In most cases, effective pressures are achieved at approximately 8 ± 3 cm H_2O. Auto-CPAP may be considered in children exceeding the manufacturer's minimum weight threshold (typically >10 kg), although current evidence does not demonstrate clear superiority over fixed-pressure CPAP in pediatrics (5, 34).

For **BiPAP**, initial settings generally start with IPAP at 8 cm H_2O and EPAP at 4 cm H_2O, with subsequent adjustments guided by age, underlying disease, and clinical response (5, 37). The primary goal is to achieve a tidal volume of 6–10 mL/kg of ideal body weight. Final EPAP values typically range from 4 to 8 cm H_2O, though higher pressures may be required in the presence of structural airway anomalies predisposing to collapse (*e.g.*, pharyngomalacia, laryngomalacia, tracheomalacia). Final IPAP values are usually 10–14 cm H_2O, although higher levels (>20 cm H_2O) have been used safely (2, 5, 37).

The **backup respiratory rate** should be set slightly below the child's spontaneous rate during N3 sleep or physiologic age-based norms (38). Breath cycling—that is, the initiation of inspiration and the transition to expiration—must be carefully tailored to the underlying pathology (38, 39). The sensitivity of the **inspiratory trigger** depends on both respiratory muscle strength and the adequacy of central respiratory control. Conversely, the **expiratory trigger** should reflect the ventilatory pattern: in restrictive disorders, prolongation of the inspiratory phase is desirable, and the trigger is commonly set at 20–25% of peak inspiratory flow, whereas in obstructive disorders such as severe tracheomalacia, earlier cycling is advantageous, with settings adjusted to 50–75% of inspiratory flow (30, 40, 41).

At present, no validated reference values exist for other ventilatory parameters; most recommendations rely on expert consensus and retrospective studies (**Table 5**).

Table 4. *Advantages and limitations of different patient interfaces for pediatric home mechanical ventilation.*

Interface type	Advantages	Limitations
Nasal mask	- Wide range of sizes - Allows speech - Possibility of oral feeding	- Air leakage through the mouth - Risk of midface hypoplasia
Oronasal mask	- Prevents mouth leak - Lower risk of midface hypoplasia	- Not suitable for very young children - Interference with feeding, speaking, and secretion clearance - Risk of aspiration/asphyxia

DISCHARGE AND FOLLOW-UP

Regular clinical follow-up after discharge is essential. The first visit is recommended one month after discharge, followed by evaluations every three to six months depending on the underlying disease, type of SDB, and treatment adherence (5, 9). Each visit should include a detailed medical history, physical examination with particular attention to mask-related complications, and analysis of ventilator software. Device data provide important insights into adherence, duration of use, patient–ventilator synchrony, and air leaks (36). Importantly, careful fine-tuning of ventilator parameters based on these data can significantly improve both patient comfort and the overall effectiveness of ventilation. Some devices generate automated estimates of AHI, but these should be interpreted with caution, as most algorithms have been validated only in adults (36, 42).

Where available, transcutaneous capnometry every six months is advisable. Follow-up PSG, or respiratory polygraphy, should be performed whenever ventilator settings are modified or when interventions—such as orthodontic or orthopedic treatments—have the potential to alter airway function (43, 44).

Follow-up practices differ substantially across healthcare systems (7, 16, 45-47). In highly developed countries, most follow-up, including continuous non-invasive transcutaneous capnometry, is provided by specialized home-care services (48-50). In middle-income settings, this monitoring is more commonly hospital-based, which may limit frequency and accessibility (7, 16, 45-47).

Recent technological progress has enabled telemonitoring, whereby ventilator software data are transmitted to secure remote servers and analyzed by clinicians without the need for in-person visits. This approach has proven particularly valuable for monitoring adherence and detecting technical or clinical issues at an early stage (51, 52).

WEANING AND DISCONTINUATION OF LONG-TERM HMV

Weaning from long-term IMV via tracheostomy is a complex, stepwise process that requires careful clinical judgment and multidisciplinary collaboration (53, 54). The ultimate goal is decannulation and transition to NIV, whenever feasible, in order to minimize long-term complications and improve quality of life (53, 54).

The key steps in the weaning process include:

1. Overall assessment – evaluation of disease stability, improvement or resolution of the original indication for IMV, adequate spontaneous respiratory drive, and sufficient bulbar function to protect the airway.
2. Gradual reduction of ventilatory support – progressive shortening of IMV duration, initially maintaining nocturnal ventilation, followed by stepwise daytime trials off the ventilator.
3. Capping and tolerance trials – daytime tracheostomy capping to evaluate the child's ability to maintain adequate gas exchange without ventilatory support, with continuous monitoring of SpO_2 and $TcCO_2$.
4. Transition to NIV – initiation of mask ventilation once spontaneous breathing with capping is tolerated, typically starting during sleep and extending as feasible.

***Table 5**. Recommended BiPAP settings for long-term HMV in children.*

Parameter	Settings
IPAP	Start at 8 cm H_2O; Target tidal volume: 6–10 mL/kg/ideal body weight per breath
EPAP	Minimum: 4 cm H_2O; Typical final range: 6–10 cm H_2O
Respiratory Rate	*Controlled ventilation*: 2–3 breaths below physiologic rate for age; *Spontaneous breathing*: based on rate during N3 sleep
Inspiratory Time (Ti)	*Controlled ventilation*: Ti = 1/3 of total cycle time; *Spontaneous breathing*: Ti-min and Ti-max defined by device
Inspiratory Trigger	High sensitivity for neuromuscular disorders; Low sensitivity for central hypoventilation syndromes
Expiratory Trigger	Restrictive patterns: 20–25% of flow; Obstructive patterns: 50–75% of flow

5. Decannulation – performed once NIV is established and airway patency is confirmed (endoscopic assessment recommended), ensuring the child can maintain adequate ventilation and secretion clearance.
6. Post-decannulation monitoring – close observation in a controlled hospital setting to promptly detect respiratory compromise, followed by structured outpatient follow-up.

Children with neuromuscular disorders or residual central hypoventilation may continue to require nocturnal NIV even after successful decannulation (55). The timing of decannulation must balance the risks of prolonged tracheostomy (*e.g.*, infection, tracheal injury, psychosocial burden) against the safety of airway protection and effective ventilation (56). A multidisciplinary team—pulmonologist, intensivist, ENT surgeon, respiratory therapist, and speech/swallow therapist—should oversee the process in close cooperation with the family.

Discontinuation of home NIV may be considered in selected patients if normalization of SDB and gas exchange occurs spontaneously or following a therapeutic intervention (5). This is more commonly achievable in children treated with CPAP and less frequent in those requiring BiPAP. Before discontinuation, PSG or respiratory polygraphy with transcutaneous capnometry must be repeated. Criteria include resolution of SDB symptoms, AHI <10/h, $TcCO_2$ >50 mmHg for less than 2% of total sleep time, and SpO_2 <90% for less than 2% of total sleep time (5). Because recurrence of symptoms is possible, structured follow-up remains essential even after discontinuation.

MANAGEMENT OF LONG-TERM NIV FAILURE AND ALTERNATIVE OPTIONS

The most common cause of NIV failure in the home setting is poor adherence by the child or caregivers (57). Among adherent patients, failure may occur due to suboptimal patient–ventilator synchrony, excessive air leaks, progression of the underlying disease, or associated comorbidities (57). Alternative therapeutic options remain limited, but in selected cases, high-flow nasal cannula therapy or hypoglossal nerve stimulation may be considered as substitutes for CPAP (58, 59). Surgical or orthodontic interventions may be appropriate in children with Pierre–Robin sequence (60, 61). In the most severe cases, tracheostomy with invasive long-term ventilation remains the ultimate therapeutic option.

PREOPERATIVE USE OF LONG-TERM HOME NIV

Children with severe skeletal deformities, particularly kyphoscoliosis, should undergo preoperative evaluation for potential NIV initiation (62). While long-term NIV is clearly indicated in patients with alveolar hypoventilation and severe SDB, normal polygraphic and capnometric findings do not necessarily exclude the need for NIV (62, 63). Preoperative initiation has been shown to reduce the risk of prolonged postoperative ventilation, underscoring the importance of thorough evaluation and timely initiation of therapy (62, 63).

Risk assessment should include clinical features, non-invasive pulmonary function tests, the underlying condition (idiopathic scoliosis *versus* neuromuscular-associated scoliosis), and polygraphic/capnometric studies (62). Although reductions in FVC and FEV1 are inversely correlated with the need for postoperative ventilation, no universally accepted preoperative thresholds exist. Consequently, a low threshold for initiating NIV is advisable, particularly in patients with severe restrictive ventilatory patterns, pronounced spinal deformity, FEV1 <40% predicted, concomitant neuromuscular disease, or those scheduled for thoracotomy (5, 62).

CONCLUSION

Home mechanical ventilation has transformed the care of children with chronic respiratory failure, improving survival, reducing hospital dependence, and enabling better quality of life. The choice between invasive and non-invasive modalities must be individualized, but successful outcomes universally depend on structured initiation, careful parameter adjustment, and systematic follow-up. Future advances will rely on technology, multidisciplinary expertise, and broader access to specialized home-care services.

COMPLIANCE WITH ETHICAL STANDARDS

Conflict of interests

The Authors declare that they have no financial or personal conflicts of interest that might have influenced the work reported in this article.

Funding

There were no institutional or private funding for this article.

Author contributions

All the Authors confirmed the contribution to the manuscript's conception and approved its final version.

Ethical approval

Human studies and subjects

The study was conducted according to the guidelines of the Declaration of Helsinki and approved by the local Institutional Ethics Committee (n. 08/2014).

Animal studies

N/A.

Publication ethics

Plagiarism

Authors declare no potentially overlapping publications with the content of this manuscript and all original studies are cited as appropriate.

Data falsification and fabrication

The data is based on real-world observations.

REFERENCES

1. Tan HL, Chawla J. Home mechanical ventilation in children: evolving indications in an era of new treatment options. Eur Respir Rev. 2024;33(174):240154. doi: 10.1183/16000617.0154-2024.
2. Kwak S. Home mechanical ventilation in children with chronic respiratory failure: a narrative review. J Yeungnam Med Sci. 2022;40(2):123–35. doi: 10.12701/jyms.2022.00400.
3. Kaditis AG, Alonso Alvarez ML, Boudewyns A, Abel F, Alexopoulos EI, Ersu R, et al. ERS statement on obstructive sleep disordered breathing in 1- to 23-month-old children. Eur Respir J. 2017;50(6):1700985. doi: 10.1183/13993003.00985-2017.
4. Kaditis AG, Alonso Alvarez ML, Boudewyns A, Alexopoulos EI, Ersu R, Joosten K, et al. Obstructive sleep disordered breathing in 2- to 18-year-old children: diagnosis and management. Eur Respir J. 2016;47(1):69–94. doi: 10.1183/13993003.00374-2015.
5. Fauroux B, Abel F, Amaddeo A, Bignamini E, Chan E, Corel L, et al. ERS statement on paediatric long-term noninvasive respiratory support. Eur Respir J. 2022;59(6):2101404. doi: 10.1183/13993003.01404-2021.
6. Berry RB, Brooks R, Gamaldo C, Harding SM, Lloyd RM, Quan SF, et al. AASM Scoring Manual Updates for 2017 (Version 2.4). J Clin Sleep Med. 2017;13(5):665–6. doi: 10.5664/jcsm.6576.
7. Basa M, Minic P, Rodic M, Sovtic A. Evolution of Pediatric Home Mechanical Ventilation Program in Serbia—What Has Changed in the Last Decade. Front Pediatr. 2020;8:261. doi: 10.3389/fped.2020.00261.
8. Pocrncic S, Herzig JJ, Benning L, Mollet M, Bradicich M, Lichtblau M, et al. Bicarbonate from arterial blood gas analysis as predictor of sleep-related hypoventilation: a diagnostic accuracy study. BMJ Open Respir Res. 2025;12(1):e002591. doi: 10.1136/bmjresp-2024-002591.
9. Mayer OH, Amin R, Sawnani H, Shell R, Katz SL; RIND Study Group. Respiratory Insufficiency in Neuromuscular Disease (RIND): A Delphi Study to Establish Consensus Criteria to Define and Diagnose Hypoventilation in Pediatric Neuromuscular Disease. J Neuromuscul Dis. 2023;10(6):1075-1082. doi: 10.3233/JND-230053.
10. Bayav S, Çobanoğlu N. Indications and practice of home invasive mechanical ventilation in children. Pediatr Pulmonol. 2024;59(8):2210–5. doi: 10.1002/ppul.26342.
11. Basa M, Višekruna J, Gojsina-Parezanović B, Grba T, Anđelković M, Sovtić A. Congenital central hypoventilation syndrome: Heterogeneous clinical presentation, ventilatory modalities and outcome. Med Istraz. 2023;56(4):11–7.
12. Duijts L, Van Meel ER, Moschino L, Baraldi E, Barnhoorn M, Bramer WM, et al. European Respiratory Society guideline on long-term management of children with bronchopulmonary dysplasia. Eur Respir J. 2020;55(1):1900788. doi: 10.1183/13993003.00788-2019.
13. Ribeiro C, Jácome C, Oliveira P, Conde S, Windisch W, Nunes R. Patients experience regarding home mechanical ventilation in an outpatient setting. Chron Respir Dis. 2022;19:14799731221137082. doi: 10.1177/14799731221137082.
14. Chatwin M, Nickol AH, Morrell MJ, Polkey MI, Simonds AK. Randomised trial of inpatient versus outpatient initiation of home mechanical ventilation in patients with nocturnal hypoventilation. Respir Med. 2008;102(11):1528–35. doi: 10.1016/j.rmed.2008.06.008.
15. Zampoli M, Booth J, Gray DM, Vanker A. Home ventilation in low resource settings: Learning to do more, with less. Pediatr Pulmonol. 2024;59(8):2180–9. doi: 10.1002/ppul.26341.
16. Sovtic A, Minic P, Vukcevic M, Markovic-Sovtic G, Rodic M, Gajic M. Home mechanical ventilation in children is feasible in developing countries. Pediatr Int. 2012;54(5):676–81. doi: 10.1111/j.1442-200X.2012.03563.x.
17. Amin R, Sayal A, Syed F, Daniels C, Hoffman A, Moraes TJ, et al. How long does it take to initiate a child on long-term invasive ventilation? Results from a Canadian pediatric home ventilation program. Can Respir J. 2015;22(2):103–8. doi: 10.1155/2015/101685.
18. Chawla J, Edwards EA, Griffiths AL, Nixon GM, Suresh S, Twiss J, et al. Ventilatory support at home for children: A

joint position paper from the Thoracic Society of Australia and New Zealand/Australasian Sleep Association. Respirology. 2021;26(10):920–37. doi: 10.1111/resp.14088.

19. Tommesani C, Khirani S, Amaddeo A, Massenavette B, Bierme P, Taytard J, et al. Long term noninvasive respiratory support in children with OSA-I and OSA-II: Data of a nation-wide study. Sleep Med. 2025;126:67–72. https://doi.org/10.1016/j.sleep.2024.107472.
20. Genet L, Khirani S, Vegas N, Griffon L, Adnot P, Giuseppi A, et al. The challenge of assessing upper airway obstruction severity in infants with Robin Sequence. Sleep Med. 2025;132:106535. doi: 10.1016/j.sleep.2024.106535.
21. Remy F, Taverne M, Khonsari RH, Fauroux B, Khirani S, Martínez Abadías N, et al. Differential impact of Crouzon and Apert syndromes on upper airways morphology: implications for obstructive sleep apnoea. J Craniomaxillofac Surg. 2025;53(9):1577–91. doi: 10.1016/j.jcms.2025.06.012.
22. Fauroux B, Cozzo M, MacLean J, Fitzgerald DA. OSA type-III and neurocognitive function. Paediatr Respir Rev. 2025;53:39–43. 10.1016/j.jcms.2025.06.01210.1016/j.prrv.2023.12.004.
23. Trang H, Samuels M, Ceccherini I, Frerick M, Garcia-Teresa MA, Peters J, et al. Guidelines for diagnosis and management of congenital central hypoventilation syndrome. Orphanet J Rare Dis. 2020;15:252. 10.1016/j.jcms.2025.06.01210.1186/s13023-020-01505-5.
24. Porcaro F, Paglietti MG, Cherchi C, Schiavino A, Chiarini Testa MB, Cutrera R. How the management of children with congenital central hypoventilation syndrome has changed over time: two decades of experience from an Italian center. Front Pediatr. 2021;9:648927. doi: 10.3389/fped.2021.648927.
25. Gaboli MP. Prolonged mechanical ventilation in children with neuromuscular disease. In: Kawaguchi A, Pons Odena M, Graham RJ, editors. Prolonged and Long-Term Mechanical Ventilation in Children. Singapore: Springer Nature; 2024. p. 197–211. doi: 10.1007/978-981-97-8903-0_11.
26. Duyndam A, Ista E, Houmes RJ, van Driel B, Reiss I, Tibboel D. Invasive ventilation modes in children: a systematic review and meta-analysis. Crit Care. 2011;15(1):R24. doi: 10.1186/cc9973.
27. Bayav S, Çobanoğlu N. Indications and practice of home invasive mechanical ventilation in children. Pediatr Pulmonol. 2024;59(8):2210–5. doi: 10.1002/ppul.26342.
28. Park S, Suh ES. Home mechanical ventilation: back to basics. Acute Crit Care. 2020;35(3):131–41. doi: 10.4266/acc.2020.00101.
29. Gregoretti C, Navalesi P, Ghannadian S, Carlucci A, Pelosi P. Choosing a ventilator for home mechanical ventilation. Breathe. 2013;9(5):394–409. doi: 10.1183/20734735.000613.
30. Baumgartner J, Schmidt J, Klotz D, Schneider H, Schumann S, Fuchs H. Trigger performance of five pediatric home ventilators and one ICU ventilator depending on circuit type and system leak in a physical model of the lung. Pediatr Pulmonol. 2022;57(3):744–53. doi: 10.1002/ppul.25791.
31. Luján M, Flórez P, Pomares X. What circuits, masks and filters should be used in home non-invasive mechanical ventilation. J Clin Med. 2023;12(7):2692. doi: 10.3390/jcm12072692.
32. Castro-Codesal ML, Olmstead DL, MacLean JE. Mask interfaces for home non-invasive ventilation in infants and children. Paediatr Respir Rev. 2019;32:66–72. doi: 10.1016/j.prrv.2019.01.002.
33. Toussaint M, Chatwin M, Gonçalves MR, Gonzalez-Bermejo J, Benditt JO, McKim D, et al. Mouthpiece ventilation in neuromuscular disorders: narrative review of technical issues important for clinical success. Respir Med. 2021;180:106373. doi: 10.1016/j.rmed.2021.106373.
34. Khaytin I, Tapia IE, Xanthopoulos MS, Cielo C, Kim JY, Smith J, et al. Auto-titrating CPAP for the treatment of obstructive sleep apnea in children. J Clin Sleep Med. 2020;16(6):871–8. doi: 10.5664/jcsm.8376.
35. Khirani S, Griffon L, Dosso M, La Regina DP, Vedrenne-Cloquet M, Poirault C, et al. Does therapeutic CPAP pressure correlate with OSA severity in children? Sleep Med. 2025;129:89–93. doi: 10.1016/j.sleep.2024.106493.
36. Khirani S, Dosso M, Gerin L, Basa M, Collignon C, Vedrenne-Cloquet M, et al. Why breath-by-breath built-in software data should be used to monitor CPAP/NIV in children? Pediatr Pulmonol. 2024;59(2):506–9. doi: 10.1002/ppul.26210.
37. Fauroux B, Leroux K, Desmarais G, Isabey D, Clément A, Lofaso F, et al. Performance of ventilators for noninvasive positive-pressure ventilation in children. Eur Respir J. 2008;31(6):1300–7. doi: 10.1183/09031936.00159307.
38. Steindor M, Wagner CE, Bock C, Eckerland M, Heitschmidt L, Pichlmaier L, et al. Home noninvasive ventilation in pediatric subjects with neuromuscular diseases: one size fits all. Respir Care. 2021;66(3):410–5. doi: 10.4187/respcare.07979.
39. Khirani S, Dosso M, Collignon C, Tommesani C, Vedrenne-Cloquet M, Griffon L, et al. Why home NIV devices should have a Ti min/Ti max? Pediatr Pulmonol. 2023;58(8):2417–21. doi: 10.1002/ppul.26200.
40. Khirani S, Griffon L, Dosso M, Capriles S, Vedrenne-Cloquet M, Poirault C, et al. 100 ms matter: impact of back-up Ti on triggered breaths in pediatric NIV. Sleep Med. 2025;134:106727. doi: 10.1016/j.sleep.2024.106727.
41. Khirani S, Stehling F, Dudoignon B, Amaddeo A, Dosso M, Poirault C, et al. Trigger issues with a life support device in children. Sleep Med. 2025;131:106534. doi: 10.1016/j.sleep.2024.106534.
42. Onofri A, Pavone M, De Santis S, Verrillo E, Caggiano S, Ullmann N, et al. Built-in software in children on long-term ventilation in real life practice. Pediatr Pulmonol. 2020;55(10):2697–705. doi: 10.1002/ppul.24957.
43. Felemban O, Leroux K, Aubertin G, Miandy F, Damagnez F, Amorim B, et al. Value of gas exchange recording at

home in children receiving non-invasive ventilation. Pediatr Pulmonol. 2011;46(8):802–8. doi: 10.1002/ppul.21444.

44. Foster CC, Kwon S, Shah AV, Hodgson CA, Hird-McCorry LP, Janus A, et al. At-home end-tidal carbon dioxide measurement in children with invasive home mechanical ventilation. Pediatr Pulmonol. 2022;57(11):2735–44. doi: 10.1002/ppul.26091.
45. Nathan AM, Loo HY, de Bruyne JA, Eg KP, Kee SY, Thavagnanam S, et al. Thirteen years of invasive and noninvasive home ventilation for children in a developing country: A retrospective study. Pediatr Pulmonol. 2017;52(4):500–7. doi: 10.1002/ppul.23621.
46. van der Poel LAJ, Booth J, Argent A, van Dijk M, Zampoli M. Home ventilation in South African children: Do socioeconomic factors matter? Pediatr Allergy Immunol Pulmonol. 2017;30(3):163–70. doi: 10.1089/ped.2017.0826.
47. Yanaz M, Unal F, Hepkaya E, Yazan H, Oksay SC, Kostereli E, et al. Home mechanical ventilation in children: The experience of pediatric pulmonology divisions in Istanbul. Eur Respir J. 2022;60(suppl 66):3129. doi: 10.1183/13993003.congress-2022.3129.
48. Racca F, Berta G, Sequi M, Bignamini E, Capello E, Cutrera R, et al. Long-term home ventilation of children in Italy: a national survey. Pediatr Pulmonol. 2011;46(6):566–72. doi: 10.1002/ppul.21421.
49. Windisch W, Callegari J, Karagiannidis C. Home mechanical ventilation in Germany. Dtsch Med Wochenschr. 2019;144(11):743–7. doi: 10.1055/a-0880-2086.
50. Simonds AK. Home mechanical ventilation: an overview. Ann Am Thorac Soc. 2016;13(11):2035–44. doi: 10.1513/AnnalsATS.201605-343KV.
51. Duiverman ML, Crimi C. Telemedicine in home mechanical ventilation: promise, pitfalls and path forward. Thorax. 2025;thorax-2025-223348. doi: 10.1136/thorax-2025-223348.
52. Duiverman ML, Ribeiro C, Tonia T, Hazenberg A, van Meerloo S, van Meerloo H, et al. European Respiratory Society Clinical Practice Guideline on Telemedicine in Home Mechanical Ventilation. Eur Respir J. 2025;2500094. doi: 10.1183/13993003.00094-2025.
53. Kamalaporn H, Preutthipan A, Coates AL. Weaning strategies for children on home invasive mechanical ventilation. Pediatr Pulmonol. 2024;59(8):2131–40. doi: 10.1002/ppul.26338.
54. Dolinay T, Hsu L, Maller A, Walsh BC, Szűcs A, Jerng JS, et al. Ventilator weaning in prolonged mechanical ventilation—a narrative review. J Clin Med. 2024;13(7):1909. doi: 10.3390/jcm13071909.
55. Fauroux B, Khirani S, Griffon L, Teng T, Lanzeray A, Amaddeo A. Non-invasive ventilation in children with neuromuscular disease. Front Pediatr. 2020;8:482. doi: 10.3389/fped.2020.00482.
56. Verma R, Mocanu C, Shi J, Miller MR, Chiang J, Wolter NE, et al. Decannulation following tracheostomy in children: a systematic review of decannulation protocols. Pediatr Pulmonol. 2021;56(8):2426–43. doi: 10.1002/ppul.25458.
57. Amaddeo A, Khirani S, Griffon L, Teng T, Lanzeray A, Fauroux B. Non-invasive ventilation and CPAP failure in children and indications for invasive ventilation. Front Pediatr. 2020;8:544921. doi: 10.3389/fped.2020.544921.
58. Randerath W, Verbraecken J, De Raaff CAL, Hedner J, Herkenrath S, Hohenhorst W, et al. European Respiratory Society guideline on non-CPAP therapies for obstructive sleep apnoea. Eur Respir Rev. 2021;30(162):210200. doi: 10.1183/16000617.0200-2021.
59. Whitla L, Lennon P. Non-surgical management of obstructive sleep apnoea: a review. Paediatr Int Child Health. 2017;37(1):1–5. doi: 10.1080/20469047.2016.1248853.
60. Haas J, Yuen K, Farrokhyar F, Aminnejad M, Williams C, Choi M. Non-operative interventions for Pierre-Robin sequence: a systematic review and meta-analysis. J Craniomaxillofac Surg. 2024;52(12):1422–7. doi: 10.1016/j.jcms.2024.08.003.
61. Kosyk MS, Carlson AR, Zapatero ZD, Kalmar CL, Liaquat S, Bartlett SP, et al. Multimodal treatment of Robin Sequence utilizing mandibular distraction osteogenesis and continuous positive airway pressure. Cleft Palate Craniofac J. 2023;60(8):993–1001. doi: 10.1177/10556656231159393.
62. Lewis H, Norrington A. Paediatric preoperative assessment. BJA Educ. 2023;23(6):238–44. doi: 10.1016/j.bjae.2023.03.004.
63. Khirani S, Amaddeo A, Griffon L, Lanzeray A, Teng T, Fauroux B. Follow-up and monitoring of children needing long-term home ventilation. Front Pediatr. 2020;8:330. doi: 10.3389/fped.2020.00330.