



AENSI Journals

Australian Journal of Basic and Applied Sciences

ISSN:1991-8178

Journal home page: www.ajbasweb.com



Studies in Spina Bifida at Gezira State, Sudan

^{1,2}Amir Mohamed Ali, ³Amal Idris Hasan, ^{4,5}Moawia Gameraddin

¹Taibah University, Department of anatomy, Faculty of Medicine, Box. 3001. Madinah, Saudi Arabia

²University of Gezira, Department of anatomy, Faculty of Medicine - Sudan

³University of Gezira, Department of Nursing, Faculty of Applied Medical Science, Sudan

⁴Taibah University, College of Applied Medical Sciences, KSA

⁵AlzaiemAlazhari University, Faculty of Radiological Sciences and Medical Imaging.

ARTICLE INFO

Article history:

Received 19 August 2014

Received in revised form

19 September 2014

Accepted 29 September 2014

Available online 12 November 2014

Keywords:

spina bifida, children, neonates, disabilities, folic acid

ABSTRACT

Spina bifida (SB) is a common congenital defect of the nervous system, often resulting in severe disabilities. This prospective descriptive research aimed at study of spina bifida at the Gezira State, Sudan. The study conducted over 24 months from the period June 2009 to May 2011. The study samples included 105 children with spina bifida, 55 females and 50 males, aged between the first day of life to 9 years. The data was collected through a designed questionnaire, detailed clinical examination and patient data sheet. The results revealed that 22.7% were seen in the first day of life, forty babies (38.1%) were delivered at hospitals. Clinically, 36.2% had hydrocephalus, 39% club foot, 33.3% had lower limb paralysis and 34.3% had sensory loss. Regarding the back lesion, 45.2% were situated in the lumbosacral region, 72.1% were measured 5-10 cm, whereas 76.9% were covered with membrane. This important case series says yes spina bifida occurs in Sudan, and it is an important cause of childhood disability, morbidity and should be prevented by mandatory folic acid fortification programs in Sudan

© 2014 AENSI Publisher All rights reserved.

To Cite This Article: Amir Mohamed Ali, Amal Idris Hasan, Moawia Gameraddin., Studies in Spina Bifida at Gezira State, Sudan. *Aust. J. Basic & Appl. Sci.*, 8(18): 100-105, 2014

INTRODUCTION

Spina bifida (SB) or spinal dysraphism is the commonest central nervous system birth anomaly. It is recognized as the most complex developmental defect of the neural tube, notochord, ectoderm, mesoderm and very rarely endoderm (Pang *et al*, 1992). The general term spina bifida refers to those spinal anomalies that have incomplete midline closure of mesenchyme, osseous and neural tissue (Bankovich and Naidich, 1990). It is also one of the congenital anomalies with complex etiology. Both genetic and environmental factors are implicated (Noman, 2008). Folic acid has been found to decrease the risk of having a child with spina bifida (Sandler, 2004).

Spina bifida cystica is a form of spinal dysraphism associated with a protruding cyst made up of either meninges (meningocele) or meninges in combination with spinal cord tissue (meningomyelocele). These lesions are frequently associated with spinal cord dysfunction, hydrocephalus, and syringomyelia (Davis *et al*, 2004). Occult dyraphic defects are skin cover lesions that have no exposed neural tissue and no visible cystic mass (Korvic and Keller, 1992).

Babies born in Western Europe with SB are likely to have operations within 24 hours to repair and protect the spinal cord defect and prevent infection from reaching the brain (Miles, 2006). Physical signs of SB may include leg weakness and paralysis. Orthopedic abnormalities are club foot, hip dislocation, kyphosis and scoliosis. Bladder and bowel control problems, including urine and fecal incontinences, urinary tract infections, and poor renal function (Mitchell *et al*, 1885). In Africa, the reported incidence of SB is variable, e.g. in Malawi it was 0.47/1000; in Tunisia, was 1.05/1000; in Cape Town the prevalence of NTDs was 1.74/1000 and it was higher in the whites than the blacks (Soumaya *et al*, 2001). Spina bifida in Sudan is widespread and most frequently in rural area. The estimated incidence of SB is 4.8/1000 and 3.48/1000 in Gezira and Omdurman Maternity Hospital respectively (Nugud, 2003; Ghada and Salah, 2009), which is high compared to figures quoted from the literature. Detailed descriptions about this malformation in Sudan are hardly found in literature. This research aimed at study of the spina bifida at Gezira State, Sudan.

Corresponding Author: Amir Mohamed Ali, Taibah University, Department of anatomy, Faculty of Medicine, Box. 3001. Madinah, Saudi Arabia

MATERIAL AND METHODS

Materials:

A consecutive series of 105 Sudanese children with spina bifida has been reviewed. All were admitted and treated at Gezira National Centre for Pediatric Surgery between June 2009 and March 2011. All were examined on admission and assessed regarding general condition; they form the material for this study.

Methods:

Cross-sectional observational prospective study included 105 cases of spina bifida. The study was conducted at Gezira National Centre for Pediatric Surgery (GNPCPS), which is the only centre in Sudan outside the capital Khartoum that delivers pediatric surgery services. It is about 190 km southern to the Capital Khartoum.

The purposes of this study were discussed with the Ethical Review Committee in the Gezira University and the director of Gezira National Centre of Pediatric Surgery. A written consent and permission from the care takers of patients participating in the study was obtained. Patients with spina bifida who were seen or operated before the period of study and readmitted or visited the referred clinic for follow up were excluded from the study.

Data collection tools:

The data of this study were consisted of the following tools:

1. A developed interview questionnaire: It was designed by the researchers, and filled in an interview with the mothers or patient care taker. The questionnaire included; (i) patient demographic data (ii) presentation, back swelling and head enlargement; (iii) clinical examinations (swelling site, size and cysts covering) and neurological examinations for the lower limb deformities, and stool and urine incontinence.
2. Detailed clinical examinations were done to test lower limb sensation and motor function, each neonate was examined clinically by pediatric surgeon to assess the neurological state of the leg and the condition of the feet.
3. Photographs: Beside questionnaire and clinical examinations a digital camera (Kodak 8.2 megapixel, ISO 1250, Digital IS) was used to capture photographs to determine the feet deformities and back lesion.

The patients were analyzed with respect to their clinical presentation. The back lesions were examined for the type of skin coverage; patients were divided into two groups: those with skin coverage and those with membranous coverage. The back lesion also was measured in centimeter to determine the vertical and horizontal diameters of the lesion, three sizes of lesions were recorded: lesion less than five centimeters, lesion measures 5-10 centimeter and lesion more than 10 centimeter. Based on anatomy six types of vertebral levels of the lesion were identified: cervical, thoracic, thoracolumbar, lumbar, lumbosacral and sacral lesion.

General neurological examinations for the lower limb were done. Motor weakness and loss of sensation were examined. Upper and lower motor neuron dysfunction was assessed by neurologic examination. Patients were also examined for equino-varus deformity (clubfoot), accordingly they were divided into two groups: patients with clubfoot and patients without clubfoot. The former group was divided into two subgroups: patients who had bilateral deformity, and patients with unilateral deformity.

Statistical analysis:

For the purposes of this study the data were coded, processed and transferred to computer coding. The descriptive analysis was adopted which includes percentage, frequency distributions, tables and figures. Software Program: Statistical Package for Social Science (SPSS) version 13 was applied. Results were considered statistically significant at $p < 0.05$.

Results:

One hundred and five children with spina bifida were enrolled in this study. Their age ranged from the first day of the life to 9 years. Eighty two (78.1%) of samples were myelomeningocele, twenty two (21%) meningocele and one case (0.9%) presented with occult spina bifida. Three of infants were members of twin pairs and 47.6% of cases were male (Table 1).

Table 1: Sex incidence of different types of spina bifida in 105 children.

Type	Males		Females		Total	
	No	%	No	%	No	%
Occulta	0	0	1	0.95	1	0.95
Meningocele	8	7.6	14	13.3	22	20.9
Myelomeningocele	42	40	40	38.15	82	78.15
Total	50	47.6	55	52.4	105	100

The results reveal that most of the infants were brought to the hospital late. Only one quarter of the cases (25.7%) were seen on the first day of life, whereas (5.7%) of patients were not seen until they were more than six months age (Table 2). Forty babies (38.1%) were delivered in hospitals, 26 (65%) of them being by caesarian section. The others (64.9%) had been born at home and then only brought to the hospital on account of the malformation.

Table 2: Distribution of patients according to the age of presentation. (n=105).

Age	Frequency	Percent
1 day	27	25.7
2-7 days	28	26.7
1-4 wks	28	26.7
1-6 month	16	15.2
> 6 month	6	5.7
Total	105	100

Table (3) shows that all cases were presented with back swelling. Thirty eight had hydrocephalus. Thirty seven of myelomeningocele cases were associated with hydrocephalus. The only case of occult spinal dysraphism that was reviewed in this study, presented with subcutaneous lumbar lipoma. Another case was presented with cervical meningocele and hydrocephalus.

Table 3: Frequency of clinical presentation of spina bifida in 105 children.

Presentation	occult	meningocele	Myelomeningocele	Total
Back swelling	1 *	22	82	105
Hydrocephalus	0	1*	37	38

* One case had occult spina bifida associated with subcutaneous lumbar lipoma

* One case was presented with cervical meningocele associated with hydrocephalus

Table (4) and figure (1) show the anatomical distribution of the spina bifida deformity in all cases. The lumbosacral region was the commonest site of occurrence of spina bifida in (45.2%) of cases, followed by the lumbar region in (33.7%) of cases. In (2.9%) of patients, the defect was noticed in the cervical area.

Table 4: Location of spina bifida in 104 cases of meningocele and myelomeningocele.

Site	Meningocele	Myelomeningocele	Total	Percent
Cervical	3	0	3	2.9
Thoracic	2	3	5	4.8
Thoracolumbar	1	3	4	3.8
Lumbar	5	30	35	33.7
Lumbosacral	9	38	47	45.2
Sacral	3	7	10	9.6
Totals	22	82	104	100



Fig. 1: Spina bifida in different regions. A. Lumbar myelomeningocele. B. Very large oval thoracolumbar myelomeningocele.

The vertical diameter of the cyst was measured 5-10 centimeters in (72.1%) of cases, less than 5 centimeters in (20.2%) of and more than ten centimeters in (7.7%) of cases. Table (5) reveals that of total of 104 cysts (meningocele and myelomeningocele) twenty four (23.1%) were covered with skin, whereas (76.9%) of cysts were covered with membranes (Figure 2).

Table 5: Distribution of cyst covering in 104 cases of spina bifida cystica.

Cyst covering	Myelomeningocele	meningocele	Total	Percent
Skin	4	20	24	23.1
membrane	78	2	80	76.9
Totals	82	22	104	100

**Fig. 2:** A. Three months old male child presented with globular swelling in the sacral region which was found to be meningocele. B. five days old male infant with large myelomeningocele covered by membrane.

Individual patients often had more than one symptom. 39% of patients with spina bifida were presented with varying degree of weakness of the lower limb. 33.3% had paralysis. Sensory loss was elicited in 34.3% cases of spina bifida. Congenital talipes equinovarus was seen in 41 cases out of 105; unilateral in 16 (39%) cases and 25(61%) cases had bilateral talipes equinovarus (Figure 3).

**Fig. 3:** Talipes equinovarus (clubfoot). A: Twenty two days old boy presented with myelomeningocele associated with unilateral clubfoot and hydrocephalus. B: Bilateral clubfoot.

Discussion:

Spina bifida is a common congenital midline fusion defect of the nervous system. It is a potentially life-threatening condition affecting between two and four per thousand infants (Laurence, 1989; O'Neill *et al*, 1995) often resulting in severe disabilities. Risks are much reduced by immediate surgery and careful management.

Babies born with spina bifida are likely to have operations within 24 hours to repair and protect the spinal cord defect and prevent infection from reaching the brain (Miles, 2006). Furthermore, the longer the operation is delayed, the greater the risk of meningitis and other neurological problems. In this study the age of admission was specified, as 25.7% of infants were presented to the GNCPS in the first 24 hours of birth. This is best situation of admission age compared to the figure reported in previous study conducted in Nigeria (Mabogunje, 1990) in which one fifth of infants were admitted to the hospital in the first day of life. The incidence of spina bifida was higher in female than in male as sex ratio (male to female) was found to be 1: 1.1, this finding is consistent with a previous study conducted by Mitchell and others (Mitchell *et al*, 1985). Our finding is also similar to the study conducted in north American Caucasian and Hispanic individuals (Kit Sing *et al*, 2008). This condition is more likely to appear in females; the cause for this is unknown.

The effect of mode of delivery (vaginal vs. caesarian section) on neurological defect is controversial and remains under the study (Park, 1999). The study highlights that only 38.1% of the babies were delivered in hospitals, 26 (65%) of these born by caesarian section for distocia on account of gross hydrocephalus or a large baby. The others had been born at home and then only brought to the hospital on account of the malformation. Compared to other finding (Mabogunje, 1990), the frequency of hospital delivery is higher, this may be due to the good health services and follow up of pregnant women provided by the health centers in Gezira state.

All cases in this study were admitted to the GNCPS and presented with back swelling with or without hydrocephalus. One case of spina bifida occulta was presented first with back swelling which was found to be

subcutaneous lumbar lipoma. Of the total of 105 cases of spina bifida thirty eight (36.2%) of infants were already showing signs of hydrocephalus when first seen this represents 45.1% of myelomeningocele cases. The result was consistent to the finding of Dicianno, (2008) who stated that hydrocephalus occurred in approximately 11-90% of those individuals with spina bifida. It rates higher in agreement with the finding of Nugud *et al*, (2003) who reported 23.3% cases of spina bifida associated with hydrocephalus in the Gezira National Center for Pediatric Surgery. This percent is lower than the finding of Park, (1999) and Shaer, (2007) , who obtained very high rate, 90% and 80-90% respectively. The low incidence of hydrocephalus in this study might be due to the fact that the majority of patients were less than 3 months old at their first attendance and the risk of the development of hydrocephalus was known to continue until at least the age of a year, also hydrocephalus appears in most cases after surgical correction. In this series of patients one case presented with cervical meningocele associated with hydrocephalus in agreement with the results of Henry and Mickel, (1974) who obtained lower figure for meningocele which associated with hydrocephalus.

According to the anatomical distribution of the lesion, the results reveal that the lumbosacral region was the commonest site of occurrence of spina bifida in 47 patients (45.2%), followed by the lumbar and sacral regions. In 5 patients (4.8%), the defect was noticed in the thoracic region. This result confirms the findings of Kumar and Sing, (2003) and Bauer *et al* (1987) who reported 40% and 30-55% for the lumbosacral region respectively. It was also in agreement with the findings of Tortori-Donti (2000) who stated that, most myelomeningocele showed a predilection for lumbosacral region. On the other hand two studies reported higher rates of up to 81.5% and 86% for the lumbosacral region respectively (Danzer *et al*, 2009; Shehu, 2000).

The cyst size was an important factor in the surgical approach. The results revealed that the majority of the cysts were measured between 5 and 10 centimeters, and only 7.7% of the cysts were measured more than ten centimeters. Detailed study about the lesion size in spina bifida is hardly found in the literature. One study reported that the back lesion was quite variable in size and location; some may be confined to one or two centimeters in length. Others may be far more extensive, extending ten or more centimeters along the back (Sandler, 2004).

From this study it was found that in (76.9%) of cases, the lesion was covered with membrane. In 23.1% of cases the lesion was covered with intact skin. It was found that only two cases of meningoceles were covered with membranes, whereas most of myelomeningoceles were covered with membrane except for four cases. Regarding meningocele, this result is consistent to the finding of Kumar and Sing, (2003) who stated that meningoceles are fully skin covered. The majority of myelomeningoceles that covered with membrane were surrounded by skin at the margins of the lesions.

Neurological examination of all cases showed that children with spina bifida had variable manifestations. Individual children often had more than one symptom. In this study most of patients with spina bifida were presented with varying degree of weakness of the lower limb (39%). 33.3% had paralysis. Sensory loss was elicited in (34.3%) of cases. It was noticed that sensory and motor paralysis usually occurred together but either could be present alone. This percentage was lower than the finding of Kumar and Sing (2003) who found higher rates for the lower limb weakness, muscle atrophy and sensory loss.

Talipes equinovarus (clubfoot) was one of the most common orthopedic problems. In our study clubfoot was seen in 39% of cases; it was usually bilateral and occurred only in patients with myelomeningocele. This rate were higher than the figures reported by study conducted in the pediatric surgery center in central Sudan (Nugud, 2003). Consistently, Kumar and Sing, (2003) reported 36% in Indian patients.

In conclusion, this series serves as a baseline for comparison with future experience in this part of the world.

ACKNOWLEDGMENT

The authors are grateful to all mothers and patient's care takers who participated in this study and to the administration of the Gezira National Centre for Pediatrics Surgery for facilitating the research work.

REFERENCES

- Pang, D., M.S. Dias, M. Ahab- Barmada, 1992. Split cord malformation A unified theory of embryogenesis for double spinal cord malformation. *Neurosurgery*, 31:451-480.
- Barkovich, A.J., T.P. Naidich, 1990. Congenital anomalies of the spine; in Barkovich AJ (ed): *Pediatrics neuroimaging*. New York, Raven, 227-271.
- Norman, S.W., J.K. Christopher, P. Ronan, 2008. Baily & love's. Short practice of surgery. 25th ed. Hodder Arnold, part of Hachette live UK, 33: 483.
- Sandler Adrian, 2004. Living with spina bifida: a guide for families and professional. UNC Press Book. 3rd ed, 14-16.
- Davis, *et al.*, 2004. Textbook of Neuropathology. 2nd ed, 224-5.

- Korvic, H.E., M.S. Keller, 1992. Sonography of occult dysraphism in neonates and infants with MRI imaging correlation. *Radiographics*, 12: 297-308.
- Miles, M., 2006. Children with spina bifida: can medical, family and community recourses improve the life changes? *Disability and Society J*, 17: 643.
- Mitchell, L.E., N.S. Adzick, J. Melchionne, *et al.*, 2004. Spina bifida. *Lancet*, 20-26, 364 (9448):1885-95.
- Soumaya, S.G., M. Aida, M. Sami, *et al.*, 2001. Encephalocele: 26 retrospective cases at the maternal and neonatal center of La Rabta, Tunis. *Tunis Med J*, 79: 51.
- Nugud, A., M. Arbab and T.M. Osman, 2003. Pattern of neural tube fusion defects in Sudan. *Saudi Med J*. 24: S54.
- Ghada, E.A. and A.I. Salah, 2009. Neural Tube defects in Omdurman Maternity Hospital, *Sudan Khartoum Medical Journal*, 02(02): 185-190.
- Laurence, K.M., 1989. A declining incidence of neural tube defects in the U.K. *Z. Kinderchir*. 44(suppl 1):51.
- O'Neill, O.R., J.H. Piatt, P. Mitchell, S. Roman-Goldstein, 1995. Agenesis and dysgenesis of the sacrum: Neurosurgical implications. *Pediatr Neurosurg*, 22: 20-28.
- Mabogunje, O.A., 1990. Spina bifida cystica in northern Nigeria. *Child's Nerv Syst.*, 6: 103-106.
- Kit Sing Au, *et al.*, 2008. Characteristics of spina bifida population including north American Caucasian and Hispanic Individuals. *Birth defects Res A Clin Mol Teratol*, 82(10): 692-700.
- Park, T.S., 1999. Myelomeningocele. In: Albright L., Pollack I., Adelson D., editors. *Principles and Practice of Pediatric Neurosurgery*. New York: Thieme Medical Publishers, 291-320.
- Dicianno, B.E., B.G. Kurowski, J.M. Yang, M.B. Chancellor, G.K. Bejjani, A.D. Fairman *et al.*, 2008. Rehabilitation and Medical Management of the Adult with Spina Bifida.
- Shaer, C.M., N. Chescheir, J. Schulkin, 2007. Myelomeningocele: A Review of the Epidemiology, Genetics, Risk Factors for Conception, Prenatal Diagnosis, and Prognosis for Affected Individuals. *Obstetrical and Gynecological Survey*, 62: 471-479.
- Henry, A.P., R.E. Mickel, 1974. Spina Bifida in African and Indian babies. *The journal of bones and joint surgery*, 56: 4.
- Kumar, R., S.N. Singh, 2003. Spinal dysraphism: trends in northern India. *Pediatric Neurosurgery*. 38(3):133-145.
- Bauer, S.B., K.B. Labib, R.A. Dieppa, A.B. Retik, 1987. Urodynamic evaluation of boy with myelodysplasia and incontinence. *Urology*. 10: 354-362.
- Torti-Donati, P., A. Rossi, A. Cama, 2000. Spinal dysraphism: a review of neuroradiological correlations and proposal for a new classification. *Neuroradiol*. 42: 471-491.
- Danzer, E., M. Gerdes, M.W. Bebbington, L.N. Sutton, J. Melchionni, N.S. Adzick, *et al.*, 2009. Lower extremity neuromotor function and short-term ambulatory potential following in utero myelomeningocele surgery. *Fetal Diagn Ther*. 25(1): 47-53.
- Shehu, 2000. Spina bifida cystic: selective management in Zaria, Nigeria. *Ann Trop Paediatr*, 20(3): 239-42.