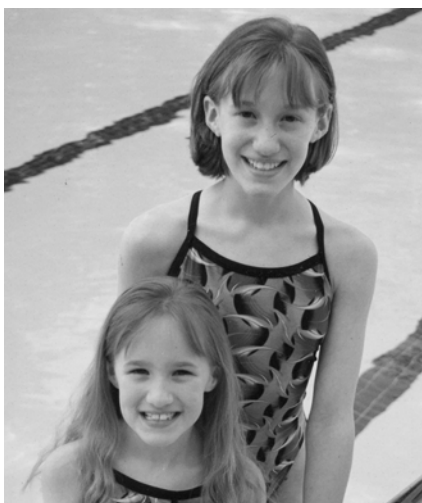


Therapeutic Strategies for Osteogenesis Imperfecta



**A Guide for Physical and
Occupational Therapists**

This publication is not copyrighted and is in the public domain. Formal permission to copy and distribute this material is not required.

For more copies, contact

National Institutes of Health
Osteoporosis and Related Bone Diseases ~ National Resource Center
2 AMS Circle
Bethesda, MD 20892-3676
Telephone: 1-800-624-BONE (toll free)
E-mail: NIAMSBONEINFO@mail.nih.gov

You also can find this publication on the
NIH Resource Center Web site at: www.niams.nih.gov/bone

Therapeutic Strategies for Osteogenesis Imperfecta

**A Guide for Physical
And Occupational Therapists**

Revised June 2005

*This publication is provided by the
National Institutes of Health
Osteoporosis and Related Bone Diseases ~ National Resource Center
in cooperation with the
Osteogenesis Imperfecta Foundation.*

The NIH Osteoporosis and Related Bone Diseases ~ National Resource Center is supported by the National Institute of Arthritis and Musculoskeletal and Skin Diseases with contributions from: National Institute on Aging, *Eunice Kennedy Shriver* National Institute of Child Health and Human Development, National Institute of Dental and Craniofacial Research, National Institute of Diabetes and Digestive and Kidney Diseases, NIH Office of Research on Women's Health, DHHS Office on Women's Health.

*The National Institutes of Health (NIH) is a component of the
U.S. Department of Health and Human Services (DHHS).*

Table of Contents

Introduction.....	1
What Causes OI?	1
The Role of Physical and Occupational Therapy in Managing OI.....	3
Safe Handling of Children and Adults with OI.....	4
Therapeutic Strategies for Osteogenesis Imperfecta.....	5
Adaptive Equipment and Aids to Independence	8
Problem Solving for Specific Self-Care Tasks	11
Resources — Publications.....	13
Resources — Suppliers of Specialized Equipment.....	14
Index.....	15

Introduction

Osteogenesis imperfecta (OI) is a genetic disorder characterized by bones that break easily, often from little or no apparent cause. OI is a highly variable disorder with signs and symptoms ranging from mild to severe. In addition to fractures, people with OI often have muscle weakness, joint laxity, and skeletal problems that can interfere with motor skills and daily function.

Physical and occupational therapists can help people with OI maximize strength and overcome functional limitations by teaching them and their families about protective handling to avoid injuries, protective positioning and movement to strengthen muscles and develop motor skills, and use of appropriate adaptive equipment. This publication provides an introduction to OI. It also presents common problems that those with OI face, and therapeutic strategies that have proven effective.

Parents of children with OI, adults with OI, and therapists can share the information in this publication as they work together to find solutions that help a particular child, teenager, or adult achieve and maintain motor and personal care skills. Chosen activities should address the individual's particular needs and interests.

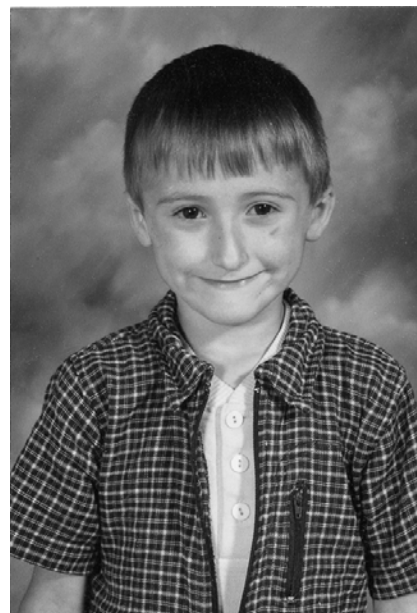
What Causes OI?

OI is caused by genetic defects in the structure or synthesis of type 1 collagen. Type 1 collagen is the major protein “building block” of bone. Individuals with Type I (mild) OI have half the normal amount of collagen, but it is all structurally normal. Individuals with Types II, III, and IV (severe and moderate) OI have structurally abnormal collagen. (The OI types are further explained below.) These defects in the collagen lead to weak bones that fracture easily.

Signs and symptoms of OI may include frequent fractures; muscle weakness; joint laxity; short stature; blue or gray sclera (whites of the eyes); thin, smooth skin; easy bruising; spinal curvature; bowing of long bones; and excessive sweating and heat intolerance. Some people with OI have a barrel-shaped rib cage and/or a triangular face. Some experience hearing loss, often beginning in early adulthood but in some cases beginning in childhood. Some also have a condition called dentinogenesis imperfecta, which affects the teeth.

A classification system of four OI types is commonly used to help define how severely a person with OI is affected. This system is referred to as the Sillence classification after the Australian doctor who developed it in the 1970s.

- People with **Type I OI**, the mildest and most common form, may have only a handful of fractures or as many as several dozen fractures in a lifetime. They may have few obvious signs of the disorder and may go undiagnosed until they begin to break bones, often as toddlers or even much later, in their teens or twenties. They usually have little or no bone malformation and are of average or near-average height for their family. Muscle weakness, joint laxity, and flat feet are common. Dislocations and sprains may occur as well as fractures. Life expectancy seems average.



Six-year-old boy with mild OI.

- **Type II OI** is the most severe form. Infants are usually quite small at birth and are born with multiple fractures, an unusually soft skull, and an unstable neck. Almost all infants with Type II OI die at or shortly after birth, often due to respiratory problems. In the newborn period, it can be difficult to distinguish between Type II and severe Type III OI. This means that some children diagnosed as Type II at birth may actually have Type III OI and have a longer life expectancy. Very rare exceptions of true Type II infants with longer survival have been reported.



Six-year-old boy with very severe OI.

- People with **Type III OI** are born with fractures, and x rays may reveal healed fractures that occurred before birth. Common signs and symptoms include short stature, progressive long bone malformation and spinal curvature, and a barrel-shaped rib cage. People with Type III OI may have anywhere from several dozen to several hundred fractures in a lifetime, and surgical correction of long bone bowing and scoliosis is common. Life expectancy varies; some people with Type III OI have severe, sometimes fatal, respiratory problems in infancy or childhood. Some children and adults with severe Type III OI may require supplemental oxygen. Some individuals succumb to respiratory problems in adulthood due to progressive rib cage and spine malformation. Others with Type III OI have a near-average life span.



The 21-month-old girl on the left in the hip spica cast has severe OI. The three-year old-girl on the right has moderate OI.

- **Type IV OI** is considered moderate. Type IV OI can be almost as mild as Type I or almost as severe as Type III. People with this type of OI may be somewhat shorter than others in their family, have frequent fractures that decrease after puberty (a common occurrence with all types of OI), and have mild to moderate bone malformation. Life expectancy appears to be average.

It is important to note that the features of OI (fracture frequency, muscle strength, bone and joint alignment) vary greatly from person to person, even among people with the same type of OI and even within the same family. Not all characteristics are evident in each case. In addition, many people with OI do not fit clearly into one of these four types. Types I to IV OI are determined based on clinical and radiographic data. Researchers are beginning to identify additional types of OI. Two new types of OI, Types V and VI, appear clinically similar to Type IV but have a distinct histology. When working with an individual who has OI, therefore, it is most important to focus on his or her particular abilities, strengths, and weaknesses rather than on his or her OI type.

OI treatment involves caring for fractures, maximizing independent mobility and function, and developing optimal bone mass and muscle strength. Surgery may be done to insert metal rods into the long bones to reduce malformations and control fractures. Medications developed for perimenopausal osteoporosis are

currently being tested in OI. They have been shown to increase vertebral bone density and decrease bone pain. Some studies have also reported a decreased fracture rate and increased mobility, but the medication's effects on ambulation and the long bones of the legs still are not clear. Use of wheelchairs, braces, walkers, and other mobility aids may be necessary for optimal independence and mobility.

Achieving and maintaining goals related to maximum function, independence, and self-care requires a team approach, with physical therapists, occupational therapists, orthopaedists, and orthotic specialists working together with the patient and his or her family.

The Role of Physical and Occupational Therapy in Managing OI

The long-term goal for people with OI is independence in all life functions (e.g., self-care, locomotion, recreation, social interaction, education, and work), with adaptive devices as needed or, in the case of very severely affected people, the ability to direct their own care.

For many years, the parents of children with OI, particularly in severe cases, were told to carry their children around on a pillow and not to expect them to achieve independent function. This led to their being treated as infants even as they grew into school age and beyond. It is now clear, however, that most people with OI can achieve some level of independent mobility and function with the help of physical and occupational therapy, appropriate exercise, surgical intervention, medications, adaptive equipment, and environmental adaptations at home, school, or the workplace.

Maximizing a person's strength and function not only improves his or her overall health and well-being. It also improves bone health, because mechanical stresses and muscle tension on bone help to increase bone density. Encouraging people with OI to adopt different body positions during the day will help strengthen different muscle groups and possibly prevent or minimize malformations, such as a flattened skull, a highly scooped lower back, or tight hip flexor muscles.

Ongoing or intermittent physical and occupational therapy is appropriate in a number of circumstances:

- **when a child with OI has delays or weakness in motor skills.** Because of fractures, muscle weakness, and joint laxity, many children with OI (even those who are mildly affected) experience delays in motor skill development, which then interfere with function. As soon as it is evident that an infant has muscle weakness or motor skill delays compared with unaffected same-age peers, therapy should begin. It should continue until a child reaches appropriate therapy goals. In some cases, an infant or young child may have delays. But after gaining sufficient strength, he or she may become able to sit, stand, and walk. After fractures or surgeries, these children may require additional intensive rehabilitation or physical therapy until they are able to regain the previous level of motor function. In other cases, certain motor skills may be unattainable due to weakness or skeletal malformations. For example, walking is not possible for some people with OI. However, the therapist can help the person maximize function by developing other skills and using adaptive equipment, energy conservation, and joint protection concepts.
- **when a person with OI is recovering from a fracture or surgery.** Because fractures and surgery are frequent for many people with OI, it is particularly important for them to regain as much function as possible during recovery, both to maximize independence and to maintain bone and muscle strength. After recovery, a person may need to relearn skills that he or she had previously mastered as well as regain strength in the affected limb(s). The therapist can help a person with OI develop alternate strategies and use appropriate equipment to make self-care skills easier. Therapists will need to teach the use of gravity-eliminated activity (e.g., aquatics) postoperatively and the principles of leverage (e.g., supporting the whole limb when giving resistance) that are safer for the individual with OI.

- **when a person with OI fears movement and trying new skills.** In some cases, the biggest obstacle to independent function is fear. A child who has had fractures may become fearful of moving or trying new things. Parents may respond fearfully to a child's attempts at independent movement, finding it difficult to allow the child to test his or her physical limits because of the pain and difficulty that a fracture will bring to the whole family. In some families, these fears can lead to the child's complete dependence on a parent for all aspects of daily function and self-care. This becomes increasingly problematic as the child enters school – and later, the adult world – unable to do most things for him- or herself and unable to rely on anyone other than the parent for help. While therapists must acknowledge these fears as understandable, they can also suggest ways that the child can practice new skills in a safe environment. This is usually successful if skills are broken down into small steps, allowing the child to succeed at something relatively easy, moving step by step through more difficult skills rather than failing repeatedly when expectations are too high. Protective equipment (e.g., clamshell splints in the pool, hinged circumferential forearm splints when doing some weightbearing), positioning, protected movement, and water therapy can also be helpful. Encouraging the child to direct some of his or her own care, transfers, and handling will also build the child's confidence.
- **when a person with OI needs to learn a new skill or a new way of performing a known skill.** Many key self-care skills, such as toileting, dressing, bathing, grooming, and preparing food, pose challenges to people with OI. Some may lack the strength to perform certain tasks or have trouble using standard household equipment because they are short-statured or use a wheelchair. Due to injury, aging, or progressive malformation, children and adults with OI often have to relearn how to do a task in an entirely different manner. Through a combination of strengthening activities, use of adaptive equipment, and creative problem-solving, many obstacles to independent self-care can be overcome. It is essential for therapists to listen to individuals with OI and their families. By respecting their problem-solving approaches, therapists may learn new ways to approach problems they may not have encountered before.

In working with individuals and families living with OI, therapists should keep two principles in mind:

- With the proper environment and equipment, most people with OI can function well in many or most areas of daily life.
- Individuals and families living with OI are truly the experts in how the disorder affects them. Listening to their concerns and ideas, building on their strengths and interests, and working with them as a team will help ensure success.

Safe Handling of Children and Adults with OI

There are some basic principles of safe handling that are important to follow anytime a therapist is working with someone who has OI. Fractures can occur simply because a part of the body was slightly twisted, pushed, pulled, or compressed. People with OI and their caregivers have extensive knowledge of what handling practices are safe for their individual cases. They should be encouraged to tell others in new situations that they are experienced in safe handling to prevent injury.

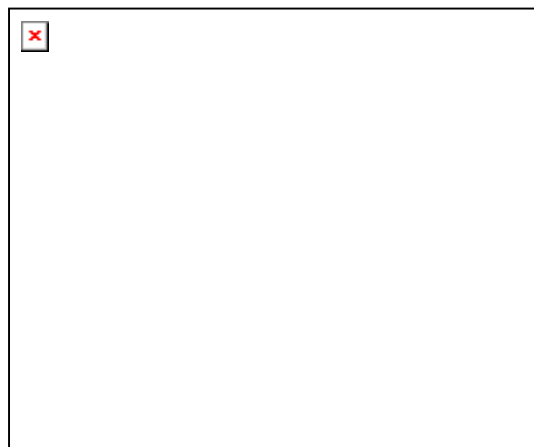
- Ask the parents or the individual with OI to show you the safe handling techniques they have developed. Before handling the person or moving a limb, state what you are going to do and how you are going to do it. If they ask you to stop, stop! For young children, using a floppy doll to demonstrate a motion and to problem-solve with parents will make new transitions easier. When the child hears, "It's your turn," and the parent helps the child with OI mirror the doll's movement, even a child as young as 10 months of age can understand and start the activity with less anxiety.



- It is often preferable to have the parent or caregiver do all handling of a young infant or child at the beginning of a therapy relationship, then gradually involve the therapist. As the child becomes secure and comfortable in the environment, he or she will gradually accept hands-on help from other individuals. The ideal is to structure the environment so it is easy for the child to perform the task without an adult’s hands-on help. This approach is safe, puts the child at minimal risk, and reinforces the child’s awareness of what he or she can do independently.
- It is critical to remember that people with OI do not have impaired coordination or sensation and do not require the complex, neurologically based interventions used for children who do. The main constraint to movement in OI is weakness. Once a child gets stronger, he or she typically figures out how to accomplish a task and often does not need the guidance of hands-on therapy to do it. This is especially true if the therapists and parents focus on constructing the environment to help the child achieve the task, rather than using external caregiver hands-on assistance.
- Never pull, push, or twist a limb. Avoid passive rotation of the arms, legs, head, or trunk.
- Lift an infant with OI with the widest base possible. Lift by placing one open hand under the buttocks and legs and the other under the shoulders, neck, and head. Do not lift the child from under the armpits, which puts pressure on fragile ribs and loose shoulders, and do not lift the buttocks by pulling on the ankles, especially during diapering.
- Be aware of where the person’s arms and legs are at all times to avoid awkward positions or getting a hand or foot caught in clothing or equipment.
- Provide adequate support when the child or adult is in a standing position so that the legs don’t “crumple” under him or her. Examples of support are straddle-riding toys, gait trainers with sling seats, and clamshell braces or splints on the legs.
- Avoid positions and motions of great leverage that stress bones, such as hip flexion (“jackknife” position with the person leaning far forward while sitting) that stresses the femur and diagonal trunk rotation that stresses the vertebrae. The “bridging” exercise (lifting the buttocks with knees flexed while lying on the back) should also be avoided because it stresses tibial bones in the same plane in which they tend to bow.

Therapeutic Strategies for Osteogenesis Imperfecta

- Before learning personal care skills, a child must first develop gross motor skills, such as reaching and sitting, which may be delayed or difficult for those with moderate to severe OI.
- Protective handling, protective positioning, and protected movement contribute to safe development of motor skills.
- Equipment, ranging from simple pillows to specialized wheelchairs, can help children and adults achieve motor and personal care goals even if they have weakness or are recovering from a fracture.
- Water therapy provides the opportunity for children with OI to develop skills in a gravity-free environment before trying them on land and for adults with OI to relearn or maintain motor skills.



A ten-year-old with very severe OI practices supported sitting with his therapist while he draws on a magnetic board.

- Patience and task analysis are both necessary to develop a successful therapy program. Therapy may progress more slowly for individuals with OI than for other therapy patients. Developmental concepts and specific skills need to be analyzed closely so that many small improvements can lead to achieving a particular therapy goal.

Protective Positioning: A key method for helping a person with OI maximize strength and function is to encourage him or her to adopt various positions throughout the day or, in the case of an infant or young child, to encourage parents and other caregivers to place the child in different positions. Position changes not only strengthen different muscle groups but also help prevent contractures and malformations that can limit mobility and increase pain. It is important to keep the hips and spine as straight as possible, prevent flattening of the back of the head from lying supine, and promote head-turning in both directions.

In many cases, everyday objects can be used to make different positions easier and safer. For example, towel rolls and padding can be used to encourage upright posture and avoid “frog-leg” positioning in a wheelchair, car seat, or stroller. An infant or child can be encouraged to try prone positioning by lying on the parent’s chest, a partially inflated beach ball, or a foam wedge.

Protected Movement: New positions and skills should be introduced gradually to allow the person to feel safe and promote gradual strengthening of muscles and bones. Providing adequate support is important to overcome weakness and prevent injury. According to Marnie King, an occupational therapist with extensive experience with children who have OI,

“Protected movement... is a hard concept because caregivers tend to protect the child from fractures by limiting potentially harmful movement experiences. But not moving creates the problems of decreased strength, bowing from constant sitting, fear of movement and being moved, and dependence in all activity... [T]he child with OI will require slow, graded introduction to being moved in progressively less supported positions.” (See Resources — Publications: King 2001, pp. 96-100.)

Therapists can begin by assessing the person’s current functional abilities. Is he or she stuck at a particular level of progression? The goal in therapy will be to gain the next level or improve within the lying, sitting, and walking levels. Thus, the goal for a very severely affected person might be to lie in a supported, inclined position. For a severely affected person, learning to sit-scoot might enhance his or her self-care skills. More mildly affected people may gain walking skills with or without braces or other aids. Very mildly affected people may function at the same level as their unaffected peers with occasional modifications or limitations, such as no high-impact activities. Activity analysis will need to be done in very small increments to assess progress. Parents of children with OI need to know that many developmental milestones might not be met but will be compensated for by building skills around them. For example, a particular child might not be able to crawl or independently get into a sitting position, but he or she can improve sitting skills and may use a power reclining wheelchair later to compensate.

King suggests a specific developmental progression for children with OI, as follows:

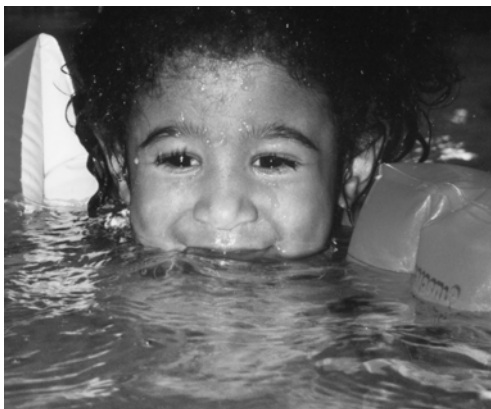
Progression	Possible Modifications Needed for People with OI
Supine positioning	Use a concave skull pad or gel pad or provide sufficient position changes to prevent flat skull. If child cannot get hands to midline, use trough-shaped foam bed pad to guide shoulders forward. Provide support or splint forearms to prevent humerus and forearm from bowing toward body.
Prone positioning	Use a chest wedge, practice on steep incline first (e.g., parent’s chest). However, a chest wedge may not work for some children; they become locked in position and can’t move.

Progression

Possible Modifications Needed for People with OI

Inclined sitting	Use a concave skull pad or gel pad. Blanket rolls along torso for support can also serve as armrests. Position to decrease wide hip abduction. Provide place for feet to rest flat. Use very wide straps or vest for trunk support.
Sidelying	Support under head and below axilla. May need support under upper arm and leg if position is used for sleeping.
Rolling	May not be comfortable for child. Start by using a blanket like a hammock and slowly tilt child or position child to reach for a ball or object. Then progress to partial rolling in a blanket on a firmer surface. Once child is able to tolerate side motion, use slight wedge to roll downhill.
Supported sitting	May be done in positioning chair as with inclined sitting (above). Provide head and neck support at first. Slowly decrease support as head turning gets better.
Unsupported sitting	Work toward positioning on chair or bench with close spotting.
Getting to a sit	Start in pool, child sitting beside parent, side-leaning on parent's thigh and trying to get to a sit. On land, side-lie on a wedge or parent/therapist thigh and forearm (not extended wrist). For adults, work on abdomen strength to use a sit-up method.
Sit-pivot, sit-scoot	Start sitting on bench in pool/tub with water to chest height and shift side-to-side to get floating toy. Lower height of water to child's hip height. Then try on land on slippery bench with close spotting. This position will be a transfer method from chair to bed to toilet during fractures and if legs are not strong enough to stand.
Crawling	Start in kneeling position with chest supported by partially inflated beach ball and aim uphill on wedge. Start static reaching for toy above child. Progress to less chest/abdomen support. If legs abduct, use "mermaid suit" of stretchy 6- to 8-inch wide tubigrip, panty hose top, or wide stockinette from child's waist to ankles.
Kneeling/pulling to a stand	"Mermaid suit" (see above) and high kneel for trunk development. In the pool, progress from high kneel to half kneel. Also in pool (water depth to waist when standing), lower to sitting and stand up again. Then try crawling in water the height of child's knees.

Some people with OI will achieve all of these skills, although interruptions in progress and reverting to previous skills are common because of fractures and surgery. Others will achieve only some of these skills. However, supplemented by equipment and environmental adaptations, any level of proficiency with these skills will increase potential for independent function and self-care.

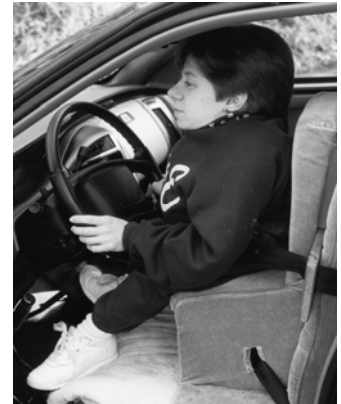


Water Therapy. Water provides an ideal environment for people with OI to practice protected movement and learn new skills. Water not only cushions bones and joints and protects the person from falls, but it also provides gentle resistance along the entire length of bones. This resistance helps to strengthen bones and muscles, and prevent fractures that are caused when too much pressure is applied to an isolated area. Swimming and other water exercise often become favorite fitness activities for older children and adults with OI.

Practicing the above-listed developmental progressions in the water can reduce fears and help make transitions from one position to another easier. Once the skills are mastered in the water, they can be tried on land. (See Resources — Publications: King 2001, p. 109 for a diagram of how progressions can be practiced in the water.)

Examples of water therapy that can promote new skills are:

- “Shimmy-sitting” and scooting by sitting on steps in the water and scooting from side to side or up and down the steps.
- Standing and walking, starting with water up to the chest. Provide support, such as lightweight splints on the legs, a foam “noodle” or kickboard for the person to hold onto, and/or a flotation vest to promote upright posture. Move into more shallow water (less buoyancy) as confidence and strength increase. Use a shoe lift during this activity if the person has a leg length discrepancy.



Custom-designed car seat cushions and pedals for a short-statured adult with OI.

Adaptive Equipment and Aids to Independence

The equipment available to help a person with OI function independently is practically unlimited if one considers both traditional adaptive equipment as well as “homemade” solutions to everyday challenges. Important concepts to consider when choosing equipment are energy conservation, joint protection, mobility, and accessibility. In considering these concepts, the goal is for the person with OI to be as independent as possible in his or her daily life.

Energy conservation: To help a person function most efficiently, evaluate what tools and environmental adaptations might be needed so that he or she can accomplish common tasks without excessive strain or fatigue. Establishing workstations, such as a homework station, toothbrush station, or hair-drying station, with all needed materials in one place and within reach, will prevent unnecessary reaching or traveling around the room searching for things. Baskets or bags attached to a wheelchair, walker, or crutches allow the person to carry things from room to room. Clothing that is easy to put on, such as pullovers and pants with elastic waists, will minimize the effort needed to dress and undress.

Joint protection: To help avoid overstretching and injury, teach people with OI to use their strongest muscle groups (usually the largest) to accomplish tasks. Suggest tools that will minimize strain during day-to-day tasks, such as jar openers and electric can openers, and low shallow shelves to help access items. Reachers must be used carefully because the added length increases the weight of the object.

Mobility: Many people with OI use a mobility aid at some point in their lives. Some may only need assistance when they are learning a new skill or recovering from a fracture or surgery, while others will use a walker, crutches, wheelchair, or other aid most of the time.

Accessibility: Physical environments at home, school, or work can be modified to allow maximum independence. While extensive structural changes – such as ramps or lowered kitchen and bathroom surfaces – are sometimes called for, some accessibility problems can be addressed with creative use of assistive devices, rearrangement of furniture and other equipment, and thoughtful consideration of how the person with OI can best use his or her home, classroom, or office. Because the world is not modified for short-statured people, using a wheelchair with a seat elevator should be considered when needed. (For more information, See Resources — Suppliers of Specialized Equipment: p. 14.)



An important byproduct of making homes as accessible as possible for children with OI is that the children can better participate not only in their own care but also in household responsibilities. It is vital, both for the child's well-being and the family's healthy functioning, that children with OI take responsibility for appropriate household tasks. To help children do their jobs safely, families may need to modify room arrangements and storage of household items.

The following table lists many commonly used types of equipment that help to maximize function for people with OI and some factors to be considered when choosing appropriate equipment. Note that this list is only a starting point. More specific ideas and recommendations can be found in the section on self-care tasks that follows the table as well as in the other publications listed at the end of this publication. It is vital to choose equipment that matches a particular individual's strengths, weaknesses, and interests as well as those of his or her family.

Type of Equipment	Common Considerations for People with OI
Walkers	Supported walking allows weight-bearing in legs, which increases strength and bone density. The person needs sufficient upper-body strength to grasp/move walker. Posterior walkers may be useful for encouraging upright posture. Some people with OI report feeling more secure/steady with an anterior walker. Baskets attached to walker can help with independence.
Crutches and canes	Supported walking allows weight-bearing in legs, which increases strength and bone density. The person needs sufficient upper-body strength to grasp/move crutches. Crutch/cane tips may need to be specially ordered if the standard tips are not sufficiently slip-resistant. They should be replaced often to maintain maximum slip-resistance.
Wheelchairs	Manual chairs can enhance upper-body strength in people whose arms are long and strong enough to push the wheels without pain or risk of fracture. Armrests should support the whole arm and flip up for pushing to avoid forearm bowing. Manual chairs are lighter and easier to transport than power chairs. Power chairs allow people with arm fractures, short arms, and/or arm deformities to move independently. Features such as a power reclining back and a power seat elevator are helpful for some people with OI.



An adult with OI using a power wheelchair with a seat elevator, which allows her to reach things on the floor or on high surfaces.

Type of Equipment

Common Considerations for People with OI

Other mobility aids

These include scooter boards, riding toys, tricycles, etc. Four wheels are best to prevent tipping/falls. Seat belts/safety harnesses are necessary. Seats with backrests provide more support and promote good posture. These aids are particularly helpful for young children who are not candidates for walkers or wheelchairs but will benefit from independent mobility.



This child is sitting in a homemade cart that she can use even when in a cast.

Braces/splints

These should be circumferential, lightweight, and perforated to avoid excessive sweating. Leg braces can help with alignment and promote standing and walking. Their role in preventing fractures is limited. Lightweight forearm splints (bivalve or hinged for support to the entire surface – 1/16" perforated Aquaplast* is perfect for children) can provide stability to people with forearm bowing, weakness, or pain, and help them with ADLs, weight-bearing, reaching, and lifting. After a short period of casting, fractured limbs are often immobilized in a lightweight splint or brace that can be removed for bathing and other activity. They may permit greater activity when worn during water therapy while a fracture is healing.

Positioning aids

These include pillows, bolsters, towel rolls, gel pads, etc. Use them to promote 90/90/90 position in car seat, wheelchair, stroller, etc., rather than "frog leg" position and to encourage sidelying and prone positions for infants and young children with OI, who often spend a lot of time on their backs.

Standers

Use these to promote vertical weight-bearing posture, which benefits bone growth and density. Supine standers are preferred to prone standers because standing can be introduced and increased gradually. Tray attachments can allow a child to use stander while coloring, doing homework and games, working on computer, etc.

Infant/child car seats

Use towel rolls, stuffed animals, or bolsters to promote good sitting posture and keep head in midline. Extra-wide padding on straps can help prevent injury in a sudden stop or traffic accident. Car bed-style safety seats may be useful for very small infants and infants or children who are unable to sit up. Look for breathable fabrics for padding and seat covers, as children with OI tend to overheat easily. Many families affix a noticeable tag or sticker to the seat indicating the OI diagnosis in case of a traffic accident.

Self-care aids

These include transfer benches, bath chairs, grab bars, reachers, etc. The goal for all people with OI is to be as independent and as safe as possible in self-care tasks. Self-care aids can help them overcome limitations due to weakness, short stature, use of a wheelchair, etc.

* Brand names included in this publication are provided as examples only, and their inclusion does not mean that these products are endorsed by the National Institutes of Health or any other Government agency. Also, if a particular brand name is not mentioned, this does not mean or imply that the product is unsatisfactory.

Problem Solving for Specific Self-Care Tasks

Toileting, bathing/grooming, dressing, and food preparation are four key self-care tasks. These tasks are often challenging for people with OI, particularly if they are short-statured, use a wheelchair, and/or are recovering from a fracture or surgery. Helping people with OI become independent or semi-independent in these key tasks will do a great deal for their overall well-being and sense of self-esteem. Therapy goals should be to assist the individual to the next higher level of independence for the greatest freedom. Levels of independence are:

1. **Independent** – able to do all aspects of self care with no modifications.
2. **Independent with modifications** – e.g., in a one-level, wheelchair-accessible home and workplace.
3. **Independent with intermittent help** – requires assistance with shopping, carrying in/out, and cleaning but can do personal care, toileting, light cooking without help.
4. **Independent with daily help** – requires assistance, as above, and also for bathing and meal setup.
5. **Dependent in most activities** – needs help for toileting, dressing, is able to do light tabletop activity, is safe alone for 2 to 3 hours.
6. **Dependent in all activities.**

Toileting: Toilet teaching a toddler often involves a “one step forward, two steps back” pattern. With children who have OI, that pattern may be even more pronounced, as a child who is learning to use the toilet may go back to diapers when recovering from a fracture or surgery. But while toilet teaching may take longer than average in some cases, people with OI can achieve independent or semi-independent toileting with the help of modified or specialized equipment.

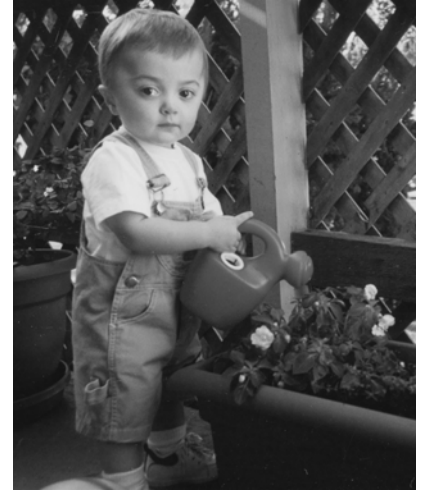
- If a standard potty chair or toilet poses problems for a child with OI (e.g., not enough sitting support or too high off the ground), a specialized potty chair can be made from a child-sized resin chair. Cut the legs so the chair sits low to the ground, cut a hole in the seat, place a bucket underneath, and add a safety harness and cushions as needed.
- Some toilet manufacturers offer models that are lower to the ground than average. Families may wish to consider installing one of these models for a short-statured person.
- A toilet-paper reacher can extend the reach of the hand if a person’s arms are particularly short. Reachers can be purchased. They also can be made using plastic-covered solid core copper wire twisted around a broom handle and finished with electrician’s tape or liquid plastic so that the reacher is washable.
- A sliding transfer bench and/or grab bars, located near the toilet, will allow for easier transferring from wheelchair to toilet. The “shimmy-sit” transfer will be used.



Bathing/Grooming: Bath time often gives a severely affected infant some of his or her first experiences of independent movement, and does so with less chance of fracture. Placing a folded towel, gel pad, or foam pad on the bottom of the tub provides a comfortable, slip-resistant surface for the infant to be bathed. Older children and adults may benefit from such adaptations as a sliding transfer bench, a shower seat, grab bars, and a hand-held shower head. Long-handled scrub brushes or sponges, washcloths, and “soap on a rope” can be hung on easy-to-reach hooks in the shower or tub enclosure.

For grooming tasks, such as brushing teeth or styling hair, it is helpful to use the “work station” concept discussed previously. For example, a hair dryer can be plugged in and mounted to the wall next to a low mirror, with combs, brushes, and styling aids in a drawer nearby.

Dressing: Infants with OI should be dressed in clothing that minimizes stretching, pushing, and pulling of limbs, such as t-shirts that snap open up the front and play suits with snaps along both legs and the torso. For older children and adults with OI, simple, easy-on clothes, such as pants with elastic waists, are useful. They may find it easiest to dress while sitting on a bed or bench. Clothes often need to be modified for people who are short-statured or are wearing a cast. For example, a seam can be cut and hook-and-loop material sewn onto the seam so it can be opened and closed around a cast. Dressing tools, such as reachers and sock donners, may be useful for some people. Closet rods can be lowered by using ropes or chains to hang a broom handle horizontally from the existing closet rod. Other clothing should be stored on shelves or in drawers that the person can easily reach.



Food Preparation: From a child who wants to fix an after-school snack to an adult living in his or her own home, people with OI benefit from having an accessible kitchen where they can prepare food. While a custom-designed kitchen with lowered counter tops and appliances, long-handled faucets, and adjustable shelving is ideal, any kitchen can be made more accessible to a person with OI. Commonly used items should be stored in low drawers or cabinets near where they will be used (e.g., cups stored near the refrigerator, pots stored near the stove, etc.). Lazy Susan turntables and pull-out shelving make items easier to reach. A loop of rope or fabric can be attached to the refrigerator door to allow a person to hook the loop onto his or her wheelchair and pull the door open. Beverage containers can be stored on the lowest door shelves for easy access, and the contents of large beverage containers can be divided into smaller containers so they are not so heavy. A miniature “ramp” can be placed in front of the microwave, so the person can slide a plate or bowl out of the microwave onto the counter without having to lift a hot, heavy item. “Heat and eat” convenience foods can be useful for people with limited mobility and strength, as they can be prepared in the microwave in lightweight plastic containers.

Resources — Publications

The NIH Resource Center provides fact sheets and other resources free of charge to families living with osteogenesis imperfecta and to professionals. (See back cover for contact information.)

For more information, the Center refers people to the Osteogenesis Imperfecta (OI) Foundation. In addition to the books listed below, which are available for purchase, the OI Foundation publishes numerous brochures and fact sheets on OI issues that are free of charge. The foundation can also connect physical and occupational therapists with other professionals who have extensive experience with OI.

Osteogenesis Imperfecta Foundation
804 West Diamond Avenue, Suite 210
Gaithersburg, MD 20878

Phone: (800) 981-2663 or (301) 947-0083
Fax: (301) 947-0456
Internet: www.oif.org E-mail: bonelink@oif.org

Other resources of interest include:

Bleakney DA, Donohoe M. Osteogenesis Imperfecta. In Campbell SK, Vander Linden DW, Palisano RJ (eds): *Physical Therapy for Children*. 2nd edition. Philadelphia: W.B. Saunders; 2000: 320-338.

Cintas H, Gerber L. Motor Performance: Succeeding Despite Brittle Bones. In Wacaster, P (ed): *Managing Osteogenesis Imperfecta: A Medical Manual*. Gaithersburg, Md.: Osteogenesis Imperfecta Foundation; 1996: 101-108.

Dollar EP (ed). *Growing Up with OI: A Guide for Families and Caregivers*. Gaithersburg, Md.: Osteogenesis Imperfecta Foundation; 2001.

This 15-chapter volume is the OI Foundation's most comprehensive resource to date on the medical, social, family, and emotional issues faced by families living with OI. It is an excellent resource for any professional seeking more information on OI. Chapters of particular interest to physical and occupational therapists are individually cited below.

Gerber L, Cintas H. Exercise and Activity: A Balance Between Work and Play. In Dollar EP (ed): *Growing Up with OI: A Guide for Families and Caregivers*. Gaithersburg, Md.: Osteogenesis Imperfecta Foundation; 2001: 131-145.

King MM. Personal Care for Lifelong Independence. In Dollar EP (ed): *Growing Up with OI: A Guide for Families and Caregivers*. Gaithersburg, Md.: Osteogenesis Imperfecta Foundation; 2001: 87-129.

McCabe M, Rosalie N. Educating the Child with Osteogenesis Imperfecta. In Dollar EP (ed): *Growing Up with OI: A Guide for Families and Caregivers*. Gaithersburg, Md.: Osteogenesis Imperfecta Foundation; 2001: 161-208.

Osteogenesis Imperfecta Foundation. *Plan for Success*. Gaithersburg, Md.: Osteogenesis Imperfecta Foundation; 1998.

Plan for Success is a video, with a companion brochure, covering issues related to educating children with OI in public schools, including mobility and accessibility concerns.

Wacaster P (ed). *Managing Osteogenesis Imperfecta: A Medical Manual*. Gaithersburg, Md.: Osteogenesis Imperfecta Foundation; 1996.

Resources — Suppliers of Specialized Equipment*

ActiveAid

Customized bathroom equipment
101 Activeaid Rd.
P.O. Box 359
Redwood Falls, MN 56283-0359
Phone: (800) 533-5330
Internet: www.activeaid.com

Hydro-Fit Aquatic Fitness Gear

Flotation vests and foam accessories for water therapy
160 Madison St.
Eugene, OR 97402
Phone: (800) 346-7295 or (541) 484-4361
Internet: www.wetvest.com

Children's Medical Ventures

Positioning aids for infants and small children (gel pads, car seat inserts)
275 Longwater Dr.
Norwell, MA 02061
Phone: (888) 766-8443
Internet: www.childmed.com

Little People of America

Information on products to aid people with short stature
5289 NE Elan Young Pkwy., Ste. F-700
Hillsboro, OR 97124
Phone: (888) LPA-2001 or (503) 846-1562
Internet: www.lpaonline.org

Maddak Inc.

ADL aids
661 Rte. 23S
Wayne, NJ 07440
Phone: (800) 443-4926 or (973) 628-7600
Internet: www.maddak.com

North Coast Medical

ADL aids, splinting products
18305 Sutter Blvd.
Morgan Hill, CA 95037-2845
Phone: (800) 821-9319 or (408) 776-5000
Internet: www.ncmedical.com

One Step Ahead

Child safety equipment, specialized clothing, toys, etc.
P.O. Box 517
Lake Bluff, IL 60044
Phone: (800) 274-8440
Internet: www.onestepahead.com

Permobil

Power wheelchairs with seat elevator and tilt
6961 Eastgate Blvd.
Lebanon, TN 37090
Phone: (800) 736-0925
Internet: www.permobilusa.com

Rifton Equipment

Mobility aids, bathroom equipment
Community Products, LLC
359 Gibson Hill Rd.
Chester, NY 10918-2321
Phone: (800) 777-4244
Internet: www.rifton.com

The Right Start

Child safety equipment, specialized clothing, toys, etc.
Phone: (888) Little-1 (548-8531)
Internet: www.rightstart.com

Sammons-Preston Rolyan

ADL aids, splinting products
4 Sammons Court
Bolingbrook, IL 60440-4995
Phone: (800) 323-5547
Internet: www.sammonspreston.com

* Organizations included in this publication are provided as examples only, and their inclusion does not mean that their products are endorsed by the National Institutes of Health or any other Government agency. Also, if a particular organization is not mentioned, this does not mean or imply that the product is unsatisfactory.

Index

A

- accessibility, 8
 - altering homes to make more accessible, 8
 - bathing and grooming, 11-12
 - dressings, 12
 - food preparation, 12
- adaptive equipment/aids, 8-10
 - braces, 10
 - canes, 9
 - car seats, 10
 - crutches, 9
 - four-wheel mobility aids, 10
 - positioning aids, 10
 - self-care aids, 10
 - splints, 10
 - standers, 10
 - walkers, 9
 - wheelchairs, 9

B

- bathing and grooming, 11-12
- braces, 10

C

- canes, 9
- car seats, 10
- crawling (part of developmental progression of children with OI), 7
- crutches, 9

D

- developmental progression for children with OI, 6-8
- dressings, 12

E

- energy conservation, 8
- equipment, 8-10

F

- fear of movement, 4
- fear of trying new skills, 4
- food preparation, 12
- four-wheel mobility aids, 10

G

- getting to a sit (part of developmental progression of children with OI), 7

I

- inclined sitting (part of developmental progression of children with OI), 7
- independence level, 11-12

J

- joint protection, 8

K

- kneeling/pulling to a stand (part of developmental progression of children with OI), 7

L

- learning new skills, 4
- learning new ways of performing known skills, 4

M

mobility, 8
modifications, physical (*See* accessibility)
motor skill delays, 3
motor skill weakness, 3
movement (*See* protective movement)

P

parents
 helping children with OI become independent, 3-4
 showing safe handling techniques to therapists, 4-5
positioning (*See* protective positioning)
positioning aids, 10
potty chairs, 11
problem solving for self-care tasks, 11-12
prone positioning (part of developmental progression of children with OI), 6
protected movement, 6
protective positioning, 6
publications, 13

R

recovery from fracture or surgery, 3
role of occupational therapy in managing OI, 3-4
role of physical therapy in managing OI, 3-4
rolling (part of developmental progression of children with OI), 7

S

safe handling, 4-5
self-care aids, 10
self-care tasks, 11-12
sidelying (part of developmental progression of children with OI), 7
Sillence classification, 1-3
sit-pivot, sit-scoot (part of developmental progression of children with OI), 7
splints, 10
standers, 10
supine positioning (part of developmental progression of children with OI), 6
suppliers of specialized equipment, 14
supported sitting (part of developmental progression of children with OI), 7

T

therapeutic strategies, 5-8
 protected movement, 6
 protective positioning, 6
 strategies for maximizing strength and function, 3-4

toileting, 11
Type I OI, 1
Type II OI, 1-2
Type III OI, 1-2
Type IV OI, 1-2
Type V OI, 2
Type VI OI, 2

U

unsupported sitting (part of developmental progression of children with OI), 7

W

walkers, 9
water therapy, 5, 7, 8
wheelchairs, 9

In Appreciation

This publication was compiled from materials previously published by the Osteogenesis Imperfecta Foundation, including several book chapters and articles by Lynn Gerber, M.D., and Holly Cintas, P.T., Ph.D., and the chapter in *Growing Up with OI: A Guide for Families and Caregivers* by Marilyn Marnie King, OTR/L. (See Resources — Publications: p. 13 for more information about these materials.)

The NIH Resource Center is grateful to the occupational and physical therapists who shared their expertise, including Tim Caruso, OT; Jane Olson, OT; Victoria Wittenberg, PT; to Ellen Painter Dollar who edited the original publication, and to the Osteogenesis Imperfecta Foundation for help in preparing and reviewing this publication.

This publication is provided by the
National Institutes of Health
Osteoporosis and Related Bone Diseases ~ National Resource Center
in cooperation with the Osteogenesis Imperfecta Foundation.

National Institutes of Health
Osteoporosis and Related Bone Diseases ~ National Resource Center
2 AMS Circle
Bethesda, MD 20892-3676
Phone: (800) 624-BONE or (202) 223-0344
Fax: (202) 293-2356, TTY: (202) 466-4315
E-mail: NIAMSBONEINFO@mail.nih.gov
Internet: www.niams.nih.gov/bone