### **Original Research Article**

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# Follow up of meningomyelocele and associated hydrocephalus, bladder and/or bowel incontinence and foot deformity in a developing country

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#### **ABSTRACT**

**Background:** Meningomyelocele (MMC) and meningocele (MC) are the two most common types of neural tube defects (NTD) seen. Some of the problems associated with care of these patients are psychological stress is parents, average financial condition of caretakers, ignorance on the part of parents, local physicians for management of this condition, very few dedicated Spina bifida clinics and social stigma associated with this condition. The aim of treatment of meningomyelocele is to the patients with maximum mobility and social continence which is possible through multidisciplinary care involving paediatric surgeon, paediatrician, nursing staff, occupational therapist, physiotherapist, social worker, parents and patients themselves. The aim of this study is to discuss our experience in initial management of meningomyelocele (MMC) and hydrocephalus and long-term management of bladder/bowel incontinence and foot deformity in a tertiary care hospital of a developing country.

**Methods:** A retrospective nonrandomized observational study was conducted in the department of pediatric surgery at a tertiary hospital, from August 2008 to October 2015. Overall 145 patients of meningomyelocele were included in the study. Patients were managed according to our institution protocol.

Results: 68.2% MMC patients were repaired primarily. 24.1% of total patients needed Y-V plasty skin advancement flap. 7.5% patients needed synthetic patch for dural closure. 24.1% patients were incontinent since birth. 19.3% patients had decreased lower limb power since birth. 5.5% patients were incontinent after MMC repair. 8.2% patients had decreased lower limb power after MMC repair. 28.9% patients needed VP shunt for gross hydrocephalus before MMC repair. 51.03% patients needed VP shunt after MMC repair. 20% patients did not require VP shunt in the present series. 13.1% patients in the present series had urinary incontinence. 11.03% patients had fecal incontinence. 73.6% patients with urinary incontinence were successfully managed with conservative treatment. 87.5% patients with fecal incontinence responded to conservative treatment. Bladder augmentation was done in 26.3% of neurogenic bladder dysfunction patients. 60% of these patients had dry interval of more than 4 hours. MACE was done in 12.5% of neurogenic bowel dysfunction patients. 2 patients required simultaneous bladder augmentation, Mitrofanoff's and MACE procedure. One of these patients achieved a dry interval of >4 hour.

**Conclusions:** Successful rehabilitation of children with MMC can be achieved with parental education and support, dedicated clinicians, trained nursing staff, regular follow-up and low threshold for diagnosing clinical deterioration and proactive management to prevent further clinical deterioration.

Keywords: Continence, Long term outcomes, MMC, Spina bifida, Social acceptance

#### INTRODUCTION

Meningomyelocele (MMC) and meningocele (MC) are the two most common types of neural tube defects (NTD) seen. The incidence of spina bifida is estimated at one to two cases per 1000 population.<sup>1</sup>

Exact etiology and specific genes responsible for the generation of NTDs such as MMC have not yet been identified. Multidisciplinary team approach right from the beginning can achieve near-normal bodily functions and purposeful life.

In a developing country like India, treatment of MMC with associated malformations needs following issues to be taken care of:

- Psychological stress in parents, average financial condition of caretakers, ignorance on the part of parents, local physicians for management of this condition
- Very few dedicated Spina bifida clinics in our country offer holistic management of this condition.
  There is lack of knowledge among parents that such facilities exist and unreliable compliance of parents to follow-up over long term at such clinics is a problem
- Social stigma associated with this condition

We are running a multidisciplinary Spina Bifida clinic since 2000 and would like to share our experience of MMC management at a tertiary care center in a developing country.

#### **METHODS**

A retrospective nonrandomized observational study was conducted in the Department of Pediatric Surgery at a tertiary hospital, from August 2008 to October 2015. Overall 145 patients of meningomyelocele were included in the study. Patients were managed according to our institution protocol.

#### Protocol of our Spina bifida clinic

- Pre-operative ultrasound (USG) of skull, spine, kidney, ureter, bladder (KUB), muscle power charting of lower limbs and fundoscopy.
- Elective correction of meningomyelocele, hydrocephalus as the day's first case in the operation theater. Patients with non-leaking MMC and hydrocephalus undergo ventriculoperitoneal shunt (VP shunt) procedure 7 days prior to MMC repair whereas those with leaking MMC and hydrocephalus are operated for both conditions at the same time. In cases of infected MMC all procedures are delayed till control of infection is achieved with antibiotics and sterile dressings.

- Pre-operative broad-spectrum antibiotic, 2 doses and post-op antibiotic course for 5 days in all cases. It is extended in case of wound infection, CSF leak.
- Lower limb muscle power charting and USG skull repeated on post-operative day 7 in all cases.
- Patients with post-operative hydrocephalus are posted for right sided ventriculoperitoneal shunt.
- Follow-up evaluation for change in head circumference, bladder and bowel habits, lower limb power and sensations.
- Patients with neurogenic bladder and bowel dysfunction (NBBD) are started on clean intermittent catheterization and bowel washes at first follow-up along with suture removal.
- Patients with NBBD are screened with 3 monthly urine routine examination, serum blood urea nitrogen, creatinine, USG KUB till 3 years of age.
- Patients with hydroureteronephrosis on USG are evaluated by Voiding cystourethrogram. Patients showing vesicoureteric reflux are started on prophylactic antibiotics.
- The mother is taught lower limb message, lower limb physiotherapy, clean intermittent catheterization, rectal wash if indicated at time of first follow-up.
- At age around 3 years patients with NBBD are evaluated for response to medical management; urodynamic study parameters and need for surgical intervention is assessed in each individual patient.
- Patients with lower limb deformity are treated in collaboration with physiotherapist, occupational therapist and paediatric orthopedic surgeons.

#### **RESULTS**

68.2% MMC patients were repaired primarily. 24.1% of total patients needed Y-V plasty skin advancement flap. 7.5% patients needed synthetic patch for dural closure. 11.7% patients had wound infection. 4.8% patients had flap necrosis. CSF leak occurred in 5.5% patients (Table 1).

24.1% patients were incontinent since birth. 19.3% patients had decreased lower limb power since birth. 5.5% patients were incontinent after MMC repair. 8.2% patients had decreased lower limb power after MMC repair (Table 2).

28.9% patients needed VP shunt for gross hydrocephalus before MMC repair. 51.03% patients needed VP shunt after MMC repair. 20% patients did not require VP shunt in our series (Table 3).

26.7% patients had VP shunt tract infection. Shunt block occurred in 11.2% patients. 4.3% patients developed intra-abdominal CSF pseudocyst. 6% patients had shunt extrusion. Shunt varix developed in 11.2% patients. Shunt revision was needed in 27.5% patients (Table 4).

Table 1: MMC repair methods and complications.

Type of MMC	Primary MMC repair	V-Y flap	Use of synthetic patch	Wound infection	Flap necrosis	CSF leak
Cervical (7)	7	0	0	1	0	0
Thoracolumbar (20)	15	4	1	2	0	1
Lumbar (34)	24	8	2	3	1	2
Lumbosacral (75)	48	20	7	9	4	4
Sacral (7)	4	2	1	1	1	1
Multiple defects (2)	1	1	0	1	1	0
Total (145)	99 (68.2%)	35 (24.1%)	11 (7.5%)	17 (11.7%)	7 (4.8%)	8 (5.5%)

Table 2: Status of continence and lower limb power before and after MMC repair.

Type of MMC	No. of patients incontinent since birth	No. of patients having lower limb power <grade 3="" birth<="" since="" th=""><th>No. of patients incontinent after MMC repair</th><th>No. of patients with decreased lower limb power after MMC repair</th></grade>	No. of patients incontinent after MMC repair	No. of patients with decreased lower limb power after MMC repair
Cervical (7)	0	0	0	0
Thoracolumbar (20)	2	3	1	2
Lumbar (34)	5	7	2	3
Lumbosacral (75)	11	13	4	4
Sacral (7)	6	4	1	2
Multiple defects (2)	2	1	0	1
Total (145)	26 (24.1%)	28 (19.3%)	8 (5.5%)	12 (8.2%)

Table 3: MMC and hydrocephalus.

Type of MMC	VP shunt before MMC repair	VP shunt after MMC repair	Patients not requiring VP shunt
Cervical (7)	0	4	3
Thoracolumbar (20)	7	9	4
Lumbar (34)	13	15	6
Lumbosacral (75)	18	42	15
Sacral (7)	3	3	1
Multiple defects (2)	1	1	0
Total (145)	42 (28.9%)	74 (51.03%)	29 (20.0%)

**Table 4: Shunt complications.** 

Shunt operated patients	VP shunt infection	Shunt block	CSF pseudocyst	Shunt extrusion	CSF varix	Shunt revision	Mortality
Cervical (4)	1	0	0	0	0	1	0
Thoracolumbar (16)	4	2	1	1	2	3	2
Lumbar (28)	5	2	1	2	3	7	2
Lumbosacral (60)	10	7	3	3	7	19	5
Sacral (6)	2	1	0	1	1	2	0
Multiple defects (2)	1	0	0	0	0	0	0
Total (116)	23 (26.7%)	12 (11.2%)	5 (4.3%)	7 (6.0%)	13 (11.2%)	25 (27.5%)	9 (7.7%)

13.1% patients in our series had urinary incontinence. 11.03% patients had fecal incontinence. 73.6% patients with urinary incontinence were successfully managed with conservative treatment. 87.5% patients with fecal incontinence responded to conservative treatment (Table 5). Bladder augmentation was done in 26.3% of

neurogenic bladder dysfunction patients. 60% of these patients had dry interval of more than 4 hours.

MACE was done in 12.5% of neurogenic bowel dysfunction patients. 2 patients required simultaneous bladder augmentation, Mitrofanoff's and MACE

procedure. One of these patients achieved dry interval of >4 hours (Table 6).

Table 5: Result of conservative management of incontinence.

No. of incontinent patients (U/F)	Urinary incontinence	Fecal incontinence
Cervical (0)	0	0
Thoracolumbar (0)	0	0
Lumbar (1/0)	1	0
Lumbosacral (12/11)	9	10
Sacral (5/4)	3	3
Multiple defects (1/1)	1	1
Total	14	14

Table 6: Result of operative intervention for bladder/bowel incontinence.

No of patients with dry interval	Bladder augmentation with/without Mitrofanoff	MACE	Bladder augmentation +Mitrofanoff +MACE
< 4 hours	2	0	1
>4 hours	3	2	1

#### **DISCUSSION**

Meningomyelocele occurs due to a defect in the embryonal formation of the neural tube. The cauda equina roots or conus medullaris contained in the sac are severely dysfunctional. As a rule, the legs are motionless, urine dribbles, keeping the patient constantly wet. An open meningomyelocele is liable to infections. The aim of treatment of meningomyelocele is to the patients with maximum mobility and social continence which is possible through multidisciplinary care.<sup>2</sup>

Birth of a baby born with MMC and hydrocephalus brings with it a lot of stress to parents. As for parents, it is equally difficult for treating paediatric surgeon to achieve satisfactory outcome in these patients. For a developing country like India where support system for these patients and parents is weak, we at our Spina bifida clinic are trying to achieve functional rehabilitation of patients and improvement in their quality of life. Our experience has shown that for an operated meningomyelocele baby to have any chance of living a fruitful life, availability of dedicated medical service will motivate parents to realize this goal.

Surgical approach for MMC repair which we follow includes following steps:

- Opening the parchment membrane close to the skin
- Careful dissection of neural placode from parchment membrane
- Dethering of cord

 Tubularization of neural placode and covering the defect with first a layer of dura and then a layer of deep fascia followed by skin.

In cases where fascial cover could not be achieved we have used synthetic patch with good results. For closure of large MMC defect a V-Y advancement flap based on subcutaneous pedicle is used. Various complications associated with MMC repair have been shown in Table 1.

MMC repair in neonatal life has shown good long-term results in terms of lower limb function and bladder-bowel continence as shown in Table 2.

MMC is associated with hydrocephalus in 70-80% of cases and in the present study we found it in 28% of cases at birth while 51% of patients developed hydrocephalus after MMC repair. 20% patients did not require shunt (Table 3). Hydrocephalus and its treatment is another important aspect in management of MMC.<sup>3</sup> Timely diagnosis and treatment of hydrocephalus will retain ability of brain parenchyma to grow and achieve near-normal intelligence to say the least.

The results of V-P shunt procedure are shown in Table 4.

For shunt tract infection, chamber tap was done and CSF was sent for culture examination. Full course of intravenous antibiotics according to sensitivity report was completed. CSF varix was managed by regular chamber compression or compression dressings in small varix. For large sized varix shunt had to be revised.

Unusual complication of shunt extrusion (6) and all cases of shunt block (12), CSF pseudocyst (5) needed revision of shunt. However, long term prognosis of these patients regarding shunt function and neurological development was dismal. In the present study shunt infection was found in 27% of patients and shunt revision was needed in all of them. Shunt related complications (e.g. shunt block, shunt infection with septicemia) mortality rate was 8%. Some patients showed tendency of repeated shunt tract infection, no specific predisposing factors could be found. The biggest concern of parents is whether their child can sit, walk and whether he/she would hold urine/feces after MMC repair. Before attempting any surgical intervention, we discuss with the parents about realistic goals that could be achieved over long term. Present study shows that 84 % of MMC patients have satisfactory continence after MMC repair. Lumbosacral and sacral type of MMC patients have highest preservation of incontinence. Same fact is observed with respect to lower limb power. Management of bladder/bowel incontinence and lower limb deformity helps in moral boosting of parents and patients as well. Our results of incontinence management are shown in Table 5 and 6.

Renal damage and renal failure are among the most severe complications of spina bifida. The majority of patients can be dry for urine by the time they go to primary school. To obtain such results, it is mandatory to treat detrusor over activity from birth onwards, as upper urinary tract changes start early in the first months of life. Current medical treatment provides the opportunity to convert a high-pressure bladder into a low-pressure reservoir that is safe for the upper urinary tracts.<sup>4-6</sup>

CIC was introduced for maintaining low pressure in neurogenic bladders in 1972.<sup>7-9</sup> We have found resolution in all patients of Grade 1 VUR and in 3 patients of Grade 2 VUR. Remaining patients showed lesser grade of VUR on subsequent VCUG.

We have not found CIC related complication in any of patient. CIC has helped in achieving dry interval in 74% of patients. Anticholinergic medication (oxybutynin) is added along with CIC for patients not having enough dry interval on CIC alone and patients showing upper tract changes on serial USG-KUB. We do urodynamic study only when child is minimum 3 years of age. Urodynamic study (UDS) of patients showing high pressure, low compliance, small capacity bladder is followed up closely. Dosage of anticholinergic medication is increased or bladder sphincter relaxants (Prazosin, Terazosin) are added as required. Chemoprophylaxis is started in these patients at risk for upper tract damage. We stress on CIC and rectal washouts in all patients to have minimum residual urine volume and empty rectum. Bowel management is important as constipation can cause difficulty in bladder emptying and increase postvoid urine volume.

This conservative management was effective in 74% of patients. Bladder augmentation (ileocystoplasty) with Mitrofanoff's procedure is considered for following group of children:

- Children on conservative management showing persistent upper tract dilation with decrease in renal parenchymal thickness
- Children showing deterioration in UDS parameters after conservative management of neurogenic bladder dysfunction.

We have encountered urine leak from incision site in 2 patients which was managed conservatively. Bladder wash with dilute sodium bicarbonate solution is started and gradually increased to clear mucous secreted by bowel segment. Blood gas analysis, renal function tests, USG-KUB are done on monthly follow-up.

Total 9 out of 12 operated patients achieved dry interval of more than 4 hours.

Patients with bowel incontinence were initially managed with rectal wash-outs with saline water prepared at home. Dose and frequency of rectal wash was determined for every child by trial and error method. Rectal wash was increased with weight of child (20 cc for each kg rise in

weight). Mothers were advised to avoid biscuits/bread, chocolates, high carbohydrate food for children. Dietary reference is taken when necessary. Patients responsive to rectal wash were given option of Malone's antegrade colonic enema (MACE) procedure. Bowel continence was achieved in 80% of patients.

Occupational therapists design custom made shoes to prevent foot ulcers, callosities and help patients walk. In addition to all this it is very important to counsel and train the parents regarding various aspects of the long-term implications, complications, treatment modules and options available for managing such children.<sup>14-15</sup>

#### **CONCLUSION**

Meningomyelocele in a child poses multiple challenges for clinicians and parents. Its management involves dedicated multispeciality clinic which should involve paediatric surgeon, pediatrician, nursing staff, occupational therapist, physiotherapist, social worker, parents and patients themselves. Successful rehabilitation can be achieved with parental education and support, dedicated clinicians and trained nursing staff, regular follow-up and low threshold for diagnosing clinical deterioration and proactive management to prevent further clinical deterioration.

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