Correction of hip joint instability in children with cerebral palsy — current state of the problem and prospects for its solution (literature review)

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Hip instability in children with cerebral palsy (CP) is a serious unresolved problem in modern orthopedics. Objective. To analyze the state of the problem of the hip joint instability in children with cerebral palsy and determine the prospects for its solution. Methods. A thematic review of 68 studies was made. Results. The basis for the prevention of instability of the hip joint should be a systematic X-ray screening at least once a year. The instability of the hip joint is based on neurological disorders, if the index of migration of the femoral head (MP) is less than 30 %, the application of selective dorsal rhizotomy or baclofen pump is promising and justified. If MP > 30–100 % dorsal rhizotomy can be used after surgical correction of abnormalities in the hip joint to reduce the recurrence rate. The existing surgeries on the pelvic and femur is sufficient to restore the stability of this joint, even in the most severe cases, but the result is not always possible to maintain due to the recurrence of the deformity. The requirement for hip intervention in a patient with cerebral palsy is a preventive focus, by which we mean not only the achievement of joint stability, but also the creation of conditions for its preservation. Temporary blockage of the medial portion of the femoral head growth area is a minimally invasive procedure and may be recommended for use alone or as an adjunct to hip soft tissue release or for hip and bone surgery. However, it is still unclear at what age it is better to block the growth plate of the femoral head, whether and how often to change the clamps and so on. A prerequisite for the treatment of patients with this pathology is an individual approach, taking into account the degree of displacement of the femoral head, the presence of pathology of adjacent joints, the age of the child and the severity of the disease. Correction of existing deviations should be performed in one step at many levels. Key words. Cerebral palsy, hip joint, dysplasia, instability, surgical correction, orthopedic correction.
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Introduction

Hip instability in children with cerebral palsy (CP) is a serious problem, far from being resolved, despite the attention it receives. In essence, anomalies of the hip joint in cerebral palsy are secondary to the underlying disease, but they often come to the fore due to a set of complex problems [1]. Their elimination leads to several surgeries, and the progressive deterioration of the joint with the instability and dislocation of the femoral head is not uncommon and in the absence of treatment or its absence becomes a «terrible phenomenon in patients experiencing pain and discomfort» [2]. In Western Europe, the incidence of cerebral palsy is 2 cases per 1,000 newborns, 1.6–6 per 1,000 full-term infants in Russia and 9–40 per 1,000 premature infants [3], from 2.8 to 4 per 1,000 newborns in the United States [4], 2.72 cases per 1,000 children in the Republic of Belarus [5], and there is no tendency to reduce the frequency of this abnormality.

The aim of the study: to analyze the state of the problem of instability of the hip joint in children with cerebral palsy and to determine the prospects for its solution.

Material and methods

A thematic review of 68 modern studies was conducted.

Results and their discussion

Abnormal mechanics of hip dislocation

In newborns with cerebral palsy, the incidence of abnormalities of the hip joint is within the average population, but steadily increases with age. The reason for this, first of all, are the wrong commands of the brain and improper muscle function, not only in the hip joint, but also the musculoskeletal system as a whole. Over time, this leads to irreversible morphological changes in muscles and ligaments, the development of contractures, subdislocations and dislocations [6, 7].

S. N. Chang et al. [8] found a strong correlation between flexion-inducing contracture in the hip joint and displacement of the femoral head, which is largely due to the anteverision of its proximal part.

Interesting data are given in the work of M. K. Chung et al. [9], who found that in cerebral palsy secondary to developing acetabular dysplasia, the circumference of the acetabulum increases, triggering a mismatch with the normal size of the femoral head, which promotes its migration. In this case, severe dysplasia of the cavity can be combined with minimal displacement of the femur and vice versa. However, to date, the abnormal mechanics of hip dislocation in cerebral palsy is not fully understood [9].

Classification

Cerebral palsy is traditionally classified as a type of motor disorder (spastic, dyskinetic, ataxic, hypotonic, and mixed), and their topographies include hemiplegia, diplegia, and quadriplegia, although terms such as monoplegia, paraplegia, triplegia, and double hemiplegia are also used [10]. The incidence of abnormalities of the hip joint in different forms of cerebral palsy differs significantly. Thus, according to F. Dobson et al. [11], it is diagnosed only in 1 % of cases with spastic hemiplegia, 5 % with diplegia, 35–55 % with quadriplegia. T. Terjesen [12] noted the migration of the femoral head by more than 33 % in 26 percent of cases, and the risk of its occurrence increased from 3 % in hemiplegia to 81 % in quadriplegia. Moreover, for types of cerebral palsy characterized by abnormal movements, this risk was significantly lower and amounted to 26 % in dyskinesia and 0 % in ataxia. The degree of displacement of the femoral head and the incidence of subluxation and dislocation have been shown to increase in cerebral palsy with age and with an increasing severity of the abnormality [13].

The existing neurological terminology undoubtedly conveys important information, but the use of different names to describe the same conditions in cerebral palsy has long hindered the understanding of physicians of different specialties and left ambiguities in the assessment of patients’ functional capabilities. Greater consistency in the community of specialists dealing with cerebral palsy has been achieved with the introduction of the descriptive classification of major motor functions (Gross Motor Function Classification System, GMFCS), proposed by R. Palisano et al. in 1997 [14]. It is based on the ability of a patient with cerebral palsy to perform certain motor functions and identifies five levels of restriction of movement in everyday life for five age groups: up to 2 years, 2 to 4, 4 to 6, 6 to 12, 12 to 18.

Some authors indicate that the higher the level of motor disorders, the more often there are deviations from the hip joint, and in children with GMFCS IV–V, in particular in 60–90 % of cases [13]. The risk of femoral head migration and severity has been found to increase from 1 to 72 % at GMFCS levels I to V, and the rate of displacement progression varies from 0.2 to 9.5 % per year [12]. At the same
time, at the I–II level of GMFCS dysplasia occurs only in 15% of cases, but the most severe disorders of the relationship in the joint, up to hip dislocation, were observed in 4–5% of cases [15].

Screening systems

Impaired stability in the hip joint, early onset of pain, loss of leaning ability and malformation of the limb significantly increase the degree of disability in children with cerebral palsy. Therefore, orthopedic treatment should be preventive in nature, which will provide better long-term results and reduce the risk of failure [16].

Unfortunately, to date, specialists treating children with cerebral palsy are not wary of the development of abnormality in the hip joint. It is not uncommon for the first X-ray examination of patients with cerebral palsy to be performed at a later date. This is especially true in patients with spastic hemiplegia, in whom severe dysplasia is usually not recognized until the onset of symptoms in adolescence [17]. We observed cases when spastic dislocation occurred secondary to seemingly completely well-being, during rehabilitation activities in children from 6–8 years of age. In such a situation, it is no longer possible to confine management to conservative measures or minimal surgical interventions, and the problem that has arisen, even if it does not put an end to the patient’s return to independent walking or mastering it, at least throws the child back in these endeavors for months and years violating the quality of life for him and his immediate environment [18].

Given the above, periodic clinical and radiological examination of the hip joints in children with cerebral palsy is absolutely expedient. Today, there are two popular screening systems, Australian and Swedish ones. The former includes only the determination of the optimal interval between examinations [19], the latter links the obtained data with the early correction of the progression of the displacement of the femoral head, starting from the age of two [20]. Although X-ray screening for hip abnormalities should be mandatory in all patients with cerebral palsy [21], the frequency of examinations is individual and is determined based on the severity of the disease, the age of the child and the ratios in the joint.

Early multilevel minimally invasive approach

It is known that the basis of orthopedic problems in cerebral palsy, including the hip joint, is spasticity and imbalance of antagonist muscles. Preventive measures, including physiotherapy, the use of orthoses and various treatments to reduce spasticity, are strongly recommended in each case. To restore proper muscle balance, including a decrease in the degree of migration of the thigh laterally and cranially, in order to avoid the subsequent development of pain in the hip joint, N. Portinaro et al., proposed the Early Multilevel Minimally-invasive Approach (EMMA) in 2009 [22]. According to them, EMMA is indicated for most patients with cerebral palsy, especially in increased tone, poor muscle control and Reimer index (RI) up to 20%. At the age of 2–4 years, the authors propose a multilevel injection of botulinum toxin in muscle hyperactivity, but without morphological changes of muscle antagonists (contractures). In the second stage, at the age of 4–6 years, a multilevel aponeurectomy is performed in case of muscle hyperactivity with morphological changes of the antagonist muscles of the pair (retraction). Unfortunately, injections of botulinum toxin into hip adductors and flexor muscle groups are not effective in preventing the progression of hip instability in cerebral palsy [23], and a positive aponeurectomy result is short-lived. Early interventions in the area of growth of the femoral head, which consist in blocking its medial part, are also a component of EMMA.

Correction goals

The right choice of goal is the most important component of the treatment of abnormalities of the hip joint in cerebral palsy. Orthopedic measures should prevent or eliminate the instability of the hip joint, maintain or create the opportunity for independent movement, and in the presence of dislocation to relieve the patient from pain.

The use of the GMFCS classification helps to build a strategic line of action aimed not only at correcting existing problems, but also at preventing the emergence of new ones. The goal for level I–II GMFCS is to maintain or improve the patient's ability to move freely; self-movement with surgical correction of deviations for level III GMFCS; creation of comfortable conditions for the patient with prevention or elimination of a pain syndrome in a hip joint and other orthopedic pathology complicating care of the patient for level V GMFCS.

Definition of indications and a choice of a technique of surgical intervention

To assess the condition of the hip joint and the choice of treatment in patients with cerebral palsy, along with conventional radiological parameters (acetabular index, Wiberg angle, anterior margin angle, Shenton line, NSA, degree of antetorsion), the Reimer’s head 24 migration index is widely used.

A common indication for surgical treatment for hip instability in children with cerebral palsy is
a femoral head migration index (MI) of more than 30 %, as this value poses a risk of dislocation [25].

In recent decades, selective dorsal rhizotomy and baclofen pumps have been actively used, which are prophylactic in patients with minimal instability of the hip joint (MP < 30 %). They are common and can significantly reduce the need for orthopedic surgery [25]. R. S. Gersztien et al. [26] showed that the number of planned orthopedic interventions decreased from 38 to 21 % after only one year of continuous infusion of baclofen. L. Krach et al. [27] reported that after a year of intracanal baclofen administration, more than 90 % of the hip joints remained stable or even improved regardless of the severity of cerebral palsy. S. Silva et al. [28] conducted a comparative analysis of the use of dorsal rhizotomy and baclofen pump and found that in patients with severe lower extremity spasticity, their effectiveness for at least 2 years was approximately the same, and reconstruction of the hip joint due to the progression of its instability was required in 25 % and 32 %, respectively.

The choice of intervention on the hip joint depends on the degree of violation of its stability, which occurs due to excessive or incorrectly directed action of the muscles: long leading, thinnest, straight thigh, iliac-lumbar. In this case, the strength of spastic muscles is almost 3 times greater than the strength of the muscles of a healthy child, and the developing imbalance leads to an abnormal setting of the thighs in the position of reduction, flexion and internal rotation [29]. Dislocation of the femoral head develops gradually and most often occurs at the age of 3 to 8 years [30, 31], but can also be observed later. To correct the asymmetry of muscle forces, soft tissue release is used, which is considered effective for preventing hip dislocation in patients with cerebral palsy [32] and is recommended to be performed at the earliest age, regardless of other factors [33]. This will provide better long-term results and reduce the risk of failure [34]. Soft tissue release in the early stages of femoral displacement helps to effectively reduce the frequency and severity of hip dislocation [20].

Adductor myotomy in the form of self-intervention is most expedient in young children with leading contracture of the thighs and uncritical displacement of the femoral head within the acetabulum. The operation, along with increased joint stability, may improve gait, increase hip abduction, facilitate hygiene, relieve pain [35].

R. Knapp pointed out that the success of adductor myotomy varies from 60 to 100 % with a femoral head migration index of less than 50 % [36]. A. Presedo reported a positive effect of soft tissue intervention on the hip in children with cerebral palsy, but recurrence of hip dislocation was often observed during child growth [32]. The best results can be achieved in patients with low levels of GMFCS and slight hip displacement [20].

It was found that the risk of soft tissue release failure is directly related to GMFCS levels, and the operation was successful in 94 % of children with GMFCS II, 49 % GMFCS III, 27 % GMFCS IV and only 14 % with GMFCS V. Therefore, the least successful early soft tissue interventions were observed in those who need it most, and the risk of failure, which the author interprets as MR > 50 % or the need for subsequent surgery to correct hip instability, is extremely high in children with GMFCS IV–V [37].

With the help of detorsion-varying osteotomy of the hip (DVOH) it is possible to change the geometry of the proximal femur and to restore the stability of the hip joint in case of subluxation or even dislocation of the femoral head with satisfactory development of the acetabulum. Osteotomy should correct anteversion and provide moderate variation (120°±10°) [38] and may be performed in conjunction with soft tissue intervention or involve shortening of the femur, which we believe is mandatory. According to B. Dohin, DVOH may result in spontaneous correction of secondary dysplasia of the acetabulum [38], but the operation does not prevent the development of dysplasia in adolescence [17].

Further development of the acetabulum is more common in patients with GMFCS II and III and rarely in GMFCS IV and V [39, 40].

It was found that in the case of unilateral femoral osteotomy in children later, in adulthood, 44 % of patients required corrective osteotomy on the opposite side [41]. Therefore, there is an opinion [42, 43] about the feasibility of primary bilateral bone surgery in case of a risk for the contralateral hip, namely MP between 10 and 30 % in GMFCS III–V.

Interesting data were presented by M. Al-Ghadir et al. [44], who compared the results of detorsion-varying osteotomy of the hip alone or in combination with pelvic osteotomy in children with cerebral palsy who had a similar clinical and radiological picture of subluxation or marginal dislocation of the hip and operated at the age of (8.1 ± 3.6) years. 4.4 years after DVOH and pelvic osteotomy, there was a significant reduction in pain and improved joint stability. After DVOH, the clinical and radiological results were significantly worse, which led to re-intervention in every fourth patient.
In a significant cranial displacement of the femoral head, the combined intervention should be supplemented by an open repositioning into the acetabulum. However, this is necessary only in cases of complete dislocation, with MR > 100 % [45], and stability is achieved by hypercovering the femoral head with an acetabulum, and the need for capsulography, the effectiveness of which is questionable, is absent. In this approach, although muscle imbalance persists, the subsequent growth of the proximal femur does not lead to recurrence of subluxation [46, 47]. We should agree with the statement of H. K. Graham, P. Selber that such tactics are the best choice for patients with spastic hip dislocation [48].

According to B. Dohin [49], Pemberton, Dega or other types of acetabuloplasty are the optimal interventions on the pelvic bones. The author does not recommend the use of Salter's operation or Le Courte's triple osteotomy due to their limited capabilities. However, we have significant positive experience in the use of pelvic osteotomy according to Salter in young children and triple osteotomy according to A. M. Sokolovsky in severe dysplasia of the acetabulum in adolescents and adults with cerebral palsy with restoration of joint stability.

Encouraging results were shown in the study of A. Christian et al. [50], reporting almost 90 % success in 13 to 71 months. after open repositioning of femoral head dislocation in combination with pelvic and femoral osteotomy in children with cerebral palsy under 3 years of age.

L. Root et al. [51] analyzed the results of open insertion and DVOH in combination with pelvic osteotomy in 31 patients with cerebral palsy (35 joints) with dislocation and severe subluxation (average migration index 74 %) on average after 7 years. Prior to surgery, 22 patients could not stand, and 13 had severe pain. At the last examination, none of the joints were painful, 7 patients had improved gait, and all children confined to a wheelchair had better seating balance. The stability of the joint has improved, the Wiberg angle has become on average 18°, the migration index was 25 %.

A complication of the combined intervention may be osteonecrosis of the femoral head, which, according to various authors, occurs in 1/3–2/3 cases, which does not add optimism to surgeons [52–54]. In our opinion, the possibility of reliable stabilization of the hip joint justifies the risks.

The principle of single event

In patients with even minimal potential for self-movement (GMFCS I-III, rarely GMFCS IV), restoration of hip stability should be considered as an extremely important component in solving the complex problem of normalizing or improving the biomechanics of the lower extremities. The condition of the hip joints should be assessed in close connection with the status of the knee and ankle, foot and spine.

In 1985, R. Norlin and H. Tkachuk concluded that most operations on the soft tissues of the lower extremities can be performed in one step and at all necessary levels [55]. Corrective osteotomies of the lower extremities and pelvis were later added to soft tissue interventions, and a similar approach was defined as Single-Event Multilevel Surgery (SEMLS). It reduces the time of treatment of a child with cerebral palsy to one hospitalization and one course of postoperative rehabilitation and provides good long-term results [56–58].

There are studies justifying a two-stage surgical correction of the position of the components of the hip joint [59], although, in our opinion, this approach is not necessary, and all existing deviations can be corrected during a single surgery due to modern advancements in orthopedics and anesthesiology. Moreover, it is advisable to simultaneously perform not only stabilizing operations on the hip joint, which may include osteotomy of both thighs, pelvis according to Salter, double or triple osteotomy of the pelvis according to A. M. Sokolovsky or one of the types of acetabuloplasty, but also interventions on the joints, foot bones and soft tissues.

Salvage surgery

Elimination of severe pain, which occurs in an average of 30 % of children [2] and 72 % of young people [14] in cerebral palsy with IV–V levels of GMFCS is a difficult problem. In such a situation, salvage surgery comes to the fore, [60] based on resection of the head or the entire proximal portion of the femur, or on the valgus of the proximal femur. Unfortunately, salvage surgery is not a heal-all recipe and is associated with a number of dangers and complications, and its results are unpredictable [48]. In this regard, if possible, we recommend that children with cerebral palsy undergo reconstructive surgery to restore joint stability while preserving the proximal femur.

It is known that recurrence of instability of the hip joint after reconstructive surgery occurs due to constant neuromuscular and biomechanical imbalance, which disrupts the normal development of the growth zone of the femoral head [61], and the risk of recurrence of dislocation increases with age [62]. Since spasticity is the main factor leading to recurrence of dislocation [41], its neurosurgical treatment can significantly reduce the incidence of abnormalities and pain.
Blocking the growth zone

Dislocation most often recur when the operation on the hip joint is performed in 5–6 years, and the subsequent jump in growth nullifies the results achieved. Exposure to the functional growth zone of the femoral head may be a deterrent to the development of recurrence of deformity or its progression. It has been established that blocking the medial part of the physis of the proximal femur can prevent the development of hip dysplasia and hip dislocations in cerebral palsy [22]. Later, based on the obtained good results, this procedure is recommended as an early and minimally invasive treatment [63]. Experts believe that such operations improve the geometry of the proximal femur and the acetabular covering of the head [64, 65]. In addition, the high social significance of the temporary blockage of the medial portion of the femoral head is also emphasized, since after such an intervention the patient can return to his normal life in the coming days [66].

Conclusions

Hip instability in children with cerebral palsy is not uncommon. Progression of displacement of the femoral head often leads to its dislocation with severe pain and impaired mobility. So far, this problem is far from being resolved.

The prevention of hip instability should be based on an X-ray screening system at least once a year, which is considered one of the most important achievements in the treatment of patients with cerebral palsy in recent times.

The instability of the hip joint is grounded on neurological disorders, so at MP < 30 % the use of selective dorsal rhizotomy or baclofen pump is not only promising, but also quite justified. This can significantly reduce the need for orthopedic interventions. At MP > 30–100 % the dorsal rhizotomy can be used after surgical correction of deviations in a hip joint for reduction of recurrence rate.

The available arsenal of interventions on the pelvic bones and thighs is sufficient to restore the stability of the hip joint, even in the most severe cases, but it is not always possible to maintain the result due to the recurrence of the deformity. The requirement for intervention on the hip joint in a patient with cerebral palsy is a preventive focus, by which we mean not only the achievement of joint stability, but also the creation of conditions for its preservation. Temporary blockage of the medial part of the growth zone of the femoral head is characterized by small invasiveness and may be recommended for use. This manipulation can be used alone, or in addition to the release of soft tissues of the hip joint, or operations on the thigh and pelvis. Today, temporary blocking looks quite promising both in terms of prevention of instability of the hip joint, and the preservation of the achieved ratios in use of other interventions. However, it remains unclear at what age it is preferable to use the block of the area of growth of the femoral head, whether and how often to change the clamps as a child grows up, until what age it is advisable to perform osteotomy of the femur or pelvis.

A prerequisite for the treatment of patients with this abnormality is an individual approach, taking into account the degree of displacement of the femoral head, the presence of pathology of adjacent joints, the age of the child and the severity of the underlying disease. Correction of existing deviations should be one-stage and multilevel.

One can fully agree with M. W. Aversano et al. [67] that the understanding of orthopedic treatment of children with cerebral palsy continues to grow and expand, but so far there is a small amount of studies with a high degree of evidence, so this area of research is one of the priorities in pediatric orthopedics.

Although surgery plays a huge role in the treatment of deformities in children with cerebral palsy [68], effective treatment of orthopedic problems, in particular hip instability, is possible only with the interaction of a neurologist, pediatrician, neurosurgeon, psychologist, physiotherapist and rehabilitologist. The best results can be obtained in specialized neuro-orthopedic departments.