

**EPIDEMIOLOGICAL CHARACTERISTICS OF PATIENTS
WITH MYELOMENINGOCELES PRESENTING TO
UNIVERSITY TEACHING HOSPITAL - LUSAKA**

By

MARTHA MWEWA LUNGU, B.Sc. (HB); MB ChB (UNZA).

*A dissertation submitted in partial fulfillment of the
requirements for the award of Master of Medicine
(Surgery) degree of the University of Zambia*

UNIVERSITY OF ZAMBIA

SCHOOL OF MEDICINE

September 2004



Thesis
M. Med
Lun
2004
C.1

APPROVAL FOR EXAMINATION

This Dissertation is ready for examination

Date 30/03/05 Supervisor T. Lambart

Mr. T.K. LAMBART

SUPERVISORS PAGE

The Dissertation of Dr Martha Mwewa Lungu is ready for examination.

Signed (Supervisor).....

Signature

MR T. K. LAMBART

Honorary Lecturer

School of Medicine

University of Zambia

Signed (Co supervisor).....

Signature

PROFESSOR ODIMBA

Honorary Professor

School of medicine

University of Zambia

Approval

The University of Zambia approves this dissertation by Dr Martha Mwewa Lungu in partial fulfilment of requirements for the award of the degree of the Master of Medicine in General Surgery.

Signature

Date

.....

.....1/6/2005

.....

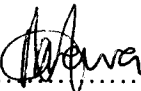
.....21/5/05

.....

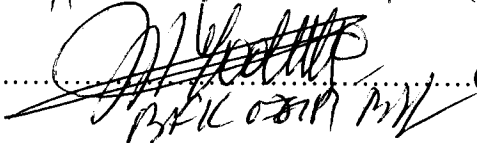
.....31/05/05

DECLARATION

I hereby declare that this dissertation herein presented for the degree of Master of Medicine (Surgery) has not been previously submitted wholly or in part for any other degree at this or any other university, nor is it being currently submitted for any other degree.

Signed.......... (Candidate)

Signed.......... (Supervisor)

Signed..... (Co-supervisor)

DEDICATION

This work is dedicated to the following people without whose assistance and encouragement this work would not have been completed: my husband, Dr Shadrick Lungu who believed that I could do it and looked after our children. My children whom I seldom saw during this period of study/research and who, I am certain, wondered why mummy was not visiting so frequently. I do not forget to thank both Mr. Lambart and Professor Odimba for the patience shown when I “troubled” them for direction and advice whenever I got stuck. Above all I thank the Almighty God for seeing me through all this work.

ABBREVIATIONS

Abbreviations

ANC	ANTENATAL CLINIC
NTD	NEURAL TUBE DEFECT
UTH	UNIVERSITY TEACHING HOSPITAL
USA	UNITED STATES OF AMERICA
MA	MASSACHEUSETTES
NY	NEW YORK
CDC	CENTRES FOR DISEASE CONTROL
M: F Ratio	MALE: FEMALE RATIO

TERMINOLOGIES

Encephalocoele

Protrusion of a variable amount of meninges and brain parenchyma through a defect in the cranial bones.

Meningocoele

Closely related to myelomeningocoele but less severe form of spinal defect in which only meninges herniated through abnormally formed vertebral arches.

Myelomeningocoele

A neural tube defect characterized by herniation of meninges and spinal cord through a posterior vertebral defect.

Neural Tube Defect

Developmental aberrations related to abnormal closure of the neural tube. Includes myelomeningocoele, meningocoele, encephalocoele, and anencephaly.

Spina Bifida

Spinal neural tube defects where there is an absence or hypoplasia of one or more vertebral arches, with variable abnormalities in the underlying meninges and/or spinal cord.

TABLE OF FIGURES

Figure		Page
Figure 1	Bar graph showing prevalence of myelomeningocoeles between 1994 and 2002	23
Figure 2	Pie chart of sex distribution of myelomeningocoeles	25
Figure 3	Pie chart of socio-economic status of myelomeningocoeles	26
Figure 4	Bar graph of maternal education	27
Figure 5	Table showing comparison of maternal tribe and myelomeningocoele	28
Figure 6	Table showing sites of occurrence	29
Figure 7	Table showing province of origin	30
Figure 8	Pie chart showing folic acid consumption	31
Figure 9	Table showing antenatal visits.	32
Figure 10	Graph showing maternal age distribution	33
Figure 11	Pie chart showing parental occupation	34

TABLE OF CONTENTS

Subject	Page
1. Introduction.	1
2. Abstract	2
3. Objectives	4
4. Rationale	5
5. Literature Review	6
6. Patients and Methods	21
7. Results (Graphs and Tables)	23
8. Discussion	35
9. Conclusion and recommendations	39
10. References	41
11. Photograph	46
12. Appendices: Appendix 1: Consent form	
Appendix 2: Unified questionnaire	

Introduction

Myelomeningocele is a complex malformation of the spinal cord, nerve roots, meninges, vertebral bodies and skin. This neural tube defect is a common congenital anomaly and typically referred to as spina bifida. It results from failure of the neural tube to close in the developing fetus (1,2). Medical, surgical and rehabilitation issues arise in the patient with myelomeningocele from birth through adulthood. Ninety per cent (90%) of these patients also have hydrocephalus. It also occurs with multiple system congenital anomalies e.g. facial clefts, heart malformations and genitourinary tract anomalies (3). Mortality rates reported for infants who are untreated for myelomeningocele range from 90-100% based on several series of studies dating from the turn of the century through recent years. Most untreated infants die within the first year of life. Death in the first 2 years of life for those untreated usually results from hydrocephalus or intracranial infection. The likelihood that a 2-month-old infant untreated for myelomeningocele lives to be 7 years is only 28% (3).

ABSTRACT

This study was carried out to highlight epidemiological characteristics that children with cranio – spinal dysraphism such as myelomeningocoeles presenting to the University Teaching Hospital in Lusaka had. Records of patients presenting to the University Teaching hospital from June 1994 to December 2002 were looked at and the data noted. Then a prospective study was carried out over a period of 10 months starting in August 2003 to June 2004. The details of how this was undertaken are described below. A total of fifty (50) patients were seen from August 2003 to June 2004. There were twenty-eight (28) females and twenty-two (22) males. Of these, thirty-six (36) had myelomeningocoeles, and encephalocoeles and meningocoeles were seven patients each respectively. Their socioeconomic status was predominantly low (86,1%). Folic acid intake by their mothers was 51.4%. The maternal age ranged was from 15 to 40 years. The average gestational month at the first ANC attendance was 4 (four) months. The educational standard of these mothers was mostly primary school education (45.7%). The maternal ethnic pattern for myelomeningocoeles was highest in the Bembas (20.0%) and seconded by the Tonga's (17.1%) whereas the Tonga tribe led overall for all the neural tube defects noted. Most patients with myelomeningocoeles were

from within Lusaka (38.9%). The sacral area was the most commonly affected site for myelomeningocele (36.1%).

OBJECTIVES

- a) To study the prevalence of myelomeningocoeles at UTH
- b) To study the disease distribution of myelomeningocoeles at UTH
- c) To determine the ethnic pattern of myelomeningocoeles.
- d) To determine the most prevalent anatomical site

RATIONALE

Myelomeningocele is one of the serious neural tube birth defects. Most children with myelomeningoceles are referred to the University Teaching Hospital, Paediatric Unit (D-Block) for specialist attention from all around Zambia. This condition presents a challenging problem with regard to clinical management. Considering the limited capacity, for-example, manpower trained to deal with this condition, it is worth trying to take preventive measures in reducing the occurrence of these defects. Preventive measures can only be started once the characteristics of the problem are identified. The rates of myelomeningoceles vary widely among countries and by geographical regions within countries (3,18). To reduce the occurrence of the birth defects (Neural Tube Defects), the Food and Drug Administration of USA authorized the fortification of all enriched cereal grain products with folic acid in March 1996, with compliance mandatory by January 1998 (6). During 1996 to 2001 a 23% decline occurred in neural tube defects (spina bifida and anencephaly combined) (6). Spina bifida declined by 24% during this period, and anencephaly declined by 21%. The United States has experienced declines in spina bifida and anencephaly cases since folic acid fortification of all enriched cereal grain products (6). The observed declines have translated into approximately 920 infants being born without these serious defects each year (6).

LITERATURE REVIEW

Myelomeningocele is the most common physically disabling birth defect in humans. It is caused by the failure of the neural tube to close and is most common in the lumbosacral area (75%) (1,2).

Myelomeningocele often occurs with multiple system congenital anomalies. Commonly associated anomalies are facial clefts, heart malformations and genitourinary tract anomalies (3,19).

Pathophysiology

Myelomeningocele is the result of a teratogenic process causing failed closure and abnormal differentiation of the embryonic neural tube during the first 4 weeks of gestation. Abnormal development of the posterior caudal neural tube produces spinal cord damage or myelodysplasia. The anatomic level of the spinal cord lesion roughly correlates with the patient's neurologic, motor, and sensory deficits (3). Abnormal development of the cephalic anterior tube gives rise to central nervous system anomalies. The Arnold Chiari Type 2 malformation impedes the flow and absorption of cerebrospinal

fluid and causes hydrocephalus, which occurs in more than 90% of infants with myelomeningocele (3).

Aetiology

In most cases the aetiology is multifactorial, involving genetic, racial, and environmental factors.

- Other offspring in a family with one affected child are at increased risk of a neural tube defect than children without affected siblings.
- The risk is 1 in 20 - 30 for subsequent pregnancies, and if 2 children are affected, the risk becomes 1 in 2.
- Most infants born with myelomeningocele are born to mothers with no previous affected children.
- A small number of cases are linked to specific aetiological factors.

Up to 10% of fetuses with a neural tube defect detected in early gestation have an associated chromosome abnormality.

Maternal risk factors include insulin dependant diabetes mellitus and hypothermia (3).

Intrauterine drug exposures to valproate, carbamazepine, and drugs to induce ovulation are identified risk factors (3,19).

- Prior to 1991, most neural tube defects occurred without identification of a specific cause.
- Research in the 1980s showed correction of folic acid deficiency as an effective means of primary and recurrent prevention.

After 1991, 50% of cases of neural tube defects are related to a nutritional deficiency of folic acid and thus, are preventable.

In September 1992, the US Public health Service (USPHS) recommended intake of folic acid at a dosage of 0.4mg/d for all women anticipating pregnancy.

In February 1996, the USPHS announced mandatory folic acid fortification of enriched cereal grain, a measure expected to increase the daily intake of folic acid in women of reproductive age by approximately 100mcg/d (3).

Frequency

In the USA, myelomeningocele is the most common major birth defect. Birth prevalence of the disease was reported to be 4.4 - 4.6 cases per 10 000 live births from 1983 - 1990. The highest estimated rates of myelomeningocele in the USA are found in Appalachia.

Internationally, the rates of myelomeningocoeles vary widely among countries and by geographic regions within countries. In the last 50 years epidemics of myelomeningocoeles have occurred in Boston, MA; Rochester, NY; Dublin, Ireland; the Peoples Republic of China and Jamaica. Lower social economic status is associated with higher risk in many populations (3,18,19).

Mortality rates reported for infants who are untreated for myelomeningocoele range from 90 to 100% based on several series of studies dating from the turn of the century through recent years (3).

Here in Zambia there is no documentation, except for a survey done by Dr L. Chikoya and the study on encephalocoeles by Professor L Munkonge. (14,15). Dr Chikoya noted that a large number of admissions are shared between two provinces in Zambia namely Southern and Northern. This data is unpublished (14). Another study on the management of encephalocoeles over a period of 5 (five) years (1985 - 1989) was conducted at the University Teaching Hospital Lusaka, Zambia. A total of 31 children with encephalocoeles were seen and of these 15 patients (48.4%) came from the maternity wing of the same hospital while 16 (51.6%) were referrals from central and district hospitals of the country. There was a high mortality rate of 81%. (15).

In the retrospective study done from June 1994 to December 2002, a total of approximately 244 (Two hundred and forty-four) cases of neural tube defects were recorded in the admission books. Only sixty (60) files of these were

traced and looked at. Of these, 40 (66.66%) were myelomeningocoeles. Southern province had 24.3% of the referrals of myelomeningocoele. And the Tonga women had the most 8 (40.0%) of myelomeningocoeles. The commonest affected site for myelomeningocoeles was lumbosacral. The sex ratio was 1.10:1 females to males.

Sex

Birth prevalence rate of myelomeningocoele was slightly higher in females than males (1.2:1), based on data from both state and national surveillance systems from 1983 – 1990 (3). A higher proportion of females than males exhibit thoracic - level malformations (3).

Physical

At birth a midline defect in the posterior elements of the vertebrae is noted with protrusion of the meninges and neural elements through an external dural sac.

The obvious physical manifestation of myelomeningocoele is paraplegia caused from spinal cord malformation (3).

A randomized double blind prevention trial with a factorial design was conducted at 33 centers in seven countries to determine whether supplementation with folic acid or a mixture of seven other vitamins (A, D,

B1, B2, B6, C and nicotinamide) around the time of conception can prevent neural tube defects (anencephaly, spina bifida, encephalocoele). A total of 1817 women at high risk of having a pregnancy were allocated at random to one of four groups- namely, folic acid, other vitamins, both or neither. The study showed a 72% protective effect in the group taking folic acid. The other vitamins showed no significant protective effect (4).

Another study was designed to determine whether folic acid also reduces the risk of first occurent neural tube defects. It was a case control study involving tertiary and birth hospitals in Boston, Massachusetts, Philadelphia, Pa, and Toronto, Ontario. The study concluded that daily periconceptional intake of 0.4mg folic acid reduced the risk of occurent neural tube defects by 60% (5).

The decrease generally has been greater for spina bifida than for other neural tube defects (6) The above study was quoted in MMWR (Morb Mortal wkly Rep 1991), which looked at the use of folic acid for the prevention of spina bifida and other neural tube defects between 1983 - 1991 and recommends that daily oral supplementation with folic acid before conception and during early pregnancy substantially reduces the recurrence of neural tube defects (7).

In September 1992 the U.S. Public Health Service recommended that all women capable of becoming pregnant should consume 400mcg of folic

acid/day on an ongoing basis to reduce their risk of having a pregnancy affected by spina bifida and anencephaly. This recommendation was preceded a year earlier by a CDC recommendation for women at high risk (i.e., those women who have had an earlier pregnancy affected by a neural tube defect (6).

The American Academy of Paediatrics endorsed the U.S. Public Health Service recommendation that all women capable of becoming pregnant consumed 400mcg of folic acid daily to prevent neural tube defects (8).

A study conducted by the Births Defects and Genetic Diseases Branch, Centers for Disease Control, Atlanta, Ga. 30333, highlighted the changing epidemiology of neural tube defects in the United States between 1968-1989. They concluded that the declining rates of neural tube defects could be partially explained in particular by the use of multivitamins and folic acid (9).

The National Center on Birth defects and Developmental Disabilities, Centers for Disease Control and Prevention, Atlanta, Georgia 30341, USA, also conducted a study to see what impact these public health policies had on the prevalence of neural tube defects. Twenty-four population – based surveillance systems were used to identify 5,630 cases of spina bifida and anencephaly from 1995-1999. Cases were divided into three temporal categories depending

on whether neural tube development occurred before folic acid fortification (January 1995 to December 1996), during optional fortification (January 1997-September 1998), or during mandatory fortification (October 1998 to December 1999). Prevalences for each birth defect were calculated for each time period. Data were also stratified by programs that did and did not ascertain prenatally diagnosed cases. The results showed that the prevalence of spina bifida decreased 31% from the pre - to the mandatory fortification period (10).

Decline in prevalence of neural tube defects in a high-risk region of the United States was noted in a study conducted over a six-year period (October 1992-September 1998). Active and passive methods were used for surveillance of NTD-affected pregnancies and births during this period. Individual genetic counseling was used to prevent NTD occurrences and a public awareness campaign was used to reduce NTD occurrences. The prevalence for NTDs decreased from 1.89 to 0.95 cases per 1000 live births. The prevalence decrease was explained primarily by decrease in cases of spina bifida. The rate of periconceptional folic acid use among women of childbearing age increased from 8% to 35% during the 6-year project period (11).

The extent to which the decline of spina bifida and other neural tube defects varies by maternal sociodemographic characteristics was undertaken in a study done from data taken from the North Carolina Birth Defects Monitoring Program. This report covered data from 1995 through 1999. The overall prevalence of spina bifida decreased by 27.2%. The magnitude of the decline varied considerably by sociodemographic characteristics of the mother. The decline was greatest among mothers who were aged more or equal to 30 years, who had more than a high school education, whose prenatal care was not paid by Medicaid, and who were non-Hispanic white. Geographically the decrease in the western and Piedmont regions of the state was almost threefold that occurring in the eastern region. The decline in spina bifida after fortification varied considerably by sociodemographic patterns. They concluded that more effort was needed to target folic acid education programs at disadvantaged populations (6).

The association of socioeconomic status, neighborhood social condition and neural tube defects was evaluated. The results showed that both lower socioeconomic status and residence in socioeconomic status – lower neighborhood increased the risk of a neural tube defect affected pregnancy (12). A study of the association of low maternal education with neural tube

defects was studied. It revealed that the single strongest predictor of having a child with a neural tube defect was low maternal education. They suggested that to further reduce the incidence of neural tube defects, interventions should target women of low educational status (13). Another study eliciting patterns by type of defect and maternal race / ethnicity was undertaken from 1990-1994 in California. The study used race / ethnicity - specific prevalence estimates of neural tube defects (NTD'S) in California using 5 years of population based data. They found that Hispanic women had the highest overall rates, followed by whites, Blacks and Asians. Hispanic women were 45% more likely than White women to have a pregnancy affected with anencephaly, while Asian women were over two times less likely to have a pregnancy affected with spina bifida. Considerable variation existed in prevalence of NTD'S by race/ethnicity and by type of NTD, with Hispanic women exhibiting the highest overall NTD rate (16).

Prevalence of spina bifida at birth in the United States, between 1983-1990 was studied by using a comparison of two surveillance systems. Data from birth defects surveillance systems in sixteen states from 1983 –1990 was compared with CDC's Birth Defects Monitoring Program for the same period. From 1983 through 1990, the birth prevalence rate for spina bifida in the 16 states was 4.6 per 10,000 births; the Birth Defects Monitoring Program rate was nearly identical (4.4 cases). State specific rates varied substantially,

ranging from 3.0 (Washington) to 7.8 (Arkansas). Both state based and Birth Defects Monitoring Program rates varied among racial / ethnic groups. In both systems, the rates were highest for Hispanics and lowest for Asians / Pacific Islanders (17).

Prevalence of neural tube defects in Cape Town, South Africa, was studied. The effects of race, gender, maternal age, parity and season of conception on the prevalence was determined. Multiple sources of ascertainment were used, including all maternity hospital records, neurosurgical and spinal defects clinic data, as well as those from the Human Genetics Department and Fetal Abnormality Group. Prevalence rates were highest for the white population group of 2.56 per 1,000 births compared to 0.95 per 1,000 for blacks and 1.05 per 1,000 for those of mixed ancestry. They reported that higher rates in the whites, who were of British and European extraction and belonged to the more affluent section of the community, would suggest that the possible effects of nutrition and infection were overshadowed by genetic factors. There was a female preponderance for both spinal bifida (M: F ratio 0.89) and anencephaly (M: F ratio 0.67). The highest NTD rates were found at both ends of the maternal age range (< 20 years and > 35 years of age). The prevalence was highest at the extremes of birth order (1.65 and 1.58 for birth order 1 and > 7, respectively, and 0.56 and 0.45 for birth order 5 and 6, respectively) (20).

Another study was done in the middle belt of Nigeria to study neural tube defects. Forty-two babies were found to have neural tube defects during a 3-year prospective study in a large cosmopolitan West African city. The commonest defect was meningocele, in 45 per cent of cases. Other defects comprised anencephaly (5 per cent), Arnold-Chiari malformation (7 per cent), encephalocele (19 per cent), occipital meningocele (14 per cent) and spina bifida occulta (10 per cent). More defects were found among females and low birth weight infants among those whose mothers were aged between 20 and 30 years, and in mothers with parity of four and above. Developmental assessments (neurological follow-up status) done at the age of 18 months on five surviving cases of meningocele showed very poor development (21).

Management of spina bifida cystica in Zaria, Nigeria was studied. Over a period of 11 years, 77 children with this defect, 54 meningoceles and 23 myelomeningoceles, 66 (86%) situated in the lumbosacral region, were treated operatively. Forty-two (55%) had surgery in the neonatal period and 91% within 6 months of birth. Postoperative complications occurred in 19 of 68 patients (28%), including mild hydrocephalus, which resolved spontaneously (six, 9%), wound infection (six, 9%), leakage of cerebrospinal fluid (four, 6%) and meningitis (three, 4%). Mortality was 3% from both meningitis and cardiac arrest. Of 32 patients followed up for 3-5 years, 20 with

meningocoeles were normal. Of 12 with myelomeningocoele, four had varying degrees of lower limb weakness, three double incontinence, two faecal incontinence, two had progressive hydrocephalus plus paralysis and double incontinence, and one had urinary incontinence. Therefore, 38% were functionally disabled and could not be adequately rehabilitated owing to poor facilities. The reported that while management of spina bifida cystica is more aggressive now in most developed countries, theirs remained selective due to difficulty with multidisciplinary care and rehabilitation. Even with their selective management, the care of patients with functional handicap remained a challenge (22).

As part of a public health campaign conducted from 1993 to 1995 in an area of China with high rates of neural-tube defects (the northern region) and one with low rates (the southern region), the outcomes of pregnancy in women who were asked to take a pill containing 400 microg of folic acid alone daily from the time of their premarital examination until the end of their first trimester of pregnancy was evaluated. . The greatest reduction in risk occurred among the fetuses or infants of a subgroup of women in the northern region with periconceptional use who took folic acid pills more than 80 percent of the time (reduction in risk, 85 percent as compared with the fetuses or infants of women who registered before their last menstrual period and who took no folic acid; 95 percent confidence interval, 62 to 94 percent) [corrected]. In the

southern region the reduction in risk among the fetuses or infants of women with periconceptional use of folic acid was also significant. They concluded that periconceptional intake of 400 microg of folic acid daily can reduce the risk of neural-tube defects in areas with high rates of these defects and in areas with low rates (23).

The reproductive history of 100 women with at least 1 child with a neural tube defect (NTD) was studied in Mexico. . A total of 204 pregnancies resulted in 205 outcomes. Of the 100 sibships, 14 (14%) had more than 1 affected member. The pregnancy was shorter than 28 weeks in 56/205 (27%) of the total outcomes. Of 104 evaluable previous outcomes, 34 corresponded to short pregnancies, positioned before an affected (23/60, 38%), a healthy (2/18, 11%), or an undiagnosed product (9/26, 35%). Short pregnancies subsequent to affected outcomes were also increased. An increased number of abortions adjacent to affected offspring and a changing fertility pattern, depending on the product diagnosis, pointed to an environmental etiological component in this high-risk NTD group of mothers (24).

Records of almost 174,000 consecutive births at six Brooklyn hospitals during the years 1968-1976 were reviewed for congenital neural tube defects. Prevalence of anencephaly, myelomeningocele and occipital encephalocele combined was significantly higher in infants delivered to mothers born in Puerto Rico than in offspring of non-Puerto Rican whites or blacks. The

association of prevalence rates with ethnicity remained significant after adjustment for several variables. No significant differences in prevalence rates between whites and blacks were observed. Sex ratios of affected infants were close to unity in each ethnic group. Statistically significant associations were found between the prevalence of neural tube defects and parity, gravidity and economic status. The patterns of these associations varied among the ethnic groups. A declining trend in the prevalence of myelomeningocele was observed for all ethnic groups (25).

Results of a study of families in the Eastern Ontario/Western Quebec region who had a child born with a neural tube defect during the years 1969-1981 was reported. Socioeconomic and ethnic influences were noted, but there was no evidence of seasonal variation or any correlation with maternal age or parity. There was support for a causative role of maternal fever in some cases. They concluded that knowledge of rates of occurrence in relatives is useful for counseling (26).

PATIENTS AND METHODS

This was a descriptive case – control study.

Patients

Retrospective Phase: The study was conducted from records collected from admission books of the University Teaching Hospital Pediatric Surgery D block. All records of infants with a neural tube defect were looked at. The data dated from June 1994 to December 2002. In all there were 244 children recorded with a diagnosis of myelomeningocele, encephalocele or meningocele, but only sixty (60) files were located.

Prospective phase: All infants diagnosed with NTDs from August 2003 to June 2004 were included. These patients were the ones directly referred to the Neurosurgeons and admitted on the paediatric surgical unit ward. The mothers of these affected infants were interviewed using a unified questionnaire (Appendix 2) after informed consent (Appendix 1) was obtained. Counselling on the current issue of folic acid prevention was done and options and advice on physiotherapy services from the Cheshire support system was given to mothers who needed the support.

Criteria of inclusion: All infants with a diagnosis of myelomeningocele, meningocele or encephalocele were considered. Out of these, the patients with myelomeningocele were then included in the study.

Criteria of exclusion:

- Conditions like dermoid cysts and lipomas were excluded
- All those who did not give consent to the study.

Ethical issues

The Ethics and Research Committee of the University of Zambia approved the research. Consent for inclusion into the study was obtained from all parents.

RESULTS

Retrospective Phase

The results of this have been included in the literature review.

The Prevalence of myelomeningocoeles between 1994 and 2002 is shown in figure 1 below

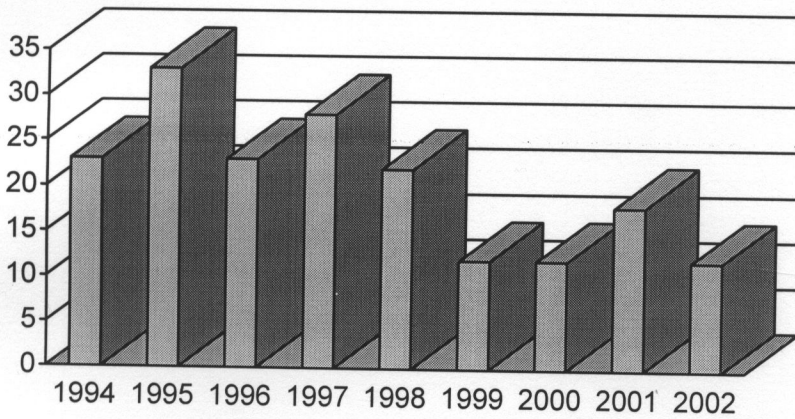


Figure 1: Prevalence of Myelomeningocoeles between 1994 and 2002.

Prospective Phase

The total number of patients included in this phase was 50 (fifty).

Encephalocoeles and meningocoeles were 14 (fourteen)

Myelomeningocoeles were 36 (thirty-six)

The patients with myelomeningoceles were 36 (thirty-six) and there were 21 (twenty-one) females and 15 males with a sex ratio of 1.4:1 females to males. This is seen in Figure 2.

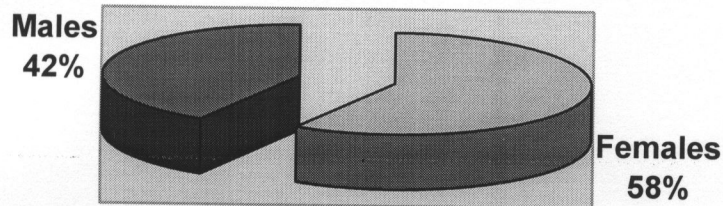


Figure 2: Sex ratio of myelomeningoceles

Figure 3: Socioeconomic status of myelomeningocele patients

The socioeconomic pattern was determined. Their status was low if their place of residence was found to be in the village / shanty compounds / if they were staying in a / servants quarter. Their status was high if they lived in a modern house and had a full time job or were earning more than K500 000. They were medium if they were civil servants employed in a government institution or were earning a salary of between K250 000 – K 500 000. The socioeconomic status was mostly in the low group status with 86.1%. This is shown in Figure 3.

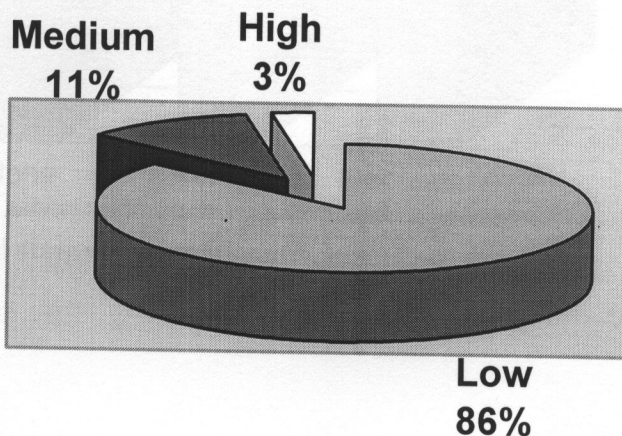


Figure 3: Socioeconomic status of myelomeningocele patients

On evaluation of the maternal education it was found that 45.7% had had a primary school education. This is shown in Figure 4.

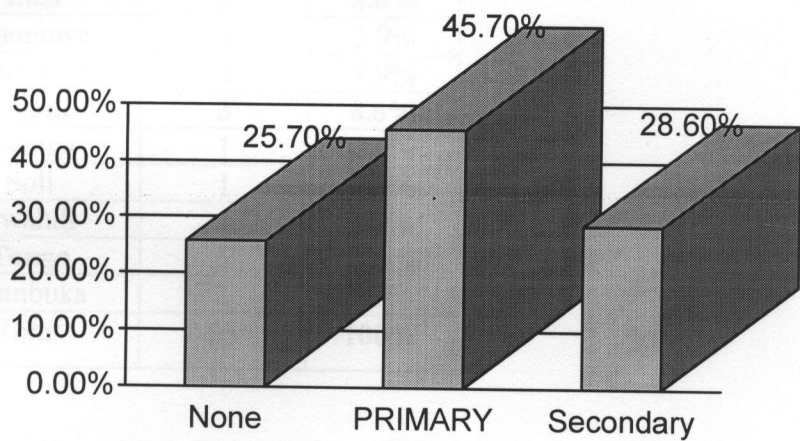


Figure 4: Educational status of mothers

Maternal tribe	Frequency	Percent
Bemba	7	20%
Chewa	2	5.5%
Indian	1	2.9%
Kalubale	1	2.9%
Kalunda	1	2.9%
Kaonde – ila	1	2.9%
Lala	1	2.9%
Lamba – lala	1	2.9%
Lenje	1	2.9%
Lozi	3	8.6%
Mambwe	1	2.9%
Namwanga	1	2.9%
Ngoni	3	8.6%
Nsenga	1	2.9%
Soli	1	2.9%
Swahili	1	2.9%
Tonga	6	17.0%
Tumbuka	2	5.5%
<i>Total</i>	35	100%

Table 5: Comparison of maternal tribe and myelomeningocoele.

Affected site most commonly seen in myelomeningocoeles was sacral (36.1%), seconded by lumbosacral (25%) and Lumbar (25%) equally. This is shown in Table 6.

Site	Frequency	Percent
Cervical	2	5.6%
Lumbar	9	25%
Lumbosacral	9	25%
Sacral	13	36.1%
Thoracolumbar	3	8.3%
Total	36	100%

Table 6: Sites of occurrence of myelomeningocoeles

Province of origin was assessed. This is seen in the Table 7 below. Most referrals were from within Lusaka (38.9%). Central had 19.4% and southern province had 8.3%.

Province	Frequency	Percent
Central	7	19.4%
Copperbelt	1	2.8%
Eastern	4	11.1%
Luapula	1	2.8%
Lusaka	14	38.9%
North Western	1	2.8%
Northern	4	11.1%
Southern	3	8.3%
Western	1	2.8%
Total	36	100.0%

Table 7: Provinces of origin

Folic acid usage by the mothers was assessed. The results are shown in the pie chart in Figure 8.

Antenatal visit was assessed. This was assessed by gestational age when attending antenatal clinic. Table 9 shows this.

First Antenatal Visit (Gestation)	Frequency	Percent
1 month	17	49%
2 months	18	51%
3 months	0	0%
4 months	0	0%
5 months	0	0%
6 months	0	0%
7 months	0	0%
8 months	0	0%
9 months	0	0%
Total	35	100%

Figure 8: Folic acid consumption by mothers

Antenatal visits were assessed. This was to record the gestational age when mothers first started attending antenatal clinic. Table 9 shows this.

First Antenatal Visit (Gestation)	Frequency	Percent
1 month	1	2.9%
2 months	2	5.7%
3 months	3	8.6%
4 months	11	31.4%
5 months	9	25.7%
6 months	2	5.7%
7 months	6	17.1%
9 months	1	2.9%
Total	35	100%

Table 9: First antenatal visit by gestational age in the mothers.

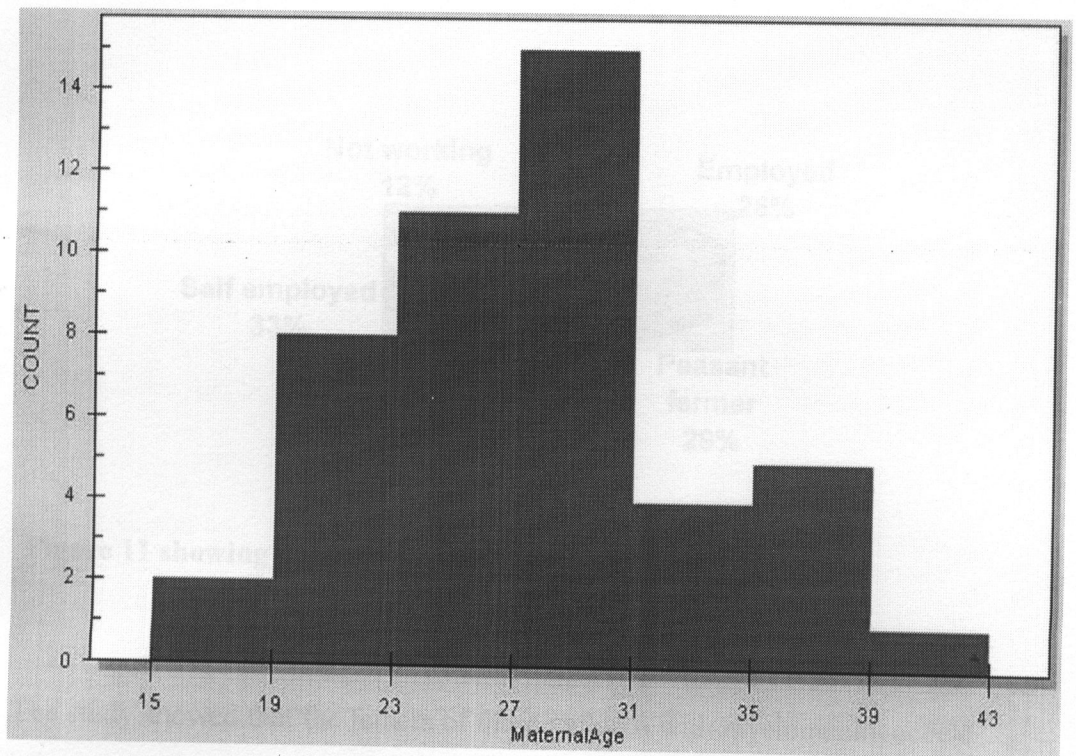


Figure 10 showing the maternal age distribution

Maternal age range was between 15 and 43 years of age. Most mothers were between the ages of 19 and 31.

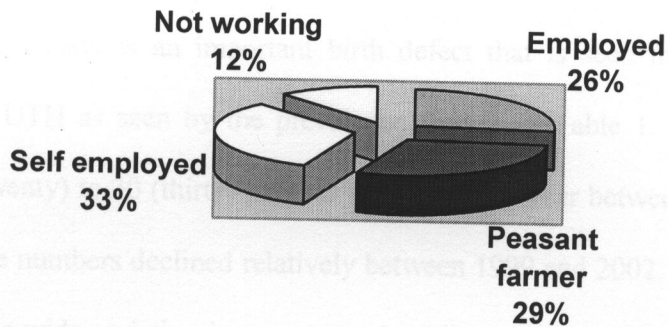


Figure 11 showing parental occupation

The study showed that the fathers of these children with myelomeningocoele were mostly self-employed. They earned a living doing odd jobs trying to make ends meet. A good number were peasant farmers who lived of their small plots of land. Those that were employed were those that had a full time paying job, that is, were working for an institution or company.

DISCUSSION

Myelomeningocele is an important birth defect that is seen in significant numbers at UTH as seen by the prevalence figures in Table 1. On average about 20 (twenty) to 30 (thirty) patients were seen per year between 1994 and 1998 and the numbers declined relatively between 1999 and 2002. It is known that there is a wide variation in prevalence in different parts of the world (18). Between 1994 and 2002 approximately 244 patients with neural tube defects were seen in UTH and over 80% of them were myelomeningoceles. Unfortunately only 60 (sixty) files were found. Of these 66.6% were myelomeningoceles. Most of these were referrals from southern province (24.3%) and Lusaka followed closely with 21.6%. The maternal tribe leading in having children with myelomeningocele was the Tonga women leading in majority 40.0% and followed closely by Bemba mothers with 35.0%. The drawback was that the records were not detailed. Judging by the numbers seen in the prevalence figures, it is important to say that these birth defects contribute significantly to the morbidity in the neurosurgical unit of the Paediatric D block.

Sex

In studies done elsewhere (3), (16), the sex ratio of myelomeningocele is slightly higher in females than males. The ratio varies considerably among racial/ethnic groups (18). In this study the sex ratio of females to male children was slightly higher in the females though it was not ascertained by ethnic type.

The rates of myelomeningocele vary widely among countries and by geographical regions within countries (3), (18,19). Prevalence rates have a wide variation in different ethnic and social groups within countries (18,19).

Socioeconomic status

On evaluation of the socioeconomic status of the patients, it was noted that most were from the low socioeconomic group (86%). This is as noted in studies done elsewhere, that neural tube defects are least common among the more prosperous, better educated, professional families and most common in the families of unskilled manual workers (18).

Maternal tribe

The Bemba women had the highest number of babies with myelomeningocele (20%), and Tonga mothers followed closely with 17.1%.

In the retrospective phase, it was the Tonga mothers that were in the majority with babies that had myelomeningocoeles.

Affected sites

The sacral region was the most common site of occurrence (36.1%), followed equally by lumbosacral with 25% and lumbar 25%. In the literature review Botto, Lorenzo, notes that the site most often affected is lumbosacral (1).

Province of Origin

Most referrals were from within Lusaka province (38.9%) followed by, Central province with 19.4% and Northern and Eastern province with 11.1% each.

Maternal education

Most mothers had a primary school education (55.1%). This is seen in the literature reviews quoted. (6), (13) and (18), that neural tube defects like myelomeningocoeles are more common in the less educated.

Folic acid consumption

Most mothers of the affected infants with myelomeningocoeles did not take folic acid (51.4%) compared to those who said they had (48.6%). The difference though is not marked but is significant for those that did not take any folic acid. Most mothers talked to have no idea why they are given folic acid and what its importance is. It could not be ascertained as to whether the clinics were deficient in the supplies or not.

Antenatal Visitation

It is noted that most mothers started antenatal clinic at about 4 (four) to 5(five) months gestation (31.4% and 25.7%). Most mothers when asked why they went this late said that either the clinic was too far from where they stayed or that they had no idea that it was necessary to start early.

Parental Occupation

Most mothers when asked what their husbands did for a living said that their husbands were unemployed and did odd jobs to make ends meet. Others were peasant farmers who lived off what they grew on small plots of land in the village. This reflects what is noted in literature that the children are born from families of unskilled manual workers (18).

CONCLUSION AND RECOMMENDATION

Of the epidemiological characteristics of patients presenting to the University Teaching Hospital, Lusaka, it is noted in this short small study that our patients mainly come from a low socioeconomic background. The mothers are mostly minimally educated, that is, they have a primary school education at the most. As to the ethnic status of our patients, the Tonga and Bemba mothers featured strongly in both aspects of the study. Most patients seen are from within Lusaka province in this study, and most reside in the highly populated and mainly low-income residential areas of the city. Myelomeningocoeles presented more in the sacral region, but lumbosacral also featured significantly.

In literature it has been quoted that neural tube defects, of which myelomeningocoele is one, are least common among the more prosperous, better educated, professional families and most common in the families of the unskilled manual workers (18).

The study may not be a true representation of the country's true picture, but it is a starting point. I would recommend another larger study of longer duration, and preferably one that would look into each province.

These babies affected were noted not to be receiving any physiotherapy while on the ward. So I would also recommend that physiotherapy be started whilst on the ward. There is a need to educate these mothers on the importance of physiotherapy in these children. The mothers would also have an opportunity to find out which physiotherapy centers they can access when at home.

A separate special outpatient clinic should be started to assist these children who need specialist attention. This clinic would be able to educate mothers on their children's condition. The mothers can also be educated on the importance of folic acid supplementation within the clinic. The clinic can also be a starting point of another larger study, where all information concerning these children is gathered. Information concerning the dietary practices of these mothers could also be looked into, including their beliefs.

To ensure all reproductive mothers get the needed requirements of folic acid, I would recommend that the issue of folic acid fortification of staple foods like mealie meal and sugar be looked into.

If the above recommendations were implemented, both clinical care and prevention of these neural tube defects will have been taken care of and reduction in mortality and morbidity will be achieved

REFERENCES

Literature Review

1. Botto LD, Moore CA, Khoury MJ, Erickson JD. Neural tube defects. *N Engl J Med* 1999; 341(20): 1509-19.
2. Marion, Robert. *Myelomeningocele. Pediatrics in Review* –August 1992
3. Kat Kolaski, MD -*Myelomeningocele: eMedicine.com*
4. Prevention of Neural tube defects: Results of the Medical Research Council Vitamin Study. MRC Vitamin Study Research Group. *Lancet* 1991 Jul 20; 338:131-7
5. Werler MM, Shapiro S, Mitchell AA. Periconceptional folic acid exposure and risk of occurrent neural tube defects. *JAMA* 1993 Mar 10; 269:1257-61
6. Meyer RE, Siega-Riz A.M. Socio-demographic patterns in spina bifida birth prevalence trends-North Carolina 1995-1999 *MMWR Recomm Rep* 2002 Sep 13; 51:12-5

7. Use of folic acid for prevention of spina bifida and other neural tube defects—1983-1991. *MMWR Morb Mortality Wkly Rep* 1991 Aug 2; 40(30): 513-6
8. Folic acid for the prevention of neural tube defects. *American Academy of Pediatrics. Committee on Genetics. Pediatrics* 1999 Aug; 104(2pt 1): 325-7
9. Yen IH, Khoury MJ, Erickson JD, James LM, Waters GD, Berry RJ The changing epidemiology of neural tube defects. United States, 1968-1989) *Am J Dis Child* 1992 Jul; 146(7): 857-61
10. Williams LJ, Mai CT, Edmonds LD et al. Prevalence of spina bifida and anencephaly during the transition to mandatory folic acid fortification in the United States- *Teratology* 2002 Jul; 66(1): 33-9
11. Stevenson RE, Allen WP, Pai GS et al. Decline in prevalence of neural tube defects in a high-risk region of the United States. *Pediatrics* 2000 Oct; 106(4) 825-7

12. Wasserman CR, Shaw GM, Selvin S, Gould JB, Syme SL. Socioeconomic status, neighborhood social condition and neural tube defects. *AM J Public Health* 1998 Nov; 88(11): 1674-80
13. Farley TF, Hambidge SJ, Daley MF. Association of low maternal education with neural tube defects in Colorado, 1989-1998. *Public Health* 2002 Mar; 116(2): 89-94
14. Unpublished Data on survey of neural tube defects. Dr Laston Chikoya
15. Munkonge L. Encephalocoele versus a General Surgeon. *African Journal of Neurological Sciences* vol. 11 Jan 1992
16. Feuchtbaum LB, Currier RJ, Riggle S et al. Neural tube defect prevalence in California (1990-1994): eliciting patterns by type of defect and maternal race ethnicity. *-Genet Test* 1999; 3(3): 265-72.
17. Lary JM, Edmonds LD. Prevalence of spina bifida at birth – United States, 1983 – 1990: a comparison of two surveillance systems. *MMWR CDC Surveill Summ* 1996 Apr 19; 45(2): 15 – 26.

18. Carys M. Bannister and Brian Tew: *Current concepts in Spina bifida and Hydrocephalus* – 1991
19. Frey L, Hanser WA. Epidemiology of neural tube defects – *Epilepsia*. 2003; 44 Suppl 3:4-13
20. Buccimazza SS, Molteno CD, Dunne TT, Viljoen DL. Prevalence of neural tube defects in Cape Town, South Africa. : *Teratology*. 1994 Sep; 50(3): 194-9.
21. Airede KI. Neural tube defects in the middle belt of Nigeria. : *J Trop Pediatr*. 1992 Feb; 38(1): 27-30.
22. Shehu BB, Ameh EA, Ismail NJ. Spina bifida cystica: selective management in Zaria, Nigeria. *Ann Trop Paediatr*. 2000 Sep; 20(3): 239-42.
23. Berry RJ, Li Z, Erickson JD, et al. Prevention of neural tube defects with folic acid in China. China-U.S. Collaborative Project for Neural Tube Defect Prevention. : *N Engl J Med* 1999 Nov 11; 341(20): 1485-90

24. Rivas F, Davalos IP, Olivares N et al . Reproductive history in mothers of children with neural tube defects. : *Gynecol Obstet Invest* 2000; 49(4): 255-60.
25. Feldman JG, Stein SC, Klein RJ, Kohl S, Casey G. The prevalence of neural tube defects among ethnic groups in Brooklyn, New York. : *J Chronic Dis* 1982; 35(1): 53-60
26. Hunter AG. Neural tube defects in Eastern Ontario and Western Quebec: demography and family data. *Am J Med Genet* 1984 Sep; 19(1): 45-63

Photograph of Myelomeningocele

EPIDEMIOLOGICAL CHARACTERISTICS OF PATIENTS WITH
MYELOMENINGOCELES PRESENTING TO THE UNIVERSITY
TEACHING HOSPITAL, CHENNAI

1. What information do you want to obtain from you concerning your child?

We are concerned in
detailed in
type of pa

2. Back

You are g
child has
characteri
infer with
the
give us di

3. Who

Dr. M. L.
the Depart
for and P
progress

The office
myelom

4. What

5. What

6. Possible problems

We may further examine you by asking you to go often



RESEARCH CONSENT FORM

EPIDEMIOLOGICAL CHARACTERISTICS OF PATIENTS WITH MYELOMENINGOCELES PRESENTING TO THE UNIVERSITY TEACHING HOSPITAL – LUSAKA

1. Why are we asking for information from you concerning your child and yourself?

We are conducting a study of patients born with the above condition and those affected in a similar way. We are asking these questions to help us know about the type of people affected and how they are affected.

2. Background information.

You are going to be asked questions concerning your child and yourself because your child has a condition that is now known to be preventable. We are getting characteristics of patients with this and similar conditions so that we can use the information gathered to find ways of helping and educating the population at large about the importance of early folic acid supplementation. We hope this study will give us distinct information.

3. Who is carrying out this study?

Dr. M.Lungu is the principal investigator in this study. Mr T.K. Lambart the Head of the Department of Neurosurgery at the University Teaching Hospital is supervising her and Professor Odimba, a Consultant General surgeon will help monitor the progress. The study being done is based in the Paediatric surgery department D01.

The official name of the study is “Epidemiological characteristics of patients with myelomeningocele presenting to the University Teaching Hospital Lusaka.

4. What happens in this research?

- ☐ A protocol form will be used to enter your child's and your personal details
- ☐ You will be reviewed as the need arises.

5. Possible problems.

We may inconvenience you by asking to see you often.

6. Benefits

You will gain more understanding about your child's condition and what you are able to do in future to reduce chances of you having another child with this condition. You will also gain more information about support groups and community-based organisations like the Cheshire Homes Society that assist disadvantaged children.

7. Your rights to participate, not to participate, or to withdraw from the study.

- ☐ Taking part in this study is optional. You need not take part in this study. You may later change your mind and withdraw from the study. You will still be counselled about this condition.
- ☐ You will suffer no penalty if you do not participate in the study and you will not lose any benefits to which you are entitled to as a patient. Your present and future medical care at the UTH; Lusaka will be the same whether or not you take part in this study.
- ☐ If there are any new findings in this study that may affect you, you will be told about them as soon as possible.

8. Confidentiality

Your name will never be made public by the investigators. The medical record of your care will be treated the same as all medical records at the University Teaching Hospital, Lusaka.

Information from this study and from you or your child's medical record may be reviewed and copied by the study investigators and examiners that may be appointed by the University of Zambia. A code number that makes it very difficult for anyone to identify you will identify the research information gathered by the study. All information will be stored in a secure place. Information from this study and from you or your child's medical record may be used for research purposes and may be published; however your name will not be made public by the investigators. It is possible that after the study is over, we may want to look again at you and your child's medical data collected during the study to help us answer another question. If this happens, your name will not be made public.

I Hereby agree to provide and allow personal information regarding my child's disease and some of my personal details to be used in this study.

I have been reliably informed by the researcher Dr M Lungu who is asking these questions that the information I shall give will be used in the strictest confidence. A copy of this form signed by me and the investigator is being given to me.

Signature; Thumb print.....

Date;

I have explained this research study to the subject. I am available to answer any questions now or in the future regarding the study and the subject's rights.

The principal investigator Dr Martha Mwewa Lungu and her supervisors can be reached at the Department of Surgery, University Teaching Hospital, Lusaka, Zambia.

.....

Signature of Investigator and Printed Name

QUESTIONNAIRE

Date: **File No:**

Name

Age..... **Sex** **D.O.B**.....

Residential Address:

Province of Origin

Place of Birth **Parity**.....

Mothers' tribe **Maternal age**

Maternal Education: (none) (Primary) (Secondary) (Tertiary)

Fathers' occupation

Antenatal clinic card yes/no

1st antenatal visit-gestational age

Supplements taken Folic acid y/n others y/n

Pregnancy multiple/single Term/premature

Birth weight.....

Type of defect

Site of defect.....

Complications neurological 1. Hydrocephalus y/n 2. orthopaedic
3. urologic 4. GIT

Socio-economic Status Low Medium High