STUDY ON SPINAL DYSRAPHISM IN TERTIARY CARE CENTRE, ANDHRA MEDICAL COLLEGE, VISAKHAPATNAM

B. Hayagriva Rao[1], K. S. Vara Prasad[2], K. Indu Sekhar[3], B. Raja Sekhar[4]

[1]Associate Professor, Department of Neurosurgery, Andhra Medical College, Visakhapatnam.
[2]Professor & HOD, Department of Neurosurgery, Andhra Medical College, Visakhapatnam.
[3]Resident, Department of Neurosurgery, Andhra Medical College, Visakhapatnam.
[4]Resident, Department of Neurosurgery, Andhra Medical College, Visakhapatnam.

ABSTRACT

INTRODUCTION

Spinal dysraphism is a broad term including heterogeneous group of congenital spinal anomalies. Defects in the early embryologic age cause spinal dysraphism. Several environmental and genetic factors are noted for causing this congenital entity. Spinal dysraphism is broadly grouped in to two groups, closed (spina bifida occulta) and open type (spina bifida aperta). Transabdominal ultrasonography is best for prenatal diagnosis. The clinical features of spinal dysraphism include more than one symptom or sign. The neurological deficits depend on malformation of the neural placode and level of the defect. This retrospective study of 54 spinal dysraphism cases was done in king George Hospital, Visakhapatnam. The study period is from August 2013 to July 2015. The craniospinal MRI was done to all patients and radiological findings and associated anomalies recorded for planning of management of these patients.

RESULTS

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 finding in closed type. The most common associated anomaly is hydrocephalus in 23(42.59%) patients and next common is Arnold chairi malformation type11 in 21(38.88%) cases. The most common postoperative 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 status quo same as in the preoperative period.

CONCLUSION

A spinal dysraphism patient should receive treatment from an experienced multidisciplinary team including neonatologist, neurosurgeon, and plastic surgeon and rehabilitation experts. Inadequate treatment at peripheral centers should be avoided. Patient should be referred to higher tertiary centre where the complete team is available. The post-operative care is equally important to avoid complications and for better outcome.

KEYWORDS

Spinal dysraphism, Myelomeningocele, Lipomyelomeningocele, Hydrocephalus.

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INTRODUCTION: Spinal dysraphism is a broad term including heterogeneous group of congenital spinal anomalies. These anomalies result from defect in spinal cord development which occurs in three stages, gastrulation (2-3 weeks), primary neurulation (3-4 weeks) and secondary neurulation (5-6 weeks).[1] Defects in the early embryologic age cause spinal dysraphism.[2]

There is herniation of neural elements, or meninges through the defective neural arch. The incidence of this congenital anatomy is 1-4 per 1,000 live births.[3,4] Incidence of spinal dysraphism is decreasing in USA and worldwide in the past 2 decades.[5] The cause of this may be due to good nutrition of women, folate intake, and better antenatal care. Data pertaining to India is uncertain. Several environmental and genetic factors are noted for causing this congenital entity.[6] Folic acid supplementation as a preventive measure is suggestible in high risk group.[7,8] Prenatal diagnosis may be an available option to mothers for termination of pregnancy.

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Corresponding Author:
Dr. B. Hayagriva Rao, Associate Professor,
Department of Neurosurgery,
Andhra Medical College, Visakhapatnam.
E-mail: bhrao64@gmail.com
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Spinal dysraphism is broadly grouped in to two groups, closed (spina bifida occulta) and open type (spina bifida aperta).[9] In open type the overlying skin is not intact, and leakage of cerebrospinal fluid may or may not be present. In occult type the defect is well covered with skin. Depending on radiological or pathological findings spina bifida aperta include myelomeningocele, meningoceles, myeloschisis and occult spinal dysraphisms include thickened filum terminale, dermal sinus, lumbosacral lipomas, lipomyelomeningocele, split cord malformation, neuroenteric cyst. The anatomical features common to the entire group are an anomaly in the midline structures of the back, absence of some of the neural arches, and defects of the skin, filum terminale, nerves and spinal cord.

Transabdominal ultrasonography is best for prenatal diagnosis. Women may undergo triple screening for detecting spina bifida, Down syndrome, and other congenital diseases in utero. Magnetic resonance imaging (MRI) is the gold standard investigation of choice in evaluating and planning for treatment of spinal dysraphism. The associated cranial lesions are well delineated by MRI. Most common lesion is meningomyelocele. Nearly all patients having myelomeningocele associate with Arnold Chiari malformation type 11 whereas 80-90% cases accompany with hydrocephalus. Syringomyelia or hydrosyringomyelia present in 40-80% of patients. The commonest site of spinal dysraphism is lumbosacral region. The clinical features of spinal dysraphism include more than one symptom or sign. The neurological deficits depend on malformation of the neural placode and level of the defect. The higher the level of defect, the worse is the prognosis. Tethered cord syndrome is low conus medullaris, thickened filum terminale, and may be associated with intra dural lipoma. The common presentation of occult type is cutaneous manifestations over the low back. Various orthopaedic manifestations like foot deformities (most common), limb-length discrepancies, vertebral abnormalities and scoliosis are present. Depending on the site and type of tethering, neurologic deficits may also involve the bladder and bowel function. Closed types of spinal dysraphism often have more than one lesion and may present with more than one symptom.[10] Timing of surgical management depends on the clinical condition of the patient and can be undertaken as soon as possible.[11]

Surgical management is aimed at freeing the neural placode and repositioning into the spinal canal. The reconstruction of the dura and coverings is done to prevent CSF leak and subsequent infections. The outcome is good if surgery done before the damage to neural tissue and using surgical microscopes.

METHODOLOGY: This retrospective study of 54 spinal dysraphism cases was done in king George Hospital, Visakhapatnam. The study period was from August 2013 to July 2015. Most of the cases were referred from gynaecology and paediatric departments. The age of patients varied from one day to seventeen years. Our aim is to study the incidence, clinical presentations, and surgical outcome in spinal dysraphism patients. Clinically patients are assessed on the basis of history, presenting symptoms and neurological examination.

The craniospinal MRI was done in all patients and radiological findings and associated anomalies are recorded for planning of management of these patients. These patients underwent appropriate surgical procedures like excision and repair of the sac, detethering of the cord and ventriculoperitoneal shunt. All these patients are followed in the postoperative period and outcome noted and assessed. The patients with follow-up of minimum three months period are included in this analysis.

RESULTS: A total of 54 patients included in this study. The age ranged from1day to 17 years. The incidence is high in females 57.40% (n=31) than males 42.60% (n=23). (Table1).

Sex	No. Cases	Percentage
Male	23	42.60%
Female	31	57.40%
Table 1: Sex Distribution		

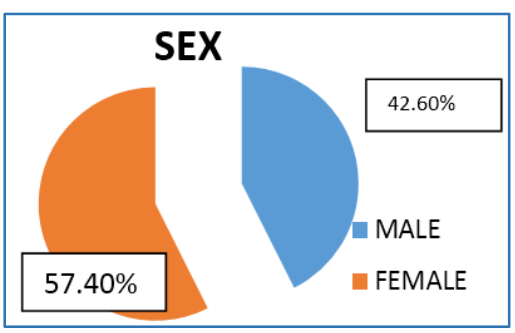

Fig. 1

In 54 cases, Spina bifida aperta (open type) was present in 39(72.22%) and more common than Spina bifida occulta (15 patients, 27.77%). (Table 2).

Type	No. Cases	Percentage
Open	39	72.22%
Closed	15	27.77%
Table 2: Type of Dysraphism		

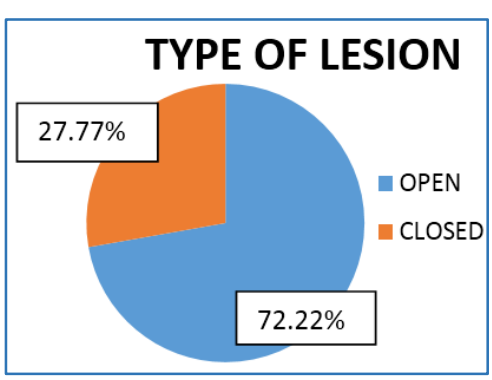

Fig. 2

In our study the age of youngest patient was one day, and the oldest was 17 years. Most of them were below one

month age, (29 patients, 53.70%), and 13 patients were in age group from one month to one year. The average age of the study group was 2.8 years. (Table 3)

Age group	No. of Cases	Percentage
1 day-1 month	29	53.70%
1 month-1 year	13	24.07%
1 year–5 years	5	9.25%
5 years–10 years	3	5.55%
10 years-15 years	3	5.55%
>15 years	1	1.85%

Table 3: Age Distribution

The occurrence of spinal dysraphism encompassed all over the spine in this study. The commonest site was the lumbo sacral region, in 28 patients (51.85%). Next to this is dorso lumbar region in 14(25.92%) patients. Six children (11.11%) present with spina bifida in the upper dorsal region and 2(3.70%) in the sacral region. Four patients (7.40%) had this anomaly in the cervical region. (Table 4)

Site	No. of Cases	Percentage
Cervical	4	7.40%
Dorsal	6	11.11%
Dorso lumbar	14	25.92%
Lumbo sacral	28	51.85%
Sacral	2	3.70%

Table 4: Site wise prevalence of the lesion

The swelling over the middle of the back was most common manifestation present in 32(59.2%) cases out of total 54 cases. 8 patients (14.81%) had ruptured myelomeningocele/myelocele sac. The skin markers which occur in spina bifida occult type like dermal sinus, Hypertrichosis, dimple of skin present in 3(5.55%), 2(3.70%), 1(1.85%) respectively. Previous operative scar was present in 5(9.25%) patients. The neuro orthopaedic foot deformities like talipes equinovarus, high-arched foot, leg length discrepancy and flat foot are present in 11(20.37%) cases. Three patients (5.55%) had significant scoliosis. Nine (16.66%) patients presented with significant back pain. Most common neurological deficit was weakness of the lower limbs present in 28(53.70%) patients. Sensory loss noted in 21(38.88%) patients. The sphincter dysfunctions like urinary or faecal incontinence were present in 17(31.48%). The other neurological dysfunctions noted were muscular atrophy (9, 16.66%), gait disturbances (7, 12.96%), and trophic ulcers in 4(7.40%) cases. (Table 5)

Clinical Features	No. of Cases	%
Cutaneous		
Mass skin covered	24	44.45%
Ruptured	8	14.81%
Tuft of hair (hypertrichisis)	2	3.70%
Dermal Sinus	3	5.55%
Dimple over skin	1	1.85%
Sub cutaneous Lipoma	4	7.40%
Scar of previous surgery	5	9.25%
Orthopedic		
Backache	9	16.66%
Foot deformities	11	20.37%
Scoliosis	3	5.55%
Neurologic		
Motor weakness	29	53.70%
Sensory deficits	21	38.88%
Sphincter dysfunction	17	31.48%
Muscular atrophy	9	16.66%
Gait disturbances	7	12.96%
Trophic Ulcerations	3	5.55%

Table 5: Clinical Presentation

Craniospinal MRI was done in all patients for evaluation of congenital anomalies associated with spinal dysraphism. All were having one or multiple anomalies. Spina bifida aperta (37) was more common than bifida occulta which is noted in17 patients. Most common finding is myelomeningocele in 33(61.11%) cases, myelocele 4(7.47%). In closed type, lipomyelomeningocele 7(12.96%) was most common finding, and meningocele 3(5.55%), lipomyelocele 2(3.70%), diastematomyelia in 2((3.70%), dermal sinus 2(3.70%), spinal lipoma in one (1.85%) cases. The most common associated anomaly was hydrocephalus in 23(42.59%) patients and next common was Arnold chairi malformation type11 in 21(38.88%) cases. The low lying tethered cord (18, 33.33%), and syringomyelia (11, 20.37%) were present in this study. Thickened filum terminale 4(7.4%), syringohydromyelia 3(5.55%), corpus callosal agenesis 2(3.70%) sacral agenesis in 2(3.70%), arachnoid cyst 1(1.85%), corpus callosal thinning 1(1.85%) were less frequently noted. (Table 6)

Anomaly	No. of Cases	%
Myelomeningocele	33	61.11%
Myelocele	4	7.47%
Lipomyelomeningocele	7	12.96%
Meningocele	3	5.55%
Lipomyelocele	2	3.70%
Diastematomyelia	2	3.70%
Dermal sinus	2	3.70%
Spinal lipoma	1	1.85%
Associated anomalies		
Hydrocephalus	23	42.59%
Arnold chairi malformation type 11	21	38.88%
Low tethered cord	18	33.33%
Syringomyelia	11	20.37%
Thickened filum terminale	4	7.40%
Syringohydromyelia	3	5.55%
Corpus callosal agenesis	2	3.70%
Sacral agenesis	2	3.70%
Arachnoid cyst	1	1.85%
Corpus callosal thinning	1	1.85%

Table 6: MRI findings

The myelomeningocele sac excision, repair and repositioning of neural elements with primary closure done in 24 cases. The remaining 9 cases needed paraspinous fascia, fascia lata or synthetic Gore-Tex graft to repair the dural

deficiency to prevent the CSF leak. Lumbar CSF drain used in 2 cases of CSF leak cases. Ventriculo peritoneal shunt for hydrocephalus was done in 27(50%) cases. Out of these 27 patients, 23 cases had hydrocephalus as an associated feature while 4 cases develop hydrocephalus during post-operative period. Detethering of cord was done in 19 patients. Three were post-operative meningomyelocele repair cases done elsewhere developing tethering of cord due to fibrosis. Complete excision of the bony spur was done in one patient and fibrous septum in one case. The subtotal or near total excision of lipoma was done due to involvement of neural structures in 10(18.5%) patients which include, lipomyelomeningocele,(7) lipomyelocele(2) and filum terminale lipoma in one patient. Dermoid cyst was present in one, and dermal sinus in 2 patients.

Findings/Procedures	No. of cases	%
Excision of sac & repair	33	61.11%
Ventriculo Peritoneal Shunt	27	50.00%
Detethering of cord	19	35.18%
Spinal lipoma excision	10	18.51%
Re-exploration	4	7.40%
Excision of the bony spur/fibrous septum	2	3.70%
Dermal sinus	2	3.70%
Dermoid Cyst	1	1.85%

Table 7: Surgical procedures/findings

The most common postoperative complication was CSF leak in 11(20.37%) patients. Eight of these patients responded to conservative management with acetazolamide therapy and prone position. Three patients could not be treated conservatively. These patients managed by lumbar drain, re-exploration and duraplasty. Pyogenic meningitis present in 4(7.40%) cases. Three patients responded to antibiotic management. One patient of severe meningitis not responded to antibiotic therapy and succumbed to death. During post-operative period, hydrocephalus developed in 4(7.40%) cases requiring ventriculo peritoneal shunt. Other complications like pseudomeningocele in 5(9.25%), wound infection in 7(12.96%), shunt infection in 3(5.55%), ventriculitis in 1(1.85%) patient were present. Two patients died in the postoperative period. (Table 8)

Complications	No. of cases	Percentage
CSF leak	11	20.37%
Surgical site infection	7	12.96%
Pseudomeningocoele	5	9.25%
Meningitis	4	7.40%
Hydrocephalus	4	7.40%
Shunt infection	3	5.55%
Ventriculitis	1	1.85%
Death	2	3.70%

Table 8: Post-operative complications

Motor weakness in the form of paraparesis or paraplegia was present preoperatively in 29 cases, out of which 12 (22.22%) patients improved. Three children (5.55%) deteriorated in postoperative period and 14 patients showed status quo. Major sensory deficits (12 in 21 cases) have not improved and remained static. In the majority of cases (11, 20.37%) sphincter function remained status quo as in the preoperative period. Three patients presented with trophic ulcers. In these, two had completely healed while one patient showed partial response to treatment. The neuro orthopaedic deformities were present in 14 cases, and did not show any improvement. All the 9 patients with back pain improved after treatment. (Table 9)

Preoperative deficits	No. of cases	Improved	Status quo	Deterioration
Pain	9	9	-	-
Motor weakness	29	12	14	3
Sensory loss	21	9	12	-
Sphincteric dysfunction	17	6	11	-
Trophic ulcer	3	2	1	-
Orthopaedic deformities	14	-	14	-

Table 9: Surgical Outcome

DISCUSSION: Spinal dysraphism include open (spina bifida aperta) and closed (spina bifida occulta) types. Neural tube defects develop during embryogenesis along the neuroaxis from the brain to the sacrum.[12] Failure of closure of caudal neuropore causes open defects while the secondary neurulation defects result in occult dysraphism. There are many risk factors for neural tube defects which include folic acid deficiency,[13] advanced mother's age,[12] lower socioeconomic status, and antiepileptic drugs. The incidence of spinal dysraphism is decreasing all over the world, mostly in developed countries. Folic acid fortification and prenatal diagnosis may contribute to this decline. In developing countries, the mothers who are illiterate and belong to low socioeconomic group do not have regular antenatal check-ups and screening procedures. There are considerable percentages of mothers who do not take folic acid supplementation during pregnancy. Supplementation of folic acid is useful to reduce the incidence of NTDs which include spinal dysraphism.[14] Most of spinal dysraphism cases are diagnosed in infancy. The symptoms may occur at later age in closed type. Swelling with or without skin covering is present in most patients. Variable neurological deficits and associated hydrocephalus may be present.[15]

The clinical features depend on the type and site of lesion and associated anomalies. The neurological features are motor, sensory deficits and sphincter dysfunctions. Spina bifida occulta present with cutaneous markers and neurological deficits. The pain may be presenting feature in tethered cord syndrome or diastematomyelia.[16] Neuro orthopaedic deformities like talipes equinus, asymmetry of the limbs and gait disturbances associated with spinal dysraphism. The high risk group of dysraphism can be diagnosed antenatally by screening of maternal serum alpha -fetoprotein. Alpha-fetoprotein crosses the placenta and can be detected maximum during 12 to 14 weeks of gestation. The increase in amniotic fluid concentration of alpha-1-fetoprotein is present with spina bifida, anencephaly and hydrocephalus.[17] The bladder function is evaluated by urodynamic studies. Spinal dysraphism diagnosed by

different imaging modalities like plain X-ray, ultrasound, CT scan, and MRI. Prenatal diagnosis of neural tube defects can be done with ultrasound. Oran S. Aaronson, MD et al[18] concluded that findings at prenatal MR imaging and prenatal US are equally valuable for the evaluating myelomeningocele in a foetus. Advantages of CT scan are excellent visualization of the osseous anatomy. MRI is the procedure of choice, to determine the spinal cord, dural sac, location of filum terminale and other associated soft tissue abnormalities. The management and the outcome of spinal dysraphism depend on various factors. Preoperative counselling of the parents is done regarding the planning of surgery, outcome, and long term care.[19]

The best age for surgical intervention is around 6 months when blood loss and anaesthesia are tolerated better compared to early age. But emergency intervention may be needed in ruptured myelomeningocele sac, or neurologically deteriorating cases. Jamil et al. demonstrated that progressive neurological deficits are only indication for surgery.[20] Prophylactic surgery can be done as the neurological deficits are not reversible and the patients with no neurological deficits benefit by not deteriorating.[21] The aim of surgical management is excision of sac, repair of the defect and preserving functional neural tissue. Complete removal of tethering lesions and prevention of retethering of the cord should be done to prevent neurological deterioration. Micro neurosurgery techniques and intra operative electrophysiological tests are useful to improve out come. Pain is the most common symptom to improve followed by motor deficits. Sensory deficits and bladder dysfunctions remain status quo post operatively. The associated hydrocephalus is an important factor for prognosis. Ventriculoperitoneal shunt insertion before the repair of lesion can avoid CSF leak and pseudo-meningocele formation.

CONCLUSION: Spinal dysraphism is common congenital cause of disability in children encountered by paediatric neurosurgeon. Open type spina bifida is more common than closed one. A spinal dysraphism patient should receive treatment from an experienced multidisciplinary team including neonatologist, neurosurgeon, plastic surgeon and rehabilitation experts. Inadequate treatment at peripheral centres should be avoided. Spinal dysraphism patients should be referred to higher tertiary centre where the complete team is available. The post-operative care is equally important to avoid complications and for better outcome. Pain is common symptom to improve. Motor deficits improve better than sensory deficits and bladder dysfunctions post operatively.

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