Extramedullary Internal Limb Lengthening

Claire Shannon, MD and Dror Paley, MD, FRCSC

Summary: Extramedullary implantable limb lengthening (EMILL) uses an implantable nail attached to the bone like and internal-external fixator. Cantilever forces can be neutralized by inserting a small diameter solid rod as a guide inside the medullary canal. EMILL expands the indications for internal limb lengthening to younger children with smaller diameter and length bones and to bones with impassable medullary canals. One must follow the same principles as with external fixation lengthening including prevention of joint subluxation and contracture by preparatory surgery (eg, pelvic osteotomy), soft tissue releases, temporary arthrodesis, and bracing. Lengthening should be restricted to amounts no >5 cm to avoid complications. A retrospective review of EMILL cases performed at the authors’ institution since 2015 was performed. Thirteen patients underwent 14 EMILL procedures; 10 femurs and 4 tibias. Twelve of 13 patients lengthened to within 5 mm of their preoperative goal. There were no mechanical nail failures. No patient had a significant axial deviation of the bone during distraction. Three patients required unplanned operations. EMILL is safe and effective in patients who would otherwise require external fixation.

Key Words: PRECICE—Stryde—limb lengthening—limb length discrepancy—pediatric—extramedullary—deformity correction—tibia femur.

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The Ellipse, PRECICE 2 nail was originally available in 10.7 and 12.5 mm diameters. A smaller diameter nail, 8.5 mm was developed and first implanted in January 2014 by one of the authors (D.P.). It was available in both piriformis and trochanteric entry femur nail version with the shortest femoral nail being 210 mm and the shortest tibial nail being 195 mm. There were 2 locking screw holes proximally and 3 distally. Ellipse together with one of the authors (D.P.) released a shorter version of the P2 nail with only one locking hole at each end reducing the nail length to 150 mm.

Sizing a femur for intramedullary use of these small diameter nails is based on the outer cortical bone diameter, not the inner canal diameter. This is different than the way nails are sized for fracture fixation. The narrowest outer diameter of the bone on anteroposterior and lateral views needs to accommodate the diameter of the nail plus 2 mm of additional reaming while leaving a cortical thickness of 2 to 3 mm circumferentially. For an 8.5 mm diameter nail, this means that the outer diameter of the bone must be at least between 14.5 and 16.5 mm. The youngest age that most children reach this outer diameter is 8 years old, while some do not reach this diameter until after age 10, meaning that they are not candidates for implantable intramedullary nailing until later childhood.

Lengthening plates date back to 1978 when Witt and colleagues designed a lengthening blade plate for the proximal femur controlled by a microprocessor chip. Unfortunately, this device was designed with a lengthening rate of 2 mm/day and could not be reprogrammed due to the cost. It was used both in animals and in humans.

A number of other surgeons and engineers have designed implantable lengthening devices. Robinson, a craniofacial plastic surgeon, designed a lengthening plate to be used in the ulna and femur, as well as in the mandible. It was actuated manually by a direct drive coming out the skin onto which was connected to a wrench. McCarthy designed a device for extramedullary lengthening and rib distraction that utilized radiofrequency or manual elongation.

More recently, in 2012, Paley and Pool, working with Ellipse Technologies, began the development of a magnetically driven plate based on the PRECICE mechanism. After acquiring Ellipse in 2016, Nuvasive Specialized Orthopedics continued this development resulting in the recently Food and Drug Administration (FDA) cleared Gro-Plate device (December 2019) which is pending first clinical use in 2020.

In the interim, 2 separate investigators (Dror Paley, MD, and Mark Dahl, MD12) have independently reviewed the results begun of the application of the PRECICE nail as an internal-external fixator to achieve extramedullary limb lengthening. One of the authors (D.P.) first applied it in this manner on February 5, 2015. This report is the first publication of extramedullary limb lengthening for the femur and tibia. The indications, method and results of extramedullary limb lengthening of the femur and tibia are presented.

INDICATIONS FOR EXTRAMEDULLARY LENGTHENING

Femur
(1) Age under 7 with open greater trochanteric physis.
(2) The diameter of bone <14.5 mm.
(3) Length of bone between physes at least 140 mm.
(4) Inability to place intramedullary implant due to impassable medullary canal.

Tibia
(1) Open proximal tibial physis with ankle joint present.
(2) The diameter of bone <14.5 mm.
(3) Length of bone between physes at least 150 mm.
(4) Inability to place intramedullary implant due to anatomy or retained implant.

Other Considerations

In the femur, the hip must be stable before lengthening. If the hip is unstable or inadequately covered, hip coverage should be achieved by a pelvic osteotomy before lengthening. The knee may remain clinically unstable as long as the patient has no functional instability and the knee achieves full extension. In the more unstable knees, ligament reconstruction with the SUPERknee...
procedure before surgery is advised. Soft tissue releases must be done before lengthening for all congenital cases. These rules are equally applicable to external fixation or intramedullary lengthening procedures as they are for extramedullary lengthening surgery. For developmental cases they should be done in selective cases based on muscle length tests (Ober test, Ely test, Popliteal Angle test), and degree of preoperative soft tissue tightness. This should include release of the iliotibial band and fascia lata, the biceps aponeurosis, and the rectus femoris tendon at the anterior inferior iliac spine as needed. During lengthening an HK-AFO (Hip-Knee-Ankle-Foot Orthosis) should be worn all night with the knee locked in full extension to prevent a flexion deformity and knee subluxation.

For tibial lengthening, stability of the ankle is important. All fibular hemimelia Paley types 2, 3, and 4 should be considered to have valgus ankle instability. The ankle should be stabilized by the SUPERankle or SHORDT procedures before lengthening if applicable. A temporary arthrodesis of the ankle joint by transarticular pins or extra-articular calcaneotibial screw can be used to prevent equinovalgus. All tibial lengthenings should wear a custom molded ankle-foot orthosis to prevent equinovalgus.

Surgical Technique: Extramedullary Internal Limb Lengthening (EMILL) of the Femur

Step 1: The patient is positioned supine on a radiolucent table. The entire lower extremity including the hemipelvis should be draped out to ensure adequate access for nail insertion.

Step 2: The osteotomy level is selected for both the tibia and fibula if it is present. In most cases, a mid-diaphyseal osteotomy is preferred. The fibula osteotomy is performed first. If a SLIM rod is used, it is inserted through a proximal starting point. The SLIM is then withdrawn and a tibial osteotomy is performed through a <1 cm incision with multiple drill holes and completed with an osteotome. The SLIM is then advanced down the tibia again. If there is room for a 4-screw nail (2 proximal and 2 distal to the osteotomy) then one may choose not to use a SLIM. In that case, the osteotomy is made later.

Step 3: The extramedullary nail can be inserted antegrade or retrograde. Retrograde insertion is easier. A 2 cm incision is made at the posteromedial portion of the tibia. Staying anterior to the deep compartment fascia, a space is tunnelled using the long drill sleeve and trochar. A docking hole is made in the proximal metaphysis by creating a hole in the metaphyseal sleeve and trochar. The drill sleeve and trochar can be used to dilate this space.

Step 4: A lateral incision is made at the level of the greater trochanter. A submuscular path is created for the nail under the vastus lateralis along the lateral border of the femur. The drill sleeve and trochar can be used to dilate this space.

Step 6: Create a docking hole in the lateral distal femoral metaphysis for the distal end of the nail. The closer the nail is positioned to the bone, the smaller the bending moment will be on the nail. Distally the nail can be docked into the distal metaphysis by creating a hole in the metaphyseal flare. Through a separate lateral incision, a wire followed by an anterior cruciate ligament reamer slightly larger than the diameter of the selected nail, such as a 10 mm reamer for an 8.5 mm nail, is used to create a 1 to 2 cm depth hole. The nail can now be advanced antegrade and nested into the distal hole. It is locked distally first and then proximally. The proximal locking guide remains on as a handle until a wire is inserted through the one proximal locking hole. The hole is drilled and the wire removed and replaced with a threaded screw. The proximal guide is removed to allow the nail to get sucked down to the bone by the lag effect of the threaded screw. If there are 4 interlocking holes then the second pair are drilled and 2 more screws added. After completing the locking, the incisions are closed (Figs. 1A, B).

Surgical Technique: EMILL of the Tibia

Step 1: The patient is positioned supine on a radiolucent table. The entire lower extremity should be draped out to ensure adequate access for nail insertion. A tourniquet is applied to the lower limb.

Step 2: The osteotomy level is selected for both the tibia and fibula if it is present. In most cases, a mid-diaphyseal osteotomy is preferred. The fibula osteotomy is performed first. If a SLIM rod is used, it is inserted through a proximal starting point. The SLIM is then withdrawn and a tibial osteotomy is performed through a <1 cm incision with multiple drill holes and completed with an osteotome. The SLIM is then advanced down the tibia again. If there is room for a 4-screw nail (2 proximal and 2 distal to the osteotomy) then one may choose not to use a SLIM. In that case, the osteotomy is made later.

Step 3: The extramedullary nail can be inserted antegrade or retrograde. Retrograde insertion is easier. A 2 cm incision is made at the posteromedial portion of the tibia. Staying anterior to the deep compartment fascia, a space is tunnelled using the long drill sleeve and trochar. A docking hole is made in the proximal metaphysis to nest the tip of the nail into the bone, as described for the femur. The nail is advanced to this hole in a retrograde fashion and locked proximally. At the distal end, a wire is inserted into the nail hole and then the nail proximal guide is removed to allow the nail to move closer to the bone. The hole is drilled and a screw inserted. The screw pulls the nail down to the bone since it acts as a lag screw. The second screw should not be inserted before the bone is broken. If a SLIM rod was used, then the tibial osteotomy is already complete. If no SLIM was used then the osteotomy is performed after drilling the hole for the screw but before inserting the screw. If there are 2 screws to insert at each end then the second pair of screws are now inserted. The tourniquet is now removed and the incisions are closed (Fig. 2).

Locking Screw Choice. The PRECICE nail offers 2 locking screw choices: (1) smooth pegs with a short threaded section near the head of the screw and (2) threaded screws with a larger diameter threaded section near the head of the screw. The larger diameter threaded section near the head of the screw was designed for fixation of the near cortex in intramedullary fixation. With extramedullary fixation it does not fit into the locking hole and therefore makes the screw head prominent. This is not as much a problem in the femur as it is in the tibia.
Another option is the new stainless steel screw for the Stryde nail. This is threaded at its distal end and again has a larger diameter thread for the near cortex. It too remains prominent and requires a step drill for insertion. It is important to use a threaded screw to prevent lateral migration of the implant, therefore either the fully threaded PRECICE titanium screws or the partially threaded Stryde screws should be used. To avoid significant screw head prominence, a metal cutting matchstick burr should be used to shave off the larger diameter threads near the screw head. This is especially important in subcutaneous locations of the screw.

Testing the Nail. In both femur and tibia applications the nail should be tested with the external remote controller device to demonstrate that it is working. This is done by distracting 1 mm in the operating room after implantation.

Postoperative Routine
All patients receive acetaminophen intravenous or oral and intravenous Toradol on a scheduled basis with breakthrough narcotics as needed for pain relief. Physical therapy starts immediately on postoperative day 1 with gentle joint motion and stretching. Braces are used for nighttime (HKAFO for the knee in congenital

FIGURE 1. A, Extramedullary internal limb lengthening technique: The SLIM rod and osteotomy are already in place. The long PRECICE drill sleeve with a trocar is used to dilate the submuscular space. The point where it contacts the lateral femoral metaphysis is marked with a wire (left). A 10 mm reamer is used to create a hole in the bone for docking of the distal portion of the nail (middle). The nail is passed into the submuscular space and inserted into the hole in the distal femur (left). B, The lateral view shows the correction of the femoral bow with the SLIM rod. The nail is intentionally located posterior to the SLIM rod. A wire is inserted into the distal femoral metaphysis and then a cannulated drill is used for the near cortex and a solid step drill for the far cortex (left). After locking distally, a wire is inserted into the proximal locking hole posterior to the SLIM rod (center left and right). The nail is affixed to the bone with Stryde screws. The larger diameter threads of the Stryde screw are shaved off to allow the nail to be less prominent as seen in the upper screw (right).
femoral deficiency and ankle-foot orthosis for the ankle for fibular hemimelia) or when the patient is just sitting or lying around. They are removed for walking and for therapy.

After discharge, formal daily physical therapy begins 5 days/week with home exercises multiple times every day. All patients with congenital limb deficiencies start lengthening on postoperative day 7 at a rate of 0.25 mm 3 times per day. This equates to 1 cm of length every 2 weeks. Clinical follow-up with anteroposterior and lateral x-rays of the affected limb are performed every 2 weeks to monitor lengthening progress.

At the completion of distraction, the patient may decrease physical therapy visits if range of motion of all joints are maintained. Monthly x-rays are obtained to evaluate consolidation. Once the bone is fully consolidated, the patient may begin full weight-bearing. The lengthening nail is left in for up to a year, or until the bone is fully consolidated.

PEARLS AND PITFALLS

- If femoral derotation is desired based on the preoperative clinical examination, Steinman or half pins and a goniometer or inclinometer should be used to ensure proper rotation correction. Correction of retroversion should only be done through a proximal to mid femoral osteotomy. Correction of anteversion can be done at any level of osteotomy.16
- Malalignment at the knee joint is best corrected gradually by guided growth using a hemiepiphysiodesis plate.
- Tibial extramedullary nails should be positioned postero medial to avoid prominence in the subcutaneous portion of the tibia.
- Temporary arthrodesis of the ankle either transarticular or extra-articular should be considered in patients who are at high risk of equinus contracture during tibial lengthening. This must be combined with bracing.15
- The SLIM rod should be left in place at the time of nail removal to prevent fracture. In the tibia, it should not be removed if the proximal physis remains open to avoid a growth arrest.

METHODS

This is a retrospective review of all cases of EILL for treatment of leg length discrepancy during past 5 years (Figs. 3A–D, 4A, B). This study was approved by the Institutional Review Board. Thirteen patients (8 male, 5 female) underwent insertion of 14 nails for EILL between 2015 and 2020. Eleven patients were skeletally immature. The diagnoses included congenital femoral deficiency (8), tibial hemimelia (2), fibular hemimelia (1), Ricketts (1), and myelomeningocele (1). The median age was 6.5 years (range: 3.5 to 20 y). Femoral lengthening was performed with 10 nails in 10 patients and tibial lengthening with 4 nails in 3 patients. One femoral lengthening and 1 tibial lengthening did not have a SLIM rod inserted.

RESULTS

The average follow-up after completing lengthening was 10 months (range: 2 mo to 5 y). Seven patients had already had the nail removed. The average lengthening amount was 48.5 mm (range: 40 to 55 mm). The regenerate bone healed in all cases. Radiographic healing was achieved in an average of 1.8 months (range: 1 to 3.75 mo). The healing index was 0.77 months/cm.17

OUTCOMES

There were 4 complications in 4 of 13 patients:1 broken screw requiring revision fixation; 2 subluxations of the hip treated by open reduction and periacetabular osteotomy, and 1 screw head that eroded through the skin but did not require any revision of the implants. There were no infections and coronal and sagittal plane mechanical axis measurements performed at initial implantation and end of lengthening showed no significant axial deviation in an undesired direction ($P > 0.05$). In 2 cases, hemiepiphyseodesis plates were used for intentional correction of a preexisting coronal plane deformity. Patients returned to previous level of function with decrease or elimination in the need for shoe lift for limb length equalization. The goal of treatment was achieved in all 13 patients.

DISCUSSION

Implantable limb lengthening is gradually replacing the use of external fixation (both monolateral and circular) for limb lengthening. It offers the advantages of reduced pain and improved mobility due to lack of tethering of skin and muscle, no pin site infections, and fewer unsightly deep scars that result from the pins. Unfortunately, implantable lengthening cannot be used in all patients. The main limitations have been the size of the bone (length and diameter) and the presence of open growth plates. Both of these factors usually relate to the age of the patient and the magnitude of the limb length discrepancy for the femur, tibia, and humerus or the caliber of the bone for the radius, ulna, metacarpals, and metatarsals.

The smallest size of currently available intramedullary lengthening nails is 8.5 mm in diameter and 150 mm in length. This limits the size of bones that can undergo intramedullary lengthening. Although there are some tricks to overcome
FIGURE 3. A, Anteroposterior, and lateral standing x-ray of a patient with right congenital femoral deficiency. The diameter of the femoral diaphysis is too narrow to accommodate an intramedullary lengthening nail, although the length of the bone is adequate. The hip is well covered and the knee was stable on clinical examination. B, Gradual (0.75 mm/d) lengthening with an extramedullary trochanteric PRECICE nail over the SLIM rod. C, Anteroposterior and lateral standing x-ray after 50 mm right femur lengthening and consolidation. D, Postoperative anteroposterior and lateral x-ray at the time of removal of an extramedullary femoral lengthening nail. Note the distal metaphyseal channel where the nail was docked into the bone.

FIGURE 4. A, A 14-year-old girl with leg length discrepancy secondary to congenital femoral deficiency and fibular hemimelia. She has a stiff knee making tibial rodding difficult. Extramedullary internal limb lengthening was chosen for this reason. Medial tibial PRECICE nail inserted along the subcutaneous border. The fibula was only fixed distally. The distal tibial osteotomy was done and fixed with a plate. To protect the ankle from going into equinu a temporary extra-articular ankle arthrodesis screw was inserted between the calcaneus and tibia. This holds the foot at 90 degrees. The temporary arthrodesis screw was removed at the end of active lengthening and the patient began active ankle range of motion. Anteroposterior and corresponding lateral views shown beneath from date of surgery (left) to during distraction (center left) to end of distraction 50 mm (center right) to bone healing after only 6 weeks of consolidation with the removal of temporary ankle arthrodesis screw to free ankle joint (left). B, Before right tibial lengthening (left). After right tibial lengthening (right). Of note is there is a slight bowing of the nail perhaps due to the usual tendency towards valgus axial deviation with tibiofibular lengthenings.
the length limitation, such as a 1 cm acute lengthening to fit the shortest available nail into the bone, this does not address the limitations imposed by small bone diameter or open physis.

The extramedullary application of lengthening nails was developed in an effort to extend internal lengthening to younger children with open growth plates whose bone caliber cannot accommodate the smallest currently available implant. This report on 13 patients demonstrated excellent results comparable to the intramedullary use of this device. Using a small diameter intramedullary nail as an adjunct to the extramedullary distractor prevented instability and axial deviation. Bone regenerate maturation was noted to occur quicker with extramedullary lengthening, in as little as 6 weeks in some cases, presumably due to lack of trauma to the intramedullary blood supply as there is minimal or no reaming of the canal. This decreases the overall treatment time, allowing earlier return to full activities. Furthermore, the soft tissue trauma imparted by implantable lengthening is significantly less than with external fixation lengthening. This results in a shorter recovery period, so the time between lengthenings can be reduced to 2 to 3 years. Both of these factors mitigate the need to perform large single stage external lengthenings (> 5 cm) in patients with very large limb length discrepancies.

Dahl et al\textsuperscript{11} presented a group of 11 skeletally immature patients treated by extramedullary application of the PRECICE nail. No supplementary intramedullary rod was used and lengthening was limited to 40 mm with an average length gained of 31.5 mm. All patients underwent femoral lengthening only. Complications included 45% of patients having deviation of the regenerate away from the axis of lengthening and 27% undergoing unplanned surgery. Our method of EMILL allowed us to lengthen to 5 cm without instability due to the use of the supplementary intramedullary SLIM rod (Pey Mei Medical), which is a modern version of the Rush rod. Lengthening over a Rush rod to prevent axial deviation dates back to 1956\textsuperscript{18} and is the forerunner of the more modern technique called lengthening over nail.\textsuperscript{19} We were also able to reduce cantilever bending forces and thereby reduce the risk of axial deviation and instability by docking the nail into one end of the femur (distal) and tibia (proximal).

New implants such as the Gro-Plate, from NuVasive Specialized Orthopedics that was FDA cleared in 2019, and others that are developed in an effort to extend internal lengthening to younger children and replace the need for external fixation lengthening in terms of stabilization of joints with preparatory surgery (eg, pelvic osteotomy, SUPERknee, SUPERankle, temporary arthrodesis of joints) and with bracing to prevent joint subluxation and contracture. Lengthening should be restricted to safe amounts such as 5 cm to reduce risks and complications. It can more easily and frequently be repeated than with external fixation reducing the need to achieve large single stage lengthenings.

**REFERENCES**