Children's Special Health
Internal Policy/Statement
Osteogenesis Imperfecta

Description

Osteogenesis Imperfecta (OI) is a congenital connective tissue disorder. Clinical characteristics include: Fragility of bone, osteopenia, variable degrees of short stature, and progressive skeletal deformities. Additionally clinical manifestations are: blue sclera, dentinogenesis imperfecta, joint laxity, and maturity-onset conductive hearing loss. OI syndromes are subdivided into 6 types (IA, IB, II, IVA, IVB):

- OI Type I (mild): most common type, characterized by osteopenia leading to fractures, distinctly blue sclera, and high incidence of adult onset hearing loss. Within OI Type I, there are two subgroups:
  - IA – characterized with normal teeth
  - IB – characterized with dentinogenesis imperfecta
- OI Type II: lethal in perinatal period
- OI Type III (progressive deforming): severe osteopenia leading to multiple fractures, progressive deformity of bones and spine, and severely decreased height
- OI Type IV (moderately severe): is rare and more marked than OI Type I, characterized by osteopenia leading to fractures, sclera are normal, short stature, deformity of long bones and spine. Within OI Type IV, there are two subgroups:
  - IVA – tend to have normal teeth
  - IVB – marked dentinogenesis imperfecta

Diagnostic Criteria

- Confirmed with collagen biochemical studies
- Severe OI can be detected as early as 16 weeks of gestation by ultrasound

CSH Coverage

- Only **providers** listed on the Eligibility Letter will be paid
- **Labs/Tests** must be performed by a Wyoming Medicaid provider
- **Well Child Checks** (coverage limited to Pediatrician) according to AAP Periodicity Schedule
- **Medications**
  - Methotrexate
- **Equipment/Supplies**
  - PRE-Authorization required

Contact CSH for questions regarding additional medication and/or equipment/supplies

Minimum Standards of Care/Care Coordination

Refer to Care Coordination Manual, Ch. 3, Pg. 8, Child and Family Assessment

- Perform **Nursing Assessment** with detailed focus on the following:
  - Muscular/Skeletal system
  - Eyes
  - Oral health (i.e. multiple dental caries)
  - Nutrition and eating patterns
  - Exercise and physical activity
  - Current medications/any side effects or reactions
  - Known food and/or drug allergies
  - Height and weight, plot on growth curve
- Encourage testing as recommended by the American Academy of Pediatrics (AAP)
- School performance and behavior
- Encourage family and child to live as "normal and active" life as possible

Contact CSH if family is Non-Compliant (i.e. repeated missed appointments, failure to follow healthcare plan)

- **Referrals** that may be recommended *(CSH prefers Pediatric Specialist, if possible)*

  Visits to Providers may be limited due to budget
  - Orthopedic Specialist
  - Otolaryngologist (ENT)
  - Genetics
  - Dentist
  - Physical Therapy
  - Mental Health
  - Link the child and family with appropriate and needed services

Specialists may or may not be covered by CSH Program

- **Well Child Checks**
  - Immunizations (including vaccinations)
  - Assess and follow-up any abnormal findings
  - Dental
  - Vision
  - Hearing

- **Emergency Preparedness Plan**
  - Medic Alert ID bracelet / necklace should be encouraged
  - Medical Emergency Plan of what to do for the child's care when away from home or with a different caregiver (i.e. extremely fragile bones – easily fractured, limited mobility and function abilities)
  - Discuss self-management of the disease
  - Encourage the family to speak with the child's school in regards to the school's policy on Osteogenesis Imperfecta and emergency plan (i.e. who will administer medical attention in the event of fracture, limited mobility and function abilities, classroom accessibility)

- **Health Record**
  - Encourage family to maintain a record of the child's health information ("Packaging Wisdom" as a suggestion) that includes:
    - Medication administration
      - Type
      - Dosage/Frequency, any side effects or response to medication
    - Hearing screens
    - Dental Health
    - Fractures/Injuries
      - Type
      - When/Where
    - Treatment/procedures and hospitalizations
    - List of providers and contact information, if available

- **Transition**

Refer to the Care Coordination Manual, Ch. 3, Pg. 10, Coordinating Care
  - Discuss with the family if the child is eligible for an IFSP, IEP, or qualify for Section 504 according to the American Disability Act (ADA)
  - Use or need for special modifications to access community resources/services