London and South East Sarcoma Network

Staging and follow-up guidelines for benign soft tissue and bone tumours, and malignant bone and soft tissue sarcomas

Introduction

This guidance covers staging and follow-up for patients diagnosed with benign and malignant soft tissue and bone sarcomas. While follow-up schedules within clinics is an important part of this, it is also well-recognised that many local tumour recurrences are detected by patients themselves. It is important therefore that patients are advised on the importance of self-monitoring between clinic appointments, and are aware of how to be able to access a clinic appointment earlier than booked if they have concerns.

Duration of follow-up:

For most of the clinical groups in these guidelines, follow-up is suggested for a duration of 10 years. However, it is recognised that there will be occasions when a patient has a good risk tumour that is expected to have a good prognosis, when shorter follow-up may be more appropriate. There are now predictive nomograms available that can provide this information for individual patients (Sarculator, PerSarc, MSKCC predictive nomograms) in order to identify patients who might be suitable for a modified follow-up schedule. Patient factors such as age, co-morbidities and performance status may also be taken into account for deciding the most appropriate follow-up schedule. Therefore, while these guidelines will be suitable for most patients, clinicians should feel able to tailor follow-up schedules to best suit an individual patient's needs.

Discharging patients from follow-up:

All patients who are discharged from follow-up should be advised of how to re-access the service if they have any concerns about possible relapse after discharge.

1. Staging and Follow-up Guidelines for Soft Tissue Tumours†

Stage of disease	Staging investigations	Follow-up schedule
1. Localised extremity/trunk post-surgery ± radiotherapy		
Benign soft tissue tumours/atypical lipomatous tumours	MRI primary tumour	
Year 1		- post-operative visit in first 6 weeks - supported discharge (discharge with documented advice on returning and re-accessing the clinical team if symptoms or a new mass appear)
Intermediate rarely metastasising tumours of uncertain malignant potential (including solitary fibrous tumour angiomatoid fibrous histiocytoma, pseudomyogenic haemangioendothelioma)	Pseudomyogenic haemangioendothelioma – CT chest abdomen and pelvis, whole body MRI	
Low grade	MRI primary (or CT if MRI contraindicated),	

	CT chest At local recurrence: MRI primary tumour, CT chest (as high rate of synchronous lung metastases)	
Year 1		- post-operative visit in first 6 weeks - 3 month clinical examination (to check function, if necessary) - 6 monthly clinical examination[1], CXR - routine MRI of primary site is not indicated unless the site is difficult to assess clinically
Year 2		- 6 monthly clinical examination, CXR
Years 3+		- annual clinical examination, CXR
Discharge at 10 years after surgery unless histiotypes associated with late recurrence or metastases, when longer follow-up should be considered: Low grade fibromyxoid sarcoma; endometrial stroma sarcoma; solitary fibrous tumour; malignant peripheral nerve sheath tumour grade 1; epithelioid haemangioendothelioma		
Intermediate and high grade	MRI primary tumour (or CT if MRI	

	contraindicated), CT chest Myxoid liposarcoma – CT chest abdomen pelvis, whole body MRI Alveolar soft part sarcoma, epithelioid sarcoma, clear cell sarcoma – CT chest, total body PET scan	
	<u>At local recurrence:</u> MRI primary tumour, CT chest (as high rate of synchronous lung metastases)	
Year 1		- post-operative visit in first 6 weeks - 3 - 4 monthly clinical examination, CXR - routine MRI of primary site is not indicated unless the site is difficult to assess clinically
Year 2		- 3 - 4 monthly clinical examination, CXR
Years 3 – 5		- 6 monthly clinical examination, CXR

Years 6 – 10		- annual clinical examination, CXR
Discharge at 10 years after surgery unless: - histiotypes associated with late recurrence or metastases, when longer follow-up should be considered: Synovial sarcoma; extraskeletal myxoid chondrosarcoma; mesenchymal chondrosarcoma (soft tissue); sclerosing epithelioid fibrosarcoma; epithelioid haemangioendothelioma; angiosarcoma - Patient has treatment-related toxicity that requires long term follow-up - Patient has a metallic prosthesis *in situ* (follow-up evaluation in physiotherapist-led endoprosthesis follow-up clinic) - Patients who were teenage or young adult at time of diagnosis will require referral into the UCH Late Effects Service: ucl-tr.LateEffectServ@nhs.net - Clinical trial patients on active follow-up		
2. Abdominal/retroperitoneal/gynaecological		*Comment*: **For retroperitoneal sarcomas**, because of the uncertainty about the timing and benefits of intervention for recurrent disease, surgical or otherwise, in this group of sarcomas follow up can take two forms: radiologically directed follow-up, or clinically

		directed follow-up. The choice of follow-up protocol is a clinical decision between clinician and patient, taking into account biological factors of the particular histological sub-type.
Radiologically directed follow-up		
Low grade	CT/MRI abdomen and pelvis, CT chest	
Year 1		- post-operative visit in first 6 weeks - 3 - 6 monthly clinical examination - baseline CT chest/abdo/pelvis[2] post surgery, then at 6 and 12 months
Year 2		- 6 monthly clinical examination - CT scan chest/abdo/pelvis[2] at 18 and 24 months
Year 3+		- annual clinical examination - annual CT chest/abdo/pelvis[2] to 10 years
Discharge at 10 years after surgery unless:		

- Histiotypes associated with late recurrence or metastases including low grade endometrial stroma sarcoma; solitary fibrous tumour		
Intermediate and high grade**	CT/MRI abdomen and pelvis, CT chest	
Year 1		- post-operative visit in first 6 weeks - 3 monthly clinical examination, CXR - baseline CT chest/abdo/pelvis[2] post surgery, then at 6 and 12 months
Year 2		- 3 monthly clinical examination, CXR - CT scan chest/abdo/pelvis[2] at 18 and 24 months
Years 3 – 5		- 6 monthly clinical examination and CXR - annual CT chest/abdo/pelvis[2]
Years 6 – 10		- annual clinical examination and CXR - annual CT chest/abdo/pelvis[2] year 5, then stop
Discharge at 10 years after surgery unless:		

- Histiotypes associated with late recurrence or metastases including solitary fibrous tumour; PEComa		
Clinically directed follow-up		
	Follow up intervals as above, with evaluation for new abdominal symptoms and clinical examination. Scanning (CT chest/abdo/pelvis[2]) is instituted for clinical suspicion of recurrence. Chest surveillance is performed at each visit by CXR.	
3. Head and neck sarcomas	MRI primary site (or CT if MRI contraindicated), CT chest	
Year 1		- 3 monthly clinical examination and CXR - post-treatment MRI of primary site at 3 months after completing treatment - surveillance MRI of primary site at 9 months
Year 2		- 3 monthly clinical examination and CXR - surveillance MRI of primary site at 15 and 21 months

Years 3 – 5		- 6 monthly clinical examination and CXR - surveillance MRI of primary site at 27 months - thereafter annual MRI of primary site
Years 6 – 10		- annual clinical examination and CXR
Discharge at 10 years after surgery unless histiotypes associated with late recurrence or metastases, when longer follow-up should be considered: Low grade fibromyxoid sarcoma; sclerosing epithelioid fibrosarcoma; synovial sarcoma; extraskeletal myxoid chondrosarcoma; mesenchymal chondrosarcoma		
4. Breast sarcomas	MRI breast (optional, booked by surgeons), CT Chest; PET scan for angiosarcoma	
Benign phyllodes		Discharge with self-managed follow up
Borderline phyllodes		Breast conserving surgery - breast imaging with 6 monthly USS, annual mammogram for 3 years, then discharge (to be done by breast surgeons)

		Mastectomy – discharge with self-managed follow-up
Malignant phyllodes, angiosarcoma, radiation-induced breast sarcoma, other breast soft tissue sarcoma		Follow up with chest imaging as high grade extremity/trunk soft tissue sarcoma (see p4). Breast conserving surgery - breast imaging with 6 monthly USS, annual mammogram for 3 years, then clinical examination.
Discharge at 10 years after surgery		
5. Chest wall/intra-thoracic sarcomas	CT chest, MRI primary tumour	
Year 1		- 3 monthly clinical examination and CXR - 6 monthly CT/MRI if primary site not adequately assessed by CXR/clinically
Year 2		- 3 monthly clinical examination and CXR - 6 monthly CT/MRI if primary site not adequately assessed by CXR/clinically

Year 3 - 5		- 6 monthly clinical examination and CXR - annual CT/MRI if primary site not adequately assessed by CXR/clinically
Years 6 – 10		- annual clinical examination and CXR *or* - annual CT/MRI if primary site not adequately assessed by CXR/clinically
Discharge at 10 years after surgery unless histiotypes associated with late recurrence or metastases, when longer follow-up should be considered: Low grade fibromyxoid sarcoma; sclerosing epithelioid fibrosarcoma; synovial sarcoma; extraskeletal myxoid chondrosarcoma; mesenchymal chondrosarcoma		
6. Dermal sarcomas	MRI primary site (or CT if MRI contraindicated) CT or chest x-ray Cutaneous angiosarcoma: CT CAP or PET	
High grade pleomorphic dermal sarcoma, grade 1 cutaneous leiomyosarcoma		- 3 monthly clinical review to 2 years, 6 monthly to 5 years, annual to 10 years - CXR 6 monthly for 2 years, annual to 10

		years
Grade 2 /3 leiomyosarcoma, MPNST, cutaneous angiosarcoma		As for high grade limb/trunk soft tissue sarcoma
7. Post pulmonary metastasectomy		
Year 1		- baseline CT scan and CXR post surgery (within 3 months) - 3 monthly clinical examination, CXR - thereafter 6 monthly CT scans
Year 2		- 3 monthly clinical examination, CXR - CT scans at 18 and 24 months
Years 3 – 5		- 6 monthly clinical examination and CXR - continue CT scans at clinician's discretion if felt to be at high risk or recurrence
Years 6 – 10		- annual clinical examination and CXR

Discharge at 10 years after surgery		
8. Locally advanced or metastatic disease		
Year 1+		- 3 monthly clinical examination and CXR (or more frequently as clinically indicated) - imaging of disease sites as clinically appropriate at clinician's discretion

† Patients treated within clinical trials should be followed up according to the trial protocol.

[1] Routine imaging of primary site at clinician's discretion, if clinical detection of recurrence is anticipated to be difficult, e.g. deep tumours; large tumours; post-radiotherapy, particularly if positive margin excision.

[2] Alternatives to CT chest abdomen and pelvis include:

- CT abdomen and pelvis, CXR
- MRI abdomen and pelvis, CXR
- Abdominal ultrasound scan, transvaginal ultrasound scan, CXR (for gynae sarcomas)

2. Staging and Follow-up guidelines for giant cell tumour of bone, conventional central cartilaginous tumours (including grade 1 chondrosarcoma), periosteoal and parosteal osteosarcoma, chordoma

Stage of disease	Staging investigations	Follow-up schedule
1. Giant cell tumour of bone		
Primary presentation	Baseline CT chest for patients presenting with pathological fracture, plain x-ray and MRI/CT primary site	
Year 1		- post-operative visit in first 6 weeks - 3 - 6 monthly clinical examination, plain films of primary site, CXR for patients presenting with pathological fracture
Years 2 - 3		- 6 monthly clinical examination, plain films of primary site, CXR for patients presenting with pathological fracture
Discharge 3 years after surgery, unless: Patient has a metallic prosthesis *in situ* (follow-up evaluation in physiotherapist-led endoprosthesis follow-up clinic)		

After local recurrence	MRI/CT primary site, CT thorax	
Year 1		- post-operative visit in first 6 weeks - 3 - 6 monthly clinical examination, plain films of primary site, CXR
Years 2 - 5		- 6 monthly clinical examination, plain films of primary site, CXR
2. Atypical cartilaginous tumour/ grade 1 chondrosarcoma	Plain x-ray and MRI primary site (chest imaging and bone scan not required)	
Localised post primary treatment – curettage +/- cementation		
Year 1		- post-operative visit in first 6 weeks - 3 monthly clinical examination, plain films of primary site[1]
Year 2		- 6 monthly clinical examination, plain films of primary site

Years 3 - 5		- annual clinical examination, plain films of primary site[1] , MRI primary site if difficult to assess clinically
Discharge at 5 years from surgery, unless: Patient has a metallic prosthesis *in situ* (follow-up evaluation in physiotherapist-led endoprosthesis follow-up clinic)		
Localised post primary treatment – on observation only		
Years 1 - 2		- interval MRI scans at 6 months and 18 months. If no change, and patient does not want curettage, discharge.
3. Grade 2 – 3 chondrosarcoma, periosteal and parosteal osteosarcoma	Plain x-ray and MRI of primary site, CT chest	
Localised post-resection		
Years 1 - 2		- post-operative visit in first 6 weeks - 3 monthly clinical examination, plain films of

		primary site[1], CXR
Years 3 – 5		- 6 monthly clinical examination, plain films of primary site[1], CXR
Years 6 - 10		- annual clinical examination, plain films of primary site[1], CXR
Discharge at 10 years from surgery, unless: Patient has a metallic prosthesis *in situ* (follow-up evaluation in physiotherapist-led endoprosthesis follow-up clinic)		
4. Chordoma	MRI of primary site (for sacral tumours, include whole pelvis), CT chest abdomen pelvis	
Localised post-resection +/- radiotherapy		
Years 1 - 2		- post-operative visit in first 6 weeks - 6 monthly clinical examination - MRI of primary site (for sacral tumours loco-regional imaging with MRI sacrum and pelvis), CXR at 6 months, 1 year, 2 years

Years 3 - 5		- 6 - 12 monthly clinical examination (depending on clinical need) - annual MRI of primary site (for sacral tumours loco-regional imaging with MRI sacrum and pelvis), CXR
Years 6 - 10		- annual clinical examination, CXR, MRI of primary site (for sacral tumours loco-regional imaging with MRI sacrum and pelvis)
Discharge at 10 years from surgery (follow-up may be continued depending on clinical need, treatment toxicity)		
Definitive radiotherapy		
Years 1 - 5		- 6 monthly clinical examination (depending on clinical need) - annual CXR, MRI of primary site (for sacral tumours loco-regional imaging with MRI sacrum and pelvis)
Years 6 - 10		- annual clinical examination, CXR, MRI of primary site (for sacral tumours loco-regional

		imaging with MRI sacrum and pelvis)
Discharge at 10 years from radiotherapy (follow-up may be continued depending on clinical need, treatment toxicity)		

[1] Plain films not required after amputation

3. Follow-up guidelines for high grade osteosarcoma/spindle cell sarcoma of bone/ dedifferentiated chondrosarcoma/ adamantinoma†

Stage of disease	Staging investigations	Follow-up schedule
1. Localised post primary treatment	Plain x-rays and MRI primary site; CT chest; whole body MRI[1], bone scan or PET scan[2]	
Year 1		- post-operative visit in first 6 weeks (if primary surgery) - end of treamtent gonadal function (males – testoterone, FSH, LH; females – oestrodiol, LH, FSH), biochemistry (U&Es, LFTs, Ca, PO4, Mg, HCO_3), audiology (if cisplatin-based chemotherapy) - echocardiogram (if anthracline-based chemotherapy) and GFR within 3 months of completing chemotherapy - 2 monthly clinical examination, CXR, plain films of primary site[3] - end of year 1 - gonadal function (males: testosoterone, LH, FSH; females: oestradiol, LH, FSH)[2]; blood biochemistry (U&E, LFT, Ca, PO_4, Mg, HCO_3)[4];

		audiology
Year 2 - 3		- 3 monthly clinical examination and CXR, plain films of primary site[3] - Year 2: blood biochemistry (Cr, Na, K, Ca, PO_4, Mg, HCO_3) [4] - Year 3: annual BP and urinalysis – if abnormal, check blood biochemistry (Cr, Na, K, Ca, PO_4, Mg, HCO_3) [4] - annual audiology if end of treatment audiology abnormal, or symptomatic - end of year 2 - echocardiogram[4]
Year 4		- 6 monthly clinical examination and CXR, plain films of primary site[1] site[3] - annual BP and urinalysis – if abnormal, check - blood biochemistry (Cr, Na, K, Ca, PO_4, Mg, HCO_3) [4] - end of year 4 - echocardiogram[4]
Year 5		- 6 monthly clinical examination and CXR, plain films of

		primary site[3] - blood biochemistry (Cr, Na, K, Ca, PO_4, Mg, HCO_3) [4] - audiology if end of treatment audiology abnormal, or symptomatic - end of year 5 - echocardiogram[4]
Years 6 - 10		- annual clinical examination and CXR, plain films of primary site[3] - annual BP and urinalysis for protein, if abnormal check blood biochemistry (Cr, Na, K, Ca, PO_4, Mg, HCO_3) [4]
Discharge at 10 years after surgery, unless: - Patient has treatment-related toxicity that requires long term follow-up - Patient has a metallic prosthesis *in situ* (follow-up evaluation in physiotherapist-led endoprosthesis follow-up clinic) - Patients who were children, teenage or young adult at time of diagnosis will require referral into the UCH Late Effects Service: ucl-tr.LateEffectServ@nhs.net - Clinical trial patients on active follow-up		

2. Post pulmonary metastatectomy		
Year 1		- post-operative visit in first 6 weeks - 3 monthly clinical examination, CXR, plain films of primary site - baseline CT scan post surgery, thereafter 6 monthly
Year 2		- 3 monthly clinical examination, CXR, plain films of primary site - 6 monthly CT scan
Years 3 – 5		- 6 monthly clinical examination, CXR, plain films of primary site
Years 6 – 10		- annual clinical examination, CXR, plain films of primary site
Discharge at 10 years after surgery		
3. Relapsed metastatic		

disease		
Year 1+		- 2 - 3 monthly clinical examination and CXR - imaging of disease sites as clinically appropriate

† Patients treated within clinical trials should be followed up according to the trial protocol.

[1] Whole body MRI for primary bone sarcoma patients receiving (neo)adjuvant chemotherapy.

[2] PET scan for primary bone sarcoma patients receiving neoaduvant chemotherapy for response assessment (particularly head and neck, pelvic sarcomas)

[3] Plain films not required after amputation

[4] Investigations to be carried out in patients who have had chemotherapy only

4. Follow-up Guidelines for undifferentiated small round cell sarcomas of bone and soft tissue (Ewing's sarcoma/round cell sarcoma with EWSR1-non-ETS fusions/CIC-rearranged sarcoma/sarcoma with BCOR genetic alterations/desmoplastic small round cell tumour/rhabdomyosarcoma/mesenchymal chondrosarcoma†

Stage of disease	Staging investigations	Follow-up schedule
1. Localised post primary treatment	Plain x-rays and MRI of primary site, CT chest, whole body MRI and/or PET scan (bone marrow aspirate and trephine no longer mandated)	
Year 1		- end of treamtent gonadal function (males – testoterone, FSH, LH; females – oestrodiol, LH, FSH), biochemistry (U&Es, LFTs, Ca, PO4, Mg, HCO_3), audiology (if cisplatin-based chemotherapy) - echocardiogram[3] (if anthracline-based chemotherapy) and GFR within 3 months of completing chemotherapy - 2 monthly clinical examination, CXR, plain films of primary bony site[1] - soft tissue tumours – baseline end of treatment MRI/CT primary site, thereafter at clinician's discretion[2]

		- radiotherapy as definitive local treatment - baseline end of treatment MRI/CT of primary site, then at 6 and 12 months - end of year 1 - gonadal function (males: testosterone, LH, FSH; females: oestradiol, LH, FSH); renal function (Cr, Na, K, Ca, PO_4, Mg, HCO_3)
Year 2 - 3		- 3 monthly clinical examination, CXR, plain films of bony primary site[1] - MRI of soft tissue primary site at clinician's discretion[2] - radiotherapy as definitive local treatment - MRI/CT of primary site at 18 and 24 months - Year 2: blood biochemistry (Cr, Na, K, Ca, PO_4, Mg, HCO_3) [4] - Year 3: annual BP and urinalysis – if abnormal, check blood biochemistry (Cr, Na, K, Ca, PO_4, Mg, HCO_3) [4] - end of year 2 – echocardiogram[3]
Year 4		- 6 monthly clinical examination, CXR, plain films of

		primary site[1] - MRI of soft tissue primary site at clinician's discretion[2] - annual BP and urinalysis – if abnormal, check - blood biochemistry (Cr, Na, K, Ca, PO_4, Mg, HCO_3) [4] - end of year 4 – echocardiogram[3]
Year 5		- 6 monthly clinical examination, CXR, plain films of primary site[1] - blood biochemistry (Cr, Na, K, Ca, PO_4, Mg, HCO_3) [4] - end of year 5 – echocardiogram[3]
Years 6 - 10		- annual clinical examination, CXR, plain films of primary site[1] - annual BP and urinalysis for protein, if abnormal check blood biochemistry (Cr, Na, K, Ca, PO_4, Mg, HCO_3) [4]
Discharge at 10 years after surgery, unless: Discharge at 10 years after surgery, unless: - Patient has treatment-related toxicity that requires long term follow-up		

- Patient has a metallic prosthesis *in situ* (follow-up evaluation in physiotherapist-led endoprosthesis follow-up clinic) - Patients who were children, teenage or young adult at time of diagnosis will require referral into the UCH Late Effects Service: ucl-tr.LateEffectServ@nhs.net - Clinical trial patients on active follow-up		
2. Relapsed metastatic disease		
Year 1+		- 2 - 3 monthly clinical examination and CXR - imaging of disease sites as clinically appropriate

† Patients treated within clinical trials should be followed up according to the trial protocol.

[1] Plain films of primary site not required after amputation

[2] If clinical detection of recurrence is anticipated to be difficult

[3] Perform only for patients who have received doxorubicin

5. Follow-up Guidelines for Gastrointestinal Stromal Tumours†

Stage of disease	Staging investigations	Follow-up schedule
1. Post-resection of localised disease	CT chest abdomen pelvis	
Very low risk[1]		No follow-up required – discharge to primary care
Low risk[1]		
Year 1		- CT abdo/pelvis[2] +/- CXR[3] at 12 months post-surgery. Then discharge.
Intermediate risk[1]		
Year 1		- baseline CT abdo/pelvis[2] +/- CXR[3] post-surgery and 6 months later
Years 2 - 5		- annual CT abdo/pelvis[2] +/- CXR[3]
Discharge after 5 years after surgery		
High risk (no adjuvant therapy)[1]		

Years 1 - 2		- 3 monthly clinical examination and CT abdo/pelvis[2] +/- CXR[3]
Years 3 - 5		- 6 monthly clinical examination and CT abdo/pelvis[2] +/- CXR[3]
Years 6 -10		- annual clinical examination - annual CT abdo/pelvis[2] +/- CXR[3] year 5, then stop
High risk (adjuvant therapy)		
Years 1 – 3 post treatment		- 3 monthly clinical examination and CT abdo/pelvis[2] +/- CXR[3]
Years 4 – 5 post treatment		- 6 monthly clinical examination and CT abdo/pelvis[2] +/- CXR[3]
Years 6 – 10 post treatment		- annual CT abdo/pelvis[2] +/- CXR[3]
Discharge at 10 years after surgery/end of adjuvant treatment		

Adjuvant imatinib		
Years 1 - 3		- 6 monthly clinical examination and CT abdo/pelvis[2] +/- CXR[3]
Years 4 - 5		- 3 – 6 monthly (at clinician's discretion) clinical examination and CT abdo/pelvis[2] +/- CXR[3]
Years 6 - 10		- annual clinical examination and CT abdo/pelvis[2] +/- CXR[3]
Discharge at 10 years after surgery		
2. Post-resection of localised disease following neo-adjuvant imatinib		As for high risk resected patients (above)
Discharge at 10 years		
3. Metastatic disease (1st line treatment)	CT chest abdomen pelvis	
Years 1 - 5		- 3 - 6 monthly clinic review, CT abdo/pelvis +/- CXR[2,3] (frequency depends on disease volume,

		gene mutation status, patient circumstances)
Years 6 – 10		- 6 monthly clinical review, CT abdo/pelvis +/- CXR[2,3,]
Year 11 onwards		- 6 monthly clinic review - annual CT abdo/pelvis +/- CXR[2,3,]
4. Metastatic disease (2st line+ treatment)		
Years 1 - 5		- 3 - 6 monthly clinic review, CT abdo/pelvis +/- CXR[2,3] (frequency depends on disease volume, gene mutation status, patient circumstances)

† Patients treated within clinical trials should be followed up according to the trial protocol.

[1] Risk grouping as defined in: Miettinen M, Lasota J. Semin Diagn Pathol. 2006 May;23(2):70-83

[2] CT may be replaced by MRI at clinician's discretion

[3] CXR at discretion of clinician; may be replaced by CT chest for syndromic and paediatric GIST