THERAPEUTICAL METHODS AND DIRECTIONS IN LIMB LENGHT DISCREPANCIES IN CHILDREN AND TEENAGERS

Gheorghe BURNEI [1]*, Ileana GEORGESCU [2], Costel VLAD[2],
Ştefan GAVRILIU[2], Dan HODOROGEA[2], Adrian PIRVAN[3],
Cristian BURNEI[3], Izabela DRĂGHICI[2], Liviu DRĂGHICI[4]

Abstract. *Introduction: This article contains notions and guidelines derived from the current therapeutic approach used in the Pediatric Orthopedics Clinic of „M. S. Curie" Hospital, Bucharest. The purpose of this work is to illustrate the clinic's experience theoretically and by means of images.*

Methods: This article is based on 25 years of clinical experience (1986-2011), gained at Mangalia Regional Hospital and the Pediatric Orthopedics Clinic of „M. S. Curie" Hospital in Bucharest, having treated more than 250 such cases. In post-traumatic shortenings with associated soft tissue injuries we used double leveled corticotomies and lengthening. The lengthening rarely exceeded 10 cm, especially for congenital deformities and repeated lengthening. From 2001 onwards, all lengthening operations included the intramedullary implantation of one or two TEN rods, with the purpose of reducing the time to fixator removal and to shrink the healing index. In the last five years we frequently used minimally invasive osteotomies after the placement of TEN implants, achieving the separation of a small fragment that sites itself between the major fragments. The lengthening rate was 1 mm per day, broken down in four steps (0.25 mm every 6 hours). For difficult cases, such as congenital pseudarthroses or the presence of scar tissue around the osteotomy site, we recommended 0.75 mm of lengthening per day (0.25 mm every 8 hours). For congenital pseudarthroses we used controlled epiphysiolysis and bone transport. For inequalities ranging 3 to 5 cm we used temporary epiphysiodesis, initially with staples, and subsequently with "8", "H" and "I" plates. Limb shortening followed by locked intramedullary fixation was reserved for those patients who did not follow through with the evaluation program and who could not benefit from temporary epiphysiodesis.

Results: The amount of lengthening per segment varied between 3 and 17 cm. The longest staged lengthening measured 20 cm, in two stages, and the greatest overall lengthening was 25 cm for an entire lower limb. An appropriate stabilization, followed by the adequate choice of osteotomy site and the postoperative weight loading of the limb ensured a quick and qualitative healing process.

[1] Professor, "Carol Davila" University of Medicine and Pharmacology, Bucharest, Romania, "Maria Sklodowska Curie" Emergency Hospital for Children, Bucharest, Romania

[2] Orthopaedyst, "Maria Sklodowska Curie" Emergency Hospital for Children, Bucharest, Romania

[3] Emergency Clinical Hospital, Bucharest, Romania

[4] "Sf. Ioan" Emergency Clinical Hospital, Bucharest, Romania

*Address for correspondence: Professor Gheorghe Burnei, "Carol Davila" University of Medicine and Pharmacology, and „Maria Sklodowska Curie" Emergency Hospital for Children, Bucharest, 20, C-tin Brâncoveanu Blvd., Bucharest, Romania. E-mail address: mscburnei@yahoo.com

Gheorghe **Burnei**, Ileana **Georgescu**, Costel **Vlad**, Ştefan **Gavriliu**, Dan **Hodorogea**,
Adrian **Pirvan**, Cristian **Burnei**, Izabela **Drăghici**, Liviu **Drăghici**

Complications: *Less than half of the patients suffered complications, most of them being minor ones. Pseudarthroses have been treated by compaction of the site, followed by distraction, and/or the injection of BMP (Bone Morphogenic Protein). We saw no complications after epiphysiodesis or limb shortening.*

Conclusions: *Limb lengthening procedures up to 5 cm lead to rapid consolidation and minimal complications. Lengthenings exceeding 5 cm require a good psychological preparation and careful monitoring. In lengthenings more than 10 cm, a faster rate of consolidation requires a double corticotomy, the use of intramedullary fixation and the immobilization of adjacent joints. In Lobstein's disease, good results can be obtained by the use of an Ilizarov external fixator. Restoring limb length equality by temporary epiphysiodesis, around the age of 10-12, is the least aggressive method and is very effective. Limb shortening by segmental resection should become obsolete.*

Keywords: Limb Length Discrepancies, Prediction, Limb Lengthening, External Fixator, Epiphysiodesis.

Rezumat. ***Introducere***: *Lucrarea cuprinde noţiuni şi orientări desprinse din atitudinea curentă aplicată în Clinica de Ortopedie Pediatrică a Spitalului „M.S. Curie" din Bucureşti. Scopul acestei lucrări este de a ilustra teoretic şi imagistic experienţa clinicii.*

Materiale şi metode: *Experienţa clinică pe care se bazează această lucrare a fost acumulată în perioada 1986 – 2011, la Spitalul Teritorial Mangalia şi în cadrul Clinicii de Ortopedie Pediatrică a Spitalului „M.S. Curie", Bucureşti, pe mai mult de 250 de cazuri. S-au aplicat corticotomii şi alungiri bifocale în scurtările posttraumatice, cu leziuni severe musculo-tegumentare. De regulă, şi mai ales în cazul malformaţiilor congenitale şi a alungirilor repetate, alungirea nu a depăşit 10 cm. Începând cu anul 2001, la toate cazurile am aplicat un splint centromedular (una sau două tije TEN) şi alungirea s-a făcut pe acest splint, permiţînd suprimarea mai rapidă a fixatorului şi micşorând indicele de vindecare. În ultimii 5 ani am practicat frecvent osteotomia pe splint prin incizie minimă, realizând eclatarea unui fragment care să se plaseze marginal în focarul de distractare. Regimul de distractare a fost de 1 mm/zi, fracţionat în 4 etape (0,25 mm la 6 ore). Pentru cazurile dificile, cu cicatrici retractile în zona de distractare şi structură osoasă displazică, regimul de distractare a fost de 0,75 mm/zi (0,25 mm la 8 ore). În scurtările din cadrul pseudartrozelor congenitale am practicat alungire prin epifizioliză şi transport osos. Pentru scurtări între 3 şi 5 cm s-au practicat epifiziodeze temporare, iniţial cu scoabe şi eclise, apoi cu plăci „în 8", „în H" sau „în I". Scurtările urmate de osteosinteză cu tijă centromedulară blocată sau placă cu şuruburi au fost practicate doar la pacienţii care nu au respectat programul de evaluare şi care nu au putut beneficia de epifiziodeza tranzitorie.*

Rezultate: *Alungirile au variat între 3 şi 17 cm, realizându-se pe un segment chiar şi 20 cm, în 2 etape, şi pe un membru pelvin până la 25 cm. O stabilizare corectă, urmată de alegerea adecvată a locului corticotomiei sau a osteotomiei, şi punerea în sarcină a membrului la care se practică elongaţia asigură o vindecare mai rapidă şi mai bună.*

Complicaţii: *La mai puţin de jumătate din pacienţi s-au înregistrat complicaţii, majoritatea fiind uşoare. Pseudartrozele au fost tratate prin compactarea în focar urmată de distractare, şi/sau prin injectarea de BMP (Bone Morphogenic Protein). Nu am constatat complicaţii după epifiziodezele tranzitorii sau după scurtări.*

Concluzii: *Alungirile până la 5 cm produc consolidări rapide şi nu dau complicaţii. Alungirile de peste 5 cm necesită obligatoriu o bună pregătire psihologică şi trebuie supravegheate atent. În cazul alungirilor de peste 10 cm, pentru o consolidare mai rapidă este necesară corticotomia bifocală, utilizarea tijelor TEN şi fixarea articulaţiilor supra- şi subiacentă. În maladia Lobstein, rezultate bune se pot obţine cu fixatorul tip Ilizarov. Egalizarea segmentelor în cadrul scurtăriilor între 2 şi 5 cm, făcută în jurul vârstei de 10-*

12 ani cu plăci „în 8" este cea mai puţin agresivă metodă şi este foarte eficientă. Rezecţiile osoase pentru egalizarea membrelor trebuie să dispară din arsenalul terapeutic.

Experienţa acumulată şi exprimată în acest rezumat îndreptăţeşte autorii să generalizeze noţiunile şi să consemneze un update despre inegalităţile de member.

Cuvinte-cheie: Membre inegale, predicţie, fixator extern , epifiziodeză.

Introduction

Limb Length Discrepancy (LLD) is defined as a difference of the sum length of the 3 bony segments: thigh, calf and foot for the lower limb and arm, forearm and hand for the upper limb, respectivelly. An inadequate length of at least one of these segments leads to LLD.

In clinical practice, LLD is found in certain well known conditions like developmental displacement of the hip, genu flexum, genu varum, muscle retraction resulting in hip or knee flexion, etc.

It should be noted, to avoid inadmissible confusion, that the difference in length up to 1 cm in case of the lower limb is not pathological, being present in approximately 75% of the population. Medical intervention is indicated only if the difference is greater than 1 cm. Moreover, it is well known that the dominant thoracic member and the same sided pelvic one are better developed, giving rise to the commonly practice of trying on gloves and shoes on the dominant side.

There are cases in which inequality is given by limb length in excess of a segment, the shorter member being actually normal configured and proportionate to the trunk. In these cases comes into question a possible gigantism, one congenital hypertrophy or an accelerated increase in length during the postnatal period. In the latter case, the mechanism is excessive stimulation of cartilage growth, as it may occur in shaft fractures treated surgically or after an infectious process, with morbid development, located in the metaphyseal area.

A severe LLD leads to static and dynamic consequences. These discrepancies cause imbalances of the pelvis in the frontal plane, which is balanced by a compensatory scoliosis. Scoliosis may become permanent, with structural changes, if the discrepancy persists in the accelerated growth period of the child.

If shortenings range between 1 and 3 cm the length discrepancy is compensated either by positioning the shorter member with the foot in equinus position or by slight flexion of the knee of the member which is longer. If during the support phase of the shorter member there is plantar positioning, after a longer period there will appear a dynamic effect of the spine. During the shorter limb support phase, the lumbar spine becomes more convex on the side of the shorter limb and during the support phase on the longer limb it gets a normal position.

In cases where the limb is shorter by 3 to 6 cm, the patient is able to compensate the difference by tilting the pelvis. If the length discrepancy exceeds 6 cm, the patient uses hip and knee flexion of the healthy limb. If no compensation is possible, the patient needs to perform a quick flexion move of the trunk towards the shorter limb during the unilateral support phase of it.

Gheorghe **Burnei**, Ileana **Georgescu**, Costel **Vlad**, Ştefan **Gavriliu**, Dan **Hodorogea**,
Adrian **Pirvan**, Cristian **Burnei**, Izabela **Drăghici**, Liviu **Drăghici**

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History

Equalization of unequal limbs may be done by lengthening, shortening or epiphysiodesis, the most commonly used method being lengthening with an external fixator. The appearance of external fixators was a revival which gave modern orthopedics the possibility of performing therapeutic acts in malformations and acquired diseases, unsolvable by classical procedures or with a very small percentage of healing.

If until the 18th century limb length discrepancies were regarded as strange diseases, being virtually ignored in a therapeutic standpoint, starting with the 18th century, the minor differences in limb length began to be treated by segmental bone resection. The lack of a proper bone fixation made these resections to be soon abandoned. In the early 19th century, the method was resumed after the appearance of firm fixation means.

Inequalities of 3-4 cm were treated only by prosthesis.

Equalization of limb length has been made possible by the early 20th century. Extemporaneous lengthenings of 2-3 cm were performed in the first three decades, leading later to extemporaneous lengthenings of maximum 5 cm.

Limb lengthening, slow and progressive, has been possible after the developing and improving of the external fixators. The first external fixators were used in trauma, for open fractures: the fixing not being a stabile one. Their later improvement allowed the transition to lengthening, leading to unimaginable ones of up to 25-30 cm in a single limb.

The study of external fixators noted along history remarkable researchers and surgeons: Lambotte, Ilizarov, Peretti, de Bastiani, Canadell, etc [1], [2]. In Romania, a special contribution in this field has been made by two brilliant surgeons: Ernest Juvara and Corneliu Zaharia [3], [4]. Professor Juvara designed an external fixator, improving Lambotte's instrumentation and using another principle: "My retainer is much simpler, smaller, stronger and easier to apply and take off than Lambotte's fixator", Juvara being quoted this way in the book "Ernest Juvara - Man and Work" by Ion Făgărăşanu. In the '70s, Professor Zaharia has developed and introduced into medical practice two devices, certified by patents in 1973, which allowed, through his prodigious activity, in Romania to be successfully practiced the lengthening the limbs. One device was applied for extemporaneous lengthening and one for progressive lengthening.

Monoplane type external fixators were first used to treat these conditions [5], and full success came at the time circular external fixator Ilizarov type appeared [6], [7], and, later, Taylor, who by joints, allows simultaneous lengthening and correction of associated malformations. Mounting is simple for the Taylor Spatial Frame, consisting of two rings connected by columns, each column with joints at the base of its insertion. The columns placed in well established points, controlled by a computer program, each point on the bone segment moving on an established trajectory, allow simultaneous lengthening, associated deformities correction and stability [8].

Thanks to these new devices, limb length discrepancies and associated deformities, limb hemimelia, congenital or acquired pseudarthroses, even the suppurated ones, may be effectively treated, the healing rate being significantly higher compared to the traditional procedures or to the use of external monoplane fixator.

Currently, all intractable diseases, accompanied by limb length discrepancies, have as the primary therapeutic solution the circular external fixator [9], [10]. According to experts, its role will be taken in the future by methods of auxological osteo-chondro-reconstruction.

In Romania, the first lengthening with a monoplane external fixator (bilateral) to correct LLDs were made to adults by I. Olaru and collaborators in 1971, obtaining calf lengthening up to 5-6 cm. The first lengthening on a circular external fixator, in our country, was made in an adult hospital, University Hospital Bucharest, by Prof. Denischi and Prof. Niculescu. In a children's hospital I have performed the first limb lengthening procedure for lengthening on a circular external fixator Ilizarov type, in Mangalia Territorial Hospital, in 1986, as a specialist in pediatric surgery and orthopedics.

In 1993, following a research work conducted in the Pediatric Surgery and Orthopedics Clinics of the "M. S. Curie" Hospital, Bucharest, clinics bearing today the name of the late Prof. Dr. Alexandru Pesamosca, I and collaborators, together with Eng. Al. Moldovanu and Eng. Gh. Stefan, from the Mechatronics Institute of Bucharest, designed and performed multiple interventions with an external fixator, which was innovative by the application of embedded columns with joints, allowing deviations up to 30°.

I supported the doctoral thesis titled "The Use of External Fixators in Pediatric Orthopedics" in 2000, and I published by "Carol Davila" University Publishing the treaty entitled "Osteosynthesis and Limb Lengthening with External Fixators in Pediatric Orthopedics", in 2002 [11].

In the past 10 years there have been made many progresses in the limb lengthening field, applied in many centers all over the world and in Romania: upgraded centering methods initiated by Wasserstein, lengthening on two TEN rods, the use of computer navigation during partial resection of premature closed growth plate, the introduction of medical archiving image, coupled with the preoperative and developmental planning, the Albizzia, FITBONE and ISKD (intramedullary Skeletal Kinetic Distractor) intramedullary rods and hybrid techniques, which combine internal and external fixation, reducing external fixation time [12].

Diagnosis

Diagnosis of LLD is determined by clinical, radiological and scanographic measurements. Radiological or scanographic measurements are required in order to establish a diagnosis of discrepancies up to 2 cm in infants and children up to three years, the clinical measurements, especially at this age and generally in children being

not very accurate. For a measurement to be accurate, these are performed under general anesthesia, if the diagnosis at this age is required: length discrepancies secondary to diseases occurring in this age, forensic cases, or upon the request of parents. In assessing radiological measurements we have corrected the augmentation index of the device. Electronic measurements require scanographic setting benchmarks to avoid incorrect determination of the acquired dimensions. The most accurate method of assessing LLD remains the CT scanographic electronic evaluation, because it respects the scale 1/1 of the image playback. For large differences in length, the diagnosis is established easily, remaining to be determined by electronic measurements the accurate values.

Prenatal diagnosis

Limb length differences can be detected as early as the prenatal period through a routine ultrasound scan, starting with the second trimester of pregnancy *(Figure no. 1)*.
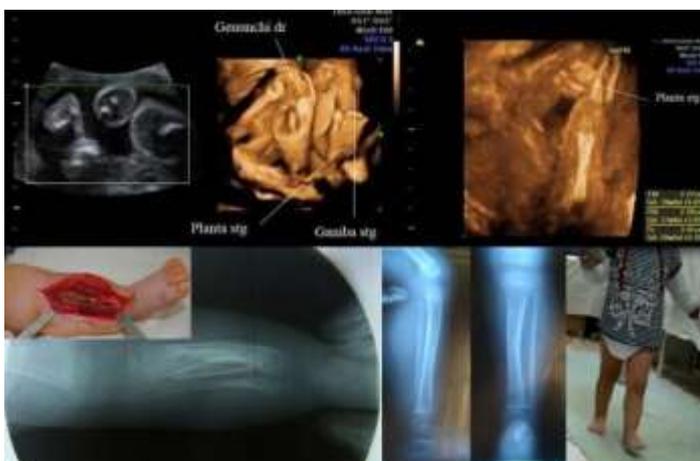

Figure no. 1:
24 weeks pregnancy: hypoplastic left tibia and fibula (of 4 weeks behind biometrics in normogram [25 mm] compared to healthy calf [35 mm]). When performing the pediatric orthopedic surgeon-gynecologist interdisciplinary consultation, parents have opted to maintain pregnancy if the disease is curable and there are no risks to induce physical or psychological damage. 4 days postpartum the patient was diagnosed with congenital curvature type Crawford I. Evaluation of the tibia at 30 and 45 days showed an increased curvature, gracility in the maximum of curvature area and fracture risk. To avoid a congenital pseudarthrosis at walking age, surgical intervention has been practiced by allografting from bone bank. The curvature area disappeared and the bone regained normal shape and structure. The child began to walk at the age of 11 months.

LLD, defined by size under the first percentile according to gestational age is associated with several chondrodistrophic syndromes. Among these are: camptomelic dysplasia, chondroplasia punctata, severe hypophosphatemia, osteogenesis imperfecta, thanatophoric dysplasia, achondroplasia of all types.

Up to 2005 there were no predictions of LLD performed on antenatal findings. Subsequently, studies were conducted and published works popularized the multiplier method described by Dror Paley, which can make predictions for cases diagnosed since the 12th week of pregnancy.

Etiopathogenic Diagnosis

Changes in limb length may be due to congenital or acquired causes.

Congenital disorders are the result of genetic diseases, inherited or appeared by "de novo" mutations, under the action of teratogenic factors. Usually, in case of malformations due to genetic causes, the defect is located only to a certain segment [13], [14]. The most common and important congenital diseases presenting with LLD are:

1) *Femoral hypoplasia*. Cases presenting only with femoral hypoplasia are very rare. The femur is reduced in size compared to the opposite one, but is normally configured and properly proportioned. A constant feature is the presence of a femoral head hypoplasia, which is smaller than the acetabulum, tending to be subluxated. The acetabulum is normal configured, but smaller than the healthy side. Affected limb segments, including the pelvis, are smaller, sometimes excluding the calf bones.

2) *Agenesis and hypoplasia of the fibula*. Congenital absence of the fibula is more common than the tibia, in a ratio of 4:1, and is more common in girls. Fibular agenesis, partial or total, and its hypoplasia are always accompanied by hypoplasia of the tibia. In total fibular agenesis, it is replaced by a relatively well-defined fibrous cord, which may present the insertion of some muscle groups. Deformity occurs alone only rarely, usually being accompanied by agenesis of the tarsal and metatarsal bones of the fibular foot ray, being outlined as a mesoacromelic hemimelia. The hypoplastic tibia is curved ventrally or laterally, further reducing the length of the calf.

3) *Agenesis and hypoplasia of the tibia*. Total absence as a congenital deformity of the tibia is a rare issue, affecting boys more frequently and is often associated with other deformities. Anatomical studies and clinical experience have enabled us to observe changes in muscle, more frequent absence of the calf muscles, and, rarely, aberrant insertions on the femur or on the fibrous cord that can replace all or part of the tibia. The fibula is usually initially increased in size to compensate for the missing tibia. During development, dynamic abstention and partial weight loading make the fibula become more unfit for the substitution of the tibia, unless the patient has exceeded 6 years. In case of partial agenesis of the tibia, usually missing are the 2/3 or 1/3 distal parts. The remaining segment presents in most cases a marked hypoplasia and ends with some sharp, prominent spur that can be palpated.

4) *Congenital pseuarthrosis of the tibia* occurs most often unilateral, mostly in boys. It is often associated with other congenital and/or genetic disorders, the most important being von Recklinghausen's neurofibromatosis. This malformation is present at birth, and in some cases it becomes apparent in the 2nd or 3rd year of life. The lesion is localized in most cases in the middle or

distal third of the tibia. The most obvious clinical sign is axial deviation of the calf, which appears as a ventro-medial concave curvature. The site of nonunion is determined by endosteal and periosteal bone formation changes, induced by a vascular disorder, and may be formed during embryonic or later period, under the influence of postnatal mechanical factors [15].

5) *Proximal Femoral Focal Deficiency*. This anomaly may affect one or both lower limbs. It may be evident at birth or may become visible in evolution. The deformity can involve all anatomical structures: skeleton, muscles, vessels and nerves, as in some forms being very severe. In all cases muscle contraction and tendons retraction of the thigh appears. In the missing segment of the femur there is a cord of connective tissue, muscular fascia being inserted on it. Shortening is considerable and the thigh shows a large increase in volum*e (**Figure no. 2**).

Figure no. 2:
Focal dysplasia of the proximal femur.
The patient underwent femoral-iliac sinostosis to stabilize the hip
and lengthening of the femur on a monoplane external fixator.

6) *Transverse hemimelia of the calf (calf peromelia).* It is a congenital amputation of the calf and may be associated with that of the forearm *(**Figure no. 3**)*. This deformity occurs most frequently only in one member, usually the distal third of the calf. In literature we may find described bilateral and symmetrical transverse hemimelia. Our data highlight the statistical association of this malformation with amniotic constriction bands of the fingers of hands and feet.

Figure no. 3:
*Transverse hemimelia of the forearm.
A severe malformation of the humeral-
radial-ulnar joint, The radius and ulna
present a proximal synostosis to the distal
extremity of the humerus.*

7) Metaphyseal-epiphyseal dischondroplasia. These are genetically induced diseases accompanied by biochemical changes that affect the local structures, shortening and axial deviations are due to lower mechanical strength. Manifestations may be present at birth or may appear later in childhood or adolescence. In these conditions, shortening are asymmetric, especially in the thighs, axial deviations occur frequently and are sometimes accompanied by dislocation of the hip. These dislocations are similar to pathological dislocation occurred after osteoarthritis in infant and toddler. Growth defects may lead to stature-weight hypertrophy or dwarfism. These events are characteristic to hemimelic epiphyseal dysplasia and Ollier's disease.

8) Phocomelia is characterized by the absence or shortening of proximal limb segment, the distal presenting with variable dimensions. In clinical practice we encounter the following forms: phocomelia with femoral agenesis, phocomelia with agenesis of the tibia and the fibula, phocomelia with agenesis of the femur, tibia and fibula, the foot being directly articulated to the hip.

Traumatic disorders

Pelvic limb fractures in children, particularly those involving the growth cartilage may cause LLD, sometimes accompanied by axial deviations. Metaphyseal-epiphyseal fractures type Salter-Harris III, IV *(Figure no. 4)* and V *(Figure no. 5)* lead more often to LLD.

Figure no. 4:

A serious complication of a Salter-Harris IV fracture, left femur distal end, resulting in a thigh shortening of 5 cm, in a valgus deviation of the femur associated with distortion and a 1 cm shortening of the tibia. Equalization of the limbs was achieved by lengthening the thigh and calf on a complex assembly of external fixation, allowing simultaneous correction of associated malformations. There has been performed functional and anatomical axis correction of the limb.

Figure no. 5:

A Salter-Harris V fracture of distal tibia with considerable destruction of the growth plate treated by immobilisation in plaster device. After 4 years of development, radiographic and MRI, CT and 3D-CT show the disappearance of growth plate in the distal extremity of the tibia and ankle and foot deformity in varus equinus and the appearance of calf and hind foot LLD. Fibula, with normal growth in relation to the shortened tibia, got bowed due to foot deviation in varus, pulled by the ligaments that keep the external stability of the ankle. Transient hemiepiphysiodesis of the distal fibula corrected slowly the bowing and ankle position. Lengthening will ensure final lower limbs equalization.

That is why these types of fractures require anatomic reduction, which may be performed in open surgery, too [16]. Our clinical experience, acquired during the past 20 years, emphasized the need for open reduction of metaphyseal-epiphyseal fractures after Ogden's classification: I C; II B, D; III A, B, C; IV A, B; V; VI; VII *(Figure no. 6)*.
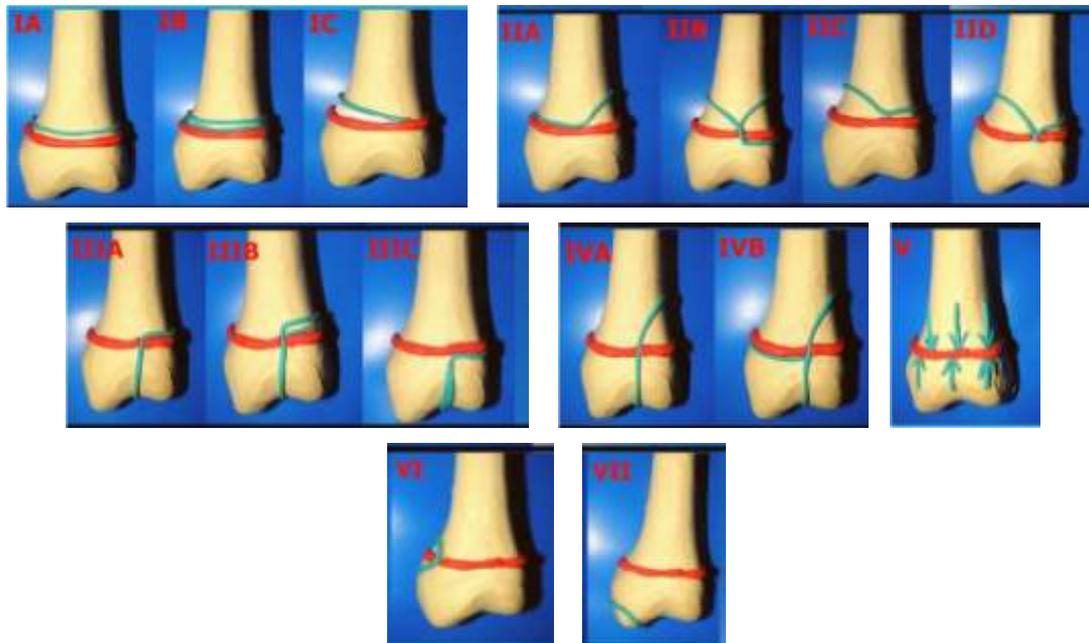
Figure no. 6:
Ogden classification of fractures involving the growth plate.

Bone infections

Infectious processes that may lead to LLD occur most frequently in the ranges 6-8 years and 10-12 years. The most common infection of bone that may induce limb shortening is osteomyelitis, rarely osteitis being involved. In osteomyelitis, the gateway is represented by a primitive source, such as a trivial infectious process (rhinitis, sinusitis, otitis, etc.) or a neglected and infected old wound. Bacteriaemia occurs after 2-3 weeks with the outbreak of germs from the primitive focus, which causes septic embolization to the metaphyseal vessels, where it forms a primitive abscess, initially diffuse, infiltrative, then located. Abscess extends first in the medullary canal and then broadcasts in the periosteum, forming a subperiosteal abscess. It is important to emphasize the role of the growth plate as a barrier in the path of infection extension.

Osteomyelitis located in the proximal humerus and femur transform into osteoarthritis after 3 days of evolution, since the appearance of the metaphyseal abscess. In these cases the growth plate is located intra-articular. In other joints, growth plate is partly loose, and septic arthritis occurs only by piercing it, which occurs after 12 days of evolution of the metaphyseal abscess.

Entire growth plate damage produces shortening and axial deviation results in partial damage (most common in the knee); it may lead to premature epiphyseal fusion.

Infections that do not affect growth cartilage can stimulate growth as a result of hyperemia during the inflammatory process. In case of minor damage to the growth plate, LLD may become evident much later, during adolescence *(Figure no. 7)*.

Figure no. 7:
Serious complication after acute osteomyelitis of the femur and tibia in a child who was in coma for two weeks. After the treatment of suppurated nonunion, thigh and calf limb lengthening has been made to correct a 22-cm shortening.

Dismetabolic anomalies

Rickets is now much more rarely, in cases where pregnant women and child less than 1 year old neglect treatment with vitamin D. In case of light and medium forms of rickets, associated to deficient and absent rickets prevention, there is only genu varum present. The deformity, diagnosed in infancy, disappears after 1-2 years of reasonable exposure to the sun. Severe deformities occur in forms of vitamin-resistant rickets, caused by inherited abnormalities of calcium-phosphorus metabolism. The vitamin-resistant rickets is accompanied by skeletal deformations of the chest, spine, pelvis and, mainly, thoracic and pelvic limb. Femurs are curved, metaphyses are increased in volume, genu varum is evident and coxa vara occurs less frequent. Intensive sun baths, made up to the age of 7 years, may lead to healing, but persistent rickets after this age require surgery. LLD appears less frequently and are always associated with the bowing that emphasizes the shortened segment.

Bone tumors, malignant and benigne, next to the growth plate may influence limb growth.

Benign bone tumors influence growth through changes in hypo- or hipervascularity. An example is the aneurysmal bone cyst or solitary bone cyst:

their structure and metaphyseal location determine segmental shortening, perhaps by hipovascularity. Sometimes we find, after the removal of bone cysts, increased growth and the emergence of inequality in the opposite direction. In our medical practice we encountered 3 cases of localized giant osteoid osteoma located metaphyseal-diaphyseal, which induced hypertrophy of the segment, the LLD reaching 6 cm *(Figure no. 8)*.
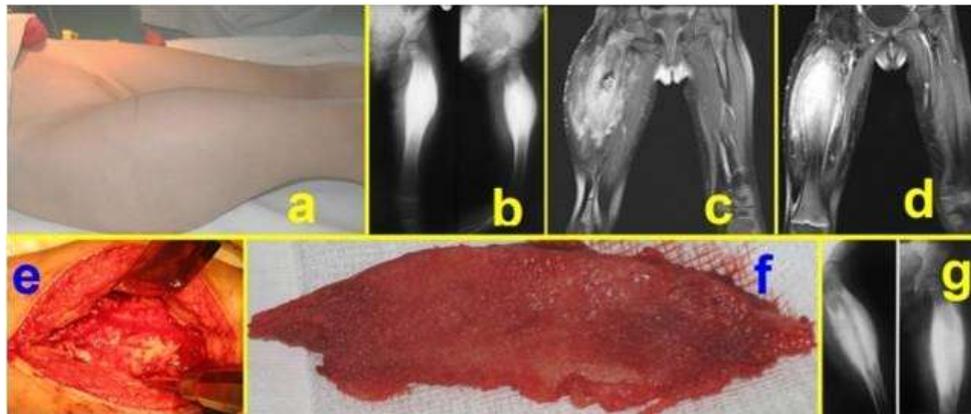

Figure no. 8:
Giant osteoid osteoma of the femoral shaft: (a) Clinical massive volume of the right thigh. (b) X-rays of the femur shows a fusiform mass of bone sclerosis. (c) The location of the nidus by MRI in the proximal femur. (d) MRI of the right femur: pandiaphyseal massive sclerosis. (e) Intraoperative macroscopic appearance of the tumoral formation. (f) Longitudinal segmental "en bloc" resection of the tumoral formation. (g) Postoperative X-ray of the femur shows central longitudinal segmental resection with restoration of bone medullar channel.

Malignant bone tumors treatment should be radical, if the tumor exceeded the growth plate. Oncological en-bloc resection, made after preoperative chemotherapy, aims the removal of epiphysis, followed by osteoarthroplastic endoprosthesis reconstruction [17] *(Figure no. 9)*. Expandable endo-prostheses allow lengthening by repeated extensions. If inextensibile prostheses are used, the best and most effective method for correcting LLD is temporary epiphysiodesis with "8", "H" or "I" plates.

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Figure no. 9:
Shortening of the femur after osteosarcoma in the 1/3 distal part, treated by "en block" resection and osteoarthroplastic reconstruction by a modular endoprosthesis in a 9 years patient. A periprosthetic fracture occurred after 1 year which has been reduced and stabilized by a Dall Miles plate. The 2.5 cm LLD will be treated by transient epiphysiodesis with a "8" plate.

Neurologic lesions

Various neurological damages may delay the increase in length of limbs and cause structural changes.

In children with poliomyelitis, growth disorders occur in 99% of the cases. Firstly the growth in length of the lower limb is affected, and next structural changes and the altering in volume of muscle occur. LLD, especially in the lower limbs, is increasing throughout childhood.

In the lower limb paralysis, the most affected segment is the distal femoral metaphysis. Shortening is due to a bone ischemic process, given the poor vascular motility related to injuries of the neurons in the spinal lateral horn *(Figure no. 10)*. The degree of shortening is proportional to the degree of ischemia.

Figure no. 10:
Sequel after poliomyelitis: lower limb shortening by affecting the thigh, calf and foot, axial deviation and genu valgum recurvatum.

Currently, the incidence of polio has decreased significantly. All cases reported in the last 5 years in Romania appeared due to incorrectly performed vaccinations or in communities with children left unvaccinated.

Brachial plexus palsy, due to obstetrical or posttraumatic causes, occurring in young children causes structural bone changes, sometimes the LLD between the two sides reaching 5-8 cm by the age of 16 years. The L'Episcopo-Rigault operation carried out between 4 and 6 years of age significantly improves the scapular-humeral joint mobility and reduces the extent of limb shortening.

Vascular malformations

In arterial-venous vascular and/or lymph malformations, bone length and thickness are increased. Hypertrophy affects both skeleton and soft tissues. Significant in case of these malformations is that the epiphyseal volume is increased in proportion to the metaphysis and shaft. The LLD is variable and increases as the children get older.

In *Klippel-Trenaunay* syndrome, vascular malformations consist in the presence of abnormal veins and shunt arranged especially in the distal extremity of the thigh and popliteal space.

Proteus Syndrome, a condition known as the "Elephant Man disease" affects the body by abnormal, asymmetric growth, and various abnormalities involving the skin, face, eyes, ears, lungs, muscles and nerves. So far there is no evidence that Proteus syndrome may be transmitted hereditarily [18]. Proteus syndrome was identified in 1986 by Michael Cohen of the University in Halifax, Canada. Research studies are currently performed in Massachusetts General Hospital and American Research Center for Human Genome *(Figure no. 11)*.

Figure no. 11:
Huge second and third fingers of the left hand in the Proteus syndrome. Radiological image shows bone malformations clinodactily of the third finger. Angio-MRI images confirmed multiple arteriovenous malformations. The surgical intervention consisted in suppressing aberrant vasculature, clinodactily correction by osteotomy and plate with screws, epiphysiodesis of the proximal and distal phalanges and the middle phalanx hemiepiphysiodesis.

Gheorghe **Burnei**, Ileana **Georgescu**, Costel **Vlad**, Ștefan **Gavriliu**, Dan **Hodorogea**,
Adrian **Pirvan**, Cristian **Burnei**, Izabela **Drăghici**, Liviu **Drăghici**

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Profound unaesthetic impact manifestations occur in children with enlarged monstrous fingers. Angio-CT investigation reveals vascular abnormalities, both arterial and venous. Operative interventions are directed to correct excessive length of the phalanges, ligatures and / or excision of vascular malformations, plus, depending on age, epiphysiodesis.

Hemihypertrophia and *hemihypotrophia* are conditions that occur within the first weeks of life, being characterized by discrepancies between the two halves of the body. These problems are more evident at the lower limbs. LLD ranges 2-5 cm. To differentiate between hemihypertrophia and hemihypotrophia the limb dimensions must be assessed for the age and sex of a normal child.

Slipped capital femoral epiphysis

In this condition, shortening is a terrible and relatively frequent sequel, present in all cases in which the treatment consists of epiphysiodesis. After Dunn and Southwick operations, shortening is variable in relation to the patients' age. If shortening is small, 2-3 cm, an opposite transient distal femoral epiphysiodesis may be applied [19].

Femoral head necrosis after surgical treatment is a feared complication. LLD is always present and can be corrected either by expandable endoprostheses or by transient epiphysiodesis.

Limb lengthening in this disease is a relative indication and upon request. Indication of choice is the LLD of about 5 cm, with normal or close to normal hip mobility.

Blount disease

LLD in this disease occurs in unilateral forms (approximately 50% of cases). Curved and shorter tibia become evident after the first three years of life, the medial half of the proximal growth plate shows irregularity and proximal epiphysis alters its shape.

Osteogenesis imperfecta

Osteogenesis imperfecta is a genetic disorder characterized by qualitative and quantitative deficiency of collagen type I. Among the 12 types described by Shapiro in literature, type I, II, III and IV are more common and transmitted autosomal dominant, except type III, which may be transmited autosomal recessive, too. Genes responsible for this disease's transmission are COL1A1 and COL1A2.

Repeated fractures occurred when the child begins to walk, creating inequalities predispose to LLD. Limb shortening appears more frequently after the orthopedic treatment of fractures. The bowing degree and LLD increase with the accumulation of fractures on the same segment. In the worst forms, represented by osteogenesis imperfecta type Vrolik and types I and type III, after Shapiro,

fractures may occur even in intrauterine life, the newborn period or infancy, and often complicated by nonunion. Normal height size occurs in about 1 in 20 patients with osteogenesis imperfecta. Lengthening limbs for LLD in these patients is followed by fixation with intramedullary rods locked static. Application of bone stimulators in the lengthening focus hastens callus formation, the healing index get lower.

Limb lengthening in osteogenesis imperfecta is usually performed with the Ilizarov type external fixator, to maintain stable fragments and to ensure good consolidation of the lengthening foci. The first limb lengthening made in Romania for Lobstein disease has been performed at the Hospital "M. S. Curie", in Bucharest, in November 2002, in a patient aged 16 years *(Figure no. 12)*. At that time, in the world there had been communicated only two similar procedures. The most difficult case to cure applied lengthening and concomitant nonunion treatment was represented by a patient with osteogenesis imperfecta type Vrolik, who presented more than 100 fractures and a thickness of long bones of the lower limb shafts of about 0.5-0.7 cm. This exceptional case is unique in literature. Currently, the patient walks with support using a walking frame *(Figure no. 13)*.
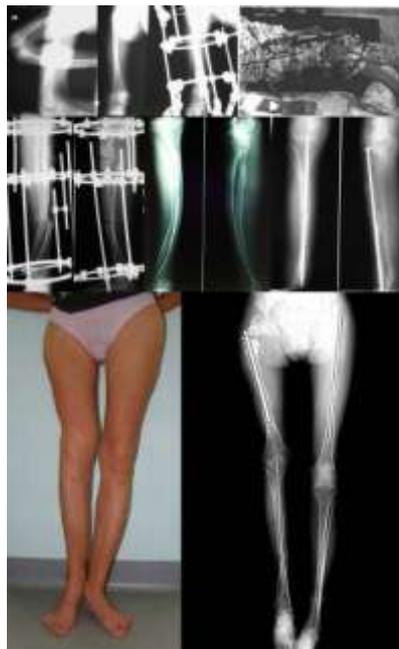

Figure no. 12:

First time limb lengthening in a patient with osteogenesis imperfecta was made in Romania in November 2002: First a lengthening of the femur was performed simultaneously with the deflection of the knee, next tibial lengthening in double level (7 cm) being performed. To increase strength and prevent fracture, there were applied a Sheffield rod to the right tibia shaft and Kuentscher rods to the femurs.

Figure no. 13:
An exceptional case of a patient with Lobstein's disease, which at the time of the consultation had already suffered over 100 fractures. The X-rays of long bone extremities showed a cystic form and a nonunion of the left femur. A 7-cm left femoral lengthening has been performed. After external fixator removal, nonunion has been cured and the femoral and tibia shafts were enforced with Sheffield telescopic rod. Currently this child walks with help of a walking frame and enjoys school, society and environment.

LLD prediction at growth ending

Prediction of LLD gained interest with the discovery of the possibility to control the growth by different surgical techniques: stopping the growth of the longer limb, lengthening the shorter limb, combining the two processes. In 1933, Phemister describes epiphysiodesis as a therapeutic solution which consists in stopping the growth of one or two epiphyses so that the global growth slowing of the longer limb will lead to the lower limbs equalisation at the end of growth. Progressive limb lengthening was firstly described by Codivilla in 1905. Calculation of LLD at the end of growth is actually the calculation of the timing for longer limb epiphysiodesis or the calculation of the lengthening amount of shorter limb.

In dwarfs, to determine the discrepancy in relation to normal size, a simple formula is applied, given for guidance: $H = 5 \times V + 80$ (in which H is the body height, V is the patient age. It is considered that dwarf patients at the end of growth does not exceed a height of 140 cm. Assessment can be done in relation to annual growth.

Prediction of LLD is based on the principles of Hootnik and Amstutz; according this principle the ratio between the longer limb length and the shorter limb length remains constant during growth and is applicable to type I of LLD only, according to Shapiro's classification. The timing of epiphysiodesis could be established for those cases where the difference between the two limbs lengths remains constant during growth. Calculation accuracy depends on the quality of bone length measure and bone age assessment [20].

LLD may be assessed clinically and radiologically. The difference between the clinical and radiological measures of LLD may exceed 1 cm. The classical method for assessing radiological lower limb length, teleroentgenography, presents the disadvantage of radiological image augmentation with up to 3 cm from the actual length of the measured segment, because of the parallax effect.

Orthoroentgenography and scanography eliminated this disadvantage but they don't provide an overview of the examined skeletal segments. CT-scanography allows more accurate measurements of limbs lengths, but it presents the disadvantage that the image acquisition cannot be done with the patient in orthostatic position and the mechanical axes analysis. EOS technique, recently introduced in the medical practice, eliminates all the disadvantages of other methods, while also decreasing the X-ray dose administered to the patient.

Starting from the principle of Amstutz, the length of the shorter limb may be calculated by simple mathematical formulas, at the end of growth, if known the lengths of the two limbs at a given time and the length of the normal limb at the end of growth: if $L_o/S_o=L_f/s_f$, it results that that $S_f=(S_o \times L_f)/L_o$, where L_o is the present normal limb length, S_o is the present shorter limb length, L_f is the normal limb length at the end of growth, S_f is the shorter limb length at the end of growth. From this equation, Lo and So are measurable values by imaging methods, while Lf can be calculated from growth charts described by Anderson and Green.

The next step is to calculate LLD at the end of growth:

$$I = L_f - S_f = L_f - (S_o \times L_f)/L_o = L_f \times (1 - S_o/L_o).$$

Anderson and Green's residual growth charts will allow then to identify the timing of epiphysiodesis, and the epiphysis to be fused.

One problem of growth curves is represented by the reference age, which has to be the bone age. According to Cundy, 10% of children have a discrepancy of more than two years between bone age and chronological age and 50% of children have a 1-2 years discrepancy between the chronological age and bone age. The calculation of bone age is performed using the Greulich and Pyle catalog, which takes into account the ossification degree of the epiphyses in the non-dominant hand. Another method for calculating the bone age is the Sauvegrin method, which calculates a score based on the degree of epiphyseal ossification at the elbow. To calculate the timing of epiphysiodesis, Menelaus proposes an empirical but effective method. According Menelaus the residual growth is 9 mm / year for the distal femur, and 6 mm/year for proximal tibia epiphysis. Based on these principles the error was less than 6 mm at the end of growth in Menelaus series.

Another method is proposed by Moseley who transformed the curves of Anderson and Green into straight lines by mathematical calculation.

In 2000, based on the same studies of Anderson and Green, Paley proposed a simplified method of calculation. He calculated an age-specific multiplier, which applies to the current length of the member: $L_f = L_o \times M_o$, where L_f is the final length, L_o is the present length, M_o is the multiplier corresponding to the patient's bone age at the present time.

Treatment principles

Operative interventions for limb lengthening may be done to dwarfs, in individuals with normal size, but LLD, or in exceptional cases, to patients who want a larger size. Lengthening "on demand" for patients with size greater than 140 cm has to be made with caution, after careful psychological evaluation, after the patient is informed of the risks and complications he is exposed to *(Figure no. 14)*.

Figure no. 14:
A 10 cm Lengthening by request, made simultaneously in two levels, on both thighs.

In dwarfs, lower limb lengthening is a necessity or the indications may be optional. Patients with hypotrophic stature or dwarfs present the necessity indication for limb lengthening when associating axial deviations *(Figure no. 15)*.
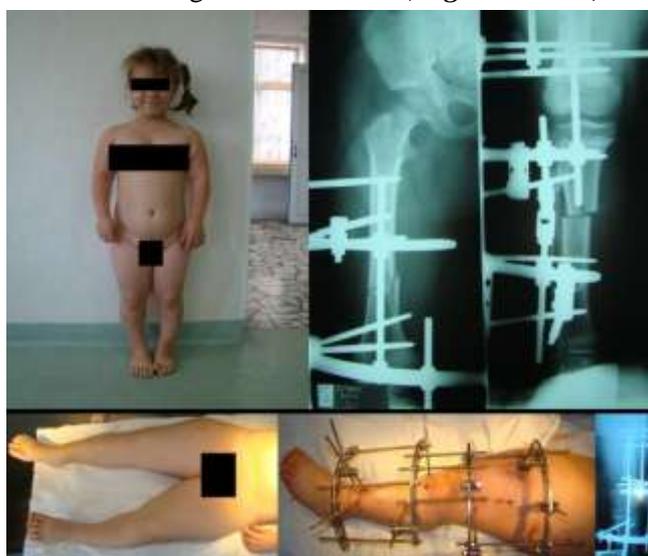

Figure no. 15: *Achondroplasia: concomitant lengthening of the thigh (5 cm) and calf (3 cm), with correction of genu varum knee deviation, using an Ilizarov external fixator with hemieliptic arches: the mounting is complex and protects the knee.*

In other forms of dwarfism, in the absence of axial deviation, surgical intervention may be made "on demand", aiming to ensure proportionality between limb and trunk segments. Decision on surgical intervention depends on the patient, parents and surgeon. Express indication in dwarfs for limb lengthening is

the disharmonic dwarfism and is usually contraindicated in the harmonic one. Lengthening limbs in the latter situation can be practiced if the patient wishes to exceed a height of about 140 cm and reach 150 cm. Patients' and surgeons' option to perform lengthening for harmonic dwarfism is a questionable and objectionable decision, given the need for performing a four staged surgery, over a long period, with an extremely high risk of complications and decrease of the healing index after the first or second intervention. The best results are obtained achondroplasic dwarfs, as lower limb bones are short, but well set up and easy to lengthen.

Asymmetric discrepancies are operative indications of need or can be made "on demand". Intervention must take into account the patient's age and the discrepancy between limbs.

Fundamental principles to be known to the orthopedic surgeon and should be discussed with parents and/or patient are:

• The difference in length of limb shortening may be caused either by a segment or the whole lower limb or excess growth of the other. The results depend on the technical elements and the physical and mental condition of the patient. It is important that patients about to be operated to have excellent health. The patient must also be in a good psychological state, to be prepared for a long period of recovery, to observe the recovery period and be aware of complications that can occur intraoperatively and especially during the lengthening. Knowledge of these problems is even more necessary, as the child is smaller.

• As a therapeutic alternative to LLD there may be performed limb shortening. This can be achieved by resection of a bone segment, temporary or definitive epiphysiodesis. Epiphysiodesis is made to limit the growth of the longer segment. This alternative used when the difference in length between the limbs is between 2 and 5 cm, must be used before the end of the growing period, but preferably not in very young patients, because epiphysiodesis may become permanent. Epiphysiodesis best time to be performed, depending on patient sex and skeletal maturity, ranges between 11 and 14 years of age. Epiphysiodesis can be done by Phemister method, or by the destruction of growth plate with a drill guided under fluoroscopic control, or by the use of in "8", "H" or "I" plates [21] ***(Figure no. 16)***.

Figure no. 16:
Congenital pseudarthrosis of the left calf, in case of a patient with the von Recklinghausen disease, operated at the age of 9 years by Patterson method. After healing of the nonunion, the

*growth rhythm differece has led to LLD. Right proximal tibia transient epiphysiodesis with "8"
plates has been performed at the age of 14.*

• All interventions for limb lengthening have absolute indication when shortening is accompanied by an axial deviation, malrotation, genu flexum, etc.

• The operation of limb lengthening may be done at any age above three years and the LLD is greater than 3 cm, because of differences over 3 cm leading to compensatory disturbances in the foot, equins being the most common deformity [22]. Compensating this difference, rightly or wrongly, compromises static balance of the spine, resulting in time to the occurrence of scoliosis.

• The position and mobility of the pelvis should be considered in adults. The adult who has never worn orthopedic shoes has a rigid pelvic position which can not be corrected.

• In severe LLD, associated to congenital malformations, limb lengthening over 10 cm, in one step, is very carefully recommended since the results are not commensurate with the suffering that patients endure [23].

• Limb lengthening may induce as an unbearable sequel the limiting of over- and underlying joint mobility and muscle strength decrease compared to the other limb.

• The treatment with external fixator in limb shortening caused by nonunion is addressed simultaneously to nonunion and shortening.

• Taylor Spatial Frame reduces the mounting complexity, requiring bone fixing only at two levels, each consisting of a ring which is fixed to a wire and one or two pins. The columns are placed in well established points, controlled by a computer program, allowing lengthening. Each point on the bone segment will move on well established paths, while ensuring lengthening, the correction of associated deformities and stability, too *(Figure no. 17)*.

Figure no. 17:
Dischondroplasic Dwarfism: lengthening and correction of associated deformities with a TSF (Taylor Spatial Frame).
The first intervention made in Romania with a TSF, May 2009, in the Department of Pediatric Surgery and Orthopedics in "M. S. Curie "Hospital, Bucharest.

• Progress has been made in the limb lengthening in the last 10 years, applied in many centers in the world and in Romania: upgrading fixing methods initiated by Wasserstein, lengthening on two TEN rods, the use of computer navigation during partial resection of growth plate if prematurely closed, the introduction of medical image archiving, coupled with the preoperative and plannig and in dynamics, Albizzia, FITBONE and ISKD (Intramedullary Skeletal Kinetic Distractor) lengthening devices and hybrid techniques that combine internal and external fixation, thereby reducing external fixation period.

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