

NCCN Continuing Education

Target Audience: This journal article is designed to meet the educational needs of oncologists, nurses, pharmacists, and other healthcare professionals who manage patients with cancer.

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Release date: December 10, 2025; Expiration date: December 10, 2026

Learning Objectives:

Upon completion of this activity, participants will be able to:

- Integrate into professional practice the updates to the NCCN Guidelines for Soft Tissue Sarcoma
- Describe the rationale behind the decision-making process for developing the NCCN Guidelines for Soft Tissue Sarcoma

Disclosure of Relevant Financial Relationships

None of the planners for this educational activity have relevant financial relationship(s) to disclose with ineligible companies whose primary business is producing, marketing, selling, re-selling, or distributing healthcare products used by or on patients.

Individuals Who Provided Content Development and/or Authorship Assistance:

The faculty listed below have no relevant financial relationship(s) with ineligible companies to disclose.

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The faculty listed below have the following relevant financial relationship(s) with ineligible companies to disclose. All of the relevant financial relationships listed for these individuals have been mitigated.

Margaret von Mehren, MD, Panel Chair, has disclosed receiving grant/research support from Cogent Biosciences, Inc., Deciphera Pharmaceuticals, LLC, IDRx, Inc., and Novartis Pharmaceuticals Corporation; and serving as a scientific advisor for Boehringer Ingelheim International GmbH and Deciphera Pharmaceuticals, LLC.

Richard F. Riedel, MD, Panel Member, has disclosed receiving grant/research support from Aadi Bioscience, AROG Pharmaceuticals, Inc., Ayala Pharmaceuticals, Inc., BioAtla, Inc., Cogent Biosciences, Inc., Daiichi Sankyo, Inc., GSK plc, Immune Design, Inhibrx Biosciences, Inc., Intensity Therapeutics, Karyopharm Therapeutics Inc., Oncternal Therapeutics, PTC Therapeutics, Servier Laboratories, SpringWorks Therapeutics, Inc., SynOx Therapeutics Ltd, and TRACON Pharmaceuticals, Inc.; and receiving consulting fees from Aadi Bioscience, Blueprint Medicines Corporation, Daiichi Sankyo, Inc., Deciphera Pharmaceuticals, LLC, GSK plc, and SpringWorks Therapeutics, Inc.

To view disclosures of external relationships for the NCCN Guidelines panel, go to [NCCN.org/guidelines/guidelines-panels-and-disclosure/disclosure-panels](https://www.nccn.org/guidelines/guidelines-panels-and-disclosure/disclosure-panels)

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Soft Tissue Sarcoma, Version 1.2025

Featured Updates to the NCCN Guidelines

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Abstract

The treatment landscape for soft tissue sarcomas (STS) is evolving constantly. These NCCN Guidelines Insights focus on recent updates to the NCCN Guidelines for STS specific to surgical management and systemic therapy recommendations for various subtypes.

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Overview

Soft tissue sarcomas (STS) are rare tumors of mesodermal origin that can arise in various tissues of the body. There are >70 subtypes of STS, each with distinct clinical behaviors, pathology, and responses to treatment.¹ STS are considered some of the most challenging tumors to diagnose and treat, because their rarity and morphologic heterogeneity significantly affect diagnostic accuracy; therefore, expertise requires access to large number of cases. Given the heterogeneity of these tumors, therapeutic strategies have evolved from a one-size-fits-all model to histology-specific approaches.²

The NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for STS provide up-to-date recommendations for the clinical management of patients with STS of the extremities, body wall, head and neck, retroperitoneum, and specific histologic subtypes, including desmoid tumors (aggressive fibromatoses), rhabdomyosarcoma, and atypical lipomatous tumor or well-differentiated liposarcoma. These guidelines have been developed and updated by a multidisciplinary panel of representatives from NCCN Member Institutions with STS-focused expertise in the fields of medical oncology, surgical oncology, radiation oncology, hematology, diagnostic and interventional

radiology, pathology, orthopedic oncology, pediatric oncology, and patient advocacy.

In recent updates to the NCCN Guidelines for STS, the panel revised recommendations for the treatment and follow-up of several STS subtypes. These NCCN Guidelines Insights focus on updates related to the surgical management of desmoid tumors and systemic therapy recommendations for leiomyosarcoma (LMS), synovial sarcoma, alveolar soft part sarcoma (ASPS), and dedifferentiated liposarcoma (DDLPS).

Role of Surgery in the Treatment of Desmoid Tumors

Desmoid tumors are rare, potentially locally invasive lesions characterized by fibroblastic proliferation that can result in significant functional morbidity. The panel noted that treatment modalities for desmoid tumors have evolved over the past 10 years. Historically, surgery was considered the primary treatment option for most cases. However, several studies have reported morbidity as well as high rates of local recurrence associated with resection, depending on the location and size of tumor and other factors.^{2–5} Accordingly, a paradigm shift toward less-invasive management has occurred over the past decade.

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*Provided content development and/or authorship assistance.

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