

OUR EXPERIENCE WITH THE MODULAR UNIVERSAL
TUMOUR AND REVISION SYSTEM IN PEDIATRIC
PATIENTS WITH EWING'S SARCOMA – REPORT
OF SIX CASES

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Abstract

Ewing's sarcoma (ES) usually affects patients between 10 and 19 years of age and has a rapid progression. The main goal of surgical treatment is to remove the tumour with at least wide margins, limb salvage being achievable in most cases. In recent years, modular tumour endoprostheses have become one of the main methods used in children.

From August 2012 to May 2019, six patients with histologically diagnosed ES underwent limb salvage surgery and reconstruction with a MUTARS[®] type endoprosthesis. The mean age of the patients is 12.6 years (from 8 to 16). For the reconstruction in all six patients we used MUTARS[®] type endoprostheses, two of which were expandable (MUTARS[®] Xpand) (33.33%).

The average follow-up period was 40 months. The longest follow-up was 96 months. The average MSTS score one month after operative treatment and rehabilitation was 62% (18/30) and the average MSTS score after 12 months was 72% (21/30). Mechanical failure of the expanding mechanism was the main complication we encountered in our series.

In conclusion limb salvage surgeries and reconstruction with modular tumour endoprostheses give patients with Ewing's sarcoma not only a chance for a disease-free life, but a potentially functional limb

Key words: sarcoma, Ewing surgery, bone neoplasms surgery, expandable endoprostheses, limb length inequality, limb salvage surgery

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Introduction. In pediatric patients Ewing's sarcoma (ES) is the second most prevalent bone tumour after osteosarcoma. However, it is a rare disease with an annual incidence of 2.9 per million with 90% of cases from the age of 10 to 20 [1-3]. ES is most commonly localized in the pelvis, proximal and distal femur, tibia, humerus, and fibula [1-3]. The most typical clinical symptoms are pain and local swelling. Usually low-grade fever is also present. ES has a variable imaging presentation, the most common being a poorly defined, infiltrative osteolysis with the cortex breached or destroyed. One of its most typical but infrequent signs is the lamellar periosteal reaction which leads to fusiform enlargement of the diaphysis. A large radiolucent soft tissue mass is usually present at the time of diagnosis [1-3]. MRI reveals the soft tissue extension and bone marrow invasion. PET/CT and isotope scan are hot and may occasionally show skeletal metastases at presentation. MRI, CT, and/or PET/CT typically show a much larger tumour extension than what appears on conventional radiographs [1-3].

The treatment of ES is multimodal comprising neoadjuvant chemotherapy, surgery, and adjuvant chemotherapy [1,4]. The main goal of the surgical treatment is to remove the tumour with at least wide margins, limb salvage being achievable in most cases. Wide tumour resection in the long bones often requires sacrifice of a physis and not infrequently of one around the knee [4-6]. Reconstructions with modular non-expanding and expanding tumour endoprostheses give these patients good function and a chance for equal limb length at skeletal maturity. We present our experience with the Modular Universal Tumour and Revision System (MUTARS®) in children with Ewing's sarcoma.

Materials and methods. From August 2012 to May 2019, six patients with histologically diagnosed ES underwent limb salvage surgery and reconstruction with a MUTARS® type endoprosthesis. Our series consists of five male and one female patients (5:1). The mean age of the patients is 12.6 years (from 8 to 16). The most common localization of the tumour was the distal femur in three patients (50%). Other localizations were the proximal femur (1), distal humerus (1) and distal tibia (1). All six patients underwent neoadjuvant chemotherapy. MRT, CT and/or PET/CT were conducted for pre- and post-chemotherapy staging. For the reconstruction in all six patients we used MUTARS® type endoprostheses, two of which were expandable (MUTARS® Xpand) (33.33%). Evaluation of the postoperative functional results was done with the Musculoskeletal tumour society score.

Procedure. Meticulous dissection of the tumour and the surrounding soft tissues was carried out before resection, which was done with an oscillating saw. We used a medial parapatellar approach in three of the patients with a localization of the tumour in the bones of the knee joint. The modular endoprosthesis was implanted and the remaining muscles were reattached using a Dacron sleeve. An expanding endoprosthesis was used in two of the patients (Fig. 1, 2). The expanding mechanism was tested before closure of the wound. Rehabilitation was initiated on the following day in all six patients.



Fig. 1. A, B: Pre-chemotherapy radiographs showing an osteolytic lesion in the distal metaphysis and diaphysis of the femur with a large soft tissue extension. C, D: Post-chemotherapy radiographs showing diffuse sclerosis and disappearance of the extraosseal extension. E, F: Post-operative weight-bearing full leg radiograph of the implanted expandable endoprosthesis

Results. The average follow-up period was 40 months. The longest follow-up was 96 months. The MSTS score of all six patients was calculated periodically during the routine physical exams. The average MSTS score one month after operative treatment and rehabilitation was 62% (18/30), and the average MSTS score after 12 months was 72% (21/30). One of our patients received a shoulder and elbow joint mega-prosthesis. His MSTS score at the one month follow-up

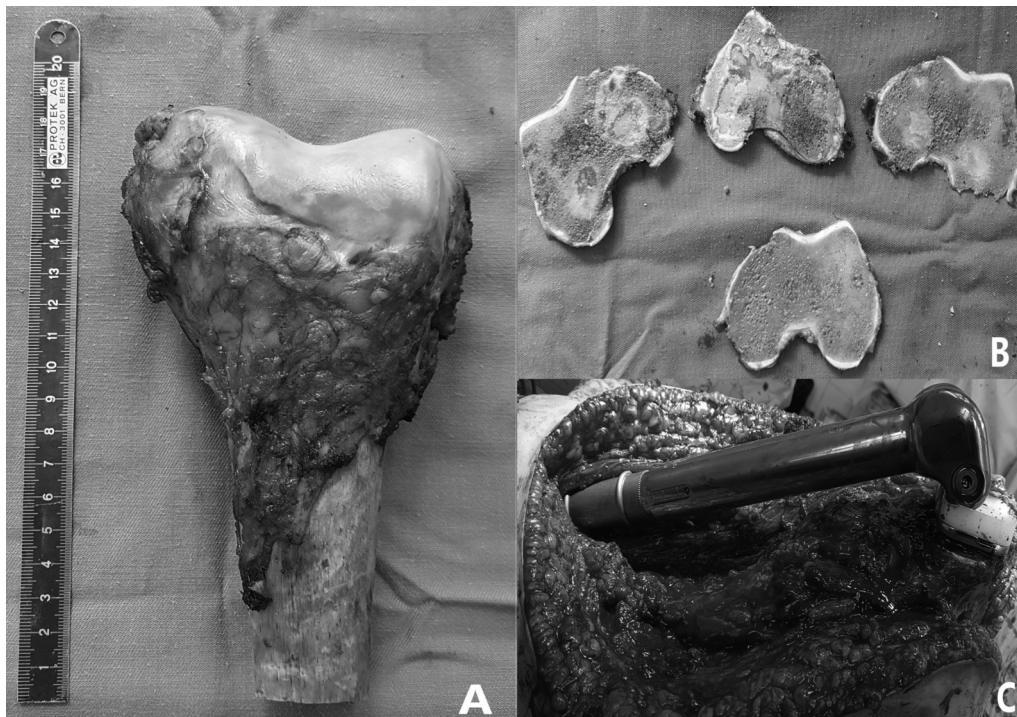


Fig. 2. A: Resected specimen. B: Serial slices of the distal femur for histological evaluation of the percentage of necrosis after chemotherapy. C: The implanted expandable endoprosthesis

was 53% (16/30), and improved drastically after vigorous rehabilitation reaching 76% (22/30) at the one year follow-up. Two of our patients died from metastatic disease, one of them two years and the other one year after operative treatment. One of our patients had local recurrence of the disease four years after surgery and is currently being treated with adjuvant chemotherapy.

We encountered complications mainly in our two patients who received an expandable endoprosthesis. In one patient (16%) mechanical failure of the expanding mechanism occurred two consecutive times in a single year and was managed with revision surgery. The same patient later developed severe reduction in the range of movement of the affected joint which did not improve after months of physical therapy. The patients MSTS score is 83% (25/30) seven years after the reconstruction. Superficial surgical site infection was encountered in the second patient one month after surgery and was successfully treated conservatively.

Discussion. Successful treatment of ES in pediatric patients is challenging as they are still in their skeletal growing years. Limb salvage surgery (LSS) together with the current advancements in chemotherapy give these patients a chance of full recovery and a potentially functional limb. There are four basic principles of LSS according to SIMON [7]:

- 1) local recurrence rate should not be greater and survival rate lower than that with amputation;
- 2) the procedure should not delay the following adjuvant chemotherapy;
- 3) reconstruction should be stable and should not be associated with local complications which require more hospital stays;
- 4) the function of the limb should approach that obtained by amputation and artificial limb prosthesis, although body image, patient preference, and lifestyle may influence the decision [7].

Some relative contraindications for LSS are infiltration of the neurovascular structures by the tumour, poor response to neoadjuvant chemotherapy, pathological fracture with violation of compartment boundaries, inappropriately performed biopsy, and predicted limb length inequality of more than 8 cm [8]. One of the main issues that arises from these procedures is the development of limb length inequality in children. This problem could be solved with the implementation of expanding endoprosthesis for the reconstruction, which offers the advantage of minimally or non-invasive elongation of the affected limb [4–8]. Usually an MRI, CT, and PET/CT are done to define the true extent of the tumour and to determine the resection length needed to achieve a wide surgical margin.

Determining the remaining skeletal growth and the expected limb length inequality at maturity is one of the first steps in preoperative planning. This can be achieved by using Anderson and Green's growth remaining charts, Moseley's graphs, the White–Menelaus arithmetic method and the multiplier method [8–11]. The decision to use an expandable endoprosthesis for the reconstruction is based on the remaining skeletal growth and the localization of the tumour. In our two cases with expanding endoprostheses it was determined that both patients had several remaining years of growth and the disease had affected the distal femur which contributes for about 70% of the total lower extremity growth.

The MUTARS Xpand system that we used in two of our patients is based on the FITBONE (WITTENSTEIN intens GmbH, Igersheim, Germany) principle of a telescopic actuator lengthening of the prosthesis. This expandable endoprosthesis allows for a non-invasive, daily lengthening which is achieved through a control unit and an antenna. After the patient has reached skeletal maturity and the endoprosthesis is fully expanded, a conversion to a definitive modular prosthesis is advised.

There are several reported complications associated with this type of reconstruction. Aseptic loosening is the primary late complication involving this type of procedure in tumours located in the distal femur [4–6, 13, 14]. Problems during elongation and mechanical failure are frequent complications most commonly associated with the expanding mechanism. Deep infection is the most serious late

complication as other late complications can be treated successfully with revision surgery [15]. Depending on its severity, an infection can lead to amputation [6]. Other complications include flexion contracture and reduced range of movement, stress shielding, nerve palsy, and peri-prosthetic fractures [6,13,14]. In the series of 32 patients with expandable prostheses, ECKARDT et al. [5] reported that more than 50% of their patients had at least one of the following complications: aseptic loosening, mechanical failure, flexion contracture, and temporary nerve palsy.

Conclusion. Limb salvage surgeries and reconstruction with modular tumour endoprostheses give patients with Ewing's sarcoma not only a chance for a disease-free life, but a potentially functional limb. The problem of limb length inequality that arises after such procedures can be successfully overcome with the implementation of expandable endoprostheses. Careful preoperative planning and precise surgical technique are essential for the success of the procedure and the good long-term results.

REFERENCES

- [1] PICCI P., M. MANFRINI, N. FABBRI, M. GAMBAROTTI, D. VANEL (2014) Ewing Sarcoma. In: Atlas of Musculoskeletal Tumors and Tumorlike Lesions: The Rizzoli Case Archive, Bologna, Springer Int. Publ., Switzerland, 203–217.
- [2] MAHESHWARI A. V., E. Y. CHENG (2010) Ewing sarcoma family of tumors, J. Am. Acad. Orthop. Surg., **18**(2), 94–107.
- [3] WIDHE B., T. WIDHE (2000) Initial symptoms and clinical features in osteosarcoma and Ewing sarcoma, J. Bone Joint Surg. Am., **82**(5), 667–674.
- [4] NYSTROM L., J. MORCUENDE (2010) Expanding endoprosthesis for pediatric musculoskeletal malignancy: Current concepts and results, Iowa Orthop. J., **30**, 141–149.
- [5] ECKARDT J. J., M. KABO, C. KELLEY, W. WARD, A. ASAVAMONGKOLKUL et al. (2000) Expandable endoprosthesis reconstruction in skeletally immature patients with tumors, Clin. Orthop. Relat. Res., **373**, 51–61.
- [6] DICAPRIO M., G. FRIEDLAENDER (2003) Malignant bone tumors: Limb sparing versus amputation, J. Am. Acad. Orthop. Surg., **11**(1), 25–37.
- [7] SIMON M. A. (1991) Limb salvage for osteosarcoma in the 1980s, Clin. Orthop., **270**, 264–270.
- [8] ANDERSON M., W. T. GREEN, M. B. MESSNER (1963) Growth and predictions of growth in the lower extremities, J. Bone Joint Surg. Am., **45-A**, 1–14.
- [9] MOSELEY C. F. (1977) A straight-line graph for leg-length discrepancies, J. Bone Joint. Surg. Am., **59**, 174–179.
- [10] MENELAUS M. B. (1966) Correction of leg length discrepancy by epiphysial arrest, J. Bone Joint Surg. Br., **48**(2), 336–339.
- [11] PALEY D., A. BHAVE, J. E. HERZENBERG et al. (2000) Multiplier method for predicting limb-length discrepancy, J. Bone Joint Surg. Am., **82**(10), 1432–1446.
- [12] COOL W. P., S. R. CARTER, R. J. GRIMER et al. (1997) Growth after extendible endoprosthetic replacement of the distal femur, J. Bone Joint Surg. Br., **79**(6), 938–942.

- [¹³] DOTAN A., S. DADIA, J. BICKELS et al. (2010) Expandable endoprosthesis for limb-sparing surgery in children: long-term results, *J. Child. Orthop.*, **4**(5), 391–400.
- [¹⁴] BUS M. P., M. A. VAN DE SANDE, M. FIOCCO et al. (2017) What are the long-term results of MUTARS[®] modular endoprostheses for reconstruction of tumor resection of the distal femur and proximal tibia?, *Clin. Orthop. Relat. Res.*, **475**(3), 708–718.
- [¹⁵] ZAJONZ D., A. ZIEME, T. PRIETZEL et al. (2016) Periprosthetic joint infections in modular endoprostheses of the lower extremities: a retrospective observational study in 101 patients, *Patient Saf. Surg.*, **10**, 6.

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