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RESULTS OF SURGICAL TREATMENT OF MYELOMENINGOCELES IN A SMALL NEUROSURGICAL UNIT IN THE ZENICA CANTONAL HOSPITAL

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Objective – To analyse the results of the surgical treatment of myelomeningocele in the Zenica Cantonal Hospital in a five-year period. **Patients and methods** – This retrospective study included 10 patients with myelomeningocele, surgically treated at the Department of Neurosurgery, Zenica Cantonal Hospital, in the 2011-2016 period. Patients were assessed based on their history, symptoms and neurological state. In all cases we performed a detailed neurological examination and craniospinal Magnetic Resonance Imaging. In most cases we performed surgery within the first 72 hours after birth. In three patients we did not perform early surgery because the parents did not give consent for the operation in this period. All patients were hospitalized at the Department of Paediatrics, Zenica Cantonal Hospital. The patients were followed up by a neurosurgeon, a paediatric surgeon and a paediatrician. **Results** – In our research, the incidence of myelomeningoceles in females and males was equal. In most cases the surgery is the most devastating congenital malformation compatible with survival. New-borns with late antenatal diagnosis must undergo surgery as soon as possible, preferably within 24-72 hours. Despite limited resources and conditions we documented a significant recovery in most patients. **Conclusion** – Myelomeningocele is the most devastating congenital malformation compatible with survival. New-borns with late antenatal diagnosis must undergo surgery as soon as possible, preferably within 24-72 hours. Despite limited resources and conditions we documented significant recovery in most patients.

Introduction

Spinal dysraphism may beopen or closed, depending on whether the neural structures and spinal meninges are covered with intact skin, or not (1). In patients with open spinal dysraphism, the meninges (meningocele) or spinal structures and nerves (myelomeningocele) are exposed to the external environment. Myelomeningocele (MMC) is the most severe birth defect compatible with survival (2, 3). Without surgical intervention, patients with open spinal dysraphism die from meningitis and/or internal hydrocephalus (2). The reported incidence of these congenital malformations ranges from 0.41 to 1.43 per 1000 live births (4). In the majority of cases, MMC is associated with hydrocephalus, Chiari I or II malformation, and other congenital malformations (5).

In this paper, we analysed the incidence of MMC in the Zenica – Doboj Canton, and also documented our single institution's experience in the treatment of these malformations.

Patients and methods

Patients characteristics

This retrospective study included 10 patients with MMC surgically treated at the Department of Neurosurgery of Zenica Cantonal Hospital. The study was conducted in the period from January 2011 to December 2016. All patients were referred from the Department of Paediatrics and Neonatology of Zenica Cantonal Hospital. The age of the patients was from 1 day to 55 days. To analyse the incidence of MMC in the Zenica – Doboj Canton, we used data from the Department of Gynaecology about the number of births in our institution in the same period.

Diagnostics

Patients were assessed on the basis of their history, symptoms and neurological status. In all cases, we performed a detailed neurological examination and cranio-spinal Magnetic Resonance Imaging (MRI) to evaluate the associated anomalies. These anomalies are very important for planning the surgical treatment.

Surgery

In most cases we performed surgery within the initial 72 hours. In three patients we did not perform early surgery because the parents gave no consent for the operation in that period. In all cases we performed meticulous MMC repair. We excised the dural sac and neural placode with neural tissue folded into the tube. The pia mater – arachnoid junction and the dura were closed in a watertight manner. In all cases, we covered the bone defect with fascia. In some patients the skin was mobilized by a blunt dissection and closed, but in cases with large or giant MMCs we used skin flaps or grafts. In most cases we diagnosed hydrocephalus and performed ventriculo-peritoneal shunting. In some cases we did not perform early surgery because the parents were not initially motivated for surgery. In these cases, the surgery was performed after a few days, but patients had meningitis that was treated with antibiotics and external ventricular drainage (EVD). After the patients' recovery, we performed ventriculo-peritoneal shunting. The results are presented in tables and figures.

Follow up

All patients were hospitalized at the Department of Paediatrics of Zenica Cantonal Hospital. The patients were followed up by a neurosurgeon, a paediatric surgeon and a paediatrician.

Results

This retrospective research included 10 newborn patients (5 female and 5 male patients). This research showed a low incidence of MMC in the Zenica – Doboj Canton. We used these data to calculate the cumulative incidence of MMC (Table 1).

In most cases the surgery was performed within 72 hours after birth. In three cases we performed delayed surgery (Fig. 1).

In these cases, after MMC reparation, purulent meningitis was diagnosed. In these patients we placed an EVD and administered antibiotics according the antibiogram. After the patients had recovered we performed ventriculoperitoneal shunting. In all cases some neurological dysfunction was detected. In 60% of new borns leg movements were present, but were weakened. In 40% patients we

Table 1 Cumulative incidence of myelomeningocele in the Zenica – Doboj Canton

Year	MMC (n)	Live births (n)	Cumulative incidence
2011	2	3157	0.633
2012	2	3230	0.619
2013	2	3034	0.659
2014	1	2939	0.34
2015	0	2899	-
2016	4	2622	1.527

MMC=Myelomeningocele.

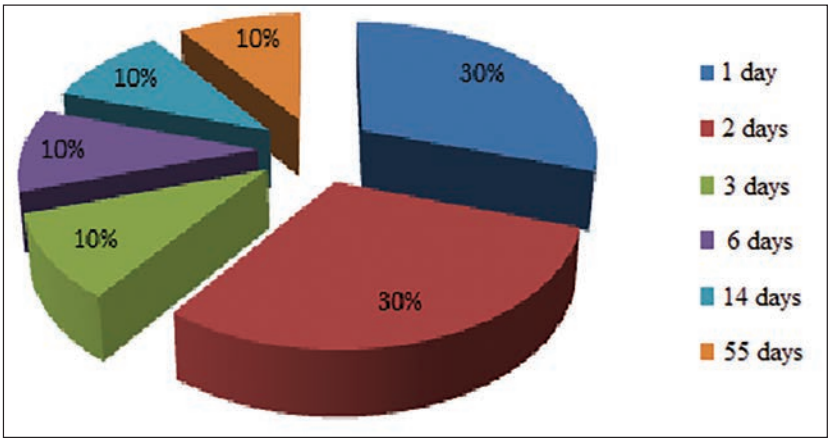

Fig. 1 *Timing of surgery.*

verified paraplegia. In all cases we noted urinary retention (extreme bladder dilatation).

All patients had postoperative follow up. In 3 cases we verified complete motor recovery. Partial motor recovery was noted in 2 patients. We verified paraplegia in 2 patients and paraparesis in 3 patients. In 5 patients we found significant neurological recovery. In all patients we found complete urinary retention before operation. In the postoperative period, 5 patients had bladder function recovery and complete bladder emptying. Hydrocephalus was diagnosed in 8 patients, who were treated with a ventriculo-peritoneal shunt. One patient showed a postoperative wound infection that was treated with antiseptics and antibiotics. After therapy, the wound healed properly. None of the patients died in the postoperative period or during the follow-up period.

Discussion

Open spina bifida or MMC is a devastating congenital defect of the central nervous system (6). This is the most common congenital neurologic malformation compatible with life. The incidence of MMC in the USA is 0.43 and 1.43 per 1000 live births (4). Worldwide, about 5 of 10,000 infants are born with spina bifida (7). The incidence of MMC in China is about 10 per 1000 pregnancies (8). The formation of MMC results from an abnormality in the fusion of the embryologic neural tube during the first month of gestation (6). Failure of neural tube closure results in a sac-like herniation of the meninges (meningocele) or a herniation of neural elements (myelomeningocele) (6, 9). There are many risk factors for neural tube defects, such as: folic acid deficiency, advanced ma-

ternal age, socioeconomic status, antiepileptic drug intake, etc. (10). Adequate folic acid administration during the periconcepcional period and early pregnancy reduces the risk of MMC (6, 11).

In our study, we found a low incidence of MMC, which is in agreement with the incidence reported in developing countries. It should be noted that our citizens live in worse socio-economic conditions than those in the developed countries. The results of our research indicate a good prevention programme in our country. MMC is associated with a skin defect, with the subsequent risk of cerebrospinal fluid (CSF) leak that increases the risk for meningitis (12). The timing of surgery mainly depends on the clinical condition of the patients. Early surgery within 24 to 72 hours is indicated in most cases (13). Delayed surgery increases the risk of meningitis and the progression of hydrocephalus. In most cases we performed early surgery within the first 72 hours. In some cases we performed delayed surgery because the parents were not initially motivated for surgery. In these cases we diagnosed purulent meningitis that was treated with antibiotics and EVD placement. After the patients recovered, we performed ventriculo-peritoneal shunt insertion. After that, we did not find any complications in these patients. In a statistically significant number of patients we found complete or incomplete motor recovery. Our result corresponds with those in the literature (4, 14, 15).

Sphincterial dysfunction was present in all patients included in this research. In a significant number of patients we registered complete bladder emptying in the postoperative period. These results correspond with those reported in the literature (2, 13, 16). A randomized trial of prenatal versus postnatal repair showed that prenatal foetal surgery for MMC reduced the need for ventriculoperitoneal shunt procedures and significantly reduced the incidence of Chiari malformations. This trial showed improved motor function of the lower limbs at 30 months of age (17, 18).

In our research, 8 patients had internal hydrocephalus, which was treated with a shunt procedure. This is also in agreement with reports in the literature (19). The most common reported postoperative complication is CSF leak, present in 24.5% patients (20). In our research, we found no cases of CSF leak. The incidence of postoperative infection is 12.2% with meningitis (15, 20). In our study we registered postoperative wound infection in only 1 case (10%). The reported mortality rate for untreated infants born with MMC ranges from 90% to 100% (21). Early mortality has been reported to be 2%, but this mortality rate increases with the patient's age, being 24% in childhood (13). No mortality occurred in our series.

Limitations of study

This is a retrospective study. Some data about preoperative clinically and neurological status were insufficient. There is a possibility that some babies were born in other hospitals and that they underwent in those institutions.

Conclusion

MMC is the most devastating congenital malformation compatible with survival. Because of its high mortality and morbidity, MMC deserves special antenatal and postnatal attention. Adequate preconceptual folic acid intake could decrease the incidence of MMC. New-borns with late antenatal diagnosis of MMC must undergo surgery as soon as possible, preferably within the first 24-72 hours. In most cases we performed the surgery in this period. In three cases we performed late surgery, because the parents were not initially motivated for surgery. Despite

our limited resources we obtained significant recovery in most patients.

Authors' contributions: Conception and design: HB, RS and AJ; Acquisition, analysis and interpretation of data: HB and MI; Drafting the article HB, AMA and FA; Revising it critically for important intellectual content: HB, AM, EB, HŠJ.

Conflict of interest: The authors declare that they have no conflict of interest.

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