Ranger Lock and Load!
From The Joint Council on In-Training Examinations
American Board of Anesthesiology-American Society of Anesthesiologists

"The following responses on the ITE suggested misconceptions in specific areas of the knowledge domain of Anesthesiology... based upon the performance of CA-3 residents taking the examination for ABA credit."

(I regard these "misconceptions" to be important because in a competitive situation questions related to them would stratify and discriminate within a group, thus meeting their goal. Go over these several times. Be careful! Expect these concepts to be disguised with distracters and land mines all around the target area. Ranger now attack!)

According to the Board:
More than 1/2 of CA-3 residents failed to assess cardiac status during preoperative assessment of a 12-year-old patient with Duchenne's muscular dystrophy who is unable to ambulate.

Duchenne's results in death, usually between 15 and 25 years of age, secondary to congestive heart failure or pneumonia. These then are the major problems confronting the anesthesiologist caring for such patients. Two major organ systems are involved and must be assessed, especially in a 12 year old, the heart and the lungs:

a. Cardiac: The heart atrophies and degenerates. Contractility is decreased and sometimes papillary muscle dysfunction leads to mitral regurgitation.
b. Pulmonary: There is decreased ability to cough, decreased respiratory reserve, and decreased ability to handle secretions. All predispose to pneumonia, which in these patients is especially difficult to treat.

Thus, both cardiac and pulmonary status must be assessed in a patient with Duchenne's.

I. Myotonic Dystrophy
1. Myotonic dystrophy is a group of diseases manifested by prolonged muscle contraction and delayed muscle relaxation following stimulation. The inability of muscle to relax after stimulation is diagnostic.
   a. Atrophy and weakness are the key features of myotonic dystrophy.
   b. Typically, involved muscles include the facial, pharyngeal, & sternocleidomastoid.
   c. An autosomal dominant disease with variable penetrance, myotonic dystrophy is marked by symptoms which appear in the second and third decades of life.
   d. As with myasthenia gravis, myotonic dystrophy strikes hardest at respiratory and cardiac muscle.

2. There are three major groups of myotonic dystrophy
   a. Myotonia dystrophica
      1) Myotonia dystrophica is the most common and most serious form of myotonic dystrophy, with symptoms usually appearing during the third decade.
      2) Besides muscle weakness (especially respiratory), the most important symptom is cardiac abnormalities, including cardiomyopathy, cardiac dysrhythmias, and cardiac conduction abnormalities. Preoperative cardiopulmonary evaluation is essential (see Lock 'n Load above).
         a) Myocardial depression from volatile anesthetics aggravates cardiomyopathy from myotonic dystrophy.
         b) Anesthesia can also worsen cardiac conduction defects. It is not infrequent to encounter RBBB or even sudden third degree heart block.

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1Stoelting, RK, Dierdorf, SF. Skin And Musculoskeletal Diseases. Anesthesia and Co-Existing Disease, Stoelting, RK, Dierdorf, SF (eds.), Churchill Livingstone.
c) Succinylcholine causes sustained muscle contraction and should be avoided. Nondepolarizers are acceptable.

d) Extubation criteria must be observed. Restrictive lung disease can be present secondary to muscle weakness. Respiratory sensitivity to barbiturates, opioids, and benzodiazepines can be extreme.

e) As with many of the other M's (Myasthenia gravis, myasthenic syndrome, myotonia) muscular dystrophy is associated with increased sensitivity to the nondepolarizers. Doses should be decreased.

b. Myotonia congenita
  1) Myotonia congenita is manifested at birth.
  2) Skeletal muscle involvement is widespread; other organs often spared.
  3) Myotonia congenita often responds to quinine.

c. Paramyotonia
  1) Paramyotonia is the rarest of the three forms of MD.
  2) Symptoms are mildest and usually develop only in response to cold.

Written Board Beach Pillbox
(Q & A at Course)

3. Neuromuscular relaxation and myotonic dystrophy?

a. Succinylcholine, a membrane active depolarizing agent, should be avoided in myotonic dystrophy as it causes sustained contraction and hyperkalemia. In terms of the later, according to Stoelting: "Succinylcholine may be associated with exaggerated potassium release leading to life-threatening cardiac dysrhythmias". [In MG sux causes an unreliable relaxation and in MD it causes a sustained contraction and hyperkalemia. In both cases therefore, it is best avoided.]

b. Sensitivity to nondepolarizers is typically increased.

c. Anticholinesterases to reverse a nondepolarizing block often precipitates myotonia.

4. IV regional and local infiltration have advantages because anesthetic goes directly to the muscle. Spinal does not ensure relaxation in these cases. In fact, sustained contraction can occur with spinal. Infiltration with quinine, tocanide, and mexiteline may also alleviate symptoms.

5. These patients are very sensitive to the depressant effects of barbiturates, benzodiazepines, and narcotics. They should only be administered with caution.

6. "Full stomach precautions", including rapid sequence induction should be taken because gastric motility is decreased and emptying delayed.

II. Muscular Dystrophy3 (Pseudohypertrophic or Duchenne Muscular Dystrophy is the most common type.)

1. Muscular dystrophy is an X-linked disease characterized by painless atrophy and degeneration of muscles.

2. The condition is pseudohypertrophic because fatty infiltration causes affected skeletal muscles to enlarge.

3. Death usually occurs between 15-25 years of age secondary to congestive heart failure or pneumonia. By remembering this, one will also remember that these are the major problems confronting the anesthesiologist caring for such patients:
   a. Cardiac: The heart atrophies and degenerates. Contractility is decreased and papillary muscle dysfunction leads to mitral regurgitation.

b. Pulmonary: Decreased ability to cough, decreased respiratory reserve, and decreased ability to handle secretions all predispose to pneumonic processes, which in these patients are especially difficult to treat.

4. Additional problems include associated kyphoscoliosis and increased risk of MH.

**A Question From Dr. Jensen's Written Course**

K type
In myotonic dystrophy, spasticity of muscles is relieved by
1. Succinylcholine
2. Spinal anesthesia
3. Neostigmine
4. Local infiltration of the muscle

Yes! Another answer directly from Big Blue. Easy kill for the Ranger! Let's review.
1. Myotonic dystrophy is a group of diseases in which there is prolonged muscle contraction and delayed muscle relaxation following stimulation. The inability of muscle to relax after stimulation is diagnostic.
   a. Atrophy and weakness are the key features of myotonic dystrophy.
   b. Typically, the muscles involved are the facial, pharyngeal, sternocleidomastoid.
   c. As with myasthenia gravis, think of impairment of the respiratory muscles and the increased risk of aspiration as well as impairment of the cardiac muscle (cardiomyopathy).

2. What about neuromuscular relaxation?
   a. Succinylcholine should not be used because it causes a sustained contraction and hyperkalemia. This is because succinylcholine is a membrane active depolarizing agent. [In MG it causes an unreliable relaxation and in MD it causes sustained contraction. In both cases, it is probably best avoided.]
   b. Sensitivity to nondepolarizers is typically increased.
   c. Anticholinesterases to reverse the nondepolarizing block might precipitate myotonia.

3. IV regional and local infiltration have advantages because anesthetic goes directly into muscle. Spinal does not ensure relaxation in these cases. In fact, sustained contraction can occur with spinal. Infiltration with quinine, tocainide, and mexiteline may also alleviate symptoms.

**NFI's Key Quotes**

Chuck Tanner, a long-time and successful baseball manager, knows what it takes to win—both on the baseball field and off from it.
1. Health is wealth.
2. Be cheerful and optimistic and opportunity will rise in front of you.
3. Don't find fault with the umpire. Nobody can be as perfect as you are.
4. If you're managing a ballclub, react to only half of what you see and one-fourth of what you hear.
5. Don't be afraid to fail. The ashes of failure give birth to victory.
6. If you feel like crying, cry.
7. Always put your family first.
Myotonic and Muscular Dystrophy Keywords
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Recent keywords of importance—most all of which are covered in Big Blue and/or at the Course:

1. Myotonic dystrophy: anesthetic implications; Abnormal drug resp
2. Duchenne’s: succinylcholine effects
3. Preop evaluation: duchenne muscular dystrophy
4. Muscular dystrophy: anesthetic implications

Other relevant keywords from recent years. The reference is first, the keyword is underlined.


Myotonic Dystrophy: Anesthetic Implications
(Big Blue will put you in a great position to answer anything regarding this topic)
1. Myotonic dystrophy is a group of diseases in which there is prolonged muscle contraction and delayed muscle relaxation following stimulation. The inability of muscle to relax after stimulation is diagnostic.
   a. Atrophy and weakness are the key features of myotonic dystrophy.
   b. Typically, the muscles involved are the facial, pharyngeal, sternocleidomastoid.
   c. This is an autosomal dominant disease with variable penetrance (as is MG). Symptoms usually occur when patients are in the second and third decades of life.
   d. As with myasthenia gravis, think of impairment of the respiratory muscles and the increased risk of aspiration as well as impairment of the cardiac muscle (cardiomyopathy).
2. There are three major groups of myotonic dystrophy
   a. Myotonia dystrophica
      1) Most common and most serious with symptoms usually appearing during the third decade.
      2) Besides muscle weakness (especially respiratory), the most important symptom is cardiac abnormalities, including cardiomyopathy, cardiac dysrhythmias, and cardiac conduction abnormalities. Therefore, in terms of management one must consider the presence of cardiomyopathy and conduction abnormalities as well as respiratory failure. Preop cardiopulmonary evaluation is essential.
      a) Myocardial depression from volatile anesthetics can aggravate cardiomyopathy.
      b) Anesthesia can worsen cardiac conduction defects. It is not infrequent to encounter RBBB or even sudden third degree heart block.
      c) Succinylcholine causes sustained contraction of muscle and should be avoided. Nondepolarizers are acceptable.
      d) Extubation criteria must be observed. A restrictive lung disease can be present secondary to weakness. In addition, there can be extreme respiratory sensitivity to barbiturates, opioids, and benzodiazepines.
      e) As with many of the other M’s (Myasthenia gravis, myasthenic syndrome, myotonia) muscular dystrophies are associated with increased sensitivity to the nondepolarizers. The dose should be decreased. Multiple sclerosis has normal sensitivity to nondepolarizers and responds to sux with hyperkalemia.
   b. Myotonia congenita
      1) Manifested at birth
      2) Skeletal muscle involvement is widespread but other organ systems are not usually involved.
      3) Responds to quinine
   c. Paramyotonia
      1) Rarest of the three
      2) Symptoms are the most mild and develop only in response to cold.
3. What about neuromuscular relaxation?
   a. Succinylcholine should not be used because it causes sustained contraction. This is because it is a membrane active depolarizing agent. [In MG, succinylcholine typically causes an unreliable relaxation; in MD it causes a sustained contraction. In both cases therefore, it is probably best avoided.]
   b. The response to nondepolarizers is usually heightened.
   c. Anticholinesterases to reverse a nondepolarizing block might precipitate myotonia.
4. IV regional and local infiltration have advantages because anesthetic goes directly to the muscle. It is not suitable for many cases, obviously. Spinal does not ensure relaxation in these cases. In fact, sustained contraction can occur with spinal. Infiltration with quinine, tocanide, and mexiteline may also alleviate symptoms.
5. These patients are very sensitive to the depressant effects of barbiturates, benzodiazepines, and narcotics. Use them with caution.
6. Take full stomach precautions because gastric motility is decreased and gastric emptying delayed.