Ambulant cerebral palsy

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Abstract

Cerebral palsy (CP) is the result of a non-progressive lesion or injury to the developing brain and has multiple causes and manifestations. This non-progressive lesion leads to progressive limb problems due to imbalance in muscle tendon unit (MTU) structure and function across joints. This leads to dynamic problems during movement and with posture that may progress over time to become fixed. Abnormal movement and posture result from contractures, lengthening and problems with tone in MTUs. Torsional problems, contractures and joint instability lead to lever arm dysfunction that has a detrimental effect on gait.

This article aims to summarize the aetiology and pathophysiology of cerebral palsy, improve understanding of gait in the context of muscle imbalance and lever arm dysfunction, and describe an approach to the assessment and management of these patients.

Keywords ambulant; cerebral palsy; gait; lever arm; movement and posture; muscle imbalance

Introduction

Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitation. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication and behaviour, by epilepsy and by secondary musculoskeletal problems.[1]

It is the result of a non-progressive lesion or injury to the developing brain and has multiple causes and clinical manifestations.[2]

This consensus on the definition of CP has evolved alongside our understanding of the condition since WJ Little originally described the condition in 1862,[3] and recognizes the large variety of issues that patients and families experience, as well as the musculoskeletal problems focussed on by orthopaedic surgeons.

Patients with CP vary widely in their functional and cognitive abilities, but are united by the 'clinical and functional onset of symptoms in early development, the high probability that the symptoms have an effect on the whole life course and the current lack of a definitive cure'.[2]

This article gives the general orthopaedic surgeon a structured approach to understand the lower limb manifestations of cerebral palsy and how these can be identified and treated by exploring the following key concepts:

The permanent, non-progressive brain lesion of CP leads to progressive lower limb problems due to imbalance in muscle tendon unit (MTU) structure and function across joints. This leads to dynamic problems during movement and posture that may progress over time to become fixed. These fixed problems may be present within MTUs, bones, or joints. MTUs often shorten, leading to contractures, and the antagonistic MTU may lengthen. Abnormal forces may cause torsional abnormalities within growing bones. They may also result in progressive joint subluxation and dislocation. These dynamic and fixed problems lead to gait abnormalities, which must be interpreted with knowledge of defined types of 'lever arm dysfunction.'

Clinical assessment of the child with cerebral palsy is complex and requires a structured approach. The reader should remember that cerebral palsy often has a predilection for biarticular muscles, that therefore require special clinical tests.

Walking is the essential functional currency of all lower limb orthopaedics. Gait is by its very nature a dynamic process and difficult to understand through conventional texts. Gait should not be thought of using the conventional list of orthopaedic 'gait patterns', nor by describing the 'phases' or 'rockers'. Instead, a functional assessment of gait is explained that will stimulate an understanding of gait not just in CP, but in all clinical settings.

Treatment may be non-surgical including multidisciplinary therapy and medical treatments, or surgical. Surgical treatments may be soft tissue or bony and these modalities are often used in combination. There is a currently poorly defined role for neurosurgical intervention. The days of 'birthday surgery' with a childhood spent in hospital undergoing annual procedures and continuous rehabilitation are behind us. Single event multi-level surgery (SEMLS) aims to return the child to function and family life for a larger proportion of their childhood, but requires an exacting approach in terms of accuracy, organization and effort from the patient, family and multidisciplinary team.

Epidemiology

CP has a prevalence of between 1.5 and 3 per 1000 live births worldwide,[4–7] a figure that has remained relatively static probably because of improved survival in premature infants.[8]

Prematurity is very strongly associated with CP, and in infants born before 28 weeks gestation is 50 times the risk of those born at term,[9] yet paradoxically most patients with CP are born at term.[10] The association of low socioeconomic status with preterm birth and low birth weight, as well as the increased risk of Rhesus disease, kernicterus and iodine deficiency leads to a higher prevalence in developing countries.[2]

Birth asphyxia is often thought to cause CP and is an area associated with significant anxiety for parents and clinicians. In fact, around 75% of CP is thought to be due to prenatal causes, with perinatal hypoxia responsible for between only 6–8% of cases. Postnatal infection or injury contributes in 10–18% of cases, however in 30% of patients there is no identifiable cause (Figure 1).

In 90% of cases, CP results from injury to healthy brain tissue rather than abnormal brain development.[11] Inflammation has been suggested as a final common pathway for all these

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Aetiology & Risk Factors for Cerebral Palsy

Prenatal (75%)	Perinatal (8%)	Postnatal (17%)
Prematurity	Hypoxia	Infection
Inflammation	Hypoglycaemia	Injury
Infection (ToRCH)	Jaundice	
Ingested toxins	Infection	
Genetic syndromes		
Congenital brain malformation		
Metabolic conditions		

(ToRCH = Toxoplasmosis, Rubella, Cytomegalovirus, Herpes simplex)

Figure 1 Prenatal, perinatal, postnatal.

causative mechanisms, with pro-inflammatory cytokines leading to the permanent brain injury.[12,13]

The type and location of brain lesion sustained may be predicted in some cases by the causative mechanism and the age at which it affects the patient. Understanding of the pathophysiology underlying the various subtypes of CP is incomplete however, and it is not possible to predict the clinical manifestations of these insults with accuracy.[14–17]

Prophylaxis and prevention of CP is a vital area of research, and there is promising evidence that using interventions such as magnesium sulphate, melatonin, whole-body cooling and steroids as well as secondary preventative strategies to reduce prematurity may reduce the risk of developing CP in at risk patients.[2]

Classification

The classification of CP is challenging, as CP is a group of conditions rather than a single disease. Because of this, a group of classification systems is required.

Classification is therefore achieved by assessing the topographical distribution, neuromuscular phenotype and motor function.

In describing topographical distribution, the Surveillance of Cerebral Palsy in Europe (SCPE) definitions should now be used,[18] with unilateral CP incorporating the terms monoplegia and hemiplegia, while bilateral CP is used instead of the previously used terms diplegia or triplegia. Quadriplegic or tetraplegic CP is now more correctly termed total- or whole-body CP (Figure 2).

Neuromuscular classification aims to describe the type of movement disorder that predominates. These are variable however and may vary or coexist in the same patient over time. The broad types are spastic, hypotonic, dyskinetic or mixed. These will be discussed further in the 'Movement & Posture' section.

The Gross Motor Function Classification System (GMFCS) is an age dependent classification system that divides children with CP into five levels according to their gross motor abilities. It is reliable and valid, can predict long term function and helps guide management. GMFCS I & II patients are ambulatory. GMFCS IV & V are non-ambulatory. GMFCS III represents a transition area between these broad groups, where the ability to walk any distance unaided may be lost without careful management (Figure 3)

Disorders of movement & posture

For most patients, and all orthopaedic surgeons, CP is predominantly a disease that affects movement and posture.

Movement and posture are affected by problems relating to tone, weakness, basal ganglia problems, poor selective motor control, muscle imbalance and balance problems.

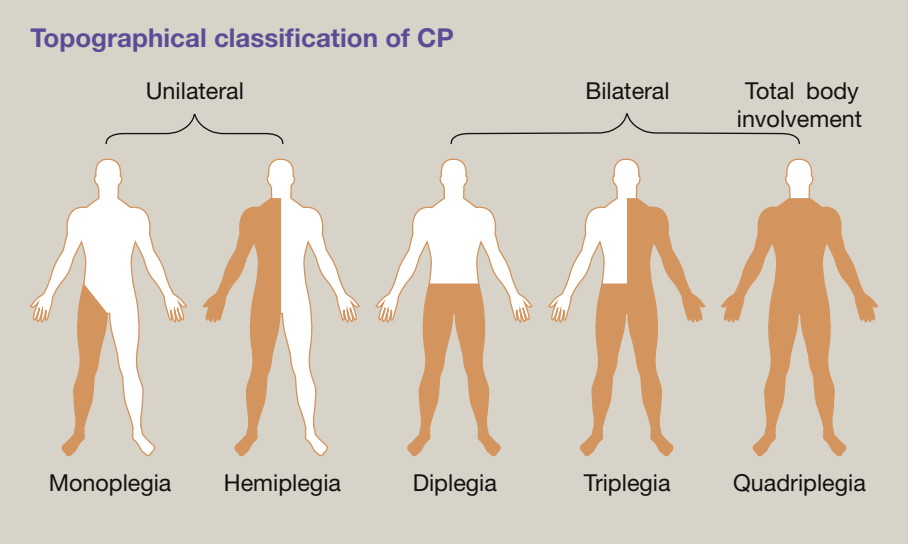

Figure 2 Topographical distribution with SCPE descriptors.

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