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Variants of pathological changes of feet in children with Klippel–Trenaunay syndrome

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ABSTRACT

Background. The severity of foot deformities in embryonic type dysplasia of the great veins of the lower extremities (Klippel–Trenaunay syndrome) varies depending on the severity of vascular damage. Hypertrophy of the limbs or individual segments can range from insignificant to extremely pronounced. Klippel–Trenaunay syndrome is often combined with orthopedic pathologies such as macro- and microdactyly, syndactyly, flat feet, and contractures.

Aim — to analyze the variants of pathological changes in the feet in children with Klippel–Trenaunay syndrome of varying severity.

Materials and methods. The work is based on the results of examining 73 patients with embryonic type dysplasia of the great veins (ETMGV) aged from 0 to 18 years, who were treated from 2014 to 2024. According to the severity of the pathology, four groups of patients were identified — with mild, moderate, severe and extremely severe degrees of ETMGV (Klippel–Trenaunay syndrome includes only moderate, severe and extremely severe degrees of ETMGV).

Data processing was performed using the STATISTICA 6.0 and Excel2007 computer programs.

Results. Patterns of increase in the length and circumference of the feet in patients with Klippel–Trenaunay syndrome were determined. Statistical analysis of the average difference in foot lengths and the asymmetry coefficient was performed in groups of patients compiled in accordance with the severity of the syndrome. An increase in the severity of foot length asymmetry was found with an increase in the severity of Klippel–Trenaunay syndrome. As a result of examination of 73 children, equinus contracture of the feet of the affected limb was detected by us in 5 patients (6.84%), of which 2 (2.7%) had a severe degree, and 3 (4.1%) had an extremely severe degree. Split foot was detected in one patient (1.4%) with a severe degree. "Spherical" foot shape was detected in 3 children (4.1%) with an extremely severe degree of the syndrome.

Conclusions. Changes in the length and circumference of the foot of the affected lower limb in patients with Klippel–Trenaunay syndrome are directly dependent on the severity of the syndrome. Correlations were determined between the severity of the syndrome and an increase in the size of the limbs (calf circumference, thigh length and overall limb length, foot length and circumference, calf length and thigh circumference). Variants of changes in the feet were identified according to the degree of expression of cosmetic changes and changes that impair their function.

Keywords: gigantism, Klippel–Trenaunay syndrome, multiple angiokeratomas, port wine spots, foot deformities

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Варианты патологических изменений стоп у детей с синдромом Клиппеля–Треноне различной степени тяжести

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РЕЗЮМЕ

Актуальность. Выраженность деформаций стоп при дисплазии магистральных вен нижних конечностей эмбрионального типа (синдром Клиппеля–Треноне) варьирует в зависимости от степени тяжести поражения сосудов. Гипертрофия конечностей или отдельных сегментов колеблется от незначительной до чрезвычайно выраженной. Синдром Клиппеля–Треноне нередко сочетается с ортопедической патологией: макро- и микродактилией, синдактилией, плоскостопием и различными контрактурами.
Цель — проанализировать варианты патологических изменений стоп у детей с синдромом Клиппеля–Треноне различной степени тяжести.
Материалы и методы. В работе использованы результаты обследования 73 пациентов с эмбриональным типом дисплазии магистральных вен (ДМВЭТ) в возрасте от 0 до 18 лет, находившихся на лечении в хирургической клинике СПбГПМУ с 2014 по 2024 год. Выделены четыре группы пациентов по степени тяжести — легкая, средняя, тяжелая и крайне тяжелая степень ДМВЭТ (синдром Клиппеля–Треноне включает только среднюю, тяжелую и крайне тяжелую степени ДМВЭТ).
Данные обрабатывали с помощью компьютерных программ STATISTICA 6.0 и Excel 2007.
Результаты. Определены закономерности увеличения длины и окружности стоп у пациентов с синдромом Клиппеля–Треноне. Статистический анализ средней разницы длин стоп и коэффициента асимметрии проводился в группах пациентов, составленных в соответствии со степенью тяжести синдрома. Выявлено увеличение выраженности асимметрии длин стоп при нарастании степени тяжести синдрома Клиппеля–Треноне. В результате обследования 73 детей эквинусная контрактура стоп пораженной конечности отмечалась у 5 (6,84%) пациентов, из них при тяжелой степени — у 2 (2,7%), и при крайне тяжелой степени у 3 (4,1%) детей. Расщепленная стопа выявлена у одного пациента (1,4%) с тяжелой степенью. «Шарообразная» форма стопы — у 3 (4,1%) детей с крайне тяжелой степенью синдрома.
Выводы. Отмечается прямая зависимость изменения длины и окружности стопы пораженной нижней конечности у пациентов с синдромом Клиппеля–Треноне от степени тяжести синдрома. Определены корреляции увеличения размера конечностей со степенями тяжести синдрома. Выделены варианты изменений стоп по степени выраженности косметических изменений и изменений, нарушающих их функцию.

Ключевые слова: гигантизм, синдром Клиппеля–Треноне, множественные ангиокератомы, «портвейные» пятна, деформации стоп

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BACKGROUND

In 1900, the French neuropathologists M. Klippel and R. Trenaunay first described the characteristic symptoms that constitute the triad of this syndrome [1]. Based on numerous studies published in the 20th century on different types of limb enlargement, in 1921, Gassel conducted a review of 53 patients with hemihypertrophy [2].

The prevalence of Klippel–Trenaunay syndrome ranges from one to five cases per 100,000 people, and sex does not influence the likelihood of its development [3]. Limb growth is proportional to the general growth of the child, and vascular anomalies persist throughout life [4]. These processes may be accompanied by anomalies of the soft tissues, bones and changes in limbs length [5, 6]. The main sign of the syndrome is a triad of symptoms: vascular stains, abnormal varicose veins, and hypertrophy of the soft tissues and bone structures, with an increase in the length and volume of the limb. This syndrome is a manifestation of embryonic-type main vein dysplasia (ETMVD) [7]. According to the classification by D. Kupatadze, ETMVD can be presented in mild, moderate, severe and extremely severe forms [7]. The mild form of ETMVD does not include all signs of Klippel–Trenaunay syndrome, since there is no increase in the volume and length of the limb, therefore, the syndrome includes ETMVD of moderate, severe, and extremely severe degrees.

The severity of deformities in the syndrome varies depending on the severity of the vascular lesions: the more serious the vascular involvement, the more pronounced the deformities. Hypertrophy of individual limb segments can be either minor or very pronounced. Since bone tissue is not involved in the pathological process, only an increase in the limb circumference is observed. Klippel–Trenaunay syndrome is often accompanied by orthopedic pathology, such as macro- and microdactyly, syndactyly, and flatfoot [8].

According to literature sources [6, 9, 10, 11], the lower limbs are affected most often: an increase in the volume and length of the foot is noted, and less often, lengthening of the entire limb. These changes may lead to secondary deformities of the spine, pelvis, adjacent joints, and symmetrical limbs, which disrupts biomechanics and functions. It is clear that, in the absence of timely correction, a number of orthopedic complications may occur.

Despite a large number of studies on the treatment of vascular malformations, orthopedic disorders are considered concomitant pathologies that receive insufficient attention, especially regarding the possibility of their orthopedic correction. Issues related to pathological changes of the feet and the planning of the scope and stages of surgical orthopedic therapy remain insufficiently covered.

Hypertrophy of the lower limbs occurs in 95% of cases [12–15] and manifests as an increase in their length and/or circumference [13]. In the literature, descriptions of foot deformities and enlargement are general in nature, without detailed specification [16–18]. In patients with moderate Klippel–Trenaunay syndrome, changes in foot configuration are observed, accompanied by moderately pronounced cosmetic imperfections. Consequently, there were no indications for surgical orthopedic correction.

Changes in foot configuration in children with severe Klippel–Trenaunay syndrome

Changes in foot configuration in children with severe degree of the disease were divided by us into two levels according to objective cosmetic criteria.

- *Moderately pronounced* changes were defined as those that do not create a significant cosmetic defect and do not interfere with the choice and use of footwear.
- *Pronounced changes* were defined as foot configurations that cause a significant cosmetic defect and reduce the child's quality of life.

In case of significant foot enlargement associated with uneven load distribution on the lower limbs and impaired weight-bearing function of the affected limb, the gait pattern changes. Over time, these changes may cause scoliotic deformation and early development of arthrosis in the lower limbs.

The most pronounced changes in foot configuration were observed in children with extremely severe Klippel–Trenaunay syndrome. In the region of the dorsal and plantar surfaces of the feet, an excess volume of soft tissues, including subcutaneous fat and vascular malformations, was observed. The soft tissues were distributed unevenly, giving the foot a spherical shape. In three patients with an extremely severe degree of the syndrome, a spherical shape of the foot on the affected limb was observed. Dysplastic changes involved all soft tissues of the foot, lower leg, and thigh. As a result, these patients experienced constant pain syndrome, and in an upright position they could remain for no more than three minutes without the use of compression hosiery (Table 1).

MATERIALS AND METHODS

This study is based on data obtained from the examination and treatment of 73 patients with embryonic-type main vein dysplasia (ETMVD) aged 0-18 years, who were treated between 2014 and 2024 in the surgical departments of Pediatric University.

The severity of embryonic-type main vein dysplasia was determined according to the classification of D.D. Kupatadze. It should be noted that this classification is aimed at systematizing vascular changes in this pathology and is widely used by pediatric vascular surgeons for planning the correction of the vascular component. However, pathological changes in the

Table 1. The frequency of cosmetic defects of the feet and the frequency of violations of the “support” of the feet, depending on the severity of vascular pathology

Таблица 1. Частота косметических дефектов стоп и частота нарушения «опорности» стоп в зависимости от степени тяжести сосудистой патологии

Степень тяжести дисплазии магистральных вен эмбрионального типа / Degree of severity embryonic type dysplasia of the great veins		Патологические изменения стоп / Pathological changes in the feet			
		частота косметических дефектов / frequency of cosmetic defects		нарушения «опорности» стопы / violations of the “support” of the foot	
		умеренно выраженные / moderate symptoms	выраженные / expressed	средней степени / medium degree	тяжелой степени / severe degree
Легкая / Light		0	0	0	0
Синдром Клиппеля–Треноне / Klippel–Trenone syndrome	Средней тяжести / Moderate severity	9 (12,3%)	0	3 (4,1%)	0
	Тяжелая / Severe degree	5 (6,2%)	17 (23,3%)	21 (28,8%)	5 (6,2%)
	Крайне тяжелая / Extremely severe degree	0	7 (9,6%)	0	7 (9,6%)

Table 2. Severity of pathology in children with embryonic type of great vein dysplasia, depending on gender and clinical picture of Klippel–Trenaunay syndrome

Таблица 2. Степени тяжести поражения у детей с эмбриональным типом дисплазии магистральных вен, в зависимости от пола и клинической картины синдрома Клиппеля–Треноне

Степень поражения / Degree of damage		Клиническая картина / Clinical picture	Пол / Gender	
Легкая / Light		1. «Гладкие» сосудистые пятна. / “Smooth” vascular spots. 2. Атипичные, эмбриональные вены (в пределах сегментов конечности). / Atypical, embryonic veins (within the segments of the limb).	м	ж
Клиническая картина синдрома Клиппеля–Треноне / Clippel–Trenaunay Syndrome clinical picture	Средняя, 3, 4, 5, 6 — дополнительно к предыдущим симптомам / Average 3, 4, 5, 6 — in addition to the previous symptoms	3. Симметричное увеличение окружности конечности / Symmetrical increase in limb circumference. 4. Несимметричная гипертрофия. / Asymmetrical hypertrophy. 5. Бугристые сосудистые пятна. / Tuberous vascular spots. 6. Местный синдром диссеминированного внутрисосудистого свертывания. / Local disseminated vascular coagulation syndrome. Соответствует клинической картине синдрома Клиппеля–Треноне / Corresponds to the clinical picture of Klippel–Trenaunay syndrome	7	4
	Тяжелая, 7, 8, 9, 10, 11, 12, 13, 14 — дополнительно к предыдущим симптомам / Severe 7, 8, 9, 10, 11, 12, 13, 14 — in addition to the previous symptoms	7. Гигантизм стоп, макрокродактилия. / Foot gigantism, macrodactyly. 8. «Элефантиаз». / “Elephantiaz”. 9. Контрактуры. / Contractures. 10. Нарушения функции конечности. / Limb function disorders. 11. Поражение тканей двух конечностей и более. / Damage to the tissues of two or more limbs. 12. Эмбриональные вены распространяются на всю конечность. / Embryonic veins spread to the entire limb. 13. Поражение сосудов органов малого таза. / Damage to the vessels of the pelvic organs. 14. Распространенный синдром диссеминированного внутрисосудистого свертывания. / A common disseminated vascular coagulation syndrome. Соответствует клинической картине синдрома Клиппеля–Треноне / Corresponds to the clinical picture of Klippel–Trenaunay syndrome	16	14
	Крайне тяжелая, 15, 16, 17, 18, 19 – дополнительно к предыдущим симптомам / Extremely severe 15, 16, 17, 18, 19 — in addition to the previous symptoms	15. Уродующая гипертрофия конечности. / Disfiguringlimb hypertrophy. 16. «Венозные соты» на голени. / “Venous honeycombs” on the lower leg. 17. Головокружение, коллаптоидные состояния, потеря сознания после кратковременной ходьбы без эластичных бинтов. / Dizziness, collaptoid states, loss of consciousness after short-term walking without elastic bandages. 18. Мелена и гематурия. / Melena and hematuria. 19. Кисты и лимфангиомы брюшной: полости и забрюшинного пространства. / Cysts and lymphangiomas of the abdominal cavity and retroperitoneal space. Соответствует клинической картине синдрома Клиппеля–Треноне / Corresponds to the clinical picture of Klippel–Trenaunay syndrome	5	2

limbs are described schematically in this classification and are not fully characterized, which limits its use for assessing orthopedic disorders.

Thus, in our study, a detailed analysis of foot pathology was performed in patients with each degree of disease severity to develop prospective plans for the scope and stages of surgical orthopedic correction.

Of 73 patients, 11 children had mild ETMVD (10.9±1.7 years), 25 children had moderate ETMVD (13.0±0.65 years), 30 children had severe ETMVD (10.2±0.7 years) and 7 children had extremely severe ETMVD (8.9±2.4 years).

In Table 2, the patients are distributed according to the D.D. Kupatadze classification depending on the severity of EMTVD and sex. The severity levels of EMTVD corresponding to the clinical presentation of Klippel–Trenaunay syndrome are also highlighted.

The circumference of the forefoot was measured using a tape at the level of the metatarsophalangeal joints. Foot length was assessed using the Fridland method: the foot was placed on a clean sheet of paper, and its contours were outlined with a pencil in a vertical position. Then, the foot length was measured along the contour from the tip of the toes to the posterior end of the heel using a ruler or measuring tape.

When analyzing the results, not only the difference between the healthy and affected foot was considered, but the asymmetry coefficient (AC) was also calculated:

$$AC = (L2 – L1) \cdot 100 / L1,$$

where L1 is the length of the affected foot (the larger measurement); L2 is the length of the normal foot (the smaller measurement). In addition to length measurement, the contourgram made it possible to determine the enlargement of individual rays and to draw conclusions about the form of foot involvement. The height of the longitudinal arch of the foot, defined as the vertical distance from the floor to the highest point of the plantar surface (area of the navicular bone), was used to assess the condition of the arch. Then, the Friedland podometric index (FPI) was calculated:

$$FPI = (\text{foot height} \cdot 100) / \text{foot length}.$$

FPI interpretation:
- more than 31 — high arch;
- from 31 to 29 — normal arch;
- from 29 to 27 — lowered arch;
- from 27 to 25 — flatfoot;
- less than 25 — pronounced flatfoot.

These measurements were obtained in adolescents with completed foot growth and without pronounced excess soft tissue in the plantar area. In preschool- and primary school-age children, normative data for arch height do not exist. All measurements were performed in the standing position with weight. The determination of hindfoot pronation (heel pronation) was carried out as follows: an axis was drawn along the posterior surface of the leg through the midline of the Achilles tendon to the middle of the calcaneal tuberosity. Deviation of the axis outward from the vertical line indicated the angle of pronation of the hindfoot, i.e., the heel. Under normal conditions, the heel axis coincides with the plumb line drawn through the middle of the Achilles tendon.

Instrumental methods of orthopedic examination

To diagnose foot pathology, different instrumental methods were used. Radiography was performed using Philips CombiDiagnost R90 (Germany). By allowing the assessment of the bone structures of the foot of the affected limb, radiography helped to identify the presence of true hypertrophy. The study was conducted on symmetrical areas of the both limbs in order to perform a comparative analysis.

The vessels were examined using a GE Innova 3100 (USA). Phlebography was necessary for a comprehensive examination of the main veins in children. Fig. 1 shows the pathological venous bed of the limb.

Ultrasound examination of the vessels and soft tissues was performed using a Philips Affiniti 50 apparatus (Netherlands). An ultrasound image of the pathological changes in the vein bed is presented in Fig. 2.

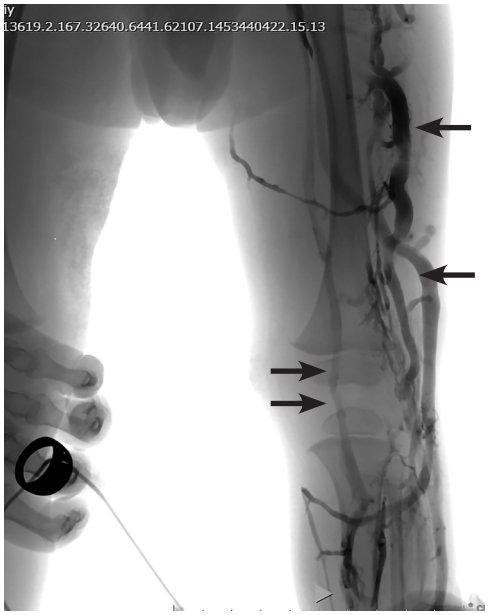

Fig. 1. Phlebography of the left lower limb of patient Ch., 1 year 8 months; Diagnosis: embryonic type dysplasia of the great veins of the left lower extremity. The lateral embryonic vein is indicated by single arrows; the hypoplasia of the popliteal segment of the deep femoral vein is indicated by a double arrow

Рис. 1. Флебография левой нижней конечности пациента Ч., 1 год 8 месяцев. Диагноз: дисплазия магистральных вен эмбрионального типа левой нижней конечности. Латеральная эмбриональная вена обозначена одинарными стрелками; гипоплазия подколенного сегмента глубокой бедренной вены обозначена двойной стрелкой

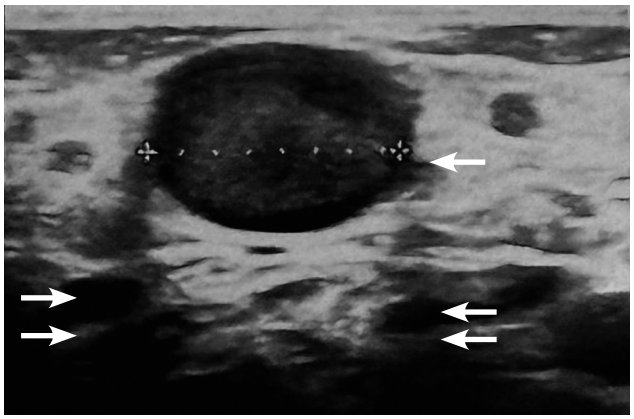

Fig. 2. Ultrasound of the veins of the upper third of the lower leg, patient Ch., 10 years old. Diagnosis: embryonic type dysplasia of the great veins of the left lower extremity. The lateral embryonic vein is indicated by a single arrow; the dilated tributaries of the embryonic vein are indicated by double arrows

Рис. 2. Ультразвуковое исследование вен верхней трети голени, пациент Ч., 10 лет. Дисплазия магистральных вен левой нижней конечности эмбрионального типа. Латеральная эмбриональная вена обозначена одинарной стрелкой; дилатированные притоки эмбриональной вены обозначены двойными стрелками

Klippel–Trenaunay syndrome may be diagnosed after birth according to phenotypic signs. To confirm the diagnosis, patients underwent ultrasound examination of the lower limb vessels and venography. The main pathological changes in the surface veins were determined during ultrasound examination, including an excessively pronounced and tortuous venous network with varicose changes and the presence of embryonic lateral veins. In the deep veins, hypoplasia of the popliteal segment of the femoral vein of varying severity was found; similar data are reported in the literature [11].

Phlebography of the affected limb was performed in all patients. The main aim of this examination was to confirm the diagnosis. Additionally, the deep veins were assessed in order to plan potential surgical operations involving the superficial veins, the lateral embryonic vein and the deep veins. The literature also reports the use of these methods for clarification and preoperative planning [12–14].

Statistical analysis

The normality of the distribution of variables was assessed using kurtosis and skewness indices, which characterize the shape of the Gaussian distribution curve. Parametric statistical methods, including the calculation of Student's t-test, were used to process the results, as the distribution of values in the samples was close to normal.

For variables with a continuous distribution, the arithmetic mean (M) and the standard error of the mean (m) were calculated, whereas for variables with discrete values, the frequency of occurrence was determined.

For correlation analysis, Pearson correlation coefficient was used, taking into account its absolute value and the level of statistical significance, which depends on the sample size and allows the assessment of sufficiently strong correlations.

Data processing was performed using STATISTICA 6.0 and Microsoft Excel 2007.

RESULTS AND DISCUSSION

During the study, pathological changes in the feet of children with various degrees of EMTVD were analyzed: mild, moderate, severe, and extremely severe.

Foot changes in mild embryonic-type main vein dysplasia

Eleven children with mild EMTVD were examined. In 6 adolescents (13 years old – 1 individual, 14 years

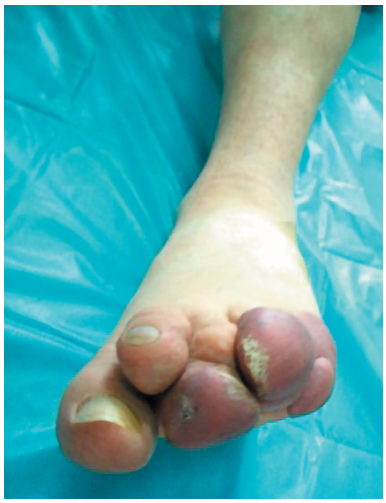

Fig. 3. Patient A., 8 years old, diagnosis: in embryonic type dysplasia of the great veins of the lower extremities moderate severity. Increase in length and circumference of toes III, IV, and V

Рис. 3. Пациент А., 8 лет, с диагнозом: дисплазия магистральных вен эмбрионального типа средней степени тяжести. Увеличение длины и окружности III, IV и V пальцев стопы

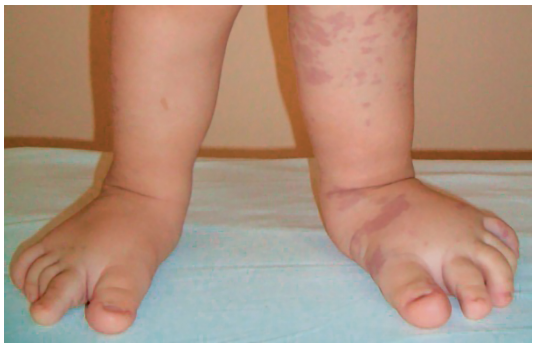

Fig. 4. Patient B., 4 years old. Diagnosis: in embryonic type dysplasia of the great veins of the lower extremities moderate severity. In patients with moderate severity, an increase in the forefoot was recorded with an increase in the circumference and length of the fingers

Рис. 4. Пациент Б., 4 года. Диагноз: дисплазия магистральных вен нижней конечности эмбрионального типа, средней степени тяжести. У пациента со средней степенью тяжести зафиксировано увеличение переднего отдела стопы, окружности и длины пальцев

old – 2 individuals, 15 years old – 2 individuals, and 16 years old – 1 person), a decrease in the height of the longitudinal arch of the foot was revealed. In the remaining five cases, no foot pathology was observed.

Foot changes in moderate embryonic-type main vein dysplasia (Klippel–Trenaunay syndrome)

Twenty-five children with moderate Klippel–Trenaunay syndrome were examined. Among them:

- in 12 adolescents, a decrease in the height of the longitudinal arch of the foot was revealed;
- in 4 patients, a valgus foot position was found;
- in 5 patients, an increase in the length and circumference of one or more toes was noted (Fig. 3);
- in 3 patients, an enlargement of the forefoot with an increase in the circumference and length of the toes was recorded (Fig. 4);
- in one child, pachydermodactyly was registered — an increase in skin thickness without a significant change in the thickness of the subcutaneous fat.

Foot changes in severe embryonic-type main vein dysplasia (Klippel–Trenaunay syndrome)

Thirty patients with severe Klippel–Trenaunay syndrome (severe ETMVD) were examined.

In children with severe Klippel–Trenaunay syndrome, pathological changes in the feet were more pronounced:

- a decrease in the height of the longitudinal arch of the affected foot was found in 4 adolescents;
- a valgus foot position was detected in 2 patients;
- in 7 children, an enlargement of the forefoot combined with a uniform increase in the toe circumference was observed (see Fig. 3);
- in 5 patients, an enlargement of individual rays of the foot was recorded (Fig. 5);

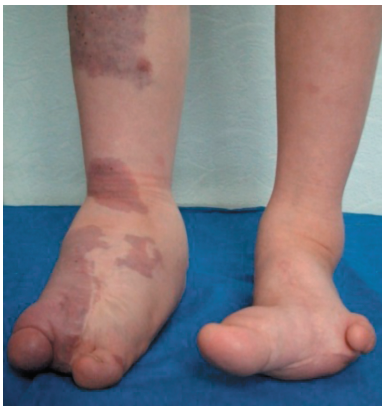

Fig. 6. Patient G., 7 years old. Diagnosis: in embryonic type dysplasia of the great veins of the lower extremities severe severity. Splitting the left foot

Рис. 6. Пациент Г., 7 лет. Диагноз: дисплазия магистральных вен нижней конечности эмбрионального типа тяжелой степени. Вариант расщепления левой стопы

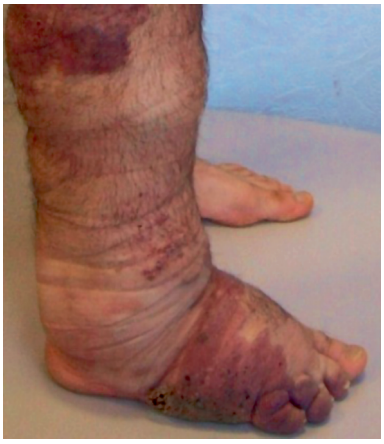

Fig. 7. Patient N., 11 years old. Diagnosis: in embryonic type dysplasia of the great veins of the lower extremities severe severity. Increase in the circumference of all parts of the right foot, with a slight increase in length and circumference of the fingers

Рис. 7. Пациент Н., 11 лет. Диагноз: дисплазия магистральных вен нижней конечности эмбрионального типа тяжелой степени. Увеличение окружности всех отделов правой стопы с незначительным увеличением длины и увеличением окружности пальцев

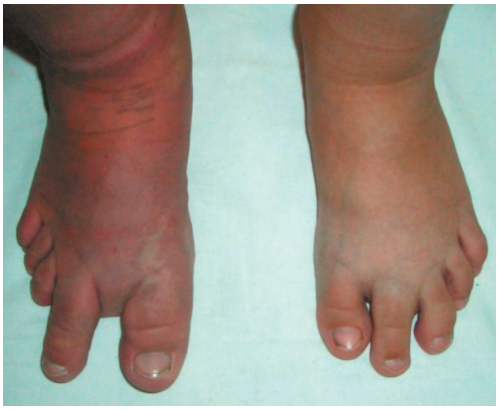

Fig. 5. Patient A., 5 years old. Diagnosis: in embryonic type dysplasia of the great veins of the lower extremities severe severity. Enlargement of rays I and II of the right foot

Рис. 5. Пациент Г., 5 лет. Диагноз: дисплазия магистральных вен нижней конечности эмбрионального типа тяжелой степени. Увеличение I и II лучей правой стопы

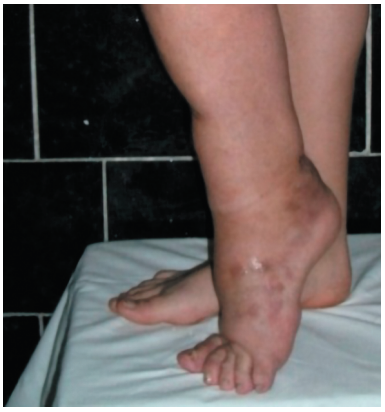

Fig. 8. Patient K., 13 years old. Diagnosis: in embryonic type dysplasia of the great veins of the lower extremities severe severity. Equinus contracture of the left foot

Рис. 8. Пациент К., 13 лет. Диагноз: дисплазия магистральных вен нижней конечности эмбрионального типа тяжелой степени. Эквинусная контрактура левой стопы

- in 4 patients, an increase in the toe circumference was noted, mainly due to vascular malformations of the soft tissues;
- in 2 children, an enlargement of individual rays of the foot combined with clinodactyly was revealed;
- in one patient, a deformity resembling a split-foot type was found (Fig. 6);
- in 3 patients, an increase in the circumference of all parts of the foot with a slight increase in length, as well as an increase in the circumference of all or several toes, was noted (Fig. 7);
- equinus contracture of the affected foot was detected in 2 children (Fig. 8).

Foot changes in extremely severe embryonic-type main vein dysplasia (Klippel–Trenaunay syndrome)

Seven patients with extremely severe Klippel–Trenaunay syndrome (extremely severe EMTVD) were examined. In this group, foot pathology was most pronounced:

- in the region of the dorsal and plantar surfaces of the feet, an excessive volume of soft tissue was observed, including subcutaneous fat and vascular malformations; the soft tissues of the foot were distributed unevenly;
- uneven enlargement of all parts of the foot combined with total syndactyly of the II, III, and IV toes was found in 1 patient (Fig. 9);
- a «spherical» shape of the foot of the affected limb was noted in 3 patients (Fig. 10);
- equinus contracture of the foot was detected in 3 patients, and the contracture was rigid in nature.

The frequency of identified foot changes in patients with EMTVD is presented in Table 3.

It was found that the circumference of the lower leg, thigh lengh and total limb length showed statistically significant positive correlations with the severity of Klippel–Trenaunay syndrome ($p \leq 0.001$). Also, lower leg length, thigh circumference, foot length and circumference were positively correlated with disease severity ($p \leq 0.01$). No statistically significant association was found between patient age and the severity of the syndrome ($r=0.13$, $p > 0.05$).

CONCLUSION

1. Thirteen types of pathological foot changes were identified;
2. Indications for surgical treatment were established in children with severe (21) and extremely severe (7) Klippel–Trenaunay syndrome.
3. Lower leg length, thigh circumference, and foot length and circumference showed significant positive correlations with disease severity ($p \leq 0.01$).
4. No statistically significant association was found between syndrome severity and patient age ($r = 0.13$, $p > 0.05$).
5. The differentiation of pathological changes depending on the severity of cosmetic and functional changes in the feet contributes to optimizing the choice of orthopedic surgical correction in the future.

Our study revealed a diversity of pathological changes in the feet of children with different severity levels of Klippel–Trenaunay syndrome. Statistically significant correlations were found between foot length and circumference, as well as between lower leg length, thigh circumference, lower leg circumference, thigh length, total limb length, and syndrome severity.

Based on the obtained results, a classification of pathological changes according to the severity of cosmetic and functional foot disorders has been proposed. This approach may serve as a guide for determining the indications and surgical strategy for future orthopedic correction.

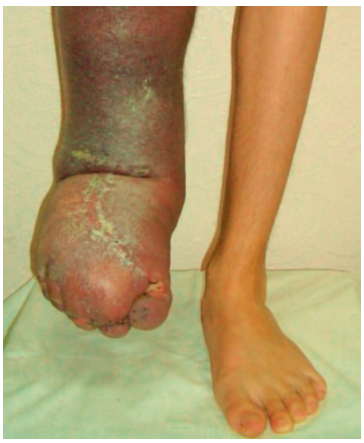

Fig. 9. Patient S., 3 years old. Diagnosis: in embryonic type dysplasia of the great veins of the lower extremities extremely severe severity. Uneven enlargement of all parts of the right foot with a total form

Рис. 9. Пациент С., 3 года. Диагноз: дисплазия магистральных вен нижней конечности эмбрионального типа крайне тяжелой степени. Неравномерное увеличение всех отделов правой стопы с тотальной формой

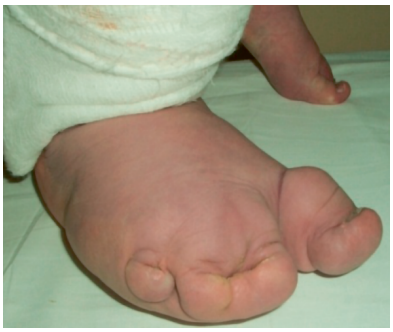

Fig. 10. Patient S., 15 years old. Diagnosis: in embryonic type dysplasia of the great veins of the lower extremities extremely severe severity. “Ball-shaped” form of the right foot

Рис. 10. Пациент Т., 15 лет. Диагноз: дисплазия магистральных вен нижней конечности эмбрионального типа крайне тяжелой степени. «Шарообразная» форма правой стопы

Table 3. Frequency of pathological changes in the feet of children with dysplasia of the great veins of the lower extremities of varying severity lippel–Trenone syndrome

Таблица 3. Частота патологических изменений стоп у детей с дисплазией магистральных вен нижней конечности эмбрионального типа различной степени тяжести Синдром Клиппеля–Треноне

Характер изменений / The nature of the changes	Степень тяжести дисплазией магистральных вен нижней конечности эмбрионального типа / Severity of embryonic type dysplasia of the main veins of the lower extremity			
	легкая / light	средняя / average	тяжелая / severe	крайне тяжелая / extremely severe
Снижение высоты продольного свода стопы / Lowering the height of the longitudinal arch of the foot	6 (8,2%)	12 (16,4%)	4 (5,4%)	*
Вальгусная установка пораженной стопы / Valgus of the affected foot	0	4 (5,4%)	2 (2,7%)	0
Увеличение длины и окружности одного или нескольких пальцев / Increasing the length and circumference of one or more fingers	0	5 (6,2%)	0	0
Пахидермодактилия / Pachydermodactyly	0	1 (1,4%)	0	0
Увеличение переднего отдела стопы с увеличением окружностей пальцев / Enlargement of the forefoot with an increase in the circumference of the fingers	0	3 (4,1%)	7 (9,6%)	0
Увеличение отдельных лучей / Magnification of individual beams	0	0	5 (6,2%)	0
Увеличение окружности пальцев за счет сосудистых мальформаций / Increased finger circumference due to vascular malformations	0	0	4 (5,4%)	0
Увеличение отдельных лучей в сочетании с клинодактилией / Magnification of individual rays in combination with clinodactyly	0	0	2 (2,7%)	0
Расщепленная стопа / Split foot	0	0	1 (1,4%)	0
Увеличение всех отделов стопы с незначительным увеличением длины и окружности пальцев / Enlargement of all parts of the foot with a slight increase in the length and circumference of the toes	0	0	3 (4,1%)	0
Эквинусная контрактура / Equinus contracture	0	0	2 (2,7%)	3 (4,1%)
Неравномерное увеличение всех отделов стопы с синдактилией нескольких пальцев / Uneven enlargement of all parts of the foot with syndactyly of several fingers	0	0	0	1 (1,4%)
«Шарообразная» форма стопы / The «spherical» shape of the foot	0	0	0	3 (4,1%)

* It was not possible to evaluate the height of the longitudinal arches of the feet in patients with extremely severe vascular pathology due to the excess volume of soft tissue in the plantar surface area.

* Провести оценку высоты продольных сводов стоп у пациентов с крайне тяжелой степенью сосудистой патологии не представлялось возможным в связи с избыточным объемом мягких тканей в области подошвенной поверхности.

ADDITIONAL INFORMATION

Author contribution. All authors made a substantial contribution to the conception of the study, acquisition, analysis, interpretation of data for the work, drafting and revising the article, final approval of the version to be published and agree to be accountable for all aspects of the study.

Competing interests. The authors declare that they have no competing interests.

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Ethical expertise. The protocol for examination and treatment of children was approved by the local ethics committee.

Consent for publication. Consent of patients (their representatives) for the processing and publication of personal data has been obtained.

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Источник финансирования. Исследование не имело спонсорской поддержки.

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