

What is it?

Osteogenesis Imperfecta (OI) is a genetic disorder characterized by easily breakable bones often from little or no apparent cause or stress. It is often called “brittle bone disease.” Strong bones usually form around collagen; the major protein of the body’s connective tissue. But in OI, the body is unable to make strong bones because of a mutation in the collagen gene. There are several types of OI and the characteristics of each can differ from person to person. Type 1 is the mildest form, while type 2 is the most severe. People with OI not only suffer broken bones, but they also can experience muscle weakness, loose joints, skeletal deformities, short stature, delayed motor skills, brittle teeth and hearing loss. Below are the four well-known types of OI, although 3 additional forms (5-8) have also been identified.



OI Type 1	Most common, mildest form. Bones fracture easily but often before puberty with minimal bone deformity. Fractures are normally spiral. Sclera (white of the eyes) is usually purple, blue or gray tinted in color. Collagen structure is normal, but less than normal amount is present.
OI Type 2	Most severe, often lethal soon after birth related to respiratory complications. Bones can appear crumpled and fractured even before birth. Severe bone deformity results. Also suffer a narrow chest with underdeveloped lungs and unusually soft skull bones. Tinted sclera similar to type 1. Collagen is improperly formed.
OI Type 3	Fractures and healed fractures are often present at birth. Tinted sclera similar to type 1. Loose joints and poor muscle development in extremities. Barrel shaped rib cage, spinal curvature, triangular face. Collagen improperly formed.
OI Type 4	Severity classified between Type 1 and 3. Bones fracture easily but often before puberty. Sclera are white or near white in color (normal). Bone deformity is mild to moderate. Barrel shaped rib cage, spinal curvature, triangular face.

What is the treatment?

There is no cure for OI. Instead, multidisciplinary treatment aims to prevent symptoms, develop muscle, strengthen bone mass and maximize independence. Fracture care is similar to any other fracture with splinting, casting and bracing for bone stabilization and healing. PT/OT services will help to strengthen muscle and promote function. For some fractures, surgical rod placement in long bones or “rodding” can be used to help with positioning and to strengthen and prevent fractures.

Some medications are prescribed to treat OI. These include bisphosphonates to increase bone mineral density. Some bisphosphonate medications have also been shown to reduce pain. Other treatments for OI include vitamin D and calcium supplements, oxygen for lung issues, dental care for teeth, and hearing aids for hearing loss.

What should monitoring include?

- Fractures
- Joint laxity
- Spinal curvature
- Muscle strength
- Pain
- Muscle spasms

Signs of fracture

- Swelling, bruising, tenderness
- Pins and needles sensation
- Child pale and sweating
- Pain (but doesn't always cry)
- Painful to bear weight
- Not moving, holding limb close to body

Suggested school accommodations

- Mobility accommodations
- Elevator access
- Extra time between class
- Hallway safety
- Assistive devices for hearing loss
- Extra set of books for home
- "Buddy" to help carry supplies
- Preferential seating in classroom
- Pace activities or offer rest break
- Keep areas clutter free
- Adapted PE
- PT/OT/Hearing consult
- 1:1 support as needed
- Training for school staff regarding condition

SHNIC school nurses information:

Specific health issues for individual health care plans

- Diagnosis
- Documentation of associated medical problems (hearing loss, scoliosis, etc.)
- Current medication list, note side effects
- Past hospitalizations including fracture and surgical history
- Nutrition orders including vitamins and supplements
- Emergency care plan for fractures, falls, injuries, etc.
- Risk assessment prior to start of school
- Orders for supervision and safety needs
- Orders for orthotics, braces, splints and other adaptive equipment
- Orders for limitation and/or mobility restrictions
- Fire evacuation plans
- Educate staff on injury or fall protocol and response plan
- Transportation accommodations
- Adaptive PE

Resources

Osteogenesis Imperfecta Foundation (OIF)

<http://www.oif.org/site/PageServer?pagename=fastfacts>

OIF: Plan for success– An educator's guide to students with OI

<http://www.oif.org/site/DocServer?docID=905>

OIF: Resources for school

<http://www.oif.org/site/PageServer?pagename=LivingSchools>

NIH: Child health and human development

<https://www.nichd.nih.gov/health/topics/osteogenesisimp/conditioninfo/Pages/treatments.aspx>

Brittle Bone Society

<http://www.brittlebone.org/>