### Optimal care pathway for people with sarcoma (bone and soft tissue sarcoma)

#### Quick reference guide

Please note that not all people will follow every step of this pathway:

<table>
<thead>
<tr>
<th>Step</th>
<th>Prevention and early detection</th>
<th>Signs and symptoms: The following should be investigated.</th>
<th>Diagnosis and staging: Work-up should include:</th>
<th>Referral: All patients with suspected sarcoma should be referred to a specialist sarcoma multidisciplinary team within two weeks and before biopsy.</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Prevention</td>
<td>• persistent non-mechanical pain in any bone lasting more than a few weeks / referred pain / unremitting pain not responsive to analgesics / nocturnal bone pain</td>
<td>• history and examination</td>
<td>All patients with suspected sarcoma should be referred to a specialist sarcoma multidisciplinary team within two weeks and before biopsy.</td>
</tr>
<tr>
<td></td>
<td>and early detection</td>
<td>• a mass, swelling, a limp</td>
<td>• staging: local – MRI, thallium or PET; systemic – bone scan, PET/CT, CT chest</td>
<td>Communication – lead clinician to:</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• limited mobility or loss of limb function</td>
<td>• image-guided needle core biopsy (NCB) performed after all imaging modalities have been completed and reviewed by the specialist</td>
<td>• explain to the patient/carer who they are being referred to and why</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• fractures with minimal trauma. For soft tissue sarcoma:</td>
<td>• examination of tumour tissue</td>
<td>• support the patient and carer while waiting for specialist appointments.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• any deep mass or superficial mass with a diameter larger than 5 cm</td>
<td>• lymph node biopsy if functional imaging is positive or clinical examination suspicious.</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>All investigations should be completed within two weeks of the first specialist assessment.</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Risk factors for soft tissue sarcoma include:**
- familial syndromes
- history of cancer
- past treatment with radiation therapy
- prolonged lymphoedema
- exposure to certain chemicals (for example, vinyl chloride and dioxin)
- age (over 50 years).

**Prevention:** The causes of sarcoma are not fully understood, and there is currently no clear prevention strategy.

**Risk factors:**
- Risk factors for bone sarcoma include:
  - family history
  - history of retinoblastoma
  - Li-Fraumeni syndrome
  - history of childhood cancer
- prior abnormalities (for example, Paget’s disease, avascular necrosis, polyostotic fibrous dysplasia)
- past treatment with chemotherapy or radiation therapy
- exposure to certain chemicals (for example, vinyl chloride and dioxin)
- age (less than 30 or over 50 years).

**Signs and symptoms:** For bone sarcoma:
- a small but growing mass
- a rapidly growing change in a mass (over months)
- a mass where there is no associated history of trauma.

**General/primary practitioner investigations should include:**
- medical history and baseline blood tests
- physical examination including assessing the physical characteristics of the mass and of the regional lymph nodes
- if there is bone pain, a plain x-ray
- if there is a soft tissue lump, refer to a specialist.

**Communication – lead clinician to:**
- explain to the patient/carer who they are being referred to and why
- support the patient and carer while waiting for specialist appointments.

**Treatment planning:** All newly diagnosed patients should be discussed in a multidisciplinary team meeting before beginning treatment.

**Research and clinical trials:** All patients should be offered the opportunity to participate in a clinical trial or clinical research if appropriate.

---

1 Lead clinician – the clinician who is responsible for managing patient care. The lead clinician may change over time depending on the stage of the care pathway and where care is being provided.
### Step 4
**Treatment:**
- Establish intent of treatment:
  - curative
  - anti-cancer therapy to improve quality of life and/or longevity without expectation of cure
  - symptomatic

**Surgery:** Surgery (resection and reconstruction) is the most common treatment option. Most patients are considered as candidates for limb salvage surgery. Appropriate vascular and plastic surgical reconstructive options should be available.

**Radiation therapy:** All patients with large, localised, soft tissue tumours should be considered for radiation therapy before or after surgery. Radiation should also be considered for smaller tumours (under 5 cm) and lower grade tumours in more difficult anatomic sites. Other than Ewing’s sarcoma, radiation therapy for bone sarcomas is mainly used for palliation.

**Chemotherapy or drug therapy:**
- All patients with osteosarcoma and Ewing’s sarcoma should be considered for protocolised pre- and/or postoperative chemotherapy.
- Other forms of bone sarcomas should be treated as per multidisciplinary team discussion.
- Rhabdomyosarcoma should be treated with protocolised pre- and/or postoperative chemotherapy.

---

### Step 5
**Care after initial treatment and recovery**

**Palliative care:** Early referral can improve quality of life. Referral should be based on need, not prognosis.

<table>
<thead>
<tr>
<th>Communication – lead clinician to:</th>
</tr>
</thead>
<tbody>
<tr>
<td>• discuss treatment options with the patient/carer (including the intent, risks and benefits)</td>
</tr>
<tr>
<td>• discuss advance care planning with the patient/carer where appropriate</td>
</tr>
<tr>
<td>• discuss the treatment plan with the patient’s general practitioner.</td>
</tr>
</tbody>
</table>

**Support:** Assess supportive care needs at every step of the pathway and refer to appropriate health professionals or organisations.

**Fertility preservation options should be discussed where appropriate.**

---

### Step 6
**Managing recurrent, residual and metastatic disease**

**Detection:** Approximately 30–40 per cent of all patients with sarcomas develop distant recurrence.

**Treatment:** Where possible, refer the patient to the original multidisciplinary team. Treatment will depend on location and extent of disease, previous management and patient preference.

**Palliative care:** Early referral can improve quality of life and in some cases survival. Referral should be based on need, not prognosis.

**Communication – lead clinician to:**
- explain the treatment intent, likely outcomes and side effects to the patient/carer
- plan for rapid re-entry to medical services for suspected recurrence.

---

### Step 7
**End-of-life care**

**Palliative care:** Consider referral to palliative care. Ensure that an advance care plan is in place.

**Communication – lead clinician to:**
- be open about the prognosis and discuss palliative care options with the patient/carer
- establish transition plans to ensure the patient’s needs and goals are addressed.

---