**CASE REPORT**

**Case Report of Speech and Ambulation Ability After Five Years Therapy in a Six-Year-Old Boy with Habilitation Sixth Lumbar Spina Bifida with Meningocele Post Resection and Hydrocephalus Post VP Shunt**

Naela Munawaroh, Lisa Nurhasanah, Rahmi Isma AP

1 Physical Medicine and Rehabilitation Department, Faculty of Medicine Diponegoro University, Indonesia
2 Physical Medicine and Rehabilitation Department, Kariadi Hospital, Indonesia

**ABSTRACT**

**Introduction:** Spina bifida, a congenital spinal cord injury, is a complex disability that results from a failure of the caudal neural tube to fuse early in embryonic development. Spina bifida affects about 300,000 newborns each year worldwide. Hydrocephalus is common in children with spina bifida, developing in 80% to 90% of children with myelomeningocele. Anatomical abnormalities in which the myelomeningocele sac occurs is associated with neurological, motor and sensory deficits. They show impaired speech and ambulation.

**Case presentation:** We report a six-year-old boy was taken to the hospital by his parents because he could not stand and walk since he was born. At birth there was a lump on the lower back, and the patient also had hydrocephalus, then surgery had been performed. The patient was diagnosed with sixth lumbar spina bifida and hydrocephalus with meningocele. He had some delay in gross motor, fine motor and language development. The patient had history of seizures. He received trunk control exercise, pelvic stability exercise, positioning and handling, coordination exercises and fine motor skills to increase independence in activities of daily life. The patient obtained 4 years exercises of speaking, comprehension, vocabulary and articulation. The child also received attention and concentration enhancement exercises.

**Conclusion:** A patient with spina bifida requires a lifelong commitment by the patient, family, and the health care personnel involved in the treatment. The medical goal is to maintain stable neurological functioning throughout the patient’s lifetime. The wide range of extensive speech and ambulation impairments and long-term disabilities in patients with spina bifida present a considerable challenge to the management of these patients.

**Keywords:** ambulation, hydrocephalus, meningocele, speech, spina bifida
ABSTRAK


Kesimpulan: Pasien dengan spina bifida memerlukan komitmen seumur hidup oleh pasien, keluarga, dan tenaga kesehatan yang terlibat dalam proses pengobatan. Tujuan pengobatan yang dilakukan adalah untuk memelihara kestabilan fungsi neurologis seumur hidup. Gangguan bicara dan ambulasi serta disabilitas jangka panjang yang dialami pasien dengan spina bifida menjadi tantangan yang cukup besar dalam proses penanganan pasien.

Kata kunci: ambulasi, bicara, hidrosefalus, meningokel, spina bifida.

INTRODUCTION

Spina bifida, a congenital spinal cord injury, is a complex disability that results from a failure of the caudal neural tube to fuse early in embryonic development. This congenital condition predisposes many areas of the central nervous system (CNS) to not develop or function adequately. In addition, all areas of development (physical, cognitive, and psychosocial) that depend heavily on central functioning will likely be impaired. The clinician must therefore be aware of the significant impact this neurological defect has on motor function as well as on a variety of related human capacities.¹

Spina bifida occurs in utero and is present at the time of birth. It is generally accepted that neural tube defects (NTDs) are caused by the failure of the neural tube to close, although it has also
been suggested that a closed tube may reopen in some cases. A variety of defects results from the failure of the neural tube to properly close. Failure of closure of the neural tube may occur at several sites and the clinical types of NTDs differ depending on the site at which closure fails.\(^2\)

Statistics about the incidence of spina bifida vary considerably in different parts of the world. Spina bifida and anencephaly, the most common forms of NTDs, affect about 300,000 newborns each year worldwide. In the United States, the most recent annual prevalence estimates that 1460 babies are born with spina bifida, and the incidence is currently 2.48 per 10,000, down from approximately 7.23 per 10,000 births from 1974 through 1979 (before the folic acid mandate). Current worldwide folic acid fortification programs have resulted in a decreased incidence of spina bifida, with annual decreases of 6600 folic acid–preventable spina bifida and anencephaly births reported since 2006. There was a 31% decline in spina bifida prevalence rates in the immediate post fortification period (October 1998 through December 1999). Additionally, there was a continued decline in spina bifida prevalence rates from 1999 to 2004 of 10%. A significant relation has also been noted between social class and spina bifida: the lower the social class, the higher the incidence.\(^2\)\(^4\)

A multifactorial genetic inheritance has been proposed as the cause of spina bifida, coupled with environmental factors of which nutrition, including folic acid intake, is key. Genetic factors seem to influence the occurrence of spina bifida. Cytoplasmic factors, polygenic or oligogenic inheritance, chromosomal aberrations, and environmental influences (e.g., teratogens) have all been considered possible causes. Many studies identified an increased risk of NTD-affected pregnancy to be associated with epidemiological findings such as maternal and paternal ages and occupations, maternal reproductive history, including maternal country of birth and country of conception, nutrition, including folic acid and vitamin B12 deficiency, hyperthermia during early pregnancy, hyperglycemia or diabetes or obesity, and maternal use of medications during early pregnancy.\(^3\)\(^5\)\(^6\)

Hydrocephalus is common in children with spina bifida, developing in 80% to 90% of children with myelomeningocele. Hydrocephalus is a pathological enlargement of the brain ventricles as a result of increased amounts of CSF. Absorption of CSF is via the arachnoid villi that are associated with venous sinuses as well as lymphatics that are associated with cranial and spinal nerves. Hydrocephalus in patients with spina bifida results from a blockage of the normal flow of CSF between the ventricles and spinal canal. Excessive pressure in the ventricles exerted by CSF compresses the nervous tissue, which causes brain damage and may result in disproportionally large head size in newborns or infants. The pressure may interfere with the function of the adjacent structures and can cause a range of impairments in brain function.\(^7\)\(^9\) A ventriculoperitoneal (VP) shunt is a cerebral shunt that drains extra cerebrospinal fluid (CSF) in hydrocephalus patients. A shunt includes a ventricular catheter related to a valve and distal catheter. The distal end of a VP shunt is positioned the peritoneal cavity.\(^10\)

Several children with spina bifida have difficulty speaking clearly. They show impaired speech due to ataxic dysarthria. The damaged nerve in the spinal canal may also affect the muscle of the patients, thus they may not able to move
their legs. Some children with spina bifida have paralyzed extremity while others can stand and walk with or without any aids or assistance. The rehabilitation therapy for speech-language and ambulation impairment are important for the future of children development.\textsuperscript{11,12}

**CASE REPORT**

A six-year-old boy was taken to the hospital by his parents because he could not stand and walk since he was born. At birth there was a lump on the lower back, and the patient also had hydrocephalus, then surgery had been performed at that time. The patient was diagnosed with sixth lumbar spina bifida and hydrocephalus with meningocele. He had some delay in gross motor, fine motor and language development.

The patient started the medical rehabilitation program when he was 1 year old. He got several physiotherapy programs such as the improvement of head control, prone and rollover ability. Until the age of 2 years, he could only prone and rollover. He received posture control exercise, sitting exercise with the improvement of trunk and pelvic. The patient had speech impairment, he could only babble. He could turn his head over when called. The eye contact was normal. He ate porridge and milk without choking. He obtained speech therapy programs include voice exercise, receptive language improvement, expressive language exercise (syntax, semantics and pragmatic). The occupation therapy program includes the improvement of concentration and attention, fine motor skills to increase independence in activities of daily life (ADL) and behavior therapy. The duration of the exercise was 60 minutes twice a week.

At the age of 3 years, the patient had seizures for the first time, after which the patient had several seizures and received treatment for the seizures. On physical examination, the head was microcephaly. there was also inferior flaccid paraplegia, fair trunk control, and bilateral inferior extremity atrophy. Furthermore, his head control and hearing function were good. The MSCT examination of the head with contrast showed non-communicating hydrocephalus, signs of increased Intra Cranial Pressure (ICP). The right and left lateral ventricles appeared parallel with the Viking helmet sign, suspicious of agenesis of the corpus callosum. The area of hypodense CSF fluid density was not well defined on the left lateral periventricular medial aspect, possibly encephalomalacia. The EEG record showed epileptogenic waves in the right frontotemporal.

Currently, the patient was seizure-free, and his last history of seizures was 1.5 years ago. After the patient got 5 years of therapy, he could sit independently. There was weakness and flaccid in his both lower limbs. He used pediatric wheelchair for his ambulation. He could speak around 20 words and arrange 2-3 words into a sentence. His ability in semantic and pragmatic started to improve but it was still limited. The mean length of utterance was still limited, the articulation was not clear. The patient could understand other people’s commands well. Eating and drinking could be done independently. There was no difficulty in chewing and swallowing. Bathing and wearing clothes need help.

The patient was still wearing pampers due to impairment in defecation and micturition. Regarding complaints about defecation and micturition, doctors provided education to parents about the importance of a Clean Intermittent Catheter.
DISCUSSION

Spina bifida occulta is characterized by a failure of one or more of the vertebral arches to meet and fuse in the third month of gestation. The spinal cord and meninges are unharmed and remain within the vertebral canal (Figure 1). The bony defect is covered with skin that may be marked by a dimple, pigmentation, or patch of hair. The most common site for this defect is the lumbosacral area, and it is usually associated with no disturbance of neurological or musculoskeletal functioning. Most individuals with spina bifida occulta are unaware that they have this defect.

Spina bifida cystica results when the neural tube and overlying vertebral arches fail to close appropriately. Cystic protrusion of the meninges or the spinal cord and meninges occurs through the defective vertebral arches and produces a cystic sac. The milder form of spina bifida cystica, called meningocele, involves protrusion of the meninges and cerebrospinal fluid (CSF) only into the cystic sac (see Figure 1B).5,8,9

The spinal cord remains within the vertebral canal, but it may exhibit structural abnormalities. Clinical signs vary (according to the extent of spinal cord anomalies) or may not be apparent. This is a relatively uncommon form of spina bifida cystica. A rare but more severe form of spina bifida cystica, called myelocoele or myelocystocele, or terminal myelocystocele, is present when the central canal of the spinal cord is dilated (syringocele) and surrounded by an expanded dural sheath, producing a large, skin-covered cyst. The neural tube appears to close normally but is distended from the cystic swelling. The CSF may ceaselessly expand the neural canal. Most born with myelocystocele have no neurological deficit; however, prompt medical attention is mandatory for repair of the cyst. The more common and severe form of the spinal bifida is known as myelomeningocele, in which both spinal cord and meninges are contained in the cystic sac (see Figure 1C).5,8,9

In this case, a patient has communication problems, ambulation impairment, urinary and pelvic incontinence.
Speech and Language Impairment

The patient could speak one sentence, but it was still limited, the articulation was not clear and he liked to imitate the words of others. He had good eye contact and could understand other people’s commands well.

Speech and language impairment in this case associated with dysfluency; ataxic dysarthria (articulatory inaccuracy, prosodic excess, and phonatory-prosodic insufficiency); and slowed speech rate. The speech fluency is limited by these motor speech deficits.

Speech therapy programs of this patient include voice exercise, receptive language improvement, expressive language exercise (syntax, semantics and pragmatic). Speech refers to the production of articulation, specifically in the daily situation. Syntax involves the study of word structure, word combinations, and sentence structure. Pragmatic communication refers to the factors involved in communication. It concerns about the ability to match the type of information to the listener.2,12

Children with spina bifida have a rather different cognitive profile than typically developing children. Impairments in the cognitive profile is related to both Arnold-Chiari II malformation and hydrocephaly. The Arnold-Chiari II malformation and hydrocephaly affect the development of brain structures of the hindbrain, midbrain, ventricular system, and subcortical gray matter. These deficits lead to impairments in the cognitive domains of executive functioning, visual-spatial working memory, intelligence, language, and learning. This children are likely to have below-average intelligence, often demonstrate learning disabilities and poor academic achievement and also show moderate to severe visual-motor perceptual deficits.12,13

The impairment of intellectual and perceptual abilities has been linked to damage to the white matter caused by ventricular enlargement. This damage to association tracts, particularly in the frontal, occipital, and parietal areas, could account for the often severe perceptual-cognitive deficits noted in children with spina bifida. Lesser involvement of the temporal areas may account for the preservation of speech, whereas the semantics of speech, which depends on association areas, is impaired. The “cocktail party speech” of children with spina bifida can be deceptive because they generally use well-constructed sentences and precocious vocabulary. A closer look, however, reveals a repetitive, inappropriate, and often meaningless use of language not associated with higher intellectual functioning. Research on learning difficulties with spina bifida and hydrocephalus suggests that many of these children experience difficulties. Tasks and skills affected include memory, reasoning, math, handwriting, organization, problem solving, attention, sensory integration, auditory processing, visual perception, and sequencing.14–16

The occupation therapy program includes the improvement of concentration and attention, fine motor skills to increase independence in activities of daily life (ADL) and behavior therapy. These exercises aim to improve visual-motor perceptual deficits and concentration attention which are expected to help improve the patient’s communication skills.

Ambulation Impairment

In this case, the patient could not stand and walk since he was born. There were weaknesses and flaccid in both lower limbs, sitting resistance was also still lacking. The clinical manifestation of spina bifida is weakness, paralysis and muscle tone abnormalities. Determining the extent
of neurological impairment is not as straightforward as assumed and requires thorough and careful examination and evaluation of the infant. At birth, two main types of motor dysfunction in the lower extremities have been identified. The first type involves a complete loss of function below the level of the lesion, resulting in a flaccid paralysis, loss of sensation, and absent reflexes. The extent of involvement can be determined by comparing the level of the lesion with a chart delineating the segmental innervation of the lower limb muscles and the examination findings. Orthopedic deformities may result from the unopposed action of muscles above the level of the lesion. This unopposed pull may lead to hip flexion, knee extension, and ankle dorsiflexion contractures, depending on the level of the lesion. When the spinal cord remains intact below the level of the lesion, the effect is an area of flaccid paralysis immediately below the lesion and possible hyperactive spinal reflexes distal to that area.\textsuperscript{17,18}

Children with lower-level spina bifida present with signs and symptoms of lower motor neuron lesions including areflexia, hyporeflexia, flaccidity, or hypotonia. Children with higher level spina bifida present with signs and symptoms of upper motor neuron (UMN) lesions including spasticity. Children with Chiari II malformation, hydrocephalus, or tethered cord syndrome (TCS) may present with spasticity, which has also been reported to occur in the upper extremities of many children with shunting. Difficulties associated with flaccidity may include inability to generate muscle force, inability to bear weight on the flaccid limb, deformities, poor limb posture, and other secondary disorders such as decreased bone strength.\textsuperscript{8,19,20} There are Strengthening exercise for core muscles, upper extremity, lower extremity that perform with play activities.

Management
The surgery for spina bifida and hydrocephalus had been performed in this patient. The key early priorities in the management of spina bifida are to repair the spinal cord and spinal nerves, to protect the exposed nerves and structures from additional trauma, and to prevent infection from developing in the exposed nerves and tissue through the spinal defect. Timing of the surgical closure and repair is important and may occur prenatally or soon after birth to minimize the risk of neural damage and infection. The aim of either surgery is to replace the nervous tissue into the vertebral canal, cover the spinal defect, and achieve a watertight sac closure. This early management has decreased the possibility of infection and further injury to the exposed neural cord.\textsuperscript{1,5,8}

Management strategies in the care of shunted hydrocephalus vary. Shunt complications occur frequently and require an average of two revisions before age 10 years. The most common causes of complications are shunt obstruction and infection. Revising the blocked end of the shunt can clear obstructions. Infections may be handled by external ventricular drainage and courses of antibiotic therapy followed by insertion of a new shunting system. The problem of separation of shunt components has been largely overcome by the use of a one-piece shunting system. The single-piece shunt decreases the complications of shunting procedures. Shunt revision is usually required as the child grows by installing a larger one.\textsuperscript{7,8,21}

In this case, the patient used pediatric wheelchair for his ambulation. Pediatric wheelchair (Figure 2) prescription is necessary for children with neuromuscular disorder.
Recommendations for the wheelchair are solid seating surface through a seat pan or cushion rigidizer to prevent hip internal rotation and adduction, a solid back provides better postural support, the wheelchair frame must allow for growth potential in frame length and width, and an adjustable axle is recommended to allow for adjustability in frame height and proper positioning for propulsion.\textsuperscript{22}

Wheelchair allows the patients to improve their independence and quality of life.\textsuperscript{23} Furthermore, the patient received trunk control exercise, pelvic stability exercise, educated the parents for positioning and handling, coordination exercises and fine motor skills to increase independence in activities of daily life. Positioning should focus on normal alignment and facilitating normal development and social skills. Positioning and handling should encourage symmetry and avoid the effects of tonic reflexes such as asymmetrical tonic neck reflex. Handling should use a variety of movement and postures to facilitate functional activities and sensorimotor development. Positioning and handling should also be taught to the family. When the child is not being handled, resting positions can be used to maintain ROM and enhance development. Positioning of the lower extremities should consider the existing limitations in ROM, muscle tone, or deformities such as clubfoot or the common contractures and deformities seen in children with spina bifida. Forced leg swaddling should be avoided to minimize the risk of hip subluxation. The presence of hydrocephalus and the increased risk of developing pressure sores are other factors to consider when positioning and handling a child with spina bifida.\textsuperscript{19,24–26}

Range of Motion (ROM) Exercises is a key determinant for function throughout rehabilitation. Adequate ROM is necessary for functional excursions of the muscle and joints, and for maintaining normal posture and alignment. Sufficient ROM is also necessary to prevent the development of contractures, deformities, and
secondary disorders. ROM exercises should be performed daily, with hands placed close to the joint being moved and holding briefly at the end of the range, thereby preventing unnecessary stress to soft tissues and joint structure. Exercises should be performed carefully for the child with hip instability or with very low muscle tone to avoid subluxation or dislocation. Positions beyond neutral or excessive hip flexion should be avoided, particularly for the child with hip instability. Range should be increased slowly and without excessive force to avoid fractures that are so often experienced in paralytic lower extremities.16–18

Normal sensorimotor experience is one of the treatment goals for young infants. Sensorimotor activities facilitate the emergence of motor and developmental skills, mobility, play, and social skills. Sensorimotor activities encourage behaviors such as reaching, handling toys, rolling, and other developmental skills. Sensorimotor training involves activities that promote normal alignment, joint stability, trunk rotation, dissociation between body segments, spatial and perceptual awareness, weight bearing, and weight shifting and transition from one position to another.16,17,27

Facilitation of normal development during the stage of early child development, the primary emphasis should be on attaining good head and trunk control, eliciting appropriate righting reactions, facilitating the ability to sit, and promoting early mobility. The normal developmental sequence may guide the progression of motor activities, ultimately preparing the child to assume the upright posture. One of the first skills to develop is head control, including ability to hold the head upright and move and maintain head positions. Head control may be difficult if the head size is large due to hydrocephalus. Rolling may be difficult or delayed in some children with spinal bifida due to large head size and weakness and paralysis in the trunk and lower extremity muscles. Creeping and crawling are also delayed due to weakness and paralysis of the trunk and lower extremities, which results in the lack of ability to stabilize and bear weight on the lower extremities. Therapists may focus on the facilitation of developmental skills. For example, the child can be seated on the therapist’s lap, facing the therapist, and alternately lowered slowly backward and side-to-side. This action helps stimulate head righting and to strengthen the neck and abdominal muscles. Weight shifting in various positions and through therapeutic handling is important to enhance the development of early head and trunk control. Developmental handling may be limited by surgical interventions that limit mobility. Active movements such as reaching to toys in different directions from different developmental positions help stimulate head and trunk control and facilitate trunk elongation. Reaching activities from sitting (supported or unsupported) to toys in different directions may assist in head control, trunk elongation and rotation, and development of sitting control.15,17,26,28

CONCLUSION

Given the range of impairments seen in patients with spina bifida, there is a clear need for coordinated interdisciplinary approach. Intervention for the patients with spina bifida in this case involves wheelchair using for his ambulation. The occupation therapy program (improvement of concentration and attention, fine motor skills) in this patient is to increase
independence on his ADL. The wide range of extensive speech and ambulation impairments and long-term disabilities in patients with spina bifida present a considerable challenge to the management of these patients.

REFERENCES


24. Doering M, Doering M. Prognosis for development of early motor milestones in a pediatric patient with myelomeningocele: A case report. Univ Iowa’s Institutional Repos [Internet]. 2018; 1-14. Available at: https://ir.uiowa.edu/cgi/viewcontent.cgi?article=1044&context=pt_casereports


