Review Article

Chronic respiratory failure in patients with neuromuscular diseases: diagnosis and treatment*

Ilma Aparecida Paschoal[1], Wander de Oliveira Villalba[2], Mônica Corso Pereira[3]

Abstract

Neuromuscular diseases affect alveolar air exchange and therefore cause chronic respiratory failure. The onset of respiratory failure can be acute, as in traumas, or progressive (slow or rapid), as in amyotrophic lateral sclerosis, muscular dystrophies, diseases of the myoneural junction, etc. Respiratory muscle impairment also affects cough efficiency and, according to the current knowledge regarding the type of treatment available in Brazil to these patients, it can be said that the high rates of morbidity and mortality in these individuals are more often related to the fact that they cough inefficiently rather than to the fact that they ventilate poorly. In this review, with the objective of presenting the options of devices available to support and substitute for natural ventilation in patients with neuromuscular diseases, we have compiled a brief history of the evolution of orthoses and prostheses used to aid respiration since the end of the 19th century. In addition, we highlight the elements that are fundamental to the diagnosis of alveolar hypoventilation and of failure of the protective cough mechanism: taking of a clinical history; determination of peak cough flow; measurement of maximal inspiratory and expiratory pressures; spirometry in two positions (sitting and supine); pulse oximetry; capnography; and polysomnography. Furthermore, the threshold values available in the literature for the use of nocturnal ventilatory support and for the extension of this support through the daytime period are presented. Moreover, the maneuvers used to increase cough efficiency, as well as the proper timing of their introduction, are discussed.

Keywords: Respiratory insufficiency/diagnosis; Chronic disease; Respiratory insufficiency/therapy; Neuromuscular diseases; Respiration; Artificial.

*Study carried out in the Pulmonology Division of the Clinical Medicine Department of the Universidade Estadual de Campinas (UNICAMP, State University at Campinas) School of Medical Sciences – Campinas (SP), Brazil.

1. Associate Professor in the Pulmonology Department of the Universidade Estadual de Campinas (UNICAMP, State University at Campinas) School of Medical Sciences – Campinas (SP), Brazil

2. Physical Therapist in the Physical and Occupational Therapy Department of the Universidade Estadual de Campinas (UNICAMP, State University at Campinas) Hospital de Clínicas – Campinas (SP), Brazil

3. PhD Professor in the Pulmonology Department of the Pontifícia Universidade Católica (PUC, Pontifical Catholic University) at Campinas School of Medicine – Campinas (SP), Brazil

Correspondence to: Ilma Aparecida Paschoal. Rua Serra da Bocaina, 178, CEP 13100-449, Campinas, SP, Brazil. E-mail: ilma@mpc.com.br

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Introduction

Neuromuscular diseases lead to alveolar hypoventilation. When the onset is slow and progressive, alveolar hypoventilation typically goes undiagnosed and untreated until an episode of acute respiratory failure occurs. This episode of decompensation is frequently seen during common upper airway infections, and it results from patient incapacity to eliminate secretions.

Respiratory muscle strength impairment can occur in various neuromuscular diseases (Chart 1). In cases of cervical spine trauma, the loss of respiratory capacity is acute and irreversible. When muscular dystrophies, neurological disorders and myoneural junction diseases affect the respiratory muscles, they present an evolution of a different sort, presenting outbursts, or progressive worsening, at varying rates of speed.

The progressive form of respiratory muscle impairment produces blood gas alterations (hypoxemia and hypercapnia, both resulting from hypoventilation). Only when the lungs present lesions caused by repeated infections as a result of inefficacious coughing and episodes of aspiration due to uncoordinated swallowing, can hypoxemia also be explained as the result of ventilation-perfusion mismatching. The initial management of a neuromuscular disease patient with hypoxemia and hypercapnia does not necessarily involve the use of oxygen but must include techniques aimed at re-establishing appropriate alveolar air exchange. Indeed, using oxygen alone in situations of hypoventilation can lead to the death of the patient.

History: the evolution of mechanical ventilators

The use of devices that perform the functions of the respiratory system constitutes an important chapter in the history of medicine and one about which physicians in general, and even pulmonologists, have little knowledge.

The interaction of the elastic forces of the chest cavity and the lung produces a subatmospheric pressure between these two structures and transforms the chest into a very peculiar organic cavity, different from the cranial cavity and abdominal cavity, which do not present this pressure-related peculiarity.

One persistent obstacle to opening the chest cavity for therapeutic purposes was that of the lower intrathoracic pressure, which made it impossible to establish communication between the interior and exterior of the cavity without compromising the lungs and thus causing their collapse.

Chart 1 - Neuro-skeletal muscle conditions that can lead to chronic hypoventilation.

Myopathies		
	Muscular dystrophies	
		Duchenne and Becker dystrophies
		Other muscular dystrophies, such as limb-girdle, Emery-Dreifuss, facioscapulohumeral, congenital, autosomal recessive, myotonic dystrophy
	Non-Duchenne myopathies	
		Metabolic or congenital myopathies
		Inflammatory myopathies (polymyositis, associated with connective tissue diseases or other systemic diseases)
		Diseases of the myoneural junction, such as myasthenia gravis
		Myopathies associated with traumas or medications
Neurological diseases		
	Spinal muscular atrophy	Neuropathies (hereditary, acquired, Guillain-Barré syndrome)
	ELA-type motor neuron diseases	Myelopathies
	Poliomyelitis	Supraspinal tonus disorders
	Multiple sclerosis	

It took a long time to reach the understanding of the very fact that the pressures are different. One of the pioneers of the physiology of ventilation, the German surgeon Sauerbruch,[1] only began to unravel this mystery at the end of the 19th century. In his attempts to open the chest, he concluded that the pressure in the interior of the chest cavity was lower than the atmospheric pressure and, in addition, that this fact was fundamental for respiration.

In order to circumvent this difficulty created by the pressure difference, Sauerbruch[1] developed a chamber for chest surgery (Figure 1). In this chamber that he created, the patient about to undergo surgery was positioned with the head outside the chamber, under the care of the anesthesiologist. The surgical team and the remaining part of the body of the patient stayed inside the chamber, where the pressure produced by air suction pumps was lower than the atmospheric pressure by approximately 7 mmHg. Communication between the interior and exterior sides of the chamber was sealed around the neck of the patient by a sort of adjustable mobile sealer clip. The head outside the chamber and the subatmospheric pressure around the chest permitted the opening of the chest without collapse of the lungs, maintaining spontaneous ventilation.

The first device designed as a substitute for spontaneous ventilation in individuals who, for some reason, no longer present adequate recirculation of air in the alveolar spaces, came to be widely used in the USA beginning in the 1930s. Built by an engineer in the Hospital of Harvard University,[2] it was dubbed the iron lung. At that time, there was a poliomyelitis pandemic, in which many people died from the paralytic respiratory form of the disease, due to the fact that there was no mechanism available that could be used as a substitute for ventilation. The need for a machine that could ventilate patients was imperative, and the knowledge of respiratory physiology available at that time led to the construction of this device (the iron lung), which was similar to the Sauerbruch chamber. It consisted of a steel cylinder, enclosing the body of the patient up to the neck, leaving only the head outside the chamber, and was powered by an electric motor that periodically generated subatmospheric pressure inside the cylinder, provoking the expansion of the chest cavity. This volume increase of the chest caused the intrathoracic pressure to drop and the air to be aspirated into the airways. Curiously, when the machine had to be temporarily

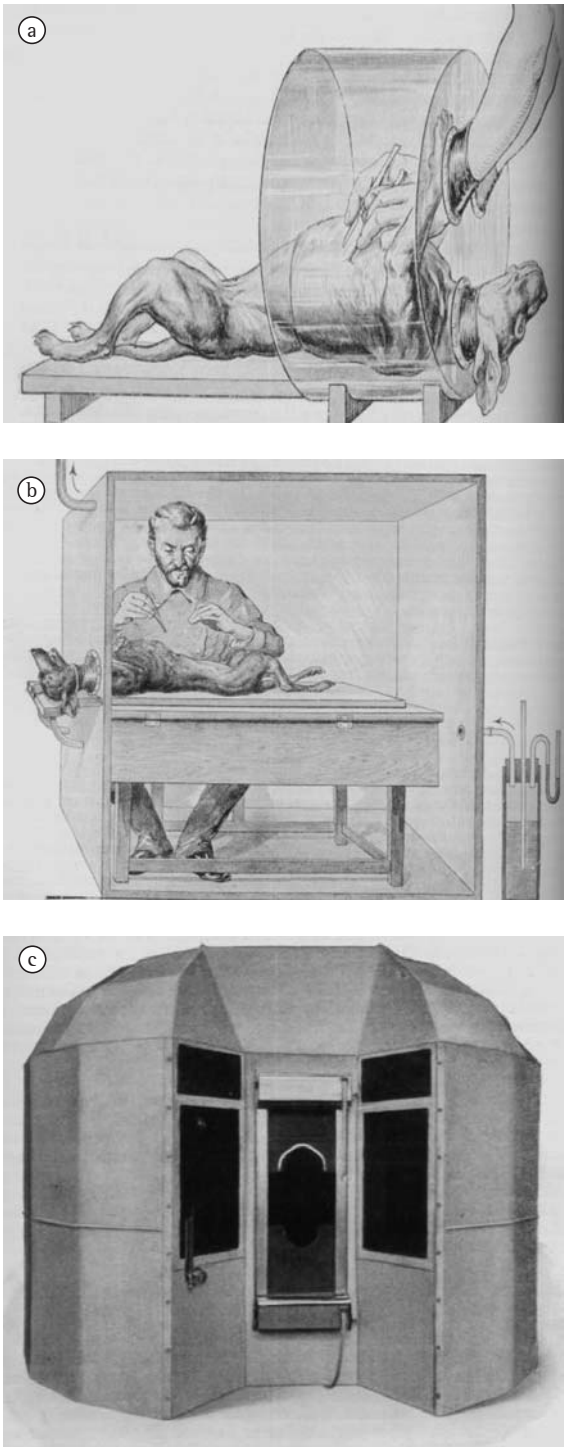

Figure 1 - Devices developed by Sauerbruch for performing thoracic surgery in a hypobaric chamber: a) Experimental negative pressure apparatus; b) Surgeon inside the hypobaric chamber; and c) Divider for the creation of the hypobaric chamber.

shut off, a transparent dome was placed over the head of the patient, within which positive pressure was produced. A similar positive pressure device was described by Sauerbruch in his book, published in 1922[1] (Figure 2), although the author frankly stated that he preferred the negative pressure chamber, his creation, for fear of the hazardous effects that positive pressure might have on the lungs.

The difficulty in providing general care, such as bathing, feeding and medicating patients in iron lungs can be foreseen. In addition, the forced immobility and the impossibility of coughing have certainly resulted in innumerable infectious pulmonary complications. Despite their shortcomings, there was a great demand for iron lungs, and their availability was limited in many hospitals.

In the summer of 1952, the poliomyelitis epidemic claimed many victims in Copenhagen, Denmark. Numerous patients presented respiratory difficulties, and the only ventilators available were a few negative pressure cuirass ventilators (iron lung-type ventilators that encircle only the chest), which are incapable of totally substituting the ventilation of the patients. The anesthesiologist Bjorn Ibsen was called upon to give his expert opinion regarding the treatment of the patients, and submitted one of them to a tracheostomy,[3] using a device known as an Ambu bag for ventilation. He proved that the invasive technique was more efficient in removing carbon dioxide than was the noninvasive technique. After the demonstration given by Ibsen, this form of ventilation became the standard treatment for the respiratory paralysis form of poliomyelitis in Denmark. Approximately 1500 Medicine and Dentistry students were summoned to take turns working six hour-shifts in the ventilation of patients with Ambu bags and logged approximately 165,000 h of service, saving the lives of many people through their considerable efforts.[3]

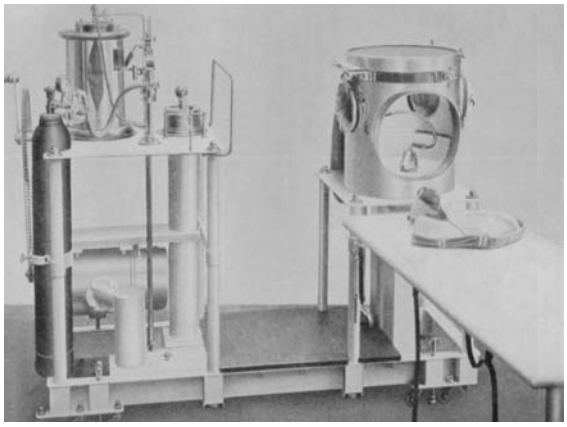

Figure 2 - Positive pressure apparatus, according to Sauerbruch.

When the poliomyelitis epidemic arrived in Sweden the following summer, the Swedes already had a mechanical ventilator that, similarly to Ambu bag, injected air under pressure into the airways, but did not require the helping hands of the volunteers. Therefore, positive pressure ventilators appeared, becoming the standard treatment in acute respiratory failure in the years to follow.

The criteria and parameters for the injection of pressurized gas into the airways have been a constant cause for concern since the 1960s. Ensuring a sufficient tidal volume for the patient, without damaging the lung due to the excessive pressure peaks, was one of the first aspects studied and inspired the development of volume control ventilators with pressure alarms. Inflating the lungs to a certain predetermined airway pressure proved insufficient to adequately ventilate the patient, especially in those with lung disease. Decreased pulmonary compliance caused cycling pressure to be reached in a very short time, insufficient for the influx of an appropriate tidal volume.

Another fundamental discovery in the area of mechanical ventilation was related to the need to maintain the alveoli open throughout the respiratory cycle, since there is a greater than normal tendency for the alveoli to collapse in some pathological conditions. Since 1969,[4] positive end-expiratory pressure has been considered a mandatory technique in artificial ventilation. Even for inspiration, it has been demonstrated that the maintenance of a pressure plateau, at the end of the gas injection, improves the distribution of the gas mixture to the millions of alveoli and favors hematosis.

Subsequently, attention was turned to the ventilation of individuals who have partially recovered from acute respiratory failure and need to be 'weaned' from mechanical ventilation. In this phase, the patients were required to be awake and cooperative, only being assisted by the machine during ventilation. Under such circumstances, it was fundamental that the process of gas injection be comfortable and that the ventilatory work could be gradually resumed by the patient. Within this context, a great advance was made in the 1980 s:

the development of the type of ventilation denominated pressure support ventilation.[5] This is a form of spontaneous ventilation that is flow-triggered, pressure-limited and flow-cycled.

The treatment of acute respiratory failure long monopolized the attention of many researchers, and few were concerned about patients with chronic respiratory failure.[6-8] The need for alternatives means of caring for such patients in environments other than intensive care units led to the development of machines with specific characteristics, to be used in places where a source of gas under pressure is not available. In 1978, the first portable ventilators, which were independent from pressurized gas networks, became available.[7] These ventilators can generate flow from room air, using compressors or turbines. In most cases, these ventilators have internal batteries or can be connected to an external, continuous power source (car battery, uninterrupted power supply, etc.).

At that time, quadriplegic patients with zero vital capacity were preferentially treated through tracheostomy and volume-cycled ventilators for home use. Tracheostomy permitted the satisfactory management of secretions, since the patient was unable to cough. In addition, it also prevented, for some time, the aspiration of the contents of the mouth or oropharynx in cases of concomitant swallowing disorders.

However, the tracheostomy maintained for years is associated with innumerable complications,[7] such as infections, increased quantity of secretion, impaired mucociliary clearance and bleeding, as well as sudden death resulting from a secretion plug or from the accidental disconnection of the device.

Other situations that lead to chronic respiratory failure, but are progressive, such as neuromuscular diseases and alterations of the chest wall, make it difficult to determine the appropriate moment at which to perform a tracheostomy, and patients would benefit considerably from an alternative method that would prevent or indefinitely postpone this intervention.

In 1976, noninvasive treatments for respiratory problems begin to be mentioned.[9] The procedure described, carried out with positive pressure, was designated continuous positive airway pressure (CPAP) and involves the use of a nasal or oronasal mask. However, although CPAP maintains the airways open and can decrease alveolar collapse, it does not provide ventilatory support. This technique became popular as a home-based treatment for obstructive sleep apnea.

Data published since 1987 provide a record of the use of noninvasive ventilation since the 1960 s in patients with post-poliomyelitis syndrome or other neurological disorders.[10-12] These patients used volume-cycled ventilators, in the assist/control mode, using mouthpieces that permitted inspiration at the desired moment.

The adaptation of pressure support to noninvasive forms of ventilation was reported in 1990, when a system was devised to be used with an oronasal mask, dependent on a pressurized gas network, a fact that limited its use to hospital environments.[13]

A pressure-cycled respirator has little chance of maintaining ventilation at an appropriate level when used in a noninvasive manner. This incapacity is due to the form of its pressure wave, which does not present any type of plateau: the pressure rises to a pre-determined value and then drops rapidly back to the baseline. In addition to this characteristic, this respirator does not compensate for air leakage around the nasal or oronasal mask. In pressure support, ventilation is pressure-limited: when the adjusted value for the support is reached, the flow does not stop, just decreases, in order to maintain constant pressure, despite the progressive increase in lung volume. This particularity creates a pressure plateau that is responsible, in great part, for the good results in the improvement of ventilation.

In 1990, some authors[14] described a new form of treating sleep apnea, using two pressure levels: a higher value for inspiratory positive airway pressure (IPAP) and a lower value for expiratory positive airway pressure (EPAP). The machine worked as a continuous flow generator, capable of detecting the respiratory stimulus of the patient, upon which it would rapidly raise the pressure in the circuit to the IPAP level chosen. This pressure was maintained during all the inspiration, and the reduced flow, back to the chosen level of expiratory pressure (EPAP), occurred concomitantly with the decreased flow demand by the patient. The system was denominated the bi-level positive airway pressure (BiPAP) ventilator, and its technical characteristics allowed it to work in a manner very similar to that of pressure support ventilators, except that it was independent from a source of gas under pressure.

The accumulation of secretion or conditions that reduce pulmonary compliance can compromise the efficiency of this type of equipment in improving ventilation, and this can be a significant limitation to its use in the pressure support of individuals with chronic respiratory failure. However, experience has shown that the increased pressure support (up to 30 cmH_2O) and efficient physical therapy techniques for the elimination of sputum broaden the applicability of the BiPAP (although the name is the trademark of a machine, it has come to signify a type of ventilatory support). Therefore, the application of BiPAP was expanded from the treatment of sleep apnea to a wide variety of situations in which its extremely comfortable standard of managing flow constitutes an advantage. This comfort is quite important whenever the patient still maintains some capacity of spontaneous ventilation, since, in these cases, square-wave flow patterns are hard to support.

In this brief history, we can observe the oscillation of basic concepts in the substitution for or aid to ventilation: 'respiratory orthoses and prostheses' were initially noninvasive, later being exclusively invasive, and currently providing a wide variety of options. The improvement of ventilation of a patient with neuromuscular disease demands appropriate knowledge from the doctor, so that they can decide what technique is better for the functional state of that patient.

Evaluation of the evolution of respiratory impairment in neuromuscular diseases

The progression of respiratory complications to chronic respiratory failure in patients with neuromuscular diseases generally occurs as a direct consequence of two principal factors: weakness/fatigue of respiratory muscles (inspiratory, expiratory and of the upper airways); and incapacity to maintain the airways free of secretions. Within a more comprehensive classification of chronic respiratory failure (Chart 2), we can characterize it as a profile of chronic restrictive respiratory failure with hypoventilation.

There have been no large prospective studies addressing this specific topic. Nevertheless, it is possible to establish a minimum routine of periodical evaluation (Figure 3) in order to introduce pertinent therapeutic measures for each stage of the disease. In addition, all therapeutic possibilities should be presented to the patient and their family, so that joint decisions can be duly implemented.

The loss of respiratory muscle strength leads to cough inefficacy and hypoventilation. Atelectasis, pneumonia and respiratory failure, initially during sleep and later on even during wakefulness, are the complications expected in this situation. The adequate frequency of evaluations of the respiratory system of these patients has not been established and depends a great deal on the speed of the progression of the symptoms and on the deterioration of pulmonary function.

Spirometry, pulse oximetry and capnography, together with measurements of peak cough flow, maximal inspiratory pressure and maximal expiratory pressure, make it possible for the physician to predict which patients will require assisted cough techniques and ventilatory support.

Functionally, patients with neuromuscular diseases present decreases in vital capacity and total lung capacity. However, these are not sensitive parameters in the evaluation of respiratory muscle strength of these individuals, since vital capacity can remain above the normal limits even if there has already been severe muscle strength impairment, which can be detected by the measurements of maximal static respiratory pressures.

Chart 2 - Classification of chronic respiratory failure.

Chronic obstructive respiratory failure	Associated diseases
Chronic restrictive respiratory failure	COPD, bronchiectasis and other forms of bronchiolitis
With hyperventilation	Interstitial diseases, such as idiopathic pulmonary fibrosis, fibrogenic pneumoconioses
With hypoventilation	Neuromuscular diseases and thoracic deformities
Chronic respiratory failure caused by pulmonary vascular disease	Primary pulmonary hypertension, other forms of precapillary pulmonary hypertension, pulmonary veno-occlusive disease, chronic pulmonary thromboembolism

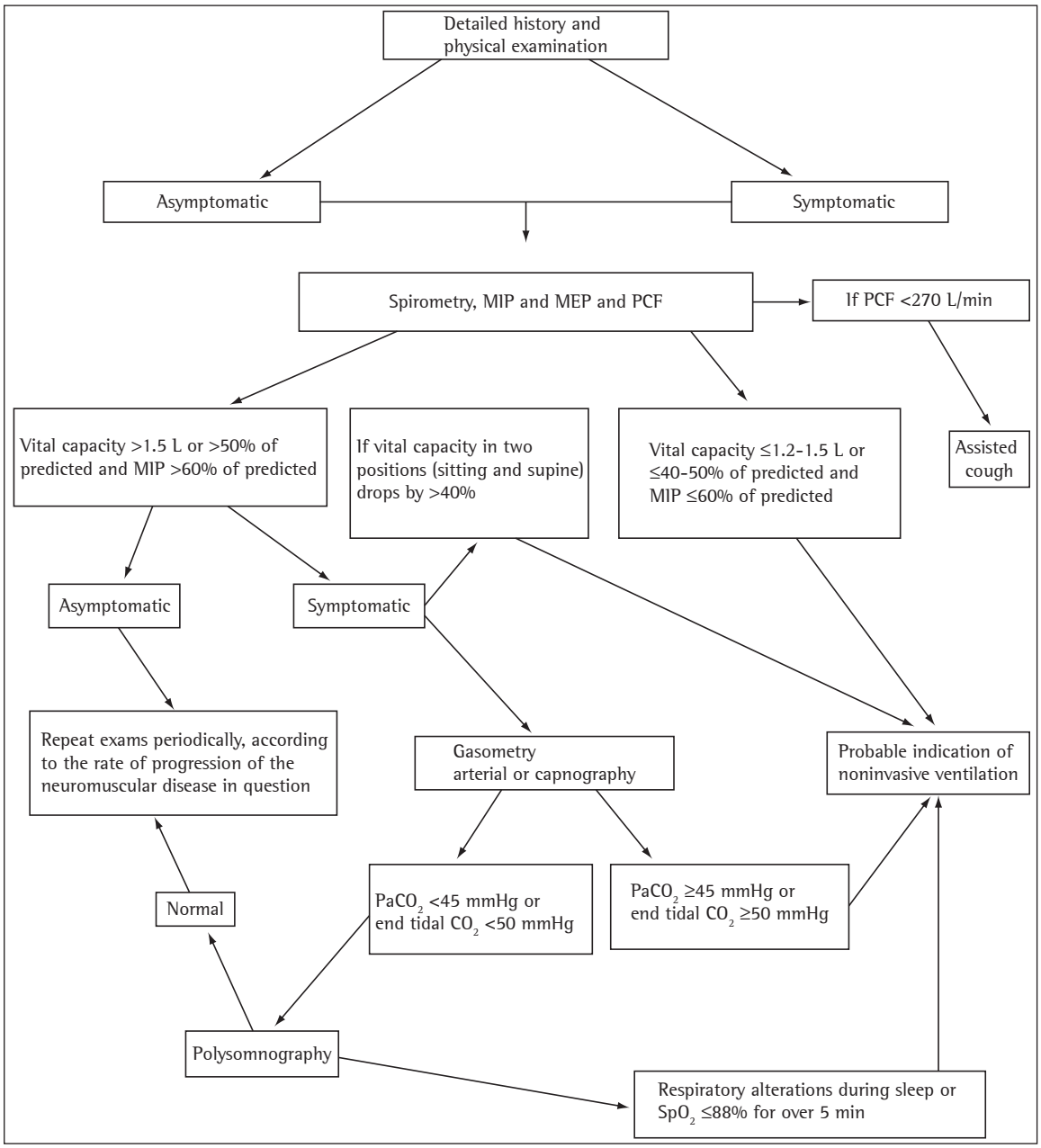

Figure 3 - Protocol for functional evaluation of patients with neuromuscular diseases. MIP: maximal inspiratory pressure; MEP: maximal inspiratory pressure; PCF: peak cough flow; $PaCO_2$: arterial carbon dioxide tension; $ETCO_2$: end-tidal carbon dioxide; SpO_2: peripheral oxygen saturation (via pulse oximetry).

Only when muscle weakness is significant (reductions in force to less than 50% of predicted) can a decrease in vital capacity be observed.[15] However, it is a useful examination in the follow-up of more critical patients whose muscle weakness is sufficient to reduce vital capacity. When vital capacity drops to below the 40–50% of predicted range, the possibility of evaluating arterial gases during wakefulness should be considered, even in asymptomatic patients.[16]

Many different values are mentioned in the literature, according to which impaired pulmonary function increases the risk of complications and death.[17] In patients with Duchenne muscular dystrophy, the variable that correlates most significantly with decreased survival is vital capacity lower than 1 L, and forced expiratory volume in one second lower than 20% of the predicted value is associated with hypercapnia during wakefulness periods.[18]

Therefore, in each respiratory evaluation, it is recommended that the following be performed: pulse oximetry; measurements of vital capacity and forced expiratory volume in one second in two positions (sitting and in the dorsal decubitus position), in order to make the test more sensitive; measurement of maximal inspiratory and maximal expiratory pressures; and the determination of peak cough flow.

When normal individuals are submitted to spirometry in the supine position, the vital capacity values obtained are equal to or even greater than those obtained when the individual is evaluated in a sitting position. The potential increase in the force of the diaphragm in the supine position can result from the muscle distention due to the movement of the abdominal content upwards. Patients whose diaphragm force is impaired by some disease would have much more difficulty in exhaling forcefully when in the dorsal decubitus position. A 40% or greater reduction in the forced vital capacity measured in the supine decubitus position is highly indicative of significantly impaired performance of the diaphragm in this position, thereby making ventilation considerably less effective during sleep. However, any drop of 20% or greater should alert the physician to the need to study respiratory performance during sleep.[19]

The measurement of peak cough flow can be made with the aid of a peak flow meter (Figure 4). Peak cough flow is directly correlated with the capacity to clear secretions from the respiratory system,[20] and values lower than 160 L/min are associated with inadequate clearance of the tracheobronchial tree.[21,22] However, values greater than, but proximal to, 160 L/min do not necessarily guarantee adequate protection of the airways, since muscle force tends to worsen during episodes of infection.[23] Therefore, a peak cough flow cut-off value of 270 L/min has been used to identify patients who would benefit from assisted cough techniques.[24]

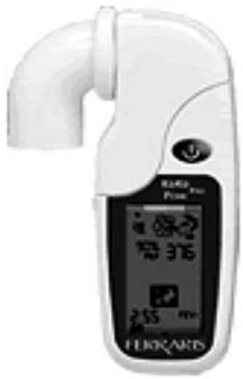

Figure 4 - Digital peak expiratory flow meter.

Maximal expiratory pressure values of 60 cmH_2O or greater are associated with the capacity to generate adequate airflows during cough, whereas measurements equal to or lower than 45 cmH_2O are correlated with inefficacious cough.[25] Pulse oximetry helps to identify situations such as atelectasis and pneumonias, in which it is necessary to intensify the secretion clearance measures.[24]

Carbon dioxide in the blood should also be determined, and capnography-based estimates are ideal for this purpose. The routine follow-up evaluation of patients with neuromuscular diseases does not include arterial blood gas analysis. When capnography is unavailable, a venous blood sample or capillary blood sample obtained by skin puncture should be used. Annual chest X rays facilitate the detection of the sequelae of parenchymal lesions, and determining hematocrit levels can further the evaluation of the severity of hypoxemia and of the efficiency of the treatment.

In addition, patients should also be tested for other respiratory or associated disorders, such as obstructive sleep apnea, aspiration, gastroesophageal reflux and asthma.

A nutritional evaluation can be helpful in controlling obesity related to low caloric expenditure, as well as in minimizing muscle mass loss related to the disease or to a sedentary life style. The body mass index should be periodically determined, as should, if possible, the distribution of lean mass and body fat, through the use of appropriate techniques (impedanciometry or nuclear medicine).

If appropriate oral nutrition cannot be guaranteed, enteral feeding or gastrostomy should be considered. Swallowing should be evaluated based on the clinical history and direct observation of the capacity of the patient to swallow food of various textures. Fluoroscopy should be used if there is a history of suffocation sensation or dysphagia.

Neuromuscular diseases can produce sleep alterations, since alveolar hypoventilation is more intense during sleep. The worsening of alveolar air exchange presents subtle symptoms that can pass unnoticed if not directly analyzed. Hypoventilation during sleep can initially manifest as a progressively increasing number of nighttime awakenings, fatigue, daytime sleepiness and morning headaches.

The phase in which polysomnography is indicated in patients with neuromuscular disease has yet to be established. Therefore, symptoms related to sleep disorders should always be investigated. In the presence of symptoms, independently of concomitant daytime hypercapnia, polysomnography should be performed. If polysomnography is unavailable, the continuous monitoring of oxygen saturation and expired carbon dioxide (oximetry and capnography) can aid in the management of these patients. In the absence of this type of monitoring, determining the levels of carbon dioxide in capillary or arterial blood upon waking can be useful. Asymptomatic patients with daytime hypercapnia or severe pulmonary function disorders should be submitted to polysomnography. It should be noted that this examination can be used in the titration of the pressure levels at the beginning of the ventilation with positive pressure support.

Techniques for lung inflation and airway clearance

If the evaluation of the peak cough flow and expiratory muscle strength suggests a situation of inadequate clearance of airway secretions, techniques aimed at facilitating the clearance of secretions should be introduced.

Maximum inflation capacity is the maximum volume of air that can be held inside the lung with a closed glottis, and it depends on the muscle force of the pharynx and larynx. The increased air volume in the lungs results in greater expiratory volume, which helps the patient generate efficacious expiratory flows for the mobilization of secretions.[26] Maximum inflation capacity can be achieved when a series of inspiratory maneuvers are performed without exhaling in between, in order to accumulate air in the lungs.

Glossopharyngeal breathing consists in using the muscle of the base of the tongue and pharynx in order to push the air contained inside the oropharynx and nasopharynx toward the larynx and trachea.

There are other techniques of pulmonary inflation and airway clearance: pulmonary inflation with an Ambu bag and mask; pulmonary inflation with noninvasive pressure support devices; and pulmonary inflation with conventional mechanical ventilators, used in an invasive or noninvasive form.

All of these techniques to increase inspiratory volume should be used with manually assisted cough, a situation in which the physical therapist or caretaker pushes the diaphragm of the patient upward with one hand and compresses the anterior aspect of the chest with the other, in synchrony with the cough efforts made by the patient, if possible.

For patients presenting little or no force to cough, it is formally recommended to use a device capable of producing inflation with positive pressure and aspiration with negative pressure immediately after lung expansion. This device is known as IN-Exsufflator or Cough Assist (Figure 5). The comparison between the peak cough flows produced with these machines and those produced by maximum inflation and assisted cough reveals the superiority of the mechanical reproduction of

Figure 5 - Cough machine (Cough Assist).

cough. Indeed, the use of one of these machines significantly facilitates the care of patients with neuromuscular disease, even in the early stages of respiratory muscle strength impairment.

Percussive ventilation, which involves high-frequency pulses of air and low tidal volume while maintaining continuous positive pressure in the airways, seems to be efficacious in reversing persistent atelectasis in children.[27] The high frequency oscillation of the chest wall is also a possible technique for the mobilization of secretions. However, like percussive ventilation, it works better in patients whose muscle strength is more preserved. Bronchoscopy should be used only if the other maneuvers fail.

Noninvasive nocturnal pressure support

The diagnosis of nocturnal hypoventilation calls for the use of noninvasive pressure support. The use of a bi-level positive pressure device, or of a conventional mechanical ventilator with nasal or oronasal mask, has proven efficacious in reversing sleep-disordered breathing in various neuromuscular diseases[28,29] The value of positive pressure capable of reversing hypoventilation during sleep should be determined in the sleep laboratory or through careful observation and monitoring at the bedside. In addition, if the underlying disease is progressive, the possible need to periodically increase the pressure values should be emphasized.

Noninvasive nocturnal pressure support can increase survival in some patients,[30,31] improve sleep quality, decrease daytime sleepiness, increase the sense of well-being/independence, improve diurnal blood gases and decrease the rate of decline in lung function.[31-36]

The complications of the noninvasive pressure support using a full face mask include conjunctivitis, skin ulcers, gastric distension and aspiration of vomit. Problems with eyes and facial skin can be prevented if the mask is appropriately fitted. Nasal obstruction, a quite common complaint among patients under noninvasive pressure support with a nasal mask, can be improved by humidifying the air supply and by the use of nasal corticosteroids. Generally speaking, when used in the support of patients with neuromuscular diseases, the machines that provide two pressure levels (BiPAP and similar devices) should necessarily offer the possibility of starting with a controlled mode respiratory frequency, in case the patient stops breathing completely (back-up ventilation). In addition, since these devices are used without alarms, nocturnal oximetry monitoring, with alarm, is recommended.

In patients with neuromuscular diseases, oxygen should never be used without appropriate ventilatory support.

Extending the pressure ventilation support to the diurnal period

Extending the ventilation to the diurnal period should be considered if the patient presents arterial carbon dioxide tension greater than 50 mmHg, or oxygen saturation by pulse oximetry lower than 92% while awake.

More comfort for the patient is achieved with intermittent ventilation with positive pressure, using a mouthpiece. Therefore, any home mechanical ventilator, capable of guaranteeing adequate tidal volumes, can be used in the assist/control mode. The machine can be adapted to a wheelchair, and the circuit, with the mouthpiece, is connected to a support near the mouth of the patient. Whenever necessary, the patient moves the neck, holds the mouthpiece with the teeth and waits for an inflation of the equipment (or triggers the inflation, if capable). Obviously, this type of ventilatory support presupposes adequate function of the bulbar and neck muscles. Mouthpieces fixed to the teeth permit the use of this same type of ventilation during the nocturnal period.

Tracheostomies should only be recommended in situations of absolute intolerance of noninvasive ventilation or in case of severe impairment of the bulbar muscles. Even in the latter, we can still insist on the noninvasive pressure support, provided that a mechanically assisted cough machine and a pulse oximeter are available. Tracheostomies hinder the normal defense mechanisms of the trachea, increase secretion, rapidly colonize with difficult to control germs, impede swallowing and impair speech (unless speaking valves, which are costly, are used).

Final considerations

Neuromuscular disease patients and their families should be presented with all of the therapeutic possibilities. Each treatment recommendation should be thoroughly discussed. There is no technical or ethical justification for making any decision against

the wishes of the patient, nor should physicians use their own values to decide what an acceptable level of quality of life is for another person.

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