

FREQUENCY OF HYDROCEPHALUS IN MYELOMENINGOCELE PATIENTS IN A TERTIARY CARE HOSPITAL

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ABSTRACT

Background: Myelomeningocele is most complex form of spina bifida and most of the times, it is associated with hydrocephalus. **Objective:** To find the frequency of hydrocephalus in Myelomeningocele patients. **Methodology:** Study design: Cross sectional Study. Setting: Department of Neurosurgery, Sheikh Zayed Hospital, Rahim Yar Khan. Study duration: From 1st January 2012 to 31st December 2013. Study subjects: 52 patients of Myelomeningocele were included in our study and were assessed for different variables like age, sex, site of lesion, presence of hydrocephalus and status of lower limbs (power, sensation and sphincter control). The data was entered and analyzed by using SPSS version 15. **Results:** 34 patients (65.38%) were male and 18 patients (34.61%) were female and common site of lesion was lumbar region (48%) followed by thoracolumbar region (42%). The power of legs was G0-G2 in majority of patients (90%). 40 patients (76%) had hydrocephalus detected by CT Scan brain. **Conclusion:** Our study suggests it is better to put VP Shunt before going for treatment of Myelomeningocele, as majority of the patients of myelomeningocele have hydrocephalus.

Key words: Myelomeningocele, Hydrocephalus, Incidence

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INTRODUCTION

Myelomeningocele is most complex and severe form of spina bifida Aperta. Its incidence varies from 0.2-2 per 1000 live births.¹ It is one of the most common congenital anomalies. It is a multifactorial disease, but genetic predisposition and folic acid deficiency make neural tube defects more likely.²⁻⁸

This anomaly often develops, before the awareness of mother about her pregnancy. Most of the symptoms of neural tube defects result from brain or spinal cord damage which may cause severe disability.^{4,5}

Anatomically spinal cord and connecting nerves are exposed through defect produced by non development of posterior vertebral canal, which result in slow hindbrain herniation. This progressive caudal migration of hindbrain, in association with low-pressure conditions created in the spine (myelomeningocele), along with loss of cerebrospinal fluid (CSF) soon after birth in meningocele sac exacerbates hindbrain hernia and associated hydrocephalus.⁸ It may lead to acute neurological deterioration, which is caused by a combination of raised intracranial pressure, ventriculomegaly and acute bulbar dysfunction (compression of the brain stem in foramen magnum). In majority of patients, ventriculomegaly gradually develops during the first few weeks or months of life. Neurological

status usually improves after ventricular shunting. Different studies showed presence of hydrocephalus in most of children with spina bifida aperta.⁹ Aqueductal stenosis has been reported in 50 per cent of these patients; an associated Chiari malformation is probably the commonest cause of hydrocephalus.¹⁰ Sometimes hydrocephalus may become significant after surgery of myelomeningocele. It was attributed to the removal of the myelocoele sac which was presumed to act as an absorbing surface for the CSF. However there is no convincing proof for this assumption. In few patients, dramatic neurological deterioration occurs after closure of defect, in such case impaction or caudal herniation of hindbrain plays a significant role. Most of children with myelomeningocele will require a shunt which has to be done soon after closure of the spinal defect. In those children who are less severely affected, frequent observation, head circumference measurements and assessment of signs of raised intra cranial pressure, will indicate the need and time of shunt insertion. However majority of children who need a shunt will require it in early child hood.^{6,7,8}

This study was carried out to observe the frequency of hydrocephalus in children with myelomeningocele in our setup. Early awareness of this association is important, as patient may needs multiple surgeries and if hydrocephalus is not treated in time it may prove fatal.

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METHODOLOGY

This cross sectional study included 52 patients of myelomeningocele who were admitted in the department of Neurosurgery of Sheikh Zayed Hospital, Rahim Yar Khan, in two years from 1st January 2012 to 31st December 2013. These patients were admitted from neurosurgery outpatient department and referred from pediatrics medicine department. It included both genders and an age ranging from 20 days to 02 years. Each patient was assessed individually; following data was recorded on a proforma: detailed history, clinical examination including measurement of the head circumference, palpation of anterior fontenella (when open), examination of the spinal swelling and lower limbs, ophthalmoscopic assessment, with a provisional diagnosis of Myelomeningocele. X-ray of the dorsal and lumbar spine with CT scan of the brain was done in all patients. Confirmed patients of myelomeningocele were included in study.

Patients diagnosed with lipomeningocele, Meningocele, or any posterior spinal swelling with or without trauma were not included in study. Similarly patients who were operated for Myelomeningocele and presented later with hydrocephalus were not included in this study.

The data was maintained on a pre-designed proforma mentioning variables like age, sex, clinical examination, diagnosis, presence of hydrocephalus, previous medical history, parents history for inter family marriages, and site of the lesion. The data was entered and analyzed by using SPSS version 15.

RESULTS

A total of 52 patients were admitted during two years. 34 patients (65.38%) were admitted through outpatient department and 18 patients (34.61%) were referred from pediatric medicine department.

Table I: Site of myelomeningocele.

Site	Number	Percentage
Cervical	1	1.92%
Thoracic	4	7.69%
Thoracolumbar	22	42.30%
Lumbar	25	48.07%
Total	52	100%

All patients were children with an age ranging from 20 days to 02 years. Majority (65%) of them were male. Most of our patients (96%) had interfamily marriages.

Table II: Presence of hydrocephalus

	Number of patients	Percentage
With Hydrocephalus	40	76.92%
Without Hydrocephalus	12	23.07%
Total	52	100 %

Table III: Status of lower limbs

Lower limbs	No	Percentage	
Power	G-0	15	28.84%
	G1-2	22	42.30%
	G3-4	15	28.84%
Sensations	Decreased	13	25%
	Absent	39	75%
Sphincter Control	Good control	05	09.61%
	Poor control	47	90.38%

The common site of myelomeningocele in our study was in lumbar region in 48% patients, followed by a thoracolumbar position in 42% patients as in shown in table I. The male patients were of age ranging from 25 days to 1 year whereas females were in 20 days to 02 years. CT scan of the brain was done in every patient to see for the presence of hydrocephalus. 40 patients (76%) had hydrocephalus as depicted in table II.

The motor status of legs of the patients ranged from G0 to G4, while majority had G to G2 power, 47 patients (90%) had poor sphincter control, absent sensations in legs were noted in 39 patients (75%) and decreased sensations were found in 13 patients (25%).

DISCUSSION

Myelomeningocele is one of the most common spinal anomaly seen at birth with an average worldwide incidence of 1–2 cases per 1000 births, though certain populations have a significantly greater risk. Its association with hydrocephalus varies markedly in various studies from 69% to 92%.¹¹ Most of studies show high incidence of this association due to displaced cerebellum which interferes with normal circulation of cerebrospinal

fluid. Early diagnosis and treatment of hydrocephalus helps in proper management of Myelomeningocele patients. Untreated hydrocephalus in these patients causes pressure effects of CSF on wound site, which results in CSF leakage or wound dehiscence after Myelomeningocele surgery. Some of these patients develop fatal hydrocephalus.

In a study done by Kojima et al, 95% of their patients with Myelomeningocele showed hydrocephalus. Patients under study were below 3 years of age, all underwent CT scan of brain to exclude hydrocephalus. Children showed associated hydrocephalus. Their study was a big as compared to current study having a sample size of 83 patients.¹²

A study done by Kumar et al, showed their incidence of hydrocephalus to 58.8% in Myelomeningocele patients. Their sample size was small as compared to current study, 9 patients with a mean age of 18 months. Diagnostic criteria for hydrocephalus was CT scan of brain.¹³

In a study done by Kawamura et al, they had incidence of hydrocephalus in their Myelomeningocele patients to an average of 65.5%. Their sample size was also small, 25 patients, with age ranging from one month to 2 ½ year. Diagnostic criteria was C.T scan of brain.¹⁴ Similarly in a study which included 47 patients of Myelomeningocele, all have C.T Scan of brain and majority patients showed associated hydrocephalus.¹⁴

It is an established fact that most of the Myelomeningocele patients have hydrocephalus which may appear early or later, before or after surgery of Myelomeningocele.

CONCLUSION

Keeping in view, results of our study and international studies done to digout the incidence of hydrocephalus in Myelomeningocele patients, our view is that most of the Myelomeningocele patients have hydrocephalus. It is suggested to put a shunt in all those patients showing ventriculomegaly before definite surgery for Myelomeningocele to have best results.

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