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ORIGINAL ARTICLE

Spinal dysraphism: A challenge continued to be faced by neurosurgeons in developing countries

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ABSTRACT

Objectives: The incidence of spinal dysraphism has significantly decreased over the last few decades, all over the world; however, still the incidence is much higher in developing countries with poor socioeconomic status.

Materials and Methods: The present study includes all patients managed for spinal dysraphism over a period of one year (January 2011-December 2011). Details including demographics, antenatal care history, site and type of lesion, neurological examination, imaging finding, associated congenital anomalies, management offered, and outcome were recorded.

Results: A total of 27 children were operated for spinal dysraphism during the study period (17 males and 11 females). Median age was 120 days (age range, 1 day to 6 years). Mothers of 15 children did not seek any regular antenatal checkup and only 13 mothers received folic acid supplementation during pregnancy. Fourteen children were delivered at home and 13 were at hospital. The most common site was lumbosacral region (67.8%). Seven patients had rupture of the sac at the time of presentation, one child had local infection, and four patients had hydrocephalus (requiring shunt before surgical repair). Two patients developed hydrocephalus at follow up, needing shunt surgery. The mean hospital stay was 7 days (range, 5 days to 31 days; median, 10 days).

Conclusion: Spinal dysraphism is still a major public health problem in developing countries. Management of patients with spinal dysraphism is complex and needs close coordination between pediatrician, neurologist, neurosurgeon, and rehabilitation experts. A large number of factors influence the outcome.

Key words: Meningocele, myelomeningocele, spinal dysraphism

Introduction

Neural tube defects are among the most common congenital malformations and a major cause of health problems in surviving children, especially in developing countries.[1-7] Neural tube defects affect 0.6 per 1,000 live births in the United States (approximately 4,000 NTD-complicated pregnancies annually)[8] and 0.5-2 per 1,000 pregnancies worldwide.[3] Although the incidence of spinal dysraphism has significantly decreased over the last few decades, all over the world; however, still the incidence is much higher in developing countries with poor socioeconomic status. Myelomeningocele derives from a failure of the neural tube[9] and it is the most common dysraphic malformation and the estimated incidence ranges from about 1-3/1,000 live births[10] to approximately 1 in 1,200 to1,400 births.[11] The social and economic impact of this disease is not well documented; however, up to 75% of adult survivors may be dependent on parents or other providers.[12]

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Materials and Methods

The present study includes all patients managed for spinal dysraphism over a period of one year (January 2011-December 2011). Clinical details of the patients including demographics, antenatal care history, site and type of lesion, neurological examination, imaging finding, associated congenital anomalies, management offered, and outcome were recorded. All patients were offered surgical excision and repair of the lesion. Ventriculoperitoneal shunt was performed when there was significant hydrocephalus.

Results

A total of 27 patients were operated for spinal dysraphism during one-year period. There were 17 males and 11 females. Median age was 120 days (age range, 1 day to 6 years). Fifteen mothers did not seek regular antenatal checkup and similar number did not receive folic acid supplementation during pregnancy. Fourteen patients underwent home delivery and 13 patients underwent hospital delivery. The most common site was lumbosacral region (67.8%) [Figure 1]. Seven patients had rupture at the time of presentation, one child had local infection, and four patients had hydrocephalus (requiring shunt before surgical repair) [Figure 2]. The mean hospital stay was 7 days (range, 5 days to 31 days; median, 10 days). Five of 7 patients who had rupture of the meningocele sac developed meningitis and succumbed to it [Figure 3]. There was no improvement in neurological function in any of the patients. Fourteen patients remained the same after surgery and 10 patients deteriorated in neurological functions. Of these 10 patients, four patients improved to preoperative neurological status at three-month follow-up. Remaining children were doing well at follow-up with variable amount of neurological deficits.

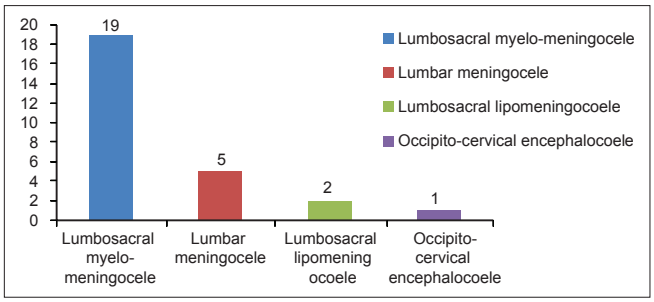

Figure 1: Site of lesion

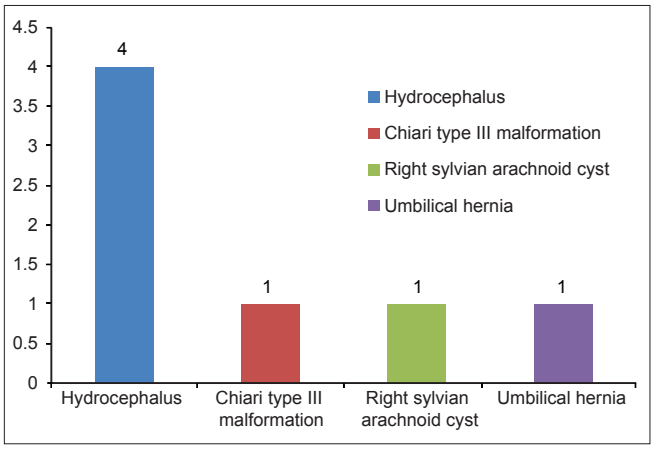

Figure 2: Associated lesions

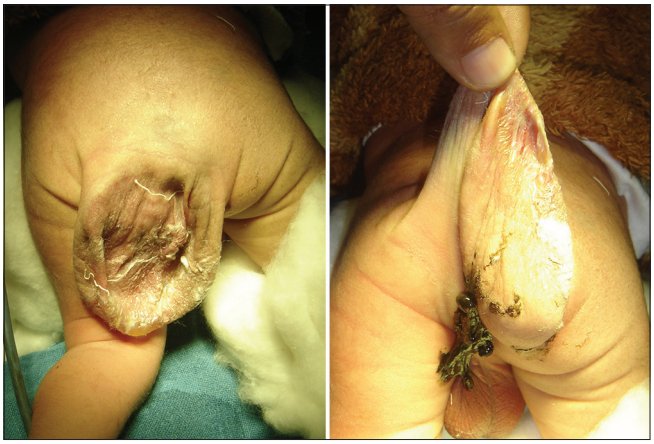

Figure 3: Clinical photograph showing ruptured meningocele sac that was repaired

Discussion

Neural tube defects can occur anywhere along the neuroaxis from the developing brain to the sacrum.[13,14] These can be divided into two main groups affecting cranial (anencephaly and encephalocele) or spinal structures (spina bifida).[7,13,15] The incidence of these lesions has significantly decreased all over the world, particularly in developed countries; however, this is not the case in resource-poor developing countries.[7,16,17] Neural tube defects are etiologically heterogeneous[2,18,19] and the epidemiology of neural tube defects is complex.[13] Many risk factors associated with increased risk for neural tube defects have been identified, including folic acid deficiency,[13] older or very young mothers,[13] modestly increased risk in primiparous,[11] previous spontaneous abortions,[20] short intervals between pregnancies,[21] multiple gestations,[22-25] maternal obesity and elevated body mass index,[26-28] maternal diabetes,[20] lower socioeconomic status,[13] tea use in the period before and during the first trimester,[29] zinc deficiency, lead, and high levels of organic matter[30-33] and drugs (including anti-epileptic medications).[13]

Intrauterine diagnosis of neural tube defects involving spine and spinal cord can be made with ultrasound, or suspected by positive screening for maternal serum alpha-fetoprotein. After birth, an obvious lesion or swelling can be seen on the back with a variable amount of the neurological deficits, with or without associated hydrocephalus.[7] Management of the children with minimal deficits where there is no involvement of neural structures can be straightforward surgical excision and repair.[7,16] The issue whether to manage or not when the child can be grossly handicapped due to neural structures involvement is greatly debated.[6,7,16,17,34-38] Most of the children who survive will have multiple system involvement, severe handicap, and a limited life expectancy.[3] Most of the untreated survivors with severe disease were severely or very severely handicapped, or to be in a 'vegetative' state.[39] In another study, it was estimated that only one in seven untreated infants would reach school age and only one in 70 would be fit to attend a normal school.[40] A policy of nonintervention results in early death of the most severely affected children.[38] It has been eloquently and sincerely recommended that life must be saved at all costs and that, however grossly malformed the infant.[41,42]

Management of patients with neural tube defects, particularly spinal dysraphism, is complex, and large number of factors influences the outcome.[7] The presence of hydrocephalus is an important and independent prognostic factor for cognitive function and insertion of ventriculoperitoneal shunt before the repair of lesion in the same sitting not only can avoid CSF leak and pseudo-meningocele formation, but also reduces the cost of treatment.[7] There are many limitations of the present study that need to be addressed, including lack of details regarding objective absence of folic acid deficiency, correlation with maternal age and parity, previous spontaneous abortions, maternal obesity, maternal diabetes, and lower socioeconomic status.

Conclusion

Spinal dysraphism occurs frequently and represents a significant public health problem in developing countries.[43] Although it is compatible with survival, most cases have moderate to severe disabilities, and may be associated with mental retardation.[15] Folic acid supplementation during pregnancy has been shown to reduce the incidence and recurrence of many congenital defects including NTDs.[13,15,33,44-48] Management of patients with spinal dysraphism is complex and needs close coordination between pediatrician, neurologist, neurosurgeon, and rehabilitation experts. A large number of factors influence the outcome.

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