**Prior Authorization Criteria**

**Evrysdi® (risdiplam) PA Criteria**

Evrysdi is a survival of motor neuron 2 (SMN2) splicing modifier indicated for the treatment of spinal muscular atrophy (SMA) in patients 2 months of age and older.

**Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.**

***Initial Authorization: 6 months***

☐ Yes ☐ No Diagnosis of SMA;

***AND***

 ☐ Yes ☐ No Genetic testing quantifying number of copies of SMN2 gene ≥ 1 but ≤ 4

***AND***

Genetic testing confirms the presence of one of the following (a, b, or c)

a. Homozygous deletions of SMN1 gene (e.g., absence of the SMN1 gene);

b. Homozygous mutation in the SMN1 gene (e.g., biallelic mutations of exon 7);

c. Compound heterozygous mutation in the SMN1 gene [e.g., deletion of SMN1 exon 7 (allele 1) and mutation of SMN1 (allele 2)];

***AND***

☐ Yes ☐ No Prescribed by or in consultation with a neurologist

***AND***

Documentation of one of the following baseline scores (a or b):

1. For age < 2 years: Children’s Hospital of Philadelphia Infant Test of Neuromuscular Disorder (CHOP-INTEND) score or Hammersmith Infant Neurological Examination (HINE) Section 2 motor milestone score;
2. For age ≥ 2 years: Hammersmith functional motor scale expanded (HFMSE) score, Revised Hammersmith Scale (RHS), Upper Limb Module (ULM), Revised Upper Limb Module (RULM), or 6-Minute Walk Test (6MWT)

***AND***

☐ Yes ☐ No Member does not require tracheostomy or invasive ventilation.

***AND***

Evrysdi is not prescribed concurrently with Spinraza® and/or Zolgensma®;

***AND***

If the member is currently on Spinraza, documentation of prescriber attestation of Spinraza discontinuation;

***AND***

Request meets one of the following (a, b, c or d):

 a. If 2 months of age or less, dose does not exceed 0.15mg/kg per day;

 b. If 2 months of age to less than 2 years of age, dose does not exceed 0.2 mg/kg per day;

c. If 2 years of age and older, weighing less than 20 kg, dose does not exceed 0.25 mg/kg per day;

d. If 2 years of age and older, weighing 20 kg or more, dose does not exceed 5 mg per day.

**Reauthorization criteria: *12 months***

Spinal Muscular Atrophy (must meet all):

☐ Yes ☐ No Member does not require tracheostomy or invasive ventilation;

***AND***

☐ Yes ☐ No Member is responding positively to therapy as evidenced by one of the following (a, b, or c):

 a. For age < 2 years, must meet one of the following (i **or** ii):

i. For CHOP-INTEND, must demonstrate score improvement or maintenance of previous score improvement of ≥ 4 points from baseline;

ii. For HINE motor milestone score, must demonstrate score improvement or maintenance of previous improvement in one or more categories AND improvement in more motor milestone categories than worsening;

b. For age ≥ 2 years, must meet one of the following (i, ii, **or** iii):

i. If first renewal since turning 2 years old, must provide submission of baseline HFMSE score, RHS score, RULM or ULM score, or 6MWT distance AND meet one of the following (1 **or** 2):

1) For CHOP-INTEND, must demonstrate score improvement or maintenance of previous score improvement of ≥ 4 points from baseline;

2) For HINE motor milestone score, must demonstrate score improvement or maintenance of previous improvement in one or more categories AND improvement in more motor milestone categories than worsening;

ii. If ≤ 2 years at therapy initiation and request is for subsequent renewal since turning 2, must meet one of the following

1) For HFMSE, RHS, ULM or RULM, must demonstrate score improvement or maintenance of previous score improvement from baseline score submitted at first renewal since turning 2 years old;

2) For 6MWT distance, must demonstrate improvement or maintenance of baseline distance;

iii. If > 2 years at therapy initiation, must meet one of the following (1, 2, 3, **or** 4):

1) For HFMSE or RHS, must demonstrate score improvement or maintenance of previous score improvement of ≥ 3 points from baseline;

2) For ULM, must demonstrate score improvement or maintenance of previous improvements in ≥ 2 points from baseline;

3) For RULM, must demonstrate score improvement or maintenance of previous improvements in ≥ 4 points from baseline;

4) For 6MWT distance, must demonstrate improvement or maintenance of baseline distance

c. Member has not had a decline in motor function test score(s) from baseline **AND** medical justification demonstrates and supports that member is responding positively to therapy;

☐ Yes ☐ No Evrysdi is not prescribed concurrently with Spinraza and/or Zolgensma;

If request is for a dose increase, request meets one of the following (a, b, c or d):

a. If 2 months of age or less, new dose does not exceed 0.15mg/kg per day;

b. If 2 months of age to less than 2 years of age, new dose does not exceed 0.2 mg/kg per day;

c. If 2 years of age and older, weighing less than 20 kg, new dose does not exceed 0.25 mg/kg per day;

d. If 2 years of age and older, weighing 20 kg or more, new dose does not exceed 5 mg per day

**See Package Insert for specific details on Contraindications/Warnings/Precautions**

**Dosing**

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**How Supplied:**