Medical Policy Bulletin

Title:
Nusinersen (Spinraza®)
Policy #:
MA08.086d

This Policy Bulletin document describes the status of CMS coverage, medical terminology, and/or benefit plan documents and contracts at the time the document was developed. This Policy Bulletin will be reviewed regularly and be updated as Medicare changes their regulations and guidance, scientific and medical literature becomes available, and/or the benefit plan documents and/or contracts are changed.

Policy

Coverage is subject to the terms, conditions, and limitations of the member's Evidence of Coverage.

When services can be administered in various settings, the Company reserves the right to reimburse only those services that are furnished in the most appropriate and cost-effective setting that is appropriate to the member's medical needs and condition. This decision is based on the member's current medical condition and any required monitoring or additional services that may coincide with the delivery of this service.

MEDICALLY NECESSARY

INITIAL THERAPY
Nusinersen (Spinraza®) is considered medically necessary and, therefore, covered when all of the following criteria, including Dosing and Frequency, are met:

- Diagnosis of spinal muscular atrophy type 1, 2, or 3 or pre-symptomatic types 1 or 2, confirmed by documentation of genetic testing for 5q-SMA by one of the following:
  - homozygous deletion of SMN1 gene (e.g., homozygous deletion of exon 7 on chromosome 5)
  - presence of a single copy of SMN1 gene with coding confirming a mutation to mimic a homozygous deletion of SMN1 gene (e.g., deletion of SMN1 exon 7 on allele 1 and mutation of SMN1 on allele 2)
- Prescribed by a neurologist or a physiatrist with subspecialty certification in neuromuscular medicine
- Dosing and Frequency: Dose does not exceed 12 mg every 4 months after the loading doses. NOTE: The first three loading doses are 12 mg administered intrathecally at 14-day intervals. The fourth loading dose is administered 30 days after the third dose. A 12 mg maintenance dose is administered once every 4 months thereafter.

CONTINUATION THERAPY
Continuation of nusinersen (Spinraza®) is considered medically necessary and, therefore, covered when all of the following criteria are met:

- All criteria under 'Initial Therapy' as stated above are met. NOTE: Initial Therapy is defined as completion of the four loading doses followed by 3 maintenance doses administered 4 months apart.
- There is documented improvement in motor function, stability in motor function or a slower rate of decline in motor function due to nusinersen (Spinraza®) treatment based on a standard measurement scale in SMA

associated symptoms (e.g., Hammersmith Infant Neurological Examination (HINE), Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND) and/or the Hammersmith Functional Motor Scale-Expanded (HFMSE) scores). It is expected that the same objective measurement scale will be used for both baseline and response to treatment.

NOT MEDICALLY NECESSARY

When molecular genetic testing reveals established benign variation(s) or wild-type genotype for 5q-SMA, nusinersen (Spinraza®) is considered not medically necessary and, therefore, not covered because the available published peer-reviewed literature does not support its use in the treatment of this disease.

EXPERIMENTAL/INVESTIGATIONAL

For all other uses, including SMA types 0 and 4, nusinersen (Spinraza®) is considered experimental/investigational and, therefore, not covered because its safety and/or effectiveness cannot be established by review of the available published peer-reviewed literature.

When molecular genetic testing reveals likely pathogenic or variations of unknown significance (VUS) for 5q-SMA, the use of nusinersen (Spinraza®) is considered experimental/investigational and, therefore, not covered because the safety and/or effectiveness of this service cannot be established by review of the available published peer-reviewed literature.

DOSING AND FREQUENCY REQUIREMENTS

The Company reserves the right to modify the Dosing and Frequency Requirements listed in this policy to ensure consistency with the most recently published recommendations for the use of nusinersen (Spinraza®). Changes to these guidelines are based on a consensus of information obtained from resources such as, but not limited to: the US Food and Drug Administration (FDA); Company-recognized authoritative pharmacology compendia; or published peer-reviewed clinical research. The professional provider must supply supporting documentation (i.e., published peer-reviewed literature) in order to request coverage for an amount of nusinersen (Spinraza®) outside of the Dosing and Frequency Requirements listed in this policy. For a list of Company-recognized pharmacology compendia, view our policy on off-label coverage for prescription drugs and biologics.

Accurate member information is necessary for the Company to approve the requested dose and frequency of this drug. If the member’s dose, frequency, or regimen changes (based on factors such as changes in member weight or incomplete therapeutic response), the provider must submit those changes to the Company for a new approval based on those changes as part of the utilization management activities. The Company reserves the right to conduct post-payment review and audit procedures for any claims submitted for nusinersen (Spinraza®).

REQUIRED DOCUMENTATION

The individual's medical record must reflect the medical necessity for the care provided. These medical records may include, but are not limited to: records from the professional provider's office, hospital, nursing home, home health agencies, therapies, and test reports.

The Company may conduct reviews and audits of services to our members, regardless of the participation status of the provider. All documentation is to be available to the Company upon request. Failure to produce the requested information may result in a denial for the drug.

When coverage of nusinersen (Spinraza®) is requested outside of the Dosing and Frequency Requirements listed in this policy, the prescribing professional provider must supply documentation (i.e., published peer-reviewed literature) to the Company that supports this request.

Guidelines

BENEFIT APPLICATION

Subject to the terms and conditions of the applicable Evidence of Coverage, nusinersen (Spinraza™) is covered under the medical benefits of the Company’s Medicare Advantage products when the medical necessity criteria and Dosing and Frequency Requirements listed in this medical policy are met.

NEUROLOGICAL TESTS TO EVALUATE MOTOR SKILLS

HAMMERSMITH INFANT NEUROLOGICAL EXAMINATION (HINE)
The Hammersmith Infant Neurological Examination (HINE) test is a motor function assessment tool designed to evaluate motor skills in infants ages 2 months to 2 years old. This exam includes 26 items that provide a comprehensive assessment of an infant's neuromuscular development. The motor milestone portion of the exam includes 8 items: voluntary grasp, ability to kick, head control, rolling, sitting, crawling, standing, and walking. Each item is scored from 0-3, with a possible 78 total points.

CHILDREN'S HOSPITAL OF PHILADELPHIA INFANT TEST OF NEUROMUSCULAR DISORDERS (CHOP INTEND) The Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND) test was developed to evaluate the motor skills of patients with SMA type 1. There are 16 items (e.g., spontaneous movement, hand grip, knee extension, head control, spinal incurvation, etc.) that are used to assess motor skills. Each of these items is graded on a scale of 0 (no response) to 4 (complete response), with a total score ranging from 0 to 64.

HAMMERSMITH FUNCTIONAL MOTOR SCALE-EXPANDED (HFMSE)
The Hammersmith Functional Motor Scale- Expanded is a scale used to evaluate motor function in individuals with later onset (types 2 and 3) SMA who have limited ambulation. The exam consists of 33 items, 13 of which come from the Gross Motor Function measure, that are scored from 0-2, for a total score range from 0-66. Higher scores indicate better motor function. Some of the evaluated items include chair sitting, supine to side lying, sitting to lying, lifting head from prone position, crawling, standing, and stepping.

US FOOD AND DRUG ADMINISTRATION (FDA) STATUS

Nusinersen (Spinraza™) was approved by the FDA on December 23, 2016 for treatment of spinal muscular atrophy (SMA) in pediatric and adult patients.

Description

Spinal muscular atrophy (SMA) is an autosomal recessive genetic disorder characterized by the loss of motor neurons in the spinal cord and lower brain stem. It is caused by a mutation in, or the absence of, survival motor neuron gene 1 (SMN1) on chromosome 5, which causes a decrease in the amount of SMN protein produced. SMN protein is critical to the function of the nerves that control muscles. Another gene, survival motor neuron gene 2 (SMN2), also produces SMN protein. Due to a splicing error, most of the SMN protein produced by SMN2 lacks exon 7, which is necessary for the production of fully functional SMN protein, thereby rendering the protein produced unusable. The number of copies of SMN2 varies from person to person, and is inversely proportional to the severity of the disease. Individuals with SMA have at least one copy of the SMN2 gene.

SMA has been estimated to occur in 1 in 10,000 live births in the United States. SMA that is genetically diagnosed prior to the onset of symptoms by the absence of the SMN1 gene is considered to be presymptomatic. There are four primary types of 5q-SMA, which are characterized by severity and age at which symptoms begin:

- Type 0 is prenatal onset SMA. At birth, individuals with SMA type 0 have severe weakness and hypotonia. These individuals never achieve any motor milestones and death occurs by 6 months, but usually by 1 month, due to respiratory failure.
- Type 1, also called infantile-onset or Werdnig-Hoffman disease, is the most severe and is the number one genetic cause of death for infants. These individuals are classified as "nonsitters." Symptoms, including progressive proximal weakness, poor head control, progressive respiratory insufficiency, hypotonia, areflexia, and tongue fasciculations, present within the first 6 months of life. The life span for these children is usually less than 2 years due to respiratory failure.
- Type 2 is an intermediate form of SMA that usually presents between 6 and 24 months of age. These individuals are classified as "sitters." In addition to the symptoms displayed in type 1, type 2 individuals also exhibit progressive scoliosis and joint contractures. The life span of these individuals varies greatly, with many individuals living into their second decade with adequate supportive care.
- Type 3, also known as Kugelberg-Welander disease, usually has symptom onset after 18 months. Symptoms seen in this population include progressive proximal weakness, causing "walkers" to eventually need a wheelchair, and hand tremors. This is a more mild form of SMA, and individuals can have a lifespan comparable to those without SMA.

- Type 4 is rare and usually presents in adulthood. Symptoms of weakness and mild motor impairment can begin as early as 18 but usually begin after 35, leading to mild motor impairment. These individuals generally have a normal life span.

Other forms of SMA exist that do not involve mutations in the SMN1 gene. These types are referred to as non-5q-SMA because the genes that cause them are not located in the SMN region of chromosome 5. These types of SMA include spinal and bulbar muscular atrophy, SMA distal type V, and SMA proximal adult.

NUSINERSEN (SPINRAZA®)

Nusinersen (Spinraza®) was approved on 12/23/16 as the only FDA-approved treatment of spinal muscular atrophy (SMA) in pediatric and adult patients. It is an antisense oligonucleotide designed to treat SMA caused by mutations in the SMN gene located on chromosome 5q. Nusinersen targets the SMN2 gene, increasing production of a complete SMN protein by enhancing exon 7 inclusion in mRNA. Antisense drugs contain synthetic genetic material that bind to RNA, and can be used to fix splicing errors in genes such as SMN2. It is dosed intrathecally in a series of 4 loading doses within the first 60 days (the first three loading doses administered at 14-day intervals, and the fourth loading dose administered 30 days after the third dose), and then maintenance doses every 4 months thereafter.

PEER-REVIEWED LITERATURE
NOTE: Nusinersen is continuing to be evaluated in several other ongoing trials.
Summary

Type 1
Efficacy and safety of nusinersen was studied in a phase III multicenter, randomized, double blind, sham procedure-controlled study (ENDEAR by Finkle et al. 2017). Beginning in August 2014, ENDEAR studied 121 individuals with infant onset (Type 1) SMA. Forty percent of the 82 eligible individuals in the nusinersen group reached the primary endpoint at the time of interim analysis of achieving a motor milestone response. This response was evaluated according to the Hammersmith Infant Neurologic Exam (HINE). In the final analysis, 51% of individuals in the nusinersen group achieved a motor milestone response (HINE), compared to 0% of individuals in the sham-controlled group. Individuals who received nusinersen showed improvement from baseline in motor skills based on the Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND), although the study was not statistically controlled for multiple comparisons at the study interim. Although in the final analysis, 71% of individuals in the nusinersen group achieved an improvement in motor skills (CHOP INTEND), compared to 3% of individuals in the sham-controlled group.

The long-term safety and tolerability of nusinersen over a five-year period is being studied in the SHINE trial by Castro et al. 2018, (NCT02594124) which is a phase III, open-label, extension study in 292 individuals who previously participated in investigational studies of nusinersen (NUTURE, ENDEAR, CHERISH, AND CS12 trials). A secondary outcome examines the long-term efficacy of nusinersen in these individuals. Interim data from October 2018 (Finkle et al 2019) was reported for those previously in ENDEAR trial. There were 65 participants who had received nusinersen in ENDEAR trial and 24 participants who received sham control in ENDEAR trial. In SHINE, all participants received nusinersen (N=89). Interim results concluded that 99% of participants had an adverse reaction (most frequently pyrexia and upper respiratory tract infection) but no participants had a treatment-related serious adverse reaction. CHOP INTEND score and WHO motor milestones have been maintained or continued to improve with treatment.

Types 2-3
The safety and efficacy of nusinersen (Spinraza®) in individuals who developed symptoms of SMA after 6 months of age (i.e., types 2-3) was evaluated in the CHERISH trial by Mercuri et al 2018. which was a multicenter, double blind, sham controlled phase 3 study. The 126 individuals in the study received either an intrathecal dose of nusinersen (Spinraza®) 12 mg or a sham procedure on days 1, 29, 85, and 274. The primary endpoint was the least squares mean change from baseline in the Hammersmith Functional Motor Scale- Expanded (HFMSE) score at 15 months. In the final analysis there was a least squares mean increase in the nusinersen group and a least squares mean decrease in the sham control group. These results favored the nusinersen group (least squares mean difference in change was 4.9 points), showing significant improvement in motor function.

CS2/CS12 are two open-label, Phase 1/2a dose-escalation longitudinal analysis across both studies (N=28) at 35 months in types 2 and 3 SMA. The results showed an increase and maintenance of motor function by HFMSE scores of 12.3 point increase from baseline in Type 2 individuals and 1.6 point increase from baseline in Type 3 individuals.

The long-term safety and tolerability of nusinersen over a five-year period is being studied in the SHINE trial by Castro et al. 2018, (NCT02594124) which is a phase III, open-label, extension study in 292 individuals who previously

participated in investigational studies of nusinersen (NUTURE, ENDEAR, CHERISH, AND CS12 trials). A secondary outcome examines the long-term efficacy of nusinersen in these individuals. Interim data from October 2018 (Darras et al 2019) was reported for those previously in CHERISH trial. There were 83 participants who had received nusinersen in CHERISH trial and 42 participants who received sham control in CHERISH trial. In SHINE, all participants received nusinersen (N=89). Interim results concluded that 91% of participants had an adverse reaction (most frequently pyrexia and upper respiratory tract infection) but no participants had a treatment-related serious adverse reaction. HFMSE scores have been maintained or continued to improve with treatment.

Types 1, 2, 3
Safety and long-term efficacy is being studied in the EMBRACE trial, which is a phase II double-blind study in individuals with infant or later-onset SMA who didn't qualify for the ENDEAR or CHERISH trials. Participants (N=21) were randomized 2:1 to receive nusinersen (Spinraza®) or placebo. The results of this study are not yet available (NCT02462759).

Presymptomatic, likely to develop Types 1 or 2

Efficacy in presymptomatic individuals, those with a genetic deletion or variant causing a likelihood to develop Types 1 or 2, was studied in a phase II, open-label, uncontrolled trial of 25 individuals, called NURTURE. Individuals were ages 3 days to 42 days at the time of the first dose. Nusinersen (Spinraza®) was administered during 4 loading doses, followed by maintenance doses every 4 months. The primary end point was time to death or respiratory intervention (invasive or noninvasive ventilation for >6 hours/day continuously over for ≥7 days or tracheostomy). At 365 days of the study visit, the mean CHOP INTEND total score was estimated as 56 and 64 in individuals with 2 and 3 copies of *SMN2*, respectively. Based on a natural history cohort of children with SMA with 2 *SMN2* copies, the highest individual CHOP INTEND score was 33 and the mean score was 19.9 over a 2-year period. At the data cutoff (July 5, 2017), all infants were alive and none required tracheostomy or permanent ventilation. Two (13%) of 15 infants with 2 *SMN2* copies required respiratory intervention for 6 or more hours per day continuously for 7 or more days during an acute, reversible viral infection, and thus met the primary end point. At the time of interim analysis (May 2018), all infants were alive and none required tracheostomy or permanent ventilation. Four (16%) of 25 infants with 2 *SMN2* copies required respiratory intervention for 6 or more hours per day continuously for 7 or more days during an acute, reversible illness. At 540 days of the study visit, the mean CHOP INTEND total score was estimated as 61 and 62.6 in individuals with 2 and 3 copies of *SMN2*, respectively.

Future trial with higher doses

DEVOTE trial (NCT04089566) will examine the clinical efficacy of intrathecal nusinersen (Spinraza®) at higher doses to participants with SMA. Secondary outcomes will examine the safety and tolerability of nusinersen, the effect of nusinersen (Spinraza®) administered at higher doses compared to the currently approved dose in participants with SMA, and the pharmacokinetic(s) (PK) of nusinersen (Spinraza®) [cerebrospinal fluid (CSF) and plasma] after intrathecal administration of nusinersen (Spinraza®) given at higher doses to participants with SMA. The four arms of the trial are:

1. Experimental: 28/28 mg Safety Group
Part A: Participants with later-onset SMA will receive loading doses of 28 mg of nusinersen (Spinraza®) on Days 1, 15 and 29 followed by maintenance doses of 28 mg on Days 149 and 269.

2. Active Comparator: 12/12 mg Randomized Control Group
Part B: Participants with infantile- or later-onset SMA will receive loading doses of 12 mg of nusinersen (Spinraza®) on Days 1, 15, 29, and 64 followed by maintenance doses of 12 mg on Days 183 and 279. Sham procedure will be administered on Day 135.

3. Experimental: 50/28 mg Randomized Treatment Group
Part B: Participants with infantile- or later-onset SMA will receive loading doses of 50 mg of nusinersen (Spinraza®) on Days 1 and 15 followed by maintenance doses of 28 mg on Days 135 and 279. Sham procedure will be administered on Days 29, 64 and 183.

4. Experimental: 12/50/28 mg Titration Group
Part C: Participants who have been receiving the approved dose of 12 mg for at least 1 year prior to entry in this study, will receive a single bolus dose of 50 mg of nusinersen (Spinraza®) on Day 1 (4 months after their most recent maintenance dose of 12 mg) followed by maintenance doses of 28 mg on Days 121 and 241.

Types 0, 4

Studies evaluating the use of nusinersen in types 0 or 4 SMA have not been identified.

OFF-LABEL INDICATIONS

There may be additional indications contained in the Policy section of this document due to evaluation of criteria highlighted in the Company's off-label policy, and/or review of clinical guidelines issued by leading professional organizations and government entities.

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Coding

Inclusion of a code in this table does not imply reimbursement. Eligibility, benefits, limitations, exclusions, precertification/referral requirements, provider contracts, and Company policies apply.

The codes listed below are updated on a regular basis, in accordance with nationally accepted coding guidelines. Therefore, this policy applies to any and all future applicable coding changes, revisions, or updates.

In order to ensure optimal reimbursement, all health care services, devices, and pharmaceuticals should be reported using the billing codes and modifiers that most accurately represent the services rendered, unless otherwise directed by the Company.

The Coding Table lists any CPT, ICD-10, and HCPCS billing codes related only to the specific policy in which they appear.

CPT Procedure Code Number(s)

N/A

ICD - 10 Procedure Code Number(s)

N/A

ICD - 10 Diagnosis Code Number(s)

G12.0 Infantile spinal muscular atrophy, type I [Werdnig-Hoffman]

G12.1 Other inherited spinal muscular atrophy

HCPCS Level II Code Number(s)

J2326 Injection, nusinersen, 0.1 mg

Revenue Code Number(s)

N/A

Policy History

Revisions From MA08.086d:

11/17/2021	This policy has been reissued in accordance with the Company's annual review process.

11/06/2019	The policy has been reviewed and reissued to communicate the Company's continuing position on nusinersen (Spinraza®).
12/17/2018	This policy has been updated to clarify: • Genetic testing associated with SMA • Coverage of pre-symptomatic types 1 or 2 • Provider types • Coverage of drug through Dosage and Frequency of Administration • Parameters for continuation of therapy • Clinical Study information

Revisions for MA08.086c

04/23/2018	This policy has undergone a routine review and the medical necessity criteria have been revised to reflect the United States Food and Drug Administration (FDA) labeling and published literature.

Revisions for MA08.086b

01/01/2018	This policy has been identified for the HCPCS code update, effective 01/01/2018. The following NOC code has been **removed** from this policy and is replaced by the following HCPCS code: **REMOVED:** J3490 Unclassified drugs **REPLACED WITH:** J2326 Injection, nusinersen, 0.1 mg

	The following HCPCS code has been **removed** from this policy: C9489 Injection, nusinersen, 0.1 mg

Revisions for MA08.086a

07/01/2017	This policy has been identified for the HCPCS code update, effective 07/01/2017. The following HCPCS code has been **added** to this policy: C9489 Injection, nusinersen, 0.1 mg

Revisions for MA08.086

04/19/2017	This new policy has been developed to communicate the Company's coverage criteria for nusinersen (Spinraza™).

Version Effective Date:
12/17/2018
Version Issued Date:
12/17/2018
Version Reissued Date:
11/17/2021