Step 1: Prevention and early detection

**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).

Step 2: Presentation, initial investigations and referral

**Signs and symptoms:** The following should be investigated.
- For bone sarcoma:
  - persistent non-mechanical pain in any bone lasting more than a few weeks / referred pain / unremitting pain not responsive to analgesics / nocturnal bone pain
  - a mass, swelling, a limp
  - limited mobility or loss of limb function
  - fractures with minimal trauma.
- For soft tissue sarcoma:
  - any deep mass or superficial mass with a diameter larger than 5 cm
  - 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.

**Referral:** All patients with suspected sarcoma should be referred to a specialist sarcoma multidisciplinary team within two weeks and before biopsy.

**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.

Step 3: Diagnosis, staging and treatment planning

**Diagnosis and staging:** Work-up should include:
- history and examination
- staging: local – MRI, thallium or PET; systemic – bone scan, PET/CT, CT chest
- image-guided needle core biopsy (NCB) performed after all imaging modalities have been completed and reviewed by the specialist
- examination of tumour tissue
- lymph node biopsy if functional imaging is positive or clinical examination suspicious.

**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.

**Communication – lead clinician to:**
- discuss a timeframe for diagnosis and treatment with the patient/carer
- explain the role of the multidisciplinary team in treatment planning and ongoing care
- provide appropriate information or refer to support services as required.

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

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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
- symptom palliation.

**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.

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

**Communication – lead clinician to:**
- discuss treatment options with the patient/carer (including the intent, risks and benefits)
- discuss advance care planning with the patient/carer where appropriate
- discuss the treatment plan with the patient’s general practitioner.


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

- Consider referral to an occupational therapist, orthotist/prosthetist and a physiotherapist or exercise physiologist.
- Healing of underlying structures, infection and other complication risks relating to skeletal implants may require input from wound nurse specialists and infection control specialists.
- Patients who have had a limb amputated require rapid access to prosthetic services.

Evaluation for postoperative rehabilitation is recommended for all patients with extremity sarcoma. Rehabilitation should be continued until maximum function is achieved and highly integrated with the treating medical team.

Provide patients with the following:

**Treatment summary** (provide a copy to the patient/carer and general practitioner) outlining:
- diagnostic tests performed and results
- tumour characteristics
- type and date of treatments(s)
- interventions and treatment plans from other health professionals
- supportive care services provided
- contact information for key care providers.

**Follow-up care plan** (provide a copy to the patient/carer and general practitioner) outlining:
- medical follow-up (tests, ongoing surveillance)
- care plans for managing late effects
- a process for rapid re-entry to medical services for suspected recurrence.

**Communication – lead clinician to:**
- explain the treatment summary and follow-up care plan to the patient/carer
- inform the patient/carer about secondary prevention and healthy living
- discuss the follow-up care plan with the GP.

**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.

**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.