

Loeys-Dietz Syndrome: Bone Health in the Pediatric Population

Some individuals with Loeys-Dietz syndrome (LDS) may have an increased fracture risk due to poor bone health. Because each person with LDS is different, it is important to talk with your doctor about what care is right for you.

WHY BONE HEALTH MATTERS?

Bone health is a key part of everyday life. Although it may not be considered daily, our bone health status significantly impacts quality of life. Healthy bones are strong and can endure more impact without breaking than bones that are less dense. Bone density measures the amount of tissue in the bone. Less bone tissue will cause bones weakness and increase likeliness of fractures (breaks). Understanding bone health and proactively improving bone health early in life can decrease risk for osteoporosis and bone fractures in the future.

LOEYS-DIETZ RISK FACTORS:

Loeys-Dietz increases your risk for poor bone health. Some risk factors include:

- Nutrition
- Chronic inflammation that occurs due to GI tract and allergic disorders.
- Steroid medications (treatment for GI and allergic disease) weaken bones
- Reduced physical activity prevents bones from becoming strong
- Genetic differences from LDS causes the bones to form differently from the typical person. The bones aren't as strong.

Poor bone health increases risk for fractures in small events that may not cause fractures in other people.

In people with LDS

- Forearm and wrist are the most common areas of injury.
- falling down is the most common way to get injured.
- 50% risk of fracture by the age of 14
- there is low bone mineral density (BMD) on specialized testing
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REGULAR CHECK-UPS AND DXA SCAN

Talk to your child's pediatrician about fracture and bone health history. They may recommend a DXA scan.

DXA (said like 'Dexa') is an acronym for "dual-energy X-ray absorptiometry." It is the preferred and most widely used tool to assess bone status in children. This is because it has very low radiation and takes little time. DXA works by shooting x-rays at the bones and measuring how fast the rays travel through the bones to determine the density. DXA is performed less often in children, so it is important to have a DXA scan evaluated by a bone health specialist (or endocrinologist) who knows how to adjust for child height. A typical radiologist may not do this adjustment. It is important the radiologist scans either the spine, or total body minus head for the DXA measurements. Other areas that are appropriate to scan in adults are not helpful to scan in children. The results of the DXA should be the Z-score, and not the T-score. A T-score indicates how BMD (bone mineral density) compares to that of a healthy 30 year old. A Z-score compares BMD to an "age-matched" control. Your doctor will then help decide how frequently this should be performed to determine if interventions are helping.

MANAGING BONE HEALTH WITH NUTRITION AND PHYSICAL ACTIVITY

The best way to manage your children's bone health is to lead by example. Your children need proper nutrition and physical activity in order to maximize bone health during development. Osteoporosis does not have a cure, so it is important to be proactive. Eat foods rich with calcium and vitamin D. If it isn't possible to eat enough calcium with food, it is okay to take supplements. Your doctor may recommend high dosing of supplements that require a prescription.

Good foods to get calcium are:

- Milk
- Cheese
- Yogurt and frozen yogurt
- Almonds
- Leafy Greens like kale (please note, for children on blood thinners, check with INR clinic before any dietary changes)
- Calcium fortified orange juice

Bones grow stronger with use, so including weight bearing activity in addition to non-impactful activities, like swimming, will help to strengthen bones. It is important to participate in activities that are safe for people with Loeys-Dietz syndrome. Weight-bearing activity does not necessitate intense cardiovascular work. It is possible to strengthen bones with gentle exercise. Because falling is the main cause of fracture, it is also important to work on balance and coordination in addition to strengthening.

Physical activities that will strengthen bones:

- Walking or hiking
- Fencing
- Activities performed for fun and in moderation such as tennis or soccer



OTHER STRATEGIES TO MANAGE BONE HEALTH

- Some medication use can increase risk for lower bone density. Anti-inflammatory medications can decrease the amount of calcium that is absorbed from foods. If steroids are needed to treat allergies or GI tract issues, then try to be on the lowest dose as possible. Talk to your doctor about your concerns.
- Limit soda intake and instead drink calcium rich beverages like chocolate milk as a treat.
- Don't smoke. It is never too early to educate your children about the dangers of smoking.
- Make sure your children aren't participating in activities that are dangerous, and will increase their risk for breaking bones.
- Limit screen time that will interfere with physical activities.
- In some children with severely decreased BMD, a type of medication called bisphosphonates may be indicated. This is typically given by an IV infusion over a specified amount of time and should be given under the care of a bone health specialist.

