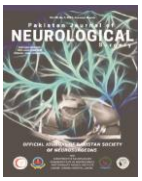

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Original Article

Incidence of Development of Hydrocephalus after Excision and Repair of Spina Bifida Aperta in Infants

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ABSTRACT

Objective: To find out the incidence of hydrocephalus after excision and repair in infants presenting with Spina Bifida Aperta.

Materials & Methods: This prospective cohort study was conducted at the Pediatric Neurosurgery Department, Children Hospital & The Institute of Child Health, Lahore, Pakistan, from January 2021 to October 2021. A total of 62 infants of both genders presenting with spina bifida Aperta undergoing repair were included. Data of the patients, i.e., name, age, gender, head circumference, location, and width of the defect, accompanying bladder, limb anomalies, radiological, laboratory findings, and diagnosis (meningocele or meningomyelocele) were noted. Patients were followed postoperatively for 1-month, and the incidence of post-surgery hydrocephalus was noted.

Results: Out of 62 children, 36 (58.1%) were male and 24 (41.9%) female. The mean age was noted to be 138.82 days. Most children, 36 (58.1%), were found to have meningocele. The most frequent local meningocele/meningomyelocele was noted to be lumbosacral, 22 (35.5%). Post-surgery hydrocephalus was noted among 11 (17.1%) cases. No significant association of gender, age, head circumference, defect size, the maximum dimension, diagnosis (meningocele or meningomyelocele), or location was noted with post-surgery hydrocephalus among study cases ($p > 0.05$). No mortality was reported.

Conclusion: Meningomyelocele and lumbosacral location of the defect were among the prominent factors affecting the incidence of post-surgery hydrocephalus.

Keywords: Spina Bifida Aperta, Meningiocele, Myelomeningocele, hydrocephalus, lumbosacral.

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INTRODUCTION

Neural tube defects are one of the most prevalent birth disorders. The incidence varies between one in 800 and one in 1000 live births depending on regional conditions, race, sex of the infant, and specific maternal factors.[1] According to studies in Pakistan, the frequency is between 38.6 and 124.1 per 10,000 births. In the first six months of life, the death rate for untreated patients is 65–70%.[2-4] This central nervous system (CNS) congenital defect affects the neural tube at the early stages of neurulation in the third or fourth week of development. Spina Bifida is a disorder caused by a malfunction in the development of an infant's spine. Spina bifida is classified as Occulta or Aperta, which includes meningocele, myelomeningocele, and myeloschisis. The most severe congenital neural tube defect in which neural tissue is exposed to the outside environment is myeloschisis form. When spina bifida affects the lumbosacral region, the clinical presentation resembles a spinal cord injury with neurogenic bowel, bladder, and lower limb paralysis.[5,6] The spinal cord and meninges may protrude through the child's back in myelomeningocele, a kind of spina bifida. In certain situations, the child's back skin covers the spinal cord and meninges. The spinal cord and meninges may also protrude through the skin in some situations. There would be some neurological deficiency beneath the location as well.[7,8] A meningocele is a kind of spina bifida in which the meninges herniate between the vertebrae with complete neurology at birth due to a developmental abnormality. Individuals with meningoceles are unlikely to have long-term health problems because the neurological system is unharmed, though incidences of tethered cords have been observed.[9] Myelomeningocele and meningocele problems range from mild to severe physical and mental difficulties, including trouble paying attention, interpreting language, and reading. The effects are dictated by the size and location of the abnormality, as well as whether it is ruptured or unruptured. The condition affects the spinal cord or nerve roots lying at the level and below to varying degrees. Surgical excision and repair are frequently performed on these patients as a therapeutic approach to improve neurological impairment.[5,6,10] According to Ghani et al, in research conducted at Lady Reading Hospital in Peshawar, 20% of the patients developed hydrocephalus following meningomyelocele excision and repair and V/P shunting.[6] In a study conducted at the Pakistan Institute of Medical Sciences (PIMS) Islamabad, postoperative hydrocephalus was observed in 22.4% of cases following meningomyelocele excision and repair.[10]

There was no relevant literature for meningocele excision and repair, and this type of study has not yet been undertaken in our setting, namely Pediatric Neurosurgery Emergency, Children Hospital & The Institute of Child Health, Lahore. The goal of this study was to determine the incidence of hydrocephalus following excision and repair in neonates with meningocele and meningomyelocele. This study was designed to assist neurosurgeons in making better decisions regarding various problems, particularly hydrocephalus.

MATERIALS & METHODS

Study Design and setting

From January 2021 to October 2021, this prospective cohort study was conducted at Pediatric Neurosurgery Emergency, Children's Hospital & The Institute of Child Health in Lahore, Pakistan. Approval from the "Institutional Ethical Committee" was acquired. Informed and written consent was taken from the parents/guardians of all study participants.

Sampling

The sample size of 62 was calculated, keeping the expected ratio of development of hydrocephalus

after excision and repair of meningomyelocele at 20%,[6] margins of error of 10%, and a confidence interval of 95%. Infants were recruited through non-probability convenience sampling.

Inclusion Criteria

The infants (aged less than one year) of both genders presenting with Spina Bifida Aperta, i.e., meningocele and meningomyelocele undergoing repair, were included. Hydrocephalus was labeled if lateral ventricle size was more than 1 cm on cranial ultrasound at the level of the foramen of Monro.

Exclusion Criteria

Exclusion criteria were patients coming with meningomyelocele-associated hydrocephalus or those cases whose parents/guardians refused to be part of this study.

Data Collection Procedure

After admission, detailed history was taken, and a thorough physical examination of all the subjects was carried out. Data of the patients, i.e., name, age, gender, head circumference, location, and width of the defect, accompanying bladder, limb anomalies, radiological, laboratory findings, diagnosis, and subtypes, i.e., meningocele or meningomyelocele, were noted. A single surgical team did the excision and repair of the defects, and standard operating procedures were followed. Patients were followed postoperatively for the development of hydrocephalous (lateral ventricular size or any new neurological deficit) after 48 hours, 7 days, and 15 days at one month. A Neurosurgeon assessed the patients with a minimum five-year experience for the development of hydrocephalous after cranial ultrasound within one month postoperatively.

Data Analysis

All study data were recorded on a predesigned proforma and analyzed in the SPSS version 26.0. The quantitative variables like age, head circumference, the width of the defect, and pre-op/post-op ventricular size were presented in the form of mean and standard deviation (SD), while the categorical variables, i.e. gender, location of defects, diagnosis, presence or absence of postoperative hydrocephalus and any new neurological deficit were shown as frequency and percentage. The normality of the data was checked using the Shapiro-Wilk test. The Chi-square test was applied for an association between gender, location of defects, diagnosis, and the presence or absence of postoperative hydrocephalus taking a p-value of ≤0.05 as significant.

RESULTS

Age and Gender Distribution

Out of 62 children, 36 (58.1%) were male and 24 (41.9%) females. The mean age was 138.83 days. Most of the children, 36 (58.1%), were found to have meningocele.

Stratification of Study Variables

The most frequent location of meningocele/ meningomyelocele was lumbosacral, 22 (35.5%). Table 1 compares study variables concerning diagnosis as meningocele or meningomyelocele.

Post-surgery hydrocephalous

All patients completed the one-month study follow-up. Post-surgery hydrocephalus was noted among 11 (17.1%) cases. The mean size of the lateral ventricle was 7.0 ± 1.54 mm (ranging from 4 – 9 mm). Ventriculoperitoneal (VP) shunting was done in all eleven cases with post-surgery hydrocephalus. No significant association of gender, age, head circumference, defect size, maximum diameter, diagnosis (meningocele or meningomyelocele), or location was noted with post-surgery hydrocephalus among study cases

Table 1: Stratification of study variables with respect to the subtypes of spina bifida aperta.

Study Variables		Meningocele	Meningomyelocele	Number (%)/ Mean ± SD (Range)
Gender	Male	21 (58.3%0	15 (57.5%)	36 (58.1%)
	Female	15 (41.7%)	11 (42.3%)	26 (41.9%)
Age in days	< 60	12 (33.3%)	7 (26.9%)	138.83 (5 – 336)
	60 – 119	6 (16.7%)	5 (19.2%)	
	120 – 179	7 (19.4%)	3 (11.5%)	
	180 – 240	2 (5.6%)	4 (15.4%)	
	> 240	9 (25.0%)	7 (26.9%)	
Head Circumference (cm)	< 40	20 (55.6%)	18 (69.2%)	39.19 ± 4.25 (31.8 – 46.8)
	≥ 40	16 (44.4%)	8 (30.8%)	
Size of Defect (cm)	1 – 4	18 (50.0%)	1 (3.8%)	6.61±2.53 (2.5 – 10.8)
	5 – 10	15 (41.7%)	19 (73.1%)	
	> 10	3 (8.3%)	6 (23.1%)	
Maximum Diameter (cm)	1 – 4	7 (19.4%)	0	(3.2 – 12.5)
	5 – 10	21 (58.3%)	17 (65.4%)	
	> 10	8 (22.2%)	9 (34.6%)	
Spinal Location of Meningiocele / Meningiomyelocele	Cervical	7 (19.4%)	4 (15.4%0	11 (17.7%)
	Dorsal	4 (11.1%)	5 (19.2%)	9 (14.5%)
	Dorsolumbar	4 (11.1%)	3 (11.5%)	7 (11.3%)
	Lumbar	6 (16.7%)	7 (26.9%)	13 (21.0%)
	Lumbosacral	15 (41.7%)	7 (26.9%)	22 (35.5%)

Table 2: Stratification of study variables with respect to post-surgery hydrocephalus.

Study Variables		Post-Surgery Hydrocephalus Yes	Post-Surgery Hydrocephalus No	P-Value
Gender	Male	7 (63.6%)	29 (56.9%)	0.680
	Female	4 (36.4%)	22 (43.1%)	
Age in days	< 60	4 (36.4%)	15 (29.4%)	0.728
	60 – 119	1 (9.150	10 (19.6%)	
	120 – 179	3 (27.3%)	7 (13.7%)	
	180 – 240	1 (9.1%)	5 (9.8%)	
	> 240	2 (18.2%0	14 (27.5%)	
Head Circumference (cm)	< 40	7 (63.6%)	31 (60.8%)	0.860
	≥ 40	4 (36.4%)	20 (39.2%)	
Size of Defect (cm)	1 – 4	4 (36.4%)	15 (29.4%)	0.786
	5 – 10	5 (45.5%)	29 (56.9%)	
	> 10	2 (18.2%0	7 (13.7%)	
Maximum Diameter (cm)	1 – 4	3 (27.3%)	4 (7.8%)	0.170
	5 – 10	6 (54.5%)	32 (62.7%)	
	> 10	2 (18.2%)	15 (29.4%)	
Subtype	Meningocele	4 (36.4%)	32 (62.7%)	0.108
	Meningomyelocele	7 (63.6%)	19 (37.3%)	0.043
Spinal Location of Meningocele / Meningomyelocele	Cervical	2 (18.2%)	9 (17.6%)	0.348
	Dorsal	1 (9.1%)	8 (15.7%)	0.29
	Dorsolumbar	1 (9.1%)	6 (11.8%)	0.56
	Lumbar	2 (18.2%)	11 (21.6%)	0.77
	Lumbosacral	5 (45.5%)	17 (33.3%)	0.0162

(p > 0.05) as shown in Table 2. No mortality was reported.

DISCUSSION

Management of meningocele or meningomyelocele is complex and requires multidisciplinary treatment. These defects should be treated surgically shortly after birth to prevent further damage to the spinal cord. In this study, we noted that the mean age of the infants was 138.83 (range: 5 – 336) days which is higher than what has been reported by another national study from Islamabad, where the mean age of the cases was recorded to be 58.58 ± 26.01 days.[10] A recent study from Karachi also reported the mean age of the cases undergoing meningomyelocele to be 2 ± 1 month.[5] Comparatively higher mean age recorded in this study points towards delay in seeking medical attention by the parents/ guardians of such patients, which highlights the need for improving awareness and knowledge about these congenital malformations.

We noted that 58.1% of the cases in our study were male. Male predominance in our study among children with meningocele/ meningomyelocele is very similar to what Alamgir et al,[10] and Rehman et al,[5] reported. A recent study from the Netherlands by Spoor et al. reported a nearly similar proportion of males to females (49.5% vs 50.5%) in their study analyzing outcomes of myelomeningocele.[11] Most common spinal location of meningocele/ meningomyelocele was lumbosacral (35.5%) followed by lumbar (21.0%). Recent data from the developed world has reported the lumbar and lumbosacral regions as the most common sites for myelomeningocele.[11] Literature has reported the most common region for myelomeningocele to be lumbar, while its frequency ranges between 60 – 70%.[12]

In the present work, the mean head circumference was found to be 39.19 ± 4.25 cm, while the mean size of the defect was 6.61 ± 2.53 cm. The mean head circumference, according to a national study by Rehman et al. among patients undergoing meningomyelocele repair, was 37.4 cm which is a little less than what we noted.[5] As the mean age among study participants of Rehman et al. was relatively lower than ours, that could be the reason why the mean head circumference of study subjects was also low in their study.[5] A study by Oncel et al. from Turkey reported mean head circumference to be 35.8 ± 3.8 cm which is lesser than what we noted and could be because Oncel et al, had enrolled newborns for meningomyelocele repairs.[13]

In the present study, the incidence of post-surgery hydrocephalus was 17.1%. Khan A et al, from Islamabad, reported the incidence of post-surgery hydrocephalus to be 22.4%.[10] Ghani et al, revealed the development of hydrocephalus after excision and repair of meningomyelocele as 20%.[6] A study from India reported the incidence of post-surgery hydrocephalus after primary repair closure of myelomeningocele to be 7.7%.[14] Another study from India reported the incidence of post-surgery hydrocephalus to be 1.5% which is again relatively low compared to what we reported in the present study.[15] In the present study, the size of the defect and maximum dimension were significantly large among children having meningomyelocele. However, this aspect was not found to have any influence in terms of significant association with the development of post-surgery hydrocephalus.

There were several limitations as well. The results of this study may not be generalizable because it was a single-center study with limited sample size. We could not evaluate socio-economic or maternal risk factors related to the presence of meningocele/meningomyelocele. We could not compare pre-surgery and post-surgery neurological outcomes as we focused on findings of the incidence of post-surgery hydrocephalus in the present study. As we had only gathered short-term follow-up data regarding the incidence of post-surgery hydrocephalus, further studies

should be conducted with long-term follow-ups to record all related complications of the surgery.

CONCLUSION

Post-surgery hydrocephalus was found to occur in 17.1% of patients. Meningomyelocele and lumbosacral location of the defect were among the prominent factors affecting the incidence of post-surgery hydrocephalus.

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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.

AUTHORS CONTRIBUTIONS

Sr.#	Author's Full Name	Intellectual Contribution to Paper in Terms of:
1.	Hassaan Zahid	1. Study design and methodology.
2.	Lubna Ijaz	2. Paper writing.
3.	Amna Malik	3. Data collection and calculations.
4.	Laeequr Rehman	4. Analysis of data and interpretation of results.
5.	Malik M. Nadeem Khan	5. Literature review and referencing.
6.	Farhan Fateh Jang	6. Editing and quality insurer.