

Guidance for the Management of Gynecological Sarcomas in Ontario: Recommendations Report

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**Ontario
Health**

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Background

Sarcomas are rare malignancies that arise in soft tissue and bone. They affect all age groups and may arise in any part of the body. Treatment is often multimodal and complex. Individuals with sarcoma frequently experience significant morbidity and mortality as a consequence of the disease and/or treatment (1,2). The recognition that sarcoma care is complex and multidisciplinary has led to the adoption of evidence-based policies in Ontario that concentrate sarcoma care in designated sarcoma centres of excellence (3). This provides patients with access to highly specialized medical expertise related to pathology, imaging, surgery, radiation, and systemic therapy required to achieve the best patient outcomes.

Pelvic sarcomas may arise directly in the female reproductive organs or in other pelvic tissues involving the reproductive organs secondarily. Sarcomas arising in female reproductive organs are referred to as gynecologic sarcomas. Ontario Health (Cancer Care Ontario) data show that in the period from 2010 to 2013, approximately 90 Ontarians were diagnosed annually with gynecologic sarcoma. This was about seven percent of all adult Ontario sarcoma cases, diagnosed during the same period (Data source: Ontario Cancer Registry (OCR). Date: July 15, 2015).

Improving accurate pre-operative recognition of primary and secondary pelvic sarcomas involving the female reproductive organs may help with the identification of gynecologic sarcomas. This is particularly important because these are frequently mistaken pre-operatively for more typical gynecologic tumours, potentially leading to inappropriate or suboptimal interventions. This is of additional concern for locally advanced tumours. For example, differentiating between fibroids (leiomyomas) and leiomyosarcomas. The clinical presentation of these two conditions is comparable, yet, the recommended treatment and prognosis differ greatly (4,5).

Rationale and Scope

Female benign pelvic tumours are frequently initially managed by general gynecologists in the community and the standard approach to investigation and management of typical uterine masses is well-established (6). However, system-level recommendations for the diagnosis and management of primary or secondary sarcomas involving female reproductive organs are lacking (7,8).

Due to the unique management challenges posed by gynecologic sarcomas in Ontario, they were not included in the Provincial Sarcoma Services Plan released in 2014 (9). Since this time, centres that provide gynecologic sarcoma and non-gynecologic sarcoma services consult each other in order to provide care for patients with pelvic sarcomas. This includes the review of cases at both sarcoma and gynecologic oncology multidisciplinary cancer conferences (MCCs) and cross-service/discipline consultations. There is a need to formalize these processes and optimize collaboration between specialties to ensure access to high-quality care for patients with gynecologic sarcomas.

The intent of this Recommendations Report is to:

- Align Ontario Health (Cancer Care Ontario)'s Sarcoma Services Host sites, Gynecologic Oncology Centres (GOCs) and general gynecologists in the care of gynecologic sarcomas,
- Improve awareness about the optimal care of pelvic sarcomas amongst general gynecologists and pathologists,
- Guide the investigation and management of suspected pelvic and extra-pelvic gynecologic sarcomas using the recommended care pathway, and
- Provide guidance on how to best direct patients who may have a suspicious pelvic sarcoma.

The scope of this report is limited to adult patients and, therefore, does not include recommendations for patients 18 years of age and younger.

The target audience of this report are community and hospital gynecologists, gynecologic oncologists and gynecologic and sarcoma health care providers, including radiologists, pathologists and surgeons.

Development of Report - Methodology

In 2016, Ontario Health (Cancer Care Ontario) convened a Gynecologic Sarcoma Working Group consisting of experts in the fields of medical oncology, gynecologic oncology, radiation oncology, surgical oncology, gynecology, and pathology. The group also included a patient representative (Appendix A).

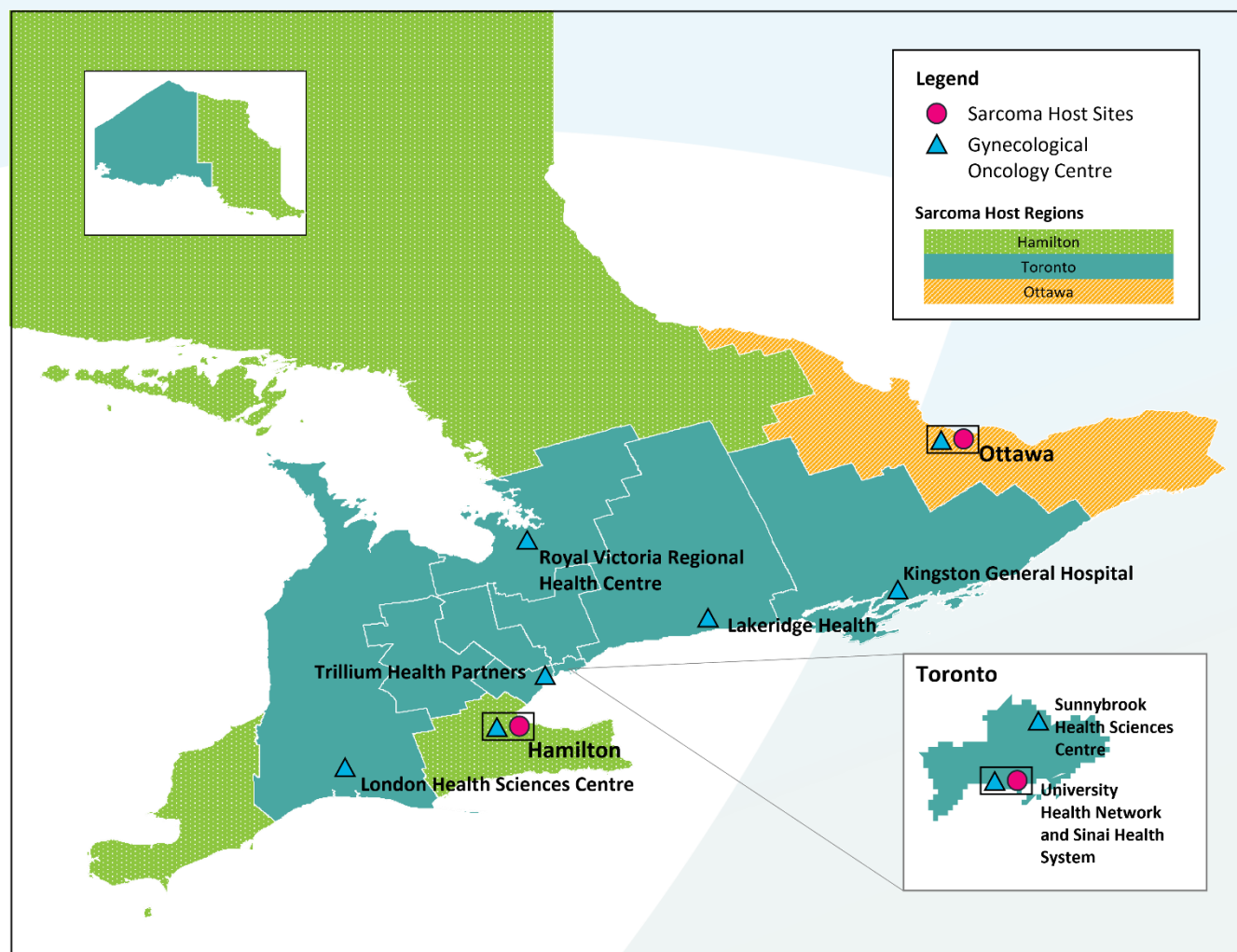
The primary aim of the Working Group was to create a guidance document for clinicians about best practices for the investigation and management of women with suspected pelvic, extra-pelvic gynecologic sarcoma, and non-gynecologic pelvic sarcomas. This work was intended to improve alignment between general gynecologists, GOCs, and Sarcoma Services Host Sites as a strategy to further improve the quality of patient care.

The Working Group identified and addressed clinical challenges faced by patients with suspected gynecologic sarcomas, and developed recommendations to improve their care. The recommendations are based on expert consensus, as there was limited evidence to guide the management of these rare tumors. The working group's clinical experience and judgment informed the recommendations.

Recommendations on the Diagnosis and Management of Gynecologic and Non-Gynecologic Pelvic Sarcomas

Since primary and secondary gynecologic sarcomas are relatively rare and the diagnosis and management are often multidisciplinary, the collaboration between general gynecologists, GOCs and Sarcoma Services Host Sites (Figure 1) is essential. GOCs are designated centres that provide surgical care, radiation therapy and systemic therapy for most invasive gynecologic oncology disease sites (Appendix B), and act as a hub for the management of invasive gynecologic cancers (Appendix C). Sarcoma treatment is provided in three multi-region collaborative sarcoma programs. In each of these programs, there is a Sarcoma Services Host Site that provides a full spectrum of specialized sarcoma services (Appendix D).

Figure 1: Map of Ontario showing location of sarcoma services host sites and gynecologic oncology sites.



Multidisciplinary Cancer Conference Review

Multidisciplinary Cancer Conferences (MCCs) are regularly scheduled meetings where healthcare providers discuss the diagnosis and treatment of individual cancer patients and have become well established in Ontario.

- All suspected or confirmed primary or secondary gynecologic sarcoma cases should be reviewed at an MCC, at a GOC or Sarcoma Services Host Site.
- The following subset of gynecologic sarcoma cases should be presented at a GOC and/ or Sarcoma Services Host Site MCCs.

Gynecologic sarcoma cases that should be presented at a Gynecologic Oncology Centre (GOC) and/or Sarcoma Services Host Site multidisciplinary cancer conferences (MCCs) and should have secondary pathology review at a Sarcoma Services Host Site.

- Histologies that may benefit from review at a Sarcoma Services Host Site MCC may include:
 - Epithelioid sarcomas, Ewing's family tumours, gastrointestinal stromal tumours (GISTs), and angiosarcomas
 - Where the differential of a soft tissue type sarcoma is suspected or remains after appropriate work-up
 - Cases requested by a pathologist with a specialty or special interest in gynecologic pathology to be reviewed by a pathologist with a specialty or special interest in sarcoma
- Locally extensive tumours where neoadjuvant therapy (radiation or chemotherapy) may lead to less morbid surgery
- Locally advanced tumours of borderline resectability that might benefit from initial radiation or chemotherapy
- Complex cases that involve multiple organs or pelvic sidewall
- Cases that are being considered for salvage treatment for recurrence that may benefit from genetic profiling, and receive chemotherapy and/or radiotherapy pre-operatively
- Metastatic cases for systemic chemotherapy and for consideration for clinical trials

Patient Referral Recommendations

Imaging

Pre-operative diagnosis of any pelvic sarcoma is optimal, but frequently does not occur. This can be a result of a low index of suspicion coupled with the use of pre-operative imaging modalities that provide inadequate information about the true nature of the tumour being imaged.

- Adequately imaged suspected fibroid cases do not require further imaging since imaging is not able to distinguish between fibroids and organ-confined leiomyosarcoma (LMS) or endometrial stromal sarcomas (ESS). These cases are frequently identified post-operatively. All completely resected uterine sarcomas should be referred to a GOC for pathology review, followed by a review by a gynecologic oncologist. Isolated pelvic masses should not be operated on without imaging as outlined below:
 - All pelvic masses with the following suspicious features on ultrasound should be followed up with magnetic resonance imaging (MRI):
 - The ultrasound does not show the contour of the mass (e.g., very large mass)
 - The patient has compressive symptoms (e.g., leg swelling, neuropathic pain)
 - There is suggestion of sidewall involvement on ultrasound
 - If extrauterine extension or pelvic side wall involvement is confirmed, the case should be referred to a soft tissue sarcoma or gynecologic oncology surgeon without further intervention.

Surgery

Patients with pre-operative abnormal bleeding should have an endometrial biopsy as some sarcomas may be diagnosed this way.

Incisional and excisional procedures should be avoided if pre-operative or intra-operative findings suggest an atypical diagnosis. These patients should be referred for diagnostic assessment at a specialist centre. Patients with suspicious or confirmed cases of gynecologic sarcomas should have surgery at GOCs or Sarcoma Services Host and Partner Sites (usually for complex cases that involve multiple organs or pelvic sidewall or pelvic sarcomas with secondary involvement of female reproductive organs).

Pathology

All suspected gynecologic sarcoma cases, regardless of topography local extent or completeness of excision, should have the pathology reviewed by appropriate pathologists located at a GOC.

Pelvic sarcomas with secondary involvement of female reproductive organs should have the pathology reviewed by pathologists located at a Sarcoma Services Host Site. Cases that may benefit from secondary pathology review at a Sarcoma Services Host Site are listed above.

Adjuvant Treatment

There is no strong evidence to support or not support the use of adjuvant therapy for gynecologic sarcomas. Thus, decisions regarding treatment should be determined with the patient on a case-by-case basis after secondary pathology review by a pathologist with a specialty or special interest in sarcoma and/or gynecologic pathology and discussion at MCC.

Palliative Care

- Early involvement of palliative care is recommended for gynecologic sarcoma patients requiring ongoing symptom management or those anticipated to require end-of-life care.
- Gynecologic sarcoma patients can experience severe pain, edema, and other symptoms that can impact on their quality of life. Timely symptom management and referral to a palliative care team should be considered.

Other

- GOCs and Sarcoma Services Host Sites should be aware of clinical trials available for gynecologic sarcoma patients. Consideration for participation in clinical trials is strongly recommended for each patient at each stage of therapy.

Quality Assurance

This recommendations report will continue to be monitored for accuracy and completeness by the Sarcoma Services Advisory Committee (Appendix A) on a biannual basis. Ongoing review of this report includes monitoring new therapy options moving through the drug approvals pipeline and updating the report based on emerging literature and best practice.

Conclusions

In conclusion, this recommendations report outlines the key components that should be instituted to increase collaboration between sarcoma service providers and gynecologic oncology centres. The report will also provide heightened awareness of pelvic sarcomas amongst general gynecologists and pathologists, and provide guidance on the investigation and management of women with suspected or proven pelvic and extra-pelvic gynecologic sarcomas. This report will also help to inform the provincial approach for planning, funding and performance management for this patient population.

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9. Sarcoma Services Advisory Committee. Provincial Sarcoma Services Plan: Version 2. 2015.

Appendices

Appendix A: Sarcoma Services Committee Members

Sarcoma Services Advisory Committee Members

The following committee members are acknowledged for their work in reviewing and maintaining the accuracy and completeness of this report.

- **Dr. Charles Catton**, Co-Chair
Princess Margaret Cancer Centre
- **Dr. Frances Wright**, Co-Chair
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- **Susan Blacker**
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- **Dr. Adriana Boyko**
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Gynecologic Sarcoma Working Group Members

The following committee members are acknowledged for their work in developing the initial version (2017) of this report.

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- **Dr. Rebecca Gladdy**

- Surgeon, Sinai Health Systems
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Medical Oncologist, University Health Network

The Working Group also wishes to thank **Dr. Sarah Ferguson**, Ontario Gynecologic Cancers Lead, Cancer Care Ontario who contributed to and supported the development of these recommendations.

Appendix B: Gynecologic Sarcoma Codes

Gynecologic sarcoma cases were identified in OCR through the following gynecologic topography codes (anatomical location).

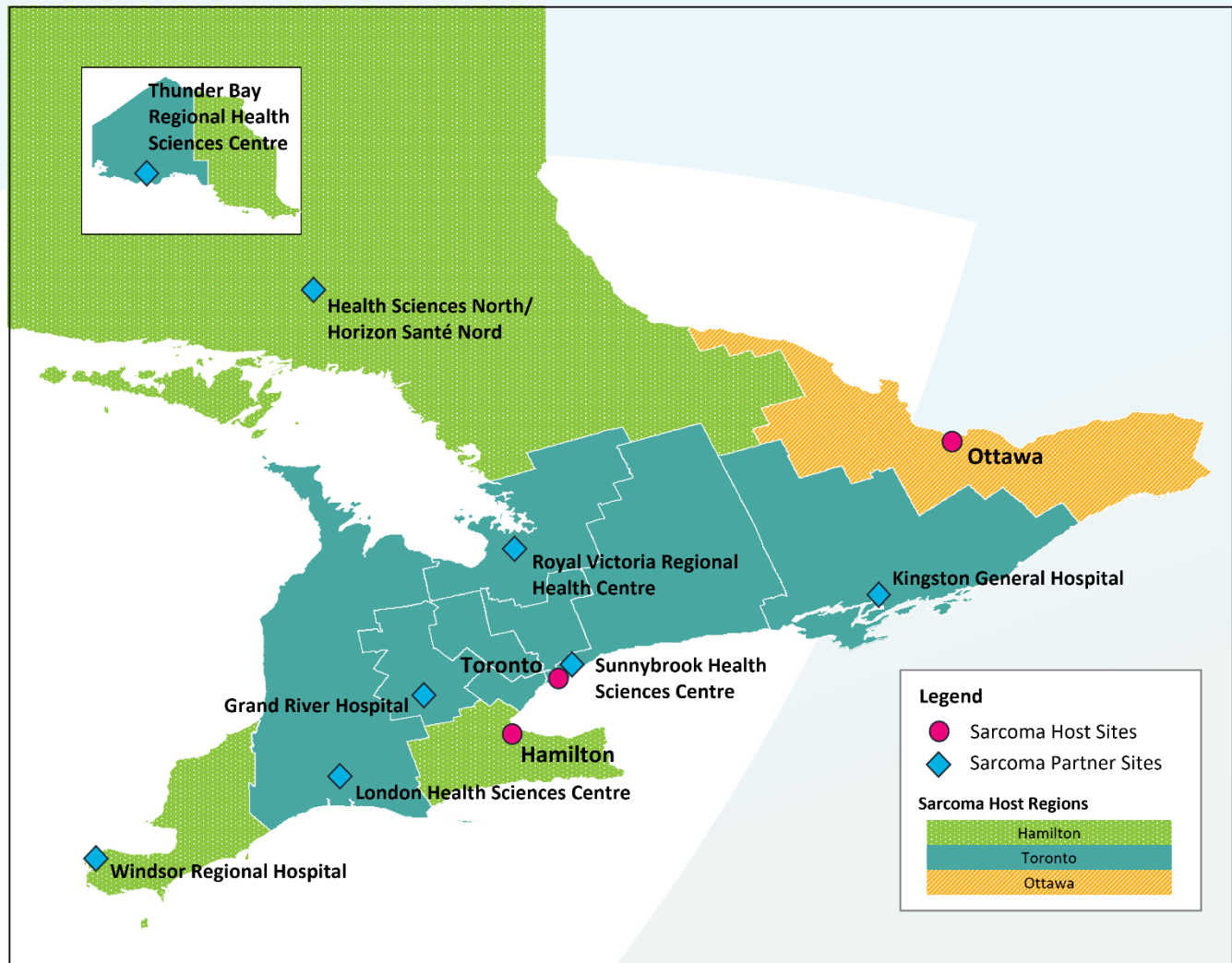
Topography Code	Topography Description
C519	Vulva NOS
C529	Vagina NOS
C530	Endocervix
C510	Labium majus
C572	Round ligament
C549	Corpus uteri
C559	Uterus NOS
C569	Ovary
C571	Broad ligament
C574	Uterine adnexa
C578	Overlapping lesion of female genital organs
C579	Female genital tract NOS
C538	Overlapping lesion of cervix uteri
C540	Isthmus uteri
C548	Overlapping region of the corpus uteri

Appendix C: List of Gynecologic Oncology Centres (GOCs)

- Hamilton Health Sciences Centre
- Kingston Health Sciences Centre
- Lakeridge Health
- London Health Sciences Centre
- Royal Victoria Regional Health Centre
- Sinai Health System
- Sunnybrook Health Sciences Centre
- The Ottawa Hospital
- Trillium Health Partners
- University Health Network

For more information regarding Gynecologic Oncology Centres, please visit:
cancercareontario.ca/en/find-cancer-services/sarcoma-service-locations

Appendix D: Location of Sarcoma Services Host and Partner Sites



For more information regarding Sarcoma Services Host and Partner Sites, please visit: cancercareontario.ca/en/find-cancer-services/sarcoma-service-locations.

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