



Original Article

The Prevalence of Chiari II Malformation in Neonates with Myelomeningocele at Ayub Teaching Hospital, KPK

Ehtisham Ahmed Khan Afridi¹, Muhammad Waleed Khan¹, Shabana Naz², Saadia Maqbool¹
Aqsa Shehzadi¹, Nusrat Shaheen¹

Departments of ¹Neurosurgery and ²Pathology, Ayub Teaching Hospital, Abbottabad

ABSTRACT

Objective: Neural tube defects (NTDs) are common in northern areas of Khyber Pakhtunkhwa (KPK) and need a lot of community education for the parents regarding this disease, which impaired the patients for their whole life. The study aimed to assess the contribution of a family history of myelomeningocele and the resulting incidence of Chiari II malformation.

Materials and Methods: A total of 131 patients were observed to determine the frequency of the Chiari II malformation in patients with myelomeningocele who presented in Ayub Teaching Hospital, Abbottabad. All neonates were sent to the radiology department for MRI. A repair procedure for meningocele was done.

Results: The mean age was 16.56 days. In 53.4% of neonates, there was a familial history of spinal dysraphism, while in 46.6% there was no familial history. Chiari II malformation was present in 23.7% of patients who presented with myelomeningocele. A significant difference (p-value < 0.00001) existed between the presence/absence of a family history of myelomeningocele and Chiari II malformation out of the total.

Conclusion: Early surgery, along with a multidisciplinary approach, provides the best opportunity for improved results and survival.

Keywords: Meningomyelocele, Neural Tissue, Maternal Folate Intake, Meningomyelocele (MMC) Repair.

Corresponding Author: Ehtisham Ahmed Khan Afridi
Department of Neurosurgery ATH
Email: ehtisham81@gmail.com

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INTRODUCTION

Myelomeningocele is the most common open neural tube abnormality. It is characterized as the failure of the lumbosacral neural tube to close during embryonic development, resulting in meningeal and spinal cord herniation through a vertebral defect.¹ Neural tube fusion begins in the hindbrain (medulla and pons) and proceeds rostrally and caudally. Incomplete caudal fusion results in the establishment of a meningocele on day 26 of pregnancy.² The

majority of open neural tube anomalies are uncommon. However, various risk factors have been associated with the development of neural tube defects.³

Females are at a 3 – 7 times greater risk than males. There are significant racial disparities; for example, the prevalence is many times greater in various sections of China, Africa, the Middle East, Thailand, and India. Folate supplementation standards in these nations might contribute to these racial disparities.⁴ The frequency of neural tube defects rises with poorer socioeconomic level and mother age. Recurrence rates in future pregnancies are around 2 – 3%.⁵ Chiari II malformation (CM-II), also known as Arnold-Chiari formation, is a very common congenital deformity characterized by beaked mi rain, tonsil downward displacement, cerebellar vermis, and spinal myelomeningocele.⁶ The clinical manifestation of CM-II varies greatly.

Defects in the neural tube After heart abnormalities, neural tube defects (NTDs) are the second most frequent congenital malformations, and folic acid deficiency is the most generally recognized risk factor, which is more prevalent in the poor socioeconomic category.⁸ Similarly, investigations in the developing world have discovered NTDs among poor socioeconomic groups.¹⁰ Preconceptional folic acid supplementation results from a Chinese supplementation program indicate that folic acid treatments can reduce NTD.¹¹ Prenatal maternal folate consumption was positive in 103 (68.7%) instances, which is lower than in Ethiopian research where only 14.4% of patients received preconception folic acid or multivitamin treatment.¹²⁻¹³

This is primarily owing to the malformation's wide range of severity and the multiple accompanying abnormalities that are not often found. However, because myelomeningocele is common, CM-II is frequently detectable at birth.⁷ The Chiari II malformation occurs in 21% of individuals with myelomeningocele.⁸ The

discovery of these anomalies, which frequently outline the basic symptoms of Chiari II malformation, raises the question of whether they are part of the illness spectrum or additional independent features with their prognosis, perhaps posing a contraindication to fetal surgery. Through the current study, we wanted to see how a family history of myelomeningocele affected the incidence of Chiari II malformation. The incidence of the Chiari II malformation in individuals with myelomeningocele who presented to Ayub Teaching Hospital in Abbottabad was determined in 131 cases.

MATERIAL AND METHODS

Study Design & Setting

From November 8, 2021, to May 8, 2022, cross-sectional research was undertaken in the Department of Neurosurgery at The Ayub Teaching Hospital in Abbottabad. A total of 131 patients were enrolled in the study.

Inclusion Criteria

The neonates of both genders were included with age ranges from 1 – 28 days.

Exclusion Criteria

The pre-term babies (<36 weeks at birth) were excluded.

Surgical Management- Repair of Meningomyelocele

Following the induction of general anesthesia, the patient was on the operating table, with the head somewhat lower than the back. The surgical site was thoroughly cleansed. Warm sterile saline was irrigated into the myelomeningocele, and the surrounding skin was washed with Betadine Draping effectively isolating the anus from the operating field. The incision is drawn on the

baby's back. Vertical extensions are aimed to undercut the skin and, if feasible, create a midline closure. The skin was incised directly over the exposed meninges, and excess skin was removed. Beginning in a non-neural area, the incision was extended down and often into the meningeal sac. The skin borders are retracted laterally. The sac's wall mobilizes nerve roots that go back into the spinal canal. Some neural components are atretic and may be sacrificed in the sac itself. The neural placode borders were folded and sutured using interrupted 6 – 0 monofilament nylon to reanimate (repair) the spinal cord structure. The filum terminale is frequently found within the spinal canal. It should be severed to remove the related spinal cord tethering. After that, the dura was separated from the subcutaneous tissue and lumbosacral fascia. The paravertebral muscles and fascia were engaged to close in the midline, if feasible, and reestablish their appropriate dorsal position relative to the vertebral components to reinforce the dural suture line. Alternatively, semilunar lumbosacral fascia flaps can be swung across the midline and sutured to the opposite side's base.⁹

Data Collection & Analysis

After receiving ethics committee approval, patients meeting the inclusion criteria from the Department of Neurosurgery/Pediatrics at Ayub Teaching Hospital in Abbottabad were included. Patients' baseline demographic information (age, gender, family history of myelomeningocele) was collected. The informed agreement was obtained from parents, safeguarding the confidentiality and the patient's safety while participating in this study. The radiology department received all newborns for MRI.

Data were analyzed with a statistical analysis program (SPSS-IBM version 25). For qualitative factors such as gender, family history of myelomeningocele, and Chiari II malformation, frequencies and percentages were calculated. The

p values were derived from Z-scores to determine if there were significant/insignificant differences in the presence/absence of a family history of myelomeningocele as well as Chiari II abnormalities.

RESULTS

Age Distribution

This research comprised 131 patients with ages ranging from 1 to 28 days. The patients in the age range 1 – 8 days were 28.2%, in 9 – 16 days they were 24.4%, in 17 – 22 days they were 29%, and in 22 – 28 days they were 18.3%.

Table 1: Information on Age Groups (n = 131)

Age of Patients	Frequency	Percentage %
1 – 8 Days	37	28.2
9 – 16 Days	32	24.4
17 – 22 Days	38	29.0
22 – 28 Days	24	18.3
Total	131	100.0

Gender Distribution

84 (64.1%) patients were male and 47(35.9%) were female.

History of Myelomeningocele and Prevalence of Chiari II Malformation

In 53.4% of neonates, there was a familial history of spinal dysraphism, while in 46.6% there was no familial history (Table 2). A significant difference

Table 2: Family History of Myelomeningocele (n = 131).

Family History of Myelomeningocele	Frequency	Percentage	P value
Yes	70	53.4	< 0.00001 (significant result)
No	61	46.6	
Total	131	100.0	

(p -value < 0.00001) existed between the presence/absence of a family history of myelomeningocele out of total. Chiari II malformation was present in 23.7% of patients who presented with myelomeningocele (Table 3). A significant difference (p -value < 0.00001) existed between the presence/absence of Chiari II malformation out of the total.

Table 3: Distribution of Chiari II Malformation (n = 131).

Chiari II Malformation	Frequency	Percentage	P value
Yes	31	23.7	< 0.00001
No	100	76.3	(significant result)
Total	131	100.0	

DISCUSSION

The average age of our patients was 16.56 days. There were 64.1% of male patients and 35.9% of female patients. There was a familial history of spinal dysraphism in 53.4% of the neonates, whereas there was no familial history in 46.6%. Chiari II malformation was seen in 23.7% of individuals with myelomeningocele. There was a significant difference between having a family history of myelomeningocele and having Chiari II malformation. Khan et al.¹⁴ reported that MMC is most commonly found in the lumbar region, with a reported frequency of 60 – 70%.¹⁴ Oncel et al,¹⁵ found that 46.6% of the lesions were lumbosacral and 40% were lumbar. Rehman et al,¹⁶ reported that 65.3% of the newborns had hydrocephalus, 9% had club foot, 2.7% had diastematomyelia, and 2% had a tethered cord. Kshetry et al,¹⁷ showed a comparable rate of hydrocephalus needing CSF diversion during the same hospital stay, with 56.6% of patients requiring shunt insertion. As a result, in addition to a thorough physical examination, all patients with MMC should be checked for the existence of additional abnormalities such as cranial and cardiac imaging examinations, as well as urinary system

ultrasonography. Early surgery is related to decreased morbidity and mortality, and one research found that surgery shortly after delivery has fewer risks and better results than fetoscopic surgery, which has major hazards for both the mother and the child.¹⁸⁻²² Kshetry et al,¹⁷ reported an in-hospital mortality rate of 1.4%. There had been no further difficulties until the previous check-up. However, spinal cord re-tethering can occur after the main surgical repair of MMC, and the injection of skin components during the initial surgery might result in the formation of intraspinal epidermoid or dermoid cysts.²¹⁻²⁴ As a result, in each situation, a close and persistent follow-up is required. A better prognosis is also influenced by increased public knowledge, prevention, excellent prenatal monitoring, and early surgical intervention.

Fractional anisotropy was substantially greater in Chiari II malformations than in age-matched fetuses with a normal CNS, while the apparent diffusion coefficient (ADC) was not significantly different. There were no variations in diffusion tensor imaging (DTI) measurements between normal controls and fetuses with hydrocephalus or ventriculomegaly. In Chiari II malformations, DTI can identify and quantify parenchymal changes in the embryonic midbrain. As a result, in situations of enlarged fetal ventricles, FA of the fetal midbrain may help distinguish Chiari II malformation from other entities.²⁵ Rapid prototyping is quickly becoming a popular and valuable approach for creating physical models of congenital abnormalities. Manufacturing models are often constructed using scan data from three-dimensional (3D) ultrasonography, computed tomography, and fetal magnetic resonance imaging (MRI).²⁶

Diffusion-weighted imaging can be used to evaluate brain maturation. When the additional assessment of the posterior fossa is necessary in cases of suspected Chiari II malformation, fetal MR imaging is employed. While aberrant white matter organization or early cerebellar

degeneration may have contributed to our findings, the most likely explanation is improper CSF outflow in the posterior fossa, with increased extracellular water perhaps accounting for this phenomenon.²⁷ Chiari II malformation is a congenital anomaly marked by a tiny posterior fossa and hindbrain downward displacement into the foramen magnum. Diffusion-weighted imaging (DWI) can be utilized to describe brain damage and physiological maturation quantitatively. The goal of this study is to assess DWI parameters of the infra- and supratentorial brain in fetuses with Chiari II malformation. In fetuses with Chiari II malformation, abnormal ADC values suggest supratentorial microstructural abnormalities. More research is needed to determine the relevance of diffusion imaging measures in detecting aberrant brain development, parenchymal damage, and the success of fetal surgery.²⁸

Prenatal diagnosis was very common, and if diagnosed before 24 weeks gestation, pregnancy termination was frequently given and accepted. A regional prenatal ultrasound program, mostly centered on primary care but with quick access to a tertiary center, can be extremely beneficial, but access to prenatal diagnostics must be addressed.²⁹ In total, 231 MMC patients were included in the study. Patients were followed for an average of 6.9 years, ranging from 1 to 20 years. Shunt implantation was shown to be most common in individuals with MMC at the highest spinal levels. Shunt failures, which were detected in 47.2% of hydrocephalus, and recurring UTIs, which were diagnosed in 55.8% of MMC, were the leading causes of morbidity and mortality. A newborn head circumference of 38 cm was discovered to be a major risk factor for shunt revision. Furthermore, lower MMC functional levels were related to less revision than higher ones. There was a strong link between recurrent UTI and thoracic functional level. Macrocephaly at birth and greater degrees of the abnormality has a negative influence on the outcome and, as a

result, provide a barrier to pediatric neurosurgical practice.³⁰

Ravindra et al, (2020) did a thorough assessment of subjects relevant to counseling in the context of myelomeningocele and present a novel paradigm for shared decision-making to assist practitioners during counseling. Fetal surgery, on the other hand, is linked with considerable maternal and newborn risks such as uterine wall dehiscence, preterm, and fetal or neonatal mortality. Expectant women must make a prompt decision among numerous viable alternatives, including pregnancy termination, postnatal surgery, or fetal surgery. Maternal health, fetal health, financial resources, social support, risk aversion, availability to care, family planning, and values all play a role in the choice. It is often a challenging decision that benefits from the advice of a pediatric neurosurgeon.³¹ Karakas et al. (2022) studied data from individuals with myelomeningocele who were followed longitudinally at a single site. Hydrocephalus occurred in 88.5% of the patients, with 90.7% necessitating a ventriculoperitoneal shunt procedure. 19.7% of patients had seizures, with 23 having hydrocephalus. The beginning of epileptic seizures has been associated with falx dysgenesis, lumbar myelomeningocele, and cortical atrophy. They concluded that individuals with myelomeningocele who suffer seizures had an overall favorable prognosis with significant long-term seizure freedom.³²

CONCLUSION

Early surgery, along with a multidisciplinary approach, provides the best opportunity for improved results and survival. It is suggested that we should explore implementing national preventative efforts to lower the prevalence of neural tube defects in our country, as well as creating a national database to set recommendations and timely referrals for better results.

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Additional Information

Disclosures: Authors report no conflict of interest.

Ethical Review Board Approval: The study was conformed to the ethical review board requirements.

Human Subjects: Consent was obtained by all patients/participants in this study.

Conflicts of Interest:

In compliance with the ICMJE uniform disclosure form, all authors declare the following:

Financial Relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.

Other Relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

AUTHOR CONTRIBUTIONS

Sr. No.	Author's Full Name	Intellectual Contribution to Paper in Terms of
1.	Ehtisham Ahmed Khan Afridi	Study Design, Methodology, and Paper Writing.
2.	Mohammad Waleed Khan	Data Calculation and Data Analysis.
3.	Ehtisham Ahmed Khan Afridi	Interpretation of Results.
4.	Shabana Naz	Statistical Analysis.
5.	Aqsa Shahzadi	Literature Review.
6.	Saadia Maqbool/ Nusrat Shaheen	Literature Review and Quality Insurer.