

## ORIGINAL ARTICLE

# Finnish children who needed long-term home respiratory support had severe sleep-disordered breathing and complex medical backgrounds

Mervi Järvelä<sup>1,2,3</sup>  | Maija Katila<sup>1,4</sup>  | Vesa Eskola<sup>1,4</sup>  | Riikka Mäkinen<sup>4</sup> | Paula Mandelin<sup>4</sup> | Outi Saarenpää-Heikkilä<sup>1,4</sup> | Eero Lauhkonen<sup>1,4</sup> 

<sup>1</sup>Tampere University, Tampere, Finland

<sup>2</sup>Department of Paediatrics, Seinäjoki Central Hospital, Seinäjoki, Finland

<sup>3</sup>Department of Anesthesiology and Intensive Care, Seinäjoki Central Hospital, Seinäjoki, Finland

<sup>4</sup>Tampere University Hospital, Tampere, Finland

## Correspondence

Mervi Järvelä, Department of Paediatrics/Anesthesiology and Intensive Care, Seinäjoki Central Hospital, Seinäjoki, Finland, Tampere University, Tampere, Finland.

Email: [mervi.jarvela@tuni.fi](mailto:mervi.jarvela@tuni.fi)

## Funding information

Hengityssairauksien Tutkimussäätiö; Lastentautien Tutkimussäätiö; Tampereen Tuberkuloosisäätiö

## Abstract

**Aim:** No studies have described long-term paediatric home respiratory support in Nordic countries. We examined the clinical characteristics and long-term outcomes of paediatric patients who received continuous positive airway pressure, non-invasive-positive-pressure ventilation and invasive ventilation from a multidisciplinary home respiratory support team.

**Methods:** Retrospective tertiary-level data were collected between 1 January 2010 and 31 December 2020 in Tampere University Hospital. These comprised patient demographics, treatment course and polysomnography-confirmed sleep-disordered breathing (SDB).

**Results:** There were 93 patients (63.4% boys). The median age at treatment initiation was 8.4 (range 0.11–16.9) years. The patients had: neuromuscular disease (16.1%), central nervous system disease (14.0%), developmental disabilities and congenital syndrome (29.0%), lung-airway conditions (11.8%), craniofacial syndrome (15.1%) and severe obesity (14.0%). More than two-thirds had severe SDB (66.7%) and the most common one was obstructive sleep apnoea in 66.7%. We found that 92.5% received long-term therapy for more than 3 months and the mean treatment duration was  $3.3 \pm 2.7$  years. A non-invasive mask interface was used in 94.7% of cases and 5.3% needed tracheostomy ventilation. More than a quarter (26.7%) achieved disease resolution during the study period.

**Conclusion:** Most children who needed long-term home respiratory support had complex conditions and severe, persistent SDB.

## KEYWORDS

continuous positive airway pressure, home respiratory support, non-invasive positive-pressure ventilation, paediatric sleep apnoea, sleep-disordered breathing

**Abbreviations:** CPAP, continuous positive airway pressure; NPPV, non-invasive positive-pressure ventilation; OSA, obstructive sleep apnoea; SDB, sleep-disordered breathing.

This is an open access article under the terms of the [Creative Commons Attribution-NonCommercial-NoDerivs](https://creativecommons.org/licenses/by-nc-nd/4.0/) License, which permits use and distribution in any medium, provided the original work is properly cited, the use is non-commercial and no modifications or adaptations are made.

© 2023 The Authors. *Acta Paediatrica* published by John Wiley & Sons Ltd on behalf of Foundation Acta Paediatrica.

## 1 | INTRODUCTION

The total prevalence of obstructive sleep apnoea (OSA) ranges from 0.1 to 13% and this is most commonly reported between 1 and 4%.<sup>1</sup> OSA is the most common type of sleep-disordered breathing (SDB).<sup>2</sup> The other type of SDB is central sleep apnoea, which is characterised by the lack of drive to breath during sleep without obstruction in the airways. The prevalence of central sleep apnoea in the paediatric population varies between 1 and 5% in healthy children.<sup>3</sup>

Central sleep apnoea is most commonly associated with brain anomalies, neurogenetic conditions, upper airway anomalies, gastroesophageal reflux, prematurity, obesity or hypothyroidism. Sometimes OSA and central sleep apnoea do present together. Central sleep apnoea or OSA can also occur in the presence or absence of hypoventilation.<sup>3</sup> This inadequate respiratory gas exchange leads to *abnormal retention of carbon dioxide in the blood and results in hypoventilation*.

In primary OSA the most common aetiology is adenotonsillar hypertrophy,<sup>1,4</sup> but sometimes OSA is secondary to complex conditions with multiple age-dependent factors resulting in heterogeneous types of sleep apnoea syndrome.<sup>5,6</sup> Congenital craniofacial or airway malformations, muscular hypotonia and central breathing disorders play a significant role in small children.<sup>3,7,8</sup> When the child gets older acquired obesity, severe lung diseases and progressive neuromuscular disorders become more prevalent.<sup>3,7,8</sup>

The first-line treatment for paediatric OSA is often an adenotonsillectomy. Weight loss, nasal steroids, montelukast or orthodontic treatments are also considered prior to treatment with continuous positive airway pressure (CPAP) or non-invasive positive pressure ventilation (NPPV).<sup>1,4</sup> Treating adult OSA with CPAP was established 40 years ago and has been rapidly increasing.<sup>9,10</sup> For example in France between 2010 and 2018, the annual incidence of CPAP treatment in adult patients increased 1.9-fold, with an average annual increase of 11.4%.<sup>11</sup> The growing awareness of SDB symptoms, increasing obesity and better diagnostic capacities are well-known reasons for the growing sleep apnoea population. Also, the development of masks and respiratory support devices has given physicians a better chance to treat a broader spectrum of SDB patients.<sup>12-14</sup> Also, home invasive ventilation in children has risen since the year 1999, especially with younger children.<sup>15,16</sup> This establishes the need to educate more physicians who can be involved in the care of these children.<sup>16</sup>

On 5 November 2015, the European Respiratory Society Task Force published guidelines on obstructive sleep apnoea for children aged 2-18 years. On 16 August 2017, the statement on obstructive sleep-disordered breathing children aged 1-23 months was accepted.<sup>1,4</sup> These guidelines suggest that treatment with a home respiratory support follows a multidisciplinary evaluation and a stepwise treatment approach with surgical and non-surgical steps.<sup>1,4</sup> Often children ending up having a home respiratory support device are those with SDB related to a complex condition background.<sup>17</sup>

### Key Notes

- No studies have described long-term paediatric home respiratory care provided by a multidisciplinary home respiratory support team in Nordic countries.
- This Finnish study found that the 93 children who needed long-term home respiratory support had complex conditions and persistent severe sleep-disordered breathing.
- The majority (94.7%) used a non-invasive mask interface, only 4.7% used tracheostomy ventilation and 26.7% achieved disease resolution during the study period.

Longitudinal descriptive data of treatment outcomes are available from specialised paediatric centres treating home mechanical ventilation via tracheostomy,<sup>18,19</sup> but also increasingly with non-invasive respiratory support via masks.<sup>12,14,16,20</sup> Still, no studies have described how modern auto-titrating devices are used in clinical practice in long-term treatment in paediatric patients who have polysomnography-confirmed SDB. Moreover, recognised issues with adherence to mask treatment in children present in up to 30-50% of patients, as recently summed in a meta-analysis.<sup>21</sup> In spite of this, clinically oriented paediatric literature is scarce. There is a need for guidance on treatment decisions, complication avoidance, practice of follow-up visits and SDB monitoring.<sup>13,14,22</sup> The aim of this study was to describe the clinical characteristics and long-term outcome of paediatric patients treated with home respiratory support.<sup>3,7,8</sup>

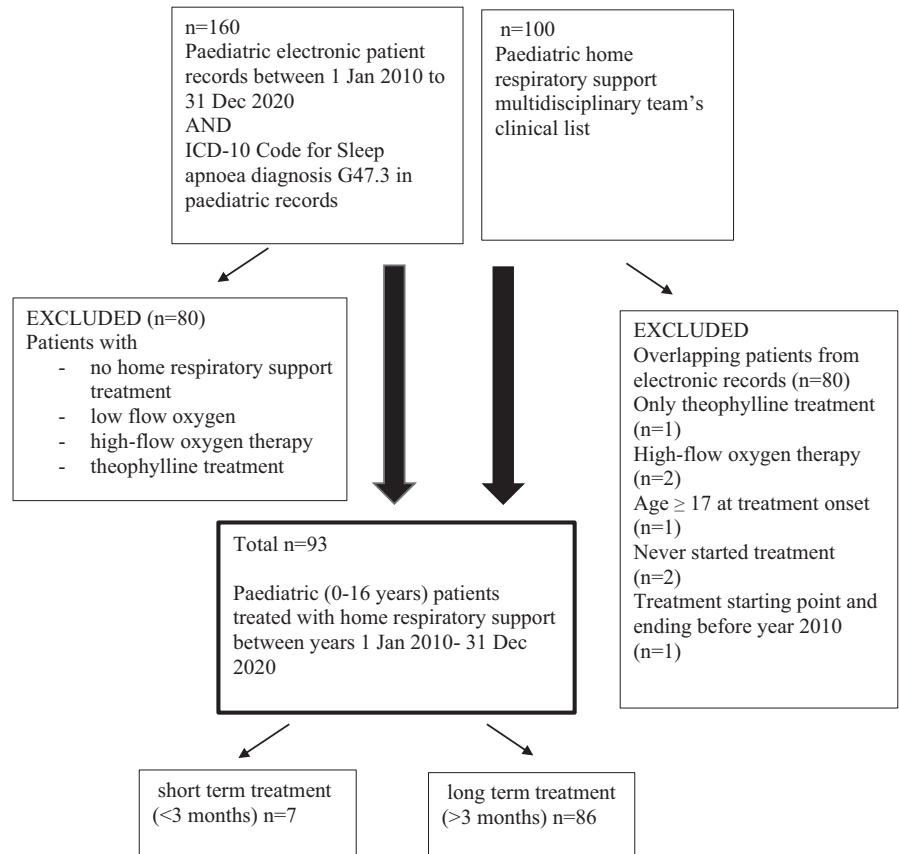
## 2 | MATERIALS AND METHODS

### 2.1 | Design and background factors

We performed a retrospective clinical data collection of patients treated by a multidisciplinary paediatric home respiratory support team from 1 January 2010 to 31 December 2020 in Tampere University Hospital, Finland. These patients had polysomnography-confirmed SDB, which was treated via continuous positive airway pressure (CPAP), non-invasive positive-pressure ventilation (NPPV) or invasive ventilation. The study group was committed to following good scientific ethics according to the Declaration of Helsinki. The study did not include any direct intervention with the patients. The study protocol was approved by the Ethics Committee of Pirkanmaa Hospital District on 1 July 2021 (study number R21086).

The individuals were identified by combining two overlapping sources (Figure 1): the team's clinical list of patients and a database search. The database search was conducted from electronic patient records from our paediatric unit. From these records, we searched for International Classification of Diseases-10th Revision diagnosis G47.3 (Sleep Apnoea). The database search found 160 cases, which were further checked manually resulting in 80 true patients who had

**FIGURE 1** Patient inclusion and exclusion criteria.



been treated with home respiratory support. The excluded patients had sleep apnoea diagnosis but never had needed home respiratory support. The team's clinical list provided 13 additional patients with mainly invasive home respiratory support. All the 93 subjects from the study group were found from the clinical list but the database search was used to confirm that no milder cases with home respiratory support would be missed. The total number of patients receiving home respiratory support was 93 patients. This included seven patients who had less than 3 months of home respiratory support treatment. However, 86 patients had long-term respiratory support meaning that they had more than 3 months of home respiratory support treatment. Patients with long-term treatment were categorised into two groups. The disease resolution group consisted of subjects whose SDB resolved before 31 December 2020 and into the non-resolution group if the SDB continued after 31 December 2020. The non-resolution group included also patients continuing the treatment elsewhere in a central hospital or an adult clinic, patients with relocation and patients with cessation of treatment or death due to underlying disease.

Data included patient demographics, underlying medical conditions and medications. The circumstances for home respiratory support onset and duration of treatment and frequency of follow-up visits were collected. Also, complications of treatment and operative interventions to treat sleep apnoea before and during periods with home respiratory support were accumulated. The type of respiratory support including device, mode and interface was recorded.

The type and severity of SDB were confirmed from polysomnography reports when applicable and reviewed by a clinical neurophysiologist. The polysomnographies were scored according to contemporary paediatric guidelines provided by American Academy of Sleep Medicine Manual for the Scoring of Sleep and Associated Events in Version 2.2 1 July 2015.<sup>23,24</sup> The version valid at the time in Tampere University Hospital, Dept. of Clinical Neurophysiology was the one that was used during different time periods. OSA was defined as obstructive apnoea-hypopnea index  $\geq 1$ /hour, central sleep apnoea as central apnoea index  $\geq 5$ /hour, mixed sleep apnoea as central apnoea index  $\geq 5$ /hour and obstructive apnoea-hypopnea index  $\geq 1$ /hour. Hypoventilation was defined by end-tidal carbon dioxide  $>50$  mmHg or  $>6.67$  kilopascal for  $>25\%$  of total sleep time.<sup>25</sup> If the patient was suffering from both sleep apnoea and hypoventilation they were categorised into the hypoventilation group.

The children were stratified into groups of mild, moderate or severe hypoventilation. Apnoea-hypopnea index 1–4 was considered mild, apnoea-hypopnea index 5–9 was considered moderate and apnoea-hypopnea index  $>10$  or hypoventilation was considered severe SDB.<sup>2,26</sup> There were some patients who had no polysomnography -data prior to treatment initiation due to acute respiratory failure. These patients were grouped as severe hypoventilation because they needed tracheostomy ventilation due to the inability to wean from ventilatory support. In cases with no polysomnography data and acute respiratory failure prior to treatment initiation, the SDB type and severity were not categorised.

SPSS version 28.00 (IBM Corp, New York, USA) was used in the statistical analyses of the data. The results are expressed as means, standard deviations (SD) and 95% confidence intervals (95% CI) for continuous variables, and numbers and frequencies for categorised variables. In the data analysis, we used a non-parametric test: Fisher's exact test or chi-square test were used to analyse categorical data and Mann-Whitney *U*-test was used for data not normally distributed.

### 3 | RESULTS

There were a total of 93 paediatric patients who received home respiratory support therapy in Tampere University Hospital from 1 January 2010 to 31 December 2020. The patient demographics and categorised patient groups are presented in Table 1. The two most common underlying main diagnoses were Down syndrome in 12.9% of the cases and severe obesity in 14.0% of the cases. Otherwise, the patient material was diverse with over 40 distinct diagnoses and can be seen in Table S1.

For clarification the patients were categorised into six groups based on the main diagnosis as follows: neuromuscular disease, central nervous system disease, developmental disabilities and congenital syndrome, lung-airway conditions, craniofacial syndrome and severe obesity (Tables 1 and S1). These groups consisted of complex conditions with multiple co-morbidities, apart from groups with severe obesity and lung-airway conditions. In the latter, there were four with adjunctive lung-airway-related diagnosis. One patient had Brune Belly syndrome, two patients had severe laryngomalacia and one patient had lung hypoplasia (Table S1).

In addition to SDB, 92.5% of the patients had severe underlying main diagnoses. There were only seven typically developing children who needed respiratory support. Of these children, six subjects had primary obstructive sleep apnoea and one subject had adenoid hypertrophy recurrence. Also, out of 23 patients with obesity, 78.3% had most commonly other co-morbidities or main diagnoses and only 21.7% of the obese children had obesity without other co-morbidities or diagnoses.

The median age at onset of treatment in 93 subjects was 8.4 years (range 0.11–16.9) and 63.4% were boys (Table 1). The two biggest co-morbidities were developmental delay in 39.8% of the patients and asthma in 23.7% of patients (Table 1). Patients were from all age groups from 1 month to 16 years old, as demonstrated in Figure 2.

The classifications of type and severity of SDB are presented in Table 2. Primary polysomnography was done and classified in Tampere University Hospital in 89.2% of the cases and another health-care unit in 5.4% of the cases. The primary polysomnography was totally lacking in six cases of which three cases were due to acute or chronic worsening of respiratory distress leading to invasive ventilation. These patients were categorised as severe hypoventilation. In addition, some three patients did not have polysomnography prior to treatment. The reasons for missing polysomnography were palliative care and in two cases treatment transfer from another unit and lack

**TABLE 1** Patient demographics and categorised patient groups for sleep-disordered breathing in 93 children treated with home respiratory support during 2010–2020.

	Study subjects N = 93	
	Median	Range SD
Age at treatment onset (years)	8.4	0.11–16.9
	N	%
Boy, n (%)	59	63.4
Girl, n (%)	34	36.6
Main diagnosis group		
Neuromuscular disease	15	16.1
Central nervous system disease	13	14.0
Developmental disabilities and congenital syndrome	27	29.0
Lung-airway disease conditions	11	11.8
Craniofacial syndrome	14	15.1
Severe Obesity (ISO-BMI $\geq 35$ kg/m <sup>2</sup> )	13	14.0
Co-morbidities		
Developmental disability	37	39.8
Asthma	22	23.7
GERD	17	18.3
Epilepsy	10	10.8
Scoliosis	8	8.6
Cleft palate/Cleft lip	11	11.8
Allergic rhinitis	5	5.4
Arnold Chiari malformation	2	2.2
CNS tumour	8	8.6
Obesity <sup>a</sup>	23	24.7
Obesity without other diagnoses or co-morbidities	5	5.4
Overweight <sup>a</sup>	8	8.6
Home medications		
Inhaled corticosteroids	22	23.7
Inhaled beta-2-agonists	31	33.3
Montelukast	8	8.6
Nasal steroids	3	3.2
Cardiovascular medication <sup>b</sup>	8	8.6

Note: Patients may have one or several co-morbidities.

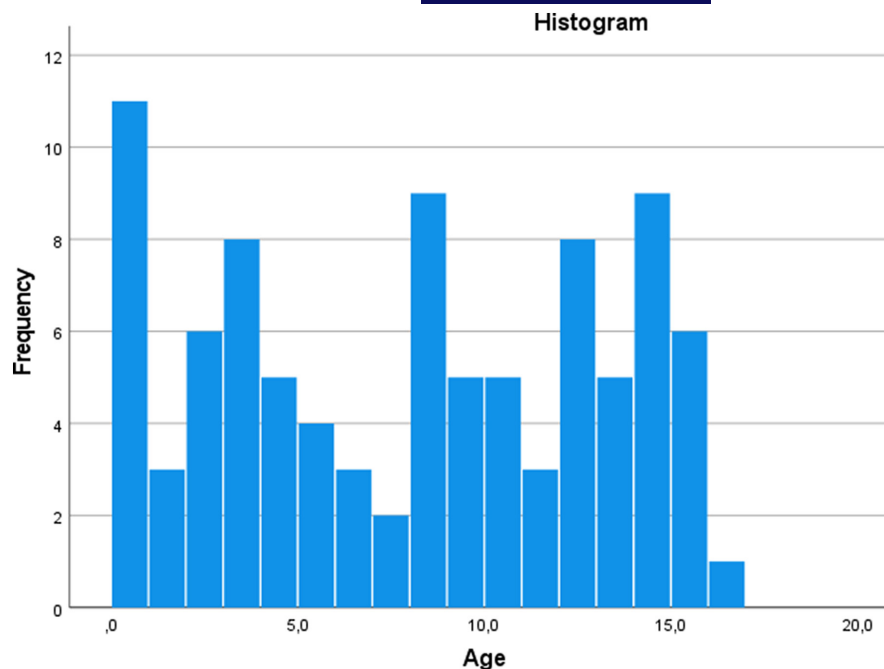
Abbreviations: CNS, central nervous system; GERD, gastroesophageal reflux disease; ISO-BMI, a body mass index corresponding to the adults' body mass index.

<sup>a</sup>Age-sex-specific weight and height data available in 80 cases (86%).

<sup>b</sup>Beta-blockers (*n* = 6), pulmonary hypertension medication (*n* = 1), (angiotensin-converting enzyme (ACE) inhibitors/angiotensin receptor blockers (*n* = 1).

of polysomnography records. Home respiratory support had been started with normal polysomnography in two cases. Both had neuromuscular disease and hypoventilation registered in a hospital during pneumonia.

**FIGURE 2** Age in years at the time of treatment initiation.



**TABLE 2** The classification of type and severity of sleep-disordered breathing in 93 children treated with home respiratory support during 2010–2020.

Study subjects N = 93	
Type of sleep-disordered breathing	N (%)
OSA	62 (66.7%)
Mixed (OSA and central sleep apnoea)	11 (11.8%)
Central sleep apnoea	2 (2.2%)
Hypoventilation	13 (14.0%)
Normal	2 (2.2%)
Not categorised (polysomnography missing/done in another unit)	3 (3.2%)
Severity of sleep-disordered breathing	
Mild	12 (12.9%)
Moderate	15 (16.1%)
Severe	62 (66.7%)
Not categorised (Polysomnography missing/done in another unit)	4 (4.3%)

Abbreviation: OSA, obstructive sleep apnoea.

The most common SDB type was OSA in 66.7% of the cases and the majority of patients were classified to have a severe SDB (Table 2). The mean apnoea-hypopnea index before treatment based on the polysomnography data was 26.4 (SD 30.0) ranging from 1 to 175 apnoeas or hypopnea per hour.

The characteristics and outcomes of home respiratory support are presented in Table 3. Nasal masks were used by 64.5% and 13.9% of children had two or more types of masks in use. Only 5.3% of the patients needed invasive home ventilation via tracheostomy and they all had severe hypoventilation. Diagnoses of these patients consisted of mitochondrial muscular disease, lung hypoplasia due to

diaphragmatic hernia, anoxic brain damage and brain tumour with pharyngeal palsy in two cases.

The frequency of follow-up visits in the long-term treatment was 3–6 months in 68.8% of the cases and there were an average of 3.5 annual visits per patient. The follow-up visits were carried out at the local central hospital in 12.9% of the cases. One patient was left without clinic visits due to the palliative care aspect of treatment. Long-term home respiratory support for over 3 months was present in 92.5% and 86 out of 93 patients. Seven patients had less than 3 months of treatment follow-up. There were five immediate transfers of treatment responsibility to another unit, one death due to underlying illness and one early treatment cessation due to poor adherence. Three patients had treatment for over 10 years, with the longest duration lasting for 11.8 years. Home respiratory support treatment had begun before 1 January 2010 in 17 cases. The mean age for treatment transfer from our paediatric unit to the adult clinic was 15.9 years ranging between 15 and 17 years in 24 cases.

There were 10 subjects with poor adherence to long-term home respiratory support use out of 86 patients. These patients did discontinue the treatment due to poor adherence. There were no statistical differences between those who continued treatment ( $n=76$ ) and those who stopped due to compliance issues ( $n=10$ ) in mean age (8.2 years,  $\pm 0.6$  vs. 6.0 years, SD 1.9,  $p=0.21$ ) or apnoea-hypopnea index (25.1/hour, SD 4.0 vs. 29.6/hour, SD 11.5,  $p=0.69$ ) at treatment onset.

Complications during home respiratory therapy were rare. Pneumothorax was not detected in any patient. Aspiration-related pneumonia was mentioned in three cases. One patient receiving home respiratory therapy over 11 years developed maxilla hypoplasia. Other type of facial shaping due to wearing a mask was registered in three cases (3.2%). The complications or complaints related to masks excluding tracheostomy ventilation are presented in Table 3.

**TABLE 3** Characteristics and long-term outcome of home respiratory support therapy in 93 children treated during 2010–2020.

Mean number of follow-up visits, frequency/year, <i>n</i> (%)	3.5 (SD 1.6)
Mask type ( <i>n</i> ): nasal/oronasal/nostril/multiple types/not specified	60/6/5/13/16
Mode of delivery	<i>n</i> (%)
CPAP	10 (10.8%)
Auto-CPAP	50 (53.7%)
NPPV	28 (30.1%)
Tracheostomy-ventilation	5 (5.3%)
Mask complications/complaints (tracheostomy ventilation not included)	<i>n</i> (%)
Nasal obstruction	21 (22.6%)
Skin irritation	10 (10.8%)
Mask pressure on face	10 (10.8%)
Unability to use mask on infection (excessive mucus)	10 (10.8%)
Sleeping disturbances	4 (4.3%)
Psychological anxiety	3 (3.2%)
Aspiration-related pneumonia	3 (3.2%)
Maxilla hypoplasia	1 (1.1%)
Pneumothorax	0
Long-term (>3 months) home respiratory support therapy	<i>N</i> =86
Duration (mean)	3.3 years (SD 2.7)
Treatment continuance in the paediatric clinic (checkpoint 31.12.2020)	18 (20.9%)
Disease resolution, <i>n</i> (%)	23 (26.7%)
Treatment cessation, <i>n</i> (%) (non-compliance and poor adherence)	10 (11.6%)
Treatment transferred to adult unit, <i>n</i> (%)	23 (26.7%)
Death due to underlying illness, <i>n</i> (%)	4 (4.7%)
Treatment continuance in another health care unit	7 (8.2%)
Unknown	1 (1.2%)

Abbreviations: CPAP, continuous positive airway pressure; NPPV, non-invasive positive pressure ventilation.

Resolution of SDB during 2010–2020 was present in 26.7% of children with a mean age of 4.3 years (SD 0.82). Resolution of SDB was a dominant feature in the groups of lung-airway conditions presenting in eight out of 11 cases (72.7%) and in craniofacial syndrome presenting eight out of 14 cases (57.1%), respectively. There were no obese or central nervous system disease patients in children with disease resolution.

In the whole study population (*n*=93) operative interventions were performed for 59.1% of the children. Adenotonsillectomy was the most common procedure in 34.4% of the cases. In addition, adenotomy was performed for 10.7%, adenotomy and tonsillectomy for 8.6%, tonsillectomy for 1.1%, orthodontic treatment for 10.7%, rapid maxillary expansion for 2.2% and craniofacial surgery for

10.7%. Some 23.7% of the subjects had more than one operative intervention previously described. Some 18.3% of the patients had also re-operations of the same surgery type. The majority of operative treatments in 73.1% of the cases had been performed prior to home respiratory support treatment and only 23.7% had been done during treatment.

A major craniofacial surgery had been performed on 10 patients referring that 71.4% of patients with the craniofacial syndrome had been operated at least once. The craniofacial surgeries were done to treat the craniofacial syndrome and SDB. The most common operative treatment was palatine closure in seven cases. Other surgeries were cranioplasty in five cases, choana atresia surgery in one case and Le Fort three osteotomy in two cases. Five patients had one craniofacial surgery and five patients had two to four craniofacial surgeries.

## 4 | DISCUSSION

There were two centres of interest in this study. First, we described detailed clinical characteristics of children needing home respiratory support. Secondly, the long-term outcomes were investigated.

The diversity of the main diagnosis shows that most children needing home respiratory support have complex medical conditions, in line with previous literature.<sup>12,15,17</sup> The prevalence of neuromuscular disease was significantly lower in our cohort (16.1% vs. 49%–52.2%) compared to previous similar studies.<sup>14,27</sup> As others have described patients needing non-invasive and invasive ventilation,<sup>14,27</sup> we also included conditions with CPAP treatment. This probably explains why we had a smaller percentage (4.7% vs. 12.3–17%) of deaths due to underlying illness compared to others.<sup>19,27</sup> Also, no deaths due to respiratory support fault or loss of airway were reported, and overall severe complications regarding home respiratory treatment were rare. Even though non-invasive methods have become the modern majority, there is still always a small number of patients needing an invasive approach with tracheostomy as 5.3% in this cohort. Like others,<sup>14,15,18,27</sup> we demonstrated, that these patients can also successfully be treated at home.

The dominance of the male sex needing home respiratory therapy found in our study has been demonstrated before as a risk factor for long-term treatment,<sup>1</sup> and invasive ventilation.<sup>14,19,27</sup> Gender differences in studies may be partly explained by the male dominance in progressive neuromuscular diseases, such as Duchenne muscular dystrophy. Also, some co-morbidities are more prevalent in boys and may add to the severity of SDB: male sex has been identified as a risk factor for OSA.<sup>2</sup> Further, the dominance of the male sex in childhood obesity has been described before.<sup>28</sup> In line, in this cohort 92.3% of the severe obesity-related SDB were boys.

The median treatment duration was longer in our study (3.3 years vs. 1.6 years) than previously presented. Also, the discontinuation of the treatment because of disease resolution was more unlikely in our study as it has been previously described (26.7% vs. 57%).<sup>12</sup>



This reflects our practice in the selection of patients with mostly severe SDB persisting after tonsillar surgery. In addition, an increased number of congenital developmental syndromes, such as Down syndrome and acquired obesity have been previously associated with treatment continuance.<sup>1</sup> Congenital conditions causing SDB are more prominent during a young age, as described in a Korean study of children needing home ventilation.<sup>27</sup> In line, the frequency of respiratory support onset was peaking bimodally in our cohort. The peaks can be seen before the age of 1 year and later after and during school age (Figure 2). This likely contributes to the overall treatment duration.

The treatment failure rate of 11.6% in this cohort was in line with previous reports and occurred due to poor adherence to the mask treatment or non-compliance.<sup>12</sup> Somewhat in contrast to a recent meta-analysis,<sup>21</sup> the level of apnoea-hypopnea index was not associated with treatment failure. This might partly be explained by the selected profile of our patients, with a minority of milder spectrum SDB, who may not benefit from CPAP treatment. The mask-interface-related issues were reported in 10–20%. The most common problems were nasal obstruction or skin irritation. These well-known problems tackle adherence and create the need for caretaker education and support. The children in our study had planned clinic visits every 3–6 months. In our experience, most input on these visits should be on adherence issues.

Disease resolution was most dominant in the group of lung-airway conditions with 72.7%. This is mainly explained by the growth and maturation of the underlying conditions such as laryngomalacia or lung hypoplasia, but also by surgical treatment interventions available to some of these patients. In paediatric OSA, adenotonsillectomy is commonly the first line of treatment. However, sometimes SDB is so severe, that home respiratory support treatment is needed briefly prior to surgery. Also, over half of patients with craniofacial syndromes resolved from home breathing support over time. This indicates that surgical interventions and growth play an important role. Thus, in selected children home respiratory support is an adjunct therapy needed only temporarily.

We had a variety of strengths in our study. Our cohort was from a single center covering a population of approximately 200 000 children and all our patients had been evaluated at treatment onset and followed up by a multi-specialty team. We had relatively high-quality data on the severity of the SDB assessed with polysomnography, used devices and complications of the treatment. This increased the accuracy of our results. The multi-specialty team with clinical expertise from different areas of paediatrics has previously been described as good clinical practice while treating children with SDB.<sup>12</sup>

The limitations of our study were related to the retrospective study design. Being a single-centre study, our results may not be generalisable to other populations. Moreover, the high complexity of the patients and severity of SDB limits the generalisation of results outside tertiary-level settings. Due to the retrospective data collection, some essential data, such as detailed device data outputs, were not available lessening the accuracy of our analysis. For instance,

we were unable to analyse objective adherence data during the treatment. Thus, modest adherence may have been present even if treatment was continued persistently. The high heterogeneity of patients also forbids more stratified analysis of factors that possibly contribute to the study outcomes. We also had to simplify the data using the main diagnosis categorisation, though some conditions have characteristics that would fit several main diagnosis groups. This may hide factors affecting the course of respiratory support treatment or distort results regarding specific patient groups.

## 5 | CONCLUSION

In conclusion, this retrospective tertiary-level single-center cohort study provided descriptive data on patients with home respiratory support, their characteristics, long-term outcomes and clinical practices, representing the era of developed device and mask variety and increased use of non-invasive methods. To our understanding, this is the first report covering children with home respiratory support from the Nordic countries.

### AUTHOR CONTRIBUTIONS

**Mervi Järvelä:** Data curation; formal analysis; investigation; methodology; project administration; resources; visualization; writing – original draft; writing – review and editing. **Maija Katila:** Conceptualization; supervision; validation; writing – review and editing. **Vesa Eskola:** Conceptualization; supervision; writing – review and editing. **Riikka Mäkinen:** Methodology; writing – review and editing. **Paula Mandelin:** Conceptualization; writing – review and editing. **Outi Saarenpää-Heikkilä:** Conceptualization; methodology; supervision; writing – review and editing. **Eero Lauhkonen:** Conceptualization; data curation; methodology; project administration; supervision; writing – original draft; writing – review and editing.

### ACKNOWLEDGEMENTS

The authors wish to thank all the children and their parents who participated in this study. The authors will also want to thank university instructor Heini Huhtala, MSc, Health Sciences, Tampere University, who helped during the statistical analyses.

### FUNDING INFORMATION

Funding was provided by Tampere Tuberculosis Foundation, The Finnish Paediatric Research Foundation and The Research Foundation of the Pulmonary Diseases in Finland.

### CONFLICT OF INTEREST STATEMENT

The authors have no conflicts of interest to declare.

### ORCID

Mervi Järvelä  <https://orcid.org/0000-0002-4800-6992>

Maija Katila  <https://orcid.org/0000-0001-6022-2498>

Vesa Eskola  <https://orcid.org/0000-0003-3489-9440>

Eero Lauhkonen  <https://orcid.org/0000-0003-4654-7602>

## REFERENCES

1. Kaditis AG, Alvarez MLA, Boudewyns A, 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:10.1183/13993003.00385-2015
2. Dehlink E, Tan HL. Update on paediatric obstructive sleep apnoea. *J Thorac Dis*. 2016;8(2):224-35. doi:10.3978/J.ISSN.2072-1439.2015.12.04
3. McLaren AT, Bin-Hasan S, Narang I. Diagnosis, management and pathophysiology of central sleep apnea in children. *Paediatr Respir Rev*. 2019;30:49-57. doi:10.1016/j.prrv.2018.07.005
4. Kaditis AG, Alvarez MLA, Boudewyns A, et al. ERS statement on obstructive sleep disordered breathing in 1- to 23-month-old children. *Eur Respir J*. 2017;50(6):1-22. doi:10.1183/13993003.00985-2017
5. Marcus CL. Pathophysiology of childhood obstructive sleep apnea: current concepts. *Respir Physiol*. 2000;119(2-3):143-54. doi:10.1016/S0034-5687(99)00109-7
6. Joosten KF, Larramona H, Miano S, et al. How do we recognize the child with OSAS? *Pediatr Pulmonol*. 2017;52(2):260-71. doi:10.1002/ppul.23639
7. Atag E, Krivec U, Ersu R. Non-invasive ventilation for children with chronic lung disease. *Front Pediatr*. 2020;8(November):1-12. doi:10.3389/fped.2020.561639
8. Hakim F, Kheirandish-Gozal L, Gozal D. Obesity and altered sleep: a pathway to metabolic derangements in children? *Semin Pediatr Neurol*. 2015;22(2):77-85. doi:10.1016/j.spen.2015.04.006
9. Sullivan CE, Berthon-Jones M, Issa FG, Eves L. Reversal of obstructive sleep apnoea by continuous positive airway pressure applied through the nares. *Lancet*. 1981;317(8225):862-5. doi:10.1016/S0140-6736(81)92140-1
10. Sanders MH, Kern N. Obstructive sleep apnea treated by independently adjusted inspiratory and expiratory positive airway pressures via nasal mask. Physiologic and clinical implications. *Chest*. 1990;98(2):317-24. doi:10.1378/CHEST.98.2.317
11. Mandereau-Bruno L, Léger D, Delmas MC. Obstructive sleep apnea: a sharp increase in the prevalence of patients treated with nasal CPAP over the last decade in France. *PLoS One*. 2021;16:e0245392. doi:10.1371/journal.pone.0245392
12. Edwards EA, Hsiao K, Nixon GM. Paediatric home ventilatory support: the auckland experience. *J Paediatr Child Health*. 2005;41:652-8.
13. Amin R, Al-Saleh S, Narang I. Domiciliary noninvasive positive airway pressure therapy in children. *Pediatr Pulmonol*. 2016;51(4):335-48. doi:10.1002/ppul.23353
14. Racca F, Berta G, Sequi M, et al. Long-term home ventilation of children in Italy: a national survey. *Pediatr Pulmonol*. 2011;46(6):566-72. doi:10.1002/PPUL.21401
15. Cancelinha C, Madureira N, Mação P, et al. Long-term ventilation in children: ten years later. *Rev Port Pneumol*. 2015;21(1):16-21. doi:10.1016/j.rppnen.2014.03.017
16. Paulides FM, Plötz FB, Verweij-Van Den Oudenrijn LP, et al. Thirty years of home mechanical ventilation in children: escalating need for pediatric intensive care beds. *Intensive Care Med*. 2012;38:847-52. doi:10.1007/s00134-012-2545-9
17. Amin R, Sayal P, Syed F, Chaves A, Moraes TJ, MacLusky I. Pediatric long-term home mechanical ventilation: twenty years of follow-up from one Canadian center. *Pediatr Pulmonol*. 2014;49(8):816-24. doi:10.1002/ppul.22868
18. Appierto L, Cori M, Bianchi R, et al. Home care for chronic respiratory failure in children: 15 years experience. *Paediatr Anaesth*. 2002;12(4):345-50. doi:10.1046/j.1460-9592.2002.00856.x
19. Gowans M, Keenan HT, Bratton SL. The population prevalence of children receiving invasive home ventilation in Utah. *Pediatr Pulmonol*. 2007;42(3):231-6. doi:10.1002/ppul.20558
20. Tibballs J, Henning R, Robertson CF, et al. A home respiratory support programme for children by parents and layperson carers. *J Paediatr Child Health*. 2010;46(1-2):57-62. doi:10.1111/j.1440-1754.2009.01618.x
21. Blinder H, Momoli F, Holland SH, Blinder A, Radhakrishnan D, Katz SL. Clinical predictors of nonadherence to positive airway pressure therapy in children: a retrospective cohort study. *J Clin Sleep Med*. 2021;17(6):1183-92. doi:10.5664/jcsm.9162
22. 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
23. Berry RB, Brooks R, Gamaldo CE, et al. AASM | Scoring Manual Version 2.2 The AASM Manual for the scoring of sleep and associated events rules, terminology and technical specifications version 2.2. 2015. Accessed 17 December 2022. [www.aasmnet.org](http://www.aasmnet.org)
24. Grigg-Damberger MM. The visual scoring of sleep in infants 0 to 2 months of age. *J Clin Sleep Med*. 2016;12(3):429-45. doi:10.5664/jcsm.5600
25. Sateia MJ. International classification of sleep disorders-third edition: highlights and modifications. *Chest*. 2014;146(5):1387-94. doi:10.1378/CHEST.14-0970
26. Savini S, Ciorba A, Bianchini C, et al. Assessment of obstructive sleep apnoea (OSA) in children: an update Valutazione critica del bambino con apnea ostruttiva notturna PAROLE CHIAVE: Apnea ostruttiva notturna • OSA • Bambini • Management • Polissonografia. *Acta Otorinolaryngol Ital*. 2019;39:289-97. doi:10.14639/0392-100X-N0262
27. Characteristics of children using HMV Pediatric Home Mechanical Ventilation. 34. doi:10.3346/jkms.2019.34.e268
28. Shah B, Cost KT, Fuller A, Birken CS, Anderson LN. Sex and gender differences in childhood obesity: contributing to the research agenda. *BMJ Nutr Prev Health*. 2020;3:387-90. doi:10.1136/bmjnph-2020-000074

## SUPPORTING INFORMATION

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

**How to cite this article:** Järvelä M, Katila M, Eskola V, Mäkinen R, Mandelin P, Saarenpää-Heikkilä O, et al. Finnish children who needed long-term home respiratory support had severe sleep-disordered breathing and complex medical backgrounds. *Acta Paediatr*. 2023;00:1-8. <https://doi.org/10.1111/apa.16981>