

FDA-Approved Therapy and ICD-10-CM Codes Now Available 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 spine⁴



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 paraspinous desmoid tumor. *World J Surg Oncol*. 2008;6:28. doi:10.1186/1477-7819-6-28 **5.** Desmoid Tumor Research Foundation. New ICD-10-CM diagnosis codes for desmoid tumors. Accessed August 9, 2023. <https://dtrf.org/icd-codes-and-desmoid-tumors/> **6.** Centers for Medicare & Medicaid Services 2024 ICD-10-CM codes. Centers for Medicare & Medicaid Services website. Accessed August 9, 2023. <https://www.cms.gov/medicare/coding-billing/icd-10-codes/2024-icd-10-cm>

IMPORTANT SAFETY INFORMATION WARNINGS AND PRECAUTIONS

- **Diarrhea:** Diarrhea occurred in 84% of patients treated with OGSIVEO. Grade 3 events occurred in 16% of patients. Monitor patients and manage using antidiarrheal medications. Modify dose as recommended.

Please see additional Important Safety Information on next page 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 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 ($>5 \times$ 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:** Oral administration of nirogacestat to pregnant rats during the period of organogenesis resulted in embryo-fetal toxicity 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 ($\geq 15\%$) adverse reactions were diarrhea, ovarian toxicity, rash, nausea, fatigue, stomatitis, headache, abdominal pain, cough, alopecia, upper respiratory tract infection, and dyspnea.
- Serious adverse reactions occurring in $\geq 2\%$ of patients were ovarian toxicity (4%).
- The most common laboratory abnormalities ($\geq 15\%$) 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.

Please [click here](#) for full Prescribing Information.