Update on the respiratory management of patients with chronic neuromuscular disease.

Background: Neuromuscular diseases include a wide range of conditions that may involve potentially life-threatening respiratory complications (infection, respiratory failure).

Surveillance and Pulmonary Function Tests: For patients with neuromuscular diseases, clinical assessment of respiratory function and regular pulmonary function tests are needed to screen for nocturnal respiratory disorders, weakness of the diaphragm and potential restrictive disorders and/or chronic hypercapnic respiratory insufficiency, possibly with cough deficiency. Management of Nocturnal Respiratory Disorders and Chronic Respiratory Failure: Nocturnal respiratory assistance is an important phase of care for nocturnal respiratory disorders and chronic respiratory failure. This may involve continuous positive airway pressure, adaptative servo-ventilation or non-invasive ventilation with a facial or nasal mask. As needed, diurnal assistance may be proposed by mouthpiece ventilation. Should non-invasive ventilation prove insufficient, or if significant swallowing disorders or recurrent bronchial obstruction develop, or in case of prolonged intubation, tracheotomy may be required.

Lower Airway Obstruction: In case of lower airway infection with ineffective cough, physical therapy, associated with air stacking, intermittent positive pressure breathing or mechanical in-exsufflation may be proposed.

Patient-centered Management: Care for swallowing disorders, nutritional counseling (cachexia, obesity), vaccinations and therapeutic education are integral elements of patient-centered management aiming to prevent the negative impact of infection and to manage respiratory failure of chronic neuromuscular disease.

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