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# Dietary modifications in thalassemia

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**Abstract**
It is estimated that there are currently about 270 million abnormal hemoglobin and thalassemia carriers in the world, of which 80 million are carriers of beta thalassemia. It is divided into two classes, called alpha and beta thalassemia. Being a hereditary disease, thalassemia is very difficult to control. It can be prevented by strong public awareness, early detection, adherence to proper rules and proper treatment. Most patients have it without symptoms, so many people do not even know for a lifetime that they are carriers of thalassemia. It is slowly spreading from country to country through intermarriage between carriers. People with thalassemia major and intermedia (in some cases) require regular or frequent blood transfusions. In addition to this type of thalassemia patient, all types of thalassemia carriers may have differences in iron overload, calcium deficiency, vitamin C, and folate modulations. Therefore, it is very important for thalassemia carriers or patients to take special care of their diet to stay healthy. This paper will provide dietary suggestions for thalassemia carriers as well as thalassemia patients.

**Keywords:** Thalassemia, thalassemia carriers, dietary modifications

## Introduction
In Greek "Thalassa" means sea and "Haema" means blood. The word "thalassemia" refers to a disorder associated with defective synthesis of α or β-globin subunits of haemoglobin (Hb) A (α2;β2), inherited as pathologic alleles of one or more of the globin genes located on chromosomes 11 (β) and 16 (α). Nowadays, thalassemia represents the most common single gene disorder in the world [1].

Hemoglobin is an iron-rich protein in red blood cells (RBCs) that carries oxygen to cells all throughout the body. This protein has two parts – one is called alpha globin and the other is called beta globin. Alpha thalassemia may be a sort of thalassemia that's acquired (passed down through families). It is a blood condition that impairs the body's ability to form healthy red blood cells and normal hemoglobin. Certain genes regulate how the body produces hemoglobin. Thalassemia arises when these genes are mutated (changed) or lacking. Alpha globin is the hemoglobin protein that is impaired in alpha thalassemia. Carriers are people who inherit hemoglobin genes that are altered or absent from one parent but normal genes from the other. Alpha thalassemia carriers normally have no symptoms. They can, however, pass on the defective genes to their offspring. People with moderate to severe alpha thalassemia have defective hemoglobin genes acquired from both parents. Alpha thalassemia is one of the world's most frequent blood diseases. Alpha thalassemia may affect anyone. Every year, thousands of babies are born with alpha thalassemia trait or hemoglobin H illness, although it is most frequent in Southeast Asia. It is also common among people of African, Greek, Italian, and Middle Eastern ancestry [1].

On the other hand, the annual incidence of symptomatic beta thalassemia individuals is estimated at 1 in 100,000 worldwide. Thalassemia cannot be prevented as it is inherited. It will be passed on from generation to generation. However, these blood disorders can be detected before birth through prenatal tests. The beta Thalassemia have four clinical features: two conditions are generally asymptomatic; a silent carrier state and a beta thalassemia trait. These Thalassemia usually result from the inheritance of one mutant beta-globin gene. Two forms of beta thalassemia require medical management, namely thalassemia major and intermedia [1].

Treatment of thalassemia is based on the type and severity of thalassemia. People who are carriers of alpha/beta thalassemia or have any type of thalassemia trait usually have mild or no symptoms and need little or no treatment. Thalassemia patients depend on transfusions for their entire lives and face severe health problems. However, thalassemia carriers and patients should follow proper dietary guidelines along with other preventive measures and treatment strategies.

**Iron overlord in thalassemia**

The required daily iron for a man over 18 years of age is 8.7 mg, while it is 14.8 mg for women aged 19 to 50 years. Consumption of more than this amount is dangerous for thalassemia patients. However, the recommended daily intake of iron in the US is 18 mg. That is, if a food label says that it contains 8% of the daily recommended iron, multiply 0.08 by 18 milligrams to get milligrams of iron from it. Outdoor foods can be checked and eaten in this way. Iron overloading is a common consequence due to its various toxic effects in thalassemia individuals.

**Iron overload is widely assessed by**

(i) serum ferritin concentration (limitation: poor reliability due to it is influenced by many other factors, including biochemicals arise from the liver damage); (ii) liver biopsy (however, it is an invasive technique and can be affected by hepatic fibrosis (due to HCV infection) and irregular iron distribution in the liver, leading to false negative results) [3]; (iii) magnetic biosusceptometry (SQUID) (however, it is an expensive technique); (iv) MRI techniques (in this case liver and heart samples are commonly used, and it is a reliable method) [4].

**Iron chelation therapies and their side effects**

Beta thalassemia major (in some cases, intermedia) individuals is the most common consequence of iron overload in beta thalassemia major (in some cases, intermedia) individuals. This also occurs in alpha thalassemia patients receiving blood from healthy persons. This can be further prevented by appropriate and adequate iron chelation therapy.

a) Desferrioxamine B (DFO): It is helpful to manage secondary effects of iron overload, thereby, consistently decrease in morbidity and mortality [5]. Low iron burden, ocular and auditory toxicity, growth restriction, renal impairment, interstitial pneumonitis, and susceptibility to *Yersinia* infections are some of the detected side effects of DFO.
b) Deferiprone: It is a bidentate oral chelator. Arthropathy, gastrointestinal symptoms, neutropenia, and agranulocytosis are some of the side effects of it [6].
c) Deferasirox: It is effective in adults and children thalassemia individuals. Gastrointestinal disorders, skin rash, mild non-progressive increase in serum creatinine concentration [7], renal failure, hepatic failure, cytopenias, and gastrointestinal hemorrhage are some of the reported side effects of this therapy.
d) Combination therapies: Combination chelation therapy, such as desferrioxamine and deferiprone have been found effective in severe iron overload individuals. It is also evident to reduce myocardial siderosis, improve cardiac and endocrine function, reduce liver iron and serum ferritin concentration, reduce cardiac mortality, and improve survival rate and reduce toxicity in patients [8, 9]. In another study, the deferasirox-desferrioxamine combination decreased liver iron accumulation in heavily loaded individuals and lowered myocardial overload in 33% of patients [10]. Deferasirox-deferiprone is another combined chelation therapy in Thalassemia [11].

**Causes of iron overloading in thalassemia individuals**

Iron overload may happen in thalassemia individuals due to (i) blood transfusions (which are responsible for the accumulation of excess iron in their bodies), and (ii) excess iron absorption and accumulation in different parts of the body (which increase the absorption of iron in the diet by the digestive system and store it in the liver and other organs). In the latter case, the amount and rate of iron absorption vary according to the degree of erythropoiesis. The hemoglobin (Hb) level and some other potential independent factors also have an influence on it. Black tea after meals may reduce dietary iron absorption, especially in thalassemia intermedia individuals [12, 13]. Excess iron accumulates in the liver as a result of regular blood circulation. When this is done, iron builds up in places like the heart and pituitary, which can cause multifactorial damage to the body. Thalassemia individuals should avoid certain foods containing high amounts of iron (e.g., liver, numerous baby foods, cereals, and multivitamin (added iron) preparations, etc.).

**Modulation of vitamin C levels**

Iron overload oxidizes vitamin C, thereby resulting in vitamin C deficiency in some thalassemia individuals. In contrast, vitamin C can increase gut iron absorption, leading to iron toxicity in some individuals [14]. Therefore, people with thalassemia should take foods rich in vitamin C, such as vegetables, fruits, etc., on the advice of a doctor.

**Effects of alcohol consumption on thalassemia**

Alcohol facilitates oxidative damage effects of iron, especially in hepatitis B virus (HBV) and hepatitis C virus (HCV) patients' liver tissue [15].

**Folate deficiency**

Thalassemia children, especially those who have taken blood from other people's bodies, are still relatively anemic, so their bodies may be experiencing iron deficiency. Children should be more careful about their diet, as most children prefer foods high in iron, such as beef (dried), liver, and fast food. Meat iron is usually absorbed much more easily from other sources, such as cereals and bread. Some thalassemia patients have an increased need for folate (folic acid), which can lead to folate deficiency [16]. In such cases, a supplement (1 mg/day) folate can be given. Some foods rich in folic acid, such as broccoli, Brussels sprouts, vegetables, etc. can be eaten [17].

**Calcium depletion in thalassemia**

Since thalassemia has many causes, it stimulates the depletion of calcium in the body of all but those who do not have nephrolithiasis, so adequate calcium-rich foods (such as milk, cheese, dairy products, and kale) are always recommended [17].

**Thalassemia carriers or patients should follow the following guidelines**

1. Avoid using cast iron cookware because it can transfer iron from the cooking pot to the food.
2. Some non-vegetarian (protein) or non-vegetarian foods should be avoided or reduced from the food list, such as oysters, liver, pork, beans, beef, peanut butter, tofu, etc.
3. Some grains should be avoided or reduced from the food list, such as flour, tortillas, baby cereals, wheat

cream, malt-o-mill, cereals, for example, most of the products 19 (breakfast made by Kellogg's including corn; this contains light sweet flakes made with oats, wheat, and rice; rich in all the vitamins and iron needed daily), total, kicks, all bran, life, resin bran, special k, 100% bran, rice checks, rice crispy, cornflakes, wheat etc.

4. Fruits or vegetables which should be avoided or omitted from the diet, such as: prune juice, prune, watermelon, spinach, green vegetables, dates, raisins, broccoli, peas, fava beans, etc.
5. Avoiding iron-rich foods and even multivitamins (which contain iron).

## Conclusion

Effective detection of iron overload and the usage of appropriate chelation therapy will be helpful to manage thalassemia along with the maintenance of quality lives, especially dietary modification in the whole lives of this class of patients.

## Conflict of interest

None declared.

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