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To Study About Clinical Manifestations and Surgical Outcome in Cases of Spinal Dysraphism in Children Coming to Dr. Bramh / DksSs Hospital, Raipur with the Aim to Improve the Preventive and Management Strategy to Provide a Better Outcome

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ABSTRACT

The aim of the present study was to study about clinical manifestations and surgical outcome in cases of spinal dysraphism in children coming to Dr. BRAMH/DKS SS Hospital, Raipur with the aim to improve the preventive and management strategy to provide a better outcome. The present study was conducted at Dr. Bhimrao Ambedkar Memorial Hospital/ DKS SS Hospital, Raipur on Patients attending OPD or emergency and admitted in Dr. BRAMH/DKS SS Hospital, Raipur for the period of one year. 35 patients were included in the study. The most common presentation of spinal dysraphism is in neonates (<1 Month) constituting 74.28%. The next common age group of presentation is infants (1 Month-12 Month) constituting 14.28%. It least commonly presents in older age group, i.e., beyond 10 YEARS, constituting 2.85%. The incidence is higher in males (60%) when compared to females (40%).71.42% of mothers of affected children did not receive folic acid during pregnancy and only 28.57% of them received folic acid. Among the patients whose mothers did not take folic acid, 37.14% were females and 34.28% were males. This relation was found to be significant at p<.05. They present with complain of swelling in the back with or without associated neurological manifestations like motor weakness, sensory loss, neurogenic bladder, trophic ulcer. They are also associated with a plethora of bony deformities including CTEV, polydactyly, hip flexion contracture, calcaneous foot and scoliosis. The presence of occult disease represented by various cutaneous markers like hemangioma, dermal sinus, dermoid and hypertrichosis as noted in our study also needs to be paid heed to.

OPEN ACCESS

Key Words

Clinical manifestations, surgical outcome, spinal dysraphism in children, preventive management

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INTRODUCTION

Spinal dysraphism or neural tube defect (NTD) is a broad term including a heterogeneous group of congenital spinal anomalies, which result from defective closure of the neural tube early in fetal life and anomalous development of the caudal cell mass[1]. Some forms of spinal dysraphism can cause progressive neurologic deterioration.

The anatomic features common to the entire group is an anomaly in the midline structures of the back, especially the absence of some of the neural arches, and defects of the skin, filumterminale, nerves and spinal cord[2]. Spinal dysraphism can be classified as closed forms or open forms. Open form of Neural Tube Defects represents a serious congenital anomaly. If the neural tube fails to fuse at the skull, the result may be that of anencephaly or encephalocele. If the tube fails to fuse along the spine, the resulting defect is an open type of dysraphism such as meningomyelocele, myelocele, meningocele. Infants with NTDs frequently have additional serious neurological, musculoskeletal, genitourinary and bowel anomalies.

The open forms are often associated with hydrocephalus and Arnold chiari malformation type II and may be classified as spina bifida aperta[3,4]. The closed form of spina bifida is termed spina bifida occulta. 5-10% of the general population may have bony spina bifida occulta with intact overlying skin. Most of these cases are found incidentally. Spina bifida occulta is characterized by variable absence of several neural arches and various cutaneuous abnormalities such as hemangioma, cutis aplasia, dermal sinus, or hairy patch and diastematomyelia or a lipoma (lipomyelomeningocele).

Patients with spina bifida occult may present with scoliosis in later years. This is often associated with low lying conus and other spinal cord anomalies. Whenever the conus lies below the L2-L3 interspaces in an infant, cord tethering should be considered. The term tethered cord implies that the cord may be attached to vertebral column or subcutaneous tissues by a thickened filum terminal or fibrousband[5]. Approximately 95% of couples that have a fetus affected with NTD have a negative family history. Most NTDs are caused by multi factorial inheritance, including genetic and environmental factors[6].

The aim of the present study was to study about clinical manifestations and surgical outcome in cases of spinal dysraphism in children coming to Dr. BRAMH/DKS SS Hospital, Raipur with the aim to improve the preventive and management strategy to provide a better outcome.

MATERIALS AND METHODS

The present study was conducted at Dr. Bhimrao Ambedkar Memorial Hospital/ DKS SS Hospital, Raipur on Patients attending OPD or emergency and admitted in Dr. BRAMH/DKS SS Hospital, Raipur for the period of one year. 35 patients were included in the study.

Inclusion Criteria:

- All the patients diagnosed as spinal dysraphism and admitted in Dr. BRAMH/DKS SS Hospital, Raipur.
- Children aged 0-18 yrs.

Exclusion Criteria:

- Patients with incomplete follow up.
- Patients not giving consent.
- Patients with associated congenital anomalies of other organs (eg. TOF).

Study Tools:

- **Clinical:** History and clinical examination.
- Laboratory and radiological Investigations.

Study Technique: 35 patients were enrolled, written informed consent was taken from parents of all the patients after which detailed history was taken including demographics, antenatal care received and recorded by intake of folic acid and antenatal USG, mode of delivery. All the patients were thoroughly examined to assess site and type of lesion, neurological examination, sensorimotor function, bowel and bladder function and orthopedic anomalies.

The patients were evaluated with routine blood parameters, serology, transfontanelle USG, USG local site, CT head, MRI spine. However, not all the investigation was possible in all the patients and in some none of the investigation could be done.

After explaining the complexity of the disease, all aspects of treatment, complications and long term problems associated with this disease to the parents, informed consent was taken and patients were operated. 13 patients in our study underwent surgical correction by anatomical repair using standard techniques.

The patients were followed up postoperatively for a period of 3 months and assessed for cosmesis, sensorimotor function to see improvement or deterioration, bladder and bowel function and complications were noted.

Data Analysis: Descriptive statistics were used to present categorical data as percentages and frequencies.

RESULTS AND DISCUSSIONS

The most common presentation of spinal dysraphism is in neonates (<1 Month) constituting 74.28%. The next common age group of presentation is infants (1 Month-12 Month) constituting 14.28%. It least

Table 1: Demographic Data

Age	Open	Closed	Total
<1 Month	17 (48.57%)	9 (25.71%)	26 (74.28%)
1 Month-12 Month	1 (2.85%)	4 (11.42%)	5(14.28%)
1 Year-10 Year	1 (2.85%)	2 (5.71%)	3 (8.57%)
>10 Year	0(0%)	1 (2.85%)	1 (2.85%)
Gender			
Male	12(34.28%)	9(25.71%)	21(60%)
Female	7(20%)	7(20%)	14(40%)
Antenatal USG			
YES	5(14.28%)	4(11.42%)	9(25.71%)
NO	14(40%)	12(34.28%)	26(74.28%)
Mode of Delivery			
NVD	14(40%)	12(34.28%)	26(74.28%)
LSCS	5(14.28%)	4(11.42%)	9(25.71%)

commonly presents in older age group, i.e., beyond 10 Years, constituting 2.85%.The incidence is higher in males (60%) when compared to females (40%). The antenatal USG was not done in mothers of 74.28% children. Out of them 40% children presented with open spinal dysraphism and 34.28% children presented with closed spinal dysraphism. Antenatal USG was done in mothers of 25.71% children of which 14.28% presented with open spinal dysraphism and 11.42% presented with closed spinal dysraphism.74.28% children were born by normal vaginal delivery. Out of them 40% had open spinal dysraphism and 34.28% had closed spinal dysraphism. Only 25.71% children were born by caeserian section out of which 14.28% had open and 11.42% had closed spinal dysraphism.

Table 2: Folic acid Intake and Type of Spinal Dysraphism

Folic Acid Intake	Male	Female	Total
Yes	9(25.71%)	1(2.86%)	10(28.57%)
No	12(34.28%)	13(37.14%)	25(71.42%)
Total	21(60%)	14(40%)	35(100%)
Type of Spinal Dysraphism			
Open	12(34.28%)	7(20%)	19(54.28%)
Closed	9(25.71%)	7(20%)	16(45.71%)

71.42% of mothers of affected children did not receive folic acid during pregnancy and only 28.57% of them received folic acid. Among the patients whose mothers did not take folic acid, 37.14% were females and 34.28% were males. This relation was found to be significant at p<.05.

Table 3: Type of lesion

Lesion	Male	Female	Total
Meningomyelocele	11(31.42%)	8(22.86%)	19(54.28%)
Meningocele	3(8.57%)	4(11.43%)	7(20%)
Lipomyelomeningocele	3(8.57%)	2(5.71%)	5(14.28%)
Lipomyelocele	1(2.86%)	0(0%)	1(2.86%)
Encephalocele	3(8.57%)	0(0%)	3(8.57%)
Total	21(60%)	14(40%)	35(100%)

The most common lesion is meningomyelocele in 54.28% patients, followed by meningocele in 20% patients, lipomyelomeningocele in 14.28%, encephalocele in 8.57% and least common being lipomyelocele in 2.86%.

Table 4: Bladder Involvement and Type of Surgery

Bladder Involvement	Open	Closed	Total
Expressible	6(17.14%)	2(5.71%)	8(22.85%)
Non Expressible	13(37.14%)	14(40%)	27(77.14%)
Type of Swelling			
Ruptured	12(34.28%)	1(2.85%)	13(37.14%)
Unruptured	7(20%)	15(42.86%)	22(62.86%)

There was no bladder involvement in 77.14% patients. In 22.85% patients bladder was expressible signifying neurogenic bladder in which 17.14% were of open type and 5.71% were of closed type. The table represents the presentation of swelling. 62.86% presented with unruptured swelling, i.e., skin overlying the swelling was intact and it was ruptured in 37.14% patients. Unruptured swelling are mostly closed spinal dysraphism constituting 42.86% and ruptured swelling are open dysraphism constituting 34.28%.

Table 5: Motor Weakness and Level of Spinal Defect

Motor Weakness	Open	Closed	Total
No Weakness	12(34.28%)	9(25.71%)	21(60%)
Weakness Present	2(5.71%)	6(17.14%)	8(22.86%)
Paralysis	5(14.28%)	1(2.86%)	6(19.35%)
Level Of Spinal Defect			
Occipital	3(8.57%)	6(17.14%)	9(25.71%)
Lumbosacral	11(31.43%)	7(20%)	18(51.42%)
Dorsolumbar	3(8.57%)	1(2.85%)	4(11.43%)
Sacral	2(5.71%)	2(5.71%)	4(11.43%)

22.86% patients presenting with motor weakness were noticed in bilateral lower limbs. 5.71% of them have open spinal dysraphism and 17.14% had closed spinal dysraphism. 14.28% patients with open spinal dysraphism presented with paralysis whereas 2.86% patients presented with complete paralysis of bilateral lower limbs. Lumbosacral area is the commonest constituting 51.42%, followed by occipital 25.71% and dorsolumbar and sacral constituting 11.43% each.

Table 6: Complications

Complications	No. of Patients	Frequency
Wound Infection	3	8.57%
CSF Collection/Leak	3	8.57%
Hydrocephalus	0	0%
Death	2	5.71%

There were 2 cases of death and wound infection and CSF collection in 3 patients each.

The term spinal dysraphism or neural tube defect (NTD) covers a diverse range of congenital spinal malformations brought on by early defects in the closure of the neural tube and abnormal caudal cell mass development[7]. Progressive neurologic decline can result from several types of spinal dysraphism. Anatomical abnormalities in the midline structures of the back, particularly the absence of some neural arches, as well as flaws in the skin, filumterminale, nerves and spinal cord are characteristics shared by the entire group[2].

In this study most common presentation of spinal dysraphism is in neonates (<1 month) constituting 74.28%. It least commonly presents in older age group, i.e., beyond 10 years, constituting 2.85%. This is similar to study by Premlal[9]. where most common age group of presentation was neonates below 1 month (37.5%) followed by infants (21.9%).In a study done by Venkatesh[10], 73% patients presented at neonatal age group. The disease is more common in males (60%) compared to females (40%) in this study. This is similar to study by Ahmad[11]. in which 61% were males and 39% were females. This is in contrast to the study done by Premlal[9]. who shows higher incidence in females (53.13%) compared to males (46.87%).

The etiology of spinal dysraphism has been proven to be closely related to folic acid deficiency. This study shows, mothers of 71.42% patients did not receive folic acid during the antenatal period. Only 28.57% of patients took folic acid supplementation during antenatal period for a variable period of 1-3 months. None of them were aware of antenatal care facility provided by anganwadi in respective localities. Also, all of them started taking folic acid after 1 month by which time the development of neural tube has already completed. This was because of delay in the knowledge of established pregnancy. The awareness of the benefits of preconception folic acid supplement is still not optimum. This study enlightens the poor compliance of females to folic acid in remote areas of Chhattisgarh and the need to devise more intensive approach for folate supplementation owing to the ignorance of population of this area. The study by Asindi[12]. reported that 75% of the mothers with affected pregnancy did not take folic acid and only 25% of them took folic acid supplementation. The most common mode of delivery in our study was normal vaginal delivery (74.28%) and LSCS was done in mothers of only 25.71% children. This is in contrast to study by Melekoglu[12] who demonstrated that the mode of delivery in 79.7% of patients with NTD's was the cesarean section and the presence of neural tube defects was the most common indication for cesarean delivery. Inconsistently, some observational studies suggesting that cesarean delivery may provide a better outcome for babies with meningomyelocele (MMC) existed. These studies suggested that cesarean delivery would be beneficial by keeping MMC sac intact, reducing bacterial contamination of the exposed neural tissue and also allow for a better preparation of the team (i.e., neonatologist and pediatric neurosurgeons).

In a study conducted by Venkatesh[10], 22 out of 74 patients presented with neural deficits, 4 patients had only lower limb motor and sensory loss, 2 out of these 4 showed Grade 2 paraparesis whereas the other 2 had complete paraplegia. 6 patients presented only with bladder bowel incontinence with no signs or symptoms suggestive of lower limb weakness. 12 out of 22 presented with a combination of paraplegia and bowel and bladder incontinence. Premlal[9] reported most common anomaly as hydrocephalus in 31.25% cases, followed by Arnold chiari malformation II in 25% cases, and low tethered cord in 15.62% cases. He also reported syringomyelia, thickened filumterminale, syringohydromyelia, corpus callosal agenesis, sacral agenesis and arachnoid cyst.

CONCLUSION

They present with complain of swelling in the back with or without associated neurological manifestations like motor weakness, sensory loss, neurogenic bladder, trophic ulcer. They are also associated with a plethora of bony deformities including CTEV, polydactyly, hip flexion contracture, calcaneous foot and scoliosis. The presence of occult disease represented by various cutaneous markers like hemangioma, dermal sinus, dermoid and hypertrichosis as noted in our study also needs to be paid heed to. Early surgical intervention is essential to achieve improvement in neurological manifestations. Operative complications like wound infection, CSF collection needs immediate attention and intervention.

REFERENCES

1. Tortori, D.P., A. Rossi and A. Cama, 2000. Spinal dysraphism: A review of neuroradiological features with embryological correlations and proposal for a new classification. Neuroradiology, 42: 471-491.
2. Williams, A.L. and R.M. Quencer, 1988. The abnormal spinal cord: Current imaging status. In 74th Scientific Assembly and Annual Meeting of RSNA,
3. Sekhar, I. and B.R. Kumar, 2022. A Study on the Incidence, Clinical Presentations, and Surgical Outcome in Spinal Dysraphism Patients. Euro Jou Molec Clin Med., 9: 1940-1955.
4. JJ, V., 1995. Neural tube formation and prosencephalic development. Neur thenew, 30-42.
5. Kumar, R. and N. Singhal, 2006. Outcome of meningomyelocele/ lipomeningomyelocele in children of northern India. Pediatr. Neurosurg., 43: 7-14.
6. Bowman, R.M., D.G. McLone, J.A. Grant, T. Tomita and J.A. Ito, 2001. Spina bifida outcome: A 25-year prospective. Pediatr. Neurosurg., 34: 114-120.

7. KV, P., 2018. A Retrospective Analysis of Clinical Profile and Surgical Outcome in Patients with Spinal Dysraphism at Tertiary Care Center. Intern Arch Integ Med, Vol. 5, No. 5.
8. Venkatesh, V., A.E. Curley and S. Stanworth, 2015. The Neonate. Disorders of Thrombosis and Hemostasis in Pregnancy: A Guide to Management. 391-409.
9. Ahmad, M., 2023. Psychosocial Effects of COVID-19 among General Population of Khyber Pakhtunkhwa. Intern Jou Psych Beha Res, Vol. 3, No. 2.
10. Asindi, A. and K. Eyong, 2021. Folic acid and brain function in childhood. Niger. J. Paediatrics, 48: 88-92.
11. Melekoglu, R., S. Eraslan, E. Celik and Y. Simsek, 2016. Perinatal and neonatal outcomes of patients who were diagnosed with neural tube defect in midtrimester fetal ultrasound scan and refused request for termination of pregnancy. BioMed Res. Int., 2016: 1-6.