



Original Article

A Spectrum of Spina Bifida: A Study of Neurosurgery Department of DHQ Teaching Hospital, Gomal Medical College, DI Khan

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ABSTRACT

Objective: To pin down the spectrum of spina bifida in infants.

Materials & Methods: This prospective cohort study was conducted at the Neurosurgery Department, DHQ teaching hospital, Gomal Medical College, D.I Khan, Pakistan, from July 2021 to July 2022. A total of 100 diagnosed infants of spina bifida of either gender, who were undergoing surgery were included in the study. Demographics like; gender, name, age, cousin marriage, region, type of spina bifida (meningocele or meningocele), associated with hydrocephalus, and width of the defect were noticed. Post-operatively the maximum follow-up was of 1 month for noticing the outcome and complications.

Results: Of a total number of 100 infants, 76 patients were male, while 24 were female. The mean age of the patients was 913.625 days. The majority of the children (n = 59) were having myelomeningocele. The lumbosacral spine was the most common location (n = 88) for myelomeningocele/meningocele. Post-operatively, there was the development of hydrocephalus in 12 patients.

Conclusion: The majority patients of with spina bifida were males. Meningomyelocele and lumbosacral location were the commonest findings. Furthermore, the lumbosacral location of the spina bifida and myelomeningocele were most commonly associated with the development of postoperative hydrocephalus.

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INTRODUCTION

Spina bifida is a congenital disorder that occurs due to defective closure (complete or partial failure) of the neural tube in the first month of gestation. On day 17, the ectoderm overlying the notochord thickens to form a neural plate.^{1,2} During 3rd week of gestation neural plate deepens to form a neural groove which then makes a neural fold. On day 22, the neural fold closed to form a neural tube. Defects in the closure of anterior neuropores lead to anencephaly and different forms of craniu-bifidum.³ The disease prevalence of spina bifida is in the range of 0.5 – 2/1000 births in the countries, that are deficient in supplementation of folic acid.^{4,5} Presentation of spina bifida varies between asymptomatic (tuft of hair or dermal pit) to severe fatal anomalies (myeloschisis). The patient may be presented with walking difficulties and sphincter disturbances. There are two main types of spina bifida; spina bifida occulta(closed) and aperta(open). The range of defects is encephalocele, anencephaly, congenital dermal sinus, and craniorachischis in the cranial region, while in case of the spine, these are myeloschisis, spina bifida cystica (meningocele and myelomeningocele), lipomyelomeningocele, congenital dermal sinus, diplomyelia, split cord malformation, amniotic band disruption, and diastematomyelia.^{6,7} The spina bifida is associated with hydrocephalus (57% to 86%),^{8,9} chiari malformation type 2 (95%), tethered cord syndrome, and foot deformities. Nowadays due to modern modalities spina bifida diagnosis is possible intrauterine. Fetal ultrasound and maternal serum alpha-fetoprotein are well-known screening tools. Prenatal MRI is an emerging advanced technology that predicts defects and other abnormalities with precision.¹⁰ The management options included intrauterine repair or repair within one week of birth. According to the MOMS (Management of Myelomeningocele Study) trial prenatal surgery decreases the need for shunting and improves motor and sensory function and reverses

hindbrain herniation but postnatal intrauterine scar and premature birth chances increases.¹¹ In the current study we analyzed different factors related to the spectrum of spina bifida.

MATERIAL AND METHODS

Study Design

This prospective cohort study was conducted at the Neurosurgery Department, DHQ teaching hospital, Gomal Medical College, D.I Khan, Pakistan, from July 2021 to July 2022. A total of 100 diagnosed infants of spina bifida of either gender, who were undergoing surgery were included in the study.

Inclusion Criteria

All the diagnosed cases of spina bifida of either gender, which were undergoing surgery for repair of spina bifida, were included.

Exclusion Criteria

Patients with active infection and not fit for General anesthesia.

Data Collection and Analysis

All the data was collected in a pre-designed performance. Different factors like; the total number of patients, sex, age, previous infants with NTD of the parents, consanguinity between the parents, location of the lesion, type of spina bifida, associated with or without hydrocephalus, and postoperative complications, were taken into account. All the patients were admitted to the Neurosurgery department for management. Before surgery, patients were optimized for GA, and all the baseline investigations and CT brain and MRI spine (to know the lesion extension and associated chiari malformation) were also done. Excision and repair were done in spina bifida, while in patients with associated hydrocephalus VP shunt was also done (before spina bifida

repair, after repair; in occult hydrocephalus and simultaneous VP shunt and repair done in selected patients). Stratification between different factors was done.

RESULTS

Age & Gender

A total of 100 patients were admitted to DHQ Teaching Hospital, Gomal Medical College, DI Khan. These were 76 males and 24 females. The age ranges from 1 day to 5 years.

Demographics

Sixty-one patients were from parents who were having consanguineous marriages (first and second cousins), while 39 patients' parents were not relatives.

Operative Findings

Spina bifida is mostly found in the lumbosacral region, in 88 patients (88%), followed by cervical (9 cases) and dorsal (3 cases). Operative findings were; myelomeningocele (39 cases) and meningocele (38 cases), which constitutes the main type of neural tube defect in our study (Table 1). One case operated on was having a single sac with dual defects at the lumbosacral region. Associated hydrocephalus cases were 60, of which 29 were overt and all were shunted. Of the remaining 31 patients, which were having covert hydrocephalus, only 12 became overt and

the VP shunt was done after spina bifida surgery. The relationship between hydrocephalus and the type of lesion was found to be insignificant (Table 3). Spina bifida surgery and VP shunt were done in the same setting in 5 cases. Surgery of spina bifida and hydrocephalus in the same setting was found to be effective and statistically significant (Table 4). Twelve patients of lumbosacral spina bifida need z-plasty for skin closure.

Complications

Postoperatively most frequent complications were wound infection and CSF leak (these were of lower levels), which were re-operated and improved, followed by tethered cord syndrome (observed after 2 years of surgery) and mortality (these were of higher levels) (Table 2).

Table 1: Operative Findings.

Operative Findings	Number of Patients	Percentage
Myelomeningocele	39	39
Meningocele	38	38
Lipo-myelomeningocele	20	20
Myeloschisis	3	3
Total	100	100

DISCUSSION

Spina bifida is one of the complicated and disabling congenital diseases that affect patients and parents physically, psychologically, and financially. Through our study, we analyze

Table 2: Complications.

Complications		Location			Total
		Cervical	Dorsal	Lumbar	
Complication	Wound infection	0	1	12	13
	CSF leak	0	1	7	8
	Tethered cord	2	2	1	5
	Mortality	4	1	0	5
	None	3	3	63	69
Total		9	8	83	100

Table 3: Stratification of Hydrocephalus with Type of the Lesion.

		Preop Hydrocephalus	Hydrocephalus Overt Hydrocephalus	No Hydrocephalus	Total	p-value
Type	Type					
	Meningocele	27	4	7	38	0.187
	Myelomeningocele	23	8	8	39	
	Lipomeningocele	8	8	4	20	
	Myeloschisis	2	0	1	3	
	Total	60	20	20	100	

Table 4: Stratification of Hydrocephalus with Surgery.

		Preop Hydrocephalus	Hydrocephalus Overt Hydrocephalus	No Hydrocephalus	Total	p-value
Surgery	Surgery					
	Repair and VP shunt in one setting	51	0	0	51	0.000 (significant result)
	Repair first and VP shunt on the second setting	9	11	2	22	
	Repair	0	9	18	27	
	Total	60	20	20	100	

maternal and environmental factors that may be responsible for these anomalies. In our study, males are mostly affected than female and the data is compatible with a national study done by Usman et al.¹² According to our study spina bifida are more common in cousin marriage (61%), and nearly the same result was found in Dughal et al, a study in which 67% mothers having cousin marriage.¹³

Through our study, we found lesion levels affected by the type of spina bifida. Verhoef et al¹⁴ also noted Level of the lesion is related to the type of spina bifida with a Spearman's correlation of 0.36. The severe type of lesion mostly affects the higher level of the spine.

As we analyzed that preoperatively hydrocephalus is present in more than 50% of patients, which is somewhat comparable with the literature, which showed 66.4% – 80% of patients with spinal dysraphism harbored hydrocephalus.^{15,16} Spina bifida repair and shunting were performed in one setting in a few cases, however in a few patients, overt hydrocephalus developed after repairing a defect

and VP shunt passed later. Joseph et al,¹⁷ quoted, "The severity of hydrocephalus usually worsens after neurosurgical repair of the lesion in the newborn, requiring placement of a ventricular shunt apparatus".

In our study, some patients with spina bifida and hydrocephalus operated in the same setting, and many researchers including Gamche et al,¹⁸ also stated the fact that both procedures can be done in the same setting. While Ibrahim et al believe that "*VPS insertion should be performed in another session following meningocele sac repair after excluding the presence of infection, especially in cases with a perforated meningocele sac*".

We analyzed the higher-level spina bifida associated with more chances of mortality, while CSF leak is common in lower levels, the more or less same results reported by Date et al,¹⁹ in their research, in which they found the higher the level the more mortality risk.

The study shows most of the patients not encountered any complications after surgery. Few of the patients developed CSF leaks and tethered cords. Caldarelli et al, stated²⁰ that performing

shunt surgery and repair of spina bifida in patients with overt hydrocephalus, results in prevention or decreased frequency of postoperative complications; CSF leak, wound infection, and meningitis. Furthermore, our study is compatible with the above statement that we performed simultaneous VP shunt and spina bifida repair in a few patients, which results in overall successful results.

CONCLUSION

The majority patients of with spina bifida were males. Meningomyelocele and lumbosacral location were the commonest findings. Furthermore, the lumbosacral location of the spina bifida and myelomeningocele were most commonly associated with the development of postoperative 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.

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AUTHORS CONTRIBUTIONS

Sr.#	Author's Full Name	Intellectual Contribution to Paper in Terms of:
1.	Shahid Nawaz	Study design and methodology.
2.	Muhammad Usman	Paper writing, Editing and quality insurer.
3.	Sarah Rehman	Data collection and calculations.
4.	Aneeta Ghazal	Analysis of data and interpretation of results, and Paper writing.
5.	Naseer Hassan	Literature review and referencing.
6.	Naeem-ul-Haq	Editing and quality insurer.