



*Original Article*

## Early intervention in NTDs in children leads to better surgical outcome

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### Abstract

**Introduction:** Neural tube defects (NTDs) are a global disease, with the largest burden falling to low-and middle-income countries. Proper management of affected children can lead to a meaningful and productive life, and poorly managed cases of NTDs can be a devastating obstacle not only for patient but also for the patient's family.

**Methodology:** This prospective observational study was conducted in the Department of Paediatric Surgery, Sylhet M.A.G. Osmani Medical College & Hospital during the period from March, 2018 to August, 2020. Thirty children with a neural tube defect during the study period were included. Associated with life threatening major congenital malformations were excluded. The surgical management protocol used for different sites of NTDs was studied.

**Results:** The ages of the patients ranged from 1 day to 9 years with the median age of 3.5 days. The types of NTDs were myelomeningocele (73.3%), encephalocele (10.0%), lipomeningocele (10.0%) and meningocele (6.7%). The nature of neural tube defects was ruptured in (60.0%) and not

ruptured in (40.0%). Associated birth defects in this study were hydrocephalus in 56.7%, club foot in 16.7%, cervical prolapse in 6.7% and kyphosis in 6.7% cases. Associated neurological defects were lower limb paresis and paralysis in 83.3%, Urinary incontinence in 50.0% and faecal incontinence abnormality in 40.0% of cases. Postoperative complications were wound infection in 10.0%, wound dehiscence in 16.7%, CSF leakage in 23.3% and meningitis in 10.0% cases. The overall mortality rate was 6.7% of cases. Outcome of this study was recovery without complication in 66.7%, recovery with complications in 26.7% and death in 6.7% of cases. Recovery without complication was more frequent than recovery with complications and death.

**Conclusion:** There was recovery without complication in most cases after operation. Early operation is associated with recovery without complication.

**Keywords:** neural tube defects, myelomeningocele, encephalocele, meningocele, surgical management of NTDs, outcome of NTDs

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### Introduction:

CNTDs account for the most common congenital anomalies of central nervous system and results from failure of the neural tube to close spontaneously between 3rd and 4th weeks of in utero development.<sup>1</sup> The prevalence rate of NTDs in India is 7.48/1000 live births.<sup>2</sup> This is one

of the serious congenital malformation responsible for infant disability, morbidity and mortality.<sup>3</sup>

In the management of myelomeningocele, the patient without overt clinical hydrocephalus, most surgeons used to repair the myelomeningocele only. But that result subsequent hydrocephalus in 65 to 85% within 6 months and also cause breakdown of repair and CSF leakage. To prevent this complication some surgeons begins to do myelomeningocele repair and introduction of VP shunt at least 3 days after the initial repair. This gives better result but increase hospital stays and imposes an additional operative trauma within a short interval. It also increases the incidences of wound complication. Significant improvement has been shown in the management of myelomeningocele by doing

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simultaneous placement of VP shunt during repair of myelomeningocele. This decreases hospital stay, prevent repair failure, prevent or delay the development of subsequent hydrocephalus.<sup>7</sup>

The closure of the back lesion within 24hrs will improve neurological status. But there may be delay in motor prognosis in most children with NTD depends on the level of paralysis and surgical intervention, but appropriate bracing, physical therapy, intervention treatment of kyphosis and scoliosis can mitigate this.<sup>8</sup> The neonatal outcomes are demonstrates as neonatal death (10.1%) those were attributable to cardiac arrest (57.1%), prematurity (28.5%), and asphyxia (14.4%). Neonatal morbidity was 36.2%. The most performed surgery was myelomeningocele closure (92%).<sup>6</sup> The overall VP shunt requirement rate was 38%. Of these patients who underwent a surgical procedure, 30.7% had paraplegia; 51.6% had a neurogenic bladder; and 6.4% had infections due to surgery.<sup>6</sup> Whereas complications seen after surgery were more frequent in patients during the first week of life than in those seen during the second, third and fourth weeks of life.<sup>1</sup> So, this study aims to outline the outcome assessment of repair of NTDs as early as possible may effect in better prognosis.

### Objectives:

**General objective:** To observe the outcome of surgical management of the neural tube defects (NTDs) in children.

**Specific objective:** To evaluate the different types of post-operative complications, to manage the different modalities of post-operative complications, to observe the overall surgical outcome of NTDs.

### Methodology:

This prospective observational study was conducted in the Department of Paediatric Surgery, Sylhet M.A.G. Osmani Medical College & Hospital during the period from March, 2018 to August, 2020. Thirty children with a neural tube defect during the study period were included. Associated with life threatening major congenital malformations were excluded.

### Study Procedure

All the admitted patients with NTDs has been assessed by gestational age, birth weight, site of the sac, type of skin cover, status of CSF leak, presence or absence of infection of the sac, extent of the neurological deficit including bladder and bowel dysfunction, associated hydrocephalus, orthopaedics defects of the spinal

column and other associated congenital malformations. Patient has been advised for CBC, BT, CT, Blood grouping and Rh typing, Urine R/M/E, X-ray lumbosacral spine both A/P and lateral view, Chest x-ray P/A view, USG of the lesion, USG of brain, USG of whole abdomen and 2D echocardiography to detect the extent of the disease, associated anomalies and to prepare the patient for surgical management needed according to types. The newborn or infant with an open NTD has been kept warm and the defect has been covered with sterile dressing. The baby has been kept prone to prevent rupture of the sac and avoid trauma to the neural placode. Inj. Ceftazidime was started as 100mg/kg body weight in 2 divided dose intravenously and continued for 7 days. Wound swab was sent for culture and sensitivity and antibiotics were given accordingly for the cases of ruptured and infected NTDs. The goal of operative repair was to preserve the neural function and prevent infection primarily by obtaining a good skin closure over the defect. Surgical management was done under general and local anaesthesia (in few cases). Closure includes partial excision of the sac, water tight closure of the dura and reconstruction. Tension-free closure of the skin has been ensured after keeping a tube drain in situ. Sterile dressing has been applied over the operative area and catheterization has been done. To avoid direct pressure on the incision, patients were kept either in prone or lateral position. Most of the patients were operated  $\leq 7$  days of admission. Preoperative antibiotic has been continued at postoperative period. Antibiotics have been changed according to culture and sensitivity of the wound swab in case of wound infection. Dressing was changed on 3rd POD or at any moment if they are wet or soaked with stools. OFC was recorded daily. Collection from drain tube was measured daily. In most of the cases drain tube was removed at 3rd to 5th POD. Catheter was removed at 3rd POD. Most of the patients were discharged on 7th POD. The follow-up schedules were on 15th POD, at monthly upto 6 months and at 1 year after operation. In the follow-up we looked for CSF leakage from the wound and wound infection, paralysis of lower limbs or club foot, incontinence of urine and faeces, hydrocephalus. Parents were counseled and advised to attend in the physiotherapy outpatient department for club foot, lower limb paralysis or paresis. VP shunt was done for hydrocephalus. The prognosis was recorded by taking detailed history from the parents and proper physical examination of the patients.

### Statistical analysis

Data were processed manually and analyzed with the help of Microsoft Office Excel and statistical package for social sciences (SPSS) version 25.

Qualitative data were presented as frequency and percentage; and quantitative data were presented as mean and standard deviation.

Chi-square ( $\chi^2$ ) test was done to find out the level of significance between two categorical variables. A probability value (p) of <0.05 was considered statistically significant.

### Ethical consideration

An approval of the study protocol was obtained from the institutional Ethical Review Committee of Sylhet M.A.G. Osmani Medical College, Sylhet before the commencement of the study.

### Results:

Table 1 showed the distribution of the patients on the basis of age group.

Age	Number	%
1-7 days	20	66.7
8-30 days	4	13.3
<30 days	6	20.0

10 (33.3%) patients, who have been admitted after 7days of life, were recovered from the primary defect with complications. The duration of hospital stay was more than that who were admitted before 7 days of life.

Table 2 showed the distribution of the patients according to type of neural tube defects. Patients with myelomeningocele have more associated birth defects.

Type of NTDs	No.	%
Meningocele	2	6.7
Myelomeningocele	22	73.3
Encephalocele	3	10.0
Lipomeningocele	3	10.0

Table 3 revealed that the nature of neural tube defects was ruptured in 18 (60.0%), among them 7 (23.3%) developed post-operative complications. Morbidity and mortality was more in ruptured cases.

Nature of NTDs	No.	%
Ruptured	18	60.0
Not-ruptured	12	40.0

Table 4 Associated congenital defects

Associated congenital defects	Number of patients	%
Hydrocephalus	17	56.7
Club foot	5	16.7
Cervical prolapse	2	6.7
Kyphosis	2	6.7
No	12	40.0

Table 5 Associated neurological defects

Associated neurological defects	Number of patients	%
Lower limb paresis/paralysis	25	83.3
Urinary incontinence	15	50.0
Faecal incontinence	12	40.0
No	5	16.7

Table 6 Age of child during surgical management (n=30)

Age of child during surgical management	No.	%
≤7 days	20	66.6
>7 days	10	33.3

Table 7 Postoperative complications

Postoperative complications	Number of patients	%
Wound infection	3	10.0
Wound dehiscence	5	16.7
CSF leakage	7	23.3
Meningitis	3	10.0
No	20	66.7

Table 8 Treatment of postoperative complications

Treatment of postoperative complications	Number of patients	%
<b>Conservative</b>		
Antibiotics	10	33.3
Regular dressing	8	26.7
<b>Surgical</b>		
Secondary closure	6	20.0

Table 9 Treatment of associated anomalies

Treatment of associated anomalies	Number of patients	%
V-P shunt	12	40.0
Physiotherapy	25	83.3
Other	4	13.3
No	5	16.7

Table 10 Duration of hospital stay (n=30)

Duration of hospital stay	Number of patients	Percentage
≤10 days	20	66.7
>10 days	10	33.3

Table 11 Follow up at 15 days after operation

Outcome	Number	%
Wound infection	3	10.0
Lower limb paresis/paralysis	23	76.7
Faecal incontinence	10	33.3
Urinary incontinence	13	43.3
Hydrocephalus	15	50.0

Table 12 Follow up at 1months, 3 months, 6months and 12 months after operation

Outcome	1m	3m	6m	12m
Lower limb paresis/paralysis	76.7%	76.7%	76.7%	70.0%
Faecal incontinence	33.3%	33.3%	33.3%	33.3%
Urinary incontinence	43.3%	43.3%	43.3%	43.3%
Hydrocephalus	60.0%	60.0%	60.0%	60.0%

Table 13 Outcome (n=30)

Outcome	Number	%
Recovery without complication	20	66.7
Recovery with complications	8	26.7
Death	2	6.7

Table 14 Association between delay in operation and outcome

Outcome	≤ 7 days	>7days	p-value
Recovery without complication	16 (80.0%)	4 (20.0%)	p=0.045
Recovery with complications & Death	4 (40.0%)	6 (60.0%)	

Early operation is significantly associated with recovery without complication (p=0.045).

### Discussion:

Neural tube defects (NTDs), those are caused by incomplete neural tube closure during the 4th week of pregnancy and result in various levels of neurological deficit. Adequate folic acid consumption during early pregnancy prevents the occurrence of NTDs. However, because the neural tube closes by week 4 of fetal development, early access to antenatal care (ANC) is crucial in the prevention or early detection of NTDs.<sup>9</sup>

In an attempt to mitigate the complications associated with open spina bifida, the standard of care in upper-middle-income countries in the acute phase includes diagnosis in utero, cesarean section rather than vaginal birth to avoid infection, surgical closure of the open lesion within 24 hours, and continued monitoring for the development

of hydrocephalus, with surgical intervention as needed. Moreover, due to the complexity of their diagnosis, patients in upper-middle-income countries are monitored by multiple highly trained specialized healthcare teams (e.g., paediatric neurosurgery, urology, orthopedics, pediatrics, physical therapy). In resource-limited settings, outcomes are greatly affected by the lack of coordinated care.<sup>5</sup>

In this study the ages of the patients ranged from 1 day to 9 years with the median age of 3.5 days. Goswami et al., (2015) found that age at presentation of neural tube defect ranged from 1 day to 5 years. In this study, there were 20 (66.7%) patients in the age group of 1-7 days, 4 (13.3%) patients in the age group of 8-30 days and 6 (20.0%) patients in the age group of above one month. Delayed presentation yet might be due to late referral, financial problems and sometimes folk cultural and religious beliefs that NTDs are a punishment from God. In the present study myelomeningocele (73.3%) was the most common neural tube defects and others were encephalocele (10.0%), lipomeningocele (10.0%) and meningocele (6.7%). Gedefaw et al., (2018) found that 54.1% were cases of anencephaly; 40.5%, spina bifida; and 5.4% encephalocele. The frequency of spina bifida was in higher proportion compared to anencephaly reported in the study of Rai et al., (2016). However, many studies have reported equal incidences of these two forms of NTD.<sup>12,13</sup>

This study revealed that the nature of neural tube defects was ruptured in 18 (60.0%) and not ruptured in 12 (40.0%) cases. This result was consistent with the study of Motah et al., (2017) who reported neural tube defects was ruptured in 52.2% of cases and un-ruptured in 47.8% of cases.

Associated congenital defects in this study were hydrocephalus in 56.7%, club foot in 16.7%, cervical prolapse in 6.7% and kyphosis in 6.7% cases. Motah et al., (2017) also found associated birth defects were hydrocephalus (65.3%), bilateral talipes calcaneovalgus (18.4%), bilateral talipes equinovarus (16.3%) and several other less frequent defects.

Associated neurological defects were lower limb paresis/ paralysis in 83.3%, Urinary incontinence in 50.0% and faecal incontinence in 40.0% of cases. Motah et al., (2017) reported neurological defects associated with NTD were lower limb paresis/ paralysis in 65.33%; and urinary incontinence and faecal incontinence deficit in 61.2% of cases Kumar and Singh, (2003) and Alatisse et al., (2006) had reported similar results. Sphincteric

dysfunctions were more frequent in patients with myelomeningocele.

The definitive procedure for NTDs is operative closure of the defect within. Closure includes partial excision of the sac, water tight closure of the dura and reconstruction. None of the patient in this study had been operated within 72hrs of age. The ages of the child during operation are  $\leq 7$  days for 20 (66.7%) patients and  $>7$  days for 10 (33.3%) patients.

Postoperative complications in the present study were wound infection in 10.0%, wound dehiscence in 16.7%, CSF leakage in 23.3% and meningitis in 10.0% cases. Treatment of postoperative complications was administration of antibiotics (33.3%), regular dressing (26.7%) and physiotherapy (10.0%) for cases as conservative treatment; and 6 (20.0%) patients required surgical management in the form of secondary closure. The 10 (33.3%) patients who had been developed postoperative complications were operated after 7 days of age, and the defect was presented in the lumbosacral region. Among them 7 (23.3%) patients had myelomeningocele and 3 (10%) patients had lipomeningocele. The overlying skin was ruptured in 7 (23.3%) patients and not ruptured in 3 (10%) patients. Motah et al., (2017) observed 55.1% of patients had complications following surgery. The most common post-operative complication was wound infections (44.9%). Other immediate post-operative complications were wound dehiscence(40.8%), CSF leakage (20.4%) and meningitis/sepsis (32.7%).

Treatment of associated anomalies was V-P shunt (40.0%), physiotherapy (83.3%) and others (13.3%). On admission 17 patients were diagnosed as hydrocephalus. Among them 2 patients were died postoperatively. Three (10%) patients had been developed hydrocephalus postoperatively. Among the total 18 patients 12 patients undergone for V-P shunt procedure within 5 months of age. From the remaining 6 patients, 3 patients had arrested hydrocephalus and the parents of other 3 patients did not give consent for shunt procedure.

Total duration of hospital stay is  $\leq 10$  days for 20 (83.3%) patients and  $>10$  days for 10 (16.7%) patients. Reasons for longer in hospital stay were due to postoperative complications like wound infection, wound dehiscence, CSF leakage and meningitis.

Outcome in this study was recovery without complication in 66.7%, recovery with complications in 26.7% and death in 6.7% of cases. Among these deceased one patient was due to prematurity, septicaemia and meningitis

and; another patient was due to encephalitis. Recovery without complication was more frequent compared to recovery with complications and death. Motah et al., (2017) found that out of the 41 patients who were discharged after surgery, 16 (39.0%) of them were completely lost to follow-up a few months to years after discharge from hospital. 10 (24.4%) of the remaining 25 patients were reported to have died a few months (some a few years) after surgery. Most of them were said to have presented symptoms of progressive hydrocephalus (enlarging cranium, sun-setting appearance of the eyes, vomiting, dilated scalp veins). The remaining 15 (36.6%) are reported to be alive with a majority of them (12; 80.0%) having gross motor and sensory deficits as well as bi-sphincteric dysfunction. The 03 others (20.0%) have completely recovered. Eleven (73.3%) of those who are alive with gross motor and sensory deficits as well as bi-sphincteric dysfunction presented with myelomeningocele.

In this study recovery without complication was in 20 (66.7%) patients which was more frequent in operation at  $\leq 7$  days of life, whereas recovery with complications and death in 10 (33.3%) patients was associated with operation at  $>7$  days of life. Recovery without complication was in 16 (80.0%) patients whose delay in operation of  $\leq 7$  days and was in 4 (20.0%) patients whose delay in operation of  $>7$  days; whereas recovery with complications and death was in 4 (40.0%) patients whose delay in operation of  $\leq 7$  days and was in 6 (60.0%) patients whose delay in operation of  $>7$  days. Early operation is significantly associated with recovery without complication ( $p=0.045$ ).

Motah et al., (2017) reported that 44 (89.8%) patients had their defects closed at birth, 25 (70%) of them within the first 48 - 72 hours following admission. The 16 (39%) others were operated after the fourth day of admission. Out the 32 (65.3%) patients who had hydrocephalus, ventriculoperitoneal shunting was performed in 25 (78.1%) of cases. 24 (64.9%) of those treated for hydrocephalus had myelomeningocele. All these ventriculoperitoneal shunts were placed after definitive repair of defects. Complications seen after surgery were more frequent in patients during the first week of life than in those seen during the second, third and fourth weeks of life.

### Conclusion:

In Neural tube defects (NTDs), the more common associated birth defect is hydrocephalus and neurologi

cal defect is lower limb paresis/ paralysis. Recovery without complication is more frequent than recovery with complications and death. Early operation is associated with recovery without complication and less morbidity.

### Limitations of the study:

This study was not without limitations. The limitations of the study were-

None of the patient have been operated before 72hrs of life, lack of opportunity for MRI facility, lack of expert neonatal neurosurgeon, lack of dedicated neonatal anaesthesiologist, several surgeons performed the operation, lack of opportunity for NICU support, lack of multidisciplinary approach, follow up period was short.

### Recommendations:

In the light of the above mentioned results, we formulate the following recommendations:

Perform definitive surgery within 24-48 hours of life, involve other specialists (Neurosurgeon, urologists, paediatricians, orthopaedicians, gynaecologists, and physiotherapists) in the management of children born with NTDs, ensure early referral of babies born with NTDs in aseptic and non-traumatic conditions to specialized health centers for proper and timely management, ensure proper NICU facilities, educate the public on the risk factors of NTDs, raised awareness about proper ANC and maternal ultrasound at mid-trimester to detect NTDs, women of child bearing age who are capable to become pregnant should consume 0.4 mg of folic acid per day to reduce their risk of having a fetus affected with spina bifida or other neural tube defects.

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