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SHORT COMMUNICATION

Low copper containing diet for Wilson disease patients

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Abstract

Copper is an essential trace mineral. It has various important functions in the body as it forms core component of proteins and metalloenzymes. Wilson disease is a genetic disorder where there is an inherited defect in the biliary excretion of copper. Wilson disease has an autosomal recessive mode of inheritance. Lifelong treatment with one of the several medications is mandatory for all confirmed Wilson disease patients. In addition to lifelong medications, to remove accumulated copper from various tissues, reduced intake of copper in diet is an important management principle. In this article, we present the content of copper in the common Indian diet and the dietary regulations for Wilson disease patients. The knowledge of dietary requirements which is important for both doctors and patients have been discussed in detail, highlighting the role of nutritionist in the management of Wilson disease.

Keywords: Wilson disease; copper diet; Indian food

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Introduction

Copper is an essential trace element for the human body. The body has complex mechanisms to ensure adequate supplies of copper without any toxic effects. It forms a core component of proteins and metalloenzymes, that play a role in essential metabolic functions. Copper is a co-factor in several important enzymes like cytochrome oxidase (mitochondrial electron transport chain), superoxide dismutase (protective against reactive oxygen species) and lysyl oxidase (cross-linking of collagen and elastin). Copper is also vitally important for brain function [1].

The source of copper for the human body is the food consumed and the absorption of copper depends on its copper content. The human gastrointestinal

system can absorb 30 to 40% of ingested copper from typical western diets. Copper absorption occurs primarily in the small intestine and a small fraction from stomach [2]. Liver maintains adequate concentrations of copper in plasma.

Recommended dietary intake

The Food and Nutrition Board at the Institute of Medicine recommends a daily intake of 2 mg/day of copper for adults. The following table (Table 1) shows the recommended dietary intake of copper for different age groups.

Table 1: Recommended dietary intake (micrograms per day).

Infants	0 to 6 months: 200 mcg/day
	7 to 12 months: 220 mcg/day
Children	1 to 3 years: 340 mcg/day
	4 to 8 years: 440 mcg/day
Adolescents and adults	9 to 13 years: 700 mcg/day
	Males and females age 14 to 18 years: 890 mcg/day
	Males and females age 19 and older: 900 mcg/day
	Pregnant females: 1,000 mcg/day
	Lactating females: 1,300 mcg/day

Note: Dietary Reference Index (DRIs): recommended dietary allowances and adequate intakes, elements (Food and Nutrition Board, Institute of Medicine, National Academies).

Discussion

Wilson disease (WD) is a genetic disorder where there is an inherited defect in the biliary excretion of copper. WD has an autosomal recessive mode of inheritance. Wilson disease is caused by mutations in a gene called ATP7B gene which has the genetic material to make copper transporting protein (ATPase). This is a trans-membrane protein of the Golgi network, responsible to remove excess copper out of the cell through ceruloplasmin. Mutations in ATP7B gene lead to an abnormal ATPase protein which is incapable of removing excess copper leading to its accumulation in different tissues such as liver, brain, kidney, bones, joints, cornea and other organs resulting in hepatic, neuropsychiatric, eye and other manifestations. The consequences can be very fatal, if not diagnosed and treated properly as it can lead to irreversible damage to brain and liver [3].

Lifelong treatment with one of several medications is mandatory for all confirmed Wilson disease patients to eliminate the excess copper accumulated in the body and to reduce absorption of copper [4]. In addition to these medications reducing the copper absorption also helps maintain normal level of copper in the body. This can be achieved by reducing the copper intake/consumption via food. Thus reduced intake of copper diet can help patients manage the disease more efficiently.

The estimated dietary intake of copper from a typical Indian diet is around 2 mg per day. Aim of diet management is to reduce the intake of copper for WD patients to less than 1 mg per day [5]. Tables 2 and 3 give the copper content of the most common food items & products available in the Indian diet. Patients with Wilson disease need to be assessed for the copper consumed which can help the treating physicians to give dietary advice to reduce the intake of copper [6]. Organ meats, nuts, seeds, chocolate and shellfish have high copper content [6]. Consumption of such foods is better to be avoided. It is advisable to consume food items that are processed and/or have a low copper content. Processed food products (e.g. wheat, maida) are almost reduced to 70% of copper of its initial content which makes them suitable for WD patients.

Another home based simple yet efficient method of removing copper is by boiling food (e.g. beans) reduces copper content by almost 50%. Another major source of copper is drinking water and hence it's important that the copper content of it is evaluated. If the water is over 0.1 ppm (parts per million) or 0.1 mg/L, an alternative water source of water needs to be considered or a good filtering system needs to be installed that is capable of removing copper. The amount of copper contained in drinking water varies highly, depending upon the natural mineral content, pH of the water and the local plumbing system. High concentrations are found in soft, acidic water conducted through a copper pipeline or in water from a system in which copper salts are added to control the growth of algae [6]. WD patients also need to understand that copper or copper alloy utensils should not be used for food preparation or consumption.

Table 2: Food items & products with high copper content in Indian diet.

Very high copper foods	High copper foods
Animal liver (lambs, pigs, cows)	Cereal grains (barley, millet)
Shell fish (oysters, crab, clams)	Pulses (Bengal gram, horse gram, red gram, lentils, kidney beans, soya bean)
Sesame seeds	Legumes (peas)
Baker's yeast	Nuts (chestnuts, pistachios, cashewnuts, walnuts)
Chocolates and cocoa powder	Oil seeds (gingelly, sunflower, pumpkin)
	Condiments and spices (green chillies, basil, black pepper)
	Lobster and fish
	Candy and fruit gums

Table 3: Food items & products with low copper content in Indian diet.

Medium copper foods	*Low copper foods*
Cereals (bajra, jowar, maize, ragi, sanwa millet, wheat)	Rice
Pulses (black gram, green gram, khesari dal, mothe beans)	Leafy vegetables (amaranthus, Brussels sprouts, cabbage, sorrel, lettuce, fenugreek, drumstick leaves, curry leaves, coriander leaves, mint, radish leaves, spinach, tamarind leaves)
Vegetables (celery, beet root, white radish, snake gourd, mushroom)	Eggs, butter, milk, milk products, cheese and yogurt
Nuts and oil seeds (almonds, arecanut, coconut, groundnut, mustard, piyal)	Roots and tubers (carrot, colocasia, onion, potato, pink radish, sweet potato, tapioca, yam)
Condiments and spices (asafetida, cardamom, cloves, coriander seeds, cumin seeds, fenugreek seeds, garlic, ginger, nutmeg, cumin, poppy seeds, turmeric)	Vegetables (bitter gourd, bottle gourd, brinjal, tomato, broad beans, cauliflower, cluster beans, cucumber, drumstick, field beans, French beans, okra, raw mango, ridge gourd)
Fruits (kiwi, lime, orange, pears, pomegranate, custard apple)	Fruits (pumpkin, apple, apricot, banana, cherries, gooseberry, dates, guvava, jack fruit, lemon, litchi, ripe mango, muskmelon, watermelon, peach, pineapple, plum, sapota)
Meat (duck, goose, rabbit)	Freshwater and marine fish
Sweets (jams, fruit cakes, pudding, peanut butter)	

Conclusion

Important points for physicians and Wilson disease patients to remember are that high copper containing diet or products should be strictly avoided. Patients can consume food products with low copper diet and should restrict themselves from consuming only the recommended products on a regular basis.

Conflicts of interest

Authors declare no conflicts of interest.

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