BACKGROUND AND OBJECTIVES

- Desmoid tumors (DT) or aggressive fibromatoses, are soft-tissue tumors with locally aggressive, infiltrative, and destructive growth. DT can arise in extra-abdominal locations, chest wall, head and neck, or involve deep-seated tumors during or following pregnancy, and intra-abdominal (either a primary or metastatic location) tumors.
- DT have a broad, negative impact on patients’ lives. Patients experience symptoms of pain and impaired mobility, which limit their everyday lives and can lead to depression in physical, social, and psychological domains.
- Treatment goals should not focus solely on clinical measures such as progression-free survival but also consider patient-relevant endpoints such as reduction in DT-specific symptom burden (e.g., pain) and DT-related complications such as functional limitation.
- Surgery remains the primary treatment for DT, but surgery can have high dependence on age, tumor extent of involvement, complexity of the case, and other factors.
- The functional impairment and high local recurrence rates observed after surgery for DT have contributed to a paradigm shift in the last 2 decades toward more conservative management.
- The aim of the research was to assess the burden of surgery in DT patients with DT, focusing on recurrence rates and functional outcomes resulting from procedures such as limb amputation. Additionally, the cost of surgery in resectable cases (2017 US dollars) and the overall cost of care was assessed to estimate the cost of surgery in DT.
- Based on feedback from several surgical oncology experts in the United States and European Union, STS may be considered a valid endpoint to estimate the costs of surgery in DT. DTs are costs of surgery that are frequently associated with significant functional loss or amputation of the involved limb.
- The average number of surgical procedures in patients with DT who were initially treated surgically was higher than previously reported.

METHODS

- To identify publications written in the English language describing recurrence rates and functional outcomes after surgery in patients with DT, a systematic review was conducted from November 2011 to November 2021 and updated in December 2022.
- During the systematic review, the reference lists of all identified articles were searched.
- Inclusion criteria were: articles published in English, including those with abstracts written in English; articles reporting on DT patients (patients with DT who have undergone surgery for their disease); and articles reporting on functional outcomes and recurrence rates.
- Exclusion criteria were: articles not written in English, articles not reporting on DT patients, and articles not reporting on functional outcomes and recurrence rates.

RESULTS

- Tumor location
- China: 49.4%
- Netherlands: 24.2%
- Extra-A: 100%
- Extra-abdominal surgery: 100%

TRENDS IN SURGICAL INTERVENTIONS

- Guidelines from the National Comprehensive Cancer Network, version 1.2023 recommend continuous surveillance with functional outcomes for all patients with DT.
- Surgery is recommended by the Desmoid Tumor Working Group for abdominal wall tumors and extra-abdominal tumors.
- Given the evidence that a cut-off for surveillance is associated with better long-term survival, surgical resection is recommended for all patients with DT. However, many patients with DT, due to surgical or clinical management that has decreased in the past 2 decades, may have undergone amputation instead of surgery.
- Figure 4: Changes in the Percentage of DT Cases Treated With Surgery as Initial Treatment

COSTS OF STS SURGERIES AND AMPUTATIONS

- Among 4 US studies reporting cost of surgery in patients with STS, cost of surgery ranged from $18,151 to $305,200.
- In 2012, the average cost of surgery for breast cancer was $28,371 in patients with STS, whereas costs of $92,588 and $107,819 were estimated for amputations below and above the knee, respectively.
- The estimated lifetime, all-cause (including any condition being treated) direct cost of surgery ranged from $92,588 in patients with coronary artery disease to $172,751 in patients with peripheral artery disease.
- Costs depend on the timing of surgery, for example, with amputations for non-traumatic amputations, total knee arthroplasty, and breast cancer patients, the average estimated cost was $29,235, $32,420, and $24,299, respectively.

CONCLUSION

- Surgery is no longer recommended as first-line treatment in most clinical situations of DT, as it is not associated with better long-term functional outcomes.
- Alternative treatments with improved long-term functional outcomes, such as selective resection and improved surgical techniques, are compared with current treatment options.
- Surgery is no longer recommended as first-line treatment in most clinical situations of DT, as it is not associated with better long-term functional outcomes.

LIMITATIONS

- Results may have been biased due to limiting the search to the last 10 years and not publishing in English.
- Two studies were available as abstracts and the functional outcomes were included in the systematic review; and in 2 studies, variables including patient characteristics and technical outcomes were reported. The authors included complete data for each study.

Burden of Surgery in Desmoid Tumors

María M Fernandez, PhD, MBA1; Timothy Bell, MHA2; Brad Tumminello, PharmD3; Shahnaz Khan, MPH4; Shengfan Zhou, MS5; Meryem Bektas, PhD, MBA6; Ana B. Oton, MD1

1RTI Health Solutions, Research Triangle Park, NC, United States; 2SpringWorks Therapeutics, Inc, Stamford, United States

Table 1: Postoperative Recurrence Rates in Patients With DT Reported in Retrospective Studies

<table>
<thead>
<tr>
<th>Reference</th>
<th>Year</th>
<th>Patient characteristics</th>
<th>Tumor location</th>
<th>Recurrence rates</th>
<th>3-year survival</th>
<th>5-year survival</th>
<th>10-year survival</th>
<th>10-year survival (2017 US dollars)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peng et al.</td>
<td>2008</td>
<td>100 patients were treated, but data on recurrences are available for only 92 patients</td>
<td>Extra-abdominal (14%)</td>
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<td>$30,200</td>
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1-3
23
33
AW = abdominal wall; EFS = event-free survival; extra-A = extra-abdominal; FAP = familial adenomatous polyposis; intra-A = intra-abdominal; NR = not reported; RFS = recurrence-specific survival.