Xermelo (telotristat ethyl)  
(Oral)

I. **Length of Authorization**

Coverage will be provided for 6 months and may be renewed.

II. **Dosing Limits**

A. **Quantity Limit (max daily dose) [NDC Unit]:**
   - Xermelo 250 tablet: 1 monthly case (84 tablets) per 28 days

B. **Max Units (per dose and over time) [HCPCS Unit]:**
   - 250 mg three times daily

III. **Initial Approval Criteria**

Coverage is provided in the following conditions:

- Patient is at least 18 years old; **AND**

**Universal Criteria**

- Patient has been receiving therapy with the FDA-approved maximum (or highest tolerated) dose of a somatostatin analog therapy (SSA) (i.e., octreotide solution/depot or lanreotide depot) for at least 3 months; **AND**
- Patient will continue to receive this SSA therapy in combination with telotristat ethyl; **AND**

**Carcinoid Syndrome Diarrhea †/Φ**

- Patient has a carcinoid/neuroendocrine tumor and has been diagnosed with carcinoid syndrome; **AND**
- Patient has had an inadequate response to antidiarrheals (e.g., loperamide); **AND**
- Patient’s baseline bowel movements per day are ≥ 4

† FDA Approved Indication(s), ‡ Compendia Recommended Indication(s); Φ Orphan Drug
IV. Renewal Criteria\textsuperscript{1,2}

Authorizations may be renewed based upon the following:

- Patient continues to meet universal and other indication-specific relevant criteria identified in section III: \textbf{AND}

- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe constipation, abdominal pain, etc.): \textbf{AND}

- Patient has responded to therapy as indicated by a reduction in the number of bowel movements per day from pre-treatment baseline

V. Dosage/Administration

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diarrhea inadequately controlled by SSA therapy</td>
<td>250 mg orally three times daily</td>
</tr>
<tr>
<td></td>
<td>\textit{When short-acting octreotide is used in combination with Xermelo, administer short-acting octreotide at least 30 minutes after administering Xermelo.}</td>
</tr>
</tbody>
</table>

VI. Billing Code/Availability Information

HCPCS code: J8499 – Prescription drug, oral, non chemotherapeutic, nos

NDC: Xermelo 250 mg tablet: monthly case of 84 tablets: 70183-0125-xx

VII. References


2. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium\textsuperscript{®}) for telotristat ethyl. National Comprehensive Cancer Network, 2020. The NCCN Compendium\textsuperscript{®} is a derivative work of the NCCN Guidelines\textsuperscript{®}. NATIONAL COMPREHENSIVE CANCER NETWORK\textsuperscript{®,} NCCN\textsuperscript{®}, and NCCN GUIDELINES\textsuperscript{®} are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed April 2020.


Appendix 1 – Covered Diagnosis Codes
<table>
<thead>
<tr>
<th>ICD-10</th>
<th>ICD-10 Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>C7A.00</td>
<td>Malignant carcinoid tumor of unspecified site</td>
</tr>
<tr>
<td>C7A.010</td>
<td>Malignant carcinoid tumor of the duodenum</td>
</tr>
<tr>
<td>C7A.011</td>
<td>Malignant carcinoid tumor of the jejunum</td>
</tr>
<tr>
<td>C7A.012</td>
<td>Malignant carcinoid tumor of the ileum</td>
</tr>
<tr>
<td>C7A.019</td>
<td>Malignant carcinoid tumor of the small intestine, unspecified portion</td>
</tr>
<tr>
<td>C7A.020</td>
<td>Malignant carcinoid tumor of the appendix</td>
</tr>
<tr>
<td>C7A.021</td>
<td>Malignant carcinoid tumor of the cecum</td>
</tr>
<tr>
<td>C7A.022</td>
<td>Malignant carcinoid tumor of the ascending colon</td>
</tr>
<tr>
<td>C7A.023</td>
<td>Malignant carcinoid tumor of the transverse colon</td>
</tr>
<tr>
<td>C7A.024</td>
<td>Malignant carcinoid tumor of the descending colon</td>
</tr>
<tr>
<td>C7A.025</td>
<td>Malignant carcinoid tumor of the sigmoid colon</td>
</tr>
<tr>
<td>C7A.026</td>
<td>Malignant carcinoid tumor of the rectum</td>
</tr>
<tr>
<td>C7A.029</td>
<td>Malignant carcinoid tumor of the large intestine, unspecified portion</td>
</tr>
<tr>
<td>C7A.090</td>
<td>Malignant carcinoid tumor of the bronchus and lung</td>
</tr>
<tr>
<td>C7A.091</td>
<td>Malignant carcinoid tumor of the thymus</td>
</tr>
<tr>
<td>C7A.092</td>
<td>Malignant carcinoid tumor of the stomach</td>
</tr>
<tr>
<td>C7A.093</td>
<td>Malignant carcinoid tumor of the kidney</td>
</tr>
<tr>
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</tr>
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<td>C7A.095</td>
<td>Malignant carcinoid tumor of the midgut, unspecified</td>
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<td>C7A.096</td>
<td>Malignant carcinoid tumor of the hindgut, unspecified</td>
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<tr>
<td>C7A.098</td>
<td>Malignant carcinoid tumors of other sites</td>
</tr>
<tr>
<td>C7A.1</td>
<td>Malignant poorly differentiated neuroendocrine tumors</td>
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<tr>
<td>C7A.8</td>
<td>Other malignant neuroendocrine tumors</td>
</tr>
<tr>
<td>C7B.00</td>
<td>Secondary carcinoid tumors, unspecified site</td>
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<tr>
<td>C7B.01</td>
<td>Secondary carcinoid tumors of distant lymph nodes</td>
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<td>C7B.02</td>
<td>Secondary carcinoid tumors of liver</td>
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<tr>
<td>C7B.03</td>
<td>Secondary carcinoid tumors of bone</td>
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<td>C7B.04</td>
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<td>Other secondary neuroendocrine tumors</td>
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<tr>
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<tr>
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<td>Benign carcinoid tumor of the ileum</td>
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<tr>
<td>D3A.019</td>
<td>Benign carcinoid tumor of the small intestine, unspecified portion</td>
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<td>ICD-10</td>
<td>ICD-10 Description</td>
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<tr>
<td>---------</td>
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<tr>
<td>D3A.020</td>
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<tr>
<td>D3A.021</td>
<td>Benign carcinoid tumor of the cecum</td>
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<tr>
<td>D3A.022</td>
<td>Benign carcinoid tumor of the ascending colon</td>
</tr>
<tr>
<td>D3A.023</td>
<td>Benign carcinoid tumor of the transverse colon</td>
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<tr>
<td>D3A.024</td>
<td>Benign carcinoid tumor of the descending colon</td>
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<td>D3A.025</td>
<td>Benign carcinoid tumor of the sigmoid colon</td>
</tr>
<tr>
<td>D3A.026</td>
<td>Benign carcinoid tumor of the rectum</td>
</tr>
<tr>
<td>D3A.029</td>
<td>Benign carcinoid tumor of the large intestine, unspecified portion</td>
</tr>
<tr>
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<td>Benign carcinoid tumor of the bronchus and lung</td>
</tr>
<tr>
<td>D3A.091</td>
<td>Benign carcinoid tumor of the thymus</td>
</tr>
<tr>
<td>D3A.092</td>
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<tr>
<td>E34.0</td>
<td>Carcinoid syndrome</td>
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<tr>
<td>Z85.020</td>
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<tr>
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<tr>
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</tbody>
</table>

**Appendix 2 – Centers for Medicare and Medicaid Services (CMS)**

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Article (LCA) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: [http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx](http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx). Additional indications may be covered at the discretion of the health plan.

**Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/LCA): N/A**

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<th>Contractor</th>
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<td>E (1)</td>
<td>CA, HI, NV, AS, GU, CNMI</td>
<td>Noridian Healthcare Solutions, LLC</td>
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<tr>
<td>F (2 &amp; 3)</td>
<td>AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ</td>
<td>Noridian Healthcare Solutions, LLC</td>
</tr>
<tr>
<td>5</td>
<td>KS, NE, IA, MO</td>
<td>Wisconsin Physicians Service Insurance Corp (WPS)</td>
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<td>6</td>
<td>MN, WI, IL</td>
<td>National Government Services, Inc. (NGS)</td>
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<tr>
<td>H (4 &amp; 7)</td>
<td>LA, AR, MS, TX, OK, CO, NM</td>
<td>Novitas Solutions, Inc.</td>
</tr>
<tr>
<td>8</td>
<td>MI, IN</td>
<td>Wisconsin Physicians Service Insurance Corp (WPS)</td>
</tr>
<tr>
<td>Jurisdiction</td>
<td>Applicable State/US Territory</td>
<td>Contractor</td>
</tr>
<tr>
<td>--------------</td>
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<tr>
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<tr>
<td>J (10)</td>
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<td>M (11)</td>
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<td>K (13 &amp; 14)</td>
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<td>15</td>
<td>KY, OH</td>
<td>CGS Administrators, LLC</td>
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