Children's Special Health
Internal Policy / Statement
Neurofibromatosis (NF)

Description

Neurofibromatosis is progressive in nature, signs and symptoms may not manifest at birth they may begin in the first few years of life. There are two types of Neurofibromatosis:

- Neurofibromatosis Type 1 (NF1)
  - NF1 (aka Von Recklinghausen) is a hereditary disease with about 50% of patients having a fresh gene mutation. It is characterized by café-au-lait spots, cutaneous and plexiform neurofibromas, pigmented iris hamartomata, macrocephaly of postnatal onset and mild short stature. Occcasional abnormalities may include optic glioma, astrocytomsa, seizures, mental deficiency, and headaches.
- Neurofibromatosis Type 2 (NF2)
  - NF2 (acoustic neurofibromatosis) usually has a later age onset during the second and third decades. NF2 is often more severe than NF1 because multiple intracranial tumors can develop in childhood, in addition to cataracts.

Diagnostic Criteria

NF1 (most common) an individual must have **two or more** of the following:

- Family history (first-degree relative) with NF1
- Six (6) or more café au lait spots
- Two (2) or more neurofibromas OR one (1) plexiform neurofibromas
- Auxiliary or inguinal area freckling
- Skeletal abnormalities (i.e. severe scoliosis, enlarged/deformed bone not including the spine)
- Optic glioma (tumor of the optic pathway)

NF2 (presumptive)

- Family history (first-degree relative) with NF2
- Unilateral Vestibular Schwannomas OR any two of the following:
  - Meningioma
  - Glioma
  - Schwannoma
  - Juvenile posterior subcapsular lenticular opacity
  - Juvenile cortical cataract

NF2 (confirmed)

- Bilateral Vestibular Schwannomas, also known as Acoustic Neuroma

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**
  - None (If child is diagnosed with seizures, refer to Convulsive Disorders policy for coverage)
- **Equipment/Supplies**
  - None

Contact CSH for questions regarding additional medications 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:
  - Development screening (i.e. learning/intellectual disabilities, behavioral)
  - Exercise and physical activity, coordination
  - Skin examination (i.e. peripheral/plexiform neurofibromas)
  - Current medications/any side effects or reactions
  - Known food and/or drug allergies
  - Skeletal growth abnormalities (i.e. macrocephaly, enlarged digits, sunken chest, ect.)
  - Height and weight, plot on growth curve

    - Hypotonia
    - Precocious puberty
    - Blood pressure (could become high due to renal artery stenosis)
- 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

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

  Visits to Providers may be limited due to budget
    - Neurologist/Neurosurgeon
    - Audiologist
    - Ophthalmologist/Optometrist
    - Geneticist
    - Dermatologist
    - Developmental Specialist
    - Mental Health
    - Endocrinologist (Pediatric preferred)
    - Cardiologist (Pediatric preferred)
    - 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 (This will vary according to disease severity)
    - Discuss self-management of the disease
    - Encourage the family to speak with the child's school in regards to the school's policy on Neurofibromatosis and emergency plan (i.e. participating in school sports or PE due to bone fragility)

- **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
        - Examine/document the size and location of masses, track developmental changes in existing nodules
        - Pain/Pain management
        - 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)
    - Encourage family to do life-care planning (i.e. medical expenses, physical limitations, sexuality)