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Special Feature

CONSENSUS STATEMENT

# 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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**This statement on the management of patients with Duchenne muscular dystrophy (DMD) undergoing procedural sedation or general anesthesia represents the consensus opinion of a multidisciplinary panel convened under the auspices of the American College of Chest Physicians. Expert recommendations on this subject are needed for several reasons. First, patients with DMD have an increased risk of complications when they undergo sedation or general anesthesia. In addition, due to improved cardiopulmonary therapies, patients with DMD are experiencing an unprecedented duration of survival. As a result, it is more common for them to require procedures involving sedation or general anesthesia. The risks related to anesthesia and sedation for DMD patients include potentially fatal reactions to inhaled anesthetics and certain muscle relaxants, upper airway obstruction, hypoventilation, atelectasis, congestive heart failure, cardiac dysrhythmias, respiratory failure, and difficulty weaning from mechanical ventilation. This statement includes advice regarding the highly interrelated areas of respiratory, cardiac, GI, and anesthetic management of patients with DMD undergoing general anesthesia or procedural sedation. The statement is intended to aid clinicians involved in the care of patients with DMD and to be a resource for other stakeholders in this field, including patients and their families. It is an up-to-date summary of medical literature regarding this topic and identifies areas in need of future research.** ***(CHEST 2007; 132:1977–1986)***

**Key words:** anesthesia; consensus statement; Duchenne muscular dystrophy; mechanical insufflation-exsufflation; neuromuscular diseases; noninvasive ventilation; sedation

**Abbreviations:** ACCP = American College of Chest Physicians; DMD = Duchenne muscular dystrophy; MEP = maximum expiratory pressure; MI-E = mechanical insufflation-exsufflation; MIP = maximum inspiratory pressure; NPPV = noninvasive positive pressure ventilation; PCF = peak cough flow; $Spo_2$ = oxyhemoglobin saturation measured by pulse oximetry

## Executive Summary

Duchenne muscular dystrophy (DMD) is a progressive neuromuscular disease transmitted by X-linked inheritance with an incidence of approximately 1 in 3,500 live male births. DMD affects the muscles of respiration and is associated with dilated cardiomyopathy, which often leads to death from cardiopulmonary causes. Patients with DMD are especially vulnerable to the adverse physiologic effects of general anesthesia and procedural sedation, prompting the need for expert recommendations on this topic. This consensus statement is the product of a panel convened under the auspices of the American College of Chest Physicians' (ACCP) Pediatric Chest Medicine and Home Care NetWorks. The

panel consisted of specialists in the areas of anesthesiology, critical care medicine, neurology, orthopedic surgery, pediatric and adult pulmonology, and respiratory therapy. The most current and relevant medical literature was identified and reviewed, obtained by querying PubMed, a service of the National Library of Medicine and the National Institutes of Health (www.pubmed.gov), which includes the MEDLINE database. Consensus of recommendations was achieved through a majority vote of the panel members, and there were no disagreements on any of the recommendations. The purposes of this statement are to aid clinicians involved in the care of patients with DMD undergoing procedures requiring sedation or general anesthesia, to be a resource for other stakeholders in this field, including patients and their families, for use as an up-to-date summary of medical literature on this topic, and to identify areas in need of future research. The statement is divided into sections on the assessment and management of patients before, during, and after procedural sedation or general anesthesia.

*Summary of Specific Suggestions for Evaluation and Management of Patients With DMD Before General Anesthesia or Procedural Sedation*

1. Obtain anesthesiology and pulmonology consultations before procedures involving general anesthesia or procedural sedation.
2. Perform a pulmonary evaluation that includes measurement of FVC, maximum inspiratory pressure (MIP), maximum expiratory pressure (MEP), peak cough flow (PCF), and oxyhemoglobin saturation measured by pulse oximetry ($Spo_2$) in room air. Measure the blood and/or end-tidal carbon dioxide level if $Spo_2$ is $< 95\%$ in room air. For DMD patients at increased risk of respiratory complications, defined by an FVC $< 50\%$ of predicted, and especially for patients at high risk for complications, defined by an FVC $< 30\%$ of predicted, consider preoperative training in the use of noninvasive positive pressure ventilation (NPPV). For patients at high risk of ineffective cough, defined in adults by PCF $< 270$ L/min or MEP $< 60$ cm $H_2O$, consider preoperative training in manual and mechanically assisted cough, emphasizing use of mechanical insufflation-exsufflation (MI-E) with a bronchial secretion clearance device (CoughAssist; Respironics; Murrysville, PA).
3. Refer the patient to a cardiologist for clinical evaluation and optimization of cardiac therapies.
4. Obtain a nutritional assessment, optimize nutritional status, and consider strategies to manage dysphagia.
5. Discuss the risks and benefits of general anesthesia or procedural sedation with the patient and guardians, and help them to decide on and implement their decisions regarding resuscitation parameters and, if applicable, advance directives.

*Summary of Specific Suggestions for Management of Patients With DMD During General Anesthesia or Procedural Sedation*

1. Consider use of a total IV anesthesia technique for induction and maintenance of general anesthesia (*eg*, propofol and short-acting opioids). The use of depolarizing muscle relaxants such as succinylcholine is absolutely contraindicated because of the risk of fatal reactions.
2. Optimize the medical setting and personnel in attendance when DMD patients undergo general anesthesia or procedural sedation, and have an ICU available for postprocedure care.
3. Options for providing respiratory support during maintenance of general anesthesia or procedural sedation for patients with DMD include endotracheal intubation, with use of NPPV to facilitate extubation for selected patients; use of the laryngeal mask airway; mechanical ventilation via a mouthpiece with leak-proof seal; and manual or mechanical ventilation (using conventional ventilators or bilevel positive pressure ventilators designed for noninvasive respiratory support) delivered via a full face mask or nasal mask interface.
4. Application of ventilation in the assisted or controlled modes should be considered for patients with DMD and an FVC $< 50\%$ of

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predicted, and strongly considered for those with an FVC < 30% of predicted, during induction of and recovery from general anesthesia and throughout procedural sedation. Options for respiratory support during induction of and recovery from general anesthesia or procedural sedation include manual ventilation using a flow-inflated manual resuscitation bag (standard "anesthesia bag") with a full face or nasal mask interface, and mechanical support using a conventional or noninvasive positive pressure ventilator via a full face or nasal mask.
5. Intraoperatively, monitor $Spo_2$ continuously and, whenever possible, blood or end-tidal carbon dioxide levels.

*Summary of Specific Suggestions for Management of Patients With DMD After General Anesthesia or Procedural Sedation*

1. Consider extubating DMD patients with FVC < 50% of predicted, and especially those with FVC < 30% of predicted, directly to NPPV. Consider delaying extubation until respiratory secretions are well controlled and $Spo_2$ is normal or baseline in room air. Continuous use of NPPV can then be weaned as tolerated. When applicable, try to utilize the patient's home interface after extubation.
2. Use supplemental oxygen therapy cautiously. Monitor $Spo_2$ continuously after general anesthesia or procedural sedation. Whenever possible, monitor blood or end-tidal carbon dioxide levels. Assess if hypoxemia is due to hypoventilation, atelectasis, or airway secretions, and treat appropriately.
3. Use manually assisted cough and MI-E postoperatively in DMD patients with impaired cough, defined in adults as PCF < 270 L/min or MEP < 60 cm $H_2O$.
4. Optimize postoperative pain control in patients with DMD. If sedation and/or hypoventilation occurs, delay endotracheal extubation for 24 to 48 h or use NPPV.
5. Obtain a cardiology consultation and closely monitor cardiac and fluid status postoperatively.
6. Initiate bowel regimens to avoid and treat constipation and consider prokinetic GI medications. Consider gastric decompression with a nasogastric tube in patients with GI dysmotility. Start parenteral nutrition or enteral feeding via a small-diameter tube if oral feeding is delayed for > 24 to 48 h postoperatively.

*Areas in Need of Further Study*

The statement identifies specific areas for future research.

## Background

DMD is a progressive neuromuscular disease transmitted by X-linked inheritance with an incidence of approximately 1 in 3,500 live male births. DMD affects the muscles of respiration and is associated with dilated cardiomyopathy, which often leads to death from cardiopulmonary causes. With current medical management, including the use of nocturnally assisted ventilation, mean survival now approximates 25 years.[1] Patients with DMD are especially vulnerable to the adverse physiologic effects of general anesthesia and procedural sedation.[2–4] For example, DMD patients have macroglossia[2] and weak upper airway dilator muscles. Sedation and general anesthesia cause relaxation of these muscles, which predisposes to upper airway obstruction. Additionally, DMD patients may have limited mobility of the mandible and cervical spine, which impedes maneuvers (such as the "jaw thrust") that restore upper airway patency. General anesthesia results in decreased functional residual capacity, which can cause lower airway closure, atelectasis, and rapid deterioration in gas exchange. Hypercapnia and hypoxemia can worsen chronic cardiopulmonary abnormalities found in some patients with DMD, such as pulmonary hypertension or cardiac conduction defects.

This statement arose from two primary considerations. First, patients with DMD and impaired pulmonary function are at high risk for death when they undergo procedures requiring sedation or general anesthesia.[2] Second, because survival of patients with DMD has reached an unprecedented level due to contemporary cardiorespiratory management, there is an increased need for these patients to undergo procedures.[5] The purposes of this statement are to aid clinicians involved in the care of patients with DMD who undergo procedures requiring sedation or general anesthesia; to be a resource for other stakeholders in this field, including patients and their families; for use as an up-to-date summary of medical literature on this topic; and to identify areas in need of future research.

While the primary purpose of this statement is to improve the management and outcomes of patients with DMD who undergo procedural sedation or general anesthesia, the committee wishes to emphasize several related points. First, that efforts should be made to optimize the timing of surgeries and procedures such as scoliosis surgery or gastrostomy placement so that patients undergo procedural sedation or general anesthesia as early in the disease course as possible, preferably when cardiopulmonary function is preserved.[6] Second, that the increasing use of glucocorticoids to treat patients with DMD

may preserve respiratory muscle strength[7] and decrease the need for procedures such as scoliosis surgery[8] and gastrostomy placement, with the potential to decrease the risk of death due to anesthesia and sedation. Finally, whenever possible, alternatives to general anesthesia and procedural sedation should be considered in patients with DMD who require procedures, such as the use of local anesthetics for pain control.[9]

## Methodology and Structure

This consensus statement is the product of a panel convened under the auspices of the ACCP Pediatric Chest Medicine and Home Care NetWorks. The panel consists of specialists in the areas of anesthesiology, critical care medicine, neurology, orthopedic surgery, pediatric and adult pulmonology, and respiratory therapy. The panel worked on this project from January 2006 to January 2007, primarily via telephone conference calls. The authors disclosed any conflicts of interest and were given complete autonomy by the ACCP. The panel was divided into working groups through which the most current and relevant medical literature was identified and reviewed, obtained by querying PubMed, a service of the National Library of Medicine and the National Institutes of Health (www.pubmed.gov), which includes the MEDLINE database. Only articles written in English were considered. There are few randomized, controlled trials involving the subject of this statement, so this document is a consensus statement derived from expert opinion rather than an evidence-based guideline. Consensus of recommendations was achieved through a majority vote of the panel members, and there were no disagreements on any of the recommendations. The statement is divided into sections on the assessment and management of patients before, during, and after procedural sedation or general anesthesia. Each section consists of a review of the subtopic, followed by a list of specific suggestions. The ACCP Health and Science Policy Committee designates that these recommendations should not be used for performance measurement or for competency purposes because they are not evidence based.

## I. Respiratory Support and Related Medical Management Before General Anesthesia or Procedural Sedation (Preoperative)

### *A. Pulmonary Assessment*

DMD is characterized by weakness of the diaphragm, intercostal muscles, and the accessory muscles of respiration, resulting in restrictive pulmonary impairment and a progressive decrease in total lung capacity and vital capacity.[10,11] These abnormalities lead to hypoventilation and impaired cough, which predisposes to atelectasis and respiratory failure. Before patients with DMD receive general anesthesia or procedural sedation, they should undergo measurement of $Spo_2$ in room air, and measurement of the patient's blood and/or end-tidal carbon dioxide level should be done if $Spo_2$ is $< 95\%$ in room air. Additionally, DMD patients should undergo measurement of the following lung function parameters to assess their risk of respiratory complications and need for perioperative and postoperative assisted ventilation or cough.

*FVC:* FVC is the pulmonary function parameter most frequently reported to have predictive value in assessing the risk of respiratory complications for patients with DMD. To determine predicted values, arm span is usually used to estimate height for patients in a wheelchair, or predictive equations based on ulnar length can be used.[12] The FVC is usually measured with the patient in a seated, upright body position. FVC $< 30\%$ of predicted has been identified as a predictor of postoperative respiratory complications and the need for postoperative ventilatory assistance among DMD patients undergoing spinal fusion surgery.[13–15] However, studies[16–18] suggest that the risks associated with spinal fusion surgery among DMD patients with FVC $< 30\%$ of predicted can be greatly reduced by facilitating postoperative extubation with NPPV and by using MI-E to assist with cough. Additionally, percutaneous endoscopic gastrostomy placement has been accomplished in DMD patients with vital capacity well $< 30\%$ of predicted through the use of NPPV during induction of and recovery from anesthesia.[19] A previous consensus conference report[20] recommended NPPV for patients with progressive neuromuscular disease and FVC $< 50\%$ of predicted and symptoms of hypoventilation. It is the consensus of this panel that DMD patients with FVC $< 50\%$ of predicted measured in the seated, upright body position are at increased risk for respiratory complications when they undergo general anesthesia or procedural sedation, and that patients with FVC $< 30\%$ of predicted are at high risk for complications. While diaphragm strength can be relatively preserved in DMD,[10,11] patients are often confined to the supine body position during and after surgery. Thus, measurement of both upright and supine FVC may be useful because patients with poor supine FVC values will be especially vulnerable to postoperative atelectasis and hypoxemia.

*MIP, MEP, and PCF:* MIP and MEP are used to assess respiratory muscle strength, and they have clinical utility in DMD.[4,10] Another useful preoperative test is the measurement of PCF.[21] Impairment of these parameters reflects an inability to generate the cough force and velocity necessary for effective clearance of respiratory secretions.[4,22] Patients with a tracheostomy tube and assisted PCF $< 160$ L/min are at increased risk for failure to achieve tracheostomy tube decannulation, and adult DMD patients who cannot generate a PCF $\geq 270$ L/min are at increased risk for pneumonia or atelectasis.[23–25] Moreover, young adults with DMD and MEP $< 60$ cm $H_2O$ are likely to have ineffective cough.[26] Therefore, it is the consensus of the panel that DMD patients with PCF $< 270$ L/min or MEP $< 60$ cm $H_2O$ are at increased risk for respiratory complications when they undergo procedural sedation or general anesthesia due to impaired cough. The data used to determine these threshold values were obtained from teenage and adult patients, and the values of MEP or PCF that predict increased risk of impaired cough in young children are unknown.

### *B. Preprocedure Initiation of Noninvasive Respiratory Aids*

It is our consensus opinion that patients with DMD and FVC $< 50\%$ of predicted, and especially those with FVC $< 30\%$ of predicted, should be considered for preoperative training in the use of NPPV due to their increased risk of respiratory complications. Preoperative training in NPPV should increase the probability of successful use of NPPV during recovery from general anesthesia or sedation and at postoperative endotracheal extubation (see sections II and III below).[4,6,18,27,28] Similarly, adult patients with PCF $< 270$ L/min or MEP $< 60$ cm $H_2O$ are at risk for ineffective cough, and preoperative training in manual and mechanically assisted cough (MI-E with a bronchial secretion clearance device) [CoughAssist; Respironics; Murrysville, PA]) is suggested, using the techniques described in the referenced articles.[24,27,29]

### *C. Cardiac Assessment*

DMD is associated with the development of dilated hypertrophic cardiomyopathy and cardiac dysrhythmias. Patients with DMD are at high risk for perioperative cardiac side effects due to hypoxemia, anemia, and other causes of impaired tissue oxygen delivery. Intravascular fluid shifts can result in congestive heart failure and impaired ventricular preload. These issues are reviewed in the recent consensus statement[30] of an expert panel convened by the American Academy of Pediatrics that states that patients with DMD should undergo a cardiac evaluation and optimization of cardiac therapies before anesthesia. Preoperative consultation with a cardiologist is advised for all patients with DMD because heart disease can be severe even among patients with only mild pulmonary involvement, and normal preoperative ECG and echocardiogram findings do not exclude the possibility of postoperative cardiac complications.[31]

### *D. Nutrition and GI Issues*

Good nutritional support is integral to the proper care of patients with DMD, and the adverse effects of malnutrition on respiratory muscle strength can be profound.[4] Therefore, preoperative nutritional status should be evaluated and optimized because poor nutrition can increase postoperative morbidity.[32,33] Optimization of preoperative nutritional status may involve the use of NPPV because patients with untreated respiratory failure may become malnourished due to increased work of breathing, or they may be unable to eat due to dyspnea. The preoperative evaluation should include measurements of serum albumin and prealbumin to identify patients who are at risk for poor healing. In addition, preoperative evaluation and therapy for dysphagia should be considered because loss of the ability to eat postoperatively can lead to malnutrition.[32]

### *E. Advance Directives*

DMD is a progressive and potentially fatal disease. Thus, advance directives (including resuscitation parameters) and attitudes toward prolonged dependency on mechanical ventilation and tracheostomy should be discussed with DMD patients and their guardians preoperatively. Furthermore, decisions regarding these issues should be clearly articulated and easily accessible in the medical record.

### *Summary of Specific Suggestions for Evaluation and Management of Patients With DMD Before General Anesthesia or Procedural Sedation*

1. Obtain anesthesiology and pulmonology consultations before procedures involving general anesthesia or procedural sedation.
2. Perform a pulmonary evaluation that includes measurement of FVC, MIP, MEP, PCF, and $Spo_2$ in room air. Measure the patient's blood and/or end-tidal carbon dioxide level if $Spo_2$ is $< 95\%$ in room air. For DMD patients at increased risk for respiratory complications, defined as FVC $< 50\%$ of predicted, and especially for patients at high risk for complications, defined as FVC $< 30\%$ of predicted, consider

preoperative training in the use of NPPV. For patients at high risk for ineffective cough, defined in adults as PCF $<$ 270 L/min or MEP $<$ 60 cm $H_2O$, consider preoperative training in manual and mechanically assisted cough, emphasizing the use of MI-E with a bronchial secretion clearance device.

3. Refer the patient to a cardiologist for clinical evaluation and optimization of cardiac therapies.
4. Obtain a nutritional assessment, optimize nutritional status, and consider strategies to manage dysphagia.
5. Discuss the risks and benefits of general anesthesia or procedural sedation with the patient and guardians, and help them to decide on and implement their decisions regarding resuscitation parameters and, if applicable, advance directives.

## II. Respiratory Support and Related Medical Management During General Anesthesia or Procedural Sedation (Intraoperative)

### *A. Choice of Pharmacologic Agents for General Anesthesia*

Patients with DMD are at increased risk for extreme hyperthermic events and rhabdomyolysis when they are exposed to certain anesthetics, especially inhaled agents such as halothane, isofluorane, and servoflurane. Such episodes can cause hyperkalemia and sudden death from cardiac arrest.[2,34] These events mimic malignant hyperthermia, but DMD and malignant hyperthermia are genetically distinct diseases. Succinylcholine, a depolarizing muscle relaxant that can disrupt unstable cell membranes, has been linked to acute rhabdomyolysis, hyperkalemia, and cardiac arrest in patients with DMD.[35] Indeed, there are numerous reports of young patients in whom previously unsuspected DMD was diagnosed after sudden death due to hyperkalemic cardiac arrest associated with general anesthesia.[36] While succinylcholine is widely recognized to be contraindicated in patients with DMD, more recently it has been suggested that inhaled anesthetic agents should also be considered contraindicated for patients with DMD.[34]

### *B. Choice of Personnel and Medical Setting*

Procedural sedation should be performed with an anesthesiologist in attendance and with full monitors and safety measures, according to the guidelines of the American Academy of Pediatrics and the American Society of Anesthesiologists.[37,38] Intraoperatively, monitor $Spo_2$ continuously and, whenever possible, blood or end-tidal carbon dioxide levels. Medical procedures involving procedural sedation or general anesthesia should be performed in the optimal medical setting (*eg*, postanesthetic care unit or operating room) and with a full complement of skilled personnel (*eg*, an anesthesiologist experienced in the management of DMD and a respiratory therapist skilled in the management of NPPV) in order to minimize the risk of respiratory complications.[6] An ICU should be available for postprocedure management.[28]

### *C. Respiratory Support Options During Maintenance of General Anesthesia or Procedural Sedation*

The options for respiratory support during maintenance of general anesthesia or procedural sedation will depend on the nature of the procedure and the type of anesthetic used (*eg*, IV vs inhaled). Options for respiratory support include endotracheal intubation, using NPPV to facilitate extubation for selected patients; use of the laryngeal mask airway; mechanical ventilation via a mouthpiece with a leak-proof seal; and manual or mechanical ventilation using either conventional ventilators or bilevel positive pressure ventilators designed for noninvasive respiratory support, delivered via a full face mask or nasal mask interface.[6,17,19,39–45]

### *D. Respiratory Support Options During Induction of and Recovery From General Anesthesia or Procedural Sedation*

While it is standard practice to provide assisted or controlled ventilation during induction of general anesthesia, DMD patients with chronic respiratory insufficiency and limited respiratory reserve will also benefit from respiratory support during recovery from general anesthesia and throughout procedural sedation. Options for respiratory support during induction of and recovery from general anesthesia or procedural sedation include manual ventilation using a flow-inflated manual resuscitation bag (standard anesthesia bag) with a full face or nasal mask interface, and mechanical ventilation using a conventional or bilevel positive pressure ventilator designed for noninvasive respiratory support, also delivered via a full face or nasal mask. Patients who have been intubated for procedures can be extubated directly to NPPV as needed (see section III, A, below).[3,6,17–19,27,28,44,46] DMD patients with FVC $<$ 50% of predicted should be considered at increased risk, and those with FVC $<$ 30% of predicted should be considered at high risk of needing

assisted or controlled ventilation during induction of and recovery from general anesthesia and throughout procedural sedation.

*Summary of Specific Suggestions for Evaluation and Management of Patients With DMD During General Anesthesia or Procedural Sedation*

1. Consider use of a total IV anesthesia technique for induction and maintenance of general anesthesia (*eg*, propofol and short-acting opioids). The use of depolarizing muscle relaxants such as succinylcholine is absolutely contraindicated because of the risk of fatal reactions.
2. Optimize the medical setting and personnel in attendance when DMD patients undergo general anesthesia or procedural sedation, and have an ICU available for postprocedure care. Intraoperatively, monitor $Spo_2$ continuously and, whenever possible, blood or end-tidal carbon dioxide levels.
3. There are several options for providing respiratory support during maintenance of general anesthesia or procedural sedation for patients with DMD. These options are outlined in Section II, C, above.
4. Application of assisted or controlled ventilation should be considered for patients with DMD and an FVC $< 50\%$ predicted, and strongly considered for those with an FVC $< 30\%$ predicted, during induction of and recovery from general anesthesia and throughout procedural sedation, using the options for respiratory support outlined in Section II, D, above.

## III. Respiratory Support and Related Medical Management After General Anesthesia or Procedural Sedation (Postoperative)

### *A. Respiratory Support*

Extubation directly to NPPV should be considered for DMD patients with baseline FVC $< 50\%$ of predicted, and should be strongly considered for those with FVC $< 30\%$ of predicted who have been endotracheally intubated for general anesthesia or procedural sedation. Extubation directly to NPPV should also be considered for any patient using NPPV preoperatively (see Section II, D, above).[27,44] Continuous use of NPPV can then be weaned as tolerated,[18] except in patients who require NPPV 24 h/d at baseline. To maximize the chance of success, consider delaying extubation until respiratory secretions are in good control and $Spo_2$ is normal or baseline in room air.[46] If NPPV has been used preoperatively, it is preferable to extubate the patient to NPPV utilizing his usual interface (home mask or mouthpiece) in order to minimize facial skin injury due to poor mask fit, optimize interface comfort, and improve the chance of successful extubation.[6,27] The best medical setting in which to extubate patients depends on the infrastructure and preferences of individual clinicians and their institutions. However, patients who require noninvasive ventilatory support at baseline may tolerate extubation to NPPV best in the ICU rather than in the operating room or postanesthetic care unit because it avoids the risk of transporting the patient to the ICU in a clinically unstable condition.[28] Postoperative use of NPPV should also be considered for DMD patients with baseline FVC $< 50\%$ of predicted, and strongly considered for those with FVC $< 30\%$ of predicted, if procedural respiratory support was accomplished using a laryngeal mask airway or with NPPV (see Section II, C, above).

### *B. Supplemental Oxygen Therapy*

Supplemental oxygen therapy should be used with caution postoperatively in patients with DMD because oxygen therapy can correct hypoxemia without treating the underlying cause (*eg*, hypoventilation or atelectasis) and oxygen therapy may impair central respiratory drive.[46,47] $Spo_2$ should be monitored continuously during and after general anesthesia or procedural sedation until cardiopulmonary status is stable. Whenever possible, assess carbon dioxide levels through blood gas sampling or end-tidal carbon dioxide monitoring by capnography. Assess if hypoxemia is due to hypoventilation, atelectasis, or airway secretions, and treat appropriately.

### *C. Assisted Cough*

Any patient with DMD and evidence of impaired cough (defined, in a teenage or adult patient, by preoperative PCF $< 270$ L/min or MEP $< 60$ cm $H_2O$) will benefit from the use of manually assisted cough maneuvers and the MI-E device postoperatively. The benefits of MI-E include cough augmentation and deep-lung insufflation to treat or prevent atelectasis. MI-E can be useful when pain prevents the patient from coughing spontaneously, such as after surgery on the spine, chest, or abdomen.[24,25,27,29] MI-E can also be used in patients who are still intubated, applied through the endotracheal tube.

### *D. Pain Control*

Adequate postoperative pain control should not be compromised because of concerns about suppression of respiratory drive. When patients are sedated

after administration of opioid analgesics, adequate ventilation can be achieved by using NPPV continuously or by delaying endotracheal extubation for 24 to 48 h. While pain control is essential, the chance of successful extubation is optimized in an awake, cooperative patient. In patients undergoing spinal fusion surgery, neuraxial techniques have been used to achieve analgesia through intermittent or continuous infusion of opioids and/or local anesthetics via epidural catheters, with minimal respiratory side effects.[48]

### *E. Cardiovascular Management*

Patients with DMD are at increased risk for intraoperative and postoperative congestive heart failure and cardiac dysrhythmias, and they have a limited ability to increase cardiac output in response to stress. After IV fluid boluses or blood transfusions, which are often required during spinal fusion procedures and other major surgeries, patients may have intravascular fluid imbalance. These issues necessitate postoperative cardiology consultation, careful attention to fluid balance, and intensive cardiopulmonary monitoring.[30]

### *F. GI and Nutritional Management*

DMD is associated with GI smooth-muscle dysfunction. Postoperatively, DMD patients may have gastroparesis, intestinal dysmotility, and constipation, all of which can be exacerbated by pain medications. GI dysfunction can impair postoperative breathing if distention of the abdomen and increased intraabdominal pressure occur, hampering diaphragmatic excursion. Gut dysmotility also increases the likelihood of gastric distention when NPPV is applied. Thus, gastric decompression may be necessary through placement of a nasogastric tube. Preoperative and postoperative bowel regimens should be employed to avoid and treat constipation,[33] and selected patients may benefit from pharmacologic therapy with GI smooth-muscle prokinetic agents. The inability to take oral nutrition postoperatively can exacerbate malnutrition and muscle weakness.[32] Therefore, any patient with DMD who cannot achieve adequate oral nutrition within 24 to 48 h after surgery should receive enteral feeding with a small-diameter nasogastric or nasoduodenal tube, or should receive parenteral nutrition if ileus is present.

### *Summary of Specific Suggestions for Management of Patients With DMD After General Anesthesia or Procedural Sedation*

1. Consider extubating DMD patients with FVC $< 50\%$ of predicted, and especially those with FVC $< 30\%$ of predicted, directly to NPPV. To optimize the chance of success, consider delaying extubation until respiratory secretions are in good control and $Spo_2$ is normal or baseline in room air. Continuous use of NPPV can then be weaned as tolerated. When applicable, try to utilize the patient's home interface after extubation.
2. Use supplemental oxygen therapy cautiously. Monitor $Spo_2$ continuously during and after general anesthesia or procedural sedation. Whenever possible, assess blood or end-tidal carbon dioxide levels. Assess if hypoxemia is due to hypoventilation, atelectasis, or airway secretions, and treat appropriately.
3. Use manually assisted cough and MI-E postoperatively in DMD patients with impaired cough, defined in a teenage or adult patient as PCF $< 270$ L/min or MEP $< 60$ cm $H_2O$.
4. Optimize postoperative pain control in patients with DMD. If sedation and/or hypoventilation occur, delay endotracheal extubation for 24 to 48 h or use NPPV.
5. Obtain a cardiology consultation and closely monitor cardiac and fluid status postoperatively.
6. Initiate bowel regimens to avoid and treat constipation and consider prokinetic GI medications. Consider gastric decompression with a nasogastric tube in patients with GI dysmotility. Start parenteral nutrition or enteral feeding via a small-diameter tube if oral feeding is delayed for $>$ 24 to 48 h postoperatively.

## IV. Areas in Need of Further Study

The area explored in this consensus statement is characterized by a lack of prospective, randomized studies. Examples of specific studies needed to develop evidence-based guidelines for the care of DMD patients undergoing general anesthesia or procedural sedation include the following: prospective studies that identify the baseline pulmonary function parameters that predict an increased risk of postoperative respiratory complications for DMD patients undergoing different types of procedures; randomized, prospective studies designed to determine which patients with DMD are most likely to benefit from the use of NPPV and/or MI-E to accomplish successful postoperative extubation; studies that assess the benefit, safety, and efficacy of noninvasive methods of respiratory support during maintenance of and recovery from general anesthesia; studies that assess the benefit, safety, and efficacy of the various techniques of assisted cough, including postoperative use of MI-E; studies of the

specific mechanical settings that optimize the efficacy of NPPV and MI-E in patients with DMD; studies that clarify the role of alternative mucus mobilization techniques, such as high-frequency chest wall oscillation and intrapulmonary percussive ventilation; and studies that better define optimal preoperative and perioperative cardiac and nutritional management in DMD patients who require procedures.

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