Non-invasive Ventilation for Neuromuscular Disease

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The Sleep Disorders Center supports patients with neuromuscular weakness through the Comprehensive Lung Center at Falk Clinic. Patients are typically referred by their neurologist, or other referring physician, following a diagnosis of neuromuscular weakness with associated concerns for respiratory insufficiency or failure and a need for noninvasive inspiratory positive pressure ventilation (NIPPV). We typically see patients with amyotrophic lateralizing sclerosis (Lou Gehrig's disease), muscular dystrophy, and Pompe's disease. Patients are readily seen with advanced neuromuscular weakness, but it is advantageous to see them at earlier stages of illness in an effort to preempt any acute loss of respiratory function that may occur as a result of insufficient home treatment resources.

First Visit

Our patients are initially assessed for spirometry, maximal inspiratory, and maximal expiratory pressures to assess the degree of respiratory muscle impairment. Medicare guidelines dictate that NIPPV cannot be initiated before the forced vital capacity (FVC) is below 50 percent or the maximal inspiratory pressure is below 60 cm H2O. The initial evaluation includes pre-and post-bronchodilator assessment to identify all reversible causes for improving lung function. Pulmonary function results are reviewed with the patient and their candidacy for NIPPV is determined.

Additionally, cough assist and suction devices are also discussed. Patients are counseled about percutaneous enterogastrostomy (PEG), or feeding tubes, and their wishes concerning the need for mechanical ventilation. Overall efforts to minimize any chance of aspiration or respiratory failure are essential in patient care.

If the patient qualifies for NIPPV, we exclusively employ average volume assured pressure support (AVAPS) in an effort to maximally support those with neuromuscular weakness non-invasively. A "sip and puff feature" provides a convenient mouthpiece for daytime and as needed AVAPS use without requiring a traditional face mask and head gear. If the patient has short-term daytime needs for NIPPV, a brief rest on ventilator support can easily be achieved with "sip and puff." Additionally, if a patient qualifies for AVAPS support, we simultaneously order a cough assist and suction device for home delivery. Technical support is provided by sleep laboratory staff and the individual vendor to educate and aid our patients during all stages of NIPPV support. Whenever possible, family members and caregivers are included in the discussions about home care and ventilator support to minimize acute respiratory events.

AVAPS Initiation

Those that qualify are assessed for the correct AVAPS settings, ideally in a supervised overnight lab assessment in our sleep lab, or while awake if daytime assessment of AVAPS is needed. Patients who are in need of an urgent start of NIPPV are prescribed AVAPS by using an industry weight-based formula. AVAPS vendors are instructed to adjust settings to comfort and expedite AVAPS initiation. Patients are then provided an AVAPS unit and instructed to return quarterly with a data download, or computer memory card, to assess effects of AVAPS use at night. The data card is very helpful in identifying patients who are not readily accommodating to AVAPS and is often communicated directly from the vendor to physician for interventions as needed.

Repeat Visits

Spirometry is assessed through repeat visits and the results are tallied in an effort to track any changes in respiratory muscle weakness with neuromuscular disease.

It may be determined that a patient needs AVAPS or they may be alternatively assessed for any slowing in the progression of the disease or improvement in pulmonary function following AVAPS initiation. Our research in NIPPV includes a data download protocol that is designed to better identify complications with NIPPV compliance and to strategize efforts to improve AVAPS compliance and benefits. Patients are frequently reminded that timing, in regard to the placement of PEG feeding tubes, in relation to decrements in lung function and anesthesia risks is complex. Overall, patients are encouraged to receive PEG tubes when relatively stable in order to better tolerate the procedure and avoid respiratory complications that may occur later, after disease progression has severely compromised respiratory function. Patients are followed throughout the course of their illness. If needed, palliative care is incorporated to better suit their needs. Our goal is to provide for all patient needs within the home with high regard for maintaining dignity and autonomy.

The Sleep Disorders Center at the Comprehensive Lung Center is fully dedicated to the care and support of patients with neuromuscular weakness and strongly encourages all those at any stage in neuromuscular weakness to present for evaluation and support.