



Medical Rehabilitation in Patient with Cerebral Palsy Spastic Diplegic GMFCS IV MACS I CFCS IV EDACS II

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Abstract: Children with cerebral palsy are present with three types of motor problems. The major disturbances in muscle tone, balance, strength, and selectivity are directly related to damage of the central nervous system. Treatment programs encompass physical and behavioral therapy, pharmacologic and surgical treatments, mechanical aids, and management of associated medical conditions. In physical, occupational, speech, and behavioral therapies, the goals include enhancing patient and caregiver interactions while providing family support. We reported a female, 7-year-old, with cerebral palsy spastic diplegic GMFCS IV MACS I CFCS IV EDACS II. The patient was treated with USD intensity 1 watt/cm², frequency 3.3 MHz in 5 minutes and continuous stretching in four extremity for 10 minutes each. It showed improvement from the popliteal angle assessment and MAS score, therefore, additional invasive therapy, such as botox injection, might be not needed in the meantime. Stiffness in lower extremities especially at gastrocnemius muscle was treated with ESWT energy of 0.15mJ/mm²; total shots dose 1500 shocks per each treated muscle (gastrocnemius); frequency 4 Hz. The patient presented with lumbar hyperlordotic and anterior pelvic tilt, and also tended to sway posteriorly during standing and walking, resulting poor protection reaction on the anterior side. It will be beneficial to add abdominal core and hip extensor strengthening exercise into the program. For walking aids, we considered about using anterior walker or posterior walker. However, since the patient was 4 years old and could not take complex instruction, it might become challenging to modify the exercise protocol. Regular daily stretching was beneficial for the home program and should be continued. Referral to surgery was not required at this moment due to no joint contracture.

Keywords: cerebral palsy; motor problem; gait rehabilitation

INTRODUCTION

Cerebral palsy (CP) is a paralysis of the brain due to a non-progressive lesion in the immature brain, resulting in paralysis of heterogeneous clinical symptoms, with characteristics of impaired muscle tone, tendon reflexes, primitive reflexes and postural reactions producing abnormal movement pattern as a special sign of cerebral palsy.¹ Cerebral palsy is the most common disability during childhood, with a prevalence of approximately 2–3.5 cases per 1000 live births, and a peak up to 65 per 1000 live births particularly in premature weighing.² In Indonesia, CP prevalence is 1-5/1000 live births where as there are around 5.000-25.000 births diagnosed with CP every 5 billion live births per year in Indonesia.³

Cerebral palsy is a clinical diagnosis based on a combination of clinical and neurological signs. Diagnosis typically occurs between age 12 and 24 months. The following four motor types exist but may emerge and change during the first two years of life: 1) spasticity (85%-91%); 2) dyskinesia (4%-7%), including dystonia and athetosis; 3) ataxia (4%-6%); and 4) hypotonia (2%), which is not classified in all countries. Dyskinesia, ataxia, and hypotonia usually affect all four limbs, whereas spasticity is categorized topographically as: 1) unilateral (hemiplegia) (38%) and 2) bilateral, including diplegia (lower limbs affected more than upper limbs) (37%) and quadriplegia (all four limbs and trunk affected) (24%).⁴ Jump gait is a type of gait spasticity of hamstrings and psoas is added. The child has equinus foot, genu flexum, and coxa flecta, so, the gait has a jumping appearance. Genu flexum is generated by spasticity of knee flexors, the most important being long head biceps femoris, semitendinosus, and semimembranosus. Without treatment, genu flexum becomes immobile, owing to muscles and posterior joint capsule shortening.⁵

Extracorporeal shock waves are sound pressure waves with a higher power level applied from outside the body. They are frequently used in the medical treatment of various problems. Radial extracorporeal shock wave therapy (rESWT) is an effective and safe physical therapy modality for reducing spasticity caused by upper motor neuron lesions in many conditions. Emara et al⁶ found a positive influence of rESWT on spasticity and an improvement in the motor skills of children with CP, leading to advances in the quality of their movements.

The goal of CP management is not to cure or to achieve normalcy but to increase functionality, improve capabilities, and sustain health in terms of locomotion, cognitive development, social interaction, and independence.⁷

CASE REPORT

A 7-year-old female with cerebral palsy spastic diplegic GMFCS IV MACS I CFCS IV EDACS II. From anamnesis we found that the patient's chief complaint was could not walk independently. Currently she could sit to stand with holding. According to the father, the patient could walk independently when she was 5 years old. The patient also could not speak well. Currently the patient could only speak specific papa-mama, and for other words were difficult to understand.

The patient had seizure at 1 year old and after that, she was unconscious. She was hospitalized for a month. After that incident, she could not stand up independently. She never had seizure after that. When she was 2 years old, the patient was able to walk, but after about 4-5 steps the patient sat or stopped to balance her body. The patient had experienced several times of upper respiratory infections. In this year the patient has been hospitalized two times. From birth until now, the patient was looked after by her sister and father at their home. Her main caregiver was her sister. There was no information of prenatal and perinatal histories. For postnatal, there was no history of head trauma, brain infection, and neonatal hyperbilirubinemia, except a seizure at 1 year old.

On admission she was alert, her vital signs were within normal limit, weight 11.5 kg, height 110 cm, and head circumference 44 cm (microcephalus). In posture examination we found that in standing position: head in the middle, symmetrical shoulder height, no windswept hip, no scissoring, no pelvic obliquity, inadequate hip control, inadequate knee locking, and ankle valgus.

The patient had not achieved developmental milestones for her age, she was still unable to walk independently, fine motor skills were also delayed demonstrated by the inability to hold



Figure 1. Examination on admission

properly and operate simple stationaries such as scissors or spoon. Moreover, the patient spoke in incomprehensible single words but could follow short and simple commands, and she was able to differentiate between strangers and family.

The patient had jump knee gait, in which there was equinus at the ankle, flexion at knee and hip, and anterior tilt and increased lumbar lordosis. On physical examination, hyperreflexia, clonus and spasticity were found in all extremities and even more marked increase in spasticity was found on the lower extremity extension muscles.⁷ The spasticity might contribute to the patient not being able to fully extend both knee joints and plantarflex her ankle joints, which is also explained when the patient achieved full ROM upon passive movement. In the supporting examination of this patient, electroencephalogram (EEG) was in normal limit, head CT scan result suggestive to paediatric idiopathic basal ganglia calcification; Fahr disease; ventriculomegaly; arachnoid cyst, dd/ mega cisterna magna and pelvic X-ray result was migration percentage: 20%/33.33%.

REHABILITATION PROGRAM

This patient had some rehabilitation problems, including: gait disturbance, weakness of gait muscle, spasticity, communication function disturbance, activities of daily living (ADL) disturbance, and low body weight. For comprehensive rehabilitation management this patient was given: balance exercise using walker; ESWT energy of 0.15mJ/mm²; total shots dose 1500 shocks per each treated muscle (gastrocnemius); frequency 4 Hz, standing exercise in standing frame; start with 5 minutes, while observing the clinical signs (pain, dizziness, hemodynamic). Increased frequently; proper positioning; stretching at iliopsoas, adductor hip, quadriceps, hamstring, gastrocnemius muscle; USD continuous 3 MHz, 1 W/cm², 5-minute regio hamstring left and right; stimulation for nonverbal communication, audio-visual stimulation; exercise to improve hand function with playing activity using toy; consult with nutritionist; and ambulation and mobilization exercise using walker and using solid ankle foot orthosis.



Figure 2. Comprehensive rehabilitation management

DISCUSSION

Cerebral palsy (CP) is a disorder of movement and posture that appears during infancy or early childhood. It caused by nonprogressive damage to the brain before, during, or shortly after birth. The CP is not a single disease but a name given to a wide variety of static neuromotor impairment syndromes occurring secondary to a lesion in the developing brain. The damage to the brain is permanent and cannot be cured but the consequences can be minimized. Pediatric rehabilitation is defined to improve the independence level of a disabled child functionally and psychologically, in the physiological, anatomic, and environmental restrictions and to increase the quality of life of children and their families. Rehabilitation process requires several disciplines to come together. In this process, inter- and intradisciplinary communication is a necessity and every discipline should act according to the needs of the child and family.⁸

A high number of children with cerebral palsy (CP) have spastic gait and consequently abnormalities in joint patterns. Several factors have been contributing to the lack of consensus on the spasticity effect in cerebral palsy gait and would be summarized and discussed in this chapter, e.g., spastic gait patterns are in constant evolution during the process of growth; there are still considerable limitations in the methodologies used to assess spasticity during gait; a wide range of rehabilitation strategies have been explored to control spasticity during gait. Spastic gait patterns are divided in hemiplegic (5 types) and diplegic (4 types) with the most prevalent joint abnormalities described in the sagittal plane.⁹ Equine foot is one of the most common deformities in patients with CP. Equine foot in children has many causes, including spasticity, altered motor control, muscle contractures, contractures of the ankle joint, and muscle weakness. Equine foot due to weakness substantially affects the gait of children with mild hemiparesis. Dorsiflexor muscle weakness leads to inefficient foot clearance, which may result in a staggering gait and falls.⁶ One of the problems in this patient was stiffness in lower extremities especially at gastrocnemius muscle, because of that, we gave ESWT energy of 0.15mJ/mm²; total shots dose 1500 shocks per each treated muscle (gastrocnemius); frequency 4 Hz.

Spasticity is a symptom of the upper motor neuron diseases characterized by the increase in tendon reflex as a result of the hyperexcitability in the stretching reflex, and the increase in the speed-related tonic stretching injury to the upper motor neuron decreases cortical input to the descending reticulospinal and corticospinal tracts, which causes weakness, loss of motor control, and reduction in the number of voluntarily active motor units. The reduction of these descending tracts removes the normal inhibition of the reflex arcs within the grey matter of the spinal cord, leading to a hyperactive reflex arc and spasticity. Muscle overactivity produces muscle shortening and then muscle shortening increases spindle sensitivity. Shortening of muscles may cause contracture of joint.¹⁰ The modified Ashworth scale is commonly used for clinical evaluation. This scale measures the tone intensity but does not evaluate the effect of spasticity on function. As CP is a life-lasting condition, spasticity also affects the growth of the individual, hampering both muscle and skeletal developments resulting in symmetric or asymmetric biomechanical deficiencies.¹¹ The methods and therapeutics that address spasticity and CP gait are numerous; some of them are well established for a long time with different degrees of success (orthopedic surgery, stretching, botulinum toxin A, orthotics), while some are more recent yet promising (selective dorsal rhizotomy, intrathecal baclofen, virtual reality, and transcranial magnetic stimulation).⁹

Spasticity has positive effects such as extensor tone in the limbs that helps standing, preserve muscle bulk, preserve bone density. However, it also has negative effects such as masks contraction in the antagonist, difficulty in movement, abnormal posture, difficulty in sitting and transfers, inhibition of muscle growth, that lead to contractures, difficulty in hygiene and dressing, pressure sores, and pain. In spasticity, more rapid movement of the joint increases the degree of resistance, and decreases its time of onset. Spastic hypertonia is increased by stress and during voluntary movement when co-contraction of agonist and antagonist muscles may occur. In the most severe form, the affected part is rigid in flexion or extension.¹¹

Although spasticity can affect the entire body, but it is generally worse in the lower limbs of

children with bilateral involvement and in the upper limbs of children with unilateral involvement. Spasticity of the trunk muscles can cause postural problems while spasticity of bulbar origin can result in difficulty in feeding and communication. Muscle weakness, spasticity, and contracture also result in abnormal skeletal forces which cause bone deformity as the child grows older. The common sites for contracture in CP are in the upper extremity (pronator, wrist and finger flexor, and thumb adductor); lower extremity (hip adductor-flexor), knee flexor, and ankle plantar flexor. The common sites for deformity are in the spine (scoliosis, kyphosis), hip (subluxation, dislocation), femur and tibia (internal and external torsion), and foot (equinus, valgus, varus).¹²

Management of spasticity includes oral medication, physiotherapy, orthosis, and surgery. There are a number of different dynamic occupational and physical therapy approaches, including the Bobath technique, sensory integration therapy, proprioceptive neuromuscular facilitation, and the Brunnstrom technique. Applying various techniques such as ice (cold), heat, positioning, stretching exercises and the use of orthotic devices for these purposes are considered. Cold inhibits spastic muscles, but the effect is short-lived, perhaps outlasting the application of the cold by about half an hour. Paradoxically, heat is also used for relaxation of a spastic muscle.¹⁰ The growth of the skeleton is associated with spasticity, reduced level of activity and weakness of muscles leads to muscle contractures. Muscle contractures should be evaluated and treated as conservatively as possible to prevent over lengthening and muscle weakness.¹³

While the main objectives in the treatment of spasticity vary depending on the site of spasticity, they primarily include increasing range of motion (ROM), reduction of pain and providing mobility. Spasticity has a wide range of treatment from conservative treatment (medicine, splint, physical treatment modalities, exercise etc.) to surgery. Among these, exercises are extremely important and passive stretching exercise is the major one among all the exercises in the treatment of spasticity. When passive stretching is applied, intramuscular tension increases and stimulates the Golgi tendon organs which represent a protective mechanism inhibiting the muscle spasm and providing muscle relaxation. So, stretching exercise is the basic treatment approach for spasticity. It is asserted that especially the cold or hot stretching applied before stretching increases the efficiency much more.¹²

The energy of ultrasound (US) from the modalities of physical treatment is absorbed by the tissue and converted into heat. Ultrasound treatment can be performed by continuous or pulse wave. Continuous wave is more suitable in cases when especially heating is preferred in the muscles and tendons. It is asserted that the stretching to be applied after heating will be more efficient. US heats the muscle and the tendon better than the other physical treatment modalities with a heating effect, and thus decreases spasticity owing to this effect. This is suggested to be provided by various mechanisms. The heat increase obtained by the US decreases the stretching sensitivity of the muscle spindles and the upper motor neuron stimulation. In the treatment area of US, by the continuous movement, the protein-oriented tissues such as the muscles and tendons absorb the ultrasound energy and convert it to heat. As a result, these tissues are heated more than the other ones and they get relax. The stimulation of the cutaneous nerve receptors is similar to the gate-control mechanism, some of the mechanisms are activated and the pain is reduced and the spasm is decreased. US can decrease the spasticity by causing viscoelastic modifications within the muscle. US also decreases the resistance against the passive prolongation by the stretching reflex inhibition. Also, through its such effects on the tendon and the muscles, it is asserted that US applied in a continuous wave mode is more efficient in providing the articular mobilization of the ROM exercises and stretching exercise combination Especially when it is applied together with the stretching exercise, it is suggested that the extensibility of the tendon and muscles is increased more. As a result, it is claimed that US provides relaxation in the spastic muscles locally by these various mechanisms.¹⁴

In this patient we gave USD continues with intensity 1 watt/cm², frequency 3.3 MHz in 5 minutes with continuous stretching in four extremity for 10 minutes each. As we can see that it showed improvement from the popliteal angle assessment and MAS score, therefore, additional invasive therapy, such as botox injection, may be not needed in the meantime. However, regular

daily stretching is beneficial for the home program and should be continued. Referral to surgery is not required at this moment due to no joint contracture.

For the functional assessment, the patient was classified as GMFCS IV (physical impairments limit voluntary control of movement, infants are unable to maintain antigravity head and trunk postures in prone and sitting, infants require adult assistance to roll). The Gross Motor Function Classification System (GMFCS) was developed to create a systematic way to describe the functional abilities and limitations in motor function of children with CP. Its purpose is to classify a child's present gross motor function. Five levels are used in the GMFCS from very mild to very severe. The Eating and Drinking Ability Classification System (EDACS) consists of an extensive manual and an algorithm, both necessary for sufficient comprehension of eating and drinking abilities. It was designed analogous to other functional classification systems for people with CP (e.g. GMFCS, MACS and CFCS). The EDACS can be used to classify how children and young people function.

According to anamnesis and physical examination, this patient was included in GMFCS IV, while the CFCS was IV and EDACS II was with CP eat and drink in everyday life using distinctions that are meaningful. The EDACS identifies key features of safety (can eat by herself or sometimes need help from caregiver), and efficiency (need more time to finish her food) linked with limitations to eating and drinking ability. Five distinct levels of ability are described in an ordinal scale ranging from Level I "eats and drinks safely and efficiently" to Level V "unable to eat and drink safely". This patient was concluded as cerebral palsy spastic diplegic GMFCS IV, CFCS IV, MACS I, EDACS II.

The deviation of pelvic alignment in standing position is a common problem in children with CP. Such children retain an anterior pelvic tilt due to the contracture of the iliopsoas muscle as well as weakness in the trunk flexors and hip extensors. Anterior pelvic tilt may lead to other problems such as femoral ante torsion and medial shift of the patella to the sagittal plane bisection of the knee joint. In spastic quadriplegic CP, the development of trunk imbalance alongside the related pelvic obliquity, asymmetric weight bearing and increased postural sway can influence both standing balance and walking ability. These children have malfunctioning movement patterns which will waste their energy and caused fatigue. Therefore, a major goal of movement training is reciprocal control of the pelvis by improving interplay among the abdominal obliques, rectus abdominalis, quadrates lumborum, and lumbar extensor muscles.¹⁰ Trunk control is impaired in children with CP thus influencing their functional balance. Based on GMFCS levels, Level II and Level III have a very strong correlation between trunk control and balance in children with spastic CP.¹⁵

The RCT by El Shamy et al¹⁶ involving 30 spastic quadriplegic CP subjects, showed that adding incremental core stability exercise into physical therapy program might significantly increase trunk flexors and extensors endurance and gait parameters (step length, walking speed, and time of support on the affected side) ($p < 0.05$). A pilot study conducted by Kim et al¹⁷ also proved that trunk-hip exercise significantly decreased anterior pelvic tilt angle and activation of the extensor spinae, rectus femoris, and semitendinosus during standing in spastic quadriplegia CP ($p < 0.05$). Muscles involved in hip extension were gluteus maximus muscle, with additional help from hamstring muscles. Additionally, standing supported by desk at the level above the weak abdomen might be beneficial for core muscle, proprioceptive, and balance.

The patient presented with lumbar hyperlordotic and anterior pelvic tilt. The patient also tended to sway posteriorly during standing and walking, resulting poor protection reaction on the anterior side. Weak abdominal and hip extensor muscles may be responsible for this problem. It will be beneficial to add abdominal core and hip extensor strengthening exercise into the program. However, since the patient was 4 years old and could not take complex instruction, it might become challenging to modify the exercise protocol. We suggested to simplify the exercises, and incorporate them into play/game activity, with low sets and repetitions. Sitting on an exercise ball might become a good alternative, as the patient would try to contract her core muscle in order to balance herself against movement of the ball. Thus, it would also serve as a great balance exercise.

Toddlers walk with wide, short steps. The foot strikes the ground with the whole sole, and the stance phase knee flexion is minimal. The legs are in external rotation throughout the swing phase and reciprocal arm swing is absent. The stance phase is longer in young children compared with the swing phase, and there is increased muscle activity. The gait pattern matures as the child grows older. Heel strike begins at approximately 3 years of age. The stance phase knee flexion and external rotation values approach normal limits. Step width narrows and reciprocal arm movements begin at approximately 4 years of age. Cadence, step length and speed reach adult values at approximately age 15 years. Longitudinal analyses of gait are necessary because of the tendency for gait patterns to change over time, and especially during growth.

This patient needed assisted device for mobilization. There is some option of walking aid for CP. In this case, we considered about using anterior walker or posterior walker. The anterior intestinal walker causes an increase in body weight against the walker and an increase in hip flexion during the walking gait. Posterior walkers in most CP children are the most appropriate because the best walking gait pattern and less spending on body energy consumption. Rehabilitation options are needed for CP children to walk in cases where extensor spasticity predominates. CP children have problems with posterior balance and tend to fall backwards.¹⁸

CONCLUSION

We reported a female, 7-year-old, with cerebral palsy spastic diplegic GMFCS IV MACS I CFCS IV EDACS II. The patient was treated with USD intensity 1 watt/cm², frequency 3.3 MHz in 5 minutes and continuous stretching in four extremity for 10 minutes each. It showed improvement from the popliteal angle assessment and MAS score, therefore, additional invasive therapy might be not needed in the meantime. Stiffness in lower extremities especially at gastrocnemius muscle was treated with ESWT energy of 0.15mJ/mm²; total shots dose 1500 shocks per each treated muscle (gastrocnemius); frequency 4 Hz. The patient presented with lumbar hyperlordotic and anterior pelvic tilt, and also tended to sway posteriorly during standing and walking, resulting poor protection reaction on the anterior side. It would be beneficial to add abdominal core and hip extensor strengthening exercise into the program. For walking aids, we considered about using anterior walker or posterior walker. However, since the patient was 4 years old and could not take complex instruction, it might become challenging to modify the exercise protocol. Regular daily stretching is beneficial for the home program and should be continued. Referral to surgery is not required at this moment due to no joint contracture.

Conflict of Interest

The authors affirm no conflict of interest in this study.

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