SpringWorks THERAPEUTICS

For people diagnosed with desmoid tumors

FINDING A WAY FORWARD WITH YOUR CARE

MAKING A PLAN THAT WORKS FOR YOU

Being diagnosed with a desmoid tumor brings important decisions you need to make about your health with your care team. Take the time you need to understand what options are available to you, the potential risks and benefits, and who can provide the best guidance—so you can ask questions and develop a plan that works for you.

STAYING INFORMED ABOUT CARE OPTIONS

Care approaches are always evolving. To stay up to date on the latest treatments, you can consult the NCCN Guidelines for Patients® which are updated frequently with the latest recommendations for desmoid tumor care.[1]

FINDING THE RIGHT CARE FROM DESMOID TUMOR EXPERTS

The NCCN Guidelines for Patients recommend seeing doctors experienced in desmoid tumors before starting treatment.[1]

It's always okay to seek additional medical opinions if you feel unsure about your care. Working closely with your care team is essential to help you choose the care option that's right for you.

Healthcare providers who specialize in desmoid tumors may be based at a sarcoma center. Find a list of sarcoma centers by visiting the website of Sarcoma Alliance for Research through Collaboration (SARC), a non-profit organization.[2,*]

*SpringWorks Therapeutics, Inc. is providing this link to help patients find a sarcoma specialist by region, but SpringWorks had no role in developing this list and inclusion on this list does not represent an endorsement or a recommendation from SpringWorks for any center or physician.

ACTIVE SURVEILLANCE AND IDENTIFYING PROGRESSION

The first step for some people may be observation. Your doctor will closely monitor any desmoid tumors to determine if they are growing or changing. You can also keep track of your symptoms to help your care team see if any changes are needed in your care plan.

ACTIVE SURVEILLANCE

Active surveillance may include regular doctor check-ups and **frequent imaging tests**.[1] This will give doctors a chance to understand if the tumor is **progressing**: if you have growth or change in your tumor and/or symptoms.[3-5]

Progression is a word used to describe how a medical condition advances or gets worse.[6]

An increase in the size of the tumor, as seen in a CT scan or MRI, may be one way to see whether a desmoid tumor is progressing.[1,5]

MONITORING SYMPTOMS

Symptoms may also signal that a desmoid tumor is progressing. Consider tracking:

- Pain[8-10]
- Other symptoms[1,5]
- Impact on daily activities[1,5]

Proactively sharing any new or changing symptoms may help to better inform your care team. Your care team may recommend starting treatment if your tumor is causing symptoms that affect your daily life.[1]

If you have symptoms, the tumor grows, and/or the tumor is in a critical location, your doctor may recommend that you begin treatment.[1]

WITH YOUR DOCTOR AND CARE TEAM, YOU MAY ALSO NEED TO CONSIDER:

- What care approach is appropriate for you
- What potential side effects you may experience after receiving a treatment
- How desmoid tumor treatment may affect other medical conditions you are managing
- The likelihood of the tumor coming back even after treatments such as surgery, radiation therapy, cryoablation, or medications

ACTIVE TREATMENT OPTIONS FOR DESMOID TUMORS

Here's a quick overview of how desmoid tumors may be treated to help you understand what is involved with each option. You may want to bring this guide with you to your next appointment to help prepare.

Results and side effects can vary from person to person. It may be helpful to talk with your care team about what to expect—and together, come up with strategies to help handle potential side effects.

MEDICAL THERAPY

Medical therapy, also known as **systemic therapy**, describes treatment with medicines that travel through the bloodstream and can affect cells throughout the body.[10] **Medical therapy (systemic) is recommended as a first-line active treatment option** for desmoid tumors that are growing and/or symptomatic in most tumor locations.[1,11,*]

The NCCN Guidelines for Patients recommend the following medical therapy options:[1,11]

Type of drug	How it is given
Gamma secretase inhibitor (GSI)	Pill
Tyrosine kinase inhibitors (TKIs)	Pill
Chemotherapy	Infusion
Nonsteroidal anti-inflammatory drugs (NSAIDs, for pain)	Pill

SURGERY

Guidelines from 2 groups of desmoid tumor experts[†] **do not recommend surgery as the first option** for treating desmoid tumors (except in certain situations).[1,12] Some of the reasons that surgery is no longer recommended are:

- Surgery may require the removal of large amounts of tissue that could cause some loss of function or changes in appearance[4,13]
- The desmoid tumor itself can be far-reaching, so completely removing the desmoid tumor can be difficult[4,13]
- Desmoid tumors often come back after surgery (recurrence).[14] They can recur from 24% to 77% of the time[15,16]
- There is a likelihood of additional surgeries for people who have had desmoid tumor surgery[17,18]

RADIATION THERAPY

Radiation therapy is only recommended for desmoid tumors in the arms, legs, outer torso, head, and neck.[1] **It can be used when surgery would not be feasible and medical therapies are not available.**[5]

CRYOABLATION

Cryoablation uses extreme cold to destroy tumor tissue.[1,19] **It can be used for small to medium tumors that are not in the abdomen (belly).**[4]
A cold gas freezes the tumor tissue. It is pumped into the tumor through a thin needle. The tissue thaws and the freezing and thawing are repeated several times to destroy the cells.[19]

Your doctor may recommend other ablation therapies to destroy tumor tissue as well.[1]

*The NCCN Guidelines for Patients also recommend ablation and radiation therapy as treatment options for certain patients. Treatment choice will depend on the location of the tumor.[1]
†National Comprehensive Cancer Network® (NCCN®) and The Desmoid Tumor Working Group (DTWG).

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FINDING THE SUPPORT YOU NEED

Support may be available to you in many forms, such as through family and friends, or finding common ground with others living with desmoid tumors. Advocacy groups may also provide helpful information. The kind of support you need may change over time—and knowing where to turn for additional resources can make you feel more in control. The important thing to remember is that you don't have to go through the journey alone. Focus on staying centered, hopeful, connected, and taking action for your health.

Start by checking out these helpful sites:

ADVOCACY GROUPS*

The Desmoid Tumor Research Foundation (DTRF)

National Organization for Rare Disorders (NORD)

Rein in Sarcoma

Sarcoma Alliance for Research through Collaboration

Global Genes

NCCN Guidelines for Patients® Soft Tissue Sarcoma

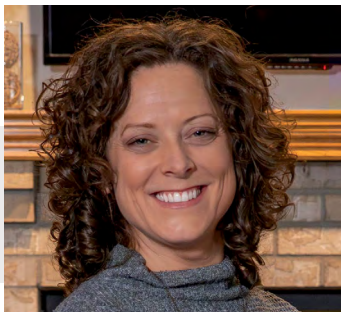
Northwest Sarcoma Foundation

Sarcoma Foundation of America

Information from these advocacy groups may be helpful but should not replace your doctor's advice. Always keep your doctor and care team informed.

FIND INFORMATION AND INSPIRING STORIES AT DESMOIDTUMORS.COM

*SpringWorks Therapeutics, Inc. is providing these links to help patients find more information about desmoid tumors, but inclusion on this list does not represent an endorsement or a recommendation from SpringWorks for any group or organization. The organizations listed are independent of SpringWorks Therapeutics.



When I meet someone who's newly diagnosed, it's important to know the resources available and to instill confidence in being able to advocate for themselves.

—Andrea, a real person living with a desmoid tumor

References: 1. Referenced with permission from the NCCN Guidelines for Patients® for Soft Tissue Sarcoma 2024. © National Comprehensive Cancer Network, Inc. 2024. All rights reserved. Accessed June 18, 2024. To view the most recent and complete version of the NCCN Guidelines for Patients, go online to NCCN.org/patientguidelines. NCCN makes no warranties of any kind whatsoever regarding their content, use, or application, and disclaims any responsibility for their application or use in any way. **2.** SARC (Sarcoma Alliance for Research through Collaboration). About SARC. Accessed June 18, 2024. https://sarctrials.org/about-sarc **3.** Kasper B, Baumgarten C, Bonvalot S, et al. Desmoid Working Group. Management of sporadic desmoid-type fibromatosis: a European consensus approach based on patients' and professionals' expertise—a sarcoma patients EuroNet and European Organisation for Research and Treatment of Cancer/Soft Tissue and Bone Sarcoma Group initiative. *Eur J Cancer.* 2015;51(2):127-136. **4.** 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. **5.** Kasper B, Baldini EH, Bonvalot S, et al; Desmoid Tumor Working Group. Current management of desmoid tumors: a review [supplementary online content]. *JAMA Oncol*. Accessed June 24, 2024. https://jamanetwork.com/journals/jamaoncology/article-abstract/2820212. **6.** NCI (National Cancer Institute). Dictionary of cancer terms: progression. Accessed June 18, 2024. https://www.cancer.gov/publications/dictionaries/cancer-terms/def/progression **7.** Cuomo P, Scoccianti G, Schiavo A, et al. Extra-abdominal desmoid tumor fibromatosis: a multicenter EMSOS study. *BMC Cancer.* 2021;21:437. **8.** Penel N, Bonvalot S, Bimbai AM, et al. Pain in desmoid fibromatosis (DF) patient: prevalence, associated factors and prognosis. Presented at the European Society of Medical Oncology Annual Meeting, 2022. Abstract 1512P. **9.** Quintini C, Ward G, Shatnawei A, et al. Mortality of intra-abdominal desmoid tumors in patients with familial adenomatous polyposis. *Ann Surg.* 2012;255:511-516. **10.** NCI (National Cancer Institute). Dictionary of cancer terms: systemic therapy. Accessed June 18, 2024. https://www.cancer.gov/publications/dictionaries/cancer-terms/def/systemic-therapy **11.** Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Soft Tissue Sarcoma V.2.2024. © National Comprehensive Cancer Network, Inc. 2024. All rights reserved. Accessed August 1, 2024. To view the most recent and complete version of the guideline, go online to NCCN.org. NCCN makes no warranties of any kind whatsoever regarding their content, use or application and disclaims any responsibility for their application or use in any way. **12.** Kasper B, Baldini EH, Bonvalot S, et al; Desmoid Tumor Working Group. Current management of desmoid tumors: a review. *JAMA Oncol*. Accessed June 24, 2024. **13.** Ballo MT, Zagars GK, Pollack A, Pisters PWT, Pollock RA. Desmoid tumor: prognostic factors and outcome after surgery, radiation therapy, or combined surgery and radiation therapy. *J Clin Oncol.* 1999;17(1):158-167. **14.** Penel N, Chibon F, Salas S. Adult desmoid tumors: biology, management and ongoing trials. *Curr Opin Oncol.* 2017;29(4):268-274. **15.** Easter DW, Halasz NA. Recent trends in the management of desmoid tumors. Summary of 19 cases and review of the literature. *Ann Surg.* 1989;210(6):765-769. **16.** Skubitz KM. Biology and treatment of aggressive fibromatosis or desmoid tumor. *Mayo Clin Proc.* 2017;92(6):947-964. **17.** Tsagozis P, Stevenson JD, Grimer R, Carter S. Outcome of surgery for primary and recurrent desmoid-type fibromatosis. A retrospective case series of 174 patients. *Ann Med Surg (Lond).* 2017;17:14-19. **18.** Fernandez M, Bell T, Tumminello B, Khan S, Zhou S, Oton A. Disease and economic burden of surgery in desmoid tumors: a review. *Expert Rev Pharmacoecon Outcomes Res*. 2023;23(6):607-618. doi: 10.1080/14737167.2023.2203915 **19.** NCI (National Cancer Institute). Dictionary of cancer terms: cryoablation. Accessed June 18, 2024. https://www.cancer.gov/publications/dictionaries/cancer-terms/def/cryoablation

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