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Respiratory Care of the Patient with Duchenne Muscular Dystrophy, ATS Consensus Statement, Approved by the ATS Board of Directors March 2004

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◆ **Consensus Statement Purpose**

This consensus statement was developed by experts in Duchenne-type Muscular Dystrophy (DMD) care to educate all practitioners on an aggressive, supportive approach to the diagnosis and management of respiratory insufficiency in DMD patients. Early and aggressive interventions make respiratory insufficiency a very treatable condition. Many of the statements recommendations can be adapted to the care of patients with other types of neuromuscular diseases.

◆ **Recommendations**

Refer to the text for a complete listing of recommendations, including nutritional support and end of life directives, as well as justifications for each recommendation.

○ **Respiratory Care**

- Children should have at least one visit with pediatric pulmonologist between 4-6 years of age and before confinement to a wheelchair to evaluate baseline pulmonary function testing and evaluate clinical status
- Bi-annual visits to a pulmonologist after confinement to a wheelchair, fall in vital capacity (VC) below 80% and /or once 12 years of age
- Pulmonologist visit every 3-6 months once assisted ventilation or airway clearance is required
- All DMD patients should undergo pulmonary and cardiac evaluations prior to surgery

○ **Routine Evaluation of Respiratory Function**

- Evaluation of oxyhemoglobin saturation by pulse oximetry (SPO₂), spirometric measurements of FVC, FEV₁ and maximal mid-expiratory flow rate, maximal inspiratory and expiratory pressures and peak cough flow on each clinic visit
- Awake CO₂ tension via capnography at least annually in conjunction with spirometry

○ **Sleep Evaluation**

- Review sleep quality and symptoms of sleep disorder breathing at every physician visit
- Annual evaluation for sleep disordered breathing (SDB) via polysomnography with continuous CO₂ monitoring after wheelchair confinement and / or when clinically indicated

○ **Long-term Care Issues and Education**

- Discussions concerning ventilatory support should occur before the need is apparent and should be repeated as the underlying disease progresses.
- Ventilatory support discussions should include patient, caregivers and medical team and all alternative options should be discussed

◆ **Recommendations (continued)**

○ **Airway Clearance**

- Teach aggressive strategies to improve airway clearance early on
- Use assistive cough technologies once peak cough flow is <270 liter per minute or maximal expiratory pressure is <60 cmH₂O or in patients whose history suggests difficulty in airway clearance
- Statement strongly supports use of mechanical insufflation-exsufflation
- Use home pulse oximetry to monitor effectiveness of airway clearance during respiratory illness and to identify need for hospitalization

○ **Noninvasive Nocturnal Ventilation**

- Use Noninvasive Positive Pressure Ventilation (NPPV) to treat sleep related airway obstruction and chronic respiratory insufficiency
- Negative pressure ventilation should be used with caution due to the risk of precipitating upper airway obstruction and hypoxemia
- Do not use oxygen to treat sleep-related hypoventilation without ventilatory assistance
- Assess adequacy of prescribed home ventilatory support via polysomnography and continuous CO₂ monitoring
- Schedule periodic reassessment based on stage of disease.
- Regularly monitor for the development of daytime hypoventilation which may necessitate daytime ventilation

○ **Daytime Noninvasive Ventilation**

- Consider daytime ventilation when awake PaCO₂ > 50 mm Hg or when SPO₂ remains < 92% while awake
- At least annual noninvasive monitoring of CO₂ and oxygen saturation once NPPV is prescribed

○ **Continuous Invasive Ventilation**

- Consider tracheostomy when NPPV is contraindicated, not feasible due to severe bulbar weakness or patient is adverse to noninvasive ventilation
- Monitor oxygen saturation with invasive ventilatory support to detect mucous plugs

◆ **What's New**

- ◆ Sleep quality and symptoms of sleep disordered breathing should be assessed at every physician visit
- ◆ Annual PSG with continuous CO₂ monitoring is indicated once the patient becomes wheelchair-bound or when clinically indicated based on symptoms

◆ **Note**

- ◆ Any form of sleep disorder breathing in DMD patients should be treated with NPPV. CPAP is likely to be of limited value due to the prevalence of nocturnal hypoventilation.
- ◆ Oxygen therapy with concurrent ventilatory support is indicated for the treatment of hypoxemia as hypoxemia in DMD is usually a manifestation of hypoventilation.
- ◆ The guidelines can be adapted for use with other types of neuromuscular diseases.

◆ **Bottom Line**

- ◆ Early intervention, regular diagnostic testing and appropriate use of a full range of respiratory services in the home make the respiratory insufficiency associated with DMD a very treatable condition.