Results and biomechanical consideration of treatment of congenital lower limb shortening and deformity using the Ilizarov method

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One of the applications of the Ilizarov apparatus is the correction of congenital shortening and deformities. Ilizarov external fixator produces biomechanical structure with surrounding tissue, which is the reason why very important is correct stability of fixator. Large distraction in the case of high value of lengthening, and large deformity correction result in shear stresses that occur additionally in the regenerate, which can potentially lead to damage of the regenerating nutritive microcirculation of bone tissue and bone fragment displacements. Our objective was to assess the results of the Ilizarov method in the treatment of congenital shortening taking into account treatment strategy and the size of the axis of lengthening and correction. Our research problems include presenting the effects of biomechanics of musculoskeletal deformations on treatment results, presenting complications and their treatment.

Between 1989 and 2009, 62 patients underwent surgery to correct congenital lower limb deficiencies at our Clinic; 33 patients were followed-up. In total, there were 70 surgeries (2.12/patient). Axial correction was performed in 26 patients (78.79%). Average age at the start of the treatment was 15.58 years. Mean follow-up was 8.58 years.

Mean lengthening per surgery was 3.17 cm with the lengthening index of 50.7 day/cm. Results were very good for 23 patients, good for 7 patients, satisfactory for 3 patients. Complications appeared in 24 patients, problems occurred in 74.42% of the cases, obstacles in 4.65% of cases, and true complications in 20.93% of the cases.

The best results were achieved in the treatment of patients with two-stage and two-segment lengthening with a total elongation of less than 7 cm, and without correction of the axis. Congenital shortening of the lower limb should be treated comprehensively because the shortening applies to all segments, and disturbs biomechanics of all lower limb. In the case of axial correction and large amount of elongation high soft tissue forces counteract the distraction forces. Hybrid construction may help to shorten treatment time, increase fixator stability and decrease rate of complications. We suggest use of hybrid Ilizarov fixator, especially when large elongation and axis correction are planned.

Key words: bone elongation, congenital deformity, hybrid external fixator, Ilizarov external fixator

1. Introduction

One of the applications of the Ilizarov apparatus is the correction of congenital shortening and deformities. Congenital shortening of the lower limb occurs very rarely, at a rate of 1–3 per 10 000 births [9], [14], [15], [29]. The aetiology is not completely clear, it is known, however, that genetic and environmental factors play a major role [3], [9], [13]–[15], [29]. The clinical picture shows a reduction in the size of the lower extremity, consisting in its shortening and thinning. The shortening applies to the pelvis and all the segments of the lower limb: femur, lower leg, and foot in any
dimension, and in different proportions in particular segments. In many cases, other abnormalities, such as valgus or varus knee deformity, the lack of certain metatarsal bones, talocalcaneal coalition, and other foot deformities co-exist [1], [4], [9], [13], [14], [16], [18], [24], [25], [28]. Ankle joint is unstable and set in a crooked position due to the spherical shape of the trochlea of the talus in the frontal plane ("ball and socket ankle joint") [1], [11], [13], [14], [18], [28]. Aside from limb lengthening, patients frequently require correction of the associated deformities. Due to lower osteogenic potential of the bone (there are alterations in bone structure that originate during the prenatal period), treatment is very difficult [6], [9], [15], [24].

Until the 1970s orthopedic footwear was the most common method of treatment, followed by prosthesis for large shortenings, and amputation if use of prosthesis was not possible [1], [4], [7], [9], [10], [14], [15], [25]. Limb lengthening with the Wagner technique or using Orthofix stabilizer was used in exceptional cases due to a large number of complications [12].

The introduction of the external circular fixator by Ilizarov and introduction of distraction osteogenesis allowed effective treatment of these patients [9], [11], [12], [16], [25]. Ilizarov external fixator produce biomechanical structure with surrounding tissue, which is the reason why very important is correct stability of fixator. Too flexible, or too rigid an aparatus causes increased rate of complications [22], [23]. In the case of congenital deformity correction, internal forces increased due to the bad condition of the soft tissues, and poor elastic properties. The amount and direction of bone fragment displacements are associated with the biomechanical conditions in the bone regenerate, which determine whether or not the treatment process will progress in an optimal manner [22], [23]. In addition to distraction, Ilizarov’s apparatus makes it possible to correct the deformity in three planes [11], [24], [25]. Large distraction in the case of high value of lengthening, and large deformity correction results in shear stresses that occur additionally in the regenerate, which can potentially lead to damage of the regenerating nutritive microcirculation of bone tissue and bone fragment displacements. Thus far, there are only a few publications in international literature on the results and treatment of congenital shortening and deformities of lower limb using the Ilizarov method [3], [9], [11], [12], [16], [25]. Hence, our objective was to assess the results of the Ilizarov method in the treatment of congenital shortening and distortion of the lower limb, taking into account treatment strategy and the size of the axis of lengthening and correction. Our research problems include presenting the effects of biomechanics of musculoskeletal deformations on treatment results, presenting complications and their treatment.

2. Materials and methods

In the years 1989–2009, 62 patients with congenital shortening and deformity of the lower limb were surgically treated by the Ilizarov method in our institution. Due to the different characteristics of defects as well as dissimilar requirements for treatment, patients with aplasia of the fibula, tibial aplasia, congenital pseudarthrosis of the tibia, Ollier’s disease, multiple exostosis, fibrous dysplasia, and other rare congenital syndromes were not included in this group.

This research project was approved by the Bioethics Committee of Wroclaw Medical University on 19.01.2012, with decision number KB–735/2011.

A retrospective study, involving 33 patients (17 males, 16 females) who had participated in the follow-up was conducted. The mean age at the start of treatment was 15.6 years (4.5–46 years of age).

Single-stage procedures were performed in patients with completed growth. In younger patients, in whom the growth process was still ongoing, multi-stage treatment was implemented due to the recurrence of limb shortening and deformity during growth or as a result of the size of the original shortening.

Fig. 1. Patient with simultaneous two-segmental surgery – lengthening of the femur and lengthening and correction of lower leg valgus deformity.
A total of 70 surgeries were performed in the group of 33 patients; an average of 2.12 operations per patient. Single-stage lengthening was performed, in 14 patients, 9 patients underwent two-stage procedures, simultaneous two-stage lengthening of femur and lower leg (Fig. 1) was implemented in 5 patients, and 5 patients were treated with multiple-stage procedures (3–6 surgeries). The treatment strategy was chosen based on the size and location of the deformity, patient’s age, and complications resulting from the treatment.

Among the 33 patients who underwent diagnostic procedures, hypoplasia of the fibula was found in 9 cases and congenital shortening of the femur in 24. The total shortening of the lower limb at the start of treatment was 5.16 cm (0.5–15.5), representing on average 6.6% of the length of the opposite, healthy limb (0.6–17.1). In addition to the shortening, other distortions were found to be present in patients (Table 1).

### Table 1. Other deformities of the limb

<table>
<thead>
<tr>
<th>Hip</th>
<th>Hip dysplasia</th>
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<tr>
<td></td>
<td>Hip subluxation</td>
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<tr>
<td>Knee</td>
<td>Valgus</td>
<td>20</td>
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<tr>
<td></td>
<td>Varus</td>
<td>9</td>
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<tr>
<td></td>
<td>Patellar instability</td>
<td>1</td>
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<td>Shin</td>
<td>Dysplastic fibula</td>
<td>9</td>
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<tr>
<td>Ankle joint</td>
<td>“ball and socet” ankle joint</td>
<td>5</td>
</tr>
<tr>
<td>Foot</td>
<td>One or more metatarsal bone agenesis</td>
<td>7</td>
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<tr>
<td></td>
<td>Equino varus</td>
<td>2</td>
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<td></td>
<td>Plano valgus</td>
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<td>Talo calcaneal coalition</td>
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<tr>
<td></td>
<td>Other</td>
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Axis correction with lengthening of the limb was performed in 26 patients (78.79%), the remaining 7 patients (21.21%) underwent lengthening without correction of the axis.

The basic fixator design for femur lengthening consisted of one proximal arch fixed with two Schanz screws, a proximal ring fixed with two K-wires and a distal ring fixed with three K-wires. The basic fixator design for shank lengthening consisted of one proximal ring with three K-wires, middle ring fixed with two K-wires, and a distal ring fixed with two K-wires. We had used a hybrid Ilizarov fixator, which has at middle or/and distal ring in femur configuration one or two Kirschner wires replaced by Shantz screws (Fig. 2). In shank fixator construction proximal ring had one Kirschner wire replaced by Shantz screws.

Corticotomy was carried out at the distal femoral metaphysis, at the proximal metaphysis of tibia, and at 1/3 proximal of fibula in the lower leg, or at the apex of the deformity.

In the case of a significant shortening of the distal end of fibula, an additional elongation of the distal metaphysis of this bone was performed in order to restore ankle stability, and to correct the proportion between tibia and fibula for improving the normal biomechanics of the ankle. Walking with partial weight bearing on the leg was initiated two days after surgery. Distraction and correction was initiated 6 days after surgery. Check-ups were carried out at intervals of 2–6 weeks. The apparatus was removed when the presence of at least 3 of 4 cortical layers was found.

The evaluation of the results of the treatment was conducted with Paley’s [27] classification of complications and a point scale for rating the lower limb developed at our Clinic, which has been adopted by treatment centers using Ilizarov method in our country [19], [20]. The following elements were taken into consideration in the point scale system: the size of segment shortening, locomotive abilities, axis deformation, lower limb muscle strength, range of motion in the joints of the lower limb, orthopedic aids, and the subjective evaluation of the patient. In assessing the results of treatment according to this scale, we compared the differences between the number of points obtained before and after treatment. In addition, an analysis of treatment outcomes including the incidence of complications was performed.

The comparison made by us was based on treatment strategy (single-stage, two-stage, multi-stage, single-segment, two-segment), the size of lengthening (<7 cm, >7 cm), and the correction or lack of correction of the axis of lengthening of the limb.
No additional sources of financing were accepted for the purposes of this work.

For the statistical analysis of the results we used: Pearson’s correlation coefficient, Cramer’s V correlation coefficient, Spearman’s rho correlation coefficient, and Kendall’s tau-b correlation coefficient. All coefficients were tested for their statistical correlation by tests available in the statistical package employed – SPSS Statistics 17.0. One-way analysis of variance ANOVA was used in the verification of the hypothesis of equality of mean values of the variable “result of improvement” in the distribution of treatment strategy. The Shapiro–Wilk and Kolmogorov–Smirnov tests were used to confirm the hypothesis of normality when the use of test data required normal distribution. For comparison of mean values of the variable “result of improvement in groups” Student’s t test was used. Wilcoxon paired test was used to compare the effect of treatment on scoring. Statistical significance was defined at the level of $\alpha = 0.1$, as $p \leq 0.05$.

3. Results

The mean follow-up was 8.58 years (2.08–22.66). Average size of lengthening during one surgery was 3.17 cm (0.5–7). The average size of the total lengthening in the same patient was 6.73 cm (0.5–19). 11 patients had total lengthening greater than 7 cm. The mean duration of one stage of treatment was 160 days. The average lengthening index was 50.7 days/cm. Average score in the evaluation of lower extremity before treatment was 69 points and 90 points after treatment. The difference was statistically significant ($p < 0.001$, Wilcoxon test).

Adverse events occurred 43 times in 24 patients (72.73%), on average 1.3 times per patient. Adverse events included the occurrence of problems 32 times (74.42% of adverse events), difficulties 2 times (4.65% of adverse events), and complications 9 times (20.93% of adverse events).

The knee joint range of motion limitation occurred 12 times (27.91% of adverse events). In 11 cases, contracture was abolished or significantly reduced following intensive rehabilitation, while one patient required surgery to lengthen the tendon of rectus femoris muscle with extensive plastic surgery and dissection of vastus intermedius muscle. Infections around implants appeared 7 times (16.28% of adverse events). In 6 cases the infection was controlled with local and systemic administration of antibiotics. One patient required surgical resection of the Kirschner wire and debridement of the site of infection. Deformation within the regenerate occurred six times (13.95% of adverse events), in 4 cases requiring the adjustment of the apparatus, and in two cases deformity correction was achieved after re-operation using Ilizarov method. Delayed union occurred 5 times (11.63% of adverse events). In these patients, union of the regenerate was achieved after compression of regenerate in 1 case, in 2 cases after removal of the apparatus and immobilization in a cast, and in 2 cases after the removal of the apparatus and limb immobilization in a brace. Fracture of regenerate occurred 5 times (11.63% of adverse events). Bone union was achieved after immobilization of the limb in a cast in 4 cases and after a closed repositioning and stabilization using Ender rods in one case. Deep vein thrombosis was observed two times (4.65% of adverse events); in each case it resolved following drug therapy. Subluxation of the knee occurred two times (4.65% of adverse events), requiring the repositioning and stabilization using the Ilizarov apparatus attached to the lower leg. Premature union of the regenerate occurred 1 time (2.33% of adverse events); this patient was treated with repeated surgical dissection of the regenerate and additional distraction. Vascular injury occurred 1 time (2.33% of adverse events) and required surgery to repair the vessel (femoral artery). There was no damage to the nerves. Breakage of Kirschner wire occurred twice (4.65% of adverse events) and in both cases required surgical re-stabilization of Ilizarov apparatus by exchanging Kirschner wire.

Depending on the treatment strategy statistically significant difference in the number of complications and outcome improved were not found in various groups ($p = 0.31$ one-way analysis of variance ANOVA).

In the group with lengthening of >7 cm results after the treatment had improved by an average of 26.73 points, while in the case of patients with lengthening of <7 cm by 18.82 points. As shown by Student’s t test, the difference between the two groups is statistically significant ($p = 0.033$). Lengthening > 7 cm had an effect on the occurrence of adverse events. Amongst the variables “total lengthening” and “number of complications” there is a negligible significant stochastic dependence at a level of 10% ($p = 0.078$) and equal to 0.311 (Spearman’s rho coefficient).

Correction of the axis also influenced the occurrence of adverse events. In the group with axis correction the outcome was improved after treatment by an average of 21.54 points, while for patients without axis correction by 21.14 points. Cramer’s V correlation coefficient for these variables is 0.348 and is sta-
4. Discussion

In previous studies, most authors separated congenital femoral shortening from congenital shortening of the fibula and congenital shortening of the tibia, presenting the results of the methods and results of treatment of these diseases in separate publications [3], [9], [10], [12], [14], [16], [25], [26].

We believe that congenital shortening of the lower limb should be treated comprehensively because the shortening applies to all segments of the limb, and disturbs biomechanics of all lower limb. Often, in patients with a hypoplastic fibula or tibia, the shortened femoral segment also requires lengthening. A similar situation occurs in the case of congenital shortening of the femur, where in many cases, the shortened lower leg also requires lengthening.

As a result of the character and extent of defect, a large proportion of patients with congenital shortening and deformity of the lower limb require two-segment or multi-stage treatment in order to restore the limb axis and achieve equal limb length to create normal, or similar to normal biomechanics conditions. Morasiewicz shows that Ilizarov patients with congenital aetiology had larger gait parameter asymmetry than patients with posttraumatic or postseptic aetiology. She suggests that patients with congenital aetiology, from birthday put compensatory mechanism, which does not allow for gait symmetry, despite complete leg deformity correction and egalisation. In the group of patients with congenital shortening and deformity, complete leg deformity correction and egalisation, do not restore primary limb function, but give new biomechanics conditions for obtaining correct limb function [21].

The best results were achieved in patients with two-stage and two-segment elongation with a total elongation of less than 7 cm and without correction of the axis.

Lengthening above 7 cm with axis correction and multi-stage elongation had slightly worse results. The greatest improvement was achieved in patients with lengthening above 7 cm. A significantly lower incidence of complications was recorded in the group without correction of the axis. After osteotomy, lower limb biomechanical configuration undergoes disturbance. Bone fragment cannot transfer limb load, muscle induced bone fragment displacements.

Distraction forces increased during rise of a value of lengthening [2], (Fig. 3a–b). Worse results in the group with lengthening above 7 cm can be due to greater soft tissue forces and stress transfer in fixator which have unfavourable influence on bone fragment displacements and regenerate formation. During elongation and axis correction the influence of soft tissue
is bigger than that of bone regenerate [5]. In the case of axial correction and large amount of elongation greater soft tissue forces counteract the distraction forces than in small elongation without axis correction. Axial deformity disturbs anatomy and biomechanics of lower limb, which effect asymmetric forces and muscle tension distribution (Fig. 4). In the group with axis correction asymmetric forces can potentially lead to damage of the regenerating nutritive microcirculation of bone tissue and bone fragment displacements (Fig. 5 a–c). Also incorrect hinge assembly can cause bone fragment displacements (Fig. 6). In the group with axis correction, velocity of bone fragment displacement was different in the case of different distance from the center of rotation. Part of regenerate had greater angular velocity, which can lead to regenerate disruption. Part of regenerate had smaller angular velocity, which can lead to premature consolidation [17], (Fig. 7). Worse results in the group with multi-stage treatment might be caused by significant shortening and axis deformity, which effect asymmetric and greater forces and muscle tension distribution (Figs. 3–5) leading to damage of the regenerating nutritive microcirculation of bone tissue and bone fragment displacements. In multi-stage treatment, with increasing total fixator time, complications and bad results occur more frequently. The differences in terms of improvement as scored by the point scale, depending on the strategy of treatment, are not statistically significant. It is difficult to determine if this is
not a type 2 error, owing to the small size of the sub-
groups. Too small numbers of individuals in the groups
did not allow statistical analysis of the dependence of
the development of adverse events based on treatment
strategy. In the future, studies with more numerous
groups are indicated.

Lengthening index is high, compared to patients
treated with the Ilizarov method due to post-trauma or
post-inflammatory shortening and distortion. This is
due to inferior quality of bone tissue, which probably
has less osteogenic potential caused by congenital
deformities.

In the group with multi-stage treatment, distraction
>7cm and correction of the axis, there were often ad-
verse events associated with limited adaptation poten-
tial of soft tissues, most common was limitation of
joint mobility (Fig. 8). Also, in this group the appara-
tus was used for a longer time period, which was as-
sociated with increased adverse events.

The most common adverse event was limitation of
joint mobility, infection around the implant, delayed
union and deformation of the regenerate. Our modi-
fication of the apparatus, which consisted in introducing
an arch mounted in the intertrochanteric area by the
Schantz screws, in the case of femur lengthening, im-
proved fixator stability, reduced the frequency of limi-
tation in hip joint mobility and led to an improvement
in patient’s functioning. Application of hybrid Ilizarov
device, which has at middle or distal ring in femur, or
proximal ring in shank, one or two Kirschner wires
replaced by Shantz screws, improved fixator stability,
increased control of deformity correction, reduced soft
tissue transfixion, made a fixator application easier,
increased patient comfort, reduced the rate of pin-site
infection and limitation of joint mobility [8]. Numerous
complications have been reported in literature with
regards to the treatment of congenital deformities of the
lower limb using Ilizarov method [3], [7], [9], [11],
[12], [16], [24], [25], [27]. Frequently, this is due to the
coeexistence of shortening and large deformations as
well as a poor quality of bone tissue. Hybrid construc-
tion may help to shorten treatment time, increase pa-
tient tolerance and decrease rate of complications. De-
spite the high percentage of complications, the Ilizarov
method is superior in the treatment of congenital short-
ening of lower limbs in comparison to other treatments
[11]. We suggest using hybrid Ilizarov fixator for cor-
rection and elongation in patients with congenital aeti-
ology, especially when large elongation and axis cor-
rection is planned.
According to our experience and results, the Ilizarov method is effective and has an acceptable rate of complications. This method allows not only to lengthen, but also to correct the axis of the limb in many planes and restore biomechanics conditions [11, 24, 25]. Modular system allows the Ilizarov apparatus, without any additional surgery, to change the design of the stabilizer during the treatment in order to perform axis corrections, whether primary or developed in the course of the treatment.

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References