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Review Article

Let the Survival Curves Speak for Themselves: Durable Function Salvage Surgery for Pediatric Bone Sarcomas

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Abstract

Oncologic outcomes following amputation and limb preservation surgery for pediatric lower extremity bone sarcomas are equivalent, therefore treatment goals have expanded to optimize patient function and quality of life. To that end, the durability and functional outcomes of a surgical reconstruction should be prioritized. Limb salvage surgery with an expandable endoprostheses provides an appealing surgical option for the skeletally immature patient, salvaging the limb's outward normal appears at the cost of construct durability and function. However, for the growing active child, functional outcomes should supersede the aesthetic. Therefore, function salvage, and not limb salvage, should be the true surgical objective. We propose the reexamination of the rotationplasty, a biologic reconstruction which sacrifices the physical appearance in an attempt to salvage patient function and quality of life.

Keywords: Pediatric; Sarcoma; Reconstruction; Limb salvage surgery; Rotationplasty; Function

Introduction

A multidisciplinary approach to the treatment of pediatric bone sarcomas has created well defined roles for each member of the therapeutic team. The primary responsibilities of the surgical oncologist are to (1) eradicate local disease and (2) provide a durable reconstruction that will minimize the functional deficits resulting from local oncologic control. Due to the relatively high incidence of pediatric bone sarcomas that occur in the lower extremity, reconstructions about the knee and in the proximal femur are performed regularly. Despite this, little consensus exists regarding the optimal reconstructive procedure for the skeletally immature patient.

The growing child poses a dynamic clinical problem- the challenge of providing a solution that will satisfy the patient's short- and long-term demands. Both rotationplasty and limb salvage surgery (LSS) with expandable endoprostheses have been championed as reconstructive options that accommodate for longitudinal growth, however both techniques have inherent deficiencies which invites critique and their comparison.

The clinical challenge

Osteosarcoma and Ewing sarcoma are the two most common bone malignancies in the pediatric population. Together they represent approximately 90% of bone sarcomas that occur in skeletally immature patients [1]. Contemporary five-year overall survival for pediatric patients with localized appendicular bone sarcoma approaches 70% [1-4]. Of these survivors, 90% will experience a durable remission of at least 15years [1]. Since equivalent oncologic outcomes have been observed with amputation and limb preserving negative margin tumor resections, surgical objectives include the optimization of post-operative function [5,6].

The functional demands of the growing patient and their anticipated long-term survival create a clinical challenge for the reconstructing surgeon. Survivors of pediatric bone sarcomas will become adolescents (if not already) and young adults (AYA), and be subject to the numerous physical and psychosocial challenges of the maturation process; all of which are accompanied by the burdensome sequelae of oncologic therapy and surveillance. Optimizing the durability of lower extremity reconstructions attempts to eliminate the looming fear of mechanical failure and minimize the time survivors spend in the healthcare system undergoing revision surgical procedures.

Currently, implant survival in LSS pales in comparison to patients' survival from their oncologic disease, with an expandable endoprostheses five-year implant survival of 35% [2,3,7,8]. Published Kaplan-Meier implant survival curves often exclude expandable endoprostheses due to their high rates of failure [9,10]. Even with the exclusion of expandable endoprostheses, which suffer a significantly higher failure rate than standard modular endoprostheses, long-term implant survival curves fail to keep up with overall patient survival (Figures 1) [7,11].

Outcomes of reconstruction

In combination with systemic therapy, equivalent oncologic outcomes have been observed following amputation and limb preserving surgery [5]. Rather than the type of surgical procedure, local recurrence rates are dependent upon the surgical margin status and the histologic response to neoadjuvant chemotherapy [6]. Therefore, limb preservation with an expandable endoprostheses or rotationplasty has replaced ablative surgery as the standard of care. Following limb preservation surgery, patients consistently demonstrate superior gait efficiency and functional outcomes when compared to those who have undergone an above-knee-amputation (AKA), but equivalent results in comparison to patients following a below-knee-amputation [12-14]. These dichotomous results suggest that significant limitations are imparted by ablation of the knee joint and not the mere addition of an external prosthesis.

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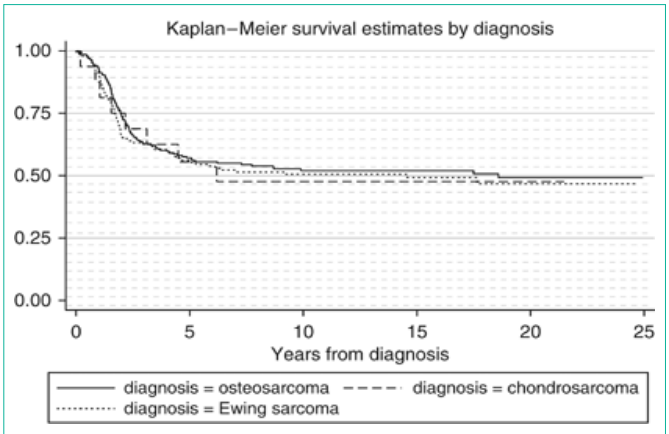

Figure 1: Kaplan-Meier curves demonstrating thelong-term survival of pediatric patients with osteosarcoma, Ewing sarcoma and high-grade chondrosarcoma. (Reprint with permission from [11]).

Complications

Despite over 30 years of clinical use and design optimization, expandable endoprostheses suffer from a high incidence of implant failure and revision surgery [5,15,16]. In a recent series Cipriano et al. reported 37 implant-related complications in 10 patients resulting in 150% reoperation rate at minimum of two years. In this series, after five years of follow up, all patients had undergone at least one endoprosthetic revision [16]. In another review of expandable endoprostheses, an implant revision rate of 38% was observed amongst 26 surviving patients, two of whom ultimately received an amputation [15]. The duration of follow up was not specified in this latter series, causing one to question the durability of these results. Predictably, the incidence of expandable endoprosthesis failures increase over time with observed three- and five-year implant survival rates of 60% and 35%, respectively [8].

As with LSS, complications occur regularly following rotationplasty, however the rates of major revision surgery are consistently lower than those observed for expandable endoprostheses [15-19]. Specifically, in a series of 70 rotationplasty patients with an average of four years of follow-up, a 50% complication rate was observed. Vascular compromise due to anastomotic failure occurred in three patients, resulting in an AKA [18]. An additional series reported a complication rate of 30% and a reoperation rate of 20% with a minimum follow-up of two years [17]. With a mean follow-up of 59 months, Sawamura et al. reported their experience of 25 rotationplasties performed in patients for whom LSS was contraindicated [19]. In this high-risk patient population, rotationplasties were performed with an ultimate success rate of 88%. Thirty-two percent of patients underwent a reoperation for anastomotic failure, fracture, nonunion, wound complications and a slipped femoral capital epiphysis [19].

Functional Outcomes

Studies consistently demonstrate superior functional results in patients who have undergone rotationplasty compared to LSS [12,20,21]. While some comparative reports demonstrate superior multimodality outcome scores in patients with rotationplasty compared to endoprostheses [12,21], others fail to corroborate statistical significance, but note that rotationplasty patients have less daily pain, are less dependent upon assisted ambulatory devices and are more engaged in recreational activities when compared to patients with endoprostheses [12,20]. Following rotationplasty, up to 85% of patients have been observed returning to sport, including high-levels of competition [22,23]. Alternatively, patients who undergo LSS with an endoprostheses are typically restricted from engaging in strenuous physical activity and mechanical failures have been observed when these recommendations are neglected [16,24].

Psychosocial Impact

In contemporary healthcare centers, the incidence of LSS with expandable endoprostheses consistently surpasses that of rotationplasty, despite the latter's superior functional outcomes. Some believe that the cosmetic consequences of a rotationplasty are so damaging that psychosocial harm is imposed upon the recipient (Figure 2). Multiple attempts have been made to assess the psychosocial consequences of limb preservation surgery whether it is with an endoprostheses or a rotationplasty. Patient quality of life (QoL) and emotional well-being are assessed through self-reporting questionnaires, such as the SF-36, which are inherently subjective and biased by patients' life experience. The unique perspective afforded a pediatric sarcoma survivor is likely responsible for SF-36 scores amongst rotationplasty patients that match, and even at times surpass, age-matched healthy controls [25]. When compared to their healthy peers, rotationplasty patients have been found to have comparable general QoL, social support and psychosocial functioning, however up to 50% of patients in one series were apprehensive regarding the initiation of social and intimate contact [26].

In an attempt to assess the psychosocial impact of an amputation or rotationplasty, Robert et al. evaluated 57 AYA patients following these procedures and LSS for lower extremity osteosarcoma [27]. They found equivalent results between treatment groups when evaluating patients' QoL, body image, self-esteem and social support. While there were no significant differences between surgical treatment groups, functional outcomes correlated directly with patients' QoL. This finding caused the authors to conclude that superior lower extremity function, independent of the specific procedure, resulted in improved psychosocial outcomes.

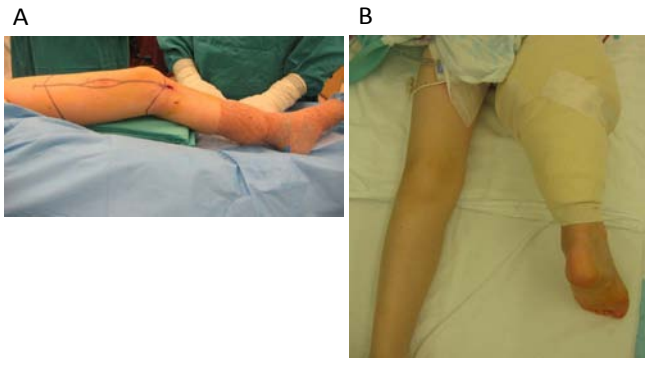

Figure 2: Intraoperative photographs of a pediatric patient (A) draped with skin markings indicating the planned region of resection for a distal femoral osteosarcoma, and (B) the appearance of the same limb, bandaged, following completion of the rotationplasty.

Conclusion

Lower extremity reconstruction options for pediatric bone sarcomas are fraught with complications and sacrifices. Limb salvage surgery with an expandable endoprosthesis allows for the approximation of an anatomically normal limb, but at the high cost of revision surgeries and functional limitations. Biologic reconstruction with rotationplasty is a function salvage surgery that sacrifices the outward appearance of the limb for superior function and durability.

In order to facilitate normalization of sarcoma survivors' lives, the treating physician would aim to minimize the time survivors spend in the health care setting. To that end, the durability and survival of reconstructive procedures should match that of the patients themselves. As five-year patient survival surpasses that of expandable endoprostheses, sarcoma survivors who receive LSS will likely experience an endoprosthetic failure and undergo revisions surgery [3,8].

Proponents of endoprostheses favor the relatively normal static outward appearance of a patient following LSS over the kinetic image of a child returning to play and sport with the use of rotationplasty prosthesis. Ignoring reoperation rates and patient functional outcomes in favor of aesthetic norms is reminiscent of the tail wagging the dog. Alternatively, if we work to decrease reoperation rates and improve patient mobility, improvements in QoL and psychosocial outcomes are sure to follow.

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