VALIDATION OF A MYELOMENINGOCELE SEVERITY SCALE

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DECLARATION

I declare this dissertation is my original work and has not been submitted for a degree in any other university.

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SB ï Spina Bifida

CSF ï Cerebrospinal fluid

CIC ï Clean Intermittent Catheterization

MMC ï Myelomeningocele

QoL ï Quality of Life

CNS ï Central nervous system

WBC ï white blood cell

SBNS ï Spina Bifida Neurologic Scale

WHO ï World Health Organization

NTD ï Neural Tube Defect

KNH ï Kenyatta National Hospital

FIM ï Functional Independence Measure

ADL ï Activities of Daily Living

KS ï Kijabe Score

UON ï University of Nairobi

Validation of a Myelomeningocele Severity Scale

ABSTRACT

Background: Myelomeningocele as a neural tube defect occurs in varying levels of severity. The ambulatory function outcome is well correlated with the MMC spectrum by existing spina bifida scores. None of the existing scores predict survival or study the effect of more than two severity markers.

Objective: To validate the proposed myelomeningocele severity scale as a predictor of survival at the 3 year end point.

Methods: This was a cross sectional institutional study that recruited 153 patients from a high volume center. The study duration was between 2010-2014, during which time data on motor function, presence or absence of a kyphus deformity, hydrocephalus, ventricular CSF WBC count, bladder function, syndromic features and Chiari II symptomatology were collected and analysed. Biostatistical analysis was done using the STATA 14 program. Results are presented in form of tables, graphs, pie charts and diagrams.

Results: A total of 153 patient charts were reviewed. The minimum and maximum scores were 7 and 24 respectively on the myelomeningocele severity scale. The new scale was found to be as reliable as the SBNS for prediction of future mobility (Pearson Correlation = -0.74). The 3 year overall mortality rate was 15% (24 patients). Thirteen of these deaths occurred in the high risk category (a score of 21-30), 11 from the intermediate risk category (12-20) and 1 within the low risk category (7-11). Low and intermediate risk groups had longer survival as opposed the high risk group patients (2/3 of the high risk mortalities occurred within the first year of life).

Conclusion: The myelomeningocele severity scale is a reliable predictor of survival 3 years post MMC closure. It is also as reliable as the already existing SBNS scale for predicting the motor outcome at 3 years post MMC repair.

CHAPTER ONE

INTRODUCTION AND LITERATURE REVIEW

1.1 Introduction

Spina bifida is the most common birth defect affecting the central nervous system (CNS) that is compatible with survival.[14] Bunch et al, first discussed this in 1972 in reference to the multiplicity of conditions that can affect a patient with myelomeningocele: spine and spinal cord, mobility, activities of daily living (ADL), continence, learning, impaired social interactions among others.

Worldwide, the incidence of neural tube defects ranges from 0.17 to 6.39 per 1000 live births[7]. Verhoef et al recruited 179 individuals and divided them into those with high-level (L2 and above), mid-level (L3-L5) and low-level (S1 and below) lesions. According to this study, most medical studies were found in the high-level lesion group, however, all subgroups suffered from health problems. The most important medical problems described are hydrocephalus and cognitive dysfunction, urinary and fecal incontinence, reduced mobility, renal failure, hypertension, pressure sores, obesity, epilepsy and decreased visual acuity. Orthopaedic problems such as scoliosis, fractures, and contractures are also often mentioned. [36, 37]

Developing countries report higher NTD incidences. A prospective study of 3500 consecutive births in Southern India found 40 babies to have NTD, bringing the incidence to 11.4/100 live births[16] in that part of the world. The incidence was 7/1000 live births in the middle belt of Nigeria [17] and 0.47/1000 live births in Blantyre, Malawi.[18]

The World Health Organization (WHO) estimates that 6% of deaths in children aged less than 5 years in Kenya are due to congenital anomalies.[26] The proportion of these deaths that are due to neural tube defects can be estimated from hospital based studies. The prevalence

would be an underestimate on account of the absence of a birth defect surveillance system in Kenya.[27]

In Kenya, Mwangôombe et al., studied 65 patients with spina bifida treated at the Kenyatta National Hospital between September 2011 and August 2012. 61.5% of these were male and 38.5% female patients.[24] A cross-sectional study conducted at the Kenyatta National Hospital in 2009 reported a hospital-based prevalence of 20 neural tube defects per 10,000 live births.[25] This survey adopted a quasi-experimental design to study a total of 7,355 births at the maternity unit of KNH, the largest referral hospital in Kenya. 4.6 NTD per 1,000 live births were counted.

This treatable spinal cord malformation occurs in varying levels of severity[3] and theoretically, mobility and mortality outcomes are dependent on the severity.

1.2 Literature Review

1.2.1 Overview

There is a wide range of neurological and functional outcomes in myelomeningocele survivors, the most critical determinants of which could be symptomatic CSF shunt failure[1] and CSF shunt infections[2]. The former study was a review of literature undertaken using the PubMed database. In the latter prospective study, 120 of 189 MMC patients (64%) experienced a first shunt failure with a median time of 303 days.

The level and size of the defect are important predictors of severity; with low level, smaller defects having better outcomes[3]. Eghwrudjakpor et al deduced this from a Nigeria-based study which examined 15 children (9 males and 6 females) following MMC closure. Although the sample was admittedly small, the authors found that level and size of MMC

defect was a major determinant of neurological impairment and a possible pointer to outcome of surgery.

In a cohort followed up for over 25 years following the initial myelomeningocele closure, mortalities were characterized by a higher rate of posterior fossa decompression[5]. In this study carried out the Childrenôs Hospital of Pittsburgh 25 children treated with MMC closure and shunting for hydrocephalus underwent sub occipital craniectomy, cervical laminectomy and dural decompression to relieve symptomatic Chiari malformations. Outcome correlated closely with the preoperative neurological status. Specifically, the presence of bilateral vocal cord paralysis was associated with a poor response to surgery ($p<0.001$ on both univariate and multivariate analyses).

CIC improves bladder continence and may potentially prevent death resulting from renal failure[4]. Bowman et al, in a 25 year prospective study additionally demonstrated that continence may decrease to 63% for patients not on CIC.

Good motor function correlates with better mobility through to teen and adult years[4]. In other words, even though some studies have demonstrated that mobility decreases from early childhood to early teen years, 75-100% of patients who remain mobile in their teens continue to ambulate in their early adult years.

Delayed wound healing and late skin breakdown with exposure of instrumentation are common problems experienced in patients a surgically corrected kyphus deformity[6]. This was described by Niall et al who studied 24 children with MMC and a kyphotic deformity treated by surgical correction. In this study further surgery to remove protruding hardware was necessary in 18 patients.

Clinical observations at the Kijabe Hospital, a peripheral hospital in Kenya with a dedicated pediatric neurosurgical unit, point to a poorer prognosis in patients with concomitant myelomeningocele, hydrocephalus, low set, externally rotated ears, cortical thumbs and telecanthus.

1.3 The Spina Bifida Neurological Scale (SBNS)

An existing Spina Bifida Neurological Scale (Table 1), scores spina bifida infants based on motor function, reflexes, bowel and bladder function[15]. It is used to foresee future daily activity of the infant. This tool does not predict survival; neither does it incorporate the severity markers outlined in the proposed Myelomeningocele Severity Scale.

Table 1: The Spina Bifida Neurological Scale

Functioning	C-Th	L1	L2	L3	L4	L5	S1
Nonfunctioning	L2		L3	L4	L5	S1	S2
Hip							
Flexion	-		+/-	+	+	+	+
Extension	-		-	-	-	-	+/-
Adduction	-		+/-	+	+	+	+
Abduction	-		-	-	-	_	+
Knee							
Extension	-		+/-	+	+	+	+
Flexion	-		-	-	-	+/-	+
Ankle							
Dorsiflexion	-		-	-	+/-	+	+

Plantarflexion	-		-	-	-	-	+/-
Inversion	-		-	-	+/-	+	+
Evertion	-		-	-	-	-	-
SBNS motor (worse side)	1		2	3	4	5	6

Scoring scale for motor function:

Functioning	C-Th ~L2	L3	L4	L5	S1	S2	S3
Nonfunctioning	L3	L4	L5	S1	S2	S3	S4
Patellar reflex	-	+/-	+	+	+	+	+
Achilles reflex	-	-	-	+/-	+	+	=
Anal reflex	-	-	-	-	_	+/-	+/-
SBNS reflex	1	2	2	3	3	4	4

Scoring scale for preserved reflexes:

Functioning	C-Th L-S1	S2	S2				S3	S3
Non-functioning	S2	S3	S3				S4	S4
Bladder control	-	-	+/-	+	+/-	+	+	+
Bowel control	-	+/-	-	+/-	+	+/-	+	+
SBNC BB control	1	2	2	3	4	4	5	5

Scoring for bladder and bowel function

1.4 Hoffer Classification

The Hoffer classification (Table 2) was described in 1973 [28] as a tool used to measure functional ambulation in patients with myelomeningocele. This classification has been used to measure the functional independence among young adults with spina bifida, in relation to hydrocephalus and level of lesion.[29]

Table 2: Hoffer's functional ambulation classification

Functional ambulation classification according to Hoffer
Community ambulators Patients walk indoors and outdoors for most activities; may need crutches, braces, or both. Wheelchair used only for long trips out of community.
Household ambulators Patients walk only indoors and with orthoses. Able to get in and out of chair and bed with little, if any, assistance. May use wheelchair for some indoor activities at home and school. Wheelchair is used for all activities in community.
Non-functional ambulators Patients walk during therapy session at home, in school, or in hospital. Wheelchair used for all other transportation.
Non-ambulators Patients are mobile only via a wheelchair but usually can transfer from chair to bed.

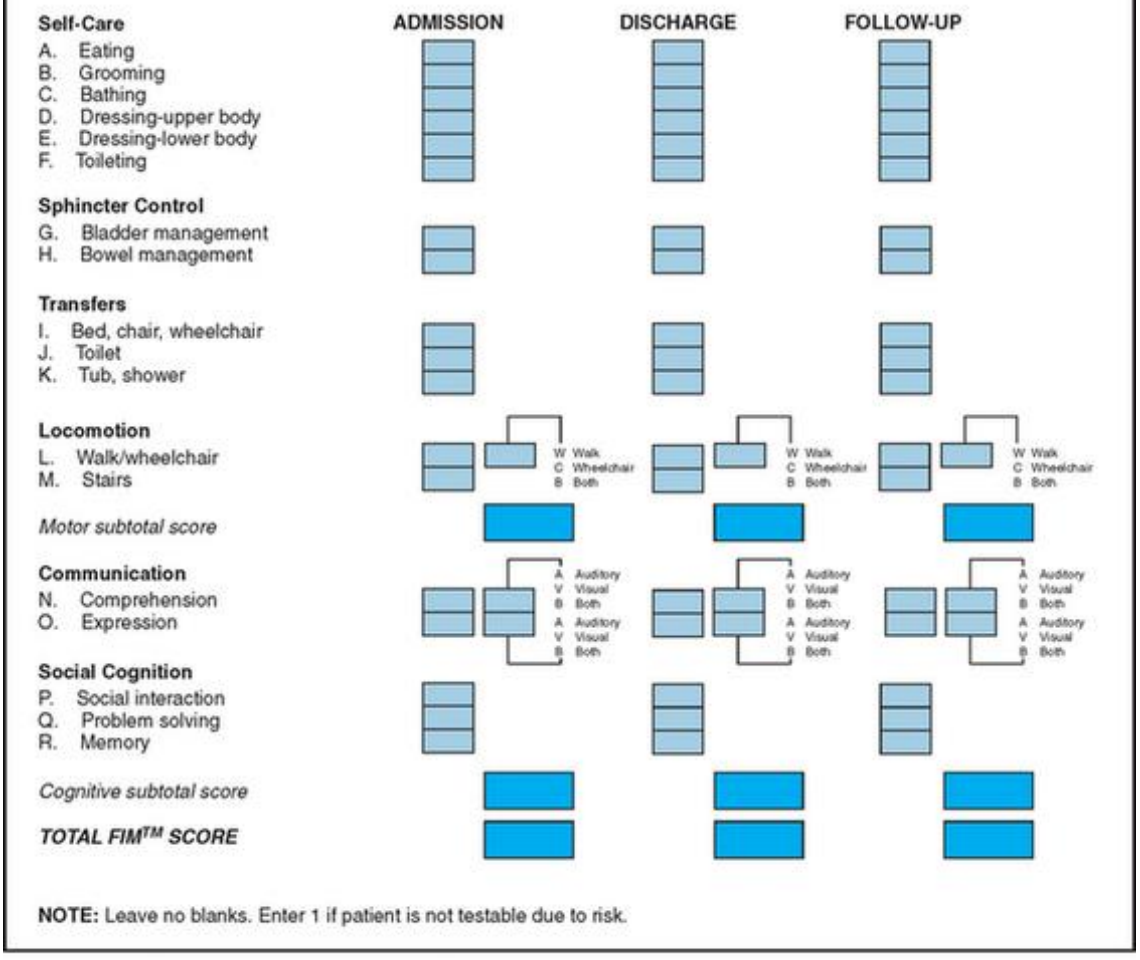
1.5 The Functional Independence Measure (FIM) instrument:

The Functional Independence Measure (FIM), an 18 item tool has been used in patients with spinal cord injury.[30] The FIM can be used as an observational instrument or as a questionnaire with strong correlation between both scores.[31] Although the FIM has been used to measure functional independence in patients with spina bifida, a few studies have revealed shortcomings of this instrument in terms of locomotion and sphincter control scales.[32, 33, 34]

Table 3: Functional Independent Measurement Instrument

FIM™ Instrument

LEVELS		
	7 Complete independence (timely, safely) 6 Modified independence (device)	NO HELPER
	Modified dependence 5 Supervision (subject = 100%) 4 Minimal assistance (subject = 75%+) 3 Moderate assistance (subject = 50%+) **Complete Dependence** 2 Maximal assistance (subject = 25%+) 1 Total assistance (subjet = less than 25%)	HELPER



NOTE: Leave no blanks. Enter 1 if patient is not testable due to risk.

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1.6 The Myelomeningocele Severity Scale:

A spina bifida severity scale (Table 3) adaptable for the local setup has been developed and used at the Bethany Kids Kijabe Hospital. It incorporates the aforementioned factors and considers in presence or absence of kyphus deformity, hydrocephalus, ventriculitis, syndromic features and chiari symptomatology. The development of this scale was necessary due to the late presentation of spina bifida for repair in the African setup.

Table 4: Myelomeningocele Severity Scale

FACTORS AFFECTING SEVERITY / QoL	**SCORE**
MOTOR	
FOOT PLANTAR FLEXION	1
FOOT DORSIFLEXION	2
KNEE FLEXION	3
KNEE EXTENSION	4
HIP FLEXION	5
PARAPLEGIA	6
KYPHUS DEFORMITY	
ABSENT	1
PRESENT	4
HYDROCEPHALUS	
ABSENT	1
PRESENT	6
VENTRICULAR CSF	
WBC <5	1
WBC 5-10	2
WBC >10	3
BLADDER	

CIC NOT NEEDED	1
CIC NEEDED	3
SYNDROMIC FEATURES	
ABSENT	1
PRESENT	3
SYMPTOMS OF CHIARI II	
ABSENT	1
PRESENT	5

1.7 Statistical Validation Techniques

Validation, according to the American Educational Research Association refers to the process of systematically collecting evidence to provide justification for the set of inferences that are intended to be drawn from scores yielded by an instrument. In validation studies, researchers seek to provide one or more of three types of evidences: content-related validity (i.e., the extent to which the items on an instrument represent the content being measured), criterion-related validity (i.e., the extent to which scores on an instrument are related to an independent external/criterion variable believed to measure directly the underlying attribute or behavior), and construct-related validity (i.e., the extent to which an instrument can be interpreted as a meaningful measure of some characteristic or quality).[38]

CHAPTER TWO

STATEMENT OF THE RESEARCH QUESTION AND JUSTIFICATION

2.1 Research Question

Can the proposed spina bifida severity scale be used as a prognostication tool?

2.2 Justification

Controversy exists as to whether all spina bifida patients should be managed aggressively or only those with a few severity markers at admission. A validated severity scale would inform clinical decision making in this subset of patients through quality of life and survival prediction.

A modified spina bifida scale, tailored to the unique African scenario (late presentation, higher infection rates at the outset and high-level of spine involvement) is important in this endeavour. The Kijabe hospital, for the years 2010 - 2014 received a large volume of MMC patients of about 200-300 cases within these 4 years. Data collection was therefore possible.

2.3 Objectives

2.3.1 Main objective

To validate the proposed spina bifida severity scale as a prognostication tool.

2.3.2 Specific objectives

1. To use the proposed Spina Bifida Severity Scale to predict 3-year survival of children with myelomeningoceles.
2. To evaluate the QoL outcomes of children with spina bifida at 3 years of age.

CHAPTER THREE

MATERIALS AND METHODOLOGY

3.1.1 Study Design

Cross-sectional Institutional study

3.1.2 Study Site

Kijabe Hospital, a dedicated pediatric neurosurgery unit caring for a large number of spina bifida cases in Kenya. On average, 200-300 cases are treated per year for the 3 years specified.

3.1.3 Study Population

All patients surgically managed for myelomeningocele between the years of 2010 ï 2014. During this duration, two pediatric neurosurgeons supervised the closure of myelomeningoceles using a standardized technique.

3.1.4 Study Endpoint

Survival at 3years post MMC repair was the primary endpoint.

The secondary endpoint was mobility at the 3 year mark.

3.2.1 Inclusion Criteria

Patients admitted and managed surgically at this institution.

Children, at least 3years following spina bifida repair.

3.2.2 Exclusion Criteria

1. Patients who died before intervention or those for whom MMC closure or VP shunting was performed at another hospital prior to presentation at the Kijabe Mission Hospital.

2. Charts with incomplete data.
3. Children with spina bifida and split cord malformation with asymmetric clinical lower extremity motor function.

3.7 Sample Size

The sample size was determined using the Fisher formula given below.

$$n = \frac{z^2 (p) (q)}{\alpha^2}$$

Where:

n = desired sample size

p = prevalence based on a similar study (Bowman R.M, McLone D.G, Grant J.A, Tomita T, Ito J.A. Spina Bifida outcome: a 25 year prospective. Pediatric Neurosurgery. 2001; 34(3), 114-120.

z = confidence level set at 95%

q = 1 ï p

α = level of significance set at 0.05

Therefore:

$$n = \frac{(1.96^2) \times 0.1 \times 0.9}{0.0025}$$

n = 138

The study was able to exceed the minimum required sample size by 15, giving a total of 153 observations.

3.8 Sampling Method

A serial sampling technique was used. Patient charts of children at least 3 years post MMC closure were scrutinized.

3.9 Ethics and Confidentiality

Approval to carry out the study was obtained from the Kijabe Hospital Research and Ethics committee.

Confidential records were maintained and client identification made anonymous.

3.10 Data Collection

Following ethical approval by the Kijabe Hospital institution, a database search was made to identify all myelomeningocele patients admitted between the months of August 2010 ï December 2014. Charts were retrieved and data concerning the demographics, level of lesion, presence or absence of a kyphus deformity, hydrocephalus and / or Symptomatic Chiari II were be obtained.

A pre-intervention score was assigned using the proposed SB scale. A SBNS score was also assigned based on the preoperative score, in order to further validate the modified score against this already existing tool. Data from mobile clinics and inpatient charts were reviewed in order to assess milestone achievement.

3.11 Data Analysis

Data has been analysed using the STATA 14 statistical research analysis program.

Validation for motor outcome was done through a criterion-related technique, i.e. by comparing the KS ambulatory score to the SBNS score for patients in this study. The Pearson Correlation and Spearmanôs Rho statistical functions were applied.

Validation for survival was achieved through a construct-related technique i.e. by drawing conclusions on survival data.

The proposed myelomeningocele severity scale was stratified into low, intermediate and high risk categories. Kaplan Meier survival estimates were obtained for each risk group category.

The results have been presented in histograms, pie charts and tabulated format.

CHAPTER FOUR

RESULTS

4.1 Results

Statistical analysis for the proposed SB scale was performed using the STATA® 14 program.

4.1.1 Descriptive Statistics

Descriptive data are summarized in Table 5.

Of the 153 observations made the mean score recorded was 14.8. The minimum and maximum scores were 7 and 24 respectively.

Table 5: Descriptive statistics

```
. summarize ks
```

Variable	Obs	Mean	Std. Dev.	Min	Max
ks	153	14.81699	4.60897	7	24

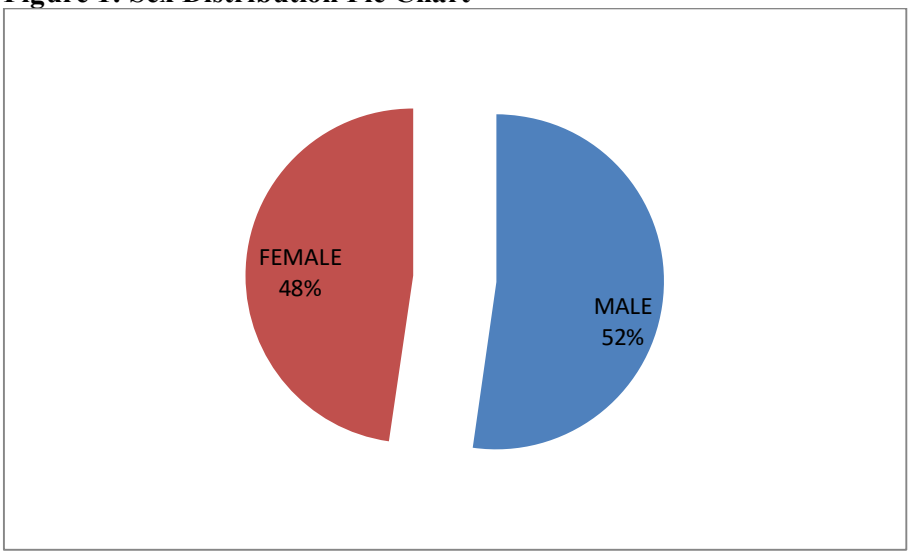
```
. summarize ks, detail
```

KIJABE SCORE

Percentiles		Smallest		
1%	7	7		
5%	7	7		
10%	8	7	Obs	153
25%	11	7	Sum of Wgt.	153
50%	14		Mean	14.81699
		Largest	Std. Dev.	4.60897
75%	19	24		
90%	22	24	Variance	21.2426
95%	22	24	Skewness	.0371908
99%	24	24	Kurtosis	2.200611

A total of 80 males (52%) and 73 females (48%) were studied, yielding a male-to-female ratio of 1.08. Figure illustrates this finding in a pie chart format.

Figure 1: Sex Distribution Pie Chart



4.1.2 Motor Outcome

In order to validate the proposed SB scale as a predictor of motor outcome, a Pearson correlation, p-value and Spearmanôs rho statistical tests were applied.

The application of Pearson correlation between the new scale and the SBNS yielded a negative coefficient of -0.74 (p value < 0.001, Spearmanôs rho = -0.73). The findings are summarized in table 6.

Table 6: Pearson Correlation Statistic

```
. pwcorr ks sbns, sig

             |       ks     sbns
-------------+------------------
          ks |   1.0000
             |
             |
        sbns |  -0.7351   1.0000
             |   0.0000
```

4.1.3 The Spearman's Rho calculation:

The Spearmanôs Rho test matched the findings of the Pearsonôs Correlation Coefficient.

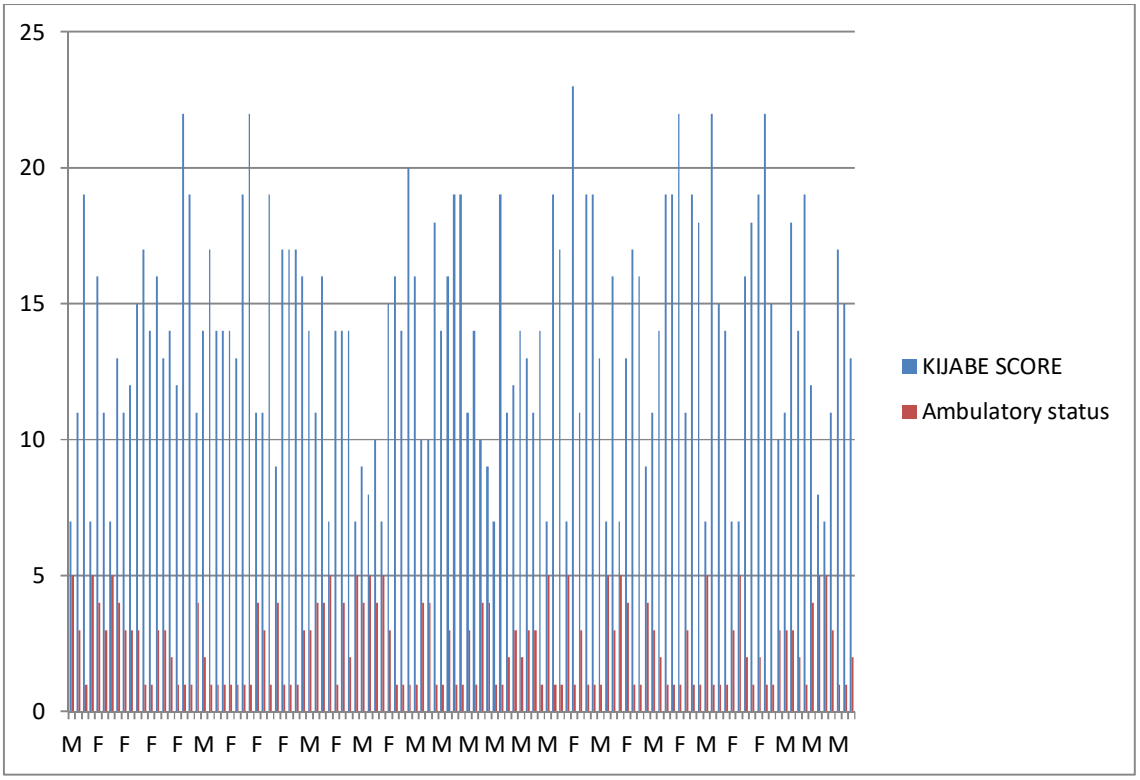
```
. spearman ks sbns

 Number of obs =     153
Spearman's rho =      -0.7329

Test of Ho: ks and sbns are independent
    Prob > |t| =       0.0000
```

A summary of the correlation between the proposed Spina Bifida Severity Scale and mobility is illustrated in Figure 2. The highest peaks on the Kijabe Score correlate with the lowest peaks on the ambulatory status.

Figure 2: Ambulatory Status Histogram



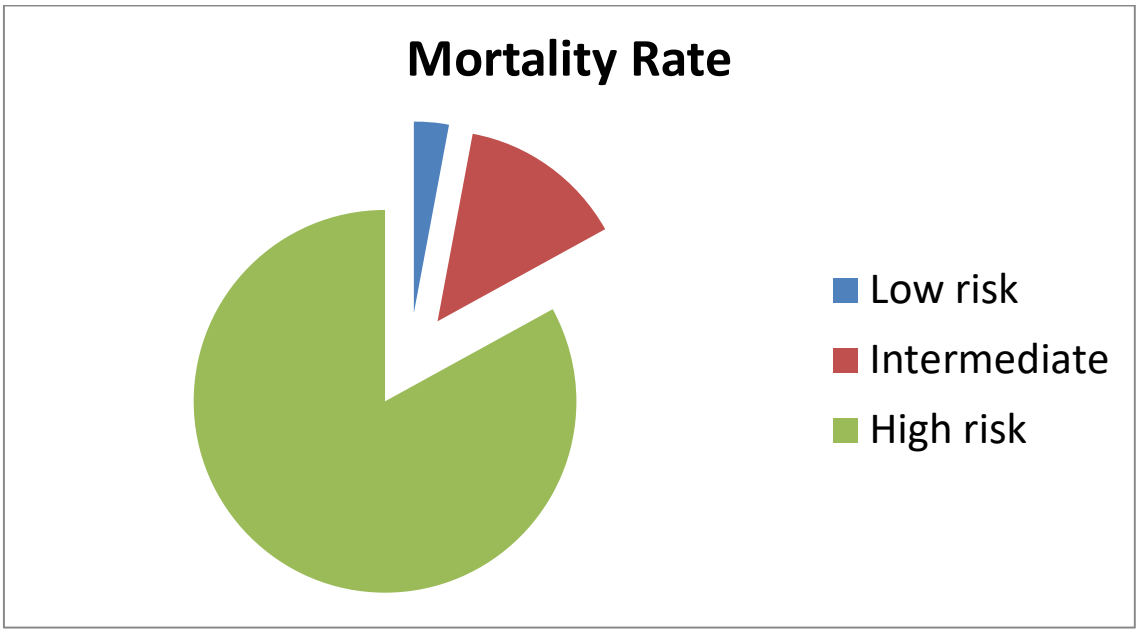
4.1.4 Survival Curves

The three year mortality rate was 15% (24 patients). The proposed SB scale was stratified into 3 risk groups; low (7-11), intermediate (12-20) and high (21-30). The mortality rate according to the risk group is tabulated in table 7 and illustrated in figure 3.

Table 7: Mortality rate according to risk group category

Low risk group	2.3% (1/42 patients)
Intermediate risk	11% (10/91 patients)
High risk	65% (13/20 patients)

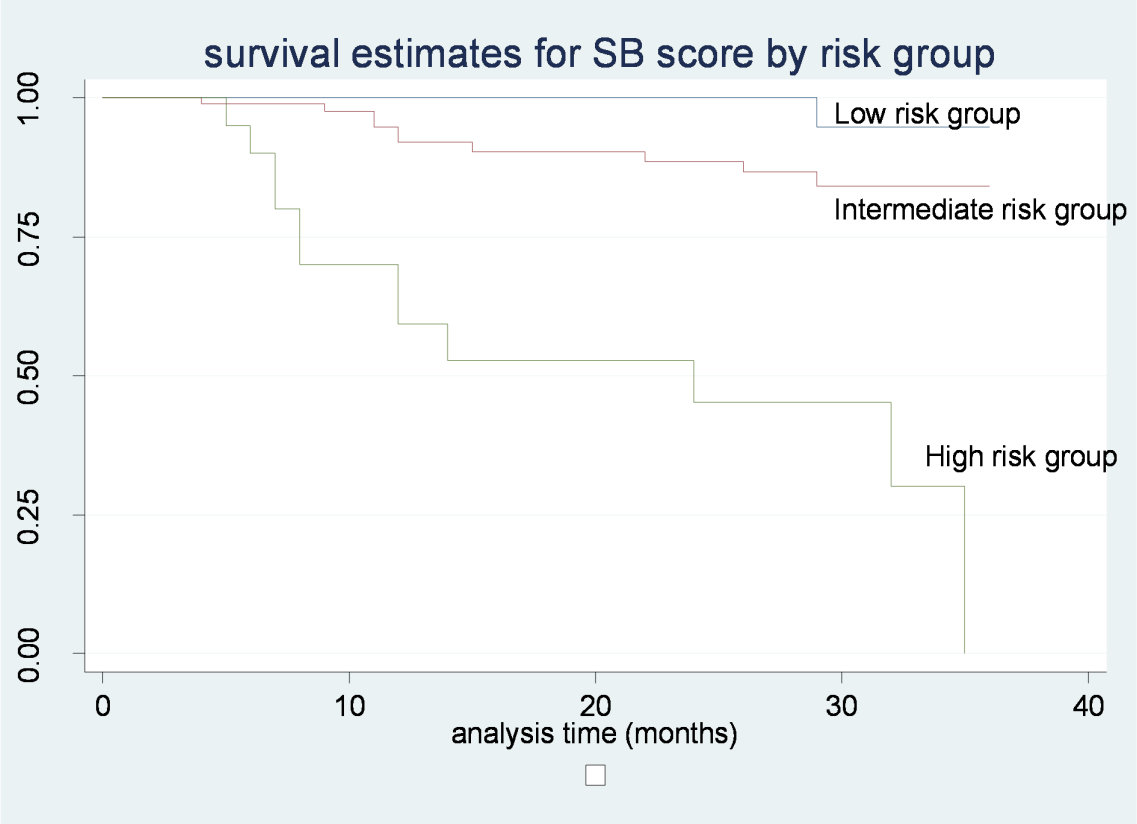
Figure 3: Mortality Rate



Low and intermediate risk groups showed better survival estimates as opposed to the high risk category, as summarized in the Kaplan Meier Survival Curves in figure 4. A single

mortality occurred in the low risk category, in the third year of the study period. Mortality within the intermediate risk group is evenly distributed over the three years as indicated in the survival curves. Sixty seven per cent (67%) of deaths within the high risk category occur by the end of the one year follow-up point.

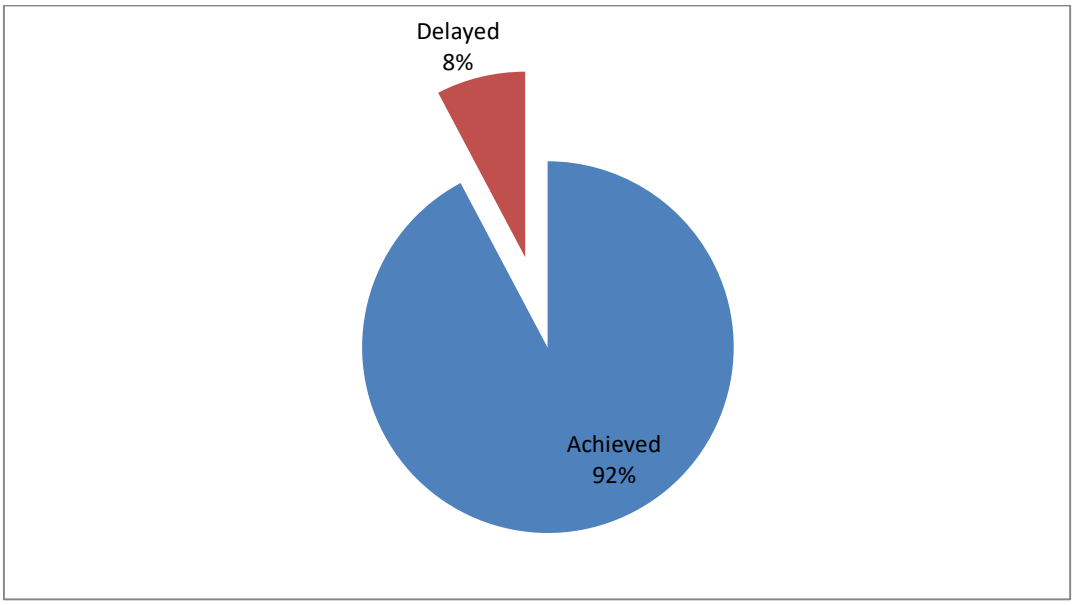
Figure 4: Survival Curves



4.1.5 Milestone Achievement

There was no identifiably correlation between the proposed scale and achievement or delay of milestones. Of the 129 patients still alive at the 3 year endpoint, 119 (92%) achieved their milestones in a timely fashion as recorded using the modified developmental milestone charts for age. Figure 5 illustrates this.

Figure 5: Percentage of milestone achievement pie chart



CHAPTER FIVE

DISCUSSION

5.1 Discussion

This study has validated a myelomeningocele severity scale as a tool that prognosticates survival 3 years following MMC closure. The severity scale also predicts future ambulatory status; however, it cannot predict the likelihood of achieving milestones.

Data from 153 patients were collected and analysed. Each patient was ascribed a KS score derived from the new myelomeningocele severity scale. This severity scale is a 30 point score that assesses an MMC infant based on motor function, kyphus deformity, presence of hydrocephalus, ventricular CSF WBC count, CIC necessity, and presence or absence of syndromic features and symptomatic chiari I. The minimum score attained was 7, while the maximum score was 24. Fifteen of 153 patients (9.8%) had a KS score of 7; these mark the proportion of infants at the lowest end of the spectrum. None of the patients achieved the maximum possible score of 30, at the highest end of the MMC severity spectrum.

The male-to-female ratio in the study population was 1.08 (52% male and 48% female). In comparison with a study done at the Kenyatta National Hospital by Mwangôombe et al, 61.5% of the spina bifida patients were male and 38.5% female. The male-to-female ratio in this case was 1.59. The differences in the ratio may be explained by the vast difference in the study population (65 patients versus 153 patients) and because 2012 study included all spina bifida patients, whereas this current study includes only with myelomeningocele.

Ambulatory status was validated by comparing the myelomeningocele severity scale against the SBNS. This comparison was done through applying the Pearson Correlation statistic and the Spearmanôs Rho test. The Pearson Correlation test yielded a result of -0.74 (p value <0.001) in this study. The negative correlation coefficient obtained indicates that the higher a

score is on the myelomeningocele scale the lower it will be in the SBNS, in a predictable fashion. The p value is statistically significant (<0.001). The Spearmanôs Rho statistically counterchecks the Pearson Correlation. In this study the Spearmanôs Rho was -0.73. The value matches that of the Pearson Correlation, indicating that the result is reliable.

The highest KS scores on Figure 2 correlate with the lowest ambulatory status scores. Appendix III indicates that a mobility score of I corresponds with normal motor function, while an ambulatory score of V indicates that an individual is wheelchair bound. This means that the patients on the higher end of the severity scale are less likely to be mobile and more likely to be wheelchair bound in the future.

The 3 year mortality rate in the present study was 15% (24 of 153) patients. The mortality rate according to risk group category was 2.3% in the low risk group, 11% in the intermediate risk group and 65% in the high risk group. It is clear that the higher the KS score on the myelomeningocele severity scale, the greater the risk of mortality by the 3 year endpoint.

Kaplan Meier estimates derived from the data in this study derive 3 distinct survival curves, one for each risk group category. More than 50% of the deaths in the high risk group occur before the end of the first year of life, whereas mortality in the intermediate risk group are evenly spread out over the 3 year study period. The mortality within the low risk group occurred within the third year of follow-up. Therefore, infants who score between 21 and 30 on the myelomeningocele severity scale not only have a 65% risk of dying, but that these deaths will likely occur within the first year of life.

Milestone achievement did not follow a predictable pattern. Of the 129 patients still alive at the 3 year endpoint, 119 (92%) achieved cognitive and behavioural milestones as recorded using the modified developmental milestone charts for age. This indicates that regardless of

severity as scored by the new scale, a child who survives has a good chance of experiencing quality of life as measured by cognitive and behavioural milestone achievement.

5.2 Study Limitations

The study aims to prognosticate spina bifida in the short and intermediate term. Long term follow up will be recommended for more comprehensive results.

The study does not consider the strength of individual severity markers. Cox regression analysis would need to be performed in a future study.

5.3 Conclusion

The myelomeningocele severity scale is reliable as a survival prognosticator at 3 years post MMC closure. It is also as reliable as the already existing SBNS scale for predicting the motor outcome at 3 years post MMC repair. The myelomeningocele severity scale therefore meets the objectives of predicting survival and future ambulatory status.

The scale is generalizable across the country and African sub-continent on account of similarities in patient profile and predominance of infection rates.

The validated severity scale offers a new benchmark for MMC patient care because the clinician learns to anticipate associated comorbidity such as symptomatic chiari II, ventriculitis, hydrocephalus and urinary tract infections.

The scores and stratification into low, intermediate and high risk categories clarify the fact that MMC can be viewed as a disease spectrum. It will therefore affect different individuals differently.

Infants falling within the high risk category have the highest risk of mortality and this risk is greatest within the first year of life.

The new scale does not predict milestone achievement, however, an infant who survives upto and beyond the 3 year mark has a good chance of experiencing quality of life as measured by cognitive and behavioural milestone achievement.

Patients on the higher end of the MMC spectrum are less likely to be mobile and more likely to be wheelchair bound in the future.

5.4 Recommendations

Care-givers should be informed that patients in the high-risk category have a high risk of death regardless of intervention.

Clinicians should tailor the surgical management of MMC infants based on risk group stratification, because the spina bifida entity is a spectrum with varying levels of severity. Infants in the high risk category will likely require multiple surgeries as opposed to those in the low risk group category.

Local hospital should set up protocols to standardize the management of spina bifida patients. For instance, the protocol could dictate when a ventricular tap will be obtained, the timing and frequency of renal function tests and bladder evaluation.

Antibiotic protocols need to be formulated in view of the anticipated risk of ventriculitis, CSF shunt infections and urinary tract infections in some MMC infants.

Folic acid supplementation policies need to be formulated at a national level in order to lower the incidence of spina bifida affected pregnancies and births.

A longer duration of follow-up would offer further enlightenment on the applicability of the new spina bifida severity scale in older patients.

The validated myelomeningocele severity scale should be adopted in neurosurgical clinics in sub-Saharan Africa where the health challenges are similar Kenya, where the study was conducted.

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APPENDICES

Appendix I: Myelomeningocele Severity Scale

FACTORS AFFECTING SEVERITY / QoL	SCORE
MOTOR	
FOOT PLANTAR FLEXION	1
FOOT DORSIFLEXION	2
KNEE FLEXION	3
KNEE EXTENSION	4
HIP FLEXION	5
PARAPLEGIA	6
KYPHUS DEFORMITY	
ABSENT	1
PRESENT	4
HYDROCEPHALUS	
ABSENT	1
PRESENT	6
VENTRICULAR CSF	
WBC <5	1
WBC 5-10	2
WBC >10	3
BLADDER	
CIC NOT NEEDED	1
CIC NEEDED	3
SYNDROMIC FEATURES	
ABSENT	1
PRESENT	3
SYMPTOMS OF CHIARI II	
ABSENT	1
PRESENT	5

Appendix II: Spina Bifida Neurological Scale

Functioning	C-Th	L1	L2	L3	L4	L5	S1
Nonfunctioning	L2		L3	L4	L5	S1	S2
Hip							
Flexion	-		+/-	+	+	+	+
Extension	-		-	-	-	-	+/-
Adduction	-		+/-	+	+	+	+
Abduction	-		-	-	-	_	+
Knee							
Extension	-		+/-	+	+	+	+
Flexion	-		-	-	-	+/-	+
Ankle							
Dorsiflexion	-		-	-	+/-	+	+
Plantarflexion	-		-	-	-	-	+/-
Inversion	-		-	-	+/-	+	+
Evertion	-		-	-	-	-	-
SBNS motor (worse side)	1		2	3	4	5	6

Scoring Scale for motor function

Functioning	C-Th	L3	L4	L5	S1	S2	S3
	~L2						
Nonfunctioning	L3	L4	L5	S1	S2	S3	S4
Patellar reflex	-	+/-	+	+	+	+	+
Achilles reflex	-	-	-	+/-	+	+	=
Anal reflex	-	-	-	-	_	+/-	+/-
SBNS reflex	1	2	2	3	3	4	4

Scoring system for preserved reflexes

Functioning	C-Th	S2	S2				S3	S3
	L-S1							
Nonfunctioning	S2	S3	S3				S4	S4
Bladder control	-	-	+/-	+	+/-	+	+	+

Bowel control	-	+/-	-	+/-	+	+/-	+	+
SBNC BB control	1	2	2	3	4	4	5	5

Scoring system for bladder and bowel function

Appendix III: Motor Scale

I	Normal
II	One crutch only
III	Two crutches only
IV	Two crutches + orthoses
V	Wheelchair bound

Appendix IV: Modified Developmental Milestone Charts

3 YEAR OLDS

Social / Emotional	Copies adults and friends
	Shows affection for friends without prompting
	Takes turns in games
	Shows concern for a crying friend
	Dresses and undresses self
	Understands the idea of ñmineò and ñhisò or ñhersò
	Shows a wide variety of emotions
	Separates easily from mom and dad
	May get upset with major changes in routine
Language / Communication	Follows instructions with 2 or 3 steps
	Can name most familiar things
	Understands words like ñin,ò ñon,ò and ñunderò
	Says first name, age and sex
	Names a friend
	Talks well enough for strangers to understand most of the time
	Says words like ñI,ò ñme,ò ñwe,ò and ñyouò and some plurals (cars, dogs, cats)
	Carries on a conversation using 2 to 3 sentences
Cognitive	Can work toys with buttons, levers, and moving parts
	Plays make-believe with dolls, animals and people
	Does puzzles with 3 or 4 pieces
	Understands what ñtwoò means

4-YEAR OLDS

Social / emotional	Enjoys doing new things
	Is more and more creative with make-believe play
	Would rather play with other children than by himself
	Cooperates with other children
	Plays ñMomò or ñDadò
	Often canôt tell whatôs real and whatôs make-believe
	Talks about what she likes and what she is interested in
Language / communication	Tells stories
	Sings a song or says a poem from memory
	Knows some basic rules of grammar, such as correctly using ñheò and ñsheò
	Can say first and last name
cognitive	Names some colors and some numbers
	Understands the idea of counting
	Starts to understand time
	Remembers parts of a story
	Understands the idea of ñsameò and ñdifferentò
	Draws a person with 2 to 4 body parts
	Uses scissors
	Starts to copy some capital letters
	Plays board or card games
	Tells you what he thinks is going to happen next in a book

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20TH AUG 2O15

DR Grace Muthoni Thiongo,

RE : VALIDATION OF A SPINAL BIFIDA SEVERITY SCALE

The institutional review board having carefully reviewed your above title proposal grant you approval to conduct this study at Kijabe hospital.

We look forward to receiving the results of the interim analysis.

We wish you all the best in the study. Kindly furnish this office with a copy of your results.

Thank you,

Sincerely,

Leland Albright, MD

Chair, Kijabe Hospital IRB

"Health Care to God's Glory"