

Caring for Children with Osteogenesis Imperfecta in a Community Program

Unified Referral and Intake System (URIS)

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Osteogenesis Imperfecta

This document was developed in consultation with health care professionals in the areas of endocrinology and community health. The Unified Referral and Intake System (URIS) wishes to acknowledge the contribution of the following individuals.

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INTRODUCTION

The Unified Referral and Intake System (URIS) is a joint initiative of the provincial Departments of Health, Family Services, and Education and Early Childhood Learning. URIS supports community programs in the care of children with specific health care needs. Community programs that are eligible for URIS support include schools, licensed child care facilities, and agencies providing respite service.

URIS provides a standard means of classifying the complexity of health care needs and establishes the level of qualification required by personnel to support children with these health care needs. Health care needs that are classified as 'Group B' can be delegated to non-health care personnel who receive training and monitoring by a registered nurse.

For children with 'Group B' health needs (e.g. osteogenesis imperfecta), the nurse provides the following support:

- develops and maintains a written health care plan;
- provides training to community program personnel that are responsible for the child; and
- monitors community program personnel that receive training.

This document provides standard clinical information that is relevant to the care of children with osteogenesis imperfecta during attendance in a community program setting.

CLINICAL INFORMATION

The following information is considered best practice in community program settings and is the basis for all osteogenesis imperfecta information contained in this document.

Osteogenesis imperfecta

Osteogenesis imperfecta (OI), also known as brittle bone disease, is a genetic disorder characterized by fragile bones that break easily. Its major feature is a fragile skeleton but many other body systems are also affected. OI is caused by a change in genes that are important for collagen and its strength. 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. Most often this is due to a new mutation to a gene and not by anything the parents did before or during pregnancy. There is no cure for OI.

In addition to fractures, the following health issues are frequently seen in children who have OI because of poor collagen quality:

- Short stature
- Weak tissues, fragile skin, muscle weakness, and loose joints
- Bleeding, easy bruising, frequent nosebleeds
- Brittle teeth (dentinogenesis imperfecta or DI)
- Hearing loss
- Vision problems including myopia and risk for retinal detachment
- Breathing problems
- Curvature of the spine

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.

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 fatal 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.

OI is classified by type and a description of the more common OI types is included in this section. More OI types continue to be discovered. OI type descriptions can provide information about the person's prognosis but does not predict functional outcome. Many people with OI do not fit clearly into one of the OI types. When providing care for a child with OI, it is important to focus on their specific abilities, strengths and weaknesses, rather than on their OI type.

Type I

- OI Type I is the mildest and most common form of the disorder. It accounts for 50% of the total OI population.
- Type I is characterized with mild bone fragility, relatively few fractures, and minimal limb deformities. The child might not experience fractures until he or she is learning to walk.
- Shoulder and elbow dislocations may occur more frequently than in healthy children.
- Some children have few obvious signs of OI or fractures. Others experience multiple fractures of the long bones, compression fractures of the vertebrae, and chronic pain.
- The intervals between fractures may vary considerably.
- After growth is completed, the incidence of fractures decreases considerably.
- The sclerae are often blue.
- Typically, a child's stature may be average or slightly shorter-than-average, but is still within the normal range for the age.
- There is a high incidence of hearing loss. Onset of hearing loss occurs primarily in young adulthood, but it may occur in early childhood.
- People with OI Type I experience the psychological burden of appearing normal and healthy despite needing to accommodate their bone fragility. The absence of obvious symptoms in some children may contribute to problems at school or with peers.
- Significant care issues that arise with OI Type I include gross motor developmental delays, joint and ligament weakness and instability, muscle weakness, the need to prevent fracture cycles, and the necessity of spine protection.

Type II

- OI Type II is the most severe form.
- At birth, infants with OI Type II have very short limbs, small chests, and soft skulls. Their legs are often in a frog-leg position.
- The sclerae are usually very dark blue or grey.
- The lungs are underdeveloped.
- Infants with OI Type II have low birth weights.
- Respiratory and swallowing problems are common.
- Infants with OI Type II usually die within weeks of delivery. A few may survive longer. Cause of death is usually respiratory and cardiac complications.
- Significant care issues that arise with OI Type II include obtaining an accurate diagnosis, getting genetic counselling, the family's need for emotional support, and management of respiratory and cardiac impairments.

Type III

- OI Type III is the most severe type among children who survive the neonatal period. The degree of bone fragility and the fracture rate vary widely.
- At birth, infants generally have mildly shortened and bowed limbs, small chests and a soft calvarium. The back of the skull can become flattened due to bone fragility. Lack of head control may also occur.
- Respiratory and swallowing problems are common in newborns.
- There may be multiple long-bone fractures at birth, including many rib fractures.
- Frequent fractures of the long bones, the tension of muscle on soft bone, and the disruption of the growth plates lead to bowing and progressive malformation. Children have a markedly short stature, and adults are usually shorter than 3 feet, 6 inches, or 102 centimetres.
- Spine curvatures, compression fractures of the vertebrae, scoliosis, and chest deformities occur frequently.
- The head is often large relative to body size.
- A triangular facial shape, due to overdevelopment of the head and underdevelopment of the face bones, is characteristic.
- The sclerae may be white or tinted blue, purple, or grey.
- Brittle teeth are common but not universal.
- Significant care issues that arise with OI Type III include the need to prevent fracture cycles, the need to develop strategies to cope with short stature and fatigue, and the family's need for emotional support, especially during infancy. It is also important to address difficulties with social integration, participation in leisure activities, and maintaining stamina.

Type IV

- People with OI Type IV are moderately affected. Type IV can range in severity from relatively few fractures, as in OI Type I, to a more severe form resembling OI Type III.
- The diagnosis can be made at birth but often occurs later.
- The child might not experience fractures until he or she is walking.
- People with OI Type IV have moderate-to-severe growth retardation, which is one factor that distinguishes them clinically from people with Type I.
- Bowing of the long bones is common, but to a lesser extent than in Type III.
- The sclerae are often light blue in infancy, but the colour intensity varies. The sclerae may lighten to white later in childhood or early adulthood.
- The child's height may be less-than-average for his or her age.
- It is common for the humerus and femur to be short.
- Long bone fractures, vertebral compression, scoliosis, and ligament laxity may also be present.

- Brittle teeth may be present or absent.
- Significant care issues that arise with OI Type IV include the need to prevent fracture cycles, the need to develop strategies for coping with short stature and fatigue, and the family's need for emotional support, especially during infancy. It is also important to address difficulties with social integration, participation in leisure activities, and maintaining stamina.

Type V

- OI Type V is moderate in severity. It is similar to OI Type IV in terms of frequency of fractures and the degree of skeletal deformity.
- The most conspicuous feature of this type is large, hypertrophic calluses in the largest bones at fracture or surgical procedure sites. Hypertrophic calluses can also arise spontaneously.
- Calcification of the interosseous membrane between the radius and ulna restricts forearm rotation and may cause dislocation of the radial head.

Type VI

- OI Type VI is extremely rare. It is moderate in severity and similar in appearance and symptoms to OI Type IV.
- This type is distinguished by a characteristic mineralization defect seen in biopsied bone.

Treatment

The goal of treatment is to minimize fractures, enhance independent function, and promote general health. Medical care for children and adults who have OI involves an interdisciplinary team which may include orthopedists, endocrinologists, geneticists, rehabilitation specialists, neurologists and respirologists.

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

- Physical and occupational therapy
- Casts, splints, or wraps for broken bones
- Braces to support legs, ankles, knees, and wrists as needed
- Orthopedic surgery, often including implanting rods to support the long bones in arms or legs
- Medications to strengthen bones
- Mobility aids such as canes, walkers, or wheelchairs and other equipment or aids

Safety precautions (Preschool age)

Lifting/Handling

- When handling a child with OI, all movements should be slow, methodical, and gentle.
- Never push, pull, twist, bend, apply pressure to, or try to straighten a child's arm or legs.
- Lift a child with OI onto your shoulder by placing one hand under the buttocks and legs and the other hand under the shoulders, neck, and head. Lean over the child so that there is a shorter distance to lift.
- Do NOT lift the child from under the armpits.
- When holding a child with OI, keep your fingers spread apart to provide a wider base of support and an even distribution of support pressure.
- Hydrocephalus occurs in a large percentage of children with Type III OI. Extra care is needed to support the head if it is oversize for the small body.
- Be aware of where the child's arms and legs are at all times to avoid awkward positions or getting a hand or foot caught.
- Babies should be repositioned frequently during the day. Beneficial positions for a baby with OI include being held, carried, held on a caregiver's shoulder and side lying.
- It is important for children with OI to be held and touched by caregivers and allowed to explore independent movement. Supporting the child in a variety of positions such as on the caregiver's shoulder and side lying develops muscles that will help with head and neck control and later on with sitting.
- When diapering, lift the child by the buttocks not the ankles. Spread your fingers apart as far as possible and put your hand under the buttocks with your forearm under the child's legs to prevent them from dangling.
- Burping a child with OI should be done very gently, with soft taps, possibly with padding over the hand or by gently rubbing the child's back.

Equipment

- A standard crib mattress is recommended.
- In general, surfaces that the child may bump including bed rails, car seat, stroller, and tub need to be padded.
- For severely affected children, it may be helpful to use a covered piece of foam rubber for transporting or holding them. Carrying a child with OI on a pillow is not recommended.
- Simple, lightweight cotton clothing is recommended. Clothes with buttons, snaps, or Velcro down the front and at the crotch are easier to put on than more complicated garments. Roll up sleeves or pants legs and gently pull the garment over the arm or leg. Do not pull the arm or leg through the sleeve or pant leg.
- An infant sized tub can be padded with towels or sponge. A molded sponge bathing aid is also helpful.

Physical Environment

- For children who are mobile, it is important to avoid cluttering the floor with toys/objects to avoid tripping and falling.

Transportation

- An approved car seat geared to the child's weight and ability to sit-up is appropriate.
- A padded washable cover for the seat is a good idea. However, it is unsafe to add extra padding that was not provided by the seat manufacturer.
- Other important car seat features include a well-padded harness and a head-hugger support pillow.
- Some severely affected infants with OI may require a car bed.

Preventing head flattening

All babies with OI have soft skulls. To prevent skull malformations, every effort should be made to reduce pressure on the back of the head. The following strategies are beneficial.

- Put gel pads under the baby's head when he or she is on his or her back.
- Position the baby in a propped, side-lying position.
- Frequently change the baby's position throughout the day.
- Carry the baby on your shoulder or in an approved sling carrier.
- Avoid leaving the baby in a car seat for long periods.
- Helmets have been used with some babies who have OI, but they are not universally recommended. In a severely affected baby, the additional weight of a helmet may make the already challenging task of gaining head and neck control even more difficult.

Safety precautions (school age)

Physical environment

For children with limited mobility, physical barriers may prevent their full participation in school activities. Common barriers include steps at school entrances or between floors, restrooms with narrow or heavy doors, high sinks and narrow stalls, 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. These barriers should be addressed if they interfere with a child's participation in the community program.

Architectural barriers can be overcome in a variety of ways:

- portable ramps and wheelchair lifts
- lowering equipment such as lockers, shelves, and soap dispensers
- providing a low desk or work surface
- providing the child with the assistance of a supportive adult in the restroom, if required

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

Some common mobility accommodations include the following:

- Allow the child to leave class several minutes early to avoid crowds and allow more 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 more accessible, 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.

Physical Education

Participation in physical education and recess is very important for children with OI. Physical activity helps strengthen their bones and muscles, provides a key opportunity for them to build friendships, develop social skills, and improve coordination. Though involving a child with OI in physical activities may require creativity and planning, it is a vital part of his/her education.

Physical education teachers should make every effort to involve the child in the same activities as their peers, with appropriate adaptations, rather than isolate the child by giving him or her different activities. Children with OI may be able to wheel or walk around the track while others run, go to bat while another child runs on their behalf, or kick a ball while sitting in a wheelchair. Some children with OI develop good upper-body strength due to using crutches or pushing a wheelchair and may be able to do push-ups or lift weights. 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 are able to do safely.

- A child with OI may be restricted from playing contact sports as injuries are difficult to avoid.
- Activities that jar or twist the spine should be avoided.
- Wearing a helmet and knee/elbow pads for sports such as bike riding and roller blading, is recommended.
- Properly fitted shoes help to support the ankles and prevent falls.

Recess/Playground

As with physical education, it is important that children with OI participate in recess activities as much as possible. Children with OI may be able to use traditional playground equipment, such as slides or play structures, with or without adult assistance. Some children with OI may require or benefit from accessible playground equipment. 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.

Evacuation plan

A child with OI may move more slowly than other students and be at risk of a fall if students are moving quickly or jostling each other to get out of the building safely. The child with IO may require the use of a wheelchair or other mobility devices. Establishing and practicing a child-specific fire/emergency evacuation plan for a child with OI is important and should be part of the school's Emergency Response Plans.

Transportation

- A child with OI may require the development of a Personalized Transportation Plan, which would be developed with the child's school support team.
- Children that ride a regular school bus may need someone to assist them when getting on and off the bus.
- It is suggested that a child with OI be seated in a section of the bus that offers the smoothest (least bouncy) ride. For some children with OI, other transportation options may need to be explored such as a wheelchair accessible bus or van.

Physical or Occupational Therapy

Some children with OI may benefit from physical or occupational therapy or assistive technology to maximize their skills and independence at school. Planning to meet the child's needs would occur through the student-specific planning process involving the child's school support team.

Emergency Response Plan (Preschool Age)

Any of the following situations may indicate a fracture:

- Child resists moving the sore body part
- Swelling or bruising over a bone
- Child has deformed limb
- Child is not using the limb
- Fussiness may be a sign of fracture

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. If staff needs to move the child, keep the affected area as still as possible and avoid jarring movements.
 - 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 anaesthesia.
 - Staff should only apply a splint if the parent has instructed them to do so or if the child must be moved before a parent or other caregiver arrives. Makeshift splints can be formed using a pillow, a towel, or even a magazine wrapped around the affected limb. Splints can be tied on using an elastic bandage or strips of cloth. Take care not to tie the splint on too tightly, as that will cause pain and decrease circulation. Minimize additional pain by applying the splint quickly but very gently, avoiding sudden or jarring movements. Most general first-aid classes instruct people in how to apply a splint.

Emergency Response Plan (School Age)

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 they 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. If community program personnel assist in moving the child, they should take care to keep the affected area as still as possible and avoid jarring movements.
 - 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 anaesthesia.
 - Staff should only apply a splint if the parent has instructed them to do so or if the child must be moved before a parent or other caregiver arrives. Makeshift splints can be formed using a pillow, a towel, or even a magazine wrapped around the affected limb. Splints can be tied on using an elastic bandage or strips of cloth. Take care not to tie the splint on too tightly, as that will cause pain and decrease circulation. Minimize additional pain by applying the splint quickly but very gently, avoiding sudden or jarring movements.

HEALTH CARE PLAN

When a community program receives URIS Group B support for children with URIS 'Group B' health care needs, a written health care plan is developed and maintained by a registered nurse minimally on an annual basis. The development and implementation of the health care plan should reflect the principles of inclusion, normalization, and independence.

- A child with osteogenesis imperfecta is foremost a child within a family, child-care facility, classroom, or other community program.
- The environment should be changed to support the child, not the child changed to suit the environment.
- Interventions should be as non-intrusive as possible and be delivered in a manner that respects the child's dignity and privacy as well as the normal routines and patterns of the community program.
- The parent/guardian and child have rights and obligations and should be encouraged to actively participate in decisions affecting them.

Consultation with the parent/guardian and community program is required to develop a health care plan that is relevant to the child's needs and the community program setting. For some children, the management of osteogenesis imperfecta within the community program may be complex and require consultation with health care professionals who are involved in the management of the child's osteogenesis imperfecta.

When a child has multiple health care needs, all relevant information should be integrated into one comprehensive health care plan. The format should use plain language and include information that is required to manage the child's osteogenesis imperfecta in a safe and appropriate manner during attendance at the community program.

The health care plan should be kept in a location that is secure and accessible. Community program personnel that are responsible for the child should be aware of its location. The health care plan should accompany the child on excursions outside the facility.

Content

The following information is included in the osteogenesis imperfecta health care plan.

Demographic information

- Child's name
- Birth date
- Community program name
- Parent(s)/guardian(s) name and phone number(s)
- Alternate emergency contact name and phone number(s)
- Specialist(s) name and phone number
- Family physician/pediatrician name and phone number

Medical information

- Medical diagnoses and other relevant conditions
- Known allergies
- Availability of Medic-Alert® identification
- Prescribed medications
 - If medication is administered at home, the name of medication is required.
 - If medication is administered at the community program, the medication name, dose, location, time, and route of administration is required.

Osteogenesis imperfecta information

- Type of OI and/or description of child's physical features (e.g. stature, muscle weakness, bleeding tendencies, brittle teeth, hearing loss, vision problems, breathing problems)
- History (e.g. when diagnosed, frequency of fractures)
- Current treatment (e.g. physical therapy, occupational therapy, mobility aids)
- Child's ability to recognize and respond to physical injury

Potential Problems

- Responding to fractures

Safety precautions

Documentation

- Template for recording interventions and actions performed by the nurse and/or community program personnel (e.g., communication, actions taken)
- Signatures and dates
 - Nurse signature and date(s) of health care plan development/review
 - Parent/guardian signature and date

TRAINING

When a community program receives URIS 'Group B' support, training is provided to community program personnel by a registered nurse. Training is provided minimally on an annual basis. The training of community program personnel should reflect the principles of adult learning.

- The learning needs of participants should be identified and integrated into the training session.
- Information should be applicable to the participants' responsibilities and focus on what is most useful to them.
- Adults have accumulated a foundation of life experiences and knowledge and need to connect learning to this knowledge base.
- An organized training session with clearly defined elements assists participants in identifying and attaining learning goals.

It is recommended that all community program personnel that may be responsible for a child with osteogenesis imperfecta attend the training session. As an example, community program personnel that may be responsible for a child with osteogenesis imperfecta in a school may include teachers, educational assistants, school administrators, office staff, substitute teachers, bus drivers, and lunch room supervisors. The community program is responsible to ensure personnel that may be responsible for a child with osteogenesis imperfecta attend the training session. It is required to keep a written record that indicates community program personnel in attendance and the date that training occurred.

Adequate time should be scheduled for training to ensure community program personnel obtain the knowledge and skill necessary to safely respond to the needs of children who have osteogenesis imperfecta within their facility. The amount of time required to train community program personnel will vary depending on factors such as the existing knowledge of community program personnel, number of persons attending the training session, and the format of training resources used (e.g. PowerPoint, Worksheet).

To ensure service is provided in an efficient manner, training should be scheduled when all community program personnel that may be responsible for the child can attend. If an adequate number of community program personnel did not attend the training session, additional training should be scheduled. If subsequent training sessions are also poorly attended, alternate strategies should be discussed with the community program to ensure training is provided in an efficient manner. If the issue remains unresolved, the URIS Provincial Coordinator and/or relevant agency (e.g. School Division Student Services Administrator, Manitoba Early Learning and Child Care,) may need to become involved.

When the community program has not received training, a child with osteogenesis imperfecta may attend the community program. In such situations, the community program's standard policy for medical emergencies is implemented, as required.

Content

The following clinical information and child-specific information is included in the training session.

Standard clinical information

- Osteogenesis imperfecta
- Types of osteogenesis imperfecta
- Treatment
- Safety precautions
- Responding to fractures

Child-specific information

- Type of osteogenesis imperfecta and/or description of child's physical features
- Current treatments
- Safety precautions specific to child
- Additional information specific to child

Training Resources

If alternate resources are used, it is the responsibility of the nurse to ensure its content is consistent with the clinical information included in this document.

MONITORING

Monitoring of trained community program personnel by a nurse is required to ensure that the knowledge and skill necessary to safely care for children with osteogenesis imperfecta has been retained. Monitoring is required minimally on an annual basis.

The frequency and timing of monitoring is based on the professional judgment of the nurse as well as the complexity of information taught, maturational issues, and the skill demonstrated by community program personnel.

The community program may also request additional monitoring if personnel have questions/concerns, and/or require additional support to ensure they are responding to the child's needs in a safe and appropriate manner.

REFERENCES

Osteogenesis Imperfecta Foundation (2007). *Guide to Osteogenesis Imperfecta for Pediatricians and Family Practice Physicians*.

Osteogenesis Imperfecta Foundation. *Plan for Success: An Educator's Guide to Students with Osteogenesis Imperfecta*. 1998.

Osteogenesis Imperfecta Foundation. *Infant Care Suggestions for Parents*

Osteogenesis Imperfecta Foundation. *Therapeutic Strategies for Imperfecta Osteogenesis: A Guide for Physical Therapists and Occupational Therapists*

RESOURCES

The following list includes resources that may be relevant to community programs in the care of children with endocrine conditions. The purpose of these agencies/organizations may not be consistent with the purpose and content of this manual.

Osteogenesis Imperfecta Foundation

www.oif.org