

Folic acid deficiency in conceptual period is still major cause of Meningomyelocele in rural area of western UP

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Abstract

Introduction: Meningomyelocele is congenital defect in backbone of children and most common neural tube closure defect and malformations affecting brain and spine of children. It has high rate of morbidity and disability in survived children. This is the second most common cause for morbidity and mortality in children. Global prevalence of Meningomyelocele has been reported to be 0.8–1 per 1,000 live births. The Hydrocephalus may affect as many as 80% of children with Meningomyelocele. Use of periconceptual folic acid supplements prevents ~50–75% of cases of neural tube defects. This study aims to evaluate the clinical profile and use of folic acid in the periconceptual period by mother in rural area of western U.P., and outcome of children with Meningomyelocele.

Methods and Materials: 226 patients with Meningomyelocele admitted to SVBP hospital, Meerut and private in private hospitals of near by area between August 2015 to Dec '18, were analyzed prospectively. The data regarding clinical profile with associated congenital anomalies and supplementation of folic acid by mother in conceptual period and post surgical disabilities were obtained by questionnaire-interview with the parents, drug history pertaining to drugs causing folic acid deficiency was unavailable as most of the patients belonged to low socio economic strata and were less literate. 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 (63.71%, n=144) associated anomaly. In 92% cases there is no history of folic acid supplementation & all patients were from low socioeconomic status. Excision & repair of Meningomyelocele is the most commonly (80% n=180) performed surgery, dural patch was used to close the larger defect in 8.35% (n=15) and 20% patients were not operated due to various regions. In our study out of 180 operated patients 80% (n=144) patients improved & discharged, and 6.6% (n=12) patients expired, 13.35% (n=24) patients left against medical advice (LAMA).

Conclusion: Low socioeconomic status & no maternal supplementation of folic acid in conceptual period are the important risk factors for the development of Meningomyelocele. Timely intervention with excision and repair gives good result. The use of Dural patch to close the neural tube dural defect was good alternative, modified z-plasty can be used to cover large defects.

Keywords: Meningomyelocele, Hydrocephalus, Folic acid supplementation, Dural patch.

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, which may be open and occulta type. 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 due to dietary and folic acid deficiency. 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 and poor dietary intake before getting pregnant and in early pregnancy increases the risk of having a foetus affected by NTDs, including Meningomyelocele.⁶ The use of periconceptual 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 devastating obstacle not only for patients but also for the patient's

family. This study aims to evaluate clinical profile and use of folic acid in periconceptual period by mother in rural area of western U.P and associated anomalies & outcome of children with Meningomyelocele.

Material and Methods

226 patients with Meningomyelocele admitted to SVBP hospital, Meerut in the department of surgery & department of paediatric medicine and operated in private hospital between August '15 to Dec-18, were analysed prospectively. The data regarding clinical profile with associated congenital anomalies and supplementation of folic acid by mother in conceptual period and post surgical disabilities were obtained by questionnaire-interview with the parents and patients themselves, drug history pertaining to drugs causing folic acid deficiency could not be available as most of the patients belonged to low socio economic strata and were less literate. Follow up was done to note for complications. MRI was the essential investigation in all patients. The Dural patch, artificial dural substitute was used in some large dural defects and having poor dural membrane. The z-plasty and s-shaped flap rotation and modified rhomboid flap technique were used to cover the defect. The data was analysed by SPSS 21.0 version.

Results

In our series of 226 patients most patients (70.8%, n=80) presented at the age of less than 6 month. (Mean =9.0months.) One patient presented at the age of > 6 year (table: 1)

Table1: The sex ratio is M:F 1.1:1(table: 2)

Age group	n	%
0-6 months	160	70.8
6-12 months	38	16.8
1-5 year	26	11.5
>5 year	2	0.9
Total	226	100

Table 2: Most of the patients (51.3%, n=116) came from villages near by to Meerut district region. (table : 3)

Sex	n	%
Male	116	51.3
Female	110	48.7
Total	226	100

Table 3: Most commonly involved site was Lumbosacral region (n=166, 73.5%). while occipital region was the least commonly (2.7%, n=6) involved. (table: 4)

Region	N	%
Meerut	116	51.3
Out side the meerut	110	48.7
Total	226	100

Table 4: In most patients (57.52%, n=130) defect was large i.e > 5 X 5 cm². (table: 5)

Site	N	%
Cervical	16	7.1
Dorsal	18	8.0
Dorsolumbar	4	1.8
Lumbosacral	166	73.5
Occipital	6	2.7
Sacral	16	7.1
Total	226	100

Table 5: Most common associated anomaly was Hydrocephalus (63.71%, n=144) followed by Arnold chiari malformation II (20.35%, n=46). (table: 6).

Size in cm ²	n	%
Large (> 5 x 5 cm ²)	130	57.52
Small (< 5 x 5 cm ²)	96	42.48
Total	226	100

Table 6: LSCS was the most common mode of delivery (n=69.9%, n=158), while 8.8%(n=20) patients were born by episiotomy. Normal vaginal delivery recorded in 21.23% (n=48) patients (table: 7)

Associated anomalies	N	%
Hydrocephalus	144	63.71
Acyanotic heart disease	14	6.19
Adenoid hypertrophy	2	0.88

Aqueductal stenosis	1	0.44
Arnold chiari mal. Ii	46	20.35
Cleft palate	2	0.88
Ctev	8	3.5
Distematomyelia	3	1.3
Syrinx	3	1.3
Tonsillar herniation	3	1.3

Table 7: Periconceptional folic acid intake was absent in 92% (n=208) patients (table:8) & all 226 patients were from low socio-economic status and from rural area of western U.P. table no.8

Mode of delivery	n	%
Episiotomy	20	8.8
Lscs	158	69.9
Nvd	48	21.23
Total	226	100

Table 8: Mean of total no. of births in a month in SVBP Hospital is 270. Mean of Neural tube defect found in unbooked patients means was not taking antenatal care is 9.7 while mean of Neural tube defect found in booked patients in a month is 1.2.

Folic acid intake in periconception period	N	%
No	208	92.0
Yes	18	8.0
Total	226	100

Most commonly performed operation was excision & repair of Meningomyelocele, and patients developed hydrocephalous are managed by Ventriculo-peritoneal shunt and some patients developed flap necrosis which was managed secondary closure of defect by modified rhomboid rotation of flap. Operation excision and repair was done in 80% (n=181). Patients who developed hydrocephalous in initial follow period were by V P shunt surgery. About 20% patients could not be operated due to various regions (n=46). (table: 9)

Table 9: Surgical site infection was the most common complication 30%(n=54) followed by CSF leak and flap necrosis. They were managed by appropriate antibiotic, acetazolamide and dressings and Ventriculoperitoneal shunt.

Operation	N	%
Excision of mmc	180	80
Not operated	46	20
Total	226	100

In our study out of 180 operated patients 80%, (n=144) patients improved & discharged, and 6.6% (n=12) patients expired, 13.35% (n=24) patients left against medical advice (LAMA). (table:10).

Table 10:

Out come	No.	Percentage
Improved and discharged	N=144	80%
Expired	N=12	6.6%
Lama	N=24	13.4%
Total operated	N=180	100%

**Fig. 1: showing lumbar meningomyelocele****Fig. 2: MRI of lumbar meningocele.**

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 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=160) 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 al¹⁰ M:F ratio is 3:2. In our series most common site of MMC is Lumbosacral () & in a series of 190 patients, Mirzai et al¹¹ also reported Lumbosacral region as a most common site for

MMC. In comparison 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 (63.71% n=144) followed by Arnold chiari malformation II (20.35%,n=46)(table:6). Chand MB et al¹⁰ 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=48) patients were found to have congenital heart disease.¹⁴ Out of 226 patients of our series 69.9% (n=158) 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=208) 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 this study drug history was unavailable because most of the attendants belonged to low socio economic status and were less literate. 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, (80%, n=180). 20%(n=46) patients not operated due to various reasons.(table:9). 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 57.52%(n=130) patients defect was

large (5X5cm²). In 180 total operated patients, 80%(n=144) patients defect was closed by primary repair while 20%(n=36) patients required Z-plasty and rotation of flap and modified rhomboid technique. Dural patch were use to close the dural defects in 15 patients, which is good alternative to dural membrane. 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 rhomboid flap rotation. Surgical site infection was the most common complication followed by VP shunt malfunctioning' CSF leak was seen in many patients. 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 out of 180 operated patients 80% (n=144) patients improved & discharged, and 6.6%(n=12) patients expired, 13.35%(n=24) patients left against medical advice. Overall results in our series were encouraging.

Conclusion

The folic acid and dietary deficiency in low socioeconomic status is the major cause and risk factor for meningocele 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.

Funding: Self.

Conflict of interest: None.

Ethical approval: The study was approved by the institutional ethics committee.

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How to cite this article: Sharma S, Sharma N, Bharti P, Folic acid deficiency in conceptual period is still major cause of Meningocele in rural area of western UP. *J Neurosci* 2019;5(1):28-31