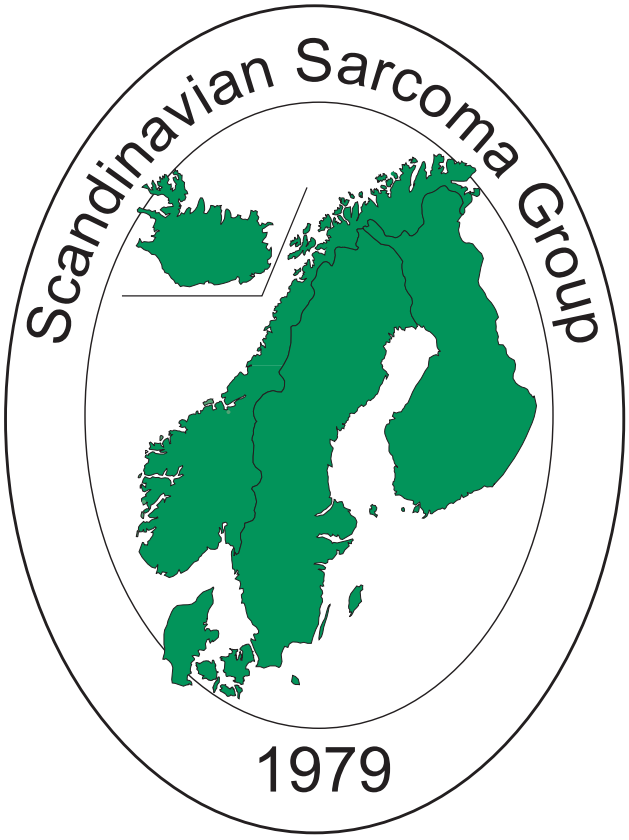
Scandinavian Sarcoma Group and
Oncologic Center, Lund, Sweden

Centralized Registration of Sarcoma Patients in Scandinavia SSG VII:4



Modified April, 2009

Scandinavian Sarcoma Group
& Oncologic Center, Lund, Sweden

CENTRALIZED REGISTRATION OF SARCOMA PATIENTS IN SCANDINAVIA
SSG VII:4

Modified April, 2009

A Scandinavian, multicentric, prospective study for evaluation of treatment results and prognostic factors in patients with soft tissue and bone sarcomas by a centralized registration. Presented by the Working Committee of the Scandinavian Sarcoma Group.

Contents

1. Introduction and background

The common registration of data allows for multicentric studies addressing treatment results and prognostic factors for local recurrence and survival in patients with soft tissue and bone sarcomas. Such studies are necessary to further define the best treatment for these patients. The close to 100% follow-up that is possible in Scandinavian countries makes our position unique.

Centralization of patients with bone and soft tissue sarcomas of the trunk wall and extremities has since long been practiced in Scandinavia. Visceral and retroperitoneal sarcomas have gathered great interest during later years due to novel techniques in the diagnosis and treatment of GIST. The multidisciplinary diagnosis and treatment require close cooperation between the surgeon, the radiologist, the cytologist, the pathologist, gynecologist and the oncologist. Thus, centralization also of patients with visceral and retroperitoneal sarcomas is mandatory. The SSG Registry of soft tissue and bone tumors was initiated March 1, 1986. All Centres in Norway and Sweden participate in the Registry, as well as certain Centres in Denmark and Finland. The yearly accrual rate is approximately 250 soft tissue and 100 bone tumor patients.

The Register gives important information on how treatment of patients with musculoskeletal tumors is evolving in the Scandinavian countries. For example, important changes in referral pattern, preoperative diagnostic techniques, surgical margin and radiotherapy have been observed.

The Register has formed the basis for several theses regarding treatment and prognosis. In depth studies of patients reported to the Registry are important for quality assurance. A theses regarding chondrosarcoma and a thesis on the quality and importance of radiotherapy in Soft Tissue sarcoma treatment are currently under preparation.

An important facet of the Registry is the histopathological re-evaluation of diagnosis performed by the SSG Pathology Board.

The forms for registration of patients to the Central Register have been modified and the new forms have been approved by the SSG working committee as of December 2008 . We have made separate forms for sarcomas of the extremity and trunk wall and for visceral and retroperitoneal sarcomas. The data collection will therefore be more appropriate for sarcomas of different sites. We have also modified the histopathological diagnoses according to the WHO classification and added SNOMED code for ambiguous classification.

For guidelines regarding surgical, medical and oncological treatment refer to ongoing SSG and collaborative study protocols.

The guidelines for surgical treatment and radiotherapy provided in SSG XX are also applicable to soft tissue sarcoma patients who are not candidates for adjuvant chemotherapy. See SSG XVII for guidelines regarding treatment of patients with visceral and retroperitoneal sarcomas.

All ongoing treatment protocols and treatment recommendations are found on the SSG site: www.ssg-org.net

Thor Alvegård, Henrik Bauer, Kirsten Sundby Hall
Chairmen of the SSG

Clement Trovik
Chairman of the SSG Central Registry

Jan Åhlén
Chairman of visceral and retroperitoneal sarcoma surgery

2. Visceral and retroperitoneal sarcomas

Guidelines for completion of forms

Primary tumor

All variables on this form refer to the **primary tumor**, whether treated before or after referral to the sarcoma center.

Date of diagnosis
Date when tissue suitable for microscopic diagnosis was **first** procured, either by needle biopsy, open biopsy or surgical treatment, before referral, or at a Centre.

Treated in accordance with SSG treatment protocol number
Indicate whether the patient has recived treatment following broadly a SSG study or a collaborative study e.g. EURAMOS, EUROBOSS etc..

Included in the SSG protocol specified above
Check “yes” if the patient is actually accepted in the protocol study.

SSG protocol patient i.d. number
If blank, the number will be added by the SSG secretariat

Referral pattern to cancer centre

Local microscopic diagnosis or excision performed before referral. Virgin implies untouched lesion. Excision implies any surgical procedure for primary tumor, e. g. open biopsy or partial or complete tumor excision. A cancer centre is defined as a sarcoma centre or a centre with defined collaboration with a sarcoma centre.

Metastasis at diagnosis of primary tumor
Refers to the diagnostic status of metastases at the time of diagnosis of the primary tumor.

Antecedents

Previous cancer, chemo- or radiotherapy, cancer-related diseases, for example neurofibromatosis. More than one can be checked.

Preoperative diagnostic procedures

How the tumor diagnosis was made preoperatively, **either before referral or at the centre**. More than one method can be checked.
Note that excision regardless of surgical procedure is not classified as a diagnostic procedure but checked as ”surgery for primary tumor”(see later). Check “none” for this variable if the surgery was done without any prior diagnostic morphology.
“Incisional biopsy” is checked when less than 50% of tumor was removed. Incomplete removal of more than 50% of tumor is classified as “intralesional surgery”

Treatment for primary tumor (does not include open biopsy)

Date of first operation
Date when first operation was performed.

Where
Whether first operation was performed before referral (**outside**) or at **centre**.

Local residual tumor
R0: Refers to no residual tumor with microscopic free margins locally within abdomen or retroperitoneum.
R1: If microscopic residual tumor is left behind
R2: If there is macroscopic tumor left behind localy within the abdomen or retroperitoneum. Does not include distant metastases at other sites, e.g. lungmetastasis (se below at"concomitant surgical treatment of metastases")
RX: Residual tumor cannot be assessed

Surgical procedure
The surgeon has to report if there has been intralesional dissection that violates the tumor pseudocapsule including resection of the tumor in pieces or draining of a cystic lesion. If the dissection somewhere around the tumor has **uncovered the tumor pseudocapsule** or if the the tumor has been removed with surrounding adequate **covering of healthy tissue.**

Last operation for primary tumor
Applies to patients operated two or more times for the *primary* tumor. For example, in a patient referred to a cancer centre for extended excision after intralesional or incomplete excision, details regarding the first procedure (outside) would be registered under the *first* operation, and the extended excision (centre) under *Last operation for primary tumor*. If **Rest tumor found** is "No", there is no need to proceed with residual tumor or procedure.

Concomitant surgical treatment of metastases
Refers to removal, or attempt to remove metastatic tumor tissue during the same operation(s) or in conjunction with treatment for the primary tumor. More than one can be checked.

All tumor removed
Metastatic tumor tissue from all locations has to be macroscopically removed to check "yes".

Number of operations for primary tumor
Total number of operations performed to remove primary tumor, normally 1 or 2. If the patient was not operated, for example because of metastatic disease, mark 0.

Other treatment
Check if the patient received radiotherapy and/or medical antitumor treatment for primary tumor. When possible give the date when radio- and/or medical antitumor treatment were started.

Tumors are classified according to clinical and histopathological evaluation
Site
Where the tumor was located or wherefrom it was considered to emanate.

Histotype, growth pattern, necrosis, vascular invasion and SNOMED
These data should be included in the histopathologic report. If a lesion can not be classified it should be referred to other SSG pathologists for consultation.

Tumor size
Largest diameter as assessed by radiological imaging or by examination of the resected specimen.

Malignancy grade
SSG 4-grade scale is standard.
(GIST should be regarded as " Malignancy grade not applicable").

Optionally grade can also be stated according to:

The French grading system

Tumor differentiation:

Score 1:	Sarcomas closely resembling normal adult mesenchymal tissue
Score 2:	Sarcomas of certain histological type (e.g. myxoid liposarcoma, myxoid MFH)
Score 3:	Embryonal and undifferentiated sarcomas, sarcomas of doubtful type, synovial sarcoma, osteosarcoma, PNET

Tumor differentiation score of sarcomas in the French Federation of Cancer Centres Sarcoma Group System*

Diagnosis	**Score**
Well-differentiated liposarcoma	1
Myxoid liposarcoma	2
Round cell liposarcoma	3
Pleomorphic liposarcoma	3
Dedifferentiated liposarcoma	3
Fibrosarcoma	2
Myxofibrosarcoma (myxoid MFH)	2
Typical storiform MFH (sarcoma, NOS)	3
Pleomorphic MFH (patternless pleomorphic sarcoma)	3
Giant cell and inflammatory MFH (pleomorphic sarcoma, NOS with giant cells or inflammatory cells)	3
Well-differentiated leiomyosarcoma	1
Conventional leiomyosarcoma	2
Poorly diff./epithelioid/pleomorphic leiomyosarcoma	3
Synovial sarcoma (biphasic, monophasic and poorly differentiated)	3
Pleomorphic rhabdomyosarcoma	3
Mesenchymal chondrosarcoma	3
Extraskeletal osteosarcoma	3
Ewing´s sarcoma/PNET	3
Malignant rhabdoid tumor	3
Undifferentiated sarcoma	3
PNET= primitive neuroectodermal tumor; MFH= malignant fibrous histiocytoma Note: Grading of malignant peripheral nerve sheath tumor, embryonal and alveolar rhabdomyosarcoma, angiosarcoma, extraskeletal myxoid chondrosarcoma, clear cell sarcoma and epithelioid sarcoma is not recommended.	

**Modified from Guillou et al. 1997 and Rubin et al. 2006.*

Mitotic rate
Should be included in the histopathologic report.

Follow-up

The length and intensity of the follow-up is decided by the treating physician and patient unless the patient is enrolled in a specific treatment protocol. The SSG recommends that patients operated for intraabdominal or retroperitoneal sarcomas should be followed every 6 months for the first 5 years and every 12 months for the next 5 years. Follow-up includes chest radiograph and a CT of the abdomen and pelvis.
Young patients who have been treated with chemotherapy and radiotherapy should have life-long follow-up due to the risk for long-term toxicity.

Treatment of first local recurrance

Only data for treatment of the first local recurrence are recorded in the Register.
The classification of the procedure is the same as that for treatment of primary tumor.

Death

Give date of death. Patients who died because of metastases or local disease died *from tumor*, those who died of non-tumor related causes but had recurrent disease died *with tumor*, and those who had no evidence of tumor disease at death died *without tumor*. If the patient dies within 2 years after metastases have occurred, the patient died from tumor (if no other clear reason).

VISCERAL AND RETROPERITONEAL SARCOMA

Primary tumor **SSG Registry**

Patient identification

This form should be sent to:
Regionala Tumörregisteret, Universitetssjukhuset i Lund
SE-221 85 LUND Tel: +46-(0)46-17 75 55

Hospital

Doctor

Treated in accordance with SSG protocol number ☐☐ ☐ No ☐ Yes

Included in protocol

Date of diagnosis year month day ☐☐☐☐☐☐

Sex ☐ Male ☐ Female

Patients protocol ID number ☐☐☐☐☐☐

Referral pattern to cancer centre

☐ Virgin ☐ FNA ☐ Core biopsy ☐ Incisional biopsy
☐ Curretage ☐ Excision ☐ Recurrence ☐ Not referred

Metastasis at diagnosis of primary tumor
☐ No ☐ Yes

Antecedents

☐ None ☐ Previous cancer ☐ Chemotherapy ☐ Radiotherapy ☐ Other, specify ……

Preoperative diagnostic procedures (either before referral or at the centre)

☐ None ☐ FNA ☐ Core biopsy
☐ Incisional biopsy ☐ Curretage ☐ Endoscopy ☐ Other

Treatment for primary tumor (does not include incisional biopsy)

Date of first operation
year month day ☐☐☐☐☐☐

Where
☐ Centre ☐ Outside

Local residual tumor
☐ R0 = No residual tumor
☐ R1 = Micro residual tumor
☐ R2 = Macro residual tumor
☐ RX = Residual tumor cannot be assessed

Surgical procedure
☐ Covering healthy tissue
☐ Uncovered tumorcapsule
☐ Intralesional

Last operation for primary tumor
year month day ☐☐☐☐☐☐

Where
☐ Centre ☐ Outside

Rest tumor found
☐ No ☐ Yes

Local residual tumor
☐ R0 = No residual tumor
☐ R1 = Micro residual tumor
☐ R2 = Macro residual tumor
☐ RX = Residual tumor cannot be assessed

Surgical procedure
☐ Covering healthy tissue
☐ Uncovered tumorcapsule
☐ Intralesional

Concomitant surgical treatment of metastases
☐ Liver surgery ☐ Lung surgery ☐ Other surgery ☐ No surgical treatment

All tumor removed
☐ No ☐ Yes

Number of operations for primary tumor ☐☐

Other treatment
☐ None ☐ Radiotherapy, start date; year month day ☐☐☐☐☐☐ Dose/fraction ☐☐.☐ Gy Number of fractions ☐☐
☐ Medical antitumor treatment, start date; year month day ☐☐☐☐☐☐ ☐ Other, specify……

optional

Site
☐ Esophagus
☐ Stomach
☐ Small intestine
☐ Colon
☐ Rectum
☐ Spleen
☐ Mesentery
☐ Liver
☐ Retroperitoneal
☐ Uterus
☐ Pelvic area
☐ Bladder
☐ Other, specify;
.....

Histotype
☐ Liposarcoma
☐ GIST
☐ Leiomyosarcoma
☐ High-grade pleomorphic sarcoma/MFH
☐ MPNST
☐ Rhabdomyosarcoma
☐ Ewing´s/PNET
☐ Solitary fibrous tumor/hemangiopericytoma
☐ Angiosarcoma
☐ Endometrial stromal sarcoma
☐ Fibromatosis
☐ Unclassified
☐ Other, specify;
……

SNOMED ☐☐☐☐☐☐

Tumor size
☐☐ cm (largest diameter) ☐ Not determinable

Growth pattern ☐ Pushing ☐ Infiltrative ☐ Not determined

Malignancy grade ☐ Not applicable
☐ Four-grade scale (1-4)
☐ FNCLCC (1–3) *optional*

Necrosis
☐ No
☐ Yes;
☐ <50% ☐ ≥50%
☐ Not determined

Vascular invasion
☐ No
☐ Yes
☐ Not determined

Mitotic rate

All STS without GIST
☐ Mitoses <10/10HPF
☐ Mitoses 10-19/10/HPF
☐ Mitoses ≥20/10HPF
☐ Not determined

GIST
☐ <5/50HPF
☐ 5-10/50HPF
☐ >10/50HPF
☐ Not determined

Genetic analysis performed
☐ No
☐ Yes

VISCERAL AND RETROPERITONEAL SARCOMA

Follow-up **SSG Registry**

This form should be sent to:
Regionala Tumörregisteret, Universitetssjukhuset i Lund
SE-221 85 LUND Tel: +46-(0)46-17 75 55

Hospital

Doctor

Patient identification

Follow-up

year month day
Date

☐ No evidence of disease (NED)

Local recurrence ☐ No ☐ Yes

Previously reported ☐ No ☐ Yes

Distant metastasis(es) ☐ No ☐ Yes ☐ Liver ☐ Lung ☐ Lymph node ☐ Skeletal ☐ Other

☐ Persistent disease

Treatment of first recurrence

Surgery ☐ No ☐ Yes, date year month day

Operation

Where ☐ Centre ☐ Outside

Local residual tumor
☐ R0 = No residual tumor
☐ R1 = Micro residual tumor
☐ R2 = Macro residual tumor
☐ RX = Residual tumor cannot be assessed

Surgical procedure
☐ Covering healthy tissue
☐ Uncovered tumorcapsule
☐ Intralesional

Other treatment
☐ None ☐ Radiotherapy ☐ Medical antitumor treatment ☐ Other, specify ……

Treatment of metastatic disease

☐ Liver surgery ☐ Lung surgery ☐ Other surgery ☐ Medical antitumor treatment ☐ Radiotherapy ☐ No treatment

All tumor removed ☐ No ☐ Yes

Death

Date of death year month day

Reason ☐ From tumor ☐ With tumor ☐ Without tumor ☐ Unknown

3. Sarcoma of Extremity and Trunk wall

Guidelines for completion of forms

Primary tumor

All variables on this form refer to the **primary tumor**, whether treated before or after referral to the sarcoma centre.

Date of diagnosis
Date when tissue suitable for microscopic diagnosis was **first** procured, either by needle biopsy, open biopsy or surgical treatment, before referral, or at a Centre.

Treated in accordance with SSG treatment protocol number
Indicate whether the patient has received treatment following broadly a SSG study or a collaborative study e.g. EURAMOS, EUROBOSS etc.

Included in the SSG protocol specified above
Check “yes” if the patient is actually accepted in the protocol study.

SSG protocol patient i.d. number
If blank, the number will be added by the SSG secretariat

Referral pattern to cancer centre

Local microscopic diagnosis or excision performed before referral. Virgin implies untouched lesion. Excision implies any surgical procedure for primary tumor, e. g. open biopsy or partial or complete tumor excision. A cancer centre is defined as a sarcoma centre or a centre with defined collaboration with a sarcoma centre.

Metastasis at diagnosis of primary tumor

Refers to the diagnostic status of metastases at the time of diagnosis of the primary tumor. When metastasis is diagnosed within 30 days from diagnostic biopsy of primary tumor, the patient is considered to have metastasis at diagnosis. Date of metastasis at diagnosis of primary tumor should *not* be recorded as date of metastasis.

Antecedents

Previous cancer, chemo- or radiotherapy, cancer-related diseases, for example neurofibromatosis. More than one can be checked.

Preoperative diagnostic procedures

How the tumor diagnosis was made preoperatively, **either before referral or at the centre**. More than one method can be checked.
Note that intralesional or marginal excision is not classified as a diagnostic procedure but checked as ”surgery for primary tumor” (see later). Check “none” for this variable if the surgery was done without any prior diagnostic morphology.
“Incisional biopsy” is checked when less than 50% of tumor was removed. Incomplete removal of more than 50% of tumor is classified as “intralesional surgery”.

Treatment for primary tumor (does not include open biopsy)

The same classification of procedures and margins applies to soft tissue as well as bone tumors.

Date of first operation
Date when first operation was performed.

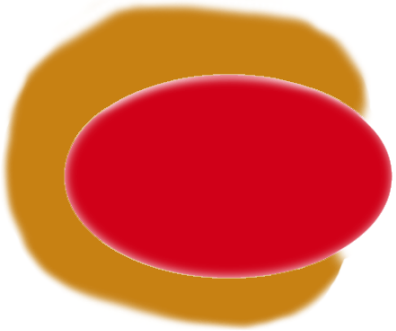
Surgical Procedure
Local excision or amputation.

Where
Whether first operation was performed before referral (**outside**) or at **centre**.

Surgical margin
As assessed at surgery and upon pathological macroscopic and microscopic examination. The most important margin is the poorest margin, i.e. the part of the specimen where the tissue coverage is poorest (qualitatively and quantitatively). In that area the pathologist should record the type of tissue (e.g. fat, connective tissue) and the thickness (mm) of tissues covering the tumor.

Two positive margins are defined:
Gross tumor left
The tumor is transected during the operation and macroscopic tumor tissue is left. This is reported by the surgeon.

Intralesional
Microscopic tumor tissue is seen at the resection border (reported by the pathologist) or leakage of fluid/tissue from the tumor into the wound occurs during surgery (reported by the surgeon).



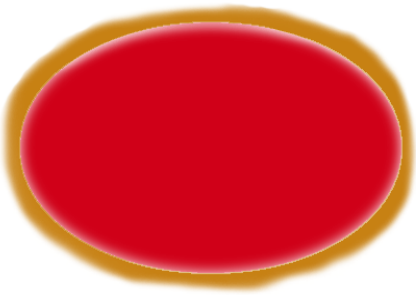
Two types of negative margins are defined:

The pathologist decides whether the margin is negative (tumor-free). In case of a negative margin the pathologist reports the shortest distance (mm) between tumor and resection border in fat, muscle or loose areolar tissue in an area where there is no fascia between the tumor and the resection border.

A fascia unengaged by the tumor is considered sufficient for a wide margin – irrespective of the distance between tumor and fascia. A total myectomy with the tumor completely surrounded by unengaged fascia needs no measurements and is by the surgeon classified as a wide margin.

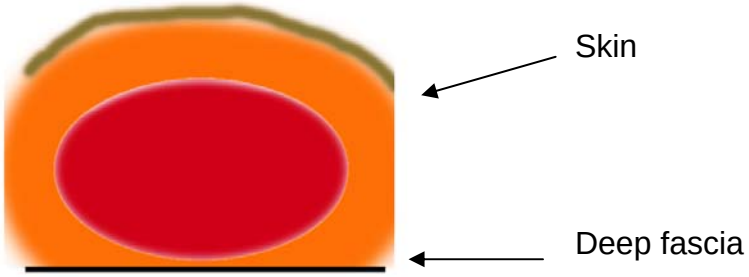
The **distinction between a *marginal* and *wide* margin is made by the surgeon** and is based on the combined information from surgery and histopathologic examination.

Marginal
The closest margin is outside but near the tumor in one or more places (irrespective of how much healthy tissue is included elsewhere) or all around the tumor (shelling out). Microscopically the margin is negative all around the tumor (otherwise the margin is intralesional), but tumor cells may be only millimetres from the margin.

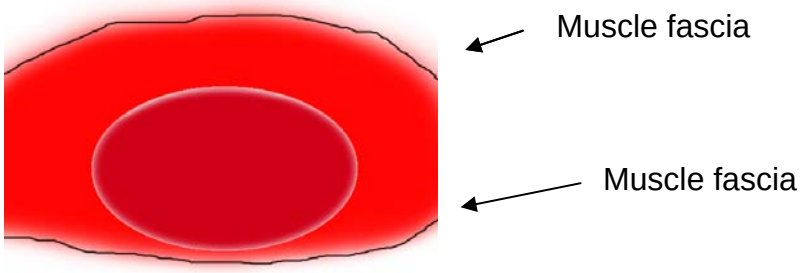


Wide
There is a cuff of healthy tissue all around the tumor. Unengaged fascia is considered a cuff regardless of the thickness of tissue between tumor and the fascia. A cuff of fatty or muscular or loose areolar tissue must be minimum 10 mm thick as measured at the histopathologic examination to qualify for a wide margin.

Subcutaneous

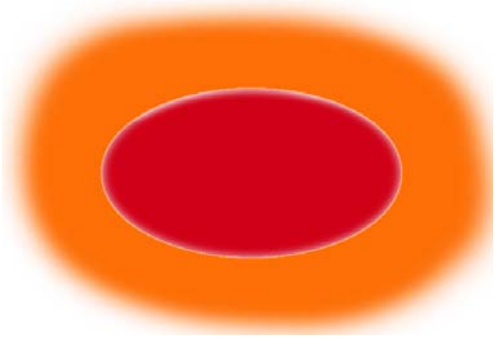


Intramuscular



A tumor within a muscle completely surrounded by an unengaged fascia is removed by total myectomy.

Deep extramuscular



At least 10 mm “cuff of healthy tissue” or unengaged fascia.

Date of last operation
Applies to patients operated two or more times of ***primary*** tumor. For example, in a patient referred to a sarcoma centre for extended excision after marginal excision of a soft tissue tumor, details regarding the first procedure (outside) would be registered under the *first* operation, and the extended excision at the centre under the *last* operation.

Number of operations for primary tumor
Total number of operations performed to remove primary tumor, normally 1 or 2. If the patient was not operated, for example because of metastatic disease, check 0.

Reconstruction (optional)
Applies to both bone and soft tissue tumors. Soft tissue reconstructions may be specified as: **Other.**

Other treatment
Give the date the patient started radiotherapy and/or chemotherapy for primary tumor, postoperatively or preoperatively. After radiotherapy give the dose pr fraction and the number of fractions received

Soft tissue tumors

Tumors are classified according to clinical and histopathological evaluation. See guidelines for surgical treatment of soft tissue sarcoma.

Site
Where the tumor was situated.

Location
Refers to whether the tumor is located within a compartment or not. Any deep tumor that originates or extends outside of a muscle is classified as extramuscular. Hence, a subcutaneous tumor with subfascial extension is classified as extramuscular.

Histotype, growth pattern, necrosis, and vascular invasion
These data should be included in the histopathological report. If a lesion can not be classified it may be referred another SSG pathologist. If a lesion is classified as a spindle cell sarcoma NOS it should be grouped together with the malignant fibrous histiocytoma/spindle cell and pleomorphic sarcoma in the data base

SNOMED code

This is optional, but should be recorded if the nomenclature in the pathology report do not correspond to SSG standard.

Below is a comparison of SSG nomenclature and corresponding WHO classification:

SSG nomenclature

1 High grade pleomorphic sarcoma/MFH

2 Myxofibrosarcoma/ Myxoid MFH

3 Low grade mal. fibromyxoid sarcoma

4 Fibrosarcoma

5 Liposarcoma

Leiomyosarcoma

Synovial sarcoma

MPNST

Angiosarcoma

Ewing´s /PNET

Rhabdomyosarcoma

Extraskel myx chond

6 Solitary fibr. tumor/ hemangiopericytoma

Clear cell sarcoma

Epithelioid sarcoma

Alveolar sarcoma

Extraskel osteosarc

Mal granular cell tumor

Dermatofibrosarcoma

Phyllodes

7 Fibromatosis

8 Unclassified

9 Other, specify

WHO Classification of Soft Tissue Tumors of Intermediate Malignant Potential and Malignant Soft Tissue Tumors

So-called Fibrohistiocytic Tumors
- Intermediate (rarely metastasizing)
  - 9 Plexiform fibrohistiocytic tumor
  - 9 Giant cell tumor of soft tissues
- Malignant
  - 1 Pleomorphic malignant fibrous histiocytoma (MFH) / Undifferentiated pleomorphic sarcoma
  - 1 Giant cell MFH / Undifferentiated pleomorphic sarcoma with giant cells
  - 1 Inflammatory MFH / Undifferentiated pleomorphic sarcoma with prominent inflammation

Fibroblastic / Myofibroblastic Tumors
- Intermediate (locally aggressive)
  - 7 Superficial fibromatoses (palmar / plantar)
  - 7 Desmoid-type fibromatoses
  - 7 Lipofibromatosis
- Intermediate (rarely metastasizing)
  - 6 Solitary fibrous tumor and hemangiopericytoma (including lipomatous hemangiopericytoma)
  - 9 Inflammatory myofibroblastic tumor
  - 9 Low grade myofibroblastic sarcoma
  - 9 Myxoinflammatory fibroblastic sarcoma
  - 4Infantile fibrosarcoma
- Malignant
  - 4Adult fibrosarcoma
  - 2 Myxofibrosarcoma
  - 3Low grade fibromyxoid sarcoma/hyalinizing spindle cell tumor
  - 4Sclerosing epithelioid fibrosarcoma

Adipocytic Tumors
- Intermediate (locally aggressive)
  - 5Atypical lipomatous tumor / Well differentiated liposarcoma
- Malignant
  - 5Dedifferentiated liposarcoma
  - 5Myxoid/round cell liposarcoma
  - 5Pleomorphic liposarcoma
  - 5Mixed-type liposarcoma
  - 5Liposarcoma, not otherwise specified

Bone Tumors

Tumors are classified according to clinical and histopathological evaluation.

Site
Where the tumor was situated.

Pathologic fracture
Whether there was a pathologic fracture at presentation.

Location
Refers to whether the tumor is located within a compartment or not. A tumor that has eroded cortical bone but the periosteum is still intact is regarded as intraosseous.

Histotype
If a lesion can not be classified it may be referred to other SSG pathologists for consultation.

SNOMED code
This is optional, but should be recorded if the nomenclature in the pathology report do not correspond to SSG standard.

Tumor size
Largest diameter as assessed by radiological imaging or pathologic examination of the resected specimen.

Malignancy grade
Check "not applicable" if tumor always has the same grade of malignancy.(i.e. Classic osteosarcoma, Ewing/PNET or GCT) Otherwise two different grading systems are applied It is mandatory to check one of them.

Scandinavian 4-grade scale
or
The French grading system

The French grade (FNCLCC grade) is optional. But it should always be reported by the review pathologist.

Tumor differentiation:

Score 1:	sarcomas closely resembling normal adult mesenchymal tissue
Score 2:	sarcomas of certain histological type (e.g. myxoid liposarcoma, myxoid MFH)
Score 3:	Embryonal and undifferentiated sarcomas, sarcomas of doubtful type, synovial sarcoma, osteosarcoma, PNET

Tumor differentiation score of sarcomas in the French Federation of Cancer Centres Sarcoma Group System*

Diagnosis	Score
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Pleomorphic liposarcoma	3
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Fibrosarcoma	2
Myxofibrosarcoma (myxoid MFH)	2
Typical storiform MFH (sarcoma, NOS)	3
Pleomorphic MFH (patternless pleomorphic sarcoma)	3
Giant cell and inflammatory MFH (pleomorphic sarcoma, NOS with giant cells or inflammatory cells)	3
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Poorly diff./epithelioid/pleomorphic leiomyosarcoma	3
Synovial sarcoma (biphasic, monophasic and poorly differentiated)	3
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Mesenchymal chondrosarcoma	3
Extraskeletal osteosarcoma	3
Ewing´s sarcoma/PNET	3
Malignant rhabdoid tumor	3
Undifferentiated sarcoma	3
PNET= primitive neuroectodermal tumor; MFH= malignant fibrous histiocytoma Note: Grading of malignant peripheral nerve sheath tumor, embryonal and alveolar rhabdomyosarcoma, angiosarcoma, extraskeletal myxoid chondrosarcoma, clear cell sarcoma and epithelioid sarcoma is not recommended.	

**Modified from Guillou et al. 1997 and Rubin et al. 2006.*

Mitotic count:
Score 1: 0-9 mitoses per 10 HPF*
Score 2: 10-19 mitoses per 10 HPF
Score 3: ≥20 mitoses per 10 HPF
* A high power field (HPF) measures 0.1734 mm^2. Standardized HPF should be used.

Tumor necrosis:
Score 0: no necrosis
Score 1: <50% tumor necrosis
Score 2: ≥50% tumor necrosis

Histological grade (FNCLCC):
Grade 1: total score 2, 3
Grade 2: total score 4, 5
Grade 3: total score 6, 7, and 8

Follow-up

The length and intensity of the follow-up is decided by the treating physician and patient unless the patient is enrolled in a specific treatment protocol. The SSG recommends that patients are followed for at least 5 years from diagnosis or last relapse. However, 10 years follow-up should be considered for patients younger than 70 years. Follow-up includes physical examination, chest radiograph and for bone tumors radiographs of the bone site. More extensive radiological examinations may be considered in individual cases.

Recommended follow-up intervals after primary treatment are as follows for:

Years after diagnosis	**Low-grade tumors**	**High-grade tumors**
0–2	6 months	3 months
3	6 months	4 months
4–5	6 months	6 months
5–10 (optional)	yearly	yearly

Young patients who have been treated with chemotherapy and radiotherapy should have life-long follow-up due to the risk for long-term toxicity.

Status at follow-up

Distant metastasis is only checked if the metastasis occurred *after* the diagnostic phase (30 days). If the patient had metastasis at diagnosis, all metastatic tumors have to be removed and a subsequent metastasis must occur for this variable to be checked.

Treatment of first local recurrence

Only data for treatment of the first local recurrence are recorded in the Register. Record the largest diameter of the local recurrence, as assessed by imaging or pathologic examination, as this may be of prognostic significance. The classification of the procedure is the same as that for treatment of primary tumor.

Death

Give date of death.
Patients who died because of metastases or rarely local disease, died *from tumor*.
Those who died of non-tumor related causes, but had recurrent disease, died *with tumor*.
Those who had no evidence of tumor disease at death, known to the sarcoma centre or to the physician issuing the death certificate, died *without tumor*. If the patient dies within 2 years after metastases have occurred, the patient died from tumor (if no other clear reason).

SARCOMA OF EXTREMITY AND TRUNK WALL

Primary tumor SSG Registry

Patient identification

This form should be sent to:
Regionala tumörregistret, Universitetssjukhuset i Lund
SE-221 85 LUND Tel: +46-(0)46-17 75 55

Hospital

Doctor

Treated in accordance with SSG protocol number ☐☐ Included in protocol ☐ No ☐ Yes

Date of diagnosis year ☐☐☐☐ month ☐☐ day ☐☐ Sex ☐ Male ☐ Female

Patients protocol ID number ☐☐☐☐☐☐☐☐

Referral pattern to cancer center

☐ Virgin ☐ FNA ☐ Core biopsy ☐ Excision ☐ Local recurrence ☐ Not referred

Metastasis at diagnosis of primary tumor

☐ No ☐ Yes

Antecedents

☐ None ☐ Previous cancer ☐ Chemotherapy ☐ Radiotherapy ☐ Other, specify . . . . . .

Preoperative diagnostic procedures (either before referral or at the center)

☐ None ☐ FNA ☐ Core biopsy ☐ Incisional biopsy

Treatment for primary tumor (does not include open biopsy)

Date of **first** operation year ☐☐☐☐ month ☐☐ day ☐☐
Where ☐ Centre ☐ Outside

Surgical procedure ☐ Local excision ☐ Amputation

Surgical margin
Positive margin: ☐ Gross tumor left ☐ Intralesional
Negative margin: ☐ Marginal ☐ Wide
☐☐ Shortest margin (mm) except unengaged fascia

Date of **last** operation year ☐☐☐☐ month ☐☐ day ☐☐
Where ☐ Centre ☐ Outside

Surgical procedure ☐ Local excision ☐ Amputation

Surgical margin
Positive margin: ☐ Gross tumor left ☐ Intralesional
Negative margin: ☐ Marginal ☐ Wide
☐☐ Shortest margin (mm) except unengaged fascia

Number of operations for primary tumour ☐ (0=none)

Reconstruction ☐ None ☐ Prosthesis ☐ Cementation ☐ Rotation plasty ☐ Other, specify . . . . . .

Other treatment
☐ None
☐ Radiotherapy, start date; year ☐☐☐☐ month ☐☐ day ☐☐ Dose/fraction ☐☐☐.☐ Gy Number of fractions ☐☐
☐ Chemotherapy, start date; year ☐☐☐☐ month ☐☐ day ☐☐ ☐ Other, specify . . . . . .

optional

Soft tissue tumors

Site
☐ Head & Neck
☐ Mamma
☐ Upper trunk
☐ Lower trunk
☐ Shoulder
☐ Upper arm
☐ Elbow
☐ Lower arm
☐ Hand
☐ Gluteal
☐ Groin
☐ Thigh
☐ Knee
☐ Lower leg
☐ Foot
☐ Other, specify . . . . . .

Histotype SNOMED ☐☐☐☐☐☐☐

Histotype
☐ High grade pleomorphic sarcoma/MFH
☐ Myxofibrosarcoma/ Myxoid MFH
☐ Liposarcoma
☐ Leiomyosarcoma
☐ Synovial sarcoma
☐ MPNST
☐ Angiosarcoma
☐ Low grade mal. fibromyxoid sarcoma
☐ Fibrosarcoma
☐ Ewing´s /PNET
☐ Rhabdomyosarcoma
☐ Extraskel myx chond
☐ Solitary fibr. tumor/ hemangiopericytoma
☐ Clear cell sarcoma
☐ Epithelioid sarcoma
☐ Alveolar sarcoma
☐ Extraskel osteosarc
☐ Mal granular cell tumor
☐ Dermatofibrosarcoma
☐ Phyllodes
☐ Fibromatosis
☐ Unclassified
☐ Other, specify. . . . . .

Location
☐ Cutaneous
☐ Subcutaneous
☐ Intramuscular
☐ Extramuscular (deep)
☐ Unclassified

Growth pattern
☐ Pushing
☐ Infiltrative
☐ Not determined

Necrosis
☐ No
☐ Yes; ☐ <50% ☐ ≥50%
☐ Not determined

Vascular invasion
☐ No
☐ Yes
☐ Not determined

Bone tumors

Site
☐ Skull/facial bones
☐ Vertebra
☐ Rib
☐ Clavicle
☐ Sacrum
☐ Pelvis, other
☐ Scapula
☐ Humerus
☐ Radius
☐ Ulna
☐ Hand
☐ Femur
☐ Tibia
☐ Fibula
☐ Foot

Pathologic fracture
☐ No
☐ Yes

Location
☐ Intraosseous
☐ Extraosseous extension
☐ Unclassified

Histotype
☐ Classic osteos
☐ Parosteal osteos
☐ Other osteos
☐ MFH
☐ GCT (benign)
☐ Ewing´s/PNET
☐ Fibrosarcoma
☐ Leiomyosarcoma
☐ Chondrosarcoma
☐ Mesen chondros
☐ Clear cell chondros
☐ Chordoma
☐ Other type, specify . . . . . .

SNOMED ☐☐☐☐☐☐☐

Long bone ☐ Proximal ☐ Mid ☐ Distal

Tumor size
☐☐☐ cm (largest diam) ☐ Not determinable

Malignancy grade ☐ Not applicable
☐ Four-grade scale (1-4) ☐ FNCLCC (1–3) *optional*

Mitotic rate ☐ Mitoses <10/10HPF ☐ Mitoses 10–19/10HPF ☐ Mitoses ≥20/10HPF ☐ Not determined

SARCOMA OF EXTREMITY AND TRUNK WALL

Follow-up **SSG Registry**

Patient identification

This form should be sent to:
Regionala tumörregistret, Universitetssjukhuset i Lund
SE-221 85 LUND Tel: +46-(0)46-17 75 55

Hospital

Doctor

Follow-up

year month day

Date

☐ No evidence of disease (NED)

Local recurrence ☐ No ☐ Yes

Previously reported ☐ No ☐ Yes

Distant metastasis(es) ☐ No ☐ Yes ☐ Lung ☐ Lymph node ☐ Skeletal ☐ Liver ☐ Other

☐ Persistent disease (previously recorded)

Treatment of first local recurrence

Where ☐ Center ☐ Outside

Tumor size ☐☐ cm (largest diameter) ☐ Not determinable

Surgery ☐ No ☐ Yes, date year month day

Surgical procedure ☐ Local excision ☐ Amputation ☐ No treatment

Surgical margin

Positive margin: ☐ Gross tumor left ☐ Intralesional

Negative margin: ☐ Marginal ☐ Wide

☐☐☐ Shortest margin (mm) except unengaged fascia

Other treatment

☐ None ☐ Radiotherapy ☐ Chemotherapy ☐ Other, specify .....

Treatment of metastatic disease

☐ Lung surgery ☐ Liver surgery ☐ Other surgery ☐ Chemotherapy ☐ Radiotherapy ☐ No treatment

Death

year month day

Date of death

Reason ☐ From tumor ☐ With tumor ☐ Without tumor ☐ Unknown

MM 2008 10 10