Limb salvage is now customary in the treatment of primary bone tumours. The proximal tibia is a frequent site for these neoplasms but reconstruction, especially in children, is a formidable challenge. We reviewed 20 children with extendible replacements of the proximal tibia, all with a minimum follow-up of five years. Five died from their disease and, of the remaining 15, four had above-knee amputations for complications. Infection occurred in seven patients; in five it was related to the lengthening procedure. Aseptic loosening is inevitable in the younger children and only two have avoided a revision, amputation or other major complication; both were aged 12 years at the time of the initial surgery. Despite this, 11 children are alive with a functioning leg and a mean Musculoskeletal Tumour Society functional score of 83%. The lengthening mechanisms used in our series required extensive open operations. We are now using a simpler, minimally invasive, technique which we hope will decrease the incidence of complications. At present, the use of extendible prostheses of the proximal tibia remains an experimental procedure.

In children, the proximal tibia is the second most common site for primary malignant tumours of bone. Our experience of osteosarcomas in children aged under 16 years has shown that 23% arise in the proximal tibia. The management of children with primary bone tumours is by chemotherapy and surgery, and survival for both Ewing’s sarcoma and osteosarcoma is now about 55% to 65%. Surgical options for tumours of the proximal tibia in children include above-knee or through-knee amputation, resection-arthrodesis, Van Nes rotationplasty, allograft replacement, bone transport or endoprosthetic replacement. Each of these techniques has advantages and disadvantages, but all must allow complete excision of the tumour with low risk of local recurrence. Limb salvage can be as safe as amputation in terms of overall survival, and is now usually considered in cases in which the tumour is resectable and when there is a reasonable response to neoadjuvant chemotherapy.

In children, the challenges of limb salvage include the additional problems of maintenance of equality of limb-length on the affected side and ensuring that any reconstruction of the limb allows function acceptable to the child and parents. Reconstruction must be durable with a low incidence of complications. Set against this must be the potential advantages of an amputation for which return of function is early and predictable. Children with artificial limbs cope well after an initial period of adjustment, but further operations for problems with the stump, non-compliance with wearing of the prosthesis and the high costs need to be considered.

The use of extendible endoprosthetic replacements has become common in recent years and their value in the distal femur is now well documented. Endoprostheses of the proximal tibia have their particular difficulties and have a higher rate of complications than endoprostheses at other sites, notably infection and problems with the extensor mechanism. In children, the risks of using an extendible endoprosthetic replacement are potentially even greater. We describe our experience of the use of these implants in children with tumours of the proximal tibia and assess whether this technique is justifiable.

Patients and Methods

Between 1983 and 1998 we inserted 30 extendible endoprostheses of the proximal tibia in children. We have followed 20 of these, carried out before June 1993, for a...
minimum of five years. There were 13 boys and seven girls with a mean age of 9.9 years (5 to 14). The diagnosis was osteosarcoma in 14, Ewing’s sarcoma in three, malignant fibrous histiocytoma in one, recurrent chondro-myxoid fibroma in one and disappearing bone disease in one.

The decision to use an endoprosthesis was made after appropriate staging and biopsy. The necessity for an extendible endoprosthesis was based on the estimated growth remaining in the resected physis, calculated using the growth charts after an estimation of bone age based on interpretation of a radiograph of the hand. In general, if there was likely to be discrepancy of less than 2 cm between the limbs after resection of the tumour and insertion of a prosthesis, then a standard adult-type prosthesis was used, but inserted up to 2 cm longer than the bone replaced. This resulted in the operated leg being temporarily longer than the normal side and usually allowed limb-length equality to be achieved by skeletal maturity. Thus, extendible prostheses were used rarely in girls over the age of 11 years and in boys over the age of 13 years unless they were skeletally immature.

The prostheses used were all designed and manufactured at the Department of Biomedical Engineering, University College, London and the types of extending mechanism have been described previously. Those used in the initial part of this series were lengthened by means of a ball-bearing mechanism, but this proved unreliable, and in 1988 it was replaced by a system which required insertion of a ‘C’ collar (Fig. 1). This needed an incision at least 2 cm longer than the length increment. Since 1991, we have used a minimally invasive extendible endoprosthesis which can be lengthened percutaneously using a customised Allen screwdriver. Six children had ball-bearing lengthening systems, ten had ‘C’ collars and four had minimally invasive devices. All of the implants were cemented on the tibial side. All patients had a passively growing uncemented component inserted on the femoral side of the prosthesis to allow continuation of some growth in the distal femoral epiphysis. This has been shown to be effective in allowing about 80% of the normal growth of the proximal tibial physis to continue when used for tumours of the distal femur.

The operative technique for resection of the proximal tibia is the same for children and adults. At the start of this series we used a synthetic mersilene mesh (Johnson & Johnson Ltd, UK) to cover the fascial defect over the tibia after resection of the tumour and a terylene rope to reattach the patellar tendon to the prosthesis. After the very high incidence of infection with this combination in 1988 we began to use gastrocnemius muscle flaps to cover the front of the prosthesis and to reconstruct the extensor mechanism.

We assessed the results using Kaplan-Meier survivorship analysis for outcomes in terms of survival of the prosthesis and the incidence of major complications. Functional assessments were done using the modified scoring system of the Musculoskeletal Tumour Society.

Results

Oncological results. Eight of the 20 children developed metastatic disease. There was local recurrence in two patients with osteosarcoma, both of whom had amputation for local control. Five patients, including one of the latter two, died as a result of metastatic disease.

Prosthesis results. The mean amount of tibia resected was 62% (40 to 89). In the smallest children the diameter of the intramedullary stem of the prosthesis was necessarily very slim (minimum 6 mm) and hence loosening was inevitable as the diameter of the tibia grew. Three patients had loosening after a mean of 54 months, and were treated by revision to a larger cemented component. On the femoral side a ‘sliding’ component was used to allow for continued growth of the physis. In older children the sliding component was contained in a polyethylene tube within the bone, while in younger children this was not possible due to the small size. The growth of the physis continued in all patients and was assessed at 63% of its anticipated normal growth. The maximum proportion of physis destroyed by the femoral component was 13% and, as in the proximal
tibial physis, there was no correlation between the surface area destroyed and continued growth.\(^{16}\)

**Complications**

*Limb salvage.* There were five amputations. Two were for local recurrence at 40 and 48 months, respectively, two for infection at 33 and 76 months and one for disappearing bone disease at 45 months.

*Infection.* Infection occurred in seven patients at a mean of 44 months from the initial operation. There were no early infections in the first three months after the insertion of the endoprosthesis. The cause was thought to be the lengthening procedure itself in five patients, secondary to an ingrowing toenail in one and to an infected fracture blister in one. Treatment was with antibiotics alone in two cases; in one the infection was successfully controlled while in the other it is ongoing and the patient is likely to require a two-stage revision. One patient had a successful one-stage revision. Two patients had two-stage revisions for infection, both of which were initially successful and one remains so. The other patient, however, developed further infection after another eight lengthening procedures some five years later and at that stage requested an amputation. Two other patients had amputation as the primary treatment of their infection. The overall risk of the prosthesis becoming infected was 68% within ten years. A total of 117 lengthening procedures was carried out and infection occurred after six of these with one patient being infected twice, a risk of infection per procedure of 5.1%. Three of these infections occurred in patients with the ball-bearing lengthening prosthesis, none of whom had a medial gastrocnemius muscle flap, and three with the ‘C’-collar device. The rate of infection for the ball-bearing lengthening prosthesis was thus 50% while for the ‘C’ collar it was 30%. There have been no infections yet after lengthenings of the minimally invasive prosthesis.

Of the seven children in whom we used a synthetic mesh to reconstruct the fascial defect at the front of their tibia, four developed infection. Only three infections occurred in the 13 children who had reconstruction using a medial gastrocnemius muscle flap. This difference was not significant.

*Fractures.* Five children had fractures of the femur. These arose after minimal trauma and were all at the level of the tip of the femoral sliding component. They were all treated conservatively, but this usually resulted in shortening of the leg and, in some, stiffening of the knee. One boy with disappearing bone disease required an amputation because of loss of bone stock after his fracture. Another developed an infected prosthesis because of infection in a fracture.
 blister while being treated by skin traction.

Other complications. Four children developed palsy of the peroneal nerve after the initial operation. All were treated conservatively and all recovered. There were no other nerve or vascular complications.

Revision procedures

Lengthening. Three patients did not have lengthening carried out since they had developed metastatic disease and died before it would have become necessary. The mean number of lengthenings in the remaining patients was 6.8. Two patients had 11 lengthenings, each gaining 66 mm as a result. Two required revision procedures because the prosthesis reached maximal extension, and one because the extension mechanism had jammed. In young patients, the original prosthesis is usually so small that the total lengthening needed to adulthood can never be achieved with one prosthesis and revision of part of the endoprosthesis is an expected procedure (Fig. 2).

Other procedures. Apart from lengthening, a wide variety of other procedures was required including manipulation, release of contractures both of the knee and ankle and treatment of periprosthetic fractures. The total number of operations, including the biopsy and initial definitive surgery, lengthenings and complications, ranged from two to 19 (mean 10).

Functional outcome. Eight of the 11 patients who were still alive reached skeletal maturity and were reviewed when aged between 16 and 26 years. The mean leg-length discrepancy in these patients was 10 mm. The mean functional score was 83%. The best patients had over 90° flexion at the knee with an intact extensor mechanism and no extensor lag. Most had a moderate extensor lag associated with some limitation of flexion as is found in adult tibial endoprosthetic replacements. 

Discussion

The management of a child with a tumour of the proximal tibia is complex. Both children and parents initially see limb salvage as the preferred option since it is thought to be more ‘normal’ than other options. We have established what ‘price’ has to be paid for achieving the goal, and in how many patients this was reached.

We identified a very high rate of complications in this small series of children with extendible endoprostheses of the proximal tibia. Of the 20 patients, five have died from their disease, including one who had an amputation for local recurrence. Four of the surviving 15 patients had amputations. There are only 11 patients still alive with intact limbs. Of these, only two have not required a revision procedure of their implant or had a major complication and both were aged 12 years at the time of their initial surgery. The complications in the other nine were infection in four, loosening in three and revisions for maximum extension in two.

We believe that in young children revision for loosening is inevitable and the patients and families must be warned of this before agreeing to surgery. Even allowing for this being an ‘acceptable’ complication, 65% of patients developed significant unexpected complications in the form of either fracture, local recurrence or infection. Of those who avoided a significant complication, three never had a lengthening since they died from their disease within two years of the initial operation. They were, however, mobile and walking throughout that time and their limb-salvage procedure was considered successful. The overall risk of developing at least one complication over the first ten years was 82% (Fig. 3).

The alternatives to the use of extendible prostheses clearly need to be assessed in this light. It is possible that the use of an uncemented endoprosthesis may secure a more durable fixation in bone thus reducing the need for revision for mechanical loosening. For tumours of the upper tibia, a through-knee or above-knee amputation gives excellent local control with early fitting of a prosthesis. Limb-length discrepancy needs to be carefully assessed before amputation to ensure that the leg will be of suitable dimension when growth is completed. Overgrowth of the stump may occur and require further surgery, possibly on more than one occasion. Children with above-knee or through-knee amputations can attain a very high level of function although most are not always so active. 

Rotation-plasty is an attractive and well-established alternative for children with tumours of the distal femur, but is rarely used for tumours of the proximal tibia.

Bone transport is another possible option and has been recently described in some detail, but only for tumours of the distal femur. 

A combination of allograft and vascularised fibular graft has been used together with the technique of intraepiphyseal resection, but does not allow lengthening of the limb.
We have previously reported the results of endoprosthetic replacement of the proximal tibia in adults and highlighted the high incidence of complications, especially infection of the prostheses. For children who are faced with repeated operations for lengthening, as is expected, the problems were worse. The overall risk of infection of 68% and the risk per lengthening of 5.1% are both clearly unacceptable. This rate of infection included data from a time when the lengthening technique was relatively unorthodox and required an extensive open approach to the prosthesis, often through thin and scarred tissue. Since we have been using the minimally invasive prosthesis with a routine medial gastrocnemius flap infection has decreased dramatically.

Other authors have highlighted the problems of extendible prostheses in the proximal tibia. After a two- to four-year follow-up of 13 children there were three infections and one amputation. One patient at another centre had an amputation for breakage of the lengthening mechanism. An infection rate of 17% was found in 28 patients with extendible replacements of the proximal tibia and distal femur, and in 15% of a series of extendible replacements around the knee. Another option would be to insert a modular type of adult prosthesis and simply to add slightly longer components as and when necessary. This has the obvious disadvantage of an extensive open procedure, although more lengthening within the tolerance of the vascular bundle can be achieved.

Despite the high incidence of complications, even in survivors, we have shown favourable outcomes in terms of quality of life, a subject which clearly warrants further investigation.

It is clear from these results that extendible replacements of the proximal tibia are fraught with problems, particularly when an open technique is needed to achieve lengthening. The advent of the minimally invasive prosthesis seems to have decreased the risk of infection, but this will need to be carefully monitored in the future. The risk of aseptic loosening may be resolved by the use of an uncemented modular type of adult prosthesis and simply to add slightly longer components as and when necessary. This has the obvious disadvantage of an extensive open procedure, although more lengthening within the tolerance of the vascular bundle can be achieved.

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References


