



Milton Rochman. *Dreams*. Acrylic on canvas, 36" × 36".
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*Limb equalization in skeletally
immature patients with
malignant bone tumors
can be accomplished with new
expandable prosthetic devices.*

Modular Endoprostheses for Children With Malignant Bone Tumors

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Background: *The treatment of malignant bone tumors in skeletally immature patients is difficult because the required surgery removes the growth plate of one extremity. To date, few techniques for limb salvage have been successful due to complications involving limb expansion and limb-length discrepancy. Newer technology is now available for prosthetic replacements for modular expansion.*

Methods: *The authors review current surgical treatment of malignant bone tumors in children, with emphasis on reconstruction with expandable modular prostheses, and they present their own experience.*

Results: *Thirty-seven children with malignant bone tumors underwent primary tumor resection and reconstruction with a modular prosthetic device. Fourteen had subsequent successful expansions with modular prostheses. A new prosthesis, in which lengthening is achieved by an external electromagnetic field rather than an open surgical procedure, is discussed.*

Conclusions: *Limb-preserving surgery in children with malignant bone tumors can be accomplished successfully with modern prosthetic devices that have expandable modules within them. The technique of expandable prostheses using electromagnetic fields rather than surgical interventions shows promise.*

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Introduction

The surgical management of skeletally immature children with malignant tumors of bone is challenging. Traditionally, amputation was the treatment of choice for these patients. Many surgeons believed that limb salvage was a nonviable option for very young patients. The combination of significant limb-length discrepancy at maturity and the child's inability to actively par-

ticipate in rehabilitation compromised the good results seen in older patients. In recent years, this view has been challenged. Custom, modular endoprostheses have been designed to allow limb lengthening and thus prevent leg-length discrepancy. Aggressive rehabilitation programs have demonstrated that even the youngest of patients can participate and succeed, resulting in a highly functional extremity. In this report, we discuss the problem in treating skeletally immature patients, review the use of modular endoprostheses, and explore future directions.

The most common primary malignant tumor of bone is osteosarcoma, followed by Ewing's sarcoma of bone.¹ Approximately 1,000 new cases of osteogenic sarcoma are diagnosed annually in the United States. The peak incidence for osteosarcoma occurs in the second decade, with 75% occurring between 10 and 25 years of age.² The peak age for females seems to be younger than that for males. There is an overall male predominance (1.3:1) in osteosarcoma,² although some histologic variants occur more often in females.

The most common anatomic location of osteosarcoma is the metaphysis around the knee³ (distal femur, 35%; proximal tibia, 20%; and proximal humerus, 10%).² The metaphysis is the location for 80% to 90% of these tumors.⁴

The incidence of Ewing's sarcoma, the second most common malignant bone tumor in childhood, is one third that of osteosarcoma.² Ewing's sarcoma is seen most commonly in the first decade of life,³ and 70%-80% occur between the ages of 5 and 25 years. The male-to-female ratio varies from 1.3-1.5:1.² Anatomically, the distribution of Ewing's sarcomas is similar to that of osteosarcoma. The most common location is in the long tubular bones, with the femur most often affected.² Metaphyseal and metadiaphyseal locations are the most common areas of involvement within the bone² contrary to the commonly held belief that Ewing's sarcoma is a mostly diaphyseal lesion.

Surgical Management of Bone Tumors

Treatment of these malignant tumors involves neoadjuvant chemotherapy for systemic control and surgery and/or radiation for local control.¹ Chemotherapy and radiation for these tumors are not considered in this report.

Surgical management of these tumors requires wide resection to achieve local control. Local control rates from current approaches are approximately 90%. As already noted, most of these tumors will occur in

patients who are skeletally immature. The location of these tumors in the growing areas of bone commonly necessitates the removal of the involved growth plate. Subsequent continued growth in the contralateral extremity results in a limb-length inequality.

The distal femoral growth plate produces 1.6 cm in longitudinal growth per year. The skeletal growth is reported to stop in boys at age 16 and in girls at age 14. Therefore, a 10-year-old boy with an osteosarcoma of the distal femur would expect at least a 9.6-cm leg-length discrepancy at skeletal maturity. This does not include the potential loss of growth due to reconstruction of the uninvolved tibial side that would result in an interruption of growth of that physis.²

From a functional standpoint, the legs should be of equal length if possible. If left untreated, limb-length inequality can result in low back pain and occasional compensatory scoliosis. Gait disturbances are also possible. In addition, the cosmetic effects of a shortened leg, as well as the increased energy and expenditures required to power an extremity with a limb-equalizing orthotic, are important considerations. Surgical options for limb-length inequalities include lengthening at the time of the initial surgery or placement of an expandable prosthesis, or both. If a nonexpandable device is used, such as an allograft or non-modular implant, then the leg-length discrepancy must be corrected later. Another option is to perform a contralateral epiphysiodesis. Bone shortening after skeletal maturity is reached has also been described for treatment of this problem.

Modular Expandable Prostheses

In an effort to prevent limb-length inequality, surgeons and engineers have sought to design devices that would provide physiologic "growth" of the reconstruction with a relatively minimal surgical procedure. This would obviate the need for surgery on the uninvolved extremity as well as revision surgeries to the affected limb.

Modular expandable prostheses have been in use as early as 1976. The original Stanmoore design utilized a screw extension mechanism.² The first expandable device widely used in the United States was the Lewis expandable adjustable prosthesis (LEAP), introduced in 1983. The mechanism consists of a fixed stem with a screw extension mechanism. Expansion is induced using a chuck key that turns the screw mechanism. The key is localized through a small incision. As the screw mechanism is advanced, the tubular portion of the construct moves and expands the overall length of the

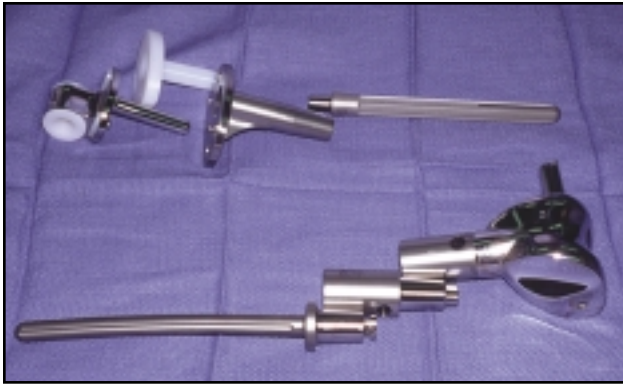


Fig 1. — Modular segmental expandable prosthesis (unassembled).



Fig 2. — Modular segmental expandable prosthesis (assembled).

prosthesis and thus the length of the extremity.⁵ More recent designs have focused on a modular system in which the original device consists of several distinct pieces (Figs 1-3). At the time of lengthening, a midsection is replaced with a longer midsection to parallel contralateral leg growth. These midsections are sequentially replaced with longer ones as growth occurs. Potential stretch injury to vital structures limits the midsection length to approximately 2 cm.

In all of the commonly used expanding mechanisms, a surgical procedure is required for expansion. The LEAP device, for example, requires minimally invasive surgery, needing only a small incision to insert the chuck key, while the modular midsection-replacing designs require major revision surgery. Occasionally, a capsulotomy is required to allow full expansion of the device. With the modular midsection-replacing system, an even longer incision is required to fully expose the prosthesis and replace the midsection. A capsulotomy is frequently used to facilitate placement of this new midsection. Some authors, however, report replacing the midsection without capsulotomy and using a smaller incision (G.D.L., personal communication, 2001).

Thus, all of the traditional lengthening mechanisms require a formal surgical procedure for expansion. Complications can be serious. Neurologic compromise, loss of motion, and vascular injury have all been reported.^{6,8} Neurologic compromise was minimal if lengthenings were 2 cm or less. Additional costs include loss of time from school, additional physical therapy, and surgical and hospital costs.

A 10-year review of 60 patients with the LEAP prosthesis indicated that most patients had satisfactory outcomes.² The authors felt that this was a viable option until a fully internal device requiring no further surgery could be designed.

Experience with a modular endoprosthetic midsection-replacing type of expandable prosthesis⁹ in

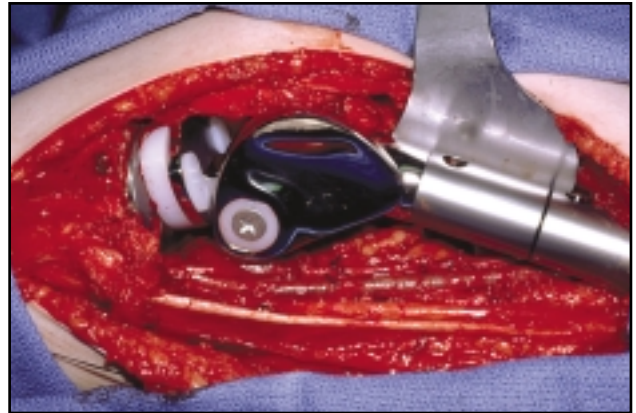


Fig 3. — Modular segmental expandable prosthesis inserted in the knee.

very young patients at St. Jude Children's Research Hospital has also been satisfactory. Over the past 5 years, we have placed 37 modular expandable prostheses. Each was a standard, rotating-hinge design manufactured by one of two companies. Twenty-seven were placed in skeletally immature patients (youngest age 6) with the anticipation of future revision surgeries to accommodate lengthening. The other 10 were placed in adolescents who were close to skeletal maturity. Segmental modular devices were placed in the event that significant leg-length inequality occurred from unexpected continued growth.

In these 37 patients, 17 expansions have been performed in 14 patients and three patients had more than one expansion. The typical expansion was 2 cm in length resulting from the placement of a new, longer midsection. In over half the procedures, an extended incision was required as well as a capsulotomy. Expansions in the remaining patients were accomplished through a smaller incision with the use of a special device to crank apart the prosthesis and allow midsection exchange. Few complications occurred in these 17 procedures. One patient developed a long-term neurapraxia after his second expansion, which has only partially resolved. Initially, most patients required a 4- or 5-day hospital stay, but with newer techniques, the

procedures are performed on an outpatient basis with physical therapy given for 4 to 6 weeks thereafter. The femoral or tibial component of the device was noted to be loose in 4 patients at the time of the expansion, and this item was revised as part of the procedure. All patients underwent extensive rehabilitation programs postoperatively. Despite the extensive nature of the surgical procedure, functional scores returned to their pre-expansion levels in all but one patient. There was little loss of motion or strength. To date, of those patients who were skeletally immature, only one has a significant limb-length inequality (4 cm). One patient has refused further surgeries and at last follow-up had a 2-cm difference. All of the patients who have reached skeletal maturity have near equal leg lengths (less than 2 cm difference). Our results indicate that the modular, expandable device is a good method for reconstruction following bone tumor resection. The expansions can be performed with little loss of function. One prosthetic infection occurred late but before the patient had an expansion. The patient had become septic after a dose of chemotherapy and was immunosuppressed. Our experience indicates that failure rates with modular systems due to loosening, infection, or implant failure are similar to rates seen with nonmodular systems.

We have begun to use an internally expandable device that does not require activation via surgical procedure. The lengthening occurs through activation of an internal device by an external electromagnetic field, thus eliminating the need for an open surgical procedure. Our indications are for children who are expected to develop a limb-length discrepancy greater than 6 cm. Bone ages are obtained at the time of diagnosis. The predicted limb-length inequality is calculated using the arithmetic method. The surgical options are then discussed with the family, and they choose either the traditional midsection lengthening device or the non-surgical internally expandable device.

We currently use the Phenix Growing Prosthesis (Phenix Medical, Paris, France). First designed in France



Fig 4. — Phenix expandable prosthesis inserted in the knee.

in the early 1980s, it has been utilized by Dr J. Dubouset for the past 10 years, and more than 100 patients have been treated. Wilkins et al^{10,11} reported the American experience in the 1999-2000 Musculoskeletal Tumor Society meeting. The Phenix prosthesis consists of two tubes with a spring mechanism buried in the larger tube (Figs 4 and 5). It is maintained compressed by a polyethylene locking mechanism. One tube is connected to the stem of the implant, and the second comprises the hinge portion of the hinge prosthesis. The uninvolved side of the joint is minimally resurfaced with a press-fit stem to attempt to preserve function of the non-involved growth plate. Expansion is achieved via exposure to an external electromagnetic field



Fig 5. — Radiograph of Phenix expandable prosthesis.



Fig 6. — Patient activating an electromagnetic device to correct leg-length discrepancy.

around the extremity (Fig 6). The coil produces heat in the center of the field. This is focused on the receiving antennae within the implant itself. This antenna is heated by the electromagnetic field. The heated element softens the surrounding polyethylene locking mechanism, which allows spring expansion. The spring expansion pushes the two tubes apart from each other, thus lengthening the overall length of the leg. As skeletal growth occurs on the contralateral side, a discrepancy in the overall length of the two legs occurs. Follow-up scanograms quantify the discrepancy. Once a .5- to 1.0-cm discrepancy has occurred, the patient is taken to the fluoroscopy suite and remains awake without sedation or anesthesia. The receiving antennae within the implant are identified under fluoroscopy guidance and the skin is marked. The electromagnetic device is then applied over the receiving antennae, and the patient activates the device for a period of approximately 15 seconds. A radiograph using magnification markers is then obtained of the level of the spring to determine how much lengthening has occurred. Once the desired amount of lengthening (typically 0.5 to 1 cm) is achieved, the process is stopped. One or two activation processes are required to achieve the desired lengthening. The procedure is halted if the patient experiences any discomfort or anxiety. Follow-up radiographs are scheduled at 2- to 3-month intervals until the next lengthening is required.

Experience with the Phenix Growing Prosthesis at St. Jude Children's Research Hospital is limited to four patients. Three patients underwent primary procedures following tumor resection. The fourth patient had a revision of a pre-existing modular expandable prosthesis with conversion to the Phenix prosthesis. The tumor locations included one total femur, two distal femurs, and one proximal tibia. To date, 13 lengthenings have been performed with no complications related to the lengthenings themselves. All but one patient underwent lengthening while awake, and they

activated the device themselves. Incremental lengthenings of .5 to 1.0 cm were achieved in all patients. The patients have experienced minimal discomfort, and none has required analgesics. Following lengthening, most patients walked out of the fluoroscopy suite and rapidly regained their motion. The neurovascular status of all patients was intact both before and after the lengthening, and no complications relating to the implant itself or the lengthening mechanism occurred. The functional results are similar to the traditional modular knees used in the past.

Conclusions

The use of modular expandable prostheses is an acceptable approach to managing limb preservation in children who have a significant expected growth discrepancy secondary to resection of malignant tumors of bone. Traditional expandable methods have included the use of a modular prosthesis, which requires an open surgical procedure for lengthening. There is some associated morbidity, but in general the technique is an effective limb salvage procedure. Current studies are underway in the United States and Europe in the use of non-invasive expandable prostheses. Early experience is promising, but further study is warranted to determine long-term structural integrity of these newer designs.

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