Bone and soft tissue sarcomas are rare mesenchymal malignancies that arise in two to four per 100,000 head of population.\(^1,2\) The limbs are the commonest sites for sarcoma, with over 50% of soft tissue and bone sarcomas occurring in the lower limb. The advent of multimodality treatment with advances in chemotherapy, radiotherapy and surgery, all supported by more sophisticated diagnostic and imaging techniques, has led to considerable improvement in long-term survival. Overall survival following treatment of primary sarcoma now approaches 75% at five years, and surgery remains the mainstay of treatment.\(^1,2\) Surgery to resect the tumour followed by reconstructions to preserve function, mobility and aesthetics (limb sparing surgery) has now replaced amputation as the primary form of surgical intervention.\(^3,5\)

### Criteria

Limb sparing surgery may be considered when specific criteria are met, including:

- Tumour resection occurs with oncologically sound margins
- Reconstruction leads to a functional limb
- All soft tissue defects can be closed primarily or with soft tissue transfers.

Previously a sensate lower limb was thought to be mandatory for limb sparing surgery. Sacrifice of the sciatic nerve traditionally led to lower limb amputation for fear of developing chronic non-healing trophic ulceration of the foot. However, with better awareness of foot hygiene and shoe wear, limb sparing surgery is now practised despite the need to include the sciatic nerve in resections of proximal thigh or pelvic tumours.\(^6,7\)

### Indications

Limb sparing surgery is indicated when:

- Tumour is resectable with oncologically sound margins
- Survival is long enough to justify complex surgery
- The patient refuses amputation and accepts the risk of local recurrence of disease from inadequate margins
- Palliating patients with limb disease that is easily and safely operated on to improve quality of life eg. impending fractures, fungation.

### Contraindications

Limb sparing surgery is contraindicated when:

- Surgical margins are expected to be inadequate for managing the primary tumour
- Survival is not expected to exceed three months
- There is gross contamination of the adjacent soft tissue compartments with tumour through poorly performed biopsy, pathologic fracture or inadvertent surgery with positive margins
- Local or systemic sepsis is a concurrent problem, or patient co-morbidities do not permit safe anaesthesia or surgery.

### Diagnosis and staging

Accurate diagnosis is critical to successful treatment. Appropriate choices of chemotherapy agents or radiotherapy depend on correct identification of the type of sarcoma.\(^8-13\) For example, chemotherapy differs between osteosarcoma, Ewing sarcoma and myxoid liposarcoma. Soft tissue sarcomas comprise a heterogeneous group and consensus on grade, type and subtype of sarcoma can be difficult to achieve.

Biopsy is fundamental to obtaining an accurate histological diagnosis. In principle, the same group that will be undertaking definitive treatment should perform biopsy of primary bone and soft tissue sarcomas. This is because the placement of the biopsy site and the avoidance of post-biopsy complications, such as haematoma or...
infection, may influence the potential for undertaking limb sparing surgery. Biopsy, whether open or closed, should always be done in the line of the operative incision to allow inclusion of the biopsy site in the definitive resection. It is our preference to perform CT-guided core needle biopsies.14

Local staging of the tumour is important for planning surgery. The tumour size, site, shape, consistency, edge, capsule and adjacent structures are important information for planning the surgical margins and reconstructions after assessing response to neoadjuvant therapies. Imaging of the limb should include plain radiographs, CT, MRI, PET or thallium scans.15, 16 In addition, CT scans of the chest are mandatory for assessing systemic spread, because pulmonary involvement is the commonest site for first metastases. Evidence of metastasis is likely to affect the nature of care and therefore all efforts to diagnose metastases should be undertaken.

**Adjuvant therapy**

Pre-operative (neoadjuvant) chemotherapy or radiotherapy is fundamental to managing bone and soft tissue sarcomas, respectively. The benefits of adjuvant therapy include:

- inducing local tumour necrosis
- reducing tumour size
- formation of a peritumoral “rind” of fibrous capsule.

These effects may aid the planning of surgical margins, improve the resectability of tumours and allow greater safety when dissecting close to vital neurovascular structures.

The response to neoadjuvant therapy may be critical for determining if limb sparing surgery or amputation should be performed. For example, there is a correlation between local recurrence and response to chemotherapy in osteosarcoma.17 Moreover, the risk for local recurrence rises substantially when margins of resection diminish.17 Therefore, if pre-operative imaging demonstrates that the response to neoadjuvant chemotherapy is poor and if planned surgical margins are expected to be close, then to avoid locally recurrent disease, amputation may be preferable over limb sparing surgery. This information is valuable in the pre-operative counselling of patients and for the obtaining of informed consent for surgery.

**Surgical margins**

Adequacy of surgical margins correlates directly with the incidence of local recurrence and relates to the quality and quantity of tissue around the tumour that is included in the resected specimen.18-20 The definition of surgical margins are as follows:

1. Intralesional margins are those where the resection enters the tumour.
2. Marginal margins are those where the surgery passes through the reactive zone of inflammation that surrounds the pseudocapsule of a tumour. “Shelling out” of a tumour is said to be marginal surgery.

3. Wide margins are those where the resected specimen includes at least two centimetres of normal tissue in the longitudinal plane and one named normal anatomic boundary in the radial plane. A named fascia, or muscle layer would represent an anatomic boundary.

4. Radical margins are those where the resected specimen includes the entire tumour bearing compartment. For example, resection of the entire quadriceps musculature from origin to insertion, and from lateral to medial intermuscular septae, may be regarded as a compartectomy because the quadriceps musculature is the sole content of the anterior compartment of the thigh.

The quality of the anatomic boundary is also relevant when determining the adequacy of the margin. The fascia lata is a very tough tissue, although it may be only a few millimetres thick. If the fascia lata is included as an uninvolved boundary, then the resection may be regarded as wide.

Intralesional and marginal margins are regarded as being inadequate surgical margins, while wide and radical margins are regarded as adequate surgical margins in the management of sarcoma. Marginal margins may be equivalent to wide margins alone when combined with radiotherapy or chemotherapy. Marginal margins are usually avoided, however may be important when having to preserve important neurovascular structures.

**Reconstructive options**

A wide variety of reconstructions are available for limb sparing surgery. These include:

- prosthetic reconstructions
- biologic reconstructions
- combination of biologic and prosthetic reconstructions.

Reconstructions may also be mobile or rigid. These refer to the preservation or fusion of a previously mobile joint at reconstruction.

Tumour prostheses take their origins from the evolution of standard joint prostheses. Advances in metallurgy, tribology and prosthetic fixation have allowed the development of modular implants that can be individualised to each patient, while exhibiting strength and durability.21-23 Prostheses for the hip and knee were the first to be developed and today, prostheses are also available for the shoulder joint, scapula, elbow, total humerus, ulnar, total femur, pelvis and the ankle.

Improvements in computer aided design of prostheses and manufacturing techniques now allow the custom creation of unique prostheses to accurately match the defect created by tumour resection. Such customised machining of prostheses is matched with computer guided surgery to ensure that the exact resection shape is created during surgery, to allow accurate matching of resection defect with the customised implant. These techniques require rigorous planning and dialogue between manufacturer, surgeon and imaging specialists.
Biologic reconstructions

One of the earliest attempts at limb sparing surgery was the arthrodesis of the knee after resection of tumours of the distal femur. In an attempt to span the gap between femur and tibia created by distal femoral resection, a segment of the ipsilateral tibia would be elevated and used to span the tibio-femoral gap in an arthrodesis procedure. Held by a long arthrodesis nail, the construct would permit full weight bearing after the bone graft united with the remnant femur and tibia.

The popularity of bone banks soon permitted harvest and storage of large structural allografts, which were then employed in place of autograft bone to fill the defect of tumour resection.\(^24\) Allograft bone had a number of advantages including:

- reducing donor site morbidity
- ready availability
- unlimited supply.

The disadvantages of allograft bone included:

- potential for disease transmission
- graft disintegration
- infection
- non-union.

The availability of modern internal fixation devices has helped to support the allograft constructs and long-term results have been acceptable.

The fibula has been a versatile resource for reconstructing defects of up to 22 centimetres. The fibula may be used as a vascularised or non-vascularised graft and has been utilised in a number of innovative ways,\(^{25, 26}\) including spanning defects, creating articulations, arthrodesis of joints and in combination with allografts.

Other innovative methods of biologic reconstruction include the role of extracorporeal radiotherapy to sterilise the tumour bearing bone immediately after resection, and then to reimplant the resected bone back into the resection site.\(^{27}\) This technique utilises established radiotherapy techniques to deliver up to 10 times the normal radiotherapy dose to tumour bearing bone in a single fraction. By performing this in an extracorporeal fashion, the anticipated toxicities of such high doses can be avoided. Moreover, the technique has the advantage of reconstructing the defect with a perfectly sized matched construct. To date, reported series have not encountered recurrent tumour following reconstruction using this technique.

Allograft prosthetic composites

Allograft prosthetic composites (APC) capitalise on the advantage of allograft bone to rebuild bone stock to the post-operative defect, while permitting the predictability of prosthetic joint replacement to regain stable joint motion. Large defects created by the resection of a joint and the adjacent diaphysis and metaphysis can be reconstructed by the use of allograft bone that includes a metaphyseal-diaphyseal segment, on to which a standard joint prosthesis may be cemented. The most common sites where APC are used include the hip (proximal femur), knee (distal femur or proximal tibia) and the shoulder (proximal humerus). Residual soft tissue attachments on the allograft allow host to allograft tendon and ligamentous reconstructions, which improve the stability and function of the reconstructed joint.

Complications specific to limb sparing surgery

Complications following limb sparing surgery may be devastating, with the potential for loss of the limb or unplanned cessation of chemotherapy.

1. Limb sparing surgery typically entails prolonged surgical time with exposure of the operative field to the external environment. The risk of infection is directly correlated with the duration of surgery, and the lowered resistance of the patient through chemotherapy and that of the tissue through radiotherapy further compound this. Infection has been reported to be as high as 30% in some series. The addition of prosthetic material also raises the risk of local infection, because foreign bodies can act as nidus for infection. Infection not only can lead to prolonged delays in wound healing, but may also delay the recommencement of chemotherapy. The use of antibiotics during and after surgery, regular irrigation of the operative field with sterile fluid and antiseptic, careful handling of tissue, minimising the creation of dead spaces and observance of sterile technique, lower the risk of infection.

2. Unplanned neurovascular injury usually results from dissection around large tumours or in confined spaces where there is a confluence of vital structures eg. popoliteal fossa. The need to create an oncologic margin may bring the dissection close to nerves and vessels, which lie adjacent to the tumour. Careful dissection and retraction can help to minimise injury. Patients undergoing chemotherapy may be more susceptible to neuropaxia during surgery because of the “priming” of nerves by the toxicity of chemotherapeutic agents. The peroneal nerve is not infrequently a victim of neuropaxia.

3. Devascularisation of soft tissue flaps is a constant threat in large and complex dissections. This may lead to skin necrosis and dehiscence of the wound, which in turn is an important antecedent factor behind infection. Wound healing problems are most common after preoperative radiotherapy, however few require reoporation for resolution. Careful dissection and preservation of muscle vascularity, avoiding narrow soft tissue flaps and skin islands, minimising tension across wounds during closure, judicious use of soft tissue transfers to obliterate dead spaces after surgery and using drains to avoid deep haematomas are ways of protecting soft tissue from necrosis.
4. Dislocations on the hip and shoulder are a risk after limb sparing surgery because of the need to remove important and significant quantities of soft tissue structures that may be critical for maintaining joint stability. For example, resection of the joint capsule and abductors of the hip predispose that joint to dislocation. Excision of the rotator cuff musculature during proximal humeral resection may predispose the humeral prosthetic reconstruction to instability. Careful reconstitution of restraining forces by soft tissue transfers or plication, and addition of pliable synthetic material around joints, may help to reduce the incidence of joint dislocation.

5. Fractures may occur after limb sparing surgery because of the potential to devascularise bone from radiotherapy, extensive ligamentous or muscular detachment or subperiosteal dissections. Almost one fifth of long bones which have undergone circumferential subperiosteal dissection of tumour after radiotherapy fracture, with the majority of these occurring within two years of the index surgery. Prophylactic internal fixations with intramedullary rods are indicated where high risk of fracture may be anticipated.

Salvaging limb sparing surgery after a complication is a complex task, but may be undertaken in certain circumstances. Careful planning and a multidisciplinary approach is required. Innovative techniques are available that may result in a functional limb.26, 29

Conclusion

Limb sparing surgery is the technique of choice for surgical management of limb sarcomas. In comparison to amputation, limb sparing surgery:

- has the same overall survival rate
- has higher patient satisfaction
- has a lower energy expenditure for walking
- has a lower cost to the community.

Limb sparing surgery is a complex procedure that requires expert knowledge of the requirements and criteria for its use. It is an important part of multidisciplinary management of sarcoma. The success of such surgery is maximised when conducted in centres with specific interest and expertise in this field.

References