Extramedullary Motorized Lengthening of the Femur in Young Children

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Background: Limb lengthening by distraction osteogenesis is now achievable via motorized intramedullary devices, mitigating many complications of lengthening by external fixation. In young patients, antegrade intramedullary nailing of the femur risks avascular necrosis of the femoral head. A method of extramedullary placement of a motorized expandable intramedullary nail has been employed by the senior author to safely achieve femoral lengthening without the use of an external fixator in young patients.

Methods: Eleven skeletally immature patients with lower limb length discrepancy were reviewed who underwent extramedullary placement of a magnetic, expandable intramedullary nail for lengthening of the femur. Surgical details, lengthening parameters, and complications were reviewed and classified according to the modified Clavien-Dindo Classification.

Results: Average lengthening was 32.3 mm (range: 27 to 40 mm) comprising an average 14.8% of femoral segment length. The average lengthening duration was 6.3 weeks, and average full weight-bearing began at 12.6 weeks. All but 1 patient underwent early removal of the device at an average of 4.5 months, and 5 had immediate plating of the femur. Complications rates were comparable to other methods of femoral lengthening, including varus or procurvatum through the regenerate, and unplanned reoperation in 3 of 11 cases. Preoperative considerations included careful planning of implant length due to short femoral segments and protection of the knee joint from contracture or iatrogenic instability.

Conclusions: Extramedullary placement of a magnetic expandable intramedullary lengthening nail can achieve lengthening of the femur without the use of external fixation. Considerations with this technique include careful planning of implant length relative to trochanteric-physeal distance, protection against knee subluxation during lengthening, and mitigating deformity of the regenerate. Off-label, extramedullary use of these devices can be considered to decrease the burdens of external fixation in young children. The technique begs the advent of future all-internal technology specifically designed for safe limb lengthening in this age group.

Level of Evidence: Level IV—retrospective case series.

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Limb lengthening was first reported in 1905 by Codivilla[1] via osteotomy, traction through a calcaneal transfixion pin, and immediate plaster application. The surgical technique evolved to the distraction osteogenesis described by Ilizarov,[2] who employed a blood-supply sparing corticotomy, circular external fixation, a regular rate of distraction, and functional limb use during the lengthening period. The learning curve for orthopaedic surgeons in performing limb lengthening is steep, with complication rates ranging from 14% to 225% regardless of the device used.[3] Adjunctive techniques have been developed to mitigate the risks of external fixation (which include difficulty with bone segment stability, pin site infection, body image issues, and difficulty with hygiene and care). These adjuncts included lengthening over intramedullary nails[4] and plate-assisted lengthening.[5,6]

Intramedullary devices that are distracted by mechanical means provided an all-internal solution,[7,8] and the first fully internal motorized lengthening nail activated by an external remote controller was introduced by Professor Rainer Baumgart of Munich, Germany in 1999.[9] This FITBONE nail (Orthofix, Lewisville, TX) is externally activated by a radiofrequency transmitter in the subdermal tissue. More recently, a magnet-driven device has also become available (PRECICE; NuVasive, Irvine, CA). At present, antegrade placement of these implants in the pediatric femur risks avascular necrosis to the femoral head because of the close proximity of the ascending cervical branches of the medial circumflex femoral artery.[10,11] Therefore, intramedullary use of femoral lengthening nails is limited to older children and adolescents, or patients with distal femoral physeal arrest by retrograde placement.

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The technique described involved off-label use of existing devices.

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The senior surgeon has developed a technique of extramedullary (EM) placement of the PRECICE magnetic lengthening nail for modest femoral lengthenings in young children (8 y and younger). The aim herein was to report on the early experience of this off-label use of an existing device, and the planning and technical considerations to achieve safe and effective lengthening. A secondary aim was to describe the complications with early experience.

METHODS

A single-surgeon review was performed of all pediatric patients who underwent EM lengthening of the femur using the PRECICE magnetic expandable nail between 2016 (first use of the technique) and 2018, with a minimum follow-up of 1 year. Informed consent was obtained for off-label EM use of the device. The investigation was approved by the institutional review board. Eleven patients underwent 11 lengthening procedures during the study period.

Demographic data collected included: underlying diagnosis, age at index procedure, limb length discrepancy, femoral starting length (the base of the trochanteric apophysis to the distal femoral physis), and overall femoral length (cephalad aspect of the femoral head to medial femoral condyle). Surgical data included: total lengthening achieved, lengthening as a percentage of initial femoral segment length, lengthening rate, lengthening duration, concomitant procedures, implant details, immobilization used, and time to weight-bearing. Complications were reviewed and classified according to the Clavien-Dindo System.[12]

Surgical Technique

A full description of the surgical technique and step-by-step images are available in the Appendix (Supplemental Digital Content 1, http://links.lww.com/BPO/A305). Nails of 8.5 mm diameter (the smallest available) were used in all patients. Multiple drill holes were used in preparation for a low-energy, diaphyseal corticotomy. An antegrade approach to the femur was made through a small incision proximal to the greater trochanter. A blunt intravastus passage for the nail was created just lateral to the femur. The nail was passed by hand, and provisional Kirschner wires were placed percutaneously in anticipation of later locking bolt placement. Sequential replacement of wires with screws was performed, with caution in tightening the 2 screws closer to the corticotomy (proximal), which could pull the femur into varus alignment. A low-energy corticotomy was then performed with a 10 to 15 mm osteotome under fluoroscopic guidance. Osteotome rotation created a slight step-off as evidence of corticotomy completion. The iliotibial band was then sectioned distally, and the magnetic nail was tested according to standard practice.

Lengthening Protocol

A 5- to 7-day latency period was utilized. The lengthening rate started at 1 mm/d divided into eight 0.125 mm increments (1/8 mm). This rate was adjusted based on the quality of regenerate visualized (ie, new bone formation was contiguous and linear) on weekly radiographs (Fig. 1). The rate was slowed if the regenerate quality was poor, or the rate was increased slightly when the excessive bone formation was apparent. A hip spica splint was maintained during the lengthening phase. Children were kept non–weight-bearing until goal length had been achieved and 3 cortices of bridging regenerate were evident on radiographs.

RESULTS

Eleven patients underwent EM femoral lengthening during the study period. Diagnoses included congenital short femur in 6 patients, fibular hemimelia in 7 patients, and proximal focal femoral deficiency in 3 patients (Table 1). The average patient age at the time of lengthening was 5.9 years (range: 4 to 8 y) including 6 females and 5 males. The average lengthening was 32.3 mm (range: 27 to 40 mm) comprising an average 14.8% of femoral segment length. The average lengthening duration was 6.3 weeks, and the average full weight-bearing began at 12.6 weeks. The average follow-up was 15 months (range: 12 to 30 mo).

Six patients had undergone pelvic or proximal femoral osteotomies as preparation for their lengthening surgery. Guided growth plates were placed at the distal medial femur or the upper medial tibia to address preexisting genu valgum in 6 patients. In 2 cases, knees were temporarily spanned with an internal small fragment plate due to severe joint instability. These implants were placed subcutaneously with screw fixation in the metaphysis and epiphysis of the distal femur and proximal tibia. All but 1 patient underwent removal of the nail at an average of 4.5 months. Five patients underwent immediate plating of the femur at the time of nail removal because of either loss of fixation or concern about patient adherence to activity limitations during maturation of the regenerate.

Complications included small varus or procurvatum through the regenerate, which was observed in most cases and is described in Table 2. There was loss of fixation or unacceptable malalignment of the regenerate requiring unplanned reoperation in 3 of 11 cases. Preoperative considerations included planning of implant length due to short femoral segments and limited implant inventory by length, as well as protection of the knee joint from flexion deformity or instability.

DISCUSSION

Motorized telescopic intramedullary nails have been safely used to achieve lower limb lengthening without the use of external fixation.[13] In congenital femoral deficiency, there may be fewer complications associated with this all-internal lengthening technology compared with circular fixators.[14] Comparisons of magnetic lengthening nails to external lengthening over a nail have also suggested that, while index implant costs are greater, overall costs due to secondary procedures may be lower with motorized nails.[15] Regardless, the antegrade femoral entry points for these intramedullary implants are at the piriformis fossa or greater trochanter, which have appreciable rates of avascular necrosis of the femoral head in skeletally immature patients (1.4% to 2.0%).[10] Until now, the only physeal-sparing technique available for

FIGURE 1. A 5-year-old boy with fibular hemimelia and a 5.5 cm limb length discrepancy underwent a 4 cm extramedullary lengthening. Because the implant is not directly apposed to the lateral femur, varus of the femur can occur because of the long "working-length" between the corticotomy and fixation. In this case, 4 degrees of varus of the regenerate was observed and had remodeled at the time of consolidation.

distraction osteogenesis in this age group has been external fixation.[16,17] To mitigate the surgical and social difficulties of external fixator–based lengthening, the senior author developed a technique of lengthening of the femur with EM placement of a magnetic lengthening nail, with modest lengthening goals (up to 4 cm) and protection of the knee. This report details safe and effective lengthening is achievable using this implant and technique.

Newer technologies do not eliminate complications common to all limb lengthening surgery. In a previous investigation of 140 procedures, complication rates with limb lengthening appeared unrelated to a particular implant and only decreased with experience after 30 procedures by the operating surgeon.[3] Complications reported with the EM technique included varus and procurvatum of the regenerate (7/11 cases) and unplanned reoperation in 3 of 11 cases. These rates are comparable to historical reportage with lengthening by a variety of techniques, even plate-assisted techniques which are designed to minimize the time in external fixators in young patients.[3,5,6,18,19] Even using techniques designed to minimize the duration of ex-fix, time in fixators for children has been reported to be a minimum of ~7 weeks.[6] Should future technology obviate the need for external fixation in select patients, we aspire to have those patients spend 0 days instead of 7 weeks in a frame.

All congenital limb lengthening requires close attention to resting joint position and absolute prevention of flexion contractures, which invariably lead to joint luxation

TABLE 1. Children Undergoing Extramedullary Femoral Lengthening

Patient No.	Age (y)	Diagnosis	LLD (cm)	Femoral Starting Length (Apophysis to Distal Femoral Physis)	Lengthening (cm)	Lengthening as % of Femoral Segment	Rate (mm/d)	Duration (d)	Concomitant Procedures	Implant*
1	5.6	CSF, fibular hemimelia	11	18	2.7	13.17	0.59	46	CBG × 2 HWR ITBR BTX-A	170 × 8.5 × 3
2	7.6	PFFD	19	14	3.3	20.00	0.92	36	HWR Knee-spanning plate ITBR BTX-A	170 × 8.5 × 3
3	5.3	PFFD	5	17	3	14.63	0.56	54	GG ITBR	170 × 8.5 × 3
4	5.1	PFFD	13	12.5	3	20.69	1	30	HWR ITBR Knee-spanning plate	170 × 8.5 × 3
5	6.8	CSF, fibular hemimelia	6	22.6	3.5	13.21	1	35	ITBR GG	195 × 8.5 × 5
6	4.8	CSF, fibular hemimelia	10	19.4	3	13.27	1	30	HWR GG ITBR	170 × 8.5 × 3
7	6.6	CSF	5.5	25.6	3	10.95	0.91	33	ITBR BTX-A	170 × 8.5 × 3
8	8.0	CSF, fibular hemimelia	8.5	22.4	3	12.24	1.3	23	ITBR BTX-A GG	170 × 8.5 × 3
9	5.3	CSF, fibular hemimelia	7	19.3	3	12.99	1	30	ITBR GG	170 × 8.5 × 3
10	5.2	Fibular hemimelia	5.5	21	4	16.19	0.95	42	ITBR STR	190 × 8.5 × 5
11	5.5	Fibular hemimelia	6	19.6	4	20.4	0.85	47	ITBR Tibial osteotomy Calcaneal osteotomy GG	190 × 8.5 × 5

*Implant specifications = length × diameter × stroke (mm × mm × cm).

BTX-A indicates botulinum toxin A injection; CBG, cortical blocking screws; CSF, congenital short femur; GG, guided growth; HWR, hardware removal; ITBR, iliotibial band release; LLD, limb length discrepancy; PFFD, proximal focal femoral deficiency; STR, soft-tissue release.

TABLE 2. Complications Associated With Extramedullary Lengthening of the Femur

Patient No.	Age (y)	Diagnosis	Lengthening (cm)	Clavien-Dindo	Complication Details
1	5.6	CSF, fibular hemi	2.7	—	N/A
2	7.6	PFFD	3.3	I	Varus regenerate, observed
				IIIB	Breakage of knee-spanning plate, early reoperation for removal of lengthening nail
3	5.3	PFFD	3	I	Varus regenerate, observed
4	5.1	PFFD	3	I	Varus regenerate, observed
5	6.8	CSF, fibular hemi	3.5	I	Varus regenerate, observed
6	4.8	CSF, fibular hemi	3	—	N/A
7	6.6	CSF	3	I	Varus regenerate, observed
8	8	CSF, fibular hemi	3	IIIB	Unplanned operation for repeat ITBR/release for KFC, BTX-A hamstrings, plating regenerate
9	5.3	CSF, fibular hemi	3	IIIB	Procurvatum regenerate, reoperation for placement of blocking screws
10	5.2	Fibular hemi	4	—	N/A
11	5.5	Fibular hemi	4	I	Varus regenerate, observed

BTX-A indicates botulinum toxin; CSF, congenital short femur; fibular hemi, fibular hemimelia; ITBR, iliotibial band release; KFC, knee flexion contracture; N/A, not applicable; PFFD, proximal focal femoral deficiency.

and ultimately joint dislocation. Any surgeon choosing to lengthen in a congenitally deficient limb is obligated to be vigilant and have a plan to maintain joint stability. With external fixation, a joint may be spanned. Herein, those with obvious knee instability were temporarily treated with external splinting or adjunctive internal plating.

The technical steps of the operation have evolved from its index use. "Blocking screws" placed outside of the nail (to capture the EM nail anteriorly and posteriorly) were initially employed to mitigate procurvatum and varus of the proximal fragment, but fewer such screws were used as the technique developed and small magnitudes of deformity were observed and often remodeled. Small lengthenings (3 cm or less) were achieved initially before progression to 4 cm lengthenings.

There are limitations to this investigation, as it is a retrospective description of off-label use of an existing implant. The technique was employed with the consent of patients' parents who wished to avoid the burdens of external fixation. Those patients who had previously undergone frame lengthenings described a subjectively better experience, although no pain or satisfaction scores are available for comparison. The senior author acknowledges external fixation as the standard of care after employing it for 35 years. Part of this experience includes the observation that adults have longstanding physical stigmata of half pins and wires traveling through the skin. Furthermore, there are deleterious effects on limb function (deep, clefted scars tethered to muscle and bone) secondary to repetitive lengthenings of 20 to 30 cm over many episodes during the lifetime of the child. It was this career of appreciating the physical and psychological scarring that was the incitement to embrace new technology and aim to improve upon the current standard of care. This risk/reward balance is the crux of the discussion with families. If families wish to pursue the off-label investigative technique to avoid a frame and (perhaps) save a future lengthening episode when older, EM lengthening is discussed.

Although all lengthenings were small in absolute magnitude (range: 2.7 to 4.0 cm), they still represented a significant proportion of overall femoral length (mean: ~15%). This was a deliberately conservative approach since the technique was new and the discrepancies were of congenital etiology (viz. higher risk). Because the lengthening nail is not designed for this use, its placement is essentially "intravastus." The consequences of such placement are unknown (muscle trauma, bleeding, prominence), even if the authors perceive such to be well-tolerated during lengthening. The larger 10.7 mm proximal end of the nail could result in iliotibial band irritation (transiently present in one smaller child during lengthening), which suggests that lower-profile implant design is necessary. The straight nail shape necessarily leaves some separation between the proximal femoral fragment and the nail, and because the nail is not intramedullary, allows for cantilever bending. Because the children and their femora were small, 8.5 mm diameter nails were employed in all cases, and the distal interlocking bolts were small diameter (3.5 mm). The initial idea of using the nail was to achieve length, but the use of a percutaneous plate at the end of lengthening (in 5 patients) was intended to minimize implant bulk when motion and weight-bearing was initiated. Finally, since unplanned reoperation rates were 25% in our series, this could increase surgical costs in addition to high implant cost.

These early results suggest that all-internal EM lengthening of the femur can be achieved safely in young children. With careful preoperative planning of implant size, low-energy corticotomy, fractionated lengthening, joint protection by splinting or bridge-plating, and careful assessment of regenerate, the use of external fixation can be minimized in young children with large lower extremity discrepancies. These results beg the advent of all-internal motorized lengthening technology specifically designed for use in this age group.

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