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# The evolution and future of respiratory care for Spinal Muscular Atrophy

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**Abstract**

Spinal Muscular Atrophy (SMA) is a hereditary neuromuscular disorder characterized by progressive weakness over time. The most common cause of death in patients with SMA is respiratory failure due to weakness of the respiratory muscles. In the past, patients with the most severe forms of SMA did not typically survive more than 2 years. However, technology developed for the short-term ventilation of patients with acute respiratory failure due to poliomyelitis, beginning in the 1920s, ultimately led to advancements in long-term ventilation in patients with SMA. In addition, advancements in artificial airways and airway clearance, also developed for short-term respiratory care for patients with polio, contributed to significant improvement in life expectancy for patients with SMA, and opened the door to advancements in other areas, such as orthopedic and nutritional care.

Now that disease modifying therapies are available, the spectrum of respiratory disease in patients with SMA continues to change. However, in moving forward, it is vital to understand the natural history of SMA and the history of the respiratory care it has required, not only to provide the best possible nuanced care for current patients with SMA, but also to learn from the advances made in SMA care and apply them to other respiratory disease processes, just as the care of patients with polio created so many advancements in the care of patients with SMA.

**Keywords:** SMA, Mechanical Ventilation, Polio epidemic

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## Introduction

Spinal Muscular Atrophy (SMA) is a group of hereditary proximal symmetrical muscular atrophies associated with degeneration of the anterior horn cells of the spinal cord, with progressive weakness over time. SMA was first described in 1891 by Guido Werdnig, and additional cases were reported a few years later by Johan Hoffman. The most severe form of SMA was often referred to as Werdnig-Hoffman Disease. Over the following decades, various classification systems were used based on severity of weakness and age of onset; the most recent and widely used classification included the highest attained motor function with type 0 (severe weakness and respiratory insufficiency at birth), type I (weakness noted in the first few months of life, never able to sit), type II (able to sit at some point during infancy), type III (able to stand), and type IV (least severe, with adult onset). [1] In this classification, types I to III were the most common phenotypes seen.

The most common cause of death in patients with SMA is respiratory failure due to weakness of the respiratory muscles, often precipitated by an acute respiratory infection, including common viral upper respiratory illnesses. Since the classification system has fluctuated over the decades since the disease was first identified, it is not possible to provide exact figures for average survival for each subcategory of SMA. However, one study published in 1961, before significant respiratory interventions were possible, described patients according to age at symptom onset. In the first group were a majority of patients who would correspond with the current category of Type I. Of the 23 patients who would currently be classified as Type I, all 23 patients died within the study timeframe of 11 years, with the average age of death 10 months. The longest survival for a patient in this group was 52 months. In the second and third groups, which would be consistent with the current diagnosis of Type II, five out of 27 patients died within the 11 year study, with the average age of death 25 months, and a range of 7-73 months. [2,3] In the past, before current respiratory interventions were available, a diagnosis of SMA Type I meant that the patient would die in early childhood, typically due to respiratory failure secondary to a viral respiratory infection that would not be fatal, or even serious, in other children.

## The development of mechanical ventilation and use with poliomyelitis patients

Patients with SMA would ultimately benefit from technological advancement in respiratory care, but none of these developments were made expressly for the purpose of SMA care. The first important development that would ultimately benefit SMA patients was the invention of the tank ventilator, or iron lung, first commercially produced in 1928 by Philip Drinker and Dr. Louis Agassiz Shaw in Boston. In October 1928, the iron lung was first used with a patient, a child with acute respiratory failure secondary to polio. The tank ventilator used negative pressure to ventilate patients in acute respiratory failure, and its use expanded greatly over the following years, prior to the development of the polio vaccine. The tank ventilator was most often used for acute respiratory failure rather than chronic respiratory failure, though some patients who did not recover adequate respiratory muscle strength did go on to use the tank ventilator for decades. However, there are no documented cases of this type of ventilator being used for patients with SMA. [4-6]

Though the tank ventilator did not directly benefit patients with SMA, its importance to the history of SMA respiratory care lies in the fact that this negative pressure ventilator led to the development of the first portable positive pressure ventilators. The tank respirator was expensive and heavy, and it was sometimes difficult or impossible to obtain a sufficient number of the devices to provide care during a localized epidemic. In addition, while the survival rate for acute respiratory failure with polio had certainly increased with the use of the tank respirator, it was still fairly low. During a polio epidemic in Los Angeles in 1948, Dr. Albert Bower and engineer V. Ray Bennett developed an attachment to the tank respirator to provide intermittent positive pressure ventilation in addition to the negative pressure ventilation provided by the tank respirator. They used this device at Los Angeles County Hospital and published a study in 1949 showing that this positive pressure attachment had increased the survival rate for acute respiratory failure secondary to polio from 21.1% in 1946 to 83.7% in 1949 in the same institution. This study therefore demonstrated the first large scale success of supplemental positive pressure ventilation over negative pressure ventilation alone. [7]

Three years after the above study was published, another polio epidemic occurred in Denmark, with a sudden influx of dozens of patients in acute respiratory failure. The primary hospital, Blegdam Hospital, only had one tank respirator at the onset of the epidemic. One of the staff anesthesiologists, Bjorn Ibsen, had trained in the United States and had some experience in using intermittent positive pressure ventilation, though only briefly for surgical patients. He proposed placing tracheostomies and trying positive pressure ventilation alone to try to save the dozens of polio patients presenting in acute respiratory failure, and the hospital agreed with his plan. At that time, positive pressure ventilation, specifically manual bagging via a tracheostomy

tube had been described in anesthesiology literature but was not a well-known or common practice. Since the normal function of the lung is ventilation via negative pressure, the prevailing medical thought at the time was that positive pressure ventilation alone could not be sufficient or safe to be used for more than a brief period. The Copenhagen hospital did not have access to the positive pressure equipment developed by V. Ray Bennett during the prior Los Angeles epidemic, and staff had to resort to manual bagging via tracheostomy to provide positive pressure ventilation. Hundreds of medical students were employed around the clock in this endeavor, with an impressive improvement in patient survival. In a study published the following year, it was noted that the mortality rate at the beginning of the epidemic at Blegdam Hospital for patients in acute respiratory failure was 87% before bag ventilation was introduced, and this dropped markedly to 22% by the end of the epidemic. While the success of positive pressure ventilation as a supplement to negative pressure ventilation in Los Angeles in 1949 did not receive much attention in the medical community, the success of positive pressure ventilation alone in Copenhagen in 1952 was well publicized and was the beginning of the era of innovations in positive pressure ventilation. [8,9]

**The development of portable positive pressure ventilators and use with chronic respiratory failure patients**

After the success of positive pressure ventilation in 1952, portable positive pressure ventilators began to appear in Europe, most commonly used by anesthesiologists for controlled ventilation during surgery. In the United States, Dr. Forrest Bird, a former World War II pilot with a medical degree, developed one of the first commercially produced mechanical ventilators to provide positive pressure. The Bird Mark 7 Respirator was first commercially produced in 1957, and the "Babybird" neonatal ventilator was first produced in 1969 and resulted in substantially decreased mortality rates for neonates. Thus began the age of portable positive pressure mechanical ventilation, which would soon change the lives of many patients with SMA. [10]

When portable positive pressure mechanical ventilators became widely available in the 1950s, they were typically used short-term for acute respiratory failure, including respiratory failure due to infections (polio and pneumonia) and trauma. The other common use for the ventilators was for controlled ventilation during surgery, with use primarily by anesthesiologists. However, as the medical world became more familiar with modern ventilators, new uses were found, including long-term ventilation for patients with various forms of hypotonia and chronic respiratory failure. Advances in ventilation were accompanied by advances in tracheostomy tubes, and the old metal tubes that often caused tracheal complications gave way to newer polyvinyl chloride (PVC) and silicone tubes in the 1970s that were safer, more comfortable, and more suited to long-term use. [11] At this point, chronic ventilation via tracheostomy became possible, and some of these patients were polio patients who never recovered enough respiratory muscle strength to breathe unassisted. However, as PVC and silicone were used to innovate tracheostomy tube design, they were also used to create new noninvasive interfaces for chronic ventilation. In 1993, Dr. John R. Bach published a groundbreaking study describing the use of noninvasive mechanical ventilation for patients with chronic respiratory failure due to hypotonia, including, for the first time, patients with SMA. Dr. Bach noted that in one medical center, "mouth IPPV [intermittent positive pressure ventilation] has been used as a principal noninvasive method of daytime ventilatory support since 1957 and for nocturnal support since 1964". [12-14] This study also describes the use of various non-invasive interfaces for chronic mechanical ventilation, including oral, nasal, and oro-nasal interfaces. The majority of patients had diagnoses of polio, myopathy, muscular dystrophy, and traumatic high-level quadriplegia, but there were also a few patients with SMA included in the study. While the type of SMA was not listed, the earliest age at which ventilatory support was begun for the SMA patients described was 7 years, which would suggest a Type II SMA diagnosis. [15] Dr. Bach published another study in 1993 which compared patient satisfaction with ventilation via tracheostomy vs noninvasive ventilation; this study again included patients with SMA, but none with SMA Type I. The conclusion was that the adult patients and their caregivers in the study considered noninvasive ventilation more convenient, with fewer negative effects on speech, appearance and comfort, and preferred overall compared to ventilation via tracheostomy. [16] Seven years after these two studies describing the use of long-term invasive and noninvasive ventilation in patients with non-type I SMA, Dr. Bach published another groundbreaking study describing the use of noninvasive mechanical ventilation in patients with SMA Type I for the first time. This study followed nine children with SMA Type I, and the conclusion was that "tracheostomy can be avoided throughout early childhood for some children with SMA Type I". [17] At this point, respiratory care for SMA patients of all types had reached a new era of options and choices, and advancements arrived in rapid succession.

**The development of mechanical airway clearance devices**

Along with the above innovations in ventilators, tracheostomy tubes, and noninvasive interfaces, advances in airway clearance techniques kept pace and allowed for more effective utilization of long-term ventilation for patients with SMA. Manual chest physiotherapy has likely existed for eons, as it is intuitive and simple, but it has continued to be widely used and perfected as part of airway clearance for a variety of patients, including patients with hypotonia. Mechanical insufflator-exsufflators, also commonly known as exsufflators or cough assist devices, were first developed in 1953, and were used initially via facemask to simulate a cough and bring up lower respiratory secretions for patients with respiratory muscle weakness. Again, this device was developed in response to the polio epidemic and the inability of polio patients to clear their lower respiratory secretions, which contributed to acute respiratory failure. With the increasing use of tracheostomy ventilation in the 1960s, the exsufflator was less commonly used. [18] The first description of the use of the exsufflator in SMA patients was published by Dr. John Bach in 1993, in his study on long-term noninvasive ventilation for hypotonic patients. He noted that the exsufflator was no longer being manufactured as of the early 1960s, and the only machines available for patients in the study were old models belonging to individual patients or to patient care networks that loaned them out to patients who needed them. Dr. Bach noted that newer models had only recently become available and published a clinical study on mechanical insufflation-exsufflation later that year using both the older Cofflater and the new Emerson exsufflator. [15] The study concluded that "the use of these noninvasive inspiratory and expiratory muscle aids can decrease the risk of pulmonary complications, intubation, and resort to tracheostomy and reduce the need for bronchoscopy in this population". [18] This 1993 study popularized the exsufflator as an airway clearance tool for patients with hypotonia, including patients with SMA of all ages and types.

Two additional advancements in airway clearance ultimately affecting SMA care were the development of IPV (intrapulmonary percussive ventilation), and of the high frequency chest wall oscillation system (Vest). The IPV was first described in 1985 as a device for delivering aerosolized medications to patients with chronic obstructive pulmonary disease, and was invented by Dr. Forrest Bird, the same physician who invented the first widely available portable ventilator. The IPV helps to mobilize lower respiratory secretions, and can be used via facemask, endotracheal tube, or tracheostomy tube. The high frequency chest wall oscillation system (Vest) was first reported in the literature in 1991, initially used only in the cystic fibrosis population. [19] These two devices have been incorporated into SMA care in the following years, though not on a uniform basis.

**Improving survival rates for SMA patients spark debate on goals of care**

With all the necessary equipment for the respiratory care of SMA patients available in the 1990s, the short-term and long-term respiratory care of all SMA patients began to change, sparking discussions regarding goals of care and the standard of care for SMA patients, particularly patients in the type I category. In 2000, Dr. John Bach published a retrospective cohort study with the objective "to determine whether spinal muscular atrophy (SMA) type 1 can be managed without tracheostomy and to compare extubation outcomes using a respiratory muscle aid protocol vs conventional management." The study employed a novel airway clearance protocol that used the exsufflator as well as manual chest physiotherapy and postural drainage. The study concluded that "although intubation may be required during intercurrent chest colds, tracheostomy can usually be avoided if respiratory muscle aids are used by highly trained and dedicated parents in both the acute and home settings, as needed". [17]

With the advent of long-term noninvasive ventilation as an option for patients with SMA, particularly SMA Type I, an ethical debate then began in the medical community regarding the options of long-term noninvasive ventilation, long-term invasive ventilation via tracheostomy, and limited or no respiratory intervention. In 2003, three years after Dr. John Bach published his study results on noninvasive ventilation for SMA Type I, an ethics article was published delineating this debate and concluded that "every child with SMA type I is unique in his or her disease process, family ideals, resources, and expectations. As such, the care of each child should be individualized to suit the child's needs". [20] A follow-up article in the same journal in 2005 noted that "current views appear polarized between those who would offer nothing, to those who would proceed as far even as tracheostomy and long term invasive ventilation for these infants." The authors discussed the ethical debate, and noted in conclusion that "the management of children with severe SMA is a highly controversial area. We believe that, if requested, NIPPV [nasal intermittent positive pressure ventilation] should be offered at night-time, and during the day for short periods of exacerbation, together with efforts to augment cough. We would discourage long term daytime NIPPV, and the use of tracheostomy". [21]

In response to this ethical debate, the National Institute of Neurological Diseases and Stroke sponsored an International

SMA Conference in 2004, with the goal of coordinating future SMA clinical trials. During this conference, it became apparent that the wide variation in medical care for SMA necessitated further discussion. Thus, the International Standard of Care Committee for Spinal Muscular Atrophy was formed in 2005, with the goal of establishing guidelines for clinical care of all aspects of SMA. The respiratory section of their consensus statement, published in 2007, included the clinical practices and study outcomes of Dr. John Bach, and the options for long-term ventilation and airway clearance described in his studies. The statement also noted that "essential to chronic management is discussion of the family's goals, which includes balancing caring for the child at home for as long as possible, long-term survival, quality of life and comfort, and the availability of resources". [22] With regards to respiratory care, the consensus statement provided a pathway for long-term survival of patients with all types of SMA but also noted that this pathway was one possible choice, and the ultimate choice depended on the individual patient and/or parents.

The Consensus Statement for Standard of Care in Spinal Muscular Atrophy included discussions not only on respiratory care, but also on gastrointestinal and nutritional care, as well as orthopedic care and rehabilitation. This multidisciplinary approach was necessitated by the fact that with improved respiratory care, patients with SMA, even type I SMA, were now surviving long enough to develop additional medical issues, such as scoliosis, contractures, and osteopenia. In addition, with type I SMA patients surviving not only months, but years, the emergent nutrition management done in the past necessarily gave way to longer term nutrition management and the resultant questions regarding optimization of this type of management. [22] In Dr. John Bach's 2000 study on noninvasive respiratory management of patients with type I SMA, the longest surviving patient was 81 months old, which was unthinkable fifty years earlier. [17] In improving respiratory care for patients with SMA, the opportunity for improving care in other areas was created, leading to major advancements in SMA care and research.

**Novel therapeutics further improve survival rates for SMA**

The next major change affecting respiratory care for patients with SMA did not arise from any additional technological advancements in ventilation or airway clearance, but in the development of systemic disease modifying therapies which improve respiratory function by maintaining respiratory muscle strength. Nusinersen (Spinraza), an antisense oligonucleotide given intrathecally every 4 months long-term, was approved by the FDA in 2016, and its mechanism of action is modification of the SMN2 gene product. Onasemnogene abeparvovec (Zolgensma), a one-time intravenous gene therapy, was approved by the FDA in 2019, but is currently only approved for patients under the age of 2 years. Risdiplam (Evrysdi) is administered enterally once daily and was approved by the FDA in 2020. This medication also modifies the SMN2 gene product and must be taken long-term. [23] All of these disease modifying therapies have the potential to preserve muscle strength, though the overall effect varies depending on SMA type and the age on administration. Many patients with SMA type I were above the age of two when gene therapy first became available, and this was not an option for them. However, many studies of various disease modifying therapies for SMA are currently ongoing, and the clinical picture will likely continue to change.

**Current trends in ventilatory support in SMA**

With the current availability of disease modifying therapies, the spectrum of respiratory muscle strength in patients with SMA is broad, ranging from normal muscle strength and lung function to very severe muscle weakness and lung function that is too low to accurately measure. The 2023 data from the Cure SMA Foundation showed that 38% of patients with SMA used some type of "breathing support." In this subset of patients, the most common type of support was bi-level positive airway pressure (BiPAP), at 66%, followed by tracheostomy with ventilator support (21%), supplemental oxygen (16%), and continuous positive airway pressure (CPAP) at 15%. [24] It is important to note that the consensus statement from the International Conference on SMA Standard of Care states that CPAP is not indicated in weaker patients, as it does not reduce the ventilatory load. The consensus statement also notes that while supplemental oxygen can be necessary in the case of acute respiratory failure, the goal is optimal ventilation, which should make supplemental oxygen unnecessary at the patient's respiratory baseline. [22] The context for the clinical use of CPAP and supplemental oxygen is not clear from this reported data, but the report does show that noninvasive BiPAP is currently the most common type of ventilatory support among patients in the United States with SMA.

This data on respiratory support from the Cure SMA Foundation discusses only positive pressure ventilation, but it should also be noted that negative pressure ventilation is still available as an option, though it is only rarely used in the pediatric neuromuscular population. The only commercially available and FDA-approved external negative-pressure ventilator in the United States currently is the Biphasic

Cuirass Ventilator (BCV) manufactured by Hayek Medical. This device has a plastic shell with foam seals which is strapped to the anterior chest and attached to the ventilator with pressure sensor tubing. A variety of modes are available, including modes similar to CPAP and BiPAP. Most recent clinical studies and case reports involving the use of the BCV focus on adult patients, with and without neuromuscular disease, and on pediatric patients without neuromuscular disease, largely due to anatomic issues in pediatric neuromuscular disease which can make the use of the BCV difficult. [25] Many pediatric patients with neuromuscular disease have significant scoliosis, and severe scoliosis is exceedingly common among patients with SMA. The presence of scoliosis can make the fitting of a BCV, which requires a tight seal, very difficult and sometimes impossible. [26] In addition, most weaker patients with SMA have gastrostomy tubes due to dysphagia, and the presence of the tube can also interfere with the placement of the cuirass. Finally, the BCV cannot relieve upper airway obstruction if this is present, and for this reason, the American Thoracic Society consensus statement on respiratory care of patients with Duchenne Muscular Dystrophy specifically states that "negative-pressure ventilators should be used with caution in patients with DMD due to the risk of precipitating upper airway obstruction and hypoxemia." [27] Thus, negative pressure ventilation currently is more useful for adult patients without significant scoliosis, and for pediatric patients without scoliosis or gastrostomy tubes, including infants with respiratory failure who can be difficult to fit with noninvasive positive pressure interfaces such as BiPAP masks. [25,28]

While positive pressure ventilation, specifically BiPAP, is currently the most common type of ventilation in SMA care, there is no clear consensus on optimal settings. In one 3-year study of 30 pediatric patients with a variety of neuromuscular disorders, the effectiveness of noninvasive ventilation in ventilatory insufficiency and sleep-disordered breathing was clearly demonstrated, with resolution of hypercapnia and hypoxemia with sleep for all study participants. 11 of these patients had SMA, 4 had Duchenne muscular dystrophy, and the remaining 15 patients had a variety of other neuromuscular disorders. While a variety of interfaces were used, all patients were placed on a BiPAP machine in pressure-assist mode, with the following settings reached after optimization for the individual patients: inspiratory positive airway pressure (IPAP) 13.9 ± 3.1 $cmH_2O$, expiratory positive airway pressure (EPAP) 4.4 ± 1 $cmH_2O$, and backup respiratory rate 19.6 ± 2.5 breaths per minute. [29] In general, as patients with neuromuscular disease typically do not have significant parenchymal lung disease, effective long-term mechanical ventilation can be achieved with a variety of pressure modes, with the simplest modes, as above, often working very well.

Thus, ventilatory techniques for patients with SMA do not appear to be significantly different from patients with other neuromuscular disorders, with the exception of one unique and important finding: in SMA patients, average $PCO_2$ values have been observed to be slightly low or in the lower end of the normal range in patients who do not yet need ventilatory support. An observational study published in 2020 of SMA patients, primarily type 2 and type 3, prior to the need for ventilatory support showed capillary $PCO_2$ levels that averaged 35.5 mmHg at patients' daytime respiratory baseline. Importantly, an increase in $PCO_2$ was observed in the year prior to requiring the initiation of mechanical ventilation (36.7 mmHg), and a further increase was noted at the start of mechanical ventilation (37.8 mmHg). The reason for this slightly low baseline $PCO_2$ is not clear at this point, though it is possible that it could be related to hyperventilation caused by altered carbon dioxide sensing in the brainstem or carotid bodies. Regardless, the study emphasized the importance of the low or low-normal range $PCO_2$ levels in SMA patients prior to requiring ventilation, as "increases of $PCO_2$ to normal levels may be a sign of pending respiratory insufficiency in some patients with SMA". [30]

## Summary

The current era of respiratory care of patients with SMA is a new frontier, with some patients unable to benefit from current disease modifying therapies, and some patients having received more than one therapy at a variety of ages, with a resultant wide spectrum of respiratory muscle strength. Currently, a pulmonologist in a neuromuscular clinic may see patients with SMA who are tracheostomy and ventilator dependent and with very little respiratory muscle strength, and may also see patients who received very early gene therapy and have no apparent respiratory muscle weakness, at least early in life. Currently, with the availability of long-term options for ventilation, supportive care, and disease modifying therapies, the annual mortality rate for SMA has dropped from 2.36 per 100 individuals in 2013 to 0.75 per 100 individuals in 2023, a decline of 68% over just a 10-year period. In addition, 2023 data from the Cure SMA Foundation showed that 50% of the individuals in their database of SMA patients in the United States are 18 years of age or older. While this data does not differentiate between types of SMA, their data also showed that 24% of the SMA patients in their database have an SMA Type 1 diagnosis, which has climbed from 20% in 2016. [24] With this

recent improvement in life expectancy for SMA, the topic of transition from pediatric to adult care, even for patients with SMA Type 1, is an important focus of current and future medical care.

Great strides have been made in the respiratory care of SMA patients since the first appearance of positive pressure ventilation in the 1940s and will continue to be made. However, in moving forward, it is vital to understand the natural history of spinal muscular atrophy and the history of the respiratory care it has required, not only to provide the best possible nuanced care for current patients with SMA on a spectrum of respiratory muscle strength, but also to learn from the advances made in SMA care and apply them to other respiratory disease processes, just as the care of patients with polio created so many advancements in the care of patients with SMA. All the physicians, engineers and inventors discussed in this review have seized the opportunity to use a new tool, or sometimes an old tool, for a new purpose, thus expanding the limits of medical care and changing the lives of hundreds of patients with SMA and their family members. As SMA Type I patients continue to reach adulthood, we will continue to learn more about SMA, and the medical frontier will continue to advance.

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