

## Emergency Department Management of Patients with OI

Osteogenesis imperfecta (OI) is a genetic disorder of connective tissue. The hallmark of OI is bones that fracture easily with little or no apparent trauma. Blue sclera; short stature; hearing loss; skeletal deformity; brittle, discolored teeth; and respiratory problems may also occur. OI ranges widely in severity. A person with mild OI may have just a few fractures in his or her lifetime, exhibit few other features of OI, and have normal stature and mobility. Those with more severe OI may have dozens or hundreds of fractures, as well as short stature, skeletal deformities, and limited mobility. Because of frequent fractures, children with OI are sometimes incorrectly assumed to be victims of child abuse or non-accidental trauma.

**Emergency Room staff members are requested to contact the patient's PCP or a physician who is familiar with the osteogenesis imperfecta as soon as possible. Adult patients with OI are often an excellent source of information about safe handling and whether a bone is likely to be broken. Patients with OI come to the emergency room most often for one of 4 reasons:**

- Fractures
- Pulmonary Problem
- Cardiovascular Problem
- Accidental Trauma – Falls and car accidents may cause more extensive injury to the person with OI than the same amount of force would inflict on others. It may be necessary to look beyond the simple complaint.

**Anesthesia consultation should be considered before any procedure using conscious or complete sedation.**

**See the OIF fact sheet “Common Medical Tests and Procedures” for more detailed information on specific procedures. It is located on the OIF website [www.oif.org](http://www.oif.org) under the For Medical Professionals Tab.**

### General Guidelines for Emergency Treatment

- People with OI often have ongoing medical and surgical needs, and have substantial experience with fracture management and orthopedic procedures. Respect the opinions, wishes, advice, or instructions of adults with OI, parents of children with OI, and older children with OI.
- Because bones are brittle, the severity of a fracture is not always directly related to the level of trauma. There may be no external sign of injury. Fractures are also unpredictable; a person with OI might have a serious fall or accidents with no fractures but then go on to fracture during normal daily activities.
- Allow parents to stay with their child at all times. This not only comforts the child, but the parent may be able to help with transferring the child or ensuring that safety precautions are used to prevent additional injury.

### Safety Precautions

- Be gentle and cautious during transfers; avoid sudden pulling of limbs, neck, or spine. Never twist, bend, apply pressure to, or try to straighten limbs. Some limbs cannot be straightened because of deformity. Parents, family members and/or older patients can provide guidance here.
- People with OI often bruise easily; therefore, IVs and blood draws should be done by the most experienced person available.
- Guard against placing a tourniquet or inflating a blood pressure cuff too tightly, which can lead to bruising or fractures when OI is severe. Automatic blood pressure cuffs may put too much pressure on the arm bone; set the cuff at the lowest possible inflation, and if possible, establish a baseline by first taking manual blood pressure measurements.
- Ask neonatal or pediatric nursing staff to help with medical procedures involving children and adults with OI who are very small.
- Some people with OI have a slightly increased risk of high urine calcium levels. Some people with OI have hyperhydrosis and may need significant fluid replacement. Otherwise, metabolic and blood

chemistries should be unremarkable. **Treatment should be per usual, with doses of medicines titrated to body weight, not age, even for adults with OI.**

- Normal-sized medical equipment may not fit people with OI who are smaller than average. Pediatric-sized equipment may be appropriate for some adults with OI. However, head size is often within normal ranges for a person's age, even if his or her body is small. Equipment to go on the head or face (such as an oxygen mask) should usually be determined by age.
- Stretchers should be padded and not have holes that a small person could slip through.
- Use caution when tightening straps on a stretcher, applying splints, using restraints, etc., to avoid causing a fracture.
- Make sure that blankets and sheets are not too tight, and be careful when removing them to avoid getting fingers, toes, etc., caught in the folds, which could cause a fracture.
- Some people with OI develop hyperthermia under general anesthesia, and some are sensitive to inhaled anesthetics.
- Some people with OI have an allergy to latex.

### **Pain Management**

- Adequate pain relief is paramount, even prior to x-ray.
- If possible, minimize handling before pain relief is administered. Babies and small children may fall asleep when held, only to awaken screaming in pain when moved slightly. Do not hesitate to use splints and wraps to reduce motion of a painful limb and to minimize spasm.
- Consider NPO until the need for surgery or surgical application of a cast is assessed by an orthopedist.

### **X-Rays**

- The decision to obtain an x-ray should be made jointly with the family or older OI patient.
- Often, the x-ray is negative until two or more weeks after the injury. Regardless of x-ray findings, symptom control using a splint, braces, medication, or sling should be provided.
- Minimize handling but ensure proper views of affected bones (i.e., it is better to properly position once than to have to repeat x-rays). "Frog leg" views make it easy to see both tibias and femurs.
- Allow a parent or other family member to assist in transferring and positioning a child. If the patient is an adult, listen to his or her advice about transferring, and allow a friend or family member to assist if the patient approves.

### **Determining Whether a Fracture Has Occurred**

- People with OI frequently have microfractures that are not visible on x-ray immediately following the injury. Due to low bone density, decreased soft tissue reaction, and bone deformity in some people with OI, nondisplaced fractures may not be discernible on early x-rays. Follow-up x-rays one to two weeks later may reveal the fracture because a callus has formed.
- If a fracture is suspected due to pain, swelling, the patient's inability to use the limb (especially in a child), and the patient's or family's insistence that it really is broken, treat the limb as if it is broken until follow-up x-rays are taken. If it is fractured, the patient will be much more comfortable with the limb immobilized. If it is not fractured, a few extra days of immobilization should not cause permanent damage.
- Soft tissue injury around a fracture is less likely in a person with OI. The bone usually cracks before the ligaments and tendons tear.

### **Immobilization of Fractures**

- If an orthopedist is not immediately available, use appropriate splinting techniques to immobilize the affected limb until definitive orthopedic treatment. Bone deformities require added consideration when immobilizing a fracture. If the patient has an orthotic, it can often serve as an effective splint.
- Prolonged (more than a few hours) of skeletal traction is usually not effective and delays definitive surgery.
- Orthopedics should be called early in the process.
- If there is internal fixation (i.e., a rod), splinting or casting may not be necessary, but adequate pain relief is still important.
- Casting should be done by the most experienced person available.