

## Myelomeningocele Associated with Arnold Chiari Malformation II & Hydrocephalus-An Institute Experience

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### Abstract:

**Introduction:** Myelomeningocele is a form of spina bifida and may affect as 1 out of 4000 infants. Hydrocephalus and Arnold chiari malformation II are the most common associations with myelomeningocele. Low levels of folic acid in a woman body before and during early pregnancy and parental consanguinity appear to play a major part in this type of congenital defects.

**Objectives:** Analyzing the incidence of myelomeningocele associated with Arnold chiari malformation II and hydrocephalus in children born out of consanguineous marriages and in children born to a mother who missed prenatal folic acid supplementation and to analyze sex predilection in children with myelomeningocele associated with Arnold chiari malformation II and hydrocephalus, retrospectively from February 2015-March 2017 in Government Medical College and Hospital, Guntur.

**Methods:** 30 patients with Myelomeningocele associated with Arnold Chiari malformation II and (or) Hydrocephalus were analysed and evaluated. Follow up of cases was done for a minimum period of 6 months.

**Results:** Out of 30 cases majority (66.6%) have myelomeningocele with both Arnold chiari II malformation and hydrocephalus. Ratio of females was 1.14 in excess than males. Prenatal folic acid supplementation was missed amongst 80% of the cases. 66.6% have positive parental consanguinity. 2 patients died due to preoperative complications.

**Conclusion:** Meningomyelocele is commonly associated with both hydrocephalus and Arnold chiari II malformation. Prenatal folic acid supplementation and counselling parents about adverse effects of parental consanguinity can prevent these malformations to a major extent.

**Keywords:** Myelomeningocele (MMC), Arnold chiari malformation II (ACM II), hydrocephalus (HCP), folic acid, consanguinity.

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### I. Introduction:

Congenital anomalies among central nervous system are very common and constitute to about 50% of all congenital anomalies and the incidence is 1-2/1000 live births and among CNS malformations myelomeningocele is the commonest spinal dysraphism in neonates<sup>1</sup> and they are often associated with Arnold chiari II malformations, hydrocephalus or both. In this study we focus to review the literature of such conditions where Myelomeningocele is associated with Arnold chiari malformation II and hydrocephalus; 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.

### II. Aims And Objectives

Analyzing the incidence of myelomeningocele associated with Arnold chiari malformation II and hydrocephalus in children born out of consanguineous marriages. Analyzing the incidence of myelomeningocele associated with Arnold chiari malformation II and hydrocephalus in children born to a mother who missed prenatal folic acid supplementation. Analyzing Sex predilection in children with myelomeningocele associated with Arnold chiari malformation II and hydrocephalus retrospectively from February 2015-March 2017 in Guntur Medical college and Hospital, Guntur.

### III. Materials And Methods

Data of all patients with myelomeningocele with Arnold chiari malformation II or hydrocephalus or both were retrospectively reviewed at government medical college and hospital, Guntur, AP. Data was collected by individual interviews after taking an informed written consent from the patients.

Factors analyzed were antenatal medical history, sex predilection, consanguinity, management and follow up for a minimum of 6months.

#### IV. Inclusion And Exclusion Criteria

Patients with myelomeningocele ruptured or unruptured, associated with Arnold chiari malformation II and or with hydrocephalus admitted to the hospital from February 2015 to march 2017 are included in the study. Patients with malformations other than Arnold chiari malformation II or Hydrocephalus associated with myelomeningocele are excluded from the study.

#### V. Results

A total of 30 patients with myelomeningocele were eligible and enrolled. Out of which 20 patients with myelomeningocele had associated Arnold chiari malformation II and hydrocephalus. 8 patients had only hydrocephalus. 2 patients had only Arnold chiari malformation II .

**Table 1:**

Associated malformation	No. of patients	Percentage
ACM II& HCP with MMC	20	66.6%
Only HCP with MMC	8	26.6%
Only ACM II with MMC	2	6.6%

Out of 30 patients 16 were female and 14 were male with a ratio of 1.14 female excess.

**Table 2:**

Malformation	Male	Female
ACM II + HCP + MMC	10	10
HCP + MMC	3	5
ACM II + MMC	1	1

Out of 30 patients mothers of 24 patients missed folic acid supplementation. 6 received supplementation periconceptionally.

**Table 3:**

Folic acid supplementation	No. of patients	Percentage
Missed supplementation	24	80%
Received	6	20%

20 patients had a positive history of parental consanguinity out of 30.

**Table 4:**

Parental Consanguinity	No. of pts	Percentage
Consanguineous marriage	20	66.6%
Non consanguineous marriage	10	33.3%

Excision of myelomeningocele and repair with V-P shunt is done in all cases with myelomeningocele with hydrocephalus. Myelomeningocele excision and repair with or without foramen magnum decompression is done in all cases with myelomeningocele with Arnold chiari malformation II. Follow up of all cases is done for a minimum period of 6 months. Neither sensorimotor improvement nor progression of sensorimotor loss was observed in all the cases.

2 out of 30 patients died due to preoperative sepsis.

#### VI. Discussion

Myelomeningocele is a defect in development that manifests as failure of vertebral arches of spine to fuse completely which results in dysplastic growth of spinal cord and its meningeal coverings<sup>2,3</sup>. When this occurs, CSF may leak around this defect and spinal cord is exposed making it susceptible for infection. Myelomeningocele has been recognized for several centuries, however effective treatment and increase in survival rate have only been achieved during last four decades. Tulp<sup>4</sup> in 1641 introduced term spina bifida to congenital malformation which is associated with a cystic protrusion from vertebral column. Morgagni<sup>5</sup> recognized important association of spina bifida and hydrocephalus. Earlier in 20<sup>th</sup> century surgeries to close the defects were performed immediately after birth, but infection around the closure site and meningitis caused mortality in perinatal period.

Hydrocephalus occurs in 90% of children affected with myelomeningocele. Hydrocephalus may not be present at birth and signs may not develop until 2-3 weeks following closure of defect.

Incidence of myelomeningocele with Arnold chiari II malformation and hydrocephalus is 66.66% in our study. The cause of hydrocephalus is related to Arnold chiari malformation. Caudal displacement of 4<sup>th</sup> 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 4<sup>th</sup> ventricle but prevents its ascension into cerebral cavity to be absorbed because the CSF flow is impeded by herniated tonsils of cerebellum. A study conducted by Mary J Saller 1987, on neural tube defects has shown an overall ratio of M:F 0.73 with female excess<sup>6</sup>. Our study has shown a similar result of female excess. There is no statistical evidence of gender predominance amongst case of meningomyelocele associated with Arnold chiari malformation II and or Hydrocephalus. Hema gupta et al., showed that maternal folate status is associated with increased incidence of neural tube defects. In 1980 Smithellser al., showed that blood levels of several micro nutrients are reduced in women with NTD child and has concluded that periconceptional intake of multivitamins containing folic acid reduces risk of having Neural tube defects in previously effected pregnancy. British medical research council 1991 concluded that 4000 microgm of dietary folate prevented 72% of recurrences of neural tube defects. Approximately 75% of spina bifida are folic acid preventable.

A study on congenital anomalies reported consanguineous marriages (44.74%) as major risk factor for neural tube defects. In our study 66.6% of patients has parental consanguinity as a positive history. According to a case control study by Nuzhat Nauman et al (2016) 60% of couples were consanguineous with a neural tube defect pregnancy as compared to 45% in controls. 30% of these Neural tube defects included meningomyelocele<sup>7</sup>. This study concluded the need for genetic counselling to the couples on risks about consanguineous marriages and their adverse 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<sup>8</sup>. 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<sup>9</sup>. 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<sup>10</sup>. 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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\*Dr. Duttaluru Seshadri Sekhar. "Myelomeningocele Associated with Arnold Chiari Malformation II & Hydrocephalus-An Institute Experience." IOSR Journal of Dental and Medical Sciences (IOSR-JDMS) 16.8 (2017): 74-76.