# SPECIALTY UPDATE What's New in Limb Lengthening and Deformity Correction

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The Limb Lengthening and Reconstruction Society (LLRS) (llrs.org) represents a distinct subspecialty within orthopaedics. We had another successful Specialty Day at the 2018 American Academy of Orthopaedic Surgeons (AAOS) meeting, where we liaised with the American Orthopaedic Foot & Ankle Society (AOFAS) for exciting discussions on the salvage of failed total ankle replacement and ankle distraction arthroplasty, to name a few topics. The official journal of the LLRS, the Journal of Limb Lengthening & Reconstruction, continued to publish meaningful articles in 2017. Additionally, the LLRS traveling fellowship has been immensely successful, immersing young attending physicians into limb deformity and limb salvage mini-apprenticeships. Our annual LLRS meeting was held in San Francisco, California, in July 2018. The focus of this Specialty Update was to provide a summary of the most impactful articles related to limb alignment, limb lengthening, and joint preservation that were published in 2017.

#### **Pediatrics**

#### **Growth Modulation**

In growing children, asymmetric tethering of the physis can lead to angular corrections in both the coronal and sagittal planes. McClure et al. reported that growth modulation was effective for deformity correction in 4 children with achondroplasia<sup>1</sup>. Leveille et al. concluded that the rebound phenomenon was common in 45 patients who underwent growth modulation and that an older age and larger initial deformity increase the risk of rebound; the authors recommended against routine overcorrection<sup>2</sup>. Welborn and Stevens described the

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successful use of growth modulation in children with focal fibrocartilaginous dysplasia<sup>3</sup>. In a retrospective study of 70 limbs with genu valgum deformity, Kang et al. reported that, in patients with multiple exostosis, temporary hemi-epiphysiodesis should be considered at an earlier age compared with those with idiopathic deformity<sup>4</sup>.

Idealized finite element models of the juvenile distal part of the femur treated with the 8-plate implant were developed for varus and valgus deformities and limb-lengthening deformity, and finite element models correlated with clinical findings<sup>5</sup>.

### **Congenital Limb Deficiencies**

In a review of 32 patients with congenital deficiency of the fibula (9 patients with an extension prosthesis and 23 who underwent amputation), Calder et al. concluded that childhood amputation for severe limb-length inequality and foot deformity offers an excellent short-term functional outcome and that an extension prosthesis without amputation offers reasonable long-term function, although the outcome scores were lower<sup>6</sup>. Kaastad et al. reviewed physical function and health-related quality of life in young adults with unilateral congenital lower-limb deficiencies and compared patients who had surgical lengthening with those who had a lengthening prosthesis<sup>7</sup>. In a radiographic review of 28 feet with congenital fibular deficiencies, Reyes et al. concluded that congenital fibular deficiency should not be viewed as a global lateral lowerlimb deficiency and the foot ray deficiency should not be viewed as being lateral<sup>8</sup>.

#### Limb Lengthening

In a review of 176 femoral lengthening procedures with external fixators, Burke et al. reported that femoral lengthening

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of >50 mm increases the risk of fracture at the regenerate<sup>9</sup>. Abdelgawad et al. provided a detailed description of a surgical technique of prophylactic Rush rod insertion through the tip of the greater trochanter following the removal of external fixators for lengthening in congenital femoral deficiency<sup>10</sup>. Georgiadis et al. described the technique of plate-assisted lengthening of the femur and tibia in 38 lengthenings<sup>11</sup>. Hammouda et al. described the trochanteric-entry approach for a magnetic femoral lengthening nail in children in a retrospective review of 31 femora<sup>12</sup>.

Bayhan et al. compared the results of 24 patients who had 8-plate application for the treatment of limb-length discrepancy with those of 48 patients who had percutaneous epiphysidesis for limb-length discrepancy and concluded that both methods are effective for the correction of limb-length discrepancy; however, the percutaneous epiphysiodesis led to greater improvement during the same follow-up time with fewer complications and less need for additional procedures<sup>13</sup>. Goote et al. reviewed the results of 40 segmental limblengthening surgical procedures in pediatric patients with Ollier disease and concluded that the enchondromatous bone appears to respond to lengthening with a structurally proper regenerate and the location of the corticotomy site, whether intralesional or extralesional, did not appear to have an impact on the healing response. Premature consolidation remains a concern in these patients<sup>14</sup>.

Scollan et al. performed a meta-analysis study to identify the complications in children with osteogenesis imperfecta who underwent lower-extremity correction, using nonelongating intramedullary nails, and reported a reoperation rate of nearly 40% of all procedures, mostly due to rod migration or bone-related complications<sup>15</sup>. In a review of children with hypophosphatemic rickets and lower-limb deformity, Horn et al. concluded that guided growth is a successful procedure to correct coronal plane deformity in these patients<sup>16</sup>. Franzone et al. reviewed 45 upper-extremity rod treatments in children with osteogenesis imperfecta and concluded that realignment and rodding reduced the fracture rate of the humerus and forearm<sup>17</sup>. van Bosse and Saldana described a detailed surgical technique of reorientation proximal femoral osteotomy in 65 children with arthrogryposis and severe hip deformity<sup>18</sup>. Iobst reported that preoperative planning, meticulous surgical technique, patient education, and close patient monitoring were all critical to minimize pin-track infection and decrease the overall 27.4% of pin-track infections reported in the litterature<sup>19</sup>. In a systematic review of the patient-reported outcomes in children with lower-limb deformities, Chhina et al. found that there were no validated patient-reported outcome instruments specifically designed to measure the quality of life in these children<sup>20</sup>. Cherkashin et al. performed an assessment of the distraction rate in hexapod compared with unilateral fixators<sup>21</sup>. Although the unilateral and Ilizarov fixators distract in the direction of bone lengthening, the hexapod fixators do not distract in the direction of bone lengthening. The authors demonstrated that the oblique orientation of the struts relative to the ring causes a discrepancy between the amount of strut and the actual bone lengthening, and that a 1-mm lengthening of each of the 6 struts always created >1-mm lengthening at the bone.

## **Adult Reconstruction**

## Intramedullary Nailing

Blocking screws, usually placed on the concave side, have been used to correct deformity during intramedullary lengthening. With the use of blocking screws, a higher degree of deformity correction was demonstrated with a slightly higher precision compared with that in patients without blocking screws<sup>22</sup>.

A systematic review was performed to compare the lengthening and then nailing technique with the conventional Ilizarov method. In the study, 183 limbs were lengthened combined with an intramedullary nail and 171 limbs were lengthened conventionally. The external fixation index and consolidation index were significantly lower for lengthening and then nailing<sup>23</sup>.

Computer hexapod-assisted orthopaedic surgery allows correction of long bone deformities using a hexapod external fixator before internal fixation with minimally invasive stabilization techniques of locked plating or intramedullary nailing. Satisfactory deformity correction and a low complication rate were reported in 49 patients who underwent femoral reconstruction<sup>24</sup>.

Review articles on acute deformity correction and lengthening with a retrograde femoral internal lengthening<sup>25</sup> and with a tibial nail<sup>26</sup> were published. Technical aspects were discussed, including design issues, preoperative planning, selection of nail length, the use of blocking screws, intraoperative external fixation assistance, osteotomy, postoperative management, and cost analysis<sup>25,26</sup>.

A review article provided a historical account of the intramedullary device and its impact on limb lengthening. The PRECICE nail (Nuvasive Specialized Orthopaedics) was considered the implant of choice for femoral lengthening because of its accuracy, robust inventory, and few complications<sup>27</sup>.

## Knee

Substantial improvements in biomechanical risk factors (mechanical axis angle and knee adduction moment) and patient-reported outcomes (Knee injury and Osteoarthritis Outcome Score [KOOS]) in 170 patients were observed 5 years after medial opening-wedge tibial osteotomy<sup>28</sup>.

The relationship between tibia vara and external tibial torsion was studied in 138 limbs in 69 adults. Almost 50% of the limbs with tibia vara also had external tibial torsion<sup>29</sup>.

Clinical evidence concerning the impact of osseous malalignment and realignment procedures in knee ligament surgical procedures was systematically reviewed and the mechanical axis was found to play an important role in knee joint stability<sup>30</sup>.

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Experience with elevation of a tibial hemiplateau or femoral condylar advancement combined with a metaphyseal osteotomy for lengthening was reported. The hemi-joint line convergence angle improved from 16° to 0.4°, the overall mechanical axis deviation improved from 70 to 9 mm, and the limb-length discrepancy improved from 6 to 1 cm in 6 patients with good outcomes<sup>31</sup>.

Tibial tubercle transfer and lateral release was compared with tibial tubercle transfer, lateral release, and medial patellofemoral ligament reconstruction. Reconstruction of the medial patellofemoral ligament combined with the tibial tubercle transfer and lateral release resulted in improved alignment parameters (patellar congruency angle and patellar tilt angle) and patient satisfaction. There were no differences in outcome scores between the 2 groups at any time period<sup>32</sup>.

Distal femoral osteotomy with an opening-wedge technique has been shown to be effective for accurately correcting femoral valgus deformity<sup>33</sup>.

In patients with valgus femoral and lateral patellar instability, the distal femoral osteotomy and lateral release served to medialize the patellar tendon insertion and decrease the Q angle. The patellar congruency angle improved from  $30.4^{\circ}$  to  $5.7^{\circ}$ . Similarly, the mechanical axis deviation, lateral distal femoral angle, visual analog scale (VAS) for pain, and Kujala score significantly improved<sup>34</sup>.

Knee joint distraction is a relatively new joint-preserving procedure with the goal of delaying total knee replacement in young and middle-aged patients. Sixty patients who were  $\leq 65$  years of age and had end-stage knee osteoarthritis were randomized to either knee joint distraction or total knee arthroplasty. All patient-reported outcome measures improved significantly over 1 year in both groups. In the knee joint distraction group, the joint space width increased significantly. Knee joint distraction did not demonstrate inferiority of outcomes at 1 year when compared with total knee arthroplasty<sup>35</sup>.

Currently, the main indication for knee arthrodesis is septic failure of a total knee arthroplasty. A study of 19 patients who underwent knee arthrodesis using the Ilizarov method was published. The Knee Society Scores were significantly improved, and 94% achieved fusion. The final limb-length discrepancy was 4 cm. Major complications occurred, mainly in patients who were >75 years of age<sup>36</sup>.

#### **External Fixation**

A simple and practical technique to improve the quality of hexapod frame radiography involves centering the beam on the designated ring. This implementation has demonstrated an improvement in the adequacy of planning radiographs and a reduction in the requirement for repeat imaging<sup>37</sup>.

Although weight-bearing is generally encouraged in patients treated with the Ilizarov apparatus, it has been demonstrated that, during the first week after the surgical procedure, patients are not able to bear full weight on the affected limb. As bone union progresses, weight-bearing is expected to increase<sup>38</sup>. Ilizarov circular frames in patients  $\geq$ 65 years who were treated for fractures, nonunions, deformity correction, and ankle fusion were studied. Mortality and complication rates were low (5% to 7%), and Short Form-12 (SF-12) results returned to normal for that age group<sup>39</sup>.

Twenty-eight patients with severe leg trauma complicated by bone loss, soft-tissue loss, and infection treated with the Ilizarov method were reviewed. The infection was eliminated in all cases. All of the soft-tissue and skeletal defects healed during bone transport, and only 2 cases required plastic surgery. All patients underwent a staged procedure at the docking site to fashion the soft tissue and to cover the bone ends. The Association for the Study and Application of the Method of Ilizarov (ASAMI) bone scores were good or excellent in 26 cases, and functional scores were good or excellent in 25 cases<sup>40</sup>.

The use of prophylactic oral antibiotics does not seem to impact postoperative pin-track infection. In a randomized controlled trial, the incidence of pin-track infection during the 90-day observation period was 46 (79%) of 58, but no differences were noted whether or not patients received prophylactic antibiotics<sup>41</sup>.

A survey was taken of external fixation surgeons to determine their current practice patterns with regard to external fixator removal. The most common criteria used for frame removal were full weight-bearing, 3 cortices on radiographs, no pain with walking, after successful dynamization, and duration of time. Routine dynamization of frames was practiced by 48% of surgeons<sup>42</sup>.

#### Upper Extremity

Although off-label in the United States at this time, the PRE-CICE internal lengthening nail has been used for humeral lengthening<sup>43</sup>. A retrospective study included a total of 6 humeri in 5 patients (mean age, 20 years) who underwent lengthening with the PRECICE nail in 2 centers in the United States. The goals of lengthening (mean, 5.1 cm) were achieved in all patients. The mean distraction index was 0.7 mm/day, and the mean consolidation index was 36 days per centimeter lengthened. No complications were observed, and patients maintained their preoperative range of motion. The abbreviated version of the Disabilities of the Arm, Shoulder and Hand (QuickDASH) and upper-extremity functional index scores showed improvement<sup>44</sup>.

Metacarpal bone lengthening can obtain functionally successful results and improvement in body image in adults with congenital hand deformity. Seven metacarpal lengthenings were studied. The mean percent lengthening was 44% and the mean distraction rate was 0.43 mm/day<sup>45</sup>.

## Hip

Hip joint instability can be secondary to congenital hip pathologies such as developmental dysplasia or can be the sequelae of an infective or neoplastic process. The Ilizarov hip THE JOURNAL OF BONE & JOINT SURGERY · JBJS.ORG VOLUME 100-A · NUMBER 16 · AUGUST 15, 2018 What's New in Limb Lengthening and Deformity Correction

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reconstruction includes a proximal femoral valgus extension osteotomy for the treatment of the unstable hip joint combined with a distal femoral varus osteotomy for limb lengthening and the correction of the mechanical axis of the leg. This reduces the limp, improves the Trendelenburg gait and leg length, and provides stability to the destroyed hip joint<sup>46</sup>.

Patients with advanced hip arthritis can present with limb-length discrepancy related to shortening from growth arrest, proximal hip migration, soft-tissue contractures, and pelvic obliquity. The patient perceives a limb-length discrepancy that is a combination of a true plus apparent limb-length discrepancy. In a series of patients who underwent primary total hip replacement and auxiliary soft-tissue procedures, the mean perceived limb-length discrepancy significantly improved from 8 to 1 cm, and all patients were walking without any assistive devices and were satisfied with their surgical outcome<sup>47</sup>.

#### **Foot and Ankle**

Ankle distraction arthroplasty was the main topic at the 2018 AAOS combined specialty day in which members of the AOFAS and the LLRS debated the results and nuances of this regenerative method. Several review articles published in 2017 addressed the increasing popularity of this treatment alternative<sup>48-50</sup>. Although popularized in the West, ankle distraction has gained momentum in China, with 3 distinct centers publishing their clinical results in a single year. In a retrospective study of 46 patients with moderate ankle osteoarthritis treated with ankle distraction arthroplasty, overall success was seen in 61% of patients. Outcomes were measured with both Ankle Osteoarthritis Scale and AOFAS scores at a mean follow-up of 43 months postoperatively. Risk factors for failure included intra-articular joint deformity of  $\geq 5^{\circ}$  and obesity. Sex was not significant<sup>51</sup>. In a separate publication, the same authors reported a case of a patient with a large, intra-articular varus deformity (who would have performed poorly with distraction arthroplasty alone according to the previous study<sup>51</sup>) treated with a combined approach of supramalleolar osteotomy and medial joint distraction aimed at the correction of all deformity. The surgeons noted that the medial joint space maintained normal alignment at a 3-year follow-up with excellent clinical results<sup>52</sup>. Zhang et al. performed a comparison study between distraction arthroplasty alone and arthroscopic ankle microfracture with concomitant ankle distraction<sup>53</sup>. In a series of 96 patients with a mean follow-up of 31 months, both groups showed mostly good and excellent results in the Ankle Activity Score and AOFAS score, with more improvement seen with the combined approach. In a separate investigation, 16 patients were followed for a mean time of 41 months after ankle distraction surgery, and outcome scores and native joint survival were recorded<sup>54</sup>. Success was seen in 56% of patients, with a correlation noted between increased radiographic joint space at 3 years and a good outcome.

End-stage ankle arthritis was treated with ankle arthrodesis using either a circular external fixator or an internal plate and screws in 59 patients<sup>55</sup>. Outcomes showed little difference except for a faster time to union with the frame. Both groups enjoyed excellent results with significant improvement in VAS and AOFAS hindfoot scores. Morasiewicz et al. studied ankle arthrodesis with circular external fixation compared with internal fixation in 62 patients and found that the external fixation cohort had more accurate alignment, higher union rates, and less adjacent-joint arthritis at the time of the latest follow-up (mean, 43 months)<sup>56</sup>. Ankle arthrodesis used to treat Charcot neuroarthropathic ankle collapse can be performed with different approaches. In a prospective cohort of 27 patients, investigators randomized patients into 2 treatment groups, tibiotalacalcaneal arthrodesis with an intramedullary nail or tibiotalar arthrodesis with an Ilizarov external fixator. Analysis showed that the fixator cohort healed faster and more reliably but that they had many more complications<sup>57</sup>. An alternative approach to manage this challenging group of patients consists of using both an ankle arthrodesis nail and an external fixator simultaneously. This strategy provides increased stability to the reconstructed ankle, shortens time in the frame, and prevents refracture after frame removal. In a series of 24 patients with diabetic Charcot arthropathy treated with both methods, the union rate was 92% with no broken implants or deep infections<sup>58</sup>.

#### **Biomechanics and In Vivo Research**

Morasiewicz et al. hypothesized that limb reconstruction with the Ilizarov method through a corticotomy would normalize lower-limb loads and balance in affected patients<sup>59</sup>. Pedobarographic scales were used to measure these metrics both preoperatively and after limb correction. Their conclusions were that limb deformity and length discrepancy correction may restore symmetric limb loads and balance to patients.

Long bone distraction osteogenesis is typically accomplished by distracting the osteotomy site at a total rate of 0.25 mm/day split into 4 times. A basic science study performed at the Kurgan Ilizarov Center used a canine model and compared automated distraction of 1 mm/day split into 60 steps with 1 mm/day of distraction applied using the standard 4 steps. Intramuscular electromyography and histological analysis of the adjacent nerve and joint cartilage demonstrated damage to the nerve tissue in the 4-step group that was absent in the 60-step group and damaging changes in articular cartilage morphology that were more profound in the 4-step group. The authors concluded that distraction of bone will have a less negative impact on the soft tissues if done in smaller increments, with 60 steps being much safer than 4 steps daily<sup>60</sup>.

Investigators tested the hypothesis that wire deflection is an indication of bone consolidation when using circular fixation with tensioned wires. By loading a bone model containing both unstable and stable fracture patterns and varying the wire tension, the authors found that wire deflection decreased as THE JOURNAL OF BONE & JOINT SURGERY • JBJS.ORG VOLUME 100-A • NUMBER 16 • AUGUST 15, 2018 What's New in Limb Lengthening and Deformity Correction

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bone stability increased and concluded that this may be another avenue for determining the ideal timing to remove external fixators<sup>61</sup>.

#### Trauma

The Ilizarov method is a reliable technique to reconstruct challenging acute and chronic trauma cases. There are unique features when performing posttraumatic reconstruction using an internal lengthening nail<sup>62</sup>. Rozbruch elucidated this further, demonstrating several case examples of femoral and tibial fractures in a supplementary article<sup>63</sup>.

Hammouda et al. reported on 17 patients who underwent posttraumatic femoral lengthening using the PRECICE intramedullary lengthening nail<sup>64</sup>. The mean age of the patients was 30 years (range, 11 to 72 years) and they had a mean lengthening of 3.8 cm (range, 2.3 to 6.0 cm). Three patients (18%) developed complications: 2 patients had premature consolidation, 1 of whom required repeat osteotomy, and 1 patient had a pressure ulceration from a postoperative splint<sup>64</sup>. The authors reported a mean consolidation index of 32 days/cm.

The motivation for posttraumatic reconstruction of large bone defects utilizing internal devices continues to evolve. Krettek and El Naga reported on their use of an "all-internal segmental bone transport" system<sup>65</sup>. This is an implant that is not approved by the U.S. Food and Drug Administration (FDA) and that attaches to an internal lengthening device. The authors reported on 1 patient who presented with distal tibial bone loss due to infection.

Building on previous integrated techniques such as lengthening and then nailing, Quinnan and Lawrie published a surgical technique describing a central cable and pulley bone transport<sup>66</sup>. Fourteen patients with a mean tibial bone defect of 11.1 cm were included. The mean patient age was 39 years. The authors reported a mean external fixation index of 1.02 month/cm. Of note, 7 patients had conversion to an intramedullary nail after completion of the bone transport. Referred to as transport and then nailing, the external fixation index was significantly reduced to 0.36 month/cm.

In another permutation of an integrated technique, Ferchaud et al. described 7 patients who underwent bone transport with a monolateral fixator over an intramedullary rod<sup>67</sup>. They reported excellent Paley-Marr scores in 6 of 7 patients.

The Ilizarov circular fixator has continued to find use in the acute trauma setting in bicondylar tibial plateau fractures<sup>68</sup> and open distal femoral fractures<sup>69</sup>. However, it is also used in post-traumatic tibial osteomyelitis, where it remains a reliable surgical technique for an extremely debilitating entity<sup>70-77</sup>. Tetsworth et al. performed a retrospective comparative study of 42 patients treated for infected tibial nonunion with a bone loss measuring between 3 and 10 cm<sup>78</sup>. They compared acute shortening and then lengthening with classic bone transport. The external fixation indexes were similar in both groups, the complication rates were lower in the acute shortening and lengthening group<sup>78</sup>.

Barger et al. reviewed the current literature and technique on antibiotic-coated interlocking intramedullary nails for the treatment of long-bone osteomyelitis<sup>79</sup>.

The treatment of a closed tibial fracture with a circular fixator remains a viable option for surgeons<sup>80</sup>. In addition, a hexapod fixator can be used accurately with minimal complications for distal tibial nonunions<sup>81</sup>.

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