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Original Research Article

Clinical Profile and Surgical Outcome in Tethered Cord Syndrome with Meningomyelocele in a Tertiary Care Centre

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Abstract:

Background: Tethered cord syndrome is a pathological anchoring of the distal spinal cord, which manifests as a neurological deficit. This results in pulling with movement and progressive damage to the spinal cord. Early diagnosis and proper surgical treatment are the keys to success in patients with tethered cord syndrome. Neural tube defect prevalence worldwide is 18.6/10,000 live births and is associated with substantial morbidity and mortality. In India prevalence is as high as 7.48/1000 live births. The present study focuses on reviewing the available literature and compares the data for assessing the profile of patients in our institute and study the outcome of the surgery.

Aim is to Study the occurrence, age, and sex distribution of tethered cord associated with myelomeningocele in our patient population, to analyze the symptoms and signs of tethered cord syndrome associated with meningomyelocele, and to study the postoperative results and follow up.

Methodology: This is a prospective study of all cases operated for myelomeningocele with a tethered cord in the Neurosurgery department in Government General Hospital, G.M.C. Guntur from the period of October- 2021 to December 2023.

Conclusions: The incidence of hydrocephalus is lower compared to western literature. Patients having associated hydrocephalus who do not present with raised ICP can be managed without shunt in 17% of patients. 75% of patients require VP shunt prior to definitive surgery to reduce the major risk of wound dehiscence. 8% of the patients require shunt after MMC repair. Clinical improvement in terms of motor power can be expected in 20% of patients and for bladder incontinence in 12.5%. Most of the patients with myelomeningocele belong to Category 2A of Yamada classification and aim of surgery is to reduce mortality and morbidity, and even though most of them may not improve in their deficits, it is important to stabilize the deficits and not allow the progression of deficits due to tethering of the cord. Much of the burden of neural tube defects is preventable before conception, and folic acid intervention programs should be implemented to increase the awareness and consumption of preconception folic acid to control the Neural tube defect associated mortality.

Keywords: Meningomyelocele, tethered cord Syndrome, Spinal cord defects, preconception folic acid.

Introduction

Tethered cord syndrome is a pathological anchoring of the distal spinal cord, which manifests as a neurological deficit. This results in pulling with movement and progressive damage to the spinal cord.

Early diagnosis and proper surgical treatment are the keys to success in patients with tethered cord syndrome.

Neural tube defect prevalence worldwide is 18.6/10,000 live births and is associated with substantial morbidity and mortality. In India prevalence is as high as 7.48/1000 live births [1] The present study focuses on reviewing the available literature and compare the data for assessing the profile of patients in our institute and study the outcome of the surgery.

Aim of the study:

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Methodology:

This is a prospective study of all cases operated for myelomeningocele with a tethered cord in the Neurosurgery department in Government General Hospital, G.M.C. Guntur from the period of October-2021 to December 2023.

Inclusion Criteria: All the patients admitted and underwent surgery at GGH, Guntur for tethered cord syndrome with meningomyelocele present now or before.

Exclusion Criteria: Patients with multiple congenital anomalies not amicable for surgical intervention.

On admission, detailed history, including antenatal history, was taken, and clinical examination was done. All the patients were investigated with Specific investigations like neurosonogram initially and MRI brain if hydrocephalus is present and MRI spine apart from general investigations like routine blood tests, chest x-ray, and also USG abdomen to look for associated anomalies.

All the cases were operated, preferably when the weight of the child was more than 3 kgs. Low birth weight and malnourished children were referred to the pediatrician for improvement in the nutrition and weight gain and were asked to review, an exception to this were babies with severe hydrocephalus manifesting with the sunset sign, irritability, poor cry, and feeding. This strategy was followed to reduce wound breakdown and the post-op infective complications. After the patient was evaluated and confirmed to be included in the study, informed consent was obtained from the parents. All the cases with symptomatic hydrocephalus initially underwent VP shunt and underwent 2nd surgery for repair of myelomeningocele. Prior placement of shunt was done to avoid the complication of CSF leak and strain on the repair.

Observations and Results

The total number of Spinal dysraphism patients operated at our hospital was 38. Out of the total number of cases, there were 24 cases of Meningomyelocele. Seven cases had Lipomyelecele, 3 had dermal sinus, 2 had Meningoceles without tethering. Two had Split cord malformation.Twenty four patients who satisfied the inclusion and exclusion criteria were taken. \

The patients in the study were grouped according to Age, Gender, MMC characteristics, Neurological deficits, including motor deficits and bladder involvement. Post-op complications and surgical outcome the age at presentation for 1st time was ranging from day one to five years, and the average age at presentation was two years six months.

Two patients presented with a history of previous surgery for MMC in childhood with features of secondary tethering at the age of two yrs and fourteen years. There were fifteen males and nine female children.In the antenatal period, most of them others started taking folic acid from the second or third month of pregnancy. Two of them did not take folic acid at all. None of the mothers had taken preconception folic acid.

None of the mothers gave a history of fever during the antenatal period, and none except one patient had a history of taking any drugs during the antenatal period. 1 one mother had been on thyroxin 150mcg supplement for hypothyroidism.

One child was born out of second-degree consanguineous marriage, and all others were not consanguineous. 10 of the patients were from Guntur, 6 form Krishna, four from Prakasam, four from West Godavari districts. All the cases presented with swelling or had a history of operated for swelling. 8 (33%) patients had the swelling limited to the lumbar spine. 13 (54%) patients had swelling in the lumbosacral region. 3 (12.5%) patients had only sacral region swelling. The size of the swelling varied from 2x1cms to 6x5cms.

Neurological presentation: Neurological examination revealed 19 out of 24 patients had motor weakness in the lower limb spf varying severity. Sixteen patients had weakness involving both the limbs and two patients had a unilateral weakness. Six patients did not have any motor deficits. Eleven of the patients had bladder incontinence elicited with a history of dribbling of urine while crying.

Amongst orthopedic problems, 3 had congenital talipes equino varus, two had bilateral, and 1 had unilateral CTEV on the right side. Two of the children had associated an inguinal hernia. Neuro imaging Spinal MRI and Neurosonogram were done in all the cases and Cranial MRI in patients with positive findings in neurosonogram for detailed evaluation.

Hydrocephalus was seen in 12 cases. 9 patients were symptomatic with an enlarged head and bulging AF and they underwent shunt before the repair of MMC, and three patients underwent MMC repair because hydrocephalus was not significant, AF was lax, and there was impending rupture due to thin covering of MMC.

One of the patients who underwent MMC repair developed symptomatic hydrocephalus post operatively and underwent shunt One month later, and two children had mild hydrocephalus with no increase in hydrocephalus on follow-up imaging and did not undergo shunt placement. One of these two children had colpocephaly. All the patients had

evidence of myelomeningocele with a variable degree of neural tissue in the sac. All the patients had tethering of the cord. Ten of the patients had syringomyelia. Six patients had Arnold Chiari malformation type 2.

Surgical procedure: The aim of the surgery was to excise the sac, release all the neural elements, and achieve anatomical closure. Sac was dissected carefully; all the adhesions of the neural placode to the sac were released and repositioned into the dural tube to preserve the function as much as possible. Five patients had improvement in motor functions, and three patients had improvement in bladder continence. Patients with no preoperative deficits had no deficits. One patient had progressive deterioration in motor power in the lower limbs and loss of continence and was evaluated with MRI and found to have tethered cord and underwent surgery for the same. The rest of the patients remained to have stable deficits as preoperative status.

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Post-operative complications: Pseudo Meningoceles developed in two patients. Following surgical repair, CSF leak developed in three patients, one of which improved with compression dressing in the prone position, two other children, required a reexploration and suturing in the operation theater under anesthesia. Two children had LRTI. One improved with antibiotics on pod 5, and another child had LRTI with klebsiella sepsis and improved completed after 20 days. One patient had retethering after One and a half years of primary surgery with deteriorating continence and motor power in lower limbs. The patient underwent a second surgery, post-op adhesions of neural placode, and roots to the scar were released.

Discussion

Age and sex distribution: The age group of the patients in this study was ranging from Day 1 of life to14 years. In a similar study conducted in north India by Raj Kumar et al. [2], the age group was ranging from 1st day of life to 19 years. The male: female ratio in our study was (5:3) 15:9 compared to ~5:4 (58:44) by Raj Kumar et al. with slight male predominance. As our hospital is a tertiary referral center for surrounding districts, the demographic location was taken into consideration, and apart from 10 patients from Guntur district, patients from neighboring districts like Krishna, Prakasam, and the west Godavari were treated at our hospital. Cases from Guntur were more because of the proximity of access. There was no significant variation in cases based on location in the neighboring districts.

Antenatal history was elicited in all the cases because antenatal risk factors like Nutritional deficiencies and antiepileptic, fever are known to be associated with neural tube defects, and none of the patients had a history of fever in the antenatal period or any antiepileptic. One patient was on thyroxin, which is not known to be associated with neural tube defects. One important consideration is there is no awareness of Preconception folic acid in our patient population as most of them started folic acid supplements after 2nd or 3rd month; by that time, neurulation is completed, and neural tube defects would have already occurred. Recommended daily supplement before pregnancy is 0.4mg with no risk factor and 4mg for women at high risk [3,6,7,8]

Clinical presentation: Twenty-Two patients in our series had swellings in Lumbo sacral regions, and two patients had old scars in the lumbosacral region. Out of Twenty-two patients, three patients had swelling limited to the sacrum. According to Yamada and Won classification, Sacral lesions belong to Category One and are more likely to show better results, and in our study, all three cases of sacral MMC showed good improvement of motor function post operatively. Motor weakness was seen in 79% of patients compared to 68.6% in the study by Raj Kumar et al.

Among 79% of patients with motor deficits, 8.3% had a unilateral weakness, and 66% had paraparesis. 45% of patients had Urinary incontinence comparable to 42% in the study by Raj Kumar et al 12.5% of cases had CTEV in our study compared to 32% of patients with the MMC category of Raj Kumar et al. study. The incidence of scoliosis reported in the same was 39%. However, there were no cases of scoliosis in our patients. Orthopedic deformities were comparatively less common in our patient population than in northern India.

Radiological findings: Hydrocephalus was there in 12 (50%) of cases compared to 59% in Raj Kumar et al. Among 12 patients, 9 (75%) underwent preop VP shunt, and 1(8%) underwent VP shunt post operatively due to increase in hydrocephalus. (2) 17% did not require a VP shunt in the follow-up.

In western population the prevalence of hydrocephalus in MMC is more than 80% as shown by study by Delacruz et al [4] and Bowmon et al [5] and 95% of the patients undergo at least 1 shunt revision in there 25 years follow-up. 41% (10) patients in the present study had Syringomyelia.

31.7% (26) of MMC patients in Raj Kumar et al. study had associated syringomyelia. (25%) Six patients had Arnold Chiari malformation type II. Raj Kumar et al., in their study, noted a 17% association of ACM II with MMC

Outcome: Among 79% (19) of patients with preop motor deficits, 20% (5) improved in motor power. 12.5% (3) among 45% (11) with incontinence had a variable degree of improvement incontinence. 4% (1) among the 22 patients who presented primarily during the study period, 18 months later, presented with the deterioration of motor power and urinary continence and underwent revision detethering for post-op adhesions.

Conclusions

The incidence of hydrocephalus in our population is lower compared to western literature. Patients having associated hydrocephalus who do not present with raised ICP can be managed without shunt in 17% of patients. 75% of patient's require VP shunt prior to definitive surgery to reduce the major risk of wound dehiscence. 8% of the patients require shunt after MMC repair. Clinical improvement in terms of motor power can be expected in 20% of patients and for bladder incontinence in 12.5%. Most of the patients with myelomeningocele belong to Category 2A of Yamada classification and aim of surgery is to reduce mortality and morbidity, and even though most of them may not improve in their deficits, it is important to stabilize the deficits and not allow the progression of deficits due to tethering of the cord. Much of the burden of neural tube defects is preventable before conception, and folic acid intervention programs should be implemented to increase the awareness and consumption of preconception folic acid to control the Neural tube defect associated mortality.

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