Prior Authorization Required From CHN

Eteplirsen (Exondys®) Guidelines

Coverage guidelines for Eteplirsen are made in accordance with the Department of Social Services (DSS) definition of Medical Necessity, as established by state law. The following factors are guidelines *only.* Coverage determinations are based on an assessment of the individual and his or her unique clinical needs. If the guidelines conflict with the definition of Medical Necessity, the definition of Medical Necessity shall prevail.

Payment will be considered for patients when the following guidelines are met:

- Patient has a diagnosis of Duchenne muscular dystrophy (DMD) with mutation amenable to exon 51 skipping confirmed by genetic testing.
  (*NOTE: physician must provide results of genetic testing*)

- Must be prescribed by or in consultation with a physician who specializes in the treatment of Duchenne muscular dystrophy.

- Patient is currently ambulatory and able to achieve an average distance of at least 180 meters while walking independently over six minutes.
  (NOTE: physician must attach a baseline 6 – Minute Walk Test [6MWT])

- Patient is currently stable on oral corticosteroid regimen for at least 6 months.

- Must be dosed on FDA approved dosing: 30mg/kg once weekly.

If guidelines for coverage are met, initial authorization will be given for 6 months *only*.

Requests for continuation of therapy will be considered at 6 month intervals when the following are met:

- Patient has demonstrated a response to therapy as evidenced by remaining ambulatory (able to walk with or without assistance, not wheelchair dependent).

- An updated 6MWT must be provided documenting the patient is able to achieve a distance of at least 180 meters.

PA requests for coverage of Eteplirsen must be submitted by the prescriber in the form of a letter of medical necessity to the Department's Medical Director. Letters of medical necessity should be faxed to (860) 424-4822 with the required documentation outlined in the Eteplirsen Coverage Guidelines.

Prior Authorization Required From CHN

Nusinersen (Spinraza®) Guidelines

Coverage guidelines for Nusinersen are made in accordance with the Department of Social Services (DSS) definition of Medical Necessity, as established by state law.

The following factors are guidelines *only.* Coverage determinations are based on an assessment of the individual and his or her unique clinical needs. If the guidelines conflict with the definition of Medical Necessity, the definition of Medical Necessity shall prevail.

The following factors will be considered in the Department's prior authorization decision

I. Type 1 Patients

SMA Type 1 patients with symptom onset at < 6 months of age are eligible for treatment if the following conditions are met:

1) The Diagnosis of SMA1 must be made by a Neurologist with expertise in diagnosing Spinal Muscular Atrophy. For purposes of this guideline, "Neurologist" refers to a Board Certified Neurologist, preferably specializing in pediatric neurology, **AND**

- Genetic testing confirming **both**:
  - Mutation/deletion in chromosome 5q with
  - Homozygous gene deletion/mutation of exon 7 at 5q13 *or* compound heterozygous mutation: deletion SMN1 exon 7 allele1 and mutation of SMN1 allele 2

- And at least two copies of SMN2; AND

2) Documentation that the patient is either **not on any artificial ventilation,** or if on artificial ventilation or other mechanical respiratory support prior to **Nusinersen,** the type, duration and degree of ventilator support in a 24 hour period must be documented; AND

3) Submission of medical records documenting a **baseline motor exam by a physician (Neurologist or PMR (Physical Medicine & Rehabilitation Specialist)) or physical therapist specializing in SMA motor exam evaluations and supervised by Neurologist or PMR physician)** experienced in treating SMA and utilizing at least one of the following exam instruments (based on patient's age), to establish this baseline motor ability**:**

o Hammersmith Infant Neurological Exam (HINE)

o Hammersmith Functional Motor Scale Expanded (HFMSE)

o Upper Limb Module Test (non-ambulatory)

o Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND); AND

4) **Nusinersen** must be **ordered by a Neurologist** experienced in treating SMA.
Initial approval of Nusinersen will be for 5 doses ONLY to be given in accordance with the current Nusinersen FDA label instructions**.** Protocol: loading doses at day 0 (dose 1), day 14 (dose 2), day 28 (dose 3), 30 days post day 28 (dose 4) and 4 months after that fourth dose (dose 5).

Prior Authorization Required From CHN

Guidelines for subsequent doses of Nusinersen:

After the 5 loading doses are completed, patient must be **re-evaluated using the same motor exam test done to establish baseline motor ability** unless it is determined that the original exam instrument is no longer age- appropriate. This re-examination must be done, whenever possible, by the same examiner as baseline exam. If this is not possible, then the re-examination must be done by another physician (Neurologist or PMR who is experienced in treating SMA or a physical therapist specializing in SMA motor exam evaluations and supervised by Neurologist or PMR physician).

• Nusinersen will be authorized for an additional 6 months if a patient is determined to be a responder by
demonstrating an **improved motor ability** in repeat motor testing after the 5th loading dose.

To be classified as a responder, the patient should receive the following score(s) on the motor

test used: HINE: a 2 point increase (or max score of 4) in ability to kick (consistent with

improvement by at
least 2 milestones) or a 1 point increase in motor milestones of head control, rolling, sitting, crawling, standing or walking (consistent with improvement by at least 1 milestone). Responder needs to exhibit improvement in more categories of motor milestones than worsening.

HFMSE: (Scored as 0, 1 or 2 with maximum of 40). Improvement of at least a 3 point increase in
score from pretreatment baseline

ULM: Improvement of at least 2 point increase in score from pretreatment baseline

CHOP INTEND: Improvement of at least 4 point increase in score from pretreatment baseline

• **Renewal authorization,** will be for 6 months. Nusinersen administration will follow the current FDA Nusinersen labelling for maintenance dosing protocol of every 4 months

• **Repeat motor testing** must be done at 6 month intervals and must show additional motor improvement or maintenance of the previously demonstrated motor improvement. If a patient becomes dependent on mechanical ventilator (defined as requiring mechanical ventilation for > 21 days) while on Nusinersen, then the Department will no longer authorize payment of Nusinersen for that patient.

The 6 month periodic re-examination must be done by the same examiner as the baseline exam whenever possible. If this is not possible, it must be conducted by another physician (Neurologist or PMR) who is experienced in treating SMA who must use the same exam instrument unless it is determined that original exam instrument is no longer age appropriate.

II. ***Other SMA Types*** **- Patients with other SMA Types are eligible for treatment if the following conditions are met:**

1) A diagnosis of SMA must be made by a Neurologist with expertise in diagnosing Spinal Muscular Atrophy; AND

2) All of the documentation listed above will be required; AND

3) **In addition,** documentation must be provided demonstrating in the opinion of the treating

neurologist, why a patient should be considered for administration of Nusinersen given the research demonstrating efficacy is limited and equivocal.

If approved for a patient with a non-Type 1 SMA diagnosis, the dosage and renewal guidelines described above will also apply.

- Pharmacy Prior Authorization Submission Process: PA requests for coverage of Nusinersen must be submitted by the prescriber in the form of a letter of medical necessity to DSS' Medical Director. Letters of medical necessity must be faxed to (860) 424-4822 with the required documentation outlined in the Nusinersen Coverage Guidelines. Outpatient Hospital

- Prior Authorization Submission Process: Effective November 1, 2017 and forward, outpatient hospitals buying and billing for Spinraza must utilize a separate PA process identified below. Outpatient hospitals must fax a completed Spinraza PA Request Form along with the required documentation to Community Health Network of CT, Inc. (CHNCT) at (203) 265-3994.

Prior Authorization Required From CHN

Palivizumab (Synagis®) Guidelines

Certain pediatric patients should be strongly considered as candidates for palivizumab (Synagis®) for respiratory syncytial virus (RSV) infection prophylaxis. Recently published (August 2014) American Academy of Pediatrics recommendations[1] for the use of palivizumab offers the following guidelines to identify those infants who may particularly benefit from monthly doses of palivizumab throughout the duration of the RSV season, which in Connecticut is typically November 1 through March 31.

Note: The following criteria are guidelines *only*. Determinations of coverage will be made in accordance with the Department of Social Services (DSS) Definition of Medical Necessity and are based on an individual assessment of the member and his or her clinical needs.

First Year of Life:

1. Infants born **before 29 weeks, 0 days'** gestation.
2. Preterm infants born **before 32 weeks, 0 days'** gestation with chronic lung disease of prematurity defined as greater than 21% oxygen for at least 28 days after birth.
3. Certain infants with hemodynamically significant heart disease. (Note: consultation with a cardiologist for decisions about prophylaxis is recommended for patients with cyanotic heart disease).

Second Year of Life:

Palivizumab prophylaxis is **not recommended in the second year of life** except for preterm infants born before 32 weeks, 0 days' gestation who required at least 28 days of supplemental oxygen after birth and who continue to require medical intervention (supplemental oxygen, chronic corticosteroid or diuretic therapy).

Other Possible Candidates:

1. Children **younger than 24 months of age** who will be profoundly immunocompromised during the RSV season may be considered for prophylaxis.
2. Children with pulmonary abnormality or neuromuscular disease that impairs the ability to clear secretions from the upper airways may be considered for prophylaxis in the **first year of life.**

NOTE: Monthly prophylaxis should be discontinued in any child who experiences a breakthrough RSV hospitalization. As noted in the current AAP recommendations, "**Clinicians may administer up to a maximum of five monthly doses of Palivizumab during the RSV season to infants who qualify for prophylaxis in the first year of life (including those in Florida). Qualifying infants born during the RSV season will require fewer doses.** For example, infants born in January would receive their last dose in March." [1]

Outpatient hospitals must fax a completed PA request form along with supporting clinical information to Community Health Network of Connecticut, Inc. (CHNCT) at 203-774-0549.

For questions regarding the prior authorization process, please contact CHNCT at 1-800-440-5071, Monday through Friday, between the hours of 8:00 a.m. and 6:00 p.m.

Updated 4/30/2020

Prior Authorization Required From CHN

Voretigene Neparvovec-rzyl (Luxturna®) Guidelines

Luxturna® Guidelines

Updated 4/30/2020