Pharmacy Policy Bulletin: J-1356 Ogsiveo (nirogacestat) - Commercial and Healthcare Reform	
Number: J-1356	**Category:** Prior Authorization
Line(s) of Business: ☒ Commercial ☒ Healthcare Reform ☐ Medicare	**Benefit(s):** **Commercial:** **Prior Authorization (1.):** **1.** Miscellaneous Specialty Drugs Oral = Yes w/ Prior Authorization **Healthcare Reform:** Not Applicable
Region(s): ☒ All ☐ Delaware ☐ New York ☐ Pennsylvania ☐ West Virginia	**Additional Restriction(s):** None
Version: J-1356-002	**Original Date:** 01/31/2024
Effective Date: 12/20/2024	**Review Date:** 12/04/2024

Drugs Product(s):	• Ogsiveo (nirogacestat)
FDA-Approved Indication(s):	• Treatment of adult patients with progressing desmoid tumors who require systemic treatment

Background:	• Ogsiveo is a gamma secretase inhibitor that blocks proteolytic activation of the Notch receptor. When dysregulated, Notch can activate pathways that contribute to tumor growth. • Desmoid tumors, also referred to as aggressive fibromatosis, are benign, locally invasive tumors of the soft tissue. They are a non-cancerous form of soft tissue sarcoma that have a high recurrence rate following surgical removal but do not have metastatic potential. Tumors can form anywhere in the body and are often found in the abdomen, as well as the shoulders, upper arms, and thighs. Superficial tumors tend to be less aggressive than intra-abdominal, extra-abdominal, or mesenteric tumors. • The incidence of desmoid tumors is estimated to be 2-4 per million people per year worldwide and, in adults, are more commonly seen (2:1) in females than males and in individuals 10-40 years of age. Because desmoid tumors rarely impact survival, data are limited to estimate mortality impact. One analysis estimated 5-year survival of patients diagnosed with stage I, II, III, and IV intra-abdominal desmoid tumor at 95%, 100%, 89%, and 76%, respectively. • Ogsiveo is the first FDA-approved therapy for desmoid tumors. Other therapies that are not FDA-approved but are supported by the National Comprehensive Cancer Network (NCCN) guidelines include Nexavar (sorafenib), Gleevec (imatinib), Votrient (pazopanib), methotrexate and either Navelbine (vinorelbine) or vinblastine, Doxil (liposomal doxorubicin), or doxorubicin with or without dacarbazine. Sorafenib has a Category 1 recommendation, while all of the others are Category 2A. • Prescribing Considerations:

	○ Though they are not cancerous, desmoid tumors are typically treated by oncologists. ○ The recommended dosage of Ogsiveo is 150 mg administered orally twice daily until disease progression or unacceptable toxicity. ○ Ogsiveo has warning and precautions for diarrhea, ovarian toxicity, hepatoxicity, non-melanoma skin cancers, electrolyte abnormalities, and embryo-fetal toxicity.

Approval Criteria

I. Initial Authorization

When a benefit, coverage of Ogsiveo may be approved when all of the following criteria are met **(A. and B.)**:

A. The member is 18 years of age or older.

B. The member has a diagnosis of desmoid tumor (ICD-10: D48.11) and meets all of the following criteria **(1. and 2.)**:

1. The desmoid tumor has progressed within the past 12 months.

2. The patient requires systemic therapy for the treatment of the desmoid tumor.

II. Reauthorization

When a benefit, reauthorization of Ogsiveo may be approved when the following criterion is met **(A.)**:

A. The prescriber attests that the member has experienced positive clinical response to therapy.

III. An exception to some or all of the criteria above may be granted for select members and/or circumstances based on state and/or federal regulations.

IV. Coverage of oncology drug(s) listed in this policy may be approved on a case-by-case basis per indications supported in the most current NCCN guidelines.

Limitations of Coverage

I. Coverage of drug(s) addressed in this policy for disease states outside of the FDA-approved indications should be denied based on the lack of clinical data to support effectiveness and safety in other conditions unless otherwise noted in the approval criteria.

II. For Commercial or HCR members with a closed formulary, a non-formulary product will only be approved if the member meets the criteria for a formulary exception in addition to the criteria outlined within this policy.

Authorization Duration

- Commercial and HCR Plans: If approved, up to a 12 month authorization may be granted.

Automatic Approval Criteria

None.

References:

1. Ogsiveo [package insert]. Stamford, CT. SpringWorks Therapeutics, Inc.; April 2024.
2. NORD. Desmoid tumor. Available at: https://rarediseases.org/rare-diseases/desmoid-tumor/. Accessed November 4, 2024.
3. Genetic and Rare Diseases Information Center. Desmoid tumor. National Institute for Advancing Translational Sciences. Available at: https://rarediseases.info.nih.gov/diseases/1820/desmoid-tumor. Accessed November 4, 2024 .

4. Jain P, Santhosh C. US FDA approves SpringWorks Therapeutics' non-cancerous tumor treatment. Reuters. Available at: https://www.reuters.com/business/healthcare-pharmaceuticals/us-fda-approves-springworks-therapeutics-non-cancerous-tumor-treatment-2023-11-27/. Accessed November 4, 2024.
5. Desmoid tumor. National Cancer Institute. Available at: https://www.cancer.gov/pediatric-adult-rare-tumor/rare-tumors/rare-soft-tissue-tumors/desmoid-tumor. Accessed November 4, 2024.
6. DRUGDEX System (Micromedex 2.0). Greenwood Village, CO: Truven Health Analytics; 2023.
7. National Comprehensive Cancer Network. NCCN Guidelines Version 2.2023 Soft tissue sarcoma. https://www.nccn.org/professionals/physician_gls/pdf/sarcoma.pdf. Accessed November 4, 2024.