Original Research Article

A Retrospective Analysis of Clinical Profile and Surgical Outcome in Patients with Spinal Dysraphism at Tertiary Care Center

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International Archives of Integrated Medicine, Vol. 5, Issue 5, May, 2018.

Available online at http://iaimjournal.com/

ISSN: 2394-0026 (P) **ISSN: 2394-0034 (O)**

Received on: 08-03-2018 **Accepted on:** 20-04-2018

Source of support: Nil **Conflict of interest:** None declared.

How to cite this article: Premlal KV. A Retrospective Analysis of Clinical Profile and Surgical Outcome in Patients with Spinal Dysraphism at Tertiary Care Center. IAIM, 2018; 5(5): 63-68.

Abstract

Background: Spinal dysraphism refers to a spectrum of congenital anomalies of the spine resulting in a defective neural arch through which meninges and / or neural elements herniated leading to a variety of clinical manifestations.

Aim: This study aimed to evaluate the incidence, clinical presentations, and surgical outcome in spinal dysraphism patients.

Materials and methods: This study was a retrospective study which consisted of 32 patients conducted in Academy of Medical Sciences, Pariyaram over a period of March 2015 to March 2017. All the patients were admitted in Department of Gynecology and Pediatrics.

Results: The incidence was high in females 54.29% than males 45.71%. Spina bifida aperta (open type) was present in 44 patients (62.86%) and more common than Spina bifida occulta (closed) which was present in only in 26 patients which constituted 37.14%. The commonest site of occurrence of spinal dysraphism was the lumbo sacral region in 32 patients which constituted to 45.7%. Most common finding was myelomeningocele in 41 (58.6%) cases; myelocele was seen in 6 cases (8.6%). Lipomyelomeningocele was seen in 8 cases which was most common finding in closed type. The most common associated anomaly was hydrocephalus in 25 patients and next common was Arnold Chairi malformation type 11 in 20 cases. Motor weakness in the form of paraparesis or paraplegia present in25 patients preoperatively, out of which only 14 improved. Major cases of sensory deficits did not improved and remained static. In the majority of cases of 11 sphincter function, 6 cases remained status quo same as in the preoperative period.

Conclusion: The most common congenital cause of disability in children is spinal dysraphism encountered by paediatric neurosurgeon. Open type spina bifida is more common than closed one .At

peripheral centres, inadequatetreatmentshouldbeavoided.Spinaldysraphismpatientsshould be referred to higher tertiary centre where all the facilities are provided to the patients.

Key words

Spinal dysraphism, Surgical outcome, Tertiary care centre, Spina bifida.

Introduction

Spinal dysraphism refers to a spectrum of congenital anomalies of the spine resulting in a defective neural arch through which meninges and / or neural elements herniated leading to a variety of clinical manifestations. They are divided in to aperta (visible lesions) and occulta (with no visible external lesion). Embryologically the abnormality manifests between 3rd and 4th week of gestation during the period called neurulation in which the neural tube is formed. The neural tube then transforms into the brain and spinal cord in two stages, the primary neurulation and secondary neurulation. Primary neurulation is the process by which the brain and spinal cord up to L1 level are formed. The portion of the spinal cord distal to L1 is formed by a process known as secondary neurulation. Disorders of primary neurulation are responsible for the various forms of open dysraphism. Disorders of secondary neurulation are responsible for the various forms of closed spinal dysraphism. The spectrum includes meningocele, mylomeningocele, lipomeningmylocele, diastmatomylia, tethered cord syndrome, dermal sinus, hydrocephalus, chiari malformation and others.

Spina bifida literally means ‘spine in two parts’ or open spine. The lesion is confirmed to bony posterior arches at one or more cords. Simple spina bifida of the lower lumbar spine is a common radiological finding, especially in children and by itself carries no significance. In contrast bring spine bifida may accompany any of several complex anomalies involving the spinal cord, nerve roots, dura and even the pelvic visceral structures, constituting a major source of disability among children and adults. There are 2 distinct syndromes of spinal dysraphism the Spinabifida cystic and Spina bifida ocutta. In Spina bifida ocutta underlying neural defect is masked by the intact overlying skin. The external figure is often subtitle. Symptoms may not develop until late childhood as a result of spinal cord tethering. Included in this group are diastemetomyelia, lipomeningomyelocele, myelocystocele and caudal regression syndromes. Early recognitions of these entities is important, become neurological function may be prescribed by early and appropriate surgical intervention. This study aimed to evaluate the incidence, clinical presentations, surgical outcome in spinal dysraphism patients.

Materials and methods

This study was a retrospective study which consisted of 32 patients conducted in Academy of Medical Sciences, Pariyaram over a period of March 2015 to March 2017. All the patients were admitted in Department of Gynecology and Pediatrics. All patients were evaluated for their complete history, presenting symptoms and examination of neurological. All patients were made to undergo cranio-spinal MRI, radiological findings and associated anomalies were recorded. All the patients underwent surgical procedures like repair and excision of the sac, cord detethering and ventriculoperitoneal shunt. These patients were followed in post-operative period and the outcomes were recorded. In the analysis, the patients who were followed up for 4 months minimum were included.

Results

A total of 32 patients were selected in this study. The age ranged from 1 day to 18 years.

Table - 1 shows that the incidence is high in females (number of cases=17), 53.13% when compared to males (number of cases=15), 46.87%. One day was the age of the youngest

patient and the oldest patient was 18 years. Most of the patients were below one month (12patients, 37.5%), 7 patients were from 1 month to 1 year,

Table - 1: Demographic distribution in this study.

Sex	**No of cases**	**%**
Males	15	46.87%
Females	17	53.13%
Age group	**No of cases**	**%**
1 day to 1 month	12	37.5%
1 month to 1 year	7	21.9%
1 year to 6 years	5	15.6%
6 years to 10 years	3	9.4%
10 years to 16 years	3	9.4%
>16 years	2	6.3%

Table - 2 shows that out of 32 cases, 19 had spina bifida aperta (open type) which constituted to 59.37% and spina bifida occulta was observed in 13 cases (closed type) which constituted to 40.63%.

Table - 2: Type of dysraphism.

Type of dysraphism	**No of cases**	**%**
Open	19	59.37%
Closed	13	40.63%

Table - 3 shows that in this study, the occurrence of spinal dysraphism was all over the spine. The most common site was the lumbo-sacral region in 11 patients (34.37%), then dorso-lumbar region in 8 patients (25%) patients. 76patients present with spina bifida in the upper dorsal region and 5 patients in cervical region and 2 patients in sacral region.

Table - 3: Site wise prevalence of the lesion.

Site	**No of cases**	**%**
Cervical	5	6.4%
Dorsal	6	18.75%
Dorso-lumbar	8	25%
Lumbo-sacral	11	34.37%
Sacral	2	6.25%

Table - 4: Clinical features in the study.

Clinical features	**No of cases**	**%**
Cutaneous		
Mass skin covered	18	56.2%
Ruptured	9	28.12%
Hypertrichisis	1	3.13%
Dermal sinus	2	6.25%
Dimple over skin	1	3.13%
Sub cutaneous lipoma	4	12.5%
Scar of previous surgery	5	15.62%

Table - 5: MRI findings.

Anomaly	**No of cases**	**%**
Myelomeningocele	15	46.87%
Myelocele	3	9.37%
Lipomyelomeningocele	4	12.5%
Meningocele	3	9.37%
Lipomyelocele	2	6.25%
Diastematomyelia	2	6.25%
Dermal sinus	2	6.25%
Spinal lipoma	1	3.12%

Table - 6: Associated anomalies.

Anomaly	**No of cases**	**%**
Hydrocephalus	10	31.25%
Arnold Chairi malfunction type 11	8	25%
Low tethered cord	5	15.62%
Syringomyelia	2	6.25%
Thickened filum terminate	2	6.25%
Syringohydromyelia	2	6.25%
Corpus callosal agenesis	1	3.12%
Sacral agenesis	1	3.12%
Arachnoid cyst	1	3.12%

Table - 4 shows that the swelling on the middle of the back was the most common manifestation present in 18 patients (56.2%) out of total of 70 cases, 1 patients had hypertrichisis which constituted to 3.13%, 2 patients had dermal sinus which constituted to 6.25%, 1 patients had dimple over skin which constituted to 3.13%, 4 patients had sub-cutaneous lipoma which

constituted to 12.5% and 5 patients had scar of previous surgery which constituted to 15.62%. The orthopaedic clinical presentation was backache which was seen in 5 patients which constituted to 15.62%, foot deformities were seen in 9 patients which constituted to 28.12%, scoliosis was seen in 2 patients which constituted to 6.25%. The most common neurological clinical presentation was motor weakness which was observed in 14 patients (43.75%), sensory deficits in 9 patients (28.12%), sphincter dysfunction in 5 patients (15.62%), muscular atrophy was seen in 2 (6.25%), 3 patients had trophic ulcerations.

Table - 5 shows that for detection of congenital anomalies associated with spinal dysraphism, craniospinal MRI was done in all patients. Most common finding was myelomeningocele which was seen in 15 patients, (46.87%); myelocele was seen in 3 patients (9.37%), lipomyelomeningocele was seen in 4 patients (12..5%), meningocele was seen in 3 patients (9.37%), lipomyelocele, diastematomyelia, dermal sinus was seen 2 in each of anomaly respectively, spinal lipoma was seen in 1 patients.

Table - 6 shows that hydrocephalus was seen in most of the patients i.e. 10 patients which constituted to 31.25%, Arnold Chairi malfunction type 11 was seen in 8 patients which constituted to 25%, low tethered cord was seen in 5 patients which constituted to 15.62%, and sacral agenesis and arachnoid cyst was seen in 1 patient each which constituted to 3.12% each.

Table - 7 shows that motor weakness was present postoperatively in 14 cases, out of which 8 patients improved, 1 patients deteriorated in post-operative period and 5 patients showed status quo. There was no improvement in major sensory deficits (7 out of 9) and remained static. Majority of the case remained status quo (3 out of 5) in the preoperative period in sphincteric dysfunction. 3 patients were presented with trophic ulcer and 9 patients with orthopedic deformities and all the patients did not show improvement in this anomaly postoperatively.

Table - 7: Surgical outcome in the study.

Preoperative deficits	No. of cases	Improved	Status quo	Deterioration
Pain	5	5	-	-
Motor weakness	14	8	5	1
Sensory loss	9	3	7	-
Sphincteric dysfuntion	5	2	3	-
Trophic ulcer	3	2	1	-
Orthopaedic deformities	9	-	9	-

Discussion

In a study done by Hyagriva Rao, et al. [6] the age ranges from one day to 17 years. The youngest was one day and oldest one 17 years in this study. The incidence is high in females 57.40% (n=31) than males 42.60% (n=23). Spina bifida aperta (open type) is present in 39 (72.22%) and more common than Spina bifida occulta (closed) which is present in only in 15 patients (27.77%). The commonest site of occurrence of spinal dysraphism is the lumbo sacral region in 28 patients (51.85%). Most common finding is myelomeningocele in 33 (61.11%) cases, myelocele 4 (7.47%). Lipomyelomeningocele 7 (12.96%) is most common findingin closed type. The most common associated anomaly is hydrocephalus in 23 (42.59%) patients and next common is Arnold Chairi malformation type 11 in 21 (38.88%) cases. The most common post-operative complication is CSF leak in 11 (20.37%) cases. Motor weakness in the form of paraparesis or paraplegia present in 29 patients preoperatively, out of which only 12 (22.22%) improved. Major

cases of sensory deficits (12 in 21 cases) did not improved and remained static. In the majority of cases (11, 20.37%) sphincter function remained statusquo same as in the preoperative period. O'Neill OR, et al. [7] reviewed 27 patients with congenital anomalies of the sacral spine. There were 16 males and 11 females with a mean follow-up of 81.1 months (range 8-211 months). Fifteen patients had sacral agenesis and 12 had sacral dysgenesis. Fifteen patients had neuroimaging of the spine. Seven patients had conus termination below the L2 vertebral body. Four patients had associated thoracic syringomyelia and 6 patients were identified with caudal or dorsal lipoma. There were only two episodes of neurological deterioration, both in a single patient who had a lipomyelomeningocele, in 182 patient-years of follow-up. Four patients with low lying conus had pre-emptive spinal cord exploration for release of tethering in order to prevent neurological deterioration. Patients with agenesis or dysgenesis of the sacrum should undergo magnetic resonance (MR) imaging of the spine in order to detect spinal cord lesions associated with progressive neurological deterioration. Findings on MR imaging are more likely to correlate with clinical course than findings on skeletal radiography. José Gilberto de Brito Henriques, et al. [8] reported that ultrasonography for diagnosis of spinal cord diseases has been used since the eighties. There are different fields for its use: traumatic spine lesions, intra-operative use in trauma and spine tumors surgery, pre-operative and follow up study for spinal dysraphism, occult spinal dysraphism of the newborn. Patients with suspected occult spinal dysraphism are the most important indication for the method. Utrasonography was performed in 292 patients of Hospital das Clínicas, Universidade Federal de Minas Gerais. Lesions such as conus medullaris lipomas, low level of conus medullaris, filum terminale tethering, cauda equina cysts and dermal sinuses can be well demonstrated. There is no difficulty in interpreting the image. The exam does not need sedation; it is safe, fast and cheap. Ideally it should be performed by the neurosurgeon as it may provide many information during the examination. The ultrasonography does not replace magnetic resonance imaging but it is an excellent method for screening. Guggisberg D, et al. [9] conducted a study to verify the diagnostic value of lumbosacral midline cutaneous lesions in asymptomatic children to detect occult spinal dysraphism (OSD) and to propose a practical approach for clinical investigations with respect to the type of cutaneous lesions observed. In this, a retrospective study was conducted in 54 children referred to the Department of Pediatric Dermatology between 1990 and 1999 for congenital midline lumbosacral cutaneous lesions. It was observed that occult spinal dysraphism was detected in 3 of 36 patients with an isolated congenital midline lesion and 11 of 18 patients with a combination of 2 or more different skin lesions. Oran S, et al. [10] conducted a study in which all fetuses underwent prenatal US. Sixty-one fetuses underwent prenatal MR imaging. Fifty fetuses underwent both postnatal spinal radiography and postnatal MR imaging, and an additional 34 fetuses underwent one postnatal study but not the other. When findings on prenatal US images were compared with those on postnatal radiographs, the findings agreed within one spinal level in 79% (55 of 70, $\kappa = 0.60$) of cases. When findings on prenatal MR images were compared with those on postnatal radiographs, the findings agreed in 82% (31 of 38, $\kappa = 0.63$) of cases. Findings on postnatal MR images and those on postnatal spinal radiographs agreed within one spinal level in 100% (50 of 50, $\kappa = 1.0$) of cases.

Conclusion

The most common congenital cause of disability in children is spinal dysraphism encountered by pediatric neurosurgeon. Open type spina bifida is more common than closed one. At peripheral centres, inadequate treatment should be avoided. Spinal dysraphism patients should be referred to higher tertiary centre where all the facilities are provided to the patients. To avoid complications, the post-operative care is equally important and for better outcome. Pain is

common symptom to improve. Post operatively, motor deficits improve better than sensory deficits and bladder dysfunctions.

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