The national Quality register for sarcomas of the extremities and trunk wall

Manual nationellt kvalitetsregister
Version 1.0.4

REGIONALA CANCERCENTRUM I SAMVERKAN

Manual version

Date	Version	Alterations in the manual
2016-04-25	Version 1.0.2	
2020-05-27	Version 1.0.3	Uppdaterad med nya variabler för FU 2020.
2021-01-18	Version 1.0.4	Uppdaterad med nya variabler för Registration och Treatment 2020.

Each variable is defined by its name (the same as the filed name on the form), thereafter a brief explanation of the variable is found. If a variable is mandatory for Swedish patients (optional for other countries) as part of the "Canceranmälan till cancerregistret" is specified here

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Innehåll

Nationella kvalitets-registret för sarkom i extremiteter och bålvägg

Sarkom är en sällsynt grupp av tumörer med olika egenskaper. De utgör ungefär 1% av alla maligniteter och utgår från bind- och stödjevävnad. De drabbar personer i alla åldrar och kan sitta nästan i vilken del av kroppen som helst. De behandlas av läkare och annan vårdpersonal inom många olika specialiteter, vilka varierar beroende på typ av tumör och var den sitter. Antalet fall av sarkom är relativt konstant över tiden. Totalt insjuknar ca 300 patienter/år i Sverige varav 70 drabbas av skelettsarkom, ett 50-tal av buksarkom (viscerala och retroperitoneala sarkom) och resterande av mjukdelssarkom i extremiteter och bålvägg. Av de cancerformer som drabbar barn och ungdomar utgör sarkom ca 10 % av alla fall

Det finns två olika register för att följa patienter som drabbats av sarkom i Sverige; ett för sarkom i extremiteter och bålvägg och ett för intraabdominella och retroperitoneala sarkom. Anledningen till denna indelning är att tumörsjukdomarna kräver olika utredningar och behandlingar och att det därför är det intressant att följa olika parametrar.

Sarkomregistret startades av Skandinaviska sarkomgruppen (SSG) redan 1986 och en separat del för viscerala och retroperitoneala sarkom startades 2008. Numera har respektive land nationella register men med ett tätt samarbete inom Skandinavien. De två svenska sarkomregistren ligger sedan 2015 på INCA. Denna manual gäller för registret för sarkom i extremiteter och bålvägg.

Registrets syfte och mål

- Att samla in tumör- och behandlingsrelaterade data om alla sarkom i extremiteter och bålvägg i Sverige.
- Att vara underlag för bedömning av resultat och följsamhet till nationella och internationella riktlinjer lokalt, regionalt och nationellt.
- Att utgöra underlag för förbättringsarbete och planering av sarkomverksamheten.
- Att möjliggöra utveckling och forskning kring sarkomsjukdomarna.

Registrets innehåll

Det nationella kvalitetsregistret för sarkom i extremiteter och bålvägg har utgått från SSG:s register och omfattar en del för anmälan av tumören (registration form), en del för behandling av primärtumören (treatment form) och en del för uppföljning (follow up form) av patienten som inkluderar behandling av eventuella lokalrecidiv eller fjärrmetastaser.

Det nationella kvalitetsregistret för sarkom i extremiteter och bålvägg utgör en del av det skandinaviska sarkomregistret och resterande del av manualen är därför gemensam med övriga länder och skriven på engelska.

Background

The Scandinavian sarcoma Group founded its registry of soft tissue and bone tumours in March 1986. Over the years the registry has evolved and now each Scandinavian country has their own national quality register for sarcomas. Their organization varies but the core variables are the same. The common registration of data allows for larger 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 possibilities to compare the registers data along with the longstanding centralization of sarcoma treatment in Scandinavia offers good possibilities for research. The yearly accrual rate is approximately 250 soft tissue and 100 bone tumour patients.

The multidisciplinary diagnosis and treatment require close cooperation between the orthopedic surgeon, the radiologist, the cytologist, the pathologist, gynecologist and the oncologist. The Register gives important information on how treatment of patients with musculoskeletal tumours 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. An important facet of the Registry is the histopathological re-evaluation of diagnosis performed by the SSG Pathology Board.

For guidelines regarding surgical, medical and oncological treatment we 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.

Organization

The national board is responsible, in collaboration with the regional cancer centers in Sweden, for the creation, running and use of the register. The register is compared to the regional cancer registers to identify missing cases in the national registry for sarcomas in the extremities and the trunk wall. Completions of missing registries are requested from reporting clinics. The data are collected annually at the Regional Cancer Centre South in Lund for statistical evaluation and inclusion in a national report.

From each region, a responsible physician at a sarcoma center is included in the national board (see http://www.cancercentrum.se/sv/Kvalitetsregister/Sarkom/). The chairman of the national board calls for meetings.

Registration

A patient diagnosed with a sarcoma of the extremities and trunk wall should be reported to the register and we recommend referral to a sarcoma center for final diagnosis and treatment. The forms for reporting a patient are the same within all of Sweden and the main variables the same within the SSG.

In Sweden, reporting to the registry can be made online using INCA. The registration form includes all parameters needed for a *Canceranmälan* and it will function as such as well (in Sweden). The variables which are mandatory for a *Canceranmälan* are marked as such in the manual.

The registration consists of two forms, the registration form, and the treatment form. When reporting a case online please note that *not all variables will be seen but only those appropriate to you prior selections*, i.e. if number of surgeries is reported as none then no data on type of surgery will be requested or if sarcoma type is reported to be a soft tissue sarcoma only variables applicable to this group of tumours will be shown

Local recurrences or distant metastases found during follow up should be reported as such though using the follow up form, not as new sarcomas. If a patient is diagnosed with a *second primary sarcoma*, either related to previous treatment or a new primary sarcoma at another site, it should be reported to the register as a new tumour.

Inclusion criteria

All patients, regardless of age at diagnosis, diagnosed with a sarcoma of the extremities and trunk wall according to the following criteria are included.

A sarcoma diagnosed at autopsy should not be included in the register; neither should a suspected but not confirmed sarcoma

Soft tissue sarcomas

Histotypes	SNOMED Codes
Alveolar soft-part sarcoma	95813
Angiosarcoma*	91203
Clear cell sarcoma of soft tissue	90443
Desmoplastic small round cell tumour	88063
Epithelioid haemangioendothelioma	91303
Epithelioid sarcoma	88043
Extra-renal rhabdoid tumour	89633
Extraskeletal Ewing	92603
Extraskeletal mesenchymal chondrosarcoma	92203
Extraskeletal myxoid chondrosarcoma	92203
Extraskeletal osteosarcoma	91803
Fibrosarcoma	88103
Infantile fibrosarcoma	88143
Leiomyosarcoma	88903, 88913, 88963
Liposarcoma (not grade 1, see below**)	88503, 88513, 88523, 88533, 88543, 88583
Low-grade-fibromyxoma	88113
Malignant granular cell tumour	95803
Malignant ossyfying fibromyxoid tumour	88423
Malignant phyllodes tumour	90203
MPNST***	95403
Myxofibrosarcoma	88113
Other STS	88003
PEC-oma, malignant	87143
Rhabdomyosarcoma	89003, 89103, 89203, 89213

Solitary fibrous tumour	88151****, 88153
Synovial sarcoma	90403, 90413, 90423, 90433
Undifferentiated pleomorphic sarcoma/unclassified sarcomas (UPS)	88013, 88023, 88303

* Tumour site C44* may be included for soft tissue angiosarcomas (see below).

** Grade 1 liposarcomas (atypical lipomatous tumours) should not be included in the national quality register (see below).

*** MPNST is a soft tissue tumour in a peripheral nerve. The ICDO-3 C47*-codes should hence be used.

**** Solitary fibrous tumour (88151) should be included in the national quality register but not connected to the *Cancerregister* (see below)

Soft tissue sarcomas at the following sites should be registered

- C490 Head and neck (may be registered, see below)
- C491 shoulder, upper arm, elbow, lower arm, hand
- C492 thigh, knee, lower leg, foot
- C495 gluteal, groin (may be registered, see below)
- C496 upper trunk, lower trunk
- C509 mamma
- C44* if the tumour is an angiosarcoma 91203 (may be registered, see below)

MPNST tumour sites

- C471 MPNST (95403) in shoulder, upper arm, elbow, lower arm, hand
- C472 MPNST (95403) in thigh, knee, lower leg, foot
- C475 MPNST (95403) in gluteal, groin (*may be registered, see below)*
- C476 MPNST (95403) upper trunk, lower trunk

Bone sarcomas

Histotypes	**SNOMED Codes**
Adamantinoma	92613, 93103
Angiosarcoma of bone	91203
Atypical cartilaginous tumour	To be defined
Benign giant cell tumour	92500*, 92501
Chondrosarcoma (grades I – III)	92203, 92433, 92213, 92423, 92403, 92313
Chordoma	93703
Clear cell chondrosarcoma	92423

Conventional osteosarcoma	91803, 91813, 91823
Dedifferentiated chondrosarcoma	92433
Epithelioid hemangioendothelioma	91333
Ewing sarcoma	92603, 93643
Leiomyosarcoma of bone	88903, 88913, 88963
Low-grade osteosarcoma	To be defined
Malignancy in giant cell tumour	92503
Mesenchymal chondrosarcoma	92403
Other osteosarcoma	91853, 91863, 91873, 91933, 91943
Other sarcoma of the bone	88003, 88123
Parosteal osteosarcoma	91923
Telangioectatic osteosarcoma.	91883
Undifferentiated high-grade pleomorphic sarcoma (UPS)	88503, 88023

* Benign giant cell tumours coded as 92500 should be included in the national quality register but not connected to the *Cancerregister* (see below)

Bone sarcomas at the following sites should be registered

- C400 scapula, humerus, radius, ulna
- C401 hand
- C402 femur, tibia, fibula
- C403 foot
- C410 head and neck (*may be registered, see below)*
- C412 vertebra
- C413 rib, clavicle or sternum
- C414 sacrum, pelvis not sacrum

Special tumour sites

No strictly cutaneous tumours (C44*) have to be registered in the national quality register but strictly cutaneous angiosarcomas *may* be registered by the physician. C44* should however not be sought after in the *Cancerregistret* or used for control of the national quality registers completeness in comparison to the *Cancerregistret.*

Head and neck sarcomas, bone (C410) and soft tissue (C490), *may* be registered in the national quality register at the physician's discretion but it is not mandatory. To avoid erroneous inclusions these cases should not be sought after in the *Cancerregistret.* C410 and C490 should not be used in the control of the national quality registers completeness in comparison to the *Cancerregistret.*

Soft tissue sarcomas in the gluteal and groin area (ICD code for pelvic area C495) *may* be registered in the national quality register at the physician's discretion but it is not mandatory. The code C495 includes many tumours located within the pelvic area and therefore only selected cases with this site code should be included in this register. To avoid erroneous inclusions these cases should not be sought after in the *Cancerregistret*. C495 should not be used in the control of the national quality registers completeness in comparison to the *Cancerregistret*

Special tumour types

Liposarcomas (soft tissue) grade 1, i.e. atypical lipomatous tumours 88501b, should not be registered in the national quality register.

Border line solitary fibrous tumour (88151) and benign giant cell tumours (coded as 92500 by the pathologist), should be included in the national quality register but NOT in the *Cancerregistret* (to the monitors: they should be saved without connect to a cancer register tumour). Benign solitary fibrous tumours (88150) should not be registered in either register.

Presentation of and access to the registry data

The register's national board is responsible for collection and presentation of data on a national level. An online annual report will be presented in cooperation with the Regional Cancer Centre South in Lund at https://cancercentrum.se/samverkan/cancerdiagnoser/sarkom/ .

Reporting clinics have access to their own data. Regional data is accessed through each regional cancer center.

National data is accessed through the Regional Cancer Centre South in Lund after application. The application should define which data is requested and include a brief presentation of the project. It should be sent to the chairman of the national board and the head of the Regional Cancer Centre South. An ethical permit to conduct the study and handle the data is needed.

To access Scandinavian data, the application should be sent to the chairman of the register's national board, the head of the Regional Cancer Centre South, and to the Scandinavian Sarcoma Group board for a review. The leading researcher should come from an active member institution and will be the first author of the publication. Before a decision is made by the board the research project will be evaluated by a group consisting of one representative from each Nordic country (two representatives from Sweden), in addition to the chairman of the central register. The members of the group will be suggested by the central register subcommittee and confirmed by the SSG board. This group will then present a proposal to the board. Authorship for pathology, imaging and translational research studies should be decided in the SSG board. All authors are expected to be involved actively in the manuscript writing, and must finally approve the version to be published.

Guidelines for completion of the forms

General information for Registration, Treatment and Follow up

Date of birth and country specific id number
The personal identity number (Sweden) or code number (other countries).

Name
The patient's name (unless it should not be reported due to national confidentiality laws).

Sex
The patient's sex.

Date of death
Date of death, automatically updated for Swedish patients. Updated in the file in Norway before transfer of the data to the register. Reported manually for other countries.

Reported date
Date when the form is sent to RCC, automatically derived in online registration.

Initiated by
Automatically derived in online registration depending who first reported the patient to the register.

Reported by
Name of the person who fills out the form.

Reporting unit: clinic/hospital/country
Automatically derived in online registration depending on the person reporting to INCA. Manually filled in on paper forms.

Responsible physician
Name of the physician responsible for the patient, the person reporting in INCA will be suggested as default but the name can be changed if needed.

Reviewers comment
Space for comments. (Online only.)

Incomplete

Online only. If variables are missing and the data cannot be found please check the incomplete box to avoid further inquiries. In such cases, you may motivate why in the field for comments to the monitor (above the actual form). Please note that you must send the form to the monitor (RCC) and not save it directly to the register if you check this box.

Registration form, diagnosis

Referral pattern to sarcoma center
Local microscopic diagnosis or excision performed before referral to a sarcoma center or a center with a defined collaboration with a sarcoma center.

- Not referred is used if the patient is identified from the sarcoma center, for example through a national cancer registry, but never referred.
- Virgin implies referred with untouched lesion.
- FNA implies referred after FNA.
- Core biopsy implies to referral after core biopsy.
- Local recurrence implies not referred for primary tumour but only later for a local recurrence.
- Excision implies any surgical procedure for primary tumour, e.g. open biopsy or partial or complete tumour excision. Patients referred after incision biopsy should be registered as referred after excision and the incisional biopsy registered as first surgery, not performed at center with R2 residual tumour.

Please note that patients referred with local recurrence or after development of metastatic disease should be registered in relation to the treatment of the primary tumour (if any) performed before referral, i.e. referral after excision or after local recurrence.

Referral date
Date when the referral letter to the sarcoma center was *written* (or first contact taken using other ways of communication).

First visit date
Date when the patients first visited the sarcoma center or was first discussed at a multidisciplinary treatment conference.

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

Diagnosed age
Automatically. (Online only.)

Patient informed date
Date when the patient **first** was informed of the diagnosis.

Date of written pathological report for diagnosis
Date when the written pathological report of the diagnostic specimen was issued, either before or after referral to a center.

NOTE: In Norway this variable is called date of diagnosis, i.e. the date that the diagnosis was established (by pathologist).

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

Patient worked up according to SVF
Indicate if the patient has been worked up according to a standardized care process (SVF) or not.

Preoperative diagnostic procedures performed before referral or at the sarcoma centre
How the tumour 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 tumour" (see later). Check:

- "None" for this variable if the surgery was done without any prior diagnostic morphology.
- "FNA"
- "Core biopsy"
- "Incisional biopsy" when less than 50 % of tumour was removed. Incomplete removal of more than 50 % of tumour is classified as "intralesional surgery".

Diagnosed based on
The clinical and/or histopathological basis of the cancer diagnosis. Mandatory for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret". Most often "Excision or surgery with histopathologic examination" (if a core biopsy and/or operation has been performed) or "Cytology" (if the diagnosis is based on fine needle aspiration only).

Choose diagnosis lab clinic
Name of the reporting pathology/cytology clinic. Mandatory for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret".

Specimen id number (PAD-number)
The id number (PAD-number) for the specimen from which the final histoype was assessed, most often the specimen from the first operation of the primary tumour. Year collected in a separate variable. Mandatory for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret".

Specimen id number, year
The year the tumour specimen leading to a sarcoma diagnosis was taken. Mandatory for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret".

Sarcoma type
Type of sarcoma, i.e. soft tissue, bone sarcoma or benign giant cell tumour of bones. Controls the rest of the online reporting form (which variables the register will inquire about).

If benign giant cell tumour of bones is selected as sarcoma type the rest of the registration form will follow the main format for bone sarcomas although no inquiries about certain variables will be done, e.g. malignancy grade, side, ICDO3-code et cetera.

Pathology of primary tumour

Mitotic rate, primary tumour
Mitotic count as reported by the pathologist.

- <10 mitoses per 10 HPF*
- 10-19 mitoses per 10 HPF*
- ≥20 mitoses per 10 HPF*

* A high power field (HPF) measures 0.1734 mm^2. Standardized HPF should be used.

Primary tumour growth pattern (soft tissue sarcoma only)
Any presence of infiltrative peripheral growth pattern should be reported (also if the majority of the circumference shows a pushing pattern). If only pushing growth pattern is seen this is reported. This data should be included in the histopathological report. If a lesion cannot be classified it may be referred another SSG pathologist.

Primary tumour vascular invasion (soft tissue sarcoma only)
Any presence of vascular invasion should be reported. This data should be included in the histopathological report. If a lesion cannot be classified it may be referred another SSG pathologist.

Necrosis in primary tumour (soft tissue sarcoma only)
Any presence of necrosis should be reported. This data should be included in the histopathological report. If a lesion cannot be classified it may be referred another SSG pathologist.

Necrosis percentage in primary tumour
Whether or not the percentage of necrosis in the tumour is below or above 50% as reported by the pathologist. This data should be included in the histopathological report.

Registration form - Bone sarcoma and benign bone tumour (GCT)

Bone sarcoma/benign bone tumour: Morphological diagnosis, histotype
Bone sarcoma morphological diagnosis. If a lesion cannot be classified it may be referred to other SSG pathologists for consultation.

Morphological diagnosis text (online only)
Morphological name of the tumour, derived automatically from morphological diagnosis for soft tissue or bone sarcomas. If "other type of sarcoma" is reported it should be defined here. Mandatory for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret".

Bone sarcoma/benign bone tumour: Primary tumour site
Where the primary bone tumour was situated.

Side (applicable for bilateral organs and body parts)
Primary tumour's location side. Mandatory for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret". Automatically derived as "Not applicable" for centrally located tumour sites (online only).

Bone sarcoma: Tumour location
Refers to whether the tumour is located within a compartment or not. A tumour that has eroded cortical bone but the periosteum is still intact is regarded as intraosseous.

Pathologic fracture at presentation
Whether there was a pathologic fracture at presentation.

Long bone
Where in a long bone, a bone sarcoma is located. Only activated when bone tumour site has been reported as one of the long bones (online only).

Size of primary tumour (largest diameter, in cm)
Largest diameter as assessed by radiological imaging or pathologic examination of the resected specimen, reported in cm.

Not possible to determine size of primary tumour
When the size of the primary tumour cannot be assessed.

Site of primary tumour, free text (on line only)
Free text for sarcoma location. Mandatory for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret". Automatically derived when from reported bone sarcoma primary tumour site and side when reporting online.

IDCO3-code for bone sarcomas
ICD-O3-code for tumour location of bone sarcomas. Mandatory for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret". Automatically derived from tumour size and location (online only).

Malignancy grade: 4 grade scale
For bone sarcoma the Scandinavian 4-tier malignancy grade scale is used. Check "not applicable" if tumour always has the same grade of malignance (i.e. Classic osteosarcoma, Ewing/PNET or GCT).

T-stage
T-stage for bone sarcomas, a mandatory variable for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret". Automatically derived from tumour size and location (online only).

- T1 if size ≤ 8cm
- T2 if size > 8cm
- T3 if discontinuous growth within the same bone
- Tx if size is not determinable or primary tumour location is unclassified.

Metastases at the time of the diagnosis of primary tumour
Refers to the diagnostic status of metastases at the time of diagnosis of the primary tumour. When metastasis is diagnosed within 30 days from diagnostic biopsy of primary tumour, the patient is considered to have metastasis at diagnosis of primary tumour. If the metastases are found later, please report them using the follow up form and report date and site of the metastases.

The N- and M-stage, mandatory variables for Swedish patients as part of the "Canceranmälan till cancerregistret", optional for others, are derived from this variable (online only).

N-stage
N-stage for bone sarcomas, a mandatory variable for Swedish patients, part of the "Canceranmälan till cancerregistret". Automatically derived as N0 (no) if no metastasis diagnosis has been reported. Must be filled in if metastasis at diagnosis has been reported. N1 if presence of regional lymph node metastases, N0 if none.

M-stage
M-stage for bone sarcomas, a mandatory variable for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret". Automatically derived as M0 (no) if no metastasis at diagnosis has been reported. Must be filled in if metastasis at diagnosis has been reported. M1 if presence of distant metastases, M0 if none.

Registration form - Soft tissue sarcoma

Soft tissue sarcoma: Morphological diagnosis, histotype
Soft tissue sarcoma morphological diagnosis. If a lesion cannot be classified it may be referred to other SSG pathologists for consultation. Please note that atypical lipomatous tumours/well differentiated liposarcomas on orthopedic locations should not be reported to the SSG register!

Morphological diagnosis text (online only)
Morphological name of the tumour, derived automatically from morphological diagnosis for soft tissue or bone sarcomas. If "other type of sarcoma" is reported it should be defined here. Mandatory for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret".

Soft tissue sarcoma: Primary tumour site
Where the primary soft tissue tumour was situated.

Side (applicable for bilateral organs and body parts)
Primary tumour's location side. Mandatory for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret". Automatically derived as "Not applicable" for centrally located tumour sites (online only).

Soft tissue sarcoma: Tumour location
Refers to whether the tumour is located within a compartment or not.

- Cutaneous, strictly cutaneous tumours.
- Subcutaneous, cutaneous tumours infiltrating the subcutaneous tissue or strictly subcutaneous tumours not invading the deep fascia.
- Intramuscular tumours not engaging the muscle fascias without any invasive growth.
- Extramuscular (deep), any deep tumour that originates or extends outside of a muscle is classified as extramuscular. Hence, a subcutaneous tumour with subfascial extension is classified as extramuscular.
- Unclassified.

Size of primary tumour (largest diameter, in cm)
Largest diameter as assessed by radiological imaging or pathologic examination of the resected specimen, reported in cm.

Not possible to determine size of primary tumour
When the size of the primary tumour cannot be assessed.

Site of primary tumour, free text (online only)
Free text for sarcoma location. Mandatory for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret". Automatically derived when from reported soft tissue sarcoma primary tumour site and side (online only).

ICDO3-code for soft tissue sarcomas

ICD-O3-code for tumour location of soft tissue sarcomas. Mandatory for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret". Automatically derived from tumour size and location (online only).

Malignancy grade

For soft tissue sarcomas the French malignancy grade system (FNCLCC grade) is used. It is based in tumour differentiation, morphological diagnosis, mitotic count and necrosis.

Histological grade (FNCLCC)

- Grade 1: total score 2, 3
- Grade 2: total score 4, 5
- Grade 3: total score 6, 7, and 8

Not applicable only applies to clear cell chondrosarcoma, chordoma, adamantinoma, atypical cartilaginous tumour, epithelioid hemangioendothelioma.

Not assessable: too limited amount of tumour tissue available to correctly assess grade.

Tumour 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.

Tumour differentiation score of sarcomas in the French Federation of Cancer Centers Sarcoma Group System (*Modified from Guillou et al. 1997 and Rubin et al. 2006)*

Diagnosis Score	
Well-differentiated liposarcoma (not recorded in the SSG)	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 (bi- or monophasic and poorly differentiated)	3
Pleomorphic rhabdomyosarcoma	3
Mesenchymal chondrosarcoma	3
Extraskeletal osteosarcoma	3
Ewing´s sarcoma/PNET	3
Malignant rhabdoid tumour	3
Undifferentiated sarcoma	3

PNET= primitive neuroectodermal tumour; MFH= malignant fibrous histoocytoma

Note: Grading of malignant peripheral nerve sheath tumour, embryonal and alveolar rhabdomyosarcoma, angiosarcoma, extraskeletal myxoid chondrosarcoma, clear cell sarcoma and epithelioid sarcoma is not recommended.

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 mm2. Standardized HPF should be used

Tumour necrosis:

- Score 0: no necrosis
- Score 1: <50% tumour necrosis
- Score 2: >50% tumour necrosis

T-stage

T-stage for soft tissue sarcomas, a mandatory variable for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret". Automatically derived from tumour size and location (online only).

- T1a = cutaneous/subcutaneous, size <=5cm,
- T1b = intramuscular/extramuscular deep, size <=5cm,
- T2a = cutaneous/subcutaneous, size >5cm,
- T2b = intramuscular/extramuscular deep, size >5cm,
- TX = size not determinable or Tumour location Unclassified

Metastases at the time of the diagnosis of primary tumour

Refers to the diagnostic status of metastases at the time of diagnosis of the primary tumour. When metastasis is diagnosed within 30 days from diagnostic biopsy of primary tumour, the patient is considered to have metastasis at diagnosis of primary tumour. If the metastases are found later, please report them using the follow up form and report date and site of the metastases.

The N- and M-stage, mandatory variables for Swedish patients, optional for others, as part of the "Canceranmälan till cancerregistret" are derived from this variable (online only).

N-stage

N-class for soft tissue sarcomas, a mandatory variable for Swedish patients, part of the "Canceranmälan till cancerregistret", optional for others. Automatically derived as N0 (no) if no metastasis diagnosis has been reported. Must be filled in if metastasis at diagnosis has been reported. N1 if presence of regional lymph node metastases, N0 if none.

M-stage

M-stage for soft tissue sarcomas, a mandatory variable for Swedish patients, optional for others, part of the "Canceranmälan till cancerregistret". Automatically derived as M0 (no) if no metastasis at diagnosis has been reported. Has to be filled in if metastasis at diagnosis has been reported. M1 if presence of distant metastases, M0 if none.

Treatment form (bone and soft tissue sarcoma)

Choose registration to connect
Select a registered tumour in the drop-list. If a previous follow up has been made, data will be loaded and new follow up data may be entered below.

Treatment decided at multi-disciplinary tumour conference (MDT)
Whether or not the treatment was decided at a multidisciplinary tumour conference.

Assigned contact nurse
Whether or not the patient had an assigned contact nurse.

Date when the patient was informed about the initial treatment plan
Date when the patient was informed about the initial treatment plan. A later adjustment in the plan does not require change of this date.

Number of surgeries for primary tumour
Total number of operations performed to remove primary tumour, normally 1 or 2. If the patient was not operated, for example because of metastatic disease, check 0. If number of surgeries is reported to be none, no further inquiries about surgical parameters will be made (online only) Reporting physician determines if the surgery is for the primary tumour or a local recurrence.

If a patient is operated in a two stage procedure where the second procedure is only reconstructive then only the tumour resection date should be entered (one surgery).

Oncologic treatment given for primary tumour
Specify whether or not the patient has received oncological treatment for the primary tumour, radiotherapy, medical antitumour treatment or other (needs specification). More than one alternative may be reported.

Other oncological treatment given for primary tumour
Specify which other type of oncological treatment the patient has received, for example isolated limb perfusion or hyperthermic treatment.

If a treatment is missing please contact RCC so we can add it

Treatment delay due to patient's choice
If the planned therapy has been postponed/delayed due to the patient's choice. Delay due to comorbidities should not be reported here.

Surgical treatment for primary tumour

Date of first surgery for primary tumour
Date of *first* surgery for the primary tumour. Note, if a date is entered more than 6 months after diagnosis of the tumour a warning will be shown to avoid typing errors. The reporting physician can ignore the warning.

First and/or last surgery for primary tumour performed
Whether the first surgery for the primary tumour was performed before referral (outside) or at the sarcoma center. The distinction between a surgery performed outside or at a sarcoma center applies to first and last (if applicable) surgery as well as to surgery of a local recurrence (if applicable).

Surgical procedure, first and/or last surgery
Local excision or amputation. The description of surgical procedure applies to first and last (if applicable) surgery as well as to surgery of a local recurrence (if applicable).

Surgical margin, first and/or last surgery
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 issues covering the tumour.

The classification of surgical margins applies to first and last (if applicable) surgery as well as to surgery of a local recurrence (if applicable).

Two positive margins are defined:
Gross tumour left (R2)
The tumour is transacted during the operation and macroscopic tumour tissue is left. This is reported by the surgeon.

Intralesional (R1)
Microscopic tumour tissue is seen at the resection border (reported by the pathologist) or leakage of fluid/tissue from the tumour into the wound occurs during surgery (reported by the surgeon).

Negative margins
Reported as R0, further specification needed (see below).

Shortest surgical margin (mm) except unengaged fascia, first and/or last surgery
The reported shortest surgical margin in mm, except in unengaged fascia. Please leave empty if not assessed or intralesional margins.

Assessed final margins on MDT (wide or marginal)
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 (as discussed on multi-disciplinary tumour conference). The pathologist decides whether the margin is negative (tumour-free). In case of a negative margin the pathologist reports the shortest distance (mm) between tumour and resection border in fat, muscle or loose areolar tissue in an area where there is no fascia between the tumour and the resection border.

R0 Marginal

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

R0 Wide
There is a cuff of healthy tissue all around the tumour. Unengaged fascia is considered a cuff regardless of the thickness of tissue between tumour 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.

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

Please note that a warning will be shown if date of last surgery for primary tumour is more than 3 months after the first surgery (online only), this warning can be ignored.

Surgery performed for recurrence should be reported on Follow-up form.

Last surgery for primary tumour performed; Surgical procedure, last surgery; surgical margin, last surgery
See definitions above for first operation.

Type of reconstruction
Applies to both bone and soft tissue tumours.
Soft tissue reconstructions may be specified as Ot**her.**

Complications
If a complication requiring re- operation within 30 days (not a planned reconstructive surgery) or a deep infection within 30 days has occurred. If neither of these complications has occurred check no complications.

Oncological treatment for primary tumour

Included in a clinical study
Whether or not the patient is included in a clinical study.

Treatment protocol
Specify which (ongoing) clinical study the patient is included in. If the patient is not included in a study choose ”No Protocol”. If a treatment is missing please contact RCC so we can add it.

Radiotherapy start date (primary tumour)
Date of radiotherapy start (primary tumour).

Dose/fraction
Radiotherapy dose/fraction.

Number of fractions
Number of fractions of radiotherapy.

Chemotherapy start date
Date of chemotherapy start.

If several variables are missing or not logical it can be explained here
If a case is missing many key variables please explain why briefly (i.e. not referred, charts lost...). This variable will be assessed by the register manager annually. Please keep the comment short, max 100 signs.

Follow up form

Choose registration to connect
Select a registered tumour in the drop-list. If a previous follow up has been made, data will be loaded and new follow up data may be entered below.

Follow up date
Date of the follow up visit/contact. Please note that the follow up date must be later than the date of diagnosis.

Follow up ended prematurely
Fill in the data from the latest follow up. If no additional FU is planned or if a patient is lost to further follow up check this box and register the reason below. If further follow up is planned leave this box un-checked.

Reason for prematurely ended follow up
Select the appropriate reason; medical reason (e.g. comorbidities which will prohibit further treatment) patient's choice or unplanned lost to follow up.

Example:

- Patient's choice: The patient no longer wishes to come in for tumour controls. Report the last follow up date, then make a new follow up form using the same date and check this box, thereafter, register the reason as patient's choice (see below).
- Unplanned lost to follow up: The patient does not show up to the appointments why the follow up is ended. Report the last follow up date, then make a new follow up form using the same date and check this box, thereafter, register the reason as unplanned lost to follow up (see below).

Status/Tumour recurrence

- Tumour status at the follow up visit/contact.
- Please report **no evidence of disease** if the patient has no tumour recurrences (local or distant).
- Please report **tumour recurrence** if the patient has any sign of tumour (local, distant or both) regardless of whether or not the recurrence is new or previously reported.
- Please report each tumour recurrence and disease free intervals to the register.
- If new recurrences/metastases are diagnosed after a tumour-free interval these should be registered later in the form as new recurrences. This can be repeated if relevant.

Patient informed date
Date when the patient was informed about the new local recurrence / distant metastases diagnosed during current follow-up period.

Local recurrence
Report yes if a local recurrence is present, else no. If you report a local recurrence here several more posts will be opened for registration. However, data on treatment of local recurrences will only be registered the first time. A total of 5 local recurrences may be recorded per patient.

Treatment of the first local recurrence can be adjusted until the patient is reported in local remission, i.e. the patient can be reported to receive radiotherapy and medical antitumoural treatment and then half a year later surgery for the same recurrence.

Date of local recurrence
Date of diagnosis of a local recurrence, clinical or radiological. Up to 5 local recurrences may be reported, see below.

Size of tumour, local recurrence not determinable
When the size of the local recurrence cannot be assessed.

Size of tumour, local recurrence (largest diameter in cm)
Largest diameter of local tumour recurrence as assessed by radiological imaging or pathologic examination of the resected specimen, reported in cm.

Surgery of local recurrence
Whether or not the patient was operated for the reported local tumour recurrence.

Surgery date local recurrence
Date of surgery of local recurrence; only activated if the previous variable reported that a surgery was performed.

Surgery for local recurrence performed

- Whether the surgery for the reported local recurrence was performed before referral (outside) or at the sarcoma center.
- The distinction between a surgery performed outside or at a sarcoma center applies to first and last (if applicable) surgery of the primary tumour as well as to surgery of a local recurrence (if applicable).

Surgical procedure, surgery local recurrence
Local excision or amputation. The description of surgical procedure applies to first and last (if applicable) surgery as well as to surgery of a local recurrence (if applicable).

Surgical margin in the surgery of the reported local recurrence
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 issues covering the tumour.

The classification of surgical margins applies to first and last (if applicable) surgery as well as to surgery of a local recurrence (if applicable).

Two positive margins are defined

Gross tumour left (R2)
The tumour is transected during the operation and macroscopic tumour tissue is left. This is reported by the surgeon.

Intralesional (R1)
Microscopic tumour tissue is seen at the resection border (reported by the pathologist) or leakage of fluid/tissue from the tumour into the wound occurs during surgery (reported by the surgeon).

Negative margins
Reported as R0, further specification needed (see below).

Surgery local recurrence: Shortest surgical margin (mm) except unengaged fascia
The reported shortest surgical margin in mm, except in unengaged fascia.

Assessment of final margin, wide or marginal, on MDT
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 (as discussed on multi-disciplinary tumour conference). The pathologist decides whether the margin is negative (tumour-free). In case of a negative margin the pathologist reports the shortest distance (mm) between tumour and resection border in fat, muscle or loose areolar tissue in an area where there is no fascia between the tumour and the resection border.

R0 Marginal
The closest margin is outside but near the tumour in one or more places (irrespective of how much healthy tissue is included elsewhere) or all around the tumour (shelling out). Microscopically the margin is negative all around the tumour (otherwise the margin is intralesional), but tumour cells may be only millimeters from the margin.

R0 Wide
There is a cuff of healthy tissue all around the tumour. Unengaged fascia is considered a cuff regardless of the thickness of tissue between tumour 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.

Other treatment of local recurrence
If the reported local recurrence has been treated with other/additional treatment to surgery. I.e. radiotherapy, medical antitumoural treatment, both or other type of treatment.

Local persistent disease
Report yes if the patient has persistent local recurrent disease, regardless of changes in size. If the patient is in local remission, report no and which date the patient became free from his/her local recurrence (see below).

Date when free of local recurrence
Date when the patient became free from his/her local recurrence.

New local recurrence
Report yes if the patient has persistent local recurrent disease (regardless of changes in size), else register no.

Date of second/third/fourth/fifth local recurrence
Date of diagnosis of a new local recurrence, clinical or radiological. Up to 5 local recurrences may be reported, see below.

Date when free of second/third/fourth/fifth local recurrence
Date when the patient became free from his/her local recurrence.

Distant metastases
Report yes if a distant metastases is present, else no. If you report a distant metastasis here several more posts will be opened for registration. Treatment of the first metastastic event can be adjusted until the patient is reported in distal remission, i.e. the patient can be reported to receive radiotherapy and medical antitumoural treatment and then half a year later surgery for the same metastasis.

A total of 5 metastatic events may be recorded per patient. However, data on treatment of distant metastases will only be registered the first time.

Date when metastases were diagnosed
Date for diagnosis of distant metastases, clinical or radiological. All metastases occurring after metastases-free interval should be reported and will be registered in this variable. Multiple recurrences of distant metastases may be registered for each patient.

Site of distant metastasis diagnosed during follow up
Site of the diagnosed distant metastasis. Please report "Multiple" if distant metastases were diagnosed in more than one location during the specific follow-up period, i.e. from previous until current follow-up visit.

Treatment of distant metastasis diagnosed during follow-up
Check boxes which are activated the first time a distant metastasis is reported. If "none" is reported, no further treatment can be registered. Else, multiple treatment modalities may be reported (surgery of liver, lung or other metastasis, radiotherapy or medical antitumoural therapy). Additional therapeutic modalities may be added until the patient is reported as being in remission from their metastatic disease.

Distant persistent disease
Report yes if the patient has persistent metastatic disease (regardless of changes in numbers of metastases and size). If the patient is in remission, report no and which date the patient became free from his/her metastasis (see below).

Date when free of distant metastases
Date when the patient became free from his/her known metastatic disease.

New distant metastasis
Report yes if a new distant metastasis is diagnosed (after a disease free interval), if no evidence of disease register no.

Date of second/third/fourth/fifth site of distant metastasis

Date of diagnosis of a distant metastasis, clinical or radiological. Up to 5 distant metastases may be reported, see below.

Date when free of second/third/fourth/fifth metastatic event

Date when the patient became free from his/her metastatic event.

Date of death

Date of death, automatically updated for Swedish patients. Updated in the file in Norway before transfer of the data to the register. Reported manually for other countries.

Cause of death

- **Dead from tumour** If the patient dies with known metastases. If the patient dies with untreated primary tumour or local recurrence causing disabling disease. If the patient has known metastases (diagnosed within two years) but exact reason for death is not known cause of death will be re-coded as Death from tumour.
- **Dead with tumour** If the patient dies with untreated primary tumour or local recurrence which does not cause disabling disease. If a patient with known metastatic disease dies of obviously unrelated cause (plane crash, murder etc.). Death from tumour should be used if in doubt for patients with generalized disease.
- **Without tumour** No evidence of recurrent disease at death.
- **Unknown** If the cause of death has not been able to establish.

Completeness of the register

A pre-defined check list for comparison of the national quality register to the *Cancerregistret* in Sweden is available for use by monitors in INCA. For further specifications, please contact the national support team at RCC Syd.

Regionala cancercentrum – regionernas nationella samverkan inom cancervården.
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