

How to Cite:

Sanasiya, K., Joshi, D., & Solanki, M. (2022). Surgical outcome of myelomeningocele repair associated with Chiari type II malformation. *International Journal of Health Sciences*, 6(S6), 8347–8352. <https://doi.org/10.53730/ijhs.v6nS6.12343>

Surgical outcome of myelomeningocele repair associated with Chiari type II malformation

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Abstract--Introduction: Meningomyelocele, more commonly known as spina bifida (open) is a congenital malformation of the central nervous system and is associated with significant morbidity. Myelomeningocele is a commonly seen congenital birth defect in children. The most common associations with myelomeningocele are hydrocephalus and ACM Type 2. Folic acid deficiency and parental consanguinity appear to play a major part in this type of congenital defects. Objectives: To analyse the clinical profile and outcome of surgery in cases of myelomeningocele associated with Arnold Chiari malformation II and hydrocephalus. Methods: A Retrospective Cohort study was done starting from January 2015 to January 2019 in the Department of Neurosurgery at SBKS Medical College. Total 18 patients with Myelomeningocele associated with Arnold Chiari malformation II and Hydrocephalus were analysed and evaluated. Follow up of cases was done for a minimum period of 6 months. Results: Out of 18 total cases, the majority of cases, i.e., 76.8%, had myelomeningocele with Chiari II malformation and hydrocephalus. Ratio of females to males was 1.14. Prenatal folic acid supplementation was missed amongst 80% of the cases. 16.6% cases had positive parental consanguinity. Mortality was seen in 2 cases due to post operative complications. Conclusion: Meningomyelocele is commonly associated with both, hydrocephalus and Arnold chiari type II malformation. Prenatal folic acid supplementation and avoiding parental consanguinity can prevent these malformations to a major extent.

Keywords---Myelomeningocele (MMC), Arnold Chiari malformation II (ACM II), hydrocephalus (HCP), folic acid.

Introduction

Myelomeningocele is the most common disorder of a developmental process called, neurulation, that results in viable infants. Its incidence in the United States is about 0.2 to 0.4 per 1000 live births. However, its incidence is up to 20-times higher in some regions of China.[1] Females are affected up to 3 to 7 times more than males. The incidence is higher amongst people belonging to lower socioeconomic status. The incidence is also high in increased maternal age. The recurrence rate in subsequent pregnancies is about 2% to 3%.[6]. Spina bifida can also be associated with Arnold Chiari type II malformation which is characterized by downward displacement of the cerebellar tonsils and medulla. This malformation leads to obstruction of the CSF flow through the posterior fossa leading to hydrocephalus.[10]

Congenital anomalies of the central nervous system are very common and constitute about 50% of all congenital anomalies. Incidence is 1-2/1000 live births. Amongst CNS malformations, myelomeningocele is the commonest spinal dysraphism in neonates. These are often associated with Arnold Chiari type II malformations, hydrocephalus or both. In this study we will focus to review and study the literature of such conditions with respect to their incidence, sex predilection, antenatal folic acid supplementation, consanguineous marriages, their management protocols and outcome in a period of 2 years in an educational institute.

Aims And Objectives

To analyse and study the clinical profile of patients with myelomeningocele associated with chiari II malformation and hydrocephalus. Various factors like perinatal folic acid deficiency, consanguinity, etc are looked for. To analyse the outcome of surgery, post operative complications, and neurological deficit.

Materials And Methods

A Retrospective Cohort study was done from January 2015- January 2019 in Department of Neurosurgery at SBKS Medical College. Total 18 patients with Myelomeningocele associated with Arnold Chiari malformation type II and Hydrocephalus were analysed and evaluated. Follow up of cases was done for a minimum period of 6 months.

Inclusion And Exclusion Criteria

- Patients with ruptured or unruptured, meningomyelocele, associated with chiari type II malformation, with or without hydrocephalus admitted in Dhiraj hospital from January 2015- January 2019 were included in the study.

- Patients with congenital malformations other than Chiari malformation II associated with myelomeningocele are excluded from the study.
- Patients not fit for surgery or patients who died preoperative were not included in the study

Results

A total of 18 patients with myelomeningocele were eligible and enrolled in this study. Out of which 14 patients with myelomeningocele had associated Chiari malformation type II as well as hydrocephalus. Two patients had only hydrocephalus. Two patients had only Chiari malformation type II. All the patients were operated at Dhiraj general hospital under Neurosurgery department attached to SBKS & Medical College. Myelomeningocele repair was done by neurosurgery team in all of the cases. In some large myelomeningocele, closure and skin flap repair was done by the plastic surgery team. In all of the cases with hydrocephalus, Ventriculo-Peritoneal & shunt was done using medium pressure Chhabra shunt. T-seal thrombin glue was used over dural closure to prevent post operative CSF leak in all cases. A subcutaneous mini-vac drain was kept in all cases and was removed between post operative day five to post operative day ten. All the patients were managed by a team of paediatricians and neurosurgeons.

Table 1

Associated malformations	Number of patients (in %)
ACM II and HCP with MMC	12
Only HCP with MMC	4
Only ACM II with MMC	2

Out of 18 patients 10 were female and 8 were male with a ratio of male : female 1: 1.25

Table 2

Folic acid supplementation	Number of patients (%)
Missed supplementation	12
Received	6

8 patients had a positive history of parental consanguinity out of 18.

Table 3

Parental consanguinity	Number of patients (in %)
Consanguineous	8
Non consanguinous	10

Table 4

Post operative complications	Number of patients (in %)
Wound infection	6
CSF leak	8

Worsening of neurodeficit	4
Meningitis	4
Mortality	2

Excision of myelomeningocele and repair with V-P shunt is done in all cases with hydrocephalus. Foramen magnum decompression was done in all cases of Chiari type II malformation. Follow up of all cases is done for a minimum period of 6 months upto 4 years. Two out of 18 patients died due to post operative sepsis.

Discussion

Myelomeningocele is a developmental defect that manifests as failure of vertebral arches of spine to fuse completely which results in dysplastic growth of spinal cord and its meningeal coverings [2,3]. When this occurs, CSF may leak around this defect and spinal cord is exposed making it susceptible for infection. Tulp[4] in 1641 introduced the term “spina bifida” for congenital malformation which is associated with a cystic protrusion from vertebral column. Morgagni[5] recognized important association of spina bifida and hydrocephalus. Myelomeningocele is well known to be associated with Arnold Chiari Malformation II & Hydrocephalus. Hydrocephalus is present in 90% of children who are affected with myelomeningocele. Hydrocephalus may not be a presenting feature at birth and signs may not develop until 2-3weeks following defect closure. In our study, the incidence rate of myelomeningocele occurring with ACM II and hydrocephalus is 66.66%. Hydrocephalus is noted due to presence of Arnold Chiari malformation.

There is caudal displacement of 4th ventricle which causes compression and thinning of upper medulla and cerebellum, which herniates through foramen magnum into upper cervical spinal cavity. This defect allows CSF to leak caudally but prevents its ascension into cerebral cavity as the CSF flow is impeded by herniated tonsils of cerebellum. A study conducted by Mary J Seller 1987, on neural tube defects has shown an overall ratio of M:F 0.73 with female excess [6]. Our study is of similar conclusion. There is no statistical evidence of gender bias amongst case of meningomyelocele associated with Arnold Chiari malformation II and/or Hydrocephalus. British medical research council in 1991 concluded that 4000microgm of dietary folate prevented 72% of recurrences of neural tube defects. Approximately 75% of spina bifida are folic acid preventable.

A congenital anomalies’ study reported that consanguineous marriages are a major risk factor for neural tube defects accounting for 44.74% cases. In our study 66.6% of patients are in consanguineous marriage. According to a case control study by Nuzhat Nauman et al (2016), 60% of couples were in consanguineous marriage, with a neural tube defect pregnancy as compared to 45% in controls. 30% of these Neural tube defects included meningomyelocele [7]. This study concluded the need for genetic counselling for the couples, educating them on risks about consanguineous marriages and their reproductive outcomes. Another study by B Shanthikumari et al, 2014 on neural tube defects: epidemiologic and demographic implication concluded consanguinity as one of the etiological factors for neural tube defects [8]. The management of

meningomyelocele with hydrocephalus may include Cerebrospinal fluid (CSF) diversion for the remainder of their lives.

Blockage of the outlets of the fourth ventricle and communication of the fourth ventricle with the central canal provides a mechanism for compensation [9]. The signs and symptoms of CSF diversion malfunction, either shunt or third ventriculostomy, can be quite subtle. A symptomatic Arnold chiari malformation II is the most common cause of death in patients with myelomeningocele who are younger than 2 years of age. The first treatment option to be considered should be cerebrospinal shunt revision because the most common symptom in this population is hydrocephalus or a failed shunt [10]. If this fails, surgical decompression with removal of the lamina of the first and second cervical vertebrae may be required.

Conclusion

Counselling the parents on risks about the adverse outcomes of consanguineous marriage, preconceptional and antenatal folic acid supplementation contributes a major role in making these malformations as preventable. Surgical intervention helps in preventing further deterioration in functional ability though there may not be significant improvement in motor and sensory functions. Early diagnosis, surgical management and rehabilitation can prevent further neurological damage and can improve quality of life in these patients.

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