Iron and hemochromatosis

Health & Medicine

IRON AND HEMOCHROMATOSIS Iron Hemochromatosis is the most common inherited condition in Caucasian populations in North America affecting about 1 in 200-400 (this varies with source) people.

(5a) Describe this condition, consequences if left untreated and treatment.

Hemochromatosis is a hereditary disease and an autosomal recessive abnormality. That creates an iron overload in the body. In other words, the body absorbs and stores too much iron (Adams & Barton, 2010). Over time, the excess iron collects and accumulates in vital organs such as the liver, joints, heart, pancreas and the pituitary. This produces general symptoms, such as fatigue. The body cannot eliminate excess iron naturally, so the excess iron begins to damage the organs.

The HFE gene controls the quantity of iron that is absorbed in the intestine from food. A defect in this gene results in a mutation that disturbs this normal function, and iron metabolism is altered so that a large amount of iron is absorbed from the small bowel, about 1mg excess daily (Sardesai, 2012). The mutation most commonly causing hereditary hemochromatosis is when tyrosine is substituted for cysteine in the C282Y protein sequence. When a child inherits C282Y from both parents, the child is most likely to have hemochromatosis 0. 5% of the United States Caucasian population has hemochromatosis (Victoria, 2011).

Consequences if not treated

If untreated, about 90% of the excess iron remains in hepatocytes, which are the main cells that function in the liver. Most remaining iron is stored in the functioning cells of the pancreas, anterior pituitary gland, joints, and heart. The endocrine disorder can cause diabetes, pigmentation, and cirrhosis in severe cases. A person can die from cirrhosis, hepatocellular carcinoma,

cardiomyopathy, or diabetes (Powell & Fischer, 2005).

Treatment

Normal life expectancy can result if the disease is diagnosed early and if the person is given phlebotomy therapy. Therapeutic phlebotomy removes 200-250 mg of elemental iron for every 450-500 ml of blood. This is done weekly so that enough body iron stores are depleted (Adams & Barton, 2010). Before each treatment, the concentration of hemoglobin should be recorded. How much therapeutic phlebotomy is needed depends on the concentration of serum ferritin and hepatic iron. These are used as estimates of treatment needs. Excess iron is completely depleted when serum ferritin concentration is 10-20 ng/ml. This takes about one year (Gropper & Smith, 2013).

5-B) Would you expect the prevalence of the symptomatic hemochromatosis to increase or decrease with the introduction of an iron fortification program or an increase in the intake of bioavailable iron. Is there research to support your answer?

The body does not have a way to eliminate excess iron naturally, and serum ferritin must be kept at less than 50 mg/mol for the rest of the person’s life. So, fortifying food with iron means the person increases chances of iron overload, especially when the diet is rich in red meat, and vitamin C (JD, Charlton RW, 1978). Vitamin C is known to increase iron absorption. For instance, expression of the high rate of iron overload as well as associated manifestations in hemochromatosis homozygotes that have been diagnosed in medical care in Australia is attributed to the high national meat consumption rate.

Several researches have been conducted that support that introduction of iron fortification increases the prevalence of the symptomatic

hemochromatosis. The Brazilian government instituted a policy of universal or mass fortification of food to help curb high incidence of iron shortage in pregnant women and children (Reeve et al 2001). As a result of that nearly every citizen of the country began to ingest iron and folic acid daily, whether needed or not. Long term medicinal ingestion of iron is a severe and rare situation. The data from the study suggest that there are cases of hemochromatosis in women whose illness were accelerated by this therapy. The amounts of iron among the Brazilian citizens have, therefore, had an appreciable increase over time (Adams & Barton, 2010). Even though, iron fortification of foods is a good strategy in combating iron deficiency, many nations have abandoned this therapy because it has accelerated iron overload among the citizens. Prolonged ingestion of medicinal iron has caused many illnesses by overload, for instance, hereditary hemochromatosis, chronic hemolytic anemia, chronic liver disease. Hereditary hemochromatosis has a high prevalence in the Northern Europe countries. Also, many Brazilian publications have reported it. The disease is caused by an inappropriate increase in the iron absorption by the intestines. The surplus metal is accumulated in the tissues hence causing severe heart diseases, liver cirrhosis, dark skin, diabetes, splenomegaly, endocrine disorders and neuropathy (Olsson et al 1988).

Other studies have also suggested that iron fortification of food increases the severity of iron overload in those that have hemochromatosis. A test conducted on the treated hemochromatosis patients to ascertain the effects of food fortification with iron revealed that doubling the iron dose results to a proportional rise in the prevalence of the symptomatic hemochromatosis. For example, research conducted by Institute for Clinical and Translational

Science indicated that increased fortification in Sweden before 1995 resulted to an increase in the rate of iron accumulation in people with hemochromatosis. Similarly, iron fortification of wheat flour in the US accelerated the initial rate of iron loading in people with hemochromatosis. Accelerated evolution of the clinical disease particularly in susceptible persons is directly relative to the quantity of fortification iron added (Adams & Barton, 2010). It is, therefore, evident that the introduction of iron fortification increases the prevalence of the symptomatic hemochromatosis.

References:

A)

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