

• In addition to a diet rich in calcium and vitamin D, your doctor may recommend taking calcium and/or vitamin D supplements. Check with your doctor about the need for these supplements.

What can be done to treat low bone mass?

Following all of the above prevention measures is important in treating low bone mass, to help insure that there is no further decrease. In addition, some doctors may prescribe a drug in the bisphosphonate family, such as Pamidronate or Etidronate. Some research trials have indicated that IV administration of Zoledronic acid may be beneficial to patients with thalassemia and osteoporosis; other trials are investigating whether increasing zinc intake may have a beneficial outcome for this patient population. Future studies will prove helpful in assessing the value of these and other options.

If you have further questions about osteoporosis and thalassemia, talk with your health care professional or contact the Cooley's Anemia Foundation at (800) 522-7222 or info@cooleysanemia.org.

The Thalassemia Centers of Excellence have the most highly trained thalassemia experts in the country. They are located at:

Children's Hospital Boston
 Children's Hospital Los Angeles
 Children's Hospital Oakland
 Children's Hospital of Philadelphia
 Children's Memorial Hospital (Chicago)
 Weill Medical College of Cornell University (New York)

Many other hospitals are satellite centers affiliated with these Centers. Please contact the Cooley's Anemia Foundation for a list of these satellite centers.

Published by the Cooley's Anemia Foundation
 330 Seventh Ave., #900, New York, NY 10001
www.cooleysanemia.org (800) 522-7222



This publication is made possible by an unrestricted educational grant from Novartis Pharmaceuticals.

The information in this publication is for educational purposes only and is not intended to substitute for medical advice. You should not use this information to diagnose or treat a health problem or disease without consulting a qualified health care provider. The Cooley's Anemia Foundation strongly encourages you to consult your health care provider with any questions you may have regarding your condition.

Low Bone Mass in Thalassemia

by Janice Beatty, RN, BSN and Craig Butler



Why is low bone mass an issue in thalassemia?

Having bones that grow and develop into strong, healthy bones is important for everyone. Low bone mass refers to weakness of the bones or bones that are not as strong as they should be.

There are many factors that may determine why any individual has low bone mass, including the genes s/he inherited from parents, dietary patterns and the amount of weight-bearing exercise the individual engages in regularly. However, people with thalassemia are also more prone to develop bone mass difficulties due to factors specifically related to thalassemia or its treatment, such as anemia, overactive bone marrow, excess iron deposited in the bones, reactions to deferoxamine and endocrine-related problems such as delayed puberty or hypogonadism.

What is osteopenia?

Osteopenia occurs when a person has low bone mass, but not so low as to be diagnosed with osteoporosis. (Sometimes osteopenia is defined as "reduced bone mass," while osteoporosis is defined as "low bone mass").

The World Health Organization defines osteopenia as occurring when a person's Bone Mineral Density (BMD) T-score is between -1 and -2.5. The WHO defines osteoporosis as occurring when a person's BMD T-score is lower than -2.5.

What is osteoporosis?

Osteoporosis is defined as a thinning of the bones, with a reduction in bone mass due to a loss of calcium and bone protein. In osteoporosis, bones become porous and brittle, are unable to support weight easily and fracture more readily.

Living with Thalassemia

Issues in Thalassemia Care

What are problems associated with low bone mass?

A person with low bone mass, especially osteoporosis, is more likely to experience fractures. Once fractured, bones may take longer to heal or heal more poorly than the bones of a person with normal bone mass.

Osteoporosis can affect a person's posture, impair physical activity and mobility and may create some physical changes as the skeletal system becomes increasingly affected.



Although any bone in the body can be affected by osteoporosis, the bones most vulnerable to fracture tend to be in the hip, spine, wrist and ribs.

How is low bone mass diagnosed?

Most people who have low bone mass are unaware of it; bone loss may occur for a long time without any visible symptoms. As a result, it is often undiagnosed until after a fracture occurs. Because it occurs with such frequency in thalassemia, individuals with thalassemia intermedia or thalassemia major should be checked regularly by having a Bone Mineral Density (BMD) test on an annual basis starting at around 8 years old. BMD is measured by a dual energy x-ray absorptiometry test, commonly called a DEXA scan. The BMD measurement will enable your doctor to determine your T-score or Z-score and to determine if you have osteopenia or osteoporosis. The doctor should also check nutritional status and vitamin levels (especially calcium and vitamin D).

What are T-scores and Z-scores?

A T-score measures a patient's BMD against that of a normal, healthy 30-year-old. A score of "0" means a patient's BMD is equal to that of a normal, healthy 30-year-old. A score above 0 means the patient's BMD is greater than normal; a score below 0 means it is lower than normal. As mentioned above, a score of -1 to -2.5 indicates osteopenia; a score lower than -2.5 indicates osteoporosis.

A Z-score measures BMD compared to a typical, healthy person whose age is the same as the patient. Because low bone mass can occur at a much younger age in thalassemia than in the general population, a Z-score may provide a physician with information that is more relevant in assessing bone mass in a person with thalassemia.

What can I do to prevent low bone mass?

Because thalassemia makes them predisposed to low bone mass, people with thalassemia should take extra efforts to keep their bone mass at healthy levels; some steps that can be taken include:

- An appropriate transfusion regimen, as determined with your doctor. As anemia and overactive bone marrow are thought to contribute to osteoporosis, keeping hemoglobin at an appropriate level can decrease the risk of developing low bone mass.
- Maintaining adequate chelation therapy. Excess iron in the bones is a factor in low bone mass, so rid the body of as much iron as possible.
- Treatment of endocrine issues that may affect bone mass, such as delayed puberty/hypogonadism. Some doctors may prescribe sex hormones to treat the latter.
- Avoiding smoking.

- Regular exercise. Patients should first discuss an appropriate exercise schedule with their doctor that takes into account any special needs before embarking on any exercise routine. For the general population, the National Osteoporosis Society recommends that a person engage in three or four 20-30 minute exercise sessions per week, with the exercise focusing on weight-bearing activities. For adults, some appropriate activities may include: brisk walking, jogging, running, aerobics, step classes, dancing, circuit training.



- Maintain a diet rich in calcium and vitamin D. This diet must also take into account restrictions that patients may have in terms of excess iron, heart issues, diabetes, or other factors; consulting a nutritionist who understands your specific issues is advised.

Following are some foods and drinks that are good sources of calcium:

- * Dairy products such as milk, yogurt and cheese (Note that skim milk actually contains more calcium than regular milk)
- * Dark green leafy vegetables such as broccoli, collard greens, spinach, turnip greens, Brussels sprouts and bok choy
- * Also tofu, okra, white beans, baked beans, rhubarb, peas, nuts, whole wheat bread

Again, remember that selecting the right mix of calcium-rich foods for your diet must also take into consideration other issues that may affect your dietary choices.