ISSN 0030-5987. Orthopaedics, traumatology and prosthetics. 2021. № 3

УДК 616.718.4+616.718.5/.6:616.147.3

DOI: http://dx.doi.org/10.15674/0030-59872021349-53

Orthopedic manifestations of congenital vascular malformations of the lower extremities

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The spectrum of orthopedic manifestations of congenital vascular malformations of lower limbs remains insufficiently studied. Objective. To analyze the results of examination of patients with orthopedic manifestations of congenital vascular malformations. Methods. 24 patients with various forms of congenital vascular malformations of the lower extremities were examined. Distribution of patients according to the Hamburg classification of vascular malformations: arteriovenous type — 14 patients; venous — 6; capillary — 2; capillary-venous — 1; lymphatic — 1 patient. The diagnosis of orthopedic pathology was performed on the basis of clinical examination, X-ray, ultrasound, X-ray densitometry. Results. All patients with congenital vascular malformations were diagnosed with orthopedic pathology: leg length discrepancy (75 %), secondary scoliotic deformity (16 %), knee flexion contracture 40° (8 %), flatfoot (12 %), equinus deformity of the foot (8 %), osteoporosis of the affected limb (16 %), areas of pathological reconstruction of the affected bone (lytic areas) (4 %), hip osteoarthritis of the affected limb (grade 2) (4 %). Leg length discrepancy was observed due to the elongation of the affected limb — in 16 patients (67 %), shortening — in 2 (8 %). Leg length discrepancy was in the range of 0.5–6 cm. Elongation of the affected limb < 2 cm was noted in 5 patients (21 %), > 2 cm — in 11 patients (46 %); shortening of the affected limb < 2 cm was observed in one patient (4 %), > 2 cm — in one patient (4 %). Among the patients with leg length discrepancy, 61 % had combinations of several orthopedic manifestations of the disease. Conclusions. Patients with congenital vascular malformations have a wide range of orthopedic pathology, among them the leading is leg length discrepancy (75 %) due to elongation (67 %) or shortening of the affected limb (8 %). Leg length discrepancy is most often observed in patients with arteriovenous type of vascular malformation (67 %), elongation of the lower limb > 2 cm (75 %) significantly predominating in this category. Among patients with a difference in the length of the lower limbs in 61 % of cases, there is a combination with other orthopedic manifestations of the disease. Systematization of orthopedic pathology and development lesions' classification of the musculoskeletal system taking into account the forms of vascular malformation will improve the diagnosis of the disease and improve the tactics of its orthopedic treatment.
Key words. Congenital vascular malformations, orthopedic manifestations, leg length discrepancy.

Спектр ортопедичних проявів, які супроводжують уроджені судинні мальформації (УСМ) нижніх кінцівок, залишається недостатньо вивченим. Мета. Проаналізувати результати обстеження пацієнтів з ортопедичними проявами уроджених судинних мальформацій. Методи. Обстежено 24 пацієнти віком від 5 до 39 років із різними формами УСМ нижніх кінцівок, а саме за Гамбургською класифікацією судинних мальформацій: артеріовенозна форма — 14 осіб, венозна — 6, капілярна — 2, капілярно-венозна — 1, лімфатична — 1. Діагностику ортопедичної патології проводили на підставі клінічного огляду, рентгенологічного, ультразвукового, рентген-денситометричного обстеження. Результати. У всіх пацієнтів із УСМ діагностовано ортопедичну патологію: різницю довжини нижніх кінцівок (75 %), вторинну сколіотичну деформацію хребта (16 %), згинальну контрактуру в колінному суглобі 40° (8 %), плоскостопість (12 %), еквінусну деформацію стопи (8 %), остеопороз кісток (16 %) і деформівний артроз кульшового суглоба II ст. (4 %) ураженої кінцівки, ділянки патологічної перебудови ураженої кістки (літичні ділянки) (4 %). Різниця довжини нижніх кінцівок (0,5–6 см) через подовження ураженої кінцівки була в 16 пацієнтів (67 %), укорочення — у 2 (8 %). Подовження враженої кінцівки до 2 см констатовано у 5 хворих (21 %), понад 2 см — в 11 (46 %); укорочення до 2 см відмічено в 1 пацієнта (4 %), понад 2 см — в 1 (4 %). Серед хворих із різницею довжини нижніх кінцівок у 61 % випадків виявлено поєднання декількох клініко-ортопедичних проявів захворювання. Висновки. У пацієнтів із УСМ визначено широкий спектр ортопедичної патології, серед якої провідною є різниця довжини нижніх кінцівок (75 %). Її найчастіше спостерігали в разі артеріовенозної форми УСМ (67 %). Різниця довжини нижніх кінцівок у 61 % випадків поєднувалася з іншими клініко-ортопедичними проявами захворювання. Розроблення класифікації ураження опорно-рухової системи з урахуванням форм судинної мальформації дозволить покращити діагностику захворювання та удосконалити тактику його ортопедичного лікування.

Key words. Congenital vascular malformations, orthopedic manifestations, leg length discrepancy

Introduction

Congenital vascular malformations (CVM) are structural abnormalities of vascular development that occur during embryonic angiogenesis as a result of incomplete resorption of primary blood vessels and complex action of teratogenic factors that lead to various forms of regional blood flow disorders, severe anatomical and functional changes and early disability of patients [1–3].

The frequency of congenital vascular disorders is 1.2 %, of which 55 % are abnormalities affecting lower extremities [4].

Clinical manifestations of the disease are extremely diverse and are characterized by polymorphism and polyorganic nature of the abnormality. One of the most common is the impact on the development of the musculoskeletal system, namely: impaired longitudinal growth of limb bones (acceleration or deceleration), resulting in differences in their length, formation of axial deformities, joint contractures, osteopenia and osteoporosis with abnormal changes. According to various information, the development of orthopedic disorders in this group of patients is up to 84 % [5, 6].

The range of orthopedic manifestations that accompany CVM remains insufficiently studied. In the practice of orthopedicians there are some difficulties in diagnosing and correcting existing changes in the musculoskeletal system, associated with the course of CVM, a variety of forms, variability of clinical manifestations and the progressive nature of the disease.

Against the background of all the achievements in the study of this abnormality and the variety of scientific publications, issues about the state of the musculoskeletal system and its changes in the CVM remain uncertain. In the orthopedic literature on the issue of this disease there are singular non-systematic descriptive reports [7, 8]. All the above determines the relevance of the study.

The purpose of the study: to analyze the results of examination of patients with orthopedic manifestations of congenital vascular malformations of the lower extremities.

Material and methods

The study is based on the assessment of the results of the examination of 24 patients with various forms of CVM of the lower extremities, who applied to the State Institution «Professor M. I. Sytenko Institute of Abnormalities of the Spine and Joints of the National Academy of Medical Sciences of Ukraine» from 2010 to 2021. The age of patients ranged from 5 to 39 years (mean age — 15.5 years). There were 11 male and 13 female patients.

All patients were previously consulted by vascular surgeons, the diagnosis of CVM was established on the basis of clinical and instrumental methods of examination (Doppler ultrasound, angiography, etc.).

CVM form was determined by the Hamburg classification (1988), according to which the distribution of patients was as follows: arteriovenous form (AVM) in 14, venous (VM) in 6, capillary (CM) in 2, capillary-venous (CM-VM) in 1, lymphatic (LM) in 1 subject.

Orthopedic abnormalities were diagnosed on the basis of clinical examination, radiological, ultrasound (US), X-ray densitometric examinations. The study involved evaluation of patients' gait, length of the lower extremities, presence of deformities ofthe spine and extremities and joint contractures. Radiographic study was performed in anterior-posterior and lateral projections depending on the location of the orthopedic disorder. Patients with joint damage underwent ultrasound examination. The state of the lumbar spine and proximal femur was assessed by an X-ray densitometry.

The materials of the study were considered and approved by the local Committee on Bioethics at the State Institution «Professor M. I. Sytenko Institute of Abnormalities of the Spine and Joints of the National Academy of Medical Sciences of Ukraine» (Protocol No. 2 of 05.04.2021).

Results and their discussion

All patients with CVM of the lower extremities were diagnosed with orthopedic disorders: the difference in the length of the lower extremities was found in 18 (75 %), secondary scoliotic deformity of the spine in 4 (16 %), flexion contracture in the knee joint (40°) in 2 (8 %), flat feet in 3 (12 %), equinus deformity of the foot in 2 patients (8 %).

X-ray examination showed osteoporosis of the bones of the affected limb in 4 (16 %), areas of abnormal reconstruction of the affected bone (lytic areas) in 1 (4 %), deforming osteoarthritis of the hip stage II in patient 1 (4 %). During US examination of the knee joint, one patient was diagnosed with chronic proliferative synovitis. Four patients (16 %) were diagnosed with osteoporosis of the affected limb by X-ray densitometric examination of the lumbar spine and proximal femur (bone mineral density; T- or Z-test),

which in one case resulted in a pathological fracture of the femur. Besides, one patient (4 %) was diagnosed with pathological reorganization (area of lysis) of the femoral neck with the formation of symptomatic coxa vara, which is the risk of pathological fracture. Clinical examples of orthopedic disorders are shown in Fig. 1 and 2.

The difference in the length of the lower extremities in the range of 0.5–6 cm was stated due to elongation of the affected limb in 16 (67%), shortening in 2 patients (8%). Elongation of the affected limb to 2 cm was recorded in 5 patients (21%), more than 2 cm in 11 (46%); shortening of the affected limb to 2 cm was observed in 1 (4%), more than 2 cm in 1 patient (4%).

The distribution of patients with orthopedic disorders depending on the form of CVM is given in the table. The difference in the length of the lower extremities was most often observed in patients with arteriovenous CVM (12-67%), with the predominance of individuals (9-75%) with elongation of the lower extremity over 2 cm.

Among 18 patients with a difference in the length of the lower extremities in 11 (61%) there was a combination of several clinical and orthopedic manifestations (with knee contracture in 2, osteoporosis in 3, scoliotic spinal deformity in 2, bone lysis in 1, proliferative in 1, deformable osteoarthritis of the hip in 1, flat feet in 1 patient).

Discussion

When discussing the results, it should be noted that the reasons for the development of orthopedic

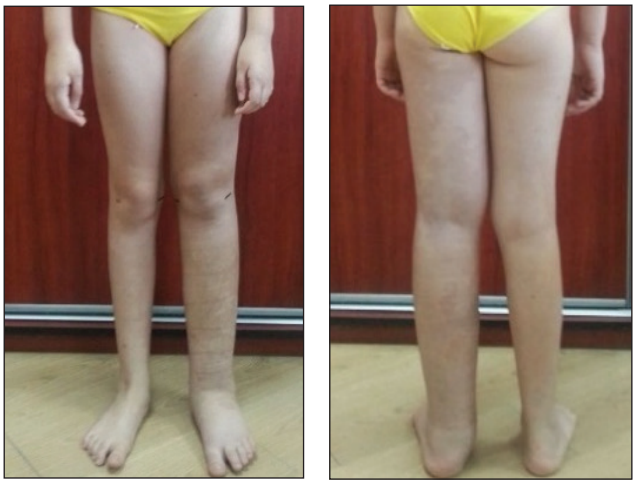

Fig. 1. Appearance of an 11-year-old patient D. Diagnosis: CVM of the left lower extremity, venous form, elongation of the left lower extremity by 3.5 cm

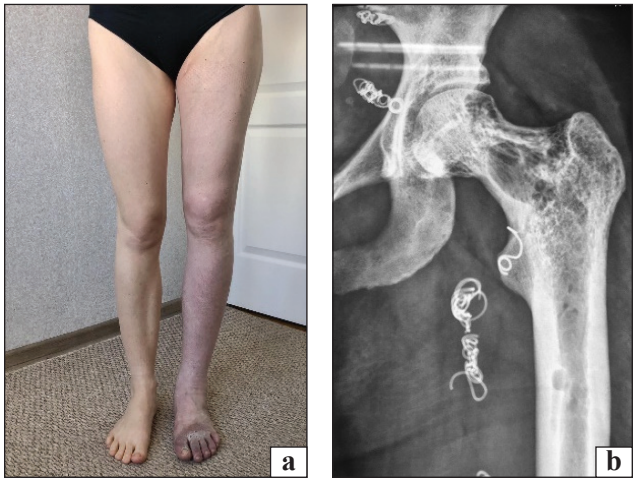

Fig. 2. A 31-year-old patient E. Diagnosis: CVM of the left lower extremity, arteriovenous microfistular form: a) appearance; b) radiography of the left hip joint in direct projection, abnormal focus of lysis of the femoral neck, *coxa vara*

Table

The number of patients with orthopedic disorders depending on CVM form

Orthopedic disorder		CVM form				
		AVM	VM	CM	CM-VM	LM
Elongation of the limb, cm	< 2	1	2	1	1	—
	> 2	9	1	1	—	—
Shortening of the limb, cm	< 2	1	—	—	—	—
	> 2	1	—	—	—	—
Scoliotic spinal deformity		2	1	1	—	—
Flatfoot		1	1	—	—	1
Contracture of the knee joint		1	1	—	—	—
Equinus deformity of the foot		1	1	—	—	—
Osteoporosis		2	2	—	—	—
Areas of bone lysis		1	—	—	—	—
Deforming osteoarthritis of the hip stage II		1	—	—	—	—
Proliferative synovitis of the knee joint		1	—	—	—	—

disorders in CVM remain not fully understood. According to various authors, this abnormality occurs due to the influence of hemodynamic, metabolic and mechanical factors, although there are some differences in the pathogenic role of some of them in different forms of CVM [9–11]. Despite the presence of similar abnormal processes that occur in CVM, patients with the same forms of the disease have different orthopedic manifestations, which indicates the variability and predominance of certain changes in each individual case.

It should be emphasized that the study of the features of orthopedic disorders in CVM allows to develop tactics for their treatment. It should be noted that the choice of methods for the treatment of orthopedic disorders in CVM depends primarily on the underlying disease (form of malformation, its location and spread). Considering the difference in the length of the lower extremities as the most common orthopedic manifestation of CVM, we note that the presence of a difference in length over 2 cm is an indication for surgical correction [12]. Patients with CVM often have a combination of several orthopedic manifestations that affect the ability of the affected limb. So, there is a key question: in which cases is it necessary to correct the difference in the length of the lower extremities by surgery as the only orthopedic manifestation of the disease and what if such patients have other orthopedic abnormalities? The answer to this question will improve the results of correction of orthopedic disorders in patients with CVM.

Thus, the obtained results encourage us to further research. Systematization of orthopedic manifestations of congenital vascular malformations needs to be improved. It is important to develop an orthopedic classification of the disease, which will be based on the choice of treatment tactics.

Conclusions

Patients with CVM have a wide range of orthopedic disorders, among which the leading one is the difference in length of the lower extremities (75 %) due to elongation (67 %) or shortening of the affected limb (8 %).

The difference in the length of the lower extremities is most often found in patients with arteriovenous vascular malformation (67 %), with this category being dominated by individuals with elongation of the lower extremity over 2 cm (75 %). Among patients with a difference in the length of the lower extremities in 61 % of cases there was a combination with other clinical and orthopedic manifestations of the disease.

Systematization of orthopedic disorders and development of classification of musculoskeletal abnormalities taking into account the forms of CVM will improve the diagnosis of the disease and improve the tactics of its orthopedic treatment.

Conflict of interest. The authors declare the absence of conflict of interest.

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The article was received by the editors 06.07.2021

ISSN 0030-5987. Orthopaedics, traumatology and prosthetics. 2021. № 3

ORTHOPEDIC MANIFESTATIONS OF CONGENITAL VASCULAR MALFORMATIONS OF THE LOWER EXTREMITIES

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