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Original Research Article

Spinal dysraphism in children: 3 years experience

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ABSTRACT

Introduction: Spinal dysraphism (SD) is a congenital malformation of the spine and spinal cord resulting from congenital aberrations during the gastrulation, primary neurulation, and secondary neurulation stages. SD is the most common congenital anomaly of the CNS. Our study outlines the demography and clinical profile of this spectrum.
Aim of the study: The aim of the study was to assess the demographic characteristics and clinical features of spinal dysraphism in children over a three-year period.
Methods: This retrospective study involved 38 children with spinal dysraphism at the Department of Pediatric Surgery, Bangabandhu Sheikh Mujib Medical University (BSMMU) from January 2021 to December 2023. Inclusion criteria included complete clinical records and surgical intervention. Diagnostic methods comprised history, clinical exams, USG, CT, and MRI. Postoperative follow-up was assessed. Data were analyzed using SPSS version 22.0.
Result: The study included 38 patients aged from 4 days to 7.5 years. The cohort exhibited a slight female predominance (57.89%). Myelomeningocele was the most predominant anomaly (52.63%), followed by lip myelomeningocele (31.58%) and meningocele (13.16%). Lesions were predominantly located in the lumbosacral region (92.11%). All patients presented with a swelling on the back. Associated conditions included hydrocephalus (34.21%), CSF leak (23.68%), and bowel and bladder incontinence (15.79%). All cases underwent an operative procedure with anatomical repair using standard techniques.
Conclusion: Among spinal dysraphism, myelomeningocele was more common than other spinal defects. All patients presented with back swelling, with or without associated symptoms.

Key words: Spinal dysraphism, Surgical repair, Neonatal spinal conditions, Retrospective study, Magnetic resonance imaging (MRI)

INTRODUCTION

Spinal dysraphism encompasses a range of congenital spinal anomalies arising from developmental defects during embryogenesis, particularly due to disruptions in the complex process of spinal axis formation between the 2nd and 6th weeks of gestation.[1] This condition results from incomplete midline closure of mesenchymal, osseous, and nervous tissue during neurulation, involving both primary and secondary stages of neural tube development. Spinal dysraphism is categorized into two types aperta, which involves visible lesions, and occulta, which does not present with external lesions. As the most predominant congenital anomaly of the central nervous system (CNS), spinal dysraphism has substantial implications for clinical outcomes, including neurogenic bladder.[2] Children with spinal dysraphism experience a wide array of complications, making it a critical public health concern that affects not only the individuals but also their families and communities.[3] These

complications include Arnold-Chiari malformation, neurogenic lower limb paralysis, neurogenic bladder and bowel dysfunction, congenital clubfoot, hip dislocation, fractures, scoliosis, hydronephrosis, and vesicourethral reflux.[4] Spinal dysraphism includes a spectrum of anomalies that can be classified into open and closed subtypes based on whether the neural elements are exposed or covered by skin.[5] In open spinal dysraphism (OSD), the neural tissues are exposed, while in closed spinal dysraphism (CSD), the neural elements remain beneath intact skin.[6] The condition can present as Spina bifida cystica or Spina bifida occulta, where the defect may involve a cystic protrusion containing abnormal meninges (meningocele) or elements of the spinal cord and nerves (myelomeningocele).[7] Spinal dysraphism (SD) refers to a spectrum of congenital malformations affecting the spine and spinal cord, with myelomeningocele being the most predominant form.[8] This defect often presents as a cystic protrusion containing meninges, cerebrospinal fluid, and neural elements.

Research has consistently identified key risk factors for SD, including female sex, and a family history of neural tube defects.[9] However, the evidence for other potential risk factors remains inconclusive, highlighting gaps in the current understanding of the condition. The occurrence of SD varies across populations, with higher rates observed among newborn females in certain groups.[10] Clinical manifestations of SD can range from overt external signs to more subtle presentations that may not be detected until later in childhood, as seen in spina bifida occulta. Early diagnosis is essential, particularly in conditions like tethered cord syndrome, to prevent irreversible neurological damage.[11] Despite the well-documented clinical manifestations and risk factors associated with spinal dysraphism, there remains a need for comprehensive studies to better understand the detailed characteristics of this condition in diverse pediatric populations.

Therefore, the purpose of this study is to evaluate the demographic characteristics and clinical features of spinal dysraphism in children over a three-year period, aiming to provide valuable insights that can inform management strategies and improve outcomes.

The aim of the study was to assess the demographic characteristics and clinical features of spinal dysraphism in children over a three-year period.

METHODS

Study type

This retrospective observational study was conducted at the Department of Pediatric Surgery, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh

Study duration

Study duration was of 3-year period from January 1, 2021, to December 31, 2023. The study included 38 children diagnosed with spinal dysraphism during this period.

Inclusion criteria

Children diagnosed with spinal dysraphism aged between 4 days and 7.5 years. Patients who underwent surgical intervention for spinal dysraphism.

Exclusion criteria

Patients with incomplete records. Children with other significant congenital abnormalities that could confound results. Informed consent was obtained from the parents or guardians, ensuring confidentiality and voluntary participation. Patients were diagnosed through detailed history-taking, clinical examination, and relevant laboratory investigations.

Ultrasonography (USG) was performed in all patients as a baseline imaging modality, while computed tomography (CT) of the skull was conducted to exclude hydrocephalus, and magnetic resonance imaging (MRI) was utilized to evaluate for tethered cord and other spinal anomalies. All cases underwent surgical procedures aimed at anatomical repair using standard techniques, including sac repair, cord detethering, and management of associated conditions such as hydrocephalus.

Postoperative care was provided to monitor recovery and address any complications. Patients were followed up to assess neurological recovery, and the need for additional interventions, with follow-up visits including clinical evaluations and, when necessary, imaging studies to monitor the integrity of the repair and the patient's overall health.

Data collection

Data were collected retrospectively from medical records, encompassing demographic characteristics, clinical presentation, imaging findings, and associated conditions.

Statistical analysis

Statistical analysis was performed using SPSS version 20, with descriptive statistics summarizing demographic characteristics, clinical presentation, and surgical outcomes.

RESULTS

The age distribution revealed that the majority of patients, 14 (36.84%), were between 4 days and 1 month old. Additionally, 8 (21.05%) patients were between 1 month and 1 year old, 6 (15.79%) were between 1 and 3

years old, and 4 (10.53%) were in both the 3 to 6 years and 6 to 7 years age groups. Only 2 (5.26%) patients were older than 7 years. Regarding gender distribution, there was a slight female predominance, with 22 (57.89%) females compared to 16 (42.11%) males, resulting in a female-to-male ratio of approximately 1.4:1.

Table 1: Demographic characteristics of patients with spinal dysraphism (n=38).

Variables		Frequency	%
Age	4 days to 1 month	14	36.84
	1 month to 1 year	8	21.05
	1 year to 3 years	6	15.79
	3 years to 6 years	4	10.53
	6 years to 7 years	4	10.53
	>7	2	5.26
Gender	Male	16	42.11
	Female	22	57.89

Table 2: MRI findings of the study patients (n=38).

Anomaly	Number of patients	%
Myelomeningocele	20	52.63
Lipomyelomeningocele	12	31.58
Meningocele	5	13.16
Total	38	100.00

The distribution of spinal dysraphism anomalies indicated that myelomeningocele was the most predominant condition, affecting 20 (52.63%) of the patients. Lipomyelomeningocele was observed in 12 (31.58%) patients, while meningocele was noted in 5 (13.16%) patients.

Table 3: Location of lesion in patients with spinal dysraphism (n=38).

Location of lesion	Number of patients	%
Lumbosacral	35	92.11
Sacral	2	5.26
Thoracic	1	2.63
Total	38	100.00

The majority of lesions were located in the lumbosacral region, with 35 (92.11%) patients affected. Sacral lesions were present in 2 (5.26%) patients, and thoracic lesions were observed in 1 (2.63%) patient.

Among the patients, 13 (34.21%) had associated hydrocephalus, while 9 (23.68%) experienced CSF leak. Bowel and bladder incontinence was noted in 6 (15.79%) patients, and hypotonia and paraparesis were present in 4 (10.53%) patients.

Table 4: Associated anomaly in patients with spinal dysraphism.

Anomaly	Number of patients	%
Associated hydrocephalus	13	34.21
CSF leak	9	23.68
Bowel and bladder Incontinence	6	15.79
Hypotonia and paraparesis	4	10.53

DISCUSSION

The findings of this study offer valuable insights into the occurrence and presentation of spinal dysraphism in children. By analyzing data collected over a three-year period, we have identified key trends and variations in how this condition manifests. This discussion will contextualize our results within the framework of existing research, comparing our observations with previously established patterns. Our goal is to deepen the understanding of spinal dysraphism and its broader implications for diagnosis and management in pediatric care.

In our study, the age distribution of spinal dysraphism patients revealed that the majority were between 4 days and 1 month old, which aligns with findings from other studies. For instance, Premlal et al observed that 37.5% of their patients were under one month old, indicating that early infancy is a common period for diagnosis.[12] Similarly, Rao et al reported a notable frequency of spinal dysraphism in patients under one year, with the youngest patient being one day old.[13] This pattern suggests that spinal dysraphism is frequently detected in the neonatal period, reinforcing the importance of early screening.

Our study also noted a slight female predominance, with 57.89% females compared to 42.11% males. This finding is consistent with the results of Rao et al, who found a female predominance of 57.40%.[10,13] Additionally, Hossain et al reported a similar female preponderance of 54% among their study subjects, and Anderson et al noted that 20 out of 27 patients were female, reflecting a notable female dominance.[14] These consistent findings across various studies suggest a possible gender predisposition in the occurrence of spinal dysraphism, highlighting the need for tailored diagnostic and management strategies for females in this population.

In our study, myelomeningocele emerged as the most predominant anomaly, affecting 20 (52.63%) of the patients, followed by lipomyelomeningocele in 12 (31.58%) and meningocele in 5 (13.16%). These findings align closely with those reported in other studies. Premlal et al also identified myelomeningocele as the most common condition, observed in 46.87% of their patients, with lipomyelomeningocele and meningocele being less frequent.[12] Similarly, another study reported that

myelomeningocele was the predominant anomaly, affecting 61.11% of their cases.[13] This consistency across studies underscores the predominance of myelomeningocele in the spectrum of spinal dysraphism, while also highlighting the relative frequency of other conditions such as lipomyelomeningocele. These results reinforce the importance of accurate diagnosis and management strategies tailored to the specific types of spinal dysraphism encountered. In our study, the majority of lesions were localized to the lumbosacral region, affecting 35 (92.11%) patients. This finding is consistent with results from other studies that also highlight the lumbosacral region as the most common site for spinal dysraphism. One study identified the lumbosacral region as the most frequent site of occurrence in 51.85% of patients, while another study reported a frequency of 34.37% in the same region.[10,12,13] Similarly, Hossain et al found that 83.33% of their study subjects had lesions in the lumbosacral area. This is in accordance with the study of Agrawal et al, where 67.8% of patients had lesions in the lumbosacral region.[15]

Additionally, Guggisberg et al, conducted a study evaluating the diagnostic value of lumbosacral midline cutaneous lesions in asymptomatic children to detect occult spinal dysraphism (OSD), proposing a practical approach for clinical investigations based on observed cutaneous lesions. These findings underscore the predominance of the lumbosacral region in spinal dysraphism cases and emphasize the need for targeted diagnostic and therapeutic approaches for lesions located in this specific region.[16]

In our study, all 38 patients (100.00%) presented with a swelling over the back. This consistent presentation highlights the critical role of visible external signs in the diagnosis of spinal dysraphism. The universal occurrence of this symptom underscores the importance of early identification of such anomalies, as back swelling can serve as a key diagnostic indicator. Timely recognition and intervention based on these external signs are crucial for improving patient outcomes and managing potential complications more effectively.

In our study, 34.21% of patients had associated hydrocephalus, 23.68% experienced CSF leak, and 15.79% exhibited bowel and bladder incontinence. These findings are consistent with other studies that have reported similar associated anomalies. Premlal et al noted that hydrocephalus was observed in 31.25% of their patients, while another study highlighted hydrocephalus as the most common associated anomaly, present in 42.59% of cases. Additionally, the latter study found CSF leak to be the most frequent post-operative complication, occurring in 20.37% of cases.[12,13] Our findings on the occurrence of bowel and bladder incontinence and hypotonia are consistent with the range of complications reported in the literature, highlighting the importance of comprehensive management strategies to effectively address these associated conditions.

In our study, all 38 patients underwent operative procedures, with anatomical repair performed using standard techniques. This uniform approach ensures that the surgical interventions align with established practices, providing a consistent basis for evaluating outcomes. By adhering to standard techniques, we aimed to optimize surgical results and minimize complications, thereby enhancing the overall management of spinal dysraphism.

This study had several limitations, small sample size may limit the generalizability of the findings. Single-center study might introduce bias in the results. Potential for selection bias due to the retrospective nature of the study.

CONCLUSION

Our study highlights that myelomeningocele was the most common spinal dysraphism among the patients, significantly outnumbering other types of spinal defects. All patients presented with a back swelling, with varying associated symptoms such as hydrocephalus, CSF leak, and bowel or bladder incontinence. These findings underline the critical importance of early diagnosis and comprehensive treatment approaches. Addressing both the primary defect and associated complications is essential for optimal management and improving outcomes for children with spinal dysraphism.

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