Lesson 267: PreAnesthetic Assessment of the Patient With Duchenne’s Muscular Dystrophy

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Needs Statement
Anesthesiologists are frequently called on to care for patients with muscular diseases whose condition may be adversely affected by the drugs used for muscle relaxation. The identification of such patients and their appropriate management have been cited by readers and the editorial board committee of Anesthesiology News as necessary information for the practicing anesthesiologist.

Target Audience
Anesthesiologists

Learning Objectives
At the end of this activity, the participant should be able to:
1. Describe the pathophysiology of Duchenne’s muscular dystrophy (DMD).
2. Identify the differences between Becker’s muscular dystrophy and DMD.
3. Discuss the organ systems affected by DMD.
4. List the preoperative considerations for the anesthetic care of the patient with DMD who requires general anesthesia.
5. Outline an appropriate preoperative evaluation.
6. Discuss the controversy surrounding the use of inhalational anesthetics in the management of patients with DMD.
7. Describe the effects of neuromuscular blockade.
8. Develop an anesthetic plan.
9. Manage postoperative anesthetic care.
10. Recognize the progressive nature of the disease.

Case History
A 15-year-old boy with severe scoliosis was admitted for arthrodesis of the spine. The levoscoliosis of his thoracolumbar spine was 58 degrees by the method of Cobb. His medical history was significant for severe DMD associated with congestive heart failure and respiratory insufficiency. The patient’s weight was 81 kg and his height approximately 56 inches, estimated by arm span. A physical examination revealed a hypertrophic tongue and a Mallampati grade II airway. Muscle strength in his lower extremities was documented at 1 to 2 (of 5) on the Medical Research Council Scale. The patient was confined to a wheelchair but was able to brush his teeth and feed himself with difficulty.

Call for Writers
If you would like to write a CME lesson for Anesthesiology News, please send an e-mail to Elizabeth A.M. Frost, MD, at ElzFrost@aoa.com.

Duchenne’s muscular dystrophy (DMD) is the most common muscular dystrophy encountered by anesthesiologists. The disease is characterized by a painless degeneration and atrophy of the skeletal muscles. DMD begins as a disorder of muscle function, but eventually it progresses to a multisystem disease that affects the heart, lungs, and other vital organs. Patients with DMD are considered to be at a high risk for perioperative complications, depending on the degree of involvement of other systems. The action of neuromuscular blocking agents may be altered, and much controversy has surrounded the selection of an appropriate anesthetic technique. It is prudent for anesthesiologists to be familiar with the characteristics of the disease and be prepared to handle the potential complications.

History
DMD was first described by Edward Meryon in 1852; however, the disease is named after Guillaume Benjamin Amand Duchenne, a French neurologist, who studied and defined many neuromuscular diseases in the mid-1800s.

Genetic Considerations
The gene for DMD, located in the p21 region of the X chromosome, encodes the protein dystrophin, which has a molecular weight of 427 kilodaltons. Dystrophin, an...
intracellular protein, forms an interface between cytoskeletal proteins and groups of transmembrane proteins that interact with the extracellular matrix. Dystrophin stabilizes the muscle surface membrane during contraction and relaxation. The dystrophin-associated complex binds intracellular actin to the extracellular basal lamina and mechanistically stabilizes the sarcolemma during muscle contraction (Figure 1). In DMD, the gene product dystrophin is absent or non-functional. In its absence, a sequence of events leads to an influx of calcium into muscle cells—eventually causing them to degenerate and die. Dead myocytes are replaced by connective and adipose tissue, which creates an appearance of muscular hypertrophy. Small, nucleated fibers are often also observed, as a result of muscle regeneration from myoblasts. Skeletal, cardiac, and smooth muscle cells are all affected, causing weakness and hypertrophy of skeletal muscle that can predispose the patient to aspiration.

Course of the Disease

DMD is characterized by a progressive course. Affected children initially behave like healthy babies and toddlers. In affected boys, weakness of the leg and pelvic muscles begins to develop at about the age of 5 years. Children who were formerly able to ambulate easily and maneuver around obstacles exhibit clumsiness, frequent falling, and difficulty climbing stairs. Pseudohypertrophy of the calves, with fatty and fibrous infiltration of the degenerating muscle, is common. While moving from a prone to an upright position, the child exhibits a classic Gower sign, “walking” his hands to, and then up, his legs (Figure 2). The muscles of the thorax and upper limbs are eventually also affected. Biochemical investigations reveal elevated levels of serum creatine phosphokinase and show minimal histologic abnormalities on muscle biopsy. However, in 2.5% of carriers, the clinical picture ranges from a pseudohypertrophy of calf muscles with cramps and mild weakness to a severe disorder that resembles DMD. Among the female population, the prevalence of manifesting carriers is approximately 1 in 100,000.

Becker’s muscular dystrophy, another X-linked disease of dystrophin deficiency, is less common than DMD, occurring in about 1 in 33,000 live births. It is often diagnosed later in life, usually at about the age of 12 years. The clinical course is more benign, although it too includes mild and slowly progressing scoliosis, cardiomyopathy, arrhythmias, and respiratory weakness. It is important to keep Becker’s muscular dystrophy in mind because many of the same anesthetic concerns that arise in DMD also apply. Other muscular dystrophies are briefly described in Table 1.

Of the muscular dystrophies, DMD and Becker’s muscular dystrophy are the most likely to be encountered by the anesthesiologist. The general concerns are similar for all the muscular dystrophies and include cardiomyopathy, respiratory insufficiency, and a weakness of pharyngeal muscle that can predispose the patient to aspiration.

Table 1. Selected Types of Muscular Dystrophy

<table>
<thead>
<tr>
<th>Type</th>
<th>Characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>Becker’s muscular dystrophy</td>
<td>X-linked recessive dystrophin disorder</td>
</tr>
<tr>
<td></td>
<td>Milder than Duchenne’s muscular dystrophy</td>
</tr>
<tr>
<td>Duchenne’s muscular dystrophy</td>
<td>Most common muscular dystrophy</td>
</tr>
<tr>
<td></td>
<td>X-linked recessive dystrophin disorder</td>
</tr>
<tr>
<td></td>
<td>Develops between ages of 3 and 7 years</td>
</tr>
<tr>
<td></td>
<td>Muscle weakness</td>
</tr>
<tr>
<td></td>
<td>Elevated level of creatine phosphokinase</td>
</tr>
<tr>
<td></td>
<td>Pseudohypertrophy of calf muscles</td>
</tr>
<tr>
<td>Emery-Dreifuss dystrophy</td>
<td>X-linked recessive disorder</td>
</tr>
<tr>
<td></td>
<td>Slow progression</td>
</tr>
<tr>
<td></td>
<td>Predominant risk for cardiac involvement,</td>
</tr>
<tr>
<td></td>
<td>with early development of atrial arrhythmia</td>
</tr>
<tr>
<td>Facioscapulohumeral</td>
<td>Autosomal dominant disorder</td>
</tr>
<tr>
<td>dystrophy</td>
<td>Classic symptom of weak pectoral and facial muscles</td>
</tr>
<tr>
<td></td>
<td>Cardiac involvement rare</td>
</tr>
<tr>
<td></td>
<td>Develops between ages of 6 and 12 years</td>
</tr>
<tr>
<td>Mitochondrial myopathy</td>
<td>Inherited as mitochondrial defect from mother; males</td>
</tr>
<tr>
<td></td>
<td>cannot transmit</td>
</tr>
<tr>
<td></td>
<td>Ophthalmoplegia</td>
</tr>
<tr>
<td>Myotonic dystrophy</td>
<td>Autosomal dominant disorder</td>
</tr>
<tr>
<td></td>
<td>Develops between ages of 20 and 30 years</td>
</tr>
<tr>
<td></td>
<td>Classic symptom of myotonia</td>
</tr>
<tr>
<td></td>
<td>Mental retardation may coexist</td>
</tr>
</tbody>
</table>

Adapted from information in reference 1.
deep Q waves in the limb leads. Short PR intervals and sinus tachycardia are also often noted. Cardiac changes parallel the impairment of skeletal muscle.

Clinically significant cardiomyopathy is uncommon before 10 years of age. Patients often have few physical signs, and frequently no symptoms, of cardiac impairment until the left ventricular ejection fraction has decreased to less than 15%, usually at about the age of 18 years. However, more than 50% of patients have some degree of dilated cardiomyopathy with a reduced ejection fraction (<45%) by age 15.

After cell death and subsequent fibrosis in the left ventricle, a dilated cardiomyopathy, decreased cardiac contractility, and concomitant heart failure can develop. Echocardiography at rest, however, may not be sufficiently sensitive to estimate the intraoperative cardiac risk in patients with DMD. Coronary insufficiency may be subclinical because of the patient’s low level of physical activity and exposed only by perioperative stress. In patients with normally functioning skeletal muscle, dilated cardiomyopathy most commonly presents as poor exercise tolerance. Patients with advanced DMD, however, place small demands on the myocardium because of their immobility. They become symptomatic only when myocardial function is severely impaired. It is important for the anesthesiologist to recognize this possibility and request additional preoperative cardiac testing.

Stress echocardiography is proposed to detect latent heart failure and to identify the inducible contraction abnormalities that may arise in many patients with DMD. In addition, right ventricular failure can result from respiratory failure and pulmonary hypertension. Ventricular dysrhythmias are common, as fibrotic tissue displaces the muscle cells in conduction pathways. Because of a protective effect in patients with cardiomyopathies, angiotensin-converting enzyme inhibitors are recommended at the early stages of disease; the administration of β-blockers may be an additional option.

A deterioration of pulmonary function should be documented carefully in patients with DMD and reviewed before any anesthetic is administered. Once patients have been confined to a wheelchair, exhibiting a drop in the vital capacity by approximately 80% of the predicted value or reaching 12 years of age, they should begin twice-yearly visits to a physician specializing in pediatric respiratory care because respiratory muscle strength decreases with age. The decline in pulmonary function is reflected first in decreases in maximal expiratory and inspiratory pressures. Later, a reduction in maximal voluntary ventilation occurs, which is followed by a progressive decrease in the forced vital capacity (FVC). The result is nocturnal and eventually daytime hypventilation, hypoxia, respiratory failure, and possibly respiratory arrest. The rate of decline is variable, but the reduction is relatively consistent over several years. Various levels of impairment have been reported and associated with an increased risk for respiratory complications and death. An FVC of less than 1 L remains the best negative predictor of survival in patients with DMD.

Historically, a preoperative FVC of less than 30% of the predicted value was a contraindication to elective surgery, because the incidence of postoperative pulmonary complications was thought to be very high. Recent studies have questioned that view; a single test is often an insufficient predictor of postoperative outcome, and advances in respiratory management—most importantly the widespread use of NPPV—have resulted in improvements in pulmonary function. NPPV, in the form of continuous or bilevel positive airway pressure, is often initiated in patients between the ages of 10 and 13 years before their FVC decreases dramatically. NPPV is often started when patients exhibit increasingly severe signs and symptoms of hypventilation: decreased FVC, nocturnal hypventilation, deteriorating arterial blood gas values, and scoliosis causing restrictive thoracic disease.

Noninvasive mechanical ventilation is usually initiated in patients who have an arterial oxygen saturation (SaO2) of less than 90% at night but normal daytime blood gas values. Aimed toward the patient’s baseline values, the use of NPPV after surgical intervention improves survival. NPPV can be used as an alternative to invasive ventilation via tracheostomy and to facilitate endotracheal extubation in patients in whom acute respiratory failure has developed. With noninvasive ventilation, the trachea can be extubated earlier, reintubation can be avoided, and the number of ventilator-associated complications can be reduced. With the aggressive use of NPPV, the postoperative course of patients whose FVC is below 30% has been found to be comparable with that of patients whose FVC is above 30%.

Weakness of the expiratory muscles, a common and prominent finding in patients with neuromuscular disease, causes the retention of bronchial mucus, atelectasis, and infection due to an ineffective cough mechanism. In addition to measurement of the FVC, the evaluation of expiratory muscle force may provide important information. Expiratory muscle force may be assessed by measuring the maximal expiratory pressure against an occluded airway after a full inspiration. To generate an effective cough, the maximal expiratory pressure must exceed 60 cm H2O, which may be impossible in cases of severe weakness. Patients may be unable to perform this test after a certain point. An acceptable alternative is measurement of the peak flow and cough peak flow, which may be useful to monitor and assess the progression of expiratory muscle weakness.

### Table 2. Progression of Disease in Patients With Duchenne’s Muscular Dystrophy

<table>
<thead>
<tr>
<th>Age Range</th>
<th>Signs and Symptoms</th>
<th>2-5 y</th>
<th>6-12 y</th>
<th>8-14 y</th>
<th>Adulthood</th>
<th>Late 20s to end of life</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;2 y</td>
<td>Healthy toddler behavior</td>
<td>First outward signs of muscular weakness, clumsiness, frequent falling, waddling gait, difficulty climbing stairs</td>
<td>Walking on toes, which is secondary to tightening of Achilles tendon and compensates for weak quadriceps, weakness of pelvic and shoulder girdles, compensatory lordosis</td>
<td>Loss of ability to walk, decrease in caloric requirements, development of obesity with even a normal diet, development of scoliosis in 95% of patients</td>
<td>Scoliosis, weakened respiratory muscles, inactivity, obesity, compromised lung expansion and function, decrease in vital capacity by approximately 50%</td>
<td>90% respiratory 10% cardiac</td>
</tr>
<tr>
<td>2-5 y</td>
<td>First outward signs of muscular weakness</td>
<td>Calf muscles begin to look enlarged</td>
<td>Weakening of pelvic and shoulder girdles, compensatory lordosis</td>
<td>Loss of ability to walk, decrease in caloric requirements, development of obesity with even a normal diet, development of scoliosis in 95% of patients</td>
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<tr>
<td>6-12 y</td>
<td>Walking on toes, which is secondary to tightening of Achilles tendon and compensates for weak quadriceps, weakness of pelvic and shoulder girdles, compensatory lordosis</td>
<td>Calf muscles begin to look enlarged</td>
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<td></td>
<td>90% respiratory 10% cardiac</td>
</tr>
</tbody>
</table>

Adapted from information in reference 1.
flow values of less than 160 L per minute are associated with ineffective airway clearance and the inability to expel airway mucus and avert respiratory problems. Patients with DMD often display signs of obstructive sleep apnea, in which complete or partial obstruction of the upper airway during sleep leads to oxygen desaturation, hypercarbia, autonomic stimulation, and sleep fragmentation. Children with severe obstructive sleep apnea are at risk for postoperative respiratory compromise, manifested by severe upper airway obstruction that may require endotracheal intubation or the use of noninvasive respiratory support, such as continuous positive airway pressure via a nasal mask. In these patients, respiratory problems are compounded and contribute to the development of pulmonary hypertension. The application of nasal continuous positive airway mucus and avert respiratory problems. 

**Preoperative Concerns**

Patients with muscular dystrophy are very sensitive to the effects of sedative agents. Spontaneous respiratory efforts may be lost after even small doses of medications as upper airway hypotonia quickly leads to airway obstruction. I.V. sedation without assisted ventilation may result in respiratory failure. Intubation is made difficult by the redundant mucosal tissue, stiff jaw, and macroglialosis found in many individuals with muscular dystrophy. A supraglottic airway device should be available as a rescue airway whenever airway manipulation is performed on these patients.

**Intraoperative Concerns: Choice of Agents**

The intraoperative management of patients with DMD is compounded by their increased incidence of cardiovascular problems, including dysrhythmia and congestive heart failure. 

The use of regional anesthesia is not contraindicated; however, neuraxial techniques—such as epidural or spinal anesthesia—may be technically difficult because of kyphoscoliosis and obesity. If a patient has undergone corrective surgery, the placement of orthopedic hardware and the fusion of bone may prevent access to the epidural or spinal space. Peripheral nerve blocks are also not contraindicated. A nerve stimulator may be used to identify the site of injection, because no evidence has been found that stimulation of an isolated muscle group significantly increases the concentration of extracellular potassium. The options for regional or local anesthesia should be considered when appropriate. Regional anesthesia avoids the risk associated with triggering agents and the risk for respiratory depression and other complications that are solely related to general anesthesia.

A malignant hyperthermia-like syndrome has been described with the use of inhalational anesthetics. Although patients with DMD are not predisposed to malignant hyperthermia, several case reports indicate that rhabdomyolysis and hyperkalemia cardiac arrest may nevertheless develop in association with the use of major inhalational agents. What all these cases have in common is cardiac arrest that occurs after an apparently innocuous intraoperative course, including maintenance with a volatile anesthetic, but without succinylcholine. It is thought that the presence of the volatile agent plus movement during recovery may trigger destabilization in the muscle cell membrane, leading to rhabdomyolysis as the muscle depolarizes and contracts.

Currently, there is no clear consensus regarding the appropriate use of inhalational agents in patients with DMD. It is suggested that caution be exercised with the continued use of inhalational agents in these patients. Given that effective I.V. agents are readily available, it may be prudent to recommend the use of a totally I.V. general anesthetic. The use of succinylcholine is contraindicated because of the risk for an exaggerated hyperkalemic response, rhabdomyolysis, and even cardiac arrest. Although succinylcholine induces a small release of potassium in normal muscle, it produces a potentially lethal efflux in patients with myopathies, which is not prevented by pretreatment with a nondepolarizing agent. The ensuing cardiac arrest is often refractory to routine cardiopulmonary resuscitation. The regenerating muscle fibers—common in patients with DMD until they are at least 8 years of age—are considered to be the most vulnerable to the effects of succinylcholine. Although patients with malignant hyperthermia and those with DMD may present with similar clinical pictures (hyperkalemia, elevated levels of creatine kinase, and cardiac arrest), in the face of a triggering agent the underlying mechanisms are different. It is speculated that an inherent membrane defect in DMD renders the muscle more susceptible to injury induced by depolarization with succinylcholine.

Nondepolarizing neuromuscular blocking agents (NMBAs) are safe to administer, although the duration of action may be prolonged. Normal and prolonged recovery together with higher sensitivity have been documented with both rocuronium and mivacurium, possibly due to a decrease in the total number of neuromuscular junctions and receptors caused by structural changes that result in the replacement of degraded muscle fibers with fatty and fibrous tissue.

The reasons for a longer duration of action of these agents than in normal controls can only be hypothesized. Regardless of the mechanism behind the altered action of NMBAs, an increased duration of action has several clinical implications. Train-of-four monitoring is mandatory because wide interpatient variability precludes an accurate estimation of dosages and the amount of time needed for complete recovery in individual patients. Clinical testing such as head lifting—documented preoperatively, if possible—should be instituted to avoid the risk for residual paralysis. Postoperative testing of muscle strength is also mandatory because of the uncertain effects of reversal agents. Recurarization may occur because the duration of action of an NMA may significantly exceed the duration of action of a reversal agent in this patient population. The future introduction of sugammadex, a modified cyclodextrin that forms a water-soluble complex with steroidal neuromuscular blocking drugs and reverses even very deep neuromuscular blockade, should eliminate the issue of prolonged paralysis in these patients—although this is speculative and further clinical studies are necessary. Sugammadex is currently undergoing Phase III clinical trials.

**Scoliosis Surgery**

Scoliosis develops in 95% of patients with DMD because of the unoppositional antagonism of the dystrophic muscles. A curve of more than 20 degrees typically develops within 3 to 4 years after the patient becomes wheelchair-bound. The combination of scoliosis, weakened respiratory muscles, inactivity, and obesity severely compromises lung expansion and function. Severe scoliosis causes profound negative effects on breathing, resulting in impaired pulmonary function, vulnerability to atelectasis and pneumonia, and pain from mechanical impairment of the musculoskeletal system. Early spinal stabilization is indicated but is associated with significant complications. Surgical correction of the spine improves nursing care and allows a more comfortable wheelchair posture, although long-term improvement of pulmonary function, and thereby increased life expectancy, is controversial. It is thought, however, that surgical correction of scoliosis can stabilize the vital capacity for up to 3 years.

**Management of the Case Presented**

In the case presented, the pulmonologist and cardiologist performed a preoperative evaluation within 2 months of the surgery. The preoperative vital capacity was 1.19 L, or 49% of the predicted value. The forced expiratory flow rate was 59% of the predicted value. The maximal inspiratory and expiratory pressures were –20 and +20 cm H₂O. Treatment included intermittent positive pressure breathing with saline 3 times per week. Echocardiography showed global systolic dysfunction with borderline dilatation and an ejection fraction of 25%.

To facilitate the interpretation of somatosensory-evoked potential monitoring, a totally I.V. technique was chosen. I.V. induction was performed with etomidate at a dose of 0.2

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**Table 3. Surgical Intervention in Patients With Advanced Duchenne’s Muscular Dystrophy**

<table>
<thead>
<tr>
<th>Common Surgical Procedures</th>
<th>Indications and Anesthetic Concerns</th>
</tr>
</thead>
<tbody>
<tr>
<td>Release of Achilles tendon</td>
<td>Prolongs ambulation, improves foot posture</td>
</tr>
<tr>
<td>Repair of fractures</td>
<td>Often emergent, full-stomach procedure; may consider regional anesthesia</td>
</tr>
<tr>
<td>Correction of scoliosis</td>
<td>Prevents deterioration of pulmonary function (controversial), improves quality of life, decreases pain of sitting, facilitates supportive care</td>
</tr>
<tr>
<td>Percutaneous/open gastrostomy</td>
<td>Despite obesity, patients often undernourished; I.V. sedation not recommended because of poor airway support; general anesthesia with endotracheal tube or laryngeal mask airway recommended</td>
</tr>
<tr>
<td>Extracorporeal shock wave lithotripsy</td>
<td>Kidney stones, neurogenic bladder</td>
</tr>
<tr>
<td>Placement of automatic implantable cardioverter-defibrillator</td>
<td>Increased frequency of arrhythmia due to cardiac muscle distortion</td>
</tr>
</tbody>
</table>

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**Intraoperative Concerns:**

- Upper airway obstruction, inadequate cough, and expiratory pressures were –20 and +20 cm H₂O.
- Forced expiratory flow rate was 49% of the predicted value. The forced expiratory flow was 59% of the predicted value. The maximal inspiratory and expiratory pressures were –20 and +20 cm H₂O. Treatment included intermittent positive pressure breathing with saline 3 times per week. Echocardiography showed global systolic dysfunction with borderline dilatation and an ejection fraction of 25%.

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To facilitate the interpretation of somatosensory-evoked potential monitoring, a totally I.V. technique was chosen. I.V. induction was performed with etomidate at a dose of 0.2
mg/kg. Muscle relaxation was achieved with rocuronium at a dose of 0.4 mg/kg. Arterial cannulation and central access were established after intubation. Anesthesia was maintained with remifentanil and propofol.

On conclusion of the procedure, the surgeon placed an epidural catheter under direct visualization; a bupivacaine-fentanyl solution was infused postoperatively. He was taken to the pediatric intensive care unit. His trachea was extubated the next day. During the initial postoperative, postextubation period, the patient was placed on nightly bilevel positive airway pressure and daily intermittent positive pressure breathing with saline. He was discharged home a week after surgery without complications.

References

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Post-test
1. Which of the following is a true statement about Duchenne’s muscular dystrophy (DMD)?
   a. Smooth muscle is the only type of muscle involved.
   b. The underlying defect is a lack of dystrophin, a component of the muscle membrane.
   c. Cardiac muscle is spared.
   d. Muscle degeneration is very painful and does not occur until the late stages of disease.

2. Which neuromuscular blocking agent is contraindicated in patients with DMD?
   a. Rocuronium
   b. Mivacurium
   c. Vecuronium
   d. Succinylcholine

3. What is the appropriate order of tests to evaluate the decline in pulmonary function?
   a. Maximal voluntary ventilation, maximal expiratory and inspiratory pressures, subjective dyspnea, forced vital capacity
   b. Maximal expiratory and inspiratory pressures, subjective dyspnea, maximal voluntary ventilation, forced vital capacity
   c. Maximal voluntary ventilation, forced vital capacity, subjective dyspnea, maximal inspiratory and expiratory pressures
   d. Maximal expiratory and inspiratory pressures, forced vital capacity, subjective dyspnea, maximal voluntary ventilation

4. Which of the following is an appropriate match of the disease with its inheritance pattern?
   a. Becker’s muscular dystrophy, mitochondrial defect
   b. Myotonic dystrophy, autosomal X-linked recessive
   c. Mitochondrial myopathies, autosomal dominant
   d. DMD, autosomal X-linked recessive

5. The features of DMD in children may include the following:
   a. normal development through 1 to 2 years of age
   b. pseudothryphody of calf muscles
   c. regression of physical motor skills (walking and climbing)
   d. all of the above

6. Which of the following is a true statement regarding patients with DMD?
   a. Preoperative testing of pulmonary function does not help predict postoperative pulmonary complications following general anesthesia with endotracheal intubation.
   b. Neuromuscular blockade should be recommended for all patients who have undergone corrective surgery for spinal scoliosis.
   c. Peripheral nerve blocks administered with a nerve stimulator are contraindicated, given that the twitches produced in a single muscle group may cause a hyperkalemic response.
   d. Preoperative assessment should include electrocardiography, echocardiography, and pulmonary function tests.

7. Which of the following is a true statement?
   a. Becker’s muscular dystrophy is the most common form of muscular dystrophy.
   b. DMD is often not diagnosed until the age of 12 years.
   c. There may be an association between a malignant hyperthermia-like reaction and the use of inhalational agents in patients with DMD.
   d. Pretreatment with a nondepolarizing muscle blocker will prevent succinylcholine-induced hyperkalemia.

8. Which of the following develop in both malignant hyperthermia and DMD, in response to triggering agents?
   a. Hyperkalemia
   b. Elevated creatine phosphokinase
   c. Cardiac arrest
   d. All of the above

9. Which of the following is a true statement regarding the diagnosis of DMD?
   a. A medical history and physical examination are needed to confirm the diagnosis.
   b. Laboratory tests may indicate elevated levels of the muscle enzyme creatine phosphokinase.
   c. Electrocardiograms typically show diminished amplitude of the Q-, R-, and S-wave deflections.
   d. Patients present with early symptoms of cardiomyopathy, including chest pain, shortness of breath, and fatigue.

10. In the management of patients with DMD for whom anesthesia and muscular agents have been indicated, the agents that should most likely be avoided are the following:
   a. propofol and opioids
   b. nitrous oxide and opioids
   c. volatile anesthetics and succinylcholine
   d. etomidate and nitrous oxide