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Evidence-Based Physical Therapy Management of the Child with Osteogenesis Imperfecta

Alicia Fernandez-Fernandez, PT, DPT, PhD
Associate Professor, Physical Therapy Dept.
Nova Southeastern University, FL
Per diem physical therapist, South Miami Hospital

Learning Outcomes

• Describe the incidence, etiology, pathophysiology and clinical presentation of osteogenesis imperfecta (OI).
• List at least 3 pharmacological or surgical approaches to management of the child with osteogenesis imperfecta.
• Describe evidence-based therapeutic strategies for the child with osteogenesis imperfecta, including exercise prescription, modifications, adaptive equipment, and parent education.
• List at least 3 considerations for safe handling of a child with osteogenesis imperfecta.
OSTEOGENESIS IMPERFECTA (OI): INCIDENCE, ETIOLOGY, PATHOPHYSIOLOGY, AND CLINICAL PRESENTATION

Osteogenesis Imperfecta

- Disorder of collagen synthesis
  - Affects bone and connective tissue
- Several types – different severity
- Very diverse clinical features!
  - Even within the same type
- Prevalence: 30,000-50,000 people in the US
- Incidence: 1 in 20,000
Etiology/Pathogenesis

• Inherited mutation of Type I collagen-forming genes
  ◦ Most commonly autosomal dominant; some forms recessive inheritance
  ◦ BUT 25% have a spontaneous gene mutation
  ◦ More than 150 different mutational variations are possible!
    • All affect genes that code for type I collagen or associated proteins that are involved in collagen production.
    • Type I collagen is a major structural component of bone, skin, and tendon

Collagen: major component of extracellular matrix in connective tissue
• Greek word meaning “glue producer”
• Tensile strength close to steel
• Triple helix (tropocollagen)
• Arranges in fibers and fascicles
• Cross-linking and “braiding” makes it very strong
• Type I collagen provides tensile strength to tissue
Osteogenesis Imperfecta (OI)

- Defects in bone ossification and bone modeling
  - Smaller cross-sectional area, thinner cortex, reduced cancellous bone mass
  - Smaller number of trabeculae
- Poor ossification of bone leads to skeletal deformity and frequent fracture.
  - LE more affected than UE.
- Musculoskeletal Signs and Symptoms
  - Short stature
  - Frequent fractures, bruising, dislocation, ligament injury (laxity)
  - Osteoporosis; bowing of long bones; spinal deformities
  - Joint hypermobility, muscle atrophy and weakness, delayed motor function
• Other areas affected:
  ◦ Hearing (deformity of bony auditory structures)
  ◦ Vision (tinted sclerae, misshaped lens)
  ◦ Teeth
  ◦ Cardiopulmonary
  ◦ Easy bruising
  ◦ Excessive sweating

Classical classification of OI

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
<th>Characteristics</th>
</tr>
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</table>
| Type I | Mildest and most common (50% of cases). Collagen production reduced by 50% | Mild to moderate bone fragility
         |                                                                | No deformity
         |                                                                | Fractures occur before puberty
         |                                                                | Hearing loss
         |                                                                | Easy bruising
         |                                                                | Normal stature |
| Type II| Most severe and perinatal lethal
         | Death occurs in utero, infancy or early childhood
         | Only 20% collagen production
         | In utero fractures and growth retardation
         | Severe bone deformity (micromelia)
         | Large, soft cranium
         | Connective tissue fragility |
| Type III| Moderately Severe
         | In utero fractures
         | Bony deformity, bowing, osteoporosis, scoliosis
         | Progressive deformity, extremely short stature
         | Triangular-shaped face
         | WC dependent by teenage years |
| Type IV| Milder; usually normal life expectancy
         | Osteoporosis
         | Skeletal deformity, more severe than type I
         | Fractures before puberty
         | Triangular-shaped face |

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Osteogenesis Imperfecta (OI)

- **Prognosis**
  - **Type I and IV**
    - Normal life span
  - **Type II**
    - Certain early death
    - No effective treatment for Type II
  - **Type III**
    - Mortality can occur from respiratory failure or intracerebral hemorrhage
Recent discoveries: New types

- Types V and VI
  - Similar to type IV, but unknown mutations, not in collagen genes themselves
  - Type V: hypertrophic calluses after fracture or surgery, ossified interosseous membrane at the forearm (restricted pronation/supination).
  - Type VI: bone has “fish-scale” appearance under microscope, elevated alkaline phosphatase. Recessive?

- Types VII and VIII
  - Recessive inheritance
  - Gene mutations associated with collagen gene translation; Varied clinical presentation

A very important caveat

- “The features of OI vary greatly from person to person [...] When working with an individual who has OI, it is most important to focus on his or her particular abilities, strengths, and weaknesses, rather than on his or her OI type.”

(Osteogenesis Imperfecta Foundation)
Orthopedic management

- Fracture prevention
  - Positioning
  - Handling precautions
  - Lightweight orthoses
    - HKAFOs preferred to KAFOs
Orthopedic management

• Fracture management
  ◦ Minimize immobilization time
  ◦ Primary strategy: rest / eliminate weight-bearing
  ◦ Pain management
  ◦ Lightweight fracture containment
    • Soft splint
    • Fiberglass
    • Air splints/casts

Surgical Management

• Telescopic IM rodding
  • For recurring fractures
  • “Grow with the bone”
  • Indications: frequent fractures, large angle deformities, unstable LE
  • Complications
• Osteotomy
  • Control of rotational deformities
• Osteotomy & intramedullary rod fixation to correct bowing
• Surgical fusion for severe scoliosis
Medical/ Pharmacological Management

- Improving bone density
  - Bisphosphonates (eg pamidronate, zoledronate)
    - Inhibit osteoclast function, improve bone density, decrease fracture incidence
  - Growth hormone
    - Some benefits, but may increase fracture incidence….
  - Bone marrow transplant; stem cell therapy
    - Increase bone mineral content and stimulate bone growth by new osteoblasts
  - Other approaches: Calcitonin, fluoride, vitamin D, monoclonal antibodies
    - Need more data

Future therapies: Gene Therapy

- Need two combined strategies:
  1. Silencing dominant mutated genes:
     Antisense gene therapy
     - RNA or DNA molecule complimentary to the mRNA for the mutant protein binds to the target mRNA to prevent its translation.
     - Still many challenges
  2. Fostering normal gene expression: Gene supplementation therapy
     - Use cell supplementation, or overexpression of the normal collagen gene
EVIDENCE-BASED THERAPEUTIC STRATEGIES TO IMPROVE FUNCTIONAL OUTCOMES IN CHILDREN WITH OI

Framework for our discussion

- International Classification of Functioning, Disability and Health (ICF)
International Classification of Functioning, Disability and Health (ICF)

- Purposely Designed
  - Focus on health & function, NOT disease & disability (words are purposely positive)
    - Two sides of same coin: functioning vs disability
    - Functioning
      - All body functions, activities, & participation that people use to have a meaningful life
  - Framework to define health and health-related outcomes
  - Biopsychosocial Model: Combined medical view & social view of disability

Views on “Disability”

- Medical view
  - Disability is feature of the person, directly caused by disease, trauma, or condition that requires medical care to “correct” problem

- Social view
  - Disability is feature of society, requiring a political response. For example, inaccessible environment (attitudes and social norms) needs policy change

- Biopsychosocial view (ICF Model)
  - Disability is complex issue involving person, environment, & society
  - Optimal patient outcomes can only be achieved if we look at the “big picture”
Interaction of Concepts in the ICF

Terminology for ICF Model

- Body function
  - physical & psychological body functions (pain, motion, strength, speed, etc)
- Body structure
  - anatomical parts of body
- Impairments
  - problems in body structure or function
- Activity
  - execution of task or action
- Participation
  - involvement in life situation
Terminology

- Activity limitation
  - Difficulty performing an activity
- Participation restriction
  - Difficulty engaging in life situations
- Contextual factors
  - Personal & environmental factors that impact outcomes
- Environmental factors
  - Physical, social, and attitudinal environment in a person's life-external to the person but impacting their function: barriers or facilitators
- Personal factors
  - Gender, age, coping style, social background, education, profession, experiences, character, & factors that influence how disability experienced

What's the end goal of therapeutic intervention?

- “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.”

(Osteogenesis Imperfecta Foundation)
Functional classification systems in OI

- Classification systems related to functional outcomes
  - Binder et al – describe expected functional outcome based on body size & limb proportions
    - For example:
      (a) largest heads, shortest trunk and bowed upper extremities = least mobile, frequently fractured UEs.
      (b) smaller heads, lower head/trunk ratios able to pull to stand and higher level of independence in self care.

Functional prognosis

- In general, prognosis for ambulation depends on severity of disease and ability to sit up independently by 9 months of age
  - Most of Type I can walk with or without assistance for community or household distances.
  - Types III and IV frequently will need power mobility
- With the proper environment and equipment, most people with OI can function well in most areas of daily life.
When are PT and OT appropriate?

- Addressing delayed motor development
- Addressing cardiorespiratory dysfunction
- Post-fracture or surgery
- Consultation about safe activities, environmental adaptations and adaptive/assistive equipment
- Learning a new skill or adapting the way a skill is performed

Motor development

- Developmental motor skills impaired due to:
  - Joint hyperlaxity
  - Fracture history
  - Muscle atrophy
  - Over time, deformity
  - BUT ALSO LACK OF OPPORTUNITY!
It’s a fine balance…

- We want to avoid fractures…
- BUT
- We want them to be active
  - Mechanical stresses and muscle tension on bone help increase bone density - “Wolff’s law”
  - Not moving reduces strength, creates bowing from lack of variation in position (sitting)
  - Not moving makes children fearful of moving and being moved, and it also makes them functionally dependent

Overall strategy

- “The child with OI will require slow, graded introduction to being moved in progressively less supported positions.” (Marnie King, OT)
- It is important to consider and apply safe handling principles when developing a plan of care for the child with OI
Safe Handling Principles

- When child with OI is old enough, before handling 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!
- Never pull, push, or twist a limb. Avoid passive rotation of the arms, legs, head, or trunk.

Safe handling principles

- 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).
Safe handling principles

- 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 weight-bearing support if placing too much stress on the LEs is a concern.
  - Straddle riding toys, gait trainers with sling seats, braces or splints on the legs.

Osteogenesis Imperfecta Foundation

Safe handling principles

- Avoid activities that place large stresses on bones.
  - Excessive hip flexion (“jack-knife” position, with the person leaning far forward in sitting)
  - Diagonal trunk rotation
  - Bridging
    - Stresses tibial bones in the same plane in which they tend to bow.

Osteogenesis Imperfecta Foundation
Safe handling principles

- Proper support during transitions and handling
  - Turns, transfers, etc. but also during burping, cuddling, holding, diaper changing.
  - Infant—fully supported, align body (midline orientation), protection against hard surfaces.
  - Do not lift by arms or legs.

- PROM to obtain hip extension safe if used in moderation
  - Avoid rotational forces, use careful myofascial stretching in straight planes. PROM/stretching possibly unsafe in infants with OI.

Safe handling and safe environment

- AROM precautions:
  - Avoid weight directly near joint lines.
  - If providing resistance, avoid long lever arms.

- When facilitating, use proximal key points of control.

- Wheelchair fit
  - Bones can bow around supporting surfaces.

- Avoid environmental dangers
  - Furniture, trip hazards, carpets, wet floors (for example around a pool).

Osteogenesis Imperfecta Foundation
Therapeutic Interventions

- Goals to address
  - Prevent or minimize deformities that interfere with function
  - Maximize active movement
    - Promote bone mass deposition
  - Promote normal motor development
  - Increase endurance
  - Prevent cardiorespiratory compromise
  - Use of orthotics & standing programs
  - Parent education

Therapeutic Interventions

- Strengthening
  - Active, playing with lightweight toys
  - Active to active resistive with older children (caution)
  - Focus on hip extensors/abductors and spinal musculature

- Endurance activities

- Water / swimming
  - Buoyancy or resistance as needed
  - Limit to 20-30 minute sessions and monitor pool temperature (elevated metabolism)
• Evidence: A hip extension, hip abduction, spinal muscular strengthening program plus twice a week swimming results in increased ability to assume and maintain erect posture and ambulation in children with OI (Goodman, 2014)

• Evidence: A 4-year follow-up study of rehabilitation in OI showed an increase in self-care and social function with all types of OI, although mobility level plateaued in moderate-severe OI (Engelbert, 2004)

• Evidence: Children with types I and IV OI who participated in a low-resistance physical training program had improved aerobic capacity (peak oxygen consumption, maximal working capacity), muscle strength, and reduced levels of subjective fatigue after 3 months. The program was safe and effective. The improvements diminished 6 months after program completion, suggesting that regular exercise of the correct intensity is important to improving OI fitness. (Van Brussel, 2008)
Therapeutic Interventions

- Whole body vibration (WBV)
  - Some evidence for effectiveness in improving bone density and strength in children and teens with OI (Semler, 2007 and 2008)
  - Tilt table, vibrating platform
  - Research shows 35° tilt angle provides best results.
  - Not appropriate for children with telescoping rods or hx of subluxation

Therapeutic interventions

- WBV plus exercise
  - 12 months of treatment with whole body vibration (for 6 months), physiotherapy, resistance training, treadmill training and 6 months follow up (Hoyer-Kuhn, 2014)
  - At 12 months, significant improvement in motor scores in GMFM, walking distance, and whole body mineral density.
  - Caveat: No control group…
  - Also: cost of WBV equipment
Therapeutic interventions

- Developmental weight-bearing activities important to keep ROM & mobility, esp. pelvic girdle
  - Prone (caveat: may be unsafe in severe OI)
  - Rolling & floor mobility
  - Ambulation with assistive device
  - Sitting & pulling to stand (with orthoses if needed)
    - Standing frame / HKAFO
<table>
<thead>
<tr>
<th>Developmental Progression</th>
<th>Modification for Child with OI</th>
</tr>
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</table>
| Supine                    | *Avoid skull flattening through gel pads or positioning changes  
*Use a foam surface to guide shoulders forward and help child get hands to midline.  
*Use splints if UEs tend to bow |
| Prone                     | *Use a chest wedge or parent chest  
*This may not be an appropriate position for some children – may be unable to transition from this position |
| Inclined sitting          | *Avoid skull flattening with pad  
*Support torso with blanket rolls.  
*Avoid wide hip abduction  
*Support trunk with wide straps or vest  
*Support feet in neutral |
| Side-lying                | *Support head and below axilla  
*May need support under upper arm and leg when sleeping |

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| Rolling                   | *Use blanket like a hammock and slowly tilt child to reach for a ball or object.  
*Then partial rolling in a blanket on a firmer surface.  
*Once child is able to tolerate side motion, use slight wedge to roll down hill. |
| Sitting                    | *Supported: Can use positioning chair. Provide head and neck support at first, then fade.  
*Unsupported: Avoid ring sit. |
| Transition into sitting   | *Try in pool first, child side-leaning on parent’s thigh and trying to get to a sit.  
*On land, side-lying on a wedge or parent/therapist thigh. Avoid excessive wrist extension. |
| Sit-pivot, 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 until water is child’s hip height. Then try on land on slippery bench (caution!).  
*Use as transfer method from chair to bed to toilet during fractures and if legs are not strong enough to stand. |
### Developmental Progression Modification for Child with OI

<table>
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</table>
| Crawling                  | *Start in kneeling position with chest supported by partially inflated beach ball and aim up hill 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, old panty hose top, or wide stockinette from child’s waist to ankles. |
| Kneel/pull to stand        | **“Mermaid suit” 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.  
*Progress to crawling in water the height of child’s knees. |

These developmental progressions are adapted from King, 2001.

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### Therapeutic Interventions

- **Independence**
  - Transfers
  - Self-care
  - UE function has huge impact (personal hygiene, clothing)

- **Mobility**
  - Assistive devices
  - Manual or power WC
    - Seating system
      - Aligned head & trunk

**continued**
Therapeutic Interventions

- Adaptive equipment and assistive device considerations:
  - Energy conservation
    - Keep things close – organize a “work station”
  - Joint protection
  - Mobility
  - Accessibility

Adaptive equipment considerations (Osteogenesis Imperfecta Foundation)

<table>
<thead>
<tr>
<th>Type of Equipment</th>
<th>Common Considerations for People with OI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Walkers</td>
<td>Supported walking allows weight bearing in legs, which increases strength and bone density. Need 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.</td>
</tr>
<tr>
<td>Crutches and canes</td>
<td>Supported walking allows weight bearing in legs, which increases strength and bone density. Need 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.</td>
</tr>
<tr>
<td>Wheelchairs</td>
<td>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.</td>
</tr>
</tbody>
</table>
Adaptive equipment considerations
(Osteogenesis Imperfecta Foundation)

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

Braces/splints
Should be circumferential, perforated (because of excessive sweating), and lightweight. Leg braces can help with alignment and promote standing and walking. Their role in preventing fractures is limited. Lightweight forearm splints (divisible 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, 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.

Adaptive equipment considerations
(Osteogenesis Imperfecta Foundation)

Positioning aids
Pillows, bolsters, towel rolls, gel pads, etc. Promote 90/90/90 position in car seat, wheelchair, stroller, etc. rather than "frog leg" position. Encourage side-lying and prone positions for infants and young children with OI, who often spend a lot of time on their back.

Standers
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, 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/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
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.
Adaptive approaches for self-care tasks

- **Bathing**
  - Slip-resistant surface
  - Towel or gel pad under buttocks
  - Transfer bench, grab bars
  - Long-handled scrub brushes
- **Toileting**
  - Lower toilets if short stature
  - Add trunk support, transfer bench or grab bars if needed
  - Toilet-paper reacher if short UEs

- **Dressing**
  - Clothes that are easy to don/doff without pushing, pulling
  - Dressing tools
  - Keeping clothes within reach
- **Food preparation**
  - Lowered counter tops and appliances, long-handled faucets, pull-out shelving
Measuring Functional Outcomes

- Many options.
  - For example, GMFM, BAMF and Bleck ambulation score have been validated in OI
- Several literature reports have also used the Pediatric Evaluation of Disability Inventory (PEDI) in this population
  - The PEDI measures current functional performance in children up to 7 ½ year old (or older if functioning at younger level).

PEDI

- Children who use mobility aids are scored with these aids: “true performance” within the environment.
- Assessment combines parent interview and observation of the child’s performance.
- Scores are scaled to 100 (full function)
**PEDI**

- Functional Skill Scale, 3 subtests:
  - **Self care**: eating, grooming, dressing, bathing, toileting
  - **Mobility**: transfers (chair, bed, toilet, tub/shower, and car), indoors and outdoors mobility
  - Social function: communication, social interaction, household and community tasks.

- Environmental modification and amount of caregiver assistance are recorded also
  - Modification Scale and Caregiver Assistance Scale

**Family Education - CRUCIAL**

- Safe handling principles
  - Fractures can occur during ordinary activities!

- Appropriate activity modifications to ensure safety

- Importance of activity to motor development
  - Avoid rough play, BUT:
  - Play is therapeutic!

- Check for fractures daily
  - Unusual limb position, swelling and tenderness, crying, child avoids using the extremity
Family Education

- Bone health
- Promoting self-care as they become older
- Preventing child abuse accusations
  - Carry documentation of the diagnosis (letter from pediatrician with weekend and night-time contact information)
  - Meet with the supervising physician of the emergency department at their local hospital
  - Plan ahead with child’s orthopedist about how to handle fractures that occur at night, or on weekends or holidays
Care throughout the lifespan

• Infant with OI
  ◦ Assess AROM (goniometer or functional)
  ◦ PROM avoided in infants due to fracture risk
  ◦ Assess strength (functionally)
  ◦ Assess gross motor development (Peabody, PEDI, Bayley Scales, Brief Assessment of Motor Function)
  ◦ Assess pain using appropriate scales (FLACC)
  ◦ Can start pool therapy as early as 6 months

Care throughout the lifespan

• Infant with OI
  ◦ Assess equipment: car seat
  ◦ Assess caregiver handling and positioning techniques
    • Safe bathing, dressing and carrying
    • Safe positioning with proper alignment
    • Protect but also provide opportunity for motor development
      • Prone play, antigravity head stability
      • Rolling, sitting
• Use a variety of holding positions
• Do not use baby walkers or jumpers
Remember to use proximal key points of control

Care throughout the lifespan

- Preschooler with OI
  - Emphasis on protected weight-bearing, self mobility to promote independence
  - More focus on active role for child in their own care
  - Standing must be promoted, actively or through standers
    - Do not want unloading-related osteoporosis superimposed on OI
    - BUT may need to start working on standing and gait in unloaded environment (pool), or use braces and assistive devices
Care throughout the lifespan

- Preschooler with OI
  - Continue active strengthening through play; esp. hip extensors, abductors. Can use light weights with short lever arms
  - Some activity ideas:
    - Straddle-roll activities with therapist facilitating or stabilizing at the pelvis
    - Scooter-board activities
    - Riding tricycles
    - Playground games (Follow the Leader)
    - Overhead reaching / modified ball throw activities
    - Pool activities with floats; breathing activities

- Must have emergency plan if fracture happens at school
- Evaluate needs for adaptive equipment and mobility
  - If needed, powered mobility used after age 2
  - Scooters, ride on toys, etc. Keep the child mobile!
  - For those who are able, gait with walker followed by crutches. Some will stay with walker.
- Continue to use developmental assessment tools
Care throughout the lifespan

• School-age and adolescent with OI
  ◦ Encourage participation in peer activities with necessary adaptations
  ◦ Maintain proper alignment, strength (including core), and endurance
  ◦ Manage spinal deformity and long bone deformity
    • Spinal fusion? Evidence shows spinal orthotics INEFFECTIVE.
    • Consider modifications to mobility equipment to accommodate deformities.
  ◦ Frequency of fracture lower after puberty
    • BUT still need to emphasize responsibility and self-care

Care throughout the lifespan

• School-age and adolescent with OI
  ◦ Adolescents who used manual wheelchair may now be able to ambulate in the household
    • But most still use wheelchair in the community
  ◦ Independent mobility is key goal of this period
  ◦ Match the teen’s interests — adaptive sports activities, aquatic activity, resistance training (with caution), customized fitness programs
    • Let them decide, but guide
Care throughout the lifespan

- Adulthood
  - Independent living skills
    - Household ambulation with assistive device
    - Community ambulation with manual or powered mobility.
  - Personal goals: work, study. Help with task-specific strategies
  - Maintain core strength; monitor health
  - Reemphasize life-long bone health principles
    - Exercise, healthy weight, avoid alcohol and tobacco, limit caffeine, healthy calcium-rich diet.
  - Good resource: Maintaining health in the adult years
    - [http://www.oif.org/site/PageServer?pagename=AdultHealth](http://www.oif.org/site/PageServer?pagename=AdultHealth)

One closing thought: Listening

- “Keep in mind that 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.” (Osteogenesis Imperfecta Foundation)
- Ask about their goals, their challenges, their successes. You can always learn something!
Resources

Osteogenesis Imperfecta Foundation

www.oif.org

- Children with OI: Strategies to Enhance Performance (book)
- Therapeutic Strategies
- Parent Tips

References

References


CONTINUED
References


References