Expert opinions

on current desmoid tumour treatments and guidelines

Insights from two renowned experts on the clinical management of DT,

Professor Bernd Kasper from Germany and Professor Alessandro Gronchi from Italy, were captured during an "Ask Your Expert" session at a recent international oncology conference. This session focused on current clinical recommendations for managing DT according to updated guidelines from the international DT Working Group (DTWG).¹²







Professor Bernd Kasper, MD, PhD

Treatment

Diagnosis using core needle biopsy performed by in soft tissue tumours

*Persistent tumour growth was ascertained by observing tumour size increase at multiple (>2) consecutive follow-up appointments, threatening quality of life (QOL), life, or function or posing a potential risk to the feasibility or effectiveness of first-line treatment.

Reproduced with permission from Kasper et al. JAMA Oncol 2024; 10:1121-1128.

Patient referral to sarcoma

NO

NO

Tumour became

Persistent tumour

NO

Key Points:



Patient Evaluation: Suspected DT should be assessed using imaging, histopathologic biopsy analysis and, whenever possible, testing for associated gene mutations.²



Disease Management: After diagnosis and specialist referral, individualised care should be provided by multidisciplinary teams that may include medical oncologists, radiologists, oncology surgeons, pain specialists, and, where necessary, physiotherapists, psychologists and social workers.¹²



Active Surveillance: Experts from the DTWG and the US National Comprehensive Cancer Network (NCCN) advocate for active surveillance as the first-line management approach in evidence-based treatment guidelines. 12

· Monitoring: Signs of tumour progression, patient symptoms, and quality of life (QoL) should be monitored during active surveillance.



Active Treatment: Individualised treatment — usually systemic therapy, surgical intervention, or both — should be initiated by the multidisciplinary team in consultation with the patient if tumour progression or worsened DT

- Systemic therapies: Medications administered orally or otherwise to reach the treatment target through the bloodstream are the recommended first-line active treatment option for most DT patients but currently, no systemic therapies have regulatory approval for the treatment of DT in Europe.
- Surgery: Surgery is not generally advocated as a first-line treatment in DT, except for potentially life-threatening tumours or abdominal tumours with complications (e.g., gastrointestinal bleeding or obstruction).
- Locoregional Therapy: Treatments such as cryoablation (using extreme cold to destroy tumour tissue) or focal radiotherapy (delivering precise radiation doses directly to tumour tissue) can be applied in certain cases.



Systemic Therapy Classes:

1.Low-dose chemotherapy.4-8

- 2.Tyrosine kinase inhibitors (TKIs).9-11
- 3.Gamma-secretase inhibitors (GSIs). 12-14
- Treatment considerations: Low-dose chemotherapy may be used for rapidly growing, debilitating, or life-threatening DT, and TKIs have shown some clinical efficacy in progressive DT; toxicity and tolerability concerns should be taken into account for long-term use with all three therapy classes.¹⁹



References: 1. Kasper B, Baldini, EH, Bonvalot S et al. Current management of desmoid tumors: a review. JAMA Oncol. 2024;10(8):1121-1128. 2. The Demoid Tumour

References: 1. Kasper B, Baldini, EH, Bonvalot S et al. Current management of desmoid tumors: a review. JAMA Oncol. 2024;10(8):1121-1128. 2. The Demoid Tumour Working Group (DTWG). The management of desmoid tumours: a joint global consensus-based guideline approach for adult and paediatric patients. Eur J Cancer. 2020;29:64-07.3. Bektas M, Bell T, Khan S et al. Desmoid tumors: a comprehensive review. Adv Ther. 2023;40(9):3697-3722. 4. Constantinidou A, Jones RL, Scurr M et al. Advanced aggressive fibromatosis: effective palliation with chemotherapy. Acta Oncol. 2011;50(3):455-461. 5. Constantinidou A, Jones RL, Scurr M et al. Advanced aggressive fibromatosis: effective palliation with chemotherapy. Acta Oncol. 2011;50(3):455-461. 5. Constantinidou A, Jones RL, Scurr M et al. Advanced aggressive fibromatosis: effective palliation with chemotherapy. Acta Oncol. 2011;50(3):455-461. 5. Constantinidou A, Jones RL, Scurr M et al. Advanced aggressive fibromatosis: effective palliation with chemotherapy for patients with deep fibromatosis: Eur J Cancer. 2010;16(9):2258-2265. 7. Skapek SX, Ferguson WS, Granowetter L et al. Vinblastine and methotrevate for desmoid fibromatosis in children: results of a Pediatric Oncology Group phase II trial. J Clin Oncol. 2007;25(5):501-506. 8. Palassini E, Frezza AM, Mariani L et al. Long-term efficacy of methotrevate plus vinblastine/vinoralbine in a large series of patients affected by desmoid-type fibromatosis. Cancer. J 2017;23(2):86-19. 8. Kasper B, Raut CP, Gronchi A. Desmoid tumors: to treat or not to treat, that is the question. Cancer. 2020;126(24):5213-5221. 10. Toulmonde M, Pulidol M, Ray-Coquard I et al. Pazopanib or methotrexate-vinblastine combination chemotherapy in adult patients with progressive desmoid tumors. N Engl J Med. 2018;379(5):2417-2428. 12. Counder M of T et al. Nirogacestata, excertase inhibitor for desmoid tumors. MEngl J Med. 2018;379(5):2417-2428. 12. Counder M of T et al. Nirogacestata, excertase inhibitor for desmoid tumors. Mengl J Med. 2018;379(5):24