Respiratory Care of Adults with Chronic Pulmonary Disease

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Introduction

The last 30 years has seen a shift in the care of chronic pulmonary disease owing in large part to the advancements in medicine and technology. Comprehensive chronic disease management, as demonstrated in cystic fibrosis (CF) and muscular dystrophy (MD), has led to improved survival and quality of life for children diagnosed with previously fatal conditions, such that now these patients are living well into adulthood. The care of patients with chronic respiratory failure (CRF) has followed suit with this changing landscape of chronic disease management. CRF is defined as a condition persisting for greater than one month and requiring mechanical ventilation for part (at least 6 h) or all of the day to provide adequate ventilation [1]. Previously, children with CRF would be confined to a hospital with personnel trained in respiratory care and ventilator management, with patients and their families choosing home only for palliative or hospice care. Today, patients are offered multiple options for management of respiratory failure at home, albeit requiring significant education, family commitment, and medical community support.

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Like many changes in medicine, the initial impetus to move toward home care for patients with (CRF) was financial in origin. However, what seems to have grown out of this movement is a paradigm for managing complex medical patients safely while improving survival and quality of life. Integral to these improvements has been the development of programs supporting the care of chronically ventilated patients with CRF at home as well as multidisciplinary teams knowledgeable in the care of these patients. In addition to these changes, the trends in parental expectations of long-term survival in children with chronic respiratory failure has led to more parents initially taking on the long-term responsibility for these patients.

Chronic Respiratory Failure

CRF can be divided into three categories, each with different expectations of progression and need for lifelong mechanical ventilation. The first is CRF related to ventilatory muscle weakness, for example, neuromuscular diseases such as Duchenne's muscular dystrophy or quadriplegia. As home care for CRF was gaining popularity, these patients comprised more than 50 % of patients offered home mechanical ventilation (HMV), although reports reviewing the trends in HMV have shown that patients with other categories of CRF are being offered home care options more often [2]. Because these conditions

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are typically chronic and progressive, it is unlikely once a patient requires mechanical ventilatory support that they would be able to be successfully weaned from such support.

The second category of CRF is related to disorders of the lung parenchyma and airways, which in turn cause an increase in work of breathing. Examples include patients with bronchopulmonary dysplasia, interstitial lung disease, or chest wall deformities. As children with such conditions age and grow, their respiratory mechanics may improve, allowing them to be potentially weaned from ventilation.

The final category of CRF is related to disorders that cause a failure of neurologic control of breathing, for example, postencephalitis, brain trauma, or rarer conditions such as congenital central hypoventilation syndrome (CCHS). These conditions typically require lifelong ventilation, although in certain conditions, such as CCHS, ventilation may only be required at night.

Mechanical Ventilation

Home mechanical ventilation (HMV) can be described in different ways: positive versus negative pressure ventilation, invasive versus noninvasive ventilation (NIV), and mode of ventilation, to name a few. As a primary care provider (PCP), it is most important to understand the difference between invasive and the noninvasive mechanical ventilation and possible complications of each. Initiation of HMV should be accomplished in coordination with a specialist in pulmonary disease. Further, the ventilator settings, regardless of whether invasive or noninvasive ventilation is being employed, should first be established with inpatient titration and monitoring, typically during a sleep study. Optimal ventilation is achieved once the partial pressure of carbon dioxide ($PaCO_2$) normalizes. If a patient remains hypoxemic at this point, then oxygen supplementation is added. Oxygen should not be used until adequate ventilation is maintained, as it can mask cyanosis, worsen hypercapnea, and delay the initiation of ventilatory support [3]. Chronic management of mechanically ventilated patients employs monitoring carbon dioxide levels, typically with venous blood gases and intermittent sleep studies to ensure appropriate ventilation. Such monitoring may be required more frequently in children, as they tend to grow and require more frequent changes in their settings than do adults.

The goals of initiating HMV are to improve quality of life, improve mobility and rehabilitation potential, and to improve growth and development. Qualitative data on patients' experiences before and after initiation of mechanical ventilation are limited but suggest that patients' improvement in dyspnea directly impacts their quality of life. Though a minority of patients report negative social impacts and low self-esteem, these outcomes do not negate the reported improvement in quality of life due to relief of dyspnea [4]. The standard outcome measure of quality of life in children has been the patient's ability to reintegrate into the school system [5–7]. In order to accomplish these goals of HMV, ventilators must be small, lightweight, portable, durable, and able to be powered by a battery. As technology is advancing, these devices are becoming more portable and user friendly [8].

Noninvasive ventilation (NIV) employs a simpler device and is accomplished using a mask or nasal pillows, making this device less cumbersome. The success of NIV is highly dependent on the patient's ability to tolerate the masks. These devices exert a pressure on the face to maintain the seal and reduce air leak potential, leading to a risk of midface hypoplasia in children or pressure ulcers in adults. Regular monitoring of the site of contact of the mask on the face is important. Additionally, NIV cannot meet high-flow demands of certain patients, causing dyssynchrony between the patient and the ventilator, which the patient would perceive as dyspnea. As such, certain patients may not be comfortably ventilated with this type of ventilator.

Invasive mechanical ventilation is accomplished with a traditional home ventilator through a tracheostomy. This type of ventilator is a more cumbersome machine but can provide

more sophisticated modes of ventilation, allowing for improved patient comfort. Despite placement of a tracheostomy, most patients will still be able to vocalize.

The decision to use invasive or noninvasive ventilation should be made only after discussion among the patient, family, pulmonologist, and, if desired, the PCP. Traditionally if a patient requires mechanical ventilation for 24 h per day, then the invasive modality is recommended. However, in children, even those requiring ventilation for 24 h per day, noninvasive ventilation is often initiated until there is failure to adequately ventilate, failure to tolerate the mask, high risk of ongoing aspiration due to changes in bulbar function, or ineffective management of airway secretions [3, 5, 9].

Tracheostomy

A tracheostomy tube is a tube surgically placed in the neck extending into the trachea to allow for adequate ventilation, both assisted and non-assisted. These tubes require special attention and care to ensure proper cleaning and use. Typically, tracheostomy care is coordinated with an otolaryngologist (ENT)—a surgeon who most commonly places these devices. Once the decision is made to pursue invasive mechanical ventilation a tracheostomy tube is placed. The choice of the type of tube to use is made based on many factors. The two most common types of tracheostomies are plastic and metal. Plastic tubes are more flexible and typically smaller in nature. They are comprised of an outer cannula and an inner removable or disposable cannula. They come in standard sizes, although custom tubes can be made if a patient needs to have a longer portion entering the trachea, for example, in individuals with significant redundant neck tissue. Metal tubes are less flexible but more easily cleaned and therefore are used when patients have thick tenacious secretions or if they frequently obstruct their plastic tube. Tubes can also be uncuffed or cuffed, meaning there is a cuff on the distal intratracheal limb that is used to prevent air leaks in patients requiring mechanical ventilation. It is important to know that cuffs, if not managed properly, can cause damage as high pressure over extended periods of time can cause tracheal scarring. Tracheostomy tube cuffs are designed to be a high-volume, low-pressure system, but cuffs should only be inflated enough to prevent air leaks during mechanical ventilation, as overinflation can cause tracheal damage over time. Tracheostomy cuff tubes should not be inflated above a maximum pressure of 20 cm of water (cmH_2O).

Patients and caregivers will need training in tracheostomy care, which should be performed once a day. This typically involves changing the dressings and tracheostomy ties as well as cleaning the inner cannula. Plastic tubes must also be removed and new ones placed periodically. The ENT physician changes the tube the first time, typically 1–2 weeks after placement. There are no well-established guidelines on how often chronic tracheostomy tubes need to be changed thereafter, but recommendations range from every 2 weeks to every 3–4 months. This decision is best made in consultation with the ENT physician [10].

Complications of tracheostomy tubes are categorized into early and late. Early complications are those complications most commonly occurring up until the first tube change. They can be directly related to placement; for example, development of a false tract or pneumothorax, or local complications including infection, bleeding, or obstruction. The tracheostomy tube is sutured in place until the first change, so decannulation is uncommon. Late complications also include infection, obstruction, and bleeding, the most concerning of which would be an innominate artery fistula, typically occurring in the first 3 weeks after placement. Chronic tracheostomy tube placement can result in other complications over time including recurrent tracheitis, granulation tissue formation, which would lead to recurrent obstruction, and permanent tracheocutaneous fistula. Routine monitoring for development of these complications is mandatory. Some patients will develop chronic or recurrent tracheitis, as such they should routinely have tracheal cultures performed to determine the nature

of the bacterial flora in their airway in the event of an infection or an exacerbation.

Airway Clearance and Humidification

Patients with CRF due to any number of reasons often have difficulty managing respiratory secretions. For example, muscle weakness in patients with MD, increased secretion production in individuals with CF, and loss of cough reflex in those with central neurologic conditions can all pose challenges in the handling of respiratory secretions. To assist patients in managing these secretions, additional respiratory therapies should be administered 2–4 times daily. These therapies include inhaled airway hydration, mechanical airway clearance techniques, and frequent suctioning. There a multiple forms of airway clearance techniques including chest physiotherapy (CPT), high-frequency chest wall compression (HFCWC), and intrapulmonary percussive ventilation, all of which can be used with both invasive and noninvasive mechanical ventilation. Consultation with the pulmonologist should be sought to determine if the patient would benefit from these therapies.

Heated humidification is the delivery of air, with or without supplemental oxygen, that has been heated and passed over sterile water to increase the humidity. Heated humidification is mandatory for all patients who use invasive mechanical ventilation. Its benefits include prevention of tracheal mucosa dryness, preventing infection, prevention of inspissated secretions by increasing hydration, decreasing the risk of hypothermia, and increasing patient comfort. Care must be taken to ensure proper cleaning of the humidifier and use of sterile water.

Emergency Action Plans

Emergency action plans should be devised early in the planning of mechanical ventilation with input from the pulmonologist, ENT, and PCP. Key topics to include would be which physician should be contacted for an acute illness or particular issue and who is responsible for ordering specific equipment and supplies. Families should be provided with the appropriate equipment in the event of an emergency. For example, they should have equipment and training to perform manual ventilation in the event of power outages and extra tracheostomy tubes for placement in the event of an accidental decannulation.

Weaning from Ventilation

Patients will ask if weaning off of the ventilator is an option. This often is not possible if the condition requiring mechanical ventilation is a chronic progressive condition. However, for children who are placed on mechanical ventilation due to bronchopulmonary dysplasia or tracheobronchomalacia, it is possible that, as they grow, their respiratory mechanics may improve such that HMV is no longer required. If weaning is a potential option, after consultation with the pulmonologist, patients are usually liberated from the ventilator for progressively longer periods of time rather than adjusting ventilator settings. Careful monitoring of vital signs and assessment of patient tolerance of being off the ventilator are critical. The final steps of weaning from a ventilator at night should be done in a monitored setting, such as an inpatient stay or during sleep study.

Caregivers

Caregivers are the most integral part of the care of a ventilated patient. It has been suggested that it is not the education level of the caregiver that predicts success but rather the commitment and dedication to appropriate home care [8]. It is a very time-consuming, physically and mentally rigorous undertaking to be a caregiver for a patient requiring mechanical ventilation. The role of home health care services is to provide the caregiver a reprieve to allow them to rest and care for themselves as well as to provide support for certain patient activities such as bathing,

Table 19.1 Caregiver Education Checklist. Adapted with permission from Preutthipan [8]

General	Understanding of underlying disease Concerning signs and symptoms Inhaled therapies/airway clearance techniques Emergency care/CPR Emergency call number Medical history document
Oxygen therapy	Understand why it is necessary Oxygen source Care/maintenance of equipment Flow rate of oxygen Complications of oxygen therapy
Tracheostomy care	Understand why it is necessary Safe activities of daily living Communication and speech Cleaning and sterilization of equipment Suctioning techniques and equipment Tube and ties change Accidental decannulation plan In-home safety Transportation safety
Ventilator care	How the ventilator works Understanding alarms/complications Problem-solving ventilator malfunction Check ventilator and humidifier Mask/tracheostomy tube interface Cleaning and sterilization of equipment In-home safety Battery and power supply Manual bag mask ventilation

CPR cardiopulmonary resuscitation

respiratory treatments, and tracheostomy care. Prior to a mechanically ventilated patient being discharged to home, the caregivers must go through extensive education to learn tracheostomy care, understand the ventilator, coordinate emergency action plans, and be instructed in basic life support, among many other things [8] (see Table 19.1).

Conclusion

Over the last decades, improvements in clinical care and technology have afforded patients with CRF increases in survival and improvements in quality of life. Encompassed in these changes is the involvement of multidisciplinary team of family and caregivers as well as pulmonary and primary care physicians. Successful home mechanical ventilation is now an option for these individuals and experience has shown that, with a multidisciplinary approach to care and dedicated family and caregivers, HMV can improve both quality of life and survival among individuals with CRF. Multiple support systems have been established to assist in these endeavors.

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