International Journal of BioMedicine 4(1) (2014) 36-39

INTERNATIONAL JOURNAL OF BIOMEDICINE

PROBLEMS OF PEDIATRICS

Neurological Disorders in Children with Cerebrospinal Hernias

Nargiza N. Ergasheva

Tashkent Pediatric Medical Institute
Tashkent, Uzbekistan

Abstract

This study presents an analysis of the neurological manifestations in 88 children with congenital cerebrospinal hernia (CCH) from the age of one day right up to 12 years old. Hernia in the forms of meningomyelocele and meningoradiculocele is most frequently localized in the lumbosacral spine. The frequency and severity of these neurological disorders are dependent upon the dimensions and the morphological forms of the hernia, as well as the nature of the complications and associated conditions.

Keywords: congenital cerebrospinal hernia, clinical symptoms, children.

Introduction

Congenital anomalies (CA) of the different organs and systems occur in 5% of the newborns. Among the various causes of perinatal, neonatal and infant mortality, the frequency of the occurrence of CA is 25%. Among the congenital malformations (CM), anomalies of the central nervous system (CNS) account for 30%. They occur most frequently, followed by complications and high mortality, with disability for long periods of time or throughout life. Most of these patients are considered virtually incurable and die at an early age or suffer from various forms of congenital dementia, lagging behind in psychomotor development. All of the above issues pose a great clinical and social problem [1, 2]. The population frequency of CM according to the WHO Expert Committee varies in different countries, from 2.7 to 16.3%, averaging 4.6%. Neural tube defects take the leading position among all human congenital anomalies (10-30%). The true incidence of CM, including the CNS, however, remains unspecified, probably due to the difficulties in the diagnosis and various approaches to their registration. Currently, methods are available to establish the presence of CNS defect in the fetus even during the early stages of pregnancy [3-5]. One of the most important problems is the issue of the treatment and rehabilitation of children with CCH because of the growth in CCH frequency, severity and outcome. CCH constitutes 65% of the neural tube defects occurring in 0.4-4.2 cases per 1000 live births [6]. Recently, many countries across the world, have recorded a steady rise in the number of newborns with CCH [7-9]. The analysis of causes for stillbirth in Uzbekistan established that the CNS anomalies occur in 78.2% among the stillborn with CM. The causes of early neonatal mortality in 17.1% of cases were due to 32 species of the CM while the second place was occupied by the malformations of the nervous system [8, 10].

The most adverse outcomes of CCH are progressive hydrocephalus, peripheral paresis and lower limb paralysis, pelvic disorders such as urinary and fecal incontinence, leading to disability in children [6, 11, 12]. The CCH survivors have pronounced neurological disorders and relate to risk groups for disruption of social adaptation [2, 5, 7, 10].

The problem of reducing child disability associated with spinal cord and spinal column malformations is an important social problem facing many countries in the world, and is awaiting a universal solution.

The purpose of our study was to determine the nature and severity of the neurological disorders in children with CCH.

Material and Methods

In this study, we examined 88 children with CCH, between one day old and 12 years of age (45/51.1% boys and 43/48.9% girls). The examination included clinical and neurological exams, clinical and biochemical analyses of blood and urine, computer-tomography of the spine and spinal cord, neurosonography and ultrasound of the internal organs. To determine the nature of the disorders of defecation and

***Corresponding author:** **Nargiza N. Ergasheva.** *Tashkent Pediatric Medical Institute. Tashkent, Uzbekistan.*
E-mail:tpmiim@gmail.com

urination, a special urological and proctologic examination was conducted.

The following clinico-morphological forms of CCH were verified (Fig.1): meningocele - in 24 (27.3%) of the cases; meningomyelocele - in 31 (35.2%) of the cases; meningoradiculocele - in 17 (19.3%) of the cases; myelocystocele - in 8 (9.1%) of the cases; rachischisis - in 3 (3.4%), of the cases; hidden cleft (spina bifida) - in 2 (2.3%) of the cases; and spina bifida complicata - in 3 (3.4%) of the cases. CCH in the cervical vertebrae were observed in 2 (2.3%) patients - in the thoracic vertebrae - in 2 (2.3%); in lumbar region - in 38 (43.2%); in the sacrum – 13 914.8%); and in the lumbosacral region - in 33 (37.5%).

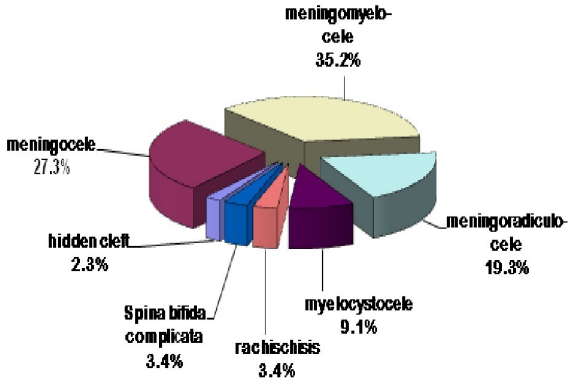


Figure 1. Clinico-morphological forms of CCH

Surgical treatment for CCH was given to 74 (84.1%) patients. In 14 (16%) children with rachischisis and severe forms of CCH combined with multiple malformations and severe manifestations, including lower sluggish paraplegia and other co-morbid conditions, surgical treatment was not performed.

Results and Discussion

Among the 88 patients, 78 (88.6%) were born at term, while 10 (11.4%) were premature infants in the 34-36 week gestation period. Body weight of the infants ranged from 1,916 to 4,600g, the average weight being 3,321.1±48.4g. A lag in the physical development was noted in 57 (64.8%) infants and a dramatic gap - in 4 (4.5%). In other cases, the patient height and weight corresponded to the age indicators.

Clinical and neurological examinations of the CCH patients revealed a combination of diffuse or focal lesions of the CNS. The nature of the pathological changes in the CNS was characterized by diversity. Focal neurological deficits were characterized with the clinical manifestations of conduction disturbance in the roots of the cauda equina and pelvic disorders. In rare cases, there were signs of spinal cord disturbance at the cone and epicone levels.

Among the 88 children surveyed, CCH was accompanied by hydrocephalus in 22 (25%) patients. In 6 (27.3%) patients, the clinical signs of hydrocephalus, which were very pronounced, appeared during the first hours of life which facilitated the diagnosis. In 11 patients (50%), the symptoms of hydrocephalus (tension of the large fontanelle, prevalence of the head circumference over that of the chest, dehiscence of the skull bones) appeared before one year of life. Progression of the hydrocephalus was observed in 5 (22.7%) patients. Increasing hydrocephalus occurred mostly in patients with a suppuration of hernia shells or a very large hernia. Comprehensive medical treatment was effective in only 6 (27.3%) children, which helped to maintain their condition in stable status. The clinical symptoms were multifarious, depending upon the location and size of the hernia defect. Deficiency symptoms of the cranial nerve innervations in CCH patients were explained with concomitant hydrocephalus and birth defects. In these patients, limited eyeball mobility was noted in four (18.2%) of the cases, convergent strabismus in three (13.6%) of the cases, smoothness of the nasolabial fold in six (27.3%) patients, and reduced swallowing reflex and choking when swallowing, in five (22.7%) patients.

Assessment of the cognitive function revealed a safety intellect with unexpressed psychological disorders in 47 (53.4%) patients. Motor development delay was noted in 41(46.6%) patients. Indicators such as the child's reaction to his surroundings, movement activity, physiological reflexes, facial expressions, movement coordination, verbal development, ability of comprehension, memory, activity, and the presence of certain symptoms of mental infantilism were estimated with age norm.

In severe forms of CCH accompanied by pelvic organ dysfunctions, in almost of all cases, signs of cerebroasthenic syndrome were evident in the form of low efficiency, high emotional lability, irritability and apathy. Among the six children of school age, 4 (66.7%) began to study at 7 years of age in a normal school and two (33.3%) children were taught at home by a special program and needed to limit the psycho-emotional load.

Violation of the functions of the pelvic organs is the most frequent and serious complication of CCH in children. General clinical and neurological examination and data regarding bladder sonography (frequency, rhythm of urination, bladder volume in the phase of accumulation and the child's ability in urine retention) and other studies formed the basis for verification of the forms of urination disorders in 84% of the patients. In children over 3 years of age, a worsening of pelvic organ dysfunction was evident. Hyper-reflex type of disorder was diagnosed in 38% patients, hypo-reflex type in 21% and detrusor-sphincter dyssynergia in 41% patients (Fig.2).

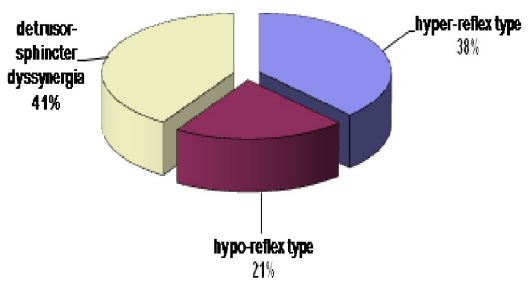


Figure 2. Disorders of urination in children with CCH

In children with meningocele (n=24), unexpressed changes were observed in the neurological status: muscular hypotonia in 1 (4.2%) patient, muscular hypertension in 4 (16.7%), hyperactivity of tendon reflexes in 4 (16.7%) and decreased sensitivity in 2 (8.3%) patients. Pelvic disorders in the form of hyper-reflex impairments were observed in 3 (12.5%) of cases and hyper-reflex bladder in 6 (25%) patients.

In patients with meningoradiculocele (n=17), meningomyelocele (n=31) and myelocystocele (n=8), pronounced impairments were observed in the nervous system. Limitation in movements of the lower limbs was observed in 16 (94.1%) patients with meningoradiculocele. In one (5.9%) case, a reduction in the anal reflex was noted. Functional disorders of the pelvic organs were mixed. Hypo-reflex type was observed in four (23.5%) patients, hyper-reflex type in eight (47.1%) patients and detrusor-sphincter dyssynergia in five (29.4%) of the cases.

In all the patients with meningomyelocele and myelocystocele restriction of active movements was noted in the lower limbs while decreased sensitivity occurred in 17 (54.8%) and 8 (100%) patients, respectively. Trophic disorders of the skin and nails were detected as pressure ulcers and venous ulcers, friability and nail discoloration. In patients with meningomyelocele, detrusor-sphincter dyssynergia was evident in 16 (51.6%) cases, while the hypo-reflex type was observed in two (6.4%) patients and the hyper-reflex type in 10 (32.2%) of the cases.

The majority of patients with myelocystocele had detrusor-sphincter dyssynergia (in 7/87.5% of the cases), whereas the hyper-reflex type was recorded only in 1(12.5%) case.

In rachischisis, marked disturbances of the neurological status, manifested as a lack of movement, numbness and trophic disorders and pelvic dysfunction as a hypo-reflex type of urination and incontinence were noted in all three patients. In one of the two children with spina bifida occulta, expressed changes in the nervous system were not noted except for the hyper-reflex type of the urinary disorders. Out of 3 children with spina bifida complicata, only one child revealed focal neurological disorders in the form of restriction of active movements, muscular hypotonia, and vegetative-trophic disorders of the skin and nails. The hyper-reflex type violation of the bladder function was noted in all the three patients.

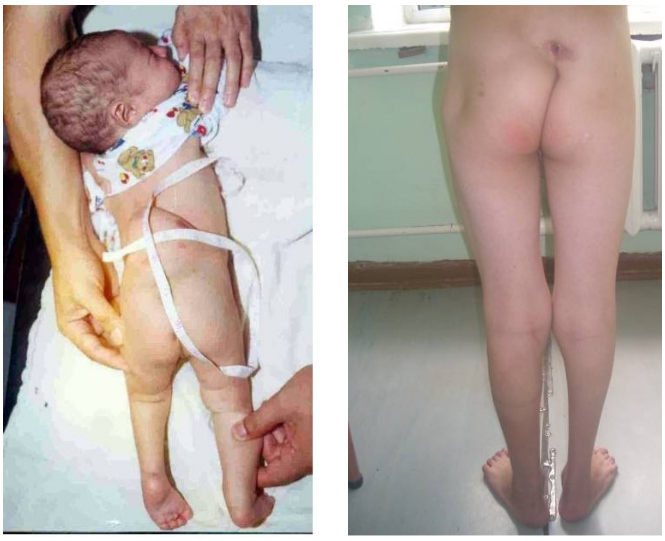
Motor impairments in the lower limbs were diagnosed in 58 (65.9%) patients. Among children under one year, these phenomena occurred in 50 (56.8%) cases. Decreased muscle tone was detected in the peripheral type. Muscle strength varied from 0 to 5 points. This indicated a limitation in leg movement up to total loss. In CCH patients, the distal regions of the lower limbs were found to be affected more often and to greater extent. Movement disorders such as monoparesis of the lower limbs were observed in 13 (14.8%) children, more often on the left side. In the proximal potions of the lower limb, more frequently, unexpressed paresis of unequal severity on both sides was noted. In all the patients, movement disorders were accompanied by numbness in the distal lower limbs, sometimes in the skin of the perineum.

Vertical stability and walking (in 18 children older than one year) were delayed by 6-12 months in four (22.2%) patients, by 13-24 months in two (11.1%) patients, and 3-4 years in two (11.1%) patients in the form of incomplete recovery of movement.

The group without motor loss included those children (n=30) with reflex disturbances in the form of moderate or severe depression of knee and Achilles reflexes, on one or both sides. Among those patients younger than one year, this observation accounted for 20%.

Sensory disturbances in the form of anesthesia and hypoesthesia were found in 39 (44.3%) cases. Anesthesia and hypoesthesia, as well as movement disorders, were noted in 37.5% in patients less than one year of age. In 12 (66.7%) children over one year of age with CCH in the form of meningocele, sensory disorders were absent. The intensity of lower limb anesthesia and hypoesthesia, in most cases, was symmetric (89.7% of the cases). Right-sided disorders were detected in one patient; left-sided in three patients. Anesthesia of the anogenital area was marked in 9 children, which indicated a spinal cord cone lesion.

A comparative analysis of the development of the lower limbs in children born normal and children with CCH showed a shortening in the length and circumference of the lower limbs in the latter that can be considered a manifestation of the trophic disorders in the form of hypotrophy or atrophy. These disorders occurred in 40 (45.4%) patients. Subsequently, they were preserved in non-operated-upon as well as in the operated-upon children. The trophic disorders of the lower limbs in 24(27.3%) patients were manifested against the background of a cooling of the distal parts and skin dryness (Fig.3a-b). In two older children, the appearance of trophic ulcers on the foot was marked.



a) ***b)***

Fig 3. Asymmetrical atrophy of the lower limbs

a) Patient N. (1 month); b) Patient Kh. (8 yrs).

In total, 14 (15.9%) children had clubfoot - on one side in 6 (42.8%) patients and on both sides in 8 (57.1%) patients. With the localization of hernias to the cervical and thoracic spine, the pattern of the most favorable clinical variant of meningocele was observed.

Conclusion

Thus, the congenital anomalies of CNS take the top position among the many congenital malformations. The frequency and severity of the neurological disorders are dependent upon the localization, morphological form of the hernia and the nature of the myelodysplasia. These disorders are characterized by flaccid paraparesis and often pronounced pelvic disorders. Pelvic dysfunctions were observed in 84% of the patients with CCH and were manifested as defecation disorders in 4% of the cases, urination disorders in 24 of the cases, with a combination of disorders in 56% of the patients. The motor, sensory and trophic disorders of the lower limbs and pelvic organ dysfunctions alone or in various combinations were redoubled with the associated vertebral-spinal abnormalities in 18.3% of the patients, hydrocephalus in 30.9%, malformations of the other organs in 69%, co-morbidities and complications associated with hernial cover in 24% of cases. The results obtained underscore the importance of improving preventive measures, the organization of which depends upon the cooperation of medical-genetic, obstetric, and pediatric services and the prenatal diagnosis of CM.

References

1. Akhmediev MM, Makhmudov ShD, Vakkasov NI, Ahmedieva Sh.R. Comprehensive treatment of congenital malformations of the primary neural tube in children. Materials of scientific and practical conference "Problems of reconstructive and restorative neurosurgery". Crimea, 7-8 October 2010; P: 11.
2. Barashnev YuI, Bakharev VA, Novikov PV. Diagnosis and treatment of congenital and hereditary diseases in children. M: Triad – X; 2004.
3. Gavryushov VV, Zubarev EA, Efimov MS. Ultrasonic diagnosis of brain damage in newborns. In: Strizhakova AN, Bunina AT, Medvedev MV, editors. *Ultrasound in obstetrics clinic*. Moscow: Medicine; 1990:190-202.
4. Nemilova TK, Voronin DV, Shapkayts VA. Antenatal diagnosis and tactics in malformations of the fetus and newborn. Handbook. SPB; 2002.
5. Syrchin EF, Zyabrov AA Voronov VG, Kutumov EB, Voronov VG, Ivanov AA, et al. Informational content of the diagnostic studies of the spinal cord and spine malformations according to evidence-based medicine. "Polenov reading". Proceedings of the All-Russian Scientific and Practical Conference. St. Petersburg; 2010:340.
6. Shodiev ASh, Mamadaliev AM, Choriyev UKh. To the issue of improving treatments for spinal hernias. "Polenov reading". Proceedings of the All-Russian Scientific and Practical Conference. St. Petersburg; 2007:296.
7. Kulakov VI, Isakov YuF, Baybarina EN. Perinatal diagnosis and treatment of congenital malformations at the present stage. Russian bulletin of perinatology and pediatrics. Moscow, 2006.
8. Kariev GM, Ahmediev MM, Ahmedieva ShR. Malformations of the spine and spinal cord at an early age. Darmon, 2010; 2 (53): 6.
9. Matveeva SP, Semenov PN, Lutsyk AA Clinico-diagnostic characteristics of congenital malformations of the spinal cord and spine. "Polenov reading". Proceedings of the All-Russian Scientific and Practical Conference. St. Petersburg; 2007:273.
10. Matchanova AT. The role of perinatal diagnosis of congenital malformations in the reduction of perinatal losses. Abstract of PhD Thesis. Tashkent; 1998.
11. Boyer KM, Yeates KO, Enrile BG. Working memory and information processing speed in children with myelomeningocele and shunted hydrocefalus: analysis of the children's paced auditory serial addition test. J Int Neuropsychol Soc 2006; 12(3):305-13.
12. Kıymaz N, Yılmaz N, Güdü BO, Demir I, Kozan A. Cervical spinal dysraphism. Pediatr Neurosurg 2010; 46(5):351-6.