



CONGENITAL MALFORMATION OF THE CENTRAL NERVOUS SYSTEM : INCIDENCE, MANAGEMENT AND OUTCOME

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ABSTRACT

INTRODUCTION

MATERIAL AND METHODS This study has been conducted on the patients suffering from the congenital malformations of the central nervous system attending/admitted in the department of Surgery/ Pediatrics /Neurosurgery, Nehru hospital, B.R.D. Medical College, Gorakhpur for a period of 12 months.

CONCLUSION

KEYWORDS : an encephaly,encephalocoe ,hydrocephalus,maningocele,maningomylocele,spina bifida occulta, .

INTRODUCTION

Congenital anomalies (birth defects) can be defined as structural, functional, and/or biochemical-molecular defects developing during foetal life caused by genetic and/or environmental factors and discovered either prenatally or after birth at any age⁽¹⁾. As congenital anomalies impose a great burden not only on health professionals and government officials but also on the society, it is necessary for the government and health care providers to be aware of the exact data on congenital anomalies in the irregion⁽²⁾.

Congenital central nervous system (CNS) malformations are highly prevalent, affecting 1 to10:1,000 live newborns⁽³⁾, they may be isolated or appear as part of a genetic syndrome or a complex congenital malformation syndrome⁽⁴⁾.

CNS congenital abnormalities can be considered one of the main causes of infant morbidity and mortality and fetal death. CNS anomalies can be divided into developmental malformations and disruptions.

Malformations carry a recurrence risk that can be calculated and sometimes avoided in the future, for example in case of maternal metabolic pathology like maternal hyper phenil alaninemia, and maternal diabetes. Disruptions do not recur, unless the exposure recurs or continues. Exposure to known teratogens and viral infections, can occur throughout pregnancy. The timing of exposure is critical for both malformations and disruptions.

The earlier the exposure, the more severe the CNS defect. For instance, fetal cytomegalovirus (CMV) infection before midgestation can be responsible for microcephaly and polymicrogyria. CMV infection in the third trimester causes an encephalitis, similar to postnatal CMV encephalitis responsible for other diseases as deafness^(5,6). The most critical period for malformations and disruptions is the third to eighth week of gestation, during which the brain and most organs develop.

Classically, brain malformations are classified according to the morphological and structural criteria.

In the past few years there have been great advances in identifying genetic and epigenetic alterations for many isolated CNS malformations and syndromes with CNS malformations. Several genomic disorders caused by copy number variation (microdeletions and microduplications) of genes whose dosage is critical for the physiological function of the nervous system have been recently identified. With recent advances in the understanding of underlying molecular mechanisms involved in the development of the brain, attempts are being made to categorize these malformations according to the underlying genetic factors⁽⁷⁾.

AIMS AND OBJECTIVES

To determine the incidence and types of Congenital Nervous System malformations cases coming to Nehru Hospital the associated teaching hospital of BRDMC, Gorakhpur.

1. To analyse the incidence of congenital central nervous system malformation cases according to age, gender, area of residence and socioeconomic status.
2. To pin point the need of specific investigations to diagnose such cases.
3. To determine the management and outcome of foresaid cases.

MATERIAL AND METHODS

This study has been conducted on the patients suffering from the congenital malformations of the central nervous system attending/admitted in the department of Surgery/ Pediatrics /Neurosurgery, Nehru hospital, B.R.D. Medical College, Gorakhpur for a period of 12 months.

INCLUSION CRITERIA:-

- Patients presenting with
- congenital malformation of central nervous system

EXCLUSION CRITERIA:-

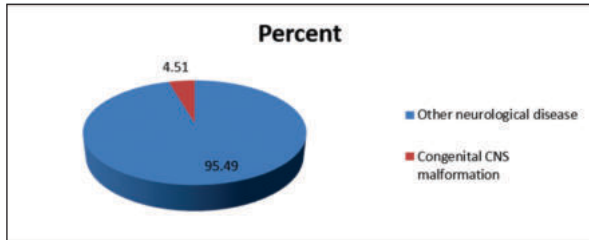
- Patients who were not giving consent

- patients (age >12 years)

OBSERVATIONS

Table-1 Congenital Cns Malformations Vs All Neurological Disease

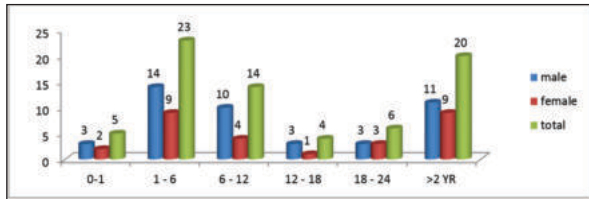
S.No	CASE	No. of cases	Percent
1	Congenital CNS malformation	72	4.51
2	other neurological disease	1524	95.49
	Total	1596	100.0



A total of 1596 patients were either attended or admitted in the neurosurgery department, 72 patients were having congenital malformation of central nervous system. i.e. 4.51%.

Table -2 Distribution Of Patients Congenital Cns Malformations According To Age And Sex

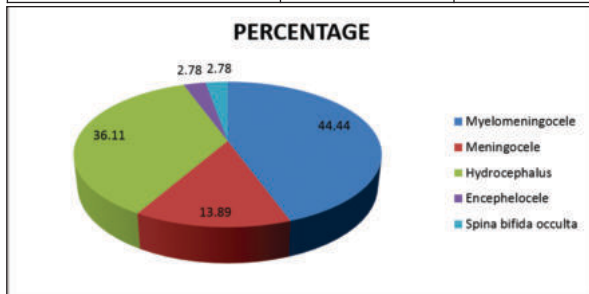
Age Grp (Month)	Male	Female	Total
0-1	3	2	5
1 - 6	14	9	23
6 - 12	10	4	14
12 - 18	3	1	4
18 - 24	3	3	6
>2 YR	11	9	20
total	44 (61.11%)	28(38.89%)	72 (100%)



In this study , 44male patients and 28 patients were females i.e. male:female ratio was 1:57:1. The maximum patients were brought to the hospital in the age group of 1-6 months (31.94%). The minimum cases 4 (5.56%) were in the age group of 12-18 months.

Table 3 Clinical Diagnosis

Disease	no of cases	Percentage
Myelomeningocele	32	44.44
Meningocele	10	13.89
Hydrocephalus	26	36.11
Encephelocele	2	2.78
Spina bifida occulta	2	2.78
Total	72	100%

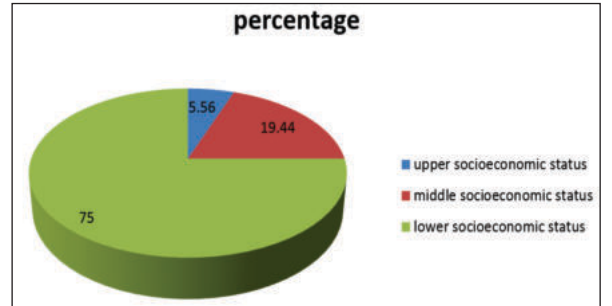


clinical diagnosis of myelomeningocele in 32 (44.44%) cases. it was followed by hydrocephalus, which was detected clinically in 26 (36.11%) patients. The minimum cases were detected of encephalocele and spina bifida occulta only in 2

(2.78%) and 2(2.78%). Meningocele as such was detected in 10(13.89%) of the total patients.

Table 4 Distribution of case on the basis of socio-economic status (Kuppuswamy scale)

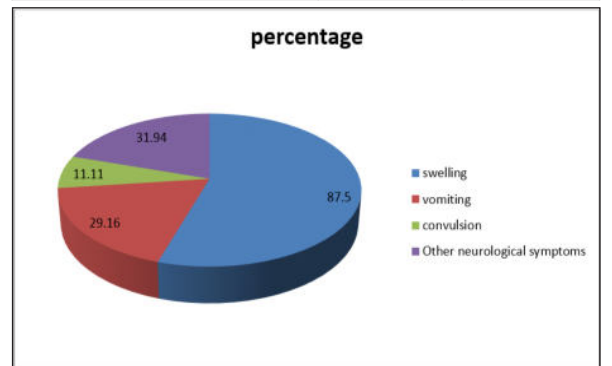
Socio economic status	No of cases	Percentage
upper socioeconomic status	4	5.56
middle socioeconomic status	14	19.44
lower socioeconomic status	54	75
Total	72	100%



Our observations shows that the congenital malformations were more prevalent in lower socio-economic status in 54(75%) patients. Only 4(5.56%) patients were belonged to upper socio-economic status. Rest of the 14(19.44%) cases was belonged to middle socio-economic status.

Table 5 Incidence Of Presenting Complaints

Complaints	No of Cases	Percentage
Swelling	63	87.5
Vomiting	21	29.16
Convulsion	8	11.11
Other Neurological Symptoms	23	31.94



Observation shows that maximum patients of congenital malformation of central nervous system presents with swelling in 63 (87.5%) patients either in the skull or spinal cord. 21 (29.16%) patients complained of vomiting. The least complaint was of convulsion, which was present in only 8(11.11%). Other neurological symptoms include weakness of the lower limb with or without the involvement of bladder and bowel, deviation of eyeball, poor development of the milestones mental retardation were present in 23 (31.94%) cases.

Table 6 Distribution Of Myelomeningocele

Myelomeningocele	No of case	Percentage
Cervical	1	3.13
Thoracic	2	6.25
Lumbar	15	46.87
Lumbo-Sacral	10	31.25
Sacral	4	12.5
total	32	100

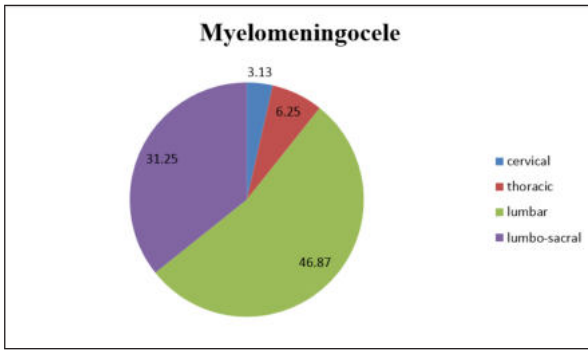


Figure 7 Myelomeningocele involves the various sites of vertebral column. The site of involvement of the vertebral column in myelomeningocele was confirmed by M. R.I. examination of vertebral column. In total 4 patients of myelomeningocele the diagnosis was confirmed by clinical examination only. Our observation shows that maximum 15(46.87%) cases were involved the lumbar area of the vertebral column and maximum cases of lumbar myelomeningocele were present between 3rd and 4th lumbar vertebra. Ten 10(31.25%) cases of myelomeningocele were present in the lumbo-sacral vertebrae. In pure sacral region only 4 (12.5%) cases of myelomeningocele were detected. Minimum cases 1 (3.13%) were found in cervical region. Rest of 2(6.25%) cases of myelomeningocele was detected in thoracic region. usually around the 10th thoracic vertebra.

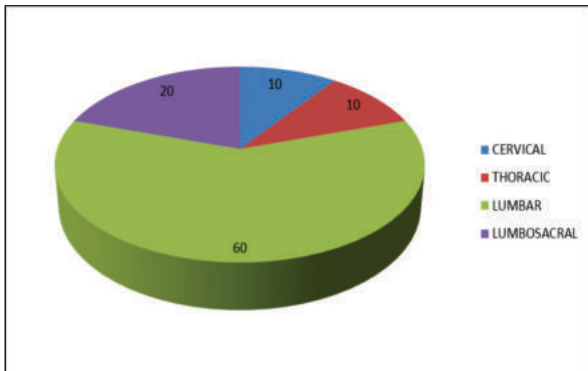
Table 7 Myelomeningocele Associated With Other Systemic Involvement

Congenital Malformation	Other Systemic Involvement			
	Anorectal Malformation (2)		Urogenital Anomalies (2)	
	Female	Male	Female	Male
Myelomeningocele (4)	AVF (anovestibular fistula) (1)	low ARM (1)	PUJ Obstruction (1)	Vesicoureteric Reflux (1)

Out of 32 Myelomeningocele patients, 4 patients associated with other systemic involvement, 2 patients associated with Anorectal Malformation on clinically diagnosed, 1 patient having AVF (Female) and 1 patient have low ARM (male) and 2 patients associated with Urogenital malformation on ultrasonography, 1 patient having PUJ Obstruction and 1 with Vesicoureteric Reflux.

Table 8 Distribution Of Meningocele

DISTRIBUTION OF MENINGOCELE		
SITE	No of Cases	percentage
CERVICAL	1	10
THORACIC	1	10
LUMBAR	6	60
LUMBOSACRAL	2	20
TOTAL	10	100



maximum cases of meningocele occurred in lumbo-sacral

region. We found 6 (60%) cases of meningocele in this area. The second commonest site of involvement in cases of meningocele was lumbar region and we found 2 (20%) cases of meningocele in lumbar area. Only 1 (10%) case was detected in thoracic region, and rest of the 1 (10%) cases of meningocele was detected in cervical region

Table 9 Distribution Of Encephalocele

SITE	Total No of Cases	Percentage
Occipital	2	100

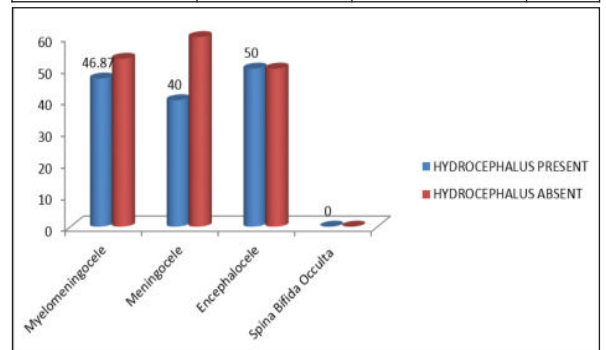
all cases of encephalocele were present in occipital region.

Table 10 Mri Finding

MRI FINDINGS		NO OF PATIENTS
Premature Disjunction	Lipomeningocele	0
	Lipomeningomyelocele	5
	Dorsal Dermal Sinus	0
	Teethering	12
	Sacral Dysgenesis	0
	Diastematomyelia	2
	Syringomyelia	6
Non Disjunction	Spina Bifida Occulta	0
	Meningocele	6
	Myelomeningocele	14

Table 11 Myelomeningocele, Meningocele, Encephalocele, Spina Bifida Occulta With And Without Hydrocephalus

CONGENITAL MALFORMATION	HYDROCEPHALUS PRESENT	HYDROCEPHALUS ABSENT	TOTAL NO
Myelomeningocele	15 (46.87%)	17 (53.12%)	32
Meningocele	4(40%)	6(60%)	10
Encephalocele	1(50%)	1(50%)	2
Spina Bifida Occulta	0	0	0
Total	20	24	44

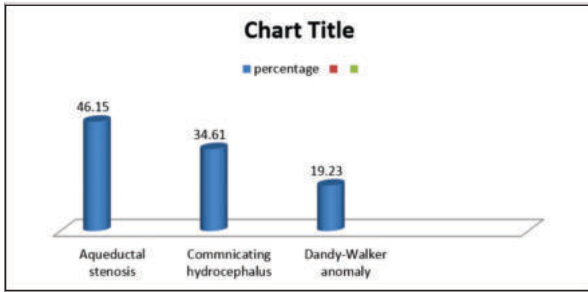


All the studied 32 patients were investigated by M.R.I for the presence of hydrocephalus and associated central nervous system anomalies as well to see the contents of the sac

Table 11 shows that the total 32 case of my elomeningocele diagnosed clinically shows the presence of hydrocephalus in association with meningomyelocele in 15 (46.87%) cases. In 10 cases of meningocele, only 4(40) cases show the presence of hydrocephalus after M.R.I. examination. In encephalocele out of 2 cases, 1(50%) cases shows the presence of hydrocephalus with encephalocele Spina Bifida Occulta not associated with hydrocephalus.

Table 13 Aetiological Incidence Of Hydrocephalus

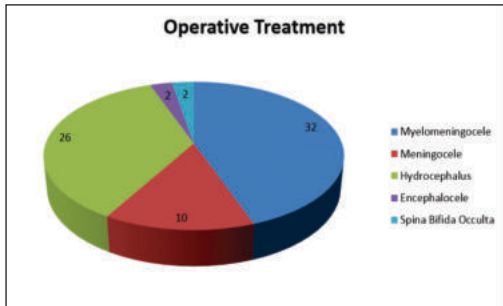
Anomaly	no of cases	percentage
Aqueductal stenosis	12	46.15
Commnicating hydrocephalus	9	34.61
Dandy-Walker anomaly	5	19.23
Total	26	100



All 26 cases hydrocephalus were investigated either by C.T scan or M.R.I. to confirm the presence of hydrocephalus or to find out the associated congenital anomalies. out of 12 cases Aqueductal stenosis was the commonest anomaly present with hydrocephalus., 12 (46.15%) cases were having aqueductal stenosis. The Dandy-walker anomaly was detected in the least, 9 (34.61%) cases of hydrocephalus. Rest of the 5 (19.23%) cases was found to have communicating type of the hydrocephalus

Table 14 Operative Treatment Given To All Patients

Type of CNS Malformation	Operative Treatment	Total
Myelomeningocele	24	32
Meningocele	10	10
Hydrocephalus	23	26
Encephalocele	2	2
Spina Bifida Occulta	2	2
Total	61	72

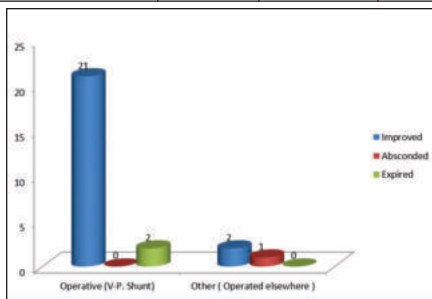


In 32cases of Myelomeningocele, 24 patients were operated for Myelomeningocele it self, hydrocephalus or for both. Maximum cases of Meningocele and hydrocephalus were given operative treatment.

10 case of Meningocele were given operative treatment out of 10 cases. .In cases of hydrocephalus 23 patients were given operative treatment .In case of Spina Bifida Occulta and Encephalocele 2 cases were operated.

Table 15 Treatment And Results Of Patients Having Hydrocephalus

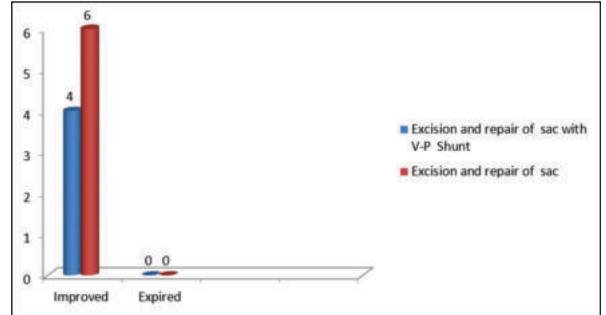
Type of Treatment	Improved	Absconded	Expired	Total
Operative (V-P. Shunt)	21	0	2	23
Other (Operated elsewhere)	02	01	0	03
Total				26



The surgical treatment was given for 23 (88.46%). Ventriculo-peritoneal shunt (V-P Shunt) was done.21 (80.77%) cases were improved with signs and symptoms of hydrocephalus and 2 cases expired after 3rd day of operation due to severe Meningitis post operatively. One case on 4th day absconded without information. The surgical mortality rate was 7.69%

Table 16 Treatment And Results Of The Patients Having Menigocele

Type of Treatment	MENINGOCELE WITH HYDROCEPHALUS		MENINGOCELE WITHOUT HYDROCEPHALUS	
	Excision and repair of sac with V-P Shunt	Excision and repair of sac	Improved	Expired
	Improved	Expired	Improved	Expired
	4	0	6	0
TOTAL	4	0	6	0

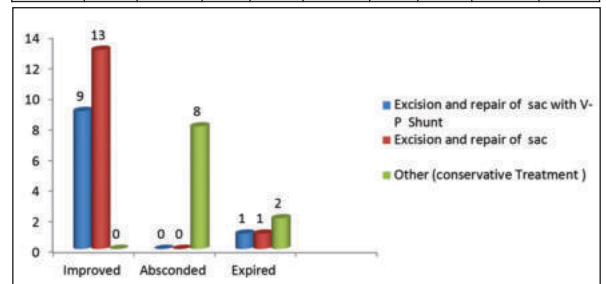


patients having Menigocele given operative treatment. 10 cases of Menigocele were given surgical treatment as excision of sac and 4 cases of Menigocele associated with hydrocephalus were given surgical treatment. In second step after 4-5 months of initial surgery. Ventriculoperitoneal shuntwas done in all 4 cases of Menigocele with hydrocephalus.

6 Cases operated for excision of sac only, were cured completely and cases having associated hydrocephalus were improved markedly. The overall mortality in all surgically treated patients was nil.

Table 17 Treatment And Result Of The Patients having myelomenIngocele

Type of Treatment	MYELOMENIN GOCELE WITH HYDROCEPHAL US			MYELOMENIN GOCELEWITH OUT HYDROCEPHAL US			OTHER (Conservative Treatment)		
	Impr oved	Absc onded	Expi red	Impr oved	Absc onded	Expi red	Impr oved	Absc onded	Expi red
	9	0	1	13	0	1	0	6	2
TOTAL	10	14	8						



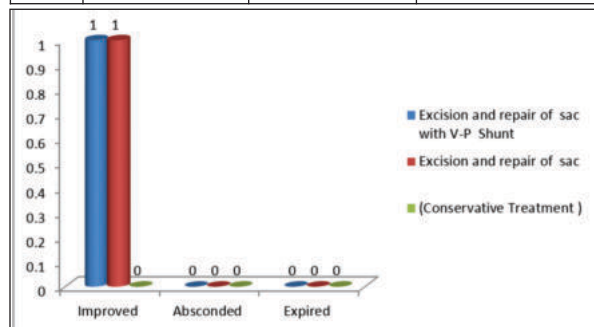
24 patients of Myelomeningocele were given surgical treatment out of total 32cases of Myelomeningocele. 14 cases were operated for excision and repair of sac and. Out of 14

cases for which excision and repair of sac were done 13 cases improved while 01 expired on 3rd post op day.

10 cases of Myelomeningocele with hydrocephalus were operated for excision and repair of sac followed by V-P Shunt after 4-5 month, 1 cases of Myelomeningocele with hydrocephalus were operated initially of V-P Shunt which was followed by excision and repair of sac 4-5 month expired, while the rest of 9 patients improved. Out of 8 cases that went for conservative treatment 6 of them absconded after knowing the poor prognosis and 2 patients expired. The overall mortality was 12.5% out of 32 case of Myelomen-ingocele. The overall surgical mortality was 8.33% out of 24 patients who were operated.

Table 18 Treatment And Results Of Patients Having Encephalocele

Type of Treatment	ENCEPHALOCELE WITH HYDROCEPHALUS			ENCEPHALOCELE WITHOUT HYDROCEPHALUS			OTHER (Conservative Treatment)		
	Improved	Absconded	Expired	Improved	Absconded	Expired	Improved	Absconded	Expired
	1	0	0	13	1	0	0	0	0
TOTAL	1	0	0	13	1	0	0	0	0



surgical treatment given in 2 patients of Encephalocele in the form of excision and repair of sac or excision of sac followed by V-P Shunt had good result.

Table 19 Treatment And Results Of Patients Having Spina Bifida Occulta

OUTCOME IN PATIENTS OF SPINA BIFIDA OCCULTA	SPINA BIFIDA OCCULTA			
	CONSERVATIVE		RELEASE OF TETHERING OF CORD	
	IMPROVED	EXPIRED	IMPROVED	EXPIRED
TOTAL PATIENT(2)	0	0	2	0

surgical treatment given in 2 patients of Spina Bifida Occulta in the form of release of tethering of cord result.

DISCUSSION

Central nervous system is relatively common site for malformations. The abnormalities in size, shape and symmetry of the face often suggest underlying congenital malformation. The present study deals with the various malformations of central nervous system involving anatomy of fetal brain and spinal cord.

Present study was conducted in the department of Neurosurgery, Nehru Hospital, B.R.D. Medical College, Gorakhpur in association with the department of Radiology. The present study was from January 2019 to December 2019. In all, 72 cases were studied; attending or admitting in the outdoor as well as indoor department of Neurosurgery and

Pediatrics department all patients underwent clinical examination to establish the diagnosis. Radiological examination was done in all doubtful cases including x-ray, ultrasound and C.T. Scan, M.R.I. Scan

The first report of ultrasound to image pathology in neonatal brain appeared in late 1970 (Hamberger et al, 1978, Jonson et al, 1979). It has become the most widely used imaging technique in neonatal period. The utility of ultrasound is due to its safety, portability, and ease of use and it has been widely adopted as a clinical tool by neonatologist. It remains the first line method for screening all intracranial problems in early infancy; the most appropriate imaging technique to be used depends on pathology or condition to be detected.

In this series the total number of patients having neurological problems were 1596, out of them 72 (4.51%) were found having congenital central nervous system malformation. Mapas P(1937) showed that approximately 1% of live births were having central nervous system malformation. Mc-Intosh et al (1954) found the incidence of congenital malformation of central nervous system was 1.0%. Carter (1960) found an incidence of 0.6%. Simpkins and Lowe (1961) found 0.1% incidence of congenital central nervous system malformation. Ghosh and Bali (1963) reported a 0.7% incidence of central nervous system malformation. Saifullah S. et al(1967) found 1.2% incidence of central nervous system malformation. In present study the incidence was taken among the patients who were having any type neurological problems and admitted or attended Neurosurgery and Pediatrics department, and the incidence was 4.51%. The relatively low socio-economic status in this area central nervous system malformation.

Due to paucity of information, it was difficult to establish a correlation between maternal history and congenital malformations of the central nervous system.

In this study the incidence of congenital malformation of the central nervous system were more in males (61.11%) than females (38.89%), i. e. male: female ratio was 1.6:1. Saifullah et al (1967) found the incidence of congenital malformation were more in males (55.6%) than females (44.4%). Mathur B.C. et al (1975) studied 1060 newborn including still birth, reported overall incidence of malformations in males: females was 2:1. In the study of Ingalls N.W. (1933) of 60 cases reviewed 80% patients were girls. Demmel (1950) studied 324 cases and he found equal ratio of males and females. In our study the slight higher incidence in males may be due to the higher importance of the male child in this area and poor health care given to the female child by parents.

Our observation showed that maximum (31.94%) patients were brought to the hospital with these malformations between 1-6 months of the age and minimum (5.6%) cases were brought between 12-18 months of age. Jess T. Schwidde (1952) studied 225 patients were brought to the hospital in first 3 weeks of their life and 82.2% in the first year of life. In the present study 6.94% patients were brought to the hospital within a month of life and 76.39% were brought within the first year of life. The delay in taking medical advice for treatment may be due to the poor attention or ignorance about the healthcare in this relatively poor and backward area.

As per our study on clinical diagnosis myelomeningocele was found in 44.45 % cases followed by Hydrocephalus in 36.1% cases. And meningocele 13.89, encephalocele 2.78, spina bifida occulta 2.78. Mathur B.O et al (1975) reported 8 cases of congenital central nervous system malformations amongst 33 cases of all types of congenital malformation and they showed the incidence of anencephaly was in 50% , hydrocephaly in 25% . meningocele in 12.5% and spina bifida .Siris (1936) found 77.4% cases of Myelomeningocele , and almost same incidence 72.6% were also found by Ingraham and swan (1943). In our study the incidence of

Myelomeningocele amongst spina bifida is 61.1%.

In our study we found that 75% cases having central nervous system malformation belong to lower socio-economic status. The reason may be as mentioned by saifullah et al (1976), anemia, malformation, infection and vitamin deficiency, which may be responsible for low birth-weight baby with multiple congenital malformation. In our study we found the maximum presenting clinical symptom was a swelling in more than two-third (87.5%) of the patients and least symptom was convulsion, only in 8 (11.11%) patients an abnormal swelling drew the attention of the parent's early. Paraplegia and other neurological symptoms drew attention of the parents relatively in later stage.

The commonest site of myelomeningocele in our study was in lumbar region (47.05%) followed by lumbo-sacral region 32.4%. The least incidence (2.9%) was noted in cervical region. In the study of Jess T. Schwidde (1952) the 52.8% of Myelomeningocele and Meningocele in lumbar region and followed by lumbo-sacral region (18.6%). He noticed only 1% cases in cervical region. Nihal Gurshinghe (1995) reported that Myelomeningocele were most commonly occupied thoraco-lumbar region 45%, lumbar 20% ,lumbar —sacral 20% and sacral 10% regions. Michel pollay also reported most of the Myelomeningocele were in lumbar region. In our study the incidence of Myelomeningoceles is correlating with the study of Jess T. Schwidde and the incidence of Meningocele was 60% in lumbar region which was followed by lumbosacral region 20% only one cases was noticed in M.R.I. Scan confirmed thoracic region.

The commonest site for Encephalocele in our study was in occipital region. We found 100% cases of Encephalocele in occipital region as per the study of Jess T. Schwidde in 1952, he also found the commonest site of Encephalocele in occipital region in 73% cases and 27% cases were in frontal region. Mealey J. Et al 1970 reported 85.0% lesions were occipital in western hemispher in contrast to the predominance of fronto-nasal in Far East.

The incidence of hydrocephalus in our study was 63% alone or in association with other central nervous system malformation. Almost half the patients of Meningocele, Myelomeningocele or Encephalocele showed hydrocephalus on C.T and M.R.I. examination. In study of Ingraham and swan (1943) found the incidence of hydrocephalus in 36.5% cases of spina bifida and Encephalocele. Jess T Schwidde (1952) found the incidence of hydrocephalus in 52.0% cases of spina bifida and Encephalocele present study hydrocephalus was present in 44.0% cases of spina bifida and Encephalocele. Our study is quite similar to the above-mentioned study.

In our study we found the commonest cause of hydrocephalus was aqueductal stenosis. It accounts about 50% cases of hydrocephalus communicating hydrocephalus in 9 cases followed. It. The least common cause of hydrocephalus was due to Dandy —Walker anomalies which occurred in 5 (19.23%) cases only. Milhorat (1972) found the cause of hydrocephalus in two-third of cases due to aqueductal stenosis. Burton in 1979 found Dandy- Walker anomalies responsible for 13% case of hydrocephalus. In our study the incidence of aqueductal stenosis is slightly lower 46.15%. It may be due to the less number of hydrocephalus patients studied.

In our study we found that C.T. and M.R.I. scan is an important and effective investigation to diagnose the presence of hydrocephalus with or without the other central nervous system malformation like Meningocele Myelomeningocele, Encephalocele and spina bifida occulta.

All the patients studied were put on the treatment surgical /conservative. The maximum cases of hydrocephalus,

Encephalocele, Meningocele, spina bifida occulta and myelomeningocele were given operative treatment. Excision and repair of the sac was done in case of Meningocele, Encephalocele and myelomeningocele. And Hydrocephalus patients were operated for V_P Shunt and only 03 case of hydrocephalus were not given surgical treatment due to gross neurological involvement with low general condition. The overall mortality rate for myelomeningocele was 12.5% and it was 8.33% in cases of surgically treated patients. Maximum mortality took place within first week of treatment. 6 patients absconded within 3-4 days of admission as per the poor prognosis that was explained to them. The surgical treatment was given for 24 patients, in the form of excision and repair of sac in 14 patients and VP shunt with excision and repair of sac done in 09 cases. Ingraham and Swan in 1943 found 12% mortality in his 188 cases of meningomyelocele.

In case of Encephalocele patients 02 patients were given surgical treatment. Prognosis was good in surgically treated patients. Mealey (1970) reported an 83% mortality rate in patients who did not undergo surgical treatment. Mahapatra A.K. (1994) reported no mortality in 30 cases of encephalocele treated surgically.

In the case of spina bifida occulta 2 patient were given surgical treatment. Prognosis was good in surgically treated patients.

CONCLUSION

In this study we have done an analysis of the cases of congenital malformations of the central nervous system admitted/ attended Neurosurgery and Pediatric Department, Nehru Hospital, B.R.D. Medical College, Gorakhpur from January 2019 to December 2019.

These patients were studied with particular reference to their clinical features, radiological examinations. Conclusion of this work is as follows:-

- The total of patients included in this study was 72. 44 patients were male and 28 patients were females. MALE: FEMALE ratio was 1.6:1
- The incidence of congenital malformations of the central nervous system among all the patients having neurological problems was 4.51
- The maximum patients of congenital malformation of the central nervous system came to the Neurosurgery and Paediatric department either directly or referred from other centres and departments, were in the age group of 1-6 months. Which was 31.9% and minimum in 12-18 months (5.56%)
- Maximum number of cases were of myelomeningocele, which was 44.45% and least were of encephalocele and spina bifida occulta (2.78%)
- The maximum cases of congenital malformation of central nervous system were of low socio-economic status. They accounted 75% and minimum were from upper socio-economic status, 5.56% only.
- Maximum patients came in department with the complaints of swelling (87.5%) since birth in midline and increased head size. Some of them were associated with convulsion (11.11%) and fever
- In maximum patients of myelomeningocele the lesions were present in lumbar region accounting 46.8% followed by lumbo-sacral region 31.25% and least were present in cervical region 3.12%
- Out of 10 cases of meningocele, the maximum 6 cases were present in lumbar area accounting 60% and least in the thoracic and cervical region (10%)
- All 2 cases of the encephalocele were present in occipital region
- On M.R.I. examination of 32 myelomeningocele patients, 14 patients showed presence of hydrocephalus, 40% patients of meningocele and 50% patients of

encephalocele were also having hydrocephalus.

- M.R.I. of all 26 patients of hydrocephalus, 46.15% showed aqueductal stenosis and 19.23% Dandy-Walker anomaly, 34.61% patients showed communicating type of hydrocephalus.
- Mostly patients required C.T. scan and M.R.I. scan for detection of hydrocephalus and to see the content of the sac
- 61 patients (84.72%) patients of all types of congenital central nervous system malformation were given surgical treatment and overall surgical mortality was 6.56%
- In all the patients of hydrocephalus either they were treated surgically or conservatively, the mortality was 11.5%. Surgical mortality was 4.34% for hydrocephalus Surgical mortality was nil in cases of meningoceles
- In cases of myelomeningocele patients out of 32 patients 24 (75%) underwent surgery. The overall mortality in the cases of myelomeningocele whether they were treated surgically or conservatively was 12.5%. Surgical mortality was 8.34%
- Surgical treatment had good outcome(100%) on the encephalocele patients who underwent surgery.
- Surgical treatment had good outcome(100%) on the spina bifida occulta patients who underwent surgery.
- The ratio of different typed of congenital malformation of the central nervous system were as follows:

Myelomeningoceles	32	44.45%
Hydrocephalus	26	36.14%
Meningoceles	10	7.2%
Encephaloceles	02	02.78%
spina bifida occulta	02	02.78%

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