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# Noninvasive Ventilation in Neuromuscular Diseases

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Additional information is available at the end of the chapter

<http://dx.doi.org/10.5772/intechopen.77173>

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## Abstract

Respiratory muscle weakness is the main contributor to respiratory imbalance in patients with neuromuscular diseases (NMD). In the advanced stages of the disease, patients develop a chronic respiratory failure due to muscle weakness, which is the principal cause of death among these patients. Respiratory muscle weakness ultimately causes alveolar hypoventilation, initially nocturnal, and later daytime respiratory failure. The signs and symptoms of early respiratory muscle weakness are discrete, namely: dyspnoea on effort, orthopnea, insomnia, frequent nocturnal awakenings, morning headache, loss of appetite, excessive daytime sleepiness, depression, anxiety, and marked fatigue. The management of respiratory failure in neuromuscular diseases requires the use of non-invasive ventilation (NIV) to assist the respiratory muscles in order to correct the alveolar hypoventilation and ameliorate gas exchange. NIV thus slows down the decline of forced vital capacity thereby improving the patient's quality of life, physical activity and hemodynamics, normalization of blood gases, slight improvement in other physiological measures, and maximal mouth pressures and increases survival. NIV support should be offered to all patients who present with early signs of ventilatory failure as it is probably the most effective among treatments in prolonging life in neuromuscular patients.

**Keywords:** non-invasive ventilation, respiratory failure, neuromuscular diseases

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## 1. Introduction

Neuromuscular diseases (NMD) are a group of diseases that affect the nerves that control voluntary muscles, including respiratory muscles in more advanced stages, that varies according to underlying disease [1]. The weakness of the respiratory muscles causes alveolar hypoventilation, initially during sleep, and then leading to respiratory insufficiency in the

daytime [1, 6]. Respiratory infections, in which neuromuscular patients are predisposed, usually aggravate the evolution of respiratory insufficiency.

Muscle weakness affects three categories of muscles involved in breathing:

- Inspiratory muscles, which contribute to the act of ventilation and voluntary inspiration.
- Expiratory muscles performing forced expiration and forced expiratory flow.
- Bulbar muscles, which have a glottic function, thus contributing to swallowing/speech, increased intra-thoracic pressure and cough [4, 9, 13].

Affection of inspiring muscles in NMD leads to dyspnoea, orthopnea, alveolar hypoventilation and hypercapnia. Alveolar hypoventilation occurs initially during rapid eyes movement sleep (REM) sleep, then in non-REM sleep, leading to morning hypercapnia and later in the evening hypercapnia as well as hypoxia [2, 3, 8, 13]. The weakness of the expiratory muscles leads to an inefficient clearance of the airway, ineffective cough, thus predisposing to respiratory infections. Affecting bulbar muscles causes swallowing and speech disturbances, aspiration secretions, reduced airway clearance, and recurrent respiratory infections [1, 6, 8, 13].

Existing studies have shown, mostly uncontrolled trials, that non-invasive ventilation (NIV) in neuromuscular diseases improved quality of life, physical activity and hemodynamic, normalization of blood gases and slight improvement in other physiological measures, such as the vital capacity and maximal mouth pressures [1, 3, 5, 6, 11, 12].

Monitoring of patients diagnosed with NMD is essential for the early detection of signs of respiratory failure and the establishment of NIV at an early stage of respiratory distress.

## **2. Signs and symptoms of respiratory failure in neuromuscular diseases**

### **2.1. Symptoms**

The symptoms of respiratory muscle weakness depend on the speed of its development. When the onset is subacute (for example, in Guillain-Barre syndrome), the predominant symptoms are dyspnoea and orthopnea, or sometimes respiratory arrest. These symptoms are often accompanied by those of bulbar weakness and inability to clear respiratory secretions. The symptoms of respiratory failure may easily be overlooked and should be specifically sought in any patient with rapidly progressive weakness, especially when the bulbar muscles and shoulder girdle are affected [4, 8, 9, 11].

When respiratory muscle weakness develops gradually, inadequate respiration usually occurs first during sleep. Symptoms of nocturnal hypoventilation include a broken sleep pattern, nightmares, nocturnal confusion, morning headache, daytime fatigue, mental clouding and somnolence [4, 8, 9, 11].

Symptoms	Signs
Dyspnea to minimal effort or speech	Tachypnea
Orthopnoea	The use of auxiliary respiratory muscles
Frequent nocturnal awakenings	Paradoxical abdominal movements
Excessive daytime sleepiness	Reducing the amplitude of the thoracic movements
Daytime fatigue	Ineffective cough
Morning headaches	Sweating
Difficulty to expectorate secretions	Tachycardia
Apathy, loss of appetite	Morning confusion, hallucination
Hypomnesic concentration deficiency and memory impairment	Weight loss
Difficulty sleeping	Dry mouth or hypersalivation

Table 1. Signs and symptoms of respiratory failure in neuromuscular diseases.

Rapidly progressive NMD	Variable progression	Slowly progressive or non-progressive
Amyotrophic lateral sclerosis (ALS)	Limb girdle muscular dystrophy	Spinal muscular atrophy
Duchene muscular dystrophy (DMD)	Miopathies Nemaline miopathy Metabolic miopathy	Poliomyelitis, post-polio syndrome
	Merosin negative congenital muscular dystrophy	Facio-scapulohumeral muscular dystrophy
		Becker muscular dystrophy

Table 2. Classification of Neuromuscular disorders (NMD) according to evolution [1].

Exertional dyspnoea is encountered less frequently in neuromuscular patients than in those with other cardiorespiratory disorders, particularly when the patient has reduced mobility. Dyspnoea when lying flat or immersed in water specifically suggests weakness of diaphragm [4, 9].

2.2. Signs

A patient with severe respiratory muscle weakness or respiratory failure may appear overtly breathless and may be using accessory muscle of respiration. The patient may be unable to speak in complete sentences or take deep breaths to command. Inability to count from 1 to 20 in a single breath indicates significant reduction of vital capacity (VC) or forced vital capacity (FVC) [4, 9, 11]. Paradoxical abdominal motion (inwards movement of the abdominal wall with inspiration) suggests significant weakness of the diaphragm. The combination

of hypoxemia and respiratory acidosis may produce mental clouding or somnolence. It is also important to assess the bulbar musculature, weakness of which can hinder clearing of respiratory secretions and so allow aspiration. Most of the patients with respiratory muscle weakness resulting from a neuromuscular condition have limb weakness. Acute respiratory failure in patients with neuromuscular disorders is often precipitated by respiratory infection [4, 8, 9, 11].

Particular attention should be paid to:

- Presence or absence of bulbar weakness.
- A tall, thin face (congenital myopathy, myotonic dystrophy)
- Ptosis of ophthalmoparesis (myasthenia)
- Fasciculation (motor neuron diseases)
- Paraspinal muscle wasting (acid maltase deficiency)
- Skin rash (dermatomyositis)

The main signs and symptoms of respiratory failure in the NMD can be found in **Tables 1** and **2**.

### 3. Course of neuromuscular disorders

Neuromuscular disorders can be divided into slowly progressive, rapidly progressive and NMD with variable progression; understanding the speed of progression of the disease is important in deciding the appropriateness of NIV [1, 2].

In the rapidly progressive NMD, the prototype of this category is Duchenne muscular dystrophy (DMD). Monitoring these patients begin in early ambulation stage, when the patient can walk independently, by using serial spirometry, sleep studies and blood gases, for capturing early FVC decline and respiratory disturbance in REM and non-REM sleep [11, 14].

In general, reducing FVC demonstrated by spirometry does not correlate very well with the occurrence of dyspnea as a symptom at these patients. Therefore, monitoring of clinical signs and symptoms of respiratory disturbance is not enough. Alveolar hypoventilation, secondary to respiratory muscular weakness, initially occurs in REM sleep, and it can be early diagnosed by using polysomnography. In a later stage, sleep disturbances occur both in REM and non-REM sleep, resulting in morning hypercapnia, and in the final stages, it also occurs during the daytime [11, 14].

The classical spirometry measuring FVC has some limitation in detecting moderate inspiratory muscle weakness; performing lung function test in supine position, could improve the value of FVC [16]. In DMD and other rapidly progressive NMD, initial evaluation using

spirometry, polysomnography, blood gases and SaO<sub>2</sub> is performed once a year, subsequently two times a year, and in advanced stages at 3 months [11, 14]. In slowly progression NMDs and those with variable progression, annual monitoring is sufficient.

#### 4. Monitoring evolution of NMD

From a functional stand point, neuromuscular patients can be classified as following:

- Ambulant patients, who can walk without any help.
- Non-ambulant patients, who cannot stand seated without any help.
- Non-ambulant patients, who can stand seated without any help, but cannot walk without any help [14].

The monitoring of the patients is in relation with the specific neuromuscular disease and the rate of progression of the disease in each patient. It is recommended that the respiratory evaluation be done every 3–6 months, less frequently for ambulant patients, and more frequently for the nonambulant patients, and where the disease progresses at a faster pace.

Methods for respiratory monitoring in NMD:

- Spirometry
- Pulse oximetry
- Blood gases or capnography
- Polysomnography and/or cardiorespiratory polygraph
- Manometry for measurement of maximum inspiratory pressure (MIP) and maximum expiratory pressure (MEP)
- Cough peak flowmetry
- Sniff nasal inspiratory pressure (SNIP)

Spirometry and lung function testing are useful for detection of reducing FVC [11, 21]. It can be applied while the patient is standing, however, classical spirometry measuring FVC has some limitations in detecting moderate inspiratory muscle weakness, spirometry in supine position is recommended. When measured FVC is in the supine position, vital capacity is lower, especially in patients with diaphragmatic weakness. Supine vital capacity may be useful in monitoring disease progression [11, 20]. A vital capacity of <1.11 liters predicted risk of chest infection with a sensitivity of 90.5% and a specificity of 70.8% [11].

Peak flowmetry during cough [cough peak expiratory flow (PEF)] allows efficient evaluation of coughing. A cough with PEF <270 ml/min suggests ineffective coughing [16].

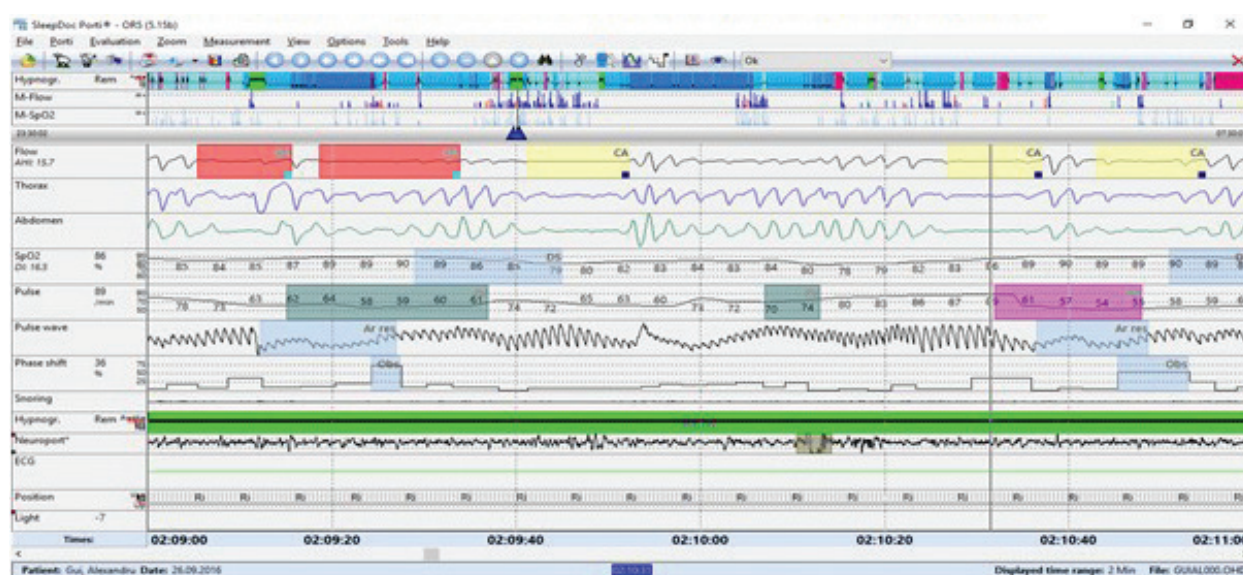


Pulse oximetry can be used for highlighting hypoxemia during day, but also for guiding during the clearance of the airways. If the  $O_2$  saturation is lower than 94%, clearance of the airways must be initiated. Continuous night pulse oximetry can be used for screening of the night time hypoxemia. Currently, it is not recommended to routinely monitor  $SaO_2$  at home, more studies being required for this issue [11, 14, 16].

Blood gases or capnography allows the assessment and evaluation of the initial morning hypercapnia; then, it becomes permanent. Blood gases should be performed if  $SaO_2 < 94\%$  and the patient do not have lung disease [16, 17]. In children with NMD, the use of capnography is preferred, a noninvasive method, in order to determine the transcutaneous  $CO_2$ , and to monitor the exchange of gases routinely [11, 14].

Polysomnography represents a diagnostical investigation option for respiratory disturbances during sleep and during alveolar hypoventilation in NMD and is the most pertinent indicator for proposing NIV [10, 21]. Polysomnography (**Figures 1 and 2**) is useful in nonambulant patients who cannot stand without any help and can be used for initiation and titration of the respiratory support, more specifically non-invasive ventilation. If polysomnography is not available, the cardiorespiratory polygraph is recommended, with a minimum of four channels:  $O_2$  saturation, cardiac frequency, nasal flow, and chest movements during sleep [11, 14, 16].

A specific evaluation of respiratory muscle strength is the measurement of maximum inspiratory pressure (MIP), maximum expiratory pressure (MEP), and sniff nasal pressure (SNIP). In some patients, specifically in NMD with bulbar determination, some discrepancies are registered between the maximum inspiratory pressure (MIP) and sniff nasal pressure (SNIP). As a consequence of the discrepancies, it is recommended to do both tests, taking note to select the highest pressure [11, 16].



**Figure 1.** Polysomnography in 9 years old patient with Central Core Myopathy. Obstructive and central apnoea, AH1 = 15.7/hour of sleep, desaturation in  $O_2$  in REM sleep.



**Figure 2.** Polysomnography in a patient aged 45 years with ALS generalized form, spastic tetraplegia, pseudobulbar syndrome, sialorrhoea, eating difficulties, dyspnoea. Epoch with predominantly hypopneic flow, accompanied by desaturations in  $O_2$ , in stage  $N_2$ , morning  $PCO_2 = 51$  mmHG.

## 5. Indications and contraindications for long-term NIV in NMD

The most frequent indications for NIV in NMD are:

- Amyotrophic lateral sclerosis (ALS)
- Duchenne's muscular dystrophy
- Becker's muscular dystrophy
- Steinert's muscular dystrophy
- Myasthenia gravis
- Spinal muscular atrophy [2, 7, 13, 15]

Depending on the natural evolution of neuromuscular disease to respiratory distress, NIV can be introduced to the management of the disease as soon as possible.

Contraindications for NIV in NMD:

- Facial burns/trauma/facial surgery or recent upper respiratory tract surgery
- Anatomical or functional obstruction
- Gastrointestinal or ileus bleeding



- Vomiting
- Hypersalivation
- Severe hypercapnia or severe respiratory acidosis ( $\text{pH} < 7.1$ )
- Without patient's consent for setting up NIV [2, 7, 15].

## 6. Neuromuscular diseases and long-term NIV: when?

There is plenty of evidence that precociously introducing NIV to the neuromuscular patient brings improvements in the quality of life and even prolongs survival. The question the clinician must ask is: when is the optimum moment for starting NIV with the neuromuscular patient?

NIV must be initiated when:

- The neuromuscular patient shows signs and symptoms of respiratory disturbances
- $\text{pCO}_2$  in the morning  $>45$  mmHg
- $\text{FVC} < 50\%$  predicted value
- MIP or SNIP  $< 60$  mmHg
- Nocturnal  $\text{SaO}_2 < 88\%$  for more than 5 min while under room air [1, 2, 14, 17].

Or

- $\text{FVC} < 80\%$  predicted value plus any symptoms or signs of respiratory impairment
- SNIP or MIP  $< 65$  cm  $\text{H}_2\text{O}$  for men or  $55$  cm  $\text{H}_2\text{O}$  for women plus any symptoms or signs of respiratory impairment, particularly orthopnoea [17].

The classical indications to start NIV, when  $\text{pCO}_2 > 45$  mmHg or when a patient has an exacerbation, as markers for respiratory failure could be too late. The history of neuromuscular diseases has a prolonged period of discrete symptoms of respiratory impairment, but with variable period of nocturnal hypoventilation. Performing polysomnography every year in a neuromuscular patient, as a routine method of disease monitoring, we can identify nocturnal hypoventilation very early and NIV could be started early [10]. Starting NIV earlier in the course of respiratory failure should be accompanied by a significant improvement in quality of life, and probably in prolonging life. There is no data for the moment to demonstrate this [10].

## 7. Introducing NIV in NMD, ventilator choices, interfaces

Non-invasive ventilation in NMD improved or corrected diurnal hypoxemia and hypercapnia, improved nocturnal hypoventilation and increases maximal respiratory pressures [7, 11, 14, 15, 22].

From the clinical point of view, NIV improved quality of life, control of symptoms of sleep related breathing, for example, headaches, sleep fragmentation, decreased daytime sleepiness, increased ability to perform daily activities, provide patients with sense of control and autonomy in advance stages, reduce hospitalizations and prolonged survival [1, 2, 3, 7].

Type of ventilators for long-term NIV:

- Pressure-support ventilator
- Volume-targeted ventilator: deliver known tidal volumes, but most machines have a limited capacity to correct leaks, leading to underventilation
- Hybrid mode ventilator: pressure-targeted volume-assured mod

Bi-level positive airway pressure ventilator (BiPAP), spontaneous/timed (S/T) mode is the most common type of pressure-support ventilator used for long-term NIV in NMD. There is a lack of study to compare different types of ventilators used for long-term NIV. But, pressure-targeted ventilators tend to be lighter and cheaper and also comfortable to the patient than volume-targeted ventilators [6, 11, 18].

Intelligent volume-assured pressure support (iVAPS) is a hybrid mode ventilator, providing constant automatic adjustment of pressure support (PS) to achieve a target ventilation determined by the patient's pathology [11, 18]. iVAPS demonstrated, in small studies, similar arterial



**Figure 3.** Choosing the interfaces: Pillow mask.

blood gases control to BiPAP, but iVAPS had higher overnight adherence, due to better patient-ventilator synchrony; there was no difference in outcome between ventilator modes for spirometry, respiratory muscle strength, sleep quality, arousals or O<sub>2</sub> desaturation index [11, 18].

### 7.1. Choosing the interfaces

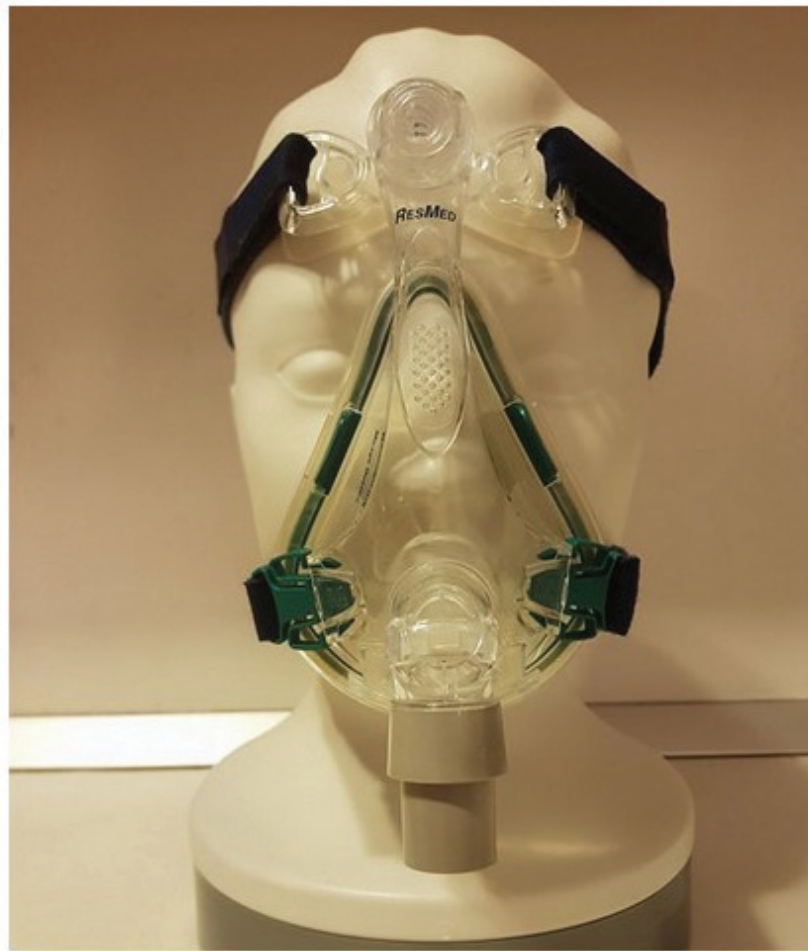
Extremely important for good compliance to NIV is choosing the right interfaces. Nasal mask and pillow mask are best suited for cooperative patients that have a lower severity of the disease, or for children, needed low to moderate pressures only (< 20 cm H<sub>2</sub>O). It also allows the patient to speak, drink, cough and clear his/her secretions while receiving the treatment. Nasal masks are more prone to leaks and the effectiveness is limited in patients with nasal obstructions, septal defects or other kind of deformities. Nasal and pillow mask are more comfortable for the patient than orofacial mask [11, 38, 39].

Orofacial mask, which encompass the mouth and nose are best suited for less cooperative patients who have more or less severe illnesses. It particularly fits patients who are mouth-breathing and edentulous and they are contraindicated in claustrophobic patients. Orofacial mask does not allow the patient to talk or eat and it is more uncomfortable for the patient than nasal or pillow mask [11, 38, 39].

Nasal mask, pillow mask and orofacial mask are illustrated in **Figures 3–5**.



**Figure 4.** Choosing the interfaces: Nasal mask.



**Figure 5.** Choosing the interfaces: Orofacial mask.

#### *7.1.1. Adverse effects and complications of NIV*

The majority of adverse events of NIV are related to the mask: discomfort, skin rash, claustrophobia, nasal ulceration nasal congestion, eyes irritation, nasal or oral dryness. This mask related adverse events could be easily resolved by changing the interface and adding humidifier for the dryness of the mucosa.

Other NIV complications are aspiration pneumonia, pneumothorax or hypotension, with a low frequency < 5% [11, 38, 39].

### **8. Monitoring NIV in NMD**

Effectiveness of NIV depended on a several factors: settings, interfaces, compliance and adherence of the patient to his ventilator. For obtaining a good compliance and adherence to NIV, monitoring NIV is crucial. The minimum requirement is a sleep study recording continuous oximetry, capnography or blood gases.

The frequency of monitoring NIV is depending of the cases; for new cases, monitoring is required to be done more often, every few weeks, until established that we obtained correction



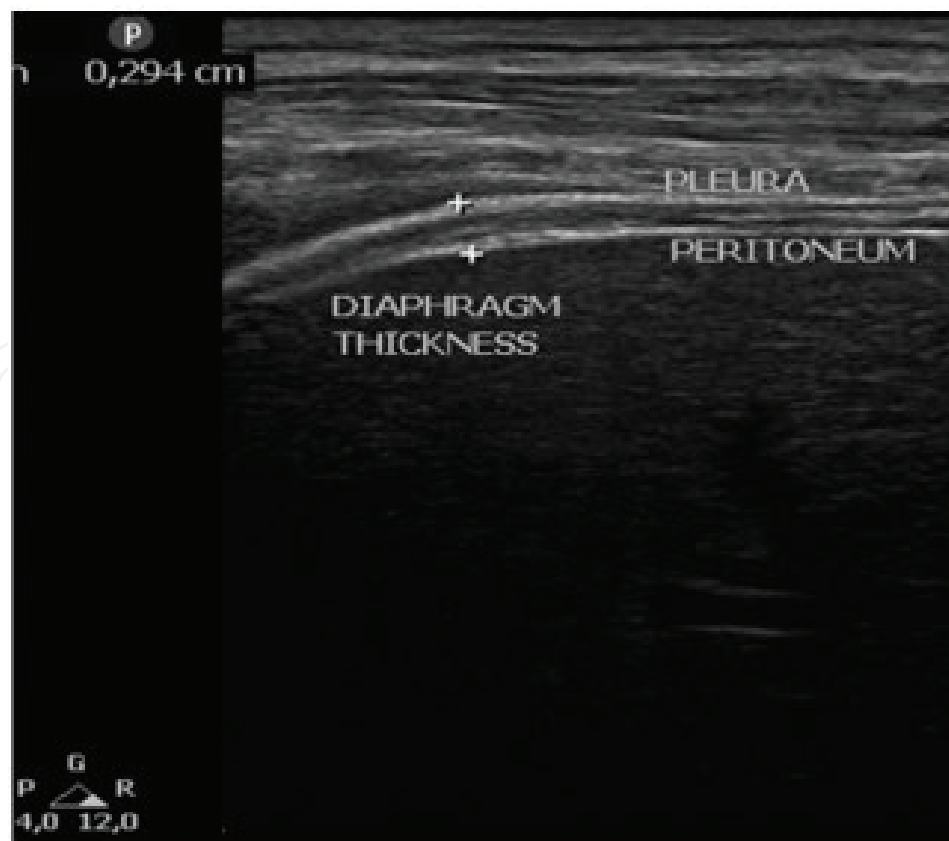
of nocturnal hypoventilation and blood gases. In stable cases on home-ventilation, with slowly progressive or non-progressive disease, annual assessment is sufficient [11, 38]. The new type of ventilators provided a compliance card, which permitted a minimum set of data to monitor: hours of usage, AHI index, leaks.

### 8.1. Using ultrasound to monitor NIV in NMD

One of the main causes of morbidity and mortality in patients with neuromuscular diseases (NMD) is respiratory failure. The diaphragm acts as the main respiratory muscle during inspiration and accounts for 70% of the inspired air volume during regular breathing [19]. The diaphragm function can indirectly be analyzed by techniques such as fluoroscopy and chest radiography, which are non-specific and also ionizing exams [23]. Ultrasound (US) as a non-invasive, radiation-free imaging tool, allows an accurate, reproducible and safe assessment of diaphragm anatomy and function at the bedside [24–27]. Ultrasound has been shown to be similar in accuracy to most other imaging modalities for diaphragm assessment [28].

#### 8.1.1. Technique of diaphragmatic ultrasound (US) assessment

With ultrasound, the diaphragm is typically identified by its deep location, curved shape and muscular echo-structure. Longitudinally it has a mixed echogenic appearance, consisting of hypo echoic (dark) muscle fibers separated by two hyper echoic (bright) layers: peritoneum and pleura (**Figure 6**).



**Figure 6.** Normal US appearance and thickness of the diaphragm.

Patients are typically examined during spontaneous respiration to help identify the diaphragm moving. The supine position of the patient is preferred, because there is less overall variability, less side-to-side variability, and greater reproducibility [29]. Also, it could identify any paradoxical movement.

The right diaphragm can be visualized through the liver window. Visualization of the left diaphragm could be sometimes more difficult because of the smaller window of the spleen.

Classically, there are two methods to evaluate the diaphragm: the analyses of the movement of diaphragmatic dome using the M mode and the measurement of diaphragmatic thickness and the thickening during inspiration in the area of apposition using the B mode.

The anterior subcostal view is preferred for evaluation of diaphragm excursion. It requires a lower frequency, ideally curvilinear, transducer (2–6 MHz) placed between the mid-clavicular and anterior axillary lines (**Figure 7**), so that the ultrasound beam could reach the posterior third of the diaphragm. B mode is used to visualize the diaphragm moving toward or away from the transducer. Imaging is then changed to M mode with the line of sight positioned in order to obtain maximum excursion (**Figure 8**). Either dome of the diaphragm can be evaluated using the liver and spleen window and the amplitude of excursion can be measured on M mode, and diaphragm velocity can be calculated (**Figure 8**).

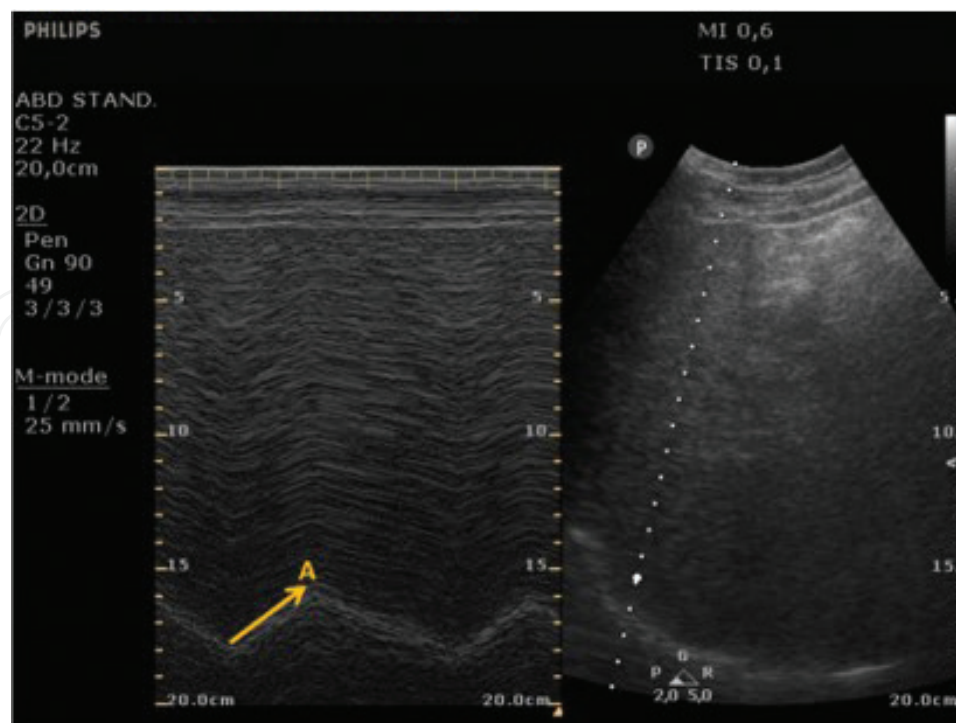
For an intercostal view, a higher frequency linear array transducer (7–18 MHz) is placed at the anterior axillary line, with the transducer positioned to obtain a sagittal image at the intercostal space between the 7th and 8th, or 8th and 9th ribs (**Figure 9**). The zone of apposition is assessed for measurement of the diaphragm thickness and echogenicity.

### 8.1.2. Measurements

Diaphragm thickness is measured at the zone of apposition during inspiration or expiration using the intercostal approach. The average thickness of the diaphragm is 0.22–0.28 cm in healthy volunteers [30]. Diaphragm thickness less than 0.2 cm, measured at the end of expiration, have been proposed as the cut-off to define diaphragm atrophy [31].



**Figure 7.** Diaphragm assessment in anterior subcostal view.



**Figure 8.** Assessment of diaphragm movement in M mode and 2D mode (A-wave amplitude).

Muscle fibers shorten with contraction and cause muscle thickening. A chronically paralyzed diaphragm is thin, atrophic, and does not thicken during inspiration. The measurement of thickness alone may miss an acutely paralyzed diaphragm with normal thickness or could incorrectly identify atrophy in a low weight individual with a healthy, yet thin, diaphragm. Therefore, the degree of diaphragm thickening has been proposed to be more sensitive than measurement of thickness alone [32]. Thickening fraction (TF) was calculated as:  $[\text{thickness at end-inspiration (TEI)} - \text{thickness at end-expiration (TEE)}] / \text{TEE}$  and expressed as a percentage (TEI thickness at end-inspiration; TEE thickness at end-expiration). Diaphragm thickening of less than 20% is proposed to be consistent with paralysis [32].

Diaphragm movement is recorded using the M mode assessment of the dome in the anterior subcostal view. The diaphragm is seen as a single thick echogenic line, and its movements with respiration can be plotted against a time curve. Measurement of the amplitude of excursion can be used to compare movement of the two hemi-diaphragms and for follow-up of diaphragmatic function (**Figure 8**). The normal range of motion from the resting expiratory position to full inspiration in adults has been reported to range from 1.9 cm in normal breathing to 9 cm in deep breathing [33]. Excursion greater than 2.5 cm in adults has been proposed as a cut-off for excluding severe diaphragm dysfunction [34]. Diaphragm weakness is indicated by less than normal amplitude of excursion on deep breathing with or without paradoxical motion on sniffing. Some variations are mentioned related to sex, age, weight or height.



**Figure 9.** Diaphragm assessment in intercostal view, using a high frequency linear probe.

#### *8.1.3. Clinical application*

Mechanical ventilation is associated with decreased muscle weight and alterations in contractile properties of the diaphragm within 48 h of intubation [35]. Diaphragm dysfunction may contribute to weaning failure, even in patients with no obvious reason to suspect phrenic nerve or diaphragm pathology. Decreased diaphragm excursion on M-mode ultrasound has been shown to predict weaning failure, with a 1.4 cm cut-off for the right hemidiaphragm and 1.2 cm for the left hemidiaphragm [36].

In critically ill patients under non-invasive ventilation, the diaphragm thickness and the thickening fraction (TF) are decreased as the level of pressure support increased (5, 10, 15 cm H<sub>2</sub>O). The measurements done in the zone of apposition during tidal ventilation showed that, during NIV, thickening of the diaphragm is due to muscle effort and not due to increase in pulmonary volume induced by ventilation [37].

TF could be used in the ICU setting to assess diaphragmatic function and could contribute to respiratory workload in various situations, including ventilator-induced diaphragmatic dysfunction and ICU-acquired paresis [32].

## **9. Conclusions**

Non-invasive ventilation in neuromuscular diseases should be introduced earlier in the evolution of respiratory failure, for obtaining maximum benefit for the quality of life, control of



symptoms, increased ability to perform daily activities, reduce hospitalizations and prolonged survival. Choosing the ventilator, the most appropriate interface, the ventilation mode, and periodic monitoring of the NIV is essential in obtaining success.

## Abbreviations

ALS	Amyotrophic lateral sclerosis
BiPAP	bi-level positive airway pressure ventilator
DMD	Duchenne muscular dystrophy
FVC	forced vital capacity
iVAPS	intelligent volume-assured pressure support
NMD	neuromuscular disease
NIV	non-invasive ventilation
pCO <sub>2</sub>	arterial pressure of CO <sub>2</sub>
PS	pressure support
REM	rapid eyes movement sleep
S/T	spontaneous/timed
SNIP	sniff nasal pressure
US	ultrasound
TF	thickening fraction
MIP	maximum inspiratory pressure
MEP	maximum expiratory pressure

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