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FRACTURES OF LOWER LIMBS IN CHILDREN WITH *SPINA BIFIDA*

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Background. *Spina bifida* is a serious defect in the development of the spine and spinal cord. It is accompanied by several orthopedic disorders of the spine and lower limbs, including fractures of long tubular bones. In *spina bifida*, osteoporosis plays an important role in the pathogenesis of motor disorders.
Aim. The objective was to determine the patterns of occurrence and the clinical and radiological features of fractures of the long tubular bones of the lower limbs in children with sequelae of *spina bifida*.
Materials and methods. From 2006 to 2017, 544 patients with *spina bifida* were examined and treated at the Turner Research Institute for Children's Orthopedics. Clinical-neurological and radiographic methods were used. The neurosegmental level of spinal cord involvement was determined using the Sharrard classification, and the motor level was assessed according to the method proposed by Melbourne Medical University.
Results. The clinical picture of a fracture of a long tubular bone in a child with *spina bifida* has many characteristics. There was no abnormal mobility in the fracture site in 56% of cases, edema was absent in 88% of children, and pain in the fracture region was observed in only 19% of cases. The radiographic features of the atypical fracture of long tubular bones in children with sequelae of *spina bifida* included lack of a fracture line, presence of a hypertrophic periosteal reaction, and sclerosis areas at the fracture site.
Conclusion. The frequency and localization of fractures of the lower limbs in children with sequelae of *spina bifida* are determined according to the neurosegmental level. The clinical picture of fracture often differs from usual fractures by the absence of pain syndrome, edema in the fracture region, and displacement of bone fragments, which must be considered for diagnosis. The peculiarities of the clinical and radiological picture are associated with the presence of osteoporosis in this pathology due to a decrease in the motor activity level of the patients.

Keywords: *spina bifida*; fracture of the femur and the tibia; osteoporosis in children.

ПЕРЕЛОМЫ НИЖНИХ КОНЕЧНОСТЕЙ У ДЕТЕЙ С ПОСЛЕДСТВИЯМИ СПИННОМОЗГОВЫХ ГРЫЖ

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Введение. Спинномозговая грыжа — это тяжелый порок развития позвоночника и спинного мозга, сопровождающийся разнообразными ортопедическими нарушениями со стороны позвоночника и нижних конечностей, в том числе переломами длинных трубчатых костей. При спинномозговых грыжах остеопороз имеет ключевое значение в патогенезе двигательных нарушений.
Цель исследования — определить закономерности возникновения и клинико-рентгенологические особенности переломов длинных трубчатых костей нижних конечностей у детей с последствиями спинномозговой грыжи.
Материалы и методы. В ФГБУ «НИДОИ им. Г.И. Турнера» Минздрава России в период с 2006 по 2017 г. проведено обследование и лечение 544 пациентов с последствиями спинномозговых грыж. Применяли кли-

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нико-неврологический и рентгенологический методы исследования. Определяли нейросегментарный уровень поражения спинного мозга, используя классификацию Sharrard, а также двигательный уровень по методике, предложенной Мельбурнским медицинским университетом.

Результаты. Клиническая картина перелома длинной трубчатой кости у ребенка со спинномозговой грыжей имеет ряд особенностей. Патологической подвижности в месте перелома не было в 56 % случаев, отек отсутствовал у 88 % детей, а боль в области перелома мы наблюдали только в 19 % случаях. Среди рентгенологических особенностей атипичного перелома длинных трубчатых костей у детей с последствиями спинномозговых грыж выделялись: отсутствие «линии» перелома, наличие гипертрофированной периостальной реакции, а также участки склероза в месте перелома.

Заключение. Частота и локализация переломов нижних конечностей у детей с последствием спинномозговой грыжи определяются нейросегментарным уровнем. Клиническая картина перелома зачастую отличается от обычных переломов отсутствием болевого синдрома, отека в области перелома, смещения костных отломков, что необходимо учитывать при диагностике. Особенности клинико-рентгенологической картины связаны с наличием при данной патологии остеопороза вследствие снижения уровня двигательной активности пациентов.

Ключевые слова: спинномозговая грыжа; перелом бедренной и большеберцовой кости; остеопороз у детей.

Introduction

Spina bifida is a malformation of the spine, which is an incomplete closure of the neural tube in the incompletely formed neural canal and is accompanied by various orthopedic disorders of the spine and lower extremities. Static and dynamic changes and fractures of the long tubular bones often occur during treatment of lower limb deformities caused primarily by muscle imbalance due to neurotrophic disorders [1–3]. In recent decades, interest in the study of osteoporosis in pediatric patients has increased significantly [4]. The high risk of bone fractures in patients with neuromuscular diseases is widely discussed in the literature. In terms of pathogenesis, there are two variants of osteoporosis, namely, primary and secondary. In primary osteoporosis, the cause is not verified, and secondary osteoporosis develops as a result of the underlying disease. The most important factors in modeling bone tissue are muscular activity and axial loads, both of which are necessary for the normal formation of bone architectonics and preservation of bone mass. In spina bifida, osteoporosis is of key importance in the pathogenesis of motor disorders [5].

Risk factors for osteoporosis in pediatric patients with consequence of spina bifida are a decrease in motor activity, a decrease in axial load, immobilization after surgical treatment, malnutrition, and low growth rates. Immobilization of a patient or limb leads to an increase in osteoclastic activity and cytokinin production [6, 7]. Accordingly, pediatric patients with the consequences of spina bifida may have fractures which may be interpreted as pathological. As a rule, these fractures occur as a result of forced movements during the rehabilitation treatment. The X-ray pattern of the fracture in this group of patients differs in a number of characteristic features, the most important of which is the formation of excess bone callus. The presence of excess bone callus in a child with consequences of spina bifida will necessitate differential diagnostics with tumor, osteomyelitis, and/or other bone diseases [8, 9].

This study aimed to determine patterns of occurrence and the clinical and radiological aspects of fractures of the long tubular bones of the lower extremities in pediatric patients with the consequences of spina bifida.

Materials and methods

We studied 544 pediatric patients aged from the neonatal period to 17 years old, treated in the Department of Neuro-Orthopedics and Systemic Diseases in the Turner Scientific and Research Institute for Children's Orthopedics, from 2006 to 2017. Patients had orthopedic deformities of the lower extremities caused by the consequences of spina bifida. The anamnestic, clinical, and radiological study methods were used.

In taking history, information about fractures of the long tubular bones of the lower extremities was specified in all patients. During clinical examination, the neurosegmental level of the consequences of spina bifida was determined according to the classification proposed by Sharrard [10]. It is based on the assessment of the strength of the primary

Table 1

Classification of motor disorders by neurosegmental level according to Sharrard

Level of motor impairments	Muscular strength	Neurosegmental level
1	Plantarflexor strength (4–5 points)	S_2
2	Plantarflexor strength (less than 3 points) Tibial flexor strength (3 points) Strength of the extensors and/or abductor muscles of the thigh (2–3 points)	S_1–L_5
3	Thigh flexor strength (4–5 points) Tibial flexor strength (less than 3 points) Strength of the extensors and abductor muscles of the thigh (1–2 points)	L_4–L_3
4	Lack of active knee extension Thigh flexor strength (adductor muscles) (less than 2 points) Raising the pelvis (3–4 points)	L_2–L_1
5	Lack of muscle activity of the lower extremities Lack of ability to raise the pelvis above the supporting surface in a horizontal position	Thoracic

muscle groups of the lower limbs (in points), which are innervated by the corresponding neurosegment (Table 1).

Motor level was also determined in all patients, according to the method proposed by the Melbourne Medical University for pediatric patients with consequences of spina bifidas [11], which includes five levels of motor activity: level 5 represents the ability to walk no worse than peers; level 4 represents walking with ankle orthoses without additional support means, the use of a wheelchair only for long distances; level 3 represents walking within the house with the use of orthoses and additional support means and the use of a wheelchair both outside and inside the house; level 2 represents walking during treatment at school and for a limited time at home using a wheelchair when moving; and level 1 represents the complete absence of walking.

Results

In total, we observed 204 fractures of the long tubular bones in 134 of the 544 pediatric patients examined. A retrospective analysis of case histories enabled us to identify the four most common causes of fractures in pediatric patients with the consequences of spina bifida (Fig. 1).

As seen in Figure 1, the greatest number of fractures was noted during and after orthopedic treatment (41%). With conservative treatment, fractures occurred during the staged plastering of foot deformities using splints, devices, and bandages to stabilize the hip joint where the presence of subluxation or dislocation of the hip was observed. Fractures occurred in the postoperative period in surgical treatment of contractures and deformities of the lower extremities, particularly during reconstructive surgery on the hip and knee joints, as well as stabilizing surgery on the feet.

We studied the dependence of the frequency of fractures in pediatric patients with the consequences of spina bifida on various factors. Thoracic and L_1–L_2 neurosegmental levels were noted in 49 of 110 (44.5%) patients with fractures of various localization and 32 out of 84 (38.1%) pediatric patients. In patients with neurosegmental S_2 level, fractures were observed in only 3 of 79 (3.8%) cases. Therefore, we determined a direct pattern of lower incidence of fracture with lower neurosegmental levels.

We also analyzed the dependence of the frequency of the lower limb injury localization (hip/lower leg) on the neurosegmental level and found that fractures of the femoral bone were more often noted in patients with thoracic and L_1–L_2

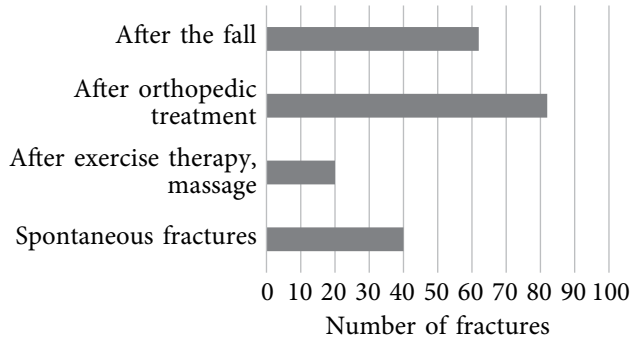

Fig. 1. Causes of fracture formation

Table 2

The distribution of patients with fractures of the long tubular bones depending on the neurosegmental level

Neurosegmental level	Total patients	Total patients with fractures	%
Thoracic	110	49	44.5
L_1–L_2	84	32	38.1
L_3–L_4	186	38	20.4
L_5–S_1	85	12	14.1
S_2	79	3	3.8

Table 3

Localization of bone fracture depending on neurosegmental level

Neurosegmental level	Femoral fracture	Tibial fracture	Total
Thoracic	65 (79.3%)	17 (20.7%)	82 (100%)
L_1–L_2	42 (76.4%)	13 (23.6%)	55 (100%)
L_3–L_4	26 (52%)	24 (48%)	50 (100%)
L_5–S_1	2 (14.3%)	12 (85.7%)	14 (100%)
S_2	–	3 (100%)	3 (100%)
Total	135 (66.2%)	69 (33.8%)	204 (100%)

Table 4

Special aspects of long tubular bone fractures in pediatric patients with the consequences of spina bifida

Symptom/sign	Typical fracture	Atypical fracture
Clinical aspects		
Pain	+	–
Post-traumatic deformity	+	–
Edema in the first h after injury	+	–
Adequacy of injury	+	–
Radiological aspects		
Fracture line	+	–
Excessive periosteal reaction	–	+
Sclerosis areas at the fracture site	–	+

neurosegmental levels in 79.3 and 76.4% of cases, respectively. In patients with L_5–S_1 neurosegmental level, fractures of the tibial bone were more common and noted in 85.7% of cases (Table 3).

Thus, the lower the neurosegmental level, the lower the incidence of femoral fractures. Taking into account the fact that in most cases, the neurosegmental level of the consequences of spina bifida determines the motor abilities of a child. Accordingly, the lower the neurosegmental level, the higher the motor ability, and we associate such features of the topical location of fractures with this fact. Thus, fractures occurred as a result of a fall during movement in 15 patients with neurosegmental levels of L_5–S_1 and S_2.

We observed 22 (16%) patients with 3 or more fractures. In 98 out of 134 (73%) pediatric patients, motor capabilities corresponded to levels

1 and 2 of motor activity. In 48 (36%) patients, fractures were observed after a long period of plaster immobilization. In 22 of 134 (16%) patients, a recurring fracture was observed in the next 6 months after the primary fracture. The fracture of the femoral bone was most frequent in its lower third.

The clinical picture of a fracture in a child with spina bifida had a number of critical aspects. First, there was often no classical history or clinical signs of damage; in 19% of cases, "spontaneous" fractures occurred without significant trauma. There was no pathological mobility at the fracture site in 56% of cases, there was no edema in 88% of pediatric patients, and we observed pain in the fracture site in only 19% of cases.

Comparative signs of typical and atypical course of the fracture are presented in Table 4.

A distinctive feature of an atypical fracture in this category of patients was also the lack of adequate injury or that the injury was not noticed by patients or parents. The absence of pain in the area of the fracture is associated with impaired sensitivity caused by underlying disease. The absence or insignificant amount of post-traumatic deformity of the limb is due to the incomplete dissociation of the bone fragments of the fracture due to low motor activity of patients. Indirect clinical signs of a fracture, as well as changes in the general blood tests (leukocytosis, thrombocytosis, and increased ESR), should be the indication for X-ray examination.

The radiological features of an atypical fracture of long tubular bones in pediatric patients with the consequences of spina bifidas include the absence of a fracture line, the presence of an excess periosteal reaction, and sclerosis at the fracture site (Fig. 2).

Hypertrophic callus was detected in 31 of 134 (23%) patients. In a retrospective analysis, it was revealed that the atypical nature of the X-ray pattern of the fracture in 10 patients required a bone biopsy to rule out osteogenic sarcoma, and in five patients, the excess callus was mistakenly interpreted as a manifestation of osteomyelitis. Knowledge of the radiological aspects of atypical fractures of the long tubular bones in patients with the consequences of spina bifidas enables us to conduct a correct differential diagnosis and develop appropriate treatment tactics.

a *b*

Fig. 2. Patient G. The diagnosis is the consequence of spina bifida. Neurosegmental level L_1–L_2. Osteoepiphysiolysis of the distal femoral epiphysis on the right: *a* — day 14 after injury; *b* —week 5 after injury, hypertrophic periosteal reaction

Conclusion

In pediatric patients with the consequences of spina bifida, various orthopedic problems manifest themselves during their lifetime. Fractures of the long tubular bones of the lower limbs in these patients become a frequent complication in the process of orthopedic treatment. This problem in the Russian and foreign literature is represented by single publications covering specific issues of diagnostics and treatment of fractures of long tubular bones. Our study confirms the high prevalence of fractures in patients with the consequences of spina bifida. It was established that their frequency and localization directly depend on the neurosegmental level, which in turn determines the level of motor activity of children. This is one of the factors in osteoporosis development. The results of the study emphasize the need for special care in order to prevent fractures in these pediatric patients at different stages of the treatment. Surgical treatment of deformities of the lower extremities should be performed while taking into account the potential motor capabilities of the child, assessing the rehabilitation potential of the patient using the Sharrard classification:

1. A varying degree of reduction in the level of motor activity and axial load on the osteoarticular apparatus is noted in pediatric patients with the consequences of spina bifida, depending on the neurosegmental level of the spinal cord lesion. This is one of the main risk factors for the development of secondary osteoporosis.

2. The frequency and localization of fractures of the long tubular bones of the lower extremities in pediatric patients with the consequences of spina bifida are directly dependent on the neurosegmental level.
3. The clinical picture of an atypical fracture of the lower extremities in a child with a consequence of spina bifida is characterized by the absence of pain syndrome, swelling in the area of the fracture, and/or displacement of bone fragments. This must be taken into account in differential diagnosis.
4. The X-ray pattern of the fracture in pediatric patients with the consequences of spina bifida is characterized by special aspects of callus formation (lack of a clear fracture line, the presence of an excessive periosteal reaction, sclerosis areas at the fracture site), which must also be considered during diagnostics and treatment.

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Ethical Review. All patients and their legal representatives agreed to the processing and publication of personal data.

Contribution of the Authors

A.C. Fedoseeva, M.A. Ugurchieva, and *T.N. Prokopenko* — collection and processing of clinical material.

V.M. Kenis and *S.V. Ivanov* wrote the results and conclusions of the article presented.

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