

ORIGINAL ARTICLE

# Short Term Complications of Myelomeningocele Repair. An Experience in Neurosurgery Department Lady Reading Hospital Peshawar

MUHAMMAD IDRIS KHAN, WEFAQ ULLAH, MUHAMMAD ISHFAQ

*Bakht Zar Khan, Mumtaz Ali*

*Department of Neurosurgery, PGMI Lady Reading Hospital, Peshawar*

## ABSTRACT

**Objective:** To determine the short term complications of Myelomeningocele repair.

**Material and Methods:** This cross sectional study was conducted at Neurosurgery Department, PGMI Lady Reading Hospital, Peshawar from January 2014 to June 2014 with total 6 months duration. All patients having age from 1 to 6 months with either gender who were operated for myelomeningocele were included and those patients having MMC with a prior VP shunt, moribund patients who do not survive for 30 days were excluded from the study. Patients age, gender and short term complications of surgery were documented on pre designed proforma. Minimum one month follow up was done. Data was analyzed by SPSS version 17 and expressed in the form of tables and charts.

**Results:** In this study 58% children were in age range 1 – 2 months and 42% children were in age range 3 – 6 months. Mean age was 2 months with  $SD \pm 1.26$ . Fifty five percent children were male and 45% children were female. VP Shunt was put in 12% children, CSF Leak was found in 15% and infection was found in 23% of cases.

**Conclusion:** Wound site infection and CSF leak are the most common short term complications of myelomeningocele repair.

**Key Words:** Ventriculoperitoneal shunt, Myelomeningocele, Spinal cord, CSF leak.

**Abbreviations:** CNS: Central Nervous System. MMC: Myelomeningocele. HDC: Hydrocephalus. CSF: Cerebrospinal Fluid. NTD: Neural Tube Defects. VP: Ventriculoperitoneal.

## INTRODUCTION

Congenital fusion defects of the spine and associated tissues are classified as spinal dysraphism. Neural tube defects vary in severity and affects tissues overlying the spinal cord; meninges, vertebral arch, muscles and skin. Amongst the congenital defects of the Central nervous system (CNS) spina bifida cystica (Myelomeningocele; MMC) is the most common<sup>1</sup>. MMC is characterized by an exposure of the nervous system and/or the meninges to the environment because of a congenital bone defect.<sup>2</sup> Failure of neural tube formation and closure may occur anywhere along the neural axis. MMC occurs in 0.4 – 1 per 1000 live birth and incidence varies with environmental and genetic

factors.<sup>3</sup>

The cause of myelomeningocele (MMC) is multifactorial and includes genetic predisposition, nutritional deficiencies, particularly foliate and zinc, use of anti-epileptic drugs like carbamazepine or valproic acid, diabetes mellitus (type – 1), pre-pregnancy obesity and possible other non-medical factors such as agricultural pesticides, radiations, hyperthermia and use of tobacco or drugs.<sup>6</sup> While the etiology of MMC remains poorly understood, primary failure of either neural tube or mesenchymal closure at the caudal neuropore in the embryonic period, results in exposure of the developing spinal cord to the uterine environment. Without protective tissue coverage, secondary des-

truction of the exposed neural tissue by trauma or amniotic fluid may occur throughout gestation.<sup>4</sup> Children with MMC who survive are likely to have life – long disabilities. Functional problems may often result from the neurologic defect or the surgical repair.<sup>2</sup>

Patients with MMC have various sensory and motor deficits such as lower limb weakness and loss of sphincter control.<sup>3,4</sup> MMC is associated with other congenital abnormalities.<sup>1-4</sup> Among associated abnormalities are Hydrocephalus, Chiari malformation, scoliosis, telipes equinovarus, flat/high arched foot and trophic ulcer.<sup>2-4</sup> Electrolyte imbalance may present as hypokalemia, hyperkalemia, hypernatremia. Children may also be anemic and malnourished.<sup>3</sup> These patients may have intraoperative complications like arrhythmias, hypotension, bronchospasm, laryngospasm, hypoxemia and hypothermia.<sup>3</sup> MMC most commonly affect the lumbosacral area, followed by lumbar and then sacral and thoracic areas.<sup>3</sup>

The traditional treatment of MMC is postnatal repair.<sup>4</sup> The surgical management is poised by post-operative complications such as post-operative cerebrospinal (CSF) leak, infection, hydrocephalus needing a ventriculoperitoneal (VP) shunt and wound dehiscence.<sup>2,4,5</sup> Hydrocephalus (HDC) develops in more than 85 – 90%<sup>5</sup> of patients with MMC.<sup>5</sup> At least 54%<sup>5</sup> to 80%<sup>4</sup> of MMC patients require placement of shunts to prevent the neurologic and intellectual compromise that accompanies significant ventriculomegaly.<sup>4</sup> Post-operative CSF leak is amongst the deadly complications following MMC surgery; increasing the morbidity and mortality of the patient.<sup>4,6</sup> The incidence of the post-operative CSF leak ranges from 18%<sup>7</sup> to 30%.<sup>5</sup>

The rationale of current study is to determine the short term complications of MMC repair in our setup. MMC repair is a commonly performed procedure in the Neurosurgery Department, infants are more prone to congenital anomalies due to lack of proper antenatal work-up in this part of the world. Care of a patient with myelomeningocele is a big headache for the family with the added burden of post-operative complications. Doing this study will help us to generate local statistics of post MMC repair complications. The results of this study will be shared with other neurosurgeons through publications to make them aware about the local magnitude of the problem and on the basis of results of this study we will be able to further rectify our management strategy for the future.

## **MATERIALS AND METHODS**

This cross sectional study was conducted at Neurosurgery Department, PGMI Lady Reading Hospital, Peshawar from January 2014 to June 2014 with total 6 months duration. All patients having age from 1 to 6 months with either gender who were operated for myelomeningocele were included and those patients with. Patients having MMC with a prior VP shunt, moribund patients who do not survive for 30 days and having meningocele were excluded from the study. Patients age, gender and short term complications of surgery were documented on pre designed proforma. Minimum one Month follow-up was done. Data was analyzed by SPSS version 17 and expressed in the form of tables and charts.

After permission from hospital ethical committee, patients with MMC admitted in Neurosurgery Department of Lady Reading Hospital was approached. Those who fulfill inclusion criteria were included in the study. Informed consent was taken from all patients. These patients were further assessed through detailed history, including personal particulars, name, age, sex, address, symptoms and clinical examination. Diagnosis of MMC was made on the basis of clinical and per-operative findings. The patient mobile number would be taken for further correspondence. The patients were followed post operatively for one month. Descriptive statistics like mean  $\pm$  standard deviation was used for age. Frequency/percentage was calculated for categorical variables like gender and short term complications (wound infection, CSF Leak and VP-Shunt). Short term complications were stratified among the age and gender to see the effect modifiers. Results were presented in tables for different variables. All results were presented in the form of charts and graphs.

## **RESULTS**

This study was conducted at Neurosurgery Department, Lady Reading Hospital, Peshawar in which a total of 106 children were observed to find the short term complication of myelomeningocele repair and the results were analyzed as. Age distribution among 106 children was analyzed as 61 (58%) children were in age range 1 – 2 months while 45 (42%) children were in age range 3 – 6 months. Mean age was 2 months with SD  $\pm$  1.26 (as shown in table 1). Gender distribution among 106 children was analyzed as 58 (55%) children were male while 48 (45%) children were female (as shown in table 2). Short term complication of myelomeningocele among 106 children was ana-

lyzed as VP Shunt was found in 13 (12%) children, CSF Leak was found in 16 (15%) children and infection was found in 24 (23%) children (as shown in table 3).

**Table 1:** Age Wise Distribution (n = 106).

Age	Frequency	Percentage
1 – 2 months	61	58%
3 – 6 months	45	42%
Total	106	100%

Mean age was 2 months with SD ± 1.26

**Table 2:** Gender Wise Distribution (n = 106).

Gender	Frequency	Percentage
Male	58	55%
Female	48	45%
Total	106	100%

**Table 3:** Frequency of Short Term Complications (n = 106).

Complications	Frequency	Percentage
VP shunt	13	12%
CSF Leak	16	15%
Infection	24	23%
Total	53	50%

Stratification of short term complication of myelomeningocele with age distribution was analyzed as in 13 cases of VP Shunt 5 children were in age range 1 – 2 months and 8 children were in age range 3 – 4 months. In 16 cases of CSF Leak 4 children were in age range 1 – 2 months and 12 children were in age range 3 – 4 months. In 24 cases of infection 10 children were in age range 1 – 2 months and 14 children were in age range 3 – 4 months (as shown in table 4). Stratification of short term complication of myelomeningocele with gender distribution was analyzed as in 13 cases of VP Shunt 7 children were male and 6 children were female. In 16 cases of CSF Leak 10 children were male and 6 children were female. In 24 cases of infection 12 children were male and 12 children were female (as shown in table 5).

**Table 4:** Stratification of Complications with Age Distribution (n = 53).

Complications / Age	1 – 2 Months	3 – 4 Months	Total
VP shunt	5	8	13
CSF Leak	4	12	16
Infection	10	14	24
Total	19	34	53

Chi square test was applied in which P Value was 0.831

**Table 5:** Stratification of Complications with Gender Distribution (n = 53).

Complications / Gender	Male	Female	Total
VP shunt	7	6	13
CSF Leak	10	6	16
Infection	12	12	24
Total	29	24	53

Chi square test was applied in which P Value was 0.902

## DISCUSSION

Neurosurgeons should focus on the neurosurgical disorders that affect a large number of children.<sup>8</sup> Third world countries are facing many problems; socio-economic, cultural, educational and nutritional that result in congenital anomalies of CNS like neural tube defects (NTD) more frequently than developed societies like the USA or Europe. NTD involves entire central nervous system and leads to disability or death. Children are the most valuable individuals in the lives of most people. Nothing else can cause the grief than the death of the child does, and few disasters cause the grief of a permanently brain damaged child.<sup>9</sup> The most common form of neural tube defect is myelomeningocele a term used synonymously with spina bifida aperta, spina bifida cystica and open neural tube defect.

As the term implies, there is some form of cyst apparent, even if it collapses shortly after birth, It occurs because spinal cord fails to fuse dorsally during primary neurulation during days 18 – 27 of human embryo genesis leaving a flat plate of neural tissue called a neural placode.<sup>10</sup> The prevalence of myelomeningo-

gocele has declined in developed countries of the world owing to both prenatal foliate supplementation and to pregnancy termination following prenatal diagnosis. In United States before 1980 prevalence of myelomeningocele was 1 – 2/1000 live births, but more recently prevalence has declined to 0.44 per 1000 live births.<sup>11</sup> Unfortunately, in third world countries prevalence is much higher, and acceptable prevalence data are not available, nor has the issue been addressed with the goal of eradication, or reduction of incidence. The cause of myelomeningocele is multifactorial and includes genetic predisposition, nutritional deficiencies, particularly foliate and zinc, use of anti-epileptic drugs like carbamazepine or valproic acid, diabetes mellitus (type – 1), pre-pregnancy obesity and other non- medical factors such as agricultural pesticides, radiation, hyperthermia and use of tobacco or drugs. The defect involve neural tissue. Skin over the cystic lesion is not fully developed. The defect size in our series was an average 30.9 cm.

In our series all patient had undergone CT-Brain i.e. 106 (100%) to determine ventricular size and other anomalies. MRI spine was done in selected cases for detecting Chiari – II malformations. In our series 106 patients having MMC were parapertic and incontinent. In our study 87% myelomeningocele were located at thoracolumbar region, 5% were located in cervical region, and 7% in thoracic region, 20% were associated with hydrocephalus or other anomalies.<sup>12</sup> In our study hydrocephalus (after repair of MMC) requiring VP Shunt was found in 13 (12%) children, CSF Leak was found in 16 (15%) children and infection was found in 24 (23%) children.

Similar findings were observed in study done by James HE et al,<sup>8</sup> in which VP Shunt was put in 10% children, CSF Leak was found in 14% children and infection was found in 20% children. As for as normal intelligence is concerned, ventriculitis is the single most important factor, diagnosed at different stages during the care of patient, either before repair of myelomeningocele, after repair, before VP shunt insertion or after placement of VP shunt. Lee SK et al<sup>9</sup> had observed 26 cases (61%) with ventriculitis related to myelomeningocele repair and in 17 cases (4%) due to VP shunt. Organisms found in these cases were usually gastro intestinal tract or skin colonizers. Almost all patients with ventriculitis suffered mental retardation and are under continued follow-up he encountered 16 (3.8%) cases with ruptured myelomeningocele either due to birth trauma, a very thin placode and massive hydrocephalus or due to mishandling by

medical professionals. One of the most important issues concerned with management protocol is the optimal timing of myelomeningocele repair.

It is generally accepted that repair ideally should be performed 72 hours after birth to avoid ventriculitis.<sup>13</sup> In his series 16 cases i.e., 3.8% were repaired within 48 – 72 hours of birth because of CSF leak. In the rest of the cases average time of repair was 28 days, due to delayed referral of the cases. All cases had cultures taken from the neural placode and only sterile cases were directly repaired; others were treated with antibiotics before repair. In all infected cases shunt surgery was delayed, until investigation and treatment for ventriculitis was successful. The main purpose of myelomeningocele repair is to protect the functional tissue in the neural placode, to prevent loss of CSF and minimize the risk of meningitis by reconstructing the neural tube and its coverings with a stable soft tissue closure. To avoid complications, use of lumbar periosteal turn over flap and tissue expansion for delayed closure of large myelomeningocele has been advocated.<sup>12</sup>

Recently, rectal monitoring during repair of myelomeningocele has been performed to preserve neural tissues.<sup>14</sup> In our series all cases were repaired with standard surgical techniques. In another series, a low pressure VP shunt was placed without repair of MMC in 52 of 155 cases (33.5%). There was 100% shrinkage of myelomeningocele, saving operation time, avoiding problems with wound closure due to very thin and inadequate skin, and minimizing hospital stay due to poor wound healing. However, neurological deficits were irreversible because of cord tethering.<sup>5</sup> All cases were repaired with standard procedure and in 365 cases (88%) VP shunt was put in.

Restoration of neural C.S.F dynamics can reduce flow into the sac resulting in meningocele size reduction. In another series there were wound healing problems including skin necrosis CSF leakage, and gross wound infection in 22 cases (5.3%). There was strong association of poor wound healing with ventriculitis. Out of total cases, 26 (6%) had ventriculitis, and in the experience the incidence of ventriculitis was strongly associated with wound infection of the myelomeningocele closure.<sup>16</sup> There were VP shunt infections in 17 cases (4%) only and the rate was more common in patients previously treated for ventriculitis. There was a close relationship between wound healing and placement of VP shunt. The incidence of myelomeningocele repair breakdown was very low because of our protocol wherein all patients with hydrocephalus had a

VP shunt before repair or simultaneously. To avoid C.S.F leak other factors were also considered like skin flaps; to avoid tension at skin edges.

In another series hydrocephalus was present in 365 cases (88% of total) and all were shunted. The incidence of ventriculitis in these patients after shunt insertion was present in 17 cases (4%). Different factors observed were responsible for shunt related infection including, age at shunt placement, local condition of scalp, duration of shunt surgery, use of prophylactic antibiotics and general condition of the patient.<sup>17</sup> Overall results were encouraging; there were 5 deaths (1.2%) due to ventriculitis, ruptured meningocele and shunt infection. All other patients are under follow up for their associated anomalies such as hydrocephalus, Chiari malformation and other bony anomalies. Following repair of myelomeningocele there is the possibility of tethered cord syndrome leading to neurological and urologic deficits.<sup>18,19</sup>

## CONCLUSION

There is strong need of public health awareness programme in all under developed countries with special reference to nutritional needs like folic acid deficiency. Prenatal diagnosis with high resolution fetal ultrasonography should be done and the site and size should be determined for a recommendation of termination of pregnancy. Early surgical repair must be done to avoid further complications. Our method constructs a secure and water tight covering over spinal cord, provides durable soft tissue closure, and may be used in conjunction with other traditional skin or muscle flap techniques.

*Address for Correspondence:*

*Dr. Wefaq Ullah*

*Department of Neurosurgery*

*PGMI Lady Reading Hospital, Peshawar*

*E-mail: wefaquallah@yahoo.co.in*

*Cell No: +923339723982*

## REFERENCES

1. Singh P, Soni A, Singh RJ. Double meningocele; case report and anatomic rationale. *J Anatom Societ Ind.* 2008; 57 (1): 43-6.
2. Idris B. Factors Affecting the Outcomes in Children Post-Myelomeningocele Repair in Northeastern Peninsular Malaysia. *Malaysian J Med Sci.* 2011; 18 (1): 52-9.
3. Chand MB, Agrawal J, Bista P. Anaesthetic Challenges and Management of Myelomeningocele Repair. *Postgrad Med J NAMS.* 2011; 11 (1): 41-6.
4. Adzick NS. Fetal myelomeningocele: natural history, pathophysiology, and in-utero intervention. *Semin Fetal Neonatal Med.* 2010; 15 (1): 9-14.
5. Radmanesh F, Khashab F, Nejat ME. Shunt complications in children with myelomeningocele; Effect of timing of shunt placement. *J Neurosurg Pediat.* 2009; 3: 516-20.
6. Hashim ASM, Ahmad S, Jooma R. Management of Myelomeningocele. *J Surgery Pakistan (Inter).* 2008; 13 (1): 7-11.
7. Sankhla S, Khan GM. Reducing CSF shunt placement in patients with spinal myelomeningocele. *J Pediatr Neurosci.* 2009; 4 (1): 20-9.
8. Ikasi. James HE, Lubinsky G. Terminal myelocystocele. *J Neurosurg.* 2005; 103: 443-5.
9. Lee SK, Gower DJ, MC Whorter JM. The role of MR imaging in the diagnosis and treatment of anterior sacral meningocele *J Neurosurg.* 1988; 69: 628-31.
10. Logan WJ. Neurological examination in infancy and childhood chapter 200 sections VII. *Pediatrics,* 3169-86.
11. MC Neely PD, Howe WJ. Ineffectiveness of dietary folic acid supplementation on the incidence of lipomyelomeningocele: pathogenic implications. *J Neurosurg.* 2004; 100: 98-100.
12. Sharif S, Allcutt D, Markes C. Tethered cord syndrome; recent clinical experience. *Br J Neurosurg.* 1997; 11: 49-51.
13. Park TS, Cail WS, Maggio WM. Progressive spasticity and scoliosis in children with myelomeningocele. Radiological investigation and surgical treatment. *J Neurosurg.* 1985; 62: 367-75.
14. Lee SK, Gower DJ, MC Whorter JM. The role of MR imaging in the diagnosis and treatment of anterior sacral meningocele *J Neurosurg.* 1988; 69: 628-31.
15. Taskaynatan MA, Izci Y, Ozgul A, Hazneci B, Dursun H, Kalyon TA. "Clinical significance of congenital lumbosacral malformations in young male population with prolonged low back pain". *Spine,* 2005; 30 (8): E210-3.
16. Steinbok P, Cochrane DD. The nature of congenital posterior cervical or cervico thoracic midline cutaneous mass lesions. *J Neurosurg.* 1991; 75: 206-12.
17. Tamaki N, Shirataki K, Kojima N. Tethered cord syndrome following repair of myelomeningocele. *J Neurosurg.* 1988; 9: 393-8.
18. Vernet O, Farmer JP, Houle AM. Impact of urodynamic studies on the surgical management of spinal cord tethering. *J Neurosurg.* 1996; 85: 555-9.
19. Warder DE, Oakes WJ. Tethered cord syndrome and the conus in a normal position. *Neurosurgery,* 1993; 33: 374-8.

**AUTHORS DATA**

<b>Name</b>	<b>Post</b>	<b>Institution</b>	<b>E-mail</b>	<b>Role of Authors</b>
Dr. M. Idris Khan		Department of Neurosurgery, PGMI Lady Reading Hospital, Peshawar		Data Collection
Dr. Wafaq Ullah			wefaqullah@yahoo.co.in	Paper Writing
Dr. Muhammad Ishfaq				Tables
Dr. Bakht Zar Khan				Discussion
Dr. Mumtaz Ali	Professor			Critical Review