

Home Mechanical Ventilation in Children: A 7-year Experience

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ABSTRACT

Aims and objectives: To review the profile of children requiring home mechanical ventilation (HMV), the diagnoses, modes of presentation, age at initiating HMV, and outcome of these children on follow-up.

Materials and methods: This is a retrospective observational study. We included all children up to 18 years of age who were started on HMV between May 2013 to April 2020 at our hospital. Source of data was the hospital records of children receiving HMV. Clinical data were captured on Excel sheet and analyzed.

Results: Fifty-seven children were started on HMV with a mean age of 6.43 years (range of 3 months to 17 years 8 months) at start of HMV; 35 (61.4%) were male and 22 (38.6%) were female. Fifteen (17.5%) of 57 presented with acute respiratory failure, and 42 (73.6%) of 57 with chronic respiratory failure. Thirty-nine (68.4%) of 57 had an established diagnosis of a neuromuscular disease (NMD) of 22 (56.41%) of 39 had an underlying diagnosis of spinal muscular atrophy 2 (SMA type II) which was also the most common diagnosis in this study. Average age at initiating ventilation in the neuromuscular group was 7.92 years (range: 4 months–17 years 8 months) and the other non-NMD group was 2.91 years (range: 3 months–15 years). Seven children were on invasive tracheostomy and 50 on noninvasive ventilation (NIV). Fifty-six children were started on BiPAP, and one child is on CPAP. Two children have been weaned of HMV and are doing well. There were three episodes of life-threatening complications in three different children, and the annual rate of hospitalization with respiratory morbidity was 0.36 per child. There has been no mortality in this study period.

Conclusion: Home mechanical ventilation improves the life expectancy and enhances the quality of care and survival. Under the appropriate clinical scenarios, HMV significantly reduces the economic, psychosocial burden on the family and improves quality of life for the child. Transition to home care is challenging, especially in developing countries with lack of nursing care/home care support, but is feasible with meticulous planning wherein parents or caregivers are the key partners. To the authors knowledge, this is the largest case series of children on HMV from India.

Keywords: Bilevel positive airway pressure, Continuous positive airway pressure, Chronic respiratory failure, Home mechanical ventilation.

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INTRODUCTION

Advances in intensive care interventions and infrastructure have improved survival in children with various causes of respiratory failure (acute and chronic). However, with this, there has been an increase in use of home mechanical ventilation (HMV) in the pediatric population.¹ This has further improved the health-related quality of life for these children.² These children often require prolonged frequent hospitalizations, but once the child is clinically stable, home care management is ideal. HMV caters to the emotional and psychological need of the child, reduces the chance of life-threatening nosocomial infections, and the financial burden on the family.³ Conditions leading to HMV dependency include congenital airway malformations, chronic lung disease, hypoventilation syndrome, neuromuscular diseases, and spinal cord injuries.⁴ Ventilation at home can be provided through tracheostomy tube or noninvasively through a mask. The family plays the most important role in providing routine care to a child on HMV. There is scarce data from developing countries on children on HMV, and this study aims to review the profile and outcome of these children.

AIMS AND OBJECTIVES

Aim of the study was to review the profile of children requiring HMV, the diagnoses, modes of presentation, age of initiation of HMV, and outcome of these children on follow-up.

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MATERIALS AND METHODS

This is a retrospective observational study. All children up to 18 years of age who were started on HMV between May 2013 and April 2020 were included in the study. Source of data was the hospital records of children receiving HMV. Clinical data were captured on excel sheet and analyzed. A multidisciplinary team that comprised of the general pediatrician, pediatric pulmonologist, pediatric neurologist, pediatric intensivist, otolaryngologist, and pediatric cardiologist along with the physiotherapy and nutrition team evaluated these children at different points of time as needed clinically.

Informed consent was taken for every patient before the start of HMV with adequate and appropriate counseling of caregivers. In all children, ventilatory support was initiated in the hospital.

Prior to discharge, ventilator settings were determined by overnight monitoring of oxygen saturation and partial pressure of carbon dioxide (PaCO₂) in blood or end tidal carbon dioxide (ETCO₂) along with titration of the ventilator settings. Supplemental O₂ was added only if the oxygen saturation by pulse oximetry (SpO₂) could not be maintained >92% overnight with optimal CO₂ control.

All tracheostomized children had portable suction machines and equipment necessary for humidification and nebulization, and caregivers were also taught chest physiotherapy, equipment care, and tracheostomy tube care. All the caregivers were taught basic cardiopulmonary resuscitation (CPR) with repeated reinforcements prior to discharge.

Tracheostomized children were also admitted at periodic intervals for tube changes. Caregivers observed and performed supervised change of tracheotomy tubes in case of an emergency or inadvertent tube removal. Counseling and psychological support were provided as part of the protocol.

The children were followed up regularly starting weekly postdischarge, then 2–3 monthly for tracheostomy patients, and 3–6 monthly for NIV patients. A regular review for change in ventilator settings individualized to each patient based on growth and development, clinical work of breathing, blood gases, end tidal carbon dioxide, and annual sleep studies wherever feasible were done.

In person follow-up was done for 42 patients. Those children from outstation locations have been followed up via video consults and liaisons with their local pediatricians.

RESULTS

Fifty-seven children were included in the study. Mean age of the children at start of HMV was 6.43 years (range: 3 months–17 years 8 months); 35 (61.4%) were male and 22 (38.6%) were female. Fifteen (17.5%) of 57 presented with acute respiratory failure, and 42 (73.6%) of 57 presented with chronic respiratory failure. The overall percentage of children with neuromuscular disease was 39 (68.4%) of 57, followed by cardiopulmonary disease 6 (10.5%) of 57, congenital anomaly in 6 (10.5%) of 57, airway involvement in 3 (5.26%) of 57, metabolic/degenerative cause in 2 (3.5%) of 57, and other causes in 2 (3.5%) of 57 (Table 1 and Figs 1 and 2).

Ten (17.54%) of 57 children had the home ventilation initiated at age less than 1 year, with the youngest child being 3 months of age with a diagnosis of distal 10q trisomy and 12p monosomy syndrome. Twenty (35%) of 57 children had HMV initiated between 1 year and 5 years, 11 (19.29%) of 57 children between 5 and 10 years, and 16 (28.07%) of 57 children were more than 10 years (Fig. 3).

In all, 22 (56.41%) of 39 children had a underlying diagnosis of spinal muscular atrophy 2 (SMA2) which was the most common diagnosis followed by 10 (25.6%) of 39 children with Duchenne Muscular Dystrophy (DMD). Average age of start of ventilation in the neuromuscular group was 7.92 years (range: 4 months to 17 years 8 months) and in the other non-neuromuscular disease was 2.91 years (range: 3 months to 15 years) (Fig. 3).

Various types of interfaces were used. Fifty children were started on noninvasive ventilation, with 5 children on nasal mask/pillow and 45 children using oronasal mask as interface.

Table 1: Underlying disease

Disease classification	Diagnosis	n	Total (n)
NMD	Spinal muscular atrophy 1	2	38
	Spinal muscular atrophy 2	22	
	Duchenne muscular dystrophy	10	
	Unknown myopathy	2	
	Myotonic dystrophy	1	
	Chronic inflammatory demyelinating polyneuropathy	1	
Cardiopulmonary	Congenital laryngomalacia, cardiomyopathy, pulmonary hypertension	1	6
	Congenital heater disease with pulmonary hypertension	1	
	Interstitial lungs disease	1	
	Chronic lung disease, pulmonary hypertension	3	
Congenital anomaly	Down syndrome	1	6
	Apert syndrome	1	
	Prune belly syndrome	1	
	Joubert syndrome	1	
	Distal 10q trisomy 12p13 deletion	1	
	Pallister-Killian syndrome	1	
Airway	Obstructive sleep apnea	2	3
	Congenital subglottic stenosis	1	
Metabolic/degenerative	Mucopolysaccharidosis 4	1	2
	Metabolic myopathy	1	
Other	Diabetic ketoacidosis with cardiac failure	1	2
	Traumatic brain injury with hydrocephalus	1	

Seven children were on invasive tracheostomy. Fifty-six children were started on BiPAP and 1 child on CPAP with a diagnosis of obstructive sleep apnea (OSA) due to enlarged tonsils and adenoids and pulmonary hypertension. Two (3.5%) patients received HMV throughout the day, whereas 55 (96.4%) received HMV only during sleep (Flowchart 1).

Follow Up

In-person follow-up was done for 42 patients.

Complications

Three children had compliance issues with using interfaces for home ventilation. The annual rate of hospitalization with respiratory morbidity was 0.36 per child. One child has had a life-threatening complication of a tube block but brought immediately to the hospital. Two children had tracheal bleed from granulation tissue due to tracheostomy tube.

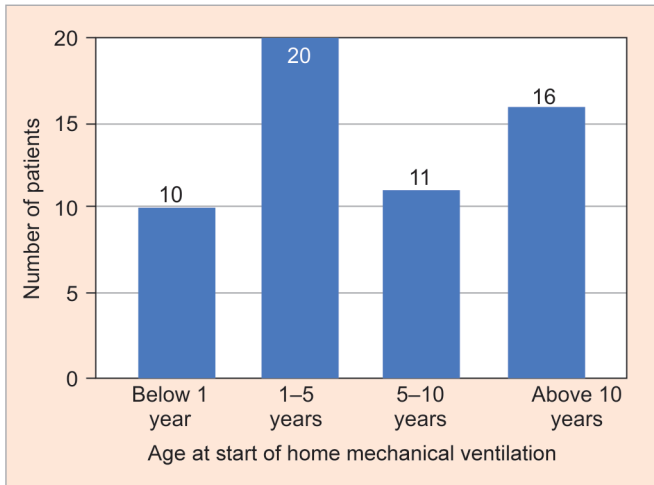


Fig. 1: Age of children at initiation of home mechanical ventilation

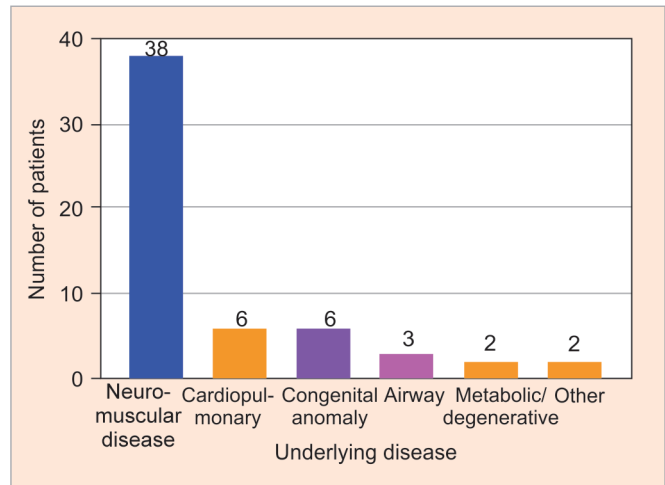


Fig. 2: Distribution of number of patients based on underlying disease

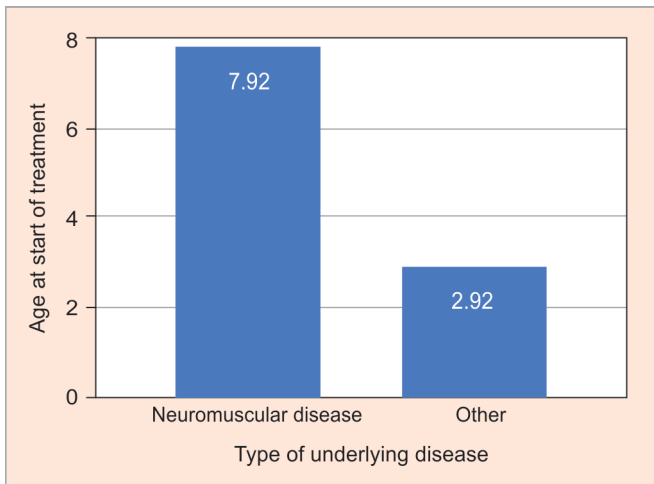


Fig. 3: Average age at start of home mechanical ventilation in the neuromuscular disease group vs other group

Flowchart 1: Type of home mechanical ventilation and interface

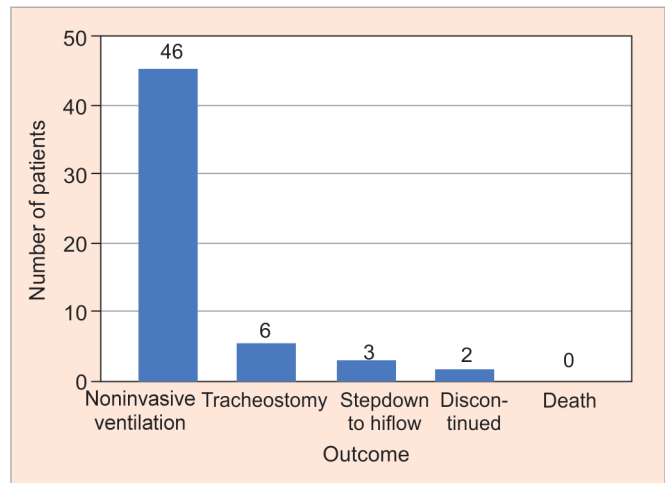
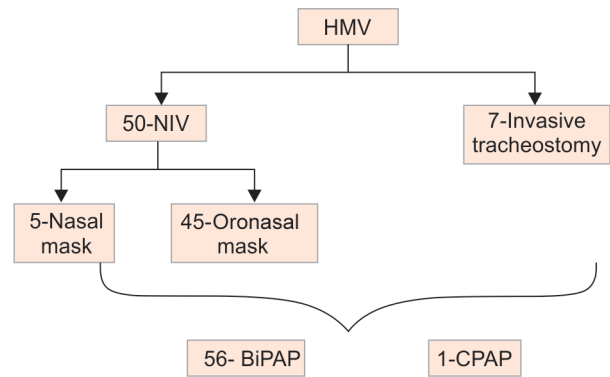


Fig. 4: Outcomes at end of this study period

Outcomes

Forty-six children are continuing HMV via noninvasive ventilation, 6 children via tracheostomy, and 3 children were weaned from BiPAP to high-flow oxygen. Two children with a diagnosis of cardiomyopathy, laryngomalacia, pulmonary arterial hypertension, and the other with chronic lung disease with congenital heart disease (COA, ASD, VSD, and PDA) pulmonary arterial hypertension are off HMV from the age of 4.5 and 5 years, respectively. There has been no mortality in our case series (Fig. 4).

DISCUSSION

The balance between the airways, the lung along with the respiratory muscles, and the central drive maintains a synchronized respiration. Respiratory insufficiency can be due to increased respiratory load due to airway or lung disease, muscle weakness due to neuromuscular or restrictive chest wall disease, and failure of central drive.¹

HMV can be initiated in a child presenting in acute or chronic respiratory failure, when there is failure to wean off ventilation, to decrease the duration of stay at hospital, reduce respiratory illnesses, morbidity, and to improve quality of life in children.

There are various clinical entities presenting with airway involvement, parenchymal lung disease, neuromuscular weakness, restrictive lung disease, and obstructive and central apnea.^{1,5}

In clinical practice, home ventilation via tracheostomy is suitable for children who are clinically stable, requiring more than 16 hours of ventilation per day, or in infants and children who cannot tolerate even brief periods of discontinuation of ventilatory support. NIV is initiated in children who can tolerate short periods



off ventilator, requiring support mainly during sleep with abnormal nocturnal oxygen and carbon dioxide levels, and in children with a high apnea–hypoapnea index on polysomnography (PSG).^{5,6}

In our study, PSG was done in 37 children before start of HMV, and these were the children presenting in chronic respiratory failure.

Children can be ventilated at home by invasive ventilation via tracheostomy or NIV via positive pressure or negative pressure. The negative pressure NIV is cumbersome and is no longer available and of historical importance.⁷ In this article, NIV refers to positive pressure NIV, which are of two types: CPAP which is used in isolated upper or lower airway involvement and BiPAP to assist weakened respiratory muscles and central drive.¹ There are three different modes of BiPAP ventilation: spontaneous (S) mode, spontaneous/times (S/T) mode, and timed (T) mode.⁸ In our study, there is one patient with a diagnosis of OSA and pulmonary hypertension who is on CPAP and rest all are on BiPAP on S/T mode.

The various types of interfaces available for NIV include nasal mask, oronasal mask, mouthpiece, total face mask, nasal pillows, and hybrid mask which is a combination of nasal pillows with oral masks.⁷ Oronasal mask was mostly used in our patients followed by nasal masks. NIV gives better patient comfort, has decreased chance of complications, and less strain on the family. Tracheostomy (invasive ventilation) requires extra care with suctioning, maintenance of the tracheostomy tube, and associated with life-threatening complications, such as tube block or tracheal bleed secondary to granulations.

The choice between invasive and noninvasive ventilation depends on the underlying etiology or clinical presentation; pathology whether airway, lung parenchymal, neuromuscular, or central drive is affected; and availability of appropriate noninvasive interfaces ability to tolerate short breaks. In addition, psychosocial and financial factors are important aspects that may determine the long-term goals for individual patients.

In our study, the most common cause for initiation of HMV was NMD which is in agreement with other studies.^{9–11} In all, 40.74% of the children had a underlying diagnosis of SMA2, which was also the commonest diagnosis in this study. There was a significant difference between the age of initiation of HMV between the neuromuscular patients and the other groups.

In all, 16.67% of the children had HMV initiated at age less than 1 year who were mostly from the non-neuromuscular disease group which is similar to other studies.¹²

The most common reason for hospitalization was respiratory infections. Life-threatening complication of tube block occurred in one child and tracheal bleed secondary to granulation tissue in two children. Respiratory infections can be avoided with hand hygiene, clean handling of tracheal tube, and stringent infection protocols. Two children have been weaned from BiPAP to high-flow oxygen. Two children have been successfully weaned of ventilation, are doing well, and going to school. One child who is off BiPAP at present, had required 16 hospital admission prior to initiation of ventilatory support, and only 2 thereafter. All patients are alive during the study period. This shows that there is a relatively low rate of life-threatening complications in our study, and children with only cardiopulmonary disease or upper airway obstruction may outgrow the need for home ventilation requirement.

Poor compliance with HMV has been reported in many articles.^{1,13,14} However, in our study, only three patients were documented to have compliance issues, and most patients are currently tolerating HMV well.

From our experience, the key factors for success of HMV are a multidisciplinary approach; careful patient selection; algorithmic approach in initiation, use, titration, and discontinuation of HMV; ensuring patient comfort and addressing dys-synchrony effectively; and monitoring for complications with a structured regular follow-up. Equal emphasis meted out to motivate and train caregivers as partners, and strengthen parental training with confidence, education and reinforcement during each and every follow-up.

The advantages of HMV are better psychosocial environment for the child and family, improvement in the families quality of life along with the child, decrease in healthcare costs, and rate of nosocomial infections.¹⁵ These advantages compared to the lack of life-threatening complications, and possibility of training caregivers without expert nursing services, form a very important aspect of care in a developing country like India, where HMV is an actual cost-effective option. Despite this, there is a lack of use of HMV and research in this area because of lack of trained expertise and knowhows of starting HMV. The biggest challenges were initiating those children with chronic respiratory failure, as most of them were not aware of the long-term implications of the disease process itself. Another crucial factor was putting these caregivers into support groups with parents who were already initiated on HMV providing feedback, education, and the added advantages of supporting these patients. Although this factor cannot be quantitated, this worked in favor for each child, where parents were extremely hesitant about the initiation of HMV and the long-term care. Needless to say, we were able to use this as advantage for these patients. In the opinion of the author, this is the largest case series from India of children on HMV.

LIMITATIONS AND FUTURE DIRECTIONS

Our study is limited by its retrospective design. The data on criteria for initiation of HMV, pre-HMV hospitalization rate, morbidity, and quality-of-life index have not been captured in this paper.

Quantitating quality of life, socioeconomic burden, and psychological factors would be ideal but very challenging, given the variable case scenarios, variable clinical presentations as well as the challenges in follow-up for these patients.

CONCLUSION

Home mechanical ventilation extends life expectancy and improves quality of care and survival. Advances in ventilator technology and a growing experience and acceptance of home care have increased the possibilities for discharging children requiring long-term ventilation from hospital to home. It has its set of challenges, however, as it significantly reduces the economic, psychosocial burden on the family and improves quality of life for many whose disease may be progressive with unpredictable hospitalizations and poor quality of life. Transition to home care is challenging, especially in developing countries with lack of trained personnel, knowledge, and additional burden of nursing care but is feasible with meticulous planning and motivated parents or caregivers are. HMV requires appropriate clinical setting, clear knowledge and understanding of the clinical course, and a dedicated team to motivate, trouble shoot, and guide the patients.

REFERENCES

1. Amaddeo A, Frapin A, Fauroux B. Long-term non-invasive ventilation in children. *Lancet Respir Med* 2016;4(12):999–1008. DOI: 10.1016/S2213-2600(16)30151-5.

2. Brookes I. Long-term ventilation in children. *Paediatr Child Health (Oxford)* 2019;4(29). DOI: 10.1016/j.paed.2019.01.013.
3. Hewitt-Taylor J. Children who require long-term ventilation: staff education and training. *Intensive Crit Care Nurs* 2004;20(2):93–102. DOI: 10.1016/j.iccn.2003.09.002.
4. Lewarski JS, Gay PC. Current issues in home mechanical ventilation. *Chest* 2007;132(2):671–676. DOI: 10.1378/chest.07-0558.
5. Amaddeo A, Moreau J, Frapin A, et al. Long term continuous positive airway pressure (CPAP) and noninvasive ventilation (NIV) in children: initiation criteria in real life. *Pediatr Pulmonol* 2016(9):51. DOI: 10.1002/ppul.23416.
6. MacLusky I, Amin R, Zielinski D. Section 2: initiation of in-hospital long-term mechanical ventilation and transition to home. *Can J Respir Crit Care, Sleep Med [Internet]* 2018;2(Suppl. 1):5–15. DOI: 10.1080/24745332.2018.1494490.
7. Hess DR. Noninvasive ventilation for neuromuscular disease. *Clin Chest Med* 2018;39(2):437–447. DOI: 10.1016/j.ccm.2018.01.014.
8. Alexiou S, Piccione J. Neuromuscular disorders and chronic ventilation. *Semin Fetal Neonatal Med* 2017;22(4):256–259. DOI: 10.1016/j.siny.2017.04.001.
9. Kennedy JD, Martin AJ. Chronic respiratory failure and neuromuscular disease. *Pediatr Clin North Am* 2009;56(1):261–273. DOI: 10.1016/j.pcl.2008.10.011xii.
10. Paulides FM, Plötz RB, van den Oudenrijn LPV, et al. Thirty years of home mechanical ventilation in children: escalating need for pediatric intensive care beds. *Intensive Care Med* 2012;38(5):847–852. DOI: 10.1007/s00134-012-2545-9.
11. Preutthipan A. Home mechanical ventilation in children. *Indian J Pediatr* 2015;82(9):852–859. DOI: 10.1007/s12098-015-1842-z.
12. Ottonello G, Ferrari I, Pirroddi IMG, et al. Home mechanical ventilation in children: retrospective survey of a pediatric population. *Pediatr Int* 2007;49(6):801–805. DOI: 10.1111/j.1442-200X.2007.02463.x.
13. Marcus CL. Adherence to and effectiveness of positive airway pressure therapy in children with obstructive sleep apnea. *Pediatrics [Internet]* 2006;117(3):e442–e451. DOI: 10.1542/peds.2005-1634 Available from: <http://pediatrics.aappublications.org/cgi/doi/10.1542/peds.2005-1634>.
14. Marcus CL, Radcliffe J, Konstantinopoulou S, et al. Effects of positive airway pressure therapy on neurobehavioral outcomes in children with obstructive sleep apnea. *Am J Respir Crit Care Med [Internet]* 2012;185(9):998–1003. DOI: 10.1164/rccm.201112-2167OC Available from: <https://pubmed.ncbi.nlm.nih.gov/22323303>.
15. Norregaard O. Noninvasive ventilation in children. *Eur Respir J [Internet]* 2002;20(5):1332–1342. Available from: <http://erj.ersjournals.com/cgi/doi/10.1183/09031936.02.00404802>.