Specialty Update

What's New in Limb Lengthening and Deformity Correction

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Limb reconstruction surgery continues to affirm itself as a distinct subspecialty worldwide. Apart from the specialty day at the annual American Academy of Orthopaedic Surgeons (AAOS) meeting, the annual Limb Lengthening and Reconstruction Society (LLRS) North American meeting, the combined Association for the Study and Application of the Methods of Ilizarov (ASAMI)/International LLRS (ASAMI/ILLRS) meeting in Brisbane, Queensland, Australia, and the very popular annual deformity courses (Baltimore, Hull, Cairo, and others), national ASAMI meetings were held in many countries. Two new issues of the *Journal of Limb Lengthening & Reconstruction* were published in 2016 with 20 manuscripts, including editorials on why deformity concepts are still not a mainstream part of orthopaedics[1], the evolution of limb lengthening and reconstruction in the post-Ilizarov era[2], and what happened to the regenerate following the numerous modifications of the original Ilizarov technique[3].

Pediatrics

Congenital Limb Deficiencies

A most comprehensive description of surgical treatment of tibial and fibular hemimelia was reported by Paley[4,5]. Eidelman et al. recommended a routine performance of pelvic osteotomy for patients with Paley Type 1B deficiency undergoing lengthening[6]. Saldanha et al. stressed the importance of systematic assessment of the knee prior to any intervention in children with congenital limb deficiencies[7]. Foot preservation and reconstruction of tibial hemimelia were reported to provide good functional outcome[8]. The Ilizarov method provided comparable results in the treatment of congenital pseudarthrosis of the tibia whether it was associated with neurofibromatosis type 1 (NF1) or was idiopathic in origin in pediatric cases[9]. The use of the Fassier-Duval rod as an adjunct procedure in the treatment of congenital pseudarthrosis of the tibia was shown to be successful in obtaining union in 4 children[10]. A new successful technique for the treatment of congenital pseudarthrosis of the tibia was described[11].

Pediatric Trauma

The role of hexapod external fixation in the treatment of tibial fractures in children was discussed by Iobst[12]. Ilizarov bone transport and fibular bone graft for reconstruction of segmental tibial defects were reported to give equally effective results[13]. Shore et al. compared the cost and complications of uniplanar external fixation with those of Taylor Spatial Frame (Smith & Nephew) external fixation in the treatment of pediatric diaphyseal fractures and found that a corrected cost analysis revealed equivalent costs for care delivery[14].

Limb-Length Discrepancy

Mills and Nelson reported on an improved spreadsheet for calculating limb-length discrepancy and epiphysiodesis timing using the multiplier method[15]. Poor efficiency and unpredictability of epiphysiodesis using Eight-Plates (Orthofix) for the treatment of limb-length discrepancy were demonstrated by Gaumétou et al.[16]. Makarov et al. reported a 7% complication rate, with angular deformity being the most common complication, in 863 children who underwent epiphysiodesis for the treatment of limb-length discrepancy[17]. Donnan et al. reported that a 2-ring tibial lengthening fixator was effective in maintaining segmental alignment and callus production during tibial lengthening[18]. In a comparative study evaluating 2 techniques of syndesmotic fixation, transverse tricortical screws and oblique quadricortical screws, the authors found no advantage of 1 technique over the other[19]. Yoshida et al. evaluated the

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What's New in Limb Lengthening and Deformity Correction

complications related to fibular resection during tibial lengthening performed with a Taylor Spatial Frame and recommended fixing the tibial fibular joints by transfixing the wire and/or cannulated screws both proximally and distally to minimize proximal and distal fibular migration[20].

Skeletal Dysplasias

Extensive limb lengthening of >50% of the initial length in patients with achondroplasia or hypochondroplasia was reported to carry a considerable risk of complications[21]. Knee malalignment was evaluated in 581 children with Down syndrome by Duque Orozco et al., who found that 5% had patellofemoral instability and other underlying knee deformities[22], a relevant finding in situations in which limb lengthening is considered. Distraction osteogenesis appears to be an effective and safe approach for the simultaneous correction of the shortness of the first ray as well as the medial angulation of the great toe in patients with Apert syndrome[23]. The mean tibial slope in patients with achondroplasia was found to be substantially more anterior, and this may predispose patients to genu recurvatum[24].

Growth Modulation

Stevens presented an excellent review on various strategies using reversible and serial guided growth in limb lengthening[25]. Farr et al. reported a high rate of radiographic recurrence of frontal plane malalignment after tension-band plating removal[26]. Funk et al. did not recommend hemiepiphysiodeses in patients with late-onset tibial vara with a body mass index of >35 kg/m^2 or a mechanical axis deviation of >80 mm of varus[27]. Hemiepiphysiodesis should be considered in the treatment strategy for limb malalignment in patients with mucopolysaccharidosis Type IV-A[28]. Kemppainen et al. reported a high prevalence of incomplete follow-up (about 12%) in patients undergoing growth modulation, and, of these, nearly one-third required a surgical procedure other than implant removal[29].

Other Pediatric Conditions

In a systematic review of the prevalence of pin-track infection using external fixation, the pediatric population was shown to be at greatest risk[30]. In a study on hexapod external fixators, Iobst et al. demonstrated that the universal joints attached to the undersurface of rings allowed more translational rotation, whereas the ball and socket joints attached to the outer surface of rings allowed more rotational correction and more correction with less strut change in patients with severe deformity[31]. The mechanisms by which isolated distal tibial rotation osteotomy corrects excessive tibial torsion in children with cerebral palsy were extensively investigated[32]. The results of a double-blind, multicenter, randomized controlled trial comparing Botox (botulinum toxin type A) injection with placebo in children undergoing lower-limb lengthening supported the use of Botox as an adjunct treatment in limb lengthening[33].

Adult Limb Reconstruction

Osteotomy About the Knee: High Tibial Osteotomy

Measurement of alignment before and after high tibial osteotomy was best performed with standing long-leg radiographs. The reproducibility and reliability of alignment measurement were better than for knee radiographs and were almost as good as a computed tomographic (CT) scan. The extra time and specialized software needed for CT scan measurement were not worthwhile[34]. Single-photon emission CT and conventional CT (SPECT/CT) were used to evaluate knee compartments after opening-wedge high tibial osteotomy for providing clinical guidance for proper correction. Optimal realignment requires overcorrection that does not exceed 3° valgus. An increased signal was noted in the patellofemoral joint when the patellar position was changed[35,36].

A systematic review of 19 high tibial osteotomy studies including 1,189 patients showed that 87% returned to sport and 85% returned to work. Of those patients who did return to work and sport, 90% did so within 1 year. At a mean of 7 years, 93% of the patients did not progress to total knee replacement[37].

Medial opening-wedge high tibial osteotomy was performed on 18 patients with malunited tibial plateau fractures (excluding complex intra-articular deformity and severe arthritis) using a plate and bone graft. Coronal and sagittal plane alignment and the Knee injury and Osteoarthritis Outcome Score (KOOS) significantly improved with surgical procedures[38].

Osteotomy About the Knee: Distal Femoral Osteotomy

Distal femoral osteotomy using an opening-wedge technique on 41 patients with valgus deformity resulted in 95% accuracy of correction using mechanical axis deviation, lateral distal femoral angle, and medial proximal tibial angle measurements[39], and outcomes using the Short Form-36 (SF-36) and AAOS lower limb module (LLM) were significantly improved. In 10 knees with patellar subluxation, the patella congruence angle significantly improved after distal femoral osteotomy[39].

Opening-wedge distal femoral osteotomy was used in 29 knees, and the angular correction was 8°, changing the mechanical axis angle from 188° to 180°. Lower-limb length was not significantly changed. At 5 years, 91% did not progress to total knee replacement[40].

In patients with hypoplasia of the lateral femoral condyle, an intra-articular osteotomy with advancement of the lateral femoral condyle led to improvement in the tibiofemoral angle from 35° to 9° and satisfactory outcomes[41].

Total Knee Arthroplasty

Outcomes at 1 year after total knee replacement with high tibial osteotomy were no different from those performed without previous high tibial osteotomy. Osteotomy is an effective means of delaying total knee replacement without compromise of outcome, especially when high tibial osteotomy techniques are opening-wedge with a plate or using external fixation[42].

What's New in Limb Lengthening and Deformity Correction

A systematic review of 8 studies examining total knee arthroplasty after medial opening and lateral closing-wedge high tibial osteotomy showed similar performance. Clinical and radiographic outcomes, including revision rates, did not differ; however, there were more surgical technical concerns in conversion to total knee arthroplasty in the lateral closing-wedge high tibial osteotomy group than in the medial opening-wedge high tibial osteotomy group[43].

Following total knee arthroplasty, functional outcomes of patients with >15 mm of postoperative limb-length discrepancy were lower than those with <15 mm of limb-length discrepancy. This calls attention to the importance of limb-length discrepancy as it relates to functional outcomes of primary total knee arthroplasty[35].

Alignment

Although 10% of patients perceive limb-length discrepancy after total knee replacement, this resolves in most within 3 months and is not correlated with mechanical alignment[44]. A systematic review of 18 studies and 2,214 patients concluded that malalignment may correlate with lower patient-reported outcome scores. Hadi et al. called for larger longitudinal studies with standardized methods for assessing alignment[45].

Remote-Controlled Internal Lengthening Nail

A magnetic, remote-controlled intramedullary nail was used to successfully lengthen the femur and tibia in 23 patients. The mean lengthening was 48 mm, the mean angular correction was 16°, and the mean consolidation index was 1.12 months/cm. Although 8 patients had overlengthening, the nails were driven back to the desired length[46]. Another study examined the experience with these nails in 10 patients with a mean limb-length discrepancy of 4.7 cm. In all patients, the limb-lengthening goals were reached within 0.5 cm after a mean time of 53 days. However, in 2 patients, mechanical failures with unintended shortening were observed. In 1 patient, nail breakage occurred[47]. In another review of 9 patients, the mean lengthening (and standard deviation) was 34.7 ± 10.7 mm. All patients reached normal alignment and normal joint orientation. An unintentional loss of the achieved length during the consolidation phase was noticed in patients with delayed bone healing in 2 cases. In the first case (loss of 20-mm distraction), the nail could be re-distracted and the goal length was achieved. In the second case (loss of 10-mm distraction), the nail broke shortly after the diagnosis and the nail was exchanged. Weight-bearing restrictions are necessary until consolidation[48].

The use of blocking screws is necessary to correct and prevent deformity when using the internal lengthening nail technique. A systematic approach to the appropriate use of blocking screws in these deformities is described, including the reverse rule of thumb as a quick reference to determine the ideal location(s) and number of blocking screws[49].

A comparison of femoral lengthening with the internal nail and a monolateral fixator showed quicker osseous union with the nail. The nailing also had fewer complications, was better tolerated by patients, and was perceived to have a better cosmetic result[50].

Strategies to avoid anterior nail impingement include the use of rigid straight reamers. In a series of 45 retrograde femoral lengthenings, the mean lengthening was 5.9 cm and the consolidation index was 1.05 months/cm. The mean distance from the osteotomy site to the intercondylar notch of the femur was 81 mm. The mean posterior cortex reaming thickness was 3.7 mm. No impingement-related complications or nail damage were observed[51].

Many complications can be avoided with attention to technical detail and with increased experience. Technique papers for antegrade femoral lengthening and retrograde femoral lengthening using the magnetic internal nail include indications, technical tips, and pitfalls to avoid. Mechanical stability with optimal nail sizing, osteotomy location or technique, reaming technique, rate and rhythm of distraction, use of blocking screws, and adjuvant soft-tissue procedures are important to achieve success[52,53].

Other Adult Reconstruction

Neglected patellar tendon rupture with massive proximal patellar migration was successfully treated in 2 cases with patellar transport using a circular external fixator and staged allograft reconstruction[54]. Neglected knee dislocation with residual deformity was successfully treated using a 2-stage strategy with gradual reduction using the Ilizarov technique and subsequent arthroscopic anterior and posterior cruciate ligament reconstruction[55]. Knee flexion contractures were treated using a circular external fixator and gradual correction, and one-third of the patients were simultaneously treated for ankle equinus. The mean range of motion at the time of the final follow-up was −10° extension, 64° flexion, 9° ankle dorsiflexion, and 29° ankle plantar flexion. The arc of motion in the knee was unchanged but was increased in the ankle. The difference between preoperative and postoperative ranges of motion was significant. All but 2 patients were able to stand and walk with fewer aids or no aids[56].

Despite numerous studies that work to derive evidence-based recommendations for the prevention of pin-site infections, substantial controversy exists with regard to the optimal protocol. Kazmers et al. comprehensively evaluated the current literature to provide an overview of factors that may influence the incidence of pin-site infections in patients undergoing treatment with external fixators and provided a description of the preferred surgical and postoperative pin-site protocols that they employed[57].

Lengthening of a short residual tibia after transtibial amputation by >100% was achieved with no complications. Moreover, a special, custom-made, external fixator-prosthesis composite was described to allow for early weight-bearing and exercising[58]. Both the Gigli saw and De Bastiani corticotomy techniques result in good bone formation following distraction

What's New in Limb Lengthening and Deformity Correction

osteogenesis of the tibia. The anterior tibial cortex consolidates more slowly than the other cortices in both groups. This is likely due to deficient soft-tissue coverage and direct periosteal damage at the time of osteotomy[59].

Trauma

Successful reconstruction of bone defects remains a surgical challenge. A critical bone defect suggests a fracture gap that will not undergo healing without some form of intervention (e.g., bone-grafting). Currently, there is no consensus on the lower-limit size of a critical bone defect. Haines et al. studied 40 patients with open tibial diaphyseal fractures to define this lower limit[60]. They included patients with gap sizes between 10 and 50 mm. Twenty-one patients (52.5%) developed a nonunion. The authors concluded that a fracture gap after intramedullary nailing of <25 mm had a reasonable probability of achieving union, whereas a larger gap likely warrants secondary bone-grafting[60]. Bone morphogenetic protein-2 (BMP-2) has been used as a bone graft in such defects. In a rat bone defect model, Glatt et al. demonstrated that, by optimizing the mechanical environment, one can enhance BMP-2's effect on bone growth[61]. They reported that either a medium-stiffness external fixator or reverse dynamization could decrease the amount of BMP-2 necessary to generate the same amount of bridging callus. Reverse dynamization was defined as beginning with a low-stiffness external fixator that changed to a stiff external fixator at 2 weeks after an osteotomy[61].

Infected nonunions with associated bone defects remain a surgical challenge. The Ilizarov method of distraction osteogenesis with circular ring fixators has been shown to be successful in eradicating infection, equalizing limb lengths, and allowing soft-tissue lesions to heal[62-65]. This is especially true in the case of periarticular bone defects in which surface implants or intramedullary nails cannot achieve the stability required for successful reconstruction. Circular frames that are mounted juxta-articularly using 1.8-mm Ilizarov wires that are tensioned to 130 kg achieve the stability required to promote bone union, to correct deformity, and to eradicate infection. Eralp et al. retrospectively reported on 13 patients who underwent distal tibial periarticular reconstruction[66]. The mean bone loss was 4.8 mm. At a mean follow-up of 36 months, 92% (12 patients) achieved union with a mean external fixation index of 29 days/cm[66]. Repo et al. published a mean 6.6-year follow-up on 16 patients who underwent tibial reconstruction with a mean bone gain of 3.8 cm[67]. All patients had use of a latissimus dorsi free flap for the soft-tissue defect. Bone transport was performed using various circular external fixators. The external fixation index was 54 days/cm. Although a lengthy and, at times, arduous process for the patient, the health-related quality-of-life measures that were reported were comparable with age-standardized population samples[67].

A hexapod circular fixator is extremely helpful in correcting complex deformities because the surgeon does not need to build a physical hinge at the apex of the deformity (as in a classic Ilizarov circular frame). Instead, a virtual hinge is created through the computer software and an algorithm with an accompanying schedule dictates how to move the struts to make the bone straight. Hughes et al. used a temporary hexapod frame intraoperatively to acutely correct complex tibial deformities followed by the insertion of an intramedullary nail[68]. Thirteen procedures were performed in 12 patients. The tibial mechanical axis was restored to anatomic values; however, 2 patients developed a common peroneal nerve palsy, and 1 patient developed a tibial artery pseudoaneurysm[68]. Although the hexapod frame can correct complex deformities, its other benefit is the ability for gradual correction. This needs to be considered for large angular corrections.

Anatomic alignment in fracture reconstruction remains dogmatic. Weinberg et al. studied 37 fractured tibiae in cadaveric skeletons[69]. Intra-articular fractures were excluded. They correlated knee arthritis with >5° of coronal malreduction ($p = 0.006$) and malrotation of >10° ($p = 0.004$)[69]. Hip arthritis was associated with tibial shortening of >10 mm ($p = 0.009$). Avilucea et al. compared 266 patients with distal tibial fractures with respect to whether postoperative angulation of >5° occurred[70]. The lateral distal tibial angle and anterior distal tibial angle were measured on postoperative tibial radiographs. Twenty-six percent of patients in the infrapatellar group compared with 3.8% of patients in the suprapatellar group had >5° of coronal or sagittal malalignment ($p < 0.0001$)[70].

Baruah published a case report on using a modified Ilizarov external fixator to gradually reduce a chronic patellar fracture nonunion[71]. Yang et al. reported on 7 patients with tibial plateau malunions who were treated with intra-articular osteotomies[72]. Preoperative planning was achieved with templating off a 3-dimensional printed model of the malunion.

Combining internal fixation (intramedullary nails or plates) with external fixation during limb lengthening and deformity correction is a strategy to decrease the amount of time that the patient needs to remain in the circular frame while the regenerate heals (latency phase). Burghardt et al. compared patients who underwent classic Ilizarov lengthening with a matched cohort who underwent lengthening over an intramedullary nail[73]. The mean external fixation time for the group that underwent lengthening over an intramedullary nail was 2.6 months compared with 7.6 months in the classic Ilizarov lengthening group. Of note, there were 4 deep infections, increased blood loss, and an increase in surgical cost in the group who underwent lengthening over an intramedullary nail[73]. Watanabe and Matsushita performed deformity correction of 21 femoral nonunions with a specific type of osteotomy at the nonunion site called the chipping technique[74]. No bone grafts were utilized, and all nonunions healed with 1 procedure (19 cases) or 2 procedures (2 cases)[74]. Azzam and El-Sayed performed lengthening over an existing femoral intramedullary nail for 13 diaphyseal femoral nonunions[75]. The mean length achieved was 4 cm, and union occurred in 11 cases (85%)[75].

What's New in Limb Lengthening and Deformity Correction

Foot and Ankle

Ankle and Hindfoot

Although supramalleolar osteotomy has been successful for treating distal tibial malalignment with ankle arthritis[76], neither supramalleolar osteotomy nor calcaneal osteotomy will correct an intra-articular varus or valgus deformity at its apex. Plafondplasty, an intra-articular realignment akin to hemiepiphyseal elevation, has been implemented to remedy the asymmetric ankle joint line deformity at the source[77]. The article by Al-Nammari and Myerson[77] was technique-based and highlighted the need for lateral ligament reconstruction for the joint line varus.

Forefoot

Ginés-Cespedosa et al. were unable to find a correlation between the presence of uncorrected hindfoot deformity and the outcome of hallux valgus corrective surgical procedures[78]. This finding undermines the teaching that a valgus hindfoot creates hallux valgus.

Brachymetatarsia is amenable to distraction osteogenesis, with a low complication rate[79], but when the regenerate does not heal, the resulting defect can be treated with bone-grafting using a bioabsorbable hydroxyapatite-collagen composite[80]. The authors emphasized the need for preservation of the periosteum and the existing thin regenerate bone during grafting. An alternative treatment for this defect could have been an injectable self-contained phosphate-releasing scaffold to promote biomineralization[81].

Basic Science

Bhave et al. studied the effect of femoral lengthening, with either lengthening over a nail or Ilizarov classic methods, determining that patients with congenital femoral shortening who underwent lengthening had a higher incidence of loss of terminal knee flexion at a minimum follow-up of 2 years[82]. Burzyńska et al. showed that, in small children, these life-sized models (10 cm long) could be used to mount external fixators and to trial deformity correction surgical procedures[83]. Morasiewicz et al. examined weight distribution and balance after Ilizarov osteotomy reconstruction surgical procedures and found that limb realignment and length equalization restored these parameters to close to normal values[84].

Adjuvant Intervention for Bone Healing

Understanding the biological principles of distraction osteogenesis is the first step toward augmenting bone formation. A fantastic review focusing on similarities between clinically observed regenerate and basic in vivo tissue engineering was published[85]. In a distraction osteogenesis model, researchers found that slower distraction rates were associated with osteogenesis originating from the bone end and a faster distraction rate created periosteal new bone with a thicker and more voluminous regenerate[86]. In a meta-analysis of clinical distraction osteogenesis publications, both low-intensity pulsed ultrasound (LIPUS) and pulsed electromagnetic fields were shown to significantly reduce time to healing by a mean time of 11 days/cm[87]. A separate meta-analysis of clinical distraction osteogenesis cases showed that LIPUS lowered healing time by a mean time of 15 days/cm[88].

Promising osteogenesis-promoting adjuvants include sclerostin antibody injection[89], BMP-2 with osteoprotegerin[90], human fetal mesenchymal stem cell secretome[91], and staphylococcal enterotoxin[92].

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WHAT'S NEW IN LIMB LENGTHENING AND DEFORMITY CORRECTION

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What's New in Limb Lengthening and Deformity Correction

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