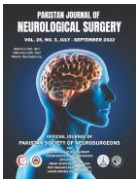

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Original Article

Comparison of Outcomes of Early and Late Surgical Interventions in Lipomeningomyelocele (LMMC) and Lipomeningocele (LMC)

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ABSTRACT

Objectives: The study compared the signs and symptoms of post-operative complications in early vs. late intervention lipomeningomyelocele (LMMC) and lipomeningocele (LMC).

Materials and Methods: We compared the clinical and surgical data between two groups i.e., lipomeningomyelocele (n = 189) and lipomeningocele (n = 64), and their early vs. late surgical interventions for 3 years from January 2018 to July 2021. We included patients of both genders (n = 253) with lipomeningomyelocele or lipomeningocele aged up to 7 years. A detailed neurological exam i.e., sensory, motor, and cerebellar signs was performed to evaluate the patients.

Results: The presentation of LMMC (74.7%) was very high compared to LMC (25.3%). 74.7% underwent detethering of the spinal cord, as they had cord tissue coming out of the defect. 25.2% had only meninges coming out of the bony deficiency and performed dural repairs. 47 patients had incontinence which was improved postoperatively. Sixty-nine patients had hydrocephalus which was treated with VP shunt or ETV. 23 patients had diastematomyelia which is a bony spur duly repaired intra-operatively. 50 presented with paraplegia and 19 cases with club feet. The majority of patients in both groups, reported for Power would fall between 3/5-4/5. For patients who underwent late intervention, 7 presented with post-operative incontinence, 12 with hydrocephalus, 12 with CSF leakage, and 13 with paraplegia.

Conclusion: If performed on time, surgical intervention in lipomeningocele and lipomeningomyelocele yields good results. Early intervention is substantially better for managing post-op CSF leakage and incontinence than late intervention.

Keywords: Lipomeningocele (LMC), Lipomeningomyelocele (LMMC), Diastematomyelia.

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INTRODUCTION

Spina bifida has an overall incidence of 1-2/1000 live births worldwide and 78/1000 live births in Pakistan. That is a huge burden both on the economy and human resources. Neural tube defects are a group of defects involving the brain, spine, and spinal cord. These defects are caused by the failure of the caudal neuropore to close in 4th week of embryonic development[1] with an incidence of 4.7:10,000 in developed countries.[1-2] The exact cause of defects is unknown but women taking enough amount of folic acid during and before the period of organogenesis are less prone however women who are obese, have diabetes mellites, family history of birth defects, or taking antiseizure drugs, anti-metabolite drugs are at increased risk of developing these defects.[3-7]

Procedures done prenatally are relatively safer and show insignificant benefits. During the prenatal period early closure shows improved motor function, and they have less likely hindbrain herniation which results reduce cerebral spinal fluid diversion.[2] Fetal ultrasound is done for diagnostics of myelomeningocele. Physicians prefer to do ultrasound manifestation during the first and second trimesters of pregnancy. It can be accurately diagnosed by physicians in 2nd trimester of life (18 – 22 weeks). A procedure like an amniocentesis can also be done by a physician to exclude any genetic disease that is associated with myelomeningocele.[7] Prenatal surgery can be done and is highly suggestive based on the severity of myelomeningocele and the health of the mother. This surgery can help prevent the risk of further damage to the spinal cord or nerves already present. Patient with prenatal complication is more likely to get prenatal complications during the early trimesters of life causing uterine dehiscence, thinning, or tearing at the incision at the uterus at the time of delivery.[7]

Low maternal intake of iron, magnesium, and niacin also causes birth defects. Defects can be divided into spina bifida occulta i.e., the defect is covered with skin having a tuft of hair, and only CSF coming out of bony deficiency is associated with arsenic exposure. Spina bifida aperta are open spinal column defects where meninges protrude out of the spine forming a tethered sac i.e., occult tethered cord syndrome.[6-10]

Symptoms of motor and sensory loss are predominantly seen in Myelomeningocele which includes changes in bladder dysfunction and paraplegia. NTDs (neural tube defect) end can cause cranial fissure malformation. The most severe manifestation of this, anencephaly, is incompatible with life. Meningomyelocele is a defect in which brain tissue itself protrudes out along with its protective coverings. Leptomeningeal where the brain protrudes out of the spinal column forms a sac under the skin and attaches itself to the adipose tissue of the back. This can be diagnosed via ultrasonography and magnetic resonance imaging and also has started to play a role antenatally or presents with neurological symptoms in the early years of life. Symptoms with which they present mostly are movement-related, learning disabilities, and speech problems.[11-12] They may also get sphincter incontinence, constipation, gastric paresis, and cord tether. Early intervention in lipomeningocele and other spinal defects can get us better outcome than seen in developed countries spinal dysraphism are being diagnosed antenatally and being operated upon intrauterine. That arrives us to the understanding of early intervention with better prognostic features post operatively.

MATERIALS AND METHODS

Study Design

We conducted an observational study at the Children Complex, Nishtar Medical University, Multan, and at the Department of Neurosurgery, Punjab Institute of Neurosciences (PINS), Lahore Pakistan. The study compared the epidemiology, demographic data, and preoperative and

postoperative variables. We compared the clinical and surgical data between two groups i.e., lipomeningomyelocele and lipomeningocele, and their early vs. late surgical interventions for 3 years from January 2018 to July 2021.

Inclusion Criteria

We included patients with ages up to 7 years, of both genders who were having lipomeningomyelocele or lipomeningocele. Patients who took vitamin supplements or those who were not in the first trimester. Patients taking any antiepileptic, anti-metabolite, or otherwise medications. Patients included had communicating hydrocephalus.

Exclusion Criteria

Patients having other congenital anomalies with lipomeningomyelocele and lipomeningocele were excluded. Patients having psychosis or sepsis were not included. Patients having low GCS, with ventriculitis, or age more than 7 years was also not included.

Clinical Management & Radiology

We devised a proforma which included the evaluation of the demographics of the patients, and postoperative symptoms i.e., focal neurological deficit, GCS, any other associated morbidity and surgical outcome, neurological examination, cerebellar signs, sunset sign, Babinski sign, or motor examination. Cranial ultrasounds and CT scans Brain to track hydrocephalus, CT scan of the lumbosacral spine, and MRI of the brain and lumbosacral spine were radiological tests performed. We monitored the ECG, chest X-ray, and hemorrhage profiles. Before the procedure, they had PT, APTT, INR, BT, and CT tests. To rule out infection, 10% of patients had their CSF completely examined and cultured. Before surgery, a CT brain scan was conducted in all instances to search for bony deformities. BP was maintained in the 140-160 mmHg range postoperatively. An MRI was performed to check for cord tethering, whether or not neural tissue is coming out of the defect, and the level at which faults are happening. The overall follow-up duration was three months through OPD. Antibiotics, pain relievers, and antiemetics are given to patients for medical care. For co-morbidities, patients were encouraged to obtain a medical check-up, physiotherapy follow-up, and neurologist consultations.

Cognitive Assessment

To check for higher mental function associated with hydrocephalus and hydrocephalus-related parameters i.e., the requirement of shunt and shunt revisions and infections were also included. A detailed neurological exam i.e., sensory, motor, and cerebellar signs was performed to evaluate the patients. We did a fontanelle examination of all our patients. For this study, we consulted a psychiatrist, anesthetist, neurosurgeon, and neurologist.

Surgical Procedure

All patients we operated on within 12 hours of admission. Neurosurgical. A standard surgical procedure under general anesthesia with noninvasive cardiac monitoring was performed. The patient was positioned prone on gel rolls to avoid pressure sores. We localized the position of spina bifida with help of a tuft of hair, step-off in the spinal column, or magnetic resonance imaging. Longitudinal incision dorsal to the placode was made superiorly up till the last intact vertebra. All the fascial planes and muscle layers were splatted. Ligamentum flavum is released with the help of a periosteal elevator. Before opening the dura, the child was tapped into the Trendelenburg position to decrease CSF flow toward the lower part of the spinal column. If a fibro-fatty tissue was presented, a carbon dioxide laser was used to identify the dura distally. The

the dural sac was opened and is continuously irrigated with normal saline. All the spinal nerves emitting laterally were also secured carefully. Tethered spinal cord or meninges were released. Sac secured and dura repaired in reverse order. If needed; the spine, bone grafted, tendon transferred and all other layer layers closed were in reverse order under aseptic conditions.

Data Analysis

SPSS version 25 was used for data entry and statistical analysis. A chi-square test was applied to see significant/ insignificant differences between the two groups (early vs. delayed repairs).

Table 1: Clinical Data Comparison Between Lipomeningomyelocele (LMMC) and Lipomeningocele (LMC) [Total: 253].

Variable	Categories	LMMC N = 189	LMC N = 64
Gender	Male	106 (56%)	34 (53%)
	Female	83 (44%)	30 (15.8%)
Incontinence	Yes	34 (18%)	13 (20.3%)
	No	155 (82%)	51 (79.6%)
Hydrocephalus	Yes	50 (26.4%)	19 (29.6%)
	No	139 (73.5%)	45 (70.3%)
Diastematomyelia	Yes	19 (10%)	4 (6.25%)
	No	170 (90%)	60 (93.7%)
Paraplegia	Yes	38 (20%)	12 (18.75%)
	No	151 (80%)	52 (81.2%)
Club foot	Yes	15 (8%)	4 (6.25%)
	No	174 (92%)	60 (93.7%)
Power	0/5	38 (20%)	12 (18.75%)
	1/5	5 (2.6%)	5 (7.81%)
	2/5	27 (14.2%)	7 (10.9%)
	3/5	48 (25.3%)	17 (25.5%)
	4/5	70 (37%)	23 (35.9%)
	5/5	1 (0.5%)	0 (0%)

RESULTS

Incidence of LMMC and LMC

A total of 253 patients performed Spina bifida repair in our center in 3 years with 189 cases of lipomeningomyelocele (LMMC) and 64 cases of lipomeningocele (LMC). We observed in our study that the presentation of LMMC (74.7%) was very high compared to LMC (25.3%) in our center. The incidence of lipomeningomyelocele is high in our ethnicity.

Age & Gender Distribution

The age of our patients was in the range of 2 days to 7 years. Among the LMMC group, M: F ratio was high and M = F was found in the case of the LMC group. When we compare (57%) fit into the category of lumbar section of spinal column abnormalities and were included, while the remainder were removed based on our inclusion criteria. 56% were males and 44% were females in the LMMC group, whereas, 53% were male and 15.8% were female patients in the LMC group. Sixty-four (25.2%) out of 253 had only meninges coming out of the bony deficiency and were performed the dural repair.

Clinical Data on LMMC and LMC

Out of 253 patients, 189 (74.7%) underwent detethering of the spinal cord, as they had cord tissue coming out of the defect. 47 patients of both genders had incontinence which was improved postoperatively. Sixty-nine patients had hydrocephalus which was treated with VP shunt or ETV depending upon whether it was obstructive or non-obstructive. Only 23 patients had diastematomyelia which is a bony spur duly repaired intra-operatively. 50 cases presented with paraplegia and 19 cases with club feet. The majority of patients in both groups, reported for Power would fall between 3/5-4/5 (See Table 1 for details).

Table 2: Comparing Early Vs. Late Interventions [Total: 253].

Variable	Categories	Early Repair N = 16	Late Repair N = 237	p-value
Post-op Incontinence (No. of Patients)	Yes	1	7	$\chi^2 = 0.531$ p-value = 0.466
	No	15	230	
Post-op Hydrocephalus (No. of Patients)	Yes	1	12	$\chi^2 = 0.0433$ P value = 0.830
	No	15	225	
Post-op CSF Leakage (No. of Patients)	Yes	0	12	$\chi^2 = 0.035$ P value = 0.356
	No	16	225	
Post-op Paraplegia (No. of Patients)	Yes	1	13	$\chi^2 = 0.0179$ P value = 0.897
	No	15	224	
Post-op Complications (No. of Patients)	Yes	0	0	–
	No	16	237	
Mean Hospital Stay	In Days	4 – 5	4 – 6	
Mean Follow up	In Days	6	6	

Early Vs. Late Intervention Outcomes

Out of 16 patients, who underwent early intervention, one presented with post-operative incontinence, one with hydrocephalus, none with CSF leakage, one with post-operative paraplegia, and their mean hospital stay was from 4 to 5 days. Out of 237 patients, who underwent late intervention, 7 presented with post-operative incontinence, 12 with hydrocephalus, 12 with CSF leakage, and 13 with paraplegia, and the mean hospital stay was 4 to 6 days. As per the Chi-square test, an insignificant difference was observed between the presence/absence of different complications in early vs. late repair groups (See Table 2 for details).

DISCUSSION

The current research evaluated the signs and symptoms of postoperative complications in patients with lipomeningomyelocele (LMMC) and lipomeningocele (LMMC) (LMC). Our patients had a history of altered state of consciousness, vomiting, fits and headaches, dementia, gait instability, and urine incontinence. Because regional hospitals lack facilities, patients always arrive with problems or late. No mortality was observed during our study. In our facility, the presentation of LMMC was much higher than that of LMC. Because patients had cord tissue coming out of the defect, 74.7% had spinal cord detethering. 25.2% had just meninges protruding from the bone deficit and underwent dural repairs. Incontinence was improved in 47 individuals after surgery. Sixty-nine individuals were diagnosed with hydrocephalus and treated with a VP shunt or ETV. Diastematomyelia is a bony spur that was treated intraoperatively in 23 cases. There were 50 occurrences of paraplegia and 19 cases of club feet. The majority of patients in both groups reported a Power score of 3/5 – 4/5. One patient with post-operative incontinence, one with hydrocephalus, and one with post-operative paraplegia presented with early management (n = 16). Out of 237 patients who had a late intervention, 7 had post-operative incontinence, 12 had hydrocephalus, 12 had CSF leakage, and 13 had paraplegia. The earliest intrauterine interventions significantly reduce complications. Prenatal intervention, which has been found to minimize shunt installation and enhance motor abilities, has become the standard

of therapy for prenatal diagnosis of myelomeningocele. However, the urological advantage of early intervention is yet unknown. Patients with lipomeningocele, on the other hand, may benefit from early surgical correction.[11] A study sought to determine if in-utero repair of an experimental neural tube lesion in a fetal lamb might protect neural tissue from subsequent harm and preserve neurologic capabilities after delivery. All lambs with the defect covered had minimal neurologic morbidity, but lambs with the defect uncovered had substantial neurologic morbidity. These findings show that long-term exposure of the open spinal cord to the intrauterine environment can cause neural tissue damage and, as a result, loss of neurologic capabilities, and that covering the defect can lead to a better neurologic outcome.[13]

Open spina bifida is a fetal abnormality that is not fatal. Over the last 75 years, significant progress has been achieved in the prevention, diagnosis, and treatment of open spina bifida. The most important therapy for preventing open spina bifida has been folic acid supplementation; however, more research into the intricate role that genetics and the environment play in metabolism is emerging.[12] Early intervention yields a significant reduction in complications. Healthcare awareness for even intrauterine interventions is encouraged. In the majority of developing countries, pregnant women have a drastically low level of antenatal care or several visits. Even awareness among mothers of folic acid supplementation and nutritional status is very poor. That is why the incidence of neural tube defects among these countries is very high the order of 7 – 8/1000 live in Pakistan as compared to 1 – 2/1000 live births overall worldwide. Patients on anti-metabolites or anti-epileptics also lack counseling about contraception or proper planning of conception. Also, in our countries, there is a lack of a multidisciplinary approach to prenatal diagnosis of spina bifida and almost no center provides the intrauterine solution to such a defect, which also counts as a hyped-up incidence of spina bifida. This is also the reason, why we lack a concept of early intervention i.e., in utero and patients mostly present with neuromuscular stigmata like significant power loss in lower limbs, club feet, bowel bladder incontinence gait disturbances, etc., and once an insult to neural tissue is done, it can't be reversed. Patients also present with hydrocephalus which is a complication of tethered cord and cerebellar tonsillar herniation as the vertebral column lengthens and tethered cord cannot ascend as it normally should. This can be decompressed and has promising results as far as intracranial pressure is concerned yet insult to neural tissue cannot be reversed.

Vora et al,[14] investigated the functional results and risk variables for outcomes following lipomyelomeningocele surgery (LMMC). Data from 109 children with LMMC who had surgery were evaluated retrospectively to assess functional results and potential risk factors for early- (at hospital discharge or within 1 month of surgery) and long-term outcomes following surgery. At long-term follow-up, radical excision of the LMMC is likely to help preserve neurological function in more than 90% of infants. Following surgery, 55% of children over the age of 2 years old with partial bladder impairment regained normal function. Early surgical surgery can correct neurological abnormalities in infants who develop symptoms after delivery. Iqbal et al,[15] reported a prospective cohort research to evaluate the early neurological outcome of surgery in newborns with lipomyelomeningocele. There was no significant difference between the research groups on day 3 and day 10 after surgery. In Group-A, early outcomes at 6-month intervals were shown to be related to significantly better neurological function grades. Lipomyelomeningocele surgery is a risk-free technique. Even in asymptomatic newborns, early therapy is suggested to avoid neurological degeneration. Bulsara et al,[16]

investigated if there is a clinical difference between individuals with lipomyelomeningocles, intraspinal lipomas, and filum terminale lipomas. The lipomyelomeningocele group showed the least improvement in these areas. Lipomyelomeningoceles patients ranged in age from 1 day to 18 years, with the majority being less than 2 years. Following operational intervention, all patients improved in motor strength after an average of 8 months of follow-up.

Traditionally, surgery has focused on the bladder and bladder neck, as well as the construction of catheterizable channels. Despite agreement on the importance of early urological intervention in the care of patients with spina bifida, there is still debate over optimal therapy. Major reconstructive urological procedures continue to play an important role in the treatment of these cases in order to protect the upper urinary system and establish continence. However, further research is needed to determine the real impact of these therapies on the quality of life of patients and their families. The transition of urological treatment to adulthood remains a key opportunity for disease management improvement.[17]

CONCLUSION

Surgical intervention in lipomeningocele and lipomeningomyelocele produces promising results if done timely. Post-op CSF leakage and incontinence are much better managed in the early intervention than in late intervention.There is a scarcity of research on the outcomes of patients with myelomeningocele and lipomeningocele. More study is required to better understand the variations in long-term urological results between these two diseases.

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Additional Information

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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:
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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.	Shahzeb Ahmad	Study Design, Methodology, and Paper Writing.
2.	Shakil Mashori	Data Calculation and Data Analysis.
3.	Syeda Kiran Zahra	Interpretation of Results.
4.	Rana Muhammad Usama	Statistical Analysis.
5.	Abdul Hanan	Literature Review.
6.	Shmama Tu Zahra	Literature Review and Quality Insurer.