The American College of Chest Physicians Consensus Statement on the Respiratory and Related Management of Patients With Duchenne Muscular Dystrophy Undergoing Anesthesia or Sedation*  

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ABSTRACT  
This is a summary of the presentation on the American College of Chest Physicians consensus statement on the respiratory and related management of patients with Duchenne muscular dystrophy undergoing sedation or anesthesia, presented as part of the program on pulmonary management of pediatric patients with neuromuscular disorders at the 30th Annual Carrell-Krusen Neuromuscular Symposium on February 20, 2008. Pediatrics 2009;123:S242–S244

The consensus statement on the respiratory and related management of patients with Duchenne muscular dystrophy (DMD) undergoing anesthesia or sedation was based on work performed by a multidisciplinary panel of experts convened by the American College of Chest Physicians.1 Permission was obtained from the journal Chest to present the contents of the statement at the Carrell-Krusen meeting and to submit this summary document for publication in Pediatrics. The statement’s writing panel consisted of specialists in the areas of anesthesiology, critical care medicine, neurology, orthopedic surgery, pediatric and adult pulmonology, and respiratory therapy. The medical literature was reviewed by querying PubMed (www.pubmed.gov), a service of the National Library of Medicine and the National Institutes of Health. Consensus of recommendations was achieved by majority vote, and there were no disagreements regarding the final recommendations.

The need for this statement derives from the fact that persons with DMD are at high risk of major medical complications when they undergo general anesthesia (GA) or procedural sedation (PS).2 In addition, survival has reached unprecedented levels among people with DMD, requiring them to undergo more procedures involving GA and PS.3

DMD causes weakness of the respiratory muscles, resulting in impaired cough and a progressive decrease in total lung capacity and vital capacity, which makes patients especially vulnerable to the adverse physiologic effects of GA on the respiratory system.4 Airway patency can be compromised during PS or GA, which makes respiratory management complex.

The American College of Chest Physicians designated these recommendations to not be used for performance measurement or for competency purposes, because they are not evidence based.

PREOPERATIVE ASSESSMENT AND MANAGEMENT

For preoperative assessment, the panel chose tests that are widely available (avoiding the need for special equipment), objective and reproducible, simple to understand, and easy to measure (avoiding the need for specialized personnel). The suggested preoperative respiratory tests assess gas exchange, lung volume, and cough effectiveness. To assess gas exchange, the panel advised measuring the patient’s oxyhemoglobin saturation in room air by using pulse oximetry (SpO₂). SpO₂ at <95% in room air was defined as a clinically significant abnormality, instigating the need for measurement of the patient’s blood and/or end-tidal carbon dioxide levels. This SpO₂ threshold level was chosen on the basis of consensus opinion and the threshold level identified by Bach et al5 in their protocols for prevention of pulmonary morbidity in patients with neuromuscular diseases. To assess lung volume, the panel advised measuring the patient’s preoperative forced vital capacity (FVC) with the patient in the seated, upright body position. FVC is simple to measure and highly reproducible and, thus, was chosen in preference to more specialized measurements of lung volume. FVC was also chosen because low levels have been shown to predict respiratory complications in persons with DMD.6,7 Therefore, a 2-tier risk-assessment method was devised to incorporate these FVC threshold levels. The panel’s consensus opinion was that patients with DMD are at increased risk of complica-
tions of GA and PS if preoperative vital capacity is <50% of that predicted and at high risk of complications if preoperative FVC is <30% of that predicted. These threshold values are used to identify patients with DMD at risk of postoperative respiratory complications; patients who will benefit from preoperative training in noninvasive positive-pressure ventilation (NPPV) to optimize the success of postoperative extubation facilitated by NPPV, and patients who need NPPV during induction of and recovery from GA throughout PS. This 2-tiered risk-assessment analyzer is one of the innovations of the consensus statement, as was the emphasis on a proactive approach to the use of assisted ventilation during recovery from GA and throughout PS in patients with DMD and low baseline pulmonary function. This strategy has the potential to be applied to patients with chronic respiratory insufficiency resulting from diagnoses other than DMD when such patients require GA or PS.

In the consensus statement, the authors advocated preoperative assessment of cough strength and effectiveness by measuring maximum expiratory pressure (MEP) and peak cough flow (PCF). For patients with a preoperative PCF of <270 L/minute or MEP of <60 cm H2O, the panel advocated preoperative training in and postoperative use of manual and mechanically assisted cough (ie, mechanical insufflation/exsufflation with the CoughAssist device [Respironics, Murrysville, PA]). The recommended preoperative evaluation also includes cardiac consultation, nutritional and gastrointestinal evaluation, and discussions with patients and their families regarding their attitudes toward prolonged mechanical ventilator dependency, tracheostomy, and advance directives.

INTRAOPERATIVE MANAGEMENT

The intraoperative use of depolarizing muscle relaxants or neuromuscular blockers, such as succinylcholine, is absolutely contraindicated in persons with DMD because of the risk of rhabdomyolysis, hyperkalemia, and cardiac arrest.

Inhalational anesthetics are associated with the risk of malignant hyperthermia-like reactions in persons with DMD, with the potential for cardiac arrest and sudden death. Thus, the clear trend is away from the use of inhaled anesthetics for persons with DMD, and a total intravenous anesthetic technique was advocated in the consensus statement.

To minimize the risk of cardiorespiratory complications, the authors also advocated the presence of an anesthesiologist when patients with DMD undergo PS, with full monitoring and safety measures adhering to the guidelines of the American Academy of Pediatrics and the American Society of Anesthesiologists. Intraoperatively, Spo2 should be monitored continuously, and whenever possible, end-tidal or blood carbon dioxide levels should be measured. Medical procedures should be performed in the optimal setting and with the presence of a full complement of skilled personnel, including a respiratory therapist skilled in the management of NPPV. An ICU should be available for postoperative monitoring and medical management.

Options for respiratory support during maintenance of GA or PS include endotracheal intubation, with use of NPPV to facilitate extubation for selected patients; use of the laryngeal mask airway; and use of mechanical or manual NPPV via lip seal, face mask, or nasal interface.

The statement’s authors identified patients with FVC of <50% of that predicted as being at risk and patients with FVC of <30% of that predicted as being at high risk of needing assisted ventilation during induction of and recovery from GA and throughout PS.

POSTOPERATIVE ASSESSMENT AND MANAGEMENT

The statement advocated that, for patients with DMD who have been intubated for GA or PS, extubation to NPPV should be considered if the patient has a preoperative FVC of <50% of that predicted and strongly considered for patients with preoperative FVC of <30% of that predicted. Patients who use NPPV chronically should also be extubated to NPPV postoperatively. Continuous NPPV can then be weaned off or back to baseline hours of use per day. Patients with subthreshold FVC whose intraoperative respiratory support was accomplished by using the laryngeal mask airway or with NPPV should also use NPPV postoperatively. Other recommendations included delaying extubation until respiratory secretions are in control andSpo2 is normal or baseline in room air; extubation to the patient’s usual home NPPV interface whenever possible; and cautious use of supplemental oxygen postoperatively, because oxygen therapy can mask hypoxemia caused by atelectasis, hypoventilation, or airway secretions without treating the underlying cause.

The authors advised that patients with preoperative MEP of <60 cm H2O or PCF at <270 L/minute should use manual and mechanically assisted cough postoperatively. Mechanical insufflation/exsufflation can augment cough, promote deep lung inflation, and treat or prevent atelectasis. Mechanical insufflation/exsufflation can be applied via the endotracheal tube before extubation.

It is important to provide patients with adequate pain control. If sedation occurs as a result of the use of narcotics, extubation can be delayed, or NPPV can be used continuously to provide respiratory assistance. When possible, it is desirable to avoid GA when patients with low pulmonary function require procedures. For example, neuraxial techniques (eg, epidural catheters) can be used in selected patients to achieve pain control while minimizing sedation and respiratory depression. Patients with DMD are at risk of postoperative congestive heart failure, dysrhythmias, and inadequate cardiac output because of fluid shifts caused by transfusions and intravenous fluid administration. Consider postoperative cardiology consultation, as well as attention to fluid balance and intensive cardiopulmonary monitoring.

Gastrointestinal, nutritional, and pulmonary management are closely interrelated. Bowel regimens should be initiated to treat or avoid constipation and associated inhibition of diaphragmatic excursion. Consider the use of prokinetic gastrointestinal medications and gastric decompression with a nasogastric tube for patients with gut dysmotility to avoid the adverse effects of gastric insufflation on the diaphragm. Parenteral nutrition or
enteral feeding via a small-diameter tube early in the postoperative course is advised to avoid malnutrition and associated respiratory muscle weakness.

The consensus statement identified areas in need for future research, because this entire area is lacking in controlled, randomized studies.

REFERENCES

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