



For people diagnosed with desmoid tumors **FINDING A WAY FORWARD WITH YOUR CARE**

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. There is a lot to learn, and a lot to consider. 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.



You and your care team

The doctor leading your care may be a medical oncologist, a physician who specializes in diagnosis and treatment of tumors. Expert doctors across different specialties can also be part of your care team. **A multidisciplinary team of doctors can share knowledge and work together to help determine your best care plan.**^{1,2}



Your care team may include:^{2,3}

- Medical oncologist
- Sarcoma specialist
- Surgeon/surgical oncologist
- Radiologist and/or radiation oncologist
- Primary care physician
- Pathologist
- Nurses
- Mental health professional

You may also want to check whether there is a sarcoma center near you. The physicians and teams there are highly experienced in the treatment of soft-tissue tumors. The Sarcoma Alliance for Research through Collaboration (SARC), a nonprofit organization, [has an online list of sarcoma centers.](#)^{4*}

*SpringWorks Therapeutics 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.

With desmoid tumors, making informed choices is important. Know that it's okay to ask questions and to speak up if there's something you don't fully understand. It's okay to seek clarity and share concerns—so you and your care team can make confident decisions together.

WHAT TO CONSIDER WHEN CHOOSING A CARE OPTION

Medical experts continue to study desmoid tumors to understand these rare tumors and how best to treat them. What we now know is that the way desmoid tumors progress can be different for each person. So, the approach to treatment has to be carefully planned. Any next steps to take may be influenced by:

Location

Where the tumor is located may influence which types of procedures or treatments are appropriate. For example, surgery is no longer recommended as a first approach for most locations.^{5,6}

Symptoms

A tumor causing symptoms that affect daily life may need to be treated sooner.⁷

Growth and risk over time

How slowly or quickly the tumor is growing and whether it poses any problems for surrounding structures (nerves, blood vessels) or organs in the body may affect the care team's recommendations.^{6,7}

If your symptoms return or start to get worse, or if you see that your tumor is growing, talk with your doctor.

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

- Whether treatment is appropriate at this time
- What approach to care you prefer (for example: watch and wait, oral medicine, or intravenous IV drugs, etc)
- What side effects to expect after receiving a treatment and how to manage them
- How desmoid tumor treatment may affect other medical conditions you are managing
- The likelihood of the tumor coming back even after surgery, radiation therapy, cryoablation, or medications

Think about these points—and openly discuss with your doctor any concerns you may have. **It's also 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. You may choose to take notes when speaking with your care team and refer back to the information later.



CURRENT CARE 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 print this out and bring it with you to your next appointment with your doctor to discuss which option may be right for you.

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 side effects if you experience them.



ACTIVE SURVEILLANCE

The first step for some people may be to “watch and wait.” This may include regular doctor check-ups and **frequent imaging tests.**⁷ This will give doctors a chance to understand if your symptoms are changing and see if or how fast the tumor is growing.^{6,8,9} **If the tumor grows quickly, symptoms get worse, or the tumor is in a critical location, your doctor may decide to begin treatment.**⁷



MEDICAL THERAPY

Currently, there are no FDA-approved treatment options **specifically for desmoid tumors.**¹⁰ However, your care team may prescribe medical therapies to help shrink and/or stabilize the tumor and manage symptoms. The recent clinical guidelines recommend the following treatment options:^{5,7}

- Tyrosine kinase inhibitors (TKIs, may include sorafenib, pazopanib, and imatinib)
- Chemotherapy (may include methotrexate and vinblastine/vinorelbine, doxorubicin-based regimens)
- Nonsteroidal anti-inflammatory drugs (NSAIDs, for pain)

Antihormonal therapies (tamoxifen or toremifene, with or without NSAIDs) do not have guideline recommendations.^{5,7}



SURGERY

The surgeon tries to remove the entire desmoid tumor and some surrounding healthy tissue to make sure they removed it all.⁵ **Surgery is generally no longer recommended as a first approach by desmoid tumor experts for most tumor locations.**^{5,6} The reasons may include:

- Surgery may require the removal of large amounts of tissue⁶ that could cause some loss of function or changes in appearance^{9,11}
- Desmoid tumors often come back after surgery (recurrence).¹² They can recur from 24% to 77% of the time^{13,14}
- Desmoid tumors may remain stable or resolve on their own⁶
- The desmoid tumor itself can be far-reaching, so completely removing the desmoid tumor can be difficult^{9,11}



RADIATION THERAPY

Radiation therapy is only recommended for desmoid tumors in the arms, legs, outer torso, head, and neck.^{3,7} **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.^{3,15} **It can be used for small to medium tumors that are not in the abdomen (belly).**⁶ 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.^{3,15}

CLINICAL TRIALS

There are other treatments being studied for desmoid tumors. Talk to your doctor to see if you may be a good candidate for a clinical trial.

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](#) ↗



[Global Genes Allies in Rare Disease](#) ↗



[Northwest Sarcoma Foundation](#) ↗



[Sarcoma Alliance for Research through Collaboration \(SARC\)](#) ↗

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

[desmoidtumors.com](https://www.desmoidtumors.com)

*SpringWorks Therapeutics 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.

HEAR FROM PATIENTS LIVING WITH DESMOID TUMORS

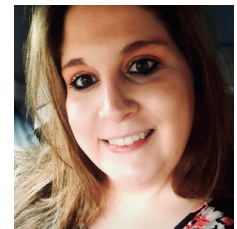
Join an upcoming Desmoid Tumor Ally Educational Program event.
Learn more and sign up at www.desmoidtumors.com/educational-program

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I had to teach myself to be my own advocate because nobody else could articulate my needs but me. I had to pull myself up by the bootstraps and start making my wishes known. I had to become a take-charge and self-reliant person.

—Stephanie, living with a desmoid tumor.

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References

1. Singhi E, Feldman J, West H. The multidisciplinary cancer conference. *JAMA Oncology*. Published online: December 22, 2022. doi:10.1001/jamaoncol.2022.4924
2. Cancer.Net. The oncology team. Accessed February 15, 2023. <https://www.cancer.net/navigating-cancer-care/cancer-basics/cancer-care-team/oncology-team>
3. Referenced with permission from the NCCN Guidelines for Patients® for Soft Tissue Sarcoma, V.2020. © National Comprehensive Cancer Network, Inc. 2020. All rights reserved. Accessed February 15, 2023. 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.
4. SARC (Sarcoma Alliance for Research through Collaboration). About SARC. Accessed February 15, 2023. <https://sarctrials.org/about-sarc>
5. Gronchi A, et al. Desmoid Tumor Working Group. The management of desmoid tumours: a joint global consensus-based guideline approach for adult and paediatric patients. *Eur J Cancer*. 2020;127:96-107.
6. 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.
7. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Soft Tissue Sarcoma V.1.2023. © National Comprehensive Cancer Network, Inc. 2023. All rights reserved. Accessed March 17, 2023. 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.
8. Cancer.Net. Desmoid tumor: types of treatment. Accessed February 15, 2023. <https://www.cancer.net/cancer-types/desmoid-tumor/types-treatment>
9. 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.
10. Riedel RF, Agulnik M. Evolving strategies for management of desmoid tumor. *Cancer*. 2022;128(16):3027-3040.
11. 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.
12. Penel N, Chibon F, Salas S. Adult desmoid tumors: biology, management and ongoing trials. *Curr Opin Oncol*. 2017;29(4):268-274.
13. 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.
14. Skubitz KM. Biology and treatment of aggressive fibromatosis or desmoid tumor. *Mayo Clin Proc*. 2017;92(6):947-964.
15. NCI (National Cancer Institute). Dictionary of cancer terms: cryoablation. Accessed February 15, 2023. <https://www.cancer.gov/publications/dictionaries/cancer-terms/def/cryoablation>