

## JOINT HYPERMOBILITY SYNDROME IN INFANTS

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### Abstract

In the article considered the problem and treatment of joint hypermobility syndrome of people in different ages, especially of children of the first year of life. The mission of the theoretical study is expected the realization of the system analysis of scientific and methodological literature on the topic, studying methods of examination and reformation of individuals with the joint hypermobility syndrome; identifying the key issues of differential diagnosis of infants with the syndrome of joints hypermobility, burdened with delayed development of general motor skills; analysis of physical rehabilitation of children in the above-mentioned age and nosological category. Methods of the analysis include the theoretical analysis, synthesis and generalization of the literary sources. In the article emphasized the concept of screening and identifying the manifestation of joints hypermobility in individuals of different ages. It demonstrates the necessity of the application of differentiated diagnostic testing of locomotors areas in infants and application of appropriate specific examinations for the objective evaluation of these patients. In particular, there is a proposed use of the following tests: the visual assessment of the child's arbitrary posture at rest and in motion, test on tone muscles, test for traction, determining whether there are «symptoms of a composite blade» and «symptom of flaccid shoulder», detection of supportive functions of legs, arms, determination and volume resistance of passive movements. Attention focused on the necessity to find a comprehensive physical rehabilitation program for the effective and rapid formation of motor functions in infants. As a result of the theoretical research, it is possible be concluded that the clinical studies of the joint hypermobility syndrome, the consequences of this pathology and directions towards its correction in the literal sources are not enough highlighted. It demands the improvement of methods of the diagnosis delay in motor areas in infants at the presence of joints hypermobility. It is necessary to optimize the programs of physical rehabilitation in infants with the developmental delay of locomotion to prevent complications of their features musculou-ligamentous apparatus.

**Key words:** joint hypermobility syndrome, musculoligamentous apparatus, diagnostics, joint and outer extra-articular symptoms, delay of overall motility, muscle tone, muscle strength and rehabilitation.

**Ольга Нагорна, Людмила Брега, Віктор Горчак. Синдром гіпермобільності суглобів у дітей першого року життя.** У статті розглянуто проблему діагностування та лікування синдрому гіпермобільності суглобів в осіб різних вікових категорій, зокрема в дітей першого року життя. Завданням теоретичного дослідження передбачено проведення системного аналізу науково-методичної літератури за темою, вивчення методів обстеження та корекції осіб із синдромом гіпермобільності суглобів; визначення основних проблем диференційованої діагностики дітей першого року життя із синдромом гіпермобільності суглобів, обтяженим затримкою розвитку загальної моторики; аналіз засобів фізичної реабілітації дітей цієї вікової та нозологічної категорії. **Методи дослідження** включали теоретичний аналіз, синтез й узагальнення літературних джерел. У статті висвітлено концепцію обстеження та виявлення гіпермобільності суглобів в осіб різного віку. Обґрунтовано необхідність застосування розмежованих діагностичних тестувань локомоторної сфери дітей першого року життя й застосування відповідних специфічних обстежень для об'єктивного оцінювання цієї категорії хворих. Зокрема, запропоновано застосування таких тестувань, як візуальна оцінка довільного положення тіла дитини в спокої та в русі, проба на тонус привідних м'язів, проба на тракцію, визначення наявності «симптому складеного ножа» й «симптому млявих плечей», виявлення опороздатності ніг, рук, визначення опірності та обсягу пасивних рухів. Акцентовано увагу на потребі пошуку комплексної програми фізичної реабілітації для ефективного й швидкого формування рухових функцій у дітей першого року життя. Унаслідок теоретичного дослідження можна зробити висновки, що клінічні дослідження щодо синдрому гіпермобільності суглобів, наслідки цієї патології та напрями корекції в літературних джерелах висвітлено достатньо. Потребує вдосконалення методика діагностики затримки моторної сфери в дітей грудного віку за наявності гіпермобільності суглобів. Постає необхідність оптимізації програми фізичної реабілітації дітей першого року життя із затримкою розвитку локомоцій для попередження в них ускладнень цієї особливості м'язово-зв'язкового апарату.

**Ключові слова:** синдром гіпермобільності суглобів, м'язово-зв'язковий апарат, діагностування, суглобові та позасуглобові прояви, затримка становлення загальної моторики, тонус м'язів, сила м'язів, реабілітація.

**Ольга Нагорная, Людмила Брега, Виктор Горчак. Синдром гипермобильности суставов у детей первого года жизни.** В статье рассматривается проблема диагностики и лечения синдрома гипермобильности суставов у лиц разных возрастных категорий, в том числе у детей первого года жизни. Задачей теоретического исследования предусматривалось проведение системного анализа научно-методической литературы по теме, изучение методов обследования и коррекции лиц с синдромом гипермобильности суставов; определение основных проблем дифференциальной диагностики детей первого года жизни с синдромом гипермобильности суставов, отягощенный задержкой развития общей моторики; анализ средств физической реабилитации детей данной возрастной и нозологической категории. **Методы исследования** включали теоретический анализ, синтез и обобщение литературных источников. В статье освещается концепция обследования и выявления гипермобильности суставов у лиц разного возраста. Обосновывается необходимость применения разграниченных диагностических тестирований локомоторной сферы детей первого года жизни и применения соответствующих специфических обследований для объективной оценки данной категории больных. В частности, предлагается применение таких тестов, как визуальная оценка произвольного положения тела ребенка в покое и в движении, проба на тонус приводящих мышц, проба на тракцию, определение наличия «симптома составленного ножа» и «симптома вялых плеч», выявление опороспособности ног, рук, определение устойчивости и объема пассивных движений. Акцентируется внимание на необходимости поиска комплексной программы физической реабилитации для эффективного и быстрого формирования двигательных функций у детей первого года жизни. Вследствие теоретического исследования можно сделать выводы, что клинические исследования по синдрому гипермобильности суставов, последствия данной патологии и направление коррекции в литературных источниках освещены достаточно. Требуется совершенствования методика диагностики задержки моторной сферы у детей грудного возраста при наличии гипермобильности суставов. Необходима оптимизация программы по физической реабилитации детей первого года жизни с задержкой развития локомоций для предупреждения у них осложнений данной особенности мышечно-связочного аппарата.

**Ключевые слова:** синдром гипермобильности суставов, мышечно-связочный аппарат, диагностирования, суставные и внесуставные проявления, задержка становления общей моторики, тонус мышц, сила мышц, реабилитация.

**Introduction.** Many sources emphasize that joint hypermobility (JHM) is a condition in which the range of motion in joints higher than normal [1; 3; 6]. Joints hypermobility syndrome (JHMS) appears when an excessive joint mobility is combined with musculo-articular symptoms and extraarticular manifestations [4].

The main feature of joint hypermobility (JHM) is fundamentally an abnormal flexor-extensor movements of the joints [1]. There is a well-known fact that the distribution of JHM among European adult population constitutes about 10–15 % and 15–25 % – for African and Asian representatives; regards gender ratio: women tend to have higher frequency and severity of JHM than men [1]. JHM is often found in athletes, especially in certain specialties, such as dancers [7].

Even though JHMS in medicine is traditionally considered as mild pathology, clinicians distinguish joint and outer joint symptoms of this condition. JHMS together with influences on the musculoskeletal system is represented by the following diagnoses: polyarthralgia [1; 6] appears in about 30–40 % of children, in the adult population – 28 %; Arthralgia appears in adults and associated with exercise-related activities and injuries [1; 6]; diagnoses recurrent arthritis, arthralgia of unknown etiology [1; 6], recurrent muscular-articular symptoms, inflammatory joint damage [1; 6]; joint subluxation, recurrent effusion, crunch («click») joints dorsalgia, symptomatic flatfoot, periarticular destruction; joint noises; functional subluxation, tendinitis, epicondylitis, enthesopathy, bursitis, tunnel syndrome, excessive flexibility of joints and spine, knees crunch in joints, back, posture, frequent dislocation, subluxation [5]. These effects lead to JHMS disorder of biomechanics musculoskeletal system, lower the tone of muscles, ligaments, tendons [5]. It is believed that the most vulnerable structure is musculoligamentous apparatus of joint ligaments, shoulder joints, lateral and medial callosity, lumbar and cervical spine [5].

Among extra-articular manifestations the frequency of appearance of prolapse of mitral valve at JHMS is 30–40 %; it appears a misbalance of leukocyte populations, reducing the number of T-lymphocytes; it observes excessive elongation, skin vulnerability, dysfunction of the autonomic nervous system, varicose veins, tendency to bruising, urogenital prolapse, Raynaud's syndrome, neuropathy, fibromyalgia, low bone density, anxiety and panic conditions, depression [1; 5; 7].

The outcomes of the research [1; 5] demonstrated that there was a presence of protrusion in lumbar spine and cervical region in 50 % of patients with JHMS. In 58,3 % of patients with connective tissue

dysplasia were detected cervical intervertebral hernia and, consequently, lumbar intervertebral hernia. Thus, there is a direct connection between hypermobility of joints and pain syndromes in back.

More objective examination of diagnosis JHMS patients of different age provided in many literary sources. In particular, there are goniometry indicators, standard criteria introduced by S. Carter J. Wilkinson (1964) with modifications by P. Beighton (1983), which are 9-point assessment of the subject's ability to perform the corresponding five movements. Criteria Rotesa is sufficiently objective and specific, and allows to assess mobility in majority of joints. When hypermobility of joints decreases, especially in the elder age, it is proposed to use Hakim and Graham's questionnaire [3; 5; 6].

P. Graham's formula (2003) is used for better understanding the relationships between physiological JHMS and pathological JHM: joint hypermobility syndrome combines actual hypermobility of joints and symptoms [1; 5].

However, there are a number of problems in the diagnosis of JHM, such as it is not proved what results of performance should be taken as an average volume rate movements; what is attributed to the unconstitutional hypermobility; often the use of standardized tests in clinical practice is limited because they are time-consuming, requiring the ability to use a goniometer; or, had not considered gender feature and age. The particular difficulty constitutes the definition of this disease in infants because their inherent physiological hypermobility, given the immaturity of the connective tissue.

If we consider the problem JHMS in pediatrics, JHM is observed in almost 50 % of children aged 2–3 years, but further decreases the proportion of about 20 years when the connective tissue condition gets stabilized. [3] Furthermore, JHM in children up to the age of three occurs with equal frequency in boys and girls, and in puberty – often in girls [3]. According to many studies [4], based on background of JHMS in children, it develops inflammatory joint damage, observed frequent injuries, especially during exercise and sports, systematic recourse to cardio rheumatologists, orthopedists and other doctors.

Thus, joint hypermobility is the cause of dysfunction of the musculoskeletal system and other systems in individuals of different gender and age. Diagnosis of the infants' health conditions is not sufficiently represented in the sources; furthermore, it requires objective evaluation and selection of physical rehabilitation.

**The aim of study** – to explore diagnostic techniques of joint hypermobility syndrome in infants and patients' physical rehabilitation with this pathology.

**Objectives of the Study:**

1. To conduct a systematic analysis of scientific literature, domestic and foreign experience in diagnostic methods and physical rehabilitation of people with joint hypermobility syndrome.

2. Identify the main problems of differential diagnosis of infants with the joint hypermobility syndrome.

3. Analyze the methods of children physical rehabilitation of this age category and nosology.

Methods involving theoretical analysis, synthesis and analysis of the literal sources.

**Material and Methods of the Study.** Due to the lack of familiarity with the above-mentioned pathology, pediatricians, neurologists and orthopedists often do not provide the correct diagnosis established even after complaints from parents and the presence of symptoms. Traditionally, doctors' attention drew from limited range of motion in joints, not the determination the excess amount. These children in their first year of life often get the attention of neurologists about the delay of the overall motor skills. Subsequently, they are turning to cardiologists, orthopedists, trauma surgeons and other physicians [3; 4; 5].

Because physiological muscle hypertonicity it is very hard to identify articular hypermobility in children of first weeks of life. During neurological or orthopedic examination, delay of motor development in infants becomes to be essential.

The main criteria for evaluation of psychomotor development in children of first year of life are tone, muscle strength, formation of motor functions, reduction of automatism, visual-motor coordination, eye contact [2].

There is a need for differentiated selection of tests to identify the causes of delays locomotions in a view of JHM presence.

To identify JHMS in infants we suggest using the following special techniques: visual assessment of the child arbitrary posture at rest and in motion, driven test tone muscles, the presence of “symptoms folded blade” and “symptom flaccid arms”, detection of feet reliance, hands, determination and resilience amount of passive movements.

The proposed tests could help detect joint hypermobility of the lower limbs, shoulder girdle, increased flexibility of the spine. However, this assessment can be used in case of change the strength and muscle tone, and thus lead to other pathology.

The concept of evaluation of infants muscle strength proposed to conduct a six-point system: 0 points – no movement; 1 – no active movement, but is determined by palpation muscle tension; 2 – passive movements are possible in full; 3 – passive movements are possible in overcoming a slight resistance; 4 – passive movements are possible in dealing with moderate resistance; 5 – muscle strength in the normal range [2].

For visual assessment of muscle tone we propose to determine the position of the child and the position of the limbs at rest and in motion, which will determine which muscle tone or their groups dominates, or is impaired at the time of the examination. An additional method of assessing muscle tone is to determine the amount of resistance and passive movements.

However, our proposed approach can also be biased because these symptoms inherent for pathological condition characterized by a decrease in muscle tone, particularly atony, hypo-, hyper- and dystonia.

However, it should be noted that at JHMS there is no reduction observed in muscle strength and muscle tone changes. In the diagnostic phase of identifying the causes of delayed motor development is important the differentiation of pathological conditions characterized by decreased muscle strength, muscle tone with joint hypermobility syndrome.

In our view, such an assessment methodology will provide impartial examination and facilitate the formation of optimal physical rehabilitation program based «fallout» in child's development.

Treatment and rehabilitation of children, including infants, from JHMS today is not completely developed. General principles of treatment and rehabilitation tactics at JHMS must fundamentally differ from those in the treatment of other diseases of the musculoskeletal system. In the role of non-medicinal therapy we offer the application of medical physical culture and massage, hydrotherapy for use exercises for muscles that surround joints hypermobility, and if necessary – correction of orthopedic artificial restriction the range of motion in the joint.

Based on the characteristics of symptoms, it is advisable to use exercises that will strengthen the muscles and do not promote active bending-extension of the joints. There are effective fitness exercises for strengthening the deep back muscles, transverse abdominal muscle, abdominal press, feet, hands and trapezius muscles. It will promote the improvement of musculo-ligamentous apparatus with use of compression joints. However, exercises, aimed at increasing the flexibility of joints and spine, in our opinion, is inappropriate in the rehabilitation of this pathology. The best way is to avoid excessive over bending of joints when performing various movements. To generate a feedback it is advisable to carry out all the exercises in front of a mirror.

It is important consider the chronological age and motor capabilities of the child at the time of rehabilitation when forming meaningful motor function. “Jump” through certain stages of the overall motor skills can only trigger abnormal situation. So, sitting near the fixation of support, or, sitting with outstretched legs without the support of the knee provokes the formation of a round back; sitting in a pose «W» – splayfoot; standing with over bending knees offset by increased lordosis of the lumbar spine.

Classes of hydrocolonotherapy will strengthen the major groups of muscles and ligaments, and, in the same time, axial load of joints and spine.

Therapeutic massage has restorative direction, as a general and periarticular, will improve the state of the musculoskeletal system, local action on the joints.

All of the above means of rehabilitation aimed at increasing muscle strength, normalize muscle tone, neuromuscular coordination and, consequently, improve the level of motor functions. In our opinion, the main argument in favor of rehabilitation program effectiveness timely phasing formation of motor functions.

**Conclusions.** The scientific literature presents enough basic clinical research on joint hypermobility syndrome, and studies the effects of disease and medical correction. However, there are no developed diagnostic tests for separated joint hypermobility syndrome in infants. This problem needs an immediate solution to create the optimum tactics and reasonable rehabilitation of children in this category and the prevention of their joint and extraarticular complications.

**Prospects for Further Research.** Diagnostic tests that would promote the differentiation of abnormalities of neurological and orthopedic in infants at JHMS remain topical. It is necessary a specific selection of physical rehabilitation of children with the joint syndrome hypermobility, burdened by developmental delay locomotions.

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