

disease develop abnormal breathing patterns, and hypoxia may occur as a result of inadequate oxygenation. Polysomnography should be performed once daytime symptoms of sleep-disordered breathing occur. The child and parents should be involved in a discussion of long-term ventilation options. Cardiac and respiratory assessment during wake/sleep cycles is imperative. Respiratory care for children with neuromuscular conditions such as SMA and DMD may involve the use of noninvasive ventilation with BiPAP on a temporary or full-time basis, mechanically assisted coughing (MAC), or tracheotomy and relief of airway obstruction with coughing and suctioning devices; the tracheotomy, however, is associated with more complications ([Simonds, 2006](#); [Young, Lowe, Fitzgerald, et al, 2007](#)). Home pulse oximetry may be used to monitor oxygenation during sleep or to aid in decision making regarding the use of MAC to clear the airways.

Several devices are available for children with neuromuscular disease to assist in clearing the airway when the cough reflex is ineffective or diminished. The mechanical in-exsufflator (MIE; also referred to as *cough assist*) has been found to be safe and effective in the daily management of respiratory function ([Kravitz, 2009](#); [Miske, Hickey, Kolb, et al, 2004](#)). The MIE delivers positive inspiratory pressures at a set rate followed by negative pressure exsufflation coordinated with the patient's own breathing rhythm. The exsufflation is designed to mimic a cough reflex so that mucus can be effectively cleared. Airway suctioning after exsufflation is accomplished as necessary to clear the airways. In children, the MIE device may be connected directly to a tracheostomy or used with a mouthpiece or face mask. [Boitano's \(2009\)](#) article provides a variety of equipment options, including various masks that can be used to deliver noninvasive positive pressure.

Manual cough-assisting techniques include glossopharyngeal breathing or air stacking (frog breathing); the abdominal thrust, which is similar to the Heimlich maneuver ([Kravitz, 2009](#)); and manual hyperinflation with a self-inflating resuscitation bag (without oxygen) and a mouthpiece. Hyperinflation may be used in conjunction with abdominal thrusts to improve peak cough flows ([Boitano, 2009](#)).

The use of routine chest physiotherapy (postural drainage) for DMD has not been adequately evaluated for its effectiveness in