Spina Bifida Cystica; features and early postoperative outcomes an experience in Kampala

Byabato S (MBChB, M.Med), **Kiryabwire J** (MBChB, M.Med, FCS), ***Galukande M** (MBChB, M.Med, MSc, FCS) Department of Surgery, College of Health Science, Makerere University **Correspondence author:* Galukande Moses, Department of Surgery,College of Health Science, Makerere University,Kampala, Uganda. Email: mosesg@img.co.ug

Abstract

Introduction: Spina Bifida is one of the most serious developmental disability of the infant. The prevalence of Spina Bifida varies across time, region, race and ethnicity. Worldwide, it accounts for as many as 4.7 in 10,000 live births. Although Spina bifida in its entirely is commonly seen in neurosurgical practice there is sparse literature on this disabling congenital anomaly in the Ugandan context. This study therefore aimed at describing the clinical features and early post operative outcomes.

Methods: A prospective descriptive study recruited children with Spina Bifida Cystica admitted to Neurosurgical ward over a period of 11 months from May 2010 to March 2011, at Uganda's National Referral Hospital. Clinical presentations of these patients were obtained by performing a general and neurological examination and using a standardized questionnaire. The patients were followed up for two weeks to establish early outcomes.

Result: A total of 51 patients with Spina Bifida Cystica were studied, representing 3.9% of all patients admitted on the unit that period. Out of those 54.9% were males and 45.1% were females. The mean age of their mothers was 23.8 years and the second birth rank was the mostly affected. Family history of Spina Bifida was present in 7.8%. Nearly all patients (98%) had myelomeningocele with a most common site being lumbosacral (47.1%). Hydrocephalus was noted in 72.5% of patients before surgery and talipes equinovarus was the other most associated congenital anomaly.

Forty three patients were operated on. The rest 8 patients died before surgery. The mean preoperative hospital stay was 17.6 days.

Conclusion: Patients had long pre operative lapse before the definitive management was offered and all the deaths occurred during this period. Exploration of factors responsible for this delay may enhance opportunities for timely intervention.

Key words: Spina Bifida Cystica, surgery, early outcomes

Introduction

Spina Bifida is a disorder of the cerebrospinal fluid system resulting from a failure of neural tube closure in the fetus and it can affect any part of the spinal cord. Over the past twenty-five years, a number of studies have demonstrated that environmental and genetic factors play an important part in Spinal Bifida and other neural tube defects etiology (1). Maternal ages, neural tube defect in previous pregnancy and low maternal folate level have been strongly associated with Spina Bifida (2,3).

The prevalence of Spina Bifida varies across time, by region, race and ethnicity (4). Worldwide, it accounts for as many as 4.7 in 10,000 live births (5, 6). The range in prevalence in Western nations is roughly 0.1 – 1 per 1,000 live births; a few non-Western studies often quote higher rates, though again widely spread (7,8). In Africa; Airede reported the incidence of 7/1000 deliveries in the middle belt of Nigeria (9).

The incidence and mortality of Spinal Bifida Cystica in Uganda is not known. This study was carried out to describe the characteristics of the Spina Bifida Cystica lesions and the early post operative outcomes at Mulago Hospital in Kampala Uganda.

Methods

This prospective descriptive study was conducted at a neurosurgery unit of Mulago hospital. Mulago hospital is the national referral and a teaching hospital for Makerere University College of health sciences. It also serves as the regional referral hospital for Kampala. Fifty one patients with Spina Bifida Cystica admitted to that unit in the period of 11 months were consecutively enrolled to this study. Patients discharged before surgery against medical advice were excluded.

Patients were admitted to ward from the emergency ward and from neurosurgery outpatients after being reviewed by a neurosurgeon. Data collection was done using a structured and pre-coded questionnaire. His-

tory was taken followed by general and neurological examination. Birth ranking was ascertained by history provided by the mother, miscarriages before 28 weeks were not considered. The lesions were examined for the content of arachnoid sac and confirmed at surgery. The state of the lesions was noted as either ruptured or intact and any ruptured lesion with cloudy/thick drainage was considered infected.

The size of the lesions were measured at their longest base diameters into small (1-3cm), medium (>3-5cm) and large (>5cm). The lesion levels were ascertained clinically into cervical, thoracic, thoracolumbar, lumbar, lumbosacral and sacral. The patients were assessed for lower limb movement into normal, weak and absent. The muscle tone was checked and graded into normal, hypotonia and hypertonia. Hydrocephalus was determined by measurement of head circumference using tape measure and compared to the normal percentile by age and sex. Cranial ultrasound scans requested for were done by senior radiologists. The treatment given was the standard of care following the established ward protocol including but not limited to antibiotic choice and use. Mortality was considered as death of the baby before or within 2 weeks of surgery, cause of death was determined by either postmortem or on clinical basis as discussed by the neurosurgical team. The questionnaires were checked for completeness by the principal investigator. Epi info 2002 computer software package was used to analyse the data.

Results

A total of 51 patients with Spina Bifida Cystica were admitted to neurosurgical ward in a period of 11 months, representing 3.9% (51/1307) of patients admitted to that ward from 1st May 2010 to 31st March 2011. There were 28 (54.9%) males and 23 (45.1%) females with a male-to-female ratio of 1.2:1. The mean age of their mothers was 23.8 years and second birth rank was the most affected with 16 (31.4%) patients. Only 4 (7.8%) patients had family history of spina bifida. Table 1 shows the clinical presentation of the patients.

Surgery was done on 43 (84.3%) patients and all the rest 8 (15.7%) patients died before surgery. Of those to whom surgery was done, 42 (97.7%) underwent standard Spina Bifida Cystica repair while 1 (2.3%) patient had standard Spina Bifida Cystica repair and VP shunt insertion at once. The mean stay of patients in the ward before surgery was 17.6 days (range 4 – 50) (fig. 1). No patient was operated on within 72hrs of admission. The mean age of patients at operation was 4.4 weeks (range 1 – 32). There was delayed admission for treatment of Spina Bifida Cystica with average age on admission being 14 days. Table 2 summarizes the post op details.

The cause of death to eight patients were identified to

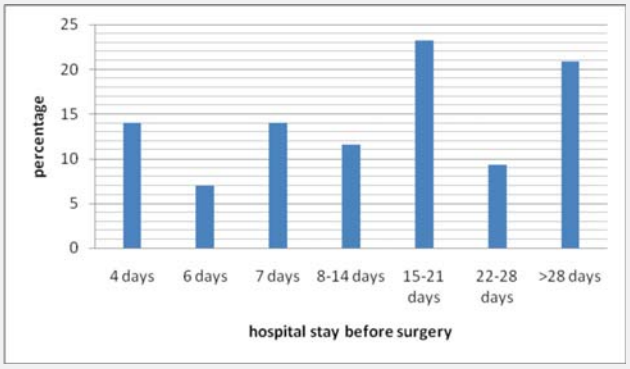

Figure 1: Distribution of hospital stay before surgery

Clinical feature	Number of patients
Clinical type	
Myelomeningocele	50 (98%)
Meningocele	01 (2%)
Site of the lesion	
Cervical	01 (2%)
Thoracic	02 (3.9%)
Thoracolumber	06 (11.8%)
Lumber	12 (23.4%)
Lumbosacral	24 (47.1%)
Sacral	06 (11.8%)
Size of defect	
Small	09 (17.7%)
Medium	35 (68.6%)
Large	07 (13.7%)
State of the lesion	
Intact	38 (74.5%)
Ruptured	13 (25.5%)
Infection status	
Not infected	36 (70.6%)
Infected	15 (29.4%)
Total	**51 (100%)**

Table 1: Clinical presentation of spina bifida patients

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Patients with post op complications and prolonged post operative hospital stay

Code	Age*	sex	Lesion characteristics	Pre ophosp. stay	Post op hosp. stay	Complications/reasons for prolonged hosp stay
01	4 weeks	M	Medium size, lumbar myelomeningocele, ruptured and infected	19 days	>14 days	Local wound infection
02	1 week	M	Medium size, lumbar myelomeningocele, ruptured and infected	9 days	>14 days	Local wound infection
03	4 weeks	F	Medium size, lumbosacral myelomeningocele, intact and not infected	28 days	14 days	CSF leak
10	9 days	F	Small size, cervical meningocele, intact and not infected	6 days	>14 days	Awaiting VP shunt
13	3 weeks	M	Medium size, lumbar myelomeningocele, intact and not infected	21 days	>14 days	Local wound infection and dehiscence
19	5 weeks	M	Medium size, thoracolumbar myelomeningocele, intact and not infected	32 days	>14 days	Awaiting VP shunt
30	11 days	M	Large size, thoracolumbar myelomeningocele, intact and not infected	4 days	>14 days	Wound dehiscence
41	8 weeks	M	Medium size, thoracolumbar myelomeningocele, intact and not infected	28 days	>14 days	Awaiting VP shunt
42	12 days	M	Small size, lumbosacral myelomeningocele, ruptured and not infected	7 days	>14 days	Awaiting VP shunt
45	5 weeks	F	Medium size thoracic myelomeningocele, intact and not infected	4 days	>14 days	Local wound infection, wound dehiscence and CSF leak
46	7 weeks	M	Medium size, lumbar myelomeningocele, intact and not infected	44 days	>14 days	Wound dehiscence and CSF leak

2 days Unknown††
** Age of patient on the day of operation or death*
*** Patient got a cardiac arrest during induction of anaesthesia. Attempt at resuscitation were futile*
† Patient had septic spina bifida lesion and malnutrition; she was weak and breastfeeding poorly. The wound was cleaned and the child was given IV Ceftriaxone. The patient was then referred to acute care for further management. The patient unfortunately died on the 4th day.
†† Patient developed difficulty in breathing on the 2nd day after birth. Parents declined a postmortem and the discussion by the Neurosurgical team was inconclusive.

Table 2: Description of Patients

Code	Age*	sex	Lesion characteristics	Hospital stay	Cause of death
05	8 weeks	F	Large size, thoracolumbar myelomeningocele, intact and not infected	8 weeks	Cardiac arrest**
11	8 weeks	F	Medium size, lumbar myelomeningocele, ruptured and infected	8 days	Septicemia
15	1 week	F	Medium size, lumbosacral myelomeningocele, ruptured and infected	7 days	Meningitis
17	2 weeks	M	Medium size, lumbosacral myelomeningocele, ruptured and infected	10 days	Septicemia
22	3 days	F	Small size, lumbar myelomeningocele, ruptured and infected	1 day	Septicemia
24	4 weeks	F	Small size, sacral myelomeningocele, ruptured and infected	3 weeks	Septicemia
25	6 weeks	F	Medium size, lumbar myelomeningocele, ruptured and infected	4 days	Septicemia†
31	2 days	M	Small size, lumbosacral myelomeningocele, intact not infected	2 days	Unknown††

Table 3: Died patients

be (table 2); a cardiac arrest during induction of anaesthesia, five patients due to septicemia, one patient due to meningitis and one patient had respiratory arrest following difficulty in breathing in 2nd day of life with unknown cause of death. Antibiotics (ceftriaxone for inpatient and ampiclox for discharged patients) were given to all 43 operated patients post operatively for a period of 3 -10 days (mean 5.4 days) and preoperatively to 17 patients who had ruptured or infected lesions.

By the end of two weeks, 33 (76.7%) operated patients had been discharged and their mean postoperative ward stay was 5.7 days. Ten (23.3%) patients stayed in the ward for more than two weeks after operation; among those, 6 had immediate postoperative complication and 4 were waiting for the VP shunt insertion. Early postoperative complication was statistically a significant cause of prolonged hospital stay. (RR of 7.71; 95% CI by Taylor series of 2.92-20.39 and P value < 0.05) There was no early postoperative death.

Seven (16.3%) patients had early postoperative complications which included; local wound infection in 4 patients, wound dehiscence in 2 patients and CSF leak in 1 patient (Table 3,4). Two out of 10 patients (20%) without hydrocephalus before surgery developed hydrocephalus within two weeks of follow up. Two patients had their lower limb movement improved from weak to

Lesion characteristics	post operative complications		
	Local wound Infection	Wound dehiscence	CSF leak
Size of defect			
Small 6 (13.9%)	0	0	0
Medium 31 (72.1%)	4	1	1
Large 6 (14.0%)	0	1	0
Total 43 (100%)	4	2	1
State of the lesion			
Intact 36 (83.7%)	2	2	1
Ruptured 7 (16.3%)	2	0	0
Total 43 (100%)	4	2	1
Infection status			
Not infected 34 (79.1%)	2	2	1
Infected 9 (20.9%)	2	0	0
Total 43 (100%)	4	2	1

Table 4: Distribution of early postoperative complication by lesion characteristics

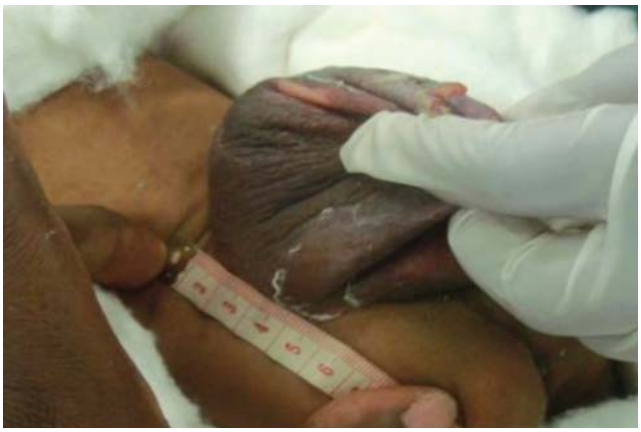

Photo 1 showing a ruptured lumbosacral lesion

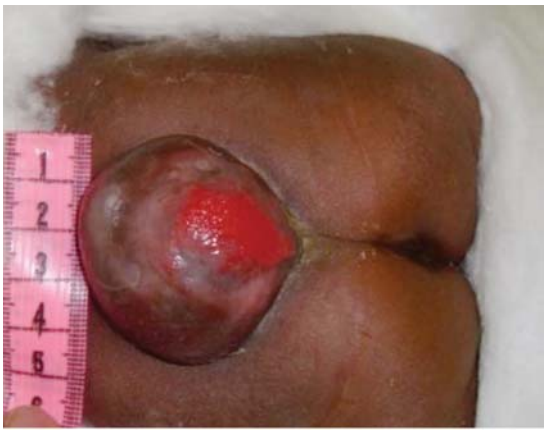

Photo 2 showing an intact lumbosacral lesion with granulation tissue on the placode

normal movements within two week of follow up and only 1 patient deteriorated from normal to week movements.

Discussion

Patients and maternal characteristics

Males were slightly more affected with Spina Bifida Cystica than female with a male to female ratio of 1.2:1. Alatise et al, found nearly the same ratio 1.1:1 (10), which was also found in Farley study (11). Most of the patients were among the 1st to 3rd born constituting more than three quarter of the patients. This is contrary to what was found by Alatise et al; 4th birth rank being mostly affected with 27.4% of patients(10) and 5th born in Kabre et al. with the reason of exhaustion of maternal folate store with repeated pregnancies(5). Maternal socio-economic level and baseline folate level were not described in this study, but most of patients who come to public hospitals in our setting are usually of low social economic status. Probably they have poor diet and thus low folate store even before starting their reproductive life, although studies may be needed to find out the possible explanation of this finding.

The mean age of the mothers of children with Spina Bifida Cystica was 23.8 years and most of them were in the age group 20–24 years. This is different from a well

studied relationship between maternal age and Spina Bifida Cystica in many studies, which shows more risk in teenage and over 35 year mothers (12,13). Lifestyle factors (ie; poor diet, smoking and illicit drug use) associated with high risk of Spina Bifida Cystica in teenage pregnancies found in other studies should be evaluated in our setting, to find out the discrepancy noted in ages of mothers giving birth to Spina Bifida Cystica children.
In this study, a family history of Spina Bifida Cystica was 7.8%. This is in correlation to other studies which showed a range of 3-8% (14, 15). Preconception genetic counseling to the couple with family history of Spina Bifida Cystica should be encouraged.

Clinical presentation

Nearly all patients 51 (98%) had myelomeningocele and one patient (2%) had meningocele. These two constitute Spina Bifidia Cystica; occulta is rarer and was not included in this study. This is in consistence with other studies, which show that Myelomeningocele is the commonest type (9, 10, 16, 17). Myelomeningocele is a more severe form of Spina Bifida Cystica as compared to meningocele due to involvement of neuro tissues in the sac. In this study, less than half (47.1%) had variable degree of lower limb weakness and this was less than what was found in Kumar et al. and Ozveren et al studies, 66.5% and 78.1% respectively (16, 18).
The most common site was lumbosacral (47.1%); again this was in consistence with other studies (10, 16). It should be noted that, neurulation is a complicated phenomenon involving multiple cells and processes and literature doesn't explain why lumbosacral is the most frequent site. Van Allen et al. postulated that, despite the cervical site there is an additional closure initiation site at the caudal end of the neural tube explaining the increased frequency of defects in this region (19). Caudal neuropore closure in human embryos is situated at about the level of the somites 32-34 (i.e., the future third to fifth sacral vertebrae) and this is the junction between primary and secondary neurulation (20). This might be the reason of more frequent defect in lumbosacral, since there is no region of overlap between primary and secondary neurulation in mammals (21).
About a quarter of these patients presented with ruptured lesions and this showed to have a higher proportion of postoperative local wound infection than the intact lesion. This association was not statistically significant in this study, probably due to small sample size. Other infected lesions were among those with exposed neuro placode. A slightly higher proportional (28.3%) of ruptured lesion were found by Alatise et al. (10).
Most of the patients (82.3%) had medium to large sized lesion and simple technique was used to cover the defects. These lesions were amenable to simple closure and probably not too large as described in Ozveren et al. study (18). There is no objective classification system that has been devised for these lesions but the literature describes 'large defects' as those not eligible for primary suture.
Hydrocephalus was noted in 72.5% before surgery and the proportion increased to 76.5% in two weeks post surgery. This was higher than what was found by Alatise et al. (10) and Kumar et al. (16) but still lower to other studies which have shown a range of 80-85% (22). Talipes equinovarus was among the mostly associated congenital malformations with 17.6% of patients, and this was also found by other studies (9, 10, 16).

Management of Spina Bifida Cystica

Out of 51 patients only 43 (84.3%) patients were offered a surgical management for Spina Bifida Cystica. The rest 8 (15.7%) patients died before surgery, septicemia being the most common cause of death. The mean hospital stay before surgery was 17.6 days; this is the probable explanation of those deaths before surgery following rupture and infection of the lesions while waiting for surgery. No patient was operated on the first 72 hours of life contrary to standard treatment and findings in many studies that; the lesions should be repaired soon after birth to decrease the risk of mortality and CNS infection which are likely to hamper mental development (23, 24). The delay in operating on these children in our study was attributed to: (i) Delayed admission to the ward (average age on admission was 14 days), (ii) Limited theatre space and (iii) Postpartum Maternal complications. Prolonged hospital stay increases the burden of disease on the hospital and the family. Prolonged bed occupancy by the same patient results into a low turnover in the healthcare service provision to the community.
The mortality rate in our study was lower compared to what was found in the study by Alatise et al whose mortality rate was 22.4%. This finding might be due short period of postoperative follow up; deaths occurring outside the 2 weeks follow up period were not recorded.

There is a need to emphasize early surgery to reduce the mortality rate by avoiding infection in the preoperative period in our setting.

VP shunt insertion was carried out in one patient simultaneously with closure of the defect despite a large proportion of patients with hydrocephalus. This patient had overt hydrocephalus with an intact non infected lumbosacral lesion. VP shunt insertion at Mulago Hospital is not routinely done simultaneously with Spina Bifida Cystica closure. This is because of the increased risk of infection in these combined operations. There is no established protocol so far regarding time interval between Spina Bifida Cystica closure and VP shunt insertion in resource scarce settings like ours and thus further studies are warranted.

Immediate postoperative outcomes and complications

Patients were followed up for two weeks and 76.7% of the operated patients had been discharged by that time. Postoperative complications were to be associated with longer postoperative hospital stay ($P<0.05$). Two lesions with wound dehiscence (4.6%) were from medium and large lesions, but were soft were closed by simple technique.

Meningitis was not found in our postoperative patients probably due to postoperative prophylactic antibiotics used to all our patients. Some of the patients stayed in the ward for more than two weeks awaiting the VP shunt insertion after Spina Bifida Cystica closure. Improvement of lower limb movement has been found in other studies, and rarely deterioration is noted (16, 18). In this study two patients had improved movement and one deteriorated from normal to weak movement.

Conclusion

Patients had long pre operative lapse before the definitive management was offered and all the deaths occurred during this period. Exploration of factors responsible for this delay may enhance opportunities for timely intervention.

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