

Adults and Fractures

Osteogenesis imperfecta (OI) is a genetic disorder characterized by bones that break easily, often from little or no trauma. OI is highly variable and symptoms range from mild to severe. In addition to fractures, people with OI also have muscle weakness, joint laxity, and skeletal deformities such as short stature, scoliosis and bowing of long bones. Hearing loss is common in adults, affecting about 50 percent.

OI During the Adult Years

The adult with OI needs to manage all of the same health issues as other adults, along with the musculoskeletal concerns associated with OI. Most adults experience a decrease in fracture rate after puberty as a result of hormonal and other metabolic changes affecting bone mass and structure. This can last into the 30's or 40's. Other medical problems, some of which are related to the basic collagen defect, may assume more importance as people age. These include tendon, muscle and joint problems as well as respiratory compromise.

OI – Osteoporosis Link

Almost all people with OI are “osteoporotic” because most people with OI do not develop normal bone mass at any age. Women and men with OI can experience additional bone loss, such as age-related bone loss, superimposed on a background of OI. Symptoms of additional bone loss, including osteoporosis may appear at a younger age than commonly seen in people who do not have OI. **This bone loss may lead to a return to the fracture cycles they experienced as children.**

Fracture Management for Adults

- The majority of fractures seen in people who have OI are nondisplaced and can be managed with immobilization.
- Displaced fractures can be managed with manipulation under general anesthetic followed by immobilization.
- Care must be taken when manipulating OI bone because of the risk of causing additional fractures.
- OI bone is fragile and can easily fracture proximal to a cast of “normal” weight. Fracture immobilization should be with the lightest materials possible.
- Adults may have intramedullary rods of different ages and types in different long bones. Their placement and condition should be evaluated if a fracture occurs in a rodded bone.
- Use of plates and screws to repair a fracture is rarely recommended for either children or adults who have OI.
 - Poor bone quality leads to screw and plate instability
 - Plate rigidity can cause bone loss underneath the plate and fractures above and/or below the plate.
 - Screw holes may add to bone fragility and predispose to new fracture.

Other Orthopedic Concerns

- Scoliosis – Curves may progress and need to be monitored. Surgery is sometimes necessary.
- Non-Union fractures – In general OI bone tends to heal at the same rate as seen in the general population. Non-unions and slow healing have been documented. Reports indicate that the incidence of non-unions in adults with OI is higher than that seen in other adults. Researchers are looking into the use of bone morphogenetic proteins (BMP) or “bone glue,” and adult stem cells as possible solutions.
- Rods – Sometimes, rods migrate and become extremely painful. These may require surgery to repair, replace or remove. Rods that have not moved and are not painful usually do not need to be removed or replaced.

- Bowing of long bones – The more bowed a bone becomes, the more it is vulnerable to slow healing or repeat fractures. This may sometimes lead to a recommendation for surgery (usually intramedullary rodding) even if a fracture itself is minimally displaced.

Fractures and Travel

Business and personal travel is often part of an adult's life. Adults who have OI and who experience a fracture away from home are advised to delay flying until the swelling has subsided. Some medical professionals suggest waiting a week or more depending on the severity of the fracture. Altitude causes swelling and aggravates inflammation. This additional swelling can make a wrap or cast dangerously tight and extremely painful. Painful muscle spasms can also occur. A person with a new fracture who absolutely must travel, can be made more comfortable by icing the limb prior to take off and throughout the flight.

Working with Your Orthopedist

- It is important to find a surgeon who is knowledgeable about OI and has experience doing procedures for OI patients.
- Adults, including young adults living away from home for extended periods (i.e. college students), must plan ahead for emergencies. Adults should meet with the orthopedist prior to having a fracture or other emergency.
 - This is an opportunity to discuss fracture history, presence of rods, anesthetic and bleeding problems, medications and their dose as related to body size, and one's general health.
 - Work out with the orthopedist clear procedures for getting x-rays, what to do when pain medication is needed, what to do on a weekend or holiday and how to reach the doctor outside of normal office hours.
- Many adults are comfortable learning how to manage new fractures by simple splinting. Then they can avoid spending time at the hospital emergency department and go directly to their orthopedist. Orthopedists who are knowledgeable about OI and their staff are usually willing to provide this type of training.
- Adults and their orthopedist should also plan ahead for the physical therapy that will be necessary for a complete recovery from a fracture.

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