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ORIGINAL RESEARCH

A Study on the Incidence, Clinical Presentations, and Surgical Outcome in Spinal Dysraphism Patients

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ABSTRACT

Background: Spinal dysraphism is common congenital cause of disability in children encountered by pediatric neurosurgeon and is a significant public health problem. Open type spina bifida is more common than closed one. Though most of these cases are diagnosed at birth or at an early infancy, some may be discovered in older children or in adults. Aim & Objective: 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.

Results: A total of 54 patients included in this study. The age ranged from 1day to 17 years. The incidence is high in females 57.40 % (n=31) than males 42.60 % (n=23). 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 myelomenigocele/ 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. 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%) case.Most of the patients associated with hydrocephalus require shunt surgery prior to the definitive surgery.The post operative care is equally important to avoid complications and for better outcome. Electro myographic studies and urodynamic studies are required to assess the sphincteric outcome following surgery. Pain is common symptom to improve. Motor deficits improve better than sensory deficits and bladder dysfunctions post operatively.Outcome of these patients are always at risk of retethering and delayed neurological deterioration.

Conclusion: Therefore, close monitoring by a multidisciplinary team is important. Inadequate treatment at peripheral centres should be avoided. Spinal dysraphism patients should be referred to higher tertiary centre where the complete team is available.

Keywords: Spinal dysraphism, Motor deficits,Prevalence, Neurological Deterioration.

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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, filum terminale, 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 occulta 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 interspace 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 terminale or fibrousband.[5]

Approximately 95% of couples that have a fetus affected with ONTD have a negative family history. Most ONTDs are caused by multi factorial inheritance, including genetic and environmental factors.[6]

The estimated incidence of spinal dysraphism is about 1−3/1,000 live births. The prevalence of spinal dysraphism has been on the decline world over in the last few decades due to better nutrition for women, folic acid supplementation, improved antenatal care and high-resolution ultrasound for prenatal screening and biochemical markers.

Aim & Objectives

- To study the incidence and clinicalfeatures of spinal dysraphism
- To study radiological features ofdifferent types ofspinal dysraphism
- To study surgical management and outcome.

MATERIALS & METHODS

This prospective study of 54 spinal dysraphism cases was done in king George Hospital,Visakhapatnam. The study period was from August 2019 to November 2021. Most of the cases were referred from gynaecology and pediatric 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 from 1day to 17 years. The incidence is high in females 57.40 % (n=31) than males 42.60 % (n=23).

Table 1: Sex Distribution

Sex	No.Cases	Percentage
Male	23	42.60%
Female	31	57.40%

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 of Dysraphism

Type	No.Cases	Percentage
Open	39	72.22%
Closed	15	27.77%

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.

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 3: Site of the Lesion

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%

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 myelomenigocele/ 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 orthopedic 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 fecal 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 4: Clinical Presentations

Clinical presentations	No. Of cases	Percentage
Cutaneous:		
Massskin 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%

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. Thickend filumterminale 4(7.4%), syringo hydromyelia 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 5: MRI Findings

Congenital Anomaly:	No. of Cases	Percentage
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 type11	21	38.88%
Low tethered cord	18	33.33%

Syringomyelia	11	20.37%
Thickend filumterminale	4	7.40%
Syringo hydromyelia	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%

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 Goretex 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.

Table 6: Surgical Procedures

Procedures	**No. Of cases**	**Percentage**
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%
Exploration and excision of Dermal sinus	2	3.70%
Excision of Dermoid Cyst	1	1.85%

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 7: Post Operative Complications

Complications	**No. of cases**	**Percentage**
CSF leak	11	20.37%
Surgical site infection	7	12.96%
Pseudomeningocele	5	9.25%
Meningitis	4	7.40%

Hydrocephalus	4	7.40%
Shunt infection	3	5.55%
Ventriculitis	1	1.85%
Death	2	3.70%

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 8: Surgical Outcome

Preoprative 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	-

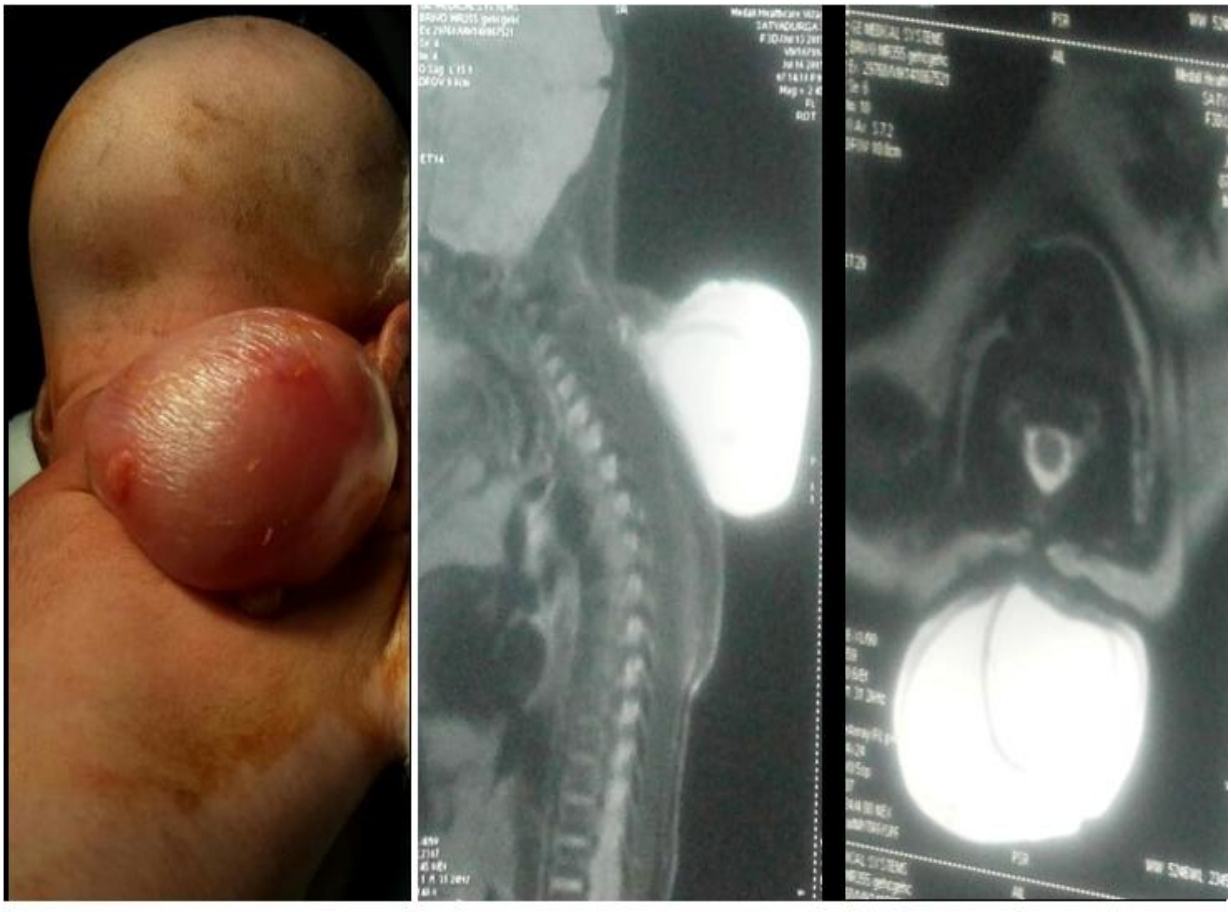

Figure 1: Cervical Meningocele

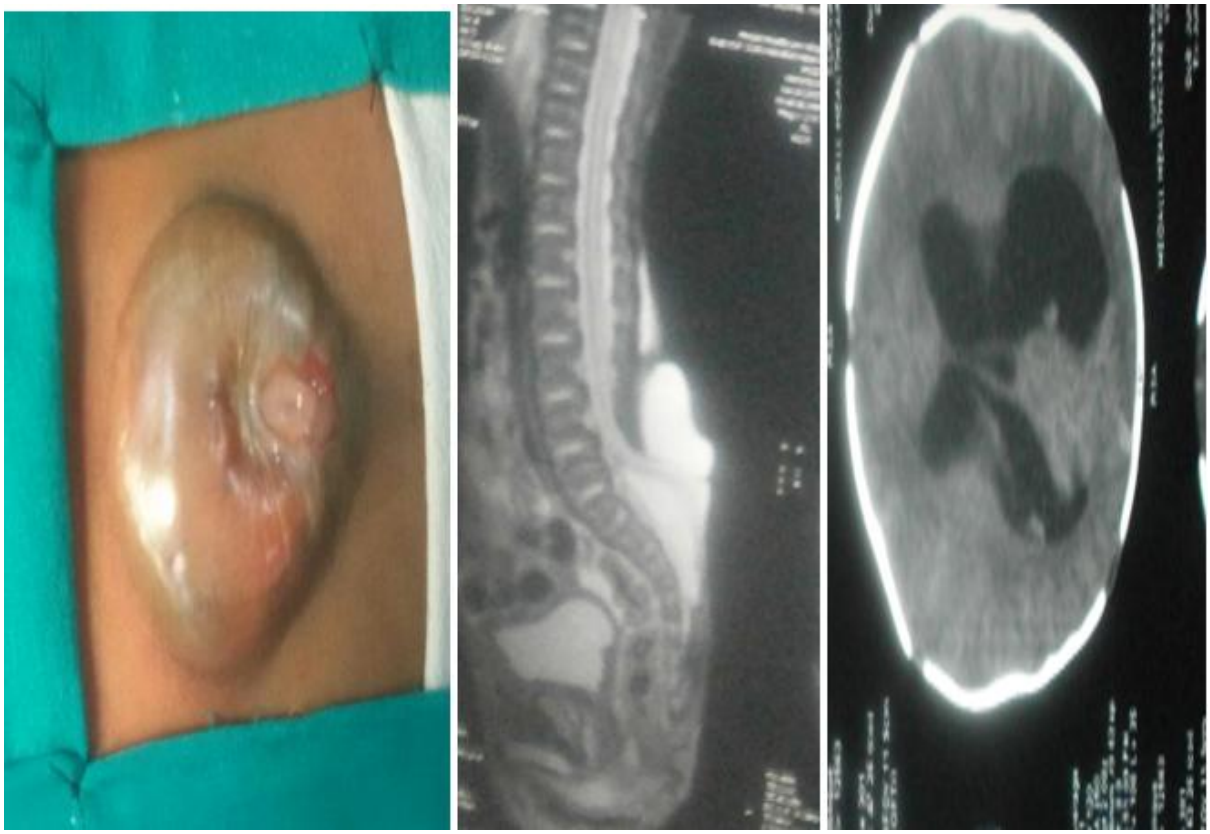

Figure 2: Lumbosacral Myelomeningocele with Hydrocephalus

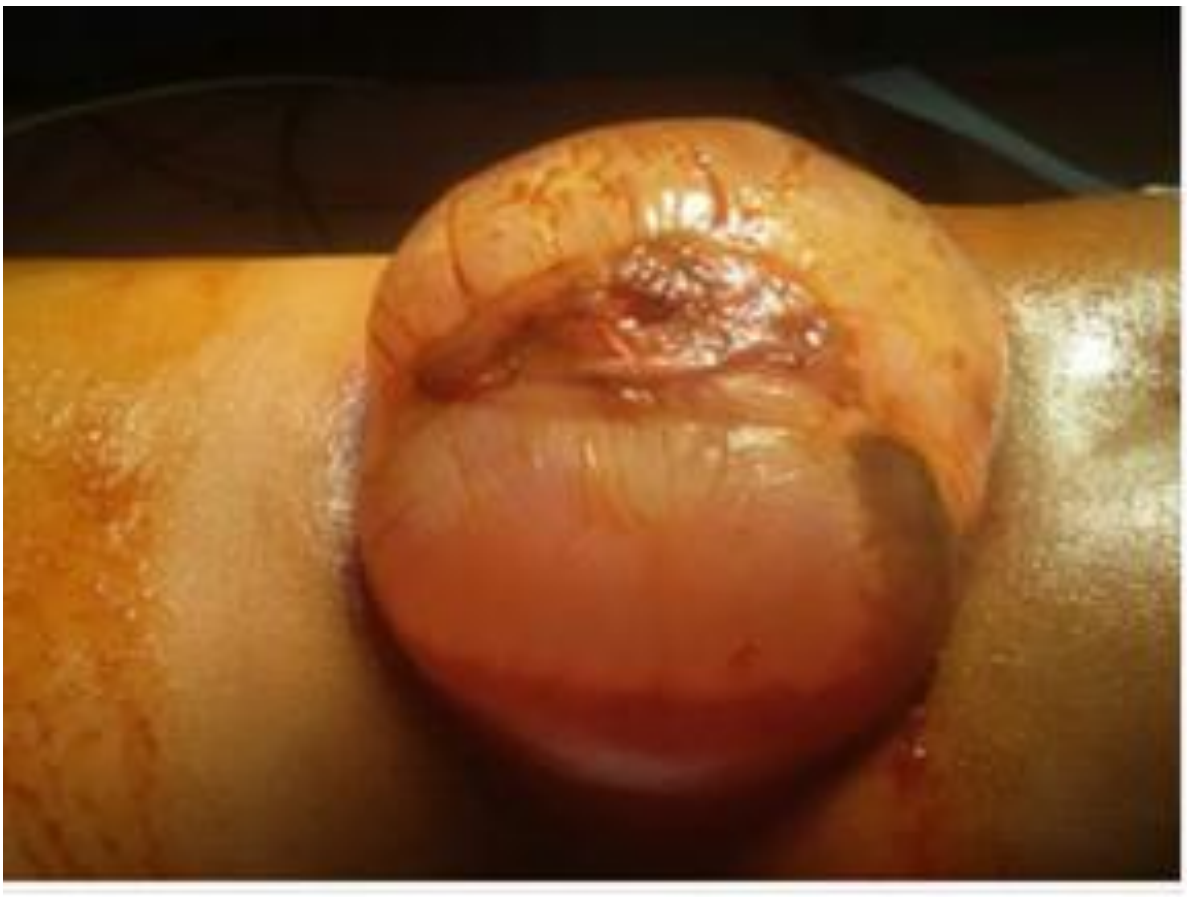

Figure 3: Lumbosacral Myelomeningocele

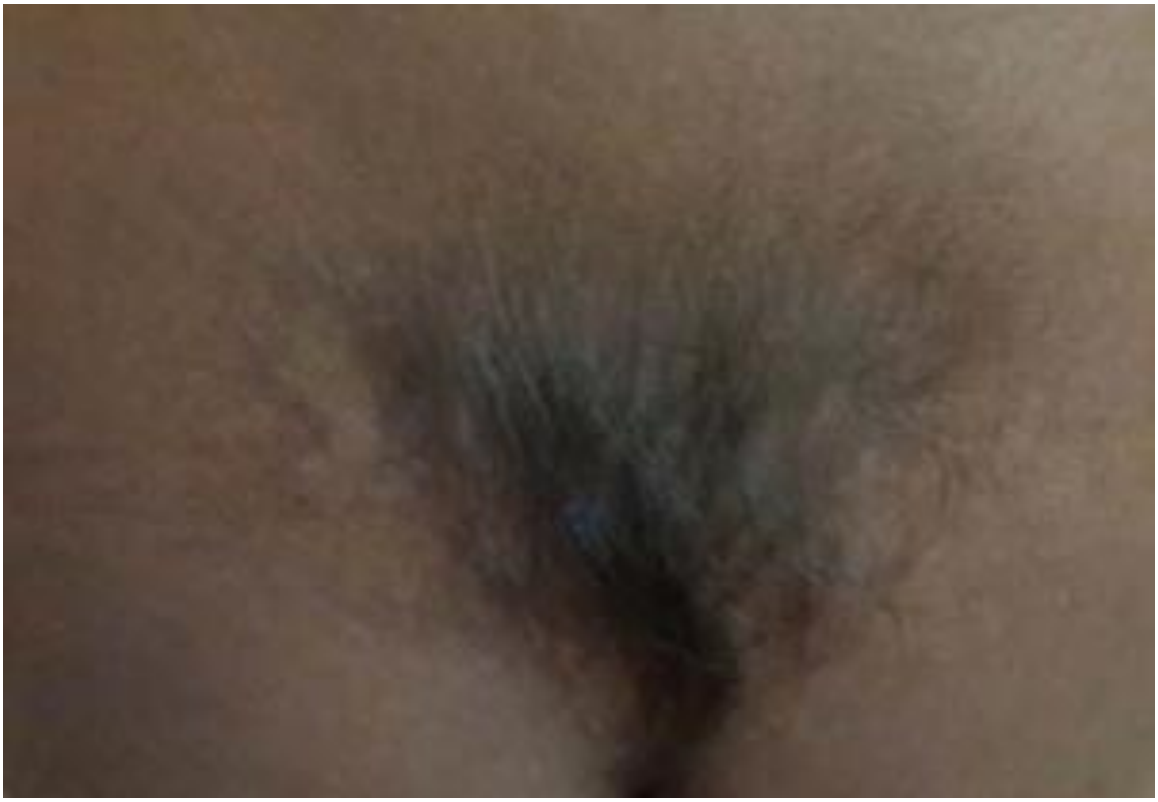

Figure 4: Hypertrichosis

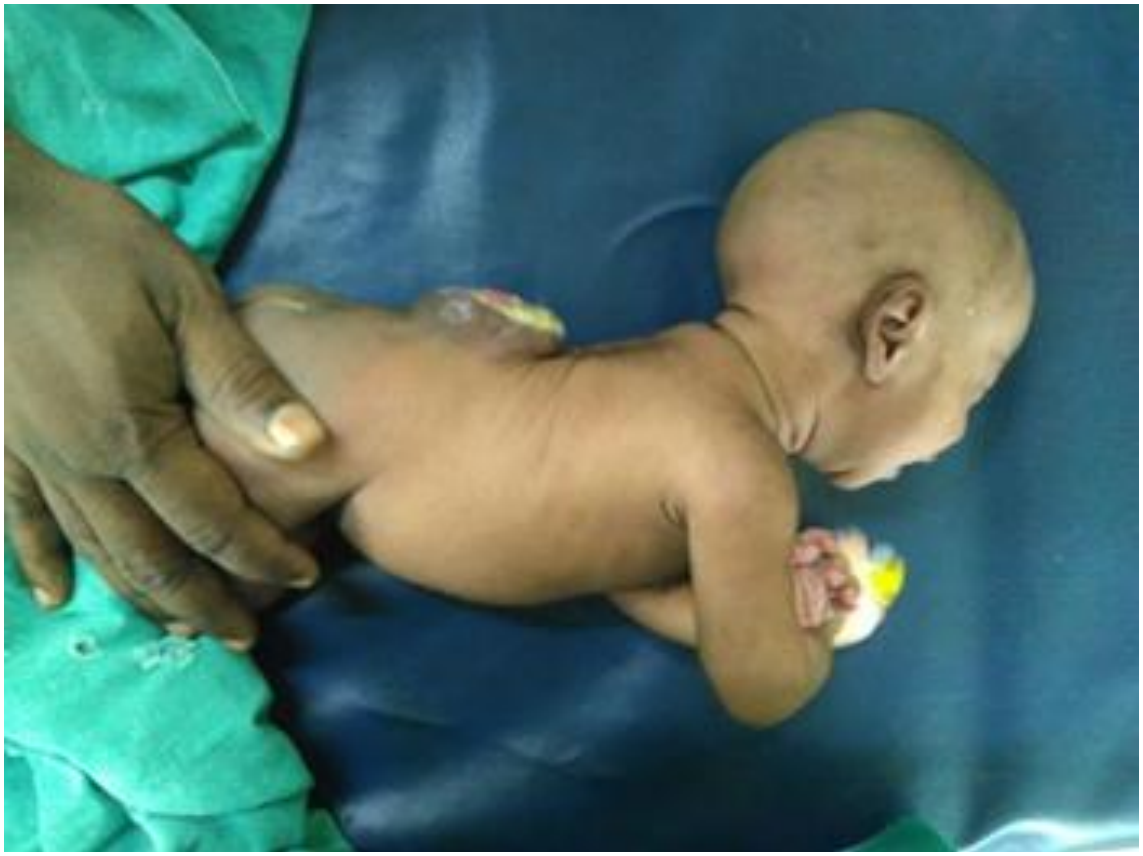

Figure 5: Dorso Lumbar Myelomeningocele

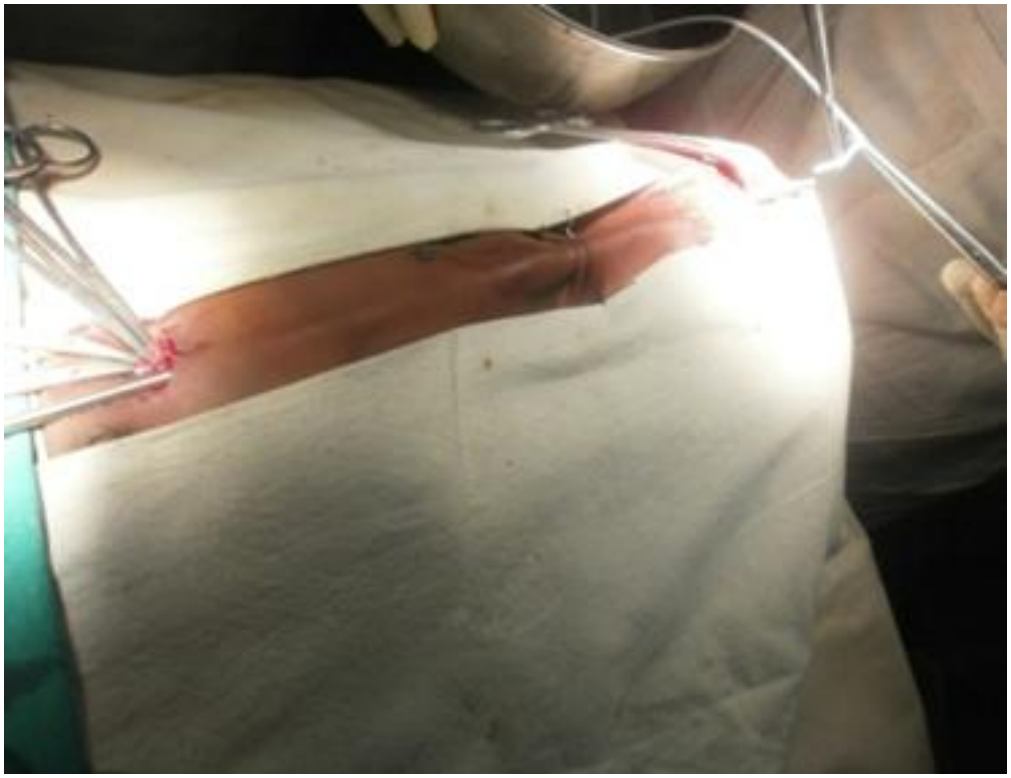

Figure 6: Procedure of Shunt Insertion

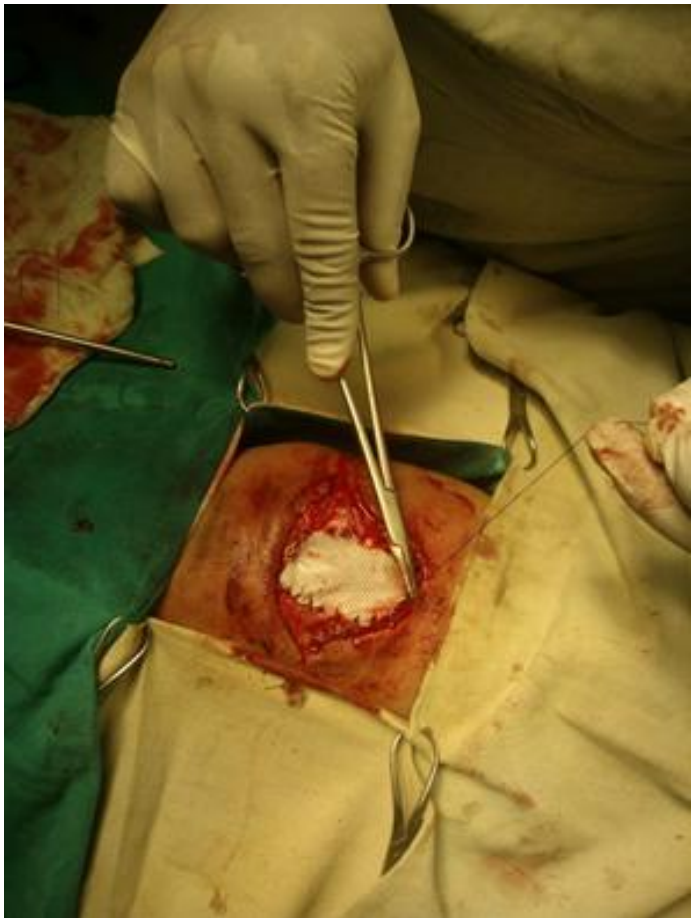

Figure 7: Dural Repair

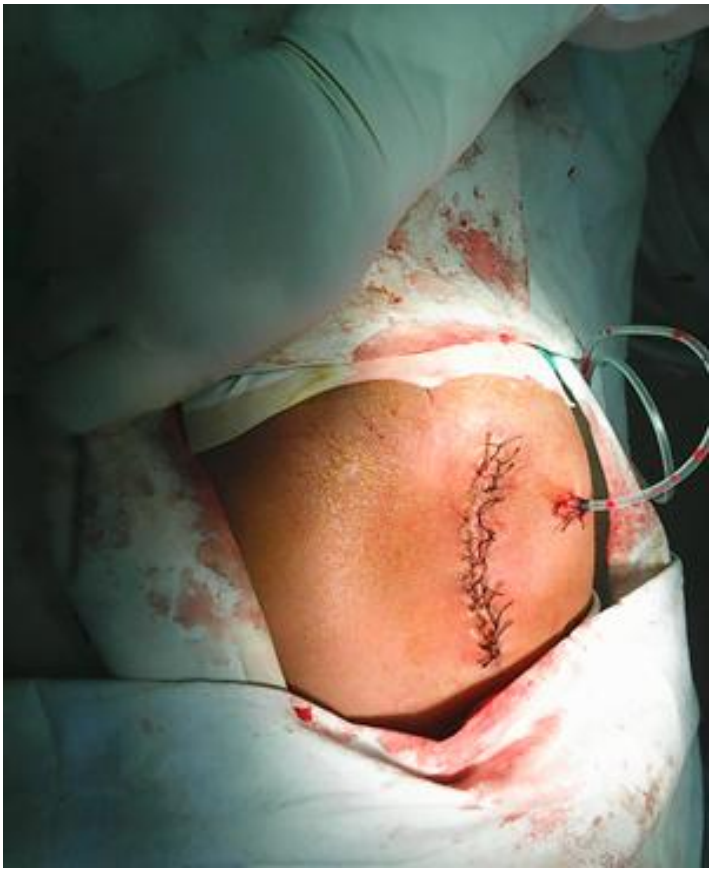

Figure 8: Wound Closure with Lumbar Drainage

DISCUSSION

The results of present series are compared with available series. Some of these are consistent with and some are in contrary to our observations. A total of 54 patients included in this study.

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. 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]

In North America, the incidence is approximately 1 per 1000 live births. In the United States the incidence is 0.6 cases per 1000 live births, but there was a gradual decline in the rate from 5.9 to 3.2 cases per 10,000 births during 1983-1990.[3] The incidence in the United Kingdom and in mainland Europe is decreasing. The disease is 2.5 times more frequent in whites than blacks; it is particularly common in Belfast, Liverpool, and Dublin and uncommon in Japan, based on the statistics of 1970s and 1980s. The incidence remains higher in Great Britain than in Japan. The trends in India are not known. The recent surveys show there is no geographic variation and a relative uniformity of incidence in all groups.[4] Rates are significantly higher in areas with a low socioeconomic status. The incidence of the spina bifida occulta increased by advent of newly diagnostic procedures.[3] In 1972, James and Lassman examined routine radiographs of 1172 consecutive autopsies and found an incidence of 5% of spina bifida occulta in adults.

The birth prevalence rate of myelomeningocele is slightly higher in girls than boys (1.2:1). There is slight predominancy of female patients in all the series in the literature.In Klenderman (1973) et al series the male and female ratio is 1:1.94.

Where as in Berman's J. Islander (2001) series the ratio is 1:1.85. In the study of Mohamed Fathy Dawoud (,2007) the ratio is 1:1.28. The incidence is high in females 57.60 % (n=31) than males 42.60 % (n=23) in present series with sex ratio 1:1.34.

In our study the age of youngest patient was one day, and the oldest was 17 years. In study of Mohamed Fathy Dawoud(2007),[3] age ranged from one day to 34 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. In a study by Choux (1993), 38% are below one year. In the study of Besnik Elshani, Basri Lenjani (2014), the incidence below one year is 90.9% and above one year is 9.1%. The mean age of the study group was 2.8 years which in comparison to other series is almost similar. In the present series 77.77% are below one year age. The least occurrence presents in more than 10 years age group in present as well as other series.

The commonest site was the lumbo sacral region in literature. In present series also commonest site was lumbo sacral region in 28 patients (51.85%) whilein Besnik Elshani&Basri Lenjani(2014) study in 72.2% cases. Low percentages of cases are noted at cervical and sacral regions in present series. In Besnik Elshani&Basri Lenjani(2014) study cervical cases notedin 5.5% andnil at sacral region.In a study of Mohamed Fathy Dawoud (2007) common site is lumbosacral in 62.5%% cases and low cases are noted at cervical (9.3%) and sacral(6.3%) cases.

Spina bifida aperta (open type) noted in 72.22% and is more common than Spina bifida occulta in the present series.Spina bifida aperta:(open type) is presented at birth with a swelling over back.The incidence of MMC is 0.2 to 0.4 per 1000 live births which is commonest in open type.[4] At birth, a midline defect in the posterior elements of the vertebrae associated with a protrusion of the meninges and cord elements through an external dural sac is noted. The neural placode is elevated in myelomeningocele due to expansion of subarachnoid space, where as in myelocele the placode is flushed with surface of back.The CSF leak or exposed spinal cord may present as the skin over swelling is poorly developed. If CSF leakage is not treated early, meningitis may occur causing significant morbidity. The extent of sensorimotor deficits would depend on the level of the meningocoele. The higher the level, worsen is prognosis. Different grades of motor weakness present in 68.6% cases.[5] Paraplegia from spinal cord malformation is common manifestation of a myelomeningocele. The segmental anatomic level of the lesion is tested to assess for neurologic deficit. It may be high at the cervical level in about 3.9% of spinal dysraphism. Thoracic myelomeningocele is 10%. Lumbosacral myelomeningoceles (80%) can be divided into a high,[6] low lumbar, and sacral groups. Scoliosis associated with a myelomeningocele can be congenital or acquired. Spinal deformities are seen more frequently in patients with higher spinal myelomeningoceles.

All most all patients with MMC have Chiari malformation.[11] Associated brainstem defects include medullary kinking, tectal beaking, and intrinsic nuclei abnormalities.In severe cases, sphincter dysfunction may present with rectal prolapsed.Nearly 80-90% of patients with MMC have hydrocephalus.The hydrocephalus may result from both obstructive and communicating components. Syringomyelia which is nonprogressive occur in 40-80% of patients.[7] More than 90% of infants with myelomeningocele have a neurogenic bladder, with 30% having lower motor neuron lesion.[8]

Myelocystocele is a localized cystic dilation of the central canal of the spinal cord.It presents as skin covered mid line masses that occur in lumbosacral area, which account 5%. Myelocystoceles are associated with cloacal exstrophy and almost two thirds (62%) of patients with cloacal exstrophy have some form of myelodysplasia. Myelocystoceles are only rarely associated with hydrocephalus or a Chiari II malformation.

Spina bifida occulta is commonly an incidental finding both in children and adults. It is usually not associated with any symptoms or signs at birth, except in lipomyelomeningocele.

The presence of cutaneous markers overlying or adjacent to the spine may be the only clue to the presence of a closed type.[9] They occur any place along the spine but most of these occur in lumbosacral area.[10] The common areDermal dimple, Hairy patch of skin (hypertrichosis, faun's tail), Smaller patch of silky hair if the dysraphism is in the cervical or upper thoracic region, Midline visible or palpable mass overlying the spine (lipoma), Dermal sinus, Capillary hemangioma.

Rudimentary tail (caudal appendage), Atretic meningocele, which is seen as a central area of thin, pearly skin surrounded by a halo of red, pink or brown (This has been likened to cigarette-burn type of skin stain).[11]

LMMs typically cause tethering of the spinal cord via the associated fibrous subcutaneous lipoma. Yamada et al demonstrated that spinal cord tethering interferes with normal energy metabolism of the spinal cord, which can lead to ischemia and progressive neural damage. Lipoma is the common form of closed spinal dysraphism, is a benign, soft, rubbery tumor composed of mature fat cells. Lipoma may lie in the dermis or in the spinal canal causing cord compression and tethered cord. A lipoma may overlay a myelomeningocele. Lipomas with a dural defect include both lipomyeloceles and lipomyelomeningoceles.

Dermal sinus usually appears as a midline dimple, with or without a tuft of hair, and may have pigmentation of the surrounding skin. The common sites are in the lumbar or lumbar-sacral area.[12] Findings include cutaneous markers, leaking of cerebral spinal fluid (CSF), and neurologic deficit or infection. Over 50% extend into the spinal canal, placing the infant at risk for meningitis.Neuroenteric cysts are rare lesions and lined with endodermal epithelium. They are commonly located in the thoracic or cervical regions and in ventral position. The cysts derive from endodermal inclusions within the spinal column and represent a persistent communication between endodermal and neuroectodermal tissues.

Prenatal sonography can help to predict karyotypically abnormal fetuses with spina bifida. However, 20% are missed if sonography is used alone in the setting of a prenatally detected spina bifida. Some authors believe that cytogenetic analysis is justified in these instances.Plain x-ray films of the spine help identify occult spinal abnormalities. This is useful for observing an associated scoliosis.

Craniospinal MRI was done to all cases in this study for evaluation of associated anomalies. In the present study myelomeningocele was commonest finding (61.11%) and also correlating with Mohamed Fathy Dawoud study in which myelomeningocele noted in 46.85% patients. Meningocele(3, 4 cases), lipomyelocele (2,1 cases), diastematomyelia in (2, 1 cases), dermal sinus (2,1 cases), spinal lipoma in (one case in both series) are less frequently present in both present and Mohamed Fathy Dawoud series.[3]

In the present series, hydrocephalus was the most common associated cranial abnormality being seen in 23/54 (42.59%) cases. This incidence is lower in comparing with western literature. In Raj Kumar et al,[13] study also hydrocephalus was the most common associated cranial abnormality noted in 60/102 (58.8%) cases. The second most common associated anomaly was Arnold chairi malformation type11 in both present and Raj kumar et al study present in 38.88% and 50.9% cases respectively. Corpus callosal agenesis or thinning as an associated feature in 5.55% of patients but in Raj kumar et al study high percentage (12.22%) of cases is noted. Tethered cord syndrome and syringo hydromyelia were other common associated anomalies present in considerable percentages.

The dura is dissected from the underlying soft tissue. The dural closure is done water tightly with or without graft. The overlying skin is dissected from underlying fascia and musculature, mobilized and approximated. If necessary, relaxing incisions or flaps may be used to close larger defects. Excision of sac & repair is done in 33 cases (61.11%) in the present study. In a study by Besnik Elshani&Basri Lenjani(2014) 76.6% patients treated by Excision of sac& repair. In the present study 25 cases (50%) needed

ventriculoperitonealshunt(VPS). Among them 24% developed hydrocephalus after repair of myelomeningocele sac while in present study 7.4% cases developed hydrocephalus requiring ventriculoperitoneal shunt. In 80% cases primary closure was possible and rest needed reinforcement with para spinal fascia in Banskota N et al study. In the present study in 24 cases (72.3%) primary closure is possible. The remaining 9 cases needed paraspinous fascia, fascia lata or synthetic Goretex graft to repair the dural deficiency to prevent the CSF leak. The associated hydrocephalus is an important factor for prognosis. Ventriculo peritoneal shunt insertion before the repair of lesion can avoid CSF leak and pseudo- meningocele formation. 21 cases of Arnold Chairi Malformation type 2 noted in the present study but none were treated by hind brain decompression which is correlating to other series.

Causative factors ofcord tethering result from a variety of conditions like anatomical (conus is placed at a lower level) physiological (tight or thick filum), developmental (secondary to congenital anomalies, terminal lipoma, intraspinal lipoma, lipomyelomeningocele, split cord malformations, sacral agenesis and other occult dysraphic states) Postoperative tethering (after surgery for myelomeningocele or recurrent adhesions from previous surgery often termed retethering. These causative factors ofcord tethering such as lipoma, epidermoid, bony spur, fibrous septum, were dealt simultaneously in single procedure. The principles of surgery for tethered cord syndrome are to complete untethering of cord and proper dural reconstruction with adequate CSF space around the spinal cord to prevent retethering.The radiological findings were confirmed intraoperatively. In a study of Raj Kumar, S.N. et al (2003),[13] the other findings like arachnoid bands atrophic conus, an associated dermoid are also encountered responsible for tethering of the cord and neurological deficits, these are dealt simultaneously with untethering of the cord. Dermal sinus could not be noted in this study.In the present study, one case of dermal sinus was treated with good outcome. In the present study dethering of cord was done in 35.18% cases. 81% of cases are treated by dethering in a study of Pang (1982).

Lipomyelomeningocele treatment is complex and has high risk of neurological deterioration. Various factors influence the outcome. These are anatomical factors (size of the lipoma, location (midline or paramedian), wide bony defect, defective muscles and fascia, poor cleavage at neurolipomatus junction) physiological (degree of traction, ability of cord to withstand the effects of traction) and pathological (vascularity of fibrolipomatous structures and associated anomalies). The principles of management include vertical skin incision, dissection of subcutaneous lipoma, identification of upper and lower laminae and performing laminectomy, identification of normal dura above and below and separation of dural tube from the thoraco lumbar fascia all around. Replacement of free fat over the dura fills the dead space, protects the soft tissue, prevents CSF leak and prevents the skin edge necrosis. Asymptomatic patients with spinal lipomas should be treated by prophylactic surgery and patients with symptomatic should be treated surgically although surgical treatment of these lesions is more complicated and carries a higher risk to the patient.[14,15] The treatment of asymptomatic patients with conus lipomas remains a topic of debate .Finally, the risks of surgical intervention appear to be declining in recent series involving newer operative technologies and the refinement of surgical goals. intraoperative monitoring decrease the complication rates and is also highly variable in reported series.[15] Available monitoring techniques include rectal EMG, urethral EMG, rectal and bladder pressure, continuous EMGs, evoked EMG, and SSEPs. In study of Choux (59%) and in Anderson study (49%) cases of lipomas are treated. In Choux study (1993), there are 59% of cases are noted. In the present study less (18.51%) cases of spinal lipomaare treated in compared to other series. In the case of split cord malformation type 1, complete excision of the bony spur and in type II fibrous septum excision done which divide the cord into two halves.[16]

After surgical repair, CSF leak from the wound site occurred in 20.37% cases in the present study and in a study of Raj Kumar (2003) there are 32.9% of cases noted. In Banskota study only 4.87% cases noted. These cases treated by medical management with acetazolamide and prone position or re-exploration and duraplasty using fascia lata graft. Lumboperitoneal shunt done in 5.55%(3 cases) in present study where as in Raj Kumar et al 9.67% of cases treated by Lumboperitoneal shunt. Pseudomeningocele developed in 9.25% cases in the present study. In Rajkumar et al study 14.1% cases of pseudomeningocele developed. In study of Bansokota et al no cases of pseudomeningocele cases noted. Post operative hydrocephalus developed in 7.4% of cases in the present study which are managed by ventriculoperitoneal shunt.[17] In contrast to this Bansokota et al study noted 24.3% of cases of hydrocephalus after meningomyelocele sac excision and repair. Ventriculoperitoneal shunt insertion before the repair of lesioncan avoid CSF leak and pseudo- meningocele formation. Ventriculitis (1 case) and meningitis (4 cases) were noted in low percentage of cases in the present study correlating to other studies

Pain is the most common symptom to improve followed by motor deficits. Sensory deficits and bladder dysfunctions remain status quo post operatively. In the present study pain improved in all 9cases (100%) correlating the same with the study of Raj Kumar et al.[17] There was significant improvement in patients presenting withsigns of upper or lower motor signs in 12 (41.37%) patients while 3 patients deteriorated in postoperative period . In study of Raj Kumar et al 51.4% patients showed no improvement or deterioration compared to 48.2% in the present study.[17] Sensory loss and sphincter dysfunction did not improve significantly in the present study which is noted in Raj Kumar et al study also.

There are many risk factors for neural tube defects which include folic acid deficiency advanced mother's age lower socioeconomic status,[18,19] 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 checkups 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.[19]

CONCLUSION

Diagnosis of closed spinal dysraphism is difficult because of the subtle appearance of some cutaneous markers and insidious onset of neurological and physical symptoms. Delayed diagnosis and management of these patients cause the multiple risks like infection, developing neurologic deficits. MRI is the investigation of choice, but high-resolution CT can be complimentary especially in split cord malformation type 1 for demonstrating bony spur. MRI is excellent in delineating spinal cord and paraspinal soft tissue. These investigations must be done in all children with cutaneous markers of occult spinal dysraphism even without any neurological deficit. Management of spinal dysraphism patients is complex and needs close coordination between, neonatologist, neurosurgeon, plastic surgeon, orthopedician, urologist and rehabilitation experts.

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