Thirteen Years of Invasive and Noninvasive Home Ventilation for Children in a Developing Country: A Retrospective Study

Anna Marie Nathan, MRCPCH,1,2* Hui Yan Loo, MBBS,3 Jessie Anne de Bruyne, MRCP,1,2 Kah Peng Eg, MMed,1,2 Sze Ying Kee, MRCPCH,4 Surendran Thavagnanam, FRCPCH,1,2 Marilyn Bouniu, MBBS,5 Jiat Earn Wong, MBBS,3 Chin Seng Gan, MMed,1,2 and Lucy Chai See Lum, MRCP1,2

Summary. Introduction: Home ventilation (HV) for children is growing rapidly worldwide. The aim was to describe (1) the sociodemographic characteristics of children on HV and (2) the indications for, means and outcome of initiating HV in children from a developing country. Methodology: This retrospective study included patients sent home on noninvasive or invasive ventilation, over 13 years, by the pediatric respiratory unit in a single center. Children who declined treatment were excluded. Results: Seventy children were initiated on HV: 85.7% on noninvasive ventilation, 14.3% on invasive ventilation. There was about a threefold increase from 2001–2008 (n = 18) to 2009–2014 (n = 52). Median (range) age of initiating HV was 11 (1–169) months and 73% of children were <2 years old. Common indications for HV were respiratory (57.2%), chest/spine anomalies (11.4%), and neuromuscular (10.0%). Fifty-two percent came off their devices with a median (interquartile range) usage duration of 12 (4.8, 21.6) months. Ten children (14.3%) died with one avoidable death. Children with neuromuscular disease were less likely to come off their ventilator (0.0%) compared to children with respiratory disease (62.1%). Forty-one percent of parents bought their equipment, whereas 58.6% borrowed their equipment from the medical social work department and other sources. Conclusion: HV in a resource-limited country is possible. Children with respiratory disease made up a significant proportion of those requiring HV and were more likely to be weaned off. The mortality rate was low. The social work department played an important role in facilitating early discharge. Pediatr Pulmonol. 2016; 9999:XX–XX.

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INTRODUCTION

Changing expectations of both parents and healthcare professionals regarding the health-related quality of life of a child, in the presence of a disability, together with technological advancement has resulted in an exponential rise in children being sent home on home ventilation (HV) throughout the world.1–6 It is estimated that the prevalence of children on HV ranges from 4.2/100,000 in Italy,1 8.1/100,000 in the United Kingdom to as high as 13.7/100,000 children in Australia and New Zealand.7

In the chronic setting, there are many groups of children who benefit from HV, for example, chronic lung disease with respiratory failure, neuromuscular disease with progressive weakness, severe kyphoscoliosis with alveolar hypoventilation, and central ventilatory control disorders.8 If not for HV, these children would have a prolonged and complicated stay in hospital, resulting in other problems like developmental delay, family

1Department of Pediatrics, University of Malaya, Kuala Lumpur, Malaysia.
2University Malaya Pediatric and Child Health Research Group, University of Malaya, Kuala Lumpur, Malaysia.
3University of Malaya, Kuala Lumpur, Malaysia.
4Department of Pediatrics, Universiti Putra Malaysia, Serdang, Malaysia.
5Ministry of Health, Malaysia.

Conflict of interest: AMN, JdB, and ST have all received some financial support, from local companies dealing with home ventilators, for the organization of continuing medical education courses in Kuala Lumpur, Malaysia.

*Correspondence to: Anna Marie Nathan, MRCPCH, Department of Pediatrics, University Malaya Medical Centre, 50603 Kuala Lumpur, Malaysia. E-mail: psr9900@hotmail.com

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separation, increased mortality from nosocomial infections, and rising hospitalization cost.9 As such, this is a service worth investing time and money as a price cannot be placed on the improved quality of life of the child.10,11 not forgetting the other tangible benefits like reduced hospitalization days, reduced intensive care days, and reduced readmission.12,13

However, there are many challenges in running this service. First, it is not inexpensive to initiate someone on HV. There are also other hidden costs, for example, maintenance of equipment and need for accessories and consumables. Second, one requires a caregiver willing and able to learn the special skills required and perhaps even give up their profession to care for their child. In addition, there is significant impact upon the rest of the family. Therefore, discharging a child on HV requires tremendous amount of organization by a multidisciplinary team and subsequent close follow-up, as recently outlined in a comprehensive document from the American Thoracic Society.14

Although many international studies have reported on their HV service, most have been from developed countries.1–3,5,15 In Malaysia, the HV program for children was first initiated in two tertiary centers: University Malaya Medical Centre (UMMC) and General Hospital of Kuala Lumpur (GHKL), in 2001.

Our hypothesis is that HV is a viable option with a favorable outcome, even in a developing country like Malaysia. Hence, the aim of this study was to determine the sociodemographic characteristics, indications for and outcome of children initiated on HV. We also wanted to highlight the ways and means of initiating HV in a developing country.

MATERIALS AND METHODS

This is a retrospective study involving all patients sent home on HV, from the program’s inception in May 1, 2001 till December 31, 2014. We included patients who were initiated on HV by the Pediatric Respiratory Unit, University Malaya Medical Centre (UMMC), Kuala Lumpur, Malaysia. HV in this study includes noninvasive ventilation (NIV) using a mask interface and invasive ventilation (IV).7 Children who were on CPAP for obstructive sleep apnea and declined treatment were excluded. Patients were identified from the Pediatric Home Ventilation database. Patient case folders were reviewed in hard copy or from the online computer database. Medical Social Work Department (MSW) case files were also reviewed. Medical Ethics approval was obtained from the local hospital ethics committee (MEC no 201312-0617). Informed consent was not obtained as this was a retrospective study; however, anonymity of the patients was preserved.

Research Setting

UMMC is a 1,000-bedded tertiary referral center, situated in Kuala Lumpur, the capital city of Malaysia. It has a separate pediatric complex, with a 100 bed capacity, including a 10-bedded pediatric intensive care unit (PICU). As a tertiary referral center, we receive referrals from all over the country. Besides the PICU, each general ward has a four-bedded high-dependency unit where patients can be initiated on NIV and at any time, a maximum of six patients can be ventilated on NIV depending on the level of nursing care required.

Health Care System and How We Do It

In Malaysia, healthcare is subsidized by the government in public hospitals. Financial support for patients who require HV can come from various government and private sources. However, there are no subsidies for private nursing care and once the patient has gone home, the other expenditure is borne by the family.

The MSW department of UMMC has an invaluable role in facilitating the HV program and provided data regarding the following: types of devices loaned, duration of loan, duration of approval from the various funding agencies, amount of money approved by the funding agencies, other sociodemographic details, for example, household income, type of housing, home visit, and return of ventilatory device to the MSW department once not required.

A patients is deemed fit for discharge once: (1) medically stable on their devices, (2) the patient has satisfactory growth and feeding practices, (3) the home environment is satisfactory, (4) the caregiver is ready,16 and (4) all equipment and necessary consumables are available and in working order.

Definitions

HV, for the purpose of this study, is defined as either IV or NIV. Ventilatory assistance was delivered via either NIV (including continuous positive airway pressure [CPAP]), via a mask interface, or IV via tracheostomy. Diagnoses were classified into the following categories: neuromuscular, respiratory (upper and lower), cardiac, abnormal ventilatory control, chest/spine anomalies, and spinal cord injury.6 Duration of HV was calculated from the time the patient was first sent home on HV until complete cessation. Partial dependency on HV was defined as the usage of ventilatory assistance for less than 12 hr a day or only at night and/or during sleep.

Data Collection

The following clinical data were collected: (1) Primary diagnosis requiring respiratory support, adapted from Wallis et al.,6 (2) age of initiation and duration of

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hospitalization before discharge home, (3) duration of
respiratory support at home (partial, i.e., <12 hr or total
≥12 hr/day), (4) type of respiratory support (IV, bilevel
positive airway pressure [BIPAP], CPAP), (5) adjunct
therapy and other devices (supplemental oxygen, suction
machine, feeding tube), (6) details of home care
providers, for example, level of education, income (7)
admission for respiratory tract infections, and (8) final
outcome, that is, alive, dead, duration of HV.

Statistics

The data were analyzed using IBM SPSS Statistics 20
software. Data were described using percentage, median,
interquartile range (IQR), and/or range, if not normally
distributed. The chi-squared test or Fisher’s exact test
(where appropriate) was used to perform univariate
analysis between categorical variables. The Mann–
Whitney U-test was used to compare nonparametric
continuous variables. A P-value of <0.05 was considered
to be statistically significant. Measure of association
was presented as odds ratios (OR) with 95% confidence
intervals (CI).

RESULTS

Seventy-nine children were initiated on HV from
May 1, 2001 till December 31, 2014. Two patients
decayed treatment and the seven who required CPAP for
OSA were excluded from the analysis. Finally data on the
seventy children were analyzed (see Fig. 1).

The racial distribution was similar to the national
demographics of Malaysia: 47 (67.1%) Malays, 15
(21.4%) Chinese, 7 (10.0%) Indians and others, 1
(1.4%).17 The median age of initiating HV was about
12 months with more than two-thirds (n = 51/70, 72.9%)
of children aged 2 years and below at the time of
initiation. Fifty-one percent of fathers (n = 23/45) and
54% (n = 25/46) mothers had tertiary education. Thirty-
nine percent of children (n = 27/69) came from single-
income families. The median (range) monthly income of
these families was USD 1,693.0 (266.0–6,000.0). Primary
caregivers for the majority of the patients were their
parents (n = 50/59, 83.0%), whereas three children
(5.1%) were cared for by domestic helpers and seven
(11.9%) by relatives. All children resided at home as there
are no long-term care facilities for patients requiring
respiratory support in Malaysia. Duration of hospitaliza-
tion (n = 60) before discharge home on HV was median
(IQR) 65 (33, 134) days. Children requiring IV stayed
significantly longer with median (IQR) of 137 (93, 217)
days than those requiring NIV who stayed median (IQR)
59 (30, 114) days (z = −2.30, P = 0.02).

Only 14.3% of children required IV, whereas the rest
were on NIV (see Table 1). Only one child on IV via a
tracheostomy used a conventional ventilator, whereas the
other nine used BIPAP as this was more economical and
equally effective.

Clinical indications for HV are as follows: increased
work of breathing (n = 38, 54.3%), hypventilation
(n = 22, 31.4%), hypoxia (n = 4, 5.7%), and uncontrolled
heart failure (n = 6, 8.6%). Forty-one percent (n = 29/70)
required oxygen therapy in addition to their ventilators.
As for other ancillary equipment, 59.0% (n = 39/66) of
children required enteral feeding, whereas 83.0% (n = 39/
47) required a suction machine for airway clearance.

Reasons for initiating HV are shown in Table 1 and the
main diagnosis was respiratory disease. On the other
hand, spinal cord injury was the commonest indication for
invasive ventilation. More than half of the children were
partially dependent (<12 hr/day) on the ventilator.

As for the outcome, about 45% of children were
weaned off their devices (n = 30/67, 44.8%), 40.3% (n = 27/67) are still on their devices, whereas 14.9%
(n = 10) have died. Median (IQR) duration of use of HV
was 12.0 (4.8, 21.6) months if they were weaned off their
device and median (IQR) 15.6 (2.0, 36.0) months if they
died. Children with neuromuscular disease were less
likely to come off their ventilator (n = 0/5 survivors)
compared to children with respiratory disease (n = 23/37
survivors, 62.2%) with a P = 0.02. Fifty-four percent of
children (n = 26/48) required readmissions for respira-
tory-related issues with a median (range) number of two
(1–6) episodes while on HV.

Of the 10 children who died, only one was an avoidable
death, due to accidental disconnection from the ventilator
while asleep. One patient died of renal failure and one
died of a brain hemorrhage, well after she had come off
HV. The other seven had progressive lung disease: SMA
type 1 (n = 2), severe lung hypoplasia (n = 4), and
worsening bronchiectasis (n = 1).

Fig. 1. Study flow chart. *Bilevel positive airway pressure*©continuous positive airway pressure.
Since 2009, there has been a significant increase in the number of children requiring HV in our institution. From 2009 to 2014, 74.3% (n = 52) of children were initiated on HV compared to 25% (n = 18) from 2001 to 2008. Although the number of children with neuromuscular disease requiring HV has remained fairly stable over the past 13 years, the number with respiratory disease has risen dramatically (see Fig. 2).

There has been an increasing usage of CPAP instead of BIPAP. In 2013–2014, CPAP was used in 43.2% (n = 19/31) of children and BIPAP in 29.0% (n = 9/31) (Fig. 3).

The indication for CPAP (2013–2014) was more for lower airway disease (n = 15/30, 50%) than upper airway disease (n = 4/30, 13.3%). Duration of hospitalization was significantly longer if one used BIPAP versus CPAP: children on BIPAP stayed median (range) 91 (1–629) days while those on CPAP stayed median (range) 47 (3–366) days, (z = −2.6, P = 0.009).

As explained earlier, due to the lack of health insurance cover for such children in our country, equipment for home use came from various sources (see Fig. 4). Fortunately, 11 families (44%, 11 out of 24 eligible families) donated their machines to the MSW department. Cost of initiating HV at discharge (including accessories like suction machine, oxygen tanks, and external battery), based on quotations from companies, was median (range) USD 7,000.0 (950.0–16,528.0): BIPAP cost median (range) USD 7,250.0 (5,875.0–16,527.0) versus IV which cost median (range) USD 9,500.0 (8,000.0–11,250.0) versus CPAP which cost median (range) USD 1,725.0 (1,475.0–2,420.0).

DISCUSSION

This 13-year review of 70 children on HV from a single center describes how young children with chronic...
respiratory failure can be sent home on HV, even in a developing country with limited financial resources and social support services.

Our patients were younger than those in studies from other parts of the world, with a median age of 11 months and more than two-thirds of the children were less than 2 years old. In other developed and developing countries, most of the children requiring HV were of school-going age.\textsuperscript{1,3,6,18–21} This difference is linked to the etiology of the respiratory failure, another noticeable difference between this study and others, as shown in Table 2. In the present study, more than 50% of children were

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**Fig. 3.** Bar chart illustrating the cumulative number of children on home ventilation (excluding those who have been weaned off and died) and type of ventilatory equipment used, especially the increasing use of continuous positive airway pressure (CPAP), from 2001 till 2014.

**Fig. 4.** Bar chart showing how patients obtained their home ventilation equipment. \textsuperscript{a}Medical Assistance Fund/Public Services Department/Lung Foundation of Malaysia/Medical Social Work/banks and private companies. \textsuperscript{b}Ventilator companies.
<table>
<thead>
<tr>
<th>Country</th>
<th>Number of patients</th>
<th>Type of study</th>
<th>Age of initiation</th>
<th>Main diagnoses</th>
<th>Duration of usage (median)</th>
<th>Came off (%)</th>
<th>IV/NIV (%)</th>
<th>Mortality (%)</th>
</tr>
</thead>
</table>
| Chile (2006)                  | 35                 | Retrospective, single-center 12-year review from 1993 | 12 months (range: 5 months–14 years) | Cardiorespiratory = 51.4%  
NMD = 34.2%  
CNS = 14.2% | 21 months | 40 | 26/9 | 17 |
| Turkey (2008)                 | 34                 | Retrospective, single-center 6-year review from 2001 | 1.8 years | Respiratory = 63.6%  
Central respiratory control = 21.2%  
NMD = 18.2% | 13 months | 8.8 | 11/23 | 32.4 |
| Thailand (2014)               | 148                | Retrospective, single-center 17-year review from 1995 | 4.6 ± 5.0 years for IV  
9.1 ± 4.4 years for NIV | Obstructive sleep apnea = 48%  
Respiratory = 17%  
NMD = 15% | NA | 13.5 | 53/95 | 13.5 |
| Serbia (2012)                 | 29                 | Retrospective, single-center 10-year review from 2001 | 9.3 years (range: 0.5–17.8 years) | NMD = 62.1%  
Respiratory/chest and spinal = 37.9% | 25 months (range: 3–119 months) | NA | 11/18 | 24 |
| Canada (2013)                 | 379                | Retrospective, single-center 22-year review from 1991 | 9.6 years (IQR: 2.9–13.9 years) | Musculoskeletal = 34.8%  
Respiratory = 32.7%  
CNS = 28.8% | 2.2 years (IQR: 0.8–4.9 years) | 8.7 | 66/313 | 15 |
| Italy (2011)                  | 362                | Cross-sectional questionnaire national in 2007 | 8 years (IQR: 4–14 years) | Musculoskeletal = 49%  
Respiratory = 18%  
Hypoxic ischemic encephalopathy = 13% | 1 year (IQR: 0–4 years) | NA | 149/213 | NA |
| United Kingdom (2015)         | 449                | Retrospective, single-center 18-year review, NIV only from 1993 | 10 years (IQR: 3–15 years) | NMD = 56%  
Respiratory = 20%  
Congenital syndrome = 14% | 74 months (range: <1–221 months) | 9 | NA | 24 |
| United States of America (2007) | 77               | Point prevalence state, Utah 1996–2004 | 6.5 months (range: 0.3–183 months) | Abnormal ventilatory control = 45%  
Respiratory = 41%  
NMD = 18% | 39 months (range: 1–216 months) | 39 | NA | 17 |

IV, invasive ventilation; NIV, noninvasive ventilation; NMD, neuromuscular disease; CNS, central nervous system disease.
ventilated for respiratory disease. In the study by Gowans et al., the age of initiation was 6.5 months, similar to ours, and the top two conditions that required HV were abnormal breathing control and respiratory disease, while neuromuscular disease contributed to only 18% of his cohort. In the study by Bertrand et al. in Chile, the main diagnostic criteria for HV was cardiorespiratory disease and the median age of initiation was also less than 2 years old. In studies where the age of initiation was older, children with a diagnosis of neuromuscular disease (NMD) or musculoskeletal disease (MSK) contributed to at least 30% of cases. Logically, other than SMA type I, respiratory compromise in children with NMD and MSK disease present later.

Being in an institution with a PICU that actively uses NIV, the number of children who survive despite severe lung disease is substantial. This could explain why respiratory disease represented 57% of our cohort, whereas NMD represented only 10%. Another possible reason is that in a developing country, lung disease from an infectious etiology is more prevalent due to overcrowding and poor hygiene, and therefore these are commoner than other chronic lung diseases.

Other important demographic characteristics to highlight are that in only 50% of families did either parent have a tertiary education, one-third of patients came from single-income families with three children from single-parent families, and the median income of these families was USD 1,693.0. Yet, they eventually went home on ventilation. Only one patient was able to afford private nursing care, whereas the others were looked after by their parents or other nonmedical personnel. This was similar to the findings of Preuthiphan et al. Therefore, contrary to common belief, in financially difficult circumstances, parents and caregivers can be trained to look after children on HV.

In this study, we excluded children who used CPAP for OSA as we wanted to include only children using CPAP as a form of ventilatory device. Currently, especially in small children, we find CPAP an economical and effective mode of HV as the latter not only helps with oxygenation by alveolar recruitment and relieves work of breathing, it also stabilizes the compliant chest wall in young children by limiting paradoxical chest movements and thus reduces the work of breathing. These are important aspects of respiratory failure that are better addressed with CPAP than oxygen. Other lauded benefits are that in Malaysia, its cost is equivalent to oxygen therapy alone and it is three times cheaper than BIPAP. It is especially suitable for infants who have high respiratory rates and small tidal volumes which may be insufficient to trigger portable ventilatory machines. With the advent of effective NIV machines with many different interfaces, invasive ventilation has become less desirable and is only used for those with very severe lung disease or spinal cord injury. However, there are side effects from the chronic use of NIV, especially mid-facial hypoplasia, although we did not objectively measure this in our study. This trend of reduced use of invasive ventilation is also seen all over the world.

With regard to the outcome of these children, nearly half were weaned off their ventilator, especially those with respiratory disease compared to neuromuscular disease. In a study by McDougall et al. who published a longitudinal 15-year review of 144 children also found that children with neuromuscular disease were less likely to come off their ventilator. The rate of children successfully being weaned off the ventilator, in this study, is similar to others (Table 2). The readmission rate was also low, demonstrating that once stable enough to go home, these children hardly get admitted for worsening respiratory failure. Finally, the mortality rate is also fairly low, comparable to the United States and Canada.

Finally, we are also fortunate that some parents donated their unused machines to the MSW department. A structured return policy facilitates early discharge by providing a pool of ventilators for loan to patients while awaiting procurement of their own machine.

There are some limitations of this study. The retrospective nature of this study has resulted in missing data especially with respect to social demographics and other finer details like duration of loan of machines, which we did not report on in this study. As this is a study from a single center, results may not be extrapolated to the rest of the country. We did not prospectively look at the quality of life of the patients and parents before and after initiating HV to prove the improved quality of life.

In conclusion, HV in children is possible even in a country with limited resources. In our institution, there has been a significant increase in the number of children requiring HV. Young children and children with respiratory disease made up a large proportion of our cohort requiring this service/HV. Many were able to come off their ventilators and the mortality rate was low and comparable to that in developed countries. We hope that this report will encourage developing countries to seriously consider setting up a HV program to allow...
these children to go home and experience a better quality of life.

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