EVALUATION OF NEONATES WITH OPEN MYELOMENINGOCELE

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ABSTRACT

Objective: Myelomeningocele is one of the most common dysraphic disorders. The aim of our study is to present the data collected from 42 babies with myelomeningocele, followed in our neonatal intensive care unit postoperatively.

Methods: Data collected from 42 babies is evaluated.

Results: Most of the cases (95%) were either not diagnosed antenatally or had a late diagnosis. The anatomical locations of the lesions were: 71% lumbar, 17% lumbosacral, 7% thoracolumbar and 5% thoracic. Seven of the babies had complete guadriplegia and 30 cases had paraparesis in their legs. Neurogenic bladder was observed in 24 cases and they were catheterised. Urinary tract infection occurred in 9 of them. Orthopedic deformities were detected in 25 neonates. In their postoperative follow-up, sepsis was diagnosed in 86% of babies operated after 24 hours of life, while only 17% of the cases operated in the first few hours developed sepsis.

Conclusion: Because of its high morbidity and mortality, open spinal dysraphism deserves special attention. Babies with late antenatal diagnosis should be delivered in a center where they can be operated soon after birth or, should be referred as soon as possible, preferably in the first 24 hours, if they were born in another hospital.

Key Words: Myelomeningocele, newborn

INTRODUCTION

Myelomeningocele is one of the most common dysraphic disorders referred to neurosurgeons during the neonatal period (1). It is associated with varying degrees of neurologic impairment with coexisting orthopedic and urologic problems in most of the babies and postnatal follow-up deserves special attention. The aim of our study is to present the data collected from 42 babies with myelomeningocele followed in our neonatal intensive care unit postoperatively.

MATERIAL AND METHODS

This is a hospital-based study, undertaken on 42 neonates with myelomeningocele over a two-year period, between January, 1996 and December, 1997 in Marmara University, School of Medicine, Divisions of Pediatric Neurosurgery and Neonatology. All babies were examined by a neonatologist before their operation and consulted by a pediatric urologist and orthopedist as well. The existing and forthcoming problems were explained to the parents and consent was taken preoperatively. Patients were operated immediately after metabolic and hematologic stabilization. Postoperatively they were followed up in the newborn intensive care unit for 3 to 30 days depending on their clinial stability. All the babies were routinely evaluated by means of cranial and renal/bladder ultrasonography. Cranio-spinal MRI was performed to detect Chiari Type II malformation, anomalies causing tethered cord syndrome and/or hydrocephalus. Sepsis work-up was held in all cases and antibiotics were started. After initial sac closure, ventriculo-peritoneal (V-P) shunt or external drainage was performed depending on the size of hydrocephalus and the existence of meningitis/ventriculitis.

RESULTS

Thirty eight of forty two babies were referred to our hospital while four of them were inborn. All but one of the babies were terms. The characteristics of the study

group are shown in Table I. Most of the cases (95%) were either not diagnosed antenatally or had a late diagnosis. Mean postnatal age of cases on admission was 3,8±2,3 (1-6) days and 85% (36/42) were older than two days. The cases operated on the first day accounted for 15% (6/42) of all the patients. The anatomical locations of the lesions were: 71% (30) lumbar, 17% (7) lumbosacral, 7% (3) thoracolumbar and 5% (2) thoracic. Seven of the babies had complete quadriplegia and 30 cases had paraparesis. All but one of the babies had hydrocephalus and Chiari Type II malformation (98%) diagnosed by magnetic resonance imaging. A V-P shunt was performed in 77% (23/30) of the cases in the first two months. Six cases died because of intractable infections (Ventriculitis and disseminated intravascular coagulation (DIC)) (6). An external drainage system was placed in four patients with ventriculitis of whom only one had a cure, three out of four died. Three patients died in the newborn period because of sepsis and disseminated intravascular coagulation.

Catheterisation was performed in 24 cases with neurogenic bladder. Urinary tract infection occurred in nine and two of them had vesicourethral reflux (VUR). Renal ultrasonography revealed unilateral renal agenesis in one and dilation of the collecting systems in three babies. Orthopedic deformities were detected in 25 (60%) neonates. Nine of them had flexed and adducted hips, hyperextension of the knees and talipes calcaneovalgus. Pes equinovarus was detected in nine cases. Other deformities observed were scoliosis (2), kyphoscoliosis (2), pectus excavatum (2) and kyphosis (1). In their postoperative follow-up (7-30 days), sepsis was diagnosed in 86% (31/36) of babies operated after 24 hours while only 17% (1/6) of the cases operated in the first few hours, developed sepsis.

Table I. Characteristics of the study group.

Variable	
Sex (Female / Male)	20/27
Mode of Delivery (Vaginal / C-section)	8/34
Mean Gestational Age (weeks) (mean±SD) (range)	39,4±2,7 (35-41)
Mean Birth Weight (grams) (mean±SD) (range)	3049±515 (2150-4100)
Mean Head Circumference (cm) (mean±SD) (range)	35,3±2,1 (33-40)
Inborn (n) (%)	4 (10)
Outborn (n) (%)	38 (90)

DISCUSSION

Myelomeningocele is a major birth defect that is a result of failure of the neural tube to close in the developing fetus (2). It is associated with varying degrees of neurologic impairment. The anatomic level of the lesion generally correlates with the neurologic motor and sensory deficit and ranges from complete paralysis to minimal or no motor deficit (2).

The prevalence of spina bifida at birth varies greatly between countries. In Europe figures have been reported ranging between 0,26-2,5/1000 and it is still the most common major birth defect (3). A study from Turkey reported that the incidence of neural tube defects was 0,27% (4).

It has been reported that girls are more frequently affected than are boys (2). Our study group did not show such a difference.

The mortality rate for untreated infants born with myelomeningocele ranges from 90% to 100% (2). It is recommended not to withhold medical or surgical interventions, that is, initial sac closure preferably in the first 24 hours and V-P shunt placement in the perinatal period. (2) In McLone's (5) series 37% of patients with myelomeningocele showed significant motor recovery shortly after surgical closure of the back and this improvement persisted throughout the follow-up period. The same authors also reported that, delay in closing more than 72 hours after birth decreased motor function in a small percentage of children. Mean postnatal age of cases on admission was 3,8±2,3 (1-6) days and 85% were older than 2 days. The cases operated on the first day accounted for 15% of all patients, because 90% of babies were referred from different hospitals. Comparing the two groups operated in the first 24 hours and later showed that the babies in the first group had a lower incidence of infection and mortality rate as indicated in the literature (6). All but one of the babies had hydrocephalus and V-P shunt was performed in 77% of the cases.

Myelomeningoceles are usually thoracolumbar, which carry the worst prognosis (7). In our study group the most common location was the lumbar (71%) region, followed by lumbosacral, thoracolumbar and thoracic regions. There is a wide range of paralysis in the legs, from none to complete. Seven of our babies had complete quadripalegia and 30 cases had paraparesis in their legs.

The infant with spinal dysraphism should be assessed for neurogenic bladder (8). Among our cases, neurogenic bladder was observed in 57% and catheterisation was performed on all of them. Two out

of nine with urinary tract infection had VUR. Renal ultrasonography revealed unilateral renal agenesis in one and dilation of the collecting systems in three babies.

Orthopedic deformities may be present at birth. They are largely due to imbalanced paralysis of muscle groups (7). Orthopedic deformities were detected in 60% of neonates.Nine of them had flexed and adducted hips, hyperextension of the knees and talipes calcaneovalgus. Pes equinovarus was detected in nine cases. Splinting for nonsevere positional foot deformities and casting for severe malformational foot deformities are begun in the early postnatal period according to our protocol as indicated in the literature (2). Other deformities observed were scoliosis, kyphoscoliosis, pectus excavatum and kyphosis.

In conclusion, because of its high morbidity and mortality, open myelomeningocele deserves special antenatal attention. Babies with late antenatal diagnosis should be delivered in a center where they can be operated on soon after birth or if born in another hospital, should be referred as soon as possible, preferably in the first 24 hours (5). All cases operated in our hospital are followed up by a special team including a pediatric neurologist and neurosurgeon, pediatric orthopedist, pediatric urologist and physiotherapist. We believe that their long term follow-up will reveal more informational data about their long term prognosis.

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