Lower Extremity Deformities in Children with Vertebromedullary Anomalies

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Abstract

The article presents the literature data, results of diagnostics and treatment of 219 children with various morphologies of spinal herniation and neuroorthopedic disorders. It characterizes in detail motion disorders and deforming arthrosis of the lower extremities in 88.1% and 26% of patients. The authors state that the genesis of disorders can include the manifestations of spinal cord fixation syndrome in patients with spinal hernias with combined congenital and acquired forms of latent spinal dysraphism. The article confirms the feasibility of early preventive and therapeutic measures corresponding to the nature of the combined pathology, with the participation of specialists.

Keywords: *Spinal cord anomalies, spinal hernias, joint deformities, children.*

Introduction

Spinal cord (SC) anomalies are essential in the structure of the neural tube pathologies in children. They can occur in the form of isolated anomalies or combined with other spinal malformations[12]. Today, they are usually referred to as spinal dysraphism, spinal malformations, or vertebromedullary abnormalities (VMA). SC open anomalies are manifested in the form of a spina bifida with the formation of cystic SH [10](spina bifida cystica), and closed anomalies - with spina bifida and soft tissue cleavage with spinal cord extension along the entire length of the spine or in its segment (spina bifida occulta) [Voronov V. G., 2002; 2016; A.V. Rudakova, S.N., Larionov 2012]. VMA can also occur in any spinal segment, but more often in the lumbosacral region, mainly combined with other malformations that are located, in particular, in the brain. VMA is mainly caused by lipomyelomeningocele, terminal filament lipomas, medullary cone, diastomatomyelia[7], dorsal band fixation for patients with spinal hernia, or dermal sinus tracts. It is also followed by tethered spinal cord (TSP), which term had been proposed by H. J. Hoffmann et al., (1976)[11]. With normal intrauterine development of the fetus, in the third month, the spinal cord occupies the entire length of the spinal canal. In the next months, the spine grows faster than the spinal cord. As a result, in a newborn's body, the spinal cord ends at the level of the third lumbar vertebra[5], and by the age of 12-18 months it reaches only the lower edge of the LI or upper edge of the LII vertebra[8]. When the cone and epiconus of the spinal cord are located below the normal level and in the presence of corresponding clinical symptoms, the pathological component is considered tethered cord syndrome (TCS). In a series of 5.5 thousand pediatric patients, the frequency of TCS was equal to 0.1 % [Fariborz Samini et al., 2014]. The pathophysiological basis of this syndrome is mechanical traction of the caudal spinal cord, which is followed by a decrease in blood flow in the involved area, and causes ischemia and depression in the electrical activity of the spinal cord[2]. At the biochemical level, it causes inhibition of oxidative phosphorylation in the neuronal mitochondria[17]. Acquired forms of TCS occur after operations for SH, in children with post-traumatic or post-infectious conditions, that may include fixation and tension of the

caudal spinal cord ('spinal cord postfixation syndrome', or SCPS). [Kushel Yu. V., Zemlyansky M. Yu. 2010; Rudakova A.V. et al., 2011;]. This pathology is dynamic in its nature[3]. So, as a child grows, it prevents the age-related displacement of the spinal cord cone, and exacerbates neurological deficits[14]. The characteristic signs of this pathology include an increase in existing function disorders in a pelvic system or appearance of new ones, as well as sensory, motion and trophic manifestations with bone-articular deformities of the lower extremities[9], autonomic disorders and internal dysfunctions[15]. Up to date, there are no reliable data on the frequency and distribution of this syndrome in large localities. According to some authors, in 30-50% of pediatric patients with effects of spinal hernias, disturbances in the muscular equilibrium of paresis or paralysis of the muscles of the lower extremities lead to subluxation or dislocation in the hip joint during their whole life[1]. In children, during the load in extremities, the neutralization of deforming efforts decreases, and the joints of the corresponding spinal cord injury area become deformed[6]. With age, the type and severity of disorders, their deterioration or improvement depend on the severity and extent of the neurosegmental lesion[4]. For example, confounding static and dynamic factors without orthopedic prophylaxis aggravate deformations of the musculoskeletal system[13] (Ivanov S.V., Kenis V.M., 2013; Erol B, et all., 2005; Baindurashvili A.G. et al. 2013[16].

The aim of the study is to determine the frequency and nature of paralytic deformities of the lower extremities in children with spinal hernias.

Materials and Method

We have examined 219 pediatric patients (116 male, 103 female) with various forms of spinal hernias (SH), aged from 1 day to 18 years, and have determined the clinical and neurological manifestations of the disease. This pathology has been verified taking into account the data of functional and instrumental studies: ultrasound, MSCT, MRI of head, spinal cord and vertebral spine; electroneuromyography (ENMG) of the lower extremity muscles. The study was carried out in the Department of Elective Surgery of the Second Children's Surgical Clinical Hospital of the city of Tashkent and the Republican Specialized Scientific and Practical Medical Center of Traumatology And Orthopedics, Tashkent, Uzbekistan, in 2014-2019. The severity of the lower extremity motion disorders in children diagnosed with the peripheral link of the reflex arc has been evaluated using Modified Rankin Scale (mRS) ranging from 1 to 5. The nature and type of deformations of the body and limbs have been evaluated in accordance with generally accepted criteria.

Results

The diagnostic criterion for SH is a hernial protrusion of various sizes and shapes without neurological deficits, urological or proctological disorders with the musculoskeletal system deformations. Age distribution of hospitalized patients with SH is the following: up to 28 days - 24 (11%), up to 1 year – 113 (51.6%), from 1 to 3 years – 31 (14.2%) from 3 to 7 years - 17 (7.8%), from 7 to 12 years – 27 (12.3%), from 12 to 18 years - 7 (3.2%). Localization of pathology in the cervicothoracic region of the vertebrae has been observed in 5 (2.3%) patients; in the thoracic – 9 (4.1%), lumbar – in 76 (34.7%), lumbosacral - in 92 (42%), and sacral – 37 (16.9%). In 158 (72.0%) patients, diseases were manifested in cystic form, as a combination of hernia with teratoid and lipomatous formations (spina bifida complicate) in 32 (14.6%) patients, Chiari malformation (MK) – in 26 (11.9%), rachisis – in 3 (1.4%). Of 219 patients, the shell molds (meningocele) have been observed in 24 (10.9%) of patients, various types of meningocerebral forms - in 192 (87.7%), rachishisis - in 3 (1.4%). In the case of MK, the hernial sac contained meningoradiculocele in 13 (50%) patients and meningomyelocele in 13 (50%).

Up to date, spinal hernia or manifestations of latent spinal dysraphism are a marker for potential malformations of the central nervous and other body systems. Children should be examined in order to identify combined anomalies of the central nervous system and other organs and systems. Skin markers (lipomas, dermal sinus, hemangiomas, hypertrichoses and asymmetric gluteal folds) were signs of spinal dysraphism in 20 (6.3%) patients. In 10 (50%) patients, one or more skin stigmas were located in the zone of reticular protrusion, in 10 (50%) - in various areas along the spine. In 140 (63.9%) patients, diseases formed isolated malformations, in 79 (36.1%) mainly with meningoradiculocele - 53 (67.1%) and MK - 26 (32.9%). With the expanded applicability of special examination method, the detectability of combined abnormalities of the spinal cord, spine and other organs and systems increased. In 38 (48.1%) patients, signs of spinal cord fixation syndrome have been observed, in 41 (51.9%) - specific forms of myelodysplasia: hydromyelia - 9

(11.4%), diastematomy - 8 (10.1%), syringomyelia - 17 (21.5%), the combination of several forms - in 7 (8.8%). In some of these patients, the hidden myelodysplastic syndrome had been established long time after the surgery, at the rehabilitation therapy stage. This is due to the insufficient volume for preoperative examinations or surgery complications. Combined anomalies of other organs and systems have been revealed in 89 (40.6%) of 219 patients, of the musculoskeletal system – in 73 (82%), in the form of various vertebral disorders determined by radiation diagnostic method. In 219 patients, clinical and neurological manifestations of various forms of cystic spinal dysraphism depended on the location, size and anatomy of the defect. Their frequency was the following: cutaneous stigma - 4.5%, internal abnormalities - 14.6%, autonomic disorders - 19.2%, cranial nerve injuries - 22.4%, pelvic system dysfunctions - 76, 7%, sensitivity disorders - 53.9%, motion disorders - 88.1%, trophic-orthopedic disorders - 39.7%.

Discussion

According to MRS, in 18 pediatric patients (9.3%), motion impairment score was 4; in 119 (61.7%) – 3, in 18 (9.3%) – 1-2, in 38 (19.7%) - 0. In severe isolated hernias, abnormalities have been found in 94 (48.7%) children; in combination of SH with other spinal cord abnormalities - in 79 (40.9%); with spina bifida complicate - in 20 (10.4%). In 164 (85%) patients, abnormalities were bilateral in nature, in 29 (15%) - unilateral. Of 193 patients, 87 (45 %) had motion disorders followed by other abnormalities: 30 (34.5 %) - trophic disorders, 35 (40.2 %) - trophic disorders and joint deformities, 22 (25.3 %) - joint deformities and trophic ulcers. This is due to the fact, that in severe and combined osteoneural anomalies, spinal cord malformations increase with the corresponding increase in the frequency and severity of disorders. Motion disturbances - from the complete loss of functions or weakness of some muscle groups to an increased tone of other ones - cause deformations of different fixation and severity of the individual components. In 57 (26%) of 193 patients with motion disorders, various types of deforming arthrosis in the lower extremity joints have been observed, more often in the distal direction. Combined deformity of the hip, knee and foot joints has been observed in 4 (7%) patients; gluteal deformity - in 4 (7%) cases. In these patients, dislocation or subluxation in the hip joint has been also revealed. Knee joint pathologies, in the form of flexion contracture in the range of 150°-90°, have been observed in 2 (3.5%) patients. In 47 (82.5%) cases, polymorphic paralytic foot deformities have been revealed, in 28 (59.6%) cases - bilateral deformities with the prevailing equinus component of the heel-valgus or flat-valgus deformity. In 19 (40.4%) patients, unilateral pathology has been diagnosed. Musculoskeletal disorders combined with deformation in the lower extremity joints indicates a common pathogenetic mechanisms, aggravated by associated spine cord and vertebral anomalies. Early treatment and preventive measures can improve the patient quality of life in some way. The revealed disorders and their combinations depended on the localization and severity of myelodysplasia in the segmental innervation area of the spinal cord, to a certain extent. Of 57 patients with various deformities, 24 (42.1%) had movement disorders. In 6 (10.53%) children with a lumbar lesion due to the absence or weakness of the extensor and abductor muscles of the thigh, a subluxation or dislocation of the femur has been revealed, combined with the absence of arbitrary movements of the lower extremities and wheelchair ambulation. 18 (31.58%) patients with severe peripheral paresis (injuries to all lower extremity muscles with the knee jerk preserved and the Achilles and plantar reflexes absent, in some cases) were able to stand up with support for the upper limbs and bear weight on their knee joints, moving with crutches or other devices. 33 (57.89%) children with various feet deformations were able to move independently, even with limping.. Of 219 patients with severe and prolonged osteoneural anomalies in the thoracolumbar spine, 8 (3.6%) children with unproportioned body parts were bedridden (Fig. 4.16). The data presented indicate that static and dynamic disorders contribute to the formation of musculoskeletal system pathologies in the form of joints deformations. This occurs depending on the level and severity of the neurosegmental lesion, as a result of muscle traction and antagonist-synergist muscles imbalance. Electromyography performed in 98 (30.5%) patients have showed that the prevalence of neuromuscular disorders depended on the severity of spinal motion segment injuries. The topography of these disorders was determined by comparing the electromyography data and the scheme of segmental innervation of the lower extremity muscles, taking into account the proximal border of the spinal cord lesion. It was largely determined by the upper segment, which innervated the muscles with the complete function loss.

In such a way, we may conclude that a frequent combination of musculoskeletal and trophic disorders

combined with deformation in the lower extremity joints indicates a common pathogenetic mechanism, aggravated by associated vertebromedullary anomalies. Early treatment and preventive measures can improve the patient quality of life in some way. A comprehensive examination of patients is aimed at establishing the nature of spinal and combined pathology, revealing latent spinal dysraphism and related functional neuroorthopedic disorders. Early preventive and therapeutic measures should be carried out with the participation of specialists with the combined pathology competencies.

Conclusions

In 26% of patients diagnosed with meningeal forms of SH against the background of motion disorders, neuro-orthopedic manifestations have been observed. They are considered associated with the spinal cord fixation syndrome due to congenital or acquired latent spinal dysraphism.

Localization, nature of neuroorthopedic manifestations, and severity of disorders in dynamics depend on the level and extent of the neurosegmental lesion and myelodysplasia severity. For example, confounding static and dynamic factors without orthopedic prophylaxis aggravate deformations of the musculoskeletal system.

Paralytic deforming arthrosis with myelodysplasia has been mainly observed in the distal parts of the lower extremities, with the ankle joint affected in most of cases.

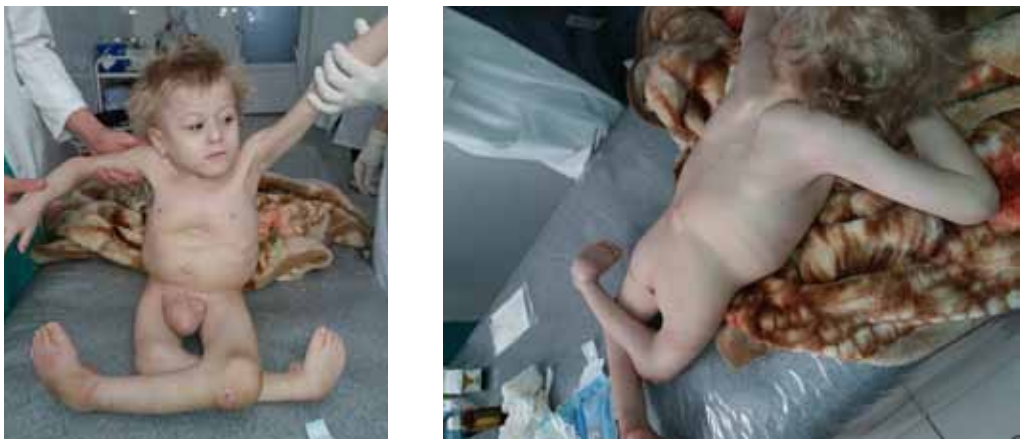

Fig. 1: Unproportioned body parts with contractures and deformities in the lower extremity joints in case of combined vertebromedullary anomaly: a) anterior view b) posterior view.

Fig. 2. Various types of lower extremity deformities with SH: a) bilateral talipes equinovarus; b) unilateral talipes equinovarus; c) lower extremity disproportion with the hip dysplasia.

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References

1. Baindurashvili A. G. ISV,KV. Hip subluxation and dislocation with effects of spinal hernias in children (literature review). Journal Traumatology and Orthopedics of Russia. 2013; 70(4): p. 97-102.

2. A.V. B. Complex treatment of flat-valgus deformity of the feet in children and adolescents, taking into account the state of the lower extremity neuromuscular apparatus. Ph. D. Thesis Abstract, Moscow. 2015; 22.
3. Voronov V. G. TEG. Vertebromedullary malformations in children: spinal malformation diagnosis and treatment manual. Moscow: Ekon-Inform Publishing House. 2016;: p. 395.
4. Guk M.Yu. SAT,OYA. Neuroorthopedic manifestations of spinal hernias depending on the lesion form and severity. Bulletin of Orthopedics, Traumatology and Prosthetics. 2008; 1: p. 17-22.
5. Degtyareva E.I. BAG,KMP. Types of locomotory dysfunction in children with the feet paralytic deformities and affects of spinal hernias of lumbosacral region. Traumatology and Orthopedics of Russia. 2009; 2(52): p. 81-87.
6. Ivanov S.V. KVM. Radiological features of the hip joint in children with the spinal hernia affects. Genius of Orthopedics. 2011; 1: p. 93-97.
7. Kuschel YV, ZMY. Postfixation spinal cord' syndrome after correction of various forms of spinal dysraphism in children. Neurosurgery. 2010; 2: p. 41-46.
8. M SD. Surgical treatment of patients with deformity of the spine against the spinal dysraphia. Ph.D. Thesis Abstract, - Kurgan. 2016; 22.
9. Sysoev K.V. ZEN, ZYMea. Latent fixed spinal cord syndrome in children (clinical observation and literature review). Neurosurgery. 2016; 2: p. 53-58.
10. Khachatryan V.A. SKV. Topical problems of the pathogenesis, diagnosis and treatment of fixed spinal cord syndrome (analytical review). Pediatric Neurosurgery and Neurology. 2014; 3: p. 76-87.
11. Ergashev N.Sh. OFA, ENN, NDS. Spinal anomalies in children with anorectal malformations//Journal of Theoretical and Clinical Medicine. 2018; 1: p. 75-89.
12. N.N E. Clinical and paraclinical characteristics of neurological disorders in children with spinal cord pathologies and new approaches to their diagnosis and treatment. PhD Thesis Abstract, Tashkent. 2018; 66.
13. Fariborz Samini HMMKMS. Orthopedic Lesions in Tethered Cord Syndrome: The Importance of Early Diagnosis and Treatment on Patient Outcome. Arch Bone Joint Surg. 2014; 2: p. 93-97.
14. Cools M.J AHWNSWRJWTJMKMIMea. Filum terminale lipomas: imaging prevalence, natural history, and conus position. J Neurosurg Pediatr. 2014; 13: p. 93-97.
15. Glenn C CAASASea. Spinal cord detethering in children with tethered cord syndrome and Chiari type 1 malformations. J Clin Neurosci. 2015; 22: p. 1749–1752.
16. Mustafa Barutcuoglu MSASUMMSGGDS. Scoliosis may be the first symptom of the tethered spinal cord//Indian journal of orthopaedics. 2016; 1: p. 80-86.
17. Yamada S. ZDE,SD. Pathophysiology of «tethered cord syndrome». J. Neurosurg. 1981; 54: p. 494-503.