"Cerebral Palsy is not a choice, but acceptance is"

Chow Wang**1**

1Consultant
Chief, Division of Paediatric Orthopaedics
Department of Orthopaedics & Traumatology
The Duchess of Kent Children's Hospital & Queen Mary Hospital
Honorary Clinical Associate Professor
Department of Orthopaedics & Traumatology
The University of Hong Kong
Email:wchow527@ortho.hku.hk

Cerebral Palsy (CP) is caused by a non-progressive lesion to an immature brain and results in different types of neurological problems including spasticity, motor weakness, poor coordination, dystonia and ataxia. Spasticity is the commonest type of problem seen in CP.

The primary neurological damage in CP results in abnormal tones, selective control, coordination and motor imbalance. The mainstay of treatment in the first 6-7 years of age is tone management and physical therapy. The primary neurological problem is static but the muscles and bones are under the influence of abnormal muscle tone and muscle imbalance during growth, secondary problem (musculoskeletal) such as torsional deformities of long bones and contractures of muscle and joint develops with time. Contractures, joint stiffness and subluxation becomes more obvious and compromise walking after the age of 7-8 years. Orthopaedic intervention to correct these secondary skeletal deformities starts to play a more important role after this age.

This article may be cited as: Wang C. "Cerebral Palsy is not a choice, but acceptance is"[Editorial].J Pak Orthop Assoc.2020;32(1):

Gross Motor Function Classification System (GMFCS)[1] was designed to aid comparability of patients' level of mobility, expected to remain at a constant level throughout childhood. Patients are graded into one of five groups under the following general headings:

Level I - Walks without Limitations
Level II - Walks with Limitations
Level III - Walks Using a Hand-Held Mobility Device
Level IV - Self-Mobility with Limitations; May Use Powered Mobility
Level V - Transported in a Manual Wheelchair

The Classification system focuses on the functional level achieved by the patient, not on the gait patterns or the neurological status. The level a child can attain is determined by the severity of the primary neurological damage but the subsequent development of the secondary musculoskeletal deformity will increase the difficulty in walking and a child with GMFCS Level II function will drop to level III. Correction of the musculoskeletal deformity carries the best chance to bring the patient back to the previous functional level as dictated by the extent of neurological involvement.

Physical examination of children with CP is a large topic that has been covered in many texts. Training on how to perform proper and detailed assessment is needed to ensure accurate documentation of the spasticity, motor power, voluntary control, muscle contracture and range of motion of the joints. Differentiating whether the tightness is coming from the Gastronemius or Triceps surae using Silverskiold test in patient with ankle equinus or the appreciation of Hamstring shift to avoid overestimation of Hamstring tightness are examples of how proper skills in assessment will help to avoid overlengthening of the muscles. Overlengthening of Triceps surae will result in crouch gait and overlengthening of Hamstring will result in stooping or recurvatum knee.

Normal gait relies on the presence of normal neurological control of muscle and musculoskeletal system. Coordinated muscle contraction will generate the appropriate forces through different lever arms across the skeleton and produce sequence of movement necessary for normal walking. The common deformities causing lever arm dysfunction in CP are femoral antetorsion, tibial external tibial torsion and breakage of midfoot.[2] The deformities occur as result of the failure of remodeling (in femoral antetorsion), abnormal growth stimulation (in tibial external torsion) and muscle imbalance (in midfoot breakage). The long bone torsional deformity and breakage of midfoot shorten the effective lever arm in the sagittal plane and has detrimental effect on knee extension. Lever arm dysfunction is a common cause in knee flexion deformity or crouch gait in CP.

Historically CP was classified on the anatomic pattern of the disability, i.e. hemiplegia, diplegia or

quadriplegia. It can also be classified as to whether there is pyramidal or extra-pyramidal involvement. The former implies injury to the cortical centres resulting in spasticity which may benefit from orthopaedic intervention. Extra-pyramidal involvement implies injury to the basal ganglia giving rise to athetoid or dystonic movement disorders or to the cerebellum, resulting in ataxia.

The first classification of gait pattern in diplegic CP was described by David Sutherland[3] in 1992. He described four patterns of gait abnormalities, crouch, recurvatum knee, jump knee and stiff knee, based on the deformity of the knee joint during walking. Rodda and Graham[4] devised a simple classification of 5 sagittal gait patterns namely true equinus, jump knee, apparent equinus, crouch gait and asymmetric gait, based on a combination of pattern recognition and kinematic data in 2004. The possible etiology and pathoanatomy were explained and interventions were suggested in both classifications. Though not all patients can be conveniently classified into one of the patterns described, recognition of this gait patterns will help to understand the causes of the specific gait problem and guide the appropriate intervention.

While the clinical examination provides important information, it provides mostly static measurements. Dynamic measures of joint motion (kinematics and kinetics) can only be achieved with 3D instrumental gait analysis. It is considered the gold standard in objective assessment of gait for outcome measures. It allows recognizable patterns to be classified and facilitates decision making.The 3D instrumental gait analysis is gaining popularity and has been shown to be a useful tool both in surgical planning and in assessing outcome,[5,6,7] but there is still little evidence in the literatures to support that the decisions based on 3D gait analysis leads to a better outcome.[8] More high quality studies are needed in this area.

Historically limb deformities and abnormalities of gait in patients with cerebral palsy were corrected with numerous small operations over a number of years, often referred to as 'birthday syndrome'. Since the introduction of 3D instrumental gait analysis, emphasis has been placed on correcting multiple deformities at one operative session, also known as Single Event Multilevel Surgery (SEMLS). SEMLS is now considered the standard of care in treating children with CP and aims to correct gait deviations. The procedures are tailored to the patient's need according to the comprehensive assessment and the deformities on both lower limbs are corrected under one operative session. It has been shown that SEMLS allows the patient to spend less time in hospital and require one single period of rehabilitation. The common procedures are muscle tendon lengthening, tendon transfer, rotational osteotomies and joints stabilization to correct the lever arm dysfunction. Indications of the surgical procedure has been outlined in different texts.[9] A systematic review on SEMLS has shown that significant improvement in gait are seen in majority of the studies.[10] A prospective study by Thomason and Selber[11] has shown that SEMLS results in significant improvement in gait and function. The changes were maintained for at least 5 years after surgery but follow-up procedures for fine tuning are frequently required. These are usually relatively minor procedures that require shorter hospitalization and rehabilitation.

Management of diplegic cerebral palsy is challenging and requires multidisciplinary approach. Comprehensive preoperative assessment, individualized surgical plan with SEMLS and intensive rehabilitation is the key to achieve a favorable outcome.

Conflict of Interests: None

Grants/Funding: None

REFERENCES

1. Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. Dev Med Child Neurol. 1997;39(4):214-23.
2. Theologis T. Lever arm dysfunction in cerebral palsy gait J Child Orthop.2013; 7:379-382.
3. Sutherland DH, Davids JR. Common gait abnormalities of the knee in cerebral palsy. Clin Orthop.1993;288:139-47.
4. Rodda JM, Graham HK, Carson L, Galea MP, Wolfe R.Sagittal gait patterns in spastic diplegia J Bone Joint Surg Br.2004;86-B:251-8.
5. Gage JR. The identification and treatment of gait problems in cerebral palsy. In: Clinics in Developmental Medicine. 2nd ed, vol. 18. London: Mac Keith Press, 2009. XIV, p 644.
6. Gage JR, Novacheck TF. An update on the treatment of gait problems in cerebral palsy. J Pediatr Orthop.2001; 10: 265-74.
7. Novacheck TF, Stout JL, Gage JR, Schwartz MH. Distal femoral extension osteotomy and patellar tendon advancement to treat persistent crouch gait in cerebral palsy. Surgical technique. J Bone Joint Surg Am. 2009; 91(2): 271-86.

8. Narayanan, Unni G.The role of gait analysis in the orthopaedic management of ambulatory cerebral palsy.Current Opinion in Pediatrics.2007;19(1):38-43.
9. Gage JR, Koop SM, Novacheck TF, editors. The identification and treatment of gait problems in cerebral palsy.2nd ed, MacKeith Press; 2009.
10. McGinley J, Dobson F, Ganeshalingam R, Shore BJ, Rutz E, Graham HK. Single Event Multilevel Surgery for children with cerebral palsy: A systematic review. Developmental Medicine and Child Neurology.2012;54:117-28.
11. Thomason P, Selber P, Kerr GH. Single Event Multilevel Surgery in children with bilateral spastic cerebral palsy: A 5 year prospective cohort study. Gait and Posture. 2013;23-28.