# MANAGEMENT OF NEURAL TUBE DEFECTS

*Mirsad HODŽIĆ, Hasan ALTUMBABIĆ, Mirza MORANJKIĆ, Zlatko ERCEGOVIĆ, Selma JAKUPOVIĆ*

Department of Neurosurgery, University Clinical Center Tuzla, Tuzla, Bosnia and Herzegovina

Correspondence to:
Mirsad Hodžić
Department of neurosurgery
University Clinical Center of Tuzla
Trnovac bb, 75000 Tuzla, Bosnia and Herzegovina, mirsad@bih.net.ba

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**Objective** The aim of this study was to present our experience in managed neural tube defect in a consecutive series of 18 patients.

**Material and methods** In the period between October 2003 and October 2007 eighteen patients with neural tube defect (NTD) were operated at Department of neurosurgery of University clinical center Tuzla. Each patient with suspected NTD was evaluated by pediatricians, radiologists and neurosurgeons with clinical findings and ultrasound investigation, computerized tomography and/or magnetic resonance imaging scans of the spine and head.

**Results** The neurosurgical data of 18 patients with NTD who were admitted and treated at our department over the four year period were studied retrospectively. The average age at the time of presentation was 15.7 months, ranging from 1 day to 8 years. Spinal NTD was noticed in 13 (72.2%) patients and cranial in 5 (27.8%) patients. Surgery was performed on emergency basis immediately upon birth for CSF leakage in five patients (38,5%) with spinal dysraphism. No postoperative cerebrospinal fluid leakage was noticed and there was no mortality.

**Conclusion** Patients born with neural tube defect need an interdisciplinary team of specialists to oversee their developmental progress. Once born, a child with NTD must be operated in order to avoid fluid infection.

**Key words:** Neural tube defect ▪ Management ▪ Outcome

## Introduction

Neural tube defects (NTD) are malformations of the neuroectoderm associated with abnormalities of the surrounding

mesodermal structures (1, 2). The incidence has declined significantly over the last 30 years and NTD's now occur in approximately 0.8 out of 1000 total births (3). Spinal NTD encompasses defective closure of the neural tube early in fetal life and anomalous development of the caudal cell mass. Anatomic features common to the spinal NTD are anomalies in the midline structures of the back (particularly absence of some of the neural arches), along with defects of the skin, filum terminale, nerves, and spinal cord (4). Cranial NTD encompasses absence of cranial vault and protrusion of cranial contents (meninges and/or cerebral tissue) beyond the normal confines of the skull, through a defect in the cranium. Cranial dysraphism, particularly encephalocele, is far less common compared to its spinal counterpart, namely, myelomeningocele, accounting for only 8-19% of all dysraphisms (5).

In this retrospective review, we present our experience in a consecutive series of 18 patients managed successfully in our institution.

## Patients and methods

In the period between April 2002 and October 2007 of a database search, we identified 19 patients with spinal or cranial neural tube defect. One patient with cranial vault encephalocele and other severe associated anomalies died second day upon birth and he was excluded from this study. Eighteen patients were operated on at the Department of neurosurgery of University clinical center Tuzla. Diagnosis was confirmed at surgery and all of them were included in this study.

Demographic, clinical, radiological and operative data of this patient population were reviewed from the hospital charts and computerized data bank. In addition we describe the outcome and follow up after discharge from the hospital.

Each patient with clinically suspected NTD was evaluated by pediatricians, radiologists and neurosurgeons with clinical findings and ultrasound investigation, computerized tomography (CT) and/or magnetic resonance imaging (MRI) scans of the spine and head. The size and anatomical location of the lesion were noted. Associated conditions, such as, large head size, suggesting underlying hydrocephalus, were also noted. Detailed neurological examination was performed noting specifically presence or absence of any neurological deficit.

All patients with neural tube defect were routinely offered surgery. Surgical treatment of spinal NTD included dissection of neural elements with reduction the size if the fatty tumor was present in order to untether the spinal cord and repair soft tissue defect. Depending upon the anatomical location of cranial NTD, either direct excision and repair of the lesion or craniotomy and repair from inside was done. The goal of surgery was to repair the defect with no injury to existing nerve or brain function.

## Results

The neurosurgical data of 18 patients with neural tube defect who were admitted and treated at our department over the four year period were studied retrospectively. There were nine males and nine females.

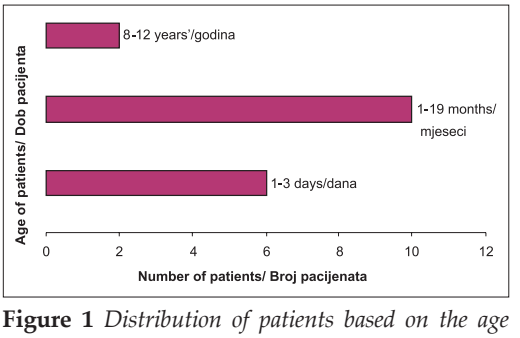


**Figure 1** *Distribution of patients based on the age groups*

**Slika 1** *Distribucija pacijenata prema dobnim skupinama*

The average age at the time of presentation was 15.7 months, ranging from 1 day to 8 years. Figure 1 summarizes the distribution of patients on the basis of age groups. Spinal neural tube defect was noticed in 13 (72.2%) patients and cranial in 5 (27.8%) patients. Table 1 summarizes localization of defect in spinal and cranial tube defects.

Distribution of NTD's according to the type was summarized in Table 2. Additional anomalies were noticed in 7 (38.8%) patients (Table 3).

**Table 1**. *Anatomical location of neural tube defect*
**Tabela 1** *Anatomska lokacija defekata neuralne cijevi*

| Location/Lokacija | No. of cases/Broj slučajeva | % |
|---|---|---|
| Spinal neural defect/Spinalni neuralni defekti | | |
| Lumbosacral/Lumbosakralni | 6 | 33.4 |
| Lumbar/Lumbalni | 4 | 22.3 |
| Sacral/Sakralni | 2 | 11.2 |
| Thoracolumbal/Torakolumbalni | 1 | 5.5 |
| Cranial neural tube defect/Kranijalni defekti neuralne cijevi | | |
| Occipital/Okcipitalni | 3 | 16.6 |
| Parieto-occipital/Parijeto-okcipitalni | 1 | 5.5 |
| Fronto-ethmoidal/Frontoetmoidalni | 1 | 5.5 |
| Total/ Ukupno | 18 | 100.0 |

**Table 2** *Distribution of NTD's according to the type*
**Tabela 2** *Distribucija defekata neuralne cijevi prema tipu*.

| Type/Tip | No. of cases/Broj slučajeva | % |
|---|---|---|
| Encephaloceles/Encefalokele | 3 | 16.7 |
| Myelomeningoceles/Mijelomeningokele | 12 | 66.7 |
| Ocult spinal dysraphism/Okultni spinalni disrafizam | | |
| Lipomyelomeningocele/Lipomijelomeningokele | 1 | 5.5 |
| Cutaneous tail/Kutani rep | 1 | 5.5 |
| Meningocele/Meningokele | 1 | 5.5 |
| Total/Ukupno | 18 | 100.0 |

**Table 3** *Additional anomalies in patients with neural tube defect*
**Tabela 3** *Udružene anomalije kod pacijenata sa defektima neuralne cijevi*

| Anomalies/Tip anomalije | No. of cases/Broj slučajeva | %* |
|---|---|---|
| Hydrocephalus/Hidrocefalus | 5 | 27.8 |
| Equinovarus/Ekvinovarus | 4 | 22.3 |
| Palatoshysis/Palatoshiza | 1 | 5.5 |
| Foramen ovale apertum/Perzistentni foramen ovale | 1 | 5.5 |

*Percent of 18 patients operated on for neural tube defect/Postotak od 18 pacijenata operiranih zbog defekta neuralne cijevi

Lower extremities palsy was present in 2 (11.1%) patients and hypotonia of lower extremities was present in 6 (33.3%) patients. An average length of hospitalization after surgery was 3.8 days (SD ± 4.75), ranging from 1 to 18 days. Wound dehiscence was noticed in one patient that underwent a revision surgery. Cranial neural tube defect was surgically approached in various ways, mainly depending on its location and type. In four patients with cranial vault encephaloceles, a direct extracranial repair was done. Frontoethmoidal defect was repaired using an extra and intradural intracranial approach with resection of encephalocele, dural plastic and bone defect reconstruction. For encephaloceles, a dural closure was performed with running sutures, in attempt to obtain watertight environment, upon resection of redundant dura. The subcutaneous layer and the skin were closed in the usual manner, using interrupted sutures.

During the closure procedure for myelomeningocele and in order to address the risk of developing subsequent dermoid tumors or lipomas, any residual tissue had been resected from the neural placode periphery. Using an interrupted monofilament sutures, pia had been closed to reconstruct neural placode into a tube. In order to reconstruct the thecal sac the dura had been closed in the midline, with nonabsorbable suture.

Surgery was performed on emergency basis immediately upon birth for CSF leakage in five patients (38,5%) with spinal dysraphism and generally within the first 24-48 hours in the absence of CSF leakage. Surgery had a goal of performing a resection of nonfunctional tissue, cord unthetering and soft tissue defect closure. Hydrocephalus associated with a myelomeningocele was treated by ventriculoperitoneal shunt.

The surgical outcome in our series was satisfactory. There were no anesthetic or procedure related complications and no cerebrospinal fluid leakage was noticed. There was no mortality. The wound dehiscence which required revision of repair was noticed in one patient. The mean follow-up period was 34 months, ranging from 4 months to 5.5 years (SD ±19 months).

## Discussion

Patients born with neural tube defect need an interdisciplinary team of specialists to oversee their developmental progress. The team includes neurosurgeons, neurologists, pediatricians, urologists and orthopedic surgeons (6, 7, 8, 9, 10). This team surveys child's development to make sure that there is no functional deterioration and that later problems, including hydrocephalus, tethered spinal cord and difficulty with bladder function, are avoided.

Once born, a child with NTD must be operated in order to avoid fluid infection (11). Occasionally, hydrocephalus associated with neural tube defect arrests spontaneously. However, in most cases, shunt surgery is required (12, 13, 14). Shunt techniques include ventriculoperitoneal, ventriculoatrial, and ventriculopleural procedures. Ventriculoperitoneal shunting is preferred modality.

The lesions of spina bifida are divided into three categories: spina bifida occulta, occult spinal dysraphic syndromes including tethered spinal cord syndrome and myelomeningocele or spina bifida aperta. Some forms of spinal dysraphism can cause progressive neurologic deterioration (15, 16, 17, 18). The incidence of clinically overt hydrocephalus at birth in patients with spinal NTD is reported to be 5-10%. In our series, 4 out of 13 (30%) patients had hydrocephalus requiring permanent cerebrospinal fluid diversion and all of these patients presented with myelomeningocele.

Encephalocele is a relatively uncommon neurosurgical entity most commonly seen in the pediatric population. An eight-year

old child in this study had a continuing, inconspicuous, unrecognized CSF leakage for frontoethmoidal encephalocele that was finally repaired at our department. Cranial vault encephaloceles may be approached without opening the cranium, while sincipital and basal encephaloceles usually require craniotomy (19, 20, 21, 22, 23, 24). The incidence of hydrocephalus in patients with encephaloceles is reported to be about 50%. In our series, two out of five (40%) patients had overt hydrocephalus requiring permanent cerebrospinal fluid diversion.

In this study we present our experience in the operative management of neural tube defects with good outcome and also share our recommendations for technical considerations in surgical approaches.

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## Sažetak

### TRETMAN DEFEKATA NEURALNE CIJEVI

*Mirsad HODŽIĆ, Hasan ALTUMBABIĆ,*
*Mirza MORANJKIĆ, Zlatko ERCEGOVIĆ, Selma JAKUPOVIĆ*

Odjeljenje neurohirurgije Univerzitetskog kliničkog centra Tuzla,
Tuzla, Bosna i Hercegovina

**Cilj** Istraživanje je obavljeno s ciljem da predstavimo naše iskustvo u tretmanu defekta neuralne tube u konsekutivnoj seriji od 18 pacijenata.
**Materijal i metode** U periodu između oktobra 2003. i oktobra 2007. 18 pacijenata je operirano na Odjeljenju neurohirurgije Univerzitetskog kliničkog centra Tuzla. Svaki pacijent sa sumnjom na defekt neuralne tube evaluiran je od strane pedijatra, radiologa i neurohirurga kliničkim i ultrazvučnim pregledom, kompjuteriziranom tomografijom i/ili magnetnom rezonancom kičme i glave.
**Rezultati** Podaci od 18 pacijenata sa defektom neuralne tube koji su primljeni i liječeni na Odjeljenju neurohirurgije Tuzla u periodu od četiri godine analizirani su retrospektivno. Prosječna dob pacijenata u vrijeme prijema bila je 15.7 mjeseci, krećući se od 1 dan do 8 godina. Spinalni defekt neuralne tube registriran je u 13 (72.2%) pacijenata, a kranijalni u 5 (27.8%) pacijenata. Hitna operacija izvedena je neposredno nakon rođenja zbog isticanja cerebrospinalne tečnosti u 5 (38.5%) pacijenata sa spinalnim disrafizmom. Nije registrirano isticanje likvora niti je bilo mortaliteta.
**Zaključak** Pacijenti rođeni sa defektom neuralne tube trebaju interdisciplinarni tim specijalista koji će pratiti njihov razvoj. Dijete rođeno sa defektom neuralne tube mora biti operirano da bi se izbjegla likvorska infekcija.

**Ključne riječi:** Defekt neuralne tube ▪ Tretman ▪ Ishod

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