"Demographic profile and outcome of meningomyelocele in government medical college of Western UP"

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Abstract

Introduction: Meningomyelocele is a birth defect in which the backbone and spinal canal do not close before birth. Neural tube defects are one of the most common congenital malformations affecting the brain and spinal cord & Meningomyelocele is one of the commonest neural tube closure defects. The global prevalence of Meningomyelocele has been reported to be 0.8–1 per 1,000 live births. Hydrocephalus may affect as many as 80% of children with Meningomyelocele. The use of periconceptional folic acid supplements, prevents ~50–75% of cases of neural tube defects. This study aims to evaluate demographic and clinical profile, associated anomalies & outcome of children with Meningomyelocele.

Materials and Methods: 113 patients with Meningomyelocele admitted to SVBP hospital, Meerut between Sept'16 to Aug'17, were analyzed prospectively. The data regarding demographic and clinical profile with associated congenital anomalies were obtained by questionnaire- interview with the parents and patients themselves. MRI was the essential investigation in all patients. The data was analyzed by SPSS 21.0 version.

Results: Mean age of presentation is 9.1 months. M:F ratio is 1.1:1. Lumbosacral region is involved in 73.5% patients. Hydrocephalus was the most common (64.6%) associated anomaly. In 92% cases there is no history of folic acid supplementation & all patients were from low socioeconomic status. Excision & repair of Meningmyelocele is the most commonly (51.3%) performed surgery while surgical site infection is the most common (19.5%) complication. In our study (91.2%, n=103) patients improved & discharged, 2.7% (n=3) patients expired, 4.4% (n=5) patients left against medical advice while 1.8% (n=2) patients referred to higher centre due to various reasons.

Conclusion: Low socioeconomic status & no maternal supplementation of folic acid are the important risk factors for the development of Meningomyelocele. Timely intervention with excision and repair gives good result. Modified rhomboid flap can be use to cover large defects.

Keywords: Meningomyelocele, Hydrocephalus, Modified Rhomboid flap.

Introduction

Meningomyelocele is a birth defect in which the backbone and spinal canal do not close before birth. At four weeks of gestation, the lateral edge of the neural plates elevate towards each other and fuse to form a tube known as the neural tube. Failure of this process results in a neural tube defect.[1] The condition is a type of spina bifida. Neural tube defects are one of the most common congenital malformations affecting the brain and spinal cord & Meningomyelocele is one of the commonest neural tube closure defects. The global prevalence of Meningomyelocele has been reported to be 0.8–1 per 1,000 live births.[2,3] Hydrocephalus may affect as many as 80% of children with meningomyelocele.[4,5] The exact causes of neural tube defects are not known. Low intake of folic acid before getting pregnant and in early pregnancy increases the risk of having a fetus affected by NTDs, including Meningomyelocele.[6] The use of periconceptional folic acid supplements, prevents ~50–75% of cases of neural tube defects. Proper management of affected children can lead to a meaningful & productive life and poorly managed cases of MMC can be a devasting obstacle not only for patients but also for the patient's family. This study aims to evaluate demographic and clinical profile associated anomalies & outcome of children with Meningomyelocele.

Materials and Methods

113 patients with Meningomyelocele admitted to SVBP hospital, Meerut in the department of surgery & department of paediatric medicine between Sept'16 to Aug' 17, were analyzed prospectively. The data regarding demographic and clinical profile with associated congenital anomalies were obtained by questionnaire- interview with the parents and patients themselves. Follow up was done to note for complications. MRI was the essential investigation in all patients. The data was analyzed by SPSS 21.0 version.

Result

In our series of 113 patients most patients (70.8%, n=80) presented at the age of less than 6 month. (Mean =9.1months.) One patient presented at the age of > 5 year. Table 1)

Table 1:

Age Group	n	%
0-6 months	80	70.8
6-12 months	19	16.8

1-2 year	3	2.7
2-3 year	4	3.5
3-4 year	4	3.5
4-5 year	2	1.8
>5 year	1	0.9
Total	113	100

Table 2: The sex ratio is M:F 1.1:1.

Sex	n	%
Male	58	48.7
Female	55	51.3
Total	113	100

Table 3: Most of the patients (51.3%, n=58) came from Meerut region

Region	n	%
Meerut	58	51.3
Out side the meerut	55	48.7
Total	**113**	**100**

Table 4: Most commonly involved site was Lumbosacral region (n=83,73.5%). While occipital region was the least commonly (2.7%, n=3) involved

Site	N	%
Cervical	8	7.1
Dorsal	9	8.0
Dorsolumbar	2	1.8
Lumbosacral	83	73.5
Occipital	3	2.7
Sacral	8	7.1
Total	113	100

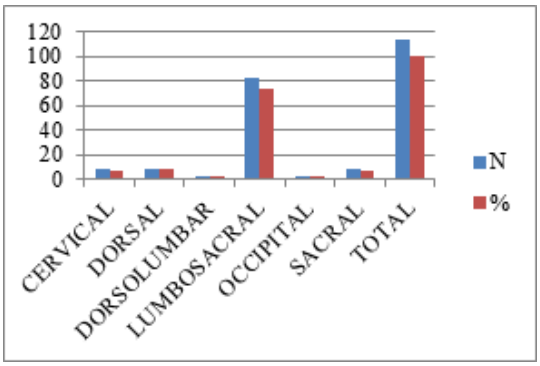

Fig. 1:

Table 5: In most patients (69.9%, n=79) defect was lagre i.e > 5 X 5 cm^2

Size in cm^2	n	%
LARGE (> 5 X 5 cm^2)	79	69.9
SMALL (< 5 X 5 cm^2)	34	30.1
Total	113	100

Most common associated anomaly was Hydrocephalus (64.6%, n=73) followed by Arnold chiari malformation II (53.1%, n=60). Aqueductal stenosis (0.9%, n=1) & colpocephaly (0.9%, n=1) were the least common associated anomaly. (Table 6)

Table 6:

Associated Anomalies	n	%
Hydrocephalus	73	64.6
Acyanotic heart disease	24	21.2
Adenoid hypertrophy	2	1.8
Aqueductal stenosis	1	0.9
Arnold chiari mal. II	60	53.1
Cleft palate	4	3.5
Colpocephaly	1	0.9
CTEV	12	10.6
Distematomyelia	3	2.7
Syrinx	8	7.1
Tethered cord	8	7.1
Tonsillar herniation	3	2.7

LSCS was the most common mode of delivery (n=68.1%, n=77), while 7.1% (n=8) patients were born by episiotomy. Normal vaginal delivery recorded in 24.8% (n=28) patients. Table 7

Table 7:

Mode of delivery	n	%
Episiotomy	8	7.1
LSCS	77	68.1
NVD	28	24.8
Total	113	100

Periconceptional folic acid intake was absent in 92% (n=104) patients (Table 8) & all 113 patients were from low socio-economic status.

Table 8:

Folic acid intake in periconception period	n	%
No	104	92.0
Yes	9	8.0
Total	113	100

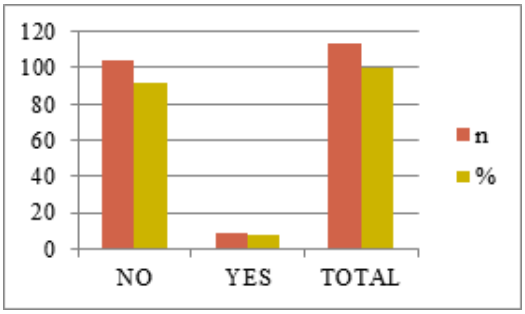

Fig. 2:

Mean of total no. of births in a month in our hospital is 270. Mean of Neural tube defect found in unbooked patients is 9.7 while mean of Neural tube defect found in booked patients in a month is 1.2.

Most commonly performed operation was excision & repair of Meningomyelocele, (51.3%, n=58) followed by VP (26.5 %, n=30) while 13.3% (n=15) patients required both excision & repair as well as VP shunting. 8.8% (n=10) patients not operated due to various reasons. (Table 9)

Table 9:

Operation	N	%
Excision of MMC	58	51.3
VP shunting	30	26.5
Both	15	13.3
Not operated	10	8.8
Total	113	100

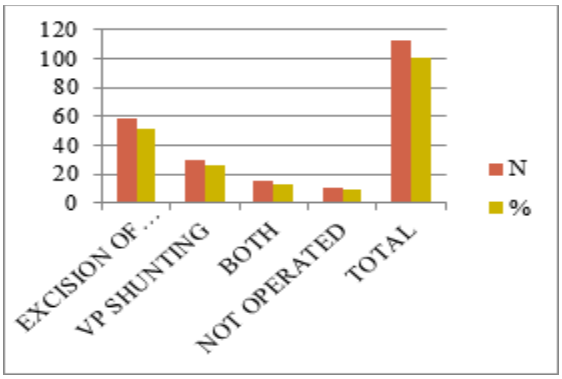

Fig. 3:

In 89.4 % cases (n=101) defect closed by primary closure while 10.6% (n=12) required Modified rombhoid repair.

Surgical site infection was the most common complication (5%, n=22) followed by blocked Ventriculoperitoneal (VP) shunt (16.8%, n=19). (Table 10)

Table 10:

Complications	n	%
Surgical site infection	22	19.5
Blocked VP shunt	19	16.8
CSF leak	9	8.0

In our study (91.2 %, n=103) patients improved & discharged, 2.7% (n=3) patients expired, 4.4% (n=5) patients left against medical advice while 1.8% (n=2) patients referred to higher centre due to various reasons. (Table 11)

Table 11:

Outcome	n	%
Expired	3	2.7
Improved and discharged	103	91.2
Lama	5	4.4
Referred to higher center	2	1.8
Total	113	100

Discussion

Myelomeningocele (MMC), one of the most common congenital malformation of the central nervous system, has been described as "the most complex treatable congenital anomaly compatible with life."[7] Failure of neural tube formation and closure may occur anywhere along the neural axis. MMC occurs in 0.8-1 per 1000 live birth and incidence varies with environmental and genetic factors.[7-9]

In our series most patients (70.8%, n=80) presented to us before the age of 6 months (mean=9.1 months) & we found an inverse relation between the age & the incidence of MMC (Table 1). In our series the M:F ratio is almost equal i.e 1.1:1 while in a other study by Chand MB et al[10] M:F ratio is 3:2. In our series most common site of MMC is Lumbosacral (73.5%, n=83) & in a series of 190 patients, Mirzai et al[11] also reported Lumbosacral region as a most common site for MMC. In comparision to other site Cervical MMC seems to have a better long-term neurological prognosis than low spinal MMC.[12,13] Most common associated anomaly was Hydrocephalus (64.6%, n=73) followed by Arnold chiari malformation II (53.1%, n=60) (Table 6). Chand MB et al[10] reported association of hydrocephalus in 67.56% cases. The cause of hydrocephalus is related to Arnold chiari malformation. Caudal displacement of 4th ventricle with compression and thinning of upper medulla and cerebellum through foramen magnum into upper cervical spinal cavity, this defect permits CSF to exit caudally displaced 4th ventricle but prevents its ascension into cerebral cavity to be absorbed because the CSF flow is impeded by herniated tonsils of cerebellum. Studies related to MMC suggests that 37% MMC are associated with congenital heart disease however in our study 21.2 % (n=24) patients were found to have congenital heart disease.[14] Out of 113 patients of our series 68.1% (n=77) patients were born by LSCS. (Table 7). As it is the well known fact that maternal folic acid intake prevents development of neural tube defect. In our series mother of 92% (n=104) patients were not supplemented by folic acid. Mean of total no. of births in a month in our hospital is 270. Mean of neural tube defect found in unbooked patients is 9.7 while mean of neural tube defect found in booked patients in a month is 1.2 it clearly states that antenatal screening and folic acid supplementation is the crucial risk factor in the development of MMC. In developed countries incidence in decreasing thanks to antenatal screening procedures, as well as dietary supplementation with folic acid to the women at risk prior to and during pregnancy. The prevalence of myelomeningocele has declined in developed countries of the world owing to both prenatal folate supplementation and to pregnancy termination

following prenatal diagnosis. In United States before 1980 prevalence of myelomeningocele was 1-2/1000 live births, but more recently prevalence has declined to 0.44 per 1000 live births.[15-18] Unfortunately, in third world countries prevalence is much higher, and acceptable prevalence data not available, nor has the issue been addressed with the goal of eradication, or reduction of incidence. The cause of MMC is multifactorial and includes genetic predisposition, nutritional deficiencies, particularly folate and zinc, use of anti-epileptic drugs like carbamazepine or valproic acid, diabetes mellitus (type-1), pre-pregnancy obesity and other non medical factors such as agricultural pesticides, radiation, hyperthermia and use of tobacco or drugs. Most commonly performed operation was excision & repair of MMC, (51.3%, n=58) followed by VP shunting (26.5 %, n=30). While 13.3% (n=15) patients required both excision & repair as well as VP shunting. 8.8% (n=10) patients not operated due to various reasons. (Table 9, Fig. 7).these patients (n=10) were not operated because, either patient referred to higher centre or left against medical advice due to various reasons including social stigma, financial issues, deficiencies related to manpower required for expertise in neonatal anesthesia & paediatric nursing care, deficiencies related to infrastructure like unavailability of paediatric ventilator, unavailability of neonatal warmer etc. In 69.9% (n=79) patients defect was large ($5X5cm^2$). In 89.4% (n=101) patients defect was closed by primary repair while 10.6% (n=12) patients required modified rombhoid repair. Closure of a large MMC defect is challenging for the reconstructive surgeon. The goals of closure are to preserve the function of the neural tissue and to prevent secondary infection.[19] If the MMC defect is small, direct repair can be performed; however, if the defect is large, various other reconstructive options are available[20] including modified rombhoid repair.

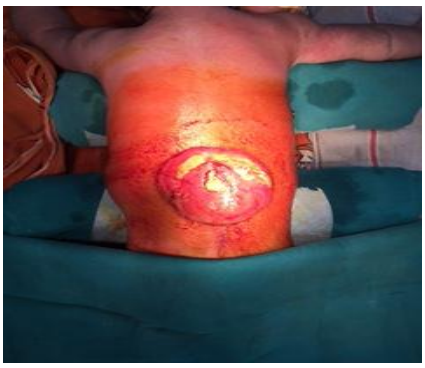

Fig. 4: Pre-op

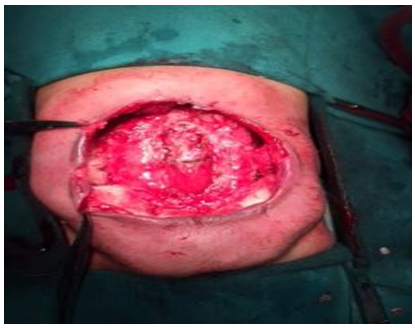

Fig. 5: Intra-op

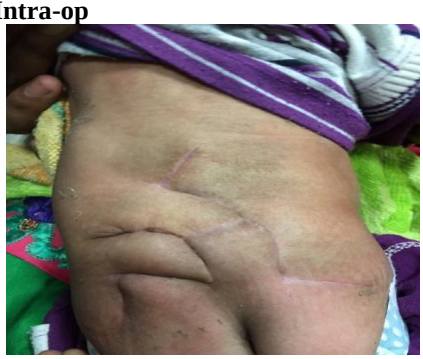

Fig. 6: Post-op

Modified Rhomboid Repair: Surgical site infection was the most common complication (5%, n=22) followed by Blocked VP shunt. CSF leak was seen in 8% (n=9) patients (Table 10, Fig. 8). Hydrocephalus affects neurocognitive outcome and result in morbidity and mortality caused by shunt malfunction and infection.[21,22] Significant Hydrocephalus may complicate the management of airway, reduced response to hypoxia and hence susceptible to post-operative apnoeic episodes.[23] In our study 91.2% (n=103) patients improved & discharged death reported in 2.7% (n=3) patients. (Table 11, Fig. 9) Over all results in our series were encouraging.

Conclusion

There is need of strong recommendation of antenatal screening of the mother specially in low socioeconomic area, as low socioeconomic status and lack of folic acid supplementation are the important risk factors for the development of MMC. Timely intervention with operative repair are encouraging. In our opinion simple primary closure should be the treatment of choice for small defects and modified rhomboid flap can be use to cover large defect.

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Conflict of Interest: None declared
Ethical Approval: The study was approved by the institutional ethics committee.

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