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Mini Review

Does Neuromuscular Disease Pose a Challenge to Respiratory Therapists?

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Abstract

Neuromuscular disorders can be of different types. Mechanical ventilation and respiratory care play a major role in maintaining the ventilation and patency of the airways. Ventilatory failure may range from mild to moderate and severe. Skilled Respiratory Therapists along with the team of neurologists and nurses and Physiotherapists helps the patients to return back to their life.

Keywords: *NIV; Rett Syndrome; Myasthenia Crisis; Respiratory Therapist*

Introduction

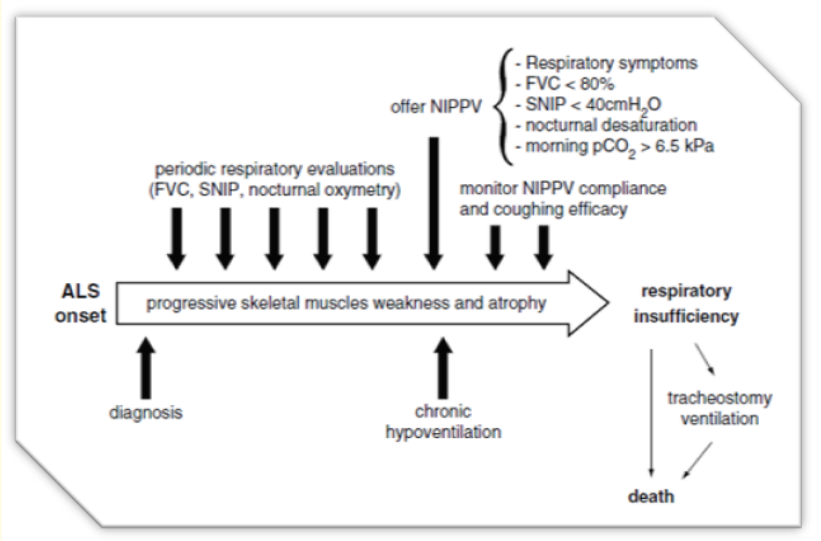
Neuromuscular disorders are many and they may be myelopathies which include muscular dystrophies such as Duchenne, limb girdle, facioscapulohumeral, myotonic dystrophy and non-Duchenne myopathies. Metabolic or congenital myopathies, inflammatory myopathies (polymyositis, linked with connective tissue diseases or other systemic diseases), myopathies from drugs or trauma and diseases of the myoneural junction such as myasthenia gravis are examples of non-Duchenne myopathies [1].

Amyotrophic lateral sclerosis

Amyotrophic lateral sclerosis (ALS) affects approximately 1.5 individuals per 100,000 every year who are 55 - 75-year-old [2]. Respiratory paradox, rapid shallow breathing, or accessory muscle contraction are clinical features. There are patients who may not show any symptoms, which may delay in detecting ALS at an earlier stage [3,4].

FVC correlated with survival and is linear decrease during the course of the disease, but with a clear inconsistency from patient to patient limitations of FVC are the low sensitivity in patients with bulbar involvement, as of reduced buccal strength, or cognitive involvement, and the relative insensitivity to detect mild or moderate diaphragmatic dysfunction. NIV is found to be successful in ALS patients [5]. MIP and MEP are other sensitive measurements. Arterial blood gas analysis may also be of help in the evaluation of patients with ALS. Sniff nasal inspiratory pressure (SNIP), which is considered as a good measure of diaphragmatic strength. Prevention of aspiration and pneumonia and adequate management of bronchial secretions should also be taken care. NIV should be used when indicated for patients with ALS.

Pathway of the progression of respiratory dysfunction in patients with amyotrophic lateral sclerosis (ALS) and the respiratory management that is recommended. FVC forced vital capacity, NIPPV noninvasive positive pressure ventilation, SNIP sniff nasal inspiratory pressure.



The effectiveness of cough is determined by peak cough flows (PCFs), if the OCF is less than 2.7 L/s then it indicates ineffective cough. PCF decreases during respiratory infection and if it is below 4.5 L/s then there is a high chance of pulmonary complications especially when there is bulbar dysfunction. Coughing techniques such as anterior chest compression and abdominal thrust are effective ways of eliminating airway secretions in patients with neuromuscular disease (NMD). Mechanical in sufflator Exsufflator also can be used effectively in patients with NMD. Positive pressure is applied through the face mask and it provides high expiratory flow rates and high expiratory pressure gradient which are generated between the mouth and alveoli.

Home ventilation is the best choice in these conditions but will need participation and commitment from a trained caretaker and dedicated respiratory therapist [1].

NMD patients may require tracheostomy, it is a major step and need good level of care, affects the quality of life and increase the ventilator dependence. Some patients do not tolerate NIV and in some it is not feasible and these are the common reasons why they end up in tracheostomy. NMD patients who have worsening of the respiratory failure in conjunction with low vital capacity, persistent hypercarbia and the need to increase the ventilator time.

In patients who does not tolerate the NIV, mouthpiece ventilation should be used and mainly in patients who use the NIV many hours a day and presenting skin lesion, gastric distension or eye irritation, sometimes alternating with the nasal or full-face mask; it is used also in weaning from tracheostomy tube [6].

Rett syndrome

A genetic disorder which affects the neurological system is Rett Syndrome and it mainly affects girl children. It is characterized by a slow growth of the brain causing a progressive loss of motor skills and speech. Over time children with Rett syndrome experience increasing problems with the use of muscles that control movement, coordination and communication.

These patients also may have problems with the coordination of breathing and seizures. Main issues are with breath holding, hyperventilation, forceful exhalation of air and the swallowing of air. They may also experience shallow breathing or periodic breathing during

sleep. NIV is the ventilator modality used through face mask. Ventilators such as Vivi 65 is useful in such patients. Vivo 65 ventilator offers a unique patented trigger technology called "eSync". Information from the flow sensor detects the start of patient effort or diaphragmatic contraction, the end-tidal CO_2 monitoring and pulse oximetry functions of the Vivo 65 will facilitate the day-to-day monitoring in the home environment [7].

Patients with neuromuscular disease frequently show piecemeal deglutition and weakening of the breathing-swallowing interaction [8].

Myasthenia gravis

Estimated prevalence of Myasthenia Gravis is 0.5 - 12.5/100,000 and incidence is 0.4/100,000 in the general population. It also has a male to female ratio of 2:3 [9]. Consequences of respiratory failure on lung mechanics include reduced capacity to expand rib cage, decreased lung inflation, decreased lung compliance, increased work of breathing, altered ventilation to perfusion ratio, diffuse lung micro atelectasis. Mechanical ventilation and immunotherapy have helped to reduce the mortality rate to 4 - 8%. NIV is the first line of treatment in myasthenia crisis since the concern is to avoid tracheal intubation and the followed complications.

Duchenne muscular dystrophy

Duchenne muscular dystrophy (DMD) is due to mutations in the gene names as dystrophin gene in X chromosome at Xp21. Treatments such as noninvasive ventilation (NIV) and cardio protection have enhanced survival and quality of life for patients with DMD [10].

Summary

Patients with neuromuscular disorder poses a great importance in the management of the ventilator and their respiratory needs. Respiratory Therapists play a very prominent role with other allied health professionals and neurologists in taking care of NMD patients. Improving the quality of life and survical is the major goal of treatment.

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