Chapter 12
Long-Term Ventilation and Home Care

Shekhar T. Venkataraman

As survival from PICU with critical illness has increased, many children are being discharged from the hospital who are technology-dependent. Respiratory care, in these children, can range from simple supplemental oxygen to mechanical ventilation, which can range from partial ventilation (e.g., night time ventilation) to 24-h ventilator assistance. Long-term care in a tertiary care center is expensive and it can alternatively be provided in a specialized chronic care center or at home. Home care is psychosocially more acceptable to the families, less expensive, and may provide a better quality of life for technology-dependent children. This chapter will review the definitions of chronic respiratory failure, options for support with chronic respiratory failure and the logistics of discharge of these patients.

12.1 Definitions and Causes

Chronic respiratory failure (CRF) is defined as respiratory failure that persists after the primary process has resolved or it is due to a cause that will remain for the foreseeable future even for the entire life span. CRF may be due to persistent or progressive hypoxemia and/or hypercarbia requiring respiratory support. The conditions that lead to CRF can be classified into those that are likely to improve, remain static, or continue to progress resulting in end-stage respiratory failure. Additionally, there are patients who require care related to obstructive sleep apnea,

S. T. Venkataraman (✉)
Professor, Departments of Critical Care Medicine and Pediatrics, University of Pittsburgh School of Medicine, Pittsburgh, PA, USA
e-mail: venkataramanst@upmc.edu

Medical Director, Respiratory Care Services, Children's Hospital of Pittsburgh, 4401 Penn Avenue, Faculty Pavilion 2117, Pittsburgh, PA 15224, USA

A. P. Sarnaik et al. (eds.), *Mechanical Ventilation in Neonates and Children*,
https://doi.org/10.1007/978-3-030-83738-9_12

Table 12.1 Causes of chronic respiratory failure

Cause of respiratory failure	Diseases
Respiratory pump failure	Static muscle weakness/Loss of function Spinal cord injury Phrenic nerve injury Skeletal/Chest wall deformities Scoliosis Congenital skeletal malformations Progressive muscle weakness Neuropathies Myopathies Mitochondrial disorders Spinal muscular atrophy
Respiratory drive	Congenital central hypoventilation syndrome Brain/brainstem injury Central nervous system tumors Degenerative CNS disorders
Structural abnormalities	Airway malformations Craniofacial malformations Tracheomalacia Bronchomalacia Acquired airway diseases Obstructive sleep apnea
Pulmonary parenchymal and vascular disorders	Chronic lung disease of infancy (bronchopulmonary dysplasia) Recurrent aspiration syndromes Cystic fibrosis Congenital heart disease Post-inflammatory or post-infectious Lung hypoplasia

and those who require support as part of their palliative care (Table 12.1). Ventilator dependence is defined as the requirement for mechanical ventilation for longer than a month.

Individual patients can have more than one cause for chronic ventilator dependency. For example, patients with neuromuscular weakness can develop kyphoscoliosis which produces restriction to breathing and exacerbation of hypercarbia. Patient with anoxic encephalopathy may not only have an abnormal respiratory drive but also develop recurrent aspiration and obstructive sleep apnea from bulbar dysfunction. The approach to chronic respiratory care will be different depending on the natural course of the underlying disease as well associated complications.

12.2 Pathophysiological Considerations of Chronic Respiratory Failure

Respiratory Pump Disorders—Weakness or Absence of Muscle Function

Neuromuscular Diseases

Most neuromuscular disorders are progressive and result in worsening respiratory function due to several mechanisms and result in multiple morbidities. Included in this category are muscular dystrophies and myopathies. Respiratory muscle weakness progresses to ventilator failure requiring increasing levels of support. Airway clearance is compromised predisposing to recurrent episodes of orotracheal aspiration and recurrent episodes of atelectasis and lower respiratory tract infections. Chest wall and skeletal mechanics are also altered and often lead to kyphoscoliosis and rigidity of the thoracic cage due to ankylosis of costovertebral joints and stiffening of the ligaments and tendons, further compromising gas exchange. Lower airway mucus plugging and increased airway resistance are the consequence of small airway caliber both anatomically and functionally. These imbalances lead to a restrictive respiratory system with reduced vital capacity and total lung capacity. These factors increase the work of breathing which coupled with muscle weakness lead to fatigue and ventilatory failure. Sleep disturbance is common in these patients including obstructive sleep apnea due to bulbar weakness. Mechanical ventilation, especially noninvasive ventilation, has been shown to be beneficial and improve the quality of life in these patients.

Nerve Injury or Illness

Nerves innervating the respiratory muscles may be injured or inflamed and cause either muscle weakness or absence of muscle function. Phrenic nerve injury may occur during childbirth, cardiac surgery and other thoracic surgery. The injury may be unilateral or bilateral resulting in either paresis or total paralysis. Often this injury is irreversible and may necessitate positive pressure support. Spinal cord injury above the level of C3 causes paralysis of the diaphragm as well as the intercostal muscles. Long-term mechanical ventilation through a tracheostomy can prolong life in many of these patients. Amyotrophic lateral sclerosis is a disease that causes the death of neurons controlling voluntary muscles and results in progressive muscle weakness and ventilatory failure similar to neuromuscular disorders. Spinal muscular atrophy is much more common than ALS in children compared to adults.

Respiratory Pump Disorders—Skeletal and Chest Wall Diseases

Distortion of the thoracic cage due to scoliosis causes restrictive respiratory disease and may result from muscle weakness, nerve injury or from congenital malformations. Chronically, the load on the respiratory muscles can result in fatigue and ventilatory failure. Chest wall diseases such as asphyxiating thoracic dystrophy cause restriction to breathing and can be life threatening without mechanical ventilation and reparative surgery.

Respiratory Drive Disorders

Several congenital or acquired central nervous system disorders result in an abnormal respiratory drive characterized by an impaired response to hypercapnia. Congenital central hypoventilation syndrome, Prader Willi syndrome, and rapid-onset obesity with hypothalamic dysfunction, hypoventilation, and autonomic dysregulation (ROHHAD) are some examples of congenital disorders resulting in an abnormal respiratory drive leading to hypercarbia from chronic hypoventilation. Children with Chiari malformation can have a blunted response to hypercapnia and can manifest with both central and obstructive apnea during sleep. Acquired brain stem lesions or injury may also affect respiratory drive resulting in chronic hypoventilation leading to hypercarbia. Progressive and degenerative brain disorders such as Leigh's disease can also affect the neurons involved with respiratory drive and cause hypoventilation.

12.3 Approach to Long-Term/Home Care Ventilation

The benefits of home care include: (1) Cost-efficiency, (2) Quality of life, (3) Psychosocial aspects of the family and the patient, and (4) Family cohesiveness. On the other hand; caregiver stress, concern for inadequate care, neglect are some of the issues of home care which may increase the risk for morbidity. The success of home care depends on the willingness and ability of family caregivers to provide care in the home, and community resources. Alternate sites of chronic ventilation are not available as widely despite the need for such centers.

Options for Respiratory Support in Technology-Dependent Children

There are several options for respiratory support for technology-dependent children. These children may be with or without an artificial airway such as a tracheostomy. Supplemental oxygen may be needed either continuously or intermittently for those patients with parenchymal lung disease with intrapulmonary shunting and venous admixture. When the severity of lung disease and gas exchange abnormalities increase, then positive pressure support may be needed. Table 12.2 lists the type of respiratory support that may be needed in technology-dependent children.

HME is described in Chap. 11. The choice of respiratory support depends upon the severity of lung disease and respiratory failure as well as the desired quality of life.

Goals of Respiratory Support

The goals of long-term respiratory care include: (1) provision of comprehensive, cost-effective respiratory care, (2) enhancement of quality of life, (3) reduction of morbidity, and (4) extend life wherever possible. The setting for post-acute care will depend on the degree of medical stability of the patient and availability of resources outside the pediatric intensive care unit. These include a care site within the hospital, a subacute or long-term facility or home care. The first consideration is the

Table 12.2 Respiratory support in technology-dependent children

Without an artificial airway
1. Supplemental oxygen
a. Intermittent
b. Continuous
2. Noninvasive positive pressure support
a. Continuous positive airway pressure
i. Intermittent
ii. Continuous
b. Spontaneous ventilation with Positive pressure assistance (e.g., BiPAP)
i. Intermittent
ii. Continuous
c. Negative pressure ventilation
i. Intermittent
ii. Continuous
d. Sip-ventilation
i. Intermittent
ii. Continuous
With an artificial airway
1. No supplemental oxygen with or without a heat and moisture exchanger (HME)
2. Supplemental oxygen
a. Intermittent
b. Continuous
3. Continuous positive airway pressure
i. Intermittent
ii. Continuous
4. Spontaneous ventilation with Positive Pressure Assistance
i. Intermittent
ii. Continuous
5. Mandatory ventilation with or without Pressure Support
i. Intermittent
ii. Continuous

medical stability of the patient. The minimum requirements are a stable airway (natural or a tracheostomy), stable clinical status and care demands that can be met by the resources of the care site.

The objectives of long-term mechanical ventilation are dependent on the cause of respiratory failure. In patients with reversible causes of CRF, respiratory support is provided to support and improve gas exchange until there is resolution of respiratory failure. CRF such as bronchopulmonary dysplasia is often associated with poor somatic growth with delayed development. Mechanical ventilation, in these patients, can result in improved somatic growth as well as neurologic development.

Consideration for Mechanical Ventilation

Noninvasive ventilation by positive pressure can be delivered through nasal or oronasal interface. Positive pressure can relieve upper airway obstruction as well as improve minute ventilation and unload inspiratory muscles. If long-term ventilatory

assistance is provided in the hospital setting, then either intensive care ventilators or portable home ventilators may be used. Portable ventilators use pistons or turbines to generate the selected volume or pressure, and can do so at lower flow rates. Noninvasive ventilation may be used intermittently, usually at night or when the patient is sleeping. With more severe or progressive disease, NIV may be required continuously. Similar to short-term NIV, for long-term NIV the ventilator settings must relieve patient symptoms.

Long-term NIV has significant physiologic effects depending on the disease process. In general, it improves the quality of life for many patients at home. It also improves night time sleep with less disturbed sleep from respiratory distress. In many patients, chronic sleep apneas and carbon dioxide retention can predispose to the development of systemic and pulmonary hypertension. NIV, by ameliorating these manifestations, decreases the incidence of systemic and pulmonary hypertension. In patients with chronic respiratory failure who are candidates for lung transplantation, NIV can act as an effective bridge toward transplantation. Table 12.3 lists the physiologic effects and outcomes from some of the conditions for which long-term NIV is employed.

Invasive ventilation via tracheostomy is needed for children who require continuous mechanical ventilation. Tracheostomy tube should be of an optimal size, neither too large that may cause injury to the trachea nor too small so as to cause a considerable leak around the tube that can compromise minute ventilation. Intermittent use of speaking valve may facilitate speech development. The presence of a tracheostomy increases the complexity of care including training of caregivers

Table 12.3 Physiologic effects and outcomes of long-tern NIV

Neuromuscular disorders
1. Improves daytime gas exchange 2. Ameliorates hypoventilation 3. Decreases intermittent sleep apneas and other sleep-disordered breathing 4. Improves quality of life 5. Decreases hospitalization 6. Increases survival especially when coupled with assisted cough and aggressive secretion clearance 7. Improves thoracoabdominal coordination during sleep in SMA types 1 and 2
Cystic fibrosis
1. Minimizes acute on chronic respiratory failure with exacerbations 2. Ameliorates sleep disturbance 3. Serves as a bridge to transplantation
Obstructive sleep apnea
1. Reverses or reduces obstructive sleep apnea 2. Improves sleep-disordered breathing 3. Provides interim solution while allowing the patient to grow and postpone surgery

on suctioning, cleaning and changing the tracheostomy tube. The presence of a tracheostomy tube increases the risk for infection. Airway complications related to the presence of a tracheostomy tube include infection at the stoma site, granuloma formation, tracheal stenosis, and fistula formation. CPAP provided through a tracheostomy can unload respiratory muscles and enhance minute ventilation by relieving upper airway obstruction. BiPAP devices provide positive pressure assistance to spontaneous breaths and may be required in patients with chronic hypercarbia. When mandatory ventilatory assistance is required, the goal of therapy is to relieve distress and allow the patient to breathe comfortably. Care should be taken not to over-ventilate the patient, especially those with neuromuscular disorders to avoid causing respiratory muscle atrophy from disuse.

Ventilator settings must relieve patient symptoms. The approach that is generally recommended for long-term NIV is the low–high approach which consists of starting at fairly low level of support that is gradually increased until desired effect is observed. A back-up rate sufficiently high to control breathing nocturnally can rest the respiratory muscles and prevent apnea, especially in patients with neuromuscular disease. There has been an increase in the continuous use of NIV, especially in patients with neuromuscular disorders. This has resulted in prolonging survival and improving quality of life in these patients.

Transitioning from an ICU-Specific Ventilator to a Portable Home Ventilator

One of the key steps before discharge from the ICU for patients on long-term positive pressure support is to transition to a home ventilator (HV). The patient must be medically stable with the current ventilator settings. If any specialty gases such as nitric oxide or Heliox have been used, they need to be weaned off. The patient must be gaining weight and able to tolerate therapies without significant changes in gas exchange or work of breathing. Airway must be stable, whether it is the natural airway or a tracheostomy. There are no strict guidelines for ventilator settings or pressures to make the transition. Some HVs may not be able to provide the ventilator settings needed for the patient. In that case, either an alternative HV should be tried or the patient needs to be transitioned when the ventilator settings are lowered and tolerated. The ventilator settings in the HV may need to be adjusted further after the transition to ensure that the patient is breathing comfortably with adequate gas exchange. When the patient is able to tolerate HV for at least 2 weeks without any changes made to the ventilator settings, the patient is considered ready for discharge.

An ideal HV should: (1) Be light-weight and portable, (2) Have a long internal battery life, (3) Able to provide continuous flow, (4) Able to compensate for leaks, (5) Have multiple modes of ventilation available, and (6) Easy to trouble-shoot. Most of the current HVs are portable and approved for children weighing 5 kg or greater. Portability and a long internal battery life allow the patients to leave their homes and travel either for appointments or family gatherings. A long battery life may be life-saving in areas where power outages are common. The actual battery life may be different from the manufacturer's estimations since those are based on providing a low level of ventilator support in adults. Battery life is inversely

proportional to level of support needed for the patient. Higher ventilator pressures and rates will decrease the battery life. The type of support provided by HV is listed in Table 12.2.

Monitoring of Patients with Long-Term NIV

Monitoring patients who are on long-term NIV requires an assessment of their respiratory function. Pulse oximeters are useful in detecting hypoxemia especially during sleep. End-tidal CO_2 monitors are only used in the clinic or hospital. Treatment of sleep-disordered breathing improves the quality of life and may prolong survival. Polysomnography may be required to document sleep-disorders but is more expensive than nocturnal pulse oximetry. If a patient has bulbar weakness or severe obesity, there may be obstruction of the upper airway or obstructive sleep apnea in addition to sleep hypoventilation. Both problems are treated with nocturnal ventilation.

12.4 Logistics of Home Care

Home care requires a team approach with interaction between several health care personnel, viz., a primary physician, home care nurses, respiratory therapists, social workers, sometimes the State Health Agency, and the family and the patient. First, the patient must be ready for home care. In children who have a tracheostomy, a mature stoma is essential. Inspired oxygen requirement should not be greater than 35%. PaO_2 should be greater than 60–70 torr with a $PaCO_2$ less than 60 torr with a normal arterial pH at relatively low ventilator settings. The family must demonstrate not only a desire to provide home care but also a minimal ability to provide various aspects of home care. Health care personnel must be available in the local community to assist the family in providing care. The home must have adequate facilities to maintain and operate all necessary equipment and supplies. Contingency plans must be made for emergencies. The location of the home has major implications for home care. The home must be easily accessible by standard transportation. A home care provider should be available on a 24-h basis to respond to emergencies. The home must have adequate space to accommodate all the caretakers and the required equipment. A thorough knowledge of the limitations of various devices is essential for the respiratory care personnel coordinating home care. Parents who are motivated to providing home care for their children are willing to take on the cumbersome responsibility of carrying all the necessary equipment from place to place just to have the benefit of having the child at home. When a technology-dependent child is being discharged home, two adults must be willing and able to learn and assume all aspects of the child's daily care, including dosages and indications for all medications being used, feedings, airway clearance and respiratory assessment, ventilator assessment and troubleshooting, and equipment care. If the child is to receive mechanical ventilation through a tracheostomy, the family caregivers must also learn how to suction the artificial airway and

perform routine and emergency tracheostomy tube changes. In addition, there must be adequate financial support from third-party payers to provide the equipment and supplies necessary to care for the child at home. The residence in which the child will be cared for must have adequate space for the child, equipment, and visiting health care providers. The home must have running water, heat, electricity, and a working telephone. Entrances must be accessible for patients confined to a wheelchair.

The discharge plan must also include the amount of skilled nursing care the family will require. All families of children who cannot correct an airway or ventilator problem or call for help should be offered skilled nursing care for at least a portion of the day to allow caregivers to sleep with reassurance that the child's welfare is not at risk. Funding for these services, which are the most expensive component of the home care of technology-dependent children, should be guaranteed by third-party payers with periodic reassessments established to determine ongoing needs. While there are no uniform criteria for establishing the number of nursing hours provided, it should be determined by the medical needs of the child, the capabilities of the family, and other demands on family members such as work requirements, care of other children in the home etc. To allow caregivers time off from continuous medical care and monitoring of the child, funded respite care should also be built into the discharge plan as it has been repeatedly identified as an essential component of the home care plan to help relieve stress and caregiver burnout.

Suggested Readings

1. McKienan C, Chua LC, Visintainer PF, allen H. High flow Nasal Cannula therapy in infants with bronchiolitis. J Pediatr. 2010 Apr; 156(4):634–8
2. Milesi C, Pierre AF, Deho A, et al. A multicenter randomized controlled trial of a 3-l/kg/min versus 2-l/kg/min high-flow nasal canula flow rate in young infants with severe viral bronchiolitis (TRAMONTANE 2). Intensive Care Med. 2018 Nov; 44(11):1870
3. Wing R, James C, Maranda LS, Armsby CC. Use of high-flow nasal cannula in the emergency department reduces the need for intubation in pediatric acute respiratory insufficiency. Pediatr Emerg Care. 2012 Nov;28(11):1117−23
4. Franklin D, Babl FE, Schlapbsch LJ, et al. A randomized trial of high-flow oxygen therapy in infants with bronchiolitis. N Engl J Med. 2018;378:1121–31.
5. Bhashyam AR, Wolf MT, Marcinkowski AL, Saville A, Thomas K, Carcillo JA, Corcoran TE. Aerosol delivery through nasal cannulas: an in vitro study. J Aerosol Med Pulm Drug Deliv. 2008;21:181–8.
6. Corcoran TE, Sallille A, Adams PS, et al. Deposition studies of aersol delivery by nasal cannula to infants. Pediatr Pulmonol. 2019;54:1319–25.
7. Fontanari P, Burnet H, Zattara-Hartmann MC, Jammes Y. Changes in airway resistance induce by nasal inhalation of cold dry, dry, or moist air in normal individuals. J Appl Physiol. 1996;81:1739–43.
8. Byerly FL, Haithcock JA, Buchnana IB, et al. Use of high flow nasal cannula on a pediatric burn patient with inhalation injury and post-extubation stridor. Burns. 2006;32:121.

9. AL Ashry HS, Modrykamien AM. Humidification during Mechanical Ventilation in the Adult Patient. BioMed Res Internat. 2014; (715434):12
10. Baudin F, Gagnon S, Crulli, et al. Modalities and complications associated with the use of high-flow nasal cannula: experience in a pediatric ICU. Respir care 2016;61(10):1305–10
11. Sarnaik SM, Saladino RA, Manole M, Pitetti R, Arora G, Kuch BA, Orr RA, Felmet KA. Diastolic hypotension is an unrecognized risk factor for ß-agonist-associated myocardial injury in children with asthma. Pediatr Crit Care Med. 2013 Jul;14(6):e273–9.
12. Kalister H. Treating children with asthma, a review of drug therapies. West J Med. 2001;174:415–20.
13. Petersen W, Karup-Pedersen F, Friis B, Howitz P, Nielsen F, Stromquist LH. Sodium cromoglycate as a replacement of inhaled corticosteroids in mild-to-moderate childhood asthma. Allergy. 1996;51:870–87.
14. de Benedictis FM, Tuteri G, Pazzelli P, Berotto A, Bruni L, Vaccaro R. Cromolyn versus nedocromil: duration of action in exercise-induced asthma in children. J Allergy Clin Immunol. 1995;96:510–4.
15. Wisecup S, Eades S, Hashmi SS, Samuels C, Mosquera RA. Diastolic hypotension in Pediatric Patients with Asthma Receiving Continuous Albuterol. J Asthma. 2015 Sep; 52 (7):693–8
16. Davis MD, Donn SM, Ward RM. Administration of inhaled pulmonary vasodilators to the mechanically ventilated neonatal patient. Pediatr Drugs. 2017;19(3):183–92.