Anesthesia for the Patient with Neuromuscular Disease

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Disclosures

Nothing to disclose
At the conclusion of this presentation the participants will be able to:

• Understand the pathophysiology of Duchenne’s muscular dystrophy

• Manage the patient anesthetic course
A 12 y/o 50 kg male presents for an exploratory laparotomy for probable appendicitis. History is significant for a diagnosis of Duchenne muscular dystrophy made at age 3. He is wheelchair bound with significant scoliosis. Medications include morphine sulfate, pepcid and cefoxtin. P 120, BP 100/50, R20, T 38.5. He is visiting his grandparents from another state.
Muscular Dystrophy - Duchenne & Dystrophy - 1

- Sex linked recessive - boys more than girls
- Onset 2-6 years of age
- Cardiac abnormalities, respiratory infections, muscle contractures and kyphoscoliosis
Incidence and Genetic Inheritance

- 1:3500 boys - the most common form of muscular dystrophy
- One-third result from spontaneous mutation (no family Hx) - rest inherited in an X gonosomal recessive way
Pathophysiology

- DMD gene dystrophin is absent or nonfunctional in DMD patients.
- Dystrophin is present in skeletal and cardiac muscle and brain.
- Absence results in decreased sarcolemma stability in contraction/relaxation or disturbed calcium homeostasis with excessive calcium influx into the cells.
- Destruction and necrosis of muscle fibers-scar and fat.
- Compensatory hypertrophy of muscles.
Diagnosis by muscle biopsy and elevated plasma creatine kinase (30-300 times normal)

70% of female carriers have elevated plasma creatine kinase
Cardiac abnormalities occur in 50-70% of patients
- Atrial tachycardia 65%
- MVP 25%
- ECHO posterolateral hypokinesis with fibrosis of the myocardium

Respiratory abnormalities
- Reduction in all lung volumes
- Kyphoscoliosis due to muscle weakness
- Recurrent infections
- Sleep apnea
Anesthetic Precautions

- Full history - motor milestones
- PFT, ABG, Chest x-ray
- Cardiac ECG, Echocardiography
- CBC, electrolytes
Anesthetic Considerations - 1

- M weakness, respiratory insufficiency, metabolic changes (hyperkalemia and hyperthermia), cardiomyopathy (late)
- Enlarged tongue, ankylosis of TMJ
- Cervical lordosis - tracheal bronchial compression - prone
- Severe hyperthermia
- Arrhythmias - tachycardia, ventricular fibrillation, cardiac arrest
Anesthetic Considerations - 2

- Cardiac arrest - postoperative period
- Joint deformities - difficult positioning and vascular access
- Mechanical ventilation postoperative for spine surgery
Duchenne Dystrophy and Succinylcholine

- Succinylcholine - exaggerated serum potassium release, rhabdomyolysis, acidosis and cardiac arrest

- Avoid until age 8 in all children?

- Responds to dantrolene and calcium
Duchenne Dystrophy and Inhaled Anesthetic Agents

- Isoflurane resulted in elevation of creatine kinase and cardiac arrest.
  
  *Chalkeades GA Anesthesia 45;22,1990*

- Avoid combination with succinylcholine.
Malignant Hyperthermia

- DMD and MH are independent processes
- MH and DMD are the same incidence as the general population
Anesthesia Induced Rhabdomyolysis

- Absence of dystrophin - glycoprotein complex - increased permeability of the sarcolemma and increased cellular calcium levels
- Exposure to inhalational agent (or succinylcholine) stresses the M cell membrane - (K⁺ and CK) leak out
- Hypermetabolic response occurs to prevent Ca²⁺ fluxes
- Hyperkalemia, hyperthermia, tachycardia, and rhabdomyolysis
Review article

Duchenne muscular dystrophy: an old anesthesia problem revisited

J Hayes, F Veyckemans and B Bissonnette

Pediatric Anesthesia 2008; 18:100-106
Succinylcholine (absolute contraindication) and halogenated agents should be avoided - hyperkalemic response

TIVA can be used - propofol, opioids, benzodiazepines

Dantrolene must be available
**Duchenne Dystrophy - Recommendations for Anesthetic Management**

- **Titration of drugs to effect such as propofol**
- **Avoid succinylcholine**
  - Nondepolarizing relaxants may be prolonged
- **Anticholinergics may increase tachycardia**
  - Glycopyrrolate is better than atropine
- **Gastric dilatation - use nasogastric tube and full stomach precautions**
- **Watch for airway obstruction in the prone position**
- **Anesthetic agents may cause respiratory depression postoperatively**