# West Yorkshire & Humber Sarcoma Service Guidelines

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<th>West Yorkshire &amp; Humber Sarcoma Service Guidelines</th>
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<tbody>
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<tr>
<td>Reviewed</td>
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<td>June 2020 (or before if new guidance becomes available)</td>
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**Proposed Target Audience for Consultation / Final Statement**

- WY&H, and HC&V Cancer Alliances:
  - Sarcoma MDT Teams
  - Lead Cancer Nurses
  - Lead Cancer Managers
  - Lead Cancer Commissioners

**Proposed Circulation List for Final Statement**

All WY&H Cancer Alliance guidelines will be made available electronically on the Wakefield CCG website (until the WY&H CA website is up and running). No hard copies will be supplied.

**Contact details**

- West Yorkshire & Harrogate Cancer Alliance
- NHS Wakefield CCG
- White Rose House
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Valid on the date of publication
1 Introduction

The Specialist Soft Tissue Sarcoma service based at Leeds Cancer Centre is a multidisciplinary group serving a catchment population of approximately 4 million across the West Yorkshire and Humberside area.

The referring Trusts include:

- Airedale NHS Foundation Trust
- Bradford Teaching Hospitals NHS Foundation Trust
- Calderdale and Huddersfield NHS Foundation Trust
- Harrogate and Direct NHS Foundation Trust
- Hull and East Yorkshire Hospitals NHS Trust
- Leeds Teaching Hospitals NHS Trust
- Mid Yorkshire Hospitals NHS Trust
- North Lincolnshire & Goole Hospitals NHS Foundation Trust
- York Hospitals NHS Foundation Trust

Choice should be offered to patients in the NLAG catchment as to their preferred location of care: Sheffield or Leeds/Hull. The latter is of high importance in relation to continuity of care and non-surgical interventions.

Bone sarcomas are referred onward to The Royal Orthopaedic Hospital, Birmingham.

Within each locality there will be a local ‘champion’ who will triage referrals within the district. The best placed individual to carry out this service is a Consultant Musculoskeletal Radiologist. Referrals in that geographical region from General Practitioners or Secondary Care are channeled to the local Musculoskeletal Radiology Department and initially an ultrasound is performed. That ultrasound examination should allow differentiation between those patients who will still need to remain within the overall Soft Tissue Sarcoma Service sphere and many other patients who have different conditions, such as bursa, ganglions or are related to other arthritic or degenerative conditions. If the ultrasound suggests an ongoing potential for the mass to be a Soft Tissue Sarcoma then the local radiologist will proceed to arrange an MRI scan and either prior to or subsequent to that scan, depending on the overall opinion, refer the imaging for discussion at the WY&H Soft Tissue Sarcoma MDT.

A decision will be made at the MDT meeting as to the imaging findings and whether the patient needs to be seen in the Leeds clinic, undergo a biopsy or go straight to an out-patient consultation with Soft Tissue Sarcoma clinical staff. Other diagnoses, for example where the imaging suggests a simple although large lipoma, can be conveyed to the locality Musculoskeletal Radiologist and the patient’s General Practitioner with the recommendation that the patient can be referred either to their local hospital for any surgery that may be thought necessary depending on symptoms. On the feedback correspondence to the General Practitioner, we try to enclose names of the Consultant Surgeons in the local district general hospital who are most likely to take an interest in these masses and also provide follow up information to us as required.

This system has run for several years and is satisfactory. It has allowed us to minimize the number of patients who have to travel to see us where there is a very low index of suspicion that their diagnosis is of Soft Tissue Sarcoma and to perform all biopsies centrally in our clinic. We adopt the same approach of the initial triage by the Musculoskeletal Radiologist for individuals resident in Leeds. We feel that it is both efficient and equitable system.
2 WY&H Sarcoma MDT Aims

WY&HSS adopts the following principles in patient management

- Inform, involve and support the patient at all times
- Attempt cure when able
- Appropriately balance morbidity of therapy against likelihood of cure
- Maximise function at all times

These aims are written in recognition that the therapy of STS can be functionally debilitating when potentially curative interventions are undertaken. Despite treatment with curative intent many patients will relapse with subsequent life limiting disease. Knowledge of the natural history of the disease in question, its treatment, the likelihood of therapeutic success and the informed wishes of the patient are essential in decision making.

Current international opinion in the management of localised STS argues that over-aggressive local control at the expense of function is more inappropriate than local failure with preserved function. This is counter to a central tenet of oncology and must be balanced against an understanding that local therapy must be appropriate in the context of the anticipated natural history of the individual patient’s disease, the individual patient’s rehabilitative potential and any options for further local therapy on relapse.
### 3 Membership –

#### 3.1 Core Members

<table>
<thead>
<tr>
<th>Member</th>
<th>Role</th>
<th>Cover</th>
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<tbody>
<tr>
<td>Mr IM Smith</td>
<td>Consultant Surgeon and MDT Lead.</td>
<td>Mr Jay Wiper</td>
</tr>
<tr>
<td>Mr Jay Wiper</td>
<td>Consultant Surgeon</td>
<td>Mr. Ian Smith</td>
</tr>
<tr>
<td>Mr. G. Toogood</td>
<td>Consultant Surgeon</td>
<td>Mr A Barlow</td>
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<tr>
<td>Mr A Barlow</td>
<td>Consultant Surgean</td>
<td>Mr G Toogood</td>
</tr>
<tr>
<td>Dr. D. Stark</td>
<td>Consultant Medical Oncologist</td>
<td>Dr. M. Marples</td>
</tr>
<tr>
<td>Dr. M. Marples</td>
<td>Consultant Medical Oncologist</td>
<td>Dr D. Stark</td>
</tr>
<tr>
<td>Dr G Bozas</td>
<td>Consultant Medical Oncologist (Hull &amp; East Yorkshire Hospitals NHS Trust)</td>
<td>Drs Marples and Stark</td>
</tr>
<tr>
<td>Dr. R. Turner</td>
<td>Consultant Clinical Oncologist, Lead for Clinical Trials.</td>
<td>Dr. P. Dickinson</td>
</tr>
<tr>
<td>Dr. P. Dickinson</td>
<td>Consultant Clinical Oncologist</td>
<td>Dr. R. Turner</td>
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<tr>
<td>Dr Rachel Barton</td>
<td>Consultant Clinical Oncologist (Hull &amp; East Yorkshire Hospitals NHS Trust)</td>
<td>Drs Turner and Dickinson</td>
</tr>
<tr>
<td>Dr. W. Merchant</td>
<td>Consultant Histopathologist</td>
<td>Dr. S. Edwards</td>
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<tr>
<td>Dr. S. Edwards</td>
<td>Consultant Histopathologist</td>
<td>Dr. W. Merchant</td>
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<tr>
<td>Ms. L. Omand</td>
<td>Sarcoma Nurse specialist</td>
<td>Mrs C Wallis</td>
</tr>
<tr>
<td>Mrs C. Wallis</td>
<td>Sarcoma Nurse Specialist</td>
<td>Mrs Katie Preer</td>
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<tr>
<td>Mrs Katie Preer</td>
<td>Sarcoma Nurse specialist</td>
<td>Mrs C Wallis</td>
</tr>
<tr>
<td>Ms S Edwards</td>
<td>Sarcoma Nurse Specialist (Hull &amp; East Yorkshire Hospitals NHS Trust)</td>
<td>C Wallis, Preer, Omand</td>
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<tr>
<td>Dr P Robinson</td>
<td>Consultant Radiologist</td>
<td>Dr’s Gupta, Thomson, Hyland, Kaye</td>
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<tr>
<td>Dr Harum Gupta</td>
<td>Consultant Radiologist</td>
<td>Dr’s Robinson, Thomson, Hyland, Kaye</td>
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<tr>
<td>Dr Rachel Hyland</td>
<td>Consultant Radiologist</td>
<td>Dr’s Gupta, Thomson, Robinson, Kaye</td>
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<tr>
<td>Dr Elen Thomson</td>
<td>Consultant Radiologist</td>
<td>Dr’s Gupta, Robinson, Hyland, Kaye</td>
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<tr>
<td>Dr Tom Kaye</td>
<td>Consultant Radiologist</td>
<td>Dr’s Gupta, Thomson, Hyland, Robinson</td>
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<tr>
<td>Miss S Begum</td>
<td>Sarcoma MDT Co-ordinator</td>
<td>Mr T Hughes</td>
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<tr>
<td>Mr T Hughes</td>
<td>Sarcoma MDT support</td>
<td>Miss S Begum</td>
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### 3.2 Extended Member

<table>
<thead>
<tr>
<th>Member</th>
<th>Role</th>
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<tbody>
<tr>
<td>Lynn Hopcroft</td>
<td>Senior Sarcoma Physiotherapist</td>
</tr>
<tr>
<td>Debbir Artis</td>
<td>Sarcoma physiotherapist</td>
</tr>
<tr>
<td>Mandy Brown</td>
<td>Sarcoma Admin</td>
</tr>
<tr>
<td>Prof Kay</td>
<td>Consultant Plastic surgeon. Core member if major nerve MDT</td>
</tr>
<tr>
<td>Mrs Suzanne Kite</td>
<td>Palliative Care Consultant</td>
</tr>
<tr>
<td>Tim Broadhead</td>
<td>Consultant in Surgical Gynaecology</td>
</tr>
<tr>
<td>Prof Pete Sagar</td>
<td>Consultant Colo-rectal surgeon</td>
</tr>
<tr>
<td>Mr K Papagiannopoulos</td>
<td>Consultant Thoracic Surgeon</td>
</tr>
<tr>
<td>Mr Mark Liddington</td>
<td>Consultant Head and Neck Plastic Surgeon</td>
</tr>
<tr>
<td>Mr Simon Dexter</td>
<td>Consultant GI surgeon</td>
</tr>
<tr>
<td>Dr Graeme Stables</td>
<td>Consultant Dermatologist</td>
</tr>
<tr>
<td>Mr Alan Paul</td>
<td>Consultant Urologist</td>
</tr>
<tr>
<td>Mr R Achuthan</td>
<td>Consultant Breast Surgeon</td>
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## 4 Location & administration

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<tr>
<th>Function</th>
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<tr>
<td><strong>Administration</strong></td>
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<tr>
<td>Sarcoma Office</td>
<td>SJUH</td>
<td>Level 3 Bexley wing</td>
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<tr>
<td>MDT</td>
<td>SJUH</td>
<td>Monday 1pm-4pm Level 7 Bexley wing</td>
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<tr>
<td><strong>Surgery</strong></td>
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<tr>
<td>Mr Smith</td>
<td>LGI</td>
<td>Wednesday all day Other lists TBC</td>
<td>Tuesday 9am-12pm</td>
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<tr>
<td>Mr Wiper</td>
<td>LGI</td>
<td>TBC</td>
<td>Tuesday 9am-12pm</td>
<td></td>
</tr>
<tr>
<td>Mr Toogood</td>
<td>SJUH</td>
<td>TBC</td>
<td>Mon 3pm-4.30pm</td>
<td></td>
</tr>
<tr>
<td>Mr Barlow</td>
<td>SJUH</td>
<td>TBC</td>
<td>Thurs 9-12pm</td>
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<td><strong>Non-Surgical Oncology</strong></td>
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<tr>
<td>Pre/Post treatment</td>
<td>SJIO</td>
<td>Monday 09.15-12.00 Clinic suite 1</td>
<td>Ward 97</td>
<td></td>
</tr>
<tr>
<td>On treatment</td>
<td>SJIO</td>
<td>Friday 09.00-12.00 Princess Royal suite</td>
<td>Ward 97</td>
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<tr>
<td><strong>Radiology</strong></td>
<td></td>
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<tr>
<td>Biopsy</td>
<td>Chapel Allerton</td>
<td>Tuesday/Friday</td>
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5 Referral

Route of referral depends upon the level of clinical suspicion and the investigations performed up to the point of referral. Soft tissue masses SHOULD NOT be biopsied without prior discussion with the WY&HSS MDT.

1. Uncharacterised soft tissue mass with suspicious clinical features
2. Uncharacterised soft tissue mass without suspicious clinical features
3. Soft tissue masses that may be nodal
4. Suspicious soft tissue mass following imaging
5. Suspicious histology
6. Third party referrals from recognised sarcoma centres
7. Patients with isolated bone lesions suspicious for primary sarcoma of bone

5.1 Referral route

5.1.1 Uncharacterised soft tissue mass with suspicious clinical features

Established risk factors that should prompt suspicion for malignant versus benign soft tissue masses are:

- Size > 5 cm
- Deeply located (with respect to investing fascia)
- Rapid growth
- Symptomatic
- Recurrence after previous resection

The presence of one or more the above should prompt urgent referral via the ‘fast-track’ cancer waiting time (CWT) process. Patients will be seen within 14 days. These criteria are neither sensitive nor specific nor rigorously evidenced as criteria for referral (they have emerged by consensus view). Ultrasound imaging based triage will be performed locally. Patients may be reassured and discharged if imaging is unconcerning without need for review by WY&HSS. Verbal and written patient information should be provided and referral made using the referral proforma.

5.1.2 Uncharacterised soft tissue mass without suspicious clinical features

The likelihood of benign disease increases with smaller tumours that are unchanging. Reducing the 'size cut-off' may allow earlier detection (and better outcomes) for sarcomas but increases the probability that the tumour will be benign. This may have an impact upon patient anxiety and inconvenience and WY&HSS capacity to deal with the specialist requirements of true sarcomas. The following clinical features would tend to support a benign diagnosis.

- Less than 5 cm and superficial to deep fascia
- Local inflammatory change
- History and consistent signs of local trauma
- Documented no change in size over 6 weeks
- Presence of pathognomic features (punctum, transillumination, tenderness)
- Associated comorbidity predisposing to benign masses (such as inflammatory arthritis)

If a benign diagnosis is suspected but confirmation is required a non-urgent referral is required. Patients will be seen within 28 days of referral. The process described as per (1) will otherwise be followed.

5.1.3 Soft tissue masses that may be nodal

Soft tissue sarcomas very rarely involve nodal masses. Suspected nodal masses arise most commonly in the context of malignancies of the head & neck, breast, pelvis, cutaneous squamous carcinoma and melanomas, lymphoma, occult/unknown primary carcinomas and benign reactive or inflammatory conditions. Patients with suspected nodal masses should prompt a systematic review of symptoms, clinical examination and fast-track referral to the most appropriate site-specific team for work-up rather than referral to the WY&HSS. An FNAC is inadequate to exclude soft tissue sarcoma or lymphoma and a core biopsy is always required. Masses referred to the sarcoma service that are subsequently found to be nodal may result in delayed site-specific management.

5.1.4 Suspicious soft tissue mass following imaging

Patients whose images are entirely consistent with benign disease do not need to be referred to the WY&HSS. Referral to WY&HSS is needed if imaging has shown:

- Lipoma requiring further evaluation
- Indeterminate features
- Possible sarcoma

Radiological criteria are listed in the imaging guidelines. Referral to the WY&HSS should be made via the WY&HSS MDT coordinator and requires completion of a referral proforma and image transfer of appropriate images for review. Failure to provide required supporting material may delay subsequent management.

5.1.5 Suspicious histology

Inappropriate biopsy and/or resection of soft tissue sarcoma by a non-specialist team is associated with worse outcomes (tumour control and function). For cases when biopsy/resection has been performed and sarcoma is only suspected following histological review, WY&HSS review is advised. Referral to the WY&HSS should be made via the WY&HSS MDT coordinator and requires completion of a referral proforma and transfer of appropriate images and tissue blocks and slides. Written operative notes, radiology and histology reports from the referring team are also required. Failure to provide required supporting material may delay subsequent management.

5.1.6 Third party referrals from recognised cancer centres

Patients whose diagnostic pathway and/or initial treatment has been started at another recognised sarcoma centre may be referred to WY&HSS for further management. A written summary of management to date is required rather than a proforma. Histology review is not usually required but full reports are needed. Provision of imaging including written image reports as well as operative notes (when performed) is essential. Image review may be required for assessment of treatment response and/or baseline status for follow-up.
5.1.7 Patients with isolated bone lesions suspicious for primary sarcoma of bone

WY&HSS is not recognised as a bone sarcoma centre. Primary malignant bone tumours are rare. In older patients isolated destructive bone lesions are more likely to represent bony metastatic disease arising from a carcinoma at another site. Bone sarcomas are commoner (but still rare) in patients under the age of 40 years. Patients with radiologically suspected sarcoma of bone should be referred directly to the Royal Orthopaedic Hospital, Birmingham. The flow chart for management of suspected primary bone sarcoma should be used. Note that this pathway does not include referral to the WY&HSS MDT. Direct liaison with existing cancer MDTs, or ROH Birmingham or the named clinicians is required.
6 (WY&HSS) Sarcoma Referral, Diagnostic and Treatment Pathway for Primary and Secondary Care

WySH Pathway for Urgent Symptoms of Soft Tissue Sarcoma (V4 June 2017)
No metastases—surgery or pre-op radiotherapy. Metastases—care taken over by non surgical oncology

Maximum of 2 Weeks if referred as urgent suspected cancer (Cancer Waiting Times Target)

Referral may be sent as urgent if >4cm, deep/palpable, increasing in size, symptomatic, site of previous surgical resection (primary sarcoma)

Maximum of 2 Weeks if referred as urgent suspected cancer (Cancer Waiting Times Target)

GR Division referral

Local Hospital Imaging (X-ray or MRI)

Soft Tissue Tumour

Suspected Malignant

If imaging sufficient refer directly to Leeds MDT

If imaging not sufficient refer to a GP

Refer to MDT at Leeds

Local further imaging if required (MRI)

Ultrasound biopsy

MDT Meeting at Leeds

Pathology reviewed by sarcoma pathologists. All small cell sarcomas have molecular cytogenetic testing

Treatment plan discussed with patient same day

Malignant?

CT Staging

No metastases?

Care provided via Leeds Sarcoma Clinic

Metastases present?

Care taken over by Medical Oncology & Palliative Care Community Care

& GD aged 1625 refer to TV/9

Surgery

Within 8 days of any urgent referral (Cancer Waiting Times)

Radiotherapy/Chemotherapy

Within 8 days of any urgent referral (Cancer Waiting Times)

Follow agreed shared care pathways where appropriate

Local Supportive and Palliative Care Pathway followed at all appropriate stages

Refer either as a GP referral or a radiology referral (using appropriate form)

For details of laboratory, pathologists and histopathological investigations—see local R&S guidelines

Notes

If suspected bone cancer then refer directly to Royal Orthopaedic Hospital, Birmingham

Valid on the date of publication

Version 4.0
WY&H Pathway for Urgent Symptoms of Soft Tissue Sarcoma (cont'd)

Care provided via Leeds Sarcoma Clinic

Surgery and consideration of pre/post operative radiotherapy

Ward discharge for clinic appointment

Discussion at Post-op MDT Meeting at Leeds

Non Surgical Oncology Care taken over by Medical Oncology & Palliative/Community Care

(If aged 16-25 refer to TYA)

Radiotherapy/Chemotherapy

2nd line treatment if appropriate. Within 31 days of decision to treat (Cancer Waiting Times Target)

Sarcoma clinic follow up

Oncology clinic follow up

End of Life Care if appropriate

Notes

All follow up to be undertaken in accordance with LRSS Guidelines

Follow-up to be shared with referring team whenever appropriate

Local Supportive and Palliative Care Pathway followed at all appropriate stages

Valid on the date of publication
Version 4.0
6.1 Appendix 1 – Additional Pathway Information

<table>
<thead>
<tr>
<th>Version Control</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Version/Draft</strong></td>
</tr>
<tr>
<td>1.0</td>
</tr>
<tr>
<td>3.0</td>
</tr>
</tbody>
</table>

6.1.1 Referral

Cutaneous lesions should first follow the local Melanoma/non-Melanoma pathways. Suspected groin/axilla/neck lymph nodes should follow the local referral pathways to appropriate site specific assessment (see section 5.1.3).

Referral for ultrasound should be sent as urgent if:
- >5cm
- Deep/fixed
- Increasing Size
- Symptomatic
- Site of previous surgical resection (for sarcoma)

Referral criteria to be regularly audited.

6.1.2 Triage/ Diagnosis

- All imaging to be undertaken in accordance with agreed WY&HSS Guidelines
- Ultrasound to be undertaken in accordance with agreed Cancer Alliance standards
- Diagnostic machine used must be subject to 6 monthly QA for image acquisition and production
- Report to include confirmation/details of MRI and request that patient is referred to MDT
- MRI undertaken in accordance with agreed Cancer Alliance protocols.
- Images to be rapidly sent to MDT
- IOG – required Triple Assessment undertaken at this point
- Biopsy to be undertaken by core member of MDT
- Access to on site molecular/cytogenetics required for small cell sarcomas and GIST
- All biopsies should be referred either directly or for confirmatory opinion to a specialist pathologist
- All histopathological/histochemical investigations and their specific indications will be in accordance with the local WY&HSS Guidelines

6.1.3 Treatment

- Surgery to be undertaken at the agreed Specialist Treatment Centre (Leeds), unless by specific agreement with the WY&HSS MDT
- IOG – compliant membership required for MDT
- Radiotherapy and Chemotherapy to be undertaken at agreed designated Centre in Leeds or by Core MDT members based in Hull using agreed guidelines (this document)

6.1.4 Follow-Up

All follow-up to be in accordance with the agreed WY&HSS Guidelines. If in doubt, contact should be made with the Sarcoma MDT via the CNSs on 0113 2068902 or 0113 2068966.
7.1 Shared Care Pathway for non-sarcoma MDTs to access the Leeds Sarcoma MDT (excluding Gynaecology)

Internal Clinician Referral

Suspected Cancer
- Incidental Finding
- Symptomatic

Review at local MDT (Diagnostic)
- Imaging +/- Pathology

Confirmed Non Sarcoma

Local Pathway

Suspected Sarcoma

Referral to Leeds Sarcoma MDT via fax Number 0113 2068978 or for advice ring Dr Rob Turner 0113 2067406

Sarcoma MDT (follow Sarcoma Pathway)

http://nww.lhp.leedsth.nhs.uk/referral_info/sarcoma.asp

The above pathway can be used by the following MDTs

- Colorectal
- Upper GI (excl GIST)
- Head & Neck
- Urology
- Lymphoma
- Skin/Melanoma
- Lung
- Breast
- TYA
7.2 Shared Care Pathway for Gynaecology MDT to access the Leeds Sarcoma MDT (recognising high level expertise in diagnostic and local management of gynaecological sarcoma)

Internal Clinician Referral

Suspected Cancer
- Incidental Finding
- Symptomatic

Review at local MDT (Diagnostic)
- Imaging +/- Pathology

Confirmed Non Sarcoma

Local Pathway

Suspected Sarcoma

1. Manage as per WY&H CA Gynae Oncology Guidelines
2. Advise Leeds Sarcoma MDT (fax 0113 2068978) in order to register
3. On progression/inoperable refer to Leeds Sarcoma MDT as above or for advice ring Rob Turner 0113 2067406

Sarcoma MDT (follow Sarcoma Pathway)

http://nww.lhp.leedsth.nhs.uk/referral_info/sarcoma.asp
8 Referral Forms

8.1 Primary Care Urgent Referral Forms

The Leeds Teaching Hospitals NHS Trust
SUSPECTED SOFT TISSUE SARCOMA
URGENT (WITHIN 14 DAYS) APPOINTMENT REQUEST

EMAIL: leedsth-tr.FastTrackTeam@nhs.net / PHONE: 0113-2065141

N.B - Please note that up to date patient contact details and a telephone number where the patient can be reached during office hours (08.00 am - 17.00 pm) are essential to allow us to offer your patient a date within seven days of your referral.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Referrer</th>
</tr>
</thead>
<tbody>
<tr>
<td>Title</td>
<td>Surname</td>
</tr>
<tr>
<td>First Name</td>
<td></td>
</tr>
<tr>
<td>Address</td>
<td></td>
</tr>
<tr>
<td>Postcode</td>
<td>Date of Birth</td>
</tr>
<tr>
<td>Gender</td>
<td>Age</td>
</tr>
<tr>
<td>Telephone (Home)</td>
<td></td>
</tr>
<tr>
<td>Telephone (Work)</td>
<td></td>
</tr>
<tr>
<td>Telephone (Mobile)</td>
<td>Date of decision to Refer</td>
</tr>
<tr>
<td>NHS Number</td>
<td>Date of Referral</td>
</tr>
<tr>
<td>E-mail address (Please print)</td>
<td></td>
</tr>
</tbody>
</table>

| Is the patient aware of the possible diagnosis of cancer? | |
| 2 week wait patient information leaflet given? | |
| Is the patient available and willing to attend an appointment within the next 14 days? | Y N |
| If not, refer when willing and able to attend | Y N |

Tumour Details – Suspected Cancer (please tick box)

Upper Limb | Lower Limb | Thorax | Head/Neck | Abdomen | Pelvis | POSITION | Left | Right | Midline | Superficial |

Deep Size (cm) ………………

| Is the patient currently taking Warfarin or Clopidogrel? | Yes | No |

Soft Tissue Sarcoma

Ultrasound findings that are suggestive of soft tissue sarcoma or ………………

Valid on the date of publication
Version 4.0
Ultrasound findings are uncertain and clinical concern persists

IMAGING MUST BE PERFORMED PRIOR TO REFERRAL*:
Investigations

- USS
- MRI
- CT
- BX

*Considerations:
Consider an urgent direct access ultrasound scan (to be performed within 2 weeks) to assess for soft tissue sarcoma in adults with an unexplained lump that is increasing in size.

Suspected Bone Sarcomas:
LTHT will not accept suspected bone sarcoma as LTHT is not commissioned for this service. Please refer patient to the Royal Orthopaedic Hospital, Birmingham. Contact details below:

MDT Co-ordinator - 0121 6854314

Referral Details

Name: ___________________________
Role: ___________________________ Location: ___________________________

Valid on the date of publication
Version 4.0
# 8.2 Secondary Care Referral Proforma

## LEEDS SOFT TISSUE SARCOMA SERVICE

New case notification and request for further information.
Please complete the following form, ensuring all information requested is completed to avoid delays in potential treatment. Additional information that you feel appropriate is also welcomed.

### PATIENT DETAILS

<table>
<thead>
<tr>
<th>NAME</th>
<th>DOB</th>
<th>ADDRESS</th>
<th>TELEPHONE NO.</th>
<th>MOBILE NO.</th>
<th>NHS NUMBER</th>
<th>GP</th>
</tr>
</thead>
</table>

### REFERRER DETAILS

<table>
<thead>
<tr>
<th>NAME</th>
<th>DEPARTMENT</th>
<th>JOB TITLE</th>
<th>Is Patient aware of referral?</th>
<th>Has GP been informed of referral?</th>
<th>Has this been referred to Radiology as a 2ww?</th>
<th>If not are you upgrading the referral?</th>
<th>Is transport required?</th>
<th>Is the patient on any anti-coagulants?</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
</tr>
</tbody>
</table>

### TUMOUR DETAILS

<table>
<thead>
<tr>
<th>SITE</th>
<th>POSITION</th>
<th>IS IT</th>
<th>SIZE (cm)</th>
<th>SYMPTOMS (If any, e.g. pain, recent trauma)</th>
<th>INVESTIGATIONS SO FAR, DATE AND LOCATION.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Head/Neck</td>
<td>Left, Right</td>
<td>Superficial</td>
<td>...</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>Upper Limb</td>
<td></td>
<td>Deep</td>
<td>...</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>Lower Limb</td>
<td></td>
<td></td>
<td>...</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>Thorax</td>
<td></td>
<td></td>
<td>...</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>Abdo</td>
<td></td>
<td></td>
<td>...</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>Pelvis</td>
<td></td>
<td></td>
<td>...</td>
<td>...</td>
<td>...</td>
</tr>
</tbody>
</table>

Thank you for completing this form.
Please fax to the sarcoma office 0113 2068978 or e-mail: leedsth-tr.FastTrackTeam@nhs.net
9 Pathway for Patients Presenting with Apparently Isolated Bone Lesions on Imaging

WY&H Soft Tissue Sarcoma Service
Pathway for patients presenting with apparently isolated bone lesions on imaging

NOTE: DO NOT refer to Sarcoma Office at Leeds General Infirmary

Valid on the date of publication
Version 4.0
10 Imaging guidelines

10.1 Bone sarcoma

Bone sarcoma is a radiological diagnosis. Suspected bone sarcoma should be referred directly to a specialist bone sarcoma MDT. WY&HSS is not recognised as a bone sarcoma centre and referral of bone sarcomas to WY&HSS may delay referral for definitive treatment.

10.2 Soft Tissue Sarcoma

10.2.1 Diagnosis

Patient attends GP

Soft tissue Mass present and assessed clinically:

1. Observe/Discharge

2. Investigate
   a) 2 week
   b) 4 week

Ultrasound

1. Performed/supervised by clinician – FRCR/ RCR accredited to perform/report ultrasound (preferably MSK but not vital).


3. Ultrasound machine used must be of diagnostic/medical standard with at least 6 monthly QA of electrical safety, transducer, machine and monitor quality.

4. Ultrasound examination to assess – mass size, mass location (relation to fascia), echotexture, cyst/solid/mixed, Doppler characteristics.

5. If diagnostic for non sarcoma (benign) (table 1) – report to GP.

6. If diagnostic for non sarcoma (malignant) by history and appearances – report to GP to refer to local oncology.

7. If diagnostic for lipoma but concerning symptoms (table 2) – report to GP & MRI (notify sarcoma service).
8. If suspicious for sarcoma or indeterminate (table 3)–report to GP & MRI (notify sarcoma service).

**MRI – performed within 2 weeks of Ultrasound (ideally <10 days)**

2. If claustrophobic refer to sarcoma service with ultrasound.
3. If diagnostic for non sarcoma (benign) (Table 1) – report to GP.
4. If diagnostic for non sarcoma (malignant) (Table 2) – report to GP to refer local oncology.
5. If suspicious for sarcoma or indeterminate (Table 3)–report to GP & MRI (notify sarcoma service).

**Sarcoma Service**

1. Review MR and US.
2. Keep biopsy appointment or
3. Postpone biopsy and advise MDT review first

**Continue on diagnostic pathway.**

**Biopsy**

1. Performed by sarcoma MDT member and send to sarcoma histopathologist.
2. Image guided – dependant on anatomical location and expertise will be either ultrasound, fluoroscopically or CT guided.
3. Non-image guided (in clinic) - dependant on anatomical location and expertise, may be preferential for expediting management decisions if radiology shows no requirement for targeting focal areas of the mass.
4. Samples should be obtained using a core needle and as large as possible (typically greater than 16 gauge) but will ultimately depend on location, lesion type and patient co-morbidity.
5. At least 2 samples should be sent preserved to histopathology and 1 sample fresh to cytogenetics if possible – variations on the need for further fresh specimens will depend on level of suspicion for differing suspected tumour types (see relevant guidelines – link e.g. lymphoma).

**10.2.2 Staging**

1. Primary (local) staging will be typically addressed by the diagnostic ultrasound and/or MRI.
2. Staging CT thorax will be performed routinely to address metastases for highly suspicious lesions prior to known biopsy results or after positive biopsy results.
3. Routine CT/MRI liver, bone or PET scanning is unnecessary except for specific histological subtypes (see relevant guidelines) as advised by WY&HSS MDT.

10.2.3 Relapse

1. Suspected early post treatment complications (< 4 weeks) can be initially assessed by ultrasound to detect abscess, seroma and haematoma.

2. Suspected tumour persistence or tumour recurrence will be typically best evaluated by MRI (may need contrast, see appendix 4). Alternative or additional use of ultrasound, CT, isotope bone scan and PET scanning will depend on the suspected tumour type or contraindications to MRI.

3. Biopsy may be necessary to clarify the diagnosis (same criteria as above).

10.2.4 Follow-up

See section on follow-up.

10.2.5 Table 1. Benign diagnoses.

<table>
<thead>
<tr>
<th>Category</th>
<th>Name</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Normal</td>
<td>No abnormality seen on ultrasound</td>
</tr>
<tr>
<td>2</td>
<td>Benign cyst or Ganglion cyst</td>
<td>Oval lesion, hypo-echoic centrally with a well defined wall and posterior acoustic enhancement</td>
</tr>
<tr>
<td>3</td>
<td>Benign vascular lesion</td>
<td>Solid or cystic structure with minor linear vascularity demonstrated on colour or power settings</td>
</tr>
<tr>
<td>4</td>
<td>Benign Other</td>
<td>Any lesion with either inflammatory characteristics or benign soft tissue mass</td>
</tr>
<tr>
<td>5</td>
<td>Lipoma</td>
<td>Homogenous hyper-echoic lesion within the dermis or deep fat planes, no flow within it on colour or power settings and causing no or minimal mass effect to the surrounding structures</td>
</tr>
</tbody>
</table>

10.2.6 Table 2. Lipoma requiring further evaluation.

<table>
<thead>
<tr>
<th>Category</th>
<th>Name</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>6</td>
<td>Lipoma requiring further evaluation</td>
<td>(i) Clinically painful, enlarging, greater than 5cm in size, deep to fascia or (ii) Lipoma but mild heterogenicity on ultrasound</td>
</tr>
</tbody>
</table>

10.2.7 Table 3. Indeterminate and Sarcoma

<table>
<thead>
<tr>
<th>Category</th>
<th>Name</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>7</td>
<td>Indeterminate</td>
<td>(i) Clinically painful or (ii) enlarging solid mass and no Doppler flow</td>
</tr>
<tr>
<td>8</td>
<td>Possible sarcoma</td>
<td>Solid, heterogeneous lesion with distortion of surrounding anatomy and disorganized vascularity on doppler flow</td>
</tr>
</tbody>
</table>
10.2.8 Protocol 4. MRI protocol guidelines

1. Mark mass with capsule(s)

2. MR imaging performed in 2 orthogonal planes, 5-6 mm slice thickness.
   a) Axial and Sagittal or Coronal
   b) T1 weighted (without fat suppression)
   c) T2 weighted with fat suppression

3. Fast spin echo sequences can be used to reduce motion artefact.

4. Gadolinium useful
   a) To determine solid or cystic
   b) Identify necrotic tumour or haematoma
   c) Patients who have had previous surgery or radiotherapy

5. If gadolinium given T1 fat saturated sequences should be performed post injection. It is
   NOT necessary to perform T1 fat saturated sequences pre gadolinium injection.

6. Small field of view is ideal but if very focussed should perform localiser views so
   subsequent clinicians involved can determine exact body positioning.
11 MDT meeting

The MDT meeting should facilitate appropriate multidisciplinary discussion of patients referred to or managed by WY&HSS. Responsibility for the organisation of the meeting lies with the WY&HSS office staff and case listing is as defined within the WY&HSS SOP. PPM should be used to schedule an agenda and populate clinical summaries.

Discussion requires timely provision of images and histopathology specimens.

Clinical background should be available from the referral proforma or, if already known to the MDT and referred by an WY&HSS MDT member, via a summary of the question to be addressed entered into an annotation on PPM. The MDT coordinator should be made aware by telephone or, ideally, e-mail (sairah.begum@nhs.net)

The MDT meeting should be chaired.

It is the responsibility of the chair to make an appropriate signed paper entry into the MDT record. A simultaneous ‘live’ entry onto PPM summarising the MDT discussion will be made by WY&HSS office staff. The MDT record should make clear the agreed:

- Radiology results
- Histology results
- Action plan
- Responsibility for action plan

11.1 Radiology results

Radiology reports may have already been issued. When a change to the report is made this should be recorded both in the MDT record and as an addendum to the issued report.

11.2 Diagnostic studies

Should include specific comment about the likelihood of malignancy and the need for further investigations. The size of lesions and relation to fascia should be reported.

11.3 Staging studies

As diagnostic studies. Also to include relationship to neurovascular structures and viscera. Presence or absence of metastatic lesions should be recorded.

11.4 Treatment response studies

Should include specific comment about the date of the baseline study as well as response to therapy according to either RECIST or Choi (GIST only) criteria.
11.5 Histology results

Histology reports may have already been issued. When a change to the report is made this should be recorded both in the MDT record and as an addendum to the issued report.

11.6 Action plan

It is essential that an outcome is recorded for each case discussion. Although free-text will be needed for radiology and pathology comments, most MDT outcomes can be recorded using a limited number of outcome codes.

11.7 Summary of outcomes

11.7.1 Missing data
Rediscuss at MDT meeting
Update named clinician directly (no need to rediscuss at MDT meeting)

11.7.2 Benign
Further imaging for interval change
Imaging for local treatment planning
Excision biopsy
Discharge

11.7.3 Likely benign
Further imaging for interval change
Imaging for local treatment planning
Excision biopsy

11.7.4 Probably malignant
Further imaging for diagnosis and staging
Imaging for local treatment planning
Core biopsy
Excisional biopsy
Incisional biopsy (exceptional circumstances only)
Planned marginal excision
Wide local excision

11.7.5 Malignant
Further imaging for diagnosis and staging
Imaging for local treatment planning
Core biopsy
Incisional biopsy (exceptional circumstances only)
Re-excision
Wide local excision
Planned marginal excision
Planned positive excision
Radical excision/amputation
Palliative surgery
Isolated limb infusion or perfusion
Systemic therapy
11.8 Responsibility for action plan

The MDT recommendation may have been made without face-to-face clinician review of the patient. The treating clinician retains the right to modify the MDT opinion based upon his/her assessment of the case-specific clinical features, including co-morbidities and general performance status, following discussion with the patient. Changes should be fed-back to the MDT coordinator.

Responsibility for action is assigned based upon submission of a radiology request, biopsy request, an out-patient clinic booking or a specifically addressed letter. An appropriate timescale should be indicated where necessary. Radiology or biopsy submissions should generate an MDT discussion following the requested examination.

11.9 MDT meeting letters

To aid communication and to support responsibility for action planning, a summary annotation or formal letter should be sent to involved clinicians following the MDT meeting.
12 Biopsy & STS specimen reporting

12.1 Introduction

This section supplements the Data set for Cancer Histopathology Reports on Soft Tissue Sarcomas, which is under consultation with the Royal College of Pathologists.

All soft tissue tumour cases will be selected for review as per the national and local guidelines. There are two specialist sarcoma pathologists in the network reporting the specimens, one of whom is the nominated lead pathologist. They will contribute to the regional sarcoma MDT, participate in the National Soft Tissue MDT and in local audit.

All patients with soft tissue tumours assessed in a diagnostic clinic should have their pathology reported by a specialist soft tissue pathologist.

12.2 Specimen Types

12.2.1 Fine needle Aspiration Cytology

Has only very limited role in the diagnosis of soft tissue sarcomas and should be avoided if possible.

12.2.2 Needle core biopsies

12.2.3 Open biopsies

12.2.4 Resection specimens

This includes large specimens, amputated limbs, limb girdle amputations, chest wall resections, retroperitoneal sarcomas and sarcomas associated with specific organs.

12.2.5 Molecular studies

As far as possible fresh tissue should be made available to the cytogenetics department based at St. James’s University Hospital for molecular studies. When fresh tissue is not available, six unstained sections on super frost slide should be sent for FISH analysis.

Specimens should be reported to an agreed time frame so as to allow appropriate clinical decision-making at a planned MDT meeting.

Clinical information required on the consent form

In addition to the demographic data, the following information should be included in the request form.
1. Duration, site, size and plane of the tumour (subcutaneous, intramuscular etc.)
2. History of relevant past illnesses, radiotherapy, chemotherapy or surgery.

Preparation of specimen prior to dissection

Undertaken in line with existing LTHT pathology laboratory procedures.
**Needle core biopsies**
Ideally, at least two samples should be provided to the histopathology department, one in formalin for histopathological examination and the other in tumour transport medium for cytogenetics. If tumour transport medium is not available, the tissue should be sent in normal saline and it should reach the Cytogenetics laboratory within four hours of the procedure.

**Open biopsies**
Should be sent fresh. If the specimen is large enough, small pieces of tissue can be taken for cytogenetics and also to be frozen in liquid nitrogen at 80 degree centigrade.

**Resection specimens**
Should be sent fresh. The specimen should be weighed, inked and measured. The specimen is then sliced. Small samples are removed for freezing and the sample for cytogenetics should be placed in tumour transport medium. The main specimen is placed in formalin for adequate fixation. Photograph of the specimen before and after slicing is desirable.

Blocks are taken to include the nearest resection margin. Lesions smaller than 5cm should be processed in its entirety. One block per cm of the longest diameter of the tumour should be ideally taken. Areas, which appear visibly different, require adequate extra sampling. In large liposarcomas, particularly of the retroperitoneum, any area with different colour or consistency should be adequately sampled to detect dedifferentiation.

---

**12.3 Core data for soft tissue sarcoma reporting**

**12.3.1 Clinical:**
- Site
- Plane of the tumour

**12.3.2 Macroscopic Description:**
- Type of excision
  - Incisional
  - Excisional
  - Radical
- Maximum tumour dimensions. Measurements should be given in three dimensions.
- Presence or absence of necrosis and percentage of necrosis.
- Other features of note.
  - Ossification.
  - Calcification.

**12.3.3 Microscopic Description:**
- Histological type
- Soft tissue sarcomas are categorised based on WHO consensus classification of 2002.
- Grade (FNCLCC)
  - 1
  - 2
  - 3
- Immunohistochemistry results.
- Tissue planes involved
  - cutaneous
  - subcutaneous
  - deep fascia
  - subfascial
  - intramuscular
- Status of margins
  - involved
  - marginal
  - wide
  - radical
12.4 Reporting of small biopsy specimens

The report should include the histological diagnosis with grade with the caveat that the excision specimen may have a higher grade. Immunohistochemical results should be included where appropriate. Results of molecular and cytogenetic studies can be issued as supplementary reports.

Points to note:

1. Extremity pleomorphic sarcomas with myogenic differentiation have a worse prognosis compared to others. Hence immunohistochemical positivity for myogenic markers such as smooth muscle actin, Desmin, smooth muscle myosin, H-Caldesmon or nuclear positivity for Myogenin or MyoD1 should be specifically noted. The pattern of staining should be mentioned i.e. diffuse, focal or occasional.

2. Retroperitoneal high-grade sarcomas could possibly represent dedifferentiated liposarcomas, which are known to have better prognosis. In order to confirm the latter diagnosis, any fatty tissue surrounding the tumour should be sampled extensively to identify well-differentiated liposarcomatous areas.

Immunohistochemical staining for MDM2 or FISH analysis on paraffin blocks will aid in the diagnosis.
12.5 Referral for review or specialist opinion

All patients seen within the West Yorkshire & Harrogate Cancer Alliance and Humber Coast and Vale Cancer Alliance with suspicion or diagnosis of sarcoma must be reviewed by the WY&HSS MDT.

The complete histopathology report should be available at the MDT meeting. The reports, slides and blocks should be requested at least five days prior to the meeting and should be available at least three days before the meeting for review.

A formal report should be issued by the reviewing pathologist to the clinician responsible for the patient and also to the original pathologist.

12.6 References


12.7 Pathology appendices

1. SNOMED codes
2. FNCLCC grading
3. Translocations and other genetic abnormalities in sarcomas
APPENDIX A

HISTOLOGICAL TYPES OF SARCOMA AND SNOMED CODING

Adipocytic Tumours
Intermediate (locally aggressive)
  Adipocytic lipomatous tumour/Well-differentiated liposarcoma  8851/3
Malignant
  Dedifferentiated liposarcoma  8858/3
  Myxoid liposarcoma  8852/3
  Round cell liposarcoma  8853/3
  Pleomorphic liposarcoma  8854/3
  Mixed-type liposarcoma  8855/3
  Liposarcoma, not otherwise specified  8850/3

Fibroblastic/Myofibroblastic Tumours
Intermediate (locally aggressive)
  Fibromatosis  8821/1
  Solitary fibrous tumour and
c  Hemangiopericytoma  9150/1
  Inflammatory myofibroblastic tumour  8825/1
  Low-grade myofibroblastic sarcoma  8825/3
  Myxoinflammatory fibroblastic sarcoma  8811/3
  Infantile fibrosarcoma  8814/3
Malignant
  Adult fibrosarcoma  8810/3
  Myxofibrosarcoma  8811/3
  Low-grade fibromyxoid sarcoma/lysinizing
  spindle cell tumour  8811/3
  Sclerosing epithelioid fibrosarcoma  8810/3

So-called Fibrohistiocytic Tumours
Intermediate (rarely metastasizing)
  Plexiform fibrohistiocytic tumour  8835/1
  Giant cell tumour of soft tissue  9251/1

Vascular Tumours
Intermediate (locally aggressive)
  Kaposiform hemangioendothelioma  9130/1
Intermediate (rarely metastasizing)
  Retiform hemangioendothelioma  9135/1
  Papillary intralymphatic angioendothelioma  9135/1
  Composite hemangioendothelioma  9130/1
  Kaposi sarcoma  9140/3
Malignant
  Epithelioid hemangioendothelioma  9133/3
  Angiosarcoma of soft tissue  9120/3

Tumours of Peripheral Nerves
Malignant
  Malignant peripheral nerve sheath tumour  9540/3
  Epithelioid malignant peripheral nerve sheath tumour  9540/3

Chondro-osseous Tumours
Malignant
  Mesenchymal chondrosarcoma  9240/3
  Extraskeletal osteosarcoma  9180/1

Tumours of Uncertain Differentiation
Intermediate (rarely metastasizing)
  Angiomatoid fibrous histiocytoma  8836/1
| Ossifying fibromyxoid tumour (including atypical/malignant) | 8842/0 |
| Mixed tumour/Myoepithelioma/parachordoma | 8582/1 |
| Malignant | |
| Synovial sarcoma | 9040/3 |
| Epithelioid sarcoma | 8804/3 |
| Alveolar soft part sarcoma | 9581/3 |
| Clear cell sarcoma of soft tissue | 9044/3 |
| Extraskeletal myxoid chondrosarcoma | 9231/3 |
| Extraskeletal pPNET/Ewing tumour | |
| peripheral primitive neuroectodermal tumour | 9364/3 |
| Extraskeletal Ewing tumour | 9260/3 |
| Desmoplastic small round cell tumour | 8806/3 |
| Extrarenal rhabdoid tumour | 8863/3 |
| Malignant mesenchymoma | 8990/3 |
| Neoplasms with perivascular epithelioid cell differentiation (PEComa) | none |
| Intimal sarcoma | 8800/3 |
APPENDIX B

FRENCH FEDERATION OF CANCER CENTRES SYSTEM OF GRADING

Tumour differentiation
Score
1 Sarcoma histologically very similar to normal adult mesenchymal tissue
2 Sarcoma for defined histological subtype (e.g. myxofibrosarcoma)
3 Sarcoma of uncertain type, embryonal and undifferentiated sarcomas

Mitosis count
Score
1 0–9/10 HPF
2 10–19/10 HPF
3 >20/10 HPF

Microscopic tumour necrosis
Score
0 No necrosis
1 <50% tumour necrosis
2 >50% tumour necrosis

Histological grade
Grade
1 Total score 2 or 3
2 Total score 4 or 5
3 Total score 6, 7 or 8

Tumour differentiation scores
<table>
<thead>
<tr>
<th>Tumour type</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Well-differentiated liposarcoma</td>
<td>1</td>
</tr>
<tr>
<td>Well-differentiated fibrosarcoma</td>
<td>1</td>
</tr>
<tr>
<td>Well-differentiated MPNST</td>
<td>1</td>
</tr>
<tr>
<td>Well-differentiated chondrosarcoma</td>
<td>1</td>
</tr>
<tr>
<td>Myxoid liposarcoma</td>
<td>2</td>
</tr>
<tr>
<td>Conventional fibrosarcoma</td>
<td>2</td>
</tr>
<tr>
<td>Conventional MPNST</td>
<td>2</td>
</tr>
<tr>
<td>Well-differentiated malignant Haemangiopericytoma</td>
<td>2</td>
</tr>
<tr>
<td>Myxoid MFH (myxofibrosarcoma)</td>
<td>2</td>
</tr>
<tr>
<td>Typical stromal/pleomorphic MFH</td>
<td>2</td>
</tr>
<tr>
<td>Conventional leiomyosarcoma</td>
<td>2</td>
</tr>
<tr>
<td>Extraskeletal myxoid chondrosarcoma</td>
<td>2</td>
</tr>
<tr>
<td>Conventional angiosarcoma*</td>
<td>2</td>
</tr>
<tr>
<td>Round cell liposarcoma</td>
<td>3</td>
</tr>
<tr>
<td>Pleomorphic liposarcoma</td>
<td>3</td>
</tr>
<tr>
<td>Dedifferentiated liposarcoma</td>
<td>3</td>
</tr>
<tr>
<td>Poorly differentiated fibrosarcoma</td>
<td>3</td>
</tr>
<tr>
<td>Epithelioid malignant schwannoma</td>
<td>3</td>
</tr>
<tr>
<td>Poorly differentiated MPNST*</td>
<td>3</td>
</tr>
<tr>
<td>Malignant Triton tumour*</td>
<td>3</td>
</tr>
<tr>
<td>Conventional malignant haemangiopericytoma</td>
<td>3</td>
</tr>
<tr>
<td>Giant cell and inflammatory MFH</td>
<td>3</td>
</tr>
<tr>
<td>Poorly differentiated/epithelioid/pleomorphic leiomyosarcoma</td>
<td>3</td>
</tr>
<tr>
<td>Synovial sarcoma</td>
<td>3</td>
</tr>
<tr>
<td>Rhabdomyosarcoma*</td>
<td>3</td>
</tr>
<tr>
<td>Mesenchymal chondrosarcoma</td>
<td>3</td>
</tr>
</tbody>
</table>

Valid on the date of publication
Version 4.0
<table>
<thead>
<tr>
<th>Tumour Type</th>
<th>Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epithelioid angiosarcoma</td>
<td>3</td>
</tr>
<tr>
<td>Extraskeletal osteosarcoma*</td>
<td>3</td>
</tr>
<tr>
<td>Extraskeletal Ewing's sarcoma/PNET *</td>
<td>3</td>
</tr>
<tr>
<td>Alveolar soft part sarcoma*</td>
<td>3</td>
</tr>
<tr>
<td>Malignant rhabdoid tumour</td>
<td>3</td>
</tr>
<tr>
<td>Clear cell sarcoma*</td>
<td>3</td>
</tr>
<tr>
<td>Undifferentiated sarcoma</td>
<td>3</td>
</tr>
</tbody>
</table>

*Grading of malignant peripheral nerve sheath tumour, embryonal and alveolar rhabdomyosarcoma, angiosarcoma, extraskeletal myxoid chondrosarcoma, alveolar soft part sarcoma, clear cell sarcoma, and epithelioid sarcoma is not recommended. In practice, the following tumours are graded by definition as below:

1. Atypical lipomatous tumour/ well-differentiated liposarcoma, dermatofibrosarcoma protuberans, infantile fibrosarcoma and angiomatoid 'MFH' are Grade 1.

2. Ewing's sarcoma/PNET, rhabdomyosarcoma (except spindle cell and botryoid variants), angiosarcoma, pleomorphic liposarcoma, soft tissue osteosarcoma, mesenchymal chondrosarcoma, desmoplastic small round cell tumour, and extra-renal malignant rhabdoid tumour are Grade 3.

3. Alveolar soft part sarcoma, clear cell sarcoma, epithelioid sarcoma, and low-grade fibromyxoid sarcoma are not graded but are usually considered as high grade for management purposes.
### APPENDIX C.

**TRANSLOCATIONS AND OTHER GENETIC ABNORMALITIES IN SARCOMAS**

<table>
<thead>
<tr>
<th>Histologic Type</th>
<th>Translocation or rearrangement</th>
<th>Fusion gene or other feature</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alveolar soft part sarcoma</td>
<td>t(X;17)(p11;q25)</td>
<td>ASPL-TEF3</td>
</tr>
<tr>
<td>Angiomatoid fibrous histiocytoma</td>
<td>t(12;22)(q13;q12)</td>
<td>EWSR1-ATF1</td>
</tr>
<tr>
<td></td>
<td>t(12;16)(p13;q11)</td>
<td>FUS-ATF1</td>
</tr>
<tr>
<td></td>
<td>t(2;22)(q33;q12)</td>
<td>EWSR1-CREB1</td>
</tr>
<tr>
<td>Clear cell sarcoma (GIST)</td>
<td>t(2;22)(q33;q12)</td>
<td>EWSR1-CREB1</td>
</tr>
<tr>
<td>Dermatofibrosarcoma protuberans</td>
<td>t(17;22)(p21;q13)</td>
<td>COL1A1-PDGFB</td>
</tr>
<tr>
<td>Desmoplastic SRCT</td>
<td>t(11;22)(p13;q12)</td>
<td>EWSR1-WT1</td>
</tr>
<tr>
<td>Epithelioid sarcoma</td>
<td>Abnormalities of 22q</td>
<td>INI1 inactivation</td>
</tr>
<tr>
<td>Ewing sarcoma/PNET</td>
<td>t(11;22)(q24;q12)</td>
<td>EWSR1-FLI1</td>
</tr>
<tr>
<td></td>
<td>t(21;22)(q12;q12)</td>
<td>EWSR1-ERG</td>
</tr>
<tr>
<td></td>
<td>t(2;22)(q33;q12)</td>
<td>EWSR1-FEV</td>
</tr>
<tr>
<td></td>
<td>t(7;22)(p22;q12)</td>
<td>EWSR1-ETV1</td>
</tr>
<tr>
<td></td>
<td>t(17;22)(q12;q12)</td>
<td>EWSR1-E1AF</td>
</tr>
<tr>
<td></td>
<td>inv(22)(q12;q12)</td>
<td>EWSR1-ZSG</td>
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<tr>
<td>Extraskeletal myxoid chondrosarcoma</td>
<td>t(9;22)(q22;q12)</td>
<td>EWSR1-NRA3</td>
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<tr>
<td></td>
<td>t(9;17)(q22;q11)</td>
<td>TAF1168-NRA3</td>
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<tr>
<td>Fibrosarcoma, infantile</td>
<td>t(12;16)(p13;q26)</td>
<td>TCF12-NRA3</td>
</tr>
<tr>
<td>Inflammatory myofibroblastic</td>
<td>2p23 rearrangement</td>
<td>ALK fusions with various genes</td>
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<tr>
<td>tumour</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Leiomyosarcoma</td>
<td>Deletion of 1p</td>
<td></td>
</tr>
<tr>
<td>Liposarcoma:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Well-differentiated</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Myxoid/round cell</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pleomorphic</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Complex</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Low-grade fibromyxoid sarcoma</td>
<td>t(7;16)(q33;p11)</td>
<td>FUS-CREB3L2</td>
</tr>
<tr>
<td></td>
<td></td>
<td>FUS-CREB3L1 (rare)</td>
</tr>
<tr>
<td>Malignant rhabdoid tumour</td>
<td>Deletion of 22q</td>
<td>INI1 inactivation</td>
</tr>
<tr>
<td>MPNST</td>
<td></td>
<td></td>
</tr>
<tr>
<td>MYF11</td>
<td></td>
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<tr>
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<td>MYF11</td>
<td>Ring form of chromosome 12</td>
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<td>Rhabdomyosarcoma:</td>
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<tr>
<td>Embryonal</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alveolar</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Trisomes 2q, 8, and 20</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>t(1;13)(p36;q14)</td>
<td>LOH at 11p15</td>
</tr>
<tr>
<td></td>
<td>t(2;13)(q35;q14)</td>
<td>PAX7-FKHR</td>
</tr>
<tr>
<td></td>
<td></td>
<td>PAX3-FKHR</td>
</tr>
<tr>
<td>Synovial sarcoma</td>
<td>t(X;18)(p11;q11)</td>
<td>SS18-SSX1</td>
</tr>
<tr>
<td></td>
<td>t(X;20)(p11;q13)</td>
<td>SS18-SSX2</td>
</tr>
<tr>
<td></td>
<td></td>
<td>SS18-SSX4 (rare)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>SS1BL1-SSX1</td>
</tr>
</tbody>
</table>

GIT indicates gastrointestinal tract; SRCT, small round cell tumour; PNET, primitive neuroectodermal tumour; MFH, malignant fibrous histiocytoma; MPNST, malignant peripheral nerve sheath tumour; LOH, loss of heterozygosity
13 Staging

Preoperative staging should be completed for all tumours using the current version of the UICC TNM staging system. This may need to be updated based upon operative findings.

13.1 Tumour (T)

- There are 3 variables for local tumour extent.
- Tumour size
- Tumour relationship to deep fascia
- Tumour grade

13.2 Nodes (N)

Nodal disease is uncommon in adult type STS but should be carefully looked for in paediatric-type STS and selected adult-type STS histological subgroups.

13.3 Metastases (M)

Metastatic disease should be actively sought in all but low-grade primary tumours. Cross-sectional CT imaging of the thorax should be performed as a minimum, with additional series (abdomen/pelvis/neck) performed based upon the site of the primary. Supplementary imaging using alternate modalities may be required. The routine role of PET is unclear.

The sites of metastatic disease should be recorded.
## Stage Grouping

<table>
<thead>
<tr>
<th>Stage</th>
<th>G</th>
<th>T</th>
<th>N</th>
<th>M</th>
</tr>
</thead>
<tbody>
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<td>IA</td>
<td>1, 2</td>
<td>1a</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>IB</td>
<td>1, 2</td>
<td>1b</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
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</tr>
<tr>
<td>IIB</td>
<td>3, 4</td>
<td>2b</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>IIC</td>
<td>3, 4</td>
<td>1a</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>IIB</td>
<td>3, 4</td>
<td>1b</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>III</td>
<td>3, 4</td>
<td>2a</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
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<td>Any T</td>
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</tr>
<tr>
<td></td>
<td>Any G</td>
<td>Any T</td>
<td>Any N</td>
<td>M1</td>
</tr>
</tbody>
</table>

## Summary

### Soft Tissue Sarcoma

<table>
<thead>
<tr>
<th>T1</th>
<th>≤5 cm</th>
</tr>
</thead>
<tbody>
<tr>
<td>T1a</td>
<td>Superficial</td>
</tr>
<tr>
<td>T1b</td>
<td>Deep</td>
</tr>
<tr>
<td>T2</td>
<td>&gt;5 cm</td>
</tr>
<tr>
<td>T2a</td>
<td>Superficial</td>
</tr>
<tr>
<td>T2b</td>
<td>Deep</td>
</tr>
<tr>
<td>N1</td>
<td>Regional</td>
</tr>
<tr>
<td>G1</td>
<td>Well differentiated</td>
</tr>
<tr>
<td>G2</td>
<td>Moderately differentiated</td>
</tr>
<tr>
<td>G3</td>
<td>Poorly differentiated</td>
</tr>
<tr>
<td>G4</td>
<td>Undifferentiated</td>
</tr>
</tbody>
</table>
A well validated prognostic scoring tool exists for determining the risk of sarcoma-specific death at 10-12 years post resection for non-metastatic soft tissue sarcoma. This tool may be of value in aiding MDT decision making in respect of balancing the functional consequences of local therapy and in guiding intensity of follow-up. It may also aid patient discussions, but its limitations should be made clear. It is not intended that survival prognosis be forced upon patients who do not wish to receive it. For higher risk patients it may serve as a starting point for discussion of adjuvant systemic therapy, though this is not an approach used routinely at present.

Several versions of the tool exist. The original Memorial Sloan Kettering Cancer Centre (MSKCC) tool has been validated for all tumours sites and uses the grading system proposed by Hadju and used by the NCI. Additional tools have been developed to look at limb sarcomas using the FNCLCC grading system used by WY&HSS and separately for adipocytic tumours which behave markedly differently when either low versus high grade and which represent a greater proportion of retroperitoneal sarcomas. A tool for post-local relapse survival estimation is also available.

In time ready-reckoners will be developed to aid more rapid use within the MDT meetings.

Four tools are shown:

1. Kattan 2002: all sites, all histologies, NCI grading
2. Dalal 2006: all sites, adipocytic only, NCI grading
3. Mariani 2004: limb only, all histologies, FNCLCC grading
4. Kattan 2003: all sites, post-relapse, NCI grading

An on-line tool is available: http://www.mskcc.org/nomograms/sarcoma
### Postoperative Nomogram for 12-Year Sarcoma-Specific Death

**Points**

<table>
<thead>
<tr>
<th>Points</th>
<th>0</th>
<th>10</th>
<th>20</th>
<th>30</th>
<th>40</th>
<th>50</th>
<th>60</th>
<th>70</th>
<th>80</th>
<th>90</th>
<th>100</th>
</tr>
</thead>
<tbody>
<tr>
<td>Size (cm)</td>
<td>&lt;=5</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Deep</td>
<td></td>
<td></td>
<td></td>
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<td></td>
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<td></td>
<td></td>
<td></td>
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<tr>
<td>Depth</td>
<td>Superficial</td>
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<td></td>
</tr>
<tr>
<td>Site</td>
<td>Lower Extremity</td>
<td>Thoracic/Trunk</td>
<td>Head/Neck</td>
<td>Upper Extremity</td>
<td>Visceral</td>
<td>Retro/Intra-abdominal</td>
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<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Histology</td>
<td>Fibro</td>
<td>Lipo</td>
<td>Leiomyo</td>
<td>Synovial</td>
<td>MPH</td>
<td>Other</td>
<td>MUPNT</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age (years)</td>
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<td>20</td>
<td>30</td>
<td>40</td>
<td>50</td>
<td>60</td>
<td>70</td>
<td>80</td>
<td>90</td>
<td></td>
<td></td>
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<tr>
<td>Total Points</td>
<td>0</td>
<td>20</td>
<td>40</td>
<td>60</td>
<td>80</td>
<td>100</td>
<td>120</td>
<td>140</td>
<td>160</td>
<td>180</td>
<td>200</td>
</tr>
<tr>
<td>12yr Low Gr. SSD</td>
<td>0.04</td>
<td>0.06</td>
<td>0.08</td>
<td>0.1</td>
<td>0.15</td>
<td>0.2</td>
<td>0.3</td>
<td>0.4</td>
<td>0.5</td>
<td>0.6</td>
<td>0.7</td>
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<tr>
<td>12yr High Gr. SSD</td>
<td>0.04</td>
<td>0.06</td>
<td>0.08</td>
<td>0.1</td>
<td>0.15</td>
<td>0.2</td>
<td>0.3</td>
<td>0.4</td>
<td>0.5</td>
<td>0.6</td>
<td>0.7</td>
</tr>
</tbody>
</table>

**Instructions for Physician**: Locate the patient's tumor size on the Size axis. Draw a line straight upwards to the Points axis to determine how many points towards sarcoma-specific death the patient receives for his tumor size. Repeat this process for the other axes, each time drawing straight upward to the Points axis. Sum the points achieved for each predictor and locate this sum on the Total Points axis. Draw a line straight down to either the Low Grade or High Grade axis to find the patient's probability of dying from sarcoma within 12 years assuming he or she does not die of another cause first.

**Instruction to Patient**: "If we had 100 patients exactly like you, we would expect between <predicted percentage from nomogram - 6%> and <predicted percentage + 6%> to die of sarcoma within 12 years if they did not die of another cause first, and death from sarcoma after 12 years is still possible."

**Figure 1.** Postoperative nomogram for 12-Year sarcoma-specific death based on 2,163 patients treated at MSKCC. Abbreviations: Fibro=Fibrosarcoma, Lipo=Liposarcoma, Leiomyo=Leiomyosarcoma, MPH=Malignant Fibrous Histiocytoma, MUPNT=Malignant Peripheral-Nerve Tumor, Gr=Grade, SSD=Sarcoma-Specific Death.

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Validity: Valid on the date of publication

Version: 4.0
14.1 Dalal 2006: All sites, adipocytic sarcoma, NCI grading

Mariani 2006: Limb only, all histologies, FNCLCC grading

Instructions: The nomogram allows obtaining the 10-year probability of death for sarcoma corresponding to a patient's combination of covariates, assuming the patient does not die of another cause first. Locate the patient's tumor size and draw a line straight upwards to the Points axis to determine the score associated to that size. Repeat the process for depth, site, histology, age and grade. Sum the scores achieved for each covariate, and locate this sum on the Total Points axis. Draw a line straight down to the sarcoma-specific death axis to find the probability.
14.2 Kattan 2003: All sites, post-local relapse, NCI grading

Instructions for Physician: Locate the patient's age on the Age axis. Draw a line straight upwards to the Points axis to determine how many points towards sarcoma-specific death the patient receives for his age. Repeat this process for the other axes, each time drawing straight upward to the Points axis. Sum the points achieved for each predictor and locate this sum on the Total Points axis. Draw a line straight down to find the patient's probability of dying from sarcoma each year within 5 years.

Instruction to Patient: "If we had 100 patients exactly like you, we would expect [predicted percentage from nomogram] to die of sarcoma within X years."
15 Surgery

15.1 Surgical intent

Surgical intent should be clear preoperatively following MDT work-up and discussion. The extent of surgery should reflect the anticipated future behaviour of the tumour (local and systemic), the place of planned adjuvant therapy, the functional (and cosmetic) impact and surgery-specific risks.

15.1.1 Incisional biopsy

Resection of lesional tissue for diagnosis. This is a procedure of last resort (multiple non-diagnostic core-biopsies) in the context of suspected STS. An excisional biopsy may be preferable as residual disease is inevitable. Biopsy should be planned to facilitate en-bloc resection of biopsy site when definitive surgery performed.

15.1.2 Excisional biopsy

Resection of tumour with narrow margin of normal tissue when either biopsy or imaging is strongly supportive of benign histology. Functional compromise should be minimised.

15.1.3 Wide local excision

Resection of tumour surrounded by a non-involved cuff of normal tissue (notionally 2cm; see surgical principles described below). Performed when malignant tumour is proven or suspected on histology or imaging. Functional compromise possible.

15.1.4 Planned marginal excision

As WLE but when one or more margin is at higher risk of histological positivity as a wider margin would have a more major impact upon function.

15.1.5 Planned positive excision

As planned marginal excision but when tumour is consciously left as resection would have a more major impact upon function. Such margins should be clearly marked and described to facilitate post-operative radiotherapy. Planned positive margin resection is not the same as surgical debulking which should usually be avoided (see palliative surgery).

15.1.6 Re-excision

Should aim to convert an unplanned positive excision into a marginal (or ideally a wide) excision by re-excising the tumour bed. Functional compromise is likely.

15.1.7 Radical excision/amputation

Radical removal of an entire compartment, limb or part of limb. May be required if tumour crosses several compartments or surgery (and/or tumour) critically disrupts distal neurological or vascular supply in order to attain clear margins. Functional compromise certain.

15.1.8 Palliative surgery

Non-curative debulking surgery is not indicated unless there is an over-whelming clinical need such as severe pain, fungation, vascular compromise (haemorrhage or ischaemia) or luminal obstruction. Limb tumours operated upon in this context are usually best managed with an amputation even when metastatic disease is present.
15.2 Surgical principles

Specific surgical techniques remain at the discretion of the operator. Sarcomas are un-encapsulated and should be excised en-bloc with an intact cuff of non-compromised muscle, fascial plane, periosteum/bone or organ/viscera. Tumour spillage should be avoided and, if occurs, clearly recorded in the operative notes. The planning of adjuvant radiotherapy is aided if clips are placed at the cranial and caudal extent of the tumour bed and where the surgeon feels that the tumour resection margin is likely to be close and/or involved. The position of clips and any suspected residual (R1 or R2) tumour should be clearly recorded in the operative note. It is also helpful if resected/sacrificed muscles, vessels and/or organs are clearly described. If periosteal stripping is performed this should be recorded.

15.3 Wound closure

Wound closure should be planned, ideally preoperatively, with due regard to received, planned or potential adjuvant radiotherapy. Primary direct closure should be avoided following pre-operative radiotherapy. When post-operative radiotherapy is likely, choice of closure technique should facilitate radiotherapy planning within 4 weeks of surgery and commencement of radiotherapy within 6 weeks of surgery. Split-skin grafting should be avoided when radiotherapy is anticipated.
16 Radiotherapy

16.1 Role of radiotherapy

Radiotherapy (RT) is used as a means of improving local control and facilitating function preserving surgery for resectable STS. For non-resectable or metastatic disease it may be useful in improving localised symptoms and delaying progression. RT volumes and doses should be disease and site appropriate according to agreed local protocols.

16.2 Radiotherapy for resectable disease

Pre- or post-operative radiotherapy should be used for appropriate cases following discussion at the sarcoma MDT meeting. Indications for radiotherapy follow European Consensus Guidelines. These guidelines do not apply for retroperitoneal STS or sarcomas arising from viscera when decisions must be made case-by-case.

The MDT should discuss and agree local therapy (both primary and adjuvant therapy) prior to surgery to optimise outcomes. Consideration should be given to the functional deficits and complications expected from all modalities used for local control. Pre-operative radiotherapy will not render unresectable disease resectable. If pre-operative RT is to be used there is an increased risk of surgical complications and consideration should be given to the method of wound closure, with vascularised flaps favoured. If planned positive or very marginal resection is anticipated in order to preserve function, post-operative RT may be favoured. Placement of surgical clips and detailed operative notes are essential in planning effective post-operative RT.

Patients should be seen by the Clinical Oncologist within 2 weeks of surgery to facilitate timely RT planning (with simulation after operative swelling has settled) in order to start post-operative RT within 12 weeks to be consistent with VORTEX of surgery.

When bone that has been stripped of periosteum is irradiated the fracture risk is increased. Consideration should be given to prophylactic pinning of long bones long-bones 8-12 weeks post-RT.

<table>
<thead>
<tr>
<th>European consensus on adjuvant RT for resected STS</th>
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<tr>
<td>Characteristic</td>
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<td>Locally recurrent</td>
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16.3 Radiotherapy for locally recurrent disease

Radiotherapy should be considered in all cases of locally recurrent disease. Previously irradiated tissues are poorly tolerant of re-irradiation using conventional external beam RT and risk of complication may outweigh clinical benefit. Techniques such as HDR-afterloaded brachytherapy, I-131 mesh implants, image-guided and intensity modulated RT and dose-planned electron solutions are in development but will likely require tailoring to individual circumstances rather than general application to class-solutions.

16.4 Radiotherapy for unresectable or metastatic disease

Volume and dose selection should be disease and site appropriate and be made with due consideration of the patient’s general performance status (PS) and anticipated natural history and overall survival. Unless the patient’s PS is poor or the patient is clearly at or nearing the very late stages of their illness it is suggested that a high-dose palliative approach is adopted. STS is radio-sensitive but not particularly radio-responsive and tumour shrinkage may take months (though clinical benefit in terms of symptom relief may come within weeks).
17 Chemotherapy

Context

a) Adult type soft tissue sarcoma (specific subtypes)

Some of the systemic therapy options discussed below are not specifically licenced for use in soft tissue sarcoma. NICE approved, or funded via the Cancer Drugs Fund.

Histological subtypes of soft tissue sarcoma vary in their sensitivity to traditional cytotoxic chemotherapy agents, and whether (or not) they are sensitive to new or unusual chemotherapy agents which do not have activity in other sarcomas.

This should be brought together with:

- the natural history of the disease in the individual,
- the tumour burden,
- the risk of progression at the local site to morbidity and mortality,
- the stage of the tumour and
- the patient's individual characteristics in terms of symptoms, co-morbidities and prior cancer treatments

in order to come to a treatment goal, agree that with the patient and decide on management. In this section I will discuss specific subtypes of adult soft tissue sarcoma requiring unusual management.¹

The following types of soft tissue sarcoma are highly refractory to cytotoxic chemotherapy, meaning more than 80% will have worsened at three months despite using all identifiable systemic treatment regimens and therefore active supportive care may be preferable to chemotherapy administration in these types:

- clear cell sarcoma
- extra-skeletal myxoid chondrosarcoma
- well differentiated liposarcoma.

The following subtypes of adult soft tissue sarcoma have a unique or unusual sensitivity to specific therapies and should be managed distinctly from other adult soft tissue sarcomas:

- gastrointestinal stromal tumour (see elsewhere),
- angiosarcoma,
- myxoid round cell liposarcoma with confirmed translocation,
- synovial sarcoma,
- endometrial stromal sarcoma,
- dermatofibrosarcoma protubersans/tenosynovial giant cell tumour,
- solitary fibrous tumour/hemangiopericytoma,
- alveolar soft part sarcoma,

• soft tissue Ewing sarcoma (see elsewhere),
• alveolar rhabdomyosarcoma and embryonal rhabdomyosarcoma (see elsewhere), inflammatory myofibroblastic tumour, and
• desmoplastic small round cell tumour

**Angiosarcoma (see also Vascular Sarcomas)**

In metastatic or locally advanced (i.e. no benefit from resection) cutaneous angiosarcoma, the agents paclitaxel and liposomal doxorubicin have unusual activity. Paclitaxel should be considered in the first line and second-line therapy alongside other agents active in soft tissue sarcoma, but those agents are also active. Pegylated liposomal doxorubicin is also active in the same angiosarcoma subtypes.

**Myxoid round cell liposarcoma (with a FUS:CHOP translocation in particular)**

This subtype of sarcoma is particularly sensitive to the agent Trabectadin. It has an unusual pattern of metastasis to visceral and non-visceral sites, requiring specific staging. Some believe it has a more indolent natural history because of repeated chemo sensitivity and re-challenge. Trabectadin should be considered alongside other standard treatments for soft tissue sarcoma but perhaps given particular priority. Trabectadin is also approved as second-line or further treatment for all sarcoma subtypes (NICE TA 185).

**Synovial sarcoma**

Synovial sarcoma is particularly sensitive to ifosfamide. It should be considered to add ifosfamide into neo-adjuvant, any adjuvant, and advanced disease regimens in synovial sarcoma in combination with doxorubicin, or as second-line treatment as a single-agent, alongside other standard soft tissue sarcoma treatments.

**Endometrial stromal sarcoma (hormone receptor positive and therefore well differentiated)**

These tumours are sensitive to aromatase inhibition. This should be considered alongside other chemotherapy-based treatments in first or later lines of management on the basis of hormone receptor positivity.

**Dermatofibrosarcoma protuberans**

This is a chemotherapy resistant tumour but with documented sensitivity to Imatinib which should be considered rather than cytotoxic chemotherapy in local advanced metastatic disease.

**Tenosynovial giant cell tumour**

This is a chemotherapy resistant subtype, (also known as pigmented villonodular synovitis) and documented to be sensitive to Imatinib.

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Solitary fibrous tumour (also known hemangiopericytoma in the past)
This subtype can be sensitive to combination chemotherapy as for other adult soft tissue sarcomas but is also sensitive to anti-angiogenic approaches. Bevacizumab in combination with Temozolomide should be considered in first or second-line therapy or later.\(^9\) There is evidence of the use of Sunitinib in this type also\(^10\) and some centres also use Pazopanib on an individual patient application basis. There are series reporting that single agent dacarbazine may have a role in the management of SFTs (Stacchiotti et al Clin Can Res (2013) 19(18)1-10).

Alveolar soft part sarcoma
This is a chemotherapy resistant subtype, with documented sensitivity to anti-angiogenic approaches including Sunitinib and cediranib.\(^11\)

This is not a cancer drugs found or NHS-approved indication and therefore will require individual patient funding application.

Inflammatory myofibroblastic tumour (with an ALK rearrangement)
There is evidence of activity of crizotinib in this as well as other ALK rearranged cancers.\(^12\)

Leiomyosarcoma
In the management of leiomyosarcomas there has been some discussion of specific approaches. These are relatively chemo sensitive soft tissue sarcomas but in addition single-agent gemcitabine can be used in patients of poor performance status, the combination gemcitabine and docetaxel can be used in first-line therapy in selected cases and cases with strong positivity for oestrogen and progesterone receptors have been documented to respond to aromatase inhibition when the leiomyosarcoma is of a uterine primary. Leiomyosarcomas are also relatively sensitive to Trabectedin, and may be relatively resistant to ifosfamide.\(^13\)

Desmoplastic small round cell tumour
This subtype can be managed according to the protocols for Ewings sarcoma.\(^14\) There is some efficacy for cisplatin and etoposide regimens in relapsed disease where previous chemo-sensitivity has been notable and durable whereas sporadic cases of sensitivity to Trabectedin also reported.\(^15\)

\(^8\) Cassiea et al cancer 2012; 118:1649 - 55.

\(^9\) Park et al Cancer (2011); 117:4939 - 47.


\(^11\) Kumar et al J clin oncol 2013; 31:2296-302
\(^12\) Butrynskin N. Engl J med 2010;363:1727 - 33


\(^15\) Frezza Clin Sarcoma Res (2014); 4:3.
b) **Bone sarcoma**

Bone sarcomas are a very heterogeneous group of tumours ranging from highly metastatic and chemo sensitive entities such as Ewing sarcoma through to entirely chemotherapy resistant and localised tumours such as low-grade chondrosarcomas.

_The Leeds Cancer Centre is not a reference centre for bone tumours and does not host a bone tumour MDT. All such decisions are carried out in collaboration with the specialist bone sarcoma multidisciplinary team based at the Royal Orthopaedic Hospital in Birmingham._

For the management of the Ewings sarcoma family of tumours the approach is embodied within the Euro-Ewings 99 and the Euro Ewings 2012 protocols.

For the osteosarcoma spectrum of illnesses the approach is embodied within the principles of the EURAMOS 1 protocol. For high-grade osteosarcomas within the license parameters, mifamurtide should be considered in this group.

In both Ewing sarcoma and high-grade osteosarcomas there is clear evidence that dose density and dose intensity correlate with outcome. Reductions in dose density and dose intensity have been shown to reduce the efficacy of treatment. Treatment at maximum doses will cause substantial toxicity acutely and in the long-term. However to optimise outcomes this form of dose density and dose intensity needs to be maintained therefore this requires very extensive and experienced supportive care.

There are very specific second and third line options in high-grade bone sarcomas of either the osteosarcoma or Ewing’s family of tumour type. In the Ewing’s family of tumours there are indications for high-dose chemotherapy with stem cell rescue in relapsed but chemo-sensitive disease. There are other subtypes of bone sarcoma which require very specific approaches.

Pigmented villonodular synovitis can be sensitive to Imatinib rather than chemotherapy. (see above)

Giant cell tumour of bone can be sensitive to Denosumab rather than chemotherapy.16

Chordoma can be sensitive to Imatinib, to doxorubicin or doxorubicin-cisplatin combinations and to antiangiogenic throughout therapies including Sunitinib or Sirolimus.17

c) **Borderline and non-malignant mesenchymal disease in the sarcoma spectrum**

17 Stacchiotti et al J clin oncol (2012); 30:914-20; Stacchiotti et al annals of oncology 2009; 20:1886-94
Desmoid and aggressive deep fibromatosis
This is a locally advanced non-metastatic disease, presenting at many sites, which should be managed according to its resectability, natural history, patient characteristics and patient wishes.

This should be done by an experienced multidisciplinary sarcoma team. After unplanned resection there is a very high incidence of local recurrence. However after planned resection very bulky tumours can be removed with very low risk of local recurrence in specific circumstances, notably in the abdominal wall after pregnancy.

Un-resectable disease or disease that is morbid upon resection, or where patients do not wish to undergo resection can be managed with hormonal approaches including Tamoxifen and a non-steroidal (sulindac or naproxen preferred)\(^\text{18}\)

In young people with fibromatosis the combination of methotrexate and vinblastine has efficacy\(^\text{19}\)

Pegulated liposomal doxorubicin can be affective particularly in the early relief of pain.\(^\text{20}\)

These systemic interventions can be used as neoadjuvant treatments, palliative treatments, and the place of radiotherapy should also be carefully considered.

PEComa. locally advanced or metastatic disease may respond to combination chemotherapy as with other soft tissue sarcomas, but may also respond to sirolimus.\(^\text{21}\) There is early data that responds to mTOR inhibition may be molecularly predicted but at this time not sufficient data to be selective.\(^\text{22}\)

Vascular Sarcomas
Vascular sarcomas span a broad spectrum of behaviours and require differing management. It should be noted that leiomyosarcoma arising from vessel (usually venous) walls should be managed as for leiomyosarcoma.

Epithelioid Haemagioendothelioma
Usually a low grade neoplasm with hepatic or pulmonary origin. When localised, surgery can be effective. Even when disseminated, surgery can be effective palliation and for isolated areas of symptomatic progression. Progression is often slow and interval observation, in the first instance, is recommended. Rapid progression can suggest transformation into a high-grade form and should be managed as angiosarcoma. Optimal therapy is undefined beyond doxorubicin as gold-standard first-line therapy. Metronomic cyclophosphamide is minimally

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\(^{18}\) Hannsman et al cancer 2004; 100:612 - 20
\(^{19}\) Azzarelli et al cancer 2001; 92:1259 - 64.
\(^{22}\) Dixon et al Int J cancer (2013); 132:1711 to 7.
toxic and yields a median PFS of ~9 months, whereas other approaches using RTKIs such as sorafenib and sunitinib are more toxic, often require dose reduction and have median response duration of 4 months.

**Cyclophosphamide 50mg po od and Vinblastine 3mgm⁻² iv weekly until PD**
Clin Cancer Res 2006;12:3092-3098

**Sorafenib 400mg po bd until disease progression**
See JCO doi/10/1200/JCO.2008.20.4495 (May 18th 2009)

**Sunitinib 37.5mg po od until PD**
JCO doi/10/1200/JCO.2008.20.9890 (May 18th 2009)

**Kaposi sarcoma**

Although closely associated with HIV infection, HIV-associated KS is falling in incidence since the advent of highly active anti-retroviral therapy (HAART). Risk stratification, using the protocol set out in the British HIV Association (BHIVA) Guidelines, is effective in identifying either those cases that may benefit from chemotherapy in addition to HAART. This benefit is through inducing more rapid remission than through HAART where disease is very symptomatic or in suppressing the effects of immune response inflammatory syndrome (IRIS) at tumour level. There is no data to suggest that chemotherapy effects a survival gain over HAART alone. HAART should be supervised by a recognised HIV treatment centre.

First line therapy is with liposomal doxorubicin, usually delivered to a maximum of 6 cycles. Second-line paclitaxel q2w, again for 6 cycles, can be repeated if required. Selumetinib has been trialled as a potential further intervention, though results are pending.

**Liposomal Doxorubicin 20mg/m² q28**

**Paclitaxel 100mg/m² q14**


Caution should be exercised in HIV patients with lymphadenopathy (+/- KS) that they do not have concurrent squamous cell carcinoma, lymphoma or multi-centric Castleman’s disease. The latter (through its association with HHV8 infection) is often associated with KS. Enlarged nodes should undergo core biopsy (not FNAC) and management should be through the appropriate site-specialist cancer or lymphoma MDT. All care should be delivered in close association with the patients’ supervising infectious diseases (ID) team.

The classical and endemic forms of KS are usually cutaneous only and managed with local palliative interventions (surgery, intra-lesional injection or radiotherapy).

**Angiosarcoma**

Angiosarcoma is usually an aggressive neoplasm prone to early local relapse (unless very widely excised) and metastatic spread. Despite this, there is no proven role for adjuvant systemic therapy. Un-resectable locally advanced disease can, on a case-by-case basis, be considered for primary systemic therapy followed by radiotherapy or surgery as salvage for local control.

Palliative chemotherapy is with either single agent doxorubicin or paclitaxel. Although doxorubicin is gold-standard, paclitaxel is preferred for primary sites above the clavicle or where prior anthracycline use (usually post breast-cancer) constrains the use of further
anthracycline. For frailer patients, a weekly regimen may be better tolerated but the evidence base for efficacy is lacking.

**Doxorubicin 75mg/m² q21**

**Doxorubicin 25mg/m² weekly**

**Paclitaxel 70mg/m² d1,d8,d15 q28**  
See JCO 26, no. 32 (November 2008) 5269-5274.

**Metronomic Methotrexate and Vinblastine weekly plus Propranolol bd**

**a) Intervention**

**Doxorubicin (conventional)**

**Indication**  
Doxorubicin is indicated in all moderately or chemosensitive type soft tissue sarcomas and the majority of paediatric type soft tissue sarcomas. Its efficacy varies enormously from case to case. Prior to its administration it should be documented that:-

1. The patient is not in clinical cardiac failure

2. The patient does not have severe jaundice. This would be defined as a bilirubin above 85 mmols/l.

There are relative cautions in patients with poor cardiac function, poor bone marrow reserve, increased bleeding tendency or active infection.

For patients being treated with doxorubicin within an intent of achieving cure or long-term disease control, a baseline echocardiogram is required ahead of treatment.

Patients of WHO performance status above two should be considered for weekly rather than three-weekly schedules.

Patients with bilirubin above the upper limit of normal but below the contraindication level (85mmols/l) should be considered for weekly fractionation of dose also.

Commencing treatment patients should have neutrophils greater than 1, platelets greater than 100.

Doses should be modified according to existing trial protocols where available, but also considered for grade 3 neutropenia complicated by sepsis, grade 3 thrombocytopenia complicated by bleeding, and any grade 4 non-haematological toxicity due to doxorubicin. The first dose reduction is usually 20%.

Mucositis should be expected and proactively managed with mouthwashes. Diarrhoea is rare but will respond to loperamide.
Response assessment should be guided by the individual case but take place after either two or three cycles depending upon intent and toxicity.

On completion of curative chemotherapy including doxorubicin a repeat echocardiogram should be performed. In high-risk cases echocardiogram should be performed after every two cycles of doxorubicin. In patients with normal echocardiograms on completion of doxorubicin with curative intent, echocardiogram should be repeated at five years from completion of treatment also. This should be done earlier if clinical or earlier cardiac assessments indicate greater concern. The predominant late effect of doxorubicin is cardiomyopathy.

Doxorubicin may contribute to impaired fertility. Appropriate patients should be referred to a specialist late effect service for assessment and ongoing monitoring once risk of recurrence is felt to have plateaued.

Men in the fertile age must choose effective contraception during this treatment and for five months afterwards.

There is current patient information on conventional doxorubicin on ppm.

**Liposome pegylated doxorubicin (PLD)**
This is approved by the cancer drugs fund in the following indications:-
- Sarcomas of soft tissue or bone in patients with cardiac impairment requiring Santhracycline in first or second-line indication
- .

It is note these are not necessarily licensed indications and therefore Trust Governance Frameworks are required.

PLD has
- similar or higher efficacy to single-agent doxorubicin in blood vessel tumours,
- good efficacy to control pain in the vast majority of patients with aggressive fibromatosis although responses are fewer.

This agent is also indicated in the treatment of AIDS-related Kaposi sarcoma in combination with specialist infectious diseases opinion.

The dose varies from 20 mg meter squared to 50 mg meter squared in different indications. The treatment interval varies from fortnightly to four weekly in different indications.

It is important ascertain adequate haematological hepatic and renal function for administration dose reductions are required for bilirubin above 20 as outlined in the summary of product characteristics. On liver function improvement the dose can be escalated.

ECG and echocardiograms are recommended at baseline and after every two doses in patients receiving the therapy with the expectation of prolonged response or cure.
However, there is good evidence of reduced cardiac toxicity from this agent than traditional doxorubicin.

The agent is often administered with a glucose solution so care should be taken in diabetic patients.

The agent is occasionally associated with histamine type allergic reactions requiring typical supportive care and therefore slow infusion may sometimes be required for additional monitoring.

**Patients require particular specific advice about skin toxicity.** Sites of pressure on the skin including tight clothing, sites of trauma during leisure activities (in the hands if walking the dog, or the buttocks if horse riding or bicycle riding, for example) will be complicated by quite marked erythema and even swelling and desquamation. These require dose delays until entirely healed, and then dose reductions unless the activity can be changed. Occasionally they will require discontinuation of this agent.

Mouth ulcers are also seen requiring dose delay and dose reduction.

There are specific parameters for management of bone marrow suppression and a dose delay for day 1 platelets less than 75 and neutrophils less than 1.5 recommended. Clearly this may have to be modified in curative treatment.

There is up to date patient information on PPM.

Patient review is required before every cycle, and response assessment after every two to three doses depending upon toxicity and intent. There are no documented late effects. However cardiac monitoring as per conventional doxorubicin would be recommended.

Men in the fertile age must choose effective contraception during this treatment and for five months afterwards.

**Trabectedin.**
This is administered and monitored according to the schedule protocol below.

The indications are third line treatment (or selected second-line cases where ifosfamide is not indicated, unlikely to be tolerated or acceptable to patients) in adult soft tissue sarcoma, and in particular of the subtypes leiomyosarcoma myxoid and round cell liposarcoma.

There are early suggestions it may be indicated in desmoplastic small round cell tumour, and even osteosarcoma.

The efficacy of Trabectedin varies by performance status, specific sarcoma subtype, and phase of use, but disease stabilisation is much commoner than disease shrinkage.
They will wish to continue treatment after a fully informed discussion of advantages and disadvantages and have WHO performance status 0-1, unresectable metastatic disease, at relapse, resistant to both ifosfamide and doxorubicin, with no active clinical trial open locally or elsewhere they are prepared to travel including London centres and the North of England (Sheffield, Manchester, Newcastle) where we refer patients for study in sarcoma with some regularity, or unsuitable for both of these agents due to:

- Cardiac impairment and doxorubicin (Fractional shortening < 24%)
- Renal impairment and ifosfamide (GFR by Cockcroft and Gault <60 or established electrolyte replacement requirement documented as due to renal tubular leak by urinary TmP/GFR assay)

In a case of previous treatment with ifosfamide, the patient will be rechallenged with ifosfamide (rather than trabectedin) as second or third line treatment if toxicity was acceptable to the patient, the patient is free of enduring end-organ toxicities or disease sites (pelvic mass and renal impairment) precluding further ifosfamide treatment and the interval free of disease progression from the last ifosfamide to first findings suggesting relapse was >26 completed weeks.

**Safety tests** for Trabectedin will include

**Full blood count**- Hb>9, Neuts >1.5, Plts >100

**Renal biochemistry**- Creatinine<2.5*ULN GFR >30. DTPA only necessary for calculated GFR <40/unreliable.

**Hepatic biochemistry** – Bilirubin N, and ALT and ALP <2.5 * ULN at inclusion (consider bone isotypes if uncertain re ALP). Elective dose reductions to 0.9 mg/m2 have been used in minor degrees of pre-treatment hepatic impairment, but are outside the existing datasheets and for consultant decision only.

**Creatinine kinase** will also be assayed at baseline and before each treatment is administered and a history of muscular complaints clearly documented at each clinical assessment.

Patients with heavy pre-treatment or a history of previous treatment delay due to myelosuppression should start at a 1.2 mg/m2 dose.

Drug interactions- cytochrome p450 inhibitors and activators significantly alter trabectedin exposure. A list of these is available on [http://www.fda.gov/cder/drug/drugInteractions/tablessubstrates.htm#4](http://www.fda.gov/cder/drug/drugInteractions/tablessubstrates.htm#4)

**Delivery**

Once informed consent, safety tests and staging imaging (within 4 weeks of day 1 of treatment) are in place, Trabectedin will be administered at a dose of 1.5 mg/m2 intravenously as a 24 hour continuous infusion every 3 weeks using a central venous line. With a Baxter pump this can be an out-patient procedure with arrangements to return for disconnection or district nurse to disconnect where training permits. The total dose has to be diluted in 500 mL of 0.9% saline. Antiemesis prophylaxis includes ondansetron 8 mg on day 1, dexamethasone 20 mg iv 30 mins prior to
Trabectedin, then 4 mg orally daily for 3 days, and metoclopramide 10mg qds/prn for 7 days.

**Leeds** patients will receive verbal and written information about contacting Bexley Wing urgently for 24-hour advice from nursing matrons and on-call oncology doctors in the event of fever >38.5, bleeding, bruising or purpura, or other uncontrolled symptoms. This information is standard for all new chemotherapy starters in Bexley Wing. Less than 10% of patients experience severe emesis. Alopecia is uncommon, as is mucositis.

**Review**
Clinical review will be weekly, in Monday morning or Wednesday morning clinic, with other 24 hours bloods ahead or chemo ~24 hours after review.

*All SpRs please discuss any complications they are aware of with Dan Stark/Maria Marples asap*

**Haematological Toxicity**
Drug re-administration on day 22 will be postponed by 1-3 weeks if there was no full hematologic recovery (ANC granulocytes > 1.5 *10^9/L; platelets >100 *10^9/L) from the previous course of treatment.

Drug doses will be adjusted according to the nadir blood counts:
- 1.5 mg/m2 is still appropriate in case of uncomplicated grade 4 neutropenia for less than 5 days with more than 50 *10^9/L platelets
- 1.2 mg/m2 in case of febrile neutropenia and/or less than 50 *10^9/L platelets

**Liver toxicity**
It is of note that the use of creatine kinase to identify patients at risk of hepatic toxicity is critical to the effective and safe administration of Trabectadin.

Treatment in the face of unresolved liver toxicity can increase the odds of severe sepsis. Drug administration will be postponed by 1-3 weeks if ALT and ALP are not below NCIC grade 1. This will be best achieved by blood testing at a practice or hospital near the patient's home on the Friday before a Monday morning clinical review and treatment administration, to avoid patient delays.

The dose of Trabectedin given (upon full biochemical recovery) will be reduced to 1.35 mg/m2 if there was not recovery of liver toxicity to NCIC grade 1 after a 1-week delay. Dose will be reduced 1.2 mg/m2 in cases of reversible NCIC grade 3 to 4 rises in bilirubin between cycles. This will require blood testing on days 8 and 15 after treatment.

**Other toxicities**
Emesis- escalate by Bexley Protocol
Rhabdomyolysis- hydration, urine alkalinisation, and dialysis may be required

**Treatment Duration and Restaging**
Patients will continue treatment until disease progression, manifest as a >25% increase in cross-sectional area of a target metastatic lesion or a new metastatic
lesion, or grade 3 toxicity despite optimal supportive measures and dose reduction to 1.35mg/m².

Radiological assessment of efficacy will be by clinical assessment of symptoms performance status and functional capacity, and radiologically by CT scan with contrast, protocolled to standardise the contrast uptake in Hounsfield units. Routine radiological assessments will be at 6 and 12 weeks. If patient has stable disease at 12 weeks, or better, I then scans will be repeated every 12 weeks (4 cycles). This will be managed through the radiology members of the Sarcoma MDT. The facility to measure the enhancement of lesions is available routinely within PACS in Bexley Wing.

It is subject to NICE approval TA185. There is some early evidence that the Choi criteria maybe helpful in assessing response rather than more traditional RECIST-type radiological criteria.\textsuperscript{23}

The attached protocol provides the baseline clinical safety data and safety tests, the common and important toxicity and its management. There is patient information for the administration of this agent as an inpatient or through ambulatory care on PPM.

Patient’s should be reviewed prior to each cycle. The schedule is three weekly.

Dose-adjustments are required for
- complicated in-cycle neutropenia less than 0.5 complicated by fever or infection,
- thrombocytopenia less than 25,
- bilirubin more than 2.5 times the upper limit of normal, increasing hepatic transaminases more than 2.5 times the upper limit of normal

Dose reductions are from 1.5 mg to meter squared to 1.2 and then to 1 mg per meter squared with no data on lower doses.

Patients with renal insufficiency with creatinine clearance less than 30 ml per minute are cautioned in the administration of this drug.

Day 1 creatinine kinase greater than 2.5 times the upper limit of normal is a contraindication to the use of this drug - retreatment prior to resolution of creatine kinase to normal is associated with idiosyncratic and at times life-threatening bone marrow toxicity, liver toxicity, renal or multi-organ failure. Clinical rhabdomyolysis may occasionally be seen.

There is little data on late effects but this agent is not currently being used in a curative or radical intent indication.

**Gemcitabine/docetaxel**

\textsuperscript{23} Taieb Eur J cancer (2014) PMID: 25499439
Gemcitabine & docetaxel is indicated in the treatment of leiomyosarcomas, particularly of uterine origin, and there is some use in the treatment of recurrent osteosarcomas, Ewing’s family tumours and in the second and third line treatment of adult type soft tissue sarcomas.

Patient should be of WHO performance status 2 or better.

At full dose of 1200 mg meter squared day 1 and 8 of gemcitabine and docetaxel 100mgs per metre squared on day eight in the original study, filgrastim was used but thrombocytopenia was still dose limiting. Patients with prior pelvic irradiation should start at 25% dose reduction.

**Contra-indications**
Prior to its administration haematology, hepatic and renal functional assessments are required.
Existing peripheral neuropathy greater than grade 1
Bone marrow suppression, platelets below 75, neutrophils below 1
Bilirubin or transaminases higher than 5* the upper limit of normal
Alkaline phosphatase higher than 2.5 *the upper limit of normal creatinine greater than 175 mol per litre.
Prior history of drug allergy should be treated with care as docetaxel causes hypersensitivity.

The expected toxicities are bone marrow suppression and its complications, fatigue, although emesis is rare, as is mucositis. The treatment is scheduled every 21 days. The patient should be reviewed after every cycle. Dose reductions of 25% are indicated for
- Grade 3 or higher in-course neutropenia
- Grade 2 neuropathy
- Grade 3 hepatic biochemical toxicity
- Any other grade three non-haematological toxicity.

Dose response assessment should be after two to three cycles depending upon toxicity, patient characteristics and indication.

There is little data on the late effects of these agents but this is not currently being used in a radical or curative setting.

**Imatinib**
Imatinib revolutionised the treatment of gastrointestinal stromal tumour when it first became available in 2001/2. It is now indicated in this condition, as well as evidence of efficacy in chordoma, desmoid/aggressive fibromatosis, dermatofibrosarcoma protuberans, and pigmented villonodular synovitis/germ-cell tumour of tendon.

In each of these the available data indicates certain mutation subgroups of these conditions are more or less sensitive to Imatinib and mutation analysis should be considered. It is established in gastrointestinal stromal tumour that efficacy varies by mutation type.
Prior to administration normal haematological and renal and hepatic biochemical function is required.

Imatinib can be started at low-dose in patients of poor performance status and dose escalated upon response in selected cases with informed consent. Even patients with very advanced disease with jaundice and poor performance status can be salvaged with Imatinib therapy in cases of GIST.

Imatinib is indicated in the adjuvant treatment of high risk GIST according to the NICE guidance of 2014. This is indicated for three years. The dose is 400 mg once daily.

Imatinib is well tolerated. Whilst in haematological conditions it is associated with bone marrow suppression, this is very uncommon in the management of GIST although occasionally seen. Dose reductions to 300 mg may be required in cases of established toxicity resistant to supportive care. It causes peri-orbital oedema, rash, with some photosensitivity, diarrhoea, and fatigue. Each of these can be managed by dose delay modification or supportive medication as appropriate. The minority of patients with very small body surface area may require elective dose reductions.

There is current patient information on Imatinib on PPM.

Imatinib can be provided by home delivery once a dose and toxicity has been established at the level of the individual patient and symptoms are controlled.

Initially review should be at 2, 3, 4 and 6 weeks from starting a patient on Imatinib (modified to individual patient requirements).

Response assessment should be individualised. For patients with advanced disease clinical palliative response can be apparent within only days of starting the medication in some cases. There are specific follow-up guidelines for monitoring after Imatinib in the adjuvant setting and metastatic disease setting based upon mutation type in the resected tumour and clinical characteristics and intent. PET scans before and after Imatinib can be used in selected cases to evaluate response, but can at times be misleading, in particular in cases of bleeding.

There is little data on the late effects of Imatinib although patients have been managed with it for 12 years or more continuously in GIST and no significant late effects are documented.

Imatinib can be interrupted during the response phase at any point beyond one year, and restarted upon progression without detriment to overall survival. Patients will relapse once Imatinib is stopped, but each will re-respond to re-challenge if treated promptly. A tiny minority of patients who were approaching the point of Imatinib resistance when Imatinib was stopped will not re-respond.

Imatinib can be used to the neoadjuvant indication, to attempt to make a marginally resectable or unresectable mass become resectable. Reassessment should be at six weeks and then three months, six months and nine months. Surgical re-referral should be at nine months with surgery planned for twelve months. Imatinib should be
withheld for a minimum of one week before and one week after surgery, depending upon the risks of wound progress with wound healing.

**Paediatric-type TYA protocols**

**Ewing sarcoma**

Management of this family of tumour is based on the principles embodied within the Euro Ewings 1999 and Euro Ewings 2012 trials. Dose modifications may be required in patients over the age of 45. As noted above, dose-density is critical. There is an indication for high-dose chemotherapy and stem cell rescue in relapsed disease. This is a specialist treatment, requiring integration with sarcoma MDT, often a bone sarcoma MDT, requiring combination chemo-radiotherapy and intensive therapies with intensive supportive care.

**Osteosarcoma**

Management of osteosarcoma is according to the principles embodied by the EURAMOS 1 protocol. The treatment used is according to the standard arm, MAP.

Patients within the NICE ruling criteria aged 2 to 30 years with high-grade localised osteosarcoma should be considered for treatment with Mifamurtide in the post-operative phase. Prior to Mifamurtide administration renal and hepatic function require measurement. There is some unpredictability of pharmacokinetics of Mifamurtide in moderate hepatic impairment.

Mifamurtide is associated with rigors and chills during the early administrations which can be effectively managed with supportive care agents. Corticosteroid should be avoided as they may aggravate the effectiveness of Mifamurtide upon macrophage-monocyte function. There is no evidence of other excess toxicities from the addition of Mifamurtide to combination chemotherapy.

Patient information for the administration of Mifamurtide can be found on [www.medicines.org.uk/EMC](http://www.medicines.org.uk/EMC).

Mifamurtide is administered initially twice a week and then weekly. Avoid concurrent administration with doxorubicin although this is a theoretical rather than a documented concern. It is a Liposome product and therefore in principle doxorubicin could change its toxicity schedule as a result.

Patient review should be alongside the administration of chemotherapy. Once the chemotherapy is completed the patient should receive consultant review once every four weeks on the ward or in clinic.

There is no indication for dose modification. There is no indication for response assessment outside that which would routinely be required from the osteosarcoma being treated.

There is extensive evidence of late long-term follow-up and cure after Mifamurtide with no significant late effects identified. However of course these patients with osteosarcoma will be undergoing extensive late effects monitoring the administration
of cisplatin, doxorubicin and methotrexate that was also required and the surgical issues.

**Important drug interactions**
It is important to note that high-dose Methotrexate has clinically important interactions with
- penicillins,
- non-steroidals,
- proton pump inhibitors, and
- potentially some other antibiotics.

The patients excreting methotrexate or about to start methotrexate treatment developing neutropenic fever penicillin-containing antibiotic should be avoided (including tasasin) and meropenem is greatly preferred. Doses of non-steroidals, proton pump inhibitors and other antibiotics should not be adjusted during a course of methotrexate, and if they need to be adjusted changes in the excretion of methotrexate at later points after its administration should be expected.

**Aggressive fibromatosis regimens - Tamoxifen and non-steroidals**
Tamoxifen with non-steroidal agents controls the growth of desmoid fibromatosis in 10 to 20% of patients. Tamoxifen dose is high, 80 mg once daily and associated with adverse effects from oestrogen withdrawal. Patients should be reviewed monthly in the first instance. Non-steroidals adverse effects should be minimised with gastric protection.

There is no patient information for the administration of Tamoxifen on PPM.

Dose modifications may be necessary due toxicity, and dose breaks. They should be individualised to the patient. Response assessment should be by symptom relief, and then by cross-sectional reassessment after two to three months depending upon intent symptoms and toxicity.

There are clearly documented long-term adverse effects of Tamoxifen including bone density, endometrial neoplasms and thrombotic effects, patient should be counselled on these at the start of the treatment, informed consent taken, but routine post-treatment screening is not subject to clear evidence.

**Methotrexate/vinblastine**
This is a long-term weekly outpatient regimen which can reduce the size and delay the growth of unresectable desmoid/aggressive fibromatosis and very occasionally cause shrinkage.

Prior to its administration documentation of bone marrow, renal, and hepatic function is mandatory as is documentation of the extent of any existing neuropathy. There are indications for dose delay for marrow suppression, mucositis and renal or hepatic impairment. Bone marrow suppression is seen in up to half of patients, and abnormal liver biochemistry in about a quarter.

Response assessment is every twelve weeks by MRI of the primary site.
Patient information in the administration of this regimen is available on PPM.

Clinical review should be every four weeks, whilst the doses and toxicity are evaluated. There is evidence of late peripheral and autonomic neuropathy from the prolonged administration of vinblastine. This may be earlier in onset the older the patient is. There is evidence of late long-term hepatic and renal toxicity from methotrexate administered peripherally although this is a conservative dose and that is relatively unlikely.

For the administration of the other agents used in aggressive fibromatosis (Imatinib, percolated liposomal doxorubicin) see above.

**Gastro-Intestinal Stromal Tumour (GIST)**

Patients with GIST typically present in the following ways:

1. Mass in GI tract or peritoneum, which may or may not be resectable:
   a. Symptomatic – e.g. anaemia, pain, GI bleed, obstruction.
   b. Incidental finding.
2. Metastatic disease
3. Recurrence following resection of primary GIST

The management of these conditions is considered below.

1. **Resectable primary GIST**

These patients usually have resection of the primary and are discussed in the sarcoma MDTM. Risk of recurrence is determined by the Miettinen classification:

<table>
<thead>
<tr>
<th>Mitotic Index</th>
<th>Size</th>
<th>Risk for Progressive Disease* (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤ 5 per 50 hpf</td>
<td>≤ 2 cm</td>
<td>Gastric: None (0%)</td>
</tr>
<tr>
<td></td>
<td>&gt; 2 ≤ 5 cm</td>
<td>Duodenum: V-low (1.9%)</td>
</tr>
<tr>
<td></td>
<td>&gt; 5 ≤ 10 cm</td>
<td>Low (3.6%) (Insuff)</td>
</tr>
<tr>
<td>&gt; 5 per 50 hpf</td>
<td>&gt; 10 cm</td>
<td>Jejunum/ ileum: Low (4.3%)</td>
</tr>
<tr>
<td></td>
<td>≤ 2 cm</td>
<td>Rectum: Low (0.5%)</td>
</tr>
<tr>
<td></td>
<td>&gt; 2 ≤ 5 cm</td>
<td>Mod (16%) (Insuff)</td>
</tr>
<tr>
<td></td>
<td>&gt; 5 ≤ 10 cm</td>
<td>High (50%)</td>
</tr>
<tr>
<td></td>
<td>&gt; 10 cm</td>
<td>High (55%) (Insuff)</td>
</tr>
<tr>
<td></td>
<td>≤ 2 cm</td>
<td>High (86%)</td>
</tr>
<tr>
<td></td>
<td>&gt; 2 ≤ 5 cm</td>
<td>High (86%) (Insuff)</td>
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<tr>
<td></td>
<td>&gt; 5 ≤ 10 cm</td>
<td>High (85%)</td>
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<tr>
<td></td>
<td>&gt; 10 cm</td>
<td>High (90%) (Insuff)</td>
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<td>High (90%) (Insuff)</td>
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<tr>
<td></td>
<td>&gt; 5 ≤ 10 cm</td>
<td>High (90%)</td>
</tr>
<tr>
<td></td>
<td>&gt; 10 cm</td>
<td>High (71%)</td>
</tr>
</tbody>
</table>

*Insuff = insufficient data; V-low = very low; Mod = moderate

Patients with a **moderate or high risk** of recurrence have mutation analysis performed on their tumour.
All patients are assessed in the medical oncology sarcoma clinic, given patient information, have a blood test (FBC, U&Es, LFTs, calcium, d-dimer), and have a post-operative baseline CT scan of abdomen/pelvis (and chest if not previously scanned).

Follow-up depends on risk of recurrence, and can be shared with surgeons if no adjuvant treatment being given. Bloods as above at each visit:

**Very low risk:** 3-monthly review for 1 year, no further scans, then annual review and discharge at 3 years.

**Low risk:** 3-monthly review, CT abdo/pelvis around anniversary of surgery for 3 years, then 6-monthly review and discharge at 5 years.

**Moderate risk:** 3-monthly review, CT abdo/pelvis at 6, 12, 24 36 months following surgery, then 6-monthly review and discharge at 5 years.

**High risk:** 3-monthly review, CT abdo/pelvis at 6, 12, 24, 30, 36 months following surgery, then 6-monthly review and discharge at 5 years.

**Familial/syndromic GIST:** lifelong follow-up may be offered. Patients may also wish to be referred for advice to the PAWS-GIST clinic which takes place quarterly in Cambridge, https://www.pawsgistclinic.org.uk/.

Patients with moderate or high risk GIST are offered **adjuvant imatinib** 400 mg daily for 3 years. Visit schedule is as for patients with metastatic disease. CT scans are performed 6-monthly during treatment, patients then revert to CT schedule appropriate to their baseline risk of recurrence.

### 2. Unresectable primary GIST

Tumours may be unresectable for technical reasons or because resection would result in unacceptable morbidity. In these circumstances, patients are treated with imatinib as below. If there is a response to treatment, patients may be re-considered for resection after 6-12 months' imatinib. Imatinib should be stopped 1 week before surgery. Following resection, patients may be considered for adjuvant imatinib.

N.B. Imatinib is not approved as a neo-adjuvant therapy.

**Metastatic and recurrent GIST**

These patients may present de novo, when they are often unwell, or after a period of surveillance following resection of primary GIST. They may be unwell, as they may have a heavy tumour burden, and may be anaemic from acute or chronic bleeding.

The **diagnosis of GIST** should generally be confirmed by biopsy. In patients where biopsy is impossible or non-contributory and where treatment needs to be started, a PET-CT scan may be performed (high-grade GIST is usually FDG-avid, although low-grade GIST often is not). Imatinib may be started, and the PET-CT scan repeated 2 weeks later. FDG activity should reduce significantly if the lesion is truly a GIST.

Patients may need emergency surgery for metastatic GIST. However, the mainstay of treatment is with tyrosine kinase inhibitors. Imatinib is used first line (unless an imatinib-resistant mutation is found), followed by sunitinib and then regorafenib. Clinical trials should be considered if available.

**Imatinib**
Prior to administration, adequate haematological and renal and hepatic biochemical function is required.

Imatinib is usually started at the approved dose of 400 mg daily. Patients with an unresected primary are at risk of bleeding if the tumour responds promptly, and the patient should be advised to seek urgent medical help for symptoms of bleeding, but the risk is not reduced by starting imatinib at a lower dose. Patients with poor performance status or abnormal bloods, however, may need to start imatinib at a lower dose (100-200 mg daily). The dose may be escalated upon response as tolerated.

Imatinib is well tolerated. It causes peri-orbital oedema, rash, photosensitivity, diarrhoea, and fatigue. Each of these can be managed by dose delay modification or supportive medication as appropriate. Whilst in haematological conditions it is associated with bone marrow suppression, this is very uncommon in the management of GIST. Dose reductions to 300 mg may be required in cases of established toxicity resistant to supportive care. The minority of patients with very small body surface area may require elective dose reductions.

There are few drug interactions with imatinib, but paracetamol metabolism may be slowed down. Patients should therefore be advised not to take more than 2 g paracetamol in 24 hours while taking imatinib.

There is current patient information on Imatinib on PPM.

Review should be at 2, 3, 4 and 6 weeks from starting a patient on Imatinib (modified to individual patient requirements). Imatinib can be provided by home delivery once a dose has been established and toxicity is controlled. Patients can be registered for home delivery in clinic, and prescriptions generated at each clinic visit to ensure continuity of supply.

Once patients are established on imatinib treatment, they are reviewed in clinic every 3 months with blood tests on each visit. CT scans of abdomen and pelvis are performed every 3 months for the first year. After this, patients with a good response and a mutation in KIT exon 11 may be scanned every 6 months as they are more likely to have a prolonged response, but patients with other mutations should remain on 3-monthly scans. Patients with a lower chance of response to imatinib (e.g. wild-type GIST) may have an initial CT scan at 6 weeks.

Imatinib can be interrupted during the response phase at any point beyond one year, and restarted upon progression without detriment to overall survival. Patients will relapse once Imatinib is stopped, but more than 80% will re-respond to re-challenge if treated promptly. The patients who do not re-respond are likely to have been approaching the point of imatinib resistance.

Patients with progressive disease on imatinib should be discussed in the sarcoma MDTM. A single site of progression may be amenable to surgical or radiological intervention (resection, RFA etc.) which may prolong the time to imatinib failure. However, recurrence that cannot be controlled by other means should prompt a switch to sunitinib.

**Sunitinib**

The chance of response to sunitinib depends on the mutation status of the tumour. Patients with KIT exon 9 mutations have the best chance of response, followed by patients with wild-type GIST. Patients with KIT exon 11 mutations have a low chance of response to sunitinib,
and it may be more appropriate for them to remain on imatinib while it is controlling their symptoms, although this is not an indication that is approved by NICE.

Sunitinib is usually dosed at 50 mg daily on a 4 week on, 2 week off schedule. Patients are reviewed after 2 and 4 weeks of the first cycle, and then at the beginning of the second cycle. If sunitinib is well-tolerated, they are reviewed 6-weekly (at the start of each cycle), with 3-monthly CT scans. Patients who do not tolerate this schedule may have the dose reduced, and some may find continuous dosing more manageable.

Similarly to imatinib, sunitinib causes peri-orbital oedema, rash, photosensitivity, diarrhoea, and fatigue. Severe, blistering or necrotising rash may require hospital admission and permanent discontinuation of imatinib. Sunitinib may also cause hypertension, proteinuria, hypothyroidism, hand-foot syndrome, yellow pigmentation of the skin, and changes in hair pigmentation (which can result in a stripy look that some patients find distressing). Patients should therefore have blood pressure and urinalysis checked at each visit, and 6-weekly thyroid function tests.

Patients who progress on, or do not tolerate, sunitinib may be considered for regorafenib.

**Regorafenib**

Regorafenib is available from the cancer drugs fund as third-line therapy for metastatic GIST. A cancer drugs fund form must therefore be completed and emailed to leedsth-tr.medicinesfundingrequest@nhs.net before treatment is initiated; this can be done on the same day as treatment is started.

The dose of regorafenib is 160 mg daily for 3 weeks followed by a week’s break. Patients are reviewed and have liver function tests checked 2-weekly for the first two cycles, and are then seen 4-weekly.

The side-effect profile of regorafenib is similar to that of sunitinib. Patients therefore have the usual GIST blood tests, blood pressure and urinalysis checked on each visit. TFTs should be checked monthly.

**Progression after regorafenib**

There is no approved treatment for metastatic GIST following regorafenib. Some patients respond to re-challenge with imatinib, for which an individual funding request would have to be submitted.

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17.1 Adjuvant systemic therapy

The following subtypes of adult-type STS are known to be more chemo-sensitive:

- Synovial sarcoma
- Leiomyosarcoma
- Myxoid liposarcoma

‘Adjuvant’ chemotherapy for patients with these subtypes may be considered for fit patients who present with either potentially resectable metastatic disease or who are at very high risk of developing metastatic disease following standard resection and adjuvant local therapy of
the primary. Even in the absence of clear data supporting routine adjuvant therapy it is felt that early delivery of systemic therapy to these patients may at least prolong relapse-free survival and maximises likelihood of cure for a small proportion (probably less than 15%) by selecting patients who are shown to be at high risk for future systemic failure, have more chemosensitive subtypes and who are less likely to develop treatment related toxicity (by virtue of low tumour load and better overall fitness).

A schema for the management of these patients is presented following ‘Palliative systemic therapy’

17.2 Potentially resectable metastatic disease

Patients with 3 or fewer resectable metastatic lesions should be considered for resection or ablative therapy for the metastases (via interventional radiology or Stereotactic Radiotherapy within the NHS England Commissioning through Evaluation scheme). Consideration should be given to ‘induction’ chemotherapy followed by radical treatment. If surgery is being considered, a surgical opinion should be sought prior to chemotherapy and, if resectable, the patient may be offered 2 cycles of chemotherapy prior to surgery. As tumour response is the primary goal, multi-agent chemotherapy (if this offers a superior likelihood of response over monotherapy) should be considered. Those patients with tumours showing significant necrosis histologically could be offered 2-4 further cycles of post-operative adjuvant chemotherapy.

Those with otherwise resectable disease where a decision is made not to offer systemic therapy, a 3 month interval CT should be considered; if there is no progression (no new lesions) on interval scanning surgery should proceed. Progressive disease should be treated as for palliative.

Borderline resectable disease (where progression of disease on chemotherapy may render it unresectable) in patients who are fit for chemotherapy should undergo resection and then be considered for ‘adjuvant’ chemotherapy as described previously.

18 Malignant soft tissue tumours by site

18.1 Limb & trunk

18.1.1 Clearly operable (limb preservation)

Wide local excision should be attempted in all cases. If WLE feasible based upon pre-operative staging consider pre-operative radiotherapy. Pre-operative RT should not be used if the lesion is likely low grade or if resection likely to be marginal. If pre-operative RT is to be used, consideration should be given to the method of defect closure and the downstream functional consequences of post-operative wound complications. Consideration should be given to re-excision if margins following primary excision are R1 or R2 (ie. an overall R0 excision should be the goal). If pre-op RT is not used consideration should be given to post-op RT given resected tumour grade, margins, size and relationship to deep fascia. European consensus guidelines should be followed. It is helpful for post-op RT planning if surgical clips are placed at the cranial and caudal extent of the surgical bed for limb tumours and in addition laterally for truncal tumours.
18.1.2 Borderline operable (limb preservation)
If WLE not deemed possible on pre-operative staging, consideration should be given to high risk limb preservation or planned marginal resection.

18.1.3 Planned marginal resection
A planned marginal resection may carry relapse risk only slightly higher than WLE, but clip placement at the site of anticipated close margins and careful operative and histopathology reporting are needed to guide a higher dose of adjuvant RT. Higher dose RT carries a greater risk of late functional morbidity and this should be considered if this combined modality approach is to be deployed. Pre-operative RT is not known to improve resectability and should not be used as optimal management of positive surgical margins in this context is unclear.

18.1.4 Amputation
For synovial sarcomas and myxoid liposarcomas chemotherapy response rates are such that neo-adjuvant combination systemic therapy with doxorubicin and ifosphamide can be considered. If there is no objective response after 4 cycles of treatment (or if progressive disease is identified at any stage) the patient should proceed directly to surgery. In selected cases isolated limb perfusion may have a role. Post-operative RT is usually required. Functional implications of aggressive CMT should be considered. Prior to amputation consideration should be given to the likelihood of metastatic failure.

18.1.5 Metastatic
Aggressive local surgery should be avoided in patients with known inoperable metastatic disease. Systemic chemotherapy should be considered. If systemic therapy not deliverable or disease becomes refractory consider palliative radiotherapy. Palliative surgery should be reserved for control of difficult local symptoms.

18.2 Pelvic

18.2.1 Resectable non-metastatic
Tumour resection should be undertaken by the appropriate site specific team after discussion with the sarcoma MDT. There is no role for neo-adjuvant therapy but post-operative radiotherapy should be considered after review within the sarcoma MDT. Technical RT delivery should be undertaken in discussion with the appropriate site specific team.

18.2.2 Unresectable or metatstatic
See 18.1.x

18.3 Supraclavicular

18.3.1 Resectable non-metastatic
Tumour resection should be undertaken by the appropriate site specific team after discussion with the sarcoma MDT. There is no role for routine neck nodal dissection. There is no role for neo-adjuvant therapy but post-operative radiotherapy should be considered after review within the sarcoma MDT. Resection margins are expected to be narrow and/or involved. Standard RT planning margins are not likely to be attainable but due care in considering tissue planes that have not been compromised should be employed. RT doses as for trunk/limb. Technical RT delivery should be undertaken by the appropriate site specific team in collaboration with the sarcoma team.
18.3.2 Unresectable or metastatic

Unresectable but non-metastatic disease should be treated with primary RT using pseudoradical doses. Neo-adjuvant combination chemotherapy may be considered for synovial sarcoma and myxoid liposarcoma as described in limb/trunk. Neo-adjuvant therapy is not otherwise recommended. Metastatic disease should be managed as described in limb/trunk. If unfit for chemotherapy and minimal systemic disease (ie locally progressive disease will/may be unacceptably morbid within the patient's expected life-span) then consider surgical resection and/or high dose RT for local control.

18.4 Breast

18.4.1 Operable non-metastatic

Often arises on a background of soft-tissue field change post-whole breast RT. Mastectomy with a deep cuff of pectoral fascia and/or pectoral muscle is required. If not previously given, followed by wide-field adjuvant RT.

Malignant phylodes should be managed per sarcoma. Mastectomy is indicated. If WLE undertaken and completion mastectomy is not possible then full-dose adjuvant RT is required. Benign and borderline phylodes may be managed with WLE without need for RT.

18.4.2 Unresectable or metastatic

Systemic therapy is indicated. Photography of index lesion (with measurement) may be helpful for cutaneous disease. Palliative radiotherapy as described for limb/trunk. Doses may be limited by prior RT. Pulmonary metastases are commonly sub-pleural and cystic with a higher than average risk of spontaneous pneumothorax. Consider intervention (VATS pleurodesis or palliative RT) for such lesions.

18.5 Cutaneous

18.5.1 Operable non-metastatic

Often arises on a background of soft-tissue field change in the context of prior RT, lymphoedema or solar exposure but may be spontaneous. Excisions should be wide and include periosteum if on the scalp. For lesions arising in areas of field-change, wider than usual margins should be taken and flap reconstruction is often required.

Low grade lesions (including dermatofibrosarcoma protuberans, DFSP) will not require adjuvant RT. Adequate wide margins may spare the need for adjuvant RT even if high grade tumour.

If anatomically restrictive site (facial) and low grade, close observation may be appropriate.

18.5.2 Unresectable or metastatic

Systemic therapy is indicated.
18.6 Lipomata

18.6.1 Operable non-metastatic

18.6.1.1 Superficial (including intrafacial)

Symptomatic lipomas or lipomata > 7 cm should be marginally excised if possible. This is non-sarcoma activity and may be undertaken by any competent surgeon. Adjuvant therapy is not indicated. All resected lipomata should be submitted for both histopathological and cytogenetic analysis. MDM-2 positivity will convert the diagnosis to atypical lipoma which carries a higher local recurrence risk but is NOT a sarcoma diagnosis. Specific additional action is not required unless reoperation on relapse would risk functional deficit. In this case, review by the WY&HSS is appropriate.

18.6.1.2 Deep

Lipomatous tumours deep to deep facia include benign lipoma through to high grade liposarcoma. These cases should all be managed via the sarcoma MDT. Benign imaging may obviate the need to undertake biopsy (by moving straight to excisional biopsy or interval imaging), though a small number of benign looking tumours will be morphologically well differentiated liposarcomas or express MDM-2 (upgrading them to well differentiated liposarcoma). Outcomes are usually excellent, though adjuvant RT may be indicated in complex sites where recurrence (~10%) would pose a functional threat and there will be impact upon cancer pathway performance metrics if an initially downgraded tumour is found to be malignant on excision (usually as part of an 18 week pathway). Deep lipomas are more appropriately excised by a sarcoma surgeon.

19 Malignant bone tumours

Bone tumours fall outside the remit of the Leeds Regional Soft Tissue Sarcoma Group. All suspected primary tumours of bone should be referred to a recognised Bone Sarcoma MDT. Current practice is to refer cases to the Royal Orthopaedic Hospital in Birmingham where a comprehensive rapid access staging, biopsy and surgical service operates. It is recognised that there are links with the Glasgow Bone Tumour Service for bone tumours arising within the facial skeleton and base-of-skull. The Leeds Regional Soft Tissue Sarcoma Group CANNOT provide recommendations for a definitive overall management plan for bone sarcomas as this is the role of the specialist Bone Sarcoma MDTs but it may participate in delivering integrated non-surgical aspects of treatment.
20 Palliative & End of Life Care

20.1 Definitions

Palliative care is part of supportive care. It embraces many elements of supportive care.

Palliative & End of life care is care that helps all those with advanced, progressive, incurable illness to live as well as possible until they die. It enables the supportive and palliative care needs of both patient and family to be identified and met throughout the last year(s) of life and into bereavement. It includes management of pain and other symptoms and provision of psychological, social, spiritual and practical support.

The Department of Health (2008) definition of end of life care states that it includes:

- Adults with advanced, progressive, incurable illness (e.g. advanced cancer, heart failure, COPD, stroke, chronic neurological conditions, dementia);
- Care given in all settings (e.g. home, acute hospital, ambulance, residential/nursing care home, hospice, community hospital, prison);
- Care given in the last year(s) of life
- Patients, carers and family members (including bereavement care).

End of Life Strategy, Department of Health 2008
National Council for Palliative Care Services 2006

20.2 Who Provides Palliative / End of Life Care?

Palliative / end of life care is provided by two distinct categories of health and social care professionals:

- All health care and social care professionals providing the day-to-day care to patients and carers in any care setting
- Those who specialize in palliative care (consultants in palliative medicine and clinical nurse specialists in palliative care, for example) who care for palliative care patients who have complex needs

Those providing day-to-day care should be able to:

- Assess the care needs of each patient and their families across the domains of physical, psychological, social spiritual and information needs
- Meet those needs within the limits of their knowledge, skills, competence in palliative care
- Know when to seek advice from or refer to specialist palliative care services

Training and education in the skills required for palliative / end of life care should be available to and undertaken by all health and social care professionals.

The national strategy Ambitions for Palliative and End of Life Care 2015-2020 sets out the vision to improve end of life care through partnership and collaborative action between organisations at local level throughout England.

More information can be found at: http://endoflifecareambitions.org.uk/
For more information about local improvements, frameworks, tools to support best practice please contact your local End of Life Care Lead or Specialist Palliative Care Team. One aspect of care is to discuss with individuals, if they wish, their preferences regarding the type of care they would wish to receive and where they wish to be cared for in case they lose capacity or are unable to express a preference in the future. This is the process of Advance Care Planning (ACP).

An ACP discussion might include:
- the individual’s concerns and wishes,
- their important values or personal goals for care,
- their understanding about their illness and prognosis,
- their preferences and wishes for types of care or treatment that may be beneficial in the future and the availability of these.

Such discussions can also inform shared decision-making regarding treatments with palliative intent. Local arrangements for recording this information for each individual patient will differ. Many services are developing/have developed Electronic Palliative Care Coordination Systems (EPaCCS) where this information can be shared across professionals and settings (e.g. on SystmOne). Contact your local specialist palliative care team for more information.

20.3 Specialist Palliative Care

Is provided by specialist multidisciplinary palliative care teams in services or units whose core specialty is palliative care (for example hospices, community or hospital palliative care teams). The specialist teams should include palliative medicine consultants and palliative care nurse specialists together with a range of expertise provided by physiotherapists, occupational therapists, dieticians, pharmacists, social workers and those able to give spiritual and psychological support.

Eligibility for referral to specialist palliative care services is based on patient need not diagnosis. The agreed criteria for referral are as follows:

1. The patient has active, progressive and usually advanced disease for which the prognosis is limited (although it may be several years) and the focus of care is quality of life.

2. The patient has unresolved complex needs that cannot be met by the caring team, for example:
   - Uncontrolled or complicated symptoms (e.g. symptoms not adequately controlled within 48 hours by the referring team, or sooner if causing overwhelming distress).
   - Complex psychological/emotional difficulties.
   - Complex social or family issues.
   - Difficult decision making about appropriate future care.

Patients fulfilling these criteria should be referred to and assessed by a member of the specialist palliative care team.

The subsequent care package will be dependent on this assessment and should be made in agreement with the patient, carer(s) and referring team. It is not appropriate for specialist palliative care services to be committed to patients by professionals outside these services. Equally, specialist services should ensure that the assessment process is accessible and responsive to patients in need.
The level of specialist palliative care support required may fluctuate. Shared care of patients between the specialist palliative care professionals and the referring team (for hospital patients) or the primary care team (for patients at home / care home) is usually appropriate. Timely and effective communication is essential in these situations. For these patients advice from specialist palliative care services on a 24 hour basis should be available in all care settings.

Sometimes the specialist palliative care consultant and team may take the lead role in patient care, usually in a specialist in-patient unit (hospice) or designated specialist palliative care beds.

Referral systems for specialist palliative care services vary in different areas. They should be clear to all local referring consultants and primary care teams.

20.4 Further Links and Information
Contact the local Specialist Palliative Care Team for further information

20.5 Directory of West Yorkshire & Harrogate Cancer Alliance Specialist Palliative Care Services
The Directory has been checked and updated in May 2017

Bradford, Airedale, Wharfedale and Craven
Bradford Teaching Hospitals NHS Foundation Trust
Airedale NHS Foundation Trust
NHS Bradford, Airedale, Wharfedale and Craven
Website: www.palliativecare.bradford.nhs.uk

| Airedale General Hospital Palliative Care Team | Tel       | 01535 292184 |
|                                             | Tel       | 01535 295016 |
|                                             | Fax       | 01535 295036 |
| Sue Ryder Care – Manorlands Hospice (Oxenhope) | Tel       | 01535 642308 |
|                                             | Fax       | 01535 642902 |
| Bradford Teaching Hospitals Palliative Care Team | Tel       | 01274 364035 |
|                                             | Fax       | 01274 366851 |
| Bradford Community Palliative Care Team | Tel       | 01274 323511 |
|                                             | Fax       | 01274 215660 |
| Marie Cure Hospice (Bradford) | Tel       | 01274 337000 |
|                                             | Fax       | 01274 337095 |
| Out of Hours Advice via on-call Palliative Medicine Consultant via Marie Curie Hospice / Manorlands Hospice | Tel       | 01274 337000 |
|                                             | Tel       | 01535 642308 |
**Calderdale and Huddersfield**  
Calderdale & Huddersfield NHS Foundation Trust  
NHS Calderdale  
NHS Kirklees  

<table>
<thead>
<tr>
<th>Service</th>
<th>Tel</th>
<th>Fax</th>
</tr>
</thead>
<tbody>
<tr>
<td>Calderdale Royal Hospital &amp; Huddersfield Royal Infirmary Palliative Care Team</td>
<td>01484 342965</td>
<td>none</td>
</tr>
<tr>
<td>Calderdale Community Palliative Care Team</td>
<td>01422 310874</td>
<td>01422 378425</td>
</tr>
<tr>
<td>Overgate Hospice</td>
<td>01422 379151</td>
<td>01422 384210</td>
</tr>
<tr>
<td>Kirkwood Hospice and Community Palliative Care Team</td>
<td>01484 557906</td>
<td>01484 557918</td>
</tr>
<tr>
<td>Out of Hours Advice via Hospices</td>
<td>01422 379151</td>
<td>01484 557900</td>
</tr>
</tbody>
</table>

**Harrogate and District**  
Harrogate NHS Foundation Trust  
NHS North Yorkshire and York  
Website: [https://www.hdft.nhs.uk/services/palliative-care/](https://www.hdft.nhs.uk/services/palliative-care/)

<table>
<thead>
<tr>
<th>Service</th>
<th>Tel</th>
<th>Fax</th>
</tr>
</thead>
<tbody>
<tr>
<td>Harrogate Hospital and Community Palliative Care Team</td>
<td>01423 553464</td>
<td>01423 555763</td>
</tr>
<tr>
<td>St Michael’s Hospice</td>
<td>01423 872658</td>
<td>01423 815454</td>
</tr>
<tr>
<td>Out of Hours Advice via Hospice</td>
<td>01423 879687</td>
<td></td>
</tr>
</tbody>
</table>

**Leeds**  
**Leeds Palliative Care**  
Website: [www.leedspalliativecare.co.uk](http://www.leedspalliativecare.co.uk)

<table>
<thead>
<tr>
<th>Service</th>
<th>Tel</th>
<th>Fax</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leeds Teaching Hospitals NHS Trust Specialist Palliative Care Team</td>
<td>0113 2064563</td>
<td>0113 2064863</td>
</tr>
<tr>
<td>Sue Ryder Care - Wheatfields Hospice and Community Palliative Care Team (West Leeds)</td>
<td>0113 2787249</td>
<td>0113 2302778</td>
</tr>
<tr>
<td>St Gemma’s Hospice and Community Palliative Care Team (East Leeds)</td>
<td>0113 2185500</td>
<td>0113 2185524</td>
</tr>
<tr>
<td>Out of Hours Advice via SJUH Switchboard</td>
<td>0113 2433144</td>
<td></td>
</tr>
</tbody>
</table>
### Mid Yorkshire
Mid Yorkshire Hospitals NHS Trust  
NHS Wakefield District  
Kirklees PCT  
Website: [https://www.midyorks.nhs.uk/palliative-care1](https://www.midyorks.nhs.uk/palliative-care1)

<table>
<thead>
<tr>
<th>Organization</th>
<th>Phone 1</th>
<th>Phone 2</th>
</tr>
</thead>
</table>
| Dewsbury Hospital and Community Palliative Care Team | Tel 01924 816052  
Fax 01924 543883 | |
| Dewsbury Day Support and Drop-in (Rosewood Centre) | Tel 01924 512039 | |
| Mid Yorkshire Hospitals NHS Trust Palliative Care Team | Tel 01924 543801  
Fax 01924 543883 | |
| Pontefract Community Palliative Care Team (Prince of Wales Hospice) | Tel 01977 781456  
Fax 01977 796209 | |
| Prince of Wales Hospice (Pontefract) | Tel 01977 708 868  
Fax 01977 600097 | |
| Wakefield Hospice | Tel 01924 331400  
Fax 01924 362769 | |
| Out of Hours Advice via Pinderfields Hospital Switchboard | Tel 01924 541000 | |

### York
York Hospitals NHS Foundation Trust  
NHS North Yorkshire and York  
[https://www.yorkhospitals.nhs.uk/our_services/az_of_services/palliative_care/](https://www.yorkhospitals.nhs.uk/our_services/az_of_services/palliative_care/)

<table>
<thead>
<tr>
<th>Organization</th>
<th>Phone 1</th>
<th>Phone 2</th>
</tr>
</thead>
</table>
| York Hospital Palliative Care Team | Tel 01904 725835  
Fax 01904 726440 | |
| Community Palliative Care Team | Tel 01904 724476  
Fax 01904 777049 | |
| St Leonard’s Hospice | Tel 01904 708553  
Fax 01904 704337 | |
| Out of Hours Advice via Hospice | Tel 01904 708553 | |
21 Rehabilitation

21.1 Surgery
Surgery for sarcoma, especially sarcoma of the limbs, can result in significant early and late functional deficits. Early deficits may be overcome to be replaced by later evolving side effects of RT or compensatory changes. Appropriate assessment and input by the MDT should aim to minimize the impact of these changes.

21.2 Physiotherapy
WY&HSS has 2 dedicated sarcoma physiotherapists. Their opinion should be sought on all cases of sarcoma discussed by the MDT, though it is not anticipated that formal physiotherapy assessment and input be required for all cases.

21.2.1 General goals
To provide a seamless, patient-centred physiotherapy service for adults with bone and soft tissue sarcoma that can be accessed from initial contact and at any stage during their treatment pathway.

21.2.2 Role of the Sarcoma Physiotherapist
• To work as a core member of the Leeds Soft Tissue Sarcoma team, providing soft tissue and bone sarcoma patients (as in- and out-patients) with Specialist physiotherapy input from diagnosis and along their treatment pathway including surgery, radiotherapy, chemotherapy and during palliative treatment.
• To ensure that each sarcoma patient is able to access physiotherapy that is specific to their individual needs at the optimum time and provided by the most appropriate physiotherapist. This process involves a combination of both direct physiotherapy input and appropriate colleague liaison.
• Integrated role summarised in attached chart

21.3 Outcome measures
The sarcoma physiotherapist will take a lead in performing serial functional assessments and recording TESS (Toronto Extremity Salvage Score) outcomes.

21.4 Specialist Rehabilitation
A non-exhaustive list of services includes prosthetics and orthotics, lymphoedema services, dietetics and speech and language therapy. Access will be tailored to the patient’s needs and should be delivered as close to the patient’s home as possible as limited by the geographic availability of specialist support.
22 Patient information & support

Sarcoma is a rare disease and general awareness in its regard is limited. Diagnostic, treatment, rehabilitation and follow-up pathways are also complex and cross many multi-profession, specialty and geographic boundaries. Provision of reliable information is essential and should be provided to the patient, family/carers and other healthcare professionals in an appropriate format and timescale that meet the individual’s needs.

Whilst every core-member of the WY&HSS MDT has a responsibility to provide clear verbal and written communication at each stage of the patient pathway, the Sarcoma Clinical Nurse Specialist (CNS) has a particularly important bridging/continuity and reinforcement role.

22.1 The Sarcoma CNS

The Sarcoma CNS is responsible for the management of a defined caseload of patients with Sarcoma, providing expert nursing advice and support to those patients with Sarcoma and other health professionals in relation to this patient group. They carry continuing responsibility for the assessment of care needs, the development, implementation and evaluation of programmes of care and the setting of standards of care.

22.1.1 Role of the Sarcoma Clinical Nurse Specialist

To be present at the point the patient has first contact with the core WY&HSS team. This may be during the diagnostic phase when there is a suspicion of sarcoma and the patient is undergoing investigation. It is not possible for the sarcoma CNS to be present at all imaging (pre-diagnostic) appointments, though telephone contact will be made when feasible. It is therefore essential that the referring team provide accurate information about the diagnostic process to the patient and that they supply appropriate WY&HSS literature.

To act as the patient’s Key Worker and ensure contact details are provided to the patient and carer. To offer support and provide information (Patient Information Pathway). To direct the patient to where further information, advice and support is available.

The CNS will perform a holistic assessment and develop a care plan to meet the needs identified in the assessment. They will coordinate the patient’s movement through their care pathway.

For those patients undergoing surgery, the Sarcoma CNS will provide support and information in the pre and post-operative period. They will visit the patient whilst an in-patient and will liaise directly with the ward health care professionals ensuring follow up arrangements are in place.

The CNS will be present at the other key discussion points (Sarcoma Pathway) that take place between the patient and medical staff regarding transitions between phases in the patient pathway: adjuvant therapy, suspected relapse, palliative and end-of-life. The Sarcoma CNS will provide support and relevant information at those points and ensure that relevant contact details are still available. They will visit those patients who require in-patient treatment and will again liaise directly with the ward health care professionals to ensure continued coordination of patients care.
They will support the patient and their families at those times ensuring they receive the required information to enable them to participate in their care delivery. They will liaise with Community Services, to ensure seamless provision of care delivery. Those Community Services will include District Nurses, Macmillan Teams and Hospices.

22.2 The ‘information prescription’

Written patient and carer information is required for each step in the patient pathway. This does not replace face-to-face verbal communication and patients should also be offered copies of clinic letters if they wish. Patients who choose to receive copies of clinic letters must understand that the letters are primarily for communication between medical professionals but that they may help their recall of issues discussed during consultations and of plans agreed. Clinicians should realise that information of a potentially damaging nature (suspicions of relapse or prognostic estimates) contained in these letters should have been discussed with the patient.

22.2.1 Information checkpoints

Written information for use in the following circumstances is available or is in development. The wide spectrum of STS is such that both generic and specific information may be required, dependent upon circumstances.

**Suspected diagnosis**
- Imaging
- Biopsy

**Diagnosis**
- Malignant
- Benign

**Treatment**
- Surgery
- Radiotherapy
- Chemotherapy

**Rehabilitation**
- Follow up
- Relapse
- Palliative care
- End-of-life care

**Additional resources**
- Cancer Backup
- Sarcoma
- Gist
23 Follow-up

Follow-up for treated sarcoma patients incorporates scheduled screening for local and systemic relapse, for the early and late consequences of local and systemic therapies and the provision of psychological support. No study has clearly shown an improvement in overall outcomes as a consequence of routine follow-up, though most agencies recommend follow-up as a component of survivorship care. A careful balance must be struck between opportunity gain arising from follow-up and the patient anxiety potentially provoked by it. As symptomatic relapse may occur between scheduled follow-up appointments it is essential that patients are informed of how to make contact with the sarcoma service so that unscheduled review can be arranged.

23.1 Components of scheduled follow-up care

23.1.1 Clinical assessment
Relapse and toxicity screening enquiry (local, end-organ and systemic) and physical examination. Toxicity recording ideally with CTCv3/4

23.1.2 Imaging
Local
There is no role for baseline post-treatment local imaging unless either (a) abdominopelvic primary or (b) non-abdominopelvic site but difficult to follow clinically. Clinical circumstance will dictate modality, notionally CT for abdo/pelvis and MRI for other sites.
Systemic
Chest x-ray.

23.2 Laboratory tests

There is no specific marker for sarcoma relapse at this time.
Routine haematology, biochemistry (U&E, LFT, Bone) is indicated if:
- patient has received systemic therapy.
- patient has had radiotherapy with potential effect upon end-organ function (essentially any non-limb site).
- primary tumour was abdominopelvic

23.2.1 Specific tests of end-organ function
- TESS score should be completed for limb sarcomas.
- A post-treatment echocardiogram should be performed 3 months after completion of any adjuvant anthracycline containing regimen.
- Formal pulmonary function tests (including DLCO) should be performed 12 months after whole-lung radiotherapy.
- An endocrine profile is required if post-treatment endocrinopathy is possible.
23.2.2 Scheduling

Non-abdominopelvic tumours

<table>
<thead>
<tr>
<th>Group</th>
<th>Nature</th>
<th>Local relapse risk</th>
<th>Systemic relapse risk</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Benign without complex surgery</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>2</td>
<td>Benign with complex surgery</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>3</td>
<td>Low grade malignant</td>
<td>Intermediate</td>
<td>Low</td>
</tr>
<tr>
<td>4</td>
<td>High grade no adjuvant RT</td>
<td>Intermediate</td>
<td>Intermediate</td>
</tr>
<tr>
<td>5</td>
<td>High grade needing adjuvant RT</td>
<td>Intermediate</td>
<td>High</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Group</th>
<th>Sub-group</th>
<th>Clinical assessment</th>
<th>Local imaging frequency</th>
<th>Lung imaging frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-2</td>
<td>All</td>
<td>Discharge post surgery</td>
<td>Not needed</td>
<td>Not needed</td>
</tr>
<tr>
<td>3</td>
<td>All</td>
<td>6 monthly to 5 years</td>
<td>Not routinely required</td>
<td>Not needed</td>
</tr>
<tr>
<td>4-5</td>
<td>Limb</td>
<td>4 monthly to year 2, 6 monthly to 5 years, annual to 10 years</td>
<td>Not routinely required</td>
<td>CXR each visit</td>
</tr>
<tr>
<td></td>
<td>Abdo-pelvic</td>
<td>Per limb, based on grade</td>
<td>Baseline post treatment. No other routine imaging ? 1 year CT</td>
<td>CXR each visit if G2/3</td>
</tr>
<tr>
<td></td>
<td>Ewings</td>
<td>3 monthly years 1-2, 6 monthly years 3-4, annual to 10 years or if &lt;30 years refer to LTFUC after year 5</td>
<td>Not routinely required</td>
<td>CXR each visit</td>
</tr>
<tr>
<td></td>
<td>Osteosarcoma</td>
<td>3 monthly years 1-2, 4 monthly years 3-4, 6 monthly to end of year 5, annual to 10 years or if &lt;30 years refer to LTFUC after year 5</td>
<td>X-ray site of primary each visit to end of year 4</td>
<td>CXR each visit and CT thorax 6 monthly</td>
</tr>
</tbody>
</table>

Abdominopelvic tumours

Non-GIST tumours are at high risk for local failure (manage as for group 5 above). Aggressive imaging-based follow-up is not considered useful. Baseline CT with imaging on clinical progression.

GIST risk groupings are described

<table>
<thead>
<tr>
<th>Size</th>
<th>&lt;5cm</th>
<th>6-10cm</th>
<th>&gt;10cm</th>
<th>Any</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mitoses/50hpf</td>
<td>&lt;5</td>
<td>6-10</td>
<td>&lt;5</td>
<td>&gt;5</td>
</tr>
<tr>
<td>Risk</td>
<td>Low</td>
<td>Intermediate</td>
<td>High</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>GIST</th>
<th>Very Low Risk</th>
<th>Low</th>
<th>Intermediate</th>
<th>To coincide with CT or 6 month intervals</th>
<th>CT 6-12 monthly for 5 years</th>
<th>CT CAP imaged</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No follow up needed</td>
<td>To coincide with CT or 6 month intervals</td>
<td>CT 6-12 monthly for 5 years</td>
<td>CT CAP imaged</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Valid on the date of publication
Version 4.0
Procedure on suspected relapse

Patients with suspected relapse of the basis of either history and physical findings or screening investigation should be re-imaged with the definitive imaging method (U/S and MRI or CT for local relapse and CT for systemic relapse). If equivocal this imaging should be re-discussed at the Sarcoma MDT meeting. Confirmatory biopsy may be required.

Follow-up post-relapse

Patients follow-up should be ‘re-zeroed’ at the time of completion of relapse management and they should enter follow-up based upon the disease risk group as defined above.

Follow-up for patients who have exhausted effective anti-cancer treatment

There may be no practical benefit to patients in travelling into the specialist sarcoma clinic if there is no possibility of sarcoma specific intervention. Adequate generic supportive and palliative measures may be delivered at a community and local district hospital level. Routine appointments are not usually necessary provided adequate lines of communication and responsibility have been established closer to the patient’s home. It is recognised that sarcoma is a rare disease and that this may provoke some anxiety in the locally based palliative care teams. It is essential that a central route of access for information provision and discussion remain open and that this is clearly communicated. Scheduling of routine appointments, if still felt to be necessary, should be based upon the patient’s individual needs and anticipated disease trajectory.

It is essential that on discharge from hospital-based WY&HSS care to community-based care (with or without specialist palliative care support) that the following issues are addressed, discussed and documented:

- Anticipated course of disease, including evolving symptoms and role of palliative radiotherapy
- Estimated survival prognosis (accepting difficulties associated with accuracy)
- Clarification of futility of cardiopulmonary resuscitation for progressive malignancy or identification of circumstances when CPR attempts may be considered appropriate
- Preferred place of death
24 Audit

It is essential that the WY&HSS monitors itself with respect to patient process, experience and outcome. It seems very likely that the DoH/CQC will monitor incidence of disease, patterns of care and survival outcomes remotely via Cancer Registry submissions and routine Hospital Episode Statistics (HES) reports. Data quality will be superior for locally held and agreed data. Patient reported experience measures (PREMS) and patient reported outcomes measures (PROMS) cannot be captured centrally and must be collected by WY&HSS for its own patients.

24.1 Patient process

Data regarding cancer waiting times (CWT) tracking is already collected as is sarcoma specific activity data (via LTHT Patient Pathway Manager PPM).

24.2 Patient experience (PREMS)

This data will be collected for all patients. Patient experience (satisfaction) data will be sought directly with biannual WY&H CA Patient Questionnaires re Diagnosis, Treatment and Follow-Up. Based upon the outcomes of these surveys a more specific/targeted tool may be developed.

24.3 Patient Outcomes

This data will be collected for patients with proven STS only (ie. will exclude benign disease). Patient outcomes are objective and subjective. Data on both will be routinely collected, though well validated tools for all subjective measures are not available. The objective endpoint data can and should be maintained via the MDT records/PPM. Subjective data (PROMS) will require regular surveys of patient reported outcomes. A schedule for data collection will be agreed.

<table>
<thead>
<tr>
<th>Objective</th>
<th>Subjective</th>
</tr>
</thead>
<tbody>
<tr>
<td>R0 resection rate</td>
<td>TESS score (change from baseline)</td>
</tr>
<tr>
<td>Post treatment complication rate(*)</td>
<td>EQ-5D score (change from baseline)</td>
</tr>
<tr>
<td>Limb preservation rate</td>
<td></td>
</tr>
<tr>
<td>Local recurrence rate</td>
<td></td>
</tr>
<tr>
<td>Systemic recurrence rate</td>
<td></td>
</tr>
<tr>
<td>Sarcoma-specific non-elective admission rate(**)</td>
<td></td>
</tr>
<tr>
<td>Local and/or systemic progression free survival</td>
<td></td>
</tr>
<tr>
<td>Overall survival</td>
<td></td>
</tr>
</tbody>
</table>
* To include end-points requiring specific intervention (non-exhaustive list): infection, wound breakdown, chronic lymphoedema, chronic pain, bone fracture. Specific criteria relating to qualifying severity are to be agreed. Subjective end-points to be captured via TESS and EQ-5D.

** To include unplanned admissions arising as a consequence of either treatment related complications or treatment failure, but excluding planned admissions for treatment of said complication or failure. For example admissions for planned surgery or chemotherapy will not be included, though admissions for infections, pain control or terminal care will be.

The National Cancer Information Network (NCIN) Sarcoma Site Specific Reference Group (SSRG) is developing comparators against which to assess sarcoma outcomes. In the light of this the WY&HSS should strive to maintain a complete Sarcoma Minimum Dataset (MDS) to judge its own performance internally, to act to validate/quality assure indicators running externally (from the CQC and the NCIN) and to satisfy the requirements of IOG Peer Review.

An audit programme is in development and will feature as part of a WY&HSS annual report.
25 Clinical trials

Participation in clinical trials should be encouraged. The following portfolio is correct at the time of writing:

25.1 Open

EuroEWING
EURAMOS
VORTEX
GeDiSS
STRASS

25.2 In set-up