The two most common primary malignant bone tumors, osteosarcoma and Ewing sarcoma, are principally diseases of childhood and adolescence, with 45% of patients under the age of 16 and 17% under the age of 12 years at diagnosis.

In the last 30 years the 5-year survival rate has increased from 10% to 70%. Even in patients with metastases at diagnosis, the 5-year survival rate has reached 20% to 30% due to chemotherapy and surgery for metastases as well as the primary tumor. 

Bone tumors in children occur predominantly in the metaphyseal region, close to the growth plate, so that sacrifice of a major physis often is necessary when the tumor is excised.

Children who have primary bone sarcoma often require chemotherapy, which may have a subsequent suppressive effect on bone growth.

Limb salvage surgery for bone tumors in the immature skeleton creates unique problems.

- Maintenance of limb length after resection of one or more major growth plates
- High functional and recreational demands of young patients, which require a durable reconstruction
- At the knee, a constrained endoprosthesis is required (most commonly a fixed or rotating hinge implant), making it necessary for the prosthesis stem to breach the physeal plate on the side of the joint opposite to the tumor.
- Reconstruction with expandable endoprostheses allows the maintenance of limb length equality, allows early weight-bearing, results in predictable function, has a low risk of early complications, and is readily available.
- Disadvantages include the expense of the prostheses and the complications that are expected to increase with time in surviving patients.

### ANATOMY

About 60% to 70% of lower limb growth occurs around the knee (distal femur and proximal tibia physes), and about 80% of total growth of the humerus occurs in the proximal physis of the humerus.

Terminal branches of the diaphyseal nutrient artery form tight loops near the physis, and the epiphysis is invaded by juxta-articular vessels.

During childhood, the physis becomes an avascular structure that lies between two vascular beds, one epiphyseal and the other metaphyseal.

The epiphyseal vessels supply oxygen and nutrients; an intact epiphyseal vasculature is essential, therefore, to sustain the chondrocytes. The metaphyseal vessels interact with the physeal chondrocytes in the hypertrophic zone and must be intact to sustain normal ossification. Excessive periosteal stripping must be avoided at surgery to maintain subsequent growth.

### INDICATIONS FOR USE OF AN EXPANDABLE ENDOPROSTHESIS

- When the estimated leg-length discrepancy at skeletal maturity is more than 3 cm or when the arm-length discrepancy is more than 5 cm
- When the estimated arm length discrepancy at skeletal maturity is less than 5 cm, a prosthesis made up to 2 to 3 cm longer can be inserted. The operated upper limb initially is longer, but the opposite limb soon catches up.
- The main problem with a slight arm-length discrepancy is cosmetic.
- Problems with bimanual tasks occur only when the difference is significant.
- Patients whose estimated leg-length discrepancy is less than 3 cm can be treated with conventional “adult-type” prostheses made longer by up to 1.5 cm, and a “sliding” prosthetic component can be used across the remaining open physis.
- Girls older than 11 years or boys older than 13 years rarely require expandable prostheses, because the estimated growth discrepancy after these ages is less than 3 cm (FIG 1).

### IMAGING AND OTHER STAGING STUDIES

- Pediatric patients with suspected malignancy require the usual staging imaging studies (ie, plain radiograph and MRI scan of affected bone, chest CT scan, and isotope bone scan).
- In addition, they require:
  - Measured full-length radiograph of the affected and contralateral limb (FIG 2).
  - Hand radiograph to estimate bone age based on Greulich and Pyle’s atlas.
- The estimated limb length discrepancy at skeletal maturity traditionally has been estimated using the charts devised by Andersen and Green or Pritchett for upper and lower extremities.
- Recently, the validated multiplier method has been shown to be a simple and accurate predictor of discrepancy. It can be computed using chronologic, not bone, age, and requires only a single measurement.

### SURGICAL MANAGEMENT

In our center, the most common sites for the use of expandable endoprostheses are the distal femur (52%), proximal tibia (24%), proximal humerus (10%), and proximal femur (6%).

Surgical techniques for excision of the sarcoma are similar to those for adult tumors, which are discussed in later chapters (ON-8, ON-10, and ON-24–26). This chapter deals primarily with factors that must be considered with the use of expandable prostheses.
Part 4 ONCOLOGY • Section I SURGICAL MANAGEMENT

We currently use two main methods of lengthening expandable endoprostheses in our center. Their advantages and disadvantages are outlined in the following paragraphs (Table 1):

- The minimally invasive expandable prosthesis has been in use since 1993. It is lengthened using a worm drive mechanism (FIG 3A,B). The mechanism is encased within the prosthesis shaft, and the telescopic implant is extended using an Allen key. The operative technique for lengthening is described later in this chapter.

- The noninvasive expandable prosthesis has been in use since 2002. Surgery is not required to lengthen the prosthesis. A sealed motor unit inside the prosthesis contains a powerful magnet that can be activated by an external power source (e.g., a rotating electromagnetic field). This causes the magnet to turn, and the motor works using a very-low-ratio gearing system (13061:1) to lengthen the prosthesis. The rate of lengthening is directly proportional to the length of time that the power source is applied: lengthening of 4.6 mm takes 20 minutes (FIG 3C-E).

- The physis on the opposite side of the joint can be either preserved using a “sliding” prosthesis or sacrificed and replaced with a fixed cemented prosthesis.

- The sliding component is an uncemented, smooth component placed through a canal made centrally in the remaining preserved physis. In larger children, it is fitted inside a plastic sleeve inside the bone, which acts as a centralizer.

- This sleeve allows the component to slide inside the bone as the remaining open physis grows (FIG 3F,G).

- Care must be taken to minimize damage to the proximal growth plate by avoiding excessive periosteal stripping and carefully drilling out a cylindrical hole in the bone and, preferably, the center of the physis.

- Insertion of the sliding component destroys no more than 13% of the growth plate in the distal femur and proximal
Table 1

<table>
<thead>
<tr>
<th>Advantages</th>
<th>Minimally Invasive Prosthesis</th>
<th>JTS Noninvasive Prosthesis</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>• Prosthesis relatively inexpensive ($14,100)</td>
<td>• No surgery required for lengthening</td>
</tr>
<tr>
<td></td>
<td>• Can have subsequent MRI scans</td>
<td>• No risk of infection</td>
</tr>
<tr>
<td></td>
<td>• Reliable, with published long-term results for all sites (used since 1993)</td>
<td>• No anesthesia risk</td>
</tr>
<tr>
<td></td>
<td>• Available in uncemented versions</td>
<td>• Reduced scarring</td>
</tr>
<tr>
<td></td>
<td>• Can revise easily to another expandable prosthesis without disturbing bone implant interface</td>
<td>• Painless</td>
</tr>
<tr>
<td>Disadvantages</td>
<td>• Requires percutaneous operation to lengthen</td>
<td>• Outpatient procedure with reduced hospital costs</td>
</tr>
<tr>
<td></td>
<td>• Increased risk of infection</td>
<td>• Prosthesis is expensive ($26,500)</td>
</tr>
<tr>
<td></td>
<td>• Increased anesthesia risk</td>
<td>• Cannot have subsequent MRI scans (will damage magnet of both prosthesis and MRI)</td>
</tr>
<tr>
<td></td>
<td>• Requires day case admission with increased hospital costs</td>
<td>• Recent advance; therefore, no long-term results available</td>
</tr>
<tr>
<td></td>
<td>• Scarring with slight pain after procedure</td>
<td>• Not available in uncemented version (forceful impaction damages motor)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Currently unable to exchange lengthening module without removing whole prosthesis (development in progress)</td>
</tr>
</tbody>
</table>

FIG 3 • Minimally invasive prosthesis showing the port used for lengthening (A) and demonstrating the worm drive mechanism (B). C. Internal design of the shaft of the JTS noninvasive prosthesis (Stanmore Implants Worldwide). D. Gearbox and magnet component of a noninvasive prosthesis. E. Patient undergoing lengthening using an electromagnetic coil. Lengthening of 4.6 mm will take 20 minutes but can be done in the outpatient department. F. Schematic illustration of a sliding component in the proximal tibia. The proximal tibial physis is preserved, and the uncemented smooth prosthesis slides within the polyethylene sleeve as the physis grows. G. Illustration of the amount of growth of the proximal tibial physis 6 years after a distal femoral replacement with a sliding tibial component. The amount of growth can be seen by the growth arrest line formed at the time of chemotherapy. Growth on the affected side is only slightly less than that on the normal side. H. The use of hydroxyapatite-coated collars has been shown to reduce the rate of aseptic loosening by encouraging bone ongrowth.
tibia. There is no correlation between the surface area destroyed and continued growth of the physis.\(^3\)\(^,\)\(^4\)\(^,\)\(^7\)
- Animal models of transphyseal pediatric anterior cruciate ligament reconstruction have shown similar results.\(^2\)
- Physis with “sliders” grow at a slower rate, achieving about 80% of normal growth in the proximal tibia and about 60% of normal growth in the distal femur compared with the contralateral limb.\(^4\) (Fig 3G).
- Other methods of lengthening prostheses are in use worldwide.
  - A device developed by Kotz allows the prosthesis to be lengthened by a ratchet system that uses knee movement to cause it to lengthen.\(^9\)
  - The Phenix system (Phenix Medical, Paris, France) is a noninvasive system that relies on a coiled spring located inside the prosthesis, contained within a shield of wax. When a power source is applied to the prosthesis, the wax melts and the spring extends. When the power source is removed, the wax solidifies and the spring is fixed in its new position.\(^1\)\(^2\)
- The following factors are important when using expandable prostheses:
  - Resect the tumor with a wide margin and divide the bone at the predetermined level of transection.
  - When an expandable replacement is being used, plan to replace the exact amount of bone that has been removed.
  - If cement is being used, the intramedullary canal must be prepared adequately. We advocate the use of antibiotic-impregnated cement.
  - The use of hydroxyapatite-coated collars and preservation of periosteal sleeve significantly reduces the long-term risk of aseptic loosening by encouraging bony ingrowth to the prosthesis and also avoids the risk of stress shielding seen in some uncemented types of implants (FIG 3H).

Preoperative Planning
- Calculate estimated limb-length discrepancy at skeletal maturity
- Allow for reduced growth from any growth plates disturbed by surgery.
- Consider whether there any surgical options other than an expandable prosthesis are feasible. These include:
  - The use of a shoe lift if the difference is less than 2 cm
  - Inserting an adult prosthesis (with or without a sliding component) longer than the resected bone at the time of initial surgery.
- Planned epiphyseodesis of the opposite limb
- Decide whether to use an invasive or noninvasive prosthesis, based on factors described previously (Table 1)
- Accurate measured radiographs of the bones to be resected are sent to the engineers, together with clear information about the planned level of transection of the bone (from MRI scan or, in complex cases, cross-sectional imaging of the resection level).
- Screen and treat patients for potential infective foci: dental hygiene review; methicillin-resistant Staphylococcus aureus (MRSA) screening; inspection for common sites of infection such as central venous lines, throat, ingrowing toenails, and fungal skin infections.
- Ensure adequate neutrophil and platelet counts prior to surgery if patient has had recent chemotherapy (our unit requires a neutrophil count greater than 1000/mm\(^3\) and platelet count of 75,000/mm\(^3\) or higher).

Positioning
- Positioning is chosen according to the usual technique and approach that the surgeon is familiar with for adult prostheses.
- We favor double skin preparation with chlorhexidine followed by an alcohol-based solution. The limb should be draped in such a way as to be left free and mobile during the procedure.
- The standard positions we use for common sites of reconstruction are as follows:
  - Distal femur: supine with removable sterile leg support
  - Proximal tibia: supine with removable sterile leg support
  - Proximal humerus: “beach chair” position, with arm supported on small side table and head turned away supported on head ring
  - Proximal femur: Lateral position

Approach
- Resection of the tumor should be carried out by the usual technique and approach that the surgeon is familiar with for adult prostheses.
- We favor an anteromedial approach to tumors around the knee. We routinely open the knee joint and reflect the extensor mechanism laterally unless there is evidence of frank knee joint invasion (which is a relative contraindication to limb salvage).
- If the knee joint is involved, consider an extra-articular resection if sufficient soft tissue will be left to cover the prosthesis.
- For tumors around the hip we favor a direct lateral approach and for tumors of the proximal humerus we favor the expansive approach of Henry.

SURGICAL TECHNIQUE FOR IMPLANTATION OF EXPANDABLE PROSTHESES

- Surgical technique for the implantation of expandable prostheses is described for each common anatomic site—
  - in greater detail for the distal femur, but with only the salient specific points for each of the other sites.
TECH FIG 1 • Operative technique series for the distal femur. A. MRI scan of distal femoral osteosarcoma. Although the tumor appears to stop short of the physis, subperiosteal extension is present below the physis. The proximal limit of tumor will be more clearly visible on T1-weighted images. B. Anteromedial approach to the distal femur, showing excision of the biopsy tract and incision of the tendon of the rectus femoris. C,D. Dissection through the knee joint, dissecting the popliteal vessels from the posterior femur. E. Resected tumor with components. F. Tibial plateau resected 10 mm below joint line and perpendicular to the ankle joint. A central hole is reamed carefully to accept a sliding component. G,H. A polyethylene sleeve is inserted, uncemented, into the tibial plateau, and the metallic stemmed tibial implant is trialled. I. The distal femoral component is cemented into place and the two prostheses secured with the bushes. J. The range of flexion is tested on the table. K. The resected specimen showing the extent of the tumor, with closest margin indicated by the inked circle. Following neoadjuvant chemotherapy 98% of the tumor was necrotic.
If the deltoid is damaged in any way, we often use the medial gastrocnemius flap in place of the rotator cuff to prevent skin necrosis.

Great care must be taken to minimize damage to the proximal humerus head when carrying out lengthening procedures. Limb salvage with an expandable humeral head has a high failure rate, with a significant risk of late head subluxation in children.

Attempts are made to preserve the coracoacromial ligament to reduce the risk of proximal subluxation with lengthening.

Care must be taken to prevent proximal migration of the humeral head when carrying out lengthening procedures.

Proximal Femoral Expandable Prosthesis

Inserting an expandable prosthesis into the proximal femur is a challenge, because the hip abductors must be detached from the greater trochanter. They can be reattached to the fascia lata with the leg in slight abduction, which results in reasonable abduction power.

The type of femoral head to use is the subject of ongoing debate.

Uni- or bipolar femoral head replacements are used most frequently.

Both have high failure rates, with a significant risk of late head subluxation in children.

Small-sized femoral heads have a significant risk of dislocation.

Consider a large bearing total hip arthroplasty once the patient has reached skeletal maturity. We currently favor a large bearing metal-on-metal articulation for the increased wear characteristics and reduced dislocation rates.

Proximal Tibial Expandable Prosthesis

The proximal tibia is a challenging site for limb salvage, with an above-average incidence of complications.

The tibial osteotomy should be done perpendicular to the long axis of the tibia so as to be parallel with the ankle joint, removing 1 cm of bone from the proximal tibia.

A trial reduction should be performed to check the soft tissue tension, because acute over-lengthening may cause neurologic impairment and fixed flexion deformities, with subsequent stiffness.

Once the prosthesis is cemented into place, the site of the screw mechanism can be marked on the skin by a stab incision, to make subsequent percutaneous lengthening easier if a minimally invasive prosthesis is being used.

The skin is closed in layers over a drain, and dressings are applied.

Percutaneous Lengthening for Minimally Invasive Prostheses

The patient is placed in the supine position with access to the lengthening port.

In older children, the lengthening can be performed under a local anesthetic, but general anesthesia is preferred in younger children.

The procedure is performed under radiographic control to keep the incision as small as possible.

After double skin preparation and intravenous antibiotic prophylaxis, a stab incision is made over the jack point down to the prosthesis cavity.

The jack point is identified radiologically, and the Allen key engaged into the mechanism.

Soft tissue occasionally may need to be cleared from the jack point with a small periosteal elevator.

The screwdriver is rotated to lengthen the prosthesis: 10 revolutions of the screw driver lengthen the prosthesis by 0.1 cm; therefore, 100 revolutions are required to lengthen by 1 cm.

Intermittent single-shot images are taken to confirm lengthening.

Most lengthening procedures are done 10 mm at a time. If lengthening of much more than this is attempted, it can lead to complications such as the development of a fixed flexion deformity or occasionally a neuropraxia (eg, a foot drop).
PEARLS AND PITFALLS

Growth potential
- Adequately estimate expected growth potential.
- If family members are very tall, the use of standard growth charts may not be adequate.
- Do not forget that chemotherapy causes a delay of normal growth.
- Is it possible to use a longer adult prosthesis and slider?

Soft tissue handling for sliders
- Avoid excessive surgical insult to the physis and periosteum.

How much to lengthen?
- Usually 10 mm will be sufficient. More may cause stiffness and neurologic compromise.

How often to lengthen?
- Most patients notice when leg-length discrepancy reaches more than 15 mm.
- Children receiving chemotherapy rarely need lengthening.
- Children who are having a growth spurt may require lengthening every 6 weeks.

Joint subluxation
- Proximal humerus: use Mersilene mesh as pseudocapsule
- Proximal femur: convert unipolar head to total joint replacement when triradiate cartilage is fused

Pathological fractures
- Increased risk of femoral fracture above sliding component with tibial prostheses
- Internal fixation is best treatment
- If prosthesis is loose, revise to longer stem, bypassing the fracture at time of internal fixation

Infection
- Dreaded complication of minimally invasive prostheses
- Recorded infection rate is 1% per lengthening procedure
- Noninvasive procedure should reduce risk of infection.
- Treat acute infection with washout and 6 weeks intensive antibiotics. Chance of cure is only 20%, however.
- Two-stage revision is needed for chronic infection

Fixed flexion deformities
- Joint stiffness can be a problem after lengthening, especially with tibial prostheses.
- Try to lengthen in smaller increments and more frequently than used to correct large limb discrepancies.
- May be due to low-grade infection

POSTOPERATIVE CARE
- We advocate 24 hours of intravenous broad-spectrum prophylactic antibiotics postoperatively.
- We advocate early removal of surgical drains (within 48 hours).
- Patients with distal femoral replacements:
  - Are allowed to mobilize partial weight bearing at 48 hours
  - Are begun on both active and passive knee exercises
  - Usually can achieve active straight leg raise by day 5 and knee flexion to 90 degrees within 10 days prior to discharge.
- Patients with proximal tibial replacements:
  - Are allowed to mobilize partial weight bearing at 48 hours but can wear an extension brace to protect the extensor mechanism for 4 weeks
  - During that time they are allowed to flex to about 45 degrees but are not permitted to perform active knee extension.
- Patients with proximal femoral replacements are kept on bed rest with the leg abducted for 5 to 7 days before getting up, after which they are permitted partial weight bearing for 6 weeks.
- Patients with humeral replacements are kept in a sling for 6 weeks but with active exercises of the elbow, wrist and hand.
- At 6 weeks, all patients start intensive physical therapy and hydrotherapy, at which time they commence full weight bearing and active exercises to maximize functional recovery.

Patient Information
- Warn patients that they have an artificial implant, which can fail.
- Caution them to avoid contact sports.
- Walking, swimming, cycling and other non-contact sports are encouraged.
- Any infective process in the body can lead to infection of the prosthesis, and early treatment with antibiotics is recommended.
- Antibiotics for prophylaxis during dental procedures is required only if infection is present.
- Patients with noninvasive endoprostheses cannot have an MRI scan.

OUTCOMES
- Over the past 30 years our unit has operated on 615 children under the age of 16 with primary malignant bone tumors.
- 74 patients (12%) required amputation and the remainder (408 patients) had limb salvage using a prosthesis.
- Of the 176 patients with an expandable prosthesis, 117 are still alive and 89 have reached skeletal maturity.
- 60 patients never had a lengthening procedure carried out, either because they developed recurrent disease (metastases or local recurrence) or a complication such as infection.
- 116 patients had one or more lengthening procedures, with a mean of 5.3 lengthening procedures per patient (range 0–17 procedures) with an average total length increase of 32 mm (range 0–120 mm).
- Nineteen patients needed an amputation, either because of local recurrence (11 cases) or infection (8 cases).
- The overall limb salvage using Kaplan Meier survival curves was 83.9% at 20 years from insertion.
FIG 4 • A, B. Aseptic and rotational loosening in the distal femur after 14 years. C. Acute shortening due to displacement of a lengthening ring medially, which required revision to an adult prosthesis. D. Hip subluxation has been a problem in young patients receiving proximal femoral replacements.
COMPLICATIONS

Infection
- Deep prosthetic infection is a major concern in children who have expandable prostheses because of the need for multiple operative procedures.
- The cumulative risk of infection in our series was 21% at 10 years.
- It was related to site (proximal tibia) and previous highly invasive prosthesis designs.
- The use of gastrocnemius flaps and minimally invasive implants has reduced the infection rate to 8% at 10 years.
- The risk of infection has decreased from 3% per lengthening to about 1% with minimally invasive prostheses.
- Noninvasive prostheses should decrease the risk of infection over time.

Loosening
- The use of hydroxyapatite collars has significantly reduced the incidence of aseptic loosening. Revision usually is fairly straightforward, and the prosthesis is changed to an adult model (FIG 4A,B). Always consider the possibility that the loosening may be caused by low-grade infection.

Unplanned Shortening or Lengthening
- An unusual complication due to mechanism failures, which usually requires revision of the implant (FIG 4C).

Stiffness
- Stiffness is a common problem in younger children with prostheses around the knee, or if the prosthesis inserted is longer than the resected bone to try to gain some extra length. In some children, excessive scar tissue builds up around the prosthesis; in such cases, removal of the scar tissue can be helpful, combined with intensive physical therapy. In cases of fixed flexion deformities, intensive physical therapy, including the use of serial plaster casting, may be useful.

Subluxation of Hip or Shoulder
- Subluxation at the shoulder can be reduced by the use of Mersilene mesh. Femoral head subluxation is far more of a problem in younger children with proximal femoral replacements. We have found that in children under age 12 there is an increasing tendency for the superior margin of the acetabulum not to develop properly, and in these children the femoral head will sublux. We have tried several techniques to prevent subluxation, without success, and now revise the unipolar head to a large bearing surface uncemented cup when the triradiate cartilage is fused or subluxation is apparent (FIG 4D).

Outgrowing the Available Extension
- The maximum lengthening available in a prosthesis is 120 mm, but in many cases less than that is needed. Revision usually involves replacing only one component of the prosthesis.

Implant Breakage
- Implant breakage is rare in patients who have reached maturity with a child’s prosthesis still in place. The most common site for a fracture is at the junction of the thinner lengthening portion with the main component. Revision is required in all cases.

Periprosthetic Fractures
- Periprosthetic fractures are rare, but there does seem to be an increased risk of femoral fractures above a sliding femoral prosthesis used in conjunction with a proximal tibial growing prosthesis.

REFERENCES