

# Rethinking Desmoid Tumors:

## MEDIA FACT SHEET

### About Desmoid Tumors

Desmoid tumors are rare, locally aggressive tumors that form in the connective tissues of the body.<sup>1,2</sup> Sometimes referred to as aggressive fibromatosis, or desmoid fibromatosis, desmoid tumors can cause severe pain, limited function and mobility, disfigurement, and compromised quality of life.<sup>1-6</sup> Desmoid tumors can be difficult to control and sometimes debilitating for people living with them.<sup>2-4</sup> They can have an unpredictable nature and high likelihood of recurrence and can have a significant impact on people's lives.<sup>4,7,8</sup> These invasive, tendrill-like growths can wrap around nearby tissue and compress vital organs, muscles, vessels, and nerves.<sup>2,3,9</sup> When vital organs are impacted, desmoid tumors can be life-threatening.<sup>2,9</sup>

### Quick Facts: Desmoid Tumors



Desmoid tumors are commonly diagnosed in people between 20 to 44 years of age.<sup>7,10</sup>



Women are 2 to 3 times more likely to be diagnosed than men.<sup>7,10</sup>



Recent pregnancy, injury, or surgery may increase the risk of developing desmoid tumors.<sup>11,12</sup>



Early and accurate diagnosis may help improve outcomes for people living with desmoid tumors.

- Approximately 30 to 40% of desmoid tumors are initially misdiagnosed, in part due to their rarity and similarities to other diseases.<sup>13</sup>
- More than half of people do not receive an accurate diagnosis for more than a year, which may prolong time living with pain and other debilitating symptoms and delay needed care.<sup>4,5,13,14</sup>



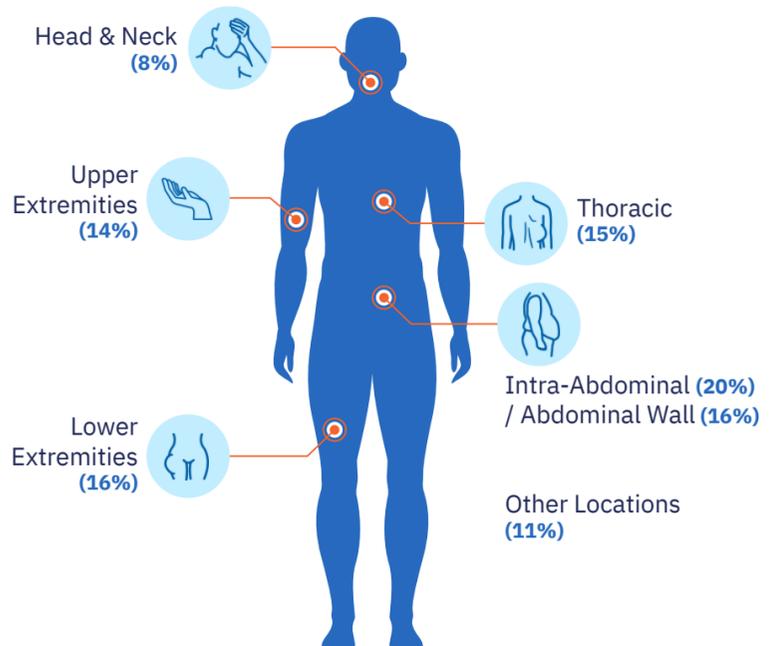
There are approximately 1,000 to 1,650 new cases of desmoid tumors diagnosed annually in the U.S.<sup>10,15,16</sup>



According to recent guidelines, people with desmoid tumors should be evaluated by a multi-disciplinary care team with experience managing desmoid tumors.<sup>17</sup>

### Desmoid Tumors Can Infiltrate Almost Any Part of the Body

While desmoid tumors can grow in almost any part of the body, the most common sites are:<sup>3,6,10,13</sup>



### Desmoid Tumor Symptoms Can Include<sup>3,5,6</sup>

- ▶ Pain
- ▶ Changes to body shape and physical function
- ▶ Limited range of motion
- ▶ Change in sleep
- ▶ Shortness of breath
- ▶ Fatigue

For some people, the pain and symptom burden can have psychological consequences, including:<sup>5,6</sup>

- ▶ Anxiety
- ▶ Depression
- ▶ Fear
- ▶ Worries about altered appearance
- ▶ Concerns about a lack of knowledge among healthcare professionals

### Treatment of Desmoid Tumors

- ▶ Desmoid tumor experts and guidelines now recommend systemic treatment as first-line intervention instead of surgery for most tumor locations requiring treatment.<sup>17,18</sup>
- ▶ Surgical resection is associated with increased rates of tumor recurrence, with up to 77% of patients experiencing recurrence following surgery.<sup>7,8\*</sup>

\*Based on retrospective, observational data. Factors associated with local recurrence postsurgery include tumor location, age of the participant, tumor size, margin status, and prior recurrence.<sup>19,20</sup>

For more information, visit

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

#### References

1. Sbaraglia M, Bellan E, Dei Tos AP. The 2020 WHO Classification of Soft Tissue Tumours: news and perspectives. *Pathologica*. 2021;113(2):70-84. doi:10.32074/1591-951X-213.
2. Penel N, Chibon F, Salas S. Adult desmoid tumors: biology, management and ongoing trials. *Curr Opin Oncol*. 2017;29(4):268-274. doi:10.1097/CCO.0000000000000374.
3. Constantinidou A, Scurr M, Judson I, Litchman C. Clinical presentation of desmoid tumors. In: Litchman C, ed. *Desmoid Tumors*. Springer; 2012:chap 2. Accessed August 2023. <https://www.researchgate.net/publication/226455135>. doi:10.1007/978-94-007-1685-8\_2.
4. Bektas, M, et al. Desmoid Tumors: A Comprehensive Review. *Adv Therapeutics* 2023. doi.org/10.1007/s12325-023-02592-0.
5. Husson O, Younger E, Dunlop A, et al. Desmoid fibromatosis through the patients' eyes: time to change the focus and organisation of care? *Support Care Cancer*. 2019;27(3):965-980. doi:10.1007/s00520-018-4386-8.
6. Gounder MM, Maddux L, Paty J, Atkinson TM. Prospective development of a patient-reported outcomes instrument for desmoid tumors or aggressive fibromatosis. *Cancer*. 2020;126(3):531-539. doi:10.1002/cncr.32555.
7. Skubitz KM. Biology and treatment of aggressive fibromatosis or desmoid tumor. *Mayo Clin Proc*. 2017;92(6):947-964. doi:10.1016/j.mayocp.2017.02.012.
8. 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. doi:10.1097/00000658-198912000-00012.
9. Joglekar SB, Rose PS, Sim F, Okuno S, Petersen I. Current perspectives on desmoid tumors: the Mayo Clinic approach. *Cancers (Basel)*. 2011;3(3):3143-3155. doi:10.3390/cancers3033143.
10. van Broekhoven DLM, Grünhagen DJ, den Bakker MA, van Dalen T, Verhoef C. Time trends in the incidence and treatment of extra-abdominal and abdominal aggressive fibromatosis: a population-based study. *Ann Surg Oncol*. 2015;22(9):2817-2823. doi:10.1245/s10434-015-4632-y.
11. Lopez R, Kemalyan N, Moseley HS, Dennis D, Vetto RM. Problems in diagnosis and management of desmoid tumors. *Am J Surg*. 1990;159(5):450-453. doi:10.1016/s0002-9610(05)81243-7.
12. Fiore M, Coppola S, Cannell AJ, et al. Desmoid-type fibromatosis and pregnancy: a multi-institutional analysis of recurrence and obstetric risk. *Ann Surg*. 2014;259(5):973-978. doi:10.1097/SLA.0000000000000224.
13. Kasper B, Baumgarten C, Garcia J, et al. Desmoid-type fibromatosis and pregnancy: a multi-institutional analysis of recurrence and obstetric risk. *Ann Surg*. 2014;259(5):973-978. doi:10.1097/SLA.0000000000000224.
14. Mercier KA, Hernandez L, Boulanger V, Seebald A, Rossow S, Milligan K. Quality of life and tumor location in patients with desmoid tumors: data from the desmoid tumor research foundation natural history study. Accessed August 2023. [https://ascopubs.org/doi/abs/10.1200/JCO.2019.37.15\\_suppl.e18291](https://ascopubs.org/doi/abs/10.1200/JCO.2019.37.15_suppl.e18291).
15. Orphanet Report Series: Rare Diseases collection. Prevalence and incidence of rare diseases: bibliographic data. Number 1, January 2022. Accessed August 2023. [https://www.orpha.net/Orpha.com/cahiers/docs/GB/Prevalence\\_of\\_rare\\_diseases\\_by\\_alphabetical\\_list.pdf](https://www.orpha.net/Orpha.com/cahiers/docs/GB/Prevalence_of_rare_diseases_by_alphabetical_list.pdf)
16. U.S. Department of Commerce. News Blog. U.S. population estimated at 332,403,650 on Jan. 1, 2022. Accessed August 2023. <https://www.commerce.gov/news/blog/2022/01/us-population-estimated-332403650-jan-1-2022#:~:>
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18. 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. doi:10.1016/j.ejca.2019.11.013.
19. Crago AM, Denton B, Salas S, et al. A prognostic nomogram for prediction of recurrence in desmoid fibromatosis. *Ann Surg*. 2013;258(2):347-353. doi: 10.1097/SLA.0b013e31828c8a30.
20. 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. doi:10.1016/j.amsu.2017.03.023.