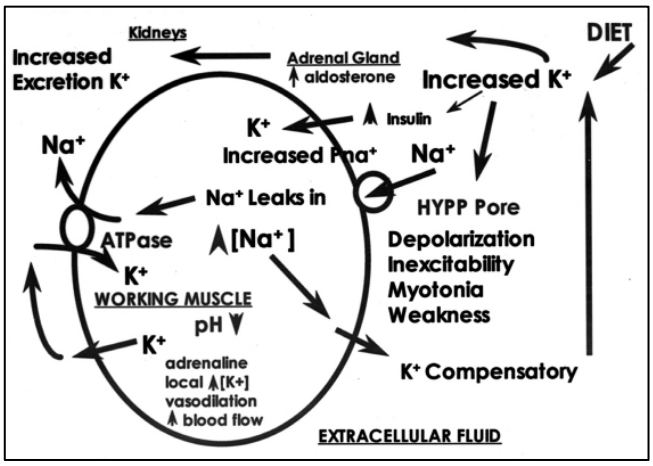
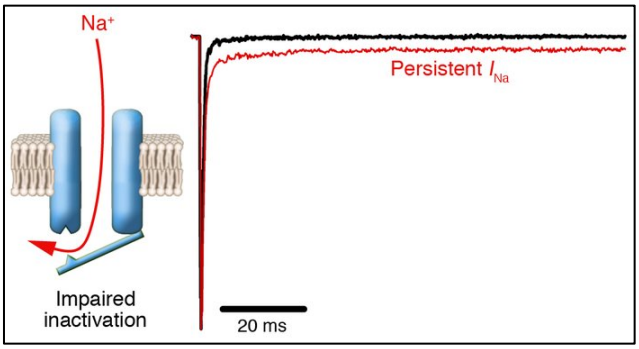
Hyperkalemic Periodic Paralysis

Anesthetic Pearls: Anesthetic Management and Implications of Hyperkalmenic Periodic Paralysis



Hyperkalemic periodic paralysis (HyperKPP) is a genetic disorder which occurs in both humans and horses. It is an inherited autosomal dominant disorder that affects sodium channels in muscle cells and alters the ability to regulate serum potassium levels. The condition is most commonly associated with horses, but also occurs in humans, where it may be called "**Gamstorp episodic adynamy**". Hyperkalemic periodic paralysis is characterized by muscle hyper-excitability or weakness which is exacerbated by potassium or cold and can lead to uncontrolled shaking followed by paralysis. Onset in humans usually occurs in the twenties.



The mutation which causes this disorder is dominant on "SCN4A" gene with linkage to the sodium channel expressed in muscle tissue. The single amino acid mutation causes changes in parts of the sodium channel that are important for conduit inactivation. In the presence of high potassium levels, including those induced by diet, the sodium channel fails to activate properly. Hyperkalemic Periodic Paralysis can therefore lead to hyperkalemia. In other cases, attacks are associated with normal blood potassium levels. Ingesting potassium can often trigger attacks in affected individuals, even if blood potassium levels do not rise in response.

The disorder causes episodes of extreme muscle weakness with attacks often beginning in infancy. Depending on the type and severity of the HyperKPP, it can increase until the fourth or fifth decade. Muscle strength often improves between attacks, although many affected people may have increasing bouts of muscle weakness as the disorder progresses (abortive attacks). Sometimes persons affected with HyperKPP may experience degrees of muscle stiffness and spasms (myotonia) in the affected muscles. Many attacks are not severe enough to need therapy. Some people have several attacks a day that can typically last about 1 - 2 hours (some can last as long as a day).

Triggering Factors:

A. Stress
B. Fatigue
C. Rest after exercise
D. Potassium-rich foods
E. Weather changes
F. Environmental pollutants (cigarette / cigar smoke)
G. Periods of fasting

Treatment of HyperKPP:

1. High-carbohydrate diet (recommended)
2. Glucose or other carbohydrates can be given during an attack to reduce the severity and duration.
3. IV Calcium decreases activity of sodium channels (may attenuate sudden attacks).
4. IV diuretics (Furosemide, Acetazolamide, Thiazides) can be effective at stopping sudden attacks.
5. IV Glucose and Insulin stimulates potassium uptake into the cell by the Na-K ATPase (may reduce weakness without a loss of total body potassium).
6. Avoidance of known attack triggers