Volume 10, Issue 08, August 2024,
Publish Date: 12-08-2024
Doi https://doi.org/10.55640/ijmsdh-10-08-05

International Journal of Medical Science and Dental Health (Open Access)

LATE REPAIR OF MYELOMENINGOCOELE: DOES TIMING OF PRESENTATION AFFECT OPERATIVE OUTCOME?

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ABSTRACT

Background: Myelomeningocele (MMC) is the most severe form of spina bifida. While delayed surgical closure is not associated with a higher prevalence of long-term morbidity, early closure is known to reduce the risk of meningitis and ventriculitis. However, most of the patients in our practice setting either presented late or presented under septic condition both precluding the possibility of early repair. This later subset had the benefit of antibiotic treatment and wound care prior to operative repair.

Aim of the study: To compare those that prsented early with those that presented late.

Patients and Methods: This is a prospective cohort study involving patients with myelomenigocoele managed operatively at the Regional Centre for Neurosurgery, Sokoto between September, 2015 and November, 2016. Demographic and post-operative clinical outcomes during in-patient admission and six months follow-up period including superficial wound infection, deep wound infection, cerebrospinal fluid leaks, hydrocephalus and timing of VP shunt insertion were studied and analysed.

Results: A total of 90 patients were studied with 45 recruited for each arm of the study (early presenters versus late presenters) respectively. The overall mean age at presentation was 101.86 days with male to female ratio of 1:1. The mean time from birth to first VP shunt was 2.7±1.4 months for the early presenters and 6.0±2.2 months for the late presenters with $p < 0.001$). Sixteen of the patients (35.6%) who presented early had superficial wound infection compared to 12 (26.7%) of those late presented late with $p = 0.362$. Six patients. Eight of the patients (17.8%) who presented early had CSF leaks while 10 (22.2%) of patients that presented late had CSF leaks with $p = 0.598$. Eighteen (40.0%) of patients that presented early developed hydrocephalus while 20 (44.4%) of patients that presented late developed hydrocephalus with $p = 0.03$. Twenty seven (60.0%) of the patients who presented late had poor gross motor development compared to 16 (35.6%) of patients that presented early with $p = 0.020$. Six (13.3%) of patients that presented early died compared to 1 (2.2%) patient that presented late.

Conclusion: The mean time from birth to first ventriculo-peritoneal shunt insertion for those who presented early was 2.7+1.4 months compared to 6.0+2.2 months for patients that presented late This association had strong statistical correlations with P < 0.001. Superficial wound infection was commoner among those that presented early. Conversely, cerebrospinal fluid leak and hydrocephalus were more common among patients that presented late.

INTRODUCTION

Myelomeningocele (MMC) is the most severe form of spina bifida.[1] Before 1960, less than 10% of patients with MMC survived, most dying of infection and hydrocephalus.[2,3,4] The management of MMC has undergone significant evolution over the years from a conservative attitude to a more active intervention.[2] There is the need for multidisciplinary approach in the management of patients with MMC.[2] Although early surgical intervention on all babies with MMC was carried out in the past to minimize injuries to the nervous tissues and its attendant worsening of neurological dysfunction, long term follow up of such patients has demonstrated unpleasant morbidities among survivors, necessitating the need for preoperative selection criteria to optimize the benefits of operative intervention in resource poor countries such as Nigeria, where access to comprehensive medical care is limited to a negligible proportion of the population.[5]

While delayed surgical closure is not associated with a higher prevalence of long-term morbidity, early closure is known to reduce the risk of meningitis and ventriculitis. Audu et al in 2003 showed mortality rate of 16.7% after delayed repair. Presence of major congenital abnormalities contributes significantly to neonatal death. Managing patients with MMC in Sub-Saharan Africa can be potentially devastating due to prevailing low socioeconomic status, harmful taboos, religious beliefs, paucity of medical man power and inadequate facilities.[6]

The development of modern neurosurgery (closure of the lesion and treatment of hydrocephalus) and the advent of treatment for the neuropathic bladder addressed the major causes of mortality.[7]

Outcomes following surgery depend on interplay of several factors. A study by Eghwrudjakpor et al in Port Harcourt, Nigeria confirmed that the most important predictors of post-operative outcome are the spinal level and size of the defect.[8] This is corroborated by another study in Zambia which found that immediate and long term outcomes are related to the site and size of the defect, associated malformations and complications.[9] Shehu et al had earlier documented functional disability in 38 percent of cases of spina bifida cystica who underwent surgery in Zaria and attributed this to inadequate facilities for postoperative rehabilitation.[10] Neural tube defects therefore constitute a burden on parents as well as community.

There has been continuing controversy on the best management option for MMC since Lobber published his treatise on the subject in 1971.[11] Broadly speaking, there are two methods of postnatal management for myelomeningocoele: early repair (within 48 hours of birth) and late repair (after 48 hours of birth). Pressure of work and lack of hospital bed spaces have forced many surgeons into delaying operation in developing countries.[12] Proponents of early surgical repair believe that reducing risk of infection and CSF leaks would reduce meningitis, verticulitis and worsening of Arnold Chiari malformations and development of syringomyelia.[12]

In our experience, most of the patients presented late thereby making early repair impossible. Even the few patients who presented early often carried high risks of potential infection because of unsupervised

delivery under septic condition, unorthodox application of non-sterile concussive materials, covering of the sac with unsterilized clothing materials or smearing of the neural placode with faecal matter. All these necessitated initial wound care before planning surgical interventions. This anecdotal evidence precluded the possibility of early repair in our setting (personal communication from expert). However, patients who presented early within the first month of delivery especially with wound complications had the benefit of antibiotic treatment and wound care prior to operative repair. For the purpose of this research, early presentation means presentation to the health facility within the first 30 days of life while late presentation means presentation to the health facility after the first 30 days of life.

OBJECTIVE

To compare outcomes of delayed repair of MMC in patients who presented within the first thirty days of life (early presentation) and those who presented after the first thirty days of life (late presentation).

PATIENTS/METHODS

Study Design

This is a prospective cohort study involving all patients with myelomenigocoele managed operatively at the Regional Centre for Neurosurgery, Usmanu Danfodiyo University Teaching Hospital Sokoto between September, 2015 to November, 2016.

Data Collection

Demographic and post-operative clinical outcomes during inpatient admission and six months follow-up period.

Inclusion Criterion

Patients with established diagnosis of myelomeningocoele who underwent surgical intervention within the study period.

Exclusion Criteria

All patients who presented with myelomeningocoele during the study period who could not have surgery.

All patients who presented with myelomeningocoele during the study period but defaulted from care or follow up clinics.

All patients who presented and managed for myelomeningocoele during the study period but refused consent for the study

All cases of lipoma, dermoid cysts or meningocoeles.

Ethical Considerations

This study was carried out after ethical and institutional clearance from the Usmanu Danfodiyo University Teaching Hospital Research and Ethics Committee. The nature of the study was carefully explained to patients, proxy or caregiver as the case may be via consent form. Consent for inclusion into the study was obtained using consent form and patients' privacy were respected and treated with confidentiality.

Sample Size

The sample size was calculated using the fisher's formula as follows:

n = z2pq/d2

Allowing for an attrition rate of 10%, 90 patients were recruited into the study. The first 45 patients who presented early during the study period and certified inclusion criteria were categorised into group A while the first 45 patients who presented late during the study period were categorised into group B respectively.

Data Management and Analysis

Data was analysed using statistical packages for the social sciences (SPSS) version 21 for windows. Data collected on proforma was entered using numerical codes. Comparisons of proportions between the two groups were made by using chi-square analysis or Fischer's exact tests for means or Mann-Whitney test for medians where appropriate. Univariate analysis was carried out to compare patient's risk factors and postoperative outcomes associated with early presentation and late presentation.

Additionally, multivariate logistic regression was used to determine significant risk factors associated with postoperative outcomes.

Peri-operative Management: All the patients presented either through the outpatient clinic or emergency Department. Those that presented with ulcerated and/or infected sacs were admitted initially for wound care and were given parenteral ceftriaxone 100mg/kg body weight per day in two divided doses for two weeks and daily wound dressings with sufratulle gauze after minimal cleaning with saline. Wound care was continued until the infection was cleared and satisfactory healing was achieved before planning for surgery. Patients without pre-operative wound complications were operated as soon as they were optimised for general anaesthesia.

All patients included in this study had surgery with excision of the MMC sac watertight dura closure under general anaesthesia. All patients had intra-operative ceftriaxone 100mg/kg body weight as single dose to reduce the risk of surgical site infection. Patients with hydrocephalus as detected by preoperative TFUSS, CT scan or MRI scan had VP shunt insertion alongside excision and repair in one sitting. However, patients who developed hydrocephalus post excision and repair of MMC had VP shunt insertion in another sitting.

Post-operatively, all the patients were nursed as in-patient for 10 days. Skin sutures were removed by the tenth day and patients discharged home except those with wound complications who had extended periods on admission. All the patients were ultimately followed up for 6 months. Outcome Measures were based on superficial wound infection, deep wound infection, cerebrospinal fluid leaks, hydrocephalus and timing of VP shunt insertion.

RESULTS

Socio-demographic Characteristics

A total of 90 patients were recruited for this study. Forty five patients with MMC who were ≤ 30 days old at the time of presentation were recruited into the group A (early presentation) arm of the study and another 45 patients who were above 30 days old at presentation were recruited into the group B

(late presentation) arm of the study respectively. The overall mean age at presentation was 101.86 days with a standard deviation of 213.32. However, the mean age for groups A and B patients were 10.0±8.1 and 193.8±276.5 days respectively. Forty-three (47.8%) patients were 28 days old or less, [41] (45.6%) were 29 – 364 days old and 8 patients were 365 days old or above. There was no sex dominance with male to female ratio of 1:1. However, there were more female patients in group A {28(60.5%)} while in group B there were more male patients {28(60.5%)}. More than half {51 (56.7%)} of the patients' mothers were age 19 – 35 years while 16 (17.8%) were < 19 years and 23 (25.6%) older than 35 years respectively. Among patients with adolescent mothers (<19 years), 10 (22.2%) were in group A while 6 (13.3%) were in group B respectively. Sixty-three (70.0%) of the patients' mothers had no formal education, of which 30 (66.7%) were in group A while 33 (73.3%) were in group B. Seventeen of the mothers (18.9%) had primary education while 10 (11.1%) secondary education or higher level of education.

Table 1 *(Summary of Socio-demographic characteristics of the patients.)*

Variable	**Time**		
	Group A (%)	**Group B (%)**	**Total n (%)**
Age			
Neonate	43 (95.6)	0 (0.0)	43 (47.8)
Infant	2 (4.4)	39 (86.7)	41 (45.6)
Child	0 (0.0)	6 (13.3)	6 (6.7)
Mean age (days)	*10.0 ± 8.1*	*193.8 ± 276.5*	
Sex			
Male	17 (37.8)	28 (62.2)	45 (50.0)
Female	28 (62.2)	17 (37.8)	45 (50.0)
Mothers' age (years)			
< 19	10 (22.2)	6 (13.3)	16 (17.8)
19 – 35	24 (53.3)	27 (60.0)	51 (56.7)
> 35	11 (24.4)	12 (26.7)	23 (25.6)
Mean maternal age (years)	*28.9±9.0*	*27.9±8.2*	
Mothers' level of education			
None	30 (66.7)	33 (73.3)	63 (70.0)
Primary	10 (22.2)	7 (15.6)	17 (18.9)
Secondary/post-secondary	5 (11.1)	5 (11.1)	10 (11.1)

Comparison of early and late presentation of MMC (Group A vs Group B)

The mean time from birth to first VP shunt for the group A was 2.7±1.4 months compared to 6.0±2.2 months for the group B with a statistically significant difference between the means ($p < 0.001$). Sixteen (35.6%) of the patients who presented early (Group A) had superficial wound infection compared to 12 (26.7%) of those who presented late (Group B) with a statistical correlation of $p = 0.362$. Six patients (13.3%) in group A had deep wound infection compared to 5 (11.1%) of patients in group B with $p = 0.748$. Eight (17.8%) of patients in group A had CSF leaks while 10 (22.2%) of patients in group B had CSF leaks with $p = 0.598$. Eighteen (40.0%) of patients in group A developed hydrocephalus while 20 (44.4%) of patients in group B developed hydrocephalus with significant statistical correlation ($p =$

0.03). Three (6.7%) of the patients in group B had tethered cord while none of the patients in group A had tethered cord. This was not statistically significant (p = 0.242). Twenty-seven (60.0%) of the patients in group B had poor gross motor development compared to 16 (35.6%) of patients in group A. This was statistically significant (p = 0.020). Six (13.3%) of patients in group A died compared to 1 (2.2%) patient in group B. This association was not statistically significant (p = 0.110) (Table 2).

Table 2 *(Comparison of early and late repair of MMC (Group A vs Group B).)*

Variable	**Time of intervention**		**Test statistic**	**p value**
	Early	**Late**		
	n (%)	**n (%)**		
Outcome after surgery				
Alive	39 (86.7)	44 (97.8)	F = 3.873	0.110
Dead	6 (13.3)	1 (2.2)		
Time to first VP shunt				
Mean time	2.7±1.4	6.0±2.2	*t* = -5.825	***< 0.001***
Superficial wound infection				
Yes	16 (35.6)	12 (26.7)	χ^2 = 0.829	0.362
No	29 (64.4)	33 (73.3)		
Deep wound infection				
Yes	6 (13.3)	5 (11.1)	χ^2 = 0.104	0.748
No	39 (86.7)	40 (88.9)		
CSF leak				
Yes	8 (17.8)	10 (22.2)	χ^2 = 0.278	0.598
No	37 (82.2)	35 (77.8)		
Hydrocephalus				
Yes	18 (40.0)	20 (44.4)	χ^2 = 0.182	**0.03**
No	27 (60.0)	25 (55.6)		
Tethered cord				
Yes	0 (0.0)	3 (6.7)	F = 3.103	0.242
No	45 (100.0)	42 (93.3)		
Gross motor activity				
Good development	29 (64.4)	18 (40.0)	χ^2 = 5.388	***0.020***

Poor development	16 (35.6)	27 (60.0)

Significant F = Fisher's exact test χ2= Chi square test t = t test

DISCUSSION

Majority of the patients on this study presented after 72 hours of birth, the time required for early operative repair of myelomeningocoele defect with a mean age of 101.86+213.[32] days at presentation. This late presentation is due to financial inability, lack of awareness of availability of treatment or time lag required to travel from place of birth to the RCNS which was the only treatment centre within the region at the time of this study. The equal sex distribution found was almost similar to the findings from two other studies in different locations in Nigeria.[8, 9] Poverty and low social statuses are known risk factors for MMC.[13] We found 88.9% of patients born to uneducated mothers. Studies by Miller P.D et al and Scort Adzick et al have shown that the single strongest predictor of having a child with a neural tube defect was low maternal education.[14, 15]

Apart from presence of MMC sac, two other common reasons for hospital presentation were the presence of hydrocephalus and infection.16 The later would ultimately lead to early onset of hydrocephalus: Infection leads to retrograde meningitis and early post meningitic hydrocephalus.[17] The mean time from birth to first ventriculo-peritoneal shunt insertion for those who presented early was 2.7+1.4 months compared to 6.0+2.2 months for patients that presented late This association had strong statistical correlations with $P < 0.001$. The majority of the patients with myelomeningocele (MMC) will need surgical treatment for associated pathologies, like hydrocephalus (HCP).[18] The insertion time of the shunt used for hydrocephalus treatment is controversial for the risk of infection in patients with myelomeningocele.[18] Many authors have reported the risk of ventriculo-peritoneal shunt infection in early myelomeningocele repaired patients.[18] Superficial wound infection was commoner among those that presented early (35.6%) compared to those who presented late (26.7%). The deep wound infection is also more common with those that presented early (13.3%) than those that presented late (11.10%). In a related study on prevalence of surgical site infection in myelomeningocoele with 109 patients, 37 (33.94%) cases of SSI were found. Among the 37 SSIs, 22 (59.46%) were classified as deep, 10 (27.03%) as superficial and five (13.51%) as organ/space.19 Conversely, the tendency for cerebrospinal fluid leak was more common among patients who presented late (22.2%) compared to those that presented early (17.8%). Previous studies considered closure techniques rather than timing of presentation and found no significant difference between incidence of csf leaks and type of closure techniques.1 The reported incidence of MMC-related hydrocephalus following postnatal closure of the MMC defect ranges from 57% to 86%.[20] In a study of 8233 patients with MMC and hydrocephalus, 2026 (24.6%) received no early hydrocephalus treatment on initial inpatient stay.20 In our study, hydrocephalus was common among patients that presented late (44.4%) compared to those that presented early (40.0%), this was because hydrocephalus has had adequate time to evolve as either late complication from infections, progressively increasing pressure from MMC sac or posterior fossa abnormality. This association was statistically significant with $P = 0.03$. More of the patients that presented early (13.3%) died compared to those that presented late (2.2%), this was similar to Eric D et al finding that mortality of NTD was increased over the general population risk in the first month of life.[21]

CONCLUSION

Majority of the patients on this study presented after 72 hours of birth, the time required for early operative repair of myelomeningocoele defect with a mean age of 101.86+213.32 days at presentation. The equal sex distribution found was almost similar to the findings from two other studies in different locations in Nigeria. The mean time from birth to first ventriculo-peritoneal shunt insertion for those who presented early was 2.7+1.4 months compared to 6.0+2.2 months for patients that presented late This association had strong statistical correlations with $P < 0.001$. Superficial wound infection was commoner among those that presented early. Conversely, cerebrospinal fluid leak and hydrocephalus were more common among patients that presented late.

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