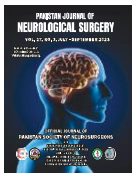

PAKISTAN JOURNAL OF NEUROLOGICAL SURGERY (QUARTERLY) – OFFICIAL JOURNAL OF PAKISTAN SOCIETY OF NEUROSURGEONS

Original Article

Incidence and Outcomes of Diastematomyelia in Spina Bifida Patients

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ABSTRACT

Objective: To determine the prevalence of diastematomyelia in spina bifida patients and to assess the efficacy of surgical intervention.

Material and Methods: This prospective research study was conducted at the Jinnah Postgraduate Medical Center in Karachi in the Neurosurgery department. We included 55 patients after fulfilling the inclusion criteria. All of the patients had craniospinal MRI, and the results, as well as any anomalies discovered, were noted for future reference during their therapy. Patients suffering from these diseases were treated surgically, which included sac excision and repair, cord detethering, and ventriculoperitoneal shunting. Throughout the postoperative period, all of these patients' outcomes were documented and assessed.

Results: The majority of patients were under 1 month old (29 patients, 53.70%), whereas 13 patients were between one month and 1 year old. The patients were 2.8 years old on average. There were 23 males (42.60%) and 32 females (58.18%). Dermal sinuses, hypertrichosis, and skin dimples (signs of spina bifida occulta), with prevalence rates of 5.55 percent, 3.70 percent, and 1.85 percent, respectively. Spina bifida occulta was less frequent (17 cases) than spina bifida aperta (37 occurrences). 33 patients (61.11%) have myelomeningocele, followed by meningocele in three (5.5%), lipomyelomeningocele in six (10.9%), diastematomyelia in six (10.9%), dermal sinus in two (3.70%), along with spinal lipoma in one (1.85%) instance.

Conclusion: The overall prevalence of Diastematomyelia in patients with spinal dysraphism is low. However vigilant assessment and management is crucial for optimal surgical benefit.

Keywords: Diastematomyelia, Spina Bifida, Spinal Dysraphism.

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Date of Submission: 10-05-2023
Date of Revision: 31-08-2023
Date of Acceptance: 01-09-2023
Date of Online Publishing: 30-09-2023
Date of Print: 30-09-2023

DOI: 10.36552/pjns.v27i3.891

INTRODUCTION

A neural tube defect, also known as spinal dysraphism, is a collective term for a variety of congenital spinal abnormalities caused by the premature or incomplete closure of the neural tube and the subsequent aberrant growth of the

caudal cell mass.[1] Some types of spinal dysraphism might result in a gradual neurologic decline. It is common among all members of the group to have some abnormality in middle back structures, including the lack of some neural arches. There can also be anomalies in the brain in addition to those in the skin, spinal cord, nerves, and filum terminale.[2] Anencephaly or encephalocele can develop if the neural tube does not close properly at the skull. Open forms of dysraphism, such as meningomyelocele, and meningocele, occur when the tube fails to fuse down the spine. Many infants with neural tube defects often have additional, severe defects in the areas of the nervous system, muscles, bones, genitourinary system, and digestive system. The open varieties, which are associated with type II Arnold Chiari malformation and hydrocephalus, could be categorized as spina bifida aperta.[3-4]

When it is closed, the condition is known as spina bifida occulta. It is estimated that between 5 and 10 percent of the population has skeletal spina bifida occulta with normal skin underlying the defect. There are a variety of cutaneous abnormalities that can identify occult spina bifida, including hemangiomas, cutis aplasia, dermal sinus, hairy patches, diastematomyelia, and lipomas. Cord tethering may be observed in infants with a conus below the L2-L3 interspace.

Clinical signs of diastematomyelia, a disorder in which two hemicords are fused together, comprise tethered cord syndrome and scoliosis. Physical examination typically reveals a tuft of hair in the back. A dural-sheathed inflexible osseocartilaginous (bony) median septum divides two hemicords of type I, each of which is contained within a distinct dural tube and has its central canal and surrounding pia. A nonrigid fibrous median septum divides two hemicords that are contained within a single dural tube in type II.[6-7] Myelomeningocele and myelocele are brought on by a problem with the main neural tube's sealing. The second is extremely uncommon. Myelomeningocele accounts for nearly nine out of 10 documented instances.

In most cases, patients have no symptoms and there are no outward indications of a tethered chord secondary to diastematomyelia.[8] Pain in the lower back and legs is a common sign of tethered cord syndrome. Hip dislocations, club feet, and high-arched feet are examples of orthopedic and urological symptoms that fall into a separate group. The majority of children with myelomeningocele also experience urodynamic abnormalities.[8] The most frequent presenting symptoms include lumbar cutaneous abnormalities, enuresis, unexplained leg or back pain, limb length discrepancies, aberrant foot development, and scoliosis. The most prevalent symptom in children with pediatric tethered cord syndrome is weakness, which is followed by gait disorders (54%), spinal scoliosis (51%), limb discomfort (32%), skeletal abnormalities (11%), and urinary troubles (6%). The plan is surgery, remove the bony spur or fibrous band first then detether the spinal cord. We have to form a single neural tube. The current study investigated how often diastematomyelia is in spina bifida patients and how effective surgical treatment is.

MATERIALS & METHODS

Study Design and Setting

This study is prospective and observational. Starting in October 2022 till April 2023, the trial was conducted in the neurosurgery department of the Jinnah Postgraduate Medical Center in Karachi. The study considered 55 patients with spinal dysraphism in total.

Inclusion Criteria

Inclusion criteria included all patients having spinal dysraphism with complete radiological evaluation.

Exclusion Criteria

Exclusion criteria were composed of patients who

did not follow up with radiological imaging and those who did not consent to be a part of the study.

Data Analysis

A predesigned proforma was used to record patient demographics, presenting complaints, neurological deficit, examination, and radiological findings of both spinal and cranial. This included the location of spina bifida, diastematomyelia, and the presence of cord tethering. Other congenital anomalies were also documented. All of the patients had craniospinal MRI, and the results, along with any abnormalities found, were recorded for future reference in the course of their care. Patients with these conditions received the appropriate surgical treatment, which included sac excision and repair, cord detethering, and ventriculoperitoneal shunting. Outcomes were recorded and evaluated for all of these individuals as they went through the postoperative period. Included in this research are patients who had at least three months of follow-up.

RESULTS

Age Distribution

In this research study, 54 patients were enrolled. Throughout our study, each participant's age ranged from one day to 17 years old. 29 patients, or 53.70%, were under one month old, while 13 patients were one year old or older. The study participants' age distribution had a mean of 2.8 years.

Gender Distribution

It has been shown that the incidence is much higher in girls (n=31, 57.40%) than in boys (n=23, 42.60%).

Types of Dysraphism

Spina bifida occulta is less common than spina bifida aperta (open forum), which affects 15 patients (27.7%) out of 54 cases. In 72.22% (39) of the instances, there was an open-form spina bifida. See Table 1.

Table 1: The Different Forms of Dysraphism.

Types	Number of Cases	Percentage
Closed	15	27.7
Open	39	72.22

Location of Spina Bifida

See Table 2. The lumbosacral area was affected in 46.29% of patients, making it the most prevalent place. The dorso-lumbar region affects 17 (31.48%) patients the next most frequently. Spina bifida is seen in the upper dorsal area of six children (11.11%) and the sacral region of two children (3.70%) of the population. This abnormality in the cervical area was found in four individuals or 7.40% of the total.

Table 2: Localization of the Lesion.

Location	No. of Case	Percentage %
Sacral	02	3.70
Cervical	04	7.40
Dorsal	06	11.11
Lumbosacral	25	46.29
Dorsolumbar	17	31.48

Clinical Features

In total, 54 cases were examined, and 33 (61.1%) of those cases had a pronounced protrusion in the middle of the back. Neuro orthopedic foot anomalies, such as talipes equinovarus, high-arched feet, leg length discrepancies, and flat feet, were seen in 11 (20.3%) of the cases 5.5% of people had scoliosis that was considered to be significant. Nine people (16.6%) reported having severe back pain. The most prevalent neurological

problem, affecting 28 (51.8%) people, was weakness in the lower limbs. Twenty people (37.0%) had sensory loss; it was discovered. Incontinence of the urine or feces was one of the sphincter dysfunctions that were discovered in 18 (33.3%) of the sample. Muscular atrophy (10, 18.51%) and trophic ulcers (3, 5.5%) were the additional neurological dysfunctions noted. See Table 3.

Table 3: The Clinical Features.

CF	Number of Cases	%age
Cutaneous		
Swelling skin covered	22	40.7
Ruptured swelling	07	12.9
Scar of previous surgery	05	9.2
Subcutaneous Lipoma	05	9.2
Dermal Sinus	03	5.5
Hypertrichosis	02	3.7
Dimple over skin	03	5.5
Orthopedic		
Scoliosis	03	5.5
Backache	09	16.6
Foot-deformities	11	20.3
Neurologic:		
Trophic Ulcerations	03	5.5
Sensory weakness	20	37.0
Motor deficits	28	51.8
Muscle atrophy	10	18.5
Sphincter disturbances	18	33.3

MRI Findings

33 cases of myelomeningocele (61.11%) have been reported. Six cases of lipomyelomeningocele (11.11%), three cases of meningocele (5.55%), six cases of diastematomyelia (11.11%), additionally, two cases of the cutaneous sinus (3.70%) and one case of spinal lipoma (1.85) have been reported. The most frequent secondary abnormality was hydrocephalus, which affected 22 patients (40.7%), followed by Arnold chiari malformation type 2 in 20 people (37.0%). The low-lying tethered chord (12; 22.2%) and syringomyelia (17) were both discovered in this sample. The less common arachnoid disorders were arachnoid cysts (1.85%), corpus callosal thinning (1.85%), arachnoid fistulas (4.74%), syringohydromyelia (3, 5.5%), corpus callosal agenesis (4, 7.4%), sacral agenesis in 2 (3.70%), and thickened filum terminale (2, 3.7%).

Table 4: Findings of MRI.

Congenital Anomaly:	Case Count	%
Spinal lipomas	01	1.85
Myelomeningocele	33	61.1
Lipomyelomeningocele	06	11.1
Meningocele	03	5.5
Diastematomyelia	06	11.1
Dermal sinus	02	3.7
Associated Anomalies:		
Corpus callosal thinning	01	1.85
Arnold chiari malformation type 11	20	37.0
Hydrocephalus	22	40.7
Syringomyelia	17	31.5
Low tethered cord	12	22.2
Syringohydromyelia	03	5.5
Corpus callosal agenesis	04	7.4
Thickened filum terminale	02	3.7
Arachnoid cyst	03	5.5
Sacral agenesis	02	3.7

Surgical Procedure

The myelomeningocele sac was excised from 24 patients, the neural components were relocated, and the sac was then closed using standard methods. 6 patients had cord detethering and bony spur excision. To stop the CSF leak in nine cases, a paraspinous fascia graft or a synthetic Goretex graft was necessary. A lumbar CSF drain was placed in response to two cases of CSF leak. A ventriculoperitoneal shunt was implemented in 29 patients (53.7%) who had hydrocephalus. Twenty-three of the 29 patients presented with concomitant hydrocephalus, and a further four developed hydrocephalus in the postoperative term. Sixteen individuals underwent detethering of the chord. Tethering of the chord owing to fibrosis occurred in three patients who had had meningomyelocele surgery elsewhere. Two

patients had a fibrous septum removed, while four had a bony spur removed. In 8 (14.8%) patients, including those with lipomyelomeningocele (5), lipomyelocele (2), and filum terminale lipoma (1), involvement of neural structures required subtotal or nearly full removal of the lipoma. Six patients had dermal sinuses, two had a dermoid cyst. See Table 5.

Table 5: Surgical Procedures.

Surgery Performed	Case Count	%
Ventriculo Peritoneal Shunt	29	53.7
Sac excision and repair	31	57.4
Excision of the bony spur/ fibrous septum	06	11.1
Detethering of cord	16	29.6
Excision of Dermoid Cyst	03	5.5
Exploration of Dermoid Sinus	05	9.2
Spinal lipoma excision	08	14.8

Surgical Complications

CSF leak was the leading cause of surgical complications in nine individuals (16.7%). Four (7.40%) of the patients were determined to have pyogenic meningitis. Four patients (7.40%) experienced postoperative hydrocephalus requiring a ventriculoperitoneal shunt. Five patients (9.2%) had pseudomeningocele, eight patients (14.8%) had wound infections, two patients (3.7%) had shunt infections, and two patients (3.7%) had ventriculitis. During the recovery phase, four patients tragically passed away.

Table 6: Surgical Complications.

Complications	Number of Cases	Percentage
Pseudomeningocele	05	9.2
Meningitis	04	7.4
Hydrocephalus	04	7.4
CSF leak	09	16.7
Wound infection	08	14.8
Ventriculitis	02	3.7
Block shunt	02	3.7
Death	04	7.4

Surgical Outcome

Twenty-nine patients presented with preoperative motor weakness; of them, 12 (22.2%) showed improvement. Only three of the kids (5.5%) became worse after surgery, while 14 stayed the same. Twelve out of twenty-one patients with significant sensory impairments showed no improvement. Sphincter function was maintained at the same level in the vast majority of instances (11, 20.37%) before surgery. Trophic ulcers were found in three cases. Two of them were entirely cured, while the third showed some improvement thanks to therapy. Fifteen patients had neuroorthopedic abnormalities, and none of them got better. After therapy, the discomfort in all nine individuals' backs decreased. See Table 7.

DISCUSSION

According to our study, girls (n=32, 57.40%) are substantially more likely than boys (n=23, 42.60%) to experience diastomyetomelia. A study by

Table 7: Surgical Outcome.

Deficits pre-op	Case Count	Improved	Same	Deteriorated
Orthopedic deformities	14	-	14	-
Pain	09	09	-	-
Trophic ulceration	03	02	01	-
Sensory weakness	20	08	11	01
Motor deficit	28	09	15	04
Sphincter disturbances	18	08	09	01

Nikaoli et al,[9] included 370 patients among which there were 170 (45.9%) male individuals and 200 (54.1%) female individuals. According to a study by Sahmat et al,[10] male patients made up 59% of all patients. Elgamal et al,[11] found that female preponderance is more pronounced in type I – diastematomyelia than in type II and that females are often affected more frequently than males. In our study, females outweigh males by a ratio of 1.2:1.0, while the ratio was 2:1 in diastematomyelia type I. In a study of 155 patients conducted in Northern India by Kumar R et al., the median age at presentation was 5.7 years, and the ratio of female to male patients was 1.5:1.[12]

In our study, the lumbosacral area was affected in 51.85% of patients, making it the most prevalent place. The dorso-lumbar region affects 14 (25.92%) patients the next most frequently. In 155 cases of spina bifida, myelomeningocele was found in 72% of cases and meningocele in 2% of patients, according to a study by Kumar R et al.[13] The Hungarian study, which included 352 cases of myelomeningocele and meningocele, found that the location was cervical (1.8%). Thoracic (4.2%), lumbar (16.8%), sacral (34.5%), cervicothoracic (0.9%), and lumbosacral (22.3%). Lumbar (55.7%) was the most prevalent location in the Nigerian study, which comprised 106 cases.

In our study, 6 cases of diastematomyelia (11.1%) were seen. Elgamal et al,[11] reported that they made up 7% of all spinal dysraphism patients (11/156) recorded in the spina bifida clinic. Clinical presentation in our study showed, that out of a total of 54 instances, 32 (59.2%) presented with a noticeable bulge in the center of the back. A flat foot, high-arched foot, leg length disparity, and talipes equinovarus are among the neuroorthopedic foot abnormalities that are present in 11 (20.37%) cases. Significant scoliosis was seen in 3.5% of individuals. Major back discomfort was reported by nine individuals (16.66%). Weakness in the lower limbs was the most often seen neurological impairment, affecting 28 (53.70%) individuals. Twenty-one individuals (38.88%) were found to have sensory loss. Sphincter dysfunctions, such as urine or fecal incontinence, were found in 17 (31.48%) of the sample. trophic ulcers (4, 7.40%), and Muscular atrophy (9, 16.66%) were the additional neurological dysfunctions reported. Clinical signs of a spinal anomaly include a hair tuft, hypertrichosis, Port-wine stain, obvious swelling, hemangiomas, dimpling, a sinus tract in the back, a short back, and irregular spinal curvature, according to research by Mehta et al. The study also discovered deviations in the gluteal furrow, human or faun tails, asymmetry in the legs and feet, subcutaneous lipomas, sensory loss in the limbs, decreased motion in the limbs, dribbling urine, and recurrent urinary tract infections. The presence of two or more back cutaneous midline lesions is the greatest predictor of occult or closed spinal dysraphism.[13]

CONCLUSION

Closed spinal dysraphism is challenging to diagnose due to the non-specific nature of its cutaneous indicators and the gradual emergence of neurological and somatic symptoms. The various hazards these patients face, including infection and the development of neurologic impairments, are directly related to the delay in diagnosis and care. Even though magnetic resonance imaging (MRI) is the gold standard, computed tomography (CT) can be beneficial, particularly in diastematomyelia type 1 for showing bony spur. The soft tissue of the spinal cord and paraspinal region can be identified pretty well by MRI. Regardless of neurological symptoms, these tests should be performed on all children who exhibit cutaneous signs of occult spinal dysraphism. Spinal dysraphism patients require close communication between neonatologists, neurosurgeons, plastic surgeons, orthopedic surgeons, urologists, and rehabilitation experts.

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Additional Information

Disclosures: Authors report no conflict of interest.

Ethical Review Board Approval: The research was a retrospective study.

Human Subjects: Consent was obtained by all patients/participants in this study.

Conflicts of Interest:

In compliance with the ICMJE uniform disclosure form, all authors declare the following:

Financial Relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.

Other Relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

AUTHOR CONTRIBUTIONS

Sr. No.	Author's Full Name	Intellectual Contribution to Paper in Terms of
1.	Sagheer Ahmed	Study Design, Methodology, and Paper Writing.
2.	Iram Bokhari	Data Calculation and Data Analysis.
3.	Tanveer Ahmed	Interpretation of Results.
4.	Rabail Akbar	Statistical Analysis & Literature Review.
5.	Sehrish Altaf	Literature Review and Quality Insurer.