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Myelomeningocoele in Nigeria: Has There Been Any Change with Improved Neurosurgery Care?

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Abstract

A 6 year retrospective study of the clinical and pathological profile of myelomeningoceles who had neurosurgical care at the neurosurgical unit of the University of Nigeria Teaching Hospital Enugu, South-East Nigeria from 2010-2015 was done. The review found 57 males (54.2%) and 47 females (45.8%). Patients' age ranged from 1 day to 7 years. More patients (59.6%) presented outside the neonatal age group. Fifty patients (48.1%) live in urban setting while 54 (51.9%) were from a rural setting. The lumbo-sacral area is the most common location, followed by thoraco-lumbar, sacral, and the thoracic regions representing 90.4%, 5.8%, 2.9%, and 1% respectively. Associated anomalies were seen in 57 patients (54.8%) with neurologic abnormalities been the most common (49% of all cases) followed by hydrocephalus (44.2%). Limb paralysis was above L2 in 45.1%. There is an association between increasing paternal age and presence of associated abnormalities (p = 0.027). Significant association was also found between place of residence and associated anomalies (p 0.009).Surgical procedures were performed in 84 patients (80.8%). The postoperative mortality was 5.9%. Ignorance of pre-conception folic acid use was demonstrated. Ante-natal preventive strategies need to be reviewed, while multi-disciplinary management with good social support is recommended post-natally.

Keywords: Myelomeningocoele; Folic acid; Limb paralysis.

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1. Introduction

Myelomeningocele (open spina bifida cystica) is the most common neural tube defect NTD, and the most severe birth defect compatible with long-term survival [1]. It has an incidence of 0.41-1.43 per 1,000 live births in the United States and is believed to be lower among African-Americans than Caucasians with an incidence of 0.1 per 1,000 live births among native Africans[2]. The incidence of the myelomeningocele in Nigeria is not known. Many hospital based studies from centers around the country, covering most of the population, provide a basis to estimate the frequency. This has become possible with the increasing number of Neurosurgeons and neurosurgical facilities in Nigeria. Management of this condition has remained challenging for neurosurgeons. The paradigm shift came from the work of John Lober who first outlined criteria for selective active management. Other pressures and improvements in available surgical and supportive facilities have made active management of all patients possible[2–6]. Myelomeningocele is often associated with other major anomalies that may impact management. It may result either from failure of closure of the neural tube or from secondary reopening of a closed tube, although most of the evidence favors the former theory[7]. Spina Bifida has been reported as the most common type of NTD in Nigeria with myelomeningocele being the most common [8,9]. It accounts for 5.7% of neurosurgical admissions in Nnewi in South-East Nigeria[10]. In Nigeria and other African countries, societal norms and other social circumstances influence management choices[11]. These factors are potentially modifiable and as care improves are likely to result in shifts in management protocols. As the society evolves, proper understanding of the clinico-pathologic profile that has become available in recent times will ensure a more holistic management of these patients. The increasing diversity in medical subspecialties and supportive disciplines now available in Nigeria has added a robust multi-disciplinary approach to the management of the patients[12]. This study reviews the changing patterns in the clinic-pathological profile and management outcomes of myelomeningocele in Nigeria.

2. Methodology

The available literature on myelomeningocele in Nigeria was reviewed from a search of PubMed and African Index Medicus and compared with data from recent records of care for these patients in the University of Nigeria Teaching Hospital UNTH, Enugu between Jan 2010 and Dec 2015. The UNTH is a major Neurosurgery referral center in the South Eastern part of Nigeria with a wide catchment area. South-East Nigeria has a projected population of approximately 16.5 million[13]. Patients are also recruited from the adjoining South-South and North-Central geopolitical regions. The study is a retrospective analysis of prospectively recorded data. Patients' and their parents' relevant information, associated anomalies and outcome were retrieved. Only patients with clinical evidence of myelomeningocele were included. Other forms of neural tube defects such as encephaloceles, myeloceles, lipomeningoceles, lipomas and meningoceles were excluded. After detailed discussion on the nature of the anomaly and the anticipated outcome of management, an informed consent was obtained from the patients' parents in all cases. Those who declined consent were excluded from the study. The patients were operated upon by the consultant neurosurgeons in the unit using a standardized protocol (microsurgical dissection, reconstitution of the spinal canal, duroplasty and layered closure). Patients had the benefit of multidisciplinary care involving orthopedic surgeons, physiotherapists, psychologists and pediatric urologists. Follow-up has continued for a minimum of nine months and when possible parents were further

interviewed during the follow-up period or by telephone (cell phones). Data was analyzed using Statistical Package for Social Sciences (SPSS version 21.0). The χ^2 test was used for level of statistical significance, with a p value <0.05.

3. Results

Over the six-year period under review, 104 patients were managed. Patients' age at first presentation ranged from birth to 7 years. Ninety one of the 104 patients (87.5%) presented after 3 days and were regarded as having presented late. Fifty seven patients (54.2%) were males and forty seven (45.8%) were females giving a male-female ratio of 1.2 to 1. Patients were from urban setting in 50 cases (48.1%), while 54 (51.9%) were living in rural areas. The most common site of myelomeningocele was the lumbo-sacral region (Figure 1.0). Ante-natal Obstetric Scan was done in 62 patients (59.6%) with 2 cases (1.9%) diagnosed pre-natally but there was no case of pregnancy termination. Most patients (93) were delivered via Spontaneous Vertex Delivery representing 89.4% of all deliveries. Only 10.6% were delivered via Caesarian Section. Associated anomalies were seen in 57 patients (54.8%) as outlined in table 3.0. These were more common in patients living in urban areas (table 6.0). Hydrocephalus was clinically obvious at presentation in 79.6% of patients. The remaining 20.4% were diagnosed with neuro-imaging (12.2% Transfrontanellar Ultrasound, 8.2% Cranial CT scan/ Magnetic resonance imaging). The mean occipito-frontal circumference was 40.9cm (SD 6.8). The frequency appears to taper with increasing birth position. (Figure 2.0) 11.5% of mothers had no form of formal education. Seventy six percent had a minimum of secondary education (Figure 3.0). Irrespective of level of education, none had knowledge of pre-conception use of folic acid but late presentation was commoner in the less educated parents who also were mostly from rural areas. The mean dimension of the lesions was 5.8 x 5.6 x 4.4cm (Length, Breadth and Height). Motor and/or sensory abnormalities were present in 51 patients (49%).

The remaining 53 patients (51%) had no clinically detected motor/ sensory levels. Eighty-four patients (80.8%) had operative treatment. Twenty patients (19.2%) were not operated upon. Forty-four (52.4%) of the patients operated on needed a ventriculo- peritoneal shunt, 25 (56.8%) of these were identified pre-operatively while 19 (43.2%) became obvious during the post-operative period. Post operatively the mortality was (6%), 3 from respiratory complications and 2 from severe post op sepsis. At the end of the follow up period mortality had increased to 34%.

Table 1.0: Age at Presentation

Age in weeks	Frequency	Percent (%)
0-3 days	13	12.5
3- 7 days	14	13.5
1-4 weeks	15	14.4
5-52	50	48.1
>52	12	11.5
Total	**104**	**100**

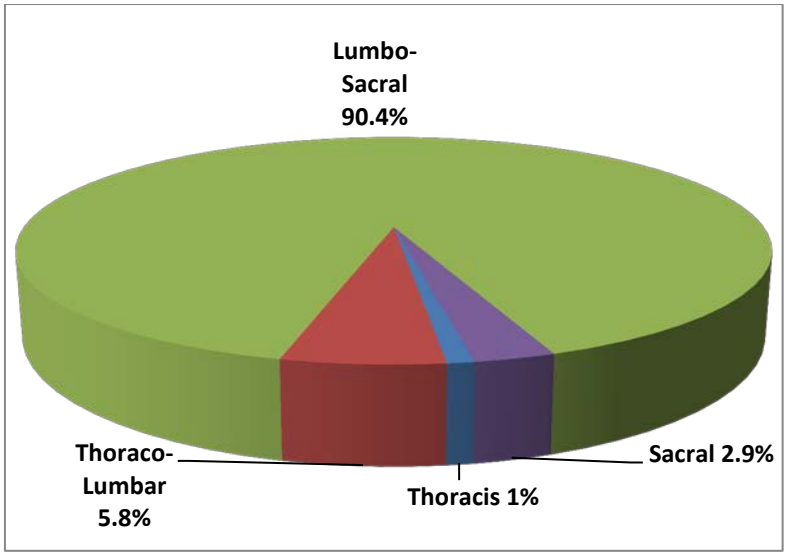

Figure 1: Location of Lesion among Patients

Table 2: State of Lesion at Presentation

	Frequency	Percent
Intact	98	94.2
Ruptured	6	5.8
Total	104	100.0

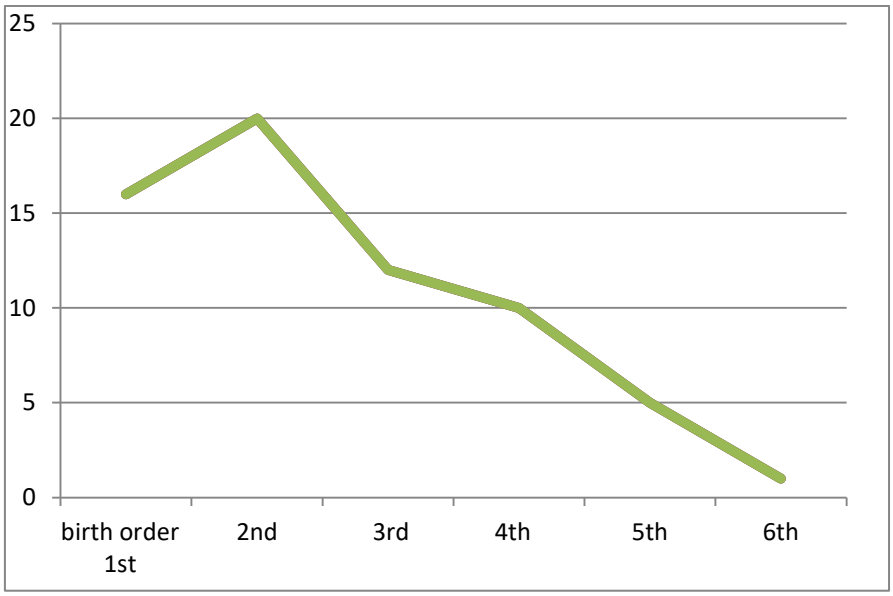

Figure 2: Birth Position of Patients

Table 3: Findings in patients with Myelomeningocele

Anomaly	**Frequency**	**Percent (%)**
Hydrocephalus	43	41.3
Club foot	11	10.6
Hydrocephalus and club foot	3	2.9
Paraplegia/Paraparesis	51	49.0
Sphincteric Dysfunction patulous anus	51	49.0
None	47	45.2

***Some patients have more than one finding**

Table 4: Maternal age distribution

Age in years	**Frequency**	**Percent (%)**
15-19	4	3.8
20-24	12	11.5
25-29	53	51.0
30-34	23	22.1
35-39	8	7.7
40-44	3	2.9
≥45	1	1.0
Total	**104**	**100.0**

Mean maternal age= 28.7 ± 5.1, median = 28.00, mode = 28

Table 5: Cross-tabulation between Paternal age and Associated Anomalies

Paternal age	**Associated Anomalies**	**Non**	**Total**	**Statistical significance**
≤ 29	2	9	11	$\chi^2 = 9.230$
30-34	14	7	21	
35-39	19	10	29	
≥40	14	16	30	df= 3
Total	**49**	**42**	**91**	p- value= 0.027

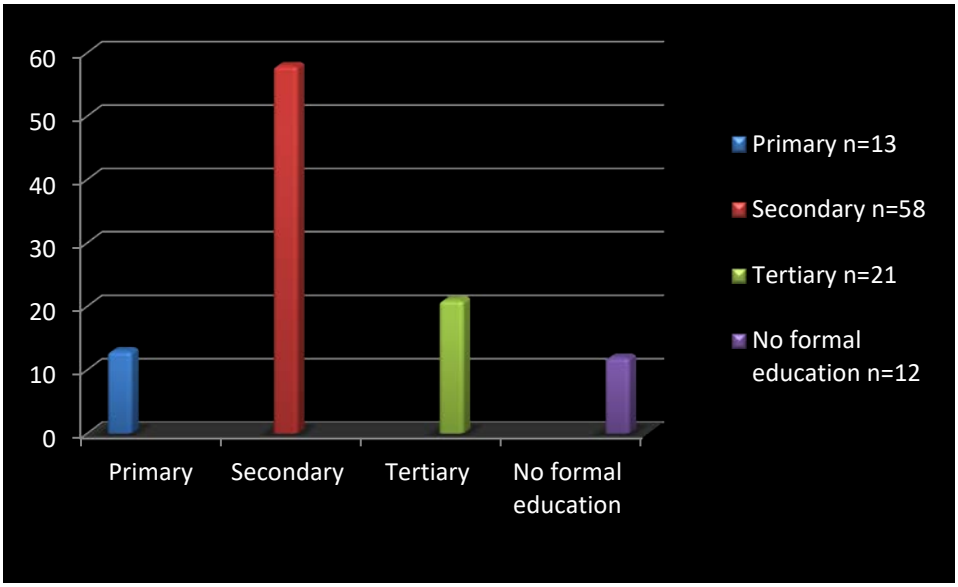

Figure 3: Distribution of Maternal Level of Education

Table 6: Association between place of residence and associated anomalies on presentation

Location	**Associated Anomalies**	**Non e**	**Total**	**Statistical significance**
Urban area	34	16	50	$\chi^2 = 6.766$
Rural setting	23	31	54	
Total	57	47	104	df = 1 p- value = 0.009

Table 7: Treatment Modalities Received By Patients

Modality	**Frequency**	**Percent (%)**
Excision and repair	59	56.7
Excision and VP shunt	25	24.0
Not operated	20	19.2
Total	**104**	**100.0**

Table 8: Causes of Mortality among Patients

Cause	Operated (n=84)	Not Operated (n=20)
Chiari Malformation + Severe Brain stem dysfunction	0.00	5.00 (25.0%)
Severe Sepsis	2.0 (2.4%)	0.00
Other Respiratory problems	3.0 (2.6%)	0.00
Total	**5.0 (6.0%)**	**5.0 (25.0%)**

4. Discussion

Myelomeningocoele was found to have a slight male preponderance among our study population with a M:F ratio of 1.2:1. This is still in keeping with an earlier publication from this center[14]. This is also consistent with findings from Port-harcourt and Jos in South-South and North Central Nigeria respectively, and from Zambia [9,15,16]. However, worldwide report, and reports from western countries have shown a female preponderance[7]. Nnadi and Singh reported a female preponderance for NTDs from Sokoto, North-West Nigeria. This finding may be due to the fact that their study was not specific for myelomeningocoele.

Only 13 (12.5%) out of the 104 patients studied presented within 72 hours of birth. This is in stark contrast to what is obtainable in developed countries where patients present in the first day of life[4]. Again, it is an accepted practice that patients are to be operated within the first day of life, however operation may be deferred for up to 72 hours without an increase in complications[17]. Even with this extended 72 hour window, early repair was not feasible in majority of our patients as 77 (74%) of them presented after 1 week of life. This delay may not be unrelated to the social stigma associated with having babies with congenital defects. Against this background parents remain in denial about affected babies until complications made it impossible to delay any longer and precipitate hospital attendance. Even then delay is often contributed to by the long chain of the referral path, most patients having to go through untrained personnel through health centers to general hospitals before reaching specialized care.

In terms of place of abode, there are about an equal number of patients from the rural and urban settings (54 and 50 patients respectively). It is worthy of note however that the patients presenting from urban areas were more likely to have associated anomalies than their rural counterparts (34 and 23 respectively). This then raises a question as to whether urbanization with its attendant pollution could have predisposed these babies to an increased risk of congenital malformations. A focused study may well throw more light on this.

The most common location of the lesion was in the lumbo-sacral area seen in 94 patients (90.4%). This is consistent with other findings from Abakaliki and Port-harcourt, both in Southern Nigeria[15,18]. In this study associated abnormalities were found in 57 cases (54.8%), the most common findings were lower limb paralysis, sphyncteric dysfunction and hydrocephalus. Similar findings have been reported in Abuja, north central

Nigeria[19]. Also the Port-Harcourt study in South-south Nigeria found lower limb abnormalities especially talipes equinovarus as the most common associated congenital anomaly[15]. This is not surprising given the embryopathogenesis of myelomeningocele. Patients with varying degree of limb paralysis and sphincteric dysfunction (49%), and those with club feet required multidisciplinary management and rehabilitation in addition to neurosurgical intervention [18–20]. Twenty three of the 51 patients with limb paralysis (45.1%) have their level above L2. It has been noted that better long term outcome in terms of independence have been shown to be associated with lesions below L2 [21,22]. This remains to be seen in the current study as our patients have not been followed up long enough to draw conclusions. Phone calls and clinical follow up visits will elucidate this.

Publications had noted that 5-10% of myelomeningocele patients have clinically overt hydrocephalus at birth, and over 80% of patients who will develop it do so before age 6 months of life [23]. We noted that the prevalence of hydrocephalus of 44.2% in our study was at variance with that clinically seen at birth from other studies [23]. But this is understandable given the delay in presentation of the patients in our study.

We found a statistically significant association was found with increasing paternal age ($p = 0.027$) and myelomeningocele. Also, patients dwelling in urban areas tended to have higher associated anomalies than those living in rural settlements ($p = 0.009$). This may be explained by more readily available folate-rich fresh vegetables and fruits in the villages. There is a need to further look in to the role of environmental factors as other studies have shown some role [16,18].

It is noteworthy to observe that educational status was not observed to influence knowledge of pre-conception folic acid use as none of the mothers in this study used it appropriately. The role of folic acid in reducing incidence and recurrence of neural tube defects among women intending to be pregnant have been established [24,25]. The poor implementation of this recommendation is disturbing as 98 mothers (94.2%) had some form of ante-natal care and yet were not aware of the use of pre-conception folic acid.

Active management was instituted for all patients but 20 (19.2%) were not fit to be operated due to severe mal-nutrition and sepsis requiring initial treatment and rehabilitation. The remaining 84 (80.8%) had excision and repair of the lesion. This was done at the same sitting with ventriculo-peritoneal shunt in 25 patients. Postoperative complications were wound dehiscence resulting in severe sepsis (2.4%), respiratory problems (2.6%). Operative mortality was 6.0%. This is comparable to mortality found in an earlier study in the same institution and also in Zaria, but lower than 16.7% reported in Abuja in Northern Nigeria[19,20]. It does appear then that not much have changed in terms of operative mortality for myelomeningocele patients. It remains to be seen if the long term mortality will improve given the improvements in ancillary services.

Among the non-operated group, mortality was 25%. This is not surprising given that these patients had other conditions which contraindicated surgery in the first instance. The likelihood was that the conditions which contraindicated surgery ab-initio viz sepsis and malnutrition added to the recorded mortality. . Immediate surgical outcome was not statistically found to be associated with level of lesion ($p = 1.000$), and surgical intervention does not appear to affect neurologic deficits. Such association between level of myelomeningocele

and survival was not demonstrated in Uganda as in our study[26].

5. Conclusion

Myelomeningocele management has improved in Nigeria. The incorporation of multidisciplinary team approach may account for some of this improvement. Many deleterious factors peculiar to developing countries such as delayed presentation, social stigmatization, inadequate care in rural areas, and need for rehabilitation after neurosurgical intervention remain a significant challenge. More needs to be done by relevant authorities to implement recommended folic acid protocols among intending mothers.

Declaration

Authors have no conflict of Interest

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