Wilson's Disease

Introduction

Wilson's disease can be termed as a genetic disorder that makes it complicated for the body to get rid of additional copper. Medical research has shown that the body of a human being requires small quantities of copper for it to be healthy. This however is not the case when the body of an individual accumulates too much copper as it results to being poisonous. Wilson's disease occurs in an individual when high levels of cooper accumulate in the body organs such as brain, eyes, liver among others. When the copper levels reach extreme levels, they become poisonous and lead to life threatening damaging of the affected organs. Wilson's disease got its name from Samuel Alexander Kinnier Wilson who was a british physician who was the one that described the condition as well as the brain and liver pathological changes. Walshe JM (1996).

Who gets Wilson's disease?

Individuals who are infected with Wilson's disease take over two abnormal copies of the ATP7B gene from both parents. People who are carriers of Wilson’s disease and have only one copy of the abnormal genes do not have symptoms of the disease. Individuals with the disease are known to have no account of the ailment in their families. One has an amplified risk of getting the illness if one or both of his parents have the disease. The chance of one getting the disease is one in forty thousand people. Wilson's disease equally affects both women and men and is usually reported in people from ages two to seventy two years.

Causes of Wilson's disease
Wilson's Disease

The cause of Wilson's disease is the accumulation of excess levels of copper in various organs of the body. In normal functioning of the body, copper that is deposited in the body is supposed to be filtered out by the liver and is then released into the bile. The copper then flows out of the system through the gastrointestinal tract. Mutation of the ATP7B gene is what causes individuals not to be able to release copper from the liver at the required rates and levels hence causing Wilson's disease. Wilson's disease also occurs in other body organs. This happens as a result of exceeding the maximum copper storage capacity of the liver, where the copper is then released into other organs of the body such as kidneys, eyes and brain through the bloodstream.

Symptoms of Wilson's disease

The first victims of Wilson's disease in the body are the central nervous system and the liver. Symptoms that are portrayed by people suffering from Wilson's disease include; swelling of the liver. The eyes become white whereas the skin adopts a yellowish color and bruises easily. Brewer GJ. (2001).

In instances where copper builds up in the central nervous system, individuals portray symptoms such as; difficulties with speech and physical coordination, where the individual has uncontrolled movements, the person starts to have behavioral changes as well as stiffness of the muscles. Anemia, low levels of white blood cells and blood clotting, high levels of protein, uric acid and arthritis are other symptoms of Wilson's disease. The most unique symptom of Wilson's disease however is the Kayser-Fleishec rings which results from the buildup of copper in the eyes. The rings occur in the eyes in form of a rusty-brown ring around the iris edge and the cornea's rim.

Diagnosis of Wilson's disease

Conducting laboratory tests and physical examination is the manner in which the disease is diagnosed. Doctors usually look for visible signs of Wilson's disease when they are conducting
physical examination. One of the equipments that are used to check for Kayser Fleischer rings in the eyes is the slit lamp. The amount of copper in the blood, urine and liver tissue of an individual is tested using laboratory tests. Individual who have the disease usually have below normal levels of copper in the blood and below normal levels of ceruloplasmin that is responsible for carrying copper in the bloodstream. In situations where there is acute liver failure the level of cooper in the blood is normally higher then usual and this leads to Wilson’s disease. Urine collections of individuals who suffer from Wilson’s disease also portray high levels of copper.

Diagnosis of Wilson’s disease can also be conducted where doctors conduct liver biopsy which is a medical procedure that extracts a small piece of the liver tissue for purposes of verifying whether it is retaining higher than normal levels of copper. In some circumstances, doctors conduct a misdiagnosis of Wilson’s disease as it has symptoms that are similar to those of other diseases. Roberts E. (2006).

Treatment of Wilson’s disease

Treatment of Wilson’s disease requires being lifelong since it involves reducing and controlling of the amount of copper in the body. At the beginning of the therapy individuals undergo procedures that are aimed at reducing the high levels of copper in the body, the reduction of the amount of copper the body intakes and treatment of any damages that may have occurred to the central nervous system as well as the liver. Various drugs are used for the purposes of releasing copper from the organs into the bloodstream. This is usually done by drugs such as d-penicillamine and trientine hydrochloride. Walshie JM (2002). Kidneys then filter out the released copper which is then excreted in urine.

Pregnant women should however consume a lower dose of d-penicillamine or trientine hydrochloride for the purposes of dropping the risk of the children having birth defects. Other treatment of the disease includes the use of zinc which is usually administered through zinc salts such as galzin. Such salts aid in the blocking of the digestive tracts that absorb copper from food. However since zinc removes copper at a very slow rate it is not used solely as a medical solution for individuals who are already suffering from the disease. It is therefore used with the combination of the other treatment drugs.
After individuals suffering from the disease have improved their reaction to the initial treatment and the levels of copper are reduced to safe levels, it is then time to begin maintenance therapy. Maintenance therapy usually involves consumption of zinc and low levels of d-penicillamine or trientine hydrochloride. Individuals undergoing maintenance therapy should ensure that they visit health care providers for checking of the copper levels in the blood and urine. People who suffer from Wilson disease and are at the maintenance stage should ensure that they reduce their consumption of diets with copper intake. They should not consume foods such as liver and shellfish since they contain high levels of copper. Brewer GJ. (2000).

Conclusion

Individuals who realize that they have symptoms that are related to Wilson’s disease should seek medical attention from professional medical practitioners. This is because the accumulation of copper in the body can reach at high levels which may turn up to be fatal. While seeking medical check for Wilson’s disease, one should ensure that he or she approaches medical practitioners that are professional to avoid a misdiagnosis.

Reference:

Walshe JM (2002). "Treatment of Wilson's disease with trientine (triethylene tetramine) dihydrochloride".

