FUNCTIONAL INDEPENDENCE OF CHILDREN BORN WITH MYELOMENINGOCELE ACCOMPANIED BY HYDROCEPHALUS

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ABSTRACT

Introduction. Myelomeningocele is one of the major causes of children motor disability. It causes paralyses or paresis of lower limbs of different extent but also incontinence as well as limb and spinal deformation depending on the level of spinal cord lesion. Majority of children born with myelomeningocele also develops hydrocephalus accompanied by abnormalities of the brain which greatly affects their functioning, mobility independence and self-care.

Aim. To present the limitation on functional independence of children who were born with myelomeningocele which is accompanied by hydrocephalus. To discuss the factors affecting ambulation and self-care capabilities as well as research results which show motor deficits areas of both motor and eye-hand coordination.

Method. Review of the world literature on the functional independence of children born with open myelomeningocele and accompanied congenital malformations.

Results. The main factors affecting the ability of ambulation in children with myelomeningocele are: the level of neurological lesion and the resulting deformities, age, weight and sex of the patient, motivation, spasticity and orthoses. The majority of patients with thoracic lesion are non-ambulators, and others lose this ability in adulthood. Children born with myelomeningocele accompanied by hydrocephalus have coexisting problems with upper limb function in activities of daily living. This causes a lack of independence in the toilet, dressing and eating. A common problem with these children is also incontinence, which largely affects their social activity and quality of live.

Conclusions. In a rehabilitation program of children born with myelomeningocele too much attention is given to motor development and at the same time fine motor skills as well as cognitive and social development are very often underestimated, while these areas of development have the greatest impact on becoming independent by children.

Key words: myelomeningocele, hydrocephalus, functional independence, mobility, self-care

Introduction

Functional independence of children born with myelomeningocele accompanied by hydrocephalus is a problem, often unseen or misunderstood by specialists responsible for rehabilitation in the broad sense. Physiotherapists, focusing mainly on improving gross motor skills, pay not much attention to impaired eye-hand coordination, which has a huge impact on children future independence. On the other hand, teachers and institutions responsible for psycho-educational support, too often state different degrees of mental retardation, because they don't understand the essence of the symptoms seen in these children.

Aim

The aim of this study was to present the most relevant information about this complex congenital defect and associated defects of the central nervous

system to those involved in the rehabilitation of children with myelomeningocele, as well as to draw attention to the problem of functional independence. Factors affecting the gait and self-care capabilities are also described, as well as, results indicate areas of deficits related to the acquisition of independence in children with myelomeningocele. This information is intended to help in the preparation of individual, comprehensive rehabilitation program of these children, preparing them for independence.

General problems associated with myelomeningocele

Meningomyelocele is a congenital malformation in which there was a failure to fuse one or more of the vertebrae arcs with associated herniation of spinal meninges along with the spinal cord and nerve roots beyond the spinal canal [1]. It is one of the most common congenital defects. By EUROCAT, incidence of spina bifida in Europe in 2000-2010 was 2.50 per 10,000 live births [2], whereas the ratio in Poland was 5.1 [3]. Despite numerous studies, no single cause of neural tube defects has been identified. It is believed that the etiology of neural tube defects is multifactorial, and the genetic factor is significant. The importance of the genetic factor can be confirmed by several facts: higher propability of birth another child with neural tube defects (2-3% if you have a child with congenital defect, 10% in the case of having two children with neural tube defects), different incidence of neural tube defects in various populations [4], higher incidence among twins (3-8%), and the occurrence of neural tube defects in combination with known genetic syndromes and chromosome aberrations [5].

In one study [6] it has been show that 15% of patients with spina bifida is homozygous of mutation 677C→T of MTHFR gene, located at 11q13 and resulting in abnormal metabolism of foliate. This is probably an important cause of neural tube defects. Taking 0,4 mg of folic acid daily by women planning pregnancy (4 mg for women who has child with neural tube defect) can prevent 72% of cases of neural tube defects [7]. The remaining 28% of the risk, indicates that there must be other causes of defects in the fetus. Environmental factors that increase the risk of neural tube defects mainly include: obesity in the mother (it has been proven that women with a BMI above 29 kg/m^2 have a higher risk of having a baby with neural tube defects [8]), hyperthermia [9], taking antiepileptic drugs during pregnancy (mainly valproic acid and carbamazepine) [10, 11], low socioeconomic status [12] and exposure to tobacco smoke [13].

About 80-90% of patients with myelomeningocele have hydrocephalus, which chance of occurrence increases with the level of spinal lesion [14]. Hydrocephalus is a pathological condition in which the imbalance between the production and absorption of CSF leads to expansion of the intracranial fluid spaces, primarily brain ventricular system [15]. Extending the brain ventricles occurs in 95% of children with Chiari II, of which 85% require shunt implantation. The cause of hydrocephalus in these patients is upward herniation of cerebellar vermis to abnormally developed tentorial notch and secondary cerebral aqueduct stenosis [16].

Most patients born with open myelomeningocele have Chiari syndrome type II. This syndrome is uniquely associated with myelomeningocele and it is presented only in this group of patients [17]. Chiari II malformation (Arnold-Chiari syndrome) is a congenital malformation of the brain, involving herniation of the cerebellar tonsils, the lower part of the cerebellar vermis, fourth ventricle and brain stem from a narrow posterior fossa through the enlarged foramen magnum with blockage of the flow of cerebrospinal fluid through holes of the IV ventricle [18]. Congenital abnormalities of the brain stem, and the pressure on the brain stem caused by the cramped posterior cranial fossa and hydrocephalus, occurring in one out of three patients with Chiari II syndrome gives a variety of symptoms such as headaches, sleep apnea, bradycardia, dysphagia, torticollis, spasticity and abnormal auditory evoked potentials [19].

The majority of patients with myelomeningocele accompanied by hydrocephalus have additional malformations of the brain: partial or complete agenesis of the corpus callosum, the lack of a septum pellucidum, increased interthalamic adhesion, colpocephaly, dysgyria and cerebellar and brainstem hypoplasia [20], as well as abnormalities of white matter pathways, mostly fornix and cingulate gyrus [21], which have an impact on the development of cognitive and functional capabilities. The degree of dysfunction of brain development is the greater, the higher the level of the spinal cord lesion is [22].

Because of the hydrocephalus and the Arnold-Chiari syndrome, about 80% of children with myelomeningocele may have different types of eye problems, such as visual acuity loss, amblyopia, atrophy of the optic nerves, cortical blindness, strabismus and abnormal eye movements. Strabismus

occurs in approximately 60% of patients and responds well to treatment, and nystagmus is found in 25% of patients [23].

Depending on the level of malformation, spinal cord lesion results in paralysis of the lower limbs, incontinence, lack of sensation of skin, as well as deformity of hip, knee and foot [24].

All children with surgically closed myelomeningocele experience anchoring of the spinal cord to the dura mater, which in MRI scans reveals as low-lying spinal cord. In 10 to 30% of them develop tethered spinal cord syndrome [25], characterized by a gradual increase of the following symptoms: muscle weakness of the lower limbs, gait disturbance, radicular pain, deformations of the foot, scoliosis, as well as sphincter dysfunction and spastic paralysis of lower limbs [26, 27]. Progressive limb spasticity and scoliosis may be the result of hydromyelia [28], which may be present in 20 to 60% of patients with myelomeningocele [29], as well as syringomyelia detected in different studies in 11 to 77 % of patients [30]. A large role in identifying early signs of these additional defects of the spinal cord has a physiotherapist.

Problems of mobility in children with myelomeningocele

Functional independence relates to activities that require motor control of the upper and lower parts of the body and relates to mobility, using the possibilities of the lower limbs, and self-care, requiring the upper limbs efficiency [31]. Because of the paralysis below the level of spinal cord lesion, as well as accompanying hydrocephalus and brain defects, children with myelomeningocele have trouble with reaching full functional independence, both in terms of mobility and self-care.

In terms of mobility, patients with myelomeningocele can be classified into one of four functional levels according to classification of Hoffer et al. [32]:

1. Community ambulators – walking with the aid of crutches or other devices for most of its activity, overcoming different types of surfaces, using a wheelchair only for long trips.
2. Household ambulators – walking only in the room using orthosis; capable of sit in a chair and move onto the bed with a little help, using a wheelchair to certain activities at home and school and all outdoor activities;
3. Non-functional ambulators – walking only during therapy sessions at home, school or hospital; using a wheelchair for most locomotion needs;
4. Non-ambulators – moving only in a wheelchair, usually capable to move from wheelchair to bed.

According to Rose et al. [33], the main problem in the treatment of patients with myelomeningocele is providing an independent gate, which has physiological benefits such as preventing of lower limb fractures, facilitates urine drainage, bowel function and lower limbs circulation as well as psychological. According to them, not every kind of movement in the upright position can be called functional gate. Functional gate requires: low energy expenditure during gate at speed 30 to 60 percent of the normal speed for the same age, ability to get in and out of a chair, and independence in setting up and removing orthoses.

De Souza and Carroll [34] list eight factors, which have the greatest ability to influence on ambulation ability in children with myelomeningocele. They are, in order of importance:

1. The level of spinal cord lesion;
2. Muscle strength at a given level of lesion;
3. The extent and degree of spinal and limb deformation;
4. Patient's age, weight and sex;
5. Motivation;
6. Spasticity;
7. The design and effectiveness of orthosis;
8. Surgical procedures.

In the case of the classification of patients according to the level of the spinal cord lesion, the most widely used is Sharrard classification [35], in which eight groups of patients were distinguished:

I. Patients with lesion below Th12 – paralysis of all muscles of the lower limbs;
II. Patients with lesion below L1 – weak or moderate hip flexor strength and palpable contraction of sartorial muscle;
III. Patients with lesion below L2 – strong hip flexor and moderately strong adductors;
IV. Patients with lesion below L3 – correct strength of hip flexors, adductors and quadriceps;
V. Patients with lesion below L4 – muscle strength such as in L3 lesion and the ability to partial hip abduction in flexion, strong foot dorsiflexion and supination;
VI. Patients with lesion below L5 – muscle strength such as in L4 lesion and moderately strong hip abductors, knee flexors and foot pronators muscles;

VII. Patients with lesion below S1 – moderately strong hip extensors, strong knee flexors and moderately strong foot plantar flexors;
VIII. Patients with lesion below S2 – weekness of deep muscle of the foot.

It turns out that the level of neurological lesion is an important factor in predicting the possibility of walking in patients with myelomeningocele. The research of Hoffer et al. [32] shows that only 40% of patients with thoracic myelomeningocele reached non-functional level of ambulation for a period of no longer than 6 months and the other moving around in a wheelchair. Among patients with lumbar lesion, 35% achieved a community ambulation, 12.5% were household ambulators, 5% reached the level of non-functional gait, and the other moved in a wheelchair. All patients with sacral lesion reached the highest level of mobility by Hoffer. Similar conclusions can be drawn from the study of Rose et al. [33], where only one patient with a thoracic lesion (2%) achieved a community ambulation, 63% of patients were therapeutic walkers, and 6% moved around in a wheelchair. On the other hand, according to research of Matuszczak et al. [36], 77% of children with myelomeningocele were moving in a wheelchair, but none of the children with the thoraco-lumbar and lumbar lesion obtained independence in a movement, like as 94% of children with lumbo-sacral lesions. In another work, Okurowska-Zawada et al. [37] presented the results of the analysis of age at the start of walking in patients with myelomeningocele. Among patients with thoracic lesion, 86% didn't walk independently in the third year of life, 10% reached the late stage of walking, and only one began to walk independently. Among patients with lumbar lesion, 50% of children did't walk independently in the third year of life.

Because very few patients with thoracic lesion achieve the ability to walk, and in adulthood, most of them lose this opportunity, some doctors are wondering whether or not to adapt to these patients only to ride in a wheelchair in early childhood. To answer this question, two groups of patients aged between twelve and twenty were examined [38]. Patients in Seattle, have used a wheelchair as the only means of transportation from earliest childhood, and patients in Melbourne were adapted for walking between two and four years old. It was shown that patients who were ambulated in early childhood were more independent in transfers from wheelchair to bed, on the floor, to toilet and back, that resulting from a better upper limbs and trunk musculature. Walking patients had also fewer fractures and pressure sores.

Directly from the levels of spinal neurosegment lesion given by Sharrard result extent and degree of orthopedic deformities:

1. Patients with lesion below the Th_{12} – lower limbs in abduction at the hips, flexion at the knees, equine foot, possible subluxation of the hip joints;
2. Patients with lesion below the L_1-L_2 – flexion contracture of hip joints with external rotation of the thigh in all patients; abduction contracture of the hips in patients with preserved L_2 activity, subluxation of the hip joints, flexion contracture of the knees, equine and excavates foot, flexion finger alignment;
3. Patients with lesion below the L_3-L_4 – flexion-abduction contracture in the hips since birth, paralytic dislocation of hip joints; extension contracture or flexion limit in knees, pes equine or clubfoot (lesion below L3) or pes calcaneus (lesion below L4);
4. Patients with lesion below L_5 – stable hips, flexion contracture of hip joint, flexion limit of knees, pes calcaneus;
5. Patients with lesion below S_1 – slight flexion contracture of hip joints, congenital flat foot or pes calcaneus with claw toe [39].

Patients with flexion contracture of the knee more than 20 degrees [40], flexion contracture of the hip more than 30 degrees [41], as well as unilateral dislocation of the hip resulting in shortening of the lower limb more than 3 cm [42] are less likely to achieve community and household level of ambulation.

It turns out that the possibility of walking is highly influenced by the age. Hoffer and col. [32] found that, despite intensive therapy, patients with thoracic lesson are not able to reach a functional level of walking, and the majority of patients with lumbal lession deteriorate between the ninth and the seventeenth year of life. The reasons for the deterioration were fractures, progressive deformities of the spine and increasing spasticity. Similar conclusions can be drawn from studies Asher and Olson [43], who showed that in patients with thoracic level lesson, the greatest factor influencing the ability to walk was an age and also knee and ankle contractures. The oldest walking patient in this group was 12 years and 8 months. The main cause of walking deterioration or improvement was patient's motivation.

Among the patients studied by Williams et al. [44], the average age to stop walking in a group

with lumbar and thoracic lesion was about 7 years, and patients with the lower lumbar lesion was 9 years. The possibility of walking is also affected by age at the start of walking. Statistically, if a child with myelomeningocele will not be able to become independent at the age of six years, it is unlikely that it will be walking. Moreover, a gait manner established to ten years of age is likely to be continued later [45]. The child's sex affects deterioration of gate ability. The study of De Souza et al. [34] showed gait deterioration with age in less than half of the boys and in two thirds of girls.

In order to ensure the ability to verticalization and ambulation, patients with myelomeningocele require a different type of orthosis, which depends on the extent of the spinal cord lesion:

1. Patients with lesion at the S_2 level initially doesn't need orthosis, although at a later stage may require foot orthoses (FO);
2. Patients with lesion at the L_{4-5} level may require only ankle-foot orthoses (AFO) or supramalleolar orthoses (SMO);
3. Patients with lesion at the L_{3-4} level can start verticalization using parapodium, and then using AFO attached to twister cables or a single lateral upright and pelvic band, or standard knee-ankle-foot orthoses (KAFO);
4. Patients with lesion above L_2 level initially require hip-knee-ankle-foot orthoses (HKAFO) as a parapodium, and then after 36 months of development age it is recommended to switch into reciprocating gait orthosis (RGO) [46].

Reciprocating gait orthosis is the only alternative to ride in a wheelchair for children with myelomeningocele at the thoracic level. It may be used in patients with no active hip flexors as well as contractures up to 35°. The only conditions to be fulfilled are: adequate upper extremity strength, coordination and motivation for verticalization and ambulation [47]. Unfortunately, ambulation using RGO is slow and exhausting, which makes the majority of patients move to wheelchair after a certain period of time. Average oxygen consumption during walking in the RGO is in fact 1.0 mL/kg/m at a speed of 0.2 to 0.3 m/s, compared to 0.176 mL/kg/m and speed of 1.28 m/s at not disabled people [48].

Problems with self-care

Children born with myelomeningocele and accompanied hydrocephalus have significant limitations in fine motor skills, resulting in problems with basic self-care activities. For this reason, these children are often diagnosed as intellectually disabled, which may misdirect their further rehabilitation. It turns out that despite the disorders in learning, memory and executive functions, these children usually have normal intelligence. Research of Lindquist et al. [49] showed that 33% of children had IQ falls in range 85-115, and 30% were within 70-85.

Because of cerebellum dysmorphology, children with myelomeningocele have motor deficits characteristic for individuals with cerebellum lesion, including the classic cerebellar triad of symptoms: ataxia, dysmetria, and dysarthria. These symptoms cause that these children have problems with the function of the upper limb and hand in daily activities, as well as the eye-hand coordination and drawing and writing [50]. Research of Dennis et al. [51] showed that individuals with myelomeningocele less accurately perform the finger-nose-finger test and rapid alternating movements of the hands, especially during the additional task load, and fare worse than the control group in the motor independence. Turner [52] conducting formal coordination and dexterity tests in 33 patients with myelomeningocele aged from 4 to 17 years, received 59% of the efficiency of use of the hand. Only two children had upper limb function in norm, and 85% of children had cerebellar ataxia. Similar results were obtained by Jansen et al. [53], who studied 25 children with spina bifida in age from 5 to 19 years. In this group, only two patients had normal hand function in neurological examination. On the other hand, Norrlin et al. [54] drew attention to the problem of poor precision and coordination of movement during reaching, especially in children with symptoms of brainstem dysfunction. Another reason for the reduced hand function in children with myelomeningocele is wrong kinesthesia, which has an impact on acquiring and performance of skills such as dressing, eating, and writing. Hwang et al. [55] tested 21 children with myelomeningocele aged from 6 to 12 years, testing the ability to map the position of hands with visual and kinesthetic hints. The task was performed correctly by 73% of children with spina bifida, compared to 87% of children in the control group. Children with myelomeningocele were also slower than children in the control group.

Factors that greatly limit activities of daily living are deformities of the spine, which develop in 90 percent of children with myelomeningocele to the tenth year of life. The most of them (82.5%) is scoliosis, and about 20% is lumbar kyphosis [56].

Progressive scoliosis, especially with curves greater than 40° and pelvis slope greater than 25° may result in loss of walking opportunities, as well as the problem of maintaining a stable sitting posture. Because of the progressive nature of the deformation and poor tolerance for orthopedic supply, conservative treatment of scoliosis is usually ineffective in these patients [57]. To improve the functional capabilities they undergo the spinal fusion, which gives good results in terms of independence in self-care activities (especially dressing and self-catheterization of the bladder) at the expense of acquired walking opportunities, especially in non-ambulators [58]. Kyphosis, with a curvature peak usually located on the second lumbar vertebra is deformation, which the most worsens prognosis of the child with myelomeningocele. In case of kyphosis progression, a child has a problem with balance in sitting, achieving comfortable position in a wheelchair, and there are problems with breathing and nutrition. The poor balance in sitting makes difficult the manual activities [59]. The increase in bending in the ventral direction can lead to difficulties with the outflow of urine, self-catheterization of the bladder and perineal hygiene, as well as cause problems with abdominal fistulas. On the other hand, compensatory thoracic and cervical lordosis, arising as a result of efforts to obtain upright position leads to further deterioration of respiratory function [60].

A significant impact on the self-care abilities has also spasticity, which can be observed in some patients with myelomeningocele. Mazur et al. [61] classified the patients according to the level of neurological lesion and the degree of spasticity, distinguished the following groups:

1. Group IA (flaccid paralysis of the lower limbs, upper limbs agility) – normal neurological functions above the level of lesion, lack of sensory, motor and reflex function below lesion.
2. Group IB (flaccid paralysis of the lower limbs, spasticity in the upper limbs).
3. Group IIA (spastic paralysis of the lower limbs) – upper motor neuron paralysis below the area of lesion, manifested by spasticity and exaggerated reflexes.
4. Group IIB (spasticity in the upper and lower extremities).

It has been shown that the upper limb spasticity affects the independence of activities of daily living: the majority of patients with normal upper limb (81% of group IA and 70% of group IIA) was independent in terms of eating, clothing and personal hygiene, as compared to a much smaller number of patients with spasticity (60% in the group IB, 29% in group IIB). Spasticity in the upper limbs significantly worsened the ability to walk due to muscle weakness and poor coordination, making difficult the use of crutches.

A common problem in patients with myelomeningocele is incontinence associated with neurogenic bladder dysfunction and neurogenic ano-rectal canal. Only 3% of children with myelomeningocele controls urination in natural way. Remaining patients suffered malfunction, the severity of which depends on the level of spinal cord neurosegment lesion. In a case of thoraco-lumbar lesion, upper motor neurone is partially damaged with intact spinal reflex. These patients outwardly control urination due to pelvic floor muscles and external urethral sphincter spasticity, but the intrabladder high pressure causes rapid damage to the upper urinary tract. On the other hand, in case of the lower lumbar and lumbo-sacral segments lesion, lower motor neuron is damaged, which is associated with flaccid sphincter and passive type of urinary incontinence. These patients have a constant leakage of urine, but a small amount of urine remains in the bladder and causes urinary tract infections [62].

An effective way to prevent kidneys damage, recurrent an infection of the urinary tract, as well as to some extent solves the problem of incontinence is the clean intermittent catheterization – CIC. It has been shown that regularly performed clean intermittent catheterization procedure with proper pharmacological therapy, allows urinary control in 93% of patients during the day and in 87% of patients during the night [63]. One of the major purposes bounded up with functional independence of the child with myelomeningocele is teaching him clean intermittent catheterization before reaching school age. This activity usually carry out every three hours a day, requires the child to achieve some early skills: preparing the necessary equipment, undressing in order to access to the urethra, washing hands, preparing and placing a catheter in the urethra, empting bladder and washing after the whole operation. According to Donlau at al. [64], independence in the field of clean intermittent catheterization has reached only 48% of children.

It turns out that the greatest factor influencing the social activity and quality of life in patients with myelomeningocele is the faeces incontinence [65], due to neurogenic bowel dysfunction. This problem affects 90% of patients with myelomeningocele and probably due to frequent episodes of relaxation of the internal anal sphincter [66]. The problem for

many reasons significantly reduces the independence and quality of life of patients: control of defecation takes a lot of time that could be spent on other activities, uncontrolled leakage of faeces causes social isolation, limited leisure options for sites with a customized toilet, and even makes a child dependent on the mother [67]. In order to increase the independence, patients with neurogenic bowel dysfunction requires the implementation of individually designed program of treatment, including diet, physical activity, equipment (toilet chair), rectal and oral medications, as well as periodically carried out by the patient's colon cleaning procedure. Such a procedure is performed three times a week and lasts for about two hours at one time [68].

So far, few studies have been published in which an assessment of the functional independence of children and adolescents with myelomeningocele using standardized tests. Tsai et al. [69] studied 63 patients with spina bifida, including 26 with myelomeningocele, using PEDI (Pediatric Evaluation of Disability Inventory). Among patients with myelomeningocele, only 18.7% were independent, and as many as 50% required total assistance with activities of daily living. Despite this, 94% of parents identified family's general life satisfaction as good or very good. Similarly own quality of life assessed children studied by King et al. [70], of whom 64% identified it as good and 30% as very good.

In another study, Flanagan et al. [71] compared two groups of children born with myelomeningocele aged 5-18 years, assessing the impact of accompanied hydrocephalus, age and BMI on their quality of life. The study used two standardized tools: PedsQL (Pediatric Quality of Life) and PODCI (Pediatric Outcomes Data Collection Instrument). In the group of 15 children with lesion at L_2 and above, it has been showed lower results in terms of transfers, basic transportation, sports and general physical and health condition compared to the 35 children with lesion at L_{3-5} level. Children with shunt treated hydrocephalus, older and with higher BMI got also worse results.

Matuszczak et al. [72] evaluated the degree of social adaptation to life in the family, kindergarten and school of children and adolescents operated on myelomeningocele. In the group of pre-school children, 63.6% had full contact with the environment, spoke in full sentences and were able to play with other children. In terms of independence, all the kids needed help with daily toilet and dressing, 63% ate independently, and 27% required assistance with the seating. In the case of mobility, 18% of the children moved around in a wheelchair, and it was only 21% patients without additional supplies. In the group of children aged 7-21 years, 87.9% had full contact with the environment and only 18% had difficulty in mastering the academic material. In the field of mobility, only 30% of the respondents walking independently, 28% walking with crutches, and 45.5% moved in a wheelchair. In this group, all of the children ate independently, 39.3% dressed without assistance and 60.6% required assistance in dressing, the daily toilet, washing and hygiene associated with incontinence.

Norrlin et al. [73] identified the factors affecting independence in mobility and activities of daily living, while they studied 32 children with myelomeningocele aged 6 to 11 years, using the PEDI. Children that needed help with activities of daily living more frequently showed early signs of brain stem dysfunction and scoliosis. It was also showed a statistically significant relationship between independence and ability to walking, level of spinal cord lesion, poor hand strength and coordination, impaired visual-spatial function and executive function, as well as low executive IQ. In the group of non-ambulators and non-functional ambulators according to Hoffer classification, the only factor significantly affecting the ability of self-care was the strength of the hand. Median hand strength in this group was 65%. Authors pointed out that the way to increase independence in the field of activities of daily living in children with myelomeningocele is to enhance the strength of the hand.

Peny-Dahlstrand et al. [74] drew attention to the lower quality of performing activities of daily living in children with myelomeningocele. They studied 50 children with spina bifida aged from 6 to 14 years with the use of standardized tool called AMPS (Assessment of Motor and Process Skills) designed to assess the quality of performing of the most well-known activities of daily living by an occupational therapist. Studies have shown that children have significantly impaired the ability to execute some of their activities of daily living in an easy, efficient and safe manner. Most of the children showed a reduction in motor skills and processing ability in relation to their peers. In the case of motor skills, the greatest difficulty is caused by motor planning and adaptation (e.g. effective body setting, the ability to reach for objects and adaptation of force during movement). The most difficult process skills were: ability to adapt activities to the task (accommodation) and initialization another steps of the task. It was also noted that children

with spina bifida usually have a strategy of asking for help, even if they do not need it.

It turns out that the problem of the functional independence in children with myelomeningocele affects also young adults. Verhoef et al. [75] studied 168 patients with myelomeningocele aged 16 to 25 years, using a functional independence measure FIM. For patients without hydrocephalus, almost all were independent in all areas of the FIM scale with the exception of the bladder control. In patients with accompanied hydrocephalus, the average score for the complete independence ranged from 2.6% in the case of bladder control to 41% for communication. In these patients it was also found a direct correlation between the level of spinal cord lesion and functional independence, in particular in the fields of locomotion (score of 0% in the case of a patients with lesion above the L2 level to 83% of the patients with lesion below S1 level), self-care (from 16,7% in patients with lesion above L2 to 83.3% in patients with lesion below S1) and transferring (from 6.1% in patients with lesion above L2 to 100% of the patients with lesion below S1). In the case of locomotion, 47.9% of patients with accompanied hydrocephalus belonged to the group of "non-ambulators" according to the Hoffer classification.

Carroll [76] described the goals of functional independence of patients with myelomeningocele, depending on the level of spinal cord lesion level. According to him, patients with thoracic level lesion should strive to achieve: a good balance in a sitting position, walking at least during the first decade of life, the ability to efficient movement in the wheelchair, self-care, social acceptance, education and access to the environment. Patients with upper lumbar segments lesion should also walking at home. On the other hand, patients with lower lumbar segments lesion have a chance to achieve a community ambulation, and should standing without the aid of crutches, be able to self-care, to attend a mainstream school, and have the motivation to get independence.

Conclusions

Myelomeningocele is a birth defect of the central nervous system resulting in reduction of functional independence in both locomotion and self-care. The previous studies have shown that patients with high level of spinal cord lesion, for the most part do not reach the functional ambulation level and gait of patients moving with orthoses is associated with high energy expenditure. In addition, most patients with high level of spinal cord lesion who have reached a functional level of ambulation lost this ability in old age. Therefore, during creation of the comprehensive physiotherapy program one should consider the advisability of striving at all costs to achieve the ambulation ability in these patients, bearing in mind the favorable influence of verticalization on overall health. In children who have poor motivation for learning to walk, and there are factors potentially reducing the likelihood of ambulation, a better solution is early adaptation to move around in a wheelchair. However, you should not give up the verticalization, which could be done in a passive way during learning or practicing eye-hand coordination.

Because of the associated defects of the brain, the majority of children with myelomeningocele demonstrate impaired eye-hand coordination, which is associated with dependence on others in the many activities of daily living. Because the independence in the self-care has the greatest impact on the future independence of the child one should strive at all costs to improve the fine motor skills. Physiotherapist, designing rehabilitation program, should pay special attention to exercises strengthening upper limb muscles, hand-eye coordination improvement training, as well as increasing the efficiency of basic self-care activities. It is important to work with occupational therapist, psychologist, teacher, and speech therapist.

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