Outcome of Patients with Meningomyelocele

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تاریخ ارائه : ۸٤/٤/۲۳ تاریخ پذیرش : ۸٤/۱۲/۲٤

پیش آگهی بیماران مبتلا به میلومننگوسل

خلاصه

مقدمه و هدف : میلومننگوسل شایع ترین ناهنجاری مادرزادی سیستم عصبی مرکزی است. در این اختلال تنها نخاع درگیر نبوده، بلکه آنومالی قسمت خلفی سیستم عصبی مرکزی (بصل النخاع و مخچه) هیدروسفالی، اختلال اسفنگتر مثانه و روده و ناهنجاریهای ارتوپدیک نیز وجود دارد بنابراین درمان آنها نیاز به همکاری رشته های مختلف پزشکی دارد. هدف از این مطالعه بررسی پیش آگهی زودرس و طولانی مدت بیمارانی است که به علت میلومننگوسل تحت عمل جراحی قرار گرفته بودند.

روش کار : در این مطالعه ٤٥ بیمار که بین سالهای ۱۹۹۰ و ۲۰۰٤ عمل جراحی شده بودند از نظر یافته های فیزیکی، عصبی، عمل جراحی و عوارض آن بررسی شدند. سن والدین و سطح معلومات آنها در صورت در دسترس بودن بررسی شده و برای تجزیه و تحلیل یافته ها از روش آماری x_2 استفاده شده است.

نتایج : بیماران شامل ۱۹ پسر (٤۲٥/۲) و ۲٦ دختر (٥۷/۸٪) بودند. شایع ترین محل میلومننگوسل در ۲۷ مورد (٦۲٥) ناحیه کمری بود. بیماران که میلومننگوسل در ناحیه ساکروم و گردن داشتند درصد بالائی از عملکرد حرکتی طبیعی برخوردار بودند ($p = ۰/۰۰۰۱$). هم چنین مشخص شد که هر چه جایگزینی میلومننگوسل در سطوح بالاتری از نخاع باشد، درصد بالاتری از کنترل طبیعی اسفنگتری وجود خواهد داشت ($p = ۰/۰۰۱۳$).

نتیجه گیری : درمان بیماران با میلومننگوسل نیاز به همکاری گروه های مختلف پزشکی دارد. چون اکثر کودکان مبتلا به این اختلال، از بهره هوشی طبیعی برخوردار بوده و عملکرد حرکتی، اجتماعی خوبی دارند درمان جراحی زودرس نتیجه بخش تر خواهد بود.

کلمات کلیدی : ناهنجاریهای مادرزادی، میلومننگوسل، دیفکت لوله عصبی، طناب نخاعی.

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Introduction :

Meningomyelocele is the most common congenital anomaly of the central nervous system (CNS) [1]. It is not only a spinal cord anomaly: associated hindbrain abnormalities, hydrocephalus, bladder and bowel disturbances, and orthopedic deformities make a team effort necessary. We reviewed our patients with meningomyelocele and evaluated their early and long-term outcomes.

Patients & Methods:

In this cross-sectional retrospective research, we included 45 patients with meningomyelocele operated on between 1990 and 2004 in Qaem hospital Mashhad University of Medical Sciences. In this study, the medical records were reviewed from the aspects of neurological and physical findings, surgery performed, and complications. The Spina Bifida Neurological Scale (SBNS) was used in the evaluation of the patients. Meningomyelocele were repaired as previously described by Mclone [2].

Parental age and education were analyzed when available. The Chi-square test was used for the statistical analyses.

Results :

Age and gender

Five patients (11.1%) were operated on for the repair of meningomyelocele on the day of their birth. Five (11.1%) and 31patients (68.8%) underwent surgery within the first month of life and between I and 12 months of age, respectively. In 4 children, the surgery was performed after 1 year of age. There were 19 boys (42.2%) and 26 girls (57.8%).

Level of meningomyelocele

The lumbar region was the site of the meningomyelocele in 19 patients (42.2%). The other locations of meningomyelocele were as follows: thoracolumbar in 8 (17.7%), Lumbosacral in 5 (11.1%), thoracic in 3 (6.6%), sacral in 6 (13.3%) and cervical in 4 (8.8%) patients (Fig.1).

Neurological status

No conspicuous neurological deficit was determined in 13 patients (28.8%). A complete neurological deficit below the level of the lesion was present in 20 (44.4%) children, and 12 (26.6%) patients had incomplete neurological involvement. The patients with cervical and sacral meningomyelocele had a higher rate of almost normal motor function than those with meningomyelocele at other levels ($X2=21.287, P=0.000$). The rate of diminished and absent anal reflexes increased with progressively more caudal location of the meningomyelocele. The anal reflex was normal in all 3 children with cervical meningomyelocele. However, this difference was not statistically significant ($X2=10.652, P=0.062$).

Hydrocephalus

Only 8 (17.7%) of 45 patients followed up by us were free of hydrocephalus. In 32 patients (71.1%), hydrocephalus was treated with a cerebrospinal fluid (CSF) shunt, and 5 patients (11.1%) were considered to have arrested hydrocephalus. There was no correlation between the level of meningomyelocele and the incidence of hydrocephalus.

Psychometric tests

Wechsler Scales test was administered to 8 children aged 6 years or older. In 5 of these patients, the intelligence quotient (IQ) was within normal limits, while 3 children had a subnormal IQ. Denver developmental screening test (DDST) was administered to 9 children younger than 6 years of age, 6 of whom (66.6%) had normal development. According to both psychometric tests, normal development or IQ were detected in 11 patients of 17 patients (64.7 %) at all.

Complications

Postoperative neurological deterioration occurred in 5 patients (11.1%). Complications related to the surgical wound, such as wound dehiscence and infection, developed in 4 patients (8.8%). CSF fistula, shunt complications occurred in 24 patients. Shunt infection (20.8%) and obstruction in 19 patients (79.1%) were the most frequent complications.

Follow-up and mortality

Follow-up in the patients ranged from 1 month to 156 months (mean 78.5 months). Of these 45 children with meningomyelocele, 17 (37.7%) died, 10 of them (22.2%) within the first year of life. CNS infection was the most common cause of death (44%).

Age and education of parents

In the medical records, the ages of the patients, mothers and fathers were available in 42, 31 cases, respectively. The youngest mother was 17 years old and the oldest was 37. The ages of the fathers ranged from 19 to 42 years. In formation on the education of the mother and father was obtained in 36 and 33 patients, respectively. In the majority of cases the social and educational levels of the parents were not high (Fig.3).

Discussion :

There was a female predominance, as [pointed out in other series [3,4,5]. In 80% of the patients, the meningomyelocele was located in the lumbar or thoracolumbar region. The incidence of cervical and thoracic myelomeningoceles is usually less than 3-5% [6,3,7 ,8 ,5].

Patients with cervical and sacral meningomyelocele had a higher rate of normal motor function than those with meningomyelocele in other locations [9]. Noetzel [8] also reported a similar observation. A more caudal location was associated with a higher rate of absence of the anal reflex. Shurtleff and Dunne [10], Steinbok et al [5]. and MCLone and Naidich [1] reported rates of ambulation of 43%, 54% and 68%, respectively, in their patients. The rate of ambulation increases with more caudal location of the lesion. However, this was not statistically significant in our series. A higher location of meningomyelocele significantly increased the rate of continence of both sphincters. Social acceptable continence have been reported in 75-85% of patients in some series [1,5], but was present in under 40% of our patients. Insufficient interest and training in urology departments account for this.

McLone and Naidich [5] reported that shunt infections had a negative impact on IQ. However, Steinbok et al [5]. And Sutton et al [11]. Did not find a significant effect of shunt infection on intelligence. We also did not detect a correlation between shunt infection and intelligence. In our series, 60% of the patients had normal IQ and development. A normal IQ rate was more frequent among the patients without hydrocephalus (76%) than among those with hydrocephalus (54%). This difference was not found to be statistically significant. In McLone s series, 70% of the patients had a normal IQ [1].

In 5 patients (11.1%), meningomyelocele repair was performed on the 1st day of life. Therefore, there are problems in comprehending the importance of early repair of meningomyelocele and referral of the patients to neurosurgical center in our country. Losing the chance of early repair may lead to a deterioration of neurological damage and a higher risk of infection, in our opinion. In McLone s series [1], the incidence of ventriculitis was affected by the promptness with which the meningomyelocele was repaired. The incidence of ventriculitis was 7% following repairs performed within 48 h of birth and 37% with repairs done later than 48 h after birth. However, in the series of Brau et al.[3]. The repair of meningomyelocele did not reduce before 48 h of age did not reduce the risk of ventriculitis.

Postoperative neurological deficits worsened in 5 patients. Wound infection and dehiscence developed in 4 children (8.8%). Large skin defects can lead to difficulties in primary closure of the skin circulation, leading skin necrosis. The incidence of skin infections was 12% in McLone s series [12,1] and 22.4% in Brau et al. s series [3]. Appropriate skin flaps and techniques can decrease the wound-related complications [13, 14, 15, 16]. The shunt infection rate was 24% in our patients, 12% in McLone's [13], 5.2% in Brau et al. [3] and 25.8% in Gamache's series [19]. Although Gamache [5] reported a 4. 19% obstruction rate, shunt obstruction occurred in 19 children (51%). The revision rates ranged from 52% to 86% in some large series [5,13,22].

Mortality rates such as 14%, 15% and 18% have been reported [9, 13, 22]. There was no mortality among the patients of Gross et al. [6] and Brau et al. [3]. In this series, 37.7% of the patients died. CNS infection was the main cause of deaths. Most of the deaths (58.8%) occurred within the 1st year following surgery. Close monitoring of the

infants with meningomyelocele could decrease the number of deaths.

The majority of parents had low social and economic levels. These data may support the hypothesis that nutritional factors play a role in the development of meningomyelocele. Low folic acid intake by the mother prior to conception has been implicated [22]. Administration of vitamin and folic acid before conception markedly reduced the expected rate of incidence of neural tube defects [23,24,25,26].

Conclusion :

The management of children with meningomyelocele needs a team approach. The majority of patients can have a normal IQ and a socially acceptable degree of continence and be able to walk. These patients should be treated with aggressive therapies whenever possible. Intrauterine recognition and prevention of these anomalies should be our goal.

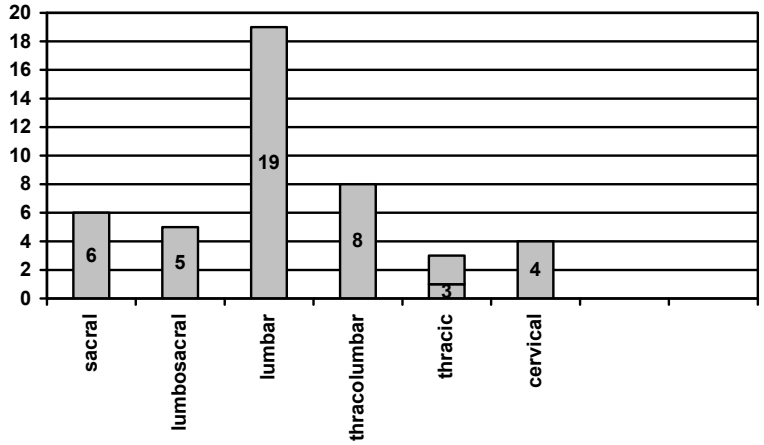

Fig .1 Locations of meningomyelocele in the45 patients

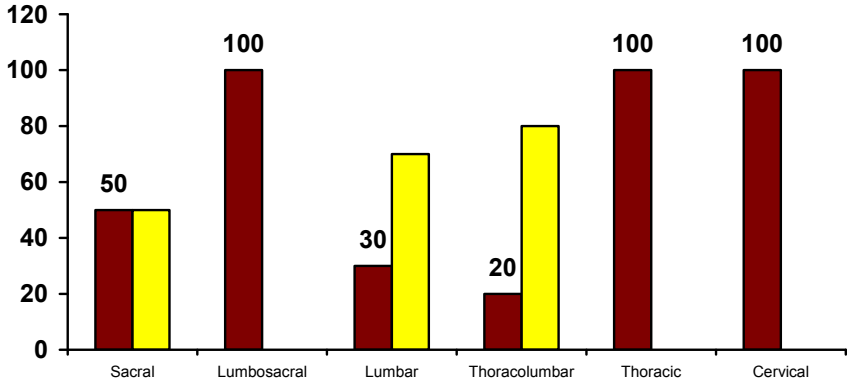

Fig 2. Bladder and bowel control in relation to the level of meningomyelocele

Table 1 : Evaluation of the patients according to the Spina bifida neurological scale

Spina Bifida Neurological Scale	No. of patients
Grade I: Normal	13(28.8%)
Grade II: Bladder and bowel problems	7(15.5%)
Grade II: Ambulatory problems	20(44.4%)
Grade IV: Nonambulatory	5(11.1%)
Grade V: Bedridden	

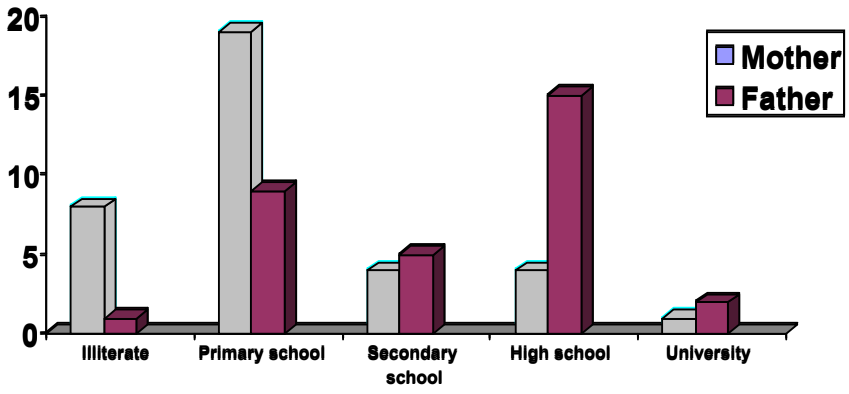

Fig. 3 : Educational status of the parents

Abstract

Introduction: Meningomyelocele is the most common congenital anomaly of the central nerves system (CNS).It is not only a spinal cord anomaly; associate hindbrain abnormalities, hydrocephalus, bladder, and bowel disturbance, and orthopedic deformities make a team effort necessary. We reviewed our patients with meningomyelocele and evaluated their early and long-term outcomes.

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Parental age and education were analyzed when available. The chiary-square test was used for the statiscal analyses.

Results: There were19 boys (42.2%) and 26 girls (57.8%). The lumbar region was the site of the meningomyelocele in 27 patients (62%).Patients with cervical and sacral meningomyelocele had a higher rate of normal motor function than those with meningomyelocele at other levels (p=0.0001).We also noted that the higher the location of meningomyelocele, the greater the control of both sphincters (p=0.0013).

Conclusion: The management of children with meningomyelocele needs a team approach. The majority of patients can have a normal IQ and a socially acceptable degree of continence and be able to walk .The patients should be treated with aggressive therapies whenever possible.

Kay Words: Congenital malformation**.** Meningomyelocele, Neural Tube Defect, Spinal Cord.

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