Spinal cord tethering in children with myelomeningocele

Zakotwiczenie rdzenia u dzieci z przepukliną oponowo-rdzeniową

Bożena Okurowska-Zawada[1], Elżbieta Gościk[2], Wojciech Kułak[1], Dorota Sienkiewicz[1], Grażyna Paszko-Patej[1]

[1]Klinika Rehabilitacji Dziecięcej z Ośrodkiem Wczesnej Pomocy Dzieciom Upośledzonym „Dać Szansę", Uniwersytet Medyczny w Białymstoku
[2]Zakład Radiologii Dziecięcej, Uniwersytet Medyczny w Białymstoku

STRESZCZENIE

Zespół zakotwiczenia rdzenia kręgowego (TCS, ang. *tethered cord syndrome*) jest zróżnicowaną jednostką chorobową, spowodowaną przez nadmierne naciąganie rdzenia kręgowego. Może występować 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 ustąpieniu dolegliwości bólowych, 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.

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).

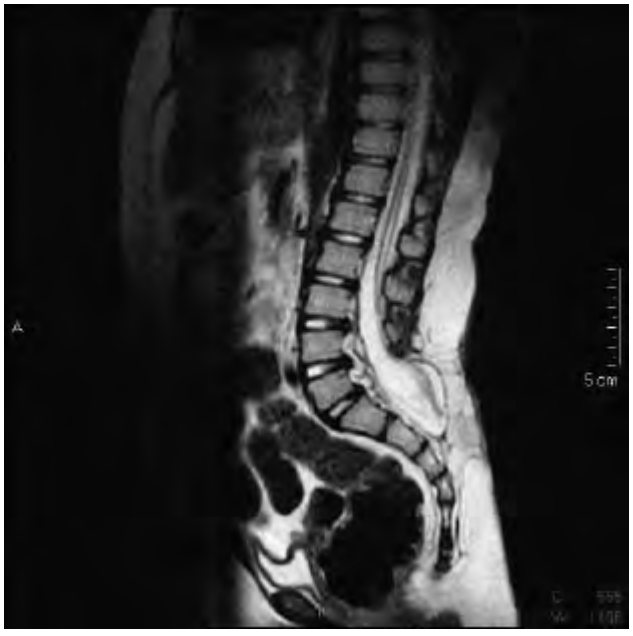

Fig. 1. Lumbosacral spine magnetic resonance imaging of patient with TCS (T1-weighted longitudinal view). *Ryc. 1. Rezonans magnetyczny kręgosłupa lędźwiowego pacjenta z zakotwiczeniem rdzenia (obraz T1-zależny).*

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 I 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 hydrosyringomyelia (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

Tab. I. Data on the clinical symptoms, the functional evaluation, and the results of the MRI of the spinal cord and head
Tab. I. Dane dotyczące objawów klinicznych, ocena funkcjonalna oraz wyniki MRI rdzenia kręgowego i głowy

No	Sex	Age (years)	Surgery	Spinal cord damage level	Motor function before surgery	Motor function after surgery	Scoliosis TH/L Cobb angle	Spinal cord MRI	Brain malformation	Ventriculo-peritoneal Shunt
1	F	16	yes	Th8-L3	II	I	87 #	no junction of L4-L5-S1 arches lipomyelomeningo-cele	Arnold-Chiari II	present
2	F	17	yes	Th8-L3	I	I	45	no junction of L4-L5-S1 arches lipomyelomeningo-cele	Arnold-Chiari II	present
3	M	10	no	Th8-L3	I	NA	40	diastematomyelia	Arnold-Chiari II	no
4	F	9	no	L4-S1	II	NA	45	no junction of L5-S arches, diastemato-myelia	Arnold-Chiari II	present
5	F	11	yes	L4-S1	II	II	28	lipomyelomeningo-cele	Hydrocehalus	present
6	M	11	no	Th10-L5	II	NA	19	no junction of L4-L5 -S arches	Hydrocepalus	present
7	F	17	yes	Th8-L3	I	I	94 #	lipomyelomeningo-cele	Hydrocepahlus	present
8	M	15	yes	L4-S1	II	II	7	no junction of L3-S2 arches	-	no
9	F	7	no	L4-S1	II	NA	5	no junction of L4-S arches Th12-L3 syringohy-dromyelia	Arnold-Chiari II	present
10	F	12	yes	L4-S2	I	I	67 #	no junction of L1-L3 arches diastemato-myelia	-	no
11	F	9	no	L4-S2	I	II	12	no junction of L2-L5-S arches diastematomyelia	Arnold-Chiari II	no
12	F	5	no	Th8-L3	I	NA	6	lipomyelomeningo-cele	Arnold-Chiari II	present
13	M	3	no	L4-S2	II	NA	38	no junction of L3-L5-S arches lipomyelomeningo-cele	Hydrocephalus	Present
14	F	12	no	Th8-L3	I	NA	30	C7-Th7-diastema-tomyelia L2-L3-dia-stematomyelia L1-S1-hydrosyringo-myelia	-	no
15	F	17	no	Th8-L3	I	NA	112 #	L1-L3 -diastemato-myelia	Arnold-Chiari II	no

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 neurological, urological, 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 refluxes 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, diastematomyelia,* 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 spurt 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 ocult 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 micturition 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

[1] Okurowska-Zawada B., Kułak W., Otapowicz D., et al.: Quality of life in children and adolescents with cerebral palsy and myelomeningocele. Pediatr Neurol 2011; 45: 163–168.

[2] Al-Holou W.N., Muraszko K.M., Garton H.J., et al.: The outcome of tethered cord release in secondary and multiple repeat tethered cord syndrome. J Neurosurg Pediatr 2009; 4: 28–36.

[3] Hudgins RJ., Gilreath C.L.: Tethered spinal cord following repair of myelomeningocele. Neurosurgical Focus 2004; 16; 7.

[4] Yamada S., Knerium D.S., Mandybur G.M., et al.: Pathophysiology of tethered cord syndrome and other complex factors. Neurol Res 2004; 26: 722–726.

[5] Lew S.M., Kothbauer K.F.: Tethered cord syndrome: an updated review. Pediatr Neurosurg 2007; 43: 236–248.

[6] Phuong L.K., Schoeberl K.A., Raffel C.: Natural history of tethered cord in patients with meningomyelocele. Neurosurgery 2002; 50: 989–993.

[7] Filippidis A.S., Kalani M.Y., Theodore N., et al.: Spinal cord traction, vascular compromise, hypoxia, and metabolic derangements in the pathophysiology of tethered cord syndrome. Neurosurg Focus 2010; 29: 9.

[8] Hoffer M.M., Feiwell E., Perry R., et al.: Functional ambulation in patients with myelomeningocele. J Bone Joint Surg 1973; 55: 137–148.

[9] Mehta V.A., Bettegowda C., Ahmadi S.A., et al.: Spinal cord tethering following myelomeningocele repair. J Neurosurg Pediatr 2010; 6: 498–505.

[10] Tamaki N., Shirataki K., Kojima N., et al.: Tethered cord syndrome of delayed onset following repair of myelomeningocele. J Neurosurg 1988; 69: 393–398.

[11] Erkan K., Unal F., Kiris T.: Terminal syringomyelia in association with the tethered cord syndrome. Neurosurgery 1999; 45:1351–1359.

[12] Erkan K., Unal F., Kiris T., et al.: Treatment of terminal syringomyelia in association with tethered cord syndrome: clinical outcomes with and without syrinx drainage. Neurosurgical Focus 2000; 8: 9.

[13] Beaumont A., Muszynski C.A., Kaufman B.A.: Clinical significance of terminal syringomyelia in association with pediatric tethered cord syndrome. Pediatr Neurosurg 2007; 43: 216–221.

[14] Rinaldi F., Cioffi F.A., Columbano L., et al.: Tethered cord syndrome. J Neurosurg Sci 2005; 49: 131–135.

[15] Perenc L., Kwolek A.: Badania nad współistnieniem przepukliny oponowo-rdzeniowej oraz innych anomalii rozwojowych mózgowia i czaszki. Przegl Med Uniwer Rzeszow 2005; 1: 23–26.

[16] Guz W., Walawska A., Samojedny A., et al.: Zespół zakotwiczenia rdzenia kręgowego – różnorodność i charakterystyka obrazów w badaniach rezonansu magnetycznego kręgosłupa piersiowo-lędźwiowo-krzyżowego. Przegl Med Uniwer Rzeszow 2007; 2: 161–174.

[17] McGirt M.J., Mehta V., Garces-Ambrossi G., et al.: Pediatric tethered cord syndrome: response of scoliosis to untethering procedures. Clinical article. J Neurosurg Pediatr 2009; 4: 270–274.

[18] Wedderkopp N., Andersen L.B., Froberg K., et al.: Back pain reporting in young girls appears to be puberty-related. BMC Musculoskeletal Disord 2005; 6: 52–57.

[19] Selçuki M., Umur A.S., Duransoy Y.K., et al.: Inappropriate surgical interventions for midline fusion defects cause secondary tethered cord symptoms: implications for natural history report of four cases. Child's Nerv Syst 2012; Feb 17.

[20] Bowman R.M., Mohan A., Ito J., et al.: Tethered cord release: a long-term study in 114 patients. J Neurosurg Pediatr 2009; 3: 181–187.

[21] Pouratian N., Elias W.J., Jane J.A Jr., et al.: Electrophysiologically guided untethering of secondary tethered spinal cord syndrome. Neurosurg Focus 2010; 29: 3.

[22] Selden N.R., Nixon R.R., Skoog S.R., et al.: Minimal tethered cord syndrome associated with thickening of the terminal filum. J Neurosurg 2006; 105(3 Suppl): 214–218.

[23] Sarwark J.F., Weber D.T., Gabrieli A.P., et al.: Tethered cord syndrome in low motor level children with myelomeningocele. Pediatr Neurosurg 1996; 25: 295–301.

[24] Van Calenbergh F., Vanvolsem S., Verpoorten C., et al.: Results after surgery for lumbosacral lipoma: the significance of early and late worsening. Childs Nerv Syst 1999; 15: 439–442.

[25] Kang H.S., Wang K.C., Kim K.M., et al.: Prognostic factors affecting urologic outcome after untethering surgery for lumbosacral lipoma. Childs Nerv Syst 2006; 22: 1111–1121.

[26] Kumar A., Mahapatra A.K., Satyarthee G.D.: J Congenital spinal lipomas: Role of prophylactic surgery J Pediatr Neurosci 2012; 7: 85–89.

[27] Okurowska-Zawada B., Sobaniec W., Kułak W., et al: Analiza rozwoju motorycznego dzieci z przepukliną oponowo- rdzeniową i stosowane metody rehabilitacji. Neurol Dziec 2007; 17: 31–38.

[28] Okurowska-Zawada B., Kułak W.: Etapy współpracy lekarza rehabilitacji i neurologa dziecięcego w terapii pacjentów z przepukliną oponowo-rdzeniową. Neurol Dziec 2011; 20: 73–79.

Adres do korespondencji:
Bożena Okurowska-Zawada, Klinika Rehabilitacji Dziecięcej z Ośrodkiem Wczesnej Pomocy Dzieciom Upośledzonym „Dać Szansę", Uniwersytet Medyczny w Białymstoku, ul. Waszyngtona 17, 15-274 Białystok, e-mail: zawada.bozena@wp.pl