PRIOR AUTHORIZATION POLICY

POLICY: Spinal Muscular Atrophy – Evrysdi Prior Authorization Policy
- Evrysdi® (risdiplam oral solution and tablets – Genentech/Roche)

REVIEW DATE: 07/22/2026

INSTRUCTIONS FOR USE

THE FOLLOWING COVERAGE POLICY APPLIES TO HEALTH BENEFIT PLANS ADMINISTERED BY CIGNA COMPANIES. CERTAIN CIGNA COMPANIES AND/OR LINES OF BUSINESS ONLY PROVIDE UTILIZATION REVIEW SERVICES TO CLIENTS AND DO NOT MAKE COVERAGE DETERMINATIONS. REFERENCES TO STANDARD BENEFIT PLAN LANGUAGE AND COVERAGE DETERMINATIONS DO NOT APPLY TO THOSE CLIENTS. COVERAGE POLICIES ARE INTENDED TO PROVIDE GUIDANCE IN INTERPRETING CERTAIN STANDARD BENEFIT PLANS ADMINISTERED BY CIGNA COMPANIES. PLEASE NOTE, THE TERMS OF A CUSTOMER'S PARTICULAR BENEFIT PLAN DOCUMENT [GROUP SERVICE AGREEMENT, EVIDENCE OF COVERAGE, CERTIFICATE OF COVERAGE, SUMMARY PLAN DESCRIPTION (SPD) OR SIMILAR PLAN DOCUMENT] MAY DIFFER SIGNIFICANTLY FROM THE STANDARD BENEFIT PLANS UPON WHICH THESE COVERAGE POLICIES ARE BASED. FOR EXAMPLE, A CUSTOMER'S BENEFIT PLAN DOCUMENT MAY CONTAIN A SPECIFIC EXCLUSION RELATED TO A TOPIC ADDRESSED IN A COVERAGE POLICY. IN THE EVENT OF A CONFLICT, A CUSTOMER'S BENEFIT PLAN DOCUMENT ALWAYS SUPERSEDES THE INFORMATION IN THE COVERAGE POLICIES. IN THE ABSENCE OF A CONTROLLING FEDERAL OR STATE COVERAGE MANDATE, BENEFITS ARE ULTIMATELY DETERMINED BY THE TERMS OF THE APPLICABLE BENEFIT PLAN DOCUMENT. COVERAGE DETERMINATIONS IN EACH SPECIFIC INSTANCE REQUIRE CONSIDERATION OF 1) THE TERMS OF THE APPLICABLE BENEFIT PLAN DOCUMENT IN EFFECT ON THE DATE OF SERVICE; 2) ANY APPLICABLE LAWS/REGULATIONS; 3) ANY RELEVANT COLLATERAL SOURCE MATERIALS INCLUDING COVERAGE POLICIES AND; 4) THE SPECIFIC FACTS OF THE PARTICULAR SITUATION. EACH COVERAGE REQUEST SHOULD BE REVIEWED ON ITS OWN MERITS. MEDICAL DIRECTORS ARE EXPECTED TO EXERCISE CLINICAL JUDGMENT WHERE APPROPRIATE AND HAVE DISCRETION IN MAKING INDIVIDUAL COVERAGE DETERMINATIONS. WHERE COVERAGE FOR CARE OR SERVICES DOES NOT DEPEND ON SPECIFIC CIRCUMSTANCES, REIMBURSEMENT WILL ONLY BE PROVIDED IF A REQUESTED SERVICE(S) IS SUBMITTED IN ACCORDANCE WITH THE RELEVANT CRITERIA OUTLINED IN THE APPLICABLE COVERAGE POLICY, INCLUDING COVERED DIAGNOSIS AND/OR PROCEDURE CODE(S). REIMBURSEMENT IS NOT ALLOWED FOR SERVICES WHEN BILLED FOR CONDITIONS OR DIAGNOSES THAT ARE NOT COVERED UNDER THIS COVERAGE POLICY (SEE "CODING INFORMATION" BELOW). WHEN BILLING, PROVIDERS MUST USE THE MOST APPROPRIATE CODES AS OF THE EFFECTIVE DATE OF THE SUBMISSION. CLAIMS SUBMITTED FOR SERVICES THAT ARE NOT ACCOMPANIED BY COVERED CODE(S) UNDER THE APPLICABLE COVERAGE POLICY WILL BE DENIED AS NOT COVERED. COVERAGE POLICIES RELATE EXCLUSIVELY TO THE ADMINISTRATION OF HEALTH BENEFIT PLANS. COVERAGE POLICIES ARE NOT RECOMMENDATIONS FOR TREATMENT AND SHOULD NEVER BE USED AS TREATMENT GUIDELINES. IN CERTAIN MARKETS, DELEGATED VENDOR GUIDELINES MAY BE USED TO SUPPORT MEDICAL NECESSITY AND OTHER COVERAGE DETERMINATIONS.

CIGNA NATIONAL FORMULARY COVERAGE:

OVERVIEW

Evrysdi, a survival motor neuron 2 (SMN2) splicing modifier, is indicated for the **treatment of spinal muscular atrophy** in pediatric patients and adults.[1]

Disease Overview

Spinal muscular atrophy is a genetic, autosomal recessive, progressive, neuromuscular disorder caused by biallelic deletions or pathogenetic variants in the survival motor neuron 1 (SMN1) gene which can be identified by genetic testing.[2-5] The subsequent reduced levels of survival motor neuron (SMN) protein impacts brainstem and spinal cord motor neurons. The estimated incidence in the US is one in 11,000.[3] The phenotypic expression of the disease is impacted by the SMN2 gene copy number.[3-5] Patients with a higher number of SMN2 gene copies often have less severe disease with milder progression. Table 1 describes disease types. Of note, various motor ability assessments and different functional motor scales are used in clinical practice to characterize impairment in patients with spinal muscular atrophy.

When motor neuron function is lost, it cannot be regained, which greatly impacts patients who have experienced progression (e.g., patients with complete paralysis of limbs or permanent ventilator dependence). A different manner of patient categorization classifies the three most common types as "non-sitters," "sitters," and "walkers."

Table 1. Types of Spinal Muscular Atrophy.[4,5]

	Age at Onset	Features/Clinical Presentation/Motor Milestones*	Lifespan*	SMN2 Gene Copy Number
Type 0 (< 1% of patients)	Birth	Severe hypotonia and weakness with respiratory failure at birth. There is no achievement of motor milestones.	A few weeks to days (< 6 months)	1
Type 1 (50% of patients)	< 6 months	Poor muscle tone and lack of movement. Respiratory assistance may be needed. Patients are never able to sit without support.	< 2 years	1 to 2 for 80% of patients
Type 2 (30% of patients)	6 to 18 months	Patients are able to sit. However, patients are unable to walk or stand without assistance.	Close to normal	2 to 3 for over 90% of patients
Type 3 (10% to 20% of patients)	> 18 months	Walks independently but may lose this ability as the disease progresses. There is loss of motor skills.	Normal	3 to 5 for most patients
Type 4 (< 1% of patients)	> 18 years	Independent walking. Fatigue and proximal muscle weakness.	Normal	4 for 75% of patients; 5 or 6 for 25% of patients

* With supportive care only; SMN2 – Survival motor neuron 2.

Clinical Efficacy

The efficacy of Evrysdi for the treatment of patients with infantile-onset (Type 1), later-onset (Type 2 and 3), and pre-symptomatic spinal muscular atrophy was evaluated in three clinical studies.[1,6-8] **FIREFISH** involved patients with Type 1 spinal muscular atrophy who had symptom onset between 28 days and 3 months of age.[1,6,7] Genetic confirmation of homozygous deletion or compound heterozygosity predictive of loss of function of the SMN1 gene was required for trial entry.[1] Patients had two SMN2 gene copies. Many patients gained improvements in the ability to sit for at least 5 seconds independently, and there was an increase in the percentages of patients who were alive without permanent ventilation. **SUNFISH** evaluated Evrysdi in patients with later-onset (Type 2 or Type 3) spinal muscular atrophy. Most patients (90%) had three SMN2 gene copie.[1,8] In Part 2 of the study, benefits of Evrysdi vs. placebo were noted at Month 12 in motor function as well as in upper limb motor performance.[1] **RAINBOWFISH** investigated Evrysdi in infants up to 6 weeks of age (at the first dose) who had been genetically diagnosed with spinal muscular atrophy but did not have symptoms. Eight patients had two SMN2 gene copies, 13 patients had three SMN2 gene copies, and five patients had four or more SMN2 gene copies. The median age at first dose was 25 days. The primary efficacy endpoint was the proportion of patients with the ability to sit without support for at least 5 seconds at Month 12, which was achieved by 87.5% of patients with two SMN2 copies (n = 7/8)

and 96.2% of patients (n = 25/26) in the full treated population. All 26 patients were alive at 12 months without permanent ventilation.

Guidelines

Evrysdi is not addressed in guidelines. According to a treatment algorithm from the Spinal Muscular Atrophy Newborn Screening Multidisciplinary Working Group (2018), immediate treatment is recommended in patients with two or three SMN2 gene copies.[9] In 2020, the Working Group updated recommendations that infants diagnosed with spinal muscular atrophy via newborn screening with four SMN2 gene copies should receive immediate treatment.[10] Patients with five (or more) SMN2 gene copies should be observed and screened for symptoms.

POLICY STATEMENT

Prior Authorization is recommended for prescription benefit coverage of Evrysdi. All approvals are provided for the duration noted below. In cases where the approval is authorized in months, 1 month is equal to 30 days. Because of the specialized skills required for evaluation and diagnosis of patients treated with Evrysdi as well as the monitoring required for adverse events and long-term efficacy, approval requires Evrysdi to be prescribed by a physician who has consulted with or who specializes in the condition. If claims history if available, verification is required for certain criteria as noted by **[verification in claims history required]**. All reviews will be forwarded to the Medical Director for evaluation.

Documentation: Documentation is required where noted in the criteria as **[documentation required]**. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information.

- **Evrysdi® (risdiplam oral solution and tablets – Genentech/Roche) is(are) covered as medically necessary when the following criteria is(are) met for FDA-approved indication(s) or other uses with supportive evidence (if applicable):**

FDA-Approved Indication

1. **Spinal Muscular Atrophy – Treatment.** Approve if the patient meets ONE of the following (A or B):
   - **A)** Initial Therapy. Approve for 4 months if the patient meets ALL of the following (i, ii, iii, iv, and v):
     - **i.** Baseline motor ability assessment that suggests spinal muscular atrophy (based on age, motor ability, and development) has been performed from ONE of the following exams (a, b, c, d, e, f, or g) **[documentation required]**:
       - **a)** Bayley Scales of Infant and Toddler Development; OR
       - **b)** Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND); OR

c) Hammersmith Functional Motor Scale Expanded (HFMSE); OR
d) Hammersmith Infant Neurological Exam Part 2 (HINE-2); OR
e) Motor Function Measure-32 Items (MFM-32); OR
f) Revised Upper Limb Module (RULM) test; OR
g) World Health Organization motor milestone scale; AND

ii. Patient has had a genetic test confirming the diagnosis of spinal muscular atrophy with bi-allelic pathogenic variants in the survival motor neuron 1 (SMN1) gene **[documentation required]**; AND
Note: Pathogenic variants may include homozygous deletion, compound heterozygous mutation, or a variety of other rare mutations.

iii. Patient meets ONE of the following (a or b):
a) Patient has two or three survival motor neuron 2 (SMN2) gene copies **[documentation required]**; OR
b) Patient meets BOTH of the following ([1] and [2]):
(1) Patient has four survival motor neuron 2 (SMN2) gene copies **[documentation required]**; AND
(2) Patient has objective signs consistent with spinal muscular atrophy Types 1, 2, or 3 **[documentation required]**; AND

iv. Patient has not received Zolgensma (onasemnogene abeparvovec-xioi intravenous infusion) or Itvisma (onasemnogene abeparvovec-brve intrathecal injection) in the past **[verification in claims history required]**; AND
Note: If no claim for Zolgensma or Itvisma is present (or if claims history is not available), the prescribing physician confirms that the patient has not previously received Zolgensma or Itvisma.

v. The medication is prescribed by a physician who has consulted with a specialist or who specializes in the management of patients with spinal muscular atrophy and/or neuromuscular disorders; OR

B) Patient Currently Receiving Evrysdi. Approve for 1 year if the patient meets ALL of the following (i, ii, iii, and iv):

i. Patient has had a genetic test confirming the diagnosis of spinal muscular atrophy with bi-allelic pathogenic variants in the survival motor neuron 1 (SMN1) gene; AND
Note: Pathogenic variants may include homozygous deletion, compound heterozygous mutation, or a variety of other rare mutations.

ii. Patient meets ONE of the following (a or b):
a) Patient has two or three survival motor neuron 2 (SMN2) gene copies; OR
b) Patient meets BOTH of the following [(1) and (2)]:
(1) Patient has four survival motor neuron 2 (SMN2) gene copies; AND
(2) Patient has objective signs consistent with spinal muscular atrophy Types 1, 2, or 3; AND

iii. The medication is prescribed by a physician who has consulted with a specialist or who specializes in the management of patients with spinal muscular atrophy and/or neuromuscular disorders; AND

iv. Patient must meet ONE of the following (a or b):
a) Patient must have had a positive clinical response (for example, improvement or stabilization) from pretreatment baseline status with

Evrysdi in ONE of the following exams [(1), (2), (3), (4), (5), (6), or (7)] **[documentation required]**:

(1) Bayley Scales of Infant and Toddler Development; OR
(2) Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND); OR
(3) Hammersmith Functional Motor Scale Expanded (HFMSE); OR
(4) Hammersmith Infant Neurological Exam Part 2 (HINE-2); OR
(5) Motor Function Measure-32 Items (MFM-32); OR
(6) Revised Upper Limb Module (RULM) test; OR
(7) World Health Organization motor milestone scale; OR

b) According to the prescribing physician, the patient has responded to Evrysdi and continues to benefit from ongoing Evrysdi therapy by the most recent physician monitoring/assessment tools **[documentation required]**.

Note: Examples include pulmonary function tests showing improvement, bulbar function test results suggesting benefits, reduced need for respiratory support, decrease in the frequency of respiratory infections or complications, and/or prevention of permanent assisted ventilation.

CONDITIONS NOT COVERED

- **Evrysdi® (risdiplam oral solution and tablets – Genentech/Roche) is(are) considered not medically necessary for ANY other use(s) including the following (this list may not be all inclusive; criteria will be updated as new published data are available):**

1. **Patient has Complete Paralysis of All Limbs.** Data are needed to determine if this patient population with advanced spinal muscular atrophy would derive benefits from Evrysdi.

2. **Patient has Permanent Ventilator Dependence.** Data are needed to determine if this patient population with advanced spinal muscular atrophy would derive benefits from Evrysdi.

3. **Concurrent Use with Spinraza (nusinersen intrathecal injection).** Further study is needed to determine if use of Evrysdi with Spinraza is efficacious and safe.

REFERENCES

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9. Glascock J, Sampson J, Haidet-Phillips A, et al. Treatment algorithm for infants diagnosed with spinal muscular atrophy through newborn screening. *J Neuromuscul Dis*. 2018;5:145-158.
10. Glascock J, Sampson J, Connolly AM, et al. Revised recommendations for the treatment of infants diagnosed with spinal muscular atrophy via newborn screening who have 4 copies of SMN2. *J Neuromuscul Dis*. 2020;7(2):97-100.

HISTORY

Type of Revision	Summary of Changes	Review Date
Annual Revision	Regarding Documentation, medical test results and prescription receipts were added as examples; the example provided of laboratory "tests" was changed to laboratory "results." In the Policy Statement, regarding verification in claims history, the phrase "if claims history is available" was added to account for situations in which claims history is not present. For Spinal Muscular Atrophy – Treatment, in criteria that the patient has not received Zolgensma in the past (with verification in claims history required), the Note was revised to account for situations in which a claims history is not available.	10/02/2024
Selected Revision	Evrysdi tablets (5 mg) were added to the policy. There were no changes to the criteria.	02/19/2025
Early Annual Revision	Regarding documentation, the exceptions were removed in which it states that in subsequent coverage reviews for a patient who has previously met the documentation requirements and related criteria in the *Spinal Muscular Atrophy – Evrysdi Prior Authorization Policy* through the Coverage Review Department, and who is requesting reauthorization, the criteria utilized do NOT require resubmission of documentation for reauthorization, except for the criterion requiring documentation of response or benefit to Evrysdi therapy. In addition, the following changes were made: **Spinal Muscular Atrophy – Treatment:** For initial therapy and for a patient currently receiving Evrysdi, the requirement was removed from approval criteria that for a patient currently receiving or who has received prior treatment with Spinraza, the prescribing physician confirms that further therapy with Spinraza will be discontinued. Also, the requirement was removed that according to the prescribing physician, a female patient of reproductive potential must not be currently pregnant and must utilize effective contraception during treatment and for 1 month after the last Evrysdi dose. In addition, the dosing criteria were removed that was based on the current (within the past 1 month) patient weight. For a patient currently receiving Evrysdi only, the documentation requirement was removed from the	07/30/2025

	following criteria: 1) patient has a genetic test confirming the diagnosis of spinal muscular atrophy with bi-allelic pathogenic variants in the survival motor neuron 1 gene; 2) patient has two or three survival motor neuron 2 gene copies; 3) patient has four survival motor neuron 2 gene copies, and 4) patient has objective signs consistent with spinal muscular atrophy Types 1, 2, or 3. For a patient currently receiving Evrysdi, the requirement was removed that the patient has not received Zolgensma in the past; the related Note and that verification was required in claims history were also deleted. **Conditions Not Covered:** Concurrent use with Spinraza was added.	
Selected Revision	**Spinal Muscular Atrophy – Treatment:** For initial therapy, Itvisma was added as a gene therapy that the patient should not have received in the past. The Note now includes that if no claim for Itvisma is present (or if claims history is not available), the prescribing physician confirms that the patient has not previously received Itvisma.	01/28/2026
Selected Revision	**Spinal Muscular Atrophy – Treatment:** For a patient currently receiving Evrysdi the duration of therapy was changed to 1 year; previously, it was 4 months.	02/18/2026
Annual Revision	No criteria changes.	07/22/2026

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