Ogsiveo® (nirogacestat) 150 mg & 100 mg tablets

FDA-Approved Therapy and ICD-10-CM Codes for Desmoid Tumors

OGSIVEO is the first and only FDA-approved treatment for adult patients with progressing desmoid tumors who require systemic treatment.[1]

Desmoid tumors are rare, locally aggressive soft tissue tumors that are characterized by high rates of initial misdiagnosis.[2,3]

Effective October 1, 2023, the list of ICD-10-CM codes for desmoid tumors included in the table below became available.

Until October 1, 2023, desmoid tumors fell under an ICD-10-CM code (D48.1) covering a variety of connective and soft tissue tumors of "uncertain behavior."[5]

Example of a large desmoid tumor proximal to the spine[4]

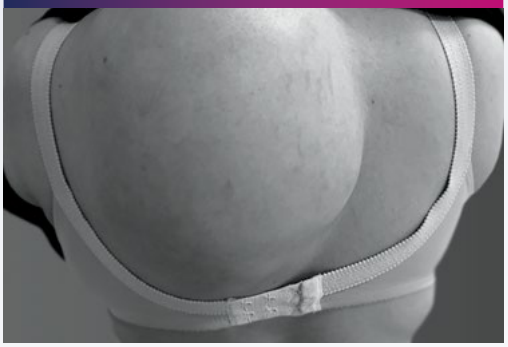

Image adapted from Cohen S et al. *World J Surg Oncol.* 2008;6:28. Reused under Creative Commons License 2.0 (https://creativecommons.org/licenses/by/2.0). Image background changed to gray.

ICD-10-CM Diagnosis Code[6]	Description[6]
D48.11	**Desmoid tumor** (category heading)
D48.110	**Desmoid tumor of head and neck**
D48.111	**Desmoid tumor of chest wall**
D48.112	**Desmoid tumor, intrathoracic**
D48.113	**Desmoid tumor of abdominal wall**
D48.114	**Desmoid tumor, intraabdominal** Desmoid tumor of pelvic cavity Desmoid tumor, peritoneal, retroperitoneal
D48.115	**Desmoid tumor of upper extremity and shoulder girdle**
D48.116	**Desmoid tumor of lower extremity and pelvic girdle** Desmoid tumor of buttock
D48.117	**Desmoid tumor of back**
D48.118	**Desmoid tumor of other site**
D48.119	**Desmoid tumor of unspecified site**

FDA=U.S. Food and Drug Administration; ICD-10-CM=*International Classification of Diseases, Tenth Revision, Clinical Modification.*

References: 1. OGSIVEO. Prescribing Information. SpringWorks Therapeutics, Inc. **2.** Constantinidou A, Scurr M, Judson I, Litchman C. Clinical presentation of desmoid tumors. In: Litchman C, ed. *Desmoid Tumors.* Springer; 2012:5-16. doi:10.1007/978-94-007-1685-8_2 **3.** Kasper B, Baumgarten C, Garcia J, et al; Desmoid Working Group. An update on the management of sporadic desmoid-type fibromatosis: a European Consensus Initiative between Sarcoma PAtients EuroNet (SPAEN) and European Organization for Research and Treatment of Cancer (EORTC)/Soft Tissue and Bone Sarcoma Group (STBSG). *Ann Oncol.* 2017;28(10):2399-2408. doi:10.1093/annonc/mdx323 **4.** Cohen S, Ad-El D, Benjaminov O, Gutman H. Post-traumatic soft tissue tumors: case report and review of the literature a propos a post-traumatic paraspinal desmoid tumor. *World J Surg Oncol.* 2008;6:28. doi:10.1186/1477-7819-6-28 **5.** Desmoid Tumor Research Foundation. Desmoid ICD-10 codes. Accessed March 26, 2024. https://dtrf.org/clinicians-researchers/desmoid-icd-10-codes/ **6.** Centers for Medicare & Medicaid Services 2024 ICD-10-CM codes. Centers for Medicare & Medicaid Services website. Accessed March 26, 2024. https://www.cms.gov/medicare/coding-billing/icd-10-codes/2024-icd-10-cm

IMPORTANT SAFETY INFORMATION

WARNINGS AND PRECAUTIONS

- **Diarrhea:** Diarrhea, sometimes severe, can occur in patients treated with OGSIVEO. Diarrhea occurred in 84% of patients treated with OGSIVEO, and included Grade 3 events in 16% of patients. Median time to first diarrhea event was 9 days (range: 2 to 434 days). Monitor patients and manage using antidiarrheal medications. Modify dose as recommended.

Please see additional Important Safety Information on reverse side and click here for full Prescribing Information.

IMPORTANT SAFETY INFORMATION (cont'd)

WARNINGS AND PRECAUTIONS (cont'd)

- **Ovarian Toxicity:** Female reproductive function and fertility may be impaired in patients treated with OGSIVEO. Impact on fertility may depend on factors like duration of therapy and state of gonadal function at time of treatment. Long-term effects of OGSIVEO on fertility have not been established. Advise patients on the potential risks for ovarian toxicity before initiating treatment. Monitor patients for changes in menstrual cycle regularity or the development of symptoms of estrogen deficiency, including hot flashes, night sweats, and vaginal dryness.
- **Hepatotoxicity:** ALT or AST elevations occurred in 30% and 33% of patients, respectively. Grade 3 ALT or AST elevations (>5 × ULN) occurred in 6% and 2.9% of patients. Monitor liver function tests regularly and modify dose as recommended.
- **Non-Melanoma Skin Cancers:** New cutaneous squamous cell carcinoma and basal cell carcinoma occurred in 2.9% and 1.4% of patients, respectively. Perform dermatologic evaluations prior to initiation of OGSIVEO and routinely during treatment.
- **Electrolyte Abnormalities:** Decreased phosphate (65%) and potassium (22%) occurred in OGSIVEO-treated patients. Phosphate <2 mg/dL occurred in 20% of patients. Grade 3 decreased potassium occurred in 1.4% of patients. Monitor phosphate and potassium levels regularly and supplement as necessary. Modify dose as recommended.
- **Embryo-Fetal Toxicity:** OGSIVEO can cause fetal harm when administered to pregnant women. Oral administration of nirogacestat to pregnant rats during the period of organogenesis resulted in embryo-fetal toxicity and death at maternal exposures below human exposure at the recommended dose of 150 mg twice daily. Advise pregnant women of the potential risk to a fetus. Advise females and males of reproductive potential to use effective contraception during treatment with OGSIVEO and for 1 week after the last dose.

ADVERSE REACTIONS

- The most common (≥15%) adverse reactions were diarrhea (84%), ovarian toxicity (75% in the 36 females of reproductive potential), rash (68%), nausea (54%), fatigue (54%), stomatitis (39%), headache (30%), abdominal pain (22%), cough (20%), alopecia (19%), upper respiratory tract infection (17%), and dyspnea (16%).
- Serious adverse reactions occurred in 20% of patients who received OGSIVEO. Serious adverse reactions occurring in ≥2% of patients were ovarian toxicity (4%).
- The most common laboratory abnormalities (≥15%) were decreased phosphate, increased urine glucose, increased urine protein, increased AST, increased ALT, and decreased potassium.

DRUG INTERACTIONS

- **CYP3A Inhibitors and Inducers:** Avoid concomitant use with strong or moderate CYP3A inhibitors (including grapefruit products, Seville oranges, and starfruit) and strong or moderate CYP3A inducers.
- **Gastric Acid Reducing Agents:** Avoid concomitant use with proton pump inhibitors and H2 blockers. If concomitant use cannot be avoided, OGSIVEO can be staggered with antacids (e.g., administer OGSIVEO 2 hours before or 2 hours after antacid use).
- Consult the full Prescribing Information prior to and during treatment for important drug interactions.

USE IN SPECIFIC POPULATIONS

- Because of the potential for serious adverse reactions in breastfed children, advise women not to breastfeed during treatment with OGSIVEO and for 1 week after the last dose.

Please click here for full Prescribing Information.

SpringWorks® THERAPEUTICS