

What Makes Bones Fragile?

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Long bones (in the arms and legs) are in a way like bamboo: a hard thick outer part and a spongy inner part. The hard outer part is called the cortex, the inner part is trabecular or webbed and is called cancellous. Bone is made up of both organic (living) and inorganic (mineral) parts. Calcium and phosphate minerals are the important inorganic components. The organic part of bone consists of bone making cells (osteoblasts) and bone destroying cells (osteoclasts) as well as their supportive matrix that holds everything together (like the atmosphere on Earth). The matrix is made up of collagen and its various linker proteins. Collagen is a twisted helix - like a rope.

Defects in any of these important components can make bone weak. A diet poor in calcium or phosphate can cause weak bone; so drink your milk or something with vitamin D and calcium! People may also be born with disorders that affect the other components of bone. One example is Osteogenesis Imperfecta (OI). People with OI have weakened bone due to various genetic defects in the bone matrix. In OI types I-IV, the genetic defect causes the collagen to be abnormal. Think of this like a frayed, weakened piece of rope that is trying to hold the bone together. It can snap easily, often with little trauma.

As we learn more about the diseases of bone, better treatments become available. One treatment now available for OI is Pamidronate. This drug inhibits osteoclasts (bone destroying cells) to allow more bone to be made which makes it stronger. Sometimes, metal rods are inserted in the hollow middle part of the bone to help support it.

The newer type of rod can telescope so that as a child grows, the rod can grow too.

For additional information about Osteogenesis Imperfecta - you can visit the Osteogenesis Imperfecta Foundation's website at www.oif.org.