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Limb lengthening and deformity correction in patients with severe fibular hemimelia: experience of the children's university hospital in Belgrade

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Abstract

Background Fibular hemimelia (FH) is a congenital disease manifested by valgus deformity and instability of the knee joint, shortening and deformity of the tibia, hypoplasia and deformities of the foot and ankle.

The aim of this paper was to determine the efficacy of the strategy of separate reconstruction of the foot along with correction of tibia deformity, and then tibial lengthening in patients with FH of types 3 and 4 according to Paley.

Methods This retrospective study is based on an analysis of the treatment of 12 children with severe FH. The patients, aged no older than 24 months, were treated for foot reconstruction along with correction of tibial deformity followed by a separate stage of tibial lengthening. Tibial lengthening was performed in the age 4.6 ± 1.2 years. The long-term result of treatment was evaluated at least 1 year after the end of tibial lengthening. Evaluation criteria after tibia lengthening were external osteosynthesis index, amount of lengthening, assessment of outcomes according to Lascombes. Walking ability was assessed using Gillette questionnaire.

Results The approach we used gave excellent and good results in 83 % of cases after the first reconstructive stage. Complications and recurrences of deformities encountered during the first stage were eliminated during subsequent planned limb lengthening. The average magnitude of lengthening was 6.4 ± 2.4 cm (37.2 ± 12.4 % of the initial segment length). The index of external osteosynthesis was 22.9 ± 12.2 days/cm. Monofocal distraction osteosynthesis was used in 9 cases and bifocal osteosynthesis in three cases. The results of lengthening were classified by Lascombes as IA in 7 cases, IB in four cases, 2B in one case.

Discussion In severe FH, the question of reconstruction or early amputation remains open. There are two opinions on the staging of reconstructive orthopedic surgery and tibial lengthening in young children with severe FH.

Conclusion The strategy of reconstruction of the foot and ankle joint at an early age (16-24 months) in children with severe FH followed by lengthening of the lower leg (at the age of 4-6 years) proved to be effective and can be used when it is chosen by the patient's parents. In 3C type cases, the use of external fixation to correct the deformity and simultaneously lengthen the tibia at the first stage is an alternative reasonable strategy option.

Keywords: fibular hemimelia; reconstruction; limb lengthening; Ilizarov method

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INTRODUCTION

Fibular hemimelia (FH) is a congenital malformation where part or all of the fibula is hypoplastic, dysplastic, or absent, and this is associated with dysplasia or hypoplasia of the tibia and foot. FH refers to the so-called postaxial anomalies, in contrast to tibial hemimelia (preaxial type of longitudinal anomaly). The incidence of FH varies between 1:40,000 and 1:50,000 newborns [1, 2]. The etiology of this anomaly remains unknown in most cases, usually classified as embryopathy [3, 4]. The main orthopedic problems in fibular hemimelia are length discrepancy of the lower limbs, instability and valgus deformity of the knee joint, tibial deformity, deformities and abnormalities of the foot and ankle joint [5, 6]. In severe FH forms, the question of reconstruction or early amputation remains open [7-10]. There is a consensus that the success of reconstructive treatment is due to the achievement of a weight-bearing position of the foot and the elimination of deformities of the ankle joint and lower leg [10, 11]. The age of the child when indications for the first stage of reconstruction (correction of deformities of the foot, ankle and tibia) are optimal is between 18-24 months [6].

Surgical treatment provides the correct foot position, ensures the function of the ankle joint, achieves vertical posture of the child, and develops independent walking with orthoses that compensate for the difference in length [12]. The operation can be combined with leg lengthening [6, 12], which, however, is accompanied by a long period of wearing an external fixation device. The decision on both the treatment strategy and the scope of primary intervention should consider the opinion of the parents [9]. Thus, the first stage of elongation can be delayed until the age of 4-8 years [13-15]. So, there are two opinions about the combination and stages of reconstructive orthopedic intervention on the limbs and leg lengthening in young children with severe FH.

This retrospective study analyzed the results of reconstructive surgical treatment and tibial lengthening in patients with severe FH (Paley types 3 and 4), when parents preferred tibial lengthening delayed for several years after primary surgical reconstruction of the foot and ankle.

Purpose To determine the effectiveness of the strategy of separate implementation of the foot reconstruction stage combined with correction of the tibial deformity and tibial lengthening stage in patients with Paley FH types 3 and 4.

MATERIALS AND METHODS

This retrospective study included 12 patients with fibular aplasia (7 boys and 5 girls).

The inclusion criteria were that the stage of foot reconstruction and correction of the tibial deformity be performed separately from the stage of the first tibial lengthening due to the choice of this treatment strategy by child's parents. Other inclusion criteria were severe FH, classified as types 3 and 4 according to Paley [15], completion of the first stage of treatment in patients no older than 24 months, completion of the second stage of treatment (tibial lengthening), the ability to evaluate the result at least 1 year after the completion of leg lengthening.

This study did not include patients with milder FH, patients whose reconstructive treatment was started at a later age, as well as cases where it was not possible to evaluate the result of lengthening one year or more after removal of the Ilizarov apparatus.

The features of the first (reconstructive) stage of surgical treatment were the following mandatory elements of the intervention: correction of tibial deformity using the resection type of closed-wedge osteotomy, corrective osteotomy through the area of the talocalcaneal coalition of the foot, resection

of the rudiment of the fibula, lengthening of the triceps using the type of aponeurotomy of the calf muscles, Z-lengthening of the peroneal tendons (or single muscle), ankle capsulotomy, internal osteosynthesis (bone locking plates or threaded pins) for fixation of tibial fragments, threaded immersion wires or external diafixation wires for fixation of the position of hindfoot and ankle joint fragments (not changing the relationship between the tibia and talus), as well as plaster immobilization. The duration of plaster fixation was 6-8 weeks; removal of the plaster cast was accompanied by the removal of the diafixation wires (in this type of fixation). Subsequently, before performing the leg lengthening stage, patients used devices with hinges inserted into shoes (AFO – ankle-foot orthosis type); walking with full load in shoes with compensation for shortening was allowed.

Tibial lengthening (the second stage of treatment) was performed using the Ilizarov apparatus in all cases, at one or two levels. Simultaneously with the application of the Ilizarov apparatus, the previously applied osteosynthesis material was removed if it interfered with external osteosynthesis. Elastic reinforcement was used in 9 cases. Lengthening of the tibia was combined with correction of deformities if necessary; osteosynthesis was carried out with protection of the knee and ankle joints, taking into account the instability of adjacent joints inherent in these severe forms.

The evaluation criteria after the first stage of treatment were the parameters of anatomical X-ray angles of orientation of the articular surfaces in the frontal (lateral distal tibial angle – aLDTA) and sagittal (anterior distal tibial angle – aADTA) planes, deviation of the biomechanical axis from the center of the knee joint (mechanical axis deviation – MAD) [16, 17], as well as the tibiotalar (TT) and tibiocalcaneal (TCC) angles in the standing position under limb loading. A clinical assessment of the position of the foot, the supportability of the foot and the ability to weight-bearing walking, and the child's motor activity was also carried out. Walking ability was examined using the Gillette questionnaire [18].

The evaluation criteria after lengthening the tibia were the index of external osteosynthesis, the amount of lengthening (cm and %). In general, the evaluation of the elongation stage was carried out according to the criteria of P. Lascombes [19]. The complications encountered, the treatment/measures applied to them and the consequences of the complications were also assessed.

Statistical assessment included descriptive parameters (mean values, standard deviations); the Attestat 12.0.5 software was used.

RESULTS

The first stage of treatment was carried out in the age from 16 to 24 months. FH cases according to the Paley classification were: type 3A (3 patients, 25 %), type 3B2 (4 patients, 33.3 %), type 3C (4 patients, 33.3 %), type 4 (1 patient, 8.3 %). In all cases, shortening of the femur was less than 1 cm, while in 3 cases there was a valgus deformity of the knee joint of no more than 10°, caused by hypoplasia of the lateral condyle of the femur.

After the first stage, the treatment goals were achieved in 10 cases (Fig. 1). Two cases had foot equinus and relapse of the hindfoot deformity and did not allow achieving good treatment results (Table 1). Among other complications of the first stage, there was local skin necrosis in three cases, which required conservative treatment for wound healing by secondary intention, and one case of superficial infection in the area of the diafixation wire.

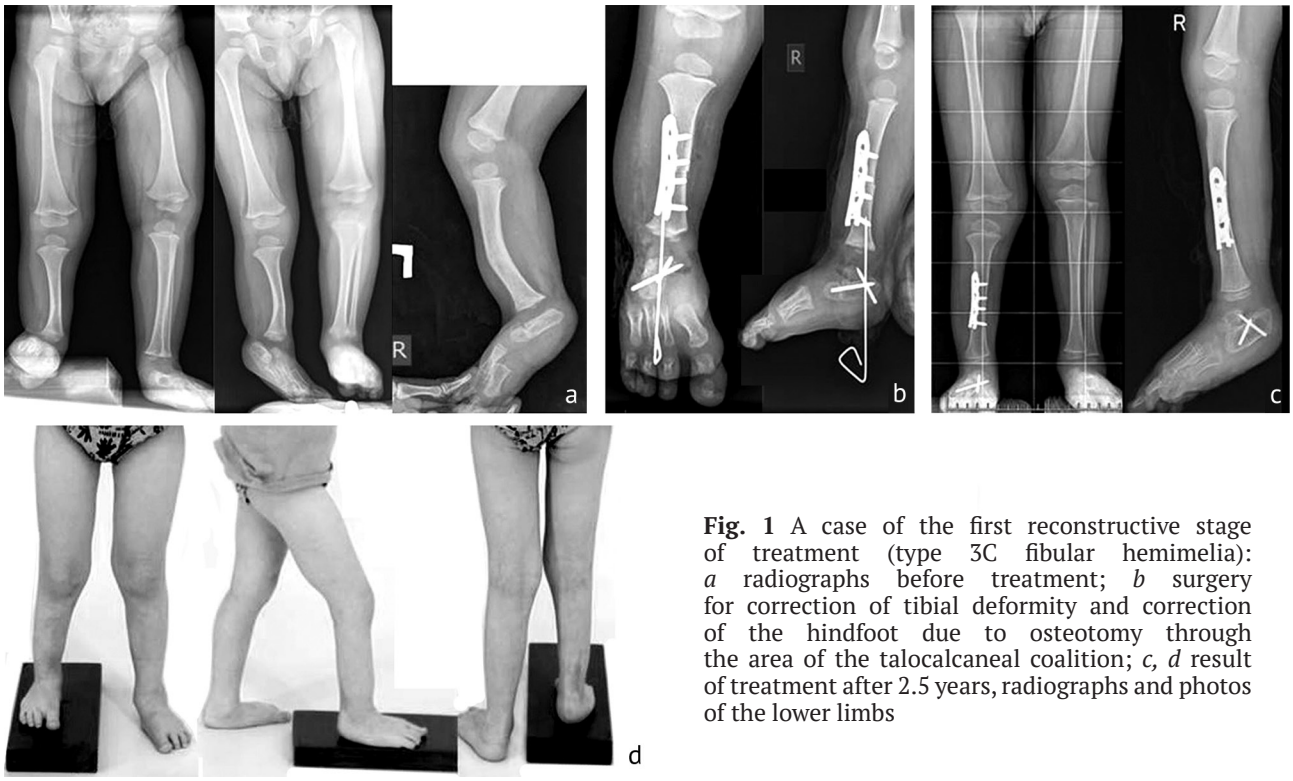

Fig. 1 A case of the first reconstructive stage of treatment (type 3C fibular hemimelia): *a* radiographs before treatment; *b* surgery for correction of tibial deformity and correction of the hindfoot due to osteotomy through the area of the talocalcaneal coalition; *c, d* result of treatment after 2.5 years, radiographs and photos of the lower limbs

Table 1

Results of radiographic studies after correction of lower limb deformities in FH patients

Patients	LDTA; °		ADTA; °		MAD; mm		TTA; °		TCCA; °	
1	68	84	84	83	12	4	120	97	80	117
2	69	86	85	83	14	3	119	97	74	105
3	74	88	84	84	12	0	117	95	69	110
4	84	87	83	82	15	3	123	99	68	106
5	83	85	84	84	15	4	130	98	70	107
6	82	85	83	87	16	5	125	95	78	99
7	84	87	85	85	18	2	130	95	67	106
8	71	85	86	85	21	2	130	111*	66	72*
9	73	90	89	84	11	0	141	98	58	108
10	72	84	85	85	18	3	134	145*	64	55*
11	68	83	89	82	22	4	142	97	65	105
12	66	86	87	84	18	6	139	99	64	116

Note: * – cases of hindfoot deformity recurrence and foot equinus

According to the Gillette questionnaire, the assessment of walking ability, which was influenced not only by anomalies of the limb development but also by age, before treatment was: level 3 – 3 patients, level 4 – 7 patients, level 5 – 2 patients. One year after the first stage of reconstructive treatment: level 7 – 6 patients, level 8 – 5 patients, level 9 – 1 patient.

The second stage of treatment to lengthen the tibia (Fig. 2) was performed in patients aged 3.5-6 years (mean age, 4.6 ± 1.2 years). The average lengthening was 6.4 ± 2.4 cm (37.2 ± 12.4 % of the initial segment length). The external osteosynthesis index averaged 22.9 ± 12.2 days/cm. Monofocal distraction osteosynthesis was used in 9 cases, bifocal in three cases. Correction of foot equinus and gradual correction of the hindfoot deformity due to osteotomy of the calcaneus were carried out simultaneously with monofocal lengthening of the tibia (Fig. 3; Fig. 4).

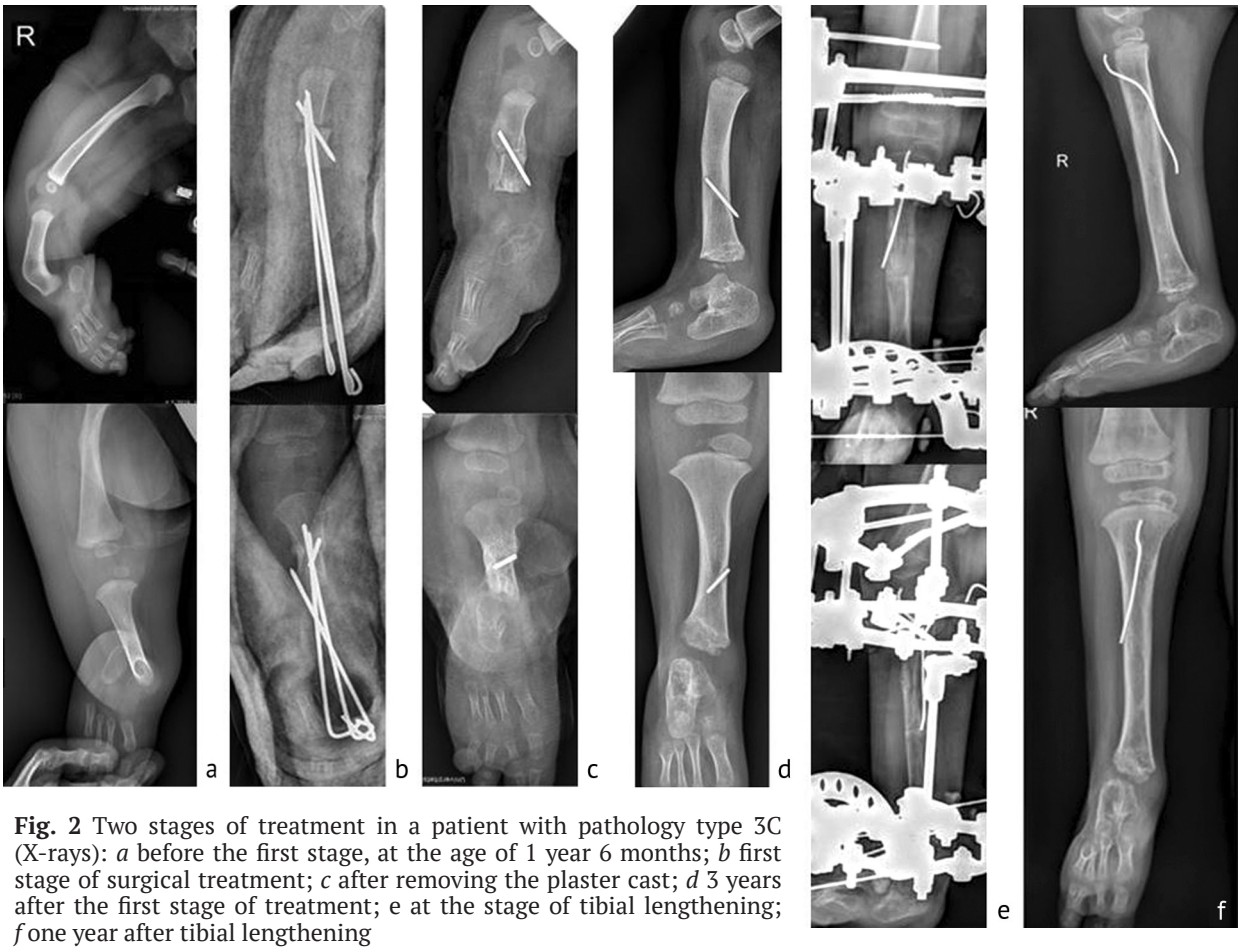

Fig. 2 Two stages of treatment in a patient with pathology type 3C (X-rays): *a* before the first stage, at the age of 1 year 6 months; *b* first stage of surgical treatment; *c* after removing the plaster cast; *d* 3 years after the first stage of treatment; e at the stage of tibial lengthening; *f* one year after tibial lengthening

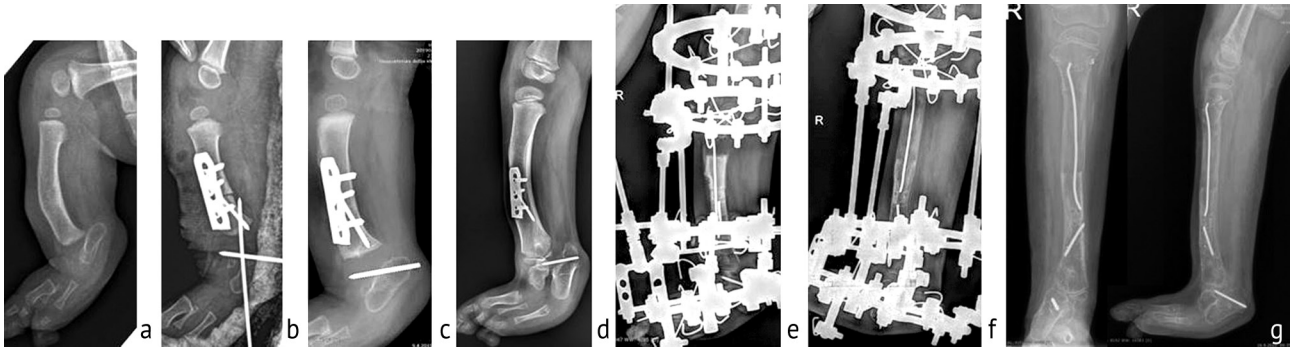

Fig. 3 A case of developed foot equinus and its correction (X-rays): *a* before surgery; *b* first stage of deformity correction; *c* after removing the plaster cast; *d* equinus of the hindfoot 2 years after surgery; *e* second stage of treatment, lengthening with simultaneous gradual closed elimination of foot deformity using the Ilizarov apparatus; *f* before removing the Ilizarov frame; *g* after removing the frame, the foot remains in a satisfactory position, close to full weight bearing

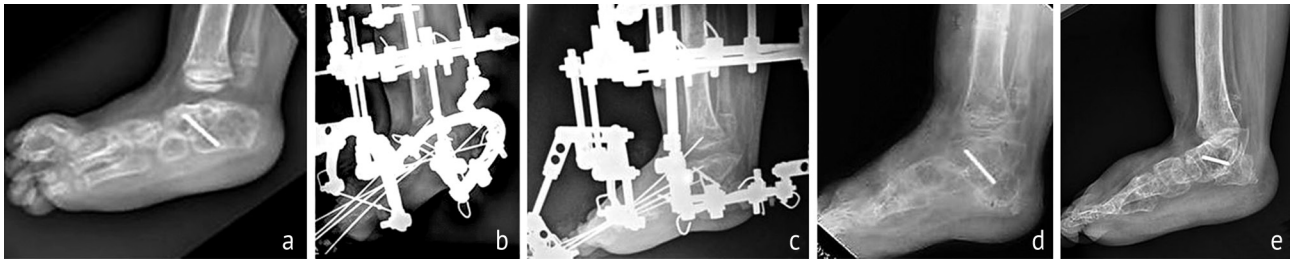

Fig. 4 Correction of recurrent deformity of the calcaneus, radiographs (FH type 3C): *a* the foot before the lengthening stage, tibial-calcaneal angle 72°; *b* at the beginning of foot deformity correction; *c* after correction of the deformity achieved, the angular divergence of the calcaneal fragments is clearly visible; *d* after removal of the Ilizarov apparatus; *e* follow-up after 1.5 years, the treatment result is maintained

Among the complications of the second stage of treatment (tibial lengthening), there was one case of a tibial fracture after removal of the Ilizarov apparatus, which required fixation of the limb in a plaster cast and was completed without loss of lengthened magnitude and secondary deformities. Four children had superficial infection near the wires. In such cases, antibiotic therapy was used, and regular dressings were effective. Thus, the results of limb lengthening using the Ilizarov apparatus were classified according to P. Lascombes as 7 cases of category IA, four cases of IB, and one case of 2B.

DISCUSSION

The most severe forms of congenital fibular hemimelia (types 3 and 4 according to the Paley classification) are accompanied by deformities of the tibia, ankle dysplasia, absence of several rays of the foot, tarsal coalition, usually in a pronounced valgus or varus position of the foot [6, 20]. The disorders are so pronounced already at an early age that they complicate or completely hinder wearing orthotic devices and become the main reason for the delay or complete absence of walking function in children [21, 22].

Therefore, early amputations and prosthetics are an alternative treatment option: Syme and Boyd methods are common interventions [23, 24]. Studies of motor activity and quality of life conducted in groups of adolescents and adults after amputation (followed by prosthetic fitting) and after reconstructive treatment showed the absence of statistically significant total indicators of gait analysis, as well as comparable socio-psychological adaptation, quality of life and physical activity [7].

We emphasize that surgical treatment in this pathology should be aimed at achieving the highest possible functional result, and may include corrective osteotomies, lengthening, and complex reconstruction of soft tissues in the treatment plan. The treatment strategy depends on both the severity of the anomaly and the family's choice [9].

If reconstructive strategy is chosen, the most important element of treatment is correction of deformities of the lower leg, ankle joint area and functional position of the foot [25]. An interesting study by EJ Morris et al. [10] compared the functional results of a 6-minute walk test between the patients who underwent foot and ankle reconstruction and a group that underwent both reconstruction and subsequent tibial lengthening. The first group showed the best functional results.

From the same point of view, we also consider the conclusions that the lack of success of reconstructive surgery in severe FH is due to residual or recurrent deformities of the ankle, foot and leg, and not to the difference in the length of the lower extremities [11, 26, 27].

Therefore, the first stage of surgical treatment, performed at an early age, determines the success of all subsequent staged surgery.

The results of our study are concordant with the above conclusions about the importance of the functional result of the first intervention. The approach we used ensured excellent and good results in 83 % of cases after the first reconstructive stage. It should be noted that the complications encountered and recurrences of stage 1 deformities were eliminated during the subsequent planned limb lengthening. However, both patients who had recurrent foot deformities had limited functional abilities (walking and wearing shoes) between treatment phases.

The reason for the recurrence of foot deformities, in our opinion, lies in the complexity of the surgical technique. Due to deficiency in the length of soft tissues, correction of tibial deformity and foot deformity requires a shortening osteotomy of the tibia, which can be significant and is not always possible under the conditions of internal osteosynthesis.

The insufficient amount of resection in our series, combined with the limited time and effectiveness of fixation with a plaster cast and diafixation wires, caused pronounced soft tissues tension, which led to a relatively rapid relapse of the foot deformity. It is obvious that the use of external fixation, when reconstructive treatment and simultaneous lengthening of the lower leg are combined, does not pose the risk of excessive shortening of the segment, short-term fixation of the foot and lower leg, and can be recommended for the most severe FH types (3C), accompanied by severe deformities of the foot and lower leg [6, 12].

We consider another disadvantage of the applied approach to be the need to remove the osteosynthesis material (as a separate stage during the tibia lengthening operation), which limits the possibilities of distraction osteosynthesis or increases the surgical burden on the patient. It is obvious that the use of transphyseal intramedullary osteosynthesis elements during the first stage of surgical treatment, over which lengthening can be carried out subsequently, may be the most rational solution [28].

However, our small series of cases showed the rationality of the approach of separating the reconstructive stage of treatment (correction of tibial and foot deformities) in children aged 16-24 months and the stage of leg lengthening with the Ilizarov apparatus. This is important both from the point of view of restoration and development of limb function, and transferring the stage of lengthening with an external fixation device, which is difficult for a child, to an older, but preschool age.

CONCLUSION

The strategy of foot and ankle reconstruction at an early age (16-24 months) in children with severe FH, followed by lengthening of the tibia at the age of 4-6 years, has proven to be effective and is used when chosen by the patient's parents. In case of significant shortening osteotomy planned at the first treatment stage to correct tibial deformity, external fixation to correct the deformity and simultaneously lengthen the tibia is an alternative reasonable strategy.

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