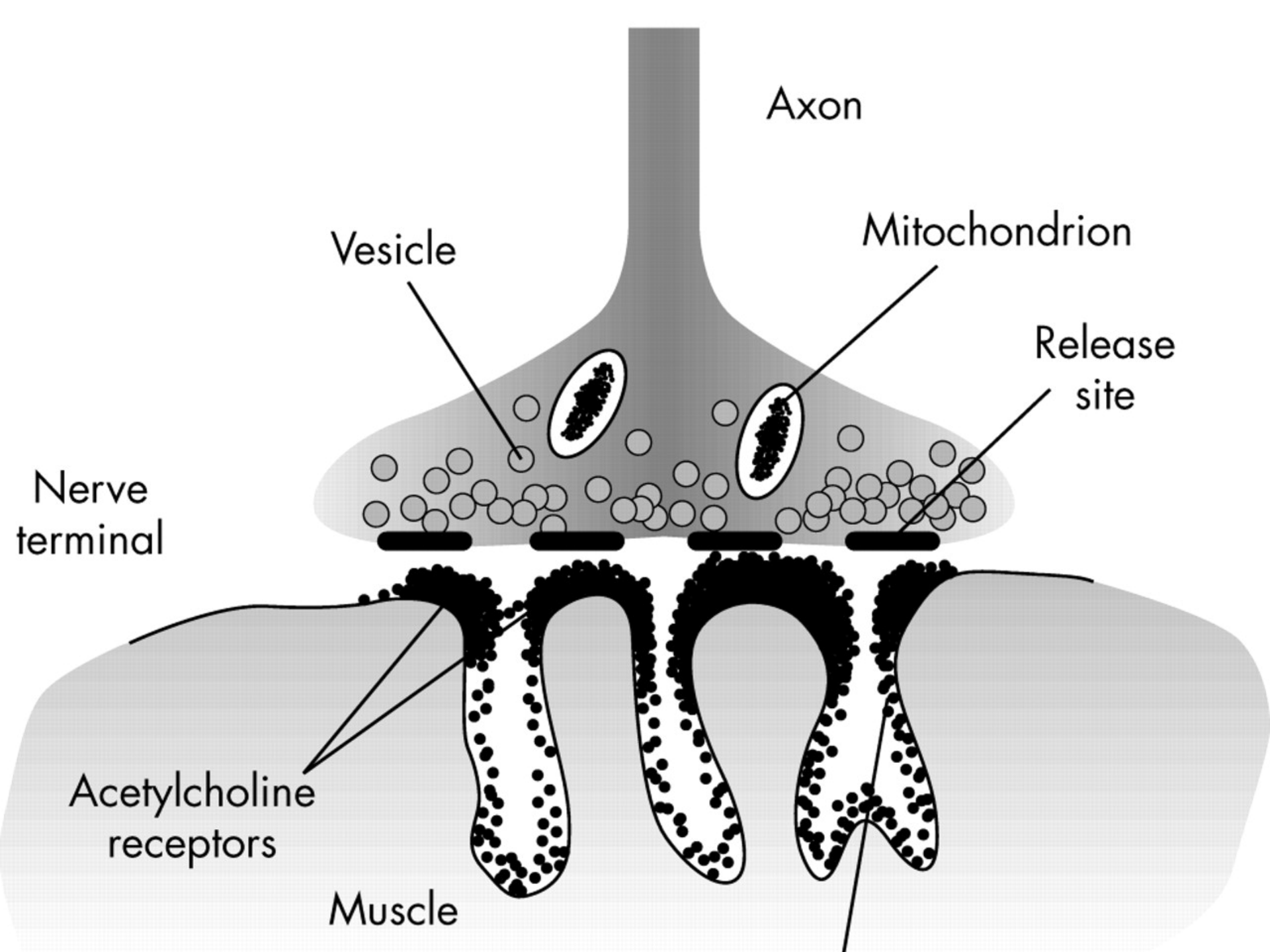
Neuromuscular Diseases

Myastenia Gravis

Antibodies created by the immune system disable the acetycholine binding sites in the neuromuscular junctions, preventing acetycholine from activating the muscle

http://highered.mcgraw-hill.com/olc/dl/120107/bio_c.swf


Axon
Vesicle
Mitochondrion
Release site
Nerve terminal
Acetylcholine receptors
Muscle

Myasthenia Gravis

Autoimmune disease

The immune system developes antibodies that block the nerve impuleses from controling the voluntary muscles.

Causes lack of muscle control and strength

Myasthenia Gravis

Litterally means "grave muscle weakness" in latin

Often can be seen as a drooping eyelid, drooling, lack of body control and inability to stand.

Muscular Dystrophy

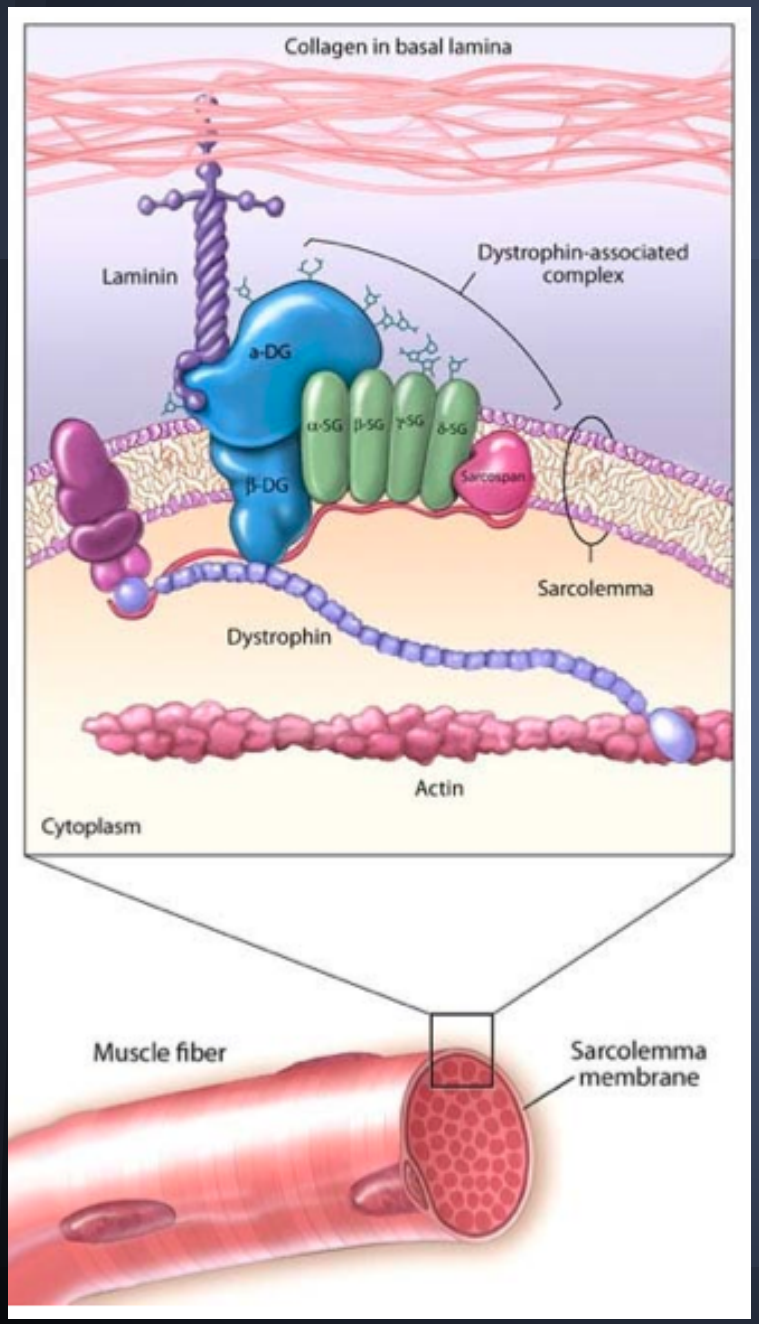
Muscular Dystrophy is a group of neuromuscular diseases causing muscular weakness

These diseases are inherited

They can occur at any age

Muscular Dystrophy

- Muscle weakness that slowly gets worse
- Delayed development of muscle motor skills
- Difficulty using one or more muscle groups
- Drooling
- Eyelid Drooping
- Frequent falls
- Loss of strength in a muscle or group of muscles as an adult
- Loss in muscle size
- Problem/Delayed walking



The exact function of Dystrophyn is not completely known but we do know that it attaches to the actin filaments in the muscle cell and provides reinforcement to the sarcolema

Muscular Dystrophy

In patients with Muscular Dystrophy (MD) genetic deffects cause dystrophyn in the muscle cell to be absent or malformed.

This causes the muscle cells to leak proteins and ensymes responsible for contraction

Muscular Dystrophy

Damaged cells lack the enzymes and proteins needed to contract, causing muscle weakness.

Extracellular substances also leak into the cell distrupting the process of contraction

Muscle cell death also occurs resulting in decreased muscle mass.

Muscular Dystrophy

There is no known cure for MD, however there are treatments for the symptoms, such as surgery for spinal problems.

MD symptoms tend to become less severe with activity rather than rest.

Rhabdomyolysis

Rhabdomyolysis is the breakdown of muscle fibers which releases myoglobin into the bloodstream.

Myoglobin a protein in muscle fibers, and is broken down into substances that are damaging to the kidneys.

Rhabdomyolysis

Alcoholism, recreational drugs, crush injuries, extreme exertion, genetic disease, heatstroke and shaking chills can be risk factors.

Anything that can damage the structure of muscle fibers causes myoglobin to be released.

Rhabdomyolysis

The kidneys filter the blood of substances including myoglobin.

Normal levels of blood myoglobin can be safely filtered by the kidneys.

When too much myoglobin enters the blood stream, the kidneys become poisoned and damaged.

Rhabdomyolysis

Symptoms include:

- Decreased and discolored urine production
- Stiff, aching muscles
- Muscle weakness
- Sometimes joint pain and even seizures can occur

Rhabdomyolysis

Patients are treated with fluids containing bicarbonate to flush out the kidneys and some patients need dialysis.

Prompt treatment is needed to prevent permanent kidney damage.

Hypocalcemic Tetany

Can be life-threatening

Results from a low level of calcium in the circulation

Low calcium results in muslce contracting and not releasing, which can damage muscles, tendons and even bones

Hypocalcemic Tetany

The lowered calcium levels cause the motor neurons to become more excitable, which can cause involuntary muscle contraction.

Lower than normal calcium levels make neural membranes more permiable to Sodium ions, which initiate an action potential

Hypocalcemic Tetany

Patients with HT have high enough calcium levels to have muscle contractrions, but not enough to release.

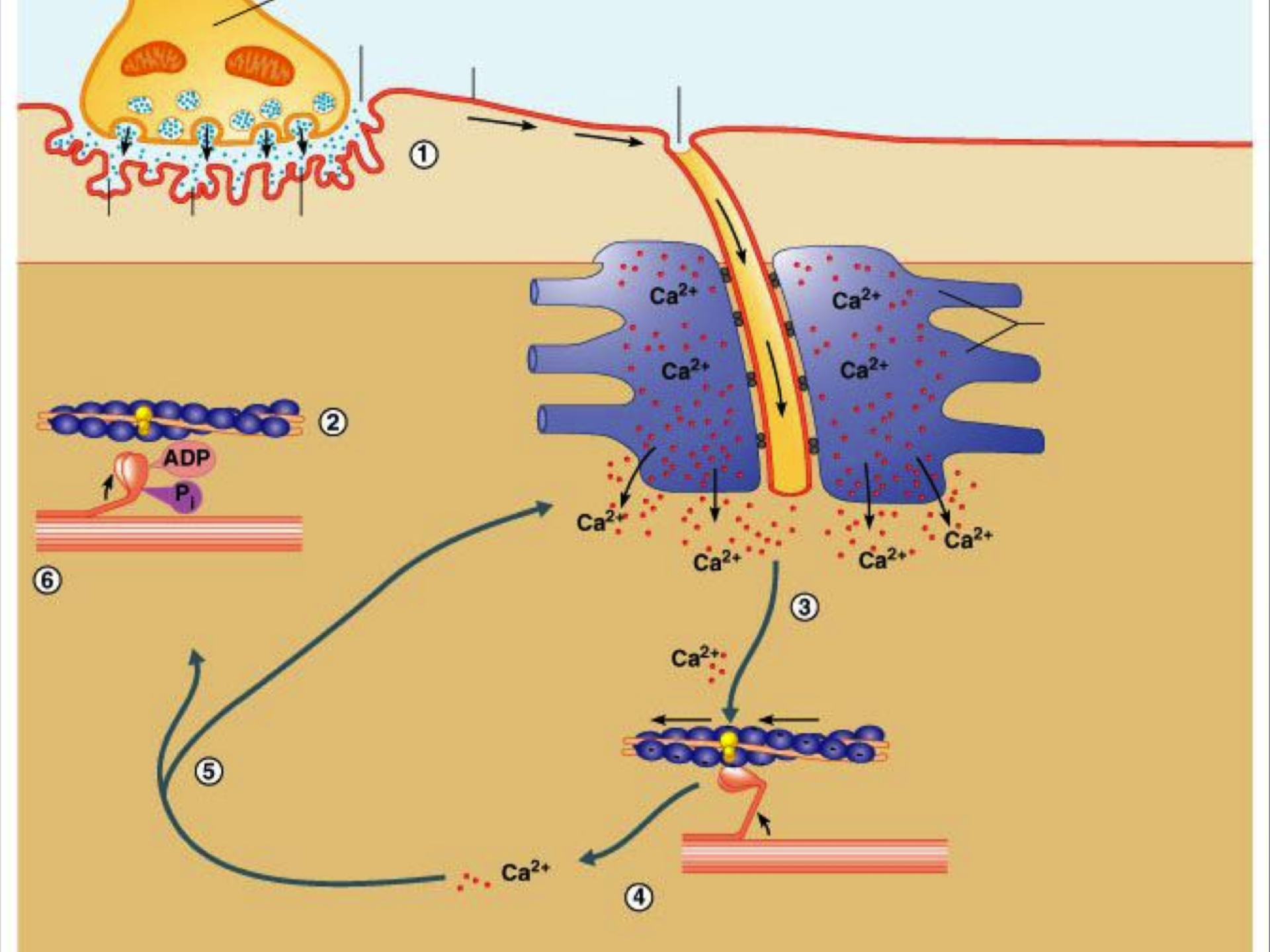
The sarcoplasmic reticulum pumps Ca+ back into the SR, to stop the contraction.

The Ca+ pump is activated by elevated Ca+ levels

Hypocalcemic Tetany

Because there is not enough Ca+ in the in the sarcoplasm, the Ca+ pumps in the SR don't activate.

The calcium levels remain just high enough to continue the contraction, which is tetany.


①
Ca2+
Ca2+
Ca2+
Ca2+
Ca2+
Ca2+
Ca2+
Ca2+
②
ADP
Pi
③
Ca2+
④
Ca2+
⑤
⑥

Rigor Mortis

A few hours after death, the skeletal muscles in the body partially contract, causing the body to tense up, joints to lock in place and often contortion of the body.

Rigor lasts for around 72 hours depending on the environment the body is in.

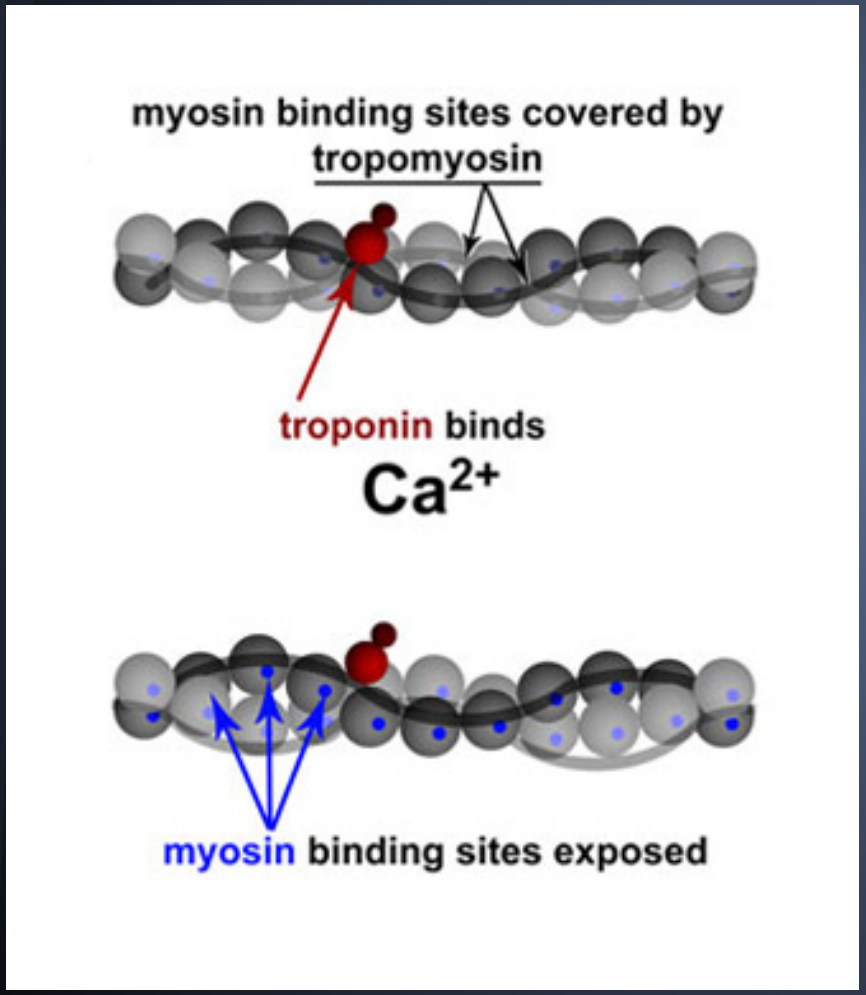
Rigor Mortis

While a body is alive, ATP is produced and used at a constant rate to pump calcium out of the muscle cell.

ATP production stops after death, stoping the calcium pumps in the sarcoplasmic membrane.

Calcium naturally fallows the concentration gradient into the sarcoplam.

Rigor Mortis



The calcium that enters the muscle cell binds with troponin and exposes the myosin binding sight, initiating the muscle contraction.

Rigor Mortis

Because ATP is required to remove the calcium and release the myofibrils, the muscles remain contracted.

The muscles only relax after the sarcoplasmic membrane breaks down allowing enzymes in to break apart the protein fibers, actin and myosin.