CHRONIC CONDITIONS

Basics of Mechanical Ventilation for the Outpatient Pediatrician

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EDUCATIONAL GAP

Advancements in medical technology have allowed a growing number of children with complex medical conditions and technology dependence to be discharged home from the hospital, including patients receiving mechanical ventilation. Familiarity with the principles of mechanical ventilator support, common modes used in the outpatient setting, and common complications is imperative in taking care of these children.

OBJECTIVES *After completing this article, readers should be able to:*

1. Describe the physiology of breathing.
2. List the different types of respiratory support.
3. Describe the basics of mechanical ventilation.
4. List the indications for mechanical ventilation and home mechanical ventilation.
5. Manage the initial settings of invasive mechanical ventilation.
6. List the complications of hospital and home mechanical ventilation.

INTRODUCTION

The respiratory system consists of the airways, which conduct gases to and from the lungs, which function as the gas exchange system, and the respiratory muscles, which act as the ventilatory pump. The respiratory system's primary function is gas exchange, which includes oxygenation and ventilation. Oxygenation refers to the transfer of oxygen from alveoli to arterial blood, and ventilation refers to removing carbon dioxide, a byproduct of cellular metabolism, from the pulmonary arterial blood. Respiratory failure ensues when the respiratory system fails to effectively supply oxygen or remove carbon dioxide, leading to hypoxemia, hypercapnia, or both. Infants and young children are at higher risk of increased work of breathing and respiratory failure due to anatomic and physiologic differences from adults (Table 1).

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ABBREVIATIONS

AC	assist-control
BiPAP	bilevel positive airway pressure
BMV	bag-mask ventilation
CMV	controlled mandatory ventilation
CPAP	continuous positive airway pressure
DP	driving pressure
e time	exhalation time
F_{IO_2}	fraction of inspired oxygen
HFNC	high-flow nasal cannula
HFOV	high-frequency oscillation ventilation
HMV	home mechanical ventilation
i time	inspiratory time
I:E ratio	inspiratory to exhalation ratio
IMV	invasive mechanical ventilation
NIV	noninvasive ventilation
NPV	negative-pressure ventilation
PARDS	pediatric acute respiratory distress syndrome
PEEP	positive end-expiratory pressure
PIP	peak inspiratory pressure
Pplat	plateau pressure
PS	pressure support
RV	right ventricular
Sa_{O_2}	saturation of arterial oxygen
SIMV	synchronized intermittent mandatory ventilation
VS	volume support
V_T	tidal volume

TABLE 1. Anatomical and Physiological Differences in the Respiratory System in Infants/Young Children

Variable	Findings in Children	Implication
Airway	Smaller diameter; obligatory nasal breathers	Increased resistance in nostrils and bronchioles when obstructed with mucous
Functional residual capacity	Lower due to smaller and fewer alveoli	Increased tendency for lungs to collapse
Chest wall compliance	Higher due to less ossification	Predisposition to chest wall collapse and respiratory distress
Rib orientation and mechanics	Horizontal, with diaphragm excursion leading to an increase in thorax diameter	Higher risk of respiratory compromise with abdominal distension
Diaphragm and intercostal muscles	Immature musculature	Weaker and easier-to-fatigue musculature

PHYSIOLOGY OF NORMAL BREATHING

Normal spontaneous breathing is negative-pressure breathing. It occurs via contraction of the diaphragm and, to a lesser extent, the external intercostal muscles. Negative-pressure breathing leads to expansion of the chest cavity, creating more negative intrapleural pressure, allowing air to fill the lungs. Normal exhalation, on the other hand, is a passive process. The inspiratory muscles relax, allowing the lung, chest wall, and abdominal structures to recoil, leading to air exhalation (Figure 1). Normal breathing constitutes a small proportion (<5%) of total oxygen consumption. The respiratory centers in the brainstem (medulla oblongata and pons) regulate rhythmic breathing. The dorsal respiratory group and the ventral respiratory group, which are present in the medulla oblongata, play a crucial role in generating rhythmic breathing patterns. The pneumotaxic and apneustic centers that are present in the pons modulate the medullary respiratory centers. Central and peripheral chemoreceptors regulate ventilation. Central chemoreceptors in the medulla oblongata and pons are very sensitive to changes in pH and $Paco_2$; in cases of chronic hypercarbia, these receptors become less sensitive. Peripheral chemoreceptors in the carotid and aortic bodies respond to changes in Pao_2, although to a lesser extent. Signals are sent from the respiratory centers to the respiratory muscles via the phrenic and intercostal nerves (Figure 2).

RESPIRATORY SUPPORT

Respiratory support may be needed either to assist with the work of breathing or to improve gas exchange. Respiratory support devices can be divided into oxygen delivery devices, noninvasive positive- or negative-pressure devices, and invasive mechanical ventilation (IMV).

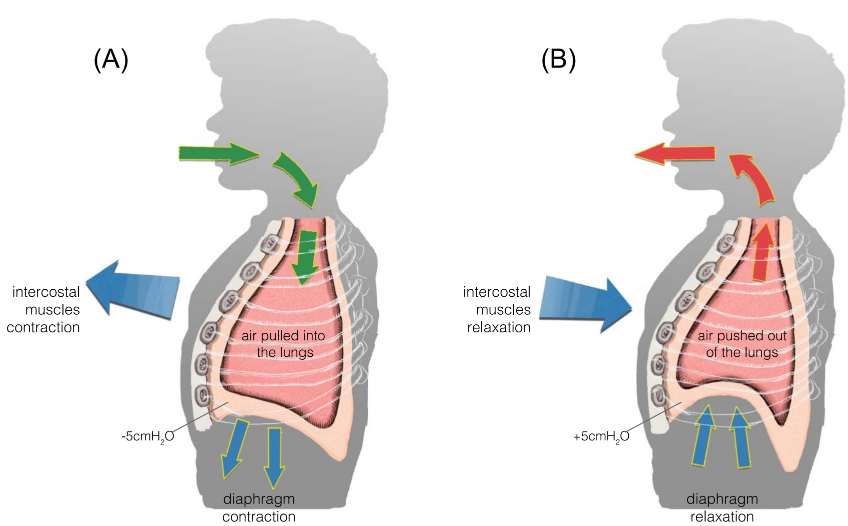

FIGURE 1. Normal breathing. Diaphragm and intercostal muscle contraction create negative intrapleural pressure, pulling air into the lungs (A). Diaphragm and intercostal muscle relaxation allow for lung recoil, generating positive intrapleural and alveolar pressure, pushing air out of the lungs (B). (Created by the authors using Adobe Illustrator).

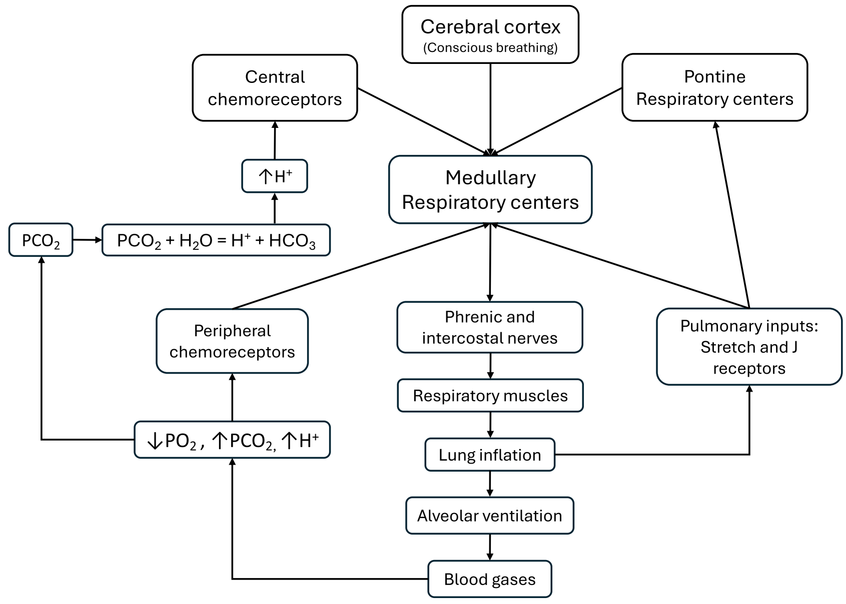

FIGURE 2. Schematic representation of the control of breathing. The medullary respiratory centers respond to signals from the cerebral cortex, peripheral and central chemoreceptors, pontine respiratory centers, and pulmonary receptors. Signals are then transmitted to the respiratory muscles via the phrenic and intercostal nerves. Central chemoreceptors are sensitive to changes in Pco_2 through changes in H^+ concentration (pH). Peripheral chemoreceptors are sensitive to changes in Po_2 and, to a lesser extent, pH and Pco_2.

Oxygen delivery devices are categorized into low and high flow, depending on the liters of oxygen delivered per minute. All these devices are connected to an oxygen source, and the fraction of inspired oxygen (Fio_2) delivered to the patient will depend on the percentage of room air mixed with the supplemental oxygen. The device that provides the most Fio_2 to a spontaneously breathing child is the non-rebreather face mask. Compared with other high-flow devices, a high-flow nasal cannula (HFNC) also delivers heated and humidified air. Oxygen delivery devices, except for HFNC, may not provide ventilatory support. When patients develop hypercapnic respiratory failure or hypoxemic respiratory failure that is refractory to supplemental oxygen, positive-pressure ventilation might be needed. Various respiratory support devices are listed in Table 2.

OVERVIEW OF MECHANICAL VENTILATION

Mechanical ventilation is a life-support technology that assists patients who cannot breathe effectively due to respiratory, neuromuscular, or neurologic conditions. Mechanical ventilation can be provided in a variety of modes and with different interfaces. Some key concepts and types of ventilation are discussed in this section.[1–3]

Positive-Pressure Ventilation

Positive-pressure ventilation refers to the delivery of air to the lungs using positive pressure via noninvasive ventilation (NIV) or IMV.

NIV refers to delivering positive pressure to the airway via nasal or naso-oral interfaces without requiring endotracheal intubation or tracheostomy. Some of the benefits of NIV include improved patient comfort, avoidance of endotracheal intubation, decreased need for sedation, and decreased risk for hospital-acquired infections. Positive pressure is delivered via continuous positive airway pressure (CPAP) or as biphasic positive airway pressure (BiPAP). For NIV to be successful, the patient requires an intact upper airway, airway protective reflexes, and an adequate seal on the mask interface to avoid pressure loss.

IMV requires endotracheal intubation or tracheostomy to provide positive pressure to the airways. IMV is indicated when the pulmonary function cannot be sustained with noninvasive modalities, as in cases of severe and rapidly progressing respiratory failure, significant neurologic or neuromuscular impairment, refractory shock with multiorgan dysfunction, or a combination of these conditions.

Negative-Pressure Ventilation

Negative-pressure ventilation (NPV) mimics normal physiologic respiration. A chest shell (cuirass) is placed external to the chest, generating subatmospheric pressure to initiate inspiration. The chest wall moves outward and induces negative pressure in the intrapleural cavity, allowing alveolar recruitment and airflow. In contrast to normal spontaneous breathing, the exhalation process is active with NPV, as the cuirass increases the intra-alveolar pressure to allow air to

TABLE 2. Respiratory Support Devices for Outpatient Use

Respiratory Support Devices	Comments
Low-flow devices	Flow limited to 1–10 L/min; F_{IO_2} will be dependent on % room air mixed with the supplemental oxygen.
Blow-by oxygen	F_{IO_2} and flow depend on the distance at which the mask is held.
Low-flow nasal cannula	Flow is limited to 6 L/min; for every 1 L/min increase in flow, the F_{IO_2} increases by 0.04 in adults, and higher in children.
Simple face mask	Flow 5–10 L/min; it can provide up to 0.5 F_{IO_2}.
High-flow devices	Allow for oxygen flow >10 L/min.
Venturi mask	Allows precise measurement of F_{IO_2} by using different sizes of ports, up to 0.6 F_{IO_2}.
High-flow nasal cannula	Can provide heated and humidified air via nasal prongs. Used with a blender to measure precise F_{IO_2}. Typical setting is 1–2 L/kg/min in young children and up to 60 L/min in adults.
Positive pressure	
NIV	
Ventilation bag (BMV)	BMV provides rescue ventilation to patients with apnea or poor respiratory effort via a face mask. Two main types of ventilation bags are available: self-inflating and non-self-inflating.
NIV via nasal cannula	Nasal prongs should be fitted to the size of the patient's nares.
NIV via nasal mask	The mask that covers the nose allows patients to speak and eat.
NIV via naso-oral mask	The mask covers both the nose and mouth. Increased risk of nasal bridge damage if fitted improperly.
NIV via full-face mask	The mask covers the eyes, nose, and mouth. There is less risk of skin breakdown in the nasal bridge. Difficult to fit in small infants, more comfortable in older children.
IMV	IMV requires endotracheal intubation or tracheostomy.
Conventional mechanical ventilation	Set tidal volumes or inspiratory pressures are delivered at physiologic rates.
NPV	Noninvasive. Chest shell generates subatmospheric pressure with NPV. It does not provide supplemental oxygen.

Abbreviations: BMV, bag-mask ventilation; F_{IO_2}, fraction of inspired oxygen; IMV, invasive mechanical ventilation; NIV, noninvasive ventilation; NPV, negative-pressure ventilation.

flow out of the lungs. Similarly to NIV, pressure delivery during NPV may be continuous or biphasic. The most common modality used with NPV is continuous negative pressure. NPV might improve cardiac output by lowering intrathoracic pressure, thus increasing right ventricular (RV) preload and decreasing RV afterload. NPV cannot provide supplemental oxygen directly, so children with hypoxemia often need additional oxygen delivery sources. In their practice, pediatricians may encounter patients receiving NPV, as it is increasingly used in children with neuromuscular disorders such as spinal muscular atrophy.

Conventional Ventilation and High-Frequency Ventilation

Conventional ventilators deliver a preset tidal volume or inspiratory pressure and a baseline positive end-expiratory pressure (PEEP) at a rate in the physiologic range for age. Inspiratory time is typically set shorter than the expiratory time to avoid hyperinflation while providing sufficient inspiratory time to ensure proper lung inflation.

High-frequency oscillation ventilation (HFOV) delivers small tidal volumes (1–3 mL/kg) at a supraphysiologic frequency of 3 to 15 Hz (1 Hz = 60 oscillations per minute), which translates to rates of 180 to 900 breaths per minute. High mean airway pressures help with alveolar recruitment and adequate oxygenation. Ventilation is maintained by diffusion, turbulence, and movement of air between lung regions (pendelluft effect). HFOV is typically used in a hospital setting when conventional ventilation fails to maintain adequate oxygenation in patients with pediatric acute respiratory distress syndrome, or bronchopleural fistulas, or in neonates with meconium aspiration syndrome or persistent pulmonary hypertension.[4]

BASICS OF A VENTILATOR

A typical mechanical ventilator consists of a control panel (for user interface), a power source, a gas supply system, a central processing unit (the brain of the ventilator), a breathing circuit with a humidification system, and a patient interface (endotracheal or tracheostomy tube for invasive ventilation and various masks and cannulas for NIV) (Figure 3).

Hospital ventilators are typically larger, more powerful, and more expensive than home ventilators. Hospital ventilators offer a wider range of adjustable parameters and monitoring capabilities to meet the patient's requirements. They typically require a high-pressure gas source at 50 psi.

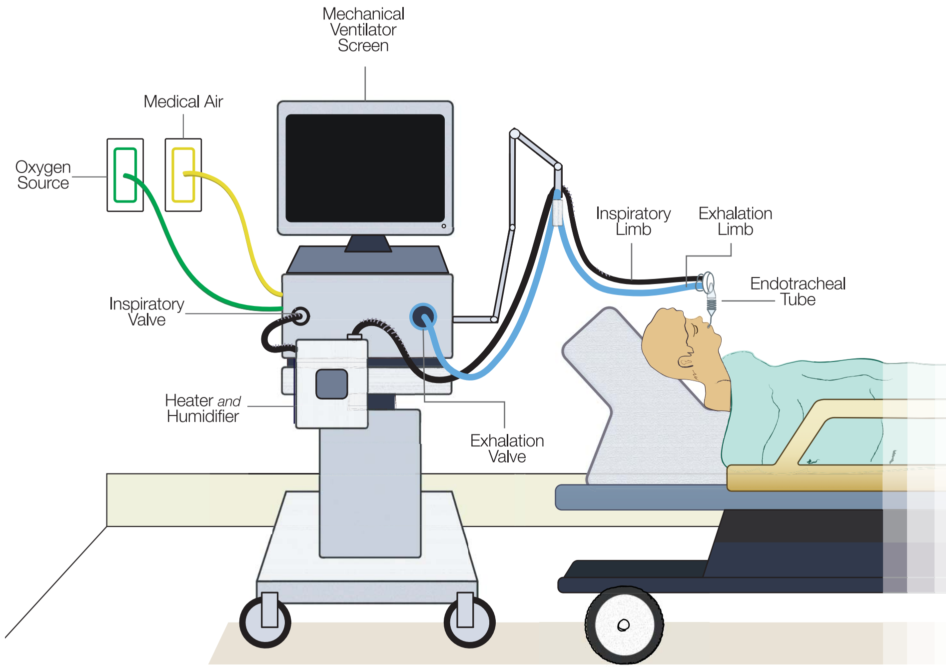

FIGURE 3. Anatomy of a ventilator. Black arrows: air flowing through a heater/humidifier, then to the patient. Blue arrows: exhaled air flowing away from the patient. (Created by the authors using Adobe Illustrator).

Home ventilators are designed to be more compact, lightweight, portable, and easier to use. The ventilator is connected to an oxygen source (usually a concentrator) and a humidifier. A high-pressure gas source is not required for home ventilators. Pediatric home ventilators must offer a more sensitive trigger, generate lower tidal volume and flow rate, and have adequate audible alarms. Home ventilators should be able to operate with an internal battery when there is no access to electricity. Unlike hospital ventilators, home ventilators have limitations, including the inability to provide high peak pressures or FiO_2 greater than 0.6. With improved technology, newer home ventilators have capabilities closer to hospital ventilators.

INVASIVE MECHANICAL VENTILATION

Basic Terminology

The most common terminology used for mechanical ventilation is listed in Table 3.

Modes of Mechanical Breaths

Various modes of mechanical breaths are described based on how inspiration is initiated, sustained (limited or controlled), and terminated. Individual brands of ventilators may deliver specific modes of mechanical breaths or use proprietary names for similar modes. In general, the following modes are available with most conventional mechanical ventilators (Figure 4).

1. Controlled mandatory ventilation: The ventilator delivers breaths at a fixed rate and set volume or pressure, regardless of the patient's respiratory efforts. The ventilator controls all phases of the breath. It is typically used in patients who are heavily sedated, chemically paralyzed, or unable to initiate their own breaths.
2. Synchronized intermittent mandatory ventilation (SIMV): The ventilator provides breaths at a fixed set rate, synchronizing with the patient's own inspiratory efforts. SIMV can be pressure-limited (preset pressure) or volume-limited (preset volume). The ventilator delivers controlled breaths if the patient's spontaneous respiratory rate is lower than the set SIMV rate. It also allows the patient to take spontaneous breaths between the synchronized mandatory ones, which can be supported with volume or pressure.
3. Assist-control (AC) ventilation: In AC ventilation, the patient can trigger their own breaths, and the ventilator will deliver a full ventilator breath, with the preset volume or pressure for each trigger, ensuring a minimum respiratory rate. Every triggered breath with AC is a full ventilator breath.
4. CPAP (CPAP/pressure support [PS]): CPAP mode provides continuous positive pressure, allowing the patient to breathe spontaneously. The patient entirely controls the breathing cycle. A PS or volume support can be added to CPAP for spontaneous breaths.

TABLE 3. Basic Terminology in Mechanical Ventilation

Ventilation Terminology	Description
F_{IO_2}	Fraction of oxygen in the inspired air; titrated down to keep acceptable oxygen saturation.
Rate	Breaths per minute; set based on the age and disease process.
Time	
i time	Time required for inspiration (and inspiratory pause).
e time	Time required for exhalation.
I:E ratio	The ratio between time for inspiration and exhalation during one breath; normally set between 1:1.5 and 1:3.
Peak pressures	
PIP	The highest level of pressure that is applied during inspiration.
Pplat	Pplat is the end-inspiratory pressure during a period of no gas flow. It reflects alveolar pressure at equilibrium.
PEEP	PEEP is the pressure maintained during exhalation to prevent alveolar collapse. It is set at 4–5 cm H_2O with normal lungs. PEEP is set at a higher level in restrictive lung diseases.
V_T	V_T is the volume of air inhaled or exhaled during a normal breath. Normally set at 5–8 mL/kg.
Support	Additional support that is provided by the ventilator for spontaneous breaths triggered by the patient.
Pressure (PS)	A predetermined pressure is provided with PS when the patient takes a spontaneous breath.
Volume (VS)	A predetermined tidal volume is provided with VS when a patient takes a spontaneous breath.
Trigger	A trigger is the mechanism by which the ventilator detects inspiratory effort and initiates a breath.
Pressure	Drop in airway pressure below a set threshold.
Flow	Change in continuous gas flow through the circuit.
Neural	Electrical activity in the diaphragm.
Inspiratory cycle-off	Cycle-off is the mechanism by which the ventilator terminates a breath (inspiration).
Volume	Volume cycle-off is when a set tidal volume has been delivered.
Pressure	Pressure cycle-off is when a set airway pressure has been achieved.
Time	Time cycle-off is when the set time for inspiration has been achieved (the most common cycling method).
Flow	Flow cycle-off is when inspiratory flow has declined to a set percentage of maximum flow, usually with supported breaths.
DP	DP is the pressure difference between Pplat and PEEP. It is typically kept below 15 cm H_2O to avoid barotrauma.

Abbreviations: DP, driving pressure; e time, exhalation time; F_{IO_2}, fraction of inspired oxygen; I:E ratio, inspiration to exhalation ratio; i time, inspiratory time; PEEP, positive end-expiratory pressure; PIP, peak inspiratory pressure; Pplat, plateau pressure; PS, pressure support; VS, volume support; V_T, tidal volume.

Control Variable

A control variable is the primary parameter the ventilator adjusts to achieve during inspiration. A limit (target) variable sets the maximum value the variable can attain during the inspiratory phase. The limit variable does not end the inspiration. A cycle variable ends the inspiration once the set variable value is achieved. A breath can be delivered by controlling the volume, pressure, time, or flow, with volume and pressure being the most used control variables.

1. Volume control: A set tidal volume is delivered, while the airway pressure generated will vary depending on the airway resistance and respiratory compliance. This mode might be used when a tight Pa_{CO_2} control is required and lung compliance is normal.
2. Pressure control: The peak pressure is set to minimize the risk of barotrauma, while the delivered tidal volume varies depending on respiratory compliance and airway resistance.

Graphics and Scalars

Ventilator graphics are useful for monitoring minute ventilation, respiratory mechanics, and improvement in respiratory function (Figures 5 and 6).

MECHANICAL VENTILATION IN SPECIFIC SETTINGS

Table 4 shows the starting settings for mechanical ventilation in the inpatient setting. In this section are some considerations for mechanical ventilation strategies in specific pathologies.

Obstructive Disease

Asthma and bronchiolitis are considered obstructive diseases due to increased airway resistance. Effective ventilation of these patients requires minimizing the risk of air trapping and hyperinflation. This can be accomplished by setting a low respiratory rate and a short inspiratory time, allowing sufficient time for complete exhalation.

Breath type	Trigger variable (breath initiation: inspiration)	Cycle variable (breath termination: exhalation)	Limit/Target Variable (maximum parameter value)
Controlled breath (CMV)	Ventilator set (time)	Ventilator set (time, flow, pressure)	Ventilator set (pressure or flow/volume)
Assisted breath (SIMV or AC)	Patient effort (flow or pressure)	Ventilator set (time, flow, pressure)	Ventilator set (pressure or flow/volume)
Supported breath (PS or VS)	Patient effort (flow or pressure)	Patient set (flow)	Ventilator set (pressure or flow/volume)
Spontaneous breath (CPAP)	Patient effort (flow or pressure)	Patient set	Patient set

FIGURE 4. The mode of ventilation dictates the way a breath will be delivered. In CMV, the ventilator strictly controls all the variables. In CPAP modes, the patient controls all the variables. Modes like SIMV, AC, or PS use a combination of both patient's effort and ventilator setting.
Abbreviations: AC, assist-control; CMV, controlled mandatory ventilation; CPAP, continuous positive airway pressure; PS, pressure support; SIMV, synchronized intermittent mechanical ventilation; VS, volume support. (Created by the authors using Adobe Illustrator).

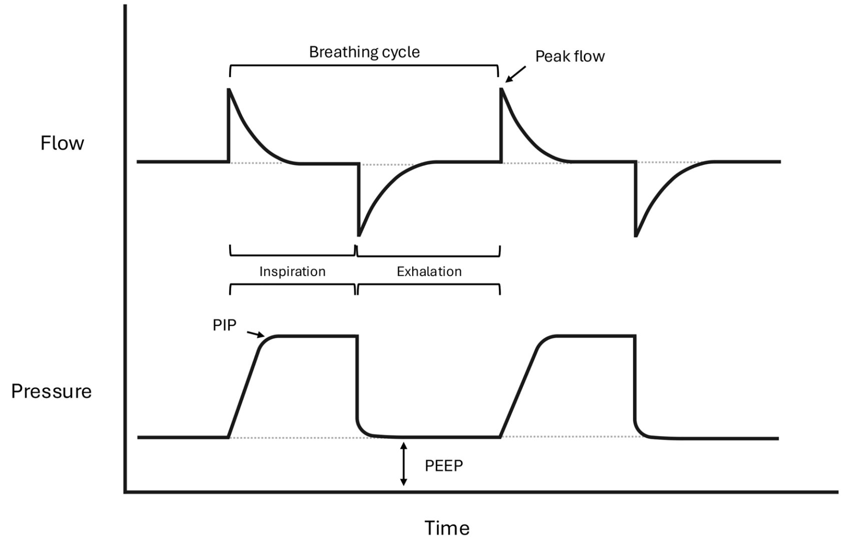

FIGURE 5. Flow and pressure waveforms in a controlled breath.
Abbreviations: PEEP, positive end-expiratory pressure; PIP, peak inspiratory pressure.

Neurologic Disorders

Patients with central nervous system dysfunction may have respiratory failure from loss of airway protection reflexes, impaired respiratory drive, diminished upper airway tone, or a combination thereof. In children with increased intracranial pressure, strict CO_2 control is essential with guaranteed minute ventilation. Additionally, avoiding lung hyperinflation is important, as elevated intrathoracic pressures can compromise venous return from the brain.

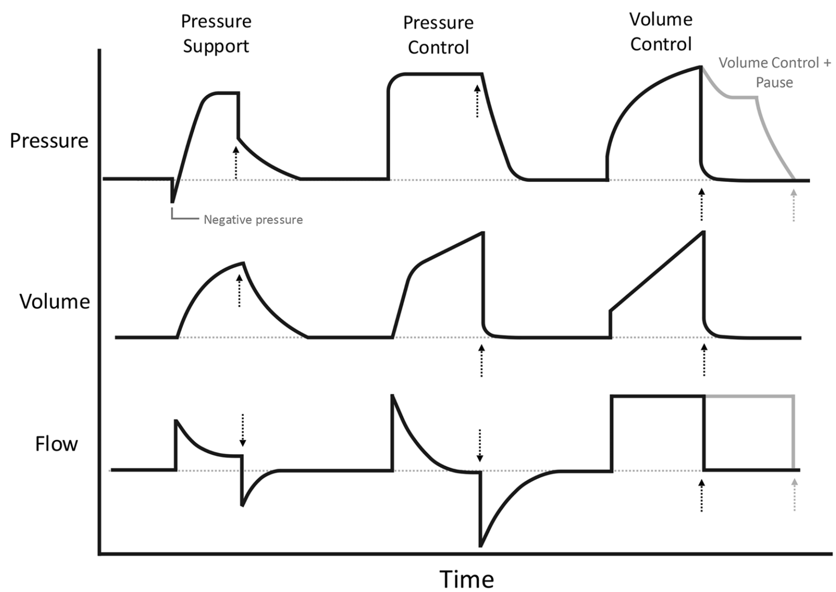

FIGURE 6. Pressure, volume, and flow curves in different ventilator modes. The negative deflection on the pressure-supported breath triggers the initiation of a breath. The arrows show the transition from inspiration to exhalation.

TABLE 4. Starting Settings for Conventional Ventilators

VT	5–8 mL per kilogram of IBW 3–6 mL/kg IBW if decreased lung compliance	Directly set on volume control mode. Set VT to maintain Pplat within range.
Pplat[a]	≤28 cm H_2O ≤32 cm H_2O with decreased chest wall compliance	Directly set on pressure control mode. Set PIP to maintain adequate VT. Adjust VT to keep Pplat below the target levels.
PEEP	5–8 cm H_2O; higher in patients with PARDS	The goal is to achieve optimal lung expansion and maintain adequate Sao_2/Pao_2 without harmful levels of Fio_2.
RR	Infants: 25–30 breaths/min 1–6 y old: 20–25 breaths/min >6 y old: 12–20 breaths/min	Adjusted to maintain $Paco_2$. A higher rate for restrictive diseases and a lower rate for obstructive diseases.
i time[b]	Infant: 0.4–0.7 s Older children: 0.7–1.2 s	Maintain I:E ratio as physiological as possible, typically 1:2 to 1:3.
Fio_2[c]	0.21–1	Maintain adequate Sao_2/Pao_2. Target Sao_2: - PARDS: 88%–92% - Biventricular physiology: 92%–96% - Single-ventricle lesions: 75%–85%, to prevent pulmonary over circulation

Abbreviations: i time, inspiratory time; IBW, ideal body weight; PARDS, pediatric acute respiratory distress syndrome; PEEP, positive end-expiratory pressure; PIP, peak inspiratory pressure; Pplat, plateau pressure; RR, respiratory rate; Sao_2, saturation of arterial oxygen; I:E ratio, inspiration to exhalation ratio; VT, tidal volume.

[a]Pplat is measured during an inspiratory hold maneuver, where there is no gas flow, and airway and alveolar pressures equilibrate.

[b]i time should be selected to allow all lung units to inflate during inspiration and to fully empty during exhalation.

[c]High Fio_2 could lead to lung injury by generating free oxygen radicals; Fio_2 <0.5 is considered safe.

Muscular Disorders

In patients with muscular disorders, muscle weakness leads to hypoventilation. When providing mechanical ventilation to these patients, encouraging spontaneous breathing is imperative to prevent muscle deconditioning and improve their chances of successful ventilator weaning. If oxygenation is not a concern, PEEP and Fio_2 should be maintained at minimal levels.

HOME VENTILATOR SUPPORT

Over the past 20 to 30 years, advances in the medical field have contributed to a growing number of children using home mechanical ventilation (HMV). A Canadian epidemiological study reported a 37% increase in the number of patients discharged home with a ventilator over 13 years.[5]

HMV aims to ensure adequate ventilation and oxygenation using stable ventilator settings without the need for

TABLE 5. Indications for Home Mechanical Ventilation[6]

Neuromuscular Weakness	Neurologic Control of Ventilation	Increased Respiratory Load
Muscular dystrophies Spinal muscular atrophy Diaphragmatic dysfunction Metabolic myopathies Spinal cord transection at high level	Congenital central hypoventilation syndrome Acquired central hypoventilation Degenerative central nervous system diseases Syringomyelia	Obstructive sleep apnea Laryngomalacia/tracheomalacia Vocal cord paralysis Chronic aspiration and bronchiectasis Scoliosis and chest wall deformities Pulmonary hypoplasia Bronchopulmonary dysplasia

Adapted from Kwak.[6]

continuous monitoring while maximizing growth and development. Ventilator mode and settings are initially established and adjusted in the inpatient setting, with close monitoring and titration to meet each patient's needs.

Indications for HMV

HMV should be considered when a patient develops progressive chronic respiratory failure or failure to wean from mechanical ventilation. Patients with chronic respiratory failure due to neuromuscular disorders, impaired neurologic control of breathing, or increased respiratory load from airway and lung pathology might benefit from HMV (Table 5).

Types of Ventilators for Home Support

The choice of ventilation type and mode is determined by the cause and severity of the respiratory failure (Table 6).

TABLE 6. Conditions That Benefit From Home Noninvasive vs Invasive Ventilation

Noninvasive Ventilation	Invasive
Obstructive sleep apnea Chronic alveolar hypoventilation Central respiratory regulation disorder Muscular dystrophies[a]	Upper airway obstruction Vocal cord paralysis Subglottic stenosis Bulbar palsy Pulmonary hypoplasia Bronchopulmonary dysplasia Patients requiring MV >16 h/d

Abbreviation: MV, mechanical ventilation.
[a]Some patients with muscular dystrophy, depending on the severity, might benefit from invasive MV.

Noninvasive. NIV can be delivered through nasal, oronasal, or full-face masks. The key to successful NIV is selecting a well-fitting mask to minimize leakage and maximize comfort. Most ventilators compensate for leaks by delivering a higher flow to reach the set pressure. Depending on the patient's condition, CPAP or biphasic mode might be required.

Some of the disadvantages of NIV are the risk of gastric content aspiration, especially in younger patients who cannot remove the mask; skin breakdown; and difficulty in clearing airway secretions. If a patient fails NIV, IMV via tracheostomy is the alternative for long-term home ventilatory support.

Invasive. Invasive HMV is delivered via tracheostomy. The stoma should be healed adequately before initiating HMV and considering discharge from the hospital. The gas delivered should be warmed and humidified to prevent the drying of the airway mucosa. Most ventilators have an active humidification device attached.

Most home ventilators can provide volume- or pressure-controlled ventilation. Volume-control ventilation delivers a set tidal volume, while pressure-control ventilation delivers a set positive pressure. The benefit of pressure-controlled ventilation is that it can adapt to the patient's irregular breathing patterns and compensate for some leaks. Some of the current home ventilators can deliver more advanced modes of ventilation.

Single-limb circuits are often used in home mechanical ventilators because they are simpler and less bulky. These circuits cannot measure tidal volumes, so they are estimated using algorithmic calculations. Some single-limb circuits have a leak port to compensate for leaks and allow some CO_2 removal. Other circuits have an exhalation valve to allow complete CO_2 elimination. Double-limb circuits have an inhalation limb and an exhalation limb connected to inhalation and exhalation ports, respectively. Accurate measurement of tidal volumes is possible with double-limb circuits.

Negative-pressure Ventilator. NPV may be used in children with neuromuscular disease for long-term support. NPV is increasingly used for managing children with respiratory failure and spinal muscular atrophy.

Management

Children who require HMV, in general, have multiple underlying comorbidities; hence, they should be cared for by a multidisciplinary team that includes a primary care physician, pulmonologist, and respiratory therapist, along with other specialists depending on the patient's comorbidities. Most children requiring home ventilation need continuous nursing and respiratory therapy care. Parents and caregivers should be trained to recognize and troubleshoot ventilator malfunction, tracheostomy dislodgement, and acute hypoxemia. The ventilator should have appropriately set alarms to alert caregivers of possible problems.

Airway clearance techniques should be used routinely to prevent mucous plugging and subsequent atelectasis and to minimize the risk of respiratory infections. These techniques range from nebulization with mucolytics to the use of chest physiotherapy and cough assist devices.

It is recommended that children be monitored with pulse oximeters, especially while asleep, alongside ventilator alarms.[7] The equipment necessary at home should include batteries, suctioning equipment, supplemental oxygen, a nebulizer, a self-inflating bag and mask, and extra tracheostomy tubes.

WEANING FROM IMV

Weaning is the gradual process of shifting the responsibility for breathing from the ventilator back to the patient as the condition requiring mechanical ventilation improves. Weaning is typically guided by the patient's respiratory effort, adequacy of ventilation and oxygenation, and decreased dependence on mechanical ventilator support. Children undergoing HMV require regular monitoring by pulmonologists to determine whether weaning off the ventilatory support is appropriate or feasible.

COMPLICATIONS OF HOME MECHANICAL VENTILATION

- The plugging of airways with thick mucus is most often seen in younger children and smaller-diameter tracheostomy cannulas. Infants have not yet developed alveolar collateral ventilation (pores of Kohn or channels of Lambert), so mucous plugging in smaller airways more frequently can lead to atelectasis. Humidification devices prevent the drying of secretions and decrease the risk of mucous plugging. Some patients might also need nebulized mucolytics.
- Accidental removal of the tracheostomy cannula happens more frequently in infants or when short tubing pulls on the cannula. Parents and caregivers should be trained before hospital discharge to manage the dislodgement of tracheostomy tubes and to perform cardiopulmonary resuscitation in case of an emergency. A spare cannula of the same and smaller size should always be available for emergent tube exchange.
- Mask leakage can occur when masks or headgear are not fitted properly. Even though a small leak is expected, larger leaks might compromise ventilation effectiveness. This may result in hypoxemia or hypercapnia and frequent nighttime awakenings, particularly in patients who require mechanical ventilation for obstructive sleep apnea.
- Midfacial deformity can occur, most commonly when masks used for NIV are improperly fitted. The high pressures exerted by the mask may interfere with normal facial bone growth. Younger children should be monitored regularly to ensure the mask fits properly, preventing skin breakdown and minimizing the risk of facial bone hypoplasia.
- Ventilator malfunctions can occur due to power failure, equipment defects, disconnection, or changes in the patient's condition. Caregivers should be trained to recognize and troubleshoot the most common causes of ventilator malfunction, such as circuit disconnection, lack of power supply, or poor battery backup. Caregivers should also be appropriately trained to provide manual ventilation until help arrives. Families should have functioning supplies, such as suction machines, appropriately sized catheters, resuscitation bags, and nebulization machines if needed.
- Children with artificial airways are at higher risk for developing infections, such as tracheitis or ventilator-associated pneumonia. Children with artificial airways are often colonized with hospital-acquired organisms, like *Pseudomonas*, *Klebsiella*, and methicillin-resistant *Staphylococcus aureus*. If there are changes in tracheal secretions, fever or hypothermia, or increased oxygen or ventilatory needs, infection should be suspected, and the patient should be evaluated accordingly (Table 7).

TABLE 7. Complications of Home Mechanical Ventilation

Ventilator Type	Complications
All ventilators	Ventilator malfunction Feeding problems Mucous plugging Hypoxemia Cardiac arrest
Noninvasive ventilators	Mask leakage Skin breakdown Facial bone hypoplasia Gastric content aspiration
Invasive ventilators	Accidental tracheostomy decannulation Stomal bleeding Stomal/tracheal granuloma Upper or lower airway infection

Summary

- When indicated, HMV should be initiated as part of a comprehensive and collaborative plan of care that includes the family members, primary caregivers, pulmonologists, and primary care physician.[8] (Strong recommendation, low quality of evidence)
- Children undergoing HMV should be monitored with noninvasive methods, such as pulse oximetry, especially when asleep.[9] (Based on expert consensus due to lack of relevant studies)
- At least two trained family caregivers who can troubleshoot common causes of ventilator malfunction should be caring for children with chronic HMV.[9] (Based on expert consensus due to lack of relevant studies)

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Scan this QR code to access references and view and save images (available March 1, 2026).