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What's new in pediatric lower limb reconstruction?

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Abstract

The last years brought many advances relevant to lower limb reconstruction. It feels like guided growth has been looked at from every angle, and still there are new emerging concepts like rotational guided growth waiting to be validated. New hexapod external devices are more accurate and easier to use, and new unilateral fixators allow for more versatile and stable fixation and lengthening. Intramedullary nail lengthening has found its place as a standard procedure for various diagnoses in children and young adults. First results of new and exciting approaches like extramedullary implantable nail lengthening and lengthening plates have been reported. Pharmaceutical treatment has changed the course of certain diseases and must be integrated and considered when making a reconstructive treatment plan. As reconstructive surgery is rapidly advancing so are the technical options for prosthetic fitting, which makes it difficult for caregivers as well as for parents to make the decision between reconstruction and amputation surgery for the most severe cases of congenital deficiencies. This review is highlighting new developments of lower limb reconstruction and is reviewing the current literature.

Keywords: Limb lengthening, deformity correction, intramedullary lengthening nails, external fixation devices, prosthetics

Guided growth

Guided growth (GG) is a continuously investigated procedure for skeletally immature patients with angular deformity and leg length discrepancy.

Rotational GG has been further investigated. It may be achieved by implantation of two obliquely oriented plates on each side of the treated growth plate. This has been shown in a cadaveric study with the use of a novel plate design.[1] Metaizeau published on a new technique with two screws and a cable in 20 patients. Results were promising with a mean derotation of 25 degrees in 22 months, despite temporary knee stiffness was an issue and further studies are needed to provide more information on concomitant effect and risks for the growth plate.[2]

With the great advantage of GG being percutaneous or minimally invasive, complications may arise. Recent studies report on failure of percutaneous drilling for limb length discrepancy (LLD) with the consequence of undercorrection and angular deformity, when leaving the central part of the physis intact.[3] Novel devices have been investigated for

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epiphysiodesis in the reduction of excessive predicted final height in tall stature. Definitive epiphysiodesis seems to be combined with a lesser risk for secondary angular deformity.[4] Treatment of LLD with either tension band plating (TBP) or transphyseal screws has been further investigated. TBP may have a greater risk of complications in temporary epiphysiodesis in comparison with the use of transphyseal screws.[5]

To treat frontal and sagittal plane malalignment, percutaneous epiphysiodesis using transphyseal screws or TBP show continuously favorable correction results in recent studies. Nevertheless, overcorrection may be an issue in the before-mentioned method and rebound may probably be slightly higher in TBP.[6,7]

Finally, a word about the so-called "sleeper plate." When rebound is expected, the metaphyseal screw in TBP may be removed to be able to percutaneously reinsert it later when needed. Two new studies have shown that this method may be complicated by bony ingrowth in the empty plate hole and thus leading to further unintended correction by tethering. In addition, re-implantation may be impossible due to further bone growth.[8,9]

External fixation devices

In recent years, we witnessed innovative approaches in the field of external fixation devices, though for a longer time they were considered to be outdated due to the rising use of intramedullary lengthening devices. The external fixators are still of importance in smaller bones and in pediatric cases with open physis and they play a crucial role for more complex deformities with additional joint problems, contractures, infections, pseudarthrosis, or soft tissue problems.

Especially for knee- or hip-bridging unilateral fixators usable for early femoral lengthening, treating fixed joint contractures or severe Perthes' cases, two modular systems are advantageous due to their versatility and multiple options in fixation: the Modular Rail System by Smith & Nephew and the DRIVE Rail by Orthopediatrics. Both provide multiple pin placement options and hybrid ring use for adding on at the lower leg or with a quarter ring at the pelvis.

The DRIVE Rail works like a cog railway with the cogs directly at the outer side of the rail and a moving drive mechanism, engaging directly with the cogs. Thus, there are less tension moments in comparison to a regular outlying distractor. It has a modular setup, adjustable to the exact length needed and it is possible to add lengthening modules on to the rail at a later stage of the treatment if necessary. In addition, there are hinging and angulation options in every dimension as well as the possibility to place multiplanar pins, thus providing for more stable constructs and better results, reducing the risk of secondary deformities.

The expiration of the hexapod fixator patent for Taylor Spatial Frame (Smith & Nephew) led to the development of several other hexapod systems (TL-Hex/ Orthofix, Maxframe/ Synthes, ORTHEX/ Orthopediatrics to name the most used) each with different modifications and advantages, thus still using the same principles of correcting the deformity with a ring fixator at a virtual hinge in a hexapod Steward platform. Regarding the hardware: the step-off or Z-plates do improve the strut placement options, helping to avoid hardware collisions or to fit in struts in very narrow spots due to highly angulated rings in high grade deformities or small bone segments. Also, the range extending dual scale struts (TL-Hex, ORTHEX) are useful improvements, minimizing the need for strut changes. Outstanding and very interesting for corrections in small children or upper extremity is the small hexapod frame by ORTHEX, which is a complete small copy of the system with all options of the regular hardware and software. Especially since there is no more equivalent as the Mini Ilizarov available, this system opens a range of indications at an increased precision and safety level.

However, the most striking changes in all these systems are the updated software and the calibration options to work with by integrating the X-ray images into the software. As shown by recent study,[10] the accuracy of measurements and correction can be improved in comparison to the conventional hexapod, although it is improved precision at an already very high level, not leading to statistically significant clinical improvements. Nonetheless, features such as correcting a given deformity by drag-and-drop, directly visualizing the expected result of a planned correction on screen (Smart TSF) is the next level and points out the way for future developments in software, which possibly may see also integration of artificial intelligence (AI)-assisted corrections. First glimpses of this sort of correction simulations can be seen in planning software systems like PeekMed, where anatomical bone landmarks will be automatically identified, measurements taken, and surgical correction suggestions based on the measurements then being visualized by the software.

Regarding the calibration of the postoperative X-rays, two systems stand out by self-calibrating tools as the X-ray marker balls of ORTHEX and the Beacon of Smart TSF. They need to be temporarily fixed at the frame after surgery for the postoperative documentation that will then allow to describe the position of the reference ring in regard to the bone in the automatically calibrated X-ray. Generally, the precision of the digitally measured mounting parameters are highly accurate and reliable in comparison to direct measurements during surgery by the surgeon.[11]

Internal nail lengthening

Motorized internal lengthening nails are increasingly used,[12] and the nails have been found to be reliable and accurate[13–17] with high patient satisfaction in pediatric lower limb lengthening.[14] However, complications still occur that warrant careful patient selection and preoperative planning in addition to close postoperative follow-up.[13–17] Complications are higher for tibial than femoral internal lengthenings in pediatric patients.[15,18] In skeletally immature patients, concerns about violating the growth plates inhibit the use of tibial and retrograde femoral nails. Antegrade femoral nails might be used in skeletally immature patients by using a trochanteric entry point; however, the age limit for avoiding avascular necrosis of the femoral head is unknown. A systematic review[19] of five studies[13,14,20–22] with 131 patients ranging from 7 to 18 years of age did not report radiographic nor clinical signs of femoral head necrosis when using the trochanteric entry in 135 femoral lengthenings. In very young patients, off-label extramedullary use of the lengthening nails has allowed for internal femoral lengthening in patients as young as 4 years of age.[23,24]

In a systematic literature search,[25] only two retrospective studies were found comparing motorized nails with external fixation in femoral lengthening in children.[21,26] The age of patients treated with motorized nails were higher than patients treated with external fixation, and more complications were observed with external fixation.[25] In a retrospective comparative study of 75 children from 10 to 18 years of age undergoing lengthening in femurs without axial deformity, fewer adverse events were observed with internal lengthening nails than external fixation.[27] In 50 children between 11 and 17 years of age, femoral lengthenings were compared between external fixation and motorized nails.[28] Patients were matched for age and indication for lengthening, and more complications were found with external fixation.[28] In a prospective study using child-specific and validated health-related quality of life measure, lengthening nails offered better health utilities and quality-adjusted life years for children compared to external fixators.[29]

In 2023, four comparative retrospective studies were published with the remaining new clinical studies being either retrospective case-series or case-reports. Despite their apparent increased costs compared with external fixators, lengthening nails were not found to be cost-prohibitive for femoral lengthening.[30] No differences were found in radiographic outcomes or complication rates between the FITBONE nail and the PRECICE nail.[31] Complications were examined in 314 nail lengthenings and the risk of sustaining a complication was higher for tibial versus femoral lengthening, for short stature compared with other etiologies, and for higher age.[32] In 420 limb lengthenings, a 10% mechanical failure rate was observed with the lengthening nail.[33] Thirteen patients were examined a minimum of 10 years after retrograde femoral nail lengthening, and it was found that all patients were free of knee pain and had full knee range of motion.[34] During tibial internal lengthening, distal migration of the proximal tibiofibular joint was greater when the proximal fibula was fixated with tether compared with screw fixation; however, knee extension was equal, and no patients reported knee pain or tightness.[35] The STRYDE nail introduced for early full weightbearing has been removed from the marked but clinical studies still emerge. In a matched pair-study, patients treated with the STRYDE nail were more likely to have osteolysis, periosteal reaction, implant breakage or pain than patients treated with the PRECICE nail.[36] Late-onset pain occurred with 10 nails out of 78 femoral STRYDE nails and the pain resolved either prior to or after nail removal.[37] Neither focal osteolysis or periosteal reactions of the bone were seen at the telescopic junction of 128 FITBONE nails.[38]

Gradual deformity correction by GG prior to or during nail lengthening was possible in skeletally immature patients; however, full deformity correction might not be achieved.[39] Acute deformity correction at the time of nail insertion provided good results; however, the bone healing during subsequent limb lengthening might be impaired.[40] Limb lengthening or bone transport with internal lengthening nails were possible after previous bone sarcoma surgery,[41–44] and magnetically driven limb lengthening could be performed in patients with pre-existing implanted programmable devices such as cardiac pacemaker, gastric pacer, or ventriculoperitoneal shunt.[45] Re-using magnetically controlled limb lengthening nails, either during the same lengthening episode or in a temporally separate treatment requiring a new corticotomy, was possible in 7 out of 12 nails.[46] However, breakage of reactivated "sleeper" nail has been reported.[47]

Despite the many advantages of motorized lengthening nails, limitations are still present. Current nail designs do not have an anatomic curvature, must be inserted through the area of an apophysis or epiphysis, and are further limited in use by diameter and length. Finally, the intermittent use of external control units does not allow for continuous distraction. These shortcomings might be overcome by future internal lengthening devices. Preliminary clinical results have shown that limb lengthening with motorized internal plates is feasible in very young children.[48] In addition, a preclinical animal study has reported on continuous lengthening by osmotic pumps that might be used for bone distraction.[49]

Achondroplasia

The most important development in recent years for patients with achondroplasia is the option of pharmaceutical treatment by means of Vosoritide, which inhibits the pathological gain of function of the mutated FGFR3 receptor aiming to enable physiological enchondral ossification. Additional annual gain of stature height of 1.57 cm has been observed in the initial randomized controlled trial with safe and persistent growth after 2 years.[50,51] However, long-term effects in combination with surgical treatment remain unclear, especially since the initial study excluded patients who underwent surgical treatment.[50]

Regarding angular deformity correction with growth modulating procedures, Ulusaloglu et al.[52] reported that tension band plates reliably correct genu varum in achondroplasia starting growth guidance at an early age (mean: 6.5 years) Makarewich et al.[53] stated that children with achondroplasia can successfully be treated with temporary hemiepiphysiodesis to correct coronal plane deformities at the distal femur and proximal tibia reducing the need for later correction osteotomies.

The general trend toward the application of intramedullary lengthening nails as an alternative to external fixators for distraction osteogenesis also accounts for patients with achondroplasia. Recently, reliable results for exclusively performed simultaneous bilateral femoral lengthening with intramedullary lengthening nails were reported,[54] whereas other studies evaluating the outcome of short stature lengthening using external and internal devices demonstrated that intramedullary lengthening nails can safely be applied for bilateral femoral and/or tibial lengthening.[55,56] Usually, intramedullary lengthening nails are applied following several consecutive fixator-controlled lengthening procedures as part of a multi-stage treatment concept for extensive lower limb lengthening.[54–56] Of course, in younger patients with bone dimensions too small for intramedullary devices, external fixators continue to be the gold standard for distraction osteogenesis.[54–56] It remains unclear if lengthening plates will contemplate the armamentarium of devices applicable for lengthening in patients with open growth plates in future.[57] In contrast to femoral lengthening with internal devices, Rovira Martí et al.[57] confirmed that prophylactic intramedullary rodding should be considered to prevent regenerate re-fracture after femoral lengthening using external fixators in patients with achondroplasia. In tibial lengthening with Ilizarov fixator, Boero et al.[58] measured proximal migration of the fibular head when proximal tibiofibular fixation was not performed as opposed to when it was performed. However, this did not lead to differences in the clinical-functional outcome of the knee, radiographic results, and quality of life. When counseling patients and families about treatment strategies for stature lengthening—regardless of the device used— one should take into account the observations made by Balci et al.[59] who found that simultaneous bilateral femoral and tibial lengthening has more physiological physeal disturbance effects than consecutive lengthening in patients with achondroplasia. Whether this effect might be mitigated by concomitant treatment with Vosoritide remains to be elucidated.

As demonstrated in the review article by Constantinides et al.,[60] the general impact of limb lengthening surgery on quality of life and physical function in patients with achondroplasia is unclear. However, some studies indicated that greater body height or upper limb length may lead to an improvement of quality of life and a gain of function. Laufer et al.[61] reported functional improvement in activities of daily living, physical appearance, and overall satisfaction after bilateral lengthening of the humerus for a minimum of 8 cm.

Orthopedic surgeons and pediatric endocrinologists should bear in mind that the condition is not sufficiently characterized when focusing on rhizomelic short stature. To date, it remains unknown whether medication with Vosoritide will positively influence other manifestations of achondroplasia such as angular deformities of the lower limb, obstructive apnea, craniocervical constriction, or lumbosacral stenosis. For the first time, a consensus statement has been published in 2022 aiming to provide an orientation on the diagnosis, multidisciplinary management of lifelong care of individuals with achondroplasia.[62]

X-linked hypophosphatemia and osteogenesis imperfecta

X-linked hypophosphatemia (XLH) is the most common inherited form of rickets, and a multisystemic disorder that should be managed by multidisciplinary teams. Gait analysis data combined with clinical and radiological data showed significantly reduced gait quality, ankle power, and plantar flexion.[63–65] Authors attributed these findings to the presence of structural changes such as subtalar ankle osteoarthritis, femoral maltorsion, and tibiofemoral angular deformities, whereas varus deformity had a greater impact on walking than valgus deformity; they conclude that the combination of radiology and gait analysis in the clinical follow-up of children with XLH may improve clinical management of these patients.

Burosumab is a relatively new human monoclonal antibody treatment for patients with XLH, and has shown to improve renal tubular phosphate reabsorption, serum phosphorus levels, linear growth, and physical function and reduced pain and the severity of rickets.[66] The effect of the antibody treatment on lower limb malalignment remains somewhat unclear. One recent study showed improvement of malalignment,[67] whereas in another study frontal axis deviation and maltorsion did not improve after 12 months of treatment with Burosumab.[68]

Osteogenesis imperfecta (OI) is a group of genetic disorders mainly resulting in low bone mass and reduced bone mineral strength, leading to increased bone fragility.

Emet et al.[69] compared intramedullary fixation alone with intramedullary fixation combined with plate and screws techniques in patients with OI. Overall complication rates were high, but significantly lower in the group combining different fixation techniques. In a retrospective cohort study of 783 patients with OI, Yang et al.[70] found that in the treatment of long bone fractures and deformities, intramedullary telescoping rods showed the lowest revision rate and longest implant survival period compared with other fixation techniques, confirming earlier results of a meta-analysis.[71]

Franzone et al.[72] demonstrated in a multicenter study that GG procedure can be effective in patients with OI. Cyclic intravenous therapy with bisphosphonates has become an established part of the treatment of moderate to severe OI.[73]

Mahmoud et al.[74] found in a systematic review that Zoledronic acid was well-tolerated in children with OI, and the treatment significantly improved bone mineral density and reduced the fracture rate. In a meta-analysis, Wehrli et al.[75] found that the quality of life in children with OI is significantly reduced compared to healthy controls and norms, emphasizing that the overall care of children with OI should not only focus on improving physical functioning rather than the emotional, school, and social functioning.

Congenital pseudarthrosis of the tibia

Congenital pseudarthrosis of the tibia (CPT) is one of the most challenging conditions in pediatric orthopedics. The new classification of CPT introduced by Dror Paley recently, provided better practical treatment guidelines[76,77] for this condition.

Congenital anterolateral bowing is a precursor of pseudarthrosis, and its treatment varies from observation and bracing to bypass allograft to the tibia and in addition recently introduced distal tibial GG.[78]

After fracture of the tibia and fibula and establishment of pseudarthrosis, cross-union between the tibia and the fibula seems to be the most effective option to achieve and maintain union.[76,77,79,80] This treatment protocol includes pre- and postoperative bisphosphonate infusion to prevent osteoclast activity and bone resorption, excision of hamartoma around pseudarthrosis of the tibia and fibula, excision of the pathologic periosteum, harvesting massive iliac bone graft and creation of the cross-union between the two bones. Intramedullary fixation of the tibia and fibula is important and rotational stability is achieved using internal or external fixation. The use of bone morphogenic protein and covering the pseudarthrosis by a normal periosteal graft may also help achieve solid union. The average rate of the union without refracture with all methods before introduction of cross-union technique was approximately 50%,[77,81] compared to near 100% in studies describing cross-union strategy.[76,77,79,80]

Parents should be informed that regular follow-up and sometimes additional intervention for exchange of intramedullary rods and correction of residual deformities might be needed till skeletal maturity. Alternative proposals to cross-union technique are vascularized fibular bone graft,[82] Masquelet technique, and in refractory cases amputation and prosthetic fitting.

Congenital deficiencies

Due to the rarity and heterogeneity of congenital femoral deficiency (CFD) and fibular hemimelia (FH) large outcome studies are missing. However, exciting articles elucidating basic-science findings and case series highlighting technical aspects of management have been published.

Differences in gene expression in patients with CFD were reported by Frydrychova and co-worker comparing tissue samples of the pseudoarthrosis area in CFD with normal bone samples;[83] how these differences correlate to, and which factors influence the pathoanatomy in CFD will need to be evaluated in future studies. Before comprehensive reconstructive surgery, anatomic variations need to be identified. Analyzing preoperative magnetic resonance imaging (MRI) in patients with CFD, Huser and co-workers showed that the femoral neurovascular bundle is closer to the anterior inferior iliac spine on the affected side when compared with the unaffected side.[84]

Lengthening in CFD and FH is complicated by joint instability[85] and re-fracture.[86] Popkov and co-worker compared titanium flexible intramedullary nailing (FIN) versus hydroxyapatite-coated FIN on the External Fixation Index (EFI) in 70 patients. They did not find a significant influence of the type of FIN on EFI. However, the refracture rate was associated with the ratio diameter nail to diameter intramedullary canal.[87]

Szymczuk and co-workers compared 32 patients with monolateral external fixation with 30 patients with internal lengthening nails. Patients treated with monolateral fixation had significantly less range of motion at the end of distraction and at consolidation, however, being similar at final follow-up.[21]

Extramedullary lengthening of the femur using a magnetic intramedullary nail in CFD and/or FH patients was reported by Dahl and co-workers. Lengthening was performed in 11 patients aged 4–8 years. Loss of fixation or unacceptable malalignment was seen in three cases.[23] Five cases of retrograde extramedullary lengthening of the femur without any encountered complications were reported by Iobst and Bafor, including three patients with CFD or FH.[24]

In another cohort of seven patients with CFD and/or FH age 3 to 10 years, lengthening was performed using a magnetic expandable plate.[48] Complications encountered included one hip subluxation, one knee flexion contracture and varus deformity in three patients.

The SUPER hip procedure was introduced by Paley to address hip instability, soft tissue contracture and deformity in CFD types 1b and 2a.[88] Sixty-eight SUPER hip procedures performed using internal fixation without BMP2, were compared with 38 procedures using internal fixation and the addition of BMP2. BMP2 significantly reduced persistent delayed ossification. Using fixed angle devices for fixation significantly reduced the incidence of recurrent varus.[89] A direct lateral approach for triple pelvic osteotomy as part of a SUPER hip procedure was shown to allow for the same amount of radiographic correction as a standard anterior approach.[90] Those surgical techniques and other techniques to correct deformity and associated pathologies in CFD were recently published in great detail with extensive illustrations.[91]

Knee subluxation and instability in CFD/FH is due to cruciate ligament aplasia or hypoplasia.[92] Symptomatic knee instability or subluxation can be treated with a SUPER knee procedure, combining various types of ligament reconstruction and soft tissue techniques.[93]

Rotationplasty is a treatment option for CFD types 3a with a functional foot. Paley reported different types of rotationplasties introducing several modifications to previous concepts.[94] Reviewing 19 rotationplasty cases revealed wound necrosis and dehiscence as the most common complication (52%). Additional complications included sciatic nerve palsy, tibial delayed union, distal femur failed epiphysiodesis, and a thigh compartment syndrome, all resolved by surgical or non-surgical means. No patient showed a late derotation of the rotationplasty at the final follow-up.[94]

Tibial hemimelia shows a big variety of clinical morphology and frequent syndrome association.[95] A recent article evaluated the results of 10 patients with 12 involved limbs treated with a staged approach including femoropedal distraction and subsequent reconstruction of the knee and foot.[96] Reconstruction resulted walking with full weight bearing with a knee–ankle–foot orthosis (KAFO). Four problems, three obstacles, but no true complications were encountered. A recent review of the different treatment approaches for reconstruction of Tibial hemimelia was published by Chong and Paley.[97]

Advances in understanding the bone and soft tissue alterations in congenital deficiencies have resulted in improved surgical techniques to reduce complications and long-term limitations of surgical reconstruction. This, together with new implants and lengthening devices, has improved the outcome of reconstructive surgery. Functional outcome studies will be necessary to prove that this directly translates into improved quality of life and function for children presenting with these pathologies.

Congenital lower limb deficiency—amputation and prosthetic function

Congenital limb deficiency remains an uncommon condition with a wide spectrum of both anatomical and clinical presentation.[98] Debate remains on the optimal treatment pathway with factors for consideration including leg length discrepancy, associated limb and foot deformity and expected functional outcome following treatment. Management can be broadly divided into limb reconstruction techniques and prosthetic support.

Fibula hemimelia can be characterized by partial or complete loss of the fibula. It can be associated with tibial, femoral, and both foot and ankle deformity and deficiency.[99–101] In cases of significant LLD and severe foot deformity, Syme amputation offers excellent short-term functional outcomes as a single surgical event compared to the use of an accommodative extension prosthesis.[102] Furthermore, a meta-analysis of amputation versus limb reconstruction in FH concluded that the cumulative evidence supported better patient satisfaction with less complications and fewer surgical procedures following amputation.[102] For balance, it should be noted that the recent article by Birch et al.[103] reported no significant differences in functional or psychological scores between groups of children who had undergone amputation versus staged limb reconstruction utilizing the SUPERankle procedure.

Attempts have been made to group patients with CFD into those suitable for limb reconstruction and those who would benefit from prosthetic support.[104] An understanding of the anatomical variants in those cases with more severe deformity has led to the development of classification systems that offer a surgical pathway to restore hip anatomy and which could, in turn, enable multiple lengthening episodes to produce a limb of equal length.[105,106] Surgery may still play a role in this management strategy and several surgical procedures have been proposed with the goal of improving limb and prosthetic function. These include the SUPERhip to restore hip stability,[89] knee fusion with Syme disarticulation alone or as part of a Van Nes Rotationplasty,[107] or simply an isolated Syme amputation to improve cosmetic design.[108] With no long-term studies, the question remains as to whether surgical intervention enhances functional outcome. Calder et al.[109] undertook an observational descriptive study in an attempt to demonstrate the effect of surgical intervention on quality of life and limb function in cases of severe CFD. They concluded that surgical intervention appeared to improve function, with scores declining into adulthood but not significantly. Their recommended strategy in severe deficiency is to create a stable hip joint, if possible. To utilize an extension prosthesis to equalize leg lengths and to consider further surgical intervention such as a Syme disarticulation with or without knee fusion to treat a painful unstable joint and improve prosthetic cosmesis and function. This could further enable a potential prosthetic knee joint to be present. Gait analysis may produce additional information on residual knee function. With restricted hip extension and the knee positioned so proximal, additional knee flexion in stance may increase stride length. This may be taken into consideration before undertaking a knee fusion.

Congenital tibial deficiency remains the rarest lower limb anomaly. Often associated with an unstable knee, relative overgrowth of the fibula, equinovarus deformity of the foot, and occasional duplication of digits, the goal of treatment is to create a limb with a stable knee and foot. In majority of cases, amputation remains the surgical procedure of choice.[110] In complete tibial absence, early knee disarticulation offers the most reliable functional outcome. With a stable knee, Syme disarticulation with a synostosis created between the distal tibial remnant and distal fibula produces a stump of good length and end-bearing surface for prosthetic fitting.

Advances in prosthetics

Prosthetic management of congenital limb deficiencies has evolved in line with prosthetic component advances and a holistic approach to patient's needs. Originally, the focus was on creating a prosthetic limb that best matched the human limb. There has been a subsequent shift toward enhancing functionality and quality of life, specifically in areas most meaningful to the prosthetic user's needs and interests. The cosmetic image and shape of a human limb replaced by a functional mechanical construct in lines of the "bionic look."[111]

Furthermore, clinical teams have moved toward an interdisciplinary sup-specialist approach whereby rare congenital deformities referred and concentrated at tertiary treatment centers enable accumulation of experience and the production of meaningful research studies. The introduction of these inter-disciplinary sub-specialist clinics can enable clear treatment algorithms to be discussed and allow families reflection and understanding in making decisions on treatment.[112]

One of the main factors affecting treatment choices has been the limited range of prosthetic pediatric components. A drive to improve the levels of energy return in prosthetic feet allowed for improved shock absorption and improved quality of gait. In particular, the options available for limited build height, such as in a Symes disarticulation, have expanded to allow the use of low-profile carbon fiber feet.

The range of pediatric prosthetic knees enables the fitting of new multiaxial prosthetic joints. These can accommodate the limited space mitigating the problem of the prosthetic knee being lower than the contralateral normal knee, as seen in cases such as knee disarticulation.[113] The new category of microprocessor controlled prosthetic knees also reduce the risk of falls and injuries, and produces a more energy-efficient gait. This has led to improved activity levels overall.[114] Prosthetic socket design and suspension methods have evolved to allow the fitting of previously unfit-able limbs. Secure suspension has improved both socket comfort and the secure attachment of the prosthesis, limiting unwanted pistoning, angular and rotational movements between the residual limb and the prosthesis.[115]

A holistic approach to the patient takes into account all aspects of their daily routine and personal interests. Prosthetic use has been traditionally associated with negative connotations causing the user to feel self-conscious which can lead to reduced activity, personal isolation and psychological problems.[116] This attitude is undergoing a major shift toward societal acceptance and personal pride. The majority of people will divide their activities between sitting and walking. Prosthetic design and surgical planning needs therefore to optimize both posture and sitting comfort alongside the enhancement of gait.

Children with prosthetic limbs are increasingly interested in sporting activities such as running and swimming. Some even choose to use their sport prosthetic limbs for daily activities.[117] There has been an emphasis on socket design, to enhance comfort and suspension, in order to manage the higher impact forces during activity. Examples of modern high activity pediatric feet include Flex-Foot Junior and Cheeta Explore by Ossur, Reykjavik, Iceland and Mini BladeXT by Blatchford, Basingstoke, United Kingdom.

Summary

Limb lengthening and congenital limb reconstruction have made enormous steps forward in the last decade. On one hand, this was possible by better understanding of pathology as well as biology and metabolic pathways. On the other hand, technical advances like lengthening nails, plates and complex external fixation devices made lengthening easier and safer for patients and surgeons alike. It will be exciting to see if another major emerging technology, AI, will have an influence on this field. AI could help in deformity planning as well as in analysis of callus patterns and or callus/bone mechanical behavior. Advances always result from pushing the barriers. To make sure that we are pushing the right barriers and that we push them into the right direction, clinical as well as basic science studies are of utmost importance for the safety and well-being of our patients.

Author contributions

All authors were involved in the writing of the article and editing.

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