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Cuirass Ventilation: An Alternative Home-Based Modality for Chronic Respiratory Failure

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Abstract

The biphasic cuirass ventilation (BCV) device is an alternative respiratory support device for patients with chronic respiratory failure. Considered by some a "forgotten" mode of supportive ventilation, the device is portable, lightweight, and easy to operate. Biphasic cuirass ventilation can also be used to rapidly resuscitate patients in acute respiratory distress and requires minimal technical skill to operate. Biphasic cuirass ventilation can be employed by the patient's caregiver in the home setting, making it a viable alternative to other forms of mechanical ventilation (e.g., BiPAP) for patients enrolled in home hospice or palliative care. The article reviews current knowledge and aims to enhance awareness and encourage further study about cuirass ventilation, particularly with regard to its use in treating patients in the palliative care setting and in the home.

Keywords

biphasic cuirass ventilation, mechanical ventilation, neuromuscular disease, critical care medicine, palliative medicine

Introduction

A cuirass is a shell-like unit that creates negative pressure on the chest using a combination of a fitted casing and a soft bladder. It is an adaptation of the sensationalized "iron lung" albeit compacted into a much smaller device. The iron lung was first used in 1928 and was one of the first negative-pressure machines used for long-term ventilation.[1] In the original model, the intrathoracic vacuum caused simultaneous expansion of both the chest and abdominal cavities, resulting in pooling of venous blood in the lower abdomen. However, it was refined and ultimately revolutionized the response to the 1940s polio epidemic. It works via a vacuum pump mechanism, which creates a negative pressure causing the chest wall to expand. This in turn causes a decrease in intrapulmonary pressure and ambient air to flow into the lungs. This process mimics the natural negative pressure in the intrathoracic space.[1]

Similarly, biphasic cuirass ventilation (BCV) offers mechanical support for patients suffering from respiratory failure. Although BCV is not a novel ventilatory device, its applications in current palliative care may expand caregivers' capability to provide airway management—particularly in patients with neuromuscular disorders (NMDs; for example, amyotrophic lateral sclerosis [ALS]) or other conditions where chronic respiratory insufficiency exists. This mode of respiratory support offers portability and comfort and, since the patient's face is not covered, conversation, eating, and even coughing can proceed unhindered.[2]

The purpose of this article is to review current knowledge, enhance awareness, and encourage further study of this useful but possibly underused modality.

Physiology and Design

The cuirass device has previously been used in negative pressure ventilation (NPV), external chest wall oscillation (ECWO), external chest wall compression (ECWC), and

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external high-frequency oscillation (EHFO).[3] The re-invented or modified cuirass negative pressure ventilator by Hayek has multiple seals and a high-pressure oscillation pump to facilitate respiration.[4] The biphasic function of the cuirass device affords users control over the inspiration:expiration ratio (I:E ratio).[3] The cuirass is a domed plastic shell that encases the patient's chest and upper abdomen and is connected to an external pump.[5] Cuirass ventilators are designed to apply equal pressure throughout the thorax, which allows for even expansion of the lungs and uniform ventilation.[6] The device generates a negative pressure within the chest during inspiration and can be programmed to perform continuous inspiratory assistance. It subsequently provides positive pressure around the thorax in order to induce expiration. This latter function makes it efficient at clearing carbon dioxide (CO_2), weaning patients from dependence on external ventilatory support, increasing cardiac output, and re-expanding areas of collapsed lung.

Modes of Cuirass Ventilation

Cuirass ventilatory devices can control both inspiration and expiration to varying degrees based on the mode in use.[7] There are five types of cuirass modes: continuous negative pressure, controlled biphasic (also known as intermittent mandatory ventilation, IMV), non-invasive synchronized BCV, high-frequency oscillation, and cardiac synchronization.[2]

Non-invasive synchronized BCV is similar to IMV except that patients initiate spontaneous breaths at their own rate and their initial inspiratory and expiratory efforts and then trigger the device to provide support that is timed with the patient's own respiratory pattern. The device is also equipped with a high-frequency oscillation mode (up to 1,200 oscillations per minute) that can assist with clearing secretions, increasing oxygenation, and reducing carbon dioxide—capabilities that essentially replace conventional chest physiotherapy. Finally, cardiac synchronized mode uses electrocardiography (ECG) to coordinate the cardiac cycle with the ventilatory cycle, also known as ECG-triggered BCV.[2]

Uses of Cuirass Ventilation

This easy-to-operate ventilation device can be used for rapid resuscitation of patients in acute respiratory distress, making it a useful tool in emergencies or combat situations. The time it takes to apply the device and then to restore adequate ventilation to the recipient is comparable to traditional intubation and positive pressure ventilation (PPV).[8] Operating the device requires minimal experience or technical skill allowing first responders and non-medically trained bystanders to employ BCV outside the hospital setting.[9] Moreover, its portable, lightweight construction can be run on rechargeable batteries, allowing for quick, easy transport into challenging situations.

One might argue that cuirass ventilation could be used in an effort to decrease the complications of invasive PPV.[10] Weaning patients from invasive PPV not only improves their quality of life but also reduces the risk of ventilator-associated pneumonia, laryngeal injury, and respiratory muscle atrophy—all of which are associated with prolonged mechanical ventilation.[2] As medical advancements prolong the survival of individuals with hitherto terminal conditions, the patient population requiring chronic ventilation will increase; having options other than tracheostomy and BiPAP would therefore be beneficial.

Cuirass Ventilation Use in Palliative Medicine

Patients in respiratory failure due to muscle weakness may use BCV as a means to postpone the need for invasive procedures such as intubation or tracheostomy. Such an intervention may enable them to communicate and assist with informed, autonomous decisions regarding their own health care trajectory.[11] Somewhat surprisingly, cuirass ventilation applications in palliative care are relatively new. However, the minimal technical skill required to operate these device makes it an ideal addition to home hospice or home-based palliative care for properly selected patients. Its non-invasive nature allows patients to continue to talk, eat, and interact with family members without the interference of face masks or tubes. Equally important is that one can quickly choose to discontinue therapy if it proves ineffective or is no longer desired, unlike chronic PPV via tracheostomy.[12]

Neuromuscular Disorders

Neuromuscular disorders (NMDs) have significant variation in their pathogenicity, clinical expression, and temporal course.[13] Most NMDs are genetic and therefore more prevalent in the descendants of specific geographic regions. However, an epidemiological review study published in 2015 showed that, in comparison to 1991, the prevalence of NMDs such as Becker muscular dystrophy, facioscapulohumeral dystrophy, myotonic dystrophy, and Charcot-Marie-Tooth disease has increased.[14]

The increased prevalence of these disorders, along with increased survival due to improvements in medical care, will likely trigger an increasing demand for minimally invasive ventilatory support.[14] Common symptoms in NMD include muscle weakness, movement limitations, difficulty swallowing, and respiratory distress. As NMDs progress, the diaphragm is often impacted, affecting the ability of patients to breathe adequately. Jackson et al[15] determined long-term nocturnal cuirass ventilation benefited those with neuromuscular and skeletal chest wall disorders. Notably, Hino et al suggested BCV as a useful alternative to non-invasive positive pressure ventilation (NIPPV)—PPV delivered through a nasal mask, face mask, or nasal plugs. Non-invasive positive pressure ventilation is still comparatively intrusive and, in this case, BCV proved advantageous in resolving a persistent air leak in a patient who was a high-risk surgical candidate and had developed tension pneumothorax while on NIPPV.[16]

Amyotrophic Lateral Sclerosis

Amyotrophic lateral sclerosis (ALS) is an incurable NMD affecting both the upper and motor neurons, causing progressive muscular weakness and contracture.[17] All patients with ALS eventually develop symptoms of respiratory insufficiency—the most common cause of death in patients suffering from this condition.[18] Nevertheless, research has shown that as many as 53% of patients with ALS make no decisions regarding ventilation prior to the onset of respiratory failure.[19] Mechanical ventilation is known to be an effective means of relieving symptoms of chronic hypoventilation and prolonging life in patients with ALS who have developed respiratory depression.[19]

Various methods are available to patients with ALS depending on their location during an acute respiratory distress episode, their personal preferences, and the presence of upper airway obstruction or heavy bronchial secretions. Because ALS is progressive and incurable, palliative therapy is typically the focus of treatment. For instance, chronic nocturnal hypoventilation is a significant source of symptom burden in this population and can long precede the onset of full-scale respiratory failure. Portable, battery-operated devices such as the cuirass system offer a relatively simple solution and may be comfortably worn by patients for extended periods of time. Beyond this, the application of BCV in the treatment of this illness has been shown to prolong survival in addition to improving the quality of life for subsets of ALS patients with normal or only moderately impaired bulbar function.[20]

Cystic Fibrosis

Cystic fibrosis is a common inherited disease in Caucasian populations, with an incidence of 1 in 2,500 newborns.[21] It is an autosomal recessive disorder caused by mutations in the cystic fibrosis transmembrane conductance regulator gene (*CFTR*).[22] The primary function of the CFTR protein is to regulate liquid volume on epithelial surfaces through chloride secretion and inhibition of sodium absorption via ion channels.[21] This impaired mechanism most commonly presents as serious mucociliary dysfunction, which prohibits cystic fibrosis patients from effectively clearing inhaled particles and mucus and thereby predisposes them to recurrent infections. With the cuirass ventilator's "secretion clearance" mode, the device uses vibration to mobilize and thin out the mucus plugging up the respiratory system of patients with cystic fibrosis. It then employs its assisted cough function to help expel the loosened secretions, thereby decreasing patient susceptibility to recurrent infections.[23] With the increase in life expectancy in cystic fibrosis into the fifth decade for at least 50% of affected individuals, cuirass ventilation may be a valuable mode of respiratory support near end of life.[3]

Other Uses

Cuirass ventilation has been reported as an alternative method of ventilation in at least two cases of central alveolar hypoventilation (CAH) syndrome, a rare disorder of respiratory and autonomic system dysregulation.[2,24] Patients with CAH are generally ventilated at night by invasive PPV via tracheostomy. However, tracheostomies can cause recurrent aspiration which fosters pulmonary infections. In these two cases, bronchoscopy was first used to rule out upper airway obstruction prior to BCV conversion as tracheomalacia may develop following prolonged periods of intubation. After a period of overnight SaO_2 and end-tidal CO_2 monitoring, both patients successfully transitioned to BCV and were ultimately able to close their tracheostomies.[2]

Another case report demonstrated successful use of cuirass ventilation in treatment of a patient with Pompe disease who had developed mediastinal and subcutaneous emphysema as a result of NIPPV.[25] Further reports still have shown BCV to be especially useful in managing patients with exacerbation of their chronic obstructive pulmonary disease (COPD) complicated by hypercarbic respiratory failure.[26]

Finally, cuirass ventilation may also act as a stand-in for endotracheal intubation in patients with decompensated heart failure. Whereas PPV decreases venous return, cuirass ventilation reduces intrathoracic pressure, allowing for increased venous return to the heart and subsequent increase in cardiac output.[27] Specifically, Shekerdemian et al[28] demonstrated that cuirass NPV had hemodynamic advantages over conventional PPV following cardiac surgery in pediatric patients, as demonstrated by a notable improvement in cardiac output.

Complications and Risk

As outlined above, PPV is an invasive approach that incurs many risks of infection and injury. Cuirass ventilation is a comparatively safer option, although not entirely risk-free. A common complication of cuirass ventilation, regardless of whether or not the device has biphasic capabilities, is skin irritation and abrasions that can become niduses for infection. The cuirass ventilator wraps tightly around the chest, in close proximity to the skin. Older cuirass devices were especially prone to falling off patients and damaging skin, making them less than ideal for long-term use.[4] However, contemporary BCV models come in 12 sizes from neonate to adult, allowing for a better fit and for easy adjustment when one treats growing children.[2]

Implications for Home Care

Since the first negative pressure device was used in the 1920s, numerous revisions and advancements have equipped patients with a ventilator that is more comfortable, convenient, and easy to operate than traditional invasive respirators.1 In addition, these devices may decrease overall expense as they minimize the cost and possible complications of PPV.[10,27,29] Biphasic cuirass ventilation has demonstrated favorable outcomes in the long-term respiratory support of patients with ALS, cystic fibrosis, NMDs, and chronic respiratory failure from other etiologies.[15,18,23] Noninvasive ventilation techniques like cuirass ventilation improve the quality of life of palliative patients by allowing for continued mobility and communication, while affording them the option of managing their condition in the home.[30]

Conclusions

This review aims to raise awareness regarding cuirass ventilation and its applications in modern health care. Currently, these devices are infrequently used, and there are patients who may benefit BCV being incorporated into their established treatment strategies. Notably, current research regarding BCV is limited to review articles and case reports, thus making it difficult to standardize BCV use. Randomized controlled trials are necessary to evaluate fully the efficacy and risks of cuirass ventilators, as well as to broaden the scope of their application in palliative medicine in the home.

Declaration of Conflicting Interests

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