Malformations of the first ray of the foot widely vary and are divided into simple and complex types. Complex malformations include abnormalities in the development of the first metatarsal or the main phalanx and refer to atypical adducted foot deformities; they are also characterized by varus location of the first ray of varying severity. The cause of deformation is damage to the longitudinal epiphyseal growth plate area of the first metatarsal, termed as longitudinal epiphyseal bracket or delta phalanx.

Over the past 5 years, we treated 37 patients (53 feet) aged 6 months to 17 years who had developmental disabilities of the first ray of the foot. The spectrum of pathologies greatly varied. Surgical treatment was often multistage. We found that complete removal of the deformity at the first stage of treatment should be performed with maximum use of the patient’s bones to restore the length and shape of the affected bone. In treating combined deformities, good results can be guaranteed only by the removal of all elements, including the excision of the fibrous bridle along the inner surface of the first ray.

Keywords: preaxial polydactyly of the foot, malformations of the first ray of the foot, congenital deformities of the great toe, longitudinal epiphyseal bracket (LEB), delta phalanx.

Malformations of the first ray of the foot comprise as much as 0.1% of all congenital disorders of the musculoskeletal system and 2%–14% of malformations of the foot. There are malformations of the entire first ray, isolated malformations of the first metatarsal bone, and toe malformations. Often, there is a combination of doubling of the first toe or ray and syndactyly of the first and second toes.

Congenital malformation of the longitudinal epiphyseal growth plate area of the first metatarsal, LEB or so-called delta phalanx, is an anomaly of the epiphyseal and diaphyseal growth of long bones [1]. The shortened first metatarsal bone acquires a trapezoidal shape. Morphology of the diaphysis is unusual and exhibits an uneven distribution chondrocytes and chondroblasts with moderate formation of rouleaux in the pathological growth zone, which is located on the medial side of the bone’s diaphysis. Delta phalanx was first described in 1921 by Pol et al.; however, the term was first used in 1964 by Jones [initials?].

The presence of several deltoid bones was described in 1999 in patients with Rubinstein-Taybi syndrome [1, 2, 3]. The literature mentions other names for this condition, diaphyseal or epiphyseal brackets, “kissing” delta phalanx, brachyphalangy, and trapezoidal phalanx. Clinodactyly at the interphalangeal joint is due to the presence of the delta phalanx and is often combined with symphalangism, brachydactyly, and split foot malformation (ectrodactyly) [5, 6]. Most often, LEB occurs as a bilateral deformation, and in some cases, it is inherited as an autosomal dominant trait. Ultrasound imaging is used to confirm the diagnosis [7]. Development defects of the first ray of the foot are among the most difficult problems associated with structural deformations of the forefoot.

These deformities affect the shape of the forefoot, impact the foot’s support function, and cause pain as well as problems in selecting and wearing shoes. The source of pain is associated with an abnormal load in the area of the metatarsal
bone heads during the metatarsophalangeal rolling phase due to a defective metatarsal parabola. The shortened, deformed first metatarsal bone is an isolated developmental defect that when present with first-ray polydactyly, adversely affects the supporting function of the foot, leading to an overload of the epiphysis of the second and third metatarsal bones that bear most of the load. This condition leads to deformations and causes “corns” and scrapes on the plantar surface of the foot.

Doubling of the first ray of the foot is a complex congenital malformation, which, in addition to increasing the number of toes and metatarsal bones, is characterized by osteoarticular deformation with uneven growth of the forefoot bones that worsens with age [8]. The deformation is bilateral in 25%–50% of cases [9]. The possible causes of polydactyly or doubling of the first ray are teratogenic effects between the 5th and 8th weeks of pregnancy [10]. Certain fibroblast growth factors activate the proliferation of mesodermal cells, leading to the formation of an extra toe [11]; however, the cellular and genetic mechanisms that control the number of toes are not fully understood [12]. Over the years, various classifications of polydactyly were proposed, mainly based on the morphology and anatomy of the defect of the forefoot. However, a classification of malformations of the first ray does not exist.

Researchers worldwide agree that early surgery is the most appropriate treatment because the structure of the foot of a young child has high plastic capabilities with potential for remodeling. Moreover, restoration of blood flow and function of the neuromuscular system occur during a short time and are accompanied by optimal functional outcomes [9, 10, 13, 14]. Surgical correction of the deformed first metatarsal bone is accomplished by excision of the pathological growth plate on the medial surface of epimetaphysis [15]. This allows lengthy (years) observations of the proportional growth of metatarsal or phalanx as well as of the correct axis of the first ray. However, according to the literature, in the 6–7 years before closing of the growing zone plate, relapse deformation develops up to 10°–15°, which requires a second surgical correction.

According to I.A. Gankin, age indications for surgery depend on the type of malformation and associated deformities. For polyphalangy of the first toe, surgery is recommended at 6 months of age [8]. For polydactyly of the first ray or toe, polymetatarsia, and in cases when the use of a free skin autograft is required, surgery is recommended after 10 months of age, and after 18 months of age, reconstruction of the foot of children with hypoplasia of the metatarsal bones is recommended. For resection of a trapezoidal metatarsal bone with modeling, and if osteosynthesis of the adjacent metatarsal bones is required, autograft surgery should be delayed until the age of 3 years. In any case, surgery must be performed no later than 5 years. Subsequent correction of angle deformations and instability can be performed when children are 8–10 years. In the case of partial or complete doubling of segments, extra toes must be removed using an elliptical incision before children reach 1 year of age [10].

Surgical correction of preaxial polydactyly is a difficult task because of the small number of good results and the large number of complications [16]. Most failures are associated with persistent varus deformity of the first toe [14]. Removing an additional rudimentary toe is uncomplicated. In the event of adequate development of both toes, surgery presents a number of difficulties—removal of the medial toe that leads to development of valgus flat-foot deformity and if a lateral toe is removed—to the development of varus deformity of the first toe.

Aim

The objective of this study was to develop a strategy for surgical treatment of malformations of the first ray of the foot in children that takes into account the type of deformation and age of the patient.

Materials and methods:

We examined and treated 37 patients (53 feet, 22 girls and 15 boys, 6 months–17 years of age) with malformations of the first ray. The patients were divided into groups, depending on the type and severity of the deformation that were determined by clinical and radiological examinations. In the present study, “longitudinal epiphyseal bracket (LEB)” of the first metatarsal bone is referred to as the “delta metatarsal bone.”
The most frequently observed deformations of the phalanges of the first toe (8 children, 8 feet) were as follows: doubling of the first toe (7 children, 12 feet); polysyndactyly of the first ray with delta metatarsal bone and delta phalanx of the main or an additional toe (17 children, 23 foot), including secondary deformations of metatarsal bones in adolescents (2 children, 4 feet). We rarely observed isolated defects of the first metatarsal bone (deformation of the growth plate observed in 2 children, 4 feet) and first- and second-ray syndactyly with clinodactyly of the phalanges (3 children, 6 feet). Structural pathology prevailed in girls, and isolated, cutaneous syndactyly of the first and second toes was present only in boys. In the present study, we used anamnestic, clinical, and radiographic methods, including X-rays and spiral multislice computed tomography (MSCT).

To assess the shape of the metatarsal parabola, condition of the growth plates and anatomy of the diaphysis radiography was performed using plantar, lateral to the load (or simulated load in children younger than 2 years), and oblique projections. MSCT was performed when it was necessary to evaluate the three-dimensional image of the malformed segment, including epiphysis, with the goal of intraoperative modeling of its shape.

Discussion:

The indications for surgery to treat malformations of the first ray of the feet of children are as follows: increased width of the forefoot due to the presence of an extra toe or ray, deformation of the first-ray axis, incorrect proportions of the first metatarsophalangeal joint, reduced ability of the foot to support weight, cosmetic defects, significant problems in selecting and wearing shoes, and pain.

A topic of debate is the recommended age for the first phase of surgery, its sequence, and degree. It is important to understand that multiple surgeries are required. The number of steps depends on the type and degree of deformation of the phalanges and metatarsals and their changes in shape with the growth of the child. The main principle of surgical treatment in our clinic of malformations of the first ray of the foot is the maximum possible use of the child’s own tissues to eliminate deformation, restoration of normal anatomy, and correction of the length of the truncated segment.

It is not difficult to perform surgery to treat doubling of the first toe with a hypoplastic first ray in the presence of delta-medial metatarsal bone as well as treatment of doubling of the first ray in the normally developed metatarsal bone. Disarticulation of additional segments is performed by circumventing incision of the additional toe on the plantar surface and transitions to the dorsal surface in the metatarsal area. Plastic surgery is performed simultaneously with reconstruction of the nail bed. With these types of malformations, tendons are normally developed on the lateral main ray and do not require correction.

According to our observations, correction of deformation of the first ray in the presence of a delta phalanx or metatarsal bone is required at the age of 10–12 months. The surgeon excises the fibrous strand on the inner surface of the deformed segment and performs wedge resection of its medial section of the meta-epiphysis and osteotomy of the metaphysis. A triangular bone graft of fragments of the removed toe is inserted simultaneously in the osteotomy to correct the deformed first ray.

The graft was then secured with cross-screws for up to 8 weeks (Fig. 1).

External fixation is accomplished using a plaster cast while the foot is in a neutral position. Such tactics extend the first column and help to orient the longitudinal growth of the ray along the correct axis. Further extension of the first metatarsal bone is performed if it is shorter than 1 cm during the growth of the child, but becomes clear no earlier than 4–6 years after the first phase of treatment.

![Figure 1. One-stage reconstruction of the first ray (A, before surgery; B, removal of the extra first toe and wedge osteotomies of the first phalanx and metatarsal bone, C, insertion of the autograft into the osteotomy)](image-url)
As an example of the second phase of surgical treatment, we present our 6-year-old patient K. (Fig. 2). She underwent surgery at 18 months of age to treat polysyndactyly of the first ray combined with delta-phalanx and trapezoid metatarsal of the first additional ray. The child had polysyndactyly of the fifth ray as well.

During the first phase of treatment, in addition to the elimination of polysyndactyly, we performed wedge osteotomy of the proximal phalanx and first metatarsal bone of the main ray and introduced wedge-shaped grafts cut from the excised segments. Further, we removed the fifth ray and performed skin grafting.

Varus deformity of the first ray formed by 6 years of age, and it caused considerable inconvenience in selecting and wearing shoes (Fig. 2A). Deformation of the fifth metatarsal bone developed concurrently, causing pain under load. To correct the deformity, we harvested a bicortical autograft from the iliac wing. The graft was excised using a “trap-door” technique without damaging the apophysis. Its size was calculated according to the results of MSCT with the goal of dividing it into three segments. We performed corrective wedge osteotomy of the proximal phalanx and first metatarsal bone of the main ray and introduced wedge-shaped grafts cut from the excised segments. Further, we removed the fifth ray and performed skin grafting.

The extent of elongation of the main ray of the first metatarsal bone was calculated according to the metatarsal parabola of a healthy foot. In the case of bilateral deformation, the calculation was based on the shape of a metatarsal parabola of the patient's parent with the greatest phenotypic similarity. The transplant was inserted in place of the osteotomy of the main metatarsal bone in the proximal metaphyseal region. An axial spoke was inserted, fixing the interphalangeal and metatarsophalangeal joints in the central position. The implanted graft was fixed with two cross-spokes (0.8-mm diameter). The proximal segments of the flexor and extensor muscles of the extra toe were attached to the main toe's interossei muscles. The distal segments of the first toe tendons, transferred to the superposition, were sutured to the proximal

Figure 2. Radiographs of patient K (6 years of age) with relapse of varus deformity of the first and fifth rays (A, radiograph of the foot before phase-2 surgery; B, radiograph of the foot after wedge osteotomies of the proximal phalanx and metatarsal bone of the first ray and fifth metatarsal bone).
segment of the corresponding tendons of the main finger (Figure 3).

We recommend two-step elimination of polysyndactyly of the first ray coupled with finger syndactyly of the first and second toes. The first step forms the first column that is performed with essential excision of the fibrous strand along the inner surface of the foot, as previously described. Elimination of syndactyly between the formed first and second toes is recommended 4–6 months after the initial surgery. A completely restructured, fully shaped metatarsal bone should be present, and the patient should walk with a normal gait.

Soft tissue syndactyly of the first and second toes during growth of a child leads to flexion contracture of the toes, causing abrasions of the interphalangeal joints, which is an indication to eliminate this defect. The Cronin method is commonly accepted. However, distinct characteristics caused by removal of syndactyly of the foot are as follows: soft tissues should be preserved, as far as possible, on the dorsal surface of the interphalangeal joints, the intertarsal ligaments should not be dissected to avoid fan-shaped spread of the toes under load, and the development of secondary deformities should be prevented. In case of persistent flexion contracture, capsulotomy of the interphalangeal joints is performed. Axial spokes are inserted in the middle positions of the toes.

The feasibility of forming the interdigital spaces in the presence of bone syndactyly is a subject of debate. In our opinion, in the case of total bone syndactyly of underdeveloped toes, the indication for surgery is, above all, pain in the first metatarsophalangeal joint, which usually develops by the age of 4 years. Pain is caused by the convergence of the metatarsal heads, hallux valgus deformation of the first toe, subluxation of metatarsophalangeal joint, and its overload due to the absence of normal instep toe rolling. Surgery includes elimination of the fusion between the first and second toes. In forming the side surfaces of the separated toes, an autograft is required, which is harvested from the dorsal and plantar surfaces of the foot. A full-thickness skin flap is moved so that the exposed bone surfaces on the distal phalanges, formed after cutting the fusion, are completely covered by the patient's own tissues. The remaining wound surfaces are closed with a split-thickness skin autograft. If there is an insufficient amount of the patient’s skin to cover the exposed bone surfaces, it is advisable to remove the nail phalanx to provide an adequate supply of tissues.

Of particular interest are adolescent patients who underwent surgery in the first year of life because of first-ray polysyndactyly combined with delta-phalanx and trapezoidal metatarsal bones. Surgery was performed without complying with the treatment principles described earlier. In addition to the severe deformation and shortening of the first ray, these patients had significant secondary changes of other metatarsal bones and varus deviation of the forefoot. The first metatarsophalangeal joint does not bear a load. The head of the second metatarsal bone is lowered onto the plantar plane where significant callus is present, and there is pain due to its overload during midstance and push-off phases of walking. Syndactyly of the first and second toes, which was not eliminated earlier, prevents adequate development of the first and

Figure 3. Patient S. (18 months of age) with first-ray polydactyly (A, image of the foot before surgery; B, MSCT of the foot showing the defect of the main and extra first ray; C, malformation of basic and extra first rays; D, the foot after surgery—the superposition of the first toe; E, image of the foot 1 year after superposition of the first ray)
second metatarsophalangeal joints.

Surgical correction requires the following procedures: elimination of the deformation and restoration of the length of the first metatarsal bone with correct parabola formation, metatarsophalangeal arthrodesis of the Lisfranc joint to eliminate the pronounced adducted position of the forefoot, corrective osteotomies of small rays, and elimination of syndactyly of the first and second rays. For maximum elongation of the first column, arthrodesis is performed after derotation, resection, and modeling of the first cuneiform bone [18]. Fragments of bone tissue obtained by resection of the cuneiform bones, which is performed to correct deformation and to shape these bones as close to normal anatomy as possible, are used for elongating arthrodesis of the first metatarsophalangeal joint (Fig. 4).

Results

Here we present the results of treatment of 37 patients (53 feet) with malformations of the first ray of the foot that we observed for 1–5 years. Clinical and radiological examinations show that patients with phalanx malformations during the period after the initial surgery did not develop changes that required surgical correction. The X-ray shows normal formation of the first and nail phalanges, epiphysis sphericity was preserved, and there was longitudinal growth of the segment. There were no deformations of the nail plate. The results of the first step of treatment were evaluated as good in 100% of patients.

The results of treatment of polydactyly with LEB of the proximal phalanx show that 3 feet of 11 patients of preschool age, who first underwent surgery between the ages of 1 and 2.5 years, had recurrent varus deformity of the proximal phalanx of the first toe with an average of 24°. Such deformation will require corrective wedge osteotomy. However, this condition is not painful but makes it difficult to select shoes. Overall, the results of treatment of polydactyly with LEB of the proximal phalanx were considered good and satisfactory in 77% and 27% of cases, respectively.

Treatment of isolated defects of the first metatarsal bone with growth plate deformity was conducted for patients aged 6 and 8 years (four feet). During the observation period, the axis of the toe was correct, range of motion of the metatarsophalangeal joint was not limited, and growth rate of the first metatarsal bone maintained the shape of the metatarsal parabola that was formed during surgery.

Correction of first and second-ray syndactyly combined with phalanx clinodactyly, which was performed for three patients (six feet), significantly reduced deformation of the fingers during growth. The absence of fusion between the nail phalanges allows use of the growth plate potential to grow in the correct direction (67% good results, 4 feet
and 33%, satisfactory results, which were severe scarring and partial fusion of the interdigital spaces).

Long-term results of treatment of complex malformations of the first ray of the foot, polysyndactyly in the presence of delta-phalangeal and metatarsal trapezoidal bones, were regarded as results of the first phase of the correction of the deformity. Fourteen patients underwent surgery at 6 months–2.5 years of age and 1 patient at 8 years of age (observation period 1 year). All results of the first phase of treatment were evaluated as good.

The intermediate outcomes of treatment of children with shortening of the first ray depend on the degree of deformation of the proximal phalanx of the first toe and the degree of shortening of the primary metatarsal bone. Here we show that the primary correction of the deformity was sufficient to maintain the correct ratio in the metatarsus-phalangeal joint for 4–5 years, as long as the correct shape of the metatarsal parabola was maintained. MSCT showed preserved, anatomically correct position of the growth plate (Fig. 5), which was considered a good result of the first phase of treatment.

At the age of 5 or 6 years, there emerges a shortening of the remaining metatarsal bone (0.8–1 cm) and proximal phalanx of the first toe (0.2–0.5 cm) with an appearance of a tilt of the epiphysis toward the medial side and varus deformity of the first toe (8°–12°). The angles between the first and second metatarsal bones ranged from 14° to 18°. Limitation of function of the first metatarsophalangeal joint did not exceed 7°. There was lateral subluxation of the first metatarsophalangeal joint with an angle of lateral deviation of up to 5° and dorsal subluxation up to 5° due to lapse of a first metatarsal bone head in the plantar direction. Use of splints during sleep in a position of overcorrection of the pathologically developed ray and wearing orthopedic shoes with a rigid internal side insert may hold the toe in the middle position but does not prevent development of pathological deformation. However, none of the patients experienced pain, and there were no significant difficulties in selecting shoes, which is regarded as a satisfactory result for all feet. None of the patients in this group required second-phase surgery.

Two patients, 15 and 16 years of age with secondary deformities of the metatarsals bones, were followed-up for 2 years. The extending arthrodesis of the first ray together with plastic resection of the first cuneiform bone, corrective osteotomy of the second and fourth metatarsal bones, and elimination of syndactyly of the first and second toes allowed formation of the forefoot, close to the anatomical norm. Metatarsus-phalangeal roll was restored, and the first metatarsal bone that carries the load was formed. The range of motion of the first metatarsophalangeal joint was 18–20° with slight limitation of plantar flexion, which did not interfere with wearing shoes with a heel.
as high as 6–8 cm. Both results were regarded as good.

Biomechanical studies show that the restoration of the length and shape of the first metatarsal bone length greatly improves the ability of the foot to support a load, and the distribution of the load on the metatarsal bones approaches normal for the patient’s age (Figure 6). Patients did not report pain, and pressure sores and abrasions were not caused by wearing various types of shoes. Further, shoes did not lose their shape with extended use.

To assess the long-term outcomes of treatment, we used a questionnaire similar to that developed by the ACFAS (Table 1).

It should be noted that the assigning nine points or fewer may indicate that a second phase of surgery will be required to correct the shape of the first ray and restore the integrity of the metatarsal parabola.

### Table 1

<table>
<thead>
<tr>
<th>Patients’ opinions</th>
<th>Number of patients</th>
<th>Number of points</th>
</tr>
</thead>
<tbody>
<tr>
<td>No pain and high physical activity</td>
<td>28</td>
<td>5</td>
</tr>
<tr>
<td>Rare pain, which did not affect physical activity</td>
<td>5</td>
<td>4</td>
</tr>
<tr>
<td>Frequent pain, reduced physical activity</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>Frequent pain, significantly reduced physical activity</td>
<td>–</td>
<td>2</td>
</tr>
<tr>
<td>Severe pain, no physical activity</td>
<td>–</td>
<td>1</td>
</tr>
</tbody>
</table>

| Evaluation of the foot shape                   |                     |                  |
| Liked it very much                             | 17                  | 5                |
| It was OK                                      | 13                  | 4                |
| Wish it was better                             | 7                   | 3                |
| Not very good                                  | –                   | 2                |
| Did not like it                                | –                   | 1                |

| Shoe selection options                         |                     |                  |
| Any kind of shoe                               | 28                  | 5                |
| Any kind of shoe, limited use                  | 5                   | 4                |
| Athletic shoe only                             | 4                   | 3                |
| Orthopedic footwear only                       | –                   | 2                |

### Conclusion

Surgical treatment of malformations of the first ray is a multistep procedure, and its goal is to maximize the reconstruction of the anatomy and function of the foot and achieve a cosmetic effect.

The purpose of the first phase of correction is to create prerequisites for normal growth by correcting the skewed position of the growth plate of the first metatarsal bone and forming an anatomically correct relationship to the metatarsophalangeal joint. The calculations to determine the required corrections were based on radiographs and MSCT. The first phase of the treatment of patients with severe deformities was performed before the age of 1 year to create the proper foot shape when the child starts to acquire erect posture. An exception was the treatment of polyphalangy, which required the second phase of surgical correction.
Corrections of the isolated malformations of the first metatarsal bone were performed in one-step surgery, elongation of the metatarsal bones and adjustment of the location of the growth plate at 5–7 years of age, because these defects were mildly to moderately severe.

Surgical correction of polysyndactyly comprised elimination of all elements of the first-ray deformation after excision of the fibrous strands. For a double first ray, additional rays and hypoplastic elements were removed, and the main ray was then formed as close to normal as possible. Correct choice of a long-term treatment strategy led to a radiological outcome that resembled the anatomical norm.

The final phase of correction of the first metatarsal or phalanx deformity was performed after growth stopped at the age of 14–15 years. Long-term results included marked improvement in gait, decrease or absence of pain, and satisfactory cosmetics.

In secondary deformations of the first ray, patients experienced pain, foot supination, cosmetic defects, deformation of the second to fifth metatarsal bones, functional impairment, and reduced ability to exert rolling support of the limb. Correction of secondary deformities of the forefoot required extending arthrodesis of the tarsometatarsal articulations and modeling resection of the first metatarsal and cuneiform bones.

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