Spinal cord tethering in children with myelomeningocele

Zakotwiczenie rdzenia u dzieci z przepuklina oponowo-rdzeniową

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STRESZCZENIE

Zespół zakotwiczenia rdzenia kręgowego (TCS, ang. tethered cord syndrome) jest zróżnicowaną jednostką chorobową, spowodowaną przez nadmierne naciąganie rdzenia kręgowego. Może wystąpić w każdej grupie wiekowej, a objawia się w zależności od stanu patologicznego i wieku bólem, zmianami skórnymi, deformacjami ortopedycznymi, neurologicznymi i urologicznymi. Wczesna interwencja chirurgiczna jest związana z poprawą obrazu klinicznego, głównie w ustapieniu doliwistości bólowej, stabilizacji funkcji neurologicznych i postępu skoliozy. Zespół rehabilitacyjny prowadzący terapię dziecka z przepukliną oponowo-rdzeniową powinien pamiętać, że TSC jest istotnym powodem pogorszenia stanu neurologicznego u dziecka z rozszczepem kręgosłupa.

Streszczenie

We retrospectively assessed tethered cord syndrome among patients with myelomeningocele in the Department of Pediatric Rehabilitation in the years 2004–2011. Fifteen (16.5% of 91 subjects with myelomeningocele) patients with tethered cord syndrome were diagnosed, aged 4–17 years (mean 11.4 ± 4.4), 11 girls and 4 boys. Reduced muscle strength and increased spasticity within the lower limbs, pain in the thoracolumbar region, and deterioration of bladder function were symptoms of tethered cord syndrome. The level of spinal cord injury in Th8–L3 prevented 9 patients from walking (use of a wheelchair); 6 children – level of spinal cord injury L4 and below – were able to walk with orthopedic equipment (crutches, walker, severely handicapped). MRI revealed the various pathologic etiologies of tethering included: lipomyelomeningocele, diastematomyelia, syringohydromyelia, and hydrosyringomyelia. A neurological or rehabilitation team working with children with myelomeningocele should remember that tethered cord syndrome is a significant cause of neurological deterioration.

Słowa kluczowe: zakotwiczenie rdzenia, przepuklina oponowo-rdzeniowa, rehabilitacja

ABSTRACT

We retrospectively assessed tethered cord syndrome among patients with myelomeningocele in the Department of Pediatric Rehabilitation in the years 2004–2011. Fifteen (16.5% of 91 subjects with myelomeningocele) patients with tethered cord syndrome were diagnosed, aged 4–17 years (mean 11.4 ± 4.4), 11 girls and 4 boys. Reduced muscle strength and increased spasticity within the lower limbs, pain in the thoracolumbar region, and deterioration of bladder function were symptoms of tethered cord syndrome. The level of spinal cord injury in Th8–L3 prevented 9 patients from walking (use of a wheelchair); 6 children – level of spinal cord injury L4 and below – were able to walk with orthopedic equipment (crutches, walker, severely handicapped). MRI revealed the various pathologic etiologies of tethering included: lipomyelomeningocele, diastematomyelia, syringohydromyelia, and hydrosyringomyelia. A neurological or rehabilitation team working with children with myelomeningocele should remember that tethered cord syndrome is a significant cause of neurological deterioration.

Key words: Tethered cord syndrome, myelomeningocele, rehabilitation

INTRODUCTION

Myelomeningocele is a congenital disorder that is a Neural Tube Defect. The incidence of neural tube defects in Poland amounted to 8.1 per 10 000 births in 2003–2004 (including 7.4 per 10 000 live births), and was significantly higher than the average for all EUROCAT countries, which amounted to 2.6 per 10 000 live births [1]. Tethered cord syndrome, described by Garceau in 1953, is a pathological anchoring of the distal spinal cord which manifests as a neurological sensory and motor deficit [2]. This results in pulling with movement and progressive damage to the spinal cord [3]. Progression of neurological symptoms is very differentiated depending on the age of the child. Most cases respond to a progressive development of excess fibrous connective tissue (fibrosis) in the filum terminale or adipose infiltration [4]. Tethered cord syndrome is often accompanied by other spinal cord and brain abnormalities and urinary tract defects [5]. Early diagnosis and proper surgical treatment may be the key to success in patients with tethered cord syndrome [5–7].

Magnetic resonance is the imaging of choice for evaluating children with suspected tethered cord, as it provides exact structural information to classify the etiology and plan the surgical approach [5, 6]. Since the introduction of MRI, early diagnosis and untethering have been used to reduce potential lifelong disabilities [5, 6]. For symptomatic cord tethering, early untethering and reconstruction of the thecal sac have been advocated [7]. Not all children operated upon for tethered cord syndrome are cured. In some, a retethering phenomenon occurs that can lead to later urological, neurological, or orthopedic changes [8]. Therefore, children who undergo tethered cord release surgery are referred to continuous clinical follow-up in these subspecialties and require rehabilitation.

The aim of this study was to analyze the symptoms of tethered cord syndrome in patients with myelomeningocele.
PATIENTS AND METHODS
This retrospective study reviewed the medical records of meningomyelocele patients who were evaluated and treated at the Department of Pediatric Rehabilitation in Białystok from 2004 through 2011. During the 8-year study period, 91 children and adolescents with meningomyelocele were evaluated at our institution. All patients had undergone repair of lumbar meningomyelocele shortly after birth.

Clinical manifestations of tethered cord syndrome in children include progressive motor and sensory deterioration (particularly motor, with difficulties with gait or running); deterioration in urologic function; developmental or orthopedic deformities of the foot, leg, and spine; and, less commonly, back and leg pain [5].

Radiologic criteria: The presence of a thickened filum terminale and a low-lying conus medullaris in symptomatic patients is indicative of tethered cord syndrome [7]. Analyzed variables included patient sex, age, level of spinal cord lesion, age at surgery, presence of lipomyelomeningoceles or lipomas of the filum terminale, and brain malformations. Neurological deficits, urological dysfunction, and brain malformations were analyzed. Clinical symptoms of tethered cord syndrome (reduced muscle strength, increased spasticity and sensory deficits within the lower limbs, pain in the thoracolumbar region, and deterioration of bladder function) were also collected.

Ambulatory function in patients with myelomeningocele was defined according to Hoffer et al. [8] in 4 categories: community ambulators, household ambulators, nonfunctional ambulators, and nonambulators – scored 4 to 1. The myelomeningocele level was defined as the lowest level on the better side at which the child was able to perform an antigravity movement through the available range of joint motion. We confirmed the diagnosis of myelomeningocele in each case. The diagnosis of cord tethering on MRI was made by a qualified neuroradiologist (E.G) and was defined as the position of the conus medullaris below L3 [5] (fig. 1).

RESULTS
The results are presented in table form and a description of selected patients. Table 1 shows data on the clinical symptoms, the functional evaluation, and the results of the MRI of the spinal cord and head.

In the years 2004–2011, 15 (16.5% of 91 subjects with myelomeningocele) patients with tethered cord syndrome were diagnosed, aged 4–17 years (mean 11.4 ± 4.4), 11 girls and 4 boys. Clinical symptoms of tethered cord (reduced muscle strength, increased spasticity and sensory deficits within the lower limbs, pain in the thoracolumbar region, and deterioration of bladder function) were found in 12 patients. Motor and sensory and deficits were noted in all patients. The level of spinal cord injury in Th8–L3 prevented 9 patients from being able to walk, Hoffer’s ambulatory function scale (use of a wheelchair); 6 children – level of spinal cord injury L4 and below – were able to walk with orthopedic equipment (crutches, walker), Hoffer’s II ambulatory function scale (severely handicapped). Untethering was achieved in 6 cases. Improvement of motor function was observed in only two patients.

The various pathologic etiologies of tethering included: lipomyelomeningocele (6), diastematomyelia (6), syringohydromyelia (2), and hydrocephrosis (1). All subjects had neurogenic bladder (data not shown) and scoliosis. Four of these patients underwent surgery for scoliosis. Arnold-Chiari II brain malformation was diagnosed in 9 patients and hydrocephalus in 4. Nine patients had ventriculoperitoneal shunts.

DISCUSSION
In our study, 16.5% of 91 patients with meningomyelocele developed tethered cord syndrome. This incidence is in agreement with the 3 to 15% incidence that is commonly cited in the literature [5, 9, 10]. In the group of 15 analyzed patients, more than half used a wheelchair, and 5 patients had great difficulty moving due to significant damage to neurosegments. Our observations indicate that the main symptoms suggesting tethered cord syndrome include: reduced muscle strength, increased spasticity and sensory deficits within the lower limbs, pain in the thoracolumbar region, and deterioration of bladder function in urodynamic test. These observations are consistent with previous reports. An increase of neurological deficits in a period of rapid growth or significant increase in body weight was the most alarming. It was found that 10% to 30% of children developed symptoms of tethered cord after treating the defect [7]. They distinguished six common clinical symptoms of tethered cord, in particular: increasing reduction in lower limb muscle strength, impaired gait function, progressive lateral curvature of the spine, pain, orthopedic deformities, and urological disorders. Mehta [8] et al. observed as the most common symptoms of tethered cord: urination disorders (87%), muscle weakening (80%), gait dysfunction (78%), and loss of sensation in

Fig. 1. Lumbosacral spine magnetic resonance imaging of patient with TCS (T1-weighted longitudinal view). Ryc. 1. Rezonans magnetyczny kręgosłupa lędźwiowego pacjenta z zakotwiczeniem rdzenia (obraz T1-zależny).
<table>
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<th>Sex</th>
<th>Age (years)</th>
<th>Surgery</th>
<th>Spinal cord damage level</th>
<th>Motor function before surgery</th>
<th>Motor function after surgery</th>
<th>Scoliosis TH/L Cobb angle</th>
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<th>Brain malformation</th>
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<td>112 #</td>
<td>L1-L3-diastematomyelia</td>
<td>Arnold-Chiari II</td>
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</table>

NA – Not applicable; # – surgery for scoliosis.
distal sections of the lower limbs (61%). However, Kushel [9] reports that tethered cord syndrome occurs in 10–75% of children after myelomeningocele and lipomyelomeningocele correction, and lists age 4 to 12 years as the main manifestation period of neurologic, urologic, proctologic and orthopedic symptoms. Our patients, aged 5 to 17 years with a significant majority of girls (11/15), also reported a deterioration in functional status between the ages of 8 to 13. Guz [10] et al. report that tethered cord syndrome is often accompanied by other spinal cord abnormalities such as: hydromyelia (19.4%), diastomatomyelia (8.3%), syringomyelia; brain abnormalities: hydrocephalus (16.6%) and Chiari malformation (5.5%); and urinary tract defects: bladder pseudodiverticula (46.2%), urinary reflexes into the ureters (30.8%), hydronephrosis (15.4%), and duplex kidney (15.4%). The neurological symptoms were dominated by: paresis (33.3%), urination and bowel movement disorders (22.2%), and spinal pain (13.9%). Our observation of spinal cord defects included: lipomyelomeningocele, syringomyelia, diastomatomyelia, brain defects, Chiari II malformation, agenesis of the corpus callosum, and hydrocephalus. Erkan [11, 12] and Beaumont [13] described syringomyelia accompanying tethered cord. Beaumont et al. [13] found no significant differences in age, sex, coexisting pathology or preoperative symptoms between tethered cord syndrome and tethered cord syndrome with terminal syringomyelia.

The neurological and rehabilitation team must take into account the fact that tethered cord syndrome may manifest itself late and deceitfully in the form of progressive gait disturbances, atrophy of various muscle groups or an entire limb, loss of reflexes, sensation in individual dermatomes, sphincter disorders, pain in the buttocks and anus, and in other areas of the pelvis [14]. In the study by Perenc [15], the incidence of the Arnold Chiari II malformation was 14.28%, and the incidence of agenesis of the corpus callosum was 21.43%. In our group of patients, more than half had Arnold Chiari II malformation. A team of specialists treating a child with myelomeningocele must consider that the morphological changes in the central nervous system impair its functioning – this may be expressed by: epilepsy, apnea, vision, speech, cognitive and mental development disturbances, the development of gross and fine motor skills, as well as pain – which should be taken into account in the differentiation in case of functional deterioration. According to Guz [16] et al. the imaging method of choice for tethered cord syndrome is MRI, and computed tomography as a complementary method, which better reflects the nature of the defects of the skeletal system. We observed rapid scoliosis progression during pubertal growth spur in our group of patients. According to McGirt [17] et al. in many patients the spine deformity still progresses after spinal cord release, but the risk factors for scoliosis progression after untethering remain unclear. Children with tethered cord syndrome scoliosis should be closely monitored for curve progression with radiographs. It is also important to perform diagnostic imaging in the event of prolonged spinal pain, especially not relieved by analgesic treatment and/or physiotherapy. Due to the complex pathogenesis, pain of the spine and surrounding structures constitutes a serious diagnostic and therapeutic difficulty. In 80-90% of the cases, the cause of pain is unknown and only in 10–20% of adult patients it is possible to determine the etiologic factor during a one-year long observation. Back pain can occur especially during puberty, due to rapid growth of the skeletal system and lack of proper hormonal balance [18]. This hypothesis is supported by papers describing the relationship between pubertal growth spurt and degeneration of the spine, increased susceptibility to incidence of microdamages and subsequent pain disclosure.

Early diagnosis improves the prognosis for patients and enables starting treatment quickly, which reduces local damage of distant pathology. Most authors [19, 20] believe that tethered cord syndrome is dynamic; the progression of changes over time is noticeable. Patients who do not undergo untethering surgery may need more orthopedic and urological procedures, and there is a risk of irreversible urinary incontinence. Intraoperative monitoring of somatosensory evoked potentials – which distinguishes sensory and motor roots, adhesions and scar tissue and assesses the integrity of the reflex arc at the core - is a significant advancement in the field and reduces iatrogenic injuries during surgery [21]. This technique is a valuable tool that increases patient safety and treatment effectiveness, even at the cost of its long duration. The earlier cord untethering treatment is performed the better the effect, but it should be kept in mind that the problem also occurs in children with occult spina bifida. In our group of patients, similarly to the observations of Bowman [20], the most effective symptoms of untethering were pain relief, improved muscle strength, reduced spasticity and improved gait function in patients moving about with the help of orthopedic equipment. Similarly to Selden [22] and Sarwark [23], we found a significant improvement in miceturition disorders in the postoperative evaluation of the bladder. However, we did not achieve suppression of scoliosis progression and increase in foot bone deformities, which required surgical treatment in patients with severe spinal cord injury. Al-Holou [2] et al. reported that long-term results of tethered cord surgical treatment are generally unsatisfactory and motor function and weakness improved in 19% of patients. To a greater extent, this improvement pertains to lower limb function rather than the sphincter.

Congenital spinal lipomas are an important cause of tethered cord syndrome. Prophylactic surgery for asymptomatic congenital spinal lipomas remains a debatable issue. In the literature one finds both proponents and opponents of prophylactic surgery. Conservative management of these lesions is associated with many drawbacks [24, 25]. Currently, with better understanding of the anatomy of spinal lipomas and with advances in microneurosurgical techniques, surgery for these lesions is safer. Over the past decade, there has been accumulating evidence in favor of prophylactic surgery. Many authors recommend prophylactic surgery, which is safe and useful [24–26]. Myelomeningocele is a cause of serious motor development. The rehabilitation methods do not effects on motor function, but effect on pathological reflexes during infancy and
stimulate psychomotor development. The ability of walking is determined mainly by the level of spine injury[27]. Patients with myelomeningocele require special care and interdisciplinary cooperation of the therapeutic team with neuropediatrist [28].

CONCLUSIONS
Early diagnosis and proper surgical treatment may be the key to success in school-age children, adolescents and young adults with tethered cord. The decision to operate a child with tethered cord syndrome should be undertaken on the basis of consistent clinical evidence. Surgery should be performed before any irreversible damage to the cord occurs in the presence or exacerbation of neurological, orthopedic and urological symptoms.

REFERENCES