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LENGTHENING OF THE ULNA BY EXTERNAL FIXATION In Children with congenital radial club hand

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Background. Congenital radial club hand is characterized by the radial deviation of the hand, the longitudinal underdevelopment of the forearm, and the dysfunction of the upper limb. The shortening of the ulna is observed in all types of congenital radial club hand. The average shortening of the ulna surgical treatment was 33.3% compared to the intact contralateral side.

Aim. This study aimed to evaluate the results of ulna elongation by the method of external fixation, depending on the level of osteotomy, in patients with congenital radial club hand type III–IV.

Materials and methods. The treatment results of 36 patients with congenital radial club hand type III–IV from 1998 to 2018 were analyzed. The average age of the patients was 7.4 years \pm 3.5 years. The patients were divided into three groups, depending on the level of ulnar osteotomy. Shortening of the ulna, correction of the angle of deformity of the ulna, radial deviation of the hand, period of correction, elongation obtained, index of fixation and osteosynthesis, and associated complications were analyzed.

Results. The observation period was an average of 5.8 years. Before surgical treatment, the ulna was 33.3% shorter, while after surgery, it was 16%. Before surgery, the angle of deformation was $20.5^{\circ} \pm 14.8^{\circ}$, while after surgery, it was $7.4^{\circ} \pm 5.6^{\circ}$; this gives an angle of deformity correction of 63.9%. The elongation of the ulna was 3.2 ± 1.1 cm. In patients who underwent proximal osteotomy, the resulting elongation was 32% and 18.4% more, respectively, than in patients who underwent an osteotomy in the middle and distal sections of the ulna. In group 1, the correction period was 24.4% and 28.9% more than in groups 2 and 3, respectively. The index of fixation in group 1 was 53.6%, which was 45.7% less than in groups 1 and 3. Postoperative complications included a false joint (15%), inflammation (10%), and forearm deformities (7.5%). **Conclusions.** In patients with congenital radial club hand type III–IV, the optimal part of an ulna osteotomy is the proximal section. With a hand deviation of more than 20° , osteotomy is performed in the distal section with simultaneous correction of the deformity.

Keywords: congenital radial club hand; external fixation; lengthening.

УДЛИНЕНИЕ ЛОКТЕВОЙ КОСТИ МЕТОДОМ Дистракционного остеосинтеза у детей с врожденной лучевой косорукостью

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Обоснование. Врожденная лучевая косорукость характеризуется лучевой девиацией кисти, укорочением предплечья и ограничением функции верхней конечности. Укорочение локтевой кости встречается при всех типах лучевой косорукости. До оперативного лечения локтевая кость была укорочена в среднем на 33,3 % по сравнению с локтевой костью интактной конечности. Цель — оценить результаты удлинения локтевой кости методом дистракционного остеосинтеза в зависимости от уровня остеотомии у пациентов с врожденной лучевой косорукостью III–IV типов.

Материалы и методы. Проведен ретроспективный анализ результатов лечения 36 пациентов с врожденной лучевой косорукостью III–IV типов в период с 1998 по 2018 г. Средний возраст пациентов составил 7,4 ± 3,5 года. Пациенты были разделены на три группы в зависимости от уровня выполнения остеотомии локтевой кости. Проанализированы основные показатели: процент укорочения и коррекция угла деформации локтевой кости, лучевая девиация кисти, период коррекции, полученное удлинение, индекс фиксации и остеосинтеза, осложнения.

Результаты. Период наблюдения составил в среднем 5,8 года. Процент укорочения локтевой кости по отношению к интактной конечности до оперативного лечения составлял в среднем 33,3 %, а после — 16 %. В дооперационном периоде угол деформации локтевой кости — $20,5 \pm 14,8^{\circ}$, а после операции — $7,4 \pm 5,6^{\circ}$, полученная коррекция угла деформации — 63,9 %. Локтевая кость была удлинена на $3,2 \pm 1,1$ см. У пациентов с остеотомией в проксимальном отделе локтевой кости достигнутое удлинение было на 32 и 18,4 % больше, чем у пациентов, которым выполняли остеотомию в средней и нижней третях. В 1-й группе период коррекции был на 24,4 и 28,9 % больше, чем во 2-й и 3-й группах соответственно. Индекс фиксации в 1-й группе был на 53,6 и 45,7 % меньше, чем во 2-й и 3-й группах. Наиболее частые осложнения — формирование ложного сустава (15 %), воспалительные процессы (10 %), вторичные деформации предплечья (7,5 %).

Заключение. Исследование показало, что для удлинения локтевой кости у пациентов с врожденной лучевой косорукостью III–IV типов оптимальной зоной остеотомии является ее проксимальный отдел. Однако при девиации кисти более 20° рекомендовано выполнение остеотомии в дистальном отделе локтевой кости с одномоментной коррекцией деформации.

Ключевые слова: врожденная лучевая косорукость; удлинение локтевой кости; компрессионно-дистракционный остеосинтез.

The characteristics of congenital radial clubhand are as follows: radial deviation of the hand, a considerable shortening of the forearm, and a pronounced limitation of the function of the entire upper limb [1–3]. In 80% of cases, this pathology is combined with hypoplasia of finger I, whereas 50% of cases contains aplasia of ray I [4]. The incidence of congenital radial clubhand varies from 1: 30,000 to 1: 100,000 newborns. Bilateral lesion occurs in 38–58% of cases [5, 6].

In clinical practice, the Bayne and Klug classification is used to describe the congenital radial clubhand, which provides four different types of clubhand [7]. All types of clubhand are characterized by shortening of the ulnar bone as compared to the intact limb, wherein underdevelopment persists during the growth of the child.

Shortening and deformity of the forearm in such patients cause severe functional and cosmetic defects of the limb [8].

Clubhand I and II are reported on an average of 21% of all cases of congenital radial clubhand [9]. In these patients, it is possible to lengthen both bones of the forearm simultaneously in order to eliminate the difference in the length of the upper limbs.

Type III–IV radial clubhand is the most common one, and occur in about 79% of cases. At the first stage, the objective of surgical treatment is to eliminate radial deviation and restore the possibility of bilateral grip by the hand [6, 7, 10–12]. Subsequently, most parents of patients complain of shortening of the forearm. The length of the ulnar bone in patients with type III–IV radial clubhand on an average range from 51.4 to 75.3% of the length of the ulnar bone on the intact limb [13, 14], which leads to severe cosmetic defects, especially in patients with unilateral lesions. In patients with bilateral lesions, a limitation of self-care is noted due to a significant shortening of the forearm.

The compression-distraction osteosynthesis method is used to eliminate the differences in the length of the forearms [15–18].

Based on the literature, the Ilizarov method is often used to restore the length of the lower extremities. A few publications are skeptical in using this technique for elongating the bones of the upper limb. These studies describe the method of distraction osteosynthesis in pediatric patients with various diagnoses, such as Madelung's deformity, multiple exostosis chondrodysplasia, lesion to the epiphyseal zone of the forearm bones, congenital radial, and ulnar clubhand [17].

The results of ulnar bone elongation in patients with congenital radial clubhand in the global literature were only obtained for small groups of patients (up to 4–15 patients), whose age ranged from 1 to 17 years [19–23]. Approximately, the same data are presented for the fixation index, osteosynthesis, and reported complications [19, 22, 24]. However, the authors performed osteotomy at different levels, some in the proximal third [17, 24], others in the middle third [20], or distal third [19, 22, 25] of the ulnar bone.

The lack of consensus and approaches to the treatment of pediatric patients with these types of radial clubhand necessitated a retrospective assessment of the results of elongation of the ulnar bone by distraction osteosynthesis based on the level of osteotomy.

The purpose of this work is to evaluate the results of ulnar elongation by the method of distraction osteosynthesis in patients with type III–IV congenital radial clubhand (according to the Bayne and Klug classification) depending on the level of osteotomy.

Materials and methods

During the period 1998-2018, 285 pediatric patients with congenital radial clubhand were examined and received treatment at the Department of Reconstructive Microsurgery and Hand Surgery of the Turner Scientific Research Institute for Children's Orthopedics. Ulnar bone elongation by distraction osteosynthesis was performed in 36 patients (40 forearms) with type III-IV congenital radial clubhand (Bayne and Klug classification), including 23 boys and 13 girls. The average age of the pediatric patients was 7.4 ± 3.5 years (3-15 years). Type III congenital radial clubhand was reported in three patients. Unilateral lesion was noted in 91.7% of cases, whereas bilateral lesion was observed in 8.3% of cases. In 15 patients, congenital radial clubhand was a part of the genetic syndrome. The Holt-Oram syndrome was reported in 10 patients, TAR syndrome was noted in three patients, and VATER syndrome was observed in two patients. Two-stage ulnar bone elongation on the affected side was performed in three patients, whereas it was performed twice on both limbs in two patients with TAR syndrome.

Before the elongation of the ulnar bone, the hand was centered on the ulnar bone in all the patients. In addition, a number of patients underwent reconstructive surgeries of various types on ray I of the hand on the affected side.

All patients were divided into three groups. Patients of group 1 underwent osteotomy in the proximal third of the ulnar bone, patients of group 2 underwent osteotomy in the middle third of the forearm at the apex of deformity, and patients in group 3 underwent osteotomy in the lower third of the ulnar bone. The follow-up period in the study groups ranged from 1 to 12 years (the average follow-up period was 5.8 ± 0.4 years).

During the planning surgical intervention, we used clinical and radiological research methods, including computed tomography.

During clinical examination, the indicators, namely the range of movement in the elbow joint and fingers, were evaluated in both the preoperative and long-term postoperative periods. We did not consider the range of movements in the elbow-carpal joint, since it was minimal in all the patients due to the previously performed centering of the hand.

Radiographic images of the forearm bones along with the elbow joint and hand were captured in two standard projections. Additionally, radiographic images of the forearms evaluated the length of the ulnar bone on the affected and intact limbs; and the percentage of shortening of the ulnar bone in relation to the healthy one was calculated. Furthermore, the angle of deformity of the ulnar bone and the angle of the hand deviation were also calculated.

In the process of preoperative planning to choose the level and type of osteotomy, factors such as the apex of the ulnar bone deformity and the angle of the hand deviation were considered.

- If the apex of deformity was in the proximal part of the ulnar bone, and the angle of deviation of the hand did not exceed 20°, then the osteotomy was performed at the level of the upper third of the ulnar bone.
- If the angle of deformity of the ulnar bone exceeded 20° and was located in the middle third of the diaphysis, then osteotomy was performed in the middle third.
- If ulnar bone deformity was present in the distal section and the hand deviation was more than 20°, then osteotomy was performed in the lower third of the forearm.

The method of osteotomy depended on the type of hand deviation. For example, Minervini osteotomy was performed in the case of radial deviation of the hand (in the sagittal plane) [26], whereas cup-and-ball osteotomy was performed if it was palmar-radial (in the sagittal and frontal planes) deviation.

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A total of 40 osteotomies were performed, 3 of which were cup-and-ball osteotomy and 2 were Minervini osteotomy. In other cases, oblique osteotomy in the sagittal plane was preferred.

To evaluate the results of ulnar bone elongation, the indicators were studied, namely distraction time, correction period, fixation and osteosynthesis index, and regenerate length. In addition, postoperative complications were analyzed according to J. Caton classification [27].

We did not evaluate the functional capabilities, as well as the cosmetic condition of the forearm, before and after the elongation of the ulnar bone.

Elbow elongation by using compressiondistraction osteosynthesis was performed according to the standard technique. Wires or rods were placed in the proximal and distal sections of the ulnar bone, followed by their fixation in two transosseous supports, and one wire was inserted through the metacarpal bones II–V with the hand fixed to the outer supports. Then, an osteotomy of the ulnar bone was performed.

Distraction began on the fifth or seventh day after surgery by 0.25 mm, for three times a day. During the entire period of osteosynthesis, the patients received conservative treatment (physiotherapeutic treatment, exercise therapy, and massage). After the maturation of the distraction regenerate, the compression-distraction apparatus was dismantled and the upper limb was immobilized with a plaster cast or cylinder plaster cast.

The results of a comprehensive examination and treatment of patients were subjected to statistical processing in the Statistica 7.0 for Windows system by using the methods of parametric and nonparametric statistics. Additionally, we determined

Table 1

Assessment of the res	ults of surgical ti	treatment of patients	with type III–IV	' congenital radial clubhand

Tra Jarra	Group 1			Group 2			Group 3		6
Index	before	after	P	before	after	P	before	after	Р
Clinical examination									
Flexion in the elbow joint, deg.	131.3 ± 15.9	129.4 ± 5.3	≥0.05	126.7 ± 17.8	128.3 ± 19.7	≥0.05	133 ± 16.4	133.6 ± 16.7	≥0.05
Extension in the elbow joint, deg.	12.5 ± 8.2	13.1 ± 9.3	≥0.05	8.3 ± 4.1	7.5 ± 2.7	≥0.05	8 ± 3.8	7.8 ± 3.9	≥0.05
Angle of the hand deviation, deg.	13.7 ± 5.8	11.2 ± 9.1	≥0.05	11.8 ± 7.2	12.8 ± 8.5	≥0.05	17.9 ± 10.9	9.3 ± 5.8	≥0.05
	X-ray examination								
Shortening of the ulnar bone in relation to healthy bone, %	35.6 ± 13.9	15.9 ± 10.7	≤0.05	34.3 ± 18.2	17.5 ± 12.4	≤0.05	31.1 ± 10.6	15.6 ± 10.4	≤0.05
The angle of the ulnar bone deformity, deg.	15.1 ± 11.8	3.5 ± 0.9	≤0.05	22.5 ± 16.0	1.8 ± 1.2	≤0.05	24.6 ± 16	1.6 ± 1.0	≤0.05
Postoperative period									
Correction period, days	46.7 ± 11.6			35.3 ± 14.0			33.2 ± 16.2		≤0.05
Resulting elongation, mm	37.6 ± 10.2			24 ± 9.7			28.8 ± 8.6		≤0.05
Resulting elongation as a percentage of the initial size of the ulnar bone	38.7 ± 12.1			35.4 ± 14.7			33.7 ± 13.8		≤0.05
Fixation index, days/cm	22 ± 9.9			47.4 ± 21.5			40.5 ± 21.2		≤0.05
Osteosynthesis index, days/cm	35.4 ± 10.5			75.5 ± 45.6			54.3 ± 20.8		≤0.05

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the mean values and standard deviations, performed the correlation analysis, and calculated the correlation coefficient.

Results

Table 1 presents the results of the study.

The range of movement in the elbow joint in patients of all groups in both the preoperative and postoperative periods did not showed a significant difference.

In patients with congenital radial clubhand, flexion-extension contractures of the elbow joint were noted, in which the average value of flexion was $131.5 \pm 16.1^{\circ}$, and its extension was $9.9 \pm 6.2^{\circ}$.

The average angle of hand deviation in different groups in the pre- and postoperative periods did not showed any statistically difference. However, before surgical treatment in the patients of groups 1 and 2, it ranged from 2 to 20°, whereas it was from 10 to 40° in the patients of group 3. In 5 patients of group 3, we simultaneously performed the correction of radial deviation of the hand, the average angle before surgery was $31 \pm 11.4^\circ$, whereas the average angle was $5.7 \pm 2.4^\circ$ in the postoperative period.

The shortening of the ulnar bone in relation to the intact limb before surgical treatment had an average value of $33.3 \pm 12.3\%$, whereas it was $16 \pm 10.5\%$ after the surgical treatment. The ulnar bone was elongated by an average of 36% as compared to its initial length.

In patients with type III–IV congenital radial clubhand, the angle of the ulnar bone deformity had an average value of $20.5 \pm 14.8^{\circ}$ (minimum 5°, maximum 40°) in the preoperative period. Significant deformity (35–40°) of the ulnar bone was reported in one patient of group 1, two patients of the group 2 with type IV of radial clubhand, and also in one patient of group 2 with type III radial clubhand. These patients underwent surgical treatment at the age of 6-7 years due to concomitant pathology, the decompensation of which was a contraindication for the surgical treatment of the orthopedic pathology. The severe secondary deformity of the ulnar bone was due to the long-term tension of the underdeveloped soft tissues and muscles along the radial surface of the forearm, along with the absence of conservative and surgical treatment. After the first stage of treatment (centering of the hand) during the child's growth, the angle of the ulnar bone deformity remained within 35-40°. In these patients, osteotomy was performed in the middle third of the ulnar bone at the deformity apex. In young patients, during their growth, we registered a decrease in the ulnar bone deformity in its middle third after the hand centering, which enabled us to perform subsequent osteotomy in the proximal or distal sections.

The angle of the ulnar bone deformity after treatment decreased, but persisted and had an average value of $7.4 \pm 5.6^{\circ}$. Moreover, the angle had an average corrected value of 63.9%.

In patients of group 1, the correction period was 24.4 and 28.9% longer than in patients of groups 2 and 3, respectively. This was due to the fact that in this group the resulting elongation rates were 32 and 18.4% greater than in patients of the groups 2 and 3, respectively.

The fixation indices in patients of group 1 were 53.6 and 45.7% lower than in the groups 2 and 3, respectively.

Table 2 presents the main complications reported during the elongation of the ulnar bone in patients with congenital radial clubhand.

Table 2

Group 1	Group 2	Group 3
2 (12.5%)	0 (0%)	2 (11.1%)
1 (6.3%)	1 (16.7%)	0 (0%)
0 (0%)	0 (0%)	1 (5.6%)
3 (18.7%)	3 (50%)	0 (0%)
0 (0%)	0 (0%)	1 (5.6%)
2 (12.5%)	0 (0%)	0 (0%)
1 (6.3%)	0 (0%)	0 (0%)
9 (56.3%)	4 (56.7%)	4 (22.3%)
	2 (12.5%) 1 (6.3%) 0 (0%) 3 (18.7%) 0 (0%) 2 (12.5%) 1 (6.3%)	2 (12.5%) 0 (0%) 1 (6.3%) 1 (16.7%) 0 (0%) 0 (0%) 3 (18.7%) 3 (50%) 0 (0%) 0 (0%) 2 (12.5%) 0 (0%) 1 (6.3%) 0 (0%)

Complications in elongation of the ulnar bone in patients with type III-IV congenital radial clubhand

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With the elongation of the ulnar bone, complications were observed in all groups of patients. Most often, complications were revealed in group 1 (56.3%) and group 2 (56.7%).

Complications of degree I (according to the Caton classification) were identified in all the groups. Soft tissue inflammation at the exit site of transosseous elements was registered in patients of groups 1 and 2 in 12.5 and 11.1% of cases, respectively. To stop the inflammatory process, oral antibiotic therapy and dressings with topical antiseptics were employed. There were no signs of deep infection in any other cases. In two cases, the patients had a fracture of the wire or the rod screw; however, this complication did not affect the final result of the treatment. One patient of group 3 showed increased contracture of the fingers during distraction. It was eliminated after a course of rehabilitation treatment, and a satisfactory (preoperative) range of motion in the fetlock joints and phalangeal joints of the hand was achieved.

Complications of degree II were most common and reported in all the groups. The greatest number of complications in the form of pseudarthrosis at the level of the distraction regenerate maturation was recorded in patients of groups 1 and 2 in 18.7 and 50% of cases, respectively. In addition, a regenerate fracture and the formation of pseudarthrosis after dismantling the compression-distraction apparatus were recorded in 5.6% of cases in patients of group 3. In all the cases, the defect grafting was performed by using a free cortical-spongious bone graft taken from the iliac bone to restore the integrity of the ulnar bone.

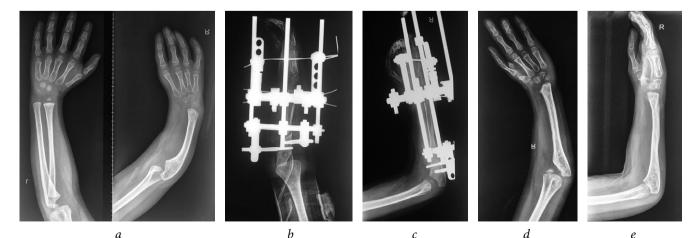
Complications of degree III were noted only in patients of group 1 in 18.8% of cases (three patients). In two cases, a dislocation of the proximal ulnar bone posteriorly with the formation of a "neojoint" was reported. These patients subsequently underwent the shortening osteotomy of the tip of the elbow with the transposition of triceps. In addition, a relapse of hand deviation occurred in one patient.

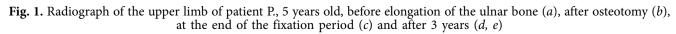
A clinical example of surgical treatment of the group 1 patient (Fig. 1)

Patient P., 5 years old, was admitted to the department with a diagnosis of congenital rightsided radial clubhand, along with aplasia of finger I. Previous surgery included the right-hand centering.

Clinical examination revealed shortening of the right forearm by 4 cm, aplasia of the radial bone, and radial deviation of the right hand of 15°. Moreover, the hand was impossible to bring to the middle position passively, and aplasia of the finger I of the right hand was reported (Fig. 1, *a*). Given the slight deviation of the hand and the apex of the ulnar deformity in its proximal part, we decided to perform osteotomy in the upper third of arm. The postoperative period was uneventful. The ulnar bone was extended by 3.7 cm, and the correction of the ulnar bone deformity was 25°. Subsequently, pollicization of the ray II of the hand was performed on the affected side.

After 3 years, the radial deviation of the hand was found to be 20° during the clinical examination, which was the state of hand after pollicization of finger II of the right hand. Ranges of movements in the elbow joint and fingers were found to be adequate. The child served himself and did not complain actively. Parents were satisfied with the result.





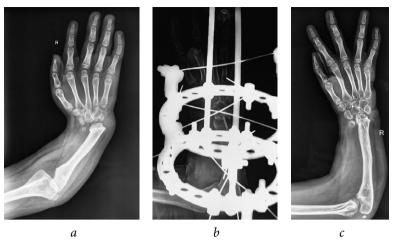


Fig. 2. Radiograph of the upper limb of patient L., 6 years old, before elongation of the ulnar bone (*a*), after osteotomy (*b*) and after 2 years (*c*)

A clinical example of surgical treatment of the group 3 patient (Fig. 2)

Patient L., 6 years old, was admitted to the department with a diagnosis of TAR syndrome, congenital bilateral radial clubhand, and hypoplasia of fingers I. The previous surgery included centering of both right and left hands. Clinical examination revealed shortening of both forearms, aplasia of the radial bones, palmar-radial deviation of the right hand by 40°, the hand was impossible to bring to the middle position passively, and hypoplasia of the fingers I was also reported (Fig. 2, a). Given the marked deviation of the hand, we decided to perform cup-and-ball osteotomy of the right ulnar bone in the lower third. The postoperative period was uneventful. The ulnar bone was elongated by 3.5 cm. The palmar-radial deviation of the hand was corrected up to 35°.

A clinical examination after two years (Fig. 2, c) revealed that radial deviation of the hand was up to 10°. The child served herself and did not complain actively. Her parents were satisfied with the result. Subsequently, the ulnar bone was elongated to the left.

Discussion

The surgical treatment of patients with congenital radial clubhand remains a complex challenge in pediatric orthopedics. As a result of surgical treatment, there is an improvement in appearance, the functional capabilities of the upper limb, and the ability to self-service. Patients begin to use this limb more often in daily life activities. These factors affect the patient's social adaptation and quality of life [19, 22, 28]. Elongation is performed preferably at the beginning of adolescence, since treatment does not interfere with the psychosocial development. In our study, the average age of patients was 7.4 ± 3.5 years, which corresponds to the data of global literature [19–23].

The limitation of the range of movements in the elbow joint is quite common in patients with congenital radial clubhand. In the presence of severe contracture in the elbow joint, the ulnar bone cannot be extended [29]. On an average, the range of motion in the elbow joint is 99° [30]. Patients in our study also revealed contracture of the elbow joint, and the range of motion had an average value of 120°.

According to some authors, the ulnar bone can be extended by 4–6 cm [17, 21, 22, 25]. In our patients, the average value of elongation was 3.2 ± 1.1 cm. Complete restoration of the forearm length is not considered to be an obligatory condition [26]. The fixation and osteosynthesis indices obtained in the study in patients of groups 1 and 3 were comparable to the literature data [17, 20, 25, 31]. A high osteosynthesis index in patients of group 2 (75.5 ± 45.6 days/cm) was associated with complications (pseudarthrosis of the ulnar bone) and attempts of conservative treatment for the formation of a regenerate.

The literature describes a considerable number of complications while using the Ilizarov method to lengthen the bones of the upper limb in up to 100% of cases. The most common complications are inflammatory processes, contractures in the joints, and fracture of the regenerate [13, 19, 21, 22, 25]. However, in our study, soft tissue inflammation at

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the exit site of transosseous elements was registered in only 10% of cases, and joint contracture and regenerate fracture were reported in 2.5% of cases.

The most common complication recorded in patients of our clinic during elongation of the ulnar bone is the formation of an atrophic regenerate with subsequent pseudarthrosis (15% of cases). A small number of studies focused on the risk factors for delayed consolidation with the elongation of the ulnar bone. Thus, the place of an osteotomy, a fast rate of distraction, a significant elongation, inflammation, as well as patient nutrition can affect the formation of atrophic regenerate [32-35]. Severe cicatricial changes, angiotrophic disorders due to malformation of the limb, damage to the endosteum, and disorder of the intraosseous circulation due to repeated surgical interventions negatively affect the trophism of the limb and, accordingly, the process of osteogenesis [36, 37].

According to Catagni [20], the formation of atrophic regenerate occurs in 20% of cases while performing an osteotomy in the middle third of the ulnar bone. However, the author notes that prolonged immobilization in an external fixation apparatus enables us to solve this problem. In our cases, this complication was recorded in patients who underwent osteotomy of the ulnar bone either in the middle third of the diaphysis (50% of cases of pseudarthrosis formation) or on the border of the upper and middle third. Circulatory failure in the middle third of the forearm due to pronounced underdevelopment of the forearm, significant discrepancy in bone fragments due to the elimination of ulnar bone deformity, and the "standard" rate of distraction resulted in the formation of an atrophic regenerate. An increase in fixation period in the compression-distraction apparatus did not produce results and required additional surgical intervention. Thus, osteotomy in the proximal or distal ulnar bone or reduction of the rate of distraction will reduce the risk of pseudarthrosis.

In addition, in 7.5% of cases, we registered complications that were not described in the literature in the treatment of patients with congenital radial clubhand. Thus, the dislocation of the elbow joint occurred in 5% of cases. This is due to the fact that patients with congenital radial clubhand have underdeveloped proximal ulnar bone (smoothness and insufficient pronouncement of the coronoid process), as well as a disorder of stability and congruency in the elbow joint, which led to the dislocation of the proximal ulnar bone posteriorly during elongation. This complication can be avoided by fixing the humerus during the period of distraction (in case of osteotomy in the proximal ulnar bone) and dismantling the fixing support during the stabilization period. In one patient (2.5%), there was a relapse of hand deviation, which was caused by insufficient stability of the hand in the compression-distraction apparatus, where the hand was fixed only with a rod installed in the metacarpal bone V. In this procedure, the hand must be fixed with a wire drawn through the four metacarpal bones.

Conclusion

When the ulnar bone is elongated in patients with type III and IV congenital radial clubhand according to the Bayne and Klug classification, the proximal bone is the zone of choice for osteotomy.

When the deviation of the hand is more than 20°, osteotomy in the distal ulnar bone with the simultaneous correction of deformity is recommended.

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Ethical consideration. The study was performed in accordance with the ethical standards of the Helsinki Declaration of the World Medical Association as amended by the Ministry of Health of Russia, approved by the Ethics Committee of the Turner Scientific Research Institute for Children's Orthopedics (protocol No. 2017/6 of 28.11.2017).

The authors received the written voluntary consent of patients (or their legal representatives) to participate in the study and publish medical data.

Contribution of authors

N.V. Avdeychik developed the examination methodology, wrote all sections of the article, collected literature data, and performed their processing. In addition, he operated on 13 patients.

S.I. Golyana exercised the leadership and participation in the development of the study

methodology and performed stepwise editing of the article text. In addition, he operated on 10 patients.

D. Yu. Grankin was involved in data processing, writing the summary, list of references, and edited the article.

A.V. Safonov took part in the study development and operated on 17 patients.

All authors made a significant contribution in the research and preparation of the article. Additionally, all the authors read and approved the final version before publication.

References

- 1. Goldberg MJ, Meyn M. The radial clubhand. Orthop Clin North Am. 1976;7(2):341-359.
- Tonkin MA. Radial longitudinal deficiency (radial dysplasia, radial clubhand). In: Green's Operative Hand Surgery. 4th ed. Ed. by D.P. Green, R.N. Hotchkiss, W.C. Pederson. Philadelphia: Churchill Livingstone; 1999. P. 345-358.
- D'Arcangelo M, Gupta A, Scheker LR. Radial club hand. In: The Growing Hand: Diagnosis and Management of the Upper Extremity in Children. Ed. by A. Gupta, S.P.J. Kay, L.R. Scheker. London: Harcourt Publishers Ltd; 2000. P. 147-170.
- Kozin SH. Upper-extremity congenital anomalies. J Bone Joint Surg Am. 2003;85(8):1564-1576. https:// doi.org/10.2106/00004623-200308000-00021.
- Ekblom AG, Laurell T, Arner M. Epidemiology of congenital upper limb anomalies in 562 children born in 1997 to 2007: a total population study from stockholm, sweden. *J Hand Surg Am.* 2010;35(11):1742-1754. https://doi.org/10.1016/j.jhsa.2010.07.007.
- Авдейчик Н.В., Говоров А.В., Голяна С.И., Сафонов А.В. Врожденная лучевая косорукость у детей в структуре генетических синдромов // Ортопедия, травматология и восстановительная хирургия детского возраста. 2015. Т. 3. № 4. С. 29–36. [Avdeychik NV, Govorov AV, Golyana SI, Safonov AV. Occurrence of radial club hand in children with different syndromes. *Pediatric Traumatology, Orthopaedics and Reconstructive Surgery*. 2015;3(4):29-36. [In Russ.)]. https://doi.org/10.17816/PTORS3429-36.
- Гребенюк Е.Б., Попков Д.А. Интрамедуллярное армирование при удлинении предплечья у больных с врожденными аномалиями развития // Гений ортопедии. – 2006. – № 3. – С. 104–109. [Grebenyuk EB, Popkov DA. Intramedullary reinforcement for forearm lengthening in patients with congenital developmental anomalies. *Genij ortopedii*. 2006;(3):104-109. (In Russ)]
- Bayne LG, Klug MS. Long-term review of the surgical treatment of radial deficiencies. J Hand Surg Am. 1987;12(2):169-179. https://doi.org/10.1016/s0363-5023(87)80267-8.
- Vergara-Amador E, Lopez Rincon L, Herrera Rodriguez S. Radial longitudinal deficiency. Analysis of clinical and radiological results. *Rev Esp Cir Ortop Traumatol.* 2019;63(3):217-226. https://doi.org/10.1016/j. recot.2018.10.004.

- 10. Paley D. The Paley ulnarization of the carpus with ulnar shortening osteotomy for treatment of radial club hand. *SICOT J.* 2017;3:5. https://doi.org/10.1051/sicotj/2016040.
- Takagi T, Seki A, Takayama S, Watanabe M. Current concepts in radial club hand. Open Orthop J. 2017;11:369-377. https://doi.org/10.2174/18743250017 11010369.
- Najd Mazhar F, Shariatzadeh H, Balvardi M, et al. Recurrence rate of radial deviation following the centralization surgery of radial club hand. *Med J Islam Repub Iran.* 2018;32:18. https://doi.org/10.14196/mjiri.32.18.
- 13. Yoshida K, Kawabata H, Wada M. Growth of the ulna after repeated bone lengthening in radial longitudinal deficiency. *J Pediatr Orthop*. 2011;31(6):674-678. https://doi.org/10.1097/BPO.0b013e318221ebe7.
- Vuillermin C, Butler L, Ezaki M, Oishi S. Ulna growth patterns after soft tissue release with bilobed flap in radial longitudinal deficiency. *J Pediatr Orthop.* 2018;38(4):244-248. https://doi.org/10.1097/ BPO.0000000000000807.
- Peterson BM, McCarroll HR, Jr., James MA. Distraction lengthening of the ulna in children with radial longitudinal deficiency. *J Hand Surg Am*. 2007;32(9):1402-1407. https://doi.org/10.1016/j.jhsa.2007.07.024.
- Бородин В.В. Лечение по Илизарову приобретенной лучевой косорукости: Автореф. дис... канд. мед. наук. – Курган, 1992. [Borodin VV. Lechenie po Ilizarovu priobretennoy luchevoy kosorukosti. Kurgan; 1992. (In Russ.)]
- Стариков О.В. Реконструктивно-восстановительное лечение врожденной косорукости по методу Г.А. Илизарова: Автореф. дис. ... канд. мед. наук. – Курган, 2002. [Starikov OV. Rekonstruktivnovosstanoviteľnoe lechenie vrozhdennoy kosorukosti po metodu G.A. Ilizarova. Kurgan; 2002. (In Russ.)]
- 18. Хмызов С.А., Гарбузняк И.Н. Эффективность комбинированного лечения детей с врожденной косорукостью в сочетании с аномалиями кисти // Травма. – 2014. – Т. 15. – № 4. – С. 95–100. [Khmyzov SA, Garbuznyak IN. The effectiveness of combined treatment of children with congenital clubhand combined with hand anomalies. *Travma*. 2014;15(4):95-100. (In Russ.)]
- Hill RA, Ibrahim T, Mann HA, Siapkara A. Forearm lengthening by distraction osteogenesis in children: a report of 22 cases. J Bone Joint Surg Br. 2011;93(11):1550-1555. https://doi.org/10.1302/0301-620X.93B11.27538.
- 20. Catagni MA, Szabo RM, Cattaneo R. Preliminary experience with Ilizarov method in late reconstruction of radial hemimelia. *J Hand Surg Am*. 1993;18(2):316-321. https://doi.org/10.1016/0363-5023(93)90367-C.
- Kawabata H, Shibata T, Masatomi T, Yasui N. Residual deformity in congenital radial club hands after previous centralisation of the wrist. Ulnar lengthening and correction by the Ilizarov method. *J Bone Joint Surg Br*. 1998;80(5):762-765. https://doi.org/10.1302/0301-620x.80b5.8839.
- 22. Pickford MA, Scheker LR. Distraction lengthening of the ulna in radial club hand using the Ilizarov technique. *J Hand Surg Br.* 1998;23(2):186-191. https://doi.org/10.1016/s0266-7681(98)80172-1.
- 23. Horii E, Nakamura R, Nakao E, et al. Distraction lengthening of the forearm for congenital and develop-

mental problems. J Hand Surg Br. 2000;25(1):15-21. https://doi.org/10.1054/jhsb.1999.0309.

- Бойчев Б., Комфорти В., Чоканов К. Оперативная ортопедия и травматология – учебник. София: Медицина и физкультура, 1961. 794 с. [Воусhev В, Котforti V, Chokanov K. Operativnaya ortopediya i travmatologiya – uchebnik. Sofiya: Meditsina i fizkul'tura; 1961. 794 p. (In Russ.)]
- 25. Caton J. L'allongement bilatéral des membres inférieurs chez les sujets de petite taille en France. Résultats de l'enquête GEOP; notre expérience: Traitement des inegalites de longueur des membres inferieurs et des sujets de petite taille chez l'enfant et l'adolescent: Symposium sous la direction de J. Caton (Lyon). *Rev Chir Orthop.* 1991;77(1):74-77.
- Raimondo RA, Skaggs DL, Rosenwasser MP, Dick HM. Lengthening of pediatric forearm deformities using the Ilizarov technique: functional and cosmetic results. *J Hand Surg Am.* 1999;24(2):331-338. https://doi. org/10.1053/jhsu.1999.0331.
- 27. Bhat SB, Kamath AF, Sehgal K, et al. Multi-axial correction system in the treatment of radial club hand. *J Child Orthop.* 2009;3(6):493-498. https://doi. org/10.1007/s11832-009-0196-3.
- Azar FM, Canale T, Beaty OP. Campbell's Operative Orthopaedics. Vol. 4. 11th ed. Mosby; 2007.
- Saini N, Patni P, Gupta S, et al. Management of radial clubhand with gradual distraction followed by centralization. *Indian J Orthop.* 2009;43(3):292-300. https:// doi.org/10.4103/0019-5413.53461.
- Masada K, Tsuyuguchi Y, Kawai H, et al. Operations for forearm deformity caused by multiple osteochondromas. *J Bone Joint Surg Br.* 1989;71(1):24-29. https:// doi.org/10.1302/0301-620X.71B1.2914999.

- 31. Farr S, Mindler G, Ganger R, Girsch W. Bone lengthening in the pediatric upper extremity. *J Bone Joint Surg Am.* 2016;98(17):1490-1503. https://doi. org/10.2106/JBJS.16.00007.
- 32. Tjernstrom B, Olerud S, Rehnberg L. Limb lengthening by callus distraction. Complications in 53 cases operated 1980-1991. *Acta Orthop Scand*. 1994;65(4):447-455. https://doi.org/10.3109/17453679408995491.
- Mader K, Gausepohl T, Pennig D. Shortening and deformity of radius and ulna in children: correction of axis and length by callus distraction. *J Pediatr Orthop B*. 2003;12(3):183-191. https://doi.org/10.1097/01. bpb.0000057485.91570.e9.
- 34. Paley D. Problems, obstacles, and complications of limb lengthening by the Ilizarov technique. *Clin Orthop Relat Res.* 1990;(250):81-104.
- 35. Li Y, Han B, Tang J, et al. Identification of risk factors affecting bone formation in gradual ulnar lengthening in children with hereditary multiple exostoses: A retrospective study. *Medicine (Baltimore)*. 2019;98(5):e14280. https://doi.org/10.1097/MD.000000000014280.
- 36. Борзунов Д.Ю., Шастова А.Л. «Ишемический» дистракционный регенерат: толкование, определение, проблемы, варианты решения // Травматология и ортопедия России. – 2019. – Т. 25. – № 1. – С. 68–76. [Воггипоv DY, Shastova AL. "Ischemic" distraction regenerate: interpretation, definition, problems and solutions. *Traumatology and Orthopedics* of Russia. 2019;25(1):68-76. (In Russ.)]. https://doi. org/10.21823/2311-2905-2019-25-1-68-76.
- 37. Kojimoto H, Yasui N, Goto T, et al. Bone lengthening in rabbits by callus distraction. The role of periosteum and endosteum. *J Bone Joint Surg Br.* 1988;70(4):543-549. https://doi.org/10.1302/0301-620X.70B4.3403595.

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