Clinical Evaluation and Management of Children with Meningomyelocoele

Dinesh Shukla[1], Sanjay Chaube[2]

[1]Associate Professor, Department of Neurosurgery, MLB Medical College, Jhansi, Uttar Pradesh.
[2]Associate Professor, Department of General Surgery, MLB Medical College, Jhansi, Uttar Pradesh.

ABSTRACT

BACKGROUND

Neural tube defects (NTDs) are one of the most common birth defects affecting the Central Nervous System (CNS) and is often associated with complex birth defects compatible with survival.[1] NTD is a general term for a congenital malformation of the central nervous system (CNS) occurring secondary to lack of closure of the neural tube and has a worldwide incidence of 1.0 to 10.0 per 1,000 births. Congenital malformations of spine and spinal cord are collectively termed as spinal dysraphism. It includes a heterogenous group of anomalies resulting from incomplete midline closure of osseous, mesenchymal and nervous tissue.[2] Most of these conditions are diagnosed at or soon after birth, but some are diagnosed late in childhood or in adulthood because of absence of clinical manifestations. We wanted to evaluate the causative factors, clinical features, management and postoperative complications of children with meningomyelocoele.

METHODS

This is a retrospective study and all children having meningomyelocoele born between April 2014 to May 2016 were included in the study. Clinical features, maternal educational status, size and location of the defect, operative details such as time to surgery and its relation to postoperative morbidity and mortality were recorded.

RESULTS

A total of 36 patients were evaluated in the study period. There were 20 (55.6%) males and 16 (44.4%) females. Most common location of MMC was in lumbar (83.3%) and followed by thoracolumbar (16.7%) regions. None of the mothers received folate supplementation. Hydrocephalus (n = 18), pelvicalyceal ectasia (n = 3), pes equinovarus (n = 7), Chiari II malformation (n = 4)), pelvicalyceal ectasia (n = 3), hip dysplasias (n = 2), accounted for additional anomalies. Short term complications following surgery were bladder dysfunction in 8 patients (22.2%), 7 patients (19.4%) had seizures, 6 patients (16.7%) had bacterial meningitis and 3 patients (8.3%) had severe hydrocephalus and 4 patients (11.1%) had ventriculitis.

CONCLUSIONS

Operating on patients with meningomyelocoele with in 72 hrs. of birth reduces complications, hospital stay and antibiotic usage.

KEYWORDS

Meningomyelocoele, Neonate, Complications

Corresponding Author:
Dr. Sanjay Chaube,
Associate Professor,
Department of General Surgery,
MLB Medical College,
Jhansi- 284128, Uttar Pradesh.
E-mail: dr.dineshshukla@gmail.com
DOI: 10.18410/jebmh/2020/34

Financial or Other Competing Interests: None.

How to Cite This Article:
Shukla D, Chaube S. Clinical evaluation and management of children with meningomyelocoele. J. Evid. Based Med. Healthc. 2020; 7(4), 164-167. DOI: 10.18410/jebmh/2020/34

Submission 13-01-2020,
Peer Review 17-01-2020,
Acceptance 24-01-2020,
Published 27-01-2020.

BACKGROUND

Neural tube defects (NTDs) are one of the most common birth defect affecting the Central Nervous System (CNS) and is often associated with complex birth defect compatible with survival.[1] NTD is a general term for a congenital malformation of the central nervous system (CNS) occurring secondary to lack of closure of the neural tube and has a worldwide incidence of 1.0 to 10.0 per 1,000 births. Congenital malformations of spine and spinal cord are collectively termed as spinal dysraphism. It includes a heterogenous group of anomalies resulting from incomplete midline closure of osseous, mesenchymal and nervous tissue.[2] Most of these conditions are diagnosed at or soon after birth, but some are diagnosed late in childhood or in adulthood because of absence of clinical manifestations.

Meningomyelocoele (MMC) is a posterior closure defect of the neural canal, which develops in the first 26 days of pregnancy.[3] Sociodemographic and epidemiologic findings have been correlated with risk for a neural tube defect affected pregnancy. It has also been shown in studies that maternal high body temperature or exposure to heat (sauna, hot tub bath) during initial 4 weeks of pregnancy, a low socioeconomic level are related to NTD development. Others factors implicated in etiopathogenesis are exposure to radiation, antiepileptic drugs (valproic acid, carbamazepine) and genetic factors. Also, maternal folic acid, zinc, selenium and methionine defeciencies as supposed to play a role in the development of NTDs.[4,5] The frequency of meningomyelocoele in 1960s was 6 per 1000 live births in England. But due to termination of pregnancies owing to early in utero diagnosis and periconceptional folate treatment, frequency of meningomyelocele has been reduced to 1 per 1,000 live births. The mechanism by which periconceptional folate prevents meningomyelocoele is unclear but it decreases its prevalence by 60-70%.

Present study aims to elucidate the importance of surgical timing to prevent early complications of meningomyelocele and to determine the clinical characteristics and additional anomalies of neonates with meningomyelocoele.

METHODS

It was a retrospective study and all the children having meningomyelocoele admitted under neurosurgery department of Maharani Laxmi Bai Medical college Hospital, Jhansi between the periods April 2014 to May 2016 were included in the study. Children with other findings such as encephalocoele, meningocele, oculo-spinal dystrophy were not included in the study because of differences in treatment regimens and early period complications. Informed consent was obtained from their families.

Patient Selection Criteria

Neonates with meningomyelocele were included in the study during the selected period. Physical examination findings such as weight, height, head circumference and type of delivery were recorded. The antenatal findings, maternal education status, maternal age and whether or not an antenatal diagnosis was present were noted. The localization and size of the meningomyelocele and presence of additional anomalies and skin defects were established. Plasma urea and creatinine levels within the first 72 hrs after birth were done to evaluate the renal functions. The patients were divided into two groups according to the time of surgical operation: group 1, in which the patients were operated with in first 72 hrs of life and group 2 in which the patients were operated after 72 hrs of life. Group 2 comprised of patients with an infected meningomyelocele sac, unstable condition, those not accepted as candidates for an emergency operation and patients who were referred to our hospital after 72 hrs. The effect of operation on antibiotic therapy and duration of hospital stay along with early morbidity were assessed.

Evaluation and Treatment

Neurological examination findings of the patients were recorded, including deep tendon and motor reflexes of lower extremities. Cranial and abdominal ultrasonography was performed to evaluate additional anomalies. CT scan was also done before the meningomyelocele correction surgery. Severe hydrocephalus was diagnosed on the basis of dilatation of all the ventricles and whose cerebral cortex thickness was <1 cm. Prophylactic antibiotics were given to neonates who were born outside the hospital and if they had an infected open sac. In cases of suspected meningomyelocele, cerebrospinal fluid was obtained using transfrontal puncture. Positive bacterial CSF culture confirmed the bacterial meningitis diagnosis.

Operative Details

Depending upon the type of lesion, skin incision was planned so as to expose the healthy skin all around. Neural placode was separated from the surrounding tissues. Neural tube was repaired by pial sutures and dura was repaired. Patients in whom adequate tissue was present, subcutaneous tissue and skin were approximated and sutured. In patients with large skin defect, with the help of plastic surgeon skin and subcutaneous flap was elevated to close the defect. If hydrocephalus was found on CT scan and in whom no CSF infection was present, Ventriculoperitoneal shunt was placed prior to the correction procedure.

RESULTS

There were 36 patients who were included in the study retrospectively. Clinical characteristics of patients who were included in the study are presented in Table 1. There were 20 (55.6%) males and 16 (44.4%) females. 61.1% were delivered by caesarean section and 31.9% were product of normal delivery. Only 27.8% were diagnosed antenatally but families of all of them decided not terminate the pregnancy. Educational status of most of the mothers of patients was

low and most of them (77.8%) had education only till elementary school. The anatomical localization, neurological findings and treatment of patients are presented in Table 2. Most common location of MMC was in lumbar (83.3%) and followed by thoracolumbar (16.7%). 4 patients (11.1%) had normal neurological findings. Additional anomalies which were noted are Hydrocephalus in 18 patients (50%) and pes equinovarus was noted in 7 patients (19.4%). None of the cases had renal failure based on urea and creatinine reports. 18 patients who had hydrocephalus without any findings of infection underwent ventriculoperitoneal shunt in the same operation. There were big skin defects in 22 patients (61.1%) requiring flap cover with the help of plastic surgeon. Short term complications following surgery were bladder dysfunction in 8 patients (22.2%), 7 patients (19.4%) had seizures, 6 patients (16.7%) had bacterial meningitis and 3 patients (8.3%) had severe hydrocephalus and 4 patients (11.1%) had ventriculitis. Patients were divided into two groups: the cases operated before and the cases operated after the third post-natal day. 24 patients were operated before and 12 patients were operated after 3 days of birth. Of 7 patients who had seizures, 5 of them developed seizures after the insertion of shunt which was placed within first 3 days after birth. 2 patients had seizures before shunt insertion which was done after third day of life.

	Group 1	Group 2	Total
No. of Patients	24	12	36
Height, cm	46.2±3.0	46.1±1.7	
Weight, g	2,950 ± 510	3050 ± 410	
Head Circumference, cm	36.5 ± 3.0	37.0± 2.5	
Gender			
Male	13	7	20 (55.6)
Female	11	5	16 (44.4)
Type of Delivery			
Caesarean	14	8	22 (61.1%)
Vaginal	10	4	14 (31.9)
Antenatally diagnosed	7	3	10 (27.8)
Maternal Education			
Uneducated	4	2	6 (16.7)
Elementary	18	10	28 (77.8)
High School	2	0	2 (5.5)
Maternal Age, years	24.8±4.9	26.9±3.0	
Table 1. Antenatal and Postnatal Characteristics of Patients			
Data presented as mean ± SD or n (%)			

Operation time, days	3.1 ± 2.7
Duration of Hospitalization, days	11.2± 8.4
Dimensions of defect (diameter, cm)	5.9± 2.4
Duration of antibiotic treatment, days	7.4±3.8
Location of MMC	
Lumbar	30 (83.3%)
Thoracolumbar	6 (16.7%)
Neurologic Examination	
Hypotonia	28 (77.8%)
Areflexia	4 (11.1%)
Normal	4 (11.1%)
Additional Anomalies	
Hydrocephalus	18 (50)
Chiari II malformation	4 (11.1%)
Hydronephrosis	5 (13.9%)
Hip dysplasia	2 (5.5%)
Pelvicalyceal ectasia	3 (8.3%)
Pes equinovarus	7 (19.4%)
Table 2. Clinical Findings and Treatment of Patients	
Data presented as means ± SD or n (%)	

None of the patients who were operated within first 3 days after birth developed bacterial meningitis or ventriculitis. 6 patients out of total 12 who were operated after 3 days of birth were diagnosed as having meningitis. 4 patients who were referred to our hospital in second and third weeks after birth were diagnosed as having ventriculitis. It was deduced from these findings that patients who had been operated within first 3 days of birth had short hospital stay and less duration of antibiotic therapy. They also had less early period complications than others.

	Time to Operation	
	< 3 days (n=24)	> 3 days (n=12)
Operation time, days	2.1±0.9	7.5± 1.7
Hospitalization period, days	7.4±9.2	12.6±4.2
Defect Dimensions (diameter,cm)	5.2±3.2	5.6±2.7
Short-term complications, n		
Seizure	5	2
Bladder dysfunction	6	2
Ventriculitis	0	4
Meningitis (Bacterial)	0	6
Severe Hydrocephalus	3	0
Total complications	14	14
Table 3. Relation of Operation Time to Morbidity		
Data presented as means ± SD or n (%)		

DISCUSSION

The prevalence of Neural tube defects (NTD) is varied ranging from 1.0 to 10.0 per 1,000 births. Various studies have shown that NTD prevalence is higher in families with low Socioeconomic status, as measured by parental occupation, education and household income.[6] In present study too, 94.4% of mothers had a low educational status. Folic acid deficiency a another known risk factor for NTDs. Folic acid supplementation of 0.4 mg/dl has been reported to reduce the incidence of NTDs by 70%.[7] As only half of this amount can be supplied by through normal adult diet, folic acid supplementation is must for prevention of NTDs. Folic acid has been added to grain products in developed countries for last 20 years.[8] In present study it was found that during pregnancy none of the mothers were taking folic acid supplementation necessitating the need to fortify the grains here too.

It is established that MMC correction surgery should be performed with in 48 hrs of birth if there are no signs of infection in CSF. The chances of ventriculitis becomes higher if the MMC correction is performed 72 hours postnatally.[9] Findings in the present study corresponds to the evidence in the Literature that neonates who underwent surgery after 72 hours of delivery had a higher chance of meningitis and ventriculitis. There is controversy regarding the timing of shunt operation in patients who have both MMC and hydrocephalus. It is believed that inserting a ventriculoperitoneal shunt at the time of MMC correction does not contribute to infection.[9] However few studies have reported that frequency of ventriculoperitoneal shunt infections increased when shunt insertion and correction operation are performed in the same sitting.[10] In present study we observed that shunt infection was found in only one patient and we suggest that the shunt insertion and correction procedure should be done together. There are reports that early and aggressive surgery reduces

complications and prevalence of death in the short term and that it also reduces the chances of urinary incontinence and duration of stay in long term follow-up.[11] In present study, there was no mortality in the neonatal period. Operating within first 3 days of life is the key to avoiding problems in early neonatal period. It was observed in present study that if surgery was performed with in first 3 days of postnatal, the prevalence of meningitis and ventriculitis as postoperative complication was low.

As the practice of routine antenatal checkups and fetal ultrasonography has become a common in most places, chances of early diagnosis and hence opportunity to terminate the pregnancy in early period has increased. However, most families rarely opt for the termination of pregnancy.[12] Although about 27.8% of patients have been diagnosed with meningomyelocele during antenatal period in our study, none of the families have opted for termination of pregnancy. In many studies, it has been found that meningomyelocoele is most commonly located in the lumbar region. We also observed that incidence of meningomyelocoele was mostly present in lumbar region (83.3%). Additional anomalies which are found are hydrocephalus and Chiari II malformation.[12] In children with MMC, increased intracranial pressure and hydrocephalus occur within the first month of life. Hydrocephalus was the most common anomaly encountered in our study (50%). Other common anomalies include Urinary system anomalies. That's why, all patients with MMC should be assessed by Ultrasonography, even though they may not be manifesting any symptoms.

It has been reported in many studies that 2-8% of patients with meningomyelocoele have seizures. In presence of hydrocephalus, the incidence of seizures increases to 14.7 - 29%.[13] It has been observed that shunt and hydrocephalus are the reasons for seizures and as they cause minor cortical damage along with CNS infection and congenital cerebral cortical dysplasia. In accordance with literature, in present study 4 children had cortical damage because of ventriculoperitoneal shunt and 3 had seizures due to infection.

CONCLUSIONS

Meningomyelocele is a disease with a relatively high prevalence of early period complications. So, strategy should be devised to build up the national folic acid supplementation program and also to educate and encourage mothers regarding folic acid supplementation for prevention of neural tube defects. An effort towards early surgery of MMC is needed so as to reduce hospitalization and avoid early period complications. Pregnant women diagnosed with baby having MMC antenatally should be promptly referred to appropriate tertiary care hospitals for better care and treatment.

REFERENCES

[1] Liptak GS, El Samra A. Optimizing health care for children with spina bifida. Dev Disabil Res Rev 2010;16 (1):66-75.

[2] French BN. The embryology of spinal dysraphism. Clin Neurosurg 1983;30:295-340.

[3] Back SA. Congenital malformations of the central nervous system. in Taeusch HW, Ballard RA, Gleason CA, eds. Avery's diseases of the newborn. 8th edn. Philadelphia: Elsevier 2005:938-959.

[4] Hambidge M, Hackshaw A, Wald N. Neural tube defects and serum zinc status. Br J Obstet Gynaecol 1993;100 (8):746-749.

[5] Essien FB, Wannberg SL. Methionine but not folinic acid or vitamin B-12 alters the frequency of neural tube defects in Axd mutant mice. J Nutr 1993;123 (1):27-34.

[6] Farley TF, Hambidge SJ, Daley MF. Association of low maternal education with neural tube defects in Colorado, 1989-1998. Public Health 2002;116 (2):89-94.

[7] Czeizel AE, Dudás I. Prevention of the first occurrence of neural-tube defects by periconceptional vitamin supplementation. N Engl J Med 1992;327 (26):1832-1835.

[8] Canfield MA, Ramadhani TA, Shaw GM, et al. Anencephaly and spina bifida among Hispanics: maternal, sociodemographic, and acculturation factors in the National Birth Defects Prevention Study. Birth Defects Res A Clin Mol Teratol 2009;85 (7):637-646.

[9] Caldarelli M, Di Rocco C. Myelomeningocele primary repair surgical technique. In: Ozek MM, Cinalli G, Maixner WJ, eds. Spina Bifida: management and outcome. Milan, Italy: Springer-Verlag 2008:143-155.

[10] Oktem IS, Menkü A, Ozdemir A. When should ventriculoperitoneal shunt placement be performed in cases with myelomeningocele and hydrocephalus? Turk Neurosurg 2008;18 (4):387-391.

[11] Stein SC, Schut J, Ames MD. Selection of early treatment of myelomeningocele: a retrospective analysis of selection procedures. Dev Med Child Neurol 1975;17 (3):311-319.

[12] Koszutski T, Kawalski H, Kudela G, et al. Babies with myelomeningocele in Poland: parents' attitudes on fetal surgery versus termination of pregnancy. Childs Nerv Syst 2009;25 (2):207-210.

[13] Isık U. Seizures in children with myelomeningocele. In: Ozek MM, Cinalli G, Maixner WJ, eds. Spina Bifida: management and outcome. Milan, Italy: Springer-Verlag 2008:297-301.