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Lower extremity growth and deformity

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Abstract The pediatric lower extremity has well known growth patterns. When deformities or growth disturbances occur, there are several methods to measure and predict the resulting discrepancy, including the Green-Anderson, Moseley, and Multiplier methods. Many techniques exist to correct leg length discrepancy and deformity such and temporary epiphysiodesis, permanent epiphysiodesis, external fixators, and internal lengthening devices. All of these methods have numerous complications and limitations; however, with careful planning and patient selection, length and alignment can be improved with high patient satisfaction.

Keywords Lower extremity · Pediatric orthopedics · Growth · Deformity

Introduction

Lower extremity deformity management in pediatric orthopedics encompasses a wide variety of congenital and acquired conditions. Understanding etiology and growth remaining is key to planning deformity correction. The growth patterns of the lower extremity, including prior knowledge, information gathered from the patient history, current assessment and growth remaining predictions allow the determination of anticipated consequences of the deformity and leg length discrepancy particularly at skeletal maturity. Guided growth, external fixation, internal fixation, or a combination may be used to correct deformities. Deformity correction and lengthening has the potential to be high risk surgery with a significant complication rates, regardless of method chosen and requires a detailed patient physical, social, and psychological evaluation. This article will cover both the principles of deformity correction and the most recent advances in the field.

Background

Lower extremity growth fluctuates over a child's life, with rapid growth rate in infancy, a plateau in early childhood, and rapid acceleration in adolescence [1] (Fig. 1). Not all limb segments grow at the same rate. The proximal femoral physis grows at approximately 4 mm/year, the distal femoral physis at 10 mm/year, proximal tibial physis at 6 mm/year, and distal tibial physis at 5 mm/year [2]. The rate of growth and growth remaining at each physis needs to be considered when considering hemiepiphysiodesis for deformity correction or epiphysiodesis for leg length correction.

Restoring the mechanical axis is one of the goals of limb deformity surgery. Proper joint alignment aids efficient locomotion and force transfer. Malaligned joints have increased isolated pressure and abnormal mechanics resulting in early osteoarthritis [3–5]. Growth arrest, trauma, and congenital anomalies can cause limb deformity and deviation of the mechanical axis. Alignment is assessed using the mechanical axis of the lower extremity, this consists of a line from the center of the femoral head to the center of the tibial plafond. The goal is to have a neutral mechanical axis with the line passing through the center of the knee. If the mechanical axis deviation is

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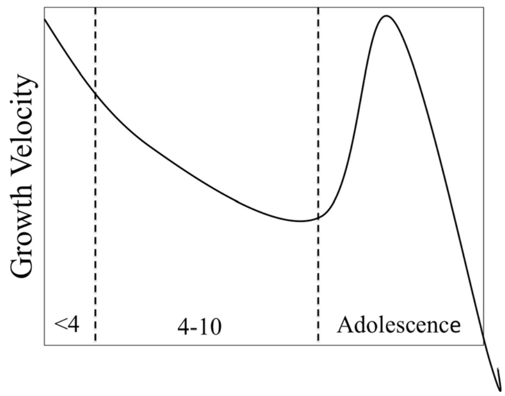

Fig. 1 Growth velocity as a function of age. The growth velocity of a child is high below the age of 4, slows down between the ages of 4 and 10, and then rapidly increases during adolescence to the velocity of an infant, with rapid decline after puberty

medial, the limb is in varus. If the mechanical axis deviation is, the limb is in valgus.

Examination

Evaluating limb length discrepancy and deformity begins with the physical exam. With the child standing, evaluate the spine for scoliosis, as occasionally an apparent leg length discrepancy is the result of scoliosis and fixed pelvic obliquity. Next, examine the height of the iliac crests and balance the iliac crests with the appropriate size block under the short leg. On gait, look for abductor weakness, a short or long leg gait with lurch or vaulting, medial or lateral thrusts, and overall limb alignment. On supine examination, measure apparent limb lengths (from umbilicus to medial malleoli) and the true leg lengths (from ASIS to medial malleoli). Look at overall limb alignment with legs together and either the medial distal femoral condyles or medial malleoli approximated. The distance between the medial distal femoral condyles (for varus) or medial malleoli (for valgus) can be measured. A Galeazzi test can determine if the limb length discrepancy is above or below the knee by assessing relative heights. Range of motion of each joint should be examined as a flexion contracture of the hip, knee, or ankle can give an appearance of a leg length discrepancy. The height of the lower leg on prone examination with the knees flexed can also determine the leg length discrepancy accounted for by the tibia or foot height difference.

Imaging

A hip-to-ankle anterior-posterior radiograph on one long film with the patellae forward is the standard imaging study to assess limb deformity and limb length. Scanogram, radiograph with radio-opaque ruler near the hip, knee and ankle on supine examination, has been compared to hip-to-ankle radiographs and are equivalent [6]. If one limb is significantly shorter, it is best to determine the height of blocks necessary to balance the pelvis under the short limb and have the radiograph taken with these blocks, as a significant short limb can cause the longer limb to bend at the knee and hip, distorting the true alignment and length. Proximal and distal joint angles should be measured using the mechanical or anatomic alignment of each bone segment [7]. The EOS® radiography imaging system (EOS Imaging, France) has been introduced in the last 8 years and reduced radiation by 50–80 %, allows for 3D reconstruction, and has similar measurement reliability to radiographs and CT scan for limb alignment but does not have the image quality of radiographs for fine bony detail [8•].

Clinical decision-making

Once a limb length discrepancy has been determined, the timing of intervention, if intervention at all, must be determined. There are several ways of attempting to predict residual limb length discrepancy at skeletal maturity. The Anderson and Green method uses charts based on a child's height, skeletal age, and growth percentile, and femur or tibia [9]. The Moseley method modified the charts of Anderson and Green into a graph to chart-predicted leg length discrepancy [10]. The multiplier method uses a multiplier of limb length based on age to the short and long limb to determine the difference [11]. Some have compared these methods in accuracy of their prediction [12, 13•]. One study recommended to temporary epiphysiodesis as early as 12 months before predicted age at which the leg length discrepancy due to the underestimation of correction [12]. Another study looked at permanent percutaneous epiphysiodesis and found the Anderson and Green method was the most accurate and had a tendency to overcorrect [13•]. This overcorrection may be due to the lag time in temporary epiphysiodesis methods in creating enough force to stunt the growth of the physis [14].

Growth modulation

There are permanent and reversible methods to modulate a limb with growth remaining. In a growing child, the Hueter-Volkman principle of compression force across a growth plate will slow growth and can be used to change the alignment of the lower extremity. Growth modulation continues to be most commonly performed around the knee, although other physes are amenable their slower growth rate provide more limited indications [15].

Permanent epiphysiodesis was originally described by Phemister involved resecting a portion of the physis, tuning it 180°, and reninserting the bony block [16]; this technique has now largely been replaced. Permanent epiphysiodesis is now more commonly achieved through percutaneous drilling or transphyseal fixation. There are many techniques used today for temporary epiphysiodesis, with no consensus on the most accurate method, as transphyseal screws, tension band plates and staples are all effective [17•]. The most common complication is knee pain, but other complications include deformity and over/under correction of limb length difference [17•]. Most patients remain undercorrected at the time of skeletal maturity after temporary epiphysiodesis [14]. Percutaneous transphyseal screws (Fig. 2) in the femur have a low complication rate but the rate of correction is progressive, with less correction in the first 3 months [14]. Tibial transphyseal screw fixation for epiphysiodesiss has a higher complication rate and tendency to induce a valgus deformity of the tibia [14]. Temporary epiphysiodesis through tension band plating or staples is possible with return of growth once the plates are removed [18]. This technique allows timing to be less precise and the leg lengths to be equalized at an earlier age, as long as the tension implant is removed in a 2-year time period [18]. Complications include inducing deformity at the joint due to placement of the tension device anterior or posterior to the center of the lateral physis and can be corrected by repositioning the implants [19]. Long-term outcomes of temporary epiphysiodesis are favorable, with deformities corrected during treatment time with removing one side of the tether [20].

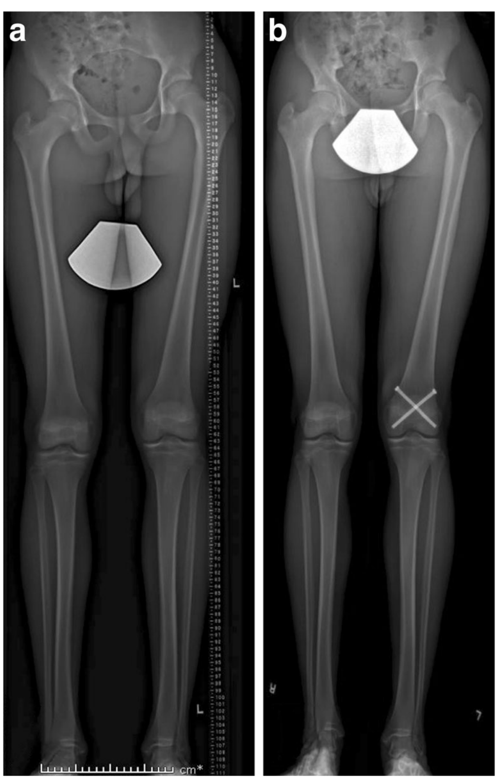

Fig. 2 Transphyseal epiphysiodesis technique for limb length discrepancy of the distal femur. **a** Pre-operative radiograph demonstrating right leg length discrepancy. **b** Radiograph 8 months post-operatively demonstrating some resolution of leg length discrepancy

Hemiephiphysiodesis uses the methods in temporary epiphysiodesis to correct angular deformity (Fig. 3). Staples, transphyseal screws, and tension band plating are all effective in correcting angular deformity. A literature review and quality of life survey determined tension band plating was the most beneficial method of deformity correction in the growing child [21].

Distraction osteogenesis and deformity correction

Long-standing principals continue to apply to limb lengthening surgery. A lengthening rate of no more than 1 mm a day is ideal to have a good regenerate. The osteotomy should be done when possible at the metaphysis due to better blood supply. The osteotomy also should be completed in a tissue-friendly manner: ensure good soft tissue envelope prior to the procedure, preserving the surrounding soft tissues and periosteum, using a drill with irrigation to template the osteotomy site, and completing with an osteotome or a Gigli saw. As a general rule, each centimeter of lengthening requires a month of support from the device (frame or nail) for healing/consolidation. Lengthening no more than 20–25 % of the bone's original is recommended [22].

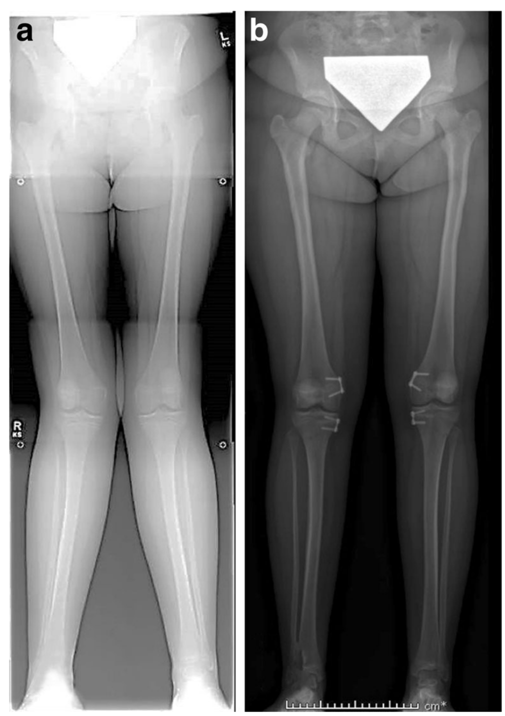

Fig. 3 Hemiepiphysiodesis using medial-based tension band plating for genu valgum. **a** Pre-operative radiographs demonstrating genu valgum. **b** One-year post-operative radiographs with good alignment of mechanical axis

External frames

There has been a move away from external fixation methods over the last 5 years with the development of fixator-assisted internal fixation for deformity correction and advances in intramedullary lengthening devices. There are still many remaining indications for their use including extensive long bone defects, multiplanar and severe deformities and patients with bony anatomy that continues to preclude intramedullary lengthening. External fixation can also be used for multilevel osteotomies, allowing for what was once a two-stage lengthening to be done in one stage, and in a recent study an average length gained of 13.5 cm [23] (Fig. 4).

When considering external fixation for torsional deformity correction, recent literature has shown acute correction of torsional deformities of the tibia and femur simultaneous with lengthening does not affect the rate of consolidation and decreases time compared with gradual torsional correction [24]. Correction of torsional deformity with external fixation and constrained hinges and motors is difficult when using the Ilizarov external fixators. Morasiewicz et al. has shown that the Z-type derotator is more accurate than the H-type or translational-derotational connectors [25]. Treating rotational deformities with external fixation has been made simpler with the Taylor spatial frame and the advent of virtual hinges [26].

Congenital limb length inequality correction continues to have a very high rate of complications (50 to 100 %) [27, 28, 29•]. The literature continues to recommended treating in multiple stages, with 6–7 cm of lengthening or <25 % of the initial length of the bone (whichever is less), as those with >6 cm or >25 % lengthening have worse outcome scores [27, 28, 29•]. Complications include knee and ankle contractures, pin site infections, deformity, delayed union, premature union, fracture of the regenerate, DVT, knee subluxation, vascular injury, and hardware breakage [27, 28]. Many patients with congenital femoral deficiencies undergo stabilization of the hip and knee prior to lengthening with consideration of joint-spanning fixators during lengthening [29•].

In skeletal dysplasias, lengthening for stature remains controversial, as lengthening has high complication rates; however, when patients request lengthening and are correctly indicated, they show good to excellent satisfaction and increased quality of life and self esteem with the results [30–32]. Complications include a high rate of peroneal nerve palsies, increased risk of paralysis due to spinal stenosis with loss of lumbar lordosis on a flat table, fractures, hip/knee/ankle contractures, and pin site infections [30, 31, 33•, 34, 35]. Techniques described are bilateral simultaneous double level tibial osteotomies, simultaneous femur and contralateral tibia, simultaneous bilateral femoral and tibial lengthenings, and staged simultaneous bilateral tibial lengthenings followed by bilateral femoral lengthenings [30, 31, 34, 35]. Prophylactic peroneal decompression, including the foot in the frame, and avoiding spinal anesthesia may reduce some of the complication [30, 36]. The appearance of the regenerate may be an indication to leave the frame on for a longer period, as lateral or central callus had a higher risk of re-fracture in achonroplastic and hypochondroplastic patients [33•]. One author suggested decreasing the rate of distraction to 0.75 mm/day due to poor bone regenerate and showed decreased post-lengthening fracture incidence [34]. There is debate on timing of lengthening in achondroplasia, as some evidence may suggest a growth acceleration in boys and growth retardation in girls; however, recent studies would suggest premature physeal closure of the distal femoral and proximal tibial physis after lengthening when compared with patients with achondroplasia who did not undergo lengthening, resulting in approximately 2 cm of growth loss [37, 38]. Those undergoing stature lengthening for achondroplasia and hypochondroplasia have fewer complications than other short-stature diagnoses, with 0.68 complications per segment [39]. Overall outcomes in cosmetic limb lengthening are good

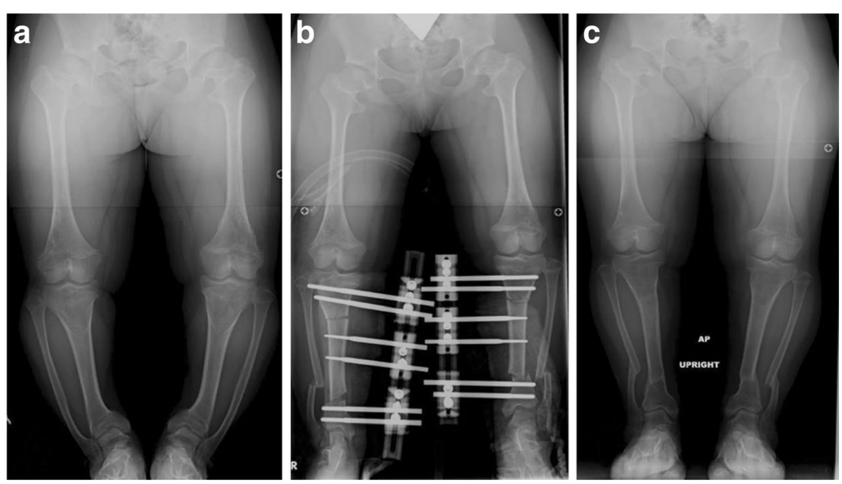

Fig. 4 Tibia vara correction in achondroplaisa using external fixation and double level osteotomy. **a** Pre-operative radiographs demonstrating multilevel varus deformity of the bilateral tibia. **b** Post-operative radiographs demonstrating bilateral two level osteotomies with external fixation. **c** Post-operative radiographs after fixator removal with improvement in mechanical alignment of the tibiae

excellent in 95 % of patients with improved self esteem in 99 % [32, 40]. Self esteem does decrease with increased complications [32]. However, it should be noted that functional scores have not been shown to improve after cosmetic limb lengthening [32].

A combination of external fixation at the time of internal fixation assistance can be used to decrease the time of the frame. One technique called plate-assisted lengthening (PAL) can decrease the time in the frame, although continues to have a high complication rate similar to traditional lengthening procedures, with at least one complication per procedure [41•]. Limbs can also be lengthened with an external fixator over an intramedullary nail, but also has a high rate of complications and concern for deep intramedullary infection due to contact with the external fixator pins [42•].

Treatment of limb length and deformity with an external fixator can be converted internal fixation prior to consolidation of the regenerate [43•]; this strategy involves early exchange of the external fixator for a submuscular plate. Care must be taken to avoid contact with the external fixation tracks when placing the internal fixation device; preparation for this method starts at the time of frame placement. Conversion to plate fixation had a lower rate of complications when compared to intramedullary fixation [43•]. Poor outcomes were associated with conversion to intramedullary devices especially with infected nonunions [43•]. Several authors have published on methods to reduce fractures after fixators; many suggest placing the leg in a cast, some even leaving the external fixator pins in place for several weeks after frame removal, or placement of intramedullary Steinman pins in case of fracture [33•, 34, 35].

Intramedullary lengthening devices

Intramedullary lengthening nails have provided some of the greatest changes in limb lengthening technology. There are several different devices being used, and mechanical oscillation devices have largely been replaced with radiofrequency and magnetic devices. Patient selection and meticulous surgical technique is critical to avoid complications associated with intramedullary devices in deformity surgery. Intramedullary lengthening offers the advantages of no external pins or frame and decreased rates of infection. The disadvantages of the intramedullary nails include the recommendation for removal after consolidation of the regenerate, a need to have or create a straight medullary canal to accommodate the nail both in sagittal and coronal planes, lack of intrinsic ability to protect adjacent joints and others which are specific to the design of the individual nails, and their lengthening mechanism as described below. To avoid anterior notching at the distal aspect of the femur by placing a straight intramedullary nail in a curved bone, Kucukkaya recommended using a rigid reamer along the posterior cortex and had no anterior notching observed using ISKD, Fitbone, or Precice [44•]. Alternate methods for joint stabilization have been described [45].

The intramedullary skeletal kinetic distractor (ISKD, Orthofix, McKinney, USA), which was recently removed from the market, was an intramedullary nail that lengthened by rotating mechanical oscillations between an inner and outer nail, requiring torsion of the leg to distract [46]. Complications included abnormal distraction (runaway nails or difficult to distract), delayed bone healing, joint contractures, with less than 40 % of patients achieving desired length and alignment at final follow-up in one study [47]. Poor regenerate and "runaway nails" were associated with <100 mm of the thick portion of the nail distal to the osteotomy and previous operations [46].

The Fitbone (Wittenstein Intens, Igersheim, Germany) is a motorized intramedullary lengthening nail with an antenna implanted subcutaneously with an external radiofrequency wave transmitter to lengthen the nail. Complications include irritation over the antennae and introduction of recurvatum due to the straight nail. Most obtain proper length and alignment in recently published studies [48, 49].

The Precice nail (NuVasive, USA) is a magnetic intramedullary nail lengthened with an external magnet that must be placed over the motor within the device, this position needs to be recorded for the patient with either a permanent marker (which needs to be refreshed frequently to avoid washing off) or using a scar or other established permanent mark on the leg as reference [50] (Fig. 5). This device mechanism is different from a Fitbone due to the lack of subcutaneous antennae. Due to the requirement for the magnetic field reaching the nail to be sufficient, some have advised limiting BMI to below 35 due to a lengthening magnet range of 5.1 cm [50]. When used, it has been shown to have good accuracy and precision; however, the range of complications observed is similar to other intramedullary devices and those associated with limb lengthening and deformity correction in general. Recent studies have documented failure of the lengthening mechanism, joint contractures above and below the lengthened segment, delayed bone healing, and premature consolidation [51]. One author noted a tendency toward varus-procurvatum malalignment after femoral osteotomies and valgus-procurvatum after tibial osteotomies [51]; however, this is likely to be minimized with appropriate patient selection and surgical technique. The Phenix nail (Phenix Medical, France) is another magnetic intramedullary nail with external magnet for lengthening, however less widely published in the literature however showed similar results to the Precice nail [52].

Few recent studies compare methods of lengthening and correcting deformity. Black et al. compared patients with congenital femoral deficiency treated with external fixator and Fitbone intramedullary lengthening, as congenital femoral deficiency is well known to have multiple complications during

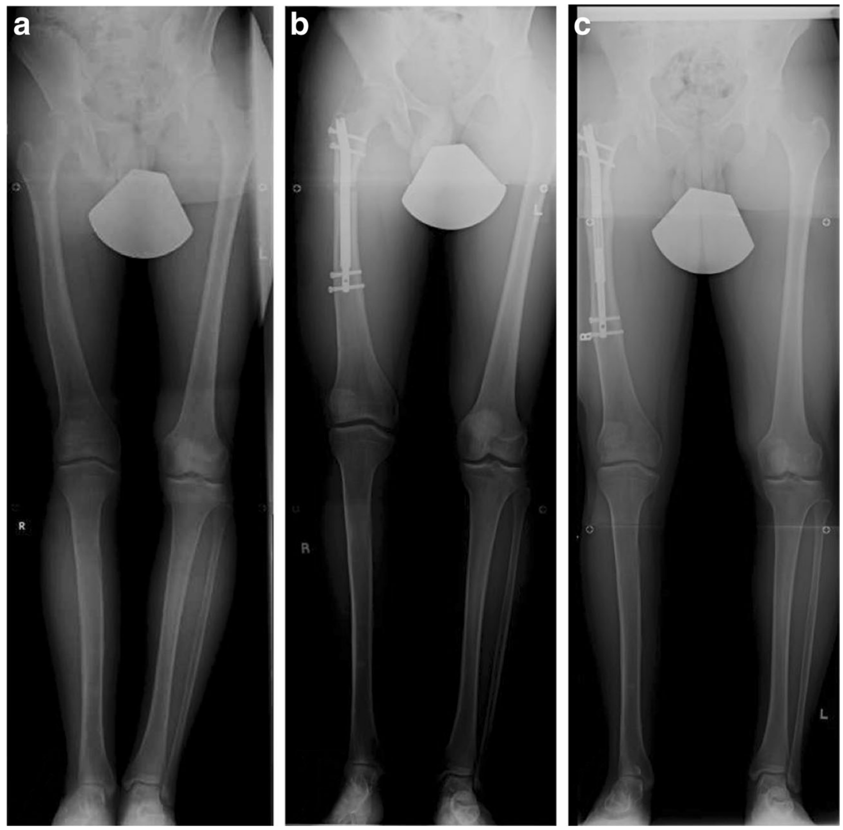

Fig. 5 Magnetic nail lengthening. **a** Pre-operative radiograph demonstrating right femorla shortening. **b** Post-operative radiographs after insertion of Precice nail. **c** Radiograph after lengthening and consolidation of the regenerate

lengthening, most notably subluxation/dislocation of the hip and/or knee joint. The external fixator group had a 100 % complication rate, whereas the intramedullary device had a 73 % complication rate with faster time to fully weight-bear on the affected extremity [53•]. Similar results were found in a heterogeneous group of limb deformity patients, with more knee stiffness requiring a Judet procedure in the external fixation group compared with the Fitbone [49].

Psychosocial considerations

Patient selection and evaluating their social context is important in limb deformity. A child who must be observed, such as in hemiepiphysiodesis or limb lengthening, must have a reliable method of attending regular clinic visits. The psychology and preparedness of the child also must be investigated, as the patient may not be able to cope with several months in an external fixator. In a recent comprehensive study into the psychosocial factors affecting pediatric external fixation, those patients with pre-existing mental health issues, previous operations, and single-parent households had more complications, longer hospital stays, more narcotic usage, and more visits [54•]. Older children were more likely to have a successful outcome, but also required more antibiotics [54•] and longer time in the external fixator. This study supports the ongoing multidisciplinary approach to limb lengthening and deformity correction.

Conclusions

Limb deformity is a complex issue that is easiest addressed in the growing child with a small deformity. For those with a large, complex deformities or shortening, the complications remain high. Although many technologies exist to lessen the complications, they need close observation and anticipation of complications specific to their underlying condition. Regardless of the mechanism chosen, limb lengthening and deformity correction surgery continues to carry significant risk and should be undertaken in the appropriate clinical setting.

Compliance with ethical standards

Conflict of interest Carley Vuillermin declares that she has no conflict of interest.

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Human and animal rights and informed consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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