

# Muscular Dystrophy

CoreNotes by Core Concepts Anesthesia Review, LLC

## What You Must Know

1. Duchenne muscular dystrophy (DMD) is the most common type of muscular dystrophy.
2. It is an X-linked disorder seen in males, appearing in childhood with progressive muscle wasting. Death usually occurs during adolescence.
3. Cardiac muscle is also affected and death usually results from cardiac and/or ventilatory failure.
4. ECG changes appear as decreased R-wave amplitude and the appearance of Q-waves.
5. Changes in the architecture of the thorax result in progressive and severe restrictive lung disease.
6. Severe hyperkalemia and rhabdomyolysis have resulted from the administration of succinylcholine. The use of succinylcholine is contraindicated in these patients.
7. Although once felt to be associated with MH, recent data suggest that this may not be true and deaths were likely the result of hyperkalemia.

DMD is the most common childhood muscular dystrophy. It is an X-linked recessive disorder appearing in between the ages of 3 - 5, with progressive weakness. It is sometimes referred to as pseudohypertrophic muscular dystrophy and is usually fatal by late adolescence as a result of ventilatory and/or cardiac failure. In addition, these young males suffer from contractures, marked scoliosis, restrictive lung disease, and cardiomyopathies; 50% of sufferers have a dilated cardiomyopathy by the age of 15.

The disease is a result of a lack of dystrophin, a protein that helps to hold muscle cells to the extra-cellular matrix. This leads to a weakening of the sarcolemma, which becomes increasingly permeable, with increased intracellular calcium levels.

Other less-common forms of muscular dystrophy also occur including: Emery-Dreifuss muscular dystrophy, limb-girdle muscular dystrophy, Becker's muscular dystrophy, oculopharyngeal muscular dystrophy, fascioscapulohumeral muscular dystrophy and congenital muscular dystrophy.

The most significant anesthetic complications in patients with DMD are the result of the actions of volatile anesthetics or neuromuscular blocking agents on the cardiac and skeletal muscle. Cases of cardiac arrest have been reported and are associated with severe hyperkalemia and rhabdomyolysis. In patients with muscular dystrophy, succinylcholine appears to damage the weakened sarcolemma causing release of intracellular contents and is absolutely contraindicated. Patients frequently have prolonged recovery from nondepolarizing relaxants. Anticholinesterase agents have also been implicated in muscle breakdown.

### Additional Reading:

Barash, PG, Cullen, BF, Stoelting, RK, Cahalan, MK, Stock, MC, and Ortega, R. Clinical Anesthesia. Philadelphia: Lippincott Williams & Wilkins, 2013:613