

Thème 3 – Corps humain et santé

5-B- La cellule musculaire et la glycémie

Glycogen storage diseases of muscle

Using the documents, explain the origin of the mechanisms responsible for GSD, a metabolic disorder of muscles.

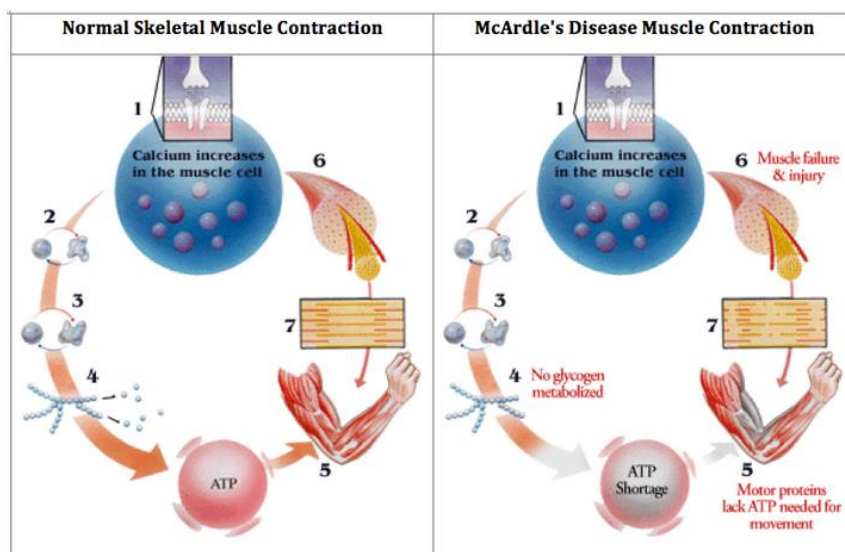
Document 1. What is glycogen and glycogen storage disease ?

The body's cells need a steady supply of fuel in order to function the right way. This fuel is a simple sugar called glucose. Glucose comes from breaking down the food we eat. The body uses as much glucose as it needs to function and stores the rest to use later. Before it can be stored, the body must combine the simple glucose units into a new, complex sugar called glycogen. The glycogen is then stored in the liver and muscle cells. When the body needs extra fuel, it breaks down the glycogen stored in the liver back into the glucose units the cells can use. Special proteins called enzymes help both make and break down the glycogen in a process called glycogen metabolism. Sometimes a person is born missing an enzyme needed for this process or it may not work right. Then the body is not able to store or break down the glycogen as it should. This can lead to very low blood glucose levels during periods of fasting*. The muscles and organs need a certain level of glucose in the blood to work properly. When the body is missing an enzyme or has a flawed enzyme and is not able to use glycogen the right way, it leads to a condition called glycogen storage disease (GSD). Many different enzymes are used by the body to process glycogen. And, as a result, there are several types of GSD.

<https://my.clevelandclinic.org/health/diseases/15553-glycogen-storage-disease-gsd>

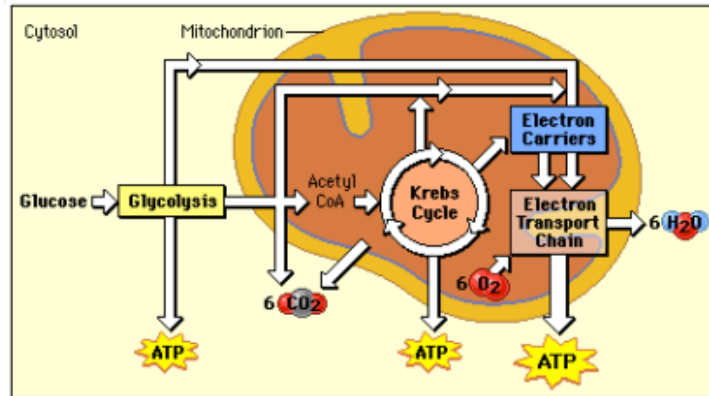
* jeûne

Document 2. Muscle contraction in an unaffected muscle compared to one with McArdle's Disease, one of the most common GSD.



https://www.physio-pedia.com/McArdle%27s_Disease

Document 3. Cellular respiration



<http://www.colby.edu/chemistry/BC368/powerpoints/CH3.pdf>