AJRCCM: 100-YEAR ANNIVERSARY

Homeward Bound: A Centenary of Home Mechanical Ventilation

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Abstract

The evolution of home mechanical ventilation is an intertwined chronicle of negative and positive pressure modes and their role in managing ventilatory failure in neuromuscular diseases and other chronic disorders. The uptake of noninvasive positive pressure ventilation has resulted in widespread growth in home ventilation internationally and fewer patients being ventilated invasively. As with many applications of domiciliary medical technology, home ventilatory support has either led or run in parallel with acute hospital applications and has been influenced by medical and societal shifts in the approach to chronic care, the creation of community support teams, a preference of recipients to be treated at home, and economic imperatives. This review summarizes the trends and growing evidence base for ventilatory support outside the hospital.

Keywords: home ventilation; noninvasive ventilation; chronic ventilatory failure

The idea of assisting ventilation has evolved through four broad eras from the middle ages and renaissance to the present time: (*1*) an understanding of cardiopulmonary anatomy and observation that positive pressure ventilation applied to the trachea could inflate the lungs (1, 2); (*2*) the development of manually applied bellows, later automated, applied via a mask to resuscitate those with acute respiratory failure due to fires, mining, and drowning accidents; (*3*) an exploration of negative pressure ventilatory support reaching its zenith in mass use of iron lungs in the poliomyelitis epidemics of the 1950s; and (*4*) a shift back to positive pressure ventilation delivered noninvasively, coupled with a greater understanding of sleep-disordered breathing and respiratory muscle physiology, enhanced by technological advances in portable ventilator design and interface technology in the 1970s and 1980s.

Evolution

A century ago there was no acute ventilatory support available for the hundreds of thousands of victims of the 1918 Spanish flu outbreak, and indeed it is arguable whether ventilatory support would have been lifesaving in the presence of overwhelming infection and intractable hypoxemia due to acute lung injury. In 1907, Heinrich Dräger (3) had patented a portable time-cycled device powered by compressed oxygen, which delivered positive pressure during inspiration and negative pressure during expiration via a mask. These were manufactured in bulk and used to resuscitate victims of gas poisoning accidents or fires in the mining industry. This development had been preceded in the late 1800s by a creative burst of invention of negative pressure devices across Europe and the new world, typically consisting of a cabinet design encircling the individual's chest or lower body and powered by variety of mechanisms—sometimes manually by the patient (4). Negative pressure devices advanced significantly with the iron lung and motorized pump designed by Drinker and colleagues, who reported prolonged support of ventilation in *The Lancet* in 1931 (5). Iron lungs (or tank ventilators) reached their apogee with the devices developed by the manufacturer J. H. Emerson (Cambridge, MA) based on the design of Drinker and predecessors. Deployment of these tank ventilators in the polio epidemics of the 1950s in the United States and Europe undoubtedly saved lives, despite their bulk and expense (Figure 1). The limitations of negative pressure ventilation were exposed by the huge polio outbreak in Denmark in 1952. Not only were there insufficient iron lungs, but these failed to adequately ventilate those with bulbar polio; the death rate rose to 80%, only to fall markedly with the introduction of tracheotomy and invasive ventilation (6). It is notable that although tracheotomy

(Received in original form February 4, 2017; accepted in final form March 13, 2017)

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Am J Respir Crit Care Med Vol 195, Iss 9, pp 1140–1149, May 1, 2017

DOI: 10.1164/rccm.201702-0285CI
Internet address: www.atsjournals.org

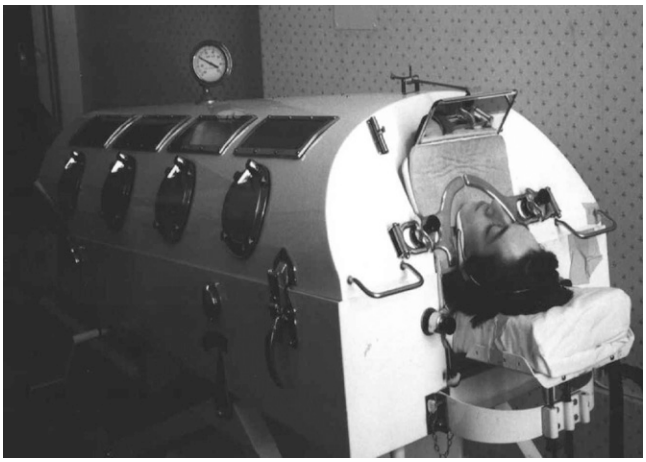

Figure 1. Iron lung.

ventilation proved the stimulus for the advance of invasive positive pressure ventilation and modern intensive care units (ICUs), some polio survivors were discharged home using more portable negative pressure devices, such as the cuirass and negative pressure jackets; these constituted some of the earliest domiciliary noninvasive ventilator (NIV) users (7).

Negative to Positive Pressure

Negative pressure devices, including iron lungs, saw a reawakening of interest in the 1970s and 1980s to treat a range of patients with chronic respiratory failure, including those with chest wall disease and chronic obstructive pulmonary disease (COPD), an application stimulated by an increased knowledge of respiratory muscle pathology (8, 9) and ventilatory changes during sleep. Sullivan and colleagues reported home continuous positive airway pressure (CPAP) therapy use for obstructive sleep apnea in 1981 (10). Although early cohort studies in patients with chronic respiratory failure due to neuromuscular and chest wall disease used volume ventilators to augment minute ventilation as opposed to delivering fixed positive pressure, cross-pollination from the development of continuous CPAP blowers and masks to treat obstructive sleep apnea led to the increasing use of smaller bilevel pressure support devices. This meant that by the early 1990s, NIV support, whether in the hospital or home, was delivered largely via positive pressure devices. Reports and case series (11–13) of patients on positive pressure noninvasive home ventilation began multiplying by the late 1980s and 1990s in patients with neuromuscular, chest wall, and obstructive lung disease, demonstrating improvements in life expectancy and quality of life in those with chest wall disease and nonprogressive (e.g., post poliomyelitis) or slowly progressive neuromuscular disorders. An illustrative timeline is given in Figure 2.

Prevalence

A first international perspective on the extent of home ventilation was seen in the Eurovent study published in 2005 (14). The majority of patients receiving home mechanical ventilation in northern European centers and university practices had neuromuscular disease, whereas in southern Europe a growing proportion of patients with lung and airway disease was reported. Prevalence rates ranged from 0.1 to 10 per 100,000 in Poland and Sweden, respectively, with only 13% receiving ventilation via tracheotomy. In Massachusetts, a census of prolonged mechanical ventilation reported by King (15) showed prevalence increasing from 2.8 per 100,000 to 7.1 per 100,000 from 1983 to 2006. But here practice differed from that in Europe, with significant numbers of long-term ventilated patients managed in hospitals or long-term acute care facilities. Some years later, in the United States, Sunwoo and colleagues (16) detailed the changing landscape of adult home ventilation; NIV was progressively replacing tracheotomy ventilation and there were widening indications, despite the fact that guidelines were lacking apart from in some neuromuscular conditions such as amyotrophic lateral sclerosis (ALS) (17) and Duchenne muscular dystrophy (DMD) (18).

Inherited Muscular Dystrophies, Myopathies, and Spinal Muscular Atrophy

Patients with DMD represent one of the first groups to receive home positive pressure ventilation, after its use in patients with polio, and are one of its greatest success stories. Early application by Rideau and

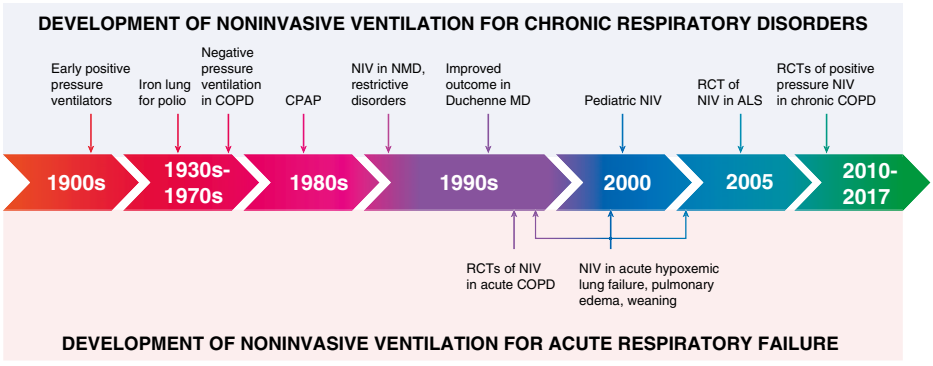

Figure 2. Timeline of development of home ventilation in the last century. ALS = amyotrophic lateral sclerosis; COPD = chronic obstructive pulmonary disease; CPAP = continuous positive airway pressure; MD = muscular dystrophy; NMD = neuromuscular disease; NIV = noninvasive ventilation; RCT = randomized controlled trial. Illustration by Jacqueline Schaffer.

colleagues (19) in France in the 1970s taken up by Bach and colleagues (20), Hill and colleagues (13), Sortor (21), and others in the United States, and Robert and colleagues (22), Leger and colleagues (11), and Carroll and Branthwaite (23) in Europe showed that mask NIV was feasible in a range of chronic neuromuscular and chest wall disorders and could also be used to wean invasively ventilated patients onto NIV to enable discharge from the ICU (24). Early case series showed marked improvement in survival (although no advantage to treatment before the development of symptoms [25]), with more recent results showing patients with DMD living into middle age. For NIV combined with cough-assist devices, there is also evidence of significant reduction in pulmonary morbidity and hospital admissions (26). Few randomized trials have been performed in these patient groups, but where these exist, evidence strongly supports use of ventilatory support (27). A notable feature of the dissemination of mechanical ventilation into the home has been the stark differences in practice, with tracheotomy ventilation being rare in the UK and United States and more common in Scandinavia, France, Japan, and the Gulf States, for example. Increasing numbers of children with neuromuscular and other disorders requiring NIV in childhood are now successfully treated and transitioning to adult care (28, 29).

Similar improvements in outcome have been seen in other relatively slowly progressive disorders, such as congenital muscular dystrophies, spinal muscular atrophy type II, and myopathies (29). Mouthpiece ventilation is an option during the day for some patients with neuromuscular disease (30). In type I spinal muscular atrophy with onset after 3 months, NIV may prolong survival or can be used to palliate symptoms (31, 32).

ALS

Ventilatory support in ALS is one of the few neuromuscular conditions subject to a randomized controlled evaluation. In a Cochrane review of mechanical ventilation for ALS in 2013 (33), the authors identified only two randomized trials of NIV, including 54 patients in total and complete data in only one trial (34). This showed an increase in average survival from 171 to 219 days; in those with mild to moderate bulbar weakness the survival increase was 205 days, and quality of life was maintained. In those patients with severe bulbar weakness, survival was not increased but symptoms improved.

Cazzolli and Oppenheimer (35) report that in patients electively started on NIV, 100% of users felt glad they chose NIV, but in 50 tracheotomy ventilated patients, many had undergone tracheotomy as a result of emergency care and fewer were satisfied with their quality of life. Failure to be involved in decision-making about institution of tracheotomy ventilation has been found to have an adverse impact on quality of life in other studies. There are still considerable national differences in care provided for patients with ALS. The American Academy of Neurology Practice Parameters (17) on drug, nutritional, and respiratory therapies in ALS recommends NIV to lengthen survival and slow decline of FVC (level of evidence B) and to improve quality of life (level of evidence C). National Institute for Clinical Evidence guidelines (36, 37) in the UK recommend referral for assessment for NIV if daytime arterial oxygen saturation is 93% or less, FVC is less than 70% predicted, maximum inspiratory pressure is less negative than -60 cm H_2O, or marked symptoms of sleep-disordered breathing or orthopnea are present. The initiation of NIV is recommended in the presence of daytime hypercapnia, symptomatic sleep-disordered breathing, and deteriorating pulmonary function. Serial measurement of respiratory muscle strength is an accurate predictor of survival (38). It is not clear whether earlier initiation of NIV offers any survival advantage.

The addition of cough-assistance techniques is particularly advantageous in ALS and may prolong duration of effective noninvasive approaches when combined with percutaneous feeding. Effective respiratory therapy to clear secretions and breath stacking with a lung volume recruitment bag should first be explored, but in those with cough peak flow less than 270 L/min, clearance of secretions is likely to be impaired during a chest infection, and cough peak flow less than 160 L/min suggests severe reduction in cough adequacy. Here, use of cough insufflation–exsufflation devices can significantly improve cough peak flow. Current scientific evidence does not support the widespread use of mechanical insufflation–exsufflation in neuromuscular disorders due to a lack of randomized controlled trials (RCTs) (39), but these devices are used clinically on a pragmatic basis and recommended as good practice by guidelines (18, 36). As with initiating NIV, setting up insufflation–exsufflation pressure may be more challenging in patients with bulbar ALS. Andersen and colleagues (40) recently compared upper airway responses in patients with ALS with no bulbar disease to those with spastic pseudobulbar and hypotonic bulbar features. The response of the larynx, aryepiglottic folds, and hypopharynx may vary considerably in these groups, suggesting careful titration of settings starting with low inspiratory positive airway pressure values in the bulbar group. In this study, optimization of settings was achieved using direct transnasal laryngoscopy, but knowledge of these variables may be helpful in the absence of such interventions, especially as Farrero and colleagues (41) have shown that with careful titration, NIV use in patients with bulbar ALS may be successful in more than 50% of cases. In those in whom NIV cannot be initiated, palliative care approaches are key.

NIV in COPD

In COPD, respiratory failure was recognized almost as soon as it became possible to measure carbon dioxide (42, 43), a facility that came after the recognition of the principle of both positive and negative pressure ventilation (44). One of the first recorded uses of mechanical ventilation in COPD was the acute application of a negative pressure device reported in 1951 (45), and it is unsurprising, therefore, that the first use of chronic home ventilation in COPD also used a negative pressure approach (46).

After pioneering work by Braun (47), Rochester (9), Gutiérrez and colleagues (48), Cropp and DiMarco (49), Zibrak and colleagues (50), and others on assessing the impact of negative pressure ventilation on respiratory muscle function, Shapiro and coworkers (46) randomized 184 patients with COPD to receive active or sham negative pressure ventilation. The study

concept would now be considered outdated, because the stated aim was to rest the respiratory muscles to relieve fatigue, which was believed to be present, and thus improve their function. In fact, subsequent studies showed that low-frequency fatigue of the respiratory muscles is in fact more difficult to elicit in patients with COPD than in normal subjects (51–53), as might be expected given both the presence of diaphragm shortening (54) and the switch to fatigue-resistant type I fibers, which was also subsequently demonstrated (55, 56). Given this subsequent knowledge, it is perhaps unsurprising that no differences were found between treatment and sham groups, especially given other issues, which were high levels of inadequate adherence or nonadherence coupled with the fact that the mean Pa_{CO_2} at entry was only 44 mm Hg (6.4 kPa).

Large trials powered for mortality or combined morbidity and mortality outcomes in which positive pressure have been used are summarized in Table 1. The most notable trends in the evolution of NIV seem to have been the following: first, more recent studies have used higher levels of inspiratory pressure support, which intuitively might permit more efficient CO_2 clearance and better oxygenation; thus, simply put, earlier studies may have failed by delivering suboptimal ventilation. This trend may continue in the future with the use of ventilators with more sophisticated modes designed to deliver a preset volume within physician-prescribed pressure limits; such devices have been evaluated in acute COPD (57), obesity (58), and mixed patient populations (59), with inconsistent results thus far. A second reason positive studies have more recently emerged likely reflects the use of strategies designed specifically to target hypercapnia rather than more general ambition, such as respiratory muscle rest or relief of dyspnea; this philosophy blends with the previous point and reflects diminishing concern regarding potentially harmful effects of high airway pressure which, although demonstrated after invasive mechanical ventilation, have never been shown to be an issue in NIV. Last, investigators have become more sophisticated in recognizing that the point of greatest need and therefore likely most easily demonstrable benefit is after acute exacerbation of COPD. In addition to accounting for more than 50% of the lifetime costs of acute care, in-patient admission is both distressing and dangerous for patients, with a typical 7% inpatient mortality and 30% 3-month readmission rate.

A range of factors may explain the divergence in results from Köhnlein and colleagues (60) showing improved survival in patients with hypercapnic COPD treated with home NIV, and those by Struik and colleagues (61), in which no difference was found in outcome in patients randomized to home NIV after an exacerbation. There were differences in COPD phenotype and selection: the Köhnlein and colleagues (60) patients had chronic stable hypercapnia and there was high mortality in the control group, whereas the postexacerbation patients in the study by Struik and colleagues (61) may still have had potential for recovery, therefore making initiation of NIV on discharge after a hypercapnic exacerbation seem premature. The study by Murphy and colleagues (62) includes patients who remained hypercapnic 2 weeks after an exacerbation rather than on discharge. Hence, it seems that patients most likely to benefit from NIV are those who require NIV for an inpatient hypercapnic COPD exacerbation and who remain hypercapnic while convalescent (62). Going forward, it is well established that physical frailty is a good marker of the likelihood of readmission (63, 64), and it may be that patient selection for future trials could also profit by including a measure of physical function and/or combining NIV with a rehabilitation program (65).

NIV in Bronchiectasis

Adoption of ventilatory support was extended to patients with bronchiectasis from experience in patients with other causes of respiratory failure, notably COPD. There are many pathophysiological similarities between COPD and bronchiectasis, and NIV offers the potential benefit of augmenting airway clearance. However, early observational studies in mixed cohorts of patients with chronic respiratory failure identified patients with bronchiectasis doing less well compared with other groups discussed below.

In 1994, Leger and colleagues (11) described 25 patients with bronchiectasis, where the 3-year dropout was 48%; they identified no change in hospital stay in the year after NIV introduction. In 1995, of a further cohort of 13 patients with bronchiectasis (12), only 20% continued NIV at 2 years, which was markedly reduced compared with patients with neuromuscular disorders (Figure 3). In 1997, Benhamou and colleagues (66) reported a case–control study of 14 patients with bronchiectasis using NIV and long-term oxygen therapy or control subjects using long-term oxygen therapy only, described benefits of reduction in days in the hospital with NIV but no change in Pa_{O_2} or mortality. Interestingly, in this study, volume ventilation was used and generally well tolerated. In 2004, Dupont and colleagues (67) described 48 patients with bronchiectasis after their first ICU admission with respiratory failure. Fifty-four percent required intubation; 27% used NIV. The 1-year mortality in this group of patients was 40%. Poor prognostic factors included age and intubation. NIV did not worsen survival, but only 10 patients continued NIV in the longer term. More recently, a 2009 series of 35 patients from Saudi Arabia with more severe bronchiectasis presenting to the ICU with respiratory failure were reported (68). Thirty-one percent required intubation, and 57% were managed with NIV. The ICU mortality was 34%, and 4-year mortality was 60%. This study highlighted premorbid activity in addition to age, premorbid condition, and ICU interventions including intubation as major determinants of mortality (68).

Unlike data in stable hypercapnic COPD, the literature on NIV in bronchiectasis consists only of small, observational studies with no RCTs. Moving forward, there are opportunities for RCTs early in the course of disease, perhaps targeting development of overnight hypercapnia to define when respiratory support may be most useful.

NIV in Cystic Fibrosis

Historically, ICU admission has been avoided in patients with cystic fibrosis (CF) and respiratory failure because of poor

Table 1. Comparison of Randomized Controlled Trials of Home Positive Pressure Ventilation in Chronic Obstructive Pulmonary Disease

Study	N	Design	Main Inclusion/Exclusion Criteria	Mean Pa_{CO_2} (*mm Hg*)	Mean Pressures Used (*cm H_2O*)	Notes	Outcome
Strumpf *et al.*, 1991 (93)	19	Randomized crossover over 3 mo	No AECOPD previous 4 wk $FEV_1 < 1$ L No OSA	46	IPAP, 15	Only 7 of 23 enrolled completed both arms of the study	No change CO_2 or other outcome measure
Meecham Jones *et al.*, 1995 (94)	18	Randomized crossover 3-mo trial	No AECOPD previous 4 wk $Pa_{CO_2} > 45$ mm Hg, LTOT No OSA	56	IPAP, 17 EPAP, 1.9	14 completers Median 6.9 h/d use	Improved ABG, no change on 6MWT
Gay *et al.*, 1996 (95)	13	NIV vs. control	No AECOPD $Pa_{CO_2} > 45$ mm Hg,	51.6	IPAP, 12 EPAP, 2	13 randomized from 35 recruited, 3 of 7 in NIV group did not complete	NIV of no benefit
Lin, 1996 (96)	12	Randomized crossover	No AECOPD previous 4 wk Patients had to be able to tolerate 2 wk NIV before entry $Pa_{CO_2} > 43$ mm Hg No OSA	50.5	IPAP, 14 EPAP, 2	2 wk of therapy	No benefit
Clini *et al.*, 2002 (97)	90	Prospective RCT. Inclusion criteria to be met 4 wk after enrollment	No AECOPD previous 4 wk $Pa_{CO_2} > 50$ mm Hg LTOT > 6 mo MRC > 1 $FEV_1 < 1.5$ L	54 in NIV group, 55.5 in LTOT group	IPAP, 14 EPAP, 2	2-yr follow-up	Improved dyspnea and HRQL but no effect on survival or admission
McEvoy *et al.*, 2009 (91)	144	Prospective RCT	No AECOPD previous 4 wk $Pa_{CO_2} > 46$ mm Hg LTOT > 3 mo $FEV_1 < 1.5$ L/50% predicted	54.4 in LTOT group 52.6 in LTOT and NIV	IPAP, 12.9 EPAP, 5.1	Powered for a 2-yr study	Improved unadjusted but not adjusted survival Worse general health and vigor
Köhnlein *et al.*, 2014 (60)	195	Prospective RCT	No AECOPD previous 4 wk $Pa_{CO_2} > 52.5$ mm Hg GOLD IV COPD (i.e., $FEV_1 < 30\%$ predicted)	58.5 in NIV group and 57.7 in control group	IPAP, 21.6 EPAP, 4.8	1-yr survival	1-yr survival advantage for NIV NIV titrated to reduce CO_2 LTOT usage ~63% of participants Rate of emergency admissions unexpectedly low in both groups
Struik *et al.*, 2014 (61)	201	Prospective RCT	GOLD III/IV Prior admission requiring NIV Convalescent $Pa_{CO_2} >$ 45 mm Hg	59.2 in NIV group and 57.7 in control group	IPAP, 19.2 EPAP, 4.8	1-yr survival	Patients had had prior ventilatory support No benefit NIV 65% readmission rate
Murphy *et al.*, 2016 (62)	116	Prospective RCT	$CO_2 > 52.5$ mm Hg within 2–4 wk of exacerbation OSA excluded	59.2	IPAP, 24 EPAP, 4	1-yr admission-free survival	NIV titrated to target hypercapnia Mortality unchanged but reduced readmission rates at 1 and 12 mo

Definition of abbreviations: 6MWT = 6-minute-walk test; ABG = arterial blood gas; AECOPD = acute exacerbation of chronic obstructive pulmonary disease; COPD = chronic obstructive pulmonary disease; EPAP = expiratory positive airway pressure; GOLD = Global Initiative for Chronic Obstructive Lung Disease; HRQL = health-related quality of life; IPAP = inspiratory positive airway pressure; LTOT = long-term oxygen therapy; MRC = Medical Research Council (UK); NIV = noninvasive ventilation; OSA = obstructive sleep apnea; RCT = randomized controlled trial.

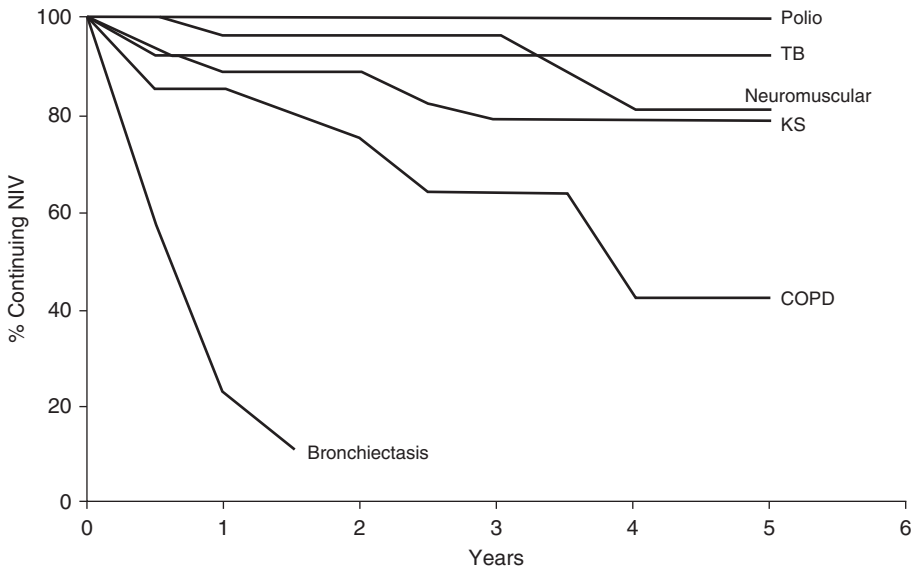

Figure 3. Probability of continuing noninvasive ventilation in neuromuscular disease, chest wall disorders, chronic obstructive pulmonary disease (COPD), and bronchiectasis (12). KS = kyphoscoliosis; NIV = noninvasive ventilation; TB = posttuberculous lung disease.

outcomes (69); the development of effective lung transplantation has challenged this view. There are now data demonstrating that when there is sudden decline with a potentially reversible cause, such as a pneumothorax or major hemoptysis, survival after ICU admission can be excellent (70–72). Invasive ventilation for worsening respiratory failure caused by respiratory infection where there is no possibility of transplantation continues to have a poor prognosis, with survival of just 10 to 30% (69).

In CF, unlike bronchiectasis, there have been a number of small studies demonstrating that NIV improves airway clearance, a facet of ventilatory support that can be very useful. In 2013, a metaanalysis identified 15 studies, only 7 of which met inclusion criteria, for a total of 106 patients (73). Of these seven trials, six were single-intervention studies and one examined nocturnal NIV over a 6-week period (eight patients). All were crossover studies. There were four trials (79 patients) using NIV against another specified method of chest physiotherapy. The authors concluded sessions of airway clearance using NIV may be easier, were preferred by the patient, and may improve some parameters of lung function. There was no change in sputum expectoration. The authors also reviewed three trials (27 patients) that examined overnight NIV and found, when used with oxygen, NIV may improve overnight gas exchange and exercise tolerance (74). Longer RCTs looking at effects of NIV on airway clearance and exercise are required.

NIV has advantages over invasive ventilation, as it allows cough and clearance of secretions and can avoid ICU admission. A retrospective analysis of 113 patients with end-stage CF, using volume preset ventilation, has been reported (75). The authors divided patients into three groups: A, awaiting lung transplantation; B, patients not yet on the transplant list; and C, patients who were not eligible for transplantation. The mean (range) duration of NIV support for groups A, B, and C was 61 (1–600) days, 53 (1–279) days, and 45 (0–379) days, respectively. In this study of patients with end-stage disease, they reported NIV with additional entrained oxygen improved hypoxia but not hypercapnia, and survival of patients who had received transplants in group A was comparable to patients with milder disease not on NIV (75). The widespread use of NIV in end-stage CF both as a bridge to transplantation and in palliation, where NIV can be delivered at home, allows a degree of autonomy for patients to make informed choices about where and how supportive care is delivered.

NIV in Obesity Hypoventilation

The association between sleepiness and obesity has been recognized in the literature since at least the early 19th century. Famously, the English satirist and polymath Charles Dickens created the character Joe, who appears in *The Pickwick Papers*. "A wonderful fat boy—habited as a serving boy standing upright on the mat, eyes closed as if asleep" (Charles Dickens, 1837). This excellent and very funny book was widely read at the time. Osler was clearly a fan, and referenced Dickens' astute observations in his 1903 medical textbook (76). Medical journals, however, lagged somewhat behind. The syndrome of alveolar hypoventilation was described in the 1950s by Auchincloss and colleagues (77) and Burwell and colleagues (78). Since the first accounts of the effectiveness of respiratory support, there has been an almost exponential increase in provision of various forms of home respiratory support. Changing population demographics with an epidemic of emerging obesity, particularly in the developing world (79), are likely to change future delivery of home ventilation, and the proportion of patients with obesity hypoventilation syndrome (OHS) using NIV is likely to increase. In many societies, middle-aged obesity is now the new norm, and the emergence of the super-obese poses significant and unique problems for healthcare delivery.

The use of NIV in this population has increased dramatically over the past 15 years, although the evidence base for the shift in respiratory support from CPAP is somewhat lacking. There is good evidence that PAP improves outcomes in OHS, but randomized controlled trials comparing CPAP and NIV have been either too short or too small to demonstrate significant differences in mortality to date (80, 81). Until those data become available, an individualized, patient-centered approach in provision of respiratory support, with a switch from CPAP to NIV in those patients who require high CPAP pressure, are in respiratory failure, or have residual OHS despite CPAP, seems sensible (82–84).

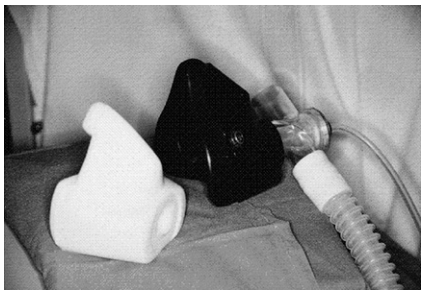

Figure 4. Early noninvasive ventilation masks used by Brochard and colleagues (98).

PAP is only one aspect of supporting the obese patient; optimal lifestyle advice must be delivered in the context of an integrated bariatric service with surgical interventions available.

Palliative NIV

In many situations, home ventilation is instituted with the aim of improving life expectancy as well as quality of life. Application with the sole intention to reduce dyspnea or symptoms of sleep-disordered breathing and enable optimal use of opiates is just as valid (85). Home NIV can therefore be used as a palliative tool in patients with ALS/motor neuron disease and has also been shown to reduce dyspnea and opiate requirements in patients with end-stage cancer and respiratory failure (86)

Interfaces

In addition to progress with ventilators, an important step toward making NIV easier to use has been the development of the interface that connects the patient to the ventilator. In one of the first studies undertaken in patients with a COPD exacerbation by Brochard and colleagues (87), a face mask had to be developed for use by the investigators (Figure 4); in contrast, a range of masks is now available (some, but not an exclusive selection, shown in Figure 5). Most typically, a patient will opt for one covering either the nose alone or the nose and mouth, but bigger devices are available that can be worn like a helmet (used for acute NIV), as well as smaller ones that use nasal cushions to insert into the nose or function purely as an oral interface retained by teeth and gums. Smaller interfaces suitable for pediatric use have been a welcome advance, although the problem of young patients developing midfacial hypoplasia from long-term use of facial masks has not been completely solved. Materials have also progressed so that normally a silicone cushion is used, which is less irritating to the skin. The interested reader is referred to Pisani and coworkers (88) for a more detailed consideration of masks currently available and potential complications and contraindications.

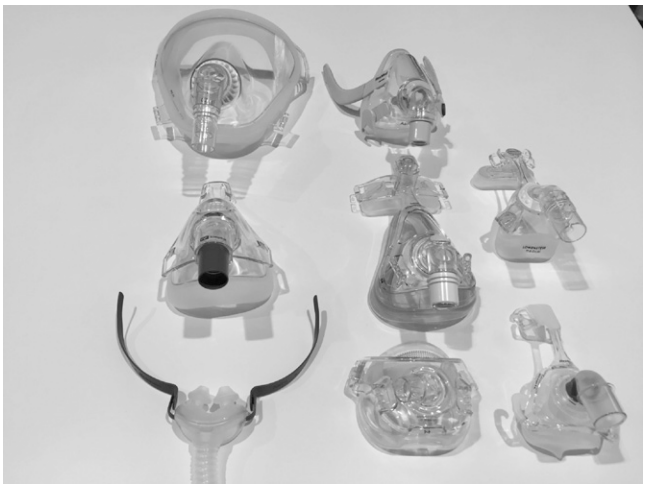

Figure 5. Range of recent noninvasive ventilation interfaces. *Top row, left to right*: Fitlife pediatric total mask (Philips Respironics, Monroeville, PA) and Airfit F20 (ResMed, Bella Vista, Australia). *Middle row, left to right*: Simplus FF (Fisher and Paykel Healthcare, Costa Mesa, CA), Amara (Respironics), and JoyceOne (Lowenstein Medical, Hamburg, Germany). *Bottom row, left to right*: Airfit P10 Nasal plugs (ResMed), Amara View (Respironics), and Mirage FX (Resmed).

Global Home Ventilation in 2017 and Beyond

International comparisons of the organization of home ventilation are difficult, as they are inevitably linked to reimbursement for healthcare and overall financial provision. Undoubtedly, however, the ability to provide ventilatory support in the home has reduced hospital and ICU stays, the move from invasive ventilation via tracheotomy to a combination of noninvasive techniques and cough-assist devices has simplified care, and quality of life for many patients with chronic respiratory failure has improved in the last century. As described in the sections above, manufacturers have assisted with the development of more portable, responsive ventilators that attempt to minimize asynchrony, have a longer battery life, and have more ergonomic interfaces. Three-dimensional printing is likely to advance the ability to personalize fit and individualize interfaces.

Downloadable data from ventilators has been available for some time, which can facilitate expert care in the home. Telemonitoring is posited as crucial to the future of home ventilation. Routine telemonitoring of CPAP now occurs in some countries, with daily transmission of data on leak, respiratory events, and adherence. Trials of early-generation telemonitoring systems in patients receiving ventilatory support have produced mixed results (89, 90). However, the utility of remote monitoring is likely to improve if telemonitoring options can focus on specific problem solving—adapting the new user to the device, optimizing settings in the home, and reducing hospital attendances for patients in remote environments or for whom mobility/travel are problematical.

Although RCTs have not been possible in some areas of rare neuromuscular disease, effective RCTs establishing the appropriate place for long-term use of NIV for COPD (60–62, 91) and the Serve-HF trial (92) showing adaptive servoventilation does not produce beneficial effects in patients with chronic systolic heart failure with central sleep apnea have been influential landmarks in the field, while at the same time increasing our understanding of the underlying pathophysiology. Research

priorities now include ascertaining the role of long-term NIV in different COPD phenotypes and its impact on quality of life, unraveling the effects of NIV on respiratory and cardiovascular consequences of OHS, understanding long-term outcomes of NIV in pediatric ventilatory disorders, determining the role of palliative NIV, and optimal use of cough assist devices, among others.

The use of physiological concepts to develop new modes, such as proportional assist ventilation (92) and volume assured pressure support ventilation, has in many ways been a stimulus to better understanding of ventilator–patient interaction and pathophysiology rather than changing outcomes. As efforts continue to produce more intelligent ventilators and assistive technology in the home, we should bear in mind that intelligent operators are required too and that training of physicians, allied health team members, and carers to select patients, optimize ventilatory control, and investigate further indications for NIV is just as crucial. ■

Author disclosures are available with the text of this article at www.atsjournals.org.

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