

OSTEOGENESIS IMPERFECTA

School Age

Osteogenesis imperfecta (OI), also known as brittle bone disease, is a genetic disorder characterized by fragile bones that break easily. OI affects both bone quality and bone mass. A person is born with this disorder and is affected throughout his/her life. People with OI have less collagen than normal or a poorer quality than normal. It is not caused by too little calcium or poor nutrition. Approximately 35% of children with OI are born into a family with no family history of OI. It is estimated that OI occurs once in every 12 000 to 15 000 births. OI occurs with equal frequency among males and females as well as across races and ethnic groups.

Treatments focus on minimizing fractures and maximizing independent function and general health. Treatments focus on minimizing fractures and maximizing independent function and general health.

Types of OI

OI is highly variable, ranging from a mild form with no deformity, normal stature and few fractures to a form that is lethal during the perinatal period (prior to and after birth). The specific medical problems a person will have depend on the degree of severity. The characteristic features of OI vary greatly from person to person, even among people with the same type of OI, and even within the same family.

SAFETY PRECAUTIONS (school-aged children)

Architectural Barriers

Common barriers include steps at school entrances or between floors; restrooms with narrow or heavy doors, high sinks, stalls too narrow for a wheelchair; play structures and hands-on work areas (e.g., science lab, woodworking classes) that are inaccessible to a child who uses a wheelchair or is of short stature.

Architectural barriers can be overcome in a variety of ways;

- providing an aide to assist a child in the restroom
- portable ramps and wheelchair lifts
- lowering lockers, shelves, soap dispensers
- providing a low desk or work surface.

Mobility Accommodations

Crowded hallways and classrooms may pose problems for children who use wheelchairs. Those who walk may have trouble using stairs, walk more slowly than their peers, and be at risk of falls in crowded hallways or on slippery floors.

Some common mobility accommodations include the following:

- Allow the child to leave class several minutes early so he or she has extra time to get to the next class or the school bus.
- In multi-level school buildings, provide the child with elevator privileges.
- Allow the child to select a seat that is easy to get to, such as near the classroom door.
- Provide an extra set of books that the child can keep at home, so he or she does not have to carry heavy loads.

Fire Evacuation

It is vital that school employees develop a fire and emergency evacuation plan for a child with OI before an emergency occurs, and that they practice this plan during routine fire drills.

- Schools may want to assign a particular staff person to accompany a child with OI during a fire drill or emergency.
- If a child is in a wheel-chair and attending a multi-level school building, a specific plan for evacuation must be made.

Transportation

- Transportation accommodation may involve a wheelchair-accessible bus or van.
- Children who walk, with or without assistance, may ride a regular school bus, but may need someone to assist them on and off the bus.
- Because most school buses do not have seat belts, children with OI who can sit in a regular bus seat may be at increased risk during an accident or if the bus stops short.

- Many children are attracted to “the back of the bus,” which often provides a bouncier ride than the front of the bus. For a child with fragile bones, however, a bouncy bus ride may prove dangerous.

Physical Education

Physical education teachers should make every effort to involve the child in the same activities as everyone else, with appropriate modifications, rather than isolate the child. If you are not certain if a child with OI should participate in a specific activity, discuss it with the parent/guardian. Children with OI often have a good sense of what they can and cannot do safely.

- A child with OI may be restricted from playing contact sports as injuries are difficult to avoid in contact sports.
- Activities that jar or twist the spine, such as jumping and games like crack-the-whip, should be avoided.
- Wearing a helmet and knee/elbow pads for sports like bike riding and roller blading, is recommended.
- Good fitting shoes help to support the ankles, and prevent tripping/slipping.

Recess/Playground

There is special playground equipment for children with disabilities, including swing sets that accommodate wheelchairs and sand tables that are at the proper height for a child in a wheelchair. Children with OI may also be able to use traditional equipment, such as slides or jungle gyms, with or without adult assistance. Even when a child with OI is able to play without assistance, adults may need to remind all children that safe and considerate play is important for preventing injury. Carelessly thrown balls or rough play can put the child with OI at risk of a fracture.

Physical or Occupational Therapy

Some children with OI may need physical or occupational therapy to maximize their skills and independence. Some children with OI will need assistive equipment to help them succeed in school.

EMERGENCY RESPONSE PLAN (school-aged children)

Any of the following situations may indicate a fracture.

- Child complains of pain in a bone that gets worse with movement
- Swelling or bruising over a bone
- Child has deformed limb
- Child is not using the limb
- Child winces or looks like that may be uncomfortable during routine play or exercises

If any of the above situations occur:

1. Contact the child’s parent/guardian.
 2. If you are unable to contact the parent/guardian or alternate contact, call 911/EMS.
 3. Inform the paramedics that the child has OI.
- Do not move the affected area unless it is absolutely necessary to move the child out of harm’s way.
 - Listen to the child’s advice. He or she may instruct you not to move a fractured limb, or tell you how to gently place a pillow under the limb with minimal movement.
 - Make the child comfortable while waiting for a parent or other designated person to arrive.
 - If the child becomes chilled or nauseated, provide a blanket, a basin, or whatever else the child might need.
 - Do not provide food or drink; if the child needs surgery to set the fracture, this will interfere with safe administration of anesthesia.
 - Staff should only apply a splint if the parent has instructed them to do so.