



## Evaluation and management of lumbosacral myelomeningoceles in children-A tertiary care centre's experience

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### Abstract

**Objective:** Myelomeningoceles are the common form of open neural tube defects that are usually associated with neurological deficits. Many techniques of repair and methods of prevention have been proposed with respect to the size of defect and the neurological condition of patient. The aim of this study was to report our experience on the management of lumbosacral myelomeningoceles in children.

**Materials and Methods:** We retrospectively analysed the data of 36 paediatric cases of lumbosacral myelomeningocele treated in our institute between 2017 and 2019. Twenty (56%) patients were female and sixteen were male, with a mean age of 4 months (ranged between 0 and 24 months). All patients had neurological deficits in the preoperative period. Computed tomography was performed in 33 (92%) patients and magnetic resonance imaging in 15 (42%) patients in the preoperative period. Repair of the myelomeningocele and closure of the skin defect were performed in all patients. The mean follow-up period was 6 months.

**Results:** Thirty (83%) patients were operated for hydrocephalus and 10 (28%) patients were re-operated for tethered cord syndrome during the follow-up period. Neurological worsening was not observed in any patient while cerebrospinal fluid Fistula was detected in 2 patients.

**Conclusion:** Surgical treatment using appropriate microsurgical techniques is crucial for lumbosacral myelomeningoceles in children. Early surgical intervention with close follow-up will improve the neurological condition of the patients.

**Keywords:** myelomeningocele, surgery, child

### Introduction

Myelomeningocele is the most common congenital malformation of the central nervous system, with a prevalence of 4.4 to 4.6 cases per 10000 live births in the United States [1]. However, there is no study in our country on the prevalence of this malformation. The myelomeningoceles may occur in any part of the spine but it is predominately observed in the lumbosacral region, the last region of the neural tube to fuse, with thoracolumbar lesions being less typical [2].

Surgery is the only treatment method of this malformation and maximal closure of the skin and dura defect with optimal preservation of the neurological functions is the main goal of the surgery [3]. Although many different techniques are in use for the closure of these malformations, clinical outcomes of the treatment are not pleasing for the families.

We report the results of a retrospective study of 36 lumbosacral myelomeningocele patients, with clinical and radiological criteria. Surgical treatment was performed in all cases and the follow-up results were done.

### Materials and Methods

A retrospective review of 36 patients, who presented with lumbosacral myelomeningocele between 2017 and 2019 was performed. There were 20 (56%) boys and 16 girls of 2 days to 2 years old at the presentation with a mean follow

up of 6 months. These patients were evaluated preoperatively and postoperatively with radiology and followed for at least 12 months. The magnetic resonance imaging (MRI), computed tomography (CT), and lumbosacral x-rays were performed for selected patients either in preoperative and postoperative periods in order to detect possible associated lesions such as split cord malformation (SCM), lipoma or hydrocephalus. A complete neurological and physical examination of each patient was also noted in detail to evaluate the benefits of surgery. The surgical repair of myelomeningocele consisted of reconstruction of neural placode and the closure of healthy skin with meticulously dissection of meningeal sac without neural damage. All epithelized tissue from the edge of the neural placode was removed using microsurgical techniques. The hypertrichosis and/or reddish-coloured skin tissue were also excised to obtain and unify the edges of normal skin. The dura was found and closed in a watertight fashion in the midline covering all neural tissues in normal appearance. The large skin defects were closed using transposition of skin flaps in a condition of adequate blood supply and all patients received prophylactic antibiotic cover in perioperative period. The infants are nursed prone in the early postoperative period for at least 48 hours. The postoperative complications such as cerebrospinal fluid (CSF) leakage and infections were recorded and the relevant treatment was performed.

## Results

The patients ranged in age from 2 days to 2 years old with a mean age of 4 months (Figure). Paraparesis was the most common presenting symptoms in 27 (75%) patients. Others were paraplegia in 6 (17%), ankle weakness in 3 (8%) and foot deformity in 7 (19%) patients. Therefore, neurological deficits were observed in all patients.

Preoperative CT was obtained in 33 patients (92%) and MRI in 15 (42%) patients. SCM was detected in 3 (8%) patients above the level of myelomeningocele. Intraspinous lipoma was also detected in 2 patients. These patients were operated subsequently for such lesions and discharged without an additional neurological deficit. Hydrocephalus was the most commonly associated lesion and detected in 30 (83%) of the 36 patients. These patients underwent ventriculoperitoneal shunt insertion during the postoperative period. Tethered cord syndrome developed in 10 patients in the follow-up and releasing of the spinal cord was performed in these patients.

All patients were operated in 1 to 10 days (mean 4.7 days) after the admission. Two patients (6%) with large skin defect (more than a diameter of 120 mm) were operated in 2 steps that the first was the expander application and the second was skin closure.

The CSF collection and CSF leak were the main complications that prolonged the hospital stay of the patient. CSF collection was detected in 6 patients and treated with conservative methods. CSF leak was observed in 2 patients and treated with re-operation.

All patients were followed-up by spinal MRI and cranial CT with 3 month-intervals and 2 (6%) of them died in the follow-up period due to pneumonia and shunt failure.



**Fig 1:** Showing Lumbar MMC in two day old neonate.

## Discussion

We reported the surgical results of 36 paediatric patients who had lumbosacral myelomeningocele in a period of 2 years. Most of them had hydrocephalus as associated clinical entity. Repair of the myelomeningocele, including reconstruction of neural placode, excision of non-functional neural structures and fibrous bands and dural closure, was performed in all patients. No neurological worsening was observed in any patient. Two patients died during the follow-up period.

Congenital posterior fusion defects of neural tube are a relatively uncommon development anomaly affecting approximately 1 in 850 live births. The more common variant of them is spina bifida aperta in which the posterior fusion failure results in the exposure of neural tissue with or without a covering meningeal sac. These myelomeningoceles are usually presented with neurological

deficits associated with spinal cord and nerve root exposure and they affect motor and sensory functions, and bladder and bowel functions [2, 7]. Although these lesions are not commonly lethal, Steinbok et al. [8] reported an 8.6 years survival of 82% in 101 children born with myelomeningocele between 1971 and 1981. Hypothalamic-pituitary dysfunction was also observed in children with myelomeningocele and treatment of these patients with growth hormone improves significantly the growth rate [9, 10]. In our series, neurological deficits were observed in all patients, but we did not analyse pituitary hormone profiles of 36 patients.

Many procedures have been described for the repair of myelomeningocele defects. Primary repair is essential only for the small skin defects. Wide undermining with primary closure after skin advancement for large defects is associated with tension, stretching of the resultant inelastic scar tissue, and inappropriate soft tissue cover. The skin flaps that are necessary for a tension-free closure can be classified as cutaneous and myocutaneous flaps. Close cooperation with plastic surgery team has allowed optimal surgical repair and minimal neurological deficit [2, 11-13]. Endoscopic coverage of foetal myelomeningocele in utero is recently and widely performed by many surgeons but its benefits on neurological recovery has not been understood yet [14, 15]. Amniotic fluid exchange was also previously reported by Olguner et al. [16] and they concluded that the exposure of myelomeningocele to amniotic fluid causes structural neural tissue damage that can be prevented by amniotic fluid exchange. This is an experimental study performed in chick eggs and not proven in humans [16]. In our series, we performed repair of myelomeningocele by reconstructing the neural placode and closing the dura in a watertight fashion. The skin defect was closed by skin flaps in 2 cases with the help of plastic surgeon.

Hydrocephalus is presented in 90% of the cases of myelomeningocele [2]. Patients with hydrocephalus associated with myelomeningocele are difficult to manage. Most patients born with myelomeningocele will need shunting. Significant controversy exists about how to select patients with stabilised ventriculomegaly who can benefit from a shunt. It is difficult to differentiate a compensated/arrested hydrocephalus and a progressive hydrocephalus when faced with one of them. If hydrocephalus is moderately severe at birth or if a delayed closure of the spinal defect is planned, a shunt system may be inserted at the same time as or prior to the myelomeningocele closure [17, 19]. In our series, we inserted ventriculoperitoneal shunt system in 30 patients (83%) having hydrocephalus on admission or detected in the postoperative period. We did not observe hydrocephalus in 6 patients during the follow-up period. The rate of hydrocephalus in our series is less than the literature. In 11 of those 30 patients, shunt malfunction occurred several times and shunt revision was performed.

In almost every instance, the children with a myelomeningocele will have some form of urinary sphincter disturbance [3]. In myelomeningocele, it is possible that the children have normal urinary function at birth, but when a tethering lesion exists on spinal cord, bladder dysfunction occurs. With rapid growth during the childhood, tethering of the cord would become obvious and the bladder function deteriorates [20]. This fact supports the importance of early neurosurgical repair in such cases. However, the reliable determination of bladder function is often not possible for

months after the operation, even though early urological assessment and urodynamic testing are completed. In our series, we did not perform urodynamic studies in preoperative period but these tests were performed in 10 patients who underwent further surgery for tethered cord syndrome. These patients had urinary disturbances in the follow-up of myelomeningocele operations.

Split cord malformations may occur in association with myelomeningocele and these are often ignored or misdiagnosed potentially causing spinal cord tethering [21]. Iskandar et al. [22] reported a ratio of 6% in their series of myelomeningocele for this association. In our series, 3 patients (8%) had split cord malformation in association with myelomeningocele. Septums of these patients were removed subsequently and the patients were discharged without an additional neurological deficit.

The electrophysiological assessment of the patients with myelomeningocele is usually neglected by neurosurgeons. SSEPs measurement, using standard surface stimulating electrodes overlying the tibial nerves is necessary in patients with neural tube defects in lumbosacral region for the comparison of neurological status of the patients either preoperatively or postoperatively. In addition, intraoperative electrophysiological monitoring is essential for a safe surgery.

The postoperative complications of myelomeningocele are discussed in detail by Pang [23] with respect to predisposing factors, diagnosis, treatment and prevention. These complications include worsened neurological level, wound dehiscence, wound infection, CSF leak, postoperative ileus, symptomatic Chiari malformation, shunt infection, necrotizing enterocolitis and problems related to kyphectomy [8, 23, 24]. In our series, CSF fistula occurred in 2 patients, and 2 patients who underwent surgery for myelomeningocele and hydrocephalus died due to pneumonia and shunt failure.

### Conclusion

Myelomeningocele is the most severe form of spina bifida in which the surgical repair is a relative neonatal emergency. The main goal of the surgery must be the preservation of all functional neural tissue and reconstruction of normal anatomic barriers with tension-free, stable and well-vascularized skin flaps. In preoperative period a complete radiological, neurophysiological and urological assessment should be performed regardless of the patient's age and neurological status. With ongoing studies, we hope to predict better results in myelomeningocele and develop new techniques to solve this problem.

### Conflict of Interest

No conflict of interest was declared by the authors

### References

- Lary JM, Edmonds LD. Prevalance of spina bifida at birth-United States, 1983-1990: a comparison of two surveillance systems. *MMWR Morb Mortal Wkly Rep* 1996; 45:15-26.
- Lanigan MW. Surgical repair of myelomeningocele. *Ann Plast Surg*. 1993; 31:514-21.
- Humpreys RP, Spinal Dysraphism. In Wilkins RH, Rengachary SS (eds): *Neurosurgery*. New York: Mc Graw-Hill, 1996, 3453-63.
- Black PM. Selective treatment of infants with myelomeningocele. *Neurosurgery*. 1979; 5:334-8.
- McLone DG. Treatment of myelomeningocele. Arguments against selection. *Clin Neurosurg*. 1986; 33:359-70.
- Sutton LN, Charney EB, Bruce DA, Schut L. Myelomeningocele. The question of selection. *Clin Neurosurg*. 1986; 33:371-81.
- Noetzel MJ. Myelomeningocele: current concepts in management. *Clin Perinatol*. 1989; 16:311-29.
- Steinbok P, Irvine B, Cochrane DD, Irwin BJ. Long-term outcome and complications of children born with myelomeningocele. *Childs Nerv Syst*. 1992; 8:92-6.
- Perrone L, Del Gaizo D, D' Angelo E, Rea L, Di Manso G, Del Gado R. Endocrine studies in children with myelomeningocele. *J Pediatr Endocrinol*. 1994; 7:219-23.
- Rotenstein D, Reigel DH. Growth hormone treatment of children with neural tube defects: Results from 6 months to 6 years. *J Pediatr*. 1996; 128:184-9.
- de Chalain TM, Cohen SR, Burstein FD, Hudgins RJ, Boydston WR, O'Brien MS. Decision making in primary surgical repair of myelomeningoceles. *Ann Plast Surg*. 1995; 35:272-8.
- Lorber J. Results of treatment of myelomeningocele. An analysis of 524 unselected cases, with special reference to possible selection for treatment. *Dev Med Child Neurol*. 1971; 18:279-303.
- Mc Lone DG. Technique for closure of myelomeningocele. *Childs Brain*. 1980; 6: 65-73.
- Bruner JP, Richards WO, Tulipan NB, Arney TL. Endoscopic coverage of fetal myelomeningocele in utero. *Am J Obstet Gynecol*. 1999; 180:153-8.
- Copeland ML, Bruner JP, Richards WO, Sundell HW, Tulipan NB. A model for in utero endoscopic treatment of myelomeningocele. *Neurosurgery*. 1993; 33:542-4.
- Olguner M, Akgür FM, Özdemir T, Aktuğ T, Özer E. Amniotic fluid exchange for the prevention of neural tissue damage in myelomeningocele: An alternative minimally invasive method to open in utero surgery. *Pediatr Neurosurg*. 2000; 33:252-6.
- Parent AD, McMillan T. Contemporaneous shunting with repair of myelomeningocele. *Pediatr Neurosurg*. 1995; 22:132-5.
- Iborra J, Pages E, Cuxart A, Poca A, Sahuquillo J. Increased intracranial pressure in myelomeningocele patients never shunted: results of a prospective preliminary study. *Spinal cord*. 2000; 38:495-7.
- Hammock MK, Milhorat TH, Baron IS. Normal pressure hydrocephalus in patients with myelomeningocele. *Dev Med Child Neurol*. 1976; 37(suppl):55-68.
- Wu HY, Kogan BA, Baskin LS, Edwards MS. Long-term benefits of early neurosurgery for lipomyelomeningocele. *J Urol*. 1998; 160:511-4.
- Kural C, Solmaz I, Pusat S et al. Surgical technique for split cord malformations: Pitfalls and solution pathways. *Gulhane Med J*. 2013; 55:77-83.
- Iskandar BJ, Mc Laughlin C, Oakes WJ. Split cord malformations in myelomeningocele patients. *Br J Neurosurg*. 2000; 14:200-3.
- Pang D. Surgical complications of open spinal dysraphism. *Neurosurg Clin N Am*. 1995; 6:243-57.
- Akalan N. Myelomeningocele (open spina bifida)-surgical management. *Adv Tech Stand Neurosurg*. 2011; 37:113-41.