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Spinal Dysraphism: Common Entity in Pediatric Neurosurgery

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

Aims and Objectives: The aim of this study is to review the demographic profile, clinical pretentions, surgical management and short term outcome of patients presenting with spinal dysraphism

Methodology: This is a prospective observational study of cases of spinal dysraphism managed surgically over the period of 2 years from March 2014 to February 2016 in Department of Neurosurgery at Tribhuvan University Teaching Hospital (TUTH), Kathmandu Nepal.

Results: Out of total 97 cases, there was male preponderance. In about 40% of population there was no history of proper ANC visit and most of them were from low economic status. Lump on the back was the commonest clinical findings. Lumbar Myelomeningocele was the commonest anatomical location of dysraphism. More than one third of patients needed CSF diversion postoperatively.

Conclusion: Myelomeningocele is a common NTDs. Open dysraphism may not always present as a lump. Delay in seeking medical attention may be due to illiteracy. None of the mother had taken folic acid prior to conception.

Key Words: hydrocephalus, myelomeningocele, spinal dysraphism

The term Spina dysraphism refers to a group of congenital anomalies of spine in which midline structure fails to fuse.[8] They are commonly known as neural tube defects (NTDs). It is among the most common congenital malformations and a major cause of health problems in surviving children.[7] Among them myelomeningocele represents the most serious form of dysraphism, a so called apert or open form involving the

vertebral column and spinal cord ,which occurs with an incidence of approximately 1 in 4000 live birth.[12]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 like Nepal. In Nepal, a demographic and health survey reported incidence of NTD is 47/10,000 live birth.[2] 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 care providers.[10] Myelomeningocele is among the common spinal dysraphism which accounts for about 75% of the cases.

The word "Spinal Dysraphism" was coined by B W Leichenstein in 1940. NTDs can be classified as "open" NTDs in which the neural tissue is exposed and "closed" NTDs with the neural tissue covered by tissue.[11] It is associated with Chiari-II malformation. Hydrocephalus in association with Chiari-II Malformation develops in at least 80% of patients with myelomeningocele. The anatomic level of the myelomeningocele sac correlates with the patients neurologic, motor, and sensory deficits.[5]

The present study aims to study the clinical profile of different types of spinal dysraphisms and short term outcome of surgical management.

The aim of this study is to review the demographic profile, clinical presentation and short term outcome of surgical management of the patient with spinal dysraphism.

Materials and Methods

This is a prospective observational study of 97 cases of spinal dysraphism managed surgically over the period of 2 years from March 2014 to February 2016 in the department of Neurosurgery at Tribhuvan University Teaching Hospital (TUTH), Kathmandu Nepal-a tertiary care centre. All the operated cases of spinal dysraphism were included in this study. Closed dysraphism which were managed with conservative treatment were excluded from the study. Detail clinical and radiological evaluation was done prior to surgery. All the cases were radiologically evaluated with magnetic Resonance imaging of the particular area with sagittal screening of brain and cervicovertebral junction. Anatomical repair was done using standard surgical technique in cases of nontethered cords. In cases with Lipomyelomeningocele, anatomical repair and excision of the lipoma was done. In cases with split cords laminectomy and anatomical repair was done. Ventriculoperitoneal shunt (VPS) was performed when indicated. Urodynamic study and neurophysiological evaluation was not done in the present study. After the surgery, all the patients were followed up routinely for 3 months. Complications and outcome were recorded.

Results

Total of 97 cases of spinal dysraphism were included in the present study. There was a male preponderance which constituted of 62% male (n=62) and 36.1% female (n=35). The age at the time of surgical intervention ranges from 6 hours to 20 years with mean age being 25 months. Most common clinical presentation was lump (79.1%) on the back followed by sphincter dysfunction as tabulated in **Table 1** and **Figure 1**.

Clinical presentation	Frequency(n)	Percentage (%)	Total
Lump	77	79.38	55
Para paresis	2	2.06	2
Sphincter Dysfunction	15	15.46	15
Cutaneous Stigmata	3	3.09	3
Total			97

Table 1: Clinical presentation

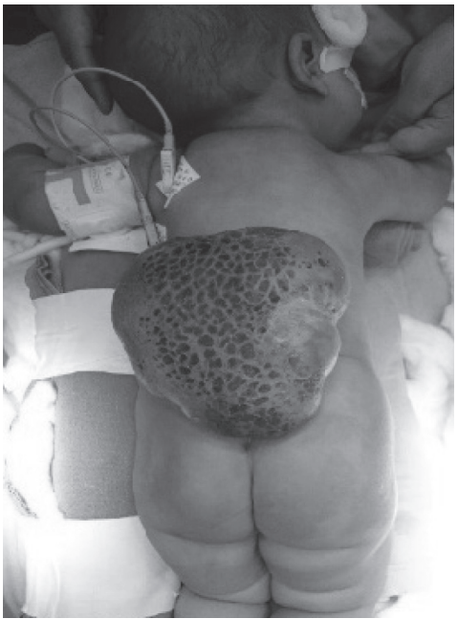

Figure 1. Three months old baby with large lumbosacral myelomeningocele

Lumbar spinal dysraphism was the commonest anatomical location which is shown in **Table 2**.

Spinal Level	Frequency(n)	Percentage (%)	Total
Cervical	9	9.3	9
Thoracic	3	3.1	3
Lumbar	40	41.2	40
Thoracolumbar	2	2.1	2
Sacral 7 7.2			7
Lumbosacral	36	37.1	36
Total			97

Table 2: Anatomical level of the spinal dysraphism

Myelomeningocele was the commonest pathology 78.4% (n=74) followed by lipomeningomyelocele14.1% (n=14), meningocele 4.1%(n=4), Split cord 2.1%(n=2) **(Table 3)**.

Pathology	Frequency(n)	Percentage (%)	Total
Myelomeningocele	74	78.1	74
Lipomyelomeningocele	14	14.4	14
Meningocele	4	4.1	4
Split cord	2	2.1	2
Mylocestocele	1	1	1
Total			97

Table 3: Pathology of spinal dysraphism

Other associated congenital anomalies were as tabulated in **Table 4**. Below is the picture **(Figure 2)** of MRI brain T2-weighted showing Chiari-II malformation with hydrocephalus.

Associated Anomalies	Frequency(n)	Percentage (%)
Hydrocephalus	26	26.80
Chiari-II Malformation	76	78.40
Leg deformities	23	23.71
Kyphoscoliosis	4	4.12

Table 4: Congenital Anomalies associated with the spinal dysraphism.

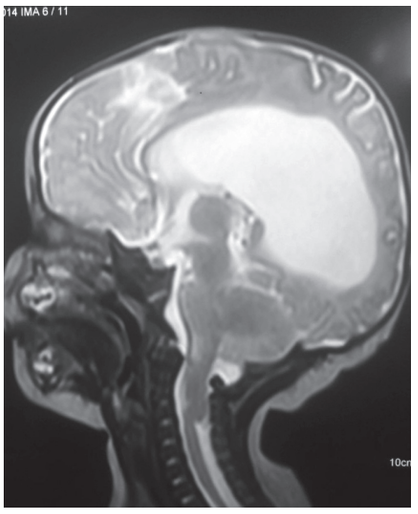

Figure 2: MRI of brain showing Chiari-II malformation with Hydrocephalus.

Regions of Nepal	Frequency(n)	Percentage (%)
Terai	57	58.8
Mountain	25	25.8
Himalaya	15	15.5

Table 5: Geographical Regional distribution of Spinal dysraphism.

Total 9 cases were operated on emergency basis. Among those who were operated in emergency, 8 cases presented in state of ruptured and one case had gross hydrocephalus so anatomical repair and CSF diversion in the form of VP shunt was done in emergency. Overall CSF diversion was done in 41.23%(n=40) cases. Preoperative hydrocephalus was found in 26.8%(n=26). Chiari -II malformation was associated in 78.4% (n=76) of the cases. Geographical distribution of the patients is shown in **Table 5**. Of the twenty-six cases with hydrocephalus, concurrent VP shunting was done in 25 cases followed by excision and repair. Additional 14 cases required VP shunting in post-operative period, 9 cases developed hydrocephalus without CSF leak and 5 developed hydrocephalus with CSF leak from surgical site so VP shunting was done.

Primary closure of the defect was possible in more than 90 % of the cases and different rotational skin flaps were done in rest of the case. Involvement of Plastic surgery team was made for the larger defect. No intervention was done for Chiari malformation in our series.

Surgical site infection was the most common postoperative complication 15.46% (n=15) followed by hydrocephalus 9.3% (n=9) and CSF leak 8.24% (n=8) as

shown in **Table 6**.

	Wound Infection	HCP	CSF leak	Shunt Mal-func-tion	Shunt infec-tion	Death
Emergency	1	0	4	0	0	1
Elective	14	9	4	6	4	3

Table 6: Post-operative Complications

Surgical site infection was managed by simple dressing in 12 cases and 3 cases required repeat debridement and primary closure. Postoperative hydrocephalus was managed with early ventriculoperitoneal shunt in 9 cases. Those cases of CSF leak(n=8) five cases required VP shunting and 3 case were managed with pressure dressing and simple measures like resuturing, avoiding supine position at most of time.

Six cases had post-operative VP shunt malfunction within 30 days and shunt revision was done in all. Shunt infection was seen in 4 cases. In these cases EVD was placed and re-shunted in those who had no infection in CSF analysis which was possible in 2 cases only. Post-operative mortality was witnessed in 4 cases. The first case had VP shunt infection who developed Ventriculitis and died. The cause of mortality in second and third case was hospital acquired pneumonia. In the fourth case, which was repaired in the emergency had surgical site infection and later on there was CSF leak for which EVD was placed and subsequently developed meningitis and later expired in PICU. The urodynamic study and neurophysiological monitoring was not done in our case series due to the unavailability of the resources in our center.

Discussion

Neural tube defects can occur anywhere along the neuroaxis from the developing brain to the sacrum.6 These can be divided into two main groups affecting cranial (anencephaly and encephalocele) or spinal structures (spina bifida).7 Although the incidence of these lesions has significantly decreased all over the world, particularly in developed countries; however, this is still a great problem in developing countries.9 Neural tube defects are etiologically heterogeneous.14 As with all the neural tube defects risk of recurrence after 1 affected child is 3-4 % and increases up to 10 % with 2 prior affected children. Nutritional and environmental factors do have role in etiology of myelomeningocele. Folate is intricately involved in the prevention and etiology of neural tube defects (NTDs.) Certain drugs which antagonize folic acid such as trimethoprim, Carbamazepine, Phenytoin, Phenobarbital and Valproic acid increased the risk of myelomeningocele.12 The US public health service has recommended that all women of child bearing age who are capable of becoming pregnant take 0.4mg of folic acid daily. If, however, a pregnancy is planned in high risk women, supplementation should be started with 4mg of folic acid daily beginning 1 mg before the time of planned conception. Maternal preconceptional use of folic acid supplementation reduces the incidences of NTDs by 50%. Fortification of food with 0.15mg folic acid /100gm was manded in United States and Canada in 1998.12

As per the data based from United State national surveillance system (1983-1990), the birth prevalence rate of myelomeningocele was slightly higher in females than in males (1.2:1).1In this present study males outnumbered female with ratio of 1.77:1. 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.4 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.1In the present study none of the cases were diagnosed in utero. In a study by Drolet B et al it was found that out of 155 cases of spinal dysraphism, 119 had open spina bifida [meningomyelocele (MMC) in 113 (72%), meningocele in 3 (2%) and myelocystocele in 3 (2%)] and 36 had occult spina bifida [split cord malformation (SCM) without overt MMC sac (pure SCM) in 29 (19%) and midline dermal sinus in 7 (4.5%)].3In the present study myelomeningocele was seen in 78.4%(n=76), lipomyelomeningocele was seen in 14.4%(n=14) ,Meningocele was seen in 4.1%(n=4), Split cord was seen in 2.1%(n=2) and Myelocestocele was seen in 1%(n=1).

In a study by Kumar R et al, lumbosacral (44%) region was the most common anatomical location of dysraphism followed by dorsolumbar (32%).13 In this study, commonest site was lumbar (41.2%), Lumbosacral regions (37.1%) and cervical 9.3% and thoracic, sacral and other comprising 12%. In about 90% of the children with myelomeningocele, hydrocephalus also occurs because the displaced cerebellum interferes with the normal flow of cerebrospinal fluid, causing an excess of the fluid to accumulate.15 In the present study about 80% cases had hydrocephalus. Significant hydrocephalus was found in only 26.8%(n=26). Total of 41.23%(n=40) cases required CSF diversion in the form of VP Shunting. Chiari -II malformation was associated in 78.4%(n=76) of the cases. In all the cases with significant hydrocephalus, concurrent VP shunting was done at the time of repair. Additional 14 cases required VP shunting in post-operative period, 9 cases developed hydrocephalus without CSF leak and

5 developed hydrocephalus with CSF leak from surgical site so VP shunting was done.

Thought the history of ANC visit was found to be positive in 56.7% (n=56). Number of mothers taking folic acid was relatively low. Only 37% (n=38.1) mother gave history of folic acid ingestion during conception. None of the pregnancy was planned. More than 70% percent of the patients were from the low economic status. They were more (58.8%) from Terai belt where arsenic is problem in drinking water **(Table 6)**. The relation between arsenic and NTDs could not be correlated due to limitation of the present study .More than 70 % of patient's father lived outside the country for earning livelihood.

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 setting not only can avoid CSF leak and pseudo-meningocele formation, but also reduces the cost of treatment.1In the modern armamentarium of neurosurgery. Endoscopic fetal surgery is performed but such facility is still questionable in developing countries like Nepal. There are many things to be addressed on the risk factor identification including objective absence of folic acid deficiency, correlation with maternal age and parity, previous spontaneous abortions, maternal obesity, maternal diabetes, and lower socioeconomic status. In the present study, socioeconomic status of the family was found to directly affect the incidence of the spinal dysraphism however large study is required to estimate the incidence. Fortification of foods with folic acid have been found to reduce the incidence of dysraphism by 50% in many studies. So, such policy if implemented in the countries like Nepal it will be a revolutionary change in the pediatric neurosurgery.

Conclusion

Spinal dysraphism is debilitating entity and represents a significant public health problem in developing countries. Although it is compatible with survival, most cases have moderate to severe disabilities, and may be associated with mental retardation. Folic acid supplementation during pregnancy has been shown to reduce the incidence and recurrence of many congenital defects including neural tube defects.

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