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SHORT TERM AUDIT OF OUTCOMES OF CHILDREN WITH NEURAL TUBE DEFECT REPAIR IN A TERTIARY CARE CENTER

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ABSTRACT **Introduction:** Neural tube defects is typically a disabling condition that results in neurologic, orthopedic, and urologic morbidity. Early diagnosis, surgical intervention and rehabilitation can prevent further neurological damage as well as improve the quality of life as well as ultimate outcome in patients with NTD. Closure of neural tube defects is a common neurosurgical procedure but outcome is highly variable and dependant on multiple factors. The aim of this study was to study outcomes and to identify predictors morbidity of surgically treated NTDs. **Aim:** Short term audit of outcomes of children with neural tube defect repair in a tertiary care center. **Results:** A total of 60 babies were included in this study. The median age at admission is 30 days with range from Day 2 -270 of life. Forty-two (70%) of the babies were male and 18 (30%) were female. The median size of swelling at admission was 5cm with range from 2 to 11 cm. Location of swelling were cervical in 4 (6.66%), upper thoracic in 1 (1.66%), dorso-lumbar in 11 (18.33%), lumbar in 22 (36.66%), lumbo-sacral in 19 (31.66%) and sacral in 3 (5%). Weakness in lower limbs was present in 22 (36.66%) patients preoperatively. Neurogenic bladder and bowel along with bilateral CTEV were present in 8 (13.33%) of patients. CSF discharge from swelling was present in 10 (16.66%) patients preoperatively. Preoperative hydrocephalous was present in 15 (25%) patients. Arnold Chiari malformation was present in 18 (30%) patients. Post operatively, wound infection was present in 19 (31.66%), CSF leak was present in 10 (16.66%), new neurological deficit developed in 11 (18.33%) and new hydrocephalous developed in 9 (15%) patients. Out of total 60 patients, ventriculo-peritoneal shunt was placed in 22(36.66%) for hydrocephalous. The median length of postoperative stay was 10 days with range from 4 to 28 days. Re-admissions were required in 17 (28.33%) patients and 2 (3.33%) patients were expired due to sepsis and aspiration pneumonia. **Conclusion:** Neural tube defect is a congenital disorder with significant morbidity. Early diagnosis, surgical intervention and rehabilitation can prevent further neurological damage as well as improve the quality of life as well as ultimate outcome in patients with NTD. There is considerable delay in surgical management in the majority of cases, requiring concentrated efforts for early surgical management of neonates with NTD to reduce the morbidity and improve the ultimate outcome.

KEYWORDS : neural tube defect, hydrocephalus, outcome, meningomyelocele, ventriculitis

INTRODUCTION

Neural tube defects (NTDs) are congenital malformations of the central nervous system [1]. They are caused by partial/incomplete closure of the neural tube during embryogenesis, between 21 and 28 days after conception [2, 3]. Disorders of primary neurulation include craniorachischisis in which the neural tube fails to initiate closure, leaving most of the brain and the entire spine open. If closure initiates successfully, then the cranial and/or spinal neural folds may fail to close generating exencephaly/anencephaly and open spina bifida (myelomeningocele). Malformations resulting from disturbance of secondary neurulation are closed (skin covered) and often involve tethering of the spinal cord, with associated ectopic lipomatous material [4, 5]. Nevertheless, recent evidences support a post-neurulation origin for encephalocele and demonstrate that brain herniation and failure of brain/spine neural tube closure can all occur as possible developmental outcomes of an identical genetic defect [6]. Infants with anencephaly are mostly stillborn, or die shortly after birth, while those with spina bifida and encephalocele may survive, although suffering from physical and developmental disabilities of various degrees of severity [7]. Worldwide average incidence is 1.8/1000 births (more than 300,000 affected infants born every year), with significant geographic (ethnic, environmental, socioeconomic) variations (> 1 to around 10/1000 births in middle and low-income countries (LMICs) and decreasing incidence in high-income countries (HICs, 0.6/1000 in USA) in the recent decades [8, 9].

NTDs are among the main causes of childhood mortality and disability worldwide, with higher rates in LMICs for limited access to specialized neonatological/pediatric and surgical care [8,9,10]. Moreover, inequities in the access to high quality care affect pregnant/childbearing age women [11], in relation to low rate of prenatal diagnosis and prevention tools (preconception folic acid supplement), thus explaining the higher prevalence of NTDs at birth.

Very few studies have been published about the short term outcomes after NTDs repair in terms of multiple variables and factors. Therefore, the aim of this study is to discuss the short term outcome of babies with NTDs repair.

MATERIAL AND METHODS

This was the retrospective study performed over past 2 year from April 2021 to April 2023 in the department of Pediatric surgery at a tertiary care hospital after approval from the institutional ethics committee. All the cases of neural tube defects operated during this period were included after obtaining written informed consent from each patient's parent. In this retrospective study, we reviewed a total number of 60 children with neural tube defect admitted to the pediatric surgery department at our center. Data of all these patients were reviewed and compared in terms of clinical and radiological findings, intraoperative surgical options and postoperative complications and its management as per patient proforma (Annexure-1). All the final data was analyzed statistically and p-value <0.05 was considered significant.

Ethical Approval

The study was approved by the institutional ethical committee.

Informed Consent

It is a retrospective observational study, so no consent was obtained.

RESULTS

A total of 60 babies were included in this study. The clinical characteristics of the babies included in the study are summarized in

Table 1 & Table 2. The median age at admission is 30 days with range from Day 2 -270 of life. Forty-two (70%) of the babies were male and 18 (30%) were female.

The median size of swelling at admission was 5cm with range from 2 to 11 cm. Location of swelling were cervical in 4 (6.66%), upper thoracic in 1 (1.66%), dorso-lumbar in 11 (18.33%), lumbar in 22 (36.66%), lumbo-sacral in 19 (31.66%) and sacral in 3 (5%). Weakness in lower limbs was present in 22 (36.66%) patients preoperatively. Neurogenic bladder and bowel along with bilateral CTEV were present in 8 (13.33%) of patients. CSF discharge from swelling was present in 10 (16.66%) patients preoperatively. Preoperative hydrocephalous was present in 15 (25%) patients. Arnold Chiari malformation was present in 18 (30%) patients. Kyphosis was present in 11 (18.33%) patients. Congenital heart disease (Atrial septal defect) was present in 1 (1.66%) patient and 1 (1.66%) patient had webbing of 4 fingers with short phalanges in right hand. Meningitis/ventriculitis observed in 9 (15%) patients.

All patients underwent excision and repair of neural tube defect with or without one or both sided rhomboid flap closure. Post operatively, wound infection was present in 19 (31.66%), CSF leak was present in 10 (16.66%), new neurological deficit developed in 11 (18.33%) and new hydrocephalous developed in 9 (15%) patients. Out of total 60 patients, ventriculo-peritoneal shunt were placed in 22(36.66%) for hydrocephalous.

The median length of postoperative stay was 10 days with range from 4 to 28 days. Re-admissions were required in 17 (28.33%) patients and 2 (3.33%) patients were expired due to sepsis and aspiration pneumonia.

DISCUSSION

The term "spina bifida" was first coined in 1641 by Nicholas Tulp [12]. The spread of surgical treatment of open NTDs and the knowledge of pathophysiology (including primary prevention through folic acid preconception supplement), epidemiology and genetics of such defects were reached much later, in the late twentieth century [9].

The study identified 60 patients with NTDs that underwent neurosurgical management in a tertiary hospital over 2 years period. Although most of the mothers had obstetric ultrasound examination, the rate of prenatal diagnosis of NTD is very low. Our study revealed that lumbo-sacral MMC is the most common type of NTD.

In the present study, the most common neural tube defect in the admitted babies was MMC 51 (85%). In nearly half of the cases, the most common site of the involvement of MMC was the lumbar area, similar to that reported in earlier studies [13].

Hydrocephalus and Arnold Chiari malformation was found to be associated with 25% and 30% cases of MMC respectively. Taking this into consideration, it is prudent that all babies with MMC should be screened with the help of a careful physical examination and cranial imaging studies.

In the present study, 22 (36.66%) babies had lower limb weakness while bowel and bladder involvement was pre- sent in 8 (13.33%) babies. The degree of involvement was stronger depending upon the level of lesion i.e. the higher the lesion the more will be the involvement. Neonates with MMC often exhibit motor and sensory neurological deficit below the level of the lesion. This may result in lower limb weakness or paralysis. Urinary and faecal incontinence also occur frequently. [14]

The median age of presentation to hospital was 30 days. Delay in presentation and ultimate intervention results in increased morbidity. An early and aggressive surgical approach in babies diagnosed with MMC has been shown to be associated with lower early morbidity rates. Yekta et al. [15] in a retrospective study on 30 babies with MMC, reported early surgical intervention (<72h after birth) to be associated with a more favourable outcome in terms of urinary symptoms in patients with MMC. They also demonstrated that patients operated upon within the first 5days of life had significantly lower durations of hospital stay while also requiring shorter courses of antibiotics ($p < .001$). They recommended to perform surgery as soon as possible, particularly in patients diagnosed prenatally.

Preoperative hydrocephalous was present in 15 (25%) patients while 9(15%) patients developed hydrocephalus post-operatively. We retrieved data of 22(36.66%) patients who ultimately needed VP shunt surgery.

The 9 (15%) meningitis/ventriculitis observed in our study is similar to the finding in a Turkish study (16.4%). Moreover, in our study 19 (31.66%) patients developed surgical wound infection almost three times of reported in the same Turkish study.[16] Wound infection rate in our study was reported to be higher than other published studies.[17] Postoperative CSF leaks were observed in 10 (16.66) cases. In a study from India, a higher percentage (12.7%) of patients developed postoperative CSF leaks.[18]

Newborns and children with NTDs need a multidisciplinary approach and follow-up, involving neonatologists, neonatal surgeons, pediatric neurologists, urologists and orthopedics, geneticists, physiatrists, physiotherapists and neurorehabilitation therapists. Our multidisciplinary team aims to guarantee to children and families, an individualized care oriented to maintain normal biologic functions and social life by preserving urinary and stool continence, lowering the possible infective risk, and planning and a neuro-developmental follow-up.

The main limitation of the present study was small sample size, hence we suggest a large study over an extended period of time that will also help to formulate a comprehensive approach for prevention and management of NTDs in future.

CONCLUSION

Further investigations and workup for risk factors determinants prior to interventions mitigate the risk of complications and mortality are highly encouraged. Early diagnosis, surgical intervention and rehabilitation can prevent further neurological damage as well as improve the quality of life as well as ultimate outcome in patients with NTD. All newborn babies with NTD should be screened for additional congenital anomalies and evaluated with more organized, multidisciplinary methods. There is a requirement for enhancement in healthcare delivery, as prenatal detection of NTDs remains significantly lower. There is considerable delay in surgical management in the majority of cases, requiring concentrated efforts for early surgical management of neonates with NTD to reduce the morbidity and improve the ultimate outcome. There was high in-hospital presence of meningitis/ventriculitis, wound infection rates, hydrocephalus, new neurological deficit. We recommend a prospective study with large sample size to determine the outcome of children with NTD in this setting.

Author Contributions

All authors contributed significantly to this work in the conception, study design, execution, data acquisition, analysis, and interpretation; participated in the drafting, revising, or critical review of the article; gave final approval of the version to be published; agreed on the journal to which the article will be submitted; and agreed to be responsible for all aspects of the work.

Disclosure

The authors declare that they have no competing interests.

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Table 1. Clinical characteristics of the babies with NTD included in the study

S. No.	Characteristics	Total number of babies (n=60)
1	Age at presentation(days) median (IQR)	30 (2-270)
2	Male, n (%)	42 (70)
3	Size(cm) median (IQR)	5 (2-11)
4	Location	C-4,Upper T-1 DL-11 L-19 LS-22 S-3
5	Preoperative Power- weak, n (%)	22 (36.66)
6	CSF Discharge , n (%)	10 (16.66)
7	Kyphosis, n (%)	11 (18.33)
8	Preoperative Hydrocephalous, n (%)	15 (25)

9	Wound Infection, n (%)	19 (31.66)
10	CSF Leak, n (%)	10 (16.66)
11	New Hydrocephalous, n (%)	9 (15)
12	VP Shunt, n (%)	22 (36.66)
13	New Neurological deficit, n (%)	11 (18.33)
14	Re-admission, n (%)	17 (28.33)
15	Length of postoperative stay (days)median (IQR)	10 (4-28)
16	Mortality, n (%)	2 (3.33)

Table 2. Types Of NTDs

Type of NTD	Number of babies
Meningomyelocele	51 (85%)
Lipo-MMC	2 (3.3%)
Encephalocele	7 (11.6%)

REFERENCES

1. Piro E, Alongi A, Domianello D, Sanfilippo C, Serra G, Pipitone L, Ballacchino A, Provenzano C, Schierz IAM, Corsello G. Malformations of central nervous system: general issues. Acta Medica Mediterranea. 2013;29:735–40..
2. Wallingford JB, Niswander LA, Shaw GM, Finnell RH. The continuing challenge of understanding, preventing, and treating neural tube defects. Science. 2013;339(6123):1222002.
3. Serra G, Antona V, Schierz M, Vecchio D, Piro E, Corsello G. Esophageal atresia and Beckwith-Wiedemann syndrome in one of the naturally conceived discordant newborn twins: first report. Clin Case Rep. 2018;6:399–401.
4. Greene ND, Copp AJ. Neural tube defects. Annu Rev Neurosci. 2014;37:221–42.
5. Lew SM, Kothbauer KF. Tethered cord syndrome: an updated review. Pediatr Neurosurg. 2007;43(3):236–48.
6. Rolo A, Galea GL, Savery D, Greene NDE, Copp AJ. Novel mouse model of encephalocele: post-neurulation origin and relationship to open neural tube defects. Dis Model Mech. 2019;12:dmm040683.
7. Wang L, Ren A, Tian T, Li N, Cao X, Zhang P, Jin L, Li Z, Shen Y, Zhang B, Finnell RH, Lei Y. Whole-exome sequencing identifies damaging de novo variants in anencephalic cases. Front Neurosci. 2019;13:1285.
8. Gandy K, Castillo H, Rocque BG, Bradko V, Whitehead W, Castillo J. Neurosurgical training and global health education: systematic review of challenges and benefits of in-country programs in the care of neural tube defects. Neurosurg Focus. 2020;48:E14.
9. Estevez-Ordonez D, Davis MC, Hopson B, Arynchyna A, Rocque BG, Fieggen G, Rosseau G, Oakley G, Blount JP. Reducing inequities in preventable neural tube defects: the critical and underutilized role of neurosurgical advocacy for folate fortification. Neurosurg Focus. 2018;45:E20.
10. Kabré A, Zabsorne DS, Sanou A, Bako Y. The cephaloceles: a clinical, epidemiological and therapeutic study of 50 cases. Neurochirurgie. 2015;61:250–4.
11. Serra G, Miceli V, Albano S, Corsello G. Perinatal and newborn care in a two years retrospective study in a first level peripheral hospital in Sicily (Italy). Ital J Pediatr. 2019;45:152.
12. Forestus P: De capitis et cerebre morbis ac symptomatic, in: Observationum et curationum medicinalium, libri III. Leiden: Officina Platiniana, 1587.
13. Ntimbani J, Kelly A, Lekgwara P. Myelomeningocele – a literature review. Interdisciplinary Neurosurgery. 2020;19:100502.
14. Northrup H, Volcik KA. Spina bifida and other neural tube defects. Curr Probl Pediatr. 2000;30(10):313–332.
15. Oncel MY, Ozdemir R, Kahilogulları G, et al. The effect of surgery time on prognosis in newborns with meningomyelocele. J Korean Neurosurg Soc. 2012; 51(6):359–362.
16. Demir N, Peker E, Gülşen İ, et al. Factors affecting infection development after meningomyelocele repair in newborns and the efficacy of antibiotic prophylaxis. *Child Nerv Syst*. 2015;31(8):1355–1359.
17. Anegbe AO, Shokunbi MT, Oyemolade TA, et al. Intracranial infection in patients with myelomeningo- cele: profile and risk factors. Childs Nerv Syst. 2019; 35(11):2205–2210.
18. Balasubramaniam C, Rao SM, Subramaniam K. Management of CSF leak following spinal surgery. *Child Nerv Syst*. 2014;30(9):1543–1547.