

DIET IN REGULARLY TRANSFUSED THALASSEMICS

Jagdish Singh
N.K. Mangal
Y.R. Khinchi

The more pressing priorities of the families of thalassemics on regular transfusions, viz., availability of safe blood, proper administration, reaction to transfusion, filters, desferal, infusion pumps, monitoring of clinical and hematological parameters, vaccinations, splenectomy, bone-marrow transplantation, carrier detection in family, ante-natal diagnosis, *etc.* have unfortunately pushed the subject of their dietary requirements into the background. This article would focus on this neglected aspect in the management of regularly transfused thalassemics.

Vitamin C and Iron

Iron overload in regularly transfused thalassemics is a well known fact. The ferric form of these large iron stores leads to oxidative catabolism of a significant

*From the Department of Pediatrics Medicine,
Sir Padampat Singhania Mother and Child
Health Institute, SMS Medical College,
Jaipur.*

*Reprint requests: Dr. Jagdish Singh, C-3,
Doctor's Enclave, Gangwal Park, Jaipur
302 004.*

proportion of available vitamin C, resulting in low levels of the vitamin in the body(1,2). This is beneficial in patients, with iron load due to four reasons: (i) Causing disproportionate increase in reticuloendothelial (RE) iron stores over parenchymal iron stores(3); (ii) Leading to deposition of iron as hemosiderin instead of as ferritin(4,5); (iii) Decreasing release of iron atoms from ferritin(6), and (iv) Attenuating damage of tissue membranes by iron(7).

This is typically seen in the Bantu tribe of South Africa where the resultant low levels of vitamin C protects them from the detrimental effect of iron overload. Excess iron over-load occurs in them due to the practice of eating maize cooked in iron pots and drinking large quantities of beer brewed in iron pots thereby ingesting as much as 100 mg of iron each day(8,9).

Vitamin C in Non-Chelators

It would thus be reasonable to achieve low levels of vitamin C in regularly transfused thalassemics not on any form of chelation therapy. In addition to the benefits mentioned above, it would also result in reduction in absorption of iron from food. At the same time care should be taken not to precipitate overt clinical vitamin C deficiency.

Vitamin C in Regulator Chelators

In patients on regular chelation therapy the story is different. Here, vitamin C administration is useful as it would release iron atoms from ferritin molecule which would then get chelated thereby greatly enhancing urinary iron excretion(10). Supplemental vitamin C should be added after few weeks of starting regular chelation,

although a few studies have advocated that vitamin C status should be restored to normal before starting chelation(11,12). Vitamin C should be stopped in case chelation therapy has to be discontinued. Furthermore, the oral dose of 200 mg of vitamin C(13) should be given an hour or two after desferrioxamine (desferal) infusion has been started to achieve optimum results. Dietary vitamin C will enhance iron absorption in these patients. However, it is likely to be relatively small compared with the substantial increase in iron excretion produced by vitamin C supplementation used in conjunction with subcutaneous desferal.

Decreasing Iron Intake

Increased rate of erythropoiesis in thalassemics leads to increased iron absorption, despite iron overload in the body(14). This calls for caution about the amount and form of iron ingested and contamination of food with iron.

Form and Absorption of Iron

There are two major forms of dietary iron: (a) Heme iron, which is derived from hemoglobin and myoglobin and contained in meat, liver, kidney, *etc.*, and enters the mucosal cells of upper GIT unchanged; (b) Non-heme iron, which is derived from cereals, fruits, vegetables, *etc.* and absorbed exclusively as ferrous form. Vitamin C increases absorption by reducing ferric to ferrous form. Other substances which increase absorption are citric acid, sugars and meat(15). Rise in iron-phosphorus ratio leads to increased absorption(14). A high phosphorous diet impairs absorption by forming insoluble ferric phosphate. Phytic acid, which is present in most cereals, converts both ferric and ferrous salts into insoluble phytates, and may thus impair absorption(16). Oxalates and carbonates simi-

larly retard absorption(17). Tannins in leaf form a complex with iron rendering it unavailable for absorption(18). This effect appears to be specific for non-heme iron(19).

Contamination of Food with Iron

Food is sometimes contaminated with iron, more so in developing countries from residual soil on vegetables and cereals or with dust which settles on the surface of foods. Iron contamination may also occur during food handling (*e.g.*, milling), or cooking in iron utensils. Water used in cooking or drinking may also be contaminated with iron(20). Although very little is known about the bioavailability of contaminating iron, avoid keeping water or cooking in iron utensils, wash vegetables and cereals properly before cooking and keep food items always covered.

Conclusion

Regularly transfused thalassemics, not on chelation therapy should restrict eating food rich in vitamin C. In case the food is excessively rich in vitamin C, it should be taken only after cooking, which destroys vitamin C. In those receiving regular chelation therapy allow diet rich in vitamin C, and after a few weeks of starting chelation add supplemental vitamin C on the days the child is on chelation.

Thalassemics whether or not on chelation therapy must avoid food with high iron content(21,22), *e.g.*, meat, liver, kidney, egg yolk, green vegetables, certain fruits and jaggery. In case it is taken, the diet should include food high in phosphorous(21,22), *e.g.*, bread, cereals, milk, moong dal, soya bean, roasted peas, *etc.* and phytic acid *e.g.*, cereals, and exclude meat, sugars and rich sources of vitamin C(21,22), *e.g.*, guava, citrus fruits and juices, amla,

tomato, *etc.* Strong tea may be taken along with or just after meals to retard iron absorption.

If the patients follow these guidelines they shall be able to significantly cut down the absorption of iron. This will enable to mitigate although to a small extent the degree of iron overload.

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