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Original Research Article

Exclusive Dorsal Spinal Dysraphism; Retrospective Institutional Analysis of the Rare Entity

Ganesh Prabhu J.[1], Hariharasuthan P.[2], Dhandayuthapani V.[3], Sachin Biradar[4]

[1]Senior Resident, Department of Neurosurgery, KAPV Medical College and MGM GH, Tiruchirappalli
[2]Assistant Professor, Department of Neurosurgery, KAPV Medical College and MGM GH, Tiruchirappalli
[3]Associate Professor, Department of Neurosurgery, KAPV Medical College and MGM GH, Tiruchirappalli
[4]Assistant Professor, Department of Neurosurgery, KAPV Medical College and MGM GH, Tiruchirappalli

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Corresponding author: Dr. Ganesh Prabhu J.
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Abstract

Introduction: Lumbosacral Meningomyelocele (MMC) is a common form of spinal dysraphism. Pure dorsal form is quite rare accounting for only 1–8% of all spinal dysraphism. There are only a few case series related to dorsal MMC in the literature. This study presents a series of dorsal meningomyelocele, reviewing its clinical features, surgical management and other management strategies.

Objective: To discuss the clinical features, surgical management and other management strategies in accordance with the literature.

Materials and Methods: We conducted a retrospective analysis from January 2019 to December 2023. 360 cases of spinal dysraphism were diagnosed during this period out of which 18 cases were dorsal meningomyelocele. All cases of open and closed spinal dysraphism admitted in our hospital were included in the study except those patients who were in sepsis or low birth weight.

Results: Out of 18 patients of dorsal MMC, 11% (n=2) were infants. The youngest patient was 5 days old and the oldest patient was 9 years of age. There was equal sex distribution. The most common sites of dorsal meningomyelocele were D5-D8 region which constituted 61% (n=11). The most common associated anomalies were hydrocephalus (22%) followed by Chiari II (16%). 50% of neural tube defect (NTD) babies were born to mothers who had hyperemesis gravidarum. 72% of mothers did NT scan and among them 61% were not diagnosed of NTD. Among 18 patients, 16 had tethering at same level and 2 had tethering at lower level for which separate incision was made and detethering of filum terminal was done. 22% had skin tag, 11% had hair and dermal sinus as neurocutaneous markers. All patients underwent microsurgical sac excision with intra operative neuro monitoring. Pre- and postoperative neurological examination of all patients were normal. Patients are still under regular follow up and showed no neurological impairment with a mean follow up of 12 – 18 months.

Conclusion: Dorsal meningomyelocele is structurally and clinically different from lumbosacral meningomyelocele and has more favourable outcomes after surgery. Preoperative magnetic resonance imaging and detailed patient evaluation are recommended to identify the dorsal meningomyelocele's sac, spinal cord structure and additional anomalies. Surgical treatment should be done early and intradural exploration is recommended in addition to resection of the sac. Folic acid supplements should be recommended within three months, for a minimum period of 100 days before pregnancy to prevent NTD.

Keywords: Meningomyelocele, Neural Tube Defects, Dorsal, tethering, Rare Entity.

Introduction

Meningomyelocele is a neural tube defect with protrusion of the neural tissue and the meninges through a vertebral defect. The embryological cause for meningomyelocele is failure of closure of neural tube during third week of gestation leading to the constellation of defects. These can occur anywhere along the spine. The lumbar meningomyelocele are more common and maybe associated with neurological deficits. On the other hand, cervical and dorsal meningomyelocele are very rare making up 1-8% of all neural tube defects [1][2][3]. Higher the meningomyelocele fewer the

neurological deficits and better outcomes after surgeries when compared to lower meningomyelocele. Diagnosis and treatment plans should be made by defining specific clinical traits. Thus, an inappropriate surgical technique may cause delayed neurological deterioration. There are not many publications about these lesions in the literature [2].

The majority of the publications investigating these lesions are limited to case report. In this study, we present 18 surgically treated cases of dorsal MMC and discuss their management strategies in accordance with the literature.

Materials and Methods

It is a retrospective study with purposive convenient sampling, conducted at K.A.P.V Government Medical College and associated MGM Government Hospital, Tiruchirappalli, Tamil Nadu. A total of 360 patients presented with congenital spinal dysraphism in our hospital from January 2019 to December 2023. Of these 360 patients, 5% (n=18) were diagnosed with dorsal spinal dysraphism.

The clinical data of the 18 patients, including their demographic profile, associated neurological malformations, radiological finding, surgical treatment strategies, retrospective results and follow-up were analysed.

Inclusion Criteria: All cases of open and closed dorsal meningomyelocele [D1-D12] presenting to our hospital in the above-mentioned period and who were willing to participate in the study.

Exclusion Criteria: Patients with low birth weight, patients in sepsis and patients not willing to participate in the study.

Neurological and radiological examinations, including magnetic resonance imaging (MRI), were performed for all patients. Following diagnosis, all patients had surgery using standard micro neurosurgery techniques.

Statistical analysis: Data were analysed using the SPSS 22.0 statistical program. Number and percentage values were given for categorical variables. Categorical variables were evaluated with the Chi-Square test. The limit of significance was taken as $p \leq 0.05$.

Surgical Procedure: The patient, with all pressure points supported, was placed on the operating table under general endotracheal anaesthesia in the prone position. In the surgery, a longitudinal skin incision was made over the mass. The skin was meticulously dissected from the incision and sac was dissected all around. Above and below laminectomies were done to visualise normal dura and sac was opened. A thick fibrous band extending to the dorsal surface of the sac was seen. After the dura was separated around the peduncle, it was observed to enter the expanded cord in the posterior midline.

The junction in the sac was sharply dissected and released using the microsurgical dissection technique. Stem amputation was performed at the cord level so that the fibro neural bands were completely free. In patients with neural tissue in the sac, after the fibrous bands were released, the neural tissues were left in place to avoid postoperative neurologic deficits. All patients underwent excision of the sac with standard micro-neurosurgery techniques followed by intradural exploration and handling of other associated anomalies.

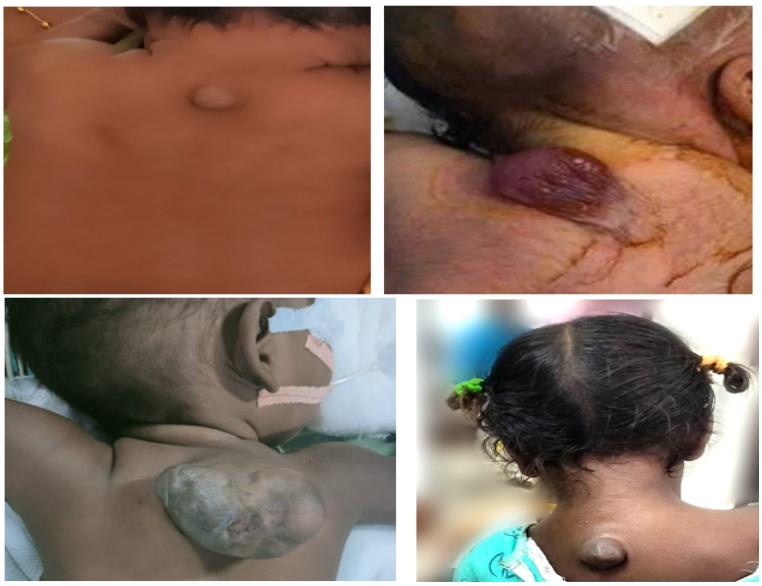

Figure 1,2,3,4: Various Locations and Sizes of Presentation (With Permission from Patient Attenders)

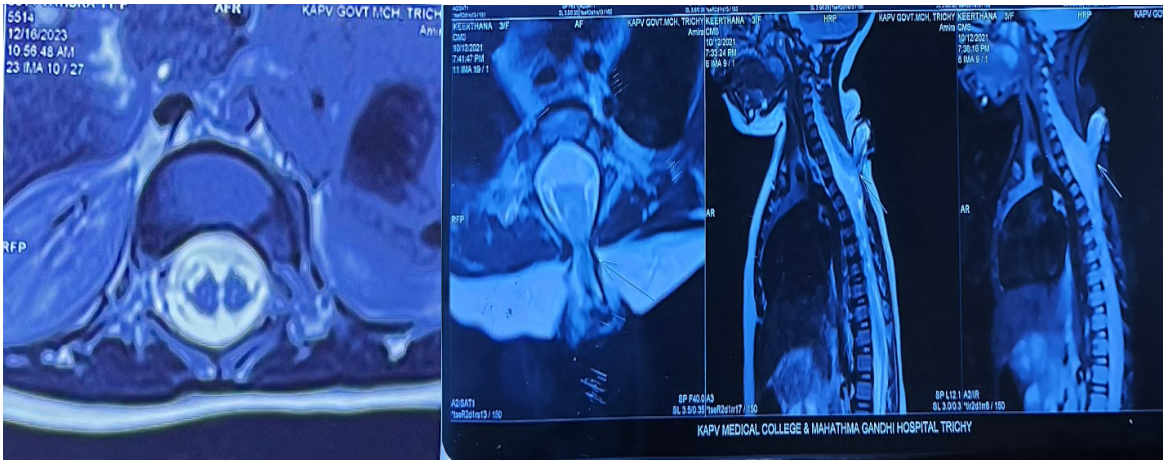

Figure 5 : MRI Findings of Few Cases

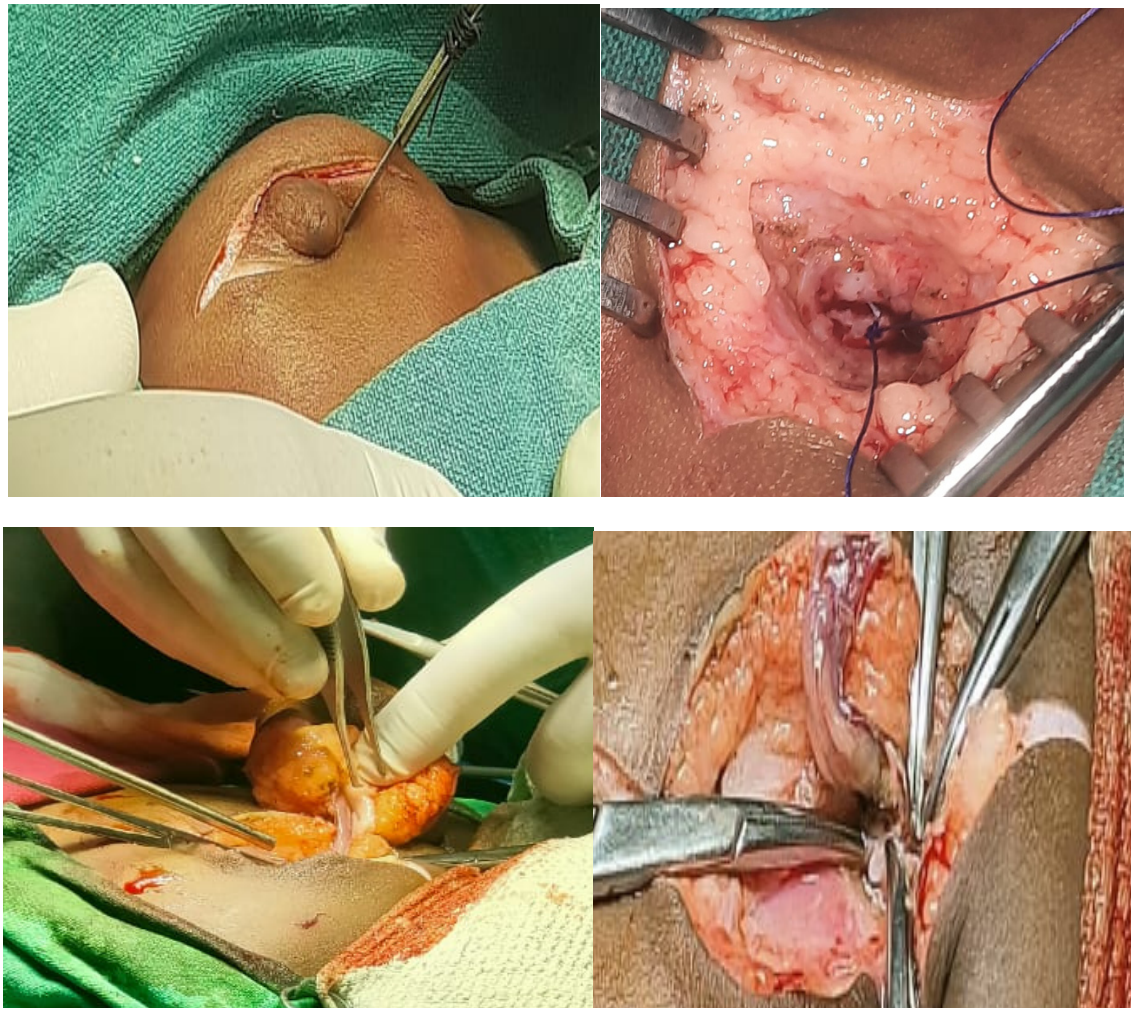

Figure 6,7,8,9: Intra Op Photos

Results

Out of 18 patients studied, 11% of the patients were infants. The youngest was 5 days old and the oldest was 9 years of age. In this 50% were male, and 50% were female. The most common site of dorsal meningomyelocele was D5-D8, 61% (n=11). Associated anomalies were present in 39% of the patients.

The most common anomaly was hydrocephalus 22% (n= 4) followed by Chiari II 16% (n=3).

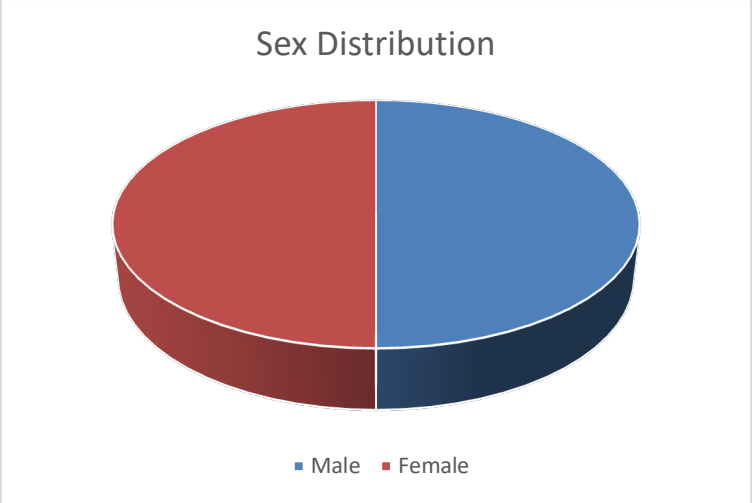

Chart 1: Sex Distribution

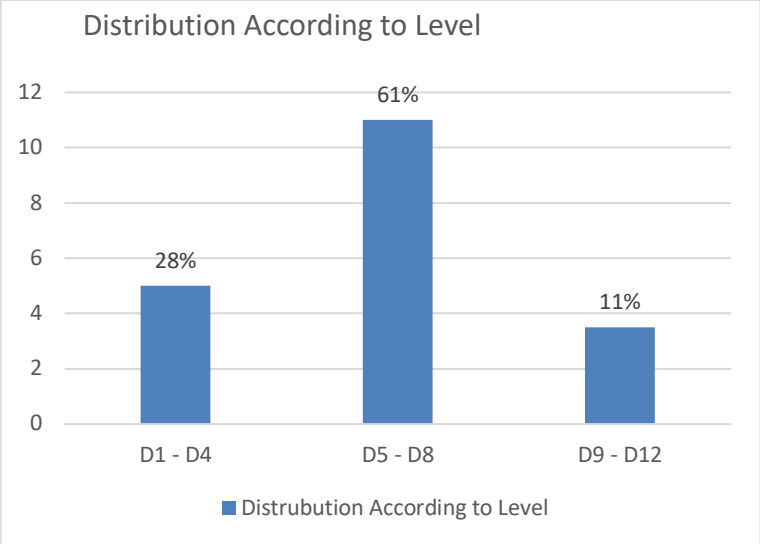

Chart 2: Distribution According to Level

All mothers who gave history of hyperemesis gravidarum in our study delivered a NTD baby with 100% association. Among 18 mothers, 6 were primigravida. The remaining 12 were second gravida whose first child was normal. 72 % (n=13) of mothers underwent NT Scan and among them 61% (n= 11) were not diagnosed of NTD. Among 18 patients, 16 had tethering at same level and 2 had tethering at lower level for which separate incision was made and detethering of filum terminal was done. 22% (n = 4) had skin tag, 11% (n =2) had hair and dermal sinus each as neurocutaneous markers. Frequency distributions according to some variables are presented in Table 1. All patients underwent sac excision. Pre- and postoperative neurological examinations of all patients were normal. Patients are under regular follow up and showed no neurological impairment.

Table 1:

Neurocutaneous Marker	Percentage
Skin Tag	22% (n=4)
Hair	11% (n=2)
Dermal Sinus	11% (n = 2)
None	56 % (n=10)

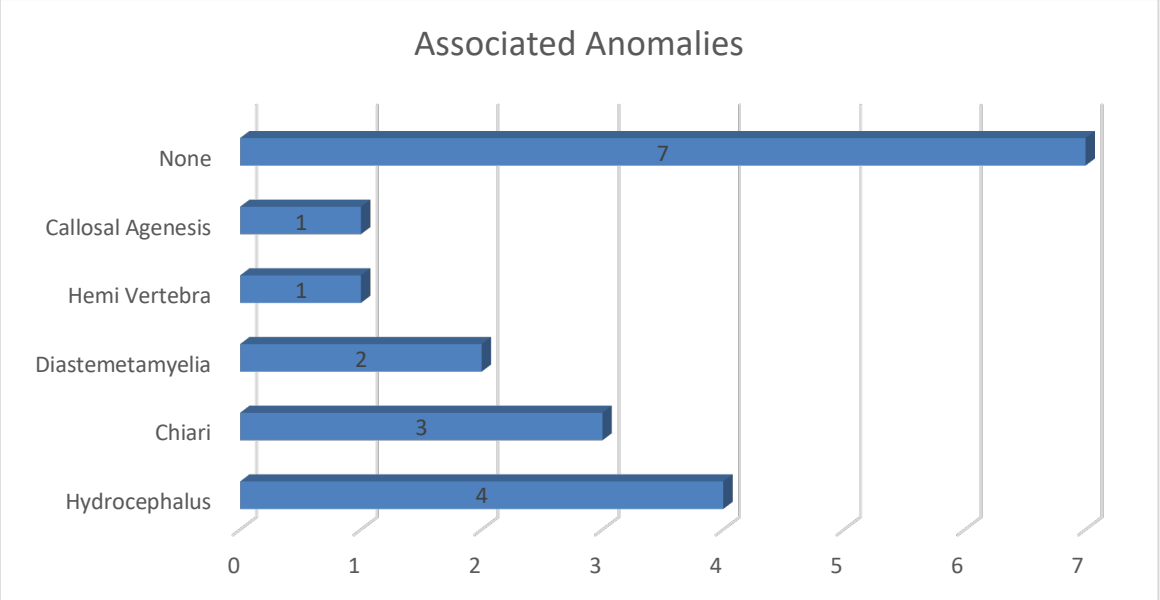

Chart 3: Associated Anomalies

In the postoperative course of all patients, no changes were noted in their neurological status. Postoperative complications such as cerebrospinal fluid leakage and infection were observed, which were managed conservatively. There was no mortality.

Discussion

Meningomyelocele is a subgroup of spinal dysraphism which most commonly occurs in the lumbar and sacral regions of the spine. Dorsal MMC is relatively rare and has a much lower incidence than the lumbosacral types [4-6]. Dorsal meningomyelocele account for only 1–8% of all cases of spinal dysraphism. [7,8,9]

Dorsal MMC clinically differs from the more widespread lumbosacral variants [10]. Lumbosacral MMC are usually fragile and covered by a thin arachnoid layer, whereas higher spinal lesions are always covered with full-thickness skin. Due to this, CSF leakage is unusual in high spinal lesions. The neural compartment of lumbosacral MMC is usually a flattened terminal neural placode, whereas in high spinal lesions there is a nearly closed neural tube. Patients harboring lumbosacral MMC often exhibit serious neurological dysfunction at or below the level of the lesion, whereas patients with high spinal lesion experience mild or no deficits. [1,11]

Recently, dorsal MMC has been classified into 3 types according to the structures found inside the cyst: Type I lesion has a stalk, either neuroglial or fibrovascular, connecting the meningocele sac and spinal cord; Type II is a non-terminal myelocystocele; and Type III is a true meningocele. [8] Our study has shown the following types according to MRI, Type I 67% (n=12), Type II 22%(n=4) and Type III 11% (n=2). Type I is associated with easy dissection and operative time is less. [23]

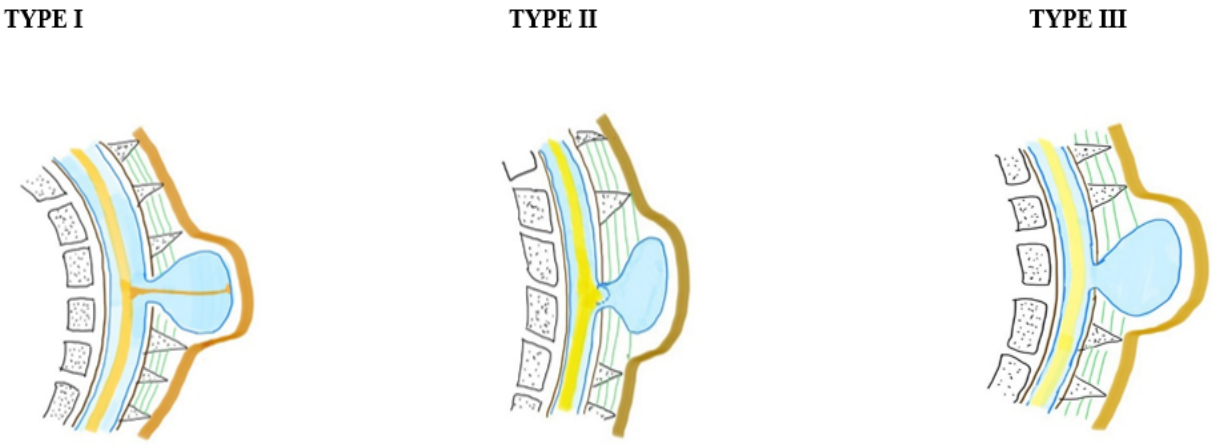

Figure 10,11,12: Types of DMC

Various theories are being considered for the origin of clinical presentations of dorsal meningomyelocele [1,8,5,12]. It was originally clarified on the basis of the regular closing theory. Accordingly, the neural tube closing stops in the dorsal region and spreads both rostrally and caudally in a Zipper-like fashion, with the cranial and caudal neuropores closing last. Van Allen improved the multisite theory to clarify the formation of neural tube defects.

The content is not a mislaid neural plate, but a neuroglial stem that originates from the posterior surface of the dorsal cord and produces tethering. These characteristics are better elucidated by the Limited dorsal myeloschisis theory (LDM) of Pang and Dias [13]. Accordingly, neurulation proceeds smoothly and the primary configuration of the neural tube is maintained, except for a subtle shift in the dorsal midline. There is never really a separation between the cutaneous ectoderm and the neuroectoderm. Even though myofascial tissues evolve into dorsal median stem of central nervous system tissue, it continues the original link between the almost closed neural tube and the still lightly spaced cutaneous ectoderm. Apart from an extension around this stem, the meninges also develop. As CSF is formed, it slowly becomes trapped in the dural fistula and slowly widens the dome, building myelomeningocele [4].

In our study, 18 dorsal MMC cases were diagnosed out of 360 spinal dysraphism cases in our hospital between January 2019-December 2023. Dorsal MMC incidence was found to be 5% and it is comparable with the available literature [1,2,3].

The general clinical presentation of dorsal MMC is a subcutaneous swelling in the posterior thoracic region without significant neurological impairment [5]. Our patients also presented with posterior midline swelling with overlying intact full thickness skin.

The most common associated abnormalities have been reported to be, in descending order of frequency, hydrocephalus and Chiari type 2 malformations [14,15]. They may have other spinal abnormalities such as syringomyelia, diastematomyelia, tethered cord, Klippel-Feil syndrome, thoracic hemivertebrae compared to caudal myelomeningoceles. Although the frequencies of the development of hydrocephalus and Chiari type 2 malformation are similar, the rate of intervention for Chiari abnormalities is much lower than that of hydrocephalus. We noted in our study no associated anomalies in 39% and associated anomalies as hydrocephalus in 22%(n=4), Chiari I Malformation in 16% (n=3), diastematomyelia in 11% (n=2) followed by thoracic hemivertebra and callosal agenesis each accounting for 5.5%. The presence of hydrocephalus may be symptomatic and may even cause deterioration of the neurological condition. VP shunt may be necessary in nearly 85% of the cases of hydrocephalus accompanying myelomeningocele. Indications for intervention for Chiari abnormalities are symptoms such as postnatal stridor, bradycardia and dysphagia due to the dysfunction of cranial nerves exiting from this level. Neonates are thought to tolerate this abnormality slightly better than hydrocephalus [15].

Another issue of discussion is the order of operations in cases of hydrocephalus accompanying myelomeningocele. Some studies have suggested that a shunt should be placed in a second operation, while some studies have demonstrated that the risk of ventriculitis and shunt infection decreases if sac repair and VP shunt placement are performed in the same operation (14,17).

We considered that the best option is to decide according to the condition of the patient and the circumstances. We decided to place the VP shunt first because we thought that the ventricle anatomy will change due to the possible draining of CSF following shunt placement and we planned sac repair one week later. For patients with Chiari malformation, we managed conservatively as these patients presented with no neurological abnormality.

The timing of the intervention for myelomeningocele is debatable. Waiting up to 72 hours can be allowed, particularly in cases of intact sac, and this period is thought to aid in the psychological preparation of the family and better assessment of the patient preoperatively. On the other hand, more satisfactory results have been reported in terms of motor development in patients with early intervention [16,17]. In addition, early intervention results in some advantages such as shorter duration of hospitalization and shorter duration of antibiotic use. Open defect should be repaired as early as possible.

We operated all 18 patients without any delay at the age of presentation to the hospital as documented in Table 2. Surgical treatment should include a bi-level laminectomy, intradural exploration to remove the band of tissue connecting the spinal cord, and resectioning of the related protruding sac.

Table 2:

Age Of Presentation	Number	Percentage
0 -3 months	11	61
4 - 6 months	5	27
7- 9 months	1	6
10 months - 1 year	0	0
≥ 1year	1	6

A microsurgical technique with intra operative neuromonitoring should be utilized to detect any spinal cord tethering or any related spinal abnormality (18). In two of our cases tethering of filum terminale was noted at lower levels. Hence separate incision was made and detethering done.

In all of our cases, intradural exploration was performed to remove the tissue bands connecting the cord and neurologic deterioration was not observed in any of the patients after sac resection. Therefore, dorsal MMC surgery should be aimed not only for cosmetic reasons but primarily at the prophylactic release of all connected cord elements and avoiding functional loss. Untreated children may develop neurological deficits later in life [4,19]. Dorsal MMC may be associated with cranial and spinal anomalies, hence MRI (magnetic resonance imaging) is required to show both fine anatomical relationships at the lesion level and to detect other spinal cord abnormalities [20,21]. Therefore, cranial and whole spinal MRI should be performed in patients diagnosed with dorsal MMC. A lack of understanding of the anatomical details of lesions and related abnormalities can lead to treatment failure and poor prognosis.

The prognosis of the dorsal MMC patient is quite when compared with patients with thoracolumbar and lumbosacral meningomyelocele. However, delayed neurological deterioration may occur in growing children because of retethering. Therefore, it is important to follow up the patients regularly after surgery. Since all mothers with hyperemesis gravidarum were associated with NTD, folic acid supplementation should be recommended three months before conception for prevention of NTDs.[22]

Conclusion

To conclude Dorsal MMC is a relatively rare disease, hence the literature is scanty. This emphasizes the need for a more vigilant case reporting and patient follow up. Dorsal meningomyelocele has a better neurological outcome compared to lower lesions. However, neurological deterioration may occur later in life due to cord tethering.

In the presence of CSF leakage or signs of infection, the lesion must be repaired urgently. Excision of the lesion with intra-dural exploration of the sac to release any potential adhesion bands, is the recommended management. In addition, identification and management of associated congenital abnormalities is inevitable as it affects prognosis.

Intra operative neuro monitoring helps in better outcome. Folic acid supplements should be recommended before conception for a minimum period of 100 days to prevent NTD.

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