



coordinated care™

Agents for Sickle Cell Anemia – Endari™ (L-glutamine)

WA.PHAR.59 Agents for Sickle Cell Anemia L-glutamine (ENDARI)

Effective: October 1, 2018

Background:

Sickle cell disease is a group of disorders that affects hemoglobin, the molecule in red blood cells that delivers oxygen to cells throughout the body. People with this disorder have atypical hemoglobin molecules called hemoglobin S, which can distort red blood cells into a sickle, or crescent, shape.

Medical necessity

| Drug | Medical Necessity |
|-----------------------|---|
| L-glutamine (Endari™) | Endari™ may be considered medically necessary when: Used to reduce the acute complications of sickle cell disease in adult and pediatric patients 5 years of age and older |

Clinical policy:

| Clinical Criteria | | | | | | | | | | | | | | | | | | | | | | | | | |
|--------------------------------|---|---------------------|------------------|-------------------|------------------|------------------|-----------------|--------------|--------------|---|----|---|---|----------|-----------|----|----|---|---|-----------------|------------------|----|----|---|---|
| Initial authorization criteria | <ol style="list-style-type: none"> 1. Diagnosis of sickle cell disease 2. Greater than or equal to (\geq) 2 painful sickle cell episodes within the past 12 months 3. History of failure after at least 3 months, contraindication or intolerance due to adverse side effects to ONE of the following: <ol style="list-style-type: none"> a. Stabilized on hydroxyurea for at least 3 months and to be continued concurrently with Endari™ b. Documentation of contraindication or intolerance due to adverse reaction to hydroxyurea 4. Prescribed by or in consultation with a hematologist or a specialist with expertise in treatment of sickle cell disease 5. Dose limits based on body weight: <table border="1" data-bbox="592 1665 1490 1829"> <thead> <tr> <th>Weight in kilograms</th> <th>Weight in pounds</th> <th>Per dose in grams</th> <th>Per day in grams</th> <th>Packets per dose</th> <th>Packets per day</th> </tr> </thead> <tbody> <tr> <td>Less than 30</td> <td>Less than 66</td> <td>5</td> <td>10</td> <td>1</td> <td>2</td> </tr> <tr> <td>30 to 65</td> <td>66 to 143</td> <td>10</td> <td>20</td> <td>2</td> <td>4</td> </tr> <tr> <td>Greater than 65</td> <td>Greater than 143</td> <td>15</td> <td>30</td> <td>3</td> <td>6</td> </tr> </tbody> </table> | Weight in kilograms | Weight in pounds | Per dose in grams | Per day in grams | Packets per dose | Packets per day | Less than 30 | Less than 66 | 5 | 10 | 1 | 2 | 30 to 65 | 66 to 143 | 10 | 20 | 2 | 4 | Greater than 65 | Greater than 143 | 15 | 30 | 3 | 6 |
| Weight in kilograms | Weight in pounds | Per dose in grams | Per day in grams | Packets per dose | Packets per day | | | | | | | | | | | | | | | | | | | | |
| Less than 30 | Less than 66 | 5 | 10 | 1 | 2 | | | | | | | | | | | | | | | | | | | | |
| 30 to 65 | 66 to 143 | 10 | 20 | 2 | 4 | | | | | | | | | | | | | | | | | | | | |
| Greater than 65 | Greater than 143 | 15 | 30 | 3 | 6 | | | | | | | | | | | | | | | | | | | | |
| | 6. Greater than or equal to (\geq) 5 years of age | | | | | | | | | | | | | | | | | | | | | | | | |

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|---------------------------------|--|
| | <p>7. Maximum dose 30 grams per day</p> <p>Approve for 12 months</p> |
| Reauthorization criteria | <p>Documentation of positive clinical response</p> <p>Approve for 12 months</p> |

References

1. Endari™ prescribing information. Emmaus Medical, Inc. Torrance, CA. July 2017
2. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2017.
3. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website
4. DrugPoints® System (electronic version). Truven Health Analytics, Greenwood Village, CO.
5. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2017