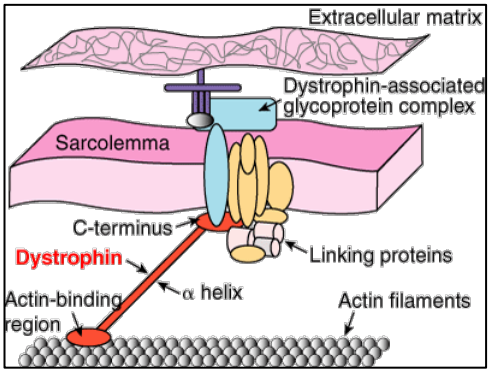
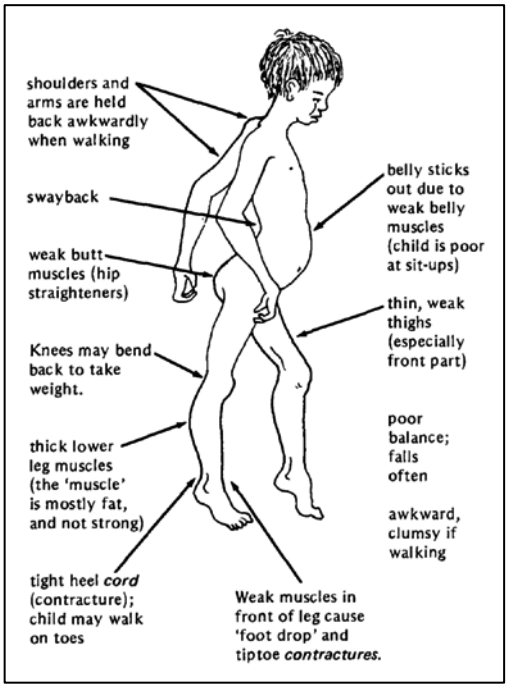
Muscular Dystrophy

Anesthetic Pearls: Anesthetic Implications and Management of Muscular Dystrophy

Duchenne's Muscular Dystrophy is an X-linked recessive disease with an incidence of 30/100,000 males. The disease is often undiagnosed until age 3-5 years. The disorder is caused by a mutation in the dystrophin gene located in humans on the X-chromosome (Xp21). The dystrophin gene codes for the protein dystrophin which is an important structural component within muscle tissue. Dystrophin provides structural stability to the dystroglycan complex (DGC) which is located on the cell membrane. Symptoms usually appear in male children before age 5 and may be visible in early infancy. Progressive proximal muscle weakness of the legs and pelvis associated with a loss of muscle mass is observed first. Eventually this weakness spreads to the arms, neck, and other areas. Early signs may include pseudohypertrophy of calf and deltoid muscles, low endurance, and difficulties in standing unaided or inability to ascend staircases. As the condition progresses, muscle tissue experiences wasting and is eventually replaced by fat and fibrotic tissue. The average life expectancy for patients afflicted with DMD varies from late teens to early to mid 20's. There have been reports of a few DMD patients surviving to the age of 40, but this is extremely rare. Peri-operative risk includes respiratory failure and prolonged mechanical ventilation. There is a potential for prolonged ventilator dependence if vital capacity is less than 30% of predicted.





Anesthetic Concerns:

1. Poor cardiac function
2. Aspiration risk
3. Hyperkalemic arrest with Succinylcholine
4. Association with MH (avoid triggering agents?)
5. History of steroid use
6. Increased sensitivity to non-depolarizing muscle relaxants (titrate to effect). Recommended to allow for spontaneous neuromuscular recovery due to variable response to reversal agents.
7. Increased propensity for mitral valve prolapse
8. Consider avoiding cardiac depressants
9. Consider NPO status as full stomach

System	Effect	H&P	Tests
CV	Decreased cardiac conduction Decreased contractile force	Tachycardia, Hx of CHF	EKG, ECHO
Resp	Decreased volume & flow OSA / pulm HTN	Snoring, apnea	PFT's, ABG, sleep study, ECHO
GI	Dysmotility, gastric dilatation, paralytic ileus	Constipation	KUB
GU	Bladder paralysis	Urinary retention	Bladder scan
MS	Scoliosis, kyphosis, contractures, muscle destruction	Progressive weakness	Spine film, myogram, increased CK level
CNS	Decreased IQ	Mental slowness	Mental status exam