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MEDICAL ABILITATION OF PATIENTS WITH PROXIMAL ECTROMELIA OF THE LOWER LIMBS

© ***V.A. Yakovleva, I.V. Shvedovchenko, A.A. Koltsov***

Federal Scientific Center of Rehabilitation of the Disabled named after G.A. Albrecht, Saint Petersburg, Russia

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One of the rarest and most severe variants of underdevelopment is proximal ectromelia of the lower extremities, in which all segments of the lower limb are affected and the degree of damage decreases from proximal to distal direction. A review of the domestic and foreign literature sources that describe the various clinic-radiological variants of pathology was carried out, from the presence of a single rudimentary foot adjacent to the trunk to the presence of all three leg segments. The terminological designations of this type of underdevelopment were then analyzed. The term "proximal ectromelia" is proposed as the most appropriate clinical and radiological features of the pathology. With a severe degree of reduction of the limb, when the femur is absent or sharply hypoplastic, prosthetics is carried out in almost all patients. In this situation, surgical treatment is used as a preparatory stage to optimize the design of the future prosthesis. In addition, surgery is the main method of treatment in cases with a lighter degree of underdevelopment, in which case the technical means of rehabilitation are auxiliary. Therefore, despite the relative rarity of this pathology, its severity and medical and social significance determine the interest of specialists of the world community in the study of the problem.

Keywords: proximal ectromelia; lower extremities; *coxa vara* congenita; phocomelia; dysmelia; medical abilitation; shortening of limb; congenital short femur; proximal femoral focal deficiency.

МЕДИЦИНСКАЯ АБИЛИТАЦИЯ ПАЦИЕНТОВ С ПРОКСИМАЛЬНЫМИ ФОРМАМИ ЭКТРОМЕЛИИ НИЖНИХ КОНЕЧНОСТЕЙ

© ***В.А. Яковлева, И.В. Шведовченко, А.А. Кольцов***

ФГБУ «Федеральный научный центр реабилитации инвалидов им. Г.А. Альбрехта» Минтруда России, Санкт-Петербург

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Проксимальная эктромелия нижних конечностей — одна из наиболее редких и тяжелых форм недоразвития, при которой затронуты все сегменты нижней конечности, а степень поражения уменьшается в дистальном направлении. Изучены отечественные и зарубежные литературные источники, в которых описаны различные клинико-рентгенологические варианты патологии: от наличия единственной рудиментарной стопы, прилежащей к туловищу, до присутствия всех трех сегментов конечности. Проанализированы используемые терминологические обозначения данного типа недоразвития, предложен термин «проксимальная эктромелия», как наиболее соответствующий клинико-рентгенологическим особенностям патологии. При тяжелой степени редукции конечности, когда бедренная кость отсутствует или резко гипопластична, протезирование проводят практически всем пациентам. Хирургическое лечение в такой ситуации выступает подготовительным этапом с целью оптимизации конструкции будущего протеза. В случае более легкой степени недоразвития оперативное вмешательство является основным методом лечения, в таком случае технические средства реабилитации

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выполняют вспомогательную функцию. Таким образом, несмотря на относительную редкость данной патологии, ее тяжесть и медико-социальная значимость определяют интерес специалистов мирового сообщества к этой проблеме.

Ключевые слова: проксимальная эктромелия; нижние конечности; врожденная варусная деформация шейки бедра; фокомелия; дизмелия; медицинская абилитация; укорочение конечности.

Background

Orthopedic diseases such as abnormalities of the lower extremities are one of the common causes of disability due to restriction in or lack of independent movement. The incidence of various congenital malformations of the lower extremities, accompanied by shortening, varies significantly. According to different authors, the incidence in the population is 0.29 per 1,000 newborns or 1 case per 3,500 newborns [1]. Higher values, 1.1 per 1,000 inhabitants, are reported in the study by Kuklik and Marzik (1999) [2, 3]. Despite the fact that, generally, developmental defects of the femur are relatively rare, aplasia of the hip joint and femur is the most common among all congenital abnormalities of the lower extremities and accounts for 1.2% of all pathologies of the musculoskeletal system [4, 5].

The etiology and pathogenesis of congenital diseases of the musculoskeletal system, including proximal forms of ectromelia of the lower extremities, cannot be considered completely resolved presently. They can develop due to environmental influences, registered in a number of chromosomal (Edwards syndrome, Orbeli syndrome, etc.) and gene (Roberts syndrome, etc.) syndromes. The mode of inheritance of the pathological isolated forms is currently understudied [4, 6, 7].

One of the common causes of congenital developmental extremity abnormalities in newborns is the teratogenic effect of a number of drugs. The most popular example of such an effect is thalidomide embryopathy manifested at birth in Western Europe, especially in Germany, in 1959–1963, in several thousand pediatric patients with gross reductions in the upper and lower extremities due to intake of thalidomide, a sedative and hypnotic drug, marketed by the Grünenthal company in 1957, by their mothers during pregnancy [3, 6, 8].

Terminology

Proximal ectromelia is one of the rarest and most severe variants of underdevelopment, in which all segments of the extremity are affected and the degree of damage decreases in the distal direction. Shturm was one of the first to use this term in Russian literature in 1960 to characterize a defect in the proximal part of the lower extremity (femur), without damage to the distal region [9]. Overseas, the proximal ectromelia definition was proposed by Henkel and Willert in 1969 and used to describe patients with the most pronounced reduction in the femur with relative preservation of the foot and lower leg [10].

In the Russian and foreign literature, other definitions denoting this pathology are more common. One of the most common terms is phocomelia (phoke, "seal"; melos, "extremity"). The name is used due to the outward resemblance of an underdeveloped extremity to a seal fin. Phocomelia is characterized by the absence of proximal or proximal and middle segments of the extremity with the distal segment (foot) attached to the trunk [11]. Frantz and O'Rahilly (1961) identified three types of pathology, depending on the severity of skeletal lesions, namely, proximal (aplasia of the femoral bone), distal (aplasia of the lower leg bones), and full (aplasia of all long tubular bones with the hip joint). In the full form of the pathology, the foot articulates directly with the body, and in most severe underdevelopment, it is represented by a single underdeveloped finger. This form of phocomelia is called peromelia [4].

The term dysmelia is also quite common and used to define a group of reduction malformations. In dysmelia, there is hypoplasia or aplasia of the tubular bones of the extremities, ranging from the underdevelopment of a particular segment to almost complete absence of an extremity [9, 12].

Regarding dysmelia, a number of authors distinguish congenital varus deformity of the femoral neck (*coxa vara*) as a symptom complex that includes shortening of the extremity by 3–25 cm and external rotation, adduction, or flexion contracture in the hip joint, often combined with damage to the distal parts of the extremities [5, 13–15].

In the 1960s, foreign authors often distinguished two variants of the pathology of the femur, namely,

proximal femoral focal deficiency (PFFD) or "local proximal femoral deficiency" and congenital short femur (CSF) or "congenital short femur." The first variant of the pathology is represented by lesions of the proximal femur, acetabulum, and inferior knee joint, while the second variant is characterized by congenital hip hypoplasia with a difference in leg length, but the hip and knee joints are functionally healthy [16]. The CSF is normal in structure but has reduced size with a decrease in the caput-collum-diaphyseal (CCD) angle and curvature of the diaphysis and is most associated with dystrophic *coxa vara* [17].

Thus, the analysis of world literature shows that the pathology described is indeed a more complex problem than damage to only the femur and includes reduction of all segments of the extremities.

Classifications

Several classifications have been proposed in the world literature, most of which are based on the analysis of the clinical and radiological image of the reduction of the femur and pelvis and differ from each other in the number of variants of underdevelopment and the detail of their description.

In the Russian literature, we found only two classifications. Thus, Mezhenina divided all variants of the abnormality into two groups: the first group included disorders with a total defect, while the second group included those with a partial hip defect. With this, the second group consisted of four subgroups according to the level of femoral damage [4]. Based on the analysis of long-term experience, Pozdeev et al. developed a classification of congenital varus deformity of the femoral neck and identified three degrees of damage depending on the CCD angle, structure of bone tissue, and degree of shortening [15].

A greater number of classifications are presented in foreign literature; one of the first was published by Mauche and Jbos (1928) and generated based on anatomical and evolutionary characteristics. The authors identified several groups: in the first group, the hip is reduced in all sizes, and *coxa vara* is almost always noted; in the second group, there are underdeveloped diaphysis and two pineal glands that exist separately; in the third group, the distal pineal gland is absent or underdeveloped; in the fourth group, the upper epiphysis is absent; in the fifth group, there is no proximal and distal femur, and the diaphysis is rudimentary; in the sixth group, the femur is completely absent [8, 18].

It seems that the classifications by Reiner (1946) and Steindler (1950) provide no clear distinction between localizations and types of extremity malformations, and therefore, these classifications are difficult to apply in daily practice. Particularly, Reiner (1946) identifies four types of underdevelopment, without investigating in detail the degree of hip underdevelopment and ratio of hip lesion to distal extremity [19].

The Aitken classification includes four classes of the pathology (*A, B, C, D*) and details the reduction of the femur based on the presence or absence of the acetabulum and femoral head [20] (Fig. 1).

Amstutz (1962) updated the above classification, dividing the class A into two types: the first type included mild forms of femur shortening with the presence of *coxa vara*; the second type was characterized by the presence of subtrochanteric pseudoarthrosis [21, 22].

The published work by Fixsen and Lloyd-Roberts proposes a prognostic X-ray model of the development of a defective femur in pediatric patients in the first year of life, using the example of 25 patients (30 extremities), and three types of underdevelopment were identified [23].

Another classification was developed by Henkel and Willert (1969). The authors described a teratological series of dysmelia, which included five types, namely, distal, axial, and proximal forms of ectromelia, phocomelia, and amelia. Each of the first three types was divided into short,

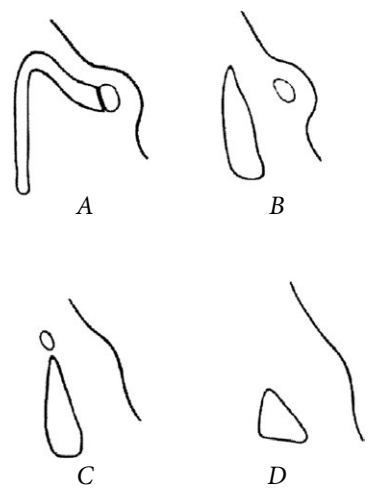

Fig. 1. Aitken classification (1959)

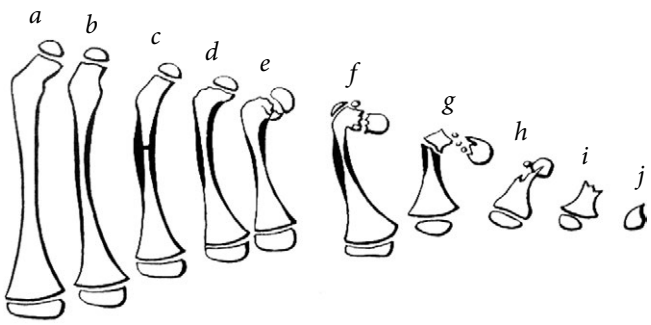

Fig. 2. Hamanishi classification (1980)

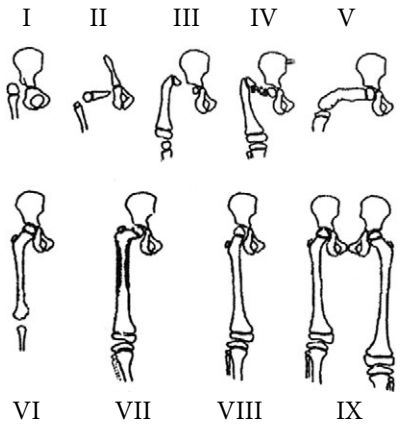

Fig. 3. Pappas classification (1983)

intermediate, and long subtypes. In the distal form, there is damage to the foot and tibia; in the axial form, there is also damage to the femur, but the fibular bone remains intact; the proximal form is characterized by varying degrees of damage to the femur only [10].

Hamanishi (1980) proposed a classification of five groups, each of which was divided into two subgroups, from hip hypoplasia with intact diaphysis to a completely absent or rudimentary hip with delayed ossification [16] (Fig. 2).

In 1983, Pappas proposed a classification of hip developmental abnormalities, consisting of nine classes of malformations, from segment aplasia to slight shortening of the femur without deformities: class P I corresponds to complete hip aplasia or with only condyles, and P IX corresponds to shortening of segments without deformities [24] (Fig. 3).

In their study, Torode and Gillespie (1991) provided a detailed description of the clinical and radiological characteristics of proximal femur underdevelopment and divided it into two groups depending on the degree of shortening of the femur and extremity as a whole: the first group (CSF) includes patients with a 20–30% deficiency of the lower extremity, while the second group (PFFD) includes those with a 35–50% deficiency of the lower extremity [25].

One of the latest classifications proposed by Paley (1998) includes four types of lesions and is based on a clinical and radiological presentation:

Type 1 shows that the femur is intact and there is a full range of movements in the hip and knee joints with normal ossification of the proximal femur (*a*) or with its delay (*b*).

Type 2 includes mobile pseudoarthrosis in the proximal femur with a full range of movements in the knee joint, while the femoral head is within the acetabulum (*a*) or the femoral head is absent or immobile in the acetabulum (*b*).

Type 3 shows underdevelopment of the femur diaphysis, with a decrease in the amplitude of movements in the knee joint of >45° (*a*) or <45° (*b*).

Type 4 shows the distal femur deficiency with the preserved proximal part, and it does not belong to the group of the pathology under consideration [26] (Fig. 4).

The state of electrical activity of the muscles and blood circulation with congenital malformations of the lower extremities, with proximal ectromelia, is not fully discussed in the literature. A number of authors noted a decrease in the electrical activity of muscles in a shortened extremity compared with that in the contralateral segment [27, 28]. The degree of reduction in the amplitude of contractions of the femur muscles in the absence of the head and neck sometimes reaches 60–70% of the muscle strength of the intact extremity [27].

The peripheral vasculature also changes with congenital underdevelopment of the femur. The rheovasography and angiography findings indicate both a decrease in volumetric blood flow and underdevelopment and atypical arrangement of vessels on the pathology side, as the diameter of the main trunks along their length is different, there are focal dilations and constriction of the vessels, the number of functioning capillaries is reduced, and their shape is disturbed [3, 28, 29].

The main methods of medical abilitation of patients with proximal forms of lower extremity ectromelia, depending on the defect severity, are surgical treatment and prosthetics. Thus, with a relatively intact extremity, surgical treatment is considered the main and effective method, and prosthetic and orthopedic supplies are used as an auxiliary method at one stage or another. In contrast,

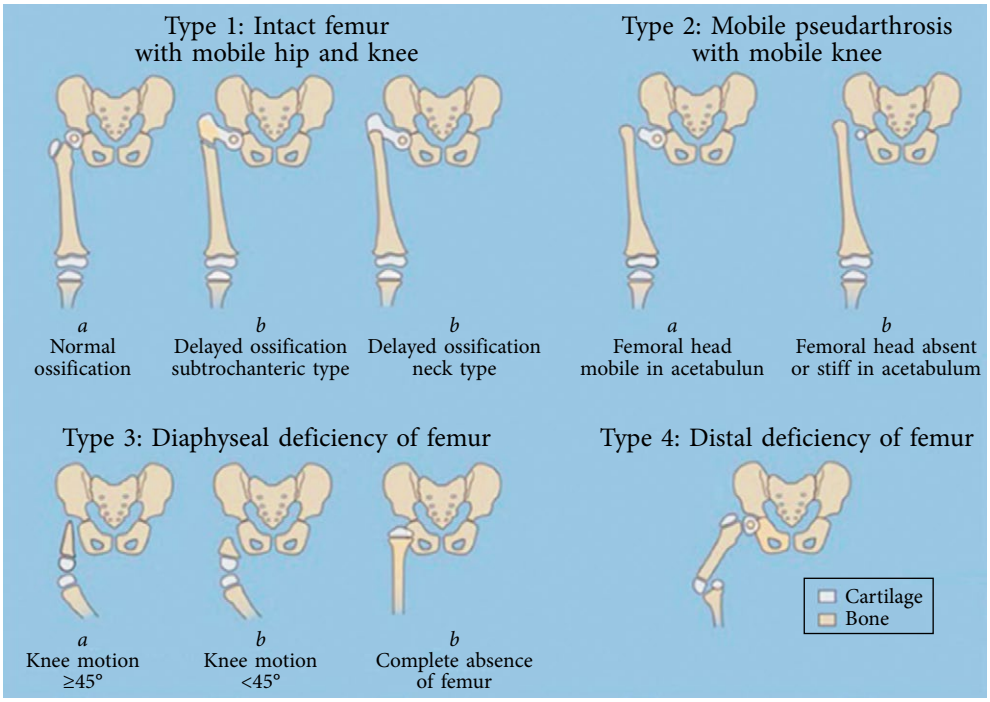

Fig. 4. Paley classification (1998)

in severe cases of extremity underdevelopment, the primary method is complex or atypical prosthetics, while surgical interventions are aimed at preparing for it or optimizing it [3, 30, 31].

Treatment

The primary aim of surgical treatment is restoration of the extremity locomotor function by eliminating the main manifestations of underdevelopment, such as shortening, deformities, and contractures [30]. Surgical intervention aimed at eliminating inequality and length was first proposed by Rizzoli in 1845, and in 1925, Deutschlander began to shorten the femur of a healthy extremity and elongate the short one simultaneously using a resected femur, for the same purpose [3]. Russian authors proposed to perform this surgery in all cases of congenital shortening of the extremities, if the difference in length exceeds 10–12 cm. Surgical treatment was performed using an auto- or allograft and performed in two stages [3]. A number of authors used surgeries that stimulate extremity growth by activating the function of the epiphysial plates. Zatsepin (1930), Moskvin (1951), and Schneiderov (1965) developed and studied the methods of stimulating extremity growth by insertion of bone pins near the epiphysial plates [30]. The abovementioned methods of surgical treatment currently have more historical value, since, with the appearance of the Ilizarov method, they lost their relevance and the method of compression and distraction osteosynthesis took a leading position [32].

In 1951, Ilizarov proposed the design of a compression and distraction apparatus. Strict adherence to the biological laws of bone tissue regeneration enabled to achieve impressive results of surgical treatment in congenital and acquired shortening of the lower extremities and shortly introduce the apparatus into practice [1, 33, 34]. The Ilizarov method can be used during reduction of the femur, both as an independent method of surgical intervention with a slight degree of reduction and as part of a complex multistage surgical treatment for more severe pathologies [32, 35, 36].

Paley proposed a stepwise surgical treatment algorithm based on his classification. The author considers it most appropriate to resort to reconstructive surgery in case of the first two types of lesions. The treatment includes corrective osteotomy to eliminate varus deformity of the femoral neck, pelvic osteotomy to correct the acetabular angle and increase the degree of coverage of the femoral head with the acetabular roof, lengthening a segment using a compression and distraction apparatus, epiphysiodesis of one or more epiphyses of a healthy contralateral extremity, and elimination of subluxation of the patella or tibia [36].

Another aim of surgical treatment of patients with gross underdevelopment of the hip and extremity as a whole consists of preparation for prosthetics by eliminating the most pronounced deformities and instability of the joints and changing the appearance and function of the extremity to optimize the prosthesis design and improve the support ability and gait stereotype [30]. According to some authors, the method of choice for proximal ectromelia P1 and P2 (according to Pappas), that is, in the presence of rudimentary condyles of the femur, is the formation of iliofemoral synostosis to stabilize the extremity in the area of its articulation with the trunk [3, 35]. Reconstruction of the proximal femur in case of intertrochanteric pseudoarthrosis or pseudoarthrosis of the femoral neck is performed for abnormalities of the development of classes P3 and P4 (according to Pappas); thus, it aims to connect the dissociated bone-cartilaginous fragments and increase the CCD angle and relative length of the extremity [3, 35, 37]. The absence of a femoral head and pronounced adduction contracture in the hip joint in pediatric patients aged >12 years and adolescents are the basis for reconstructive surgery on the proximal femur with the formation of an additional point of support of the femur in the pelvis [15].

In cases when alignment of the length of the affected and healthy extremities with the congenital absence of the femur or its pronounced growth retardation is impossible, some authors consider it appropriate to perform amputation at a certain level [38]. Particularly, one of the known methods aimed at the formation of a supportable stump of the lower leg is Syme amputation, which was proposed in 1842 and subsequently provides the patient with the possibility of lower leg prosthesis [39, 40].

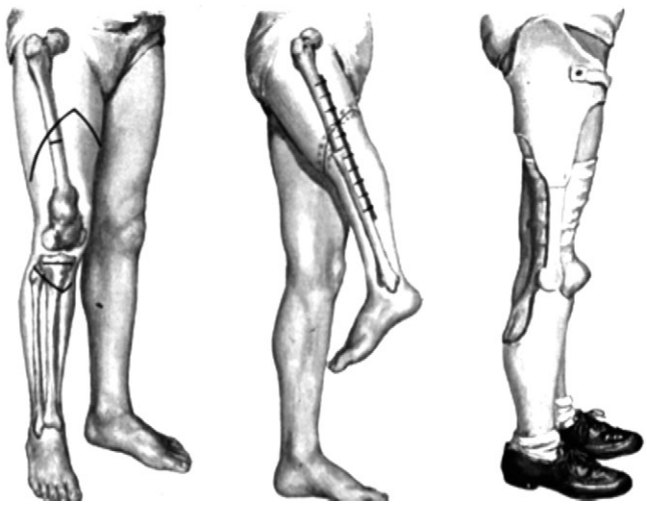

Fig. 5. Scheme of Van Nes surgery and the result of prosthetics (Frank Henry Netter)

In 1930, Borggreve, followed by Van Nes in 1950, proposed the use of rotational grafting to improve extremity function and increase the functionality of subsequent prosthetics. The surgery was initially intended to treat pediatric patients with congenital defects of the femur but was later performed for acquired defects of the femur and knee joint. During the surgery, the knee joint is resected, as well as parts of the femur and tibia, while maintaining the posterior tibial neurovascular bundle; osteosynthesis of the femur and tibia is performed after rotation of the lower leg and foot by 180°. As a result of the intervention, the level of the ankle joint of the affected extremity is displaced proximal to the level of the knee joint of the contralateral extremity, while the foot that rotated posteriorly imitates a short stump of the lower leg [41, 42]. Kostuik et al. analyzed the results of 20 surgeries and recommended their implementation in patients aged >12 years due to the high probability of repeated surgical intervention [43] (Fig. 5).

Complex or atypical prosthetic and orthopedic products are used in medical abilitation of patients with proximal forms of ectromelia of the lower extremities. The choice of the product is determined by the degree of extremity shortening and severity of anatomical changes; thus, orthopedic shoes, orthoses, and prostheses of various designs are used [31, 44, 45].

Thus, with insignificant degrees of reduction, when shortening the extremity to 5 cm, it is recommended to use orthopedic shoes or orthosis (splint) with compensation for shortening. Orthopedic shoes may include corrective elements, such as arch support, pronator, rigid bootleg, and removal of the heel and sole, which result in partial or complete correction of the foot position and fixation of the ankle joint area [31]. According to some authors, the disadvantages of orthopedic shoes are the impossibility of use with pronounced shortening of the extremity and, in some cases, a decrease in patient stability when walking [31]. In this regard, with a shortening of >5 cm and relative preservation of all extremity segments, it is possible to prescribe orthoses to the ankle joint or entire extremity with a heel lining [44].

According to the literature analysis, the approach to prosthetic and orthopedic supplies

is totally different with more severe forms of extremity underdevelopment, namely, shortening of ≥8 cm [45, 46]. In such cases, more complex products are prescribed, such as hip prostheses with a double sole, otherwise called prostheses for congenital underdevelopment of the extremity [37]. The structural property of this product consists of the presence of an artificial foot located distally with respect to the lower leg bucket. The schemes for constructing a prosthesis vary depending on the knee joint viability and degree of reduction of the extremity segments. Thus, with the relative preservation of the extremity segments, some authors used metal splints connecting the femur and lower leg buckets and providing mobility and lateral stability to the knee joint, while, with coarse forms of reduction, the entire shortened extremity was immersed in a single bucket of prosthesis [38]. In case of hip joint disability or absence, the prosthesis design included the formation of a thrust on the ischial tuberosity to unload the proximal part of the extremity under prosthetics [31].

In performing Syme amputation with simultaneous arthrodesis of the knee joint, the prospects for improving the walking function in the hip prosthesis increase due to the simplification of its scheme, namely, optimization of the shape of the bucket of prosthesis, exclusion of the "double sole" from the design, and presence of the knee joint coaxial to the knee joint of a healthy extremity [25, 47, 48].

Simultaneously, the greatest functionality when walking is provided by lower leg prostheses, which can be supplied to the patient after complex surgical preparation and Van Nes rotational grafting [42]. In such cases, the fundamental difference is the formation of the patient's own "knee joint" located at the level of the knee joint of a healthy extremity, and, thus, the appearance of additional degrees of extremity mobility. Thus, the foot that rotated posteriorly is located in the bucket of prosthesis of the lower leg and imitates a short stump of the lower leg, and the cuff on the femur ensures stable fixation of the prosthesis on the extremity [25, 49, 50].

Alman et al. analyzed the results of the clinical presentation and X-ray examination of 16 patients with proximal form of ectromelia of the lower extremities, while patients with bilateral lesions or multiple abnormalities and those who could have the femur lengthened in the future to align the length of the extremities were excluded from the study. Nine patients underwent rotational grafting, and seven patients underwent Syme amputation with knee arthrodesis. The authors note that there was no significant difference in the extremity appearance; however, the stereotype of walking with prosthetics after rotational grafting was better [32]. In 1991, another group of researchers proposed an algorithm for surgical intervention depending on the type of extremity underdevelopment: In CSF, the authors recommended performing Syme amputation in cases of fibula and/or tibia deficiency and foot instability. In PFFD, the last stage of a multistage surgical intervention has always been rotational grafting or Syme amputation, also depending on the preservation of the ankle joint and its functionality. The authors of the article noted the functional advantage of rotational grafting [24].

Conclusion

Thus, the most common terms ("phocomelia," "dysmelia," "underdevelopment of all segments of the extremity," and "axial ectromelia") characterize a whole group of gross abnormalities of the lower extremities. The term "phocomelia" implies only the appearance of the extremity, without considering its anatomical structure; "dysmelia" indicates a disorder of extremity formation, which entailed a significant change in its shape and function, while the other two definitions indicate the presence of underdevelopment of all segments of the legs without specifying their degree. The term "proximal ectromelia," from our point of view, implies most accurately the essence of the anatomical and corresponding functional changes and is optimal.

In Russian and foreign literature, there are descriptions of various clinical and radiological forms of the pathology, from the mildest (shortening with a relatively preserved femur) to the most severe reduction of all segments of the extremity. Most studies present the results of monitoring of small groups of patients with certain types of lesions. Only in rare publications, the authors provide a detailed anatomical and functional analysis on a sufficient number of patients.

Classifications presented in the world literature do not cover the entire range of proximal forms of underdevelopment of the lower extremities. However,

some of them, classifications by Aitken (1959), Pappas (1983), and Paley (1998), are used actively in clinical practice.

The analysis of the available literature shows that the main methods of abilitation of patients with proximal forms of lower extremity ectromelia are surgical treatment and complex prosthetics.

In case of severe extremity reduction, when the femur is absent or sharply hypoplastic, prosthetics is performed in almost all patients. In this situation, surgical treatment is used as a preparatory stage to optimize the design of the future prosthesis, namely, extremity derotation with excessive external rotational contracture, extremity stabilization at the level of its articulation with the pelvis with severe instability, and formation of the "knee joint" and providing the possibility of more functional prosthetics.

With a milder degree of underdevelopment, surgery is the main treatment method; in this case, technical rehabilitation tools play an auxiliary role and are used for a limited period to compensate for extremity shortening and stabilization at rest and during physical load.

Thus, despite the relative rarity of this pathology, its severity and medical and social significance determine the major interest of the global community in studying this problem, improving treatment methods, while some of the methods proposed in the last century still retain their relevance.

Concurrently, further study of the proximal forms of ectromelia of the lower extremities, such as the compilation of a complete teratological range of all forms of underdevelopment, their detailed clinical and radiological description, creation of a working classification convenient for practical use, and improvement in the concept of complex abilitation, considering the fast developing prosthetic and orthopedic industry, is a relevant task.

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Contribution of the authors

V.A. Yakovleva created the concept and design of the study, collected the data, processed the material, and wrote the initial manuscript.

I.V. Shvedovchenko performed stage and final editing of the manuscript.

A.A. Koltsov created the concept and design of the study, wrote the initial manuscript, and was involved in the stage editing.

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Information about the authors

Veronika A. Yakovleva* — MD, Orthopedic and Trauma Surgeon of the Federal Scientific Center of Rehabilitation of the Disabled named after G.A. Albrecht, Saint Petersburg, Russia. https://orcid.org/0000-0001-6534-1760. E-mail: yak_vero@mail.ru.

Igor V. Shvedovchenko — MD, PhD, D.Sc., Professor, Scientific Supervisor of the Federal Scientific Center of Rehabilitation of the Disabled named after G.A. Albrecht, Saint Petersburg, Russia. https://orcid.org/0000-0003-4618-328X. E-mail: schwed.i@mail.ru.

Andrey A. Koltsov — MD, PhD, Orthopedic and Trauma Surgeon, Head of the First Child's Traumatology-Orthopedic Department of the Federal Scientific Center of Rehabilitation of the Disabled named after G.A. Albrecht, Saint Petersburg, Russia. https://orcid.org/0000-0002-0862-8826. E-mail: katandr2007@yandex.ru.

Вероника Анатольевна Яковлева* — врач травматолог-ортопед, ФГБУ «ФНЦРИ им. Г.А. Альбрехта» Минтруда России, Санкт-Петербург. https://orcid.org/0000-0001-6534-1760. E-mail: yak_vero@mail.ru.

Игорь Владимирович Шведовченко — д-р мед. наук, профессор, научный руководитель ФГБУ «ФНЦРИ им. Г.А. Альбрехта» Минтруда России, Санкт-Петербург. https://orcid.org/0000-0003-4618-328X. E-mail: schwed.i@mail.ru.

Андрей Анатольевич Кольцов — канд. мед. наук, заведующий первым детским травматолого-ортопедическим отделением ФГБУ «ФНЦРИ им. Г.А. Альбрехта» Минтруда России, Санкт-Петербург. https://orcid.org/0000-0002-0862-8826. E-mail: katandr2007@yandex.ru.