Guideline Resource Unit
guru@ahs.ca

Desmoid Tumours

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Background

Desmoid tumours (aggressive fibromatosis, deep musculoaponeurotic fibromatosis, and formerly fibrosarcoma grade I of the desmoid type), are locally aggressive tumours which do not metastasize or differentiate. Desmoids are locally aggressive and have a high rate of recurrence even after complete resection. Tumour-related destruction of vital organs can be fatal, particularly in patients with familial adenomatous polyposis (familial adenomatous polyposis [FAP]; Gardner's syndrome).

Desmoid tumours are rare, accounting for approximately 0.03% of all neoplasms and less than 3% of soft tissue sarcomas (STS). The estimated incidence in the general population is 2-5 per million population per year.[1] Desmoids have no significant racial or ethnic predilection, occur most frequently in individuals between 15 and 60 years of age, and are more common in women than men.[2]

The risk of developing a desmoid is increased by approximately 850 times in patients with FAP versus the general population with the majority of desmoids being associated with the abdomen, although extremity desmoids can also occur.[3] Other risk factors include family history of desmoid tumour, pregnancy, and APC mutation (3' of codon 1444).[4, 5]

Guideline Questions

- What is the appropriate diagnosis and work-up strategies for patients with desmoids?
- For desmoid patients, when is Surgery/ Systemic Therapy/ Radiotherapy/ Local Ablative Therapies/ Active Surveillance appropriate and when are these treatment strategies contraindicated?
- What is the appropriate follow-up protocol for patients with desmoid tumours who have been treated or are on Active Surveillance?

Search Strategy

Relevant guidelines from the National Comprehensive Cancer Network (NCCN) and the Desmoid Tumour Working Group were considered in the development of our recommendations, as well as recommendations from recent comprehensive narrative reviews.[1, 6-10] In addition, PubMed was consulted throughout April and May of 2025 to identify any additional published research.

Target Population

Adult patients (18 years of age or older) who have been diagnosed with (or suspected) desmoid (aggressive fibromatosis) tumours.

Recommendations

Multidisciplinary care is essential for patients diagnosed with desmoid tumours at various stages including radiologists, surgeons, medical oncologists, radiation oncologists, and allied health professionals such as physical/message therapists, mental health specialists and health professionals focusing on adolescent and young adult (AYA) populations.

Diagnosis

1. The diagnosis of a desmoid tumour can only be established by histological and radiological examinations and should be confirmed by a pathologist with expertise in the area of Sarcoma. For a complete list of sarcoma biopsy recommendations, please refer to the Sarcoma Biopsy Guideline (www.ahs.ca/guru).

Staging (Evaluation/Workup)

2. Upon diagnosis (prior to initiation of therapy), all patients should be evaluated and managed by a multidisciplinary team with expertise and experience in sarcoma. Patients should have a history and physical including evaluation for Gardner's syndrome and FAP. Discussions on sigmoidoscopy/ colonoscopy or genetic counselling may be appropriate if an APC gene abnormality on biopsy. If molecular pathology is done on biopsy, those with CTNNB-1 abnormality do not need sigmoidoscopy/colonoscopy.
3. Desmoid tumours lack the propensity for regional or distant spread. Imaging of the primary site with CT or MRI are appropriate and staging for distant disease is generally not clinically indicated.
4. There is no commonly agreed upon staging system for desmoids. The American Joint Committee on Cancer (AJCC) TNM staging system specifically excludes desmoids.[11]
5. Desmoids are characterized by variable clinical behaviour. Some desmoids may grow progressively larger over time, however, it is common for desmoids tumours to experience indolent growth, or periods of growth arrest and spontaneous regression.[12-16]
6. Factors associated with increased recurrence rate include: site of disease (extremities having the worst prognosis, trunk/abdominal wall tumors have a lower rate of recurrence than either intraabdominal or extra-abdominal disease), age ≤30yr, an initial tumour size ≥5cm, and patients with an S45F mutation in CTNNB1 gene (vs other).[17-21]

Active Surveillance

7. Active Surveillance is increasingly being utilized as the initial treatment of choice (in line with recommendations from the Desmoid Tumour Working Group and NCCN),[6-8] especially for patients with at worse minimally symptomatic disease due to the fact that spontaneous growth arrest for desmoid tumour is a well-known phenomena through unknown mechanisms that are currently

under active investigation.[20, 22-25] Active Surveillance may not be appropriate if progression could be life-threatening or pose a risk for mutilation (adjacent nerves or vessels).

8. Patients should be monitored every 3-6mo initially, and if the disease is stable this can be extended.[6] Total duration should be determined through discussion between physician and patient. Spontaneous disease stabilization can take longer in patients <40yr and those with recurrent tumours.[26, 27]

Active Treatment

9. All treatment modalities beyond “wait and watch” approach in desmoid tumours can be considered in patients with progressive and/or symptomatic disease.[6] The treatment recommendation should be discussed in multidisciplinary rounds.

10. Nonsurgical approaches are feasible for patients with abdominal wall desmoid tumours <7cms, followed by surgery based on tumour growth in select cases.[13, 28] Treatment approach for mesenteric tumours should be discussed in multidisciplinary rounds.

11. Surgery can be considered as a local therapy option when complete resection with negative margins is technically feasible without undue morbidity and in symptomatic patients; however, RT may also be considered as control rates (85%) are similar to surgery (77-94%).[17, 29, 30]

Surgery

12. If R0 margins are achieved the patient should be observed.[6] If R1 margins, generally consider observation.[6] Note that a recurrence up to 25% is expected with a median time to recurrence of 16-22mo.[29, 31-34]

13. If R2 margins are achieved, multidisciplinary review and input is needed.

14. In case of progression or recurrence, reconsider all options including observation/ surveillance depending on symptoms, cryoablation, systemic therapy, resection, resection with adjuvant RT, or radical RT alone (56 Gy) if not previously irradiated; based on multidisciplinary input.[9, 17, 30]

Radiation Therapy

15. Radiotherapy is an effective treatment option, especially if patients are symptomatic and progressing.[30, 35] A dose of 50-56 Gy alone or in combination with surgery in patients with incomplete resection can achieve adequate local control at 5 years in 70–80% of patients with unresectable tumors or following incomplete resection.[7, 30, 36]

16. Post-operative RT after complete surgical resection is not typically recommended in the adjuvant setting. Utilization of RT in patients with close or positive margins is controversial, with some studies showing superior outcomes, while others fail to demonstrate any benefit of RT in this setting.[17, 33] Multidisciplinary review with input from radiation oncology is recommended for those with close or positive margins.

17. Neoadjuvant RT may be utilized to increase resectability and reduce rates of local recurrence in extra-abdominal desmoids, however, there is limited data to support this approach.[37] Currently, there is insufficient evidence to recommend neoadjuvant RT as standard of care. In select cases, it may be considered after multidisciplinary discussion.

18. For young adults, RT should be discussed individually with consideration of its late toxic effects and risk of secondary malignant neoplasms.[8, 9] Although controversial, there are some reports suggesting reduced efficacy of RT in young adult patients.[18] This should be taken into consideration when management options are being discussed.

Cryoablation

19. Cryotherapy may be considered in selected patients to achieve symptom relief and disease control through tumour stabilization or reduction.[38] Clinical benefit is most likely when at least 50% of the tumour volume is targeted, which may be difficult when the tumour is located near critical structures.[39] Tissue swelling is expected within the first 0–3 months, followed by a reduction in viable tumour volume.[40] Symptom relief, including pain reduction, typically occurs rapidly but can take several weeks. Repeat procedures may be required.

20. Cryotherapy is a highly specialized procedure and should only be performed by a well-trained expert interventional radiologist.

Systemic Therapy

21. Systemic therapy is typically reserved for those patients who have significant radiographic progression, multiple locoregional recurrences despite local therapy, symptomatic disease for which local therapy options are morbid, or intraabdominal/mesenteric desmoids tumors in patients with FAP.

22. The choice of agent largely depends on the urgency of the clinical situation (patient co-morbidities, functional status, and interpretation of risk-to-benefit ratio), where more aggressive options (such as doxorubicin based or combinational chemotherapy) should be reserved for patients with impending threat to life or function given shorter time to response, while less aggressive options (e.g. TKIs) should be used when systemic treatment is warranted without threat to life or function. Response rates are highest with anthracyclines and some targeted therapy (sorafenib, pazopanib and GSIs), and lower with single agent dacarbazine/ temozolomide or imatinib (Appendix A). Generally, those patients with non-limb disease, macroscopic nodular morphology or Gardner syndrome are associated with greater time to disease progression after systemic treatment.[41, 42]

    22.1. Several chemotherapy regimens demonstrate high disease control rates (>70%), including methotrexate-based and doxorubicin-based therapies.[42, 43] However, there are no randomized studies for any chemotherapy regimens (Appendix A). Response rate is generally higher with doxorubicin based regimens, and pegylated doxorubicin is preferred given less cardiac toxicity.[42]

22.2. Two targeted therapies (nirogacestat[44] and sorafenib[45]) have completed Phase III studies. Nirogacestat[i] significantly reduced desmoid related symptoms such as pain and have manageable side effect profile (nausea/diarrhea, hypophostemia and stomatitis). As these drugs are not publicly funded, access may be limited to compassionate use programs or private insurance. Selection should be guided by the toxicity profiles of individual agents and tailored to patient-specific factors, particularly in females of childbearing age as premature ovarian failure (ovarian toxicity) is a potential adverse effect.[8, 44] See Appendix B for targeted therapies under active investigation.

22.3. There is currently no clinical data supporting efficacy of hormonal therapy and NSAID treatment.[46, 47] Therefore, they have fallen out of the systemic treatment algorithm when other agents are accessible.[7]

23. Systemic treatment duration remains unknown for patients who have achieved response/stability. It is unclear whether treatment discontinuation is a safe strategy, and whether restarting systemic treatment upon disease progression can be potentially salvaged and achieve additional response/stability and long term favorable clinical outcome. Some studies have shed light on this issue and demonstrated that discontinuation of systemic treatments may be considered for patients who have achieved response.[48] There are insufficient data at this time to recommend discontinuation of therapy in patients with stable disease who are tolerating treatment well. Therefore, treatment discontinuation should be discussed carefully case by case according to response, tolerance, side effects, patients' preference, and quality of life.

24. Response to systemic therapy can be slow and may take many months to become apparent.[49, 50] Patients who exhibit a delayed response may even experience an initial increase in tumor size followed by an objective decrease in size.[49] Imaging is recommended every 2-3 cycles (or every 2-3 months). If the tumour is stable or has increased in size only minimally, then the response should not be considered treatment failure. Symptomatic or functional improvement warrants continued therapy even when changes to the tumour are not apparently on imaging.

Pregnancy

25. In pregnant women, desmoid tumours typically present as an abdominal wall mass which is separate from the uterus and is more common in pregnant woman with a history of desmoid tumors before conception.[51] A non-aggressive approach is advocated in women with a desmoid during pregnancy, as pregnancy-associated desmoids are typically associated with good outcomes and no obstetric complications.[51]

[i] Not approved by Health Canada

Pain Management

26. Patients with desmoids may have debilitating pain. Involvement (co-management) of a specialized pain clinic may be beneficial for the patient.

Rehabilitation

27. Patients can be referred to rehabilitation medicine (physical therapy, occupational therapy etc.) for functional and symptomatic assessment. When available, oncologic rehab/physiatry is can also be recommended.

Follow-Up

28. Patients should be monitored every 3-6 months initially, and if disease is stable this can be extended. Duration of follow-up thereafter will be subjected to the discussion between the physician and the patient.

Adolescent and Young Adult Patients

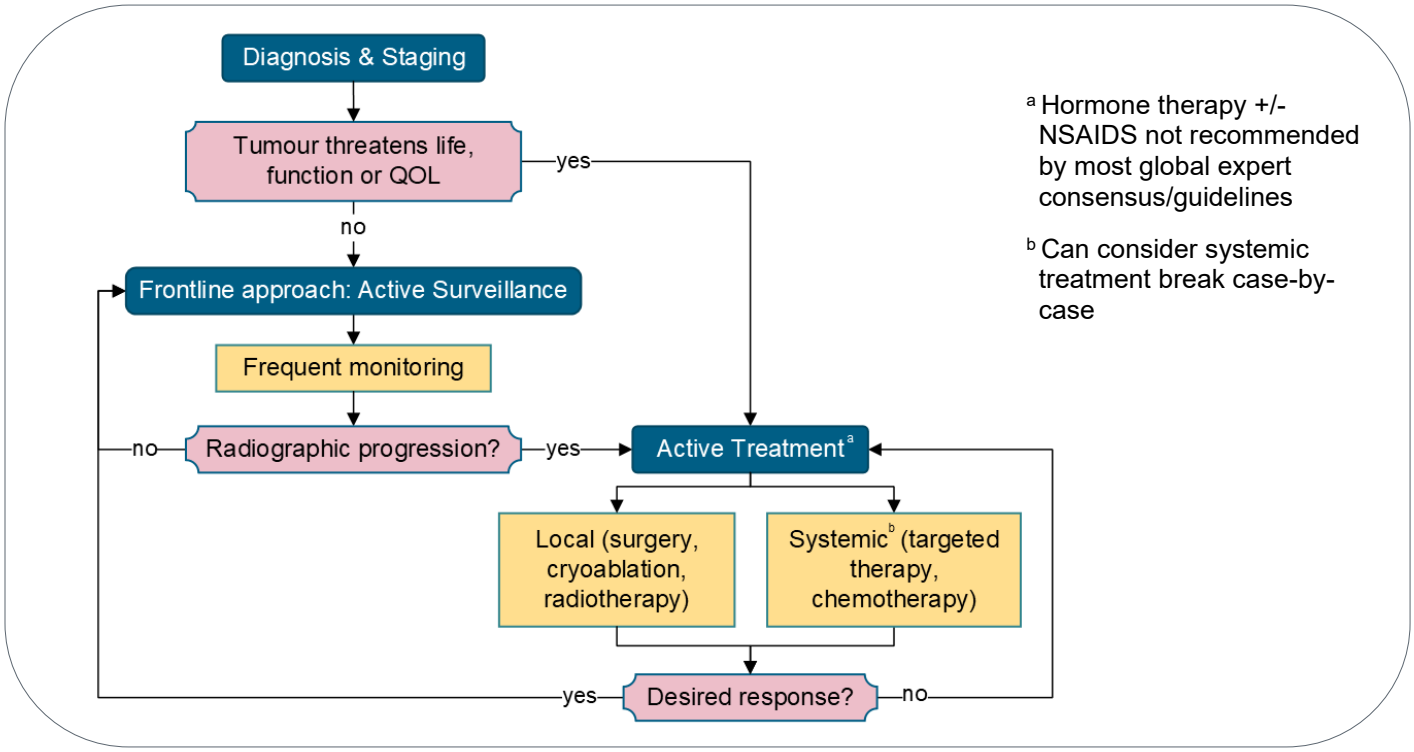
29. Given the age of diagnosis of patients with desmoid tumours, we recommend a referral to the AYA program where needed. Please refer to the Adolescent and Young Adult Guideline (www.ahs.ca/guru).

Patient Support

The Desmoid Tumor Research Foundation (DTRF) provides numerous resources for patients with desmoid tumours and continues to support desmoid tumour research (https://dtrf.org/).

Treatment Algorithm

Figure 1. Basic Treatment Flowchart for Patients Diagnosed with a Desmoid Tumour



Appendix A – Chemotherapy Treatment Regimes

Please note this is not an exhaustive list. Largely based on Tuskamoto et al. and Garbay et al.[42, 43]

Table 1. Chemotherapy Treatment Regimes for Desmoid Tumour

Agent(s)	Treatment Regime	n	Disease Control	Reference
Methotrexate (single and multi agent)	MTX 20mg/m^2 with VBL 6-10mg/m^2; 7d cycle	71	87%	Li 2017
	MTX 20-30mg/m^2 with VBL 3-6mg/m^2; 7d cycle	10	100%	Skapek 1998
	MTX 30mg/m^2 with VBL 6mg/m^2; 7d cycle	30	100%	Azzarelli 2001
	MTX 30mg/m^2 with VBL 5mg/m^2; 7-14d cycle	20	75%	Toulmonde 2019
	MTX 30mg/m^2 with VBL 6mg/m^2; 14-28d cycle	37	95%	Nishida 2020
	MTX 50mg/m^2 with VBL 10mg/m^2; 7-14d cycle	18	71%	Constantinidou 2011
	MTX 30mg/m^2 with VBL 6mg/m^2; 7d cycle	30	97%	Palassini 2017
	MTX 25mg/m^2 with VNR 25mg/m^2 (d1,8,15); 28d cycle	48	98%	Ingley 2019
	MTX 30mg/m^2 with VNR 20mg/m^2 or VBL 6mg/m^2; 7d cycle	37	95%	Napolitano 2020
	MTX 25-50mg/m^2 with VNR 10-20mg/m^2; 7d cycle	12	100%	Weis 1999
	MTX 50mg/m^2 with VNR 20mg/m^2; 7d cycle	73	100%	Palassini 2017
	MTX 30mg/m^2 (d1,18,15,21); 28d cycle	27	67%	Garbay 2012
	MTX 30mg/m^2 with VBL 6mg/m^2 (d1,18,15,21); 28d cycle			Garbay 2012
Anthracycline-based agents	DOX 20mg/m^2 with DTIC 150mg/m^2; 28d cycle	7	100%	Gega 2006
	DOX 60-90mg/m^2 with DTIC 1,000mg/m^2; 28d cycle	5	100%	Schnitzler 1997
	DOX 60-90mg/m^2 with DTIC 750-1,00mg/m^2	11	100%	Patel 1993
	DOX 60-90mg/m^2 (or CB 400rag/min) with DTIC 150mg/m^2; 28d cycle	8	100%	Poritz 2001
	DOX 60-75mg/m^2; 21d cycle	13	100%	Garbay 2012
	DOX 20mg/m^2 with DTIC 300mg/m^2 (d1-3); 21d cycle			Garbay 2012
	DOX 20mg/m^2 with IFOS 2.5g/m^2 and DTIC 300mg/m^2 (d1-3); 21d cycle			Garbay 2012
	Liposomal DOX 40-50mg/m^2; 28d cycle	10	90%	Bertagnolli 2008
	Liposomal DOX 20-50mg/m^2; 28d cycle	12	100%	Constantinidou 2011
	Liposomal DOX 32-40mg/m^2; 28d cycle	5	100%	Ananth 2017
Vinorelbine single agent	VNR 20-25mg/m^2; 7d cycle	2	100%	Palassini 2017
	VNR 20mg/m^2 (d1,8); 21d cycle	NR	NR	Garbay 2012
	VNR 90mg; 7d cycle	90	86%	Mir 2020
	Oral VNR 90mg (d1,8,15); 28d cycle	29	64%	Gennatas 2020

Other	Oral ETOP 75mg/day; 21-28d cycle	NR	NR	Garbay 2012
	Oral CCP 50mg/day; 21-28d cycle	NR	NR	Garbay 2012
	HU 20-30mg/kg/day	16	75%	Ferrari 2019
	CTX 600mg/m^2 with DOX 60mg/m^2; 21d cycle	1	100%	Okuno 2003
	IFOS 2,500mg/m^2 with ETOP 100mg/m^2 (d1-3); 21-28d cycle	4	100%	Okuno 2003
	IFOS 2,500mg/m^2 with ETOP 100mg/m^2 (d1-3); 21-28d cycle; *combined with* MMC 8mg/m^2 with DOX 40mg/m^2 and CP 50mg/m^2; 28d cycle	2	100%	Okuno 2003

CB, carboplatin; CCP, cyclophosphamide; CP, cisplatin; CYX, cyclophosphamide; DTIC, dacarbazine; DOX, docetaxel; ETOP, etoposide; *HU, hydroxyurea; IFOS, ifosfamide; MMC, mitomycin; MTX, methotrexate; NR, not reported; VBL, vinblastine; VNR, vinorelbine.*

Appendix B – Targeted Therapies in Clinical Trials

Please note this is not an exhaustive list. Adapted from Mangla et al.[52] and Timbergen et al.[53].

Table 2. Targeted Therapies for Desmoid Tumour in Clinical Trials

Type	Agent	Dose	Phase	n	Outcomes	References
Gamma-secretase inhibitors	AL102 (BMS-986115)	1.2mg once daily	2	42	Response rate of 50%	Gounder 2023 Phase 3: NCT04871282
Tyrosine kinase inhibitors	Pazopanib	800mg once daily	2	72	Response rate of 37% and 1-year PFS of 86%	Toulmonde 2019
	Imatinib	200mg – 800mg once daily	2	148	Inconsistent clinical efficacy. Response rate between 6-20% and a 1-year PFS of 37-66%	Heinrich 2006; Penel 2011; Chugh 2010; Kasper 2017
	Imatinib with vactoserib	IM: 400mg daily. VA: twice daily for 2d per week	2	27	Response rate of 70% and a 1-year PFS of 81%	Ahn 2024
	Imatinib with gemcitabine	IM: 300-400mg daily. GM: 700-800mg/m^2 weekly	1	1	Only patient with desmoid tumours withdrew due to dose-related toxicity	George 2006
	Nilotinib	800mg daily	2	8	Disease progression arrest rate of 88% in 3mo	Kasper 2017
	Sunitinib	37.5-52mg daily	2	51	Overall disease control rate of 68-75% and a 2-year PFS of 75-81	Jo 2014; Miano 2019
	Anlotinib	12mg daily	NA[1]	66	Objective response rate of 21%, a PFS of 90% (1-year) and 83% (2-year)	Xie 2024
WNT inhibitors	Tegavivint (BC2059)	5mg/kg IV once weekly	1	24	9-month PFS of 79%	Cranmer 2022
	E7386	120mg twice daily	1	2	Partial response	Kondo 2023
	Ipafricept (OMP-54F28)	Various doses	1	2	Stable disease (6mo)	Jimeno 2017
Other	Nab-paclitaxel	Weekly, 3 cycles	1	35	PFS of 85% (1-year) and 74% (2-year), and pain reduction in 90% of patients	Martin-Broto 2022

PFS, progression-free survival

[1] Retrospective cohort study

References

1. Bektas M, Bell T, Khan S, Tumminello B, Fernandez MM, Heyes C, et al. Desmoid Tumors: A Comprehensive Review. *Advances in Therapy*. 2023;40(9)
2. Mankin HJ, Hornicek FJ, Springfield DS. Extra-abdominal desmoid tumors: a report of 234 cases. *Journal of Surgical Oncology*. 2010;102(5):380-384.
3. Gurbuz AK, Giardiello FM, Petersen GM, Krush AJ, Offerhaus GJ, Booker SV, et al. Desmoid tumours in familial adenomatous polyposis. *Gut*. 1994;35(3):377-381.
4. Nieuwenhuis MH, Lefevre JH, Bülow S, Järvinen H, Bertario L, Kernéis S, et al. Family history, surgery, and APC mutation are risk factors for desmoid tumors in familial adenomatous polyposis: an international cohort study. *Diseases of the Colon and Rectum*. 2011;54(10):1229-1234.
5. Schiessling S, Kihm M, Ganschow P, Kadmon G, Büchler MW, Kadmon M. Desmoid tumour biology in patients with familial adenomatous polyposis coli. *The British Journal of Surgery*. 2013;100(5):694-703.
6. National Comprehensive Cancer Network NCCN Clinical Practice Guidelines in Oncology. Soft Tissue Sarcoma. Version 5.2024. 2025. Accessed April 28, 2025. https://www.nccn.org/professionals/physician_gls/pdf/sarcoma.pdf
7. Alman B, Attia S, Baumgarten C, Benson C, Blay J-Y, Bonvalot S, et al. The management of desmoid tumours: A joint global consensus-based guideline approach for adult and paediatric patients. *European Journal of Cancer*. 2020/03/01/ 2020;127:96-107.
8. Kasper B, Baldini EH, Bonvalot S, Callegaro D, Cardona K, Colombo C, et al. Current Management of Desmoid Tumors: A Review. *JAMA Oncology*. 2024;10(8):1121-1128.
9. Looi WS, Indelicato DJ, Rutenberg MS. The Role of Radiation Therapy for Symptomatic Desmoid Tumors. *Current Treatment Options in Oncology*. 2021/03/01 2021;22(4):34.
10. Ibrahim R, Assi T, Khoury R, Ngo C, Faron M, Verret B, et al. Desmoid-type fibromatosis: Current therapeutic strategies and future perspectives. *Cancer Treatment Reviews*. 2024/02/01/ 2024;123:102675.
11. AJJC. AJCC Staging Online - Soft Tissue Sarcoma. Accessed 2025/05/07,
12. Fiore M, Rimareix F, Mariani L, Domont J, Collini P, Le Péchoux C, et al. Desmoid-type fibromatosis: a front-line conservative approach to select patients for surgical treatment. *Annals of Surgical Oncology*. 2009;16(9):2587-2593.
13. Bonvalot S, Ternès N, Fiore M, Bitsakou G, Colombo C, Honoré C, et al. Spontaneous regression of primary abdominal wall desmoid tumors: more common than previously thought. *Annals of Surgical Oncology*. 2013;20(13):4096-4102.
14. Baumert BG, Spahr MO, Von Hochstetter A, Beauvois S, Landmann C, Fridrich K, et al. The impact of radiotherapy in the treatment of desmoid tumours. An international survey of 110 patients. A study of the Rare Cancer Network. *Radiation Oncology (London, England)*. 2007;2:12.
15. Burtenshaw SM, Cannell AJ, McAlister ED, Siddique S, Kandel R, Blackstein ME, et al. Toward Observation as First-line Management in Abdominal Desmoid Tumors. *Annals of Surgical Oncology*. 2016;23(7):2212-2219.
16. Salas S, Dufresne A, Bui B, Blay J-Y, Terrier P, Ranchere-Vince D, et al. Prognostic factors influencing progression-free survival determined from a series of sporadic desmoid tumors: a wait-and-see policy according to tumor presentation. *Journal of Clinical Oncology: Official Journal of the American Society of Clinical Oncology*. 2011;29(26):3553-3558.
17. Seinen J, Niebling M, Bastiaannet E, Pras B, Hoekstra H. Four different treatment strategies in aggressive fibromatosis: A systematic review. *Clinical and translational radiation oncology*. 2018;12
18. Bishop A, Zarzour M, Ratan R, Torres K, Feig B, Wang W, et al. Long-Term Outcomes for Patients With Desmoid Fibromatosis Treated With Radiation Therapy: A 10-Year Update and Re-evaluation of the Role of Radiation Therapy for Younger Patients. *International journal of radiation oncology, biology, physics*. 2019;103(5)
19. Crago AM, Denton B, Salas S, Dufresne A, Mezhir JJ, Hameed M, et al. A prognostic nomogram for prediction of recurrence in desmoid fibromatosis. *Annals of Surgery*. 2013;258(2):347-353.
20. Schut A-RW, Timbergen MJM, van Broekhoven DLM, van Dalen T, van Houdt WJ, Bonenkamp JJ, et al. A Nationwide Prospective Clinical Trial on Active Surveillance in Patients With Non-intraabdominal Desmoid-type Fibromatosis: The GRAFITI Trial. *Annals of Surgery*. 2023;277(4)
21. Cassidy MR, Lefkowitz RA, Long N, Qin L-X, Kirane A, Sbaity E, et al. Association of MRI T2 Signal Intensity With Desmoid Tumor Progression During Active Observation: A Retrospective Cohort Study. *Annals of Surgery*. 2020;271(4)
22. Timbergen MJM, Schut A-RW, Grünhagen DJ, Sleijfer S, Verhoef C. Active surveillance in desmoid-type fibromatosis: A systematic literature review. *European Journal of Cancer*. 2020/09/01/ 2020;137:18-29.

23. Penel N, Le Cesne A, Bonvalot S, Giraud A, Bompas E, Rios M, et al. Surgical versus non-surgical approach in primary desmoid-type fibromatosis patients: A nationwide prospective cohort from the French Sarcoma Group. *European Journal of Cancer*. 2017/09/01/ 2017;83:125-131.
24. Bonvalot S, Cozic N, Le Cesne A, Blay JY, Penel N, Fau M, et al. Initial Active Surveillance Strategy for Patients with Peripheral Sporadic Primary Desmoid-Type Fibromatosis: A Multicentric Phase II Observational Trial. *Annals of Surgical Oncology*. 2023/12/01 2023;30(13):8653-8659.
25. Colombo C, Fiore M, Grignani G, Tolomeo F, Merlini A, Palassini E, et al. A Prospective Observational Study of Active Surveillance in Primary Desmoid Fibromatosis. *Clinical Cancer Research*. 2022;28(18):4027-4032.
26. Kim Y, Rosario MS, Cho HS, Han I. Factors Associated with Disease Stabilization of Desmoid-Type Fibromatosis. *Clinics in Orthopedic Surgery*. 2020;12(1)
27. Barbier O, Anract P, Pluot E, Larouserie F, Sailhan F, Babinet A, Tomeno B. Primary or recurring extra-abdominal desmoid fibromatosis: Assessment of treatment by observation only. *Orthopaedics & Traumatology: Surgery & Research*. 2010;96(8)
28. van Houdt WJ, Husson O, Patel A, Jones RL, Smith MJF, Miah AB, et al. Outcome of Primary Desmoid Tumors at All Anatomic Locations Initially Managed with Active Surveillance. *Annals of Surgical Oncology*. 2019;26(13)
29. Timbergen MJM, Colombo C, Renckens M, Kim HS, Rosmalen Jv, Salas S, et al. The Prognostic Role of β-Catenin Mutations in Desmoid-type Fibromatosis Undergoing Resection Only: A Meta-analysis of Individual Patient Data. *Annals of Surgery*. 2021;273(6)
30. Keus RB, Nout RA, Blay JY, de Jong JM, Hennig I, Saran F, et al. Results of a phase II pilot study of moderate dose radiotherapy for inoperable desmoid-type fibromatosis--an EORTC STBSG and ROG study (EORTC 62991-22998). *Annals of Oncology: Official Journal of the European Society for Medical Oncology*. 2013;24(10):2672-2676.
31. Turner B, Alghamdi M, Henning J-W, Kurien E, Morris D, Bouchard-Fortier A, et al. Surgical excision versus observation as initial management of desmoid tumors: A population based study. *European Journal of Surgical Oncology*. 2019/04/01/ 2019;45(4):699-703.
32. Mueller C, Croner R, Klein P, Grützmann R, Vassos N. Primary and recurrent sporadic desmoids: Prognostic factors influencing recurrence-free survival after complete gross resection. *International Journal of Surgery*. 2016/07/01/ 2016;31:63-70.
33. Janssen ML, van Broekhoven DLM, Cates JMM, Bramer WM, Nuyttens JJ, Gronchi A, et al. Meta-analysis of the influence of surgical margin and adjuvant radiotherapy on local recurrence after resection of sporadic desmoid-type fibromatosis. *British Journal of Surgery*. 2017;104(4):347-357.
34. Meade A, Wisecarver I, Cho M-J, Davis JJ, Duque S, Farewell JT, Zhang AY. The Role of Reconstruction on Desmoid Tumor Recurrence: A Systematic Review and Technical Considerations. *Annals of Plastic Surgery*. 2023;90(5S)
35. Acker JC, Bossen EH, Halperin EC. The management of desmoid tumors. *International Journal of Radiation Oncology, Biology, Physics*. 1993;26(5):851-858.
36. Matsunobu T, Kunisada T, Ozaki T, Iwamoto Y, Yoshida M, Nishida Y. Definitive radiation therapy in patients with unresectable desmoid tumors: a systematic review. *Japanese Journal of Clinical Oncology*. 2020;50(5)
37. O'Dea FJ, Wunder J, Bell RS, Griffin AM, Catton C, O'Sullivan B. Preoperative radiotherapy is effective in the treatment of fibromatosis. *Clinical Orthopaedics and Related Research*. 2003;(415):19-24.
38. Taqi K, Stockley C, Wood M, Przybojewski S, Bouchard-Fortier A, Mack L. Cryotherapy in the Treatment of Extra-Abdominal Desmoid Tumors—A Review. *Current Oncology*. 2025;32(3):137.
39. Bouhamama A, Wdowik Q, Grillet F, Brahmi M, Sunyach MP, Vaz G, et al. Prognostic Factors for Local Recurrence after Cryoablation of Desmoid Tumors. *Journal of vascular and interventional radiology*. 2023;34(9):1538-1546.
40. Yan YY, Walsh JP, Munk PL, Mallinson PI, Simmons C, Clarkson PW, et al. A Single-Center 10-Year Retrospective Analysis of Cryoablation for the Management of Desmoid Tumors. *Journal of vascular and interventional radiology*. 2021;32(9):1277-1287.
41. de Camargo VP, Keohan ML, D'Adamo DR, Antonescu CR, Brennan MF, Singer S, et al. Clinical outcomes of systemic therapy for patients with deep fibromatosis (desmoid tumor). *Cancer*. 2010;116(9):2258-2265.
42. Garbay D, Le Cesne A, Penel N, Chevreau C, Marec-Berard P, Blay JY, et al. Chemotherapy in patients with desmoid tumors: a study from the French Sarcoma Group (FSG). *Annals of Oncology: Official Journal of the European Society for Medical Oncology*. 2012;23(1):182-186.
43. Tsukamoto S, Takahama T, Mavrogenis AF, Tanaka Y, Tanaka Y, Errani C, et al. Clinical outcomes of medical treatments for progressive desmoid tumors following active surveillance: a systematic review. *Musculoskeletal Surgery*. 2022;107(1)
44. Gounder M, Ratan R, Alcindor T, Schöffski P, van der Graaf WT, Wilky BA, et al. Nirogacestat, a γ-Secretase Inhibitor for Desmoid Tumors. *New England Journal of Medicine*. 2023;388(10)

45. Gounder MM, Mahoney MR, Van Tine BA, Ravi V, Attia S, Deshpande HA, et al. Sorafenib for Advanced and Refractory Desmoid Tumors. *New England Journal of Medicine*. 2018;379(25)
46. Pannier D, Cordoba A, Ryckewaert T, Robin Y-M, Penel N. Hormonal therapies in uterine sarcomas, aggressive angiomyxoma, and desmoid-type fibromatosis. *Critical Reviews in Oncology/Hematology*. 2019/11/01/ 2019;143:62-66.
47. Cho JY, Gupta S, Cho HS, Park MS, Mok SJ, Han I, Kim H-S. Role of Nonsteroidal Anti-inflammatory Drug in Treatment of Extra-abdominal Desmoid Tumors. *Clinics in Orthopedic Surgery*. 2018;10(2)
48. Noujaim J, Gupta A, Holloway C, Saleh R, Srikanthan A, Lemieux C, et al. Real-word experience of pazopanib and sorafenib in patients with desmoid tumors: A CanSaRCC multi-center study. *European journal of cancer*. 2024;205
49. Tsukada K, Church JM, Jagelman DG, Fazio VW, McGannon E, George CR, et al. Noncytotoxic drug therapy for intra-abdominal desmoid tumor in patients with familial adenomatous polyposis. *Diseases of the Colon and Rectum*. 1992;35(1):29-33.
50. Reich S, Overberg-Schmidt US, Bührer C, Henze G. Low-dose chemotherapy with vinblastine and methotrexate in childhood desmoid tumors. *Journal of Clinical Oncology: Official Journal of the American Society of Clinical Oncology*. 1999;17(3):1086.
51. Fiore M, Coppola S, Cannell AJ, Colombo C, Bertagnolli MM, George S, et al. Desmoid-type fibromatosis and pregnancy: a multi-institutional analysis of recurrence and obstetric risk. *Annals of Surgery*. 2014;259(5):973-978.
52. Mangla A, Agarwal N, Schwartz G, Mangla A, Agarwal N, Schwartz G. Desmoid Tumors: Current Perspective and Treatment. *Current Treatment Options in Oncology*. 2024;25(2)
53. Timbergen MJM, Smits R, Grünhagen DJ, Verhoef C, Sleijfer S, Wiemer EAC. Activated Signaling Pathways and Targeted Therapies in Desmoid-Type Fibromatosis: A Literature Review. *Frontiers in Oncology*. 2019;9

Development and Revision History

This guideline was developed by a multidisciplinary working group comprised of members from the Alberta Provincial Sarcoma Tumour Team, external participants identified by the Working Group Lead, and a methodologist from the Guideline Resource Unit. The draft guideline was externally reviewed and endorsed by members of the Alberta Provincial Sarcoma Tumour Team who were not involved in the guideline's development, including surgical oncologists, radiation oncologists, medical oncologists, nurses, pathologists, and pharmacists. A detailed description of the methodology followed during the guideline development process can be found in the Guideline Resource Unit Handbook.

This guideline was originally developed in 2017 and updated in 2025.

Maintenance

A formal review of the guideline will be conducted at the Annual Provincial Meeting in 2027. If critical new evidence is brought forward before that time, however, the guideline working group members will revise and update the document accordingly.

Abbreviations

AYA, Adolescents and Young Adults; CT, Computed Tomography; FAP, Familial adenomatous polyposis; GSI, Gamma-secretase inhibitor; MRI, Magnetic Resonance Imaging; NCCN, National Comprehensive Cancer Network; NSAID, Nonsteroidal anti-inflammatory drugs; QOL, Quality of Life; RT, Radiotherapy; STS, Soft tissue sarcoma.

Disclaimer

The recommendations contained in this guideline are a consensus of the Alberta Provincial Tumour Team and are a synthesis of currently accepted approaches to management, derived from a review of relevant scientific literature. Clinicians applying these guidelines should, in consultation with the patient, use independent medical judgment in the context of individual clinical circumstances to direct care.

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Conflict of Interest Statements

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