Advances in Pediatric Limb Lengthening

Part 1

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» Integrated fixation techniques continue to evolve and help to decrease the external fixator duration for pediatric patients undergoing limb lengthening.

» Intramedullary limb lengthening is available for pediatric patients. Although this technique has many advantages, the surgeon still needs to be vigilant about potential lengthening complications (contractures, joint subluxation, fractures).

» Preliminary surgery may be necessary to prepare the limb for safe lengthening.

Pediatric limb lengthening is an exciting and rapidly developing field. The primary goal is to produce healthy regenerate bone of the desired length without complications. In addition, the experience should be as easy and comfortable as possible for the patient and his or her family. Innovative surgical techniques and devices continue to push surgeons closer to being able to consistently achieve these goals. This review will provide a comprehensive summary of the latest advancements in pediatric limb lengthening in two parts. This part, Part 1, will cover advances in the preoperative assessment and methods to decrease the time spent in an external fixator. Part 2 (http://reviews.jbjs.org/content/3/9/e4), which will be published in a future issue of *JBJS Reviews*, will cover advances in techniques designed to decrease the time to union and methods to recognize and to avoid complications[1].

Preoperative Patient Evaluation

Classification

In pediatrics, it is unusual for a patient to present with only a limb-length discrepancy. Most patients present with additional pathologic conditions such as angular, rotational, and/or translational bone deformities. The stability of the joint proximal and distal to the lengthening segment may also be compromised. The surgeon then has to choose whether these additional deformities can or need to be corrected and whether to perform their correction combined with, before, or after the lengthening. Joint contractures can also complicate the picture. A comprehensive and accurate preoperative assessment of the limb deformity is essential for successful treatment. Two recent classification schemes have been described to account for the variability in deformity presentation. Manner et al. described grading the deformity on the basis of the number of dimensions involved. For example, a one-dimensional deformity is considered Type I and a four-dimensional deformity is Type IV[2]. The LLRS AIM index, a mnemonic indicating seven pretreatment domains (**L**ocation and number of deformities, **L**eg-length inequality at maturity, **R**isk factors, **S**oft-tissue coverage, **A**ngular deformity, **I**nfection/bone quality,

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and **M**otion/stability of the joints above and below), is a validated rating scale designed to assess the complexity of general lower-limb deformities[3]. It accounts for seven domains: the location of the deformity, leg-length inequality, medical risk factors, soft-tissue assessment, angular deformity, infection and bone quality, and motion and stability of the joints (Table I).

Congenital Compared with Acquired Limb-Length Discrepancy

An important first distinction in the evaluation process is to determine if the limb-length discrepancy is due to a congenital or an acquired condition. Congenital limb-length discrepancies are known to be more challenging to treat successfully and require a different thought process in their planning. Gordon et al. reported a complication rate of 44% (eleven of twenty-five) for congenital limb-length discrepancies compared with 17% (two of twelve) for acquired limb-length discrepancies[4]. Launay et al. found an increased risk of fracture in patients with achondroplasia if lengthening was performed before the patient age of nine years and if the latency period was less than seven days[5]. They also found femoral fractures in all patients undergoing lengthening for congenital femoral deficiency when the lengthening percentage exceeded 15% and the latency period was less than seven days. The pathoanatomy of the congenital long bone deficiencies (e.g., congenital femoral deficiency, tibial hemimelia, or fibular hemimelia) continues to be elucidated. A better understanding of the bone and soft-tissue deformity has allowed new reconstruction techniques to be developed. For example, a Paley Type 1b congenital femoral deficiency is described as a combination of abduction, flexion, varus, and external rotational deformities at the hip. A stepwise reconstruction that addresses each one of these abnormalities has been described[6].

Preoperative Considerations

The concept of preparatory operative treatment has been introduced[6,7]. This refers to preparing the limb for lengthening by first stabilizing the adjacent joints and removing known soft-tissue restraints. For example, lengthening for congenital femoral deficiency can lead to subluxation or dislocation of the knee and/or hip joints. Therefore, any acetabular dysplasia should be addressed with a pelvic osteotomy prior to lengthening. The knee joint may need to be protected by extending the fixator across the joint or may need to be stabilized by performing a simultaneous ligament reconstruction at the time of the lengthening. Soft-tissue restraints, such as the iliotibial band, also need to be addressed at the time of the lengthening. Similar preparation has been outlined prior to lengthening in patients with fibular hemimelia[6]. This stepwise and comprehensive approach is vital to the success of the lengthening.

The surgeon must also be cognizant that joint contractures can influence the appearance of limb-length discrepancy. The hip, knee, and ankle joints need to be tested for range of motion. Patients can have a true structural limb-length discrepancy, a functional limb-length discrepancy, or both. Adduction contracture of the hip, hip flexion contracture, and knee flexion contracture are contractures that appear to shorten the limb. Hip abduction contracture and equinus contracture can cause a limb to look longer. Sagittal plane analysis is often overlooked but is very important. A standing full-length lateral radiograph can be made to evaluate the osseous and soft-tissue deformities. This radiograph is made by having the patient stand with his or her foot facing straight ahead and the cassette parallel on the lateral aspect of the desired limb. Without moving the planted foot, the pelvis and opposite limb are then externally rotated 45° away. This maneuver allows the radiograph beam to capture the full length of the limb from the hip to the ankle without obstruction. Externally rotating the knee of the target limb about 10° also helps to get a true lateral view of the knee with this technique.

It is critical that the surgeon's preoperative assessment anticipates factors that may predispose the patient to poor bone formation. Host-related factors (the use of nonsteroidal anti-inflammatory drugs or systemic illness), local factors (scarred soft-tissue envelope, overlying infection, or prior radiation therapy), and iatrogenic factors (poor osteotomy location selection, suboptimal osteotomy technique, a >1-cm gap at the osteotomy site during the latency phase, or application of a mechanically unstable frame configuration) all contribute to potential impairment of healthy bone formation[8]. Prolonging the latency period or decreasing the distraction rate may be necessary in such cases to overcome the obstacles to producing a healthy regenerate bone.

Patient selection for limb lengthening is also extremely important. Limb lengthening can be a long, stressful process for everyone involved. A preoperative assessment of the patient's psychosocial situation is recommended before starting the lengthening[9]. Risk factors for a more challenging treatment process have been identified, such as living with a single parent, having a preexisting mental health condition, and having a history of operative treatment.

Assessing Growth

Accuracy in predicting the total limb-length discrepancy at skeletal maturity is critical when trying to make management decisions in pediatric patients. The Multiplier method calculates a coefficient for each age to represent the reciprocal of growth remaining[10]. The coefficient is independent of growth percentile, race, nationality, and generation. The Multiplier method has been shown to be reliable using chronologic age prior to the adolescent growth spurt. However, skeletal age is more accurate than chronologic age once the adolescent growth spurt has begun[11]. There are several free smartphone applications (Multiplier [Rubin Institute for Advanced Orthopedics, Baltimore, Maryland] and Paley Growth [Paley Foundation, West Palm Beach, Florida]) available that allow

TABLE I LLRS AIM Index for Limb Deformity*

Domain	Score *(points)*
Location (no. of deformities per limb of ≥10° angulation in separate planes and rotation all count as separate deformities)	
No deformity	0
One deformity	1
Two deformities	2
Three deformities	3
More than three deformities	4
Leg-length inequality (estimate at skeletal maturity)	
0 to 2 cm	0
>2 to 5 cm	1
>5 to 10 cm	2
>10 to 15 cm	3
>15 cm	4
Risk factors (assess clinically)	
None	0
Age of less than five or more than forty years	Add 1 point
Smoker	Add 1 point
Obesity	Add 1 point
Other disease (e.g., diabetes)	Add 1 point
Soft-tissue coverage	
Normal	0
Bruising or contusion	1
Scarring (open grade I)	2
Poor coverage (open grade II)	3
Inadequate coverage (open grade III)	4
Angular deformity (measure and assign greatest primary deformity)	
0° to 10°	0
>10° to 20°	1
>20° to 40°	2
>40° to 60°	3
>60°	4
Infection and bone quality (select the most severe)	
Normal	0
Osteoporotic	1
Dysplastic	2
Infection	3
Combination	4
Motion and stability of the joints above and below	
Normal	0
Decreased motion (<60% of normal)	1
Subluxation of joint	2
Dislocation of joint	3
More than one joint affected	4
LLRS AIM Index scoring (scores range from a minimum of 0 points to a maximum of 28 points)	
Normal	0
Minimal complexity	1 to 5
Moderate complexity	6 to 10
Substantial complexity	11 to 15
High complexity	16 to 28

*LLRS AIM is a mnemonic of the seven criteria that are required to determine the index.

quick, easy calculations to be performed in the office setting.

Studies of growth have also identified several useful concepts when analyzing a patient with a limb-length discrepancy[12]. The tibial length remains at a constant 80% of the femoral length throughout growth. These relative lengths remain the same regardless of the position of the child on the growth curve. Peak growth velocity in the lower limbs occurs six months earlier than spinal growth peak velocity. Growth in the lower limbs ceases two years and six months after the onset of puberty. This correlates with the closure of the distal phalangeal physes, the closure of the elbow apophysis, and Risser stage 1. Generally accepted methods for predicting the final limb-length deficit at skeletal maturity for congenital malformations have been proposed: to calculate the final limb-length discrepancy, the limb-length discrepancy can be multiplied by 5 at birth, by 3 at the patient age of one year, by 2 at the patient age of three years in girls and four years in boys, and by 1.5 at the patient age of seven years in girls and boys.

Radiographic Evaluation

Radiographic assessment of limb-length discrepancy continues to evolve and to become more precise. Children who have a clinically important limb-length discrepancy often require multiple radiographs to plan treatment and to monitor outcomes. Therefore, the ideal radiographic method for evaluating a limb-length discrepancy should be easily available, accurate, and reliable and should permit analysis of the entire lower-extremity alignment with minimal radiation exposure and no magnification error[13,14]. At present, the best combination of these traits is a standing full-length anteroposterior digital radiograph of both lower extremities, with blocks under the short leg to level the pelvis, utilizing a magnification marker. Although scanograms or orthoroentgenograms are designed to eliminate magnification errors and are readily available, they have been found to overlook vital information such as the foot or ilium height, the mechanical axis of the limb, any coexisting angular deformities, and the etiology of the leg-length discrepancy[15]. In addition, standing full-length radiographs have been found to be equally as reliable as a scanogram for measuring limb-length discrepancy[14]. Therefore, scanograms and orthoroentgenograms have fallen out of favor as the primary assessment tool for limb-length discrepancy.

New technologies have been evaluated as possible replacements for the conventional computed or digital radiographs or teleoroentgenograms for assessing limb lengths[13]. Microdose digital radiography produces a computer-generated image of the lower limbs made with the patient standing. The source and detector move together in scanning the patient so that the pencil radiograph beam is always perpendicular to the patient. A continuous series of photon beams collimated to act as a point source is projected through the patient to strike a computerized detector. This technique exposes the patient to a negligible dose of radiation (0.001 to 0.002 cGy) during the scanning process. It has been found to be more accurate than orthoroentgenograms but still is not readily available in most physicians' offices. Despite the advantage of avoiding ionizing radiation, magnetic resonance image (MRI) scanograms have not been found to be as accurate as radiographs or computed tomography (CT) scanograms and are inconvenient. CT scanograms have a lower radiation dose than conventional radiographs and are more accurate in the assessment of coronal plane length discrepancies than radiographs. However, because these are not weight-bearing images, it is difficult to accurately assess limb alignment. Although this limitation decreases the utility of this technique for standard limb-length evaluation, lateral CT scanograms may be a useful technique to assess leg lengths in patients unable to stand upright with knee flexion contractures of >30°.

One promising new technology is the EOS imaging system (Euronext, Paris, France). This low-dose, biplanar, digital radiographic imaging system uses highly sensitive gaseous photon detectors that can produce full-length, weight-bearing images of the lower extremities. Depending on the setting, an image is produced in three to eight seconds with forty-three times less radiation than a conventional radiograph and six times less radiation than a CT scanogram[16]. EOS was found to be more accurate than conventional radiographs and CT scanograms for the assessment of limb length in the coronal plane. In addition, full-length orthogonal weight-bearing lateral plane views can be obtained to allow simultaneous evaluation of any sagittal plane deformity. Biplanar EOS images can be obtained faster, with decreased turnaround between patients, compared with computed or digital radiography. However, because of the relatively long acquisition time needed (three to eight seconds), movement artifacts in the images are a potential issue.

Indications for Limb Lengthening

The indications for limb lengthening continue to evolve. Traditional teaching stipulates that 0 to 2 cm of limb-length discrepancy can be ignored, but a discrepancy between 2 and 5 cm should be treated with contralateral epiphysiodesis, and a >5-cm difference should be lengthened[17]. These guidelines provide a framework for decision making, but, in reality, each case has its own unique set of circumstances that determine the best treatment choice for the patient and family. The current guidelines do not take into account the patient's overall height (or projected height). A 2-cm discrepancy is not the same for a 1.82-m-tall (six-foot-tall) patient and a 1.52-m-tall (five-foot-tall) patient. Some patients are absolutely bothered by limb-length discrepancies of 1.5 cm and return to the clinic repeatedly until something is done. There are patients with projected short stature who value every millimeter of height and therefore do not want epiphysiodesis as a treatment option. Some families value the

choice with the fastest recovery time and want an epiphysiodesis even for large discrepancies. As the experience with limb lengthening has increased and the technology has improved, it has become possible to produce safe, reliable, and comfortable limb lengthening for patients with mild to moderate discrepancies. It is now reasonable to offer limb lengthening as the primary treatment option for many patients with discrepancies of <5 cm.

Fixator Choices

Once a decision to lengthen the limb has been made, the surgeon must choose whether a uniplanar fixator, circular fixator, or intramedullary lengthening device should be used. Uniplanar fixators have traditionally been used primarily to lengthen femora and humeri. It is less comfortable to place circular fixators on these proximal limb segments. New designs of uniplanar fixators allow half pins to be placed in multiple planes and at multiple locations, which helps to improve the stability and options of the construct. For example, the half pins are no longer limited to just placement through the clamp but can be placed out of plane or with increased spread between fixation points. There are also uniplanar fixators with attachments that allow the hip or knee joint to be spanned, to undergo range of motion movement, and to be protected during the lengthening process. A breakthrough in circular external fixators occurred with the advent of the hexapodal frame design. These fixators are based on concepts similar to flight simulators (Stewart platform) and allow six degrees of freedom of movement. The frames can rotate around a virtual point in space rather than requiring a physical hinge. These frames are paired with software programs that help the surgeon plan the deformity correction and plan the frame size and mounting location. The most powerful feature of the software is the ability to make adjustments to the deformity correction during the course of treatment without having to return to the operating room. Because these devices allow the surgeon to correct multiple planes of deformity simultaneously or sequentially, they are ideal for pediatric conditions.

Decreasing External Fixation Index

Integrated Techniques

External fixation remains the most common method of lengthening pediatric long bones worldwide. External fixation, despite its remarkable capabilities, has its shortcomings. It is cumbersome for the patient. The visible implant with pins and wires transgressing the soft tissues can create both physical and mental discomfort to the pediatric patient. The longer the treatment with the external fixator, the less well they are tolerated. The external fixation index measures the number of days that the external fixator is attached to the bone per centimeter of length gained. Integrated techniques that combine internal and external fixation have been proposed as a method to decrease the external fixator index and to improve patient comfort. In these techniques, the external fixator is removed following the distraction period and the regenerate bone is protected with internal fixation during the consolidation period. For example, lengthening over an intramedullary nail allows the fixator to be removed at the end of the distraction phase. The decreased fixator time diminishes fixator-associated issues such as pin-track infections and contractures and it improves patient tolerance and comfort. The nail provides internal stability to the regenerate bone. Consequently, the risk of deformity during lengthening and the risk of fracture or deformity after external fixator removal are decreased.

Both rigid and flexible intramedullary nails have been used in pediatric lengthenings. Gordon et al. described their experience lengthening the femur over a rigid nail in thirty-seven patients with an average age of 11.6 years[4]. They averaged 7 cm of lengthening and had the fixator removed after eighty-one days. All but one patient achieved 120° of knee flexion. Four patients had failure to distract. Gordon et al. recommended that, before leaving the operating room, surgeons ensure that the distal portion of the femur can rotate easily around the implant and that there is no binding of the half pins. It is also important that the pins are placed perpendicular to the long axis of the femur to prevent binding. Forty-nine percent of patients had a 7-mm nail inserted. Fractures occurred in five patients at an area just distal to the tip of the nail or through the distal interlocking holes. Consequently, exchanging the nail for a longer nail of appropriate length to relieve the stress riser in the mid-femur should be considered at the time of fixator removal. Eleven percent of patients developed osteomyelitis and were effectively treated by nail removal, reaming of the canal, and intravenous antibiotics.

An alternative technique is to insert a reamed nail at the time of fixator removal instead of at the time of the index procedure. The reaming of the regenerate bone appears to produce revascularization that can provide abundant new bone formation[18]. Osseous union occurred in less than one-half of the time compared with a control group lengthened with an external fixator alone[19]. A longer and larger nail can also be inserted using this technique compared with the small-diameter nails utilized in traditional techniques of lengthening over a nail.

The use of flexible intramedullary nails has also been combined with pediatric limb lengthening. This technique avoids potential physeal injuries and the risk of osteonecrosis associated with rigid intramedullary nailing in children. In the femur and tibia, nails of 1.5 to 2.0-mm diameter are bent in an arc of 40° to 50° and passed across the osteotomy site to the opposite metaphysis[20]. Rotation of the curved tip of the nail during insertion allows the nail to avoid the half pins and wires of the external fixator. The nails add stability and prevent translation. One study demonstrated fractures in six (13.6%) of forty-four bone segments in patients following lower-extremity limb lengthening with flexible

intramedullary nailing and fractures in fourteen (20.9%) of sixty-seven segments in patients without flexible intramedullary nailing[5]. Popkov et al. had 3% regenerate bone deformities between 15° and 40° angulation and 8% fractures in a control group of 194 cases of lengthening compared with no fractures and only one case of a 10° deformity of the regenerate bone in ninety-two cases of lengthening over a flexible nail[20]. The healing index was also reduced in both series with use of this technique. Launay et al. also found better union of the bone callus, shorter fixation, fewer deformities, and no infections in fourteen cases when lengthening the forearm over a flexible intramedullary nail[21]. Popkov et al. reported no deep infections in the group with lengthening over a flexible nail and recommended removing the nail electively three to eight months after the frame removal[20].

Because of concerns regarding the risk of intramedullary infection and the inability to put rigid nails in small children with open physes, an alternative integrated technique, lengthening over a plate, was developed. This technique uses a submuscular locking plate to protect the regenerate bone instead of a nail. The plate can be inserted at the index procedure or at the time of frame removal. The plate insertion does not violate the physis and therefore can be used in small children. No deep infections have been reported and any angular deformity of the regenerate can be corrected with adjustment at the time of plate locking[22-24].

Intramedullary Limb Lengthening

Although these integrated techniques have been successful in decreasing the external fixation index, they still require an external fixator for at least a portion of the treatment. Intramedullary lengthening systems have been developed in an attempt to achieve distraction without any need for external fixation. Completely internal lengthening nails have many potential advantages over external fixation, including elimination of pin-track infections, less risk of neurovascular injury from wire or half pin insertion, improved cosmesis, better body image and psychological well-being for the patient during treatment, improved range of motion, increased activity levels during lengthening, and less pain.

Two basic designs of internal lengthening devices have been utilized. One design utilizes intermittent rotation of the limb to create distraction. Two examples of this are the Albizzia Nail (DePuy, Villeurbanne, France) and the Intramedullary Skeletal Kinetic Distractor (ISKD) (Orthofix, Lewisville, Texas). The Albizzia nail has a ratchet assembly that requires approximately 15° of limb rotation to create distraction[25]; this device is not available in the United States. The ISKD was cleared by the Food and Drug Administration (FDA) for marketing in the United States in 2001[26]. It was designed to lengthen by small (3° to 9°) physiologic oscillations between two telescopic sections connected to a double-clutch mechanism. However, due to the uncontrolled lengthening rate and rhythm, the ISKD had a very high complication rate[27-32]. It was recently removed from the market and is no longer available.

The second type of intramedullary lengthening nail design uses a mechanism to push the nail in pure axial distraction without requiring rotation through the callus. There are currently two nails available with this type of design. The first, called the Fitbone (Wittenstein, Igersheim, Germany), was developed in 1991[32,33]. This steel nail has an electromagnetic motor and a subcutaneous patient-controlled antenna that allows fully implantable lengthening. The antenna powers and controls the distraction by transcutaneous transmission of radio-frequency waves. Although this device has not been approved by the FDA and is not currently available in the United States, the internationally reported results have been positive[33-37].

The PRECICE nail (Ellipse Technologies, Irvine, California), originally cleared by the FDA in July 2011, is a magnet-operated telescopic internal lengthening device[32]. The nail uses a generic rare earth magnet connected to a gear box and screw shaft assembly. An external remote control device containing two motor-driven rotating magnets interacts with the internal magnet to create distraction (or compression). The rate and rhythm of the lengthening can be precisely adjusted according to each patient's needs.

Accurate distraction control in intramedullary lengthening devices is critical because too rapid a process can lead to nonunion, nerve damage, and joint contractures, and too slow a process runs a risk of premature consolidation. Unlike the rotatory lengthening nails, the PRECICE nail has demonstrated excellent accuracy and rate control in two series of patients[38,39].

Devices that require rotation through the callus to lengthen, such as the ISKD and the Albizzia nails, have been reported to be quite painful for the patient. It is theorized that the rotation leads to friction and muscle spasm pain. In one series of lengthening with the Albizzia nail, 39% (twelve of thirty-one) of patients required readmission to perform ratcheting under general anesthesia[25]. This type of pain has been notably absent in the pure axial lengthening devices such as the Fitbone and the PRECICE nail[32,34]. In a series of thirteen patients, Herzenberg et al. compared pain scores for lengthening with the PRECICE nail and for lengthening with external fixation. They found decreased pain with internal lengthening (a visual analog scale [VAS] pain score of 7 points for external fixation and 3 points for the PRECICE nail) and decreased duration of need for pain medication (12.5 weeks for external fixation to 5.2 weeks for the PRECICE nail). There was also more comfort during physical therapy and a quicker return to full weight-bearing with the PRECICE nail[40].

The largest published case series using the PRECICE nail, 155 cases, revealed seven mechanisms that failed to lengthen, two due to operator error by the surgeon's team in applying the

external remote control device the wrong way and five after meeting excess resistance from the callus[32]. The total number of nail breakages for the first 155 PRECICE lengthenings was three. By comparison, in forty-four cases using the Fitbone, there were five cases that had failure of the lengthening mechanism and one case that had nail breakage[33-35].

Two potential failure modes were noted with the first version of the PRECICE nail: the junction of the gears to the lead screw, and the welds of the nail on either side of the drive mechanism[32]. The PRECICE 2 (P2) addresses these weaknesses and was cleared for use by the FDA in October 2013. The P2 is at least two times stronger in bending fatigue strength and has a three times stronger coupling between the gears and lead screw. The P2 is available in 50 and 80-mm stroke lengths and is also available in a smaller outer diameter (8.5 mm).

As experience with the use of intramedullary lengthening nails increases, the following recommendations have been proposed to help achieve optimal outcomes when utilizing these devices. Preoperatively, the bone size must be assessed. It must be large enough to accommodate the reaming required for insertion of the nail (1.5 to 2 mm larger than the diameter of the nail). Because the nail is straight, the osteotomy should be planned at the apex of the femoral bow to limit the amount of binding in the canal. Although short nail lengths are preferred to limit the amount of binding, the overall length should be chosen so that at least 5 cm of the thick part of the nail remains in the distal segment at the end of distraction. The nail should be inserted with minimal resistance to avoid damaging the distraction mechanism. It is not recommended to hit the nail with any forceful blows during the insertion process. If the width of the medullary canal is larger than that of the nail at the osteotomy level, blocking screws should be used. For example, while lengthening a tibia, muscular forces are known to induce valgus and apex anterior deformity into the regenerate bone. Blocking screws placed posterior and lateral to the nail in the proximal segment will resist this tendency. To confirm completion of the osteotomy, the bone should be rotated around the nail. Steinmann pins or half pins can be used to ensure rotational control of the segments during nail insertion[41].

Despite all of the described advantages over external fixation, intramedullary lengthening devices are not without their own potential complications. Joint contractures or subluxations can still occur, and there is no easy way to correct one when it appears. With external fixators, the joint can be spanned prophylactically or fixation across the joint can be added at the time early subluxation is noted. Internal lengthening nails do not have a similar rescue maneuver. Therefore, it is important that the follow-up radiographs at each visit are critically evaluated, not just for the length achieved and the quality of the regenerate bone, but also for evidence of early joint subluxation. For tibial lengthening, physical therapy should focus on ankle dorsiflexion and knee extension. The patient should wear an ankle dorsiflexion splint during the day and a knee extension brace at night. If necessary, an extra-articular tibiocalcaneal screw can be inserted to lock the ankle in neutral position during the lengthening process[42]. For femoral lengthening, therapy should focus on knee flexion or extension and hip extension. During the lengthening process, hip abduction of $<20°$ or knee flexion of $<45°$ is an indication to stop the lengthening until adequate motion is recovered[32,41]. If joint subluxation is noted, lengthening must be stopped and efforts to restore joint congruency and stability must be undertaken.

Another concern is the effect of lengthening the lower extremity along the anatomic axis rather than the mechanical axis. Intramedullary lengthening induces a lateral shift of the mechanical axis of about 1 mm for every 1 cm of lengthening. This shift should be anticipated preoperatively, especially in patients with any preexisting valgus deformity[43].

Finally, because the nails are intentionally short to avoid the bow of the femur, a stress riser is created in the mid-femur (or tibia) at the end of the nail. It is recommended to avoid the distal anteroposterior locking screw drill-hole, as it potentially increases the stress at this level in the femur and leaves a subcutaneous screw head in the tibia[32].

In contrast to external fixation, intramedullary lengthening does not allow for secondary deformity corrections during the lengthening process. Deformity correction is possible with the nail but must be done acutely. This requires meticulous preoperative planning and careful intraoperative technique. The reverse planning technique allows the deformity to be analyzed and an acute correction planned with use of a retrograde femoral nail[44]. Fixator-assisted nailing techniques allow the osteotomy to maintain the new corrected position while the reaming and nail insertion take place[45]. Blocking screws should be inserted in metaphyseal bone to guide the reaming and nail path, if necessary. It is also possible to perform deformity correction with use of a second osteotomy proximal or distal to the end of the nail and a locking plate. Alternatively, the lengthening procedure can be staged, with fixator-assisted nailing or plating initially performed to correct the deformity. After the osteotomy has healed, exchange nailing can be performed with use of a telescopic nail, with an osteotomy to lengthen the bone.

Alternative Lengthening Technique

One recently described technique produces lengthening without the use of any implants[46]. Limpaphayom and Prasongchin described stimulating growth in the short limb of patients with open physes by stripping and dividing the periosteum of the lower-extremity long bones. This technique is theorized to increase the blood flow to the physis and to reduce the tethering effect of the

periosteum on the growth plate. In eight of eleven patients, with an average preoperative discrepancy of 6 cm, the limb-length discrepancy corrected to within 2 cm within twenty-five months. However, the authors did not recommend this technique for patients with discrepancies of >6 cm.

Conclusion

Pediatric limb lengthening is a rewarding but challenging endeavor. A thoughtful and comprehensive preoperative plan will help to minimize the obstacles to success. Limb-lengthening techniques and devices continue to evolve in an effort to improve outcomes as well as patient comfort and satisfaction. New intramedullary limb-lengthening nails are extremely promising in this regard. However, the surgeon still needs to be cautious when using these nails. The process of limb lengthening, regardless of the device used to achieve it, still carries the inherent risks of fracture, infection, joint contracture, and joint subluxation or dislocation. The surgeon must be cognizant of these potential complications at all times and be prepared to manage them when they occur.

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