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ORIGINAL ARTICLE

Clinical outcomes of myelomeningocele infants without antenatal surgery: Mortality and morbidity in a tertiary ICU setting

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Abstract

Objective: To investigate the early clinical outcomes of infants with myelomeningocele

Material and methods: We included infants with myelomeningocele who had undergone treatment in our tertiary neonatal intensive care unit. We included all patients with myelomeningocele between 2016-2023. We retrospectively recorded data and assessed perinatal history, morbidity, and mortality status. A p-value of <0.05 was set for significance.

Results: Our study included 90 patients, with an equal distribution of males and females. The majority (87.4%) were delivered by cesarean section, while 12.6% were delivered vaginally. Chiari malformation was present in 7.8% of patients. The average gestational age was 38 weeks, birth weight was 3070 ± 520 grams, birth length was 48 cm, and head circumference was 35 cm. Clubfoot was observed in 31.5% of patients, and scoliosis/kyphoscoliosis in 13.3%. The average sac width was 7 cm and sac length was 6 cm. Normal foot movements were noted in 11.1% of patients. The average surgery time was 49 hours postnatally. Meningitis/CSF infection developed in 28.9% of patients, and convulsions occurred in 23.3%. Hydrocephalus was detected in 72.2% of patients. During follow-up, 59% required a VP shunt. The mortality rate was 18.9%, with an average hospital stay of 41 days.

Conclusion: The poor prognostic factors were lower gestational age, longer sac length and width for lower extremity tonus, scoliosis for mortality, and head circumference, sac length, and sac width for shunt needs.

Keywords: Myelomeningocele; hydrocephalus; ventriculoperitoneal shunt; neural tube defects

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Introduction

Spina bifida is a type of congenital spinal dysraphism, which can be simplified as an unclosed spine with two parts. The main pathology is a neural tube defect, which may contain neural tissue and meninges [1]. Myelomeningocele, a subgroup of congenital spinal dysraphism, involves the herniation of neural tissue through a congenital spinal defect [2]. Open myelomeningocele defects cause morbidity and pose a serious mortality risk. It is the most common type of spinal dysraphism, accounting for 80% of congenital neural tube defects [3]. Myelomeningocele occurs due to impaired cell migration of the neural tube, resulting in defective neural tube differentiation on the 27th day of embryogenesis, which is when the last caudal part closes during primary neurulation. Mesodermal and ectodermal cells form the posterior vertebra and skin [3]. The rate of spinal dysraphism ranges between 0.5-8 per 1,000 live births, with higher rates in developing countries [4]. The incidence has decreased with folic acid supplementation in the preconception period [1].

The treatment of infants with an open myelomeningocele defect requires a multidisciplinary approach, starting with intravenous antibiotic therapy, although some patients may still encounter life-threatening problems. Surgical closure should be performed within two days to prevent infections due to exposed neural tissue [5]. Infants with myelomeningocele may have concomitant hydrocephalus [3]. Some patients who develop hydrocephalus may need a ventriculoperitoneal shunt insertion (5), and some may even require shunt revisions [6]. Myelomeningocele can also be associated with Chiari type II malformation [3], and these infants may need suboccipital decompression [6].

The complications of a myelomeningocele defect can have devastating effects on multiple systems, such as the urinary and neurocognitive systems, as well as the extremities [3]. In-utero surgery is a secondary option for families that refuse pregnancy termination. Families must be informed about the advantages of in-utero surgery, such as the potential decrease in shunt requirements for infants who undergo the procedure [7]. However, in-utero surgery may not prevent the need for untethering operations [8], and the risk of severe complications during the surgery cannot be disregarded [9]. Consequently, families are often confused about in-utero surgery, and some may change their minds about proceeding with the surgery after being informed of the potential complications and surgical process [10].

In our homeland, in-utero surgery has recently become available for families who refuse pregnancy termination. In this study, we aimed to present the mortality and morbidity outcomes of infants who did not undergo in-utero surgery and received treatment in our tertiary intensive care unit. We believe the results will provide families and clinicians with valuable insights into the early rates and potential postnatal complications.

Materials and Methods

The study included all infants treated for an open myelomeningocele defect in a tertiary intensive care unit between 2016 and 2023. It was conducted retrospectively. We documented perinatal history, anthropometric measurements, birth week, delivery type, defect dimensions, operation timing, Chiari type II

Table 1. Desciptive status and clinical outcomes of the patients

Delivery	Vaginal delivery: 12.6% (n=11) Cesarean section: 87.4% (n=76)
Gestational age at birth	38 weeks (38-39)
Birth weight	3070 ± 520 grams
Birth length	48 cm (46-50)
Head circumference	35 cm (34-37)
Width of the sac	7 cm (6-9)
Length of the sac	6 cm (5-8)
Surgery time	49 hours (40-72)
Chiari malformation	7.8% (n=7)
Clubfoot	31.5% (n=28)
Scoliosis/Kyphoscoliosis,	13.3% (n=12)
Impaired/No foot movements	88.9% (n=80)
Meningitis/CSF infection	28.9% (n=26)
Convulsions	23.3% (n=21)
Hydrocephalus	72.2% (n=65)
VP shunt requirement	59% (n=49)
Mortality	18.9% (n=17)
Length of hospital stay	41 days (25-80)

Data is presented as %(n), mean±sd, and median (interquartile range)

defect status, congenital scoliosis and pes equinovarus deformities, hydrocephalus, meningitis, seizure status, and mortality. All patients who underwent treatment for congenital myelomeningocele were included.

Study permission was obtained from the Marmara University Ethical Committee, file number 09.2023.213.

Statistical analysis

We evaluated the normality of continuous data with the Shapiro-Wilk test, kurtosis, skewness, and Q-Q plots. We used the Mann-Whitney test to compare two groups with non-normal distributed continuous data and the chi-square test for categorical data. We presented the data as mean+/-standard deviation, median (interquartile range), and % (n). Additionally, we assessed the correlations using Kendall's tau test. We set the alpha error rate significance at $p < 0.05$. We used the Jamovi 2.3.18 software package.

Results

Our study included 90 patients. Of these patients, 50% were female and 50% were male. The descriptive features and clinical outcomes are described in table 1.

We assessed the correlations between lower extremity tonus, survival status, and shunt need. Lower extremity tone showed a positive correlation with gestational age (r=0.225, p=0.019) and a negative correlation with sac length (r=-0.213, p=0.022), sac width (r=-0.210, p=0.023), and pes equinovarus deformity (r=-0.241, p=0.024). Survival status was negatively correlated with scoliosis (r=-0.228, p=0.031). The need for shunting was associated with head circumference (r=0.225, p=0.024), sac length (r=0.244, p=0.012) and sac width (r=0.205, p=0.035) (Table 2).

The gestational age at birth was 1 week higher (39 weeks versus 38 weeks), and the sac length and sac width were each 2 cm smaller (5 cm versus 7 cm) in the normal lower extremity tonus group compared with the impaired/no tonus group (p = 0.019, p = 0.022, and p = 0.024)

Infants with scoliosis had a 58.3% survival rate, whereas those without scoliosis had an 84.6% survival rate (p = 0.03). Head circumference was 1 cm larger (36 cm versus 35 cm), sac length was 2 cm longer (7 cm versus 5 cm), and sac width was 1 cm larger (8 cm versus 7 cm) in infants who required a ventriculoperitoneal shunt compared to those who did not need a shunt (p = 0.025, p = 0.012, p = 0.035).

Table 2: Correlation results for lower extremity tonus, survival rates, and shunt need

	Lower extremity tonus		**Survival status**		**Shunt need**	
	Kendall's tau	*p*	Kendall's tau	*p*	Kendall's tau	*p*
Gestational week at birth	0.225*	0.019	-0.134	0.161	-0.152	0.125
Birth weight	0.057	0.525	0.01	0.914	-0.013	0.886
Birth length	0.175	0.069	0.097	0.315	-0.169	0.087
Head circumference	-0.017	0.863	0.005	0.957	0.225*	0.024
Sac length	-0.213*	0.022	-0.049	0.186	0.244*	0.012
Sac width	-0.210*	0.023	-0.051	0.585	0.205*	0.035
Gender (Females)	-0.141	0.182	-0.028	0.789	-0.137	0.215
Pes equinovarus deformity	-0.241*	0.024	-0.163	0.126	0.148	0.183
Scoliosis	-0.139	0.191	-0.228*	0.031	0.083	0.455

Kendall's tau test. * $p < .05$

Discussion

In this study, we aimed to present the mortality and morbidity of infants with spinal dysraphism who did not undergo in utero surgery in our tertiary intensive care unit. Our findings provide crucial insights into the early rates and possibilities of postnatal complications, aiding both families and clinicians in managing expectations and planning for postnatal care.

The rate of infant mortality linked to spina bifida has evolved over the years due to several factors, including the implementation of folic acid supplementation and food fortification programs, advancements in prenatal screening and treatments, as well as decisions regarding the termination of pregnancies. Our study found an 18.9% mortality rate, which is consistent with the higher end of the spectrum reported in previous studies on infants with spinal dysraphism [11]. The majority of our patients were delivered via cesarean section (87.4%), which is consistent with the current practice of opting for cesarean delivery in cases of severe congenital anomalies due to the reduced risk of birth trauma [12,13]. The average gestational age of 38 weeks and the average birth weight of 3070 ± 520 grams are comparable to those reported in similar studies, such as those described by Ilhan et al. (2017), who noted comparable gestational ages and birth weights in their study of infants with spinal dysraphism [14].

Our findings revealed that 31.5% of the infants had clubfoot, and 13.3% had scoliosis/kyphoscoliosis. These rates are in line with existing literature, which suggests a prevalence of orthopedic complications ranging from 30-50% for clubfoot spinal dysraphism cases [15,16]. However, contrary to the literature, our study found a lower rate of scoliosis [17].

We found that lower extremity tonus positively correlated with gestational age. This suggests that infants with more advanced gestational age might exhibit better lower extremity function. Conversely, lower extremity tonus negatively correlated with sac length, sac width, and the presence of pes equinovarus deformity. These findings indicate that more severe sac involvement and associated deformities may be linked to poorer lower extremity tonus.

Based on our findings, the association of shunt need with larger head circumference, longer sac length, and wider sac width suggests that more severe hydrocephalus and larger defects are linked to a higher likelihood of requiring a ventriculoperitoneal shunt. This emphasizes the role of these anatomical measurements in predicting the need for shunt placement and managing hydrocephalus in affected infants. Hydrocephalus was detected in 72.2% of our patients, which aligns with the high prevalence reported in the literature, where hydrocephalus affects approximately 60-80% of infants with spinal dysraphism [7]. The incidence of meningitis/CSF infection (28.9%) and convulsions (23.3%) is consistent with the rates reported in other studies, indicating significant postnatal risks [18].

The average surgery time of 49 hours postnatal is similar to that reported in other studies, suggesting that early surgical intervention remains crucial in managing spinal dysraphism [3,9]. The fact that 59% of our patients required a ventriculoperitoneal shunt further emphasizes the need for close monitoring and timely intervention for hydrocephalus, which aligns with findings from previous research indicating a high shunt requirement rate [7,19].

Conclusion

In conclusion, our study's findings contribute valuable data on the outcomes of infants with spinal dysraphism who did not receive in utero surgery. The mortality rate, prevalence of orthopedic and neurological complications, and postnatal treatment requirements are consistent with existing literature, reinforcing the need for ongoing research and improved management strategies to address the challenges faced by this vulnerable population.

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Authors' contribution: Medical practices: A.B, Concept: B.C.A, Design: B.C.A, A.Y, Data Collection or Processing: I.K, S.C.K. Analysis or Interpretation: I.K, Literature Search: B.C.A, A.Y, S.C.K, A.B, Writing: B.C.A, A.Y., I.K.

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