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Limb Salvage and Reconstruction Options in Osteosarcoma

Samuel Z. Grinberg, Abigail Posta, Kristy L. Weber, and Robert J. Wilson

Abstract

Advances in chemotherapy, sophisticated imaging, and surgical techniques over the last few decades have allowed limb-salvage surgery (LSS) to become the preferred surgical treatment for bone sarcomas of the extremities. The goal of LLS is to maximize limb functionality to allow for the maintenance of quality of life without compromising overall survival and tumor local recurrence rates. Today, limb-salvage procedures are performed on 80–95% of patients with extremity osteosarcoma, and the 5-year survival rate in extremity osteosarcoma patients is now 60–75%.

This chapter will focus on LSS for extremity osteosarcoma. Common types of surgical reconstruction techniques including endoprostheses, intercalary or osteoarticular

Medical Student, University of Pennsylvania, Perelman School of Medicine, Philadelphia, PA, USA e-mail: Samuel.Grinberg@Pennmedicine.upenn.edu

A. Posta

Undergraduate Student, Swarthmore College, Swarthmore, PA, USA e-mail: apostal@swarthmore.edu

K. L. Weber · R. J. Wilson (⊠) Penn Medicine Department of Orthopaedic Surgery, Philadelphia, PA, USA e-mail: Kristy.Weber@uphs.upenn.edu; Robert. wilson3@uphs.upenn.edu allografts, vascularized fibular autografts, and allograft prosthetic composites (APC), and their complications such as infection, local recurrence, graft fracture, implant failure, and nonunion will be discussed in detail. Anatomic locations of lesions discussed include the proximal femur, distal femur, proximal tibia, distal tibia, proximal humerus, distal humerus, and forearm bones.

Keywords

Limb salvage surgery · Osteosarcoma · Allograft · Endoprosthesis · Allograft prosthesis composite · Infection · Loosening · Wear · Femur · Local recurrence · Nonunion

Introduction

Advances in chemotherapy, imaging, and surgical techniques over several decades have allowed limb-salvage surgery (LSS) to become the preferred surgical treatment for bone sarcomas of the extremities [1, 2]. The goal of LSS is to maximize limb functionality to allow for the maintenance of quality of life without compromising overall survival and tumor local recurrence rates. Today, limb-salvage procedures are performed on 80–95% of patients with extremity osteosarcoma [1, 3]. Data suggest local

S. Z. Grinberg

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recurrence rates and overall survival are equivalent when comparing LSS to amputation, and LSS may have better function [4]. The five-year survival rate in extremity osteosarcoma patients is now 60–75%.

This chapter will focus on LSS for extremity osteosarcoma. As it pertains to this chapter, limb salvage is defined as the "successful resection of a tumor and reconstruction of a viable, functional extremity" [5]. Common types of surgical reconstruction techniques including endoprostheses, intercalary or osteoarticular allografts, vascularized fibular autografts and allograft prosthetic composites (APC), and their complications such as infection, local recurrence, graft fracture, implant failure, and nonunion will be discussed in detail [6]. Anatomic locations of lesions discussed include the proximal femur, distal femur, proximal tibia, distal tibia, proximal humerus, distal humerus, and forearm bones.

Types of Reconstruction

This review will focus on common types of reconstruction. Materials used include composite metals, cadaveric allograft, or biologic options. An endoprosthesis is a metal implant used to replace resected bone and joints that is secured to the remaining bone with a cemented or press-fit stem. Alternatively, an osteoarticular or intercalary allograft can be used to reconstruct the limb with a matched bone from a cadaver, which is commonly attached to the remaining bone with an intramedullary nail or a plate/screw construct [7]. An intercalary allograft can be used with or without a vascularized fibular graft to replace resected tumors in the diaphysis while sparing the joints. Osteoarticular allografts are an option when joint preservation is not possible; however, they are not used as commonly as endoprostheses in the United States. The remaining soft-tissue connections on the allograft allow for some functional advantages, especially when reconstructing the extensor mechanism for proximal tibia tumors [3]. However, there is often instability at the joint with increased risk for cartilaginous wear, and there are fewer size-appropriate allograft bones for pediatric patients compared to adult patients [3].

Finally, an APC is also a valid reconstruction option. An APC combines a cadaveric allograft with a hinged prosthesis to replace the resected bone and joint.

Each reconstructive method has advantages and disadvantages after a tumor resection. Endoprostheses with cemented stems often allow for weight-bearing immediately following surgery; however, there is a risk for long-term device loosening and wear [7]. A key advantage of many allografts is that tendons and ligaments remain attached to the graft bone for host soft tissue attachment. Disadvantages of osteoarticular allografts are allograft fracture risk, nonunion, joint instability, and osteoarthritis of the reconstructed joint [7]. Intercalary allografts share the nonjoint-related concerns. Lastly, APCs have combined advantages of endoprostheses and allografts. There is avoidance of the osteoarticular allograft joint and instability problems, restoration of bone stock, and tendon to tendon reconstruction of the soft tissues (rather than tendon to prosthesis). However, the risk for allograft fracture nonunion at the host graft junction remains [7].

The decision as to which type of reconstruction to use depends on multiple factors. The anatomic location of the osteosarcoma, age of the patient, and what specific type of reconstruction are the most effective issues to consider. Furthermore, with primarily retrospective clinical data and a lack of consensus among surgeons, the type of reconstruction is also based on surgeon preference, experience, and patient- and tumor-specific factors. These factors will be further explored.

Endoprosthetic Failure Classification

In 2011, Henderson et al. [8] published a literature review of failure mechanisms for endoprostheses used in tumor surgery. They also provided a classification of different failure modes. Failures were classified as: Type 1, soft-tissue failure; Type 2, aseptic loosening; Type 3, structural failure; Type 4, infection; and Type 5, tumor progression. They reported 534 failures following primary reconstructions in 2174 patients (24.5%). Of these failures, 12% were Type 1, 19% were Type 2, 17% were Type 3, 34% were Type 4, and 17% were Type 5. Throughout this chapter, the failure results from multiple studies of endoprostheses will be reported according to this classification system.

Anatomic Locations

The anatomic location of an osteosarcoma is a crucial factor in determining the feasibility and success of limb salvage surgery as well as the type of reconstruction. Along with the specific location, it is important to consider the advantages and disadvantages of each type of reconstruction as well as patient age, anticipated function, activity levels, and projected overall survival.

Proximal Femur

Reconstruction of the proximal femur is most commonly performed using an endoprosthesis or APC. Compared to the pelvis, functional outcomes tend to be better and patients frequently resume a higher level of function after reconstruction. However, functional outcomes generally are not as good as they are with distal femoral reconstruction.

In a literature review comparing endoprosthesis to APC in reconstructions of the proximal femur, Janssen et al. [9] found similar functional outcomes for both, which were described as reasonable to good, although they noted high revision surgery rates for both groups. The APC group experienced higher rates of Type 3 and Type 4 failure. Biau et al. [10] studied 32 patients who underwent reconstruction with APC and noted that, when compared to historical controls, there was no improvement over megaprostheses. Without successful union of the host bone/graft junction, the theoretical mechanical benefits of APC (improved abductor strength) are not realized when compared to an endoprosthesis.

Focusing on functional results following endoprosthetic reconstruction of the proximal femur, Hobusch et al. [11] looked at activity level and participation in sports after surgery. Of the 16 patients included, 14 participated in sports for an average of 5 hours/week before surgery. After surgery, 11 of these patients participated for an average of 2 hours/week. Additionally, there was a significant decrease in the UCLA and modified Weighted Activity Score levels from preoperative levels. While some patients were involved in higher impact sports preoperatively, following surgery patients engaged in lower impact sports such as hiking, biking, swimming, and golf.

The literature thus far appears to favor endoprosthetic reconstruction over APC for the proximal femur as APC reconstruction had higher levels of complications without offering improved functionality. Infection, aseptic loosening, and prosthetic dislocation are the primary complications encountered in this location.

Femoral Diaphysis

For osteosarcoma of the femoral diaphysis, the most common option is an intercalary allograft which maintains the native hip joint above and the native knee joint below. Aponte-Tinao et al. [12] performed 83 femoral reconstructions with intercalary allograft. The overall allograft survival rate was 85% and 76% at 5 and 10 years, respectively. Of the 83 patients, 38 experienced complications that required a follow-up surgery, and the allograft was removed in 15 of these patients. Complications included 1 infection, 14 fractures, and 20 nonunions. Of the 20 patients with nonunions, 3 received adjuvant radiation and 15 received preoperative chemotherapy. The average Musculoskeletal Tumor Society (MSTS) score was 27 of 30 for the 68 patients who retained their allograft. Ogura et al. [13] used free vascularized fibula autografts in addition to intercalary autografts in 11 patients. The mean MSTS score was 81%, and there were four complications in three patients. Complications included

two infections, one implant failure and one fracture. The graft was removed in the two patients with infections. Bone union occurred in 10 of the 11 patients.

For rare osteosarcomas that involve the majority of the femur, it may be necessary to use a total femoral replacement. Sevelda et al. [14] reviewed the results of 44 patients treated with a total femoral replacement of which 10 received an expandable prosthesis. They found overall implant survival rates of 97% for conventional prosthesis and 100% for the expandable prosthesis. There were 25 complications among the group receiving conventional implants, most commonly Type 1 and Type 4 failures. Unplanned revision rates were 50% for the conventional implant and 90% for the expandable. Overall, MSTS scores were 70% for the conventional group and 88% for the expandable group.

For osteosarcomas of the femoral diaphysis, the primary treatment is reconstruction with intercalary allograft. One important complication is nonunion at the host bone-allograft junction, and there is an increased risk for nonunion with chemotherapy and radiation. A free vascularized fibula graft can be used in conjunction with an intercalary graft to improve bone union. In rare circumstances, a total femoral replacement can be performed when reconstruction with an intercalary allograft is not possible.

Distal Femur

The distal femur is the most common location for osteosarcoma. Endoprosthesis and APC are primarily used to reconstruct the distal femur and knee. Simon et al. [15] compared amputation to limb salvage treatment and saw similar rates of overall survival and disease-free survival for patients receiving limb salvage, above the knee amputation, and hip disarticulation. Of note, endoprosthesis, APC, and osteoarticular allograft reconstruction were pooled together in the limb salvage group. In a follow-up, they found significantly improved functionality with limb salvage when compared to the two amputation groups [4].

While endoprostheses are most commonly used, results of osteoarticular allografts and APCs have still been described in the literature. Puerta-GarciaSandoval et al. [16] compared APC reconstruction in the distal femur or the proximal tibia. For the distal femur group, they saw no fractures, complete bone healing in 79% of patients, a mean MSTS score of approximately 79%, and prosthesis survival of 94% at 10 years with few complications. In 32 patients receiving an APC for tumors of the distal femur, Wang et al. [17] reported a mean MSTS score of 94% after an average follow-up of 54 months. Two patients had nonunion that healed following refixation. Wunder et al. [18] compared the results from 11 patients treated with allograft reconstruction and 64 patients treated with prosthetic reconstruction. Allografts failed 55% of the time, while prostheses failed 16% of the time. Additionally, allografts were successful in saving the limb 64% of the time compared to 95% for prosthesis. Prostheses also had better MSTS scores, 75% compared to 57%. In a retrospective review of 83 patients receiving massive distal femoral osteoarticular allografts, Mnaymneh et al. [19] saw poor functional results in 5 patients and excellent or good results 53 patients. However, complication rates were 36% and included nonunion, allograft fracture, infection, knee instability, and arthritis of the knee.

Recently, most of the literature focuses on reconstructions using endoprostheses. Options for distal femur endoprosthesis include either cemented or uncemented implants as well as fixed hinge or rotating hinge mechanisms for reconstruction of the knee. Pala et al. [20] reviewed the results of 247 rotating-hinge modular endoprostheses for distal femoral and proximal tibial reconstruction with a minimum follow-up of 2 years. Of the 247 implants, 175 were used for the primary procedures and 72 were used to revise a previously failed reconstruction. For younger patients with primary bone cancer, implants were frequently uncemented. One hundred and eighty-seven replacements were used in the distal femur. Functionally, the mean MSTS score was approximately 85% with distal femur reconstruction. The total failure rate in the distal femur group was 27%. Out of 187 distal femur implants, 7% experience Type 1 failure, 5% experienced Type 2 failure, 9% experienced Type 4 failure, and 6% experienced Type 5 failure. Of note, there were no structural failures. Overall implant survival was 60% at 8 years. Haijie et al. [21] performed a systematic review exploring implant survival and complications of endoprostheses used for distal femoral and proximal tibial replacement. For distal femoral replacements, mean implant survival rates at 5, 10, 15 and 20 years were 78%, 70%, 62% and 38%, respectively. Aseptic loosening (Type 2 failure) and infection (Type 4 failure) were the most frequent complications occurring 9% of the time each.

Based on the literature, endoprosthetic reconstruction is currently the most commonly used technique compared to APC and osteoarticular allograft. Aseptic loosening and infection continue to be the most common causes of complications with endoprosthetic reconstruction.

Case Example: Distal Femur

A 52-year-old female with Paget sarcoma of right distal femoral diaphysis treated with neoadjuvant chemotherapy followed by wide resection and cemented megaprosthesis reconstruction followed by adjuvant chemotherapy (Image 2.1).

Proximal Tibia

The proximal tibia is the second most common anatomic location for osteosarcoma after the distal femur. While reconstruction of the proximal tibia is similar anatomically to the distal femur, it tends to have higher complication rates and lower functional outcomes compared to other anatomical sites [22]. Particular challenges include limited soft tissue coverage, vascular abnormalities, and difficulty restoring the extensor mechanism. As a result, endoprosthetic survival rates are shortest while amputation rates and revision rates are highest for proximal tibia reconstruction when compared to other anatomic sites [7, 23].

Homlar et al. [7] performed an in-depth systematic review of the literature to compare postoperative complications, functional outcomes, success of limb salvage, and implant survival between endoprostheses, APCs, and osteoarticular allografts as reconstruction option in the proximal tibia. All included studies had at least 10 patients. The mean pooled MSTS score was 76% for the endoprosthesis group, 90% for the osteoarticular allograft group, and 77% for the APC group. Based on their results, each type of reconstruction had advantages and disadvantages. Endoprostheses had lower infection rates than osteoarticular allografts. Endoprostheses also had the highest rates of amputation. Osteoarticular allografts had a lower extensor mechanism failure rate than the other two reconstruction types. Local recurrence was similar among the three groups, and allograft fracture was significantly more common with osteoarticular allograft compared to APCs.

Puchner et al. [24] reviewed the results from 81 patients who underwent proximal tibia reconstruction with endoprostheses. The overall complication rate was 56%. Out of the total number of patients, 10% experienced Type 1 failure, 12% experienced Type 2 failure, 15% experienced Type 3 failure, 12% experienced Type 4 failure as their primary complication. The mean MSTS score was 83% and was not statistically different based on complication, fixed or rotating hinge prostheses, and extensor mechanism reconstructions.

Albergo et al. [25] compared the results of 88 patients who underwent reconstruction with an endoprosthesis and 44 patients who underwent reconstruction with an osteoarticular allograft. They found no difference in the probability of failure at 5 years (18% for endoprosthesis; 27% for osteoarticular allograft) and 10 years (44% for endoprosthesis; 32% for osteoarticular allograft). While there was no difference in MSTS scores between the groups, allograft reconstruction resulted in an improved range of motion and less extension lag than endoprosthetic reconstruction (13.56° for endoprosthesis; 2.41° for osteoarticular allograft). While osteoarticular allograft). While osteoarticular allograft). While osteoarticular allograft networks and less extension lag than endoprosthetic reconstruction (13.56° for endoprosthesis; 2.41° for osteoarticular allograft). While osteoarticular allograft networks reconstruction (13.56° for endoprosthesis; 2.41° for osteoarticular allograft).

Image 2.1 AP and lateral radiographs reveal an osteolytic lesion on the right distal femur, which was diagnosed as Paget's Sarcoma (**a**, **b**). Postoperatively, AP and lateral radiographs show the reconstruction with a wide megaprosthesis following resection (**c**, **d**)



motion, there are significant technical difficulties in successfully reconstructing the joint allografts.

Müller et al. [26] compared APC to megaprosthesis for reconstruction of the proximal tibia. Of the 42 patients, 23 received a megaprosthesis and 19 patients received an APC. At an average follow-up of 62 months, five megaprosthesis patients and four APC patients experienced reconstruction failure. Ten-year implant survival rates were 79% and 94% for megaprosthesis and APC, respectively. Neither failure rate nor implant survival was significantly different between the two groups, and there were no functional differences between the groups. While the difference was not statistically significant, the APC group on average had less extensor lag (7.2°) than the megaprosthesis group (11.4°) . Furthermore, two patients in the megaprosthesis group experienced extension lag of greater than 30°, whereas no patients in the APC group did. This led them to conclude that without other risk factors, APC can provide a better functional outcome.

The use of endoprostheses, osteoarticular allografts, and APCs is all supported in the literature. APC and osteoarticular allografts may provide better long-term functional outcomes through reconstruction of the extensor mechanism. However, allograft use is associated with an increased risk of fracture and infection.

Case Example: Proximal Tibia

A 16-year-old male with proximal tibia osteosarcoma treated with neoadjuvant chemotherapy followed by wide resection and megaprosthesis reconstruction followed by adjuvant chemotherapy (Image 2.2).

Distal Tibia

Osteosarcomas of the distal tibia are rare and typically have a better prognosis compared to osteosarcomas in more proximal anatomic locations [27]. They are commonly treated with below-knee amputation, limiting the clinical data of limb-sparing procedures. Like the other anatomic sites, reconstruction techniques using endoprostheses, allografts, and APCs as well as ankle arthrodeses have allowed for limb salvage. Furthermore, depending on the type of reconstruction, limb preservation can result in improved functionality compared to amputation [28]. Zhao et al. [29] saw similar MSTS scores between autograft reconstruction and below-knee amputation, which were both superior to allograft reconstruction. Autograft reconstruction was performed with nonvascularized fibular grafts, pasteurized autograft, or a combination of the two. Both types of reconstruction had more complications than amputation. In another study, Zhao et al. [28] performed a literature review comparing endoprostheses to biological reconstruction with either allograft or autograft and found that autograft performed better than allograft functionally, and both performed better than endoallografts, prostheses. Intercalary fibular autografts, and treated resected autografts were used for arthrodesis. Osteoarticular allografts were used to reconstruct the ankle. A major limitation of the study is the lack of stratification between types of reconstructions with different allografts and autografts. Each type of reconstruction had advantages and disadvantages in congruence with those previously mentioned.

Kundu et al. [30] performed ankle arthrodeses in patients using the centralization of a free fibular graft alone after resecting distal tibia tumors, resulting in a mean MSTS score of approximately 76%. While the procedure resulted in a loss of ankle mobility and varying amounts of leg length discrepancies, these did not cause significant disability for the patients. Given the limited data, the choice of a reconstruction procedure versus a



Image 2.2 AP and lateral radiographs reveal osteolytic and osteoblastic lesion in the right proximal tibia (a, b). Postoperative AP and lateral radiographs show the resection and reconstruction with a megaprosthesis (c, d)

below-knee amputation needs to be made based on the surgeon's experience and the functional needs of the patient without compromising a margin-negative resection of the tumor.

According to the literature, ankle fusions and reconstruction with either autograft or allograft are the primary methods for salvaging the distal tibia. However, below-knee amputation continues to be the primary method for treating tumors of the distal tibia and can often provide comparable functional outcomes with typically fewer complications.

Proximal Humerus

Successfully reconstructing the upper limb is important in maintaining a patient's function. Whereas a prosthetic for a lower limb amputation can allow for ambulation, upper extremity prosthetics are less able to restore normal or nearnormal function. Choosing how to reconstruct the shoulder depends on the margins of the resection as well as the soft tissue structures that are preserved during surgery [31]. Ideally, shoulder, elbow, and hand functionality should be maintained with limb salvage surgery of the proximal humerus.

Historically, preserving shoulder function has been difficult. De Wilde et al. [32] showed that utilizing a reverse total shoulder prosthesis after tumor resection allowed for glenohumeral function with the deltoid compensating for the absence of the rotator cuff. In another study, they found that functionality was maintained after a mean follow-up of 7.7 years utilizing a reverse shoulder prosthesis with irradiation of the resected humerus before being used as an autograft [33]. Reverse shoulder arthroplasty is indicated when the deltoid, axillary nerve, and enough of the glenoid are spared and when resection of the rotator cuff is required [31, 34].

When the rotator cuff is also spared, it is possible to use an endoprostheses or APC [31]. In a systematic review, Teunis et al. found no difference in outcomes between endoprostheses and APC; however, both had worse outcomes than reverse shoulder arthroplasy [31, 35]. While osteoarticular allografts have been used to treat osteosarcomas of the proximal humerus, they have high failure rates and some discourage their use given the advances in endoprostheses [36]. One exception is skeletally immature patients, where there have been high rates of complications with expandable prostheses [37]. van de Sande et al. [38] retrospectively reviewed proximal humeral endoprostheses, APCs, and osteoardetermined ticular allografts. They that endoprosthetic reconstruction had better implant survival, fewer complications, and comparable functional outcomes to APC.

The literature supports the use of endoprostheses as the most common reconstruction of the proximal humerus. Depending on whether the rotator cuff is spared, either a reverse prosthetic total shoulder prosthesis or standard endoprosthesis can be used. Resection and reconstruction decrease shoulder stability, and painless endoprosthetic subluxation is common.

Case Example: Proximal Humerus

A 17-year-old female with osteosarcoma of the right proximal humerus treated with resection and reconstruction with an APC (Image 2.3).

Distal Humerus

Tumors of the distal humerus are rare and account for only 1% of primary bone tumors [39]. Similar to the tumors of the proximal tibia, reconstruction of the distal humerus presents a unique challenge. The successful reconstruction of the elbow is important for a well-functioning upper extremity. Poor soft tissue coverage and the proximity of the neurovascular bundle to the elbow joint makes reconstruction technically difficult [40]. Due to the small number of primary bone tumors in the distal humerus, studies often pool reconstruction patients presenting with either primary bone tumors or metastatic disease. In the literature, reconstruction techniques tend to be limited to either endoprosthetic reconstruction of the elbow or reconstruction with APC. Large defects



Image 2.3 An AP radiograph of the right proximal humerus in a 17-year-old girl reveals an osteoblastic lesion (**a**). T1-weighted coronal (**b**) and T2-weighted axial MR images (\mathbf{c} - \mathbf{e}) show a large circumferential soft tissue mass that extends into the glenohumeral joint (\mathbf{b} - \mathbf{e}). AP

and lateral right humerus radiographs show the results 1 year after extra-articular resection of the osteosarcoma and reconstruction with an allograft-prosthetic composite and distal plate fixation (\mathbf{f} , \mathbf{g})

and tumors extending to the proximal humerus may require total humeral replacement as a method of reconstruction.

Weber et al. [41] reviewed the results from 23 patients who underwent complex elbow reconstructions following tumor resection. Of the 23 patients, 18 patients had tumors in the distal humerus or humeral diaphysis. They also included patients with soft tissue tumors and multiple myeloma affecting the elbow. The types of reconstruction included total humeral replacement (12 patients), prosthesis (seven patients), allograft (five patients), and segmental elbow replacement (11 patients). Of the 12 living patients at final follow-up, the mean MSTS score was 77%. While all patients had some functional restrictions, 96% had improvement in pain and greater function when compared to an amputation. Total humeral and elbow reconstruction had a mean MSTS score of 70% compared to 80% with segmental elbow reconstruction. Early complications were seen in 35% of patients. Seventeen percent of patients experienced nerve palsies, 9% had infections, and 30% experienced prosthesis or allograft complications.

Most of the literature on tumors of the distal humerus focuses on endoprosthetic reconstruction of the humerus and elbow. Aseptic loosening is a common complication when using endoprostheses. Also, given the proximity of the neurovascular bundle, patients are at risk for nerve palsies following reconstruction of the distal humerus and elbow.

Forearm

Osteosarcomas of the radius and ulna are quite rare. Little exists in the literature describing the treatment of primary forearm osteosarcoma.

Case Example: Forearm

A 14-year-old female with osteosarcoma of distal radial diaphysis treated with neoadjuvant chemotherapy followed by wide resection and free vascularized fibula reconstruction followed by adjuvant chemotherapy. Her resection specimen is shown with the skin paddle from prior open biopsy included (Image 2.4).

e

Image 2.4 AP and lateral radiographs show a lesion associated with osteosarcoma of the distal radius (**a**, **b**). T1-weighted MR image reveals the extent of the tumor

within the distal radius (c). AP and lateral radiographs show the reconstruction with a free vascularized fibula graft (d, e)

Skeletally Immature Patients

Skeletally immature patients present a unique challenge for successful reconstruction and limb salvage. Osteosarcoma most often occurs in the metaphysis of long bones near the physeal plate in skeletally immature patients. Resection of the physeal plate before physeal closure prevents future growth of the remaining portion of the resected bone. Due to the necessity for wide surgical margins in treating osteosarcoma, resection and subsequent reconstruction can lead to significant limb length discrepancies (LLD) [42]. The functional effect of the resulting LLD is largely dependent on the amount of LLD, age of the patient, and the anatomic location of the resection and reconstruction. In the upper limb, differences in length between the two limbs may result in cosmetic problems but typically do not impact function as long as the joint function and motor function of the hand is spared [42]. The major difficulties with limb preservation of skeletally immature patients occur with tumors involving the metaphysis of the lower limb. The degree to which LLD will have a clinically and functionally important effect is a product of the final difference between the affected limb's length and that of the contralateral limb. It is important to properly estimate future growth before deciding on a specific method of reconstruction.

There are a number of methods to estimate limb growth, which can be aided by computer software. The anticipated LLD is estimated assuming a normal growth rate in the contralateral limb while factoring in the patient's skeletal age and the growth remaining of the resected growth plate [3]. Levin et al. [3] suggest that when the final LLD is <2 cm, surgical procedures to accommodate the discrepancy are not necessary. For 2-5 cm, they suggest halting the growth of the contralateral side, typically via contralateral epiphysiodesis. Finally, for estimated deficits greater than 5 cm, their recommendation is to use expandable prostheses or later limb lengthening procedures. For very large predicted discrepancies, it may be necessary to consider amputation or rotationplasty.

For tumors of the diaphysis that do not involve the metaphysis, resection is often possible while sparing the growth plate. Reconstruction with allograft, vascularized autograft, or a combination of the two is the standard of care [3]. While internal fixation with plate constructs that extend to the epiphysis is often necessary to provide stability after surgery, once host-graft fusion has occurred, the epiphyseal screws can be removed to allow for the resumption of growth [3].

When resection of the growth plate is unavoidable, there are a number of reconstruction options. To preserve the articular surface and joint, Cañadell et al. [43] described the use of physeal distraction, a technique typically used for bone lengthening. As long as the epiphyseal edge of the resected bone is tumor free, they utilize external fixation for stabilization and distraction while filling the defect with a bone graft. Out of 20 patients, no patients experienced subsequent tumors in the epiphysis. Two experienced infection, one had a dislocation of the graft, one had a peroneal nerve palsy, and one had an allograft fracture. They reported mostly excellent and fair outcomes depending on the anatomic location.

Most reconstructions involving a joint in the skeletally immature are performed using endoprostheses in the United States. Implants can either be fixed length implants such as those used in adults or expandable implants that allow for later lengthening and the prevention or minimalization of LLD. Endoprostheses enable early weight-bearing and provide a stable construct. In children, implants need to be durable to prolong the need for future replacement of an endoprosthesis in the years following surgery as patients return to activities. Utilizing a slightly longer implant or fusing the contralateral growth plate is an option for patients closer to skeletal maturity [3]. In younger patients, an expandable prosthesis is often the best choice in preventing a clinically significant LLD.

There are multiple types of expandable endoprostheses on the market. Some use noninvasive magnetic expansion, which allows for expansion of a shorter length over a greater number of expansions without the need for additional surgery [37]. Most expandable endoprostheses require surgical expansion to directly lengthen the device, which increases the overall risk to the patient as a number of surgical extensions must be performed to reach the correct limb length. In a systematic literature review of the outcomes for limb-sparing surgery in pediatric patients, Groundland et al. [44] reported that patients receiving expandable implants had on average 2.95 expansions for a total expansion length of 29.9 mm in the proximal femur, 6.9 expansions for 84.8 mm for total femur, 4 expansions for 46.5 mm in the distal femur, and 5.7 expansion for 31.3 mm in the proximal tibia. They reported LLD in 24% of proximal femur patients, 0% of total femur patients and 13% of distal femur patients with no data for the proximal tibia. Additionally, a failure of the lengthening device occurred in 3.4% of patients at all locations. Futani et al. [45] reported the MSTS scores from three separate studies. The mean MSTS scores ranged from 74% to 81% with no difference based on the specific type of extendable prosthesis. However, complication rates tend be high, primarily arising from Type 4 and Type 2 failures [3, 45].

Treating skeletally immature patients requires special consideration for possible LLD following reconstruction of the lower limb. If the physeal plate is sacrificed with resection in a young patient, an extendable endoprosthesis and/or contralateral physeal ablation can be used to mitigate future LLD. However, it must be noted that some of these devices have had high rates of implant failure and may require an invasive procedure for lengthening.

Case Example: Expandable Prosthesis

A 9-year-old female with osteosarcoma of the left distal femur treated with neoadjuvant chemotherapy followed by wide resection and magnetic growing prosthesis reconstruction and adjuvant chemotherapy. Her resection specimen shows the extent of the intramedullary disease as well as the extra-osseous soft tissue component of the tumor (Image 2.5).

Case Example: Expandable Prosthesis

A 3-year-old girl who stopped using her right arm for 5-6 days due to acute onset of pain without trauma. Radiographs revealed a pathologic fracture through an osteoblastic and osteolytic lesion of the right proximal humerus consistent with an osteosarcoma. She was treated with resection and reconstruction with an expandable proximal humeral megaprosthesis (Image 2.6).

Case Example: Intercalary Graft

A 14-year-old boy with pain in the left knee after competitive biking was found to have an osteoblastic lesion in the left proximal tibia with ossification in the lateral soft tissues. He was diagnosed with osteosarcoma and was treated with resection and reconstruction with an intercalary allograft, plate fixation, and a supplemental onlay vascularized fibular graft since the tumor did not extend into the proximal tibial epiphysis. Metastasis of the vertebral body was found in the thoracic spine, and patient died despite chemotherapy and radiation (Image 2.7).

Conclusion

There are a variety of reconstruction options that can be utilized to successfully preserve affected limbs for patients with osteosarcoma. Endoprostheses, osteoarticular allografts, and APCs are common options for LSS. Each has its own advantages and disadvantages that differ among various anatomic locations. LSS has become the most common surgical treatment modality for extremity osteosarcoma. Large prospective trials comparing surgical techniques are generally not available and retrospective studies tend to have small sample sizes, limiting the evidence behind choosing one type of reconstruction over another. As a result, the type of reconstruction depends on the patient's functional needs and desires, surgeon proficiency in various techniques, the extent and anatomic location of the tumor, and the patient's age.



Image 2.5 AP and lateral radiographs show an osteoblastic and osteolytic lesion of the left distal femur consistent with osteosarcoma (a, b). The extent of the tumor can be seen on axial MR (b). The resected gross specimen can

be seen in image **d**. Postoperative radiographs show the reconstruction performed with magnetic expandable prosthesis (**e**)



Image 2.6 The pathologic fracture through an osteoblastic and osteolytic lesion on an AP radiograph of the right proximal humerus consistent with an osteosarcoma. There is a Codman's triangle at the distal medial aspect of the lesion (**a**). T1-weighted coronal and T2-weighted fat sat

MR imaging reveals the marrow and soft tissue extent of tumor (\mathbf{b} , \mathbf{c}). Five years after resection of the humeral osteosarcoma and reconstruction with an expandable proximal humeral megaprosthesis after several extensions (\mathbf{d})

Image 2.7 AP and lateral radiographs of the left tibia show an osteoblastic lesion in the proximal tibia with ossification in the lateral soft tissues (a, b). Sagittal T1-weighted and Axial T2-weighted MR images revealing the extent of tumor within the marrow and soft tissues. The tumor does not extend into the proximal tibial epiphysis, allowing a resection that spares the knee joint (c, d). AP and lateral left tibial radiographs 2 years after transepiphyseal resection and a healed reconstruction with an intercalary allograft, plate fixation, and a supplemental onlay vascularized fibular graft (e, f). Sagittal T1 weight MR image of the thoracic spine revealing a vertebral body metastasis. Despite chemotherapy and radiation, the patient died 4 months later (g)







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