



FIGURE 1. Anteroposterior radiograph of a trochanteric entry femoral intramedullary lengthening nail in a 9-year-old girl with a distal femoral growth arrest. She has consolidated her 5-cm femoral lengthening.

initially until the trochanter location is palpated with the tip of the guidewire. Once the location of the trochanter is known, the guidewire can be carefully walked more medially to the desired entry point. This technique will help to avoid inadvertent passes of the sharp guidewire in the vicinity of the vessels along the femoral neck. Use the smallest diameter entry reamer possible to avoid inadvertently making a large hole in the wrong location. Once the reaming begins, try to steer the reamer laterally in the greater trochanter to avoid cutting out the trochanter medially.

Premature Consolidation

Because the endosteal blood supply is disturbed by the reaming process during the intramedullary lengthening nail insertion, the periosteal blood supply is critical to the formation of regenerate bone. Pediatric patients have thicker, more well-developed periosteum compared with adult patients. Therefore, children have the potential to make new bone

using intramedullary lengthening nails more reliably and more quickly than adults.¹² In general, pediatric patients require a shorter latency period and a faster rate of lengthening than adult patients to minimize the risk of premature consolidation. If the regenerate bone heals too quickly, it creates increasing resistance to distraction which ultimately slows and eventually stops the intramedullary nail from further lengthening. To avoid this complication, pediatric lengthening patients need to be followed closely. A weekly assessment of the radiographs is recommended. Based on the regenerate formation seen each week, the rate and rhythm of the lengthening process can be continuously adjusted faster or slower through the external remote controller depending on the need. Blindly lengthening at the same rate for the entire distraction phase can result in poor results either from regenerate that heals prematurely or from regenerate bone that is structurally deficient. Instruct the patient's family to bring the external controller to the clinic visit each week so that it can be reprogrammed to a faster or slower rate as necessary.

Unique Issues in Congenital Conditions

Intramedullary limb lengthening in patients with a congenital limb length discrepancy (ie, congenital femoral deficiency or fibular deficiency) presents several challenges. First, some of these patients will present with large projected leg length discrepancies at skeletal maturity. Consequently, a life plan needs to be formulated for each patient and family.⁴ The plan should balance the desire to achieve equal limb lengths safely with the aim to accomplish this goal in the minimum number of surgeries over the child's lifetime. In most cases, accomplishing this task will require multiple planned (and some unplanned) surgeries. When devising the life plan, it is important to spread these surgeries out as much as possible. Maintaining several years between surgeries allows the child to avoid spending their entire childhood in the hospital or a doctor's office. These interval periods of no treatment are critical to the social, psychological, and cognitive development of the child.

Second, before any lengthening is considered in congenital patients, the hip, knee, and ankle joint stability of the limb needs to be evaluated. A thorough physical and radiographic examination is necessary to determine the range of motion, morphology, and stability of each joint. Many patients will have deficient cruciate knee ligaments, joint contractures, and/or acetabular dysplasia as part of their congenital deficiency. These findings can cause the hip and knee joints to be at risk for subluxation or dislocation during the femoral lengthening process. Preparatory surgery, such as ligament reconstruction, muscle releases, and osteotomies, should be performed as the first step to improve the stability of the joints⁴ (Figs. 2A, B). Many congenital patients also have a concomitant angular deformity that should be addressed either before lengthening or simultaneously. For example, a valgus knee with patellar instability that will be exacerbated by lengthening can be improved by restoring a normal mechanical axis.^{13,14} Once the adjacent joints are considered stable, then the lengthening process can begin. Although the concept of preparatory surgery is important in all pediatric patients undergoing lengthening, it is especially critical in patients undergoing lengthening with intramedullary lengthening nails. Unlike external fixator constructs that can be built to span an unstable joint, intramedullary lengthening nails do not inherently allow the surgeon to protect a compromised joint. Maximizing the joint stability before lengthening in conjunction with close monitoring of the joint during the lengthening process is advised when using an intramedullary lengthening nail.

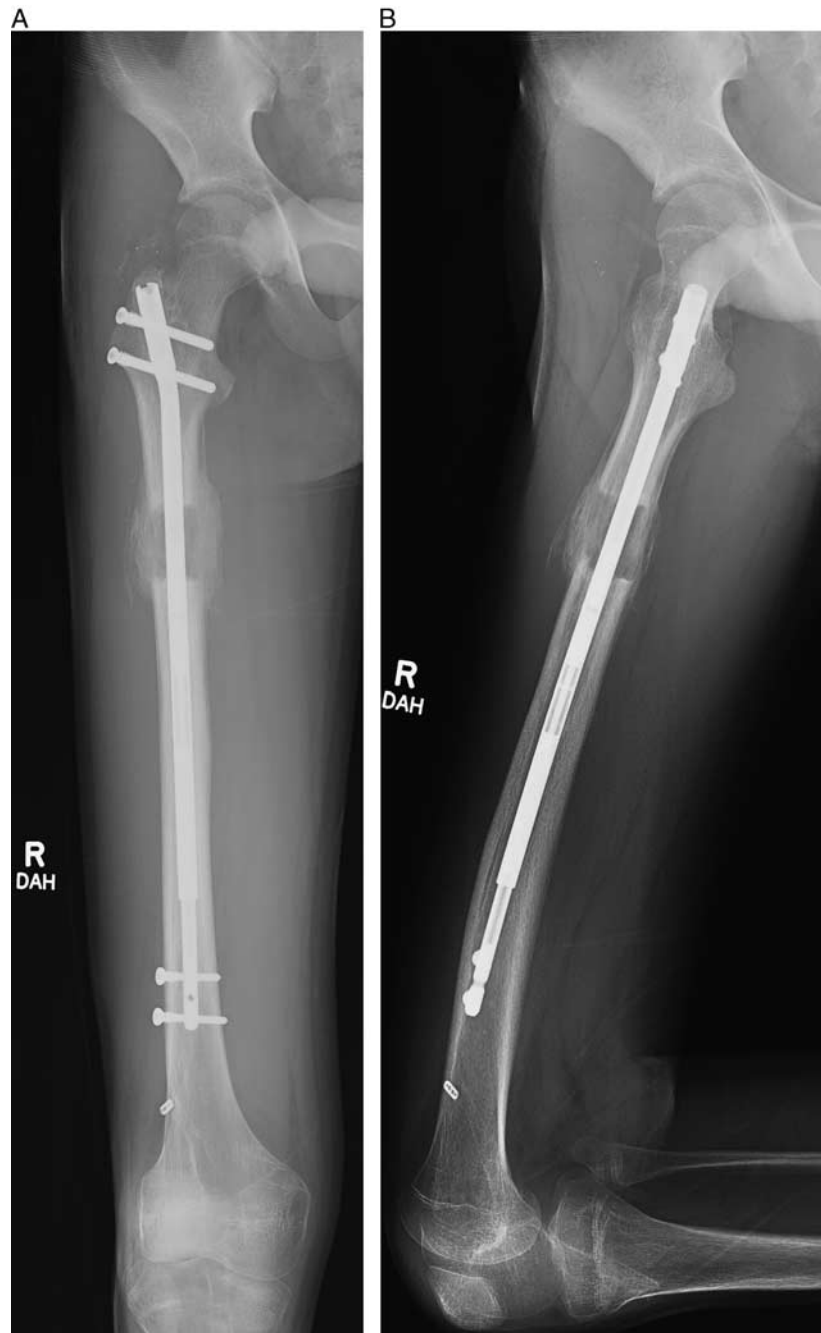


FIGURE 2. A and B, Anteroposterior and lateral radiographs of a 14-year-old boy with fibula hemimelia who has reached his target lengthening goal of 3.5 cm. Note that his anterior cruciate ligament was reconstructed before lengthening to protect his unstable knee (endo button from ligament reconstruction visible on the antero lateral distal femur).

Third, the limbs of congenital patients are not genetically programmed to be of normal size. Consequently, the soft tissue envelope is more resistant to lengthening than in patients with developmental causes of leg length discrepancy.¹⁵ This situation creates additional concerns that need to be recognized during the lengthening process. Because tight tissues will only get tighter during lengthening, the surgeon should carefully evaluate the range of motion of the limb at each joint and plan to address any contractures that are found. For example, if the quadriceps are tight, a release of the direct head of the rectus

femoris may be needed. Similarly, a gastrocnemius recession should be considered for tibial lengthening patients. For all congenital femoral lengthenings, a release of the iliotibial band is recommended.¹⁵ An additional subtle but important detail is that the lengthening rate and rhythm should be slower than a noncongenital patient. More frequent but smaller increments of lengthening (ie, 0.12 mm six times/day instead of 0.25 mm three times/day) may help to make the lengthening proceed more smoothly. Careful evaluation of the radiographs, especially the lateral of the knee, is required at each visit to monitor

for knee subluxation. If the joint range of motion is found to be decreasing, then the lengthening should be slowed or stopped until the range is recovered.

Fourth, from a practical standpoint, congenital patients tend to have small diameter and length of the lower extremity long bones. Consequently, intramedullary lengthening nails may be more difficult to use in these patients. After the preparatory surgery has been completed, the first lengthening can occur at as early as ~4 years of age. Because the bone diameter and length are too small for an intramedullary lengthening nail at this age, an external fixator or an extramedullary lengthening nail may be necessary to accomplish for the first lengthening. (See the separate article on extramedullary lengthening in this journal issue for more details about this technique.) Subsequent lengthenings may use intramedullary lengthening nails if the surgeon feels comfortable placing a trochanteric entry femoral nail in a skeletally immature patient.

Fifth, because many congenital patients have had a previous lengthening using an external fixator, there is a slightly increased risk of implant infection from colonized previous pin sites.¹⁶ If the patient had a history of pin site infections during the previous lengthening, then a STIR MRI (short-tau inversion recovery magnetic resonance imaging) of the femur or tibia may be warranted to evaluate the pin sites before using an intramedullary lengthening nail. Curettage of the affected pin sites may be necessary as a separate procedure before the introduction of the intramedullary device.

Finally, because many congenital patients require multiple lengthenings over their lifetime, the surgeon needs to decide whether to attempt to reuse the same implant or exchange it for a new one each time. Although the industry recommendation is to avoid using the same implant 2 times, there are documented cases in which this has been done successfully.¹⁷ The 2 main advantages of reusing the same implant are: (1) a cost savings to the patient if the same lengthening implant can be safely reused and (2) potentially less surgical morbidity to the patient when reusing the same implant compared with removing and inserting an entirely new device. In the past, the amount of time required to compress the nail back to its original length made this option prohibitive. However, there now exist methods to rapidly reduce the nail length so that nail compression can be done in a reasonable amount of time during one operating room setting. If reuse is considered, it is always a good idea to have a new nail available as a backup in case it is discovered that the existing nail is not working properly.

Combination With Guided Growth

Many pediatric patients with limb length discrepancy have a concomitant angular deformity. In skeletally immature patients, guided growth techniques such as tension band plating or percutaneous transphyseal screws can be used to help correct coronal plane angular deformity.^{18,19} Because the growth tethering gradually corrects the bone alignment back towards neutral without the need for an osteotomy, guided growth is the ideal method to correct distal femoral deformity in conjunction with a trochanteric entry antegrade femoral lengthening nail. For example, in a skeletally immature patient with distal femoral valgus and a leg length discrepancy, a guided growth implant can be used to tether the medial distal femoral physis in combination with a trochanteric entry antegrade intramedullary lengthening nail insertion (Figs. 3A, B). This allows simultaneous gradual correction of the angular deformity and the leg length discrepancy through a single osteotomy. Because the intramedullary nail dictates that the osteotomy location is diaphyseal or

meta-diaphyseal, it is difficult to perform acute correction around a trochanteric entry antegrade femoral nail.

The alternative to guided growth in patients close to skeletal maturity is to perform the lengthening and the deformity correction through 2 separate osteotomies in stages or to combine the 2 by performing the distal angular correction with a plate and the lengthening with a short antegrade nail. When combining guided growth with the intramedullary lengthening nail, the surgeon needs to plan the position of each implant so that the guided growth implant does not interfere with the path of the nail's distal interlocking pegs. Using a shorter nail length that stops short of the guided growth implant is the easiest solution. If there is not enough femoral length to use a short nail, then the guided growth implant can be inserted after the nail is in place with the screw(s) directed away from or between the distal interlocking pegs.

Tips for Success

There are several key elements that, if followed, increase the chances for a successful pediatric intramedullary lengthening. The first component involves proper patient selection. The management of a pediatric intramedullary limb lengthening patient can be a complex process in the best of circumstances. All patients undergoing pediatric limb lengthening require a strong family support system and the ability to diligently follow the recommended postoperative plan. Performing this procedure on a pediatric patient whose family does not have adequate resources will add additional stress that can affect the postoperative outcome. Patients with a history of anxiety or depression should have an evaluation by a behavioral health specialist before surgery.²⁰ In addition, the surgeon should make an honest assessment of his/her limb lengthening experience and gradually work up to more difficult cases. Scheduling a complicated case to be your first intramedullary limb lengthening experience is not recommended.

The second element involves meticulous preoperative planning. As previously mentioned, an assessment of the adjacent joint range of motion and stability is mandatory. A standing full-length lower extremity radiograph should undergo a comprehensive deformity analysis. Dedicated anteroposterior and lateral radiographs of the bone in question must be obtained to ensure that the implant will fit properly and the surgery can be performed safely. The level of the planned osteotomy must be in the proper location and allow smooth passage of the implant into the canal. Any postoperative immobilization devices (ie, dynamic knee extension brace, ankle-foot orthosis) should be fabricated before the surgery date.

The third element concerns the use of proper surgical techniques intraoperatively. The surgeon should closely follow the specific device's surgical technique guide and avoid attempting to cut corners. The osteotomy should be performed with minimal soft tissue injury and minimal heat. The venting holes must be placed before reaming. Patient positioning is an under-rated but critical component of the case. The patient should be lying as close to the edge of the bed as safely possible. Positioning the ipsilateral arm over the chest and placing a bump under ipsilateral the hemipelvis allows access to the proximal femur. Be careful not to place the drapes over the gluteal area that may obstruct placement of the nail insertion incision. When inserting the implant, the surgeon should be able to push it into the canal by hand without the need for forceful blows from a mallet.

Finally, the last element involves close monitoring of the patient throughout the distraction phase. After a typical latency period of 5 to 7 days, the rate and rhythm of the

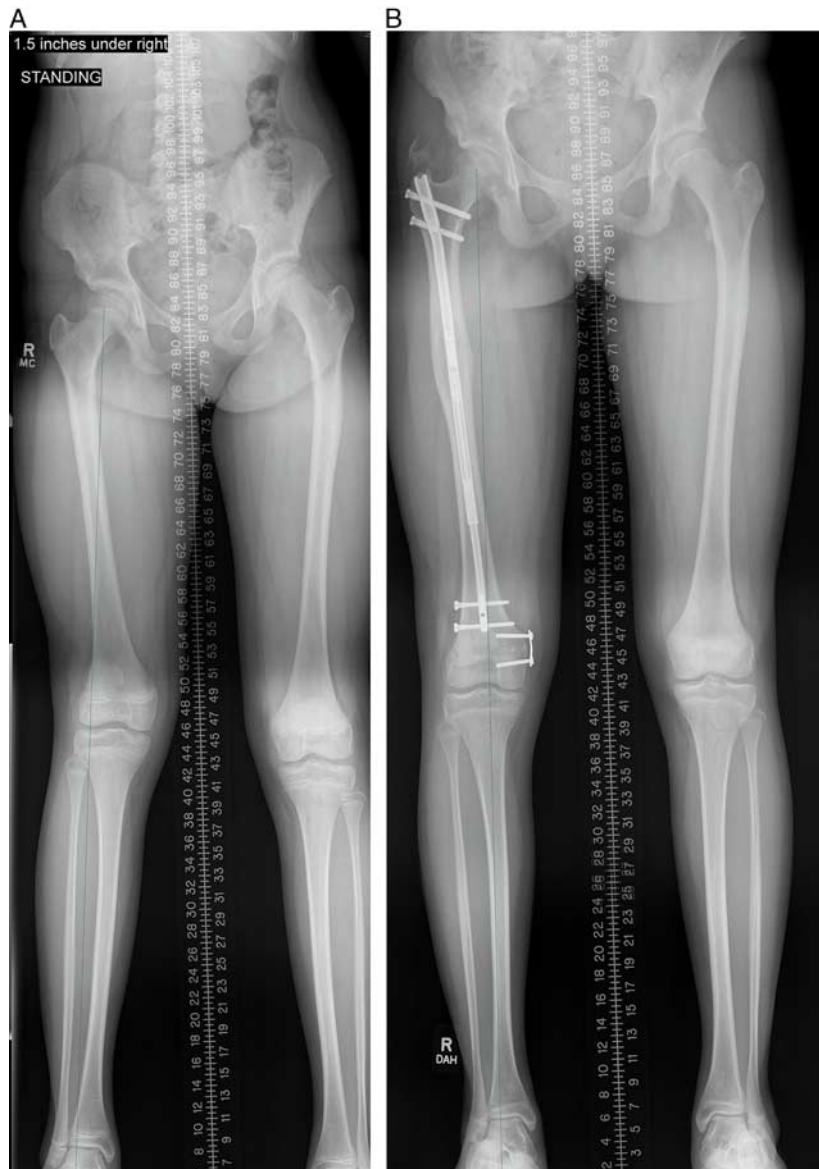


FIGURE 3. A, Preoperative standing anteroposterior bilateral lower extremity radiograph of a 13-year-old girl with genu valgum and a 4.5-cm leg length discrepancy. B, Postoperative standing anteroposterior bilateral lower extremity radiograph demonstrating correction of the limb lengths and mechanical axis using an intramedullary lengthening nail and medial distal femoral tension band plate.

femoral distraction need to be determined by the surgeon. In general, the femoral lengthening rate and rhythm should start slowly (ie, 0.75 mm/d) to ensure that there is healthy initial regenerate bone formation. Once there is confirmation that the regenerate bone is forming well, the lengthening rate can be increased if necessary.¹² Modulation of the lengthening rate and rhythm should be performed on the basis of the weekly radiographic findings. As pediatric patients tend to make the bone more quickly than adult patients, a weekly examination is essential to decrease the risk of premature consolidation. The joint range of motion should also be assessed and recorded each week. If there is clinical evidence of a decreasing range of motion, then the lengthening rate should be slowed down. If there is a decreasing range of motion and radiographic signs of joint subluxation, then the lengthening rate should be stopped immediately.

Avoiding Pitfalls

There are 3 potentially catastrophic pitfalls in pediatric intramedullary limb lengthening. The first is the development of femoral head avascular necrosis. The best option to avoid this complication is to wait until the patient has reached skeletal maturity before performing an intramedullary lengthening or to use an external fixator. As this approach is not always practical, especially in a congenital patient with a large discrepancy, meticulous surgical technique is required when inserting intramedullary lengthening nails in skeletally immature patients. There should be no false passes with the sharp tip of the guidewire into the vicinity of the femoral neck. The final position of the guidewire in the tip of the trochanter needs to be clearly visible in orthogonal planes before starting to ream.

The second potentially catastrophic pitfall is the creation of a radiographic “black hole” at the distraction site. Early

regenerate bone should be visible on both the anteroposterior and lateral radiographs by 2 to 3 weeks of lengthening. If there is no discernable regenerate bone formation at that point, then the lengthening should be stopped or reversed and restarted. Do not keep lengthening in the presence of a “black hole” hoping that it will get better. The empty space is an indication that the proper biological and mechanical environment for bone formation is not present (ie, lengthening too fast, latency too short, etc.). If you have created a large distraction gap without the presence of discernable regenerate bone, then autogenous bone grafting of the defect may be necessary. If you have flimsy regenerate bone present, then exchanging the device for a trauma nail can help stimulate healing by allowing increased weight bearing and by reaming through the regenerate before inserting the new nail.²¹

The third pitfall to avoid is producing joint subluxation or joint dislocation during lengthening. A thorough assessment of the adjacent joint stability should be performed preoperatively. If the joint is deemed to be unstable, then surgical intervention to correct it should be performed before embarking on intramedullary lengthening. It is crucial to closely examine every postoperative radiograph for subtle changes in joint alignment, especially on the lateral view of the knee. Femoral lengthening patients should use a dynamic knee extension brace 12 to 16 hours per day to help maintain full knee extension throughout the lengthening process.²² Decreases in joint range of motion are usually the first clue that there is stress on the joint. The patient may also begin to complain of knee pain. A lateral radiograph with the knee in full extension will detect subtle changes in joint alignment. The physician should react to the situation quickly by increasing the frequency of physical therapy and discontinuing any further lengthening until the joint range of motion is restored. If subluxation has already occurred, then reverse the lengthening in addition to increasing the therapy. A custom brace has been described as a nonoperative attempt to regain joint alignment.²³ Soft tissue releases may be required to achieve joint reduction if therapy and bracing are not successful.

The final pitfall to avoid is the difficult nail removal surgery. Each of the intramedullary lengthening nails should be removed from pediatric patients once the bone is fully consolidated. The typical time frame for removal is 9 to 12 months from insertion. It is important to remove the nail in a timely manner to prevent overgrowth of bone over the interlocking screws and the proximal nail end. Although endcaps for the device are available, they are generally not necessary, especially if the plan is to remove the nail within 1 year. The proximal end of the nail can be exposed percutaneously by inserting a guidewire into the proximal end of the nail under fluoroscopic guidance. A cannulated reamer equal to the proximal diameter of the nail is then placed over the wire and used to remove any bone that has grown over the end of the nail. Once the reamer is removed, it should be possible to capture the proximal end of the nail with the nail retrieval device. Removing the interlocking screws/pegs can be made easier by leaving them slightly proud of the near cortex at the time of insertion.

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