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A study of clinical profile and surgical outcome of spinal dysraphism in paediatric population an institutional study

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Abstract

Aims: To study the surgical outcome in paediatric patients presenting with spinal dysraphism in our institution.
Materials and Method: 78 paediatric cases presented between 2014 to 2019 with myelomeningocele to the neurosurgical OPD in Vijayanagara Institute of Medical Sciences, Ballari which caters to mainly rural and semi urban population. Initial clinical assessment, followed by operative work up was done. MRI Brain and spine was done as a prep operative evaluation. Post-operative outcome was noted and Post-operative regular follow up after discharge for at least 6 months.
Results: 78 patients were operated upon at our centre. The mean age of presentation of the cases 5 months +/- 6 days, ranging from 8days to 24months. Post-operative average hospital stay was 14 days. The mean post-operative follow up period was 36 months. Hydrocephalus was associated with meningomyelocele in 30 cases, but VP shunting required in same sitting in 9 cases only. Six patients found to have CSF leak and only 1 required reexploration and Dural repair.
Conclusion: The clinical profile of patients presenting to us mirror the General profile across India. Surgical outcomes at our center are on par with the standard results seen across the country. Early and prompt surgical intervention is the key in management of spinal dysraphism. The MOMS study comparing surgical outcomes with pre natal surgery shows equivalent outcomes with post natal surgery.

Keywords: Paediatric patients, spinal dysraphism, myelomeningocele

Introduction

Myelomeningocele is a congenital defect in vertebral arches with cystic dilatation of meninges and structural or functional abnormality of spinal cord or cauda equina [1].) Myelomeningocele is the commonest among the spinal dysraphism and the most common congenital malformation of the central nervous system. Incidence of spina bifida with myelomeningocele (MM) is 1-2/1000 live births (0.1-0.2%) worldwide and varies from 0.3 to 14 live births in India. Risk increases to 2-3% if there is one previous birth with MM and 6-8% after two affected children [1].

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MMC patients face long time residual disabilities like paralysis of the lower limbs; sensory loss; impaired cognition; and bladder & bowel incontinence, and sexual dysfunction [2].
Diagnosis of myelomeningocele is usually done at around 20wks of life during a routine anamoly scan and large numbers of parents opt for medical termination of pregnancy post diagnosis.
Cause is multifactorial, including periconceptional folic acid and Vitamin B12 deficiency, genetic factors, hyperthermia and intake of valproic acid. Periconceptional folic acid supplementation has reduced the incidence of myelomeningocele [3].
Management of MMC is complex, and early intervention is necessary. Usually, post-delivery the MMC defect will be closed surgically to prevent further damage to the spinal cord and neuro infection. MMC patients tend to develop other complications associated with the primary disease. A highly prevalent one is the development of hydrocephalus [4].
Hydrocephalus (HCP) develops in 65–85% of patients with MM, and 5–10% of MM patients have clinically overt HCP at birth [5]. Over 80% of MM patients who will develop HCP do so before age 6 months. Diversions like is shunting or by creating an endoscopic third ventriculostomy (ETV) can be considered.
Chiari malformation type II is another condition associated with myelomeningocele [6, 7, 8, 9]. Most patients of MM have chiari malformation type II on imaging but hardly few are symptomatic. Clinical features includes muscle weakness, apnea, neck pain, stridor and dysphagia [10].

Materials and Methods

This is a prospective study with analysis of all the cases presented to neurosurgery OPD 2014 to 2019 with myelomeningocele in Vijayanagar Institute of Medical Sciences, Bellary. Initial clinical assessment, followed by operative work-up was done. MRI Brain was done to assess the type, extent and position of the lesion. Patient posted for surgery following anaesthetic and paediatric clearance, high risk informed consent and ventilatory back-up. Post-operative intensive care management done, with regular follow up after discharge for at least 6 months.

Pre operatively following was done.

- Measure size of defect
- If ruptured antibiotics were started (Inj. Ceftriaxone was administered in our institute).
- In unruptured cases first dose of antibiotic was administered at the time of skin incision.
- Exposed lesions were covered with sponges soaked in lactated ringers or normal saline to prevent desiccation.
- Lateral position and prone position for the patient- was instructed to patient attenders(avoids pressure over the lesion)
- Neurological assessment: to look for any evidence of motor deficit in lower extremities by checking response of Lower extremities to painful stimulus.
- Neonatologist assessment for other abnormalities
- Urinary catheterization
- AP & lateral spine films to assess baseline scoliosis.

Intra operative-standard surgical approach was followed by the same team of operating surgeons keeping following basic principles

- Free placode from Dura (to avoid tethering)
- Water-tight Dural closure
- Skin closure (can be accomplished in essentially all cases)

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Post-operative assessment included

- Patient was kept off all incisions
- Bladder catheterization regimen was followed
- Daily head circumference measurement
- In patients whom shunting was not done, regular head U/S was done(weekly)
- Patient was made to lie in lateral position in order to reduce CSF pressure on incision site.

Inclusion criteria

All the myelomeningocele cases presenting neurosurgery OPD will be included.

Exclusion criteria

Patients who were already in severe sepsis.

Results

Out of 78 patients were operated at our center, the mean age of presentation of cases was 5months+/- 6 days. The average post-operative stay was for 14 days and mean post-operative follow up period was 36 months.

Out of 78 cases which were included in this study 45 of them were male (57.7%) and 33 of them were females (42.3).

Table 1: Sex distribution

Sex	No. of patients	Percentage
Male	45	57.7
Female	33	42.3

Out of 78 patients in the study population, 19 were neonates (24.5), 42 of them were infants (53.8), 15 patients were between the age of 1-5yrs (19.2) and 2 patients were more than 5yrs (2.5).

Table 2: Age Distribution

Age Distribution	No. of patients	Percentage
Neonates	19	24.5
Infants	42	53.8
1 – 5 years	15	19.2
> 5 years	2	2.5

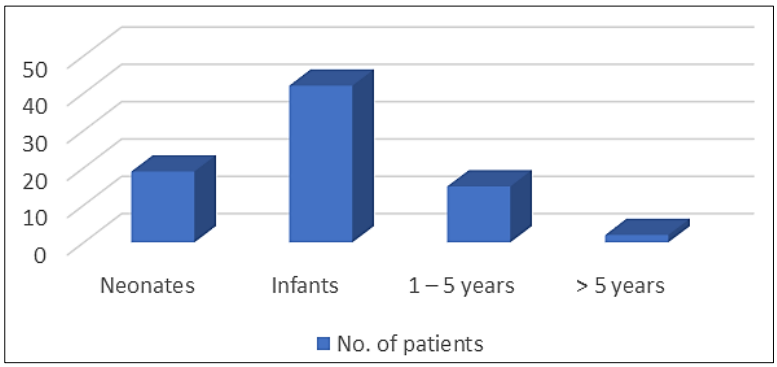

Fig 1: Age Distribution

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Out of 78 patients, 76 were cases of myelomeningocele (97.5) and 2 were

Lipomyelomeningocele (2.5%) (A subcutaneous lipoma that passes through a midline defect in the lumbodorsal fascia, vertebral neural arch, and Dura, and merges with an abnormally low tethered cord. [10]

Table 3: Type of spinal dysraphism

Type of Spinal Dysraphism	No. of patients	Percentage
Meningomyelocele	76	97.5
Lipomyelomeningocele	2	2.5

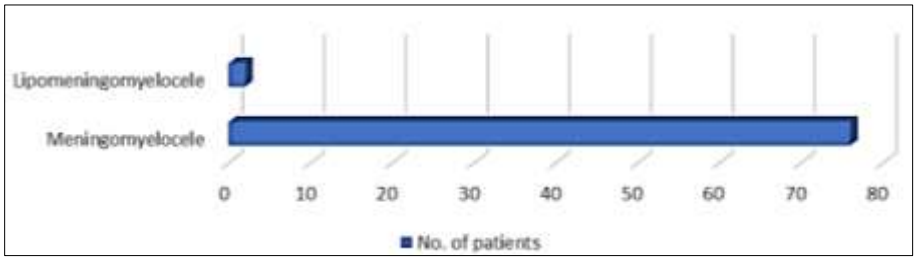

Fig 2: Type of Spinal Dysraphism

Out of 78 patients, in 64 cases myelomeningocele was present in the lumbosacral region(64%), 12 cases myelomeningocele was present in thoracolumbar region(15.4%) and in 2 cases myelomeningocele was present in cervical region(2.5%).

Table 4: Location of myelomeningocele

Location	No. of patients	Percentage
Cervical	2	2.5
Thoracolumbar	12	15.4
Lumbosacral	64	82.1

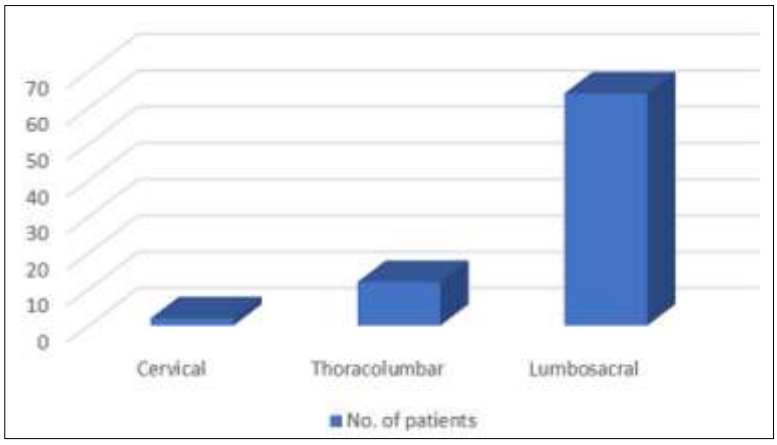

Fig 3: Location of Myelomeningocele

Out of 78 patients, 16 patients had paraparesis (20.5%) at the time of presentation to the neurosurgery OPD and no paraparesis was noted in rest 62 patients (79.5%).

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Table 5: Number of patients with paraparesis

Paraparesis	No. of patients	Percentage
Present	16	20.5
Absent	62	79.5

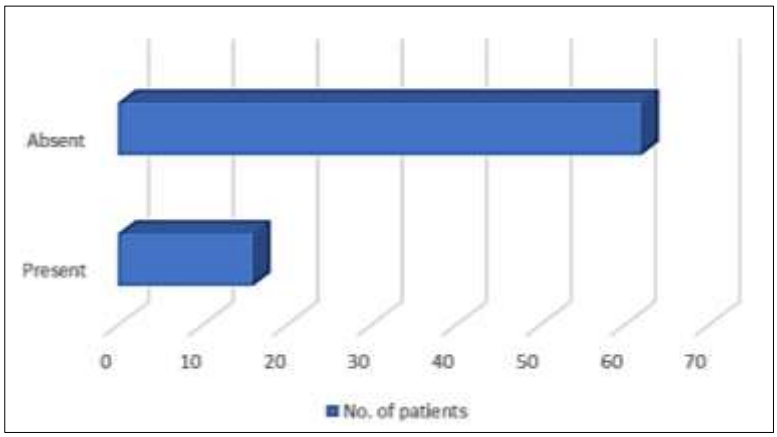

Fig 4: Paraparesis

Out of 78 patients, ruptured MMC was notes in 23 cases (29.5%) and unruptured MMC was seen in 55 cases (70.5)

Table 6: Number of patients with CSF leak

CSF leak	No. of patients	Percentage
Ruptured	23	29.5
Unruptured	55	70.5

Hydrocephalus was noted in 30 out of 78 cases (38.5%) and was absent in 48 cases (61.5%). Out of the 30 cases, 11 cases required VP shunting in the same sitting.

Table 7: Incidence of hydrocele

Hydrocephalus	No. of patients	Percentage
Present	30	38.5
Absent	48	61.5

There were multiple associated abnormalities with myelomeningocele. Out of 78 patients, 7 patients had associated cardiac anomalies (9.0%), 1 patient had ano rectal anamoly (1.3%), 3 patients had associated cutaneous anomalies (3.8%), urologic anomalies were noted in 11 patients (14.1%), in 53 patients no abnormalities were noted (67.9%).

Table 8: Associated abnormalities

Associated Anomalies	No. of patients	Percentage
Cardiac	7	9.0
Ano-rectal	1	1.3
Cutaneous	3	3.8
Urologic	11	14.1
None	53	67.9

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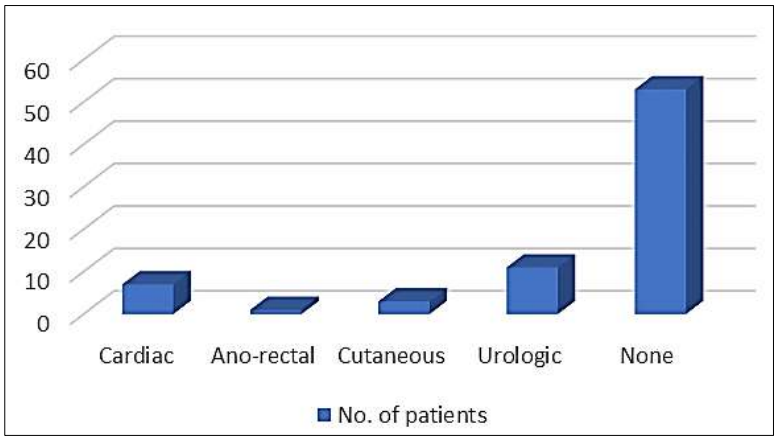

Fig 5: Associated Abnormalities

Surgical outcome

All 78 patients underwent anatomical repair by the same surgical team and no neurological worsening was noted in most of the cases in the early follow up period.
6 patients out of 78 patients (7.7%) had CSF leak in the post-operative period, of which 5 were treated conservatively, while 1 patient required re-operation for dural closure.
5 patients had wound infection (6.4%), skin necrosis was notes in 2 cases(2.5%), ventriculitis was noted in 3 cases (3.8%), new onset paraparesis was notes in 4 cases(5%) and death occurred in 2 patients (2.5%). One death occurred due to respiratory failure associated with congenital cardiac defects and the other due to septic shock.

Table 9: Various post-operative complications

Complications	No. of patients	Percentage
CSF leak	6	7.7
Wound infection	5	6.4
Skin necrosis	2	2.5
Ventriculitis	3	3.8
Paraparesis	4	5
Death	2	2.5

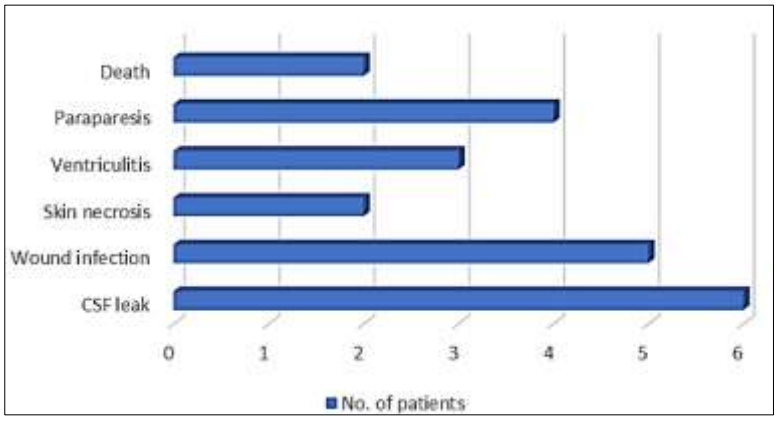

Fig 6: Various Surgical Complications

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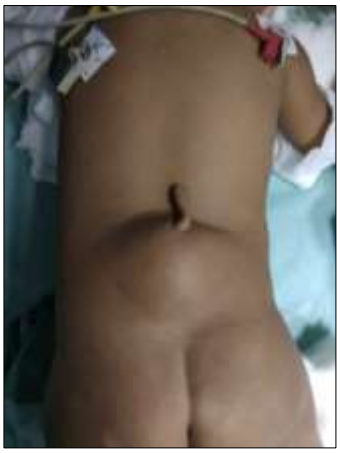

Fig 7: Case of lumbar lipomeningocele

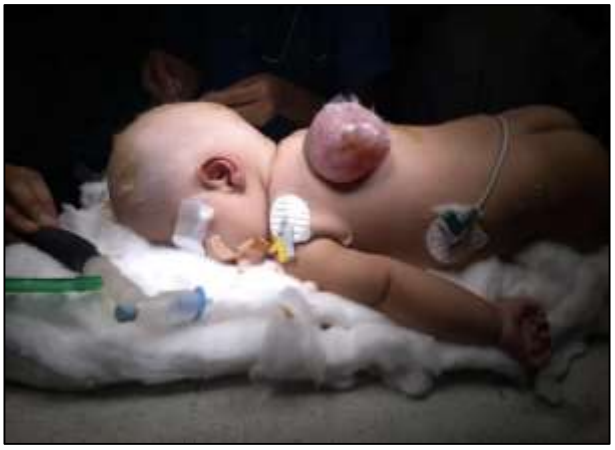

Fig 8: Case of dorsal meningomyelocoele

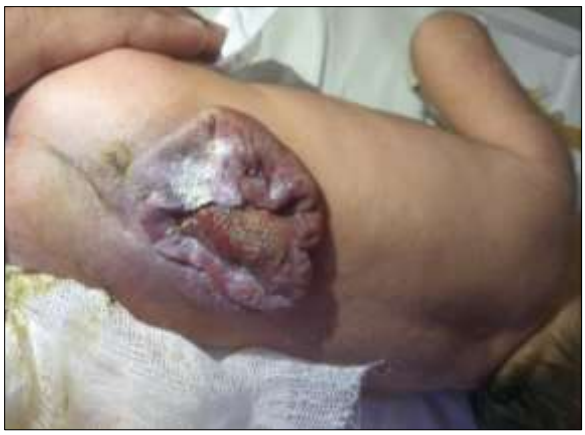

Fig 9: Case of ruptured meningomyelocoele

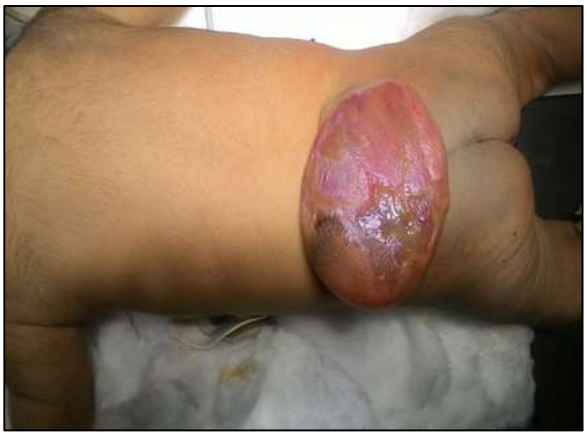

Fig 10: Case of unruptured meningomyelocoele

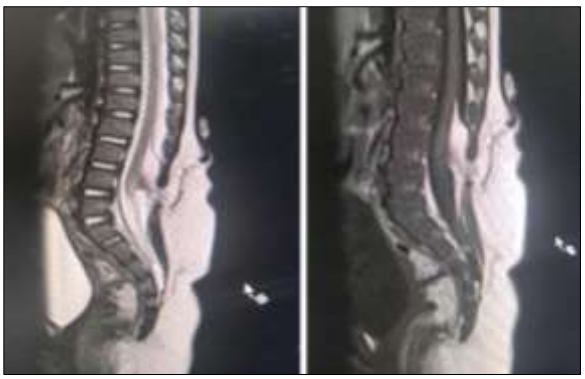

Fig 11

MRI images showing Meningomyelocele- herniation of neural elements in to Subcutaneous space intra operative pictures demonstrating dissection and separation of neural elements from sac.

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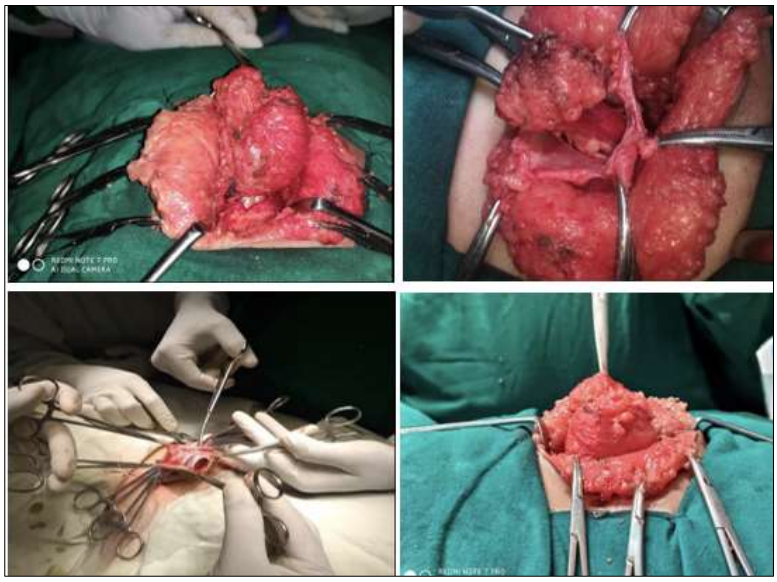

Fig 12

Discussion

We at the "Department of Neuro Surgery, Vijayanagara Institute of Medical sciences, Ballari" have made a sincere attempt to report the clinical profile and surgical outcomes among paediatric population with myelomeningocele presenting to a tertiary care semi urban centre.

Here this study also has been compared with other studies done previously. A brief outline of the studies compared is given below.

Beuriat *et al*. Conducted this study in 2017, 46 cases were included in the study out of which 27 were sacral myelomeningocele, 5 lumbar myelomeningocele and 14 were thoracic myelomeningocele. In the post of period 36 patients had orthopaedic complications and 41 patients had urological complications. 7 out of 46 cases required re- surgery.

Kumar *et al*. conducted this study in 2007 which included 102 cases of myelomeningocele. Hydrocephalus was noted in 58.8% of the cases included in the study.

Sharma *et al*. conducted the study in 2019 comprising of 11 cases, out of which 8 cases were sacral myelomeningocele; 2 lumbar myelomeningocele and 1 was thoracic myelomeningocele. Wound infection was noted in 1 case and wound dehiscence was noted in 2 out of 11 cases.

Nejat *et al*. conducted the study in 2011 comprising of 40 cases, out of which 21 were male and 19 were female. Wound complications was noted in 3 out of 40 cases.

Table 10: Comparison with other studies

Study	No. of cases	S/L/T/C	M/F	Complications	Re-surgery	Hydrocephalus
Beuriat *et al*. (2017)	46	27/5/14/0	-	Ortho-36 Uro-41	7	-
Kumar *et al*. (2007)	102	-	-	-	0	58.8%
Sharma *et al*. (2019)	11	8/2/1	-	Wound infection-1 Wound dehiscence-2	0	-
Nejat *et al*. (2011)	40	-	21/19	Wound infection-3	0	-

In the current study 78 patients underwent anatomical repair by the same surgical team and no neurological worsening was noted in most of the cases in the early follow up period.6 patients out of 78 patients (7.7%) had CSF leak in the post-operative period, of which 5 were

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Conclusion

This study provides an overview of current outcomes for infants born with an MMC managed at our center. The overall outcomes are similar to those reported in the literature. The clinical profile of patients presenting to us mirror the General profile across India. Surgical outcomes at our center, despite being a semi urban government setup, are on par with the standard results seen across the country. The MOMS study comparing surgical outcomes with pre natal surgery shows equivalent outcomes with post natal surgery.

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