Medical Nutrition Therapy Diet: Wilson's Disease

1. Purpose

a. Nutrition Indicators:

Blood tests are one way to monitor one's liver function and look for copper in the blood. Blood may also be tested for the level of a protein called ceruloplasmin, which carries copper in the bloodstream. A Doctor may also use urine tests to measure the amount of copper excreted in your urine in a 24-hour period. (Mayo Clinic, 2010) High levels of copper in the blood may indicate Wilson's Disease; yet, other diagnostic measures must be taken to officially diagnose Wilson's Disease.

b. Criteria to Assign the Diet:

A diet low in Copper; use of chelating agents; increased intake of Zn; increased intake of Vitamin E.

c. Rationale for Diet:

It is necessary to decrease a patient's copper intake because the body already cannot properly utilize/metabolize copper. Chelating agents (penicillamine or trientine), help increase urinary excretion of copper. High-dose oral zinc interferes with absorption of copper from the gastrointestinal tract and is most effective after initial decoppering with a chelating agent. Vitamin E can be used with a chelator or zinc to prevent tissue damage, particularly to the liver.

2. Population

a. Overview:

Wilson disease is an inherited disorder of copper metabolism that causes a build up of copper in the body. The disease can present with hepatic, neurologic, psychiatric disturbances, or a combination of these, in individuals ranging from age three years to over 50 years.

b. Disease Process:

It is an autosomal recessive disease that occurs in 1 to 4 per 100,000 individuals. At conception, each sib of an affected individual has a 25% chance of being affected, a 50% chance of being an asymptomatic carrier, and a 25% chance of being unaffected and not a carrier. Prenatal testing for pregnancies is available.

The main sites of copper accumulation are the liver and the brain, and consequently liver disease and neuropsychiatric symptoms are the main features that lead to diagnosis. Neurologic presentations, psychiatric disturbances, and Kayser-Fleisher rings can result from high amounts of copper in the body due to poor metabolizing ability. Liver disease can also ensue if not treated properly. Neurologic presentations include movement disorders like tremors, rigid dystonia, gait disturbances and more. Psychiatric disturbances include: depression, neurotic behaviors, disorganization of personality, and potentially intellectual deterioration.

Kayser-Fleischer rings result from copper deposition in Descemet's membrane of the cornea and reflect a high degree of copper storage in the body.

c. Biochemical and Nutrient Needs:

First and foremost, foods high in copper should be restricted since the body cannot metabolize it effectively. Furthermore, zinc can prevent the development of hepatic, neurologic, and psychiatric findings in asymptomatic affected individuals and can reduce findings in many symptomatic individuals. Lifelong treatment with chelating agents is initiated as soon as possible. Such agents are penicillamine or trientine, and they increase urinary excretion of copper. High-dose oral zinc interferes with absorption of copper from the gastrointestinal tract and is most effective after initial decoppering with a chelating agent. Antioxidants, such as vitamin E, can be used with a chelator or zinc to prevent tissue damage, particularly to the liver.

3. General Guidelines

a. Nutrition Rx:

There is no specific nutrition prescription or requirements for patients with Wilson's Disease, with the exception that food choices should contain (<1mg Cu/serving).

RDA for Vitamin E: 20mg/day

RDA for Zn: 15mg/day

RDA for Cu: 2 mg → In patients with Wilson's disease daily intake should be <1mg/day.

b. Adequacy of Nutrition Rx

All other nutritional requirements/recommendations remain the same for patients with Wilson's disease. The only one that should be closely monitored is Cu.

c. Goals

A diet low in Copper with additional supplementation of Vitamin E and Zn will help keep Cu levels somewhat in balance, help protect liver tissue damage, and decrease the absorption of Cu into the GI system.

d. Does it Meet DRI

All nutrients should meet the DRI with the exception of Cu, but this is compensated by the inadequate metabolism of the mineral.

4. Education Material

a. Nutrition Therapy:

Provide patient with a chart of acceptable foods (low-Cu containing) and non-acceptable foods. Explain the pathophysiology to patient to help them understand the vital role that a low Cu diet plays in the progression of their disease. Provide sample menus for them to comply and use the readiness to change ruler to

assess self-efficacy.

b. Ideas for Compliance

Advise patient to group sessions with other patients suffering from Wilson's Disease. Groups are available through the Wilson Disease Association. When diagnosis is new, ask patients to keep a food journal to monitor their intake and to keep a close record of their food choices.

5. Sample Menu

a. Foods Recommended:

Foods low in copper (<1mg/serving), high in vitamin E, smaller portions of meat to decrease copper consumption, non-copper utensils, and water that contains < 100 micrograms per liter to demineralizes high copper amounts.

Acceptable meats include: beef, eggs, white meat turkey & chicken, cold cuts and frankfurters that do not contain pork, dark turkey, dark chicken, or organ meats. Most vegetables besides mushrooms and most fruits besides home-dried fruits are acceptable. Recommended beverages include: coffee, tea, fruit juices, fruit-flavored beverages, and lemonade.

b. Foods to Avoid:

Liver, brain, chocolate, mushrooms, soy milk, shellfish and nuts are all high in copper and should be avoided. Alcohol should also be avoided because it can harm an already compromised liver.

c. Example of a meal plan:

Sample Menu		
Breakfast	**Lunch**	**Dinner**
▪ grapefruit 1/2 cup ▪ scrambled egg 1 ▪ white toast 1 slice ▪ margarine 1 tsp ▪ skim milk 1/2 cup ▪ coffee 1 cup ▪ salt/pepper	▪ white bread 2 slices ▪ roast beef 2 oz lettuce tomato mayonnaise 1 Tbsp ▪ carrot sticks 6 ▪ fresh orange 1 ▪ skim milk 1/2 cup	▪ chicken breast 3 oz ▪ rice 1/.2 cup ▪ green beans 1/2 cup ▪ margarine 1 tsp ▪ white bread 1 slice ▪ jelly 1 tsp ▪ apple 1 ▪ lemonade 1 cup ▪ salt/pepper

This Sample Diet Provides the Following			
Calories	1520	Sodium	1500 mg
Protein	69 gm	Potassium	2150 mg
Carbohydrates	191 gm	Copper	0.68 mg
Fat	54 gm		

Provided by: Jackson Siegelbaum Gastroenterology. Low Copper Diet for Wilson's Disease. (2008). Retrieved on April 15, 2010, from, http://gicare.com/Diets/Copper-Restriction.aspx

6. Websites

a. Organizations with Websites

Jackson Siegelbaum Gastroenterology. Low Copper Diet for Wilson's Disease. (2008). http://gicare.com/Diets/Copper-Restriction.aspx

Wilson's Disease Association International. (2003-2010). http://www.wilsonsdisease.org/wilsondisease/dietandnutrition.html

Mayo Clinic. Wilson's Disease. http://www.mayoclinic.com/health/wilsons-disease/DS00411

b. Government Websites

National Institute of Health. Wilson's Disease. http://www.ncbi.nlm.nih.gov/bookshelf/br.fcgi?book=gene&part=wilson

7. References

a. Journal articles references

Lorincz, M. (2010). Neurologic Wilson's disease. *Annals of the New York Academy of Sciences*, *1184*(1), 173-187. doi:10.1111/j.1749-6632.2009.05109.x.

Shimizu, N., Yamaguchi, Y., & Aoki, T. (1999). Treatment and management of Wilson's disease. *Pediatrics International*, *41*(4), 419-422. doi:10.1046/j.1442-200X.1999.01093.x.

NUTRIENT

1. What is the nutrient?

Sodium

2. What is the RDA/DRI for the nutrient?

There is no defined RDA for sodium because it has never lacked in the diet, but the RDA's general rule is < 2,400 mg per day.

3. How is the nutrient metabolized?

Sodium is an electrolyte that helps regulate water levels in the fluid in and around one's cells. It helps transmit nerve impulses, and influences the contraction and relaxation of muscles. The kidneys regulate the amount of sodium kept in your body. When sodium levels are low, the kidneys conserve sodium. When levels are high, they excrete the excess amount in urine.

4. What are food sources of the nutrient?

Sodium occurs naturally in some foods such as: Milk, beets, celery and even drinking water. However, the most common form of sodium is sodium chloride, which is table salt.

Sodium is added to many food products in the form of: monosodium glutamate, sodium nitrite, sodium saccharin, baking soda (sodium bicarbonate), and sodium benzoate. These are ingredients in condiments and seasonings such as Worcestershire sauce, soy sauce, onion salt, garlic salt, and bouillon cubes.

Processed meats, such as: bacon, sausage, ham, canned soups and vegetables are all examples of foods that contain added sodium.

5. What disease states alter the nutrients' metabolism?

Kidney Disease: Too much sodium can be harmful for people with kidney disease because sodium helps one's body to retain a healthy fluid balance. But with renal disease, kidneys cannot eliminate excess sodium and fluid from the body. As sodium and fluid build up in the tissues and bloodstream, blood pressure increases.

Hypertension: (High Blood Pressure) Increased salt intake causes more fluid to be retained in the blood vessels. This increased volume of blood requires the heart to work harder to pump blood to all the tissues in the body. Increasing

the blood's volume within the enclosure of the circulatory system is one way that salt increases blood pressure. Salt can also elevate blood pressure through the action of the arterioles. Arterioles are blood vessels that dilate and constrict to regulate blood pressure and blood flow. By contracting under the influence of sodium, arterioles effectively increase the resistance to blood movement and lessen the volume of blood that is returned to the heart. This action also increases blood pressure. High BP can lead to heart disease, kidney disease or stroke.

Hypotension: (Low Blood Pressure) The causes of low blood pressure can range from dehydration to problems with the way your brain signals your heart to pump blood. Low blood pressure is treatable, but it is important to find out what is causing your condition so that it can be properly treated.

6. What are the tests or procedures to assess the nutrient level in the body?

A sodium test is done by drawing blood from the patient to assess the water and electrolyte balance of the body, find the causes of high and low levels, and/or to check the progress of kidney disease or adrenal glands.

Blood pressure is another diagnostic procedure to see if a patient has high blood pressure which is directly correlated with one's sodium intake. It is a quick, painless test using a medical instrument called a sphygmomanometer and measures the systolic and diastolic pressure between heartbeats. Normal is 120/80 mmHg.

7. What is the drug –nutrient interactions?

Drugs that can potentially interfere with diclofenac sodium include: diuretics, other nonsteroidal anti-inflammatory drugs, and some herbal supplements. These interactions may increase the risk of kidney damage, reduce the activity of diuretics, or make ACE inhibitors less effective, among other things.

8. How is the nutrient measured?

The nutrient is in milligrams.

9. What is the Upper Tolerable Limits?

A tolerable upper intake level (UL) -- is set at 5.8 grams of salt (2.3 grams of sodium) per day.

10. What are the physical signs of deficiency?

Low levels of sodium in the diet (hypotension) can lead to: dizziness, fainting, dehydration, lack of concentration, blurred vision, nausea, fatigue, thirst, serious heart, endocrine or neurological disorders. Shock can result when

extremely low levels are reached; depriving the brain and other vital organs of oxygen and nutrients.

11. What are physical signs of toxicity?

Signs of sodium toxicity include swelling in the extremities and high blood pressure. Sodium causes more calcium to be lost in the urine, and high contents of sodium can also increase risks for osteoporosis.