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INTERNATIONAL DEFINITIONS OF CEREBRAL PALSIES (REVIEW)

Cerebral palsy is a non-progressive disorder of movement originating from neural lesions in the perinatal period, and is associated with a wide range of common comorbidities. The average age for diagnosis has been around 18 months; recent evidence has suggested that cerebral palsy can be detected as early as three to four months using tests such as Prechtl's Qualitative Assessment of General Movements and medical resonance imaging. At the time being, 27 existing surveillance programs on cerebral palsy apply five definitions of cerebral palsy with 46% of surveillance programs using more than one definition. As analysis showed, the definition suggested by Surveillance of Cerebral Palsy in Europe is most cited (63%), followed by the definition of Rosenbaum et al. 2007 (54%). Therefore, several most widely cited CP definitions will be outlined in this short review article.

Keywords: cerebral palsies, definition, early diagnosis, neuroimaging

Introduction.

Cerebral palsy is a non-progressive disorder of movement originating from neural lesions in the perinatal period, and is associated with a wide range of common comorbidities. Many of these respond well to interventions in early childhood, when brain plasticity is at its greatest and developmental trajectories can be altered with maximal benefit into adulthood. In large research networks, investigators have successfully decreased the age at cerebral palsy diagnosis to 19 months [1]. However, in most clinical settings the age for diagnosis of cerebral palsy is on average two years or beyond. Late diagnosis of cerebral palsy is associated with worse long-term function and participation, parental dissatisfaction, and higher rates of mental health conditions, including depression [2]. The average age for diagnosis has been around 18 months; recent evidence has suggested that cerebral palsy can be detected as early as three to four months using tests such as Prechtl's Qualitative Assessment of General Movements and medical resonance imaging [3]. Today, cerebral palsy is still a clinical description, but registries and surveillance programs, such as those in Australia, the United Kingdom, and Europe, highlight five key elements that reflect the core features of cerebral palsy, provided in definitions, and proposed by the Surveillance of Cerebral Palsy in Europe (SCPE): (i) it is an 'umbrella term'; (ii) it is permanent but not unchanging; (iii) it involves a disorder of movement, posture, motor function, or a combination; (iv) it is due to a non-progressive interference, lesion, or abnormality; and (v) the interference, lesion, or abnormality arose in the developing or immature brain [4]. Nevertheless, most existing definitions also fail to acknowledge the very clinically pertinent fact that CP is not a single disease with a clear etiology, such as Down syndrome and Duchenne muscular dystrophy, but is instead a collection of often diverse movement disorders, each of which may require different intervention strategies [5]. Particularly, the following 4 motor types exist but may emerge and change during the first 2 years of life: spasticity (85%-91%); dyskinesia (4%-7%), including dystonia and athetosis; ataxia (4%-6%); and hypotonia (2%), which is not classified in all countries (ACPR, 2016). 2 Dyskinesia, ataxia, and hypotonia usually affect all 4 limbs, whereas spasticity is categorized topographically as unilateral (hemiplegia) (38%) and bilateral, including diplegia (lower limbs affected more than upper limbs) (37%) and quadriplegia (all 4 limbs and trunk affected) (24%) [6]. Comorbidities and functional limitations are common and disabling, including chronic pain (75%), epilepsy (35%), intellectual disability (49%), musculoskeletal problems (eg, hip displacement) (28%), behavioral disorders (26%), sleep disorders (23%), functional blindness (11%), and hearing impairment (4%) [6]. This reality underscores the importance of developing a classification system that differentiates these disorders more accurately for both clinical and research purposes. According to study by Goldsmith and colleagues, 27 existing surveillance programs on CP apply 5 definitions of CP with 46% of surveillance programmes using more than one definition. The SCPE definition was most cited (63%), followed by that of Rosenbaum et al. 2007 (54%) [7]. Therefore, several most widely cited CP definitions will be outlined in this short review article.

From the mid-1940s the founding fathers of the American Academy for Cerebral Palsy and Developmental Medicine (Carlson, Crothers, Deaver, Fay, Perlstein, and Phelps) in the USA, and Mac Keith, Polani, Bax, and Ingram of the Little Club in the UK, were among the leaders who moved the concepts and descriptions of CP forward, and caused this condition to become the focus of treatment services, a advocacy, and research efforts [7]. Pertinent material was reviewed at an international symposium participated in by selected leaders in the preclinical and clinical sciences. Suggestions were made about the content of a revised definition and classification of CP that would meet the needs of clinicians, investigators, and health officials, and provide a common language for improved communication. With leadership and direction from an Executive Committee, panels utilized this information and have generated a revised Definition and Classification of Cerebral Palsy [8].

Nowadays, the definition by Bach became a classic and is still widely cited:

It stated that CP is 'a disorder of movement and posture due to a defect or lesion of the immature brain. For practical purposes it is usual to exclude from cerebral palsy those disorders of posture and movement which are (1) of short duration, (2) due to progressive disease, or (3) due solely to mental deficiency.' [9] The label does however encompass a variety of syndromes and some, therefore, prefer the term **cerebral palsies**. The heterogeneity of disorders covered by the term CP, as well as advances in understanding of development in infants with early brain damage, led Mutch and colleagues to modify the definition of CP in 1992 as follows: 'an umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of development' [10]. This definition continued to emphasize the motor impairment and acknowledged its variability, previously underscored in the Mac Keith and Polani definition; it also excluded progressive disease, a point introduced in Bax's annotation [11].

Another most widely used definition was suggested by Mutch and colleagues in 1992, resulting in a further revised definition to underline the heterogeneity of the condition: 'an umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of development'. Notably this annotation also included a revised Swedish classification system which, whilst still not perfect, offered simplicity as its major asset. The three neurological categories were spastic, ataxic, and dyskinetic; these were subcategorized in mixed ways as hemiplegia, tetraplegia, or diplegia for spastic cases; as either diplegic or congenital for ataxic cases, and as either mainly chorioathetotic or mainly dystonic for dyskinetic cases. Whilst noting that at the time it remained beyond their capability, the authors resuscitated the yearning for an aetiologically-based classification system [12].

The Gross Motor Function Classification System (GMFCS) was developed in response to the need to have a standardized system for classifying the severity of movement disability among children with CP [13]. Previous descriptive systems had included three levels, such as: (1) mild, moderate, or severe; or four levels such as (2) nonambulatory or physiological, household and community walkers; and (3) the Evans system: not walking, restricting lifestyle, functional but not fluent, or walks fluently. A five level description of children's ambulatory ability was reported by Hutton et al. in their study of factors affecting life expectancy, though they collapsed the data into only two categories of 'walking' and 'not walking' for their analyses [14]. It should be underlined, that no evaluation of the validity and reliability of any of these systems until the development of the GMFCS took place. The GMFCS describes movement ability of children with CP in one of five ordinal levels. The GMFCS currently includes descriptions of children's abilities for each level across four age bands: less than 2 years, 2 to 4 years, 4 to 6 years, and 6 to 12 years, with an adolescent age band currently under development. Children in Level I can perform all the activities of

their age-matched peers, albeit with some difficulty with speed, balance, and coordination; children in Level V have difficulty controlling their head and trunk posture in most positions and achieving any voluntary control of movement. The GMFCS has now become the principal way to describe the severity of motor disability for children with CP. The system has had good uptake internationally and across the spectrum of health care professions for use in research and clinical practice by providing a system for clearly communicating about children's gross motor function [14].

To move the scientific study of CP forward the recent definitions and classifications proposed by SCPE and Bax's group actually perform in practice were re-considered. Particularly, pertinent material was reviewed on July 11–13 at an International Workshop on Definition and Classification of Cerebral Palsy in Bethesda, Maryland, July 11–13 2004, co-sponsored by the United Cerebral Palsy Research and Educational Foundation in Washington and the Castang Foundation in the UK, with special support provided by the National Institute of Health/National Institute of Neurological Disorders and Stroke, 2004. At the workshop, it was agreed that the concept 'cerebral palsy' should be retained. Suggestions were made about the content of a revised definition and classification of CP that would meet the needs of clinicians, investigators, health officials, families and the public and would provide a common language for improved communication, so CP definition was suggested:

Cerebral palsy (CP) describes a group of disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, cognition, communication, perception, and/or behaviour, and/or by a seizure disorder [11].

The task of the participants (listing follows) was to revisit and update the definition and classification of cerebral palsy in light of emerging understanding of developmental neurobiology and changing concepts about impairments, functional status and personal 'participation'. Reassessment of the definition of CP was prompted by a host of factors: changes in delivery of care to children with disabilities; recognition that children with slowly progressive inborn errors of metabolism can present with motor difficulties that are indistinguishable from those of children with non-progressive disease; increased availability of high-quality brain imaging to identify impairments in brain structure; acknowledgment that developmental motor impairment is almost invariably associated with a range of other disabilities; and increased understanding about associated antecedents and correlates of CP. The Executive Committee for the Definition of Cerebral Palsy (CP) suggested further: this definition describes as a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. 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 [12]. According to 2007 report this definition was further revisited: "Cerebral palsy is a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain." [15] Cerebral palsy is a clinical diagnosis based on a combination of clinical and neurological signs. Diagnosis typically occurs between age 12 and 24 months [16, 17].

Neuro-imaging findings as a tool to classify CP.

Until recently, correlations between neuroimaging findings and clinical presentation in cerebral palsy were weak. However, advances both in imaging technology and in quantitative motor assessments are changing this picture. The goal of categorizing all patients based on specific neuroimaging findings will require more development before implementation. The recommendation of the American Academy of Neurology to obtain neuroimaging findings on all children with cerebral palsy should be followed whenever feasible. Imaging using various imaging modalities shows pathology in 77%, when computed tomography (CT) is used and in 89% when magnetic resonance imaging (MRI) is employed [18]. Neuroradiology is capable of defining different kinds of brain pathology including various congenital malformations and different destructive lesions in white and grey matter [19]. Although timing of an insult is the most important factor in determining the pattern of pathology, the duration and severity of the insult are other important factors. Thus profound asphyxia causes lesions different from those due to partial hypoxia in the mature brain but also in the immature brain before 34 gestational weeks [20]. Timing and pathology: The finding of a congenital malformation by MR is usually indicative of an injury during the first half of the pregnancy. Detailed classification of the malformation may further limit the period during which the insult has operated. An abnormality of cleavage, e.g. holoprosencephaly is a very early lesion, 4th to 6th week, while an abnormality of cortical organization, e.g. polymicrogyria is an example of a very late lesion which may occur as late as 20 gestational weeks or later, depending on specific type [21]. Neuroradiological demonstration of primary white matter damage, e.g. periventricular leukomalacia (PVL) or periventricular haemorrhagic infarction (PVH) represents residual from insults operating between 24 and 34 gestational weeks. While the lower limit, 24 weeks, may be difficult to define, it appears as if the later limit 34 gestational weeks is unusually well defined [22]. It is not difficult to find statements in the literature, textbooks in particular, saying that PVL can occur even later than 34 gestational weeks. However, the scientific support for this opinion is weak and most reports refer to cases in which the findings were detected and the diagnosis of PVL made after a full-length pregnancy but without solid evidence when the damage indeed occurred. When found in a neonate born at term, PVL should be considered as having occurred in utero. Systematic use of neuroimaging in populations at risk for developing CP have shown that children born preterm have neuroradiological findings of PVL in 32% while only 9% had CP [23]. Jacobson and her group have shown that children with PVL may have significant symptoms with visual cognitive defects from their brain injury even without motor deficits. At the same time precise relationships between morphological lesions and motor disability has been shown using sophisticated analysis of conventional and functional MR. In CP, the pattern of injury to a child's brain is believed to be influenced by neural development, vascular anatomy, and the many other factors that affect injury and recovery [24]. Neuroimaging studies suggest that there are particular patterns of injury that occur with relatively high frequency, including periventricular white matter injury, cerebrovascular occlusion, and selective neuronal injury [25].

Discussion.

It is increasingly apparent that cerebral palsy may result from the interaction of multiple risk factors, and in many cases, no identifiable cause may be found. Therefore, while every reasonable effort should be undertaken to investigate causes or causal pathways, clear-cut categorization by cause is unrealistic at the present time. It is possible that by looking further downstream from putative cause to common mechanisms of injury, and by grouping cases on that basis, a more salient method of classification may be developed. For the present, timing of insult should only be noted when reasonably firm evidence indicates that the causative agent, or a major component of the cause, was operative in a specific time-window, as for example, with post-natal meningitis in a previously well infant. While recording adverse events in the prenatal, perinatal and postnatal life of a child with CP is necessary, clinicians should avoid making the assumption that the presence of such events is sufficient to permit an etiologic classification that implies a causal role for these events in the genesis of CP in the affected individual. In many individuals with cerebral palsy, other impairments interfere with the ability to function in daily life, and may at times produce even greater activity limitation than the motor impairments that are the hallmark of cerebral palsy. These impairments may have resulted from the same or similar pathophysiologic processes that led to the motor disorder, but they nonetheless require separate enumeration. Examples include seizure disorders, hearing and visual problems, cognitive and attentional deficits, emotional and behavioral issues, and later-developing musculoskeletal problems. These impairments should be classified as present or absent, and if present, the extent to which they interfere with the individual's ability to function or participate in desired activities and roles should be described. In concurrence with the SCPE recommendation, the presence or absence of epilepsy (defined as two or more afebrile, non-neonatal seizures) be recorded, and IQ, hearing and vision be assessed [4]. While SCPE provides terminology for describing different degrees of cognitive, hearing and visual impairment, the IQ score, corrected vision in each eye, and decibel loss (if any) in each ear be recorded whenever this information is available [4]. Standardized instruments are available to measure IQ, vision and hearing, and categories describing specific levels of dysfunction (e.g., visual impairment, profound hearing loss, mild mental retardation) have come to be generally accepted.

Another definition, provided by ICF and WHO.

Conceptual advances regarding health are reshaping terminology, so the concept of disability is giving way to a health status construct in which biological, social, and personal attributes determine activity and participation in society. These concepts are embodied in the International Classification of Function, Disability and Health (ICF) developed by the World Health Organization [26]. The ICF is linked to causes of impairments and disabilities through the International Classification of Diseases (ICD). CP, as captured in the International Classification of Diseases (ICD)-10 with code (G80) [27]. There are four 'components' to the classification: Body structure and function, Activity, Participation, and Environmental factors. It defines 'Participation' as involvement in life situations [27]. This concept applies to all people, not just those with disabilities. It has positive, rather than negative, connotations and the difficulties are understood to reside in the interaction between the individual and their environment and not in the individual alone. The ICF recognizes that improvement may be achieved through manipulation of a child's environment and therapy requiring a change in the child's body. Therefore, the classification is in agreement with the social model of disability. The ICF also recognizes the importance of Quality of Life (QoL), a person's subjective account of how they feel about their life, including their view of their own Participation. There are now instruments such as KIDSCREEN, KINDL, TACQOL, and PEDSQL which are capable of capturing this subjective QoL in childhood. Therefore, a classification of children with CP should have clear classification on CP type, associated impairments, functional effect across trunk and limbs, child's participation as well as the child's quality of life.

ACPR definition of CP.

The Gross Motor Function Classification Scale (GMFCS) was developed to group children with CP into five levels of functional mobility based on the key function of ambulation [13]. This outcome measure is currently being used in a large randomized clinical trial for the primary prevention of CP (The 'BEAM' trial – Beneficial Effects of Antenatal Magnesium) [28]. First, the presence or absence of CP at age 2 is determined by definite findings in any two of the following areas: (1) delay in motor milestones; motor quotient of 70 or less; (2) abnormalities of tone, deep tendon reflexes, co-ordination and movement; and (3) aberration in primitive reflexes, positive support reflex, tonic labyrinthine reflex, and/or postural reactions. If CP is determined to be present, then the GMFCS is used at the final visit at age 2 years to determine level of severity of CP. In order to incorporate hand and arm function, it has been amended in this trial by adding the ability to be able to grasp and release with both hands as a requirement for scoring above level III [29]. In Australia, in its turn, any child acquiring a motor disorder as a result of a brain-damaging event before the age of 5 years is considered to have CP [16]. However, those whose neurological impairment follows a well-documented causal event after the age of 28 days and before the age of 5 years are grouped separately as postneonally acquired CP. The age of 28 days defines the end of the neonatal period and usually differentiates events related to gestation and delivery from those largely independent of it. Summary of additional criteria: To summarize, in order to refine the specificity of the generally accepted criteria for CP, it is suggested to: (1) define the lower limit of severity together with the standards on which that definition is based; (2) specify known syndromes that are included in or excluded from the data set; (3) define the age of ascertainment at which progression or resolution is decided; (4) define the minimum age of inclusion and the criteria which must be met should the child die before the age of ascertainment; and (5) specify the upper age limit of acquired brain injury to be included [16].

CP definition by SCPE.

At the beginning of the SCPE work, researchers faced with difficulties remained when pooling and comparing information from different sources. The persisting problems were mainly due to the matter of language since not all partners from the different countries were English native speakers. Not everyone had derived the same meaning from terms such as 'increased tone' and 'walking fluently'. Thus, during the next 3 years, collaborative efforts were put together, mainly between child neuro-paediatricians, in order to develop a video-based tool, the SCPE Reference and Training Manual (SCPE R&TM) [4]. The aim of this tool was to promote a shared understanding of the words and phrases used to describe the clinical, functional, and neurological features of CP. Text and video material were first discussed within the small group of child neuro-paediatricians and then proposed to illustrate these features and to discuss pitfalls in diagnosis and classification. Interobserver exercise has been performed before spreading widely the use of this SCPE R&TM [30]. After a few years of use, the hope is that it will help to improve the harmonization and standardization level between different CP registers/studies, and that it will encourage new registers in new countries to join the SCPE network [30]. During the latest years of the SCPE, collaborative researchers are working on the data quality and also toward the improvement of available information on denominators within the EURO-PERISTAT project. To summarize, SCPE suggested the following definition:

Cerebral Palsy is a group of permanent, but not unchanging, disorders of movement and/or posture and of motor function, which are due to a non-progressive interference, lesion, or abnormality of the developing/immature brain [31]. This definition specifically excludes progressive disorders of motor function, defined as loss of skills previously acquired in the first 5 years of life. For any study of CP to be valid, there must be agreement on the 'similar characteristics' of the cases eligible for inclusion. SCPE has spent time agreeing on inclusion and exclusion criteria that should accompany CP definition [30].

Inclusion criteria: Optimal age: CP is not an easy diagnosis and it requires time to be confirmed. Premature diagnosis might lead to over-ascertainment (because of transient anomalies in preterm babies) or under-ascertainment, e.g. in mild unilateral spastic cases or ataxic cases. CP, as stated above, is not an unchanging condition, with the clinical picture in some cases altering as a child develops. It was agreed that age 5 years was the optimal age for confirmation of diagnosis. What about children who die early? It is recognized that some children with severe CP are correctly diagnosed at a young age, but die before their 5th birthday. Exclusion of these children could result in under-estimation of the prevalence of CP in Europe. Also when studying the aetiology, it would be better to include these cases, for instance cases of hypoxic-ischemic encephalopathy who die early [30]. In fact, a compromise was needed, and as a group, SCPE had followed the recommendation from Hagberg that children who die too early, i.e. before the age of 2 years, and those children with clear signs of CP who die between the ages of 2 and 5 years must be included. No upper age limit of onset of CP (in children with a postneonatal cause) was identified. But it is useful to isolate CP cases of post-neonatal origin, defined as cases arising from an aetiological event 27 completed days after birth. Exclusion criteria: All progressive conditions resulting in loss of acquired skills are excluded. However, we recognize that some progressive disorders might be registered wrongly as CP, due to the delay required, in some circumstances, to confirm a diagnosis of progressive disorder. However, the proportion of these misdiagnosed CP cases does not represent more than a few per cent of all CP cases, at least in the SCPE data. Children with hypotonia as the sole clinical feature and children with isolated spinal neural tube defects should also be excluded from the CP cases.

SCPE CP classification scheme: Classification means 'the basic cognitive process of distributing children with CP into classes or categories of the same type'. Different classification systems for CP serve different functions, but for epidemiological purposes, classification systems based on clinical findings are currently the most widely used. Drawing on published work, SCPE has classified CP into three main groups, which are based on clear neurological signs indicating pathology in the cerebral motor systems, e.g. spastic, ataxic, and dyskinetic CP. All CP subtypes have an abnormal pattern of movement and posture in common. Spastic CP cases have increased tone and pathological reflexes, either increased reflexes, e.g. hyper-reflexia or pyramidal signs, such as Babinski response. Increased tone in spasticity is characterized by an increased resistance which is velocity dependent [32]. A spastic catch is felt some time after onset of movement. Clonus is often associated with hyper-reflexia. It is considered pathological when it is prolonged or does not stop spontaneously. Pathological posturing of lower limbs is characterized by: (1) internal rotation of the hip; (2) hip adduction; and (3) equinus foot, resulting in a 'scissored' position. Dyskinetic CP cases present involuntary, uncontrolled, recurring, and occasionally stereotyped movements. The primitive reflex patterns predominate, and the muscle tone is varying. SCPE uses dystonic and choreo-athetotic CP subtypes for subgrouping. Dystonic CP is dominated by abnormal postures (may give the impression of hypokinesia) and hypertonia (tone fluctuating, but easily elicitable tone increase). Characteristics are involuntary movements, distorted voluntary movements, and abnormal postures due to sustained muscle

contractions (slow rotation, extension, and flexion of bodyparts). Choreo-athetotic CP is dominated by: hyperkinesia and hypotonia (tone fluctuating, but mainly decreased). Chorea means rapid involuntary, jerky, often fragmented movements. Athetosis means slower, constantly changing, writhing, or contorting movements. In some cases, however, it may be difficult to delineate these subgroups when features are present from both. Then the term dyskinetic CP should be used. Ataxic CP cases present loss of orderly muscular coordination, so that movements are performed with abnormal force, rhythm, and accuracy. Abnormal pattern of movement in ataxic CP is characterized by: (1) Loss of orderly muscular coordination, so that movements are performed with abnormal force, rhythm, and accuracy. Typical features are trunk and gait ataxia (disturbed balance) and past pointing (over or undershooting of goal directed movements). (2) Tremor is another common sign (mainly a slow intention tremor). (3) Low tone is also a prominent feature. Mixed CP forms: When it is a mixed CP form, i.e. spasticity with ataxia and/or dyskinesia, the child should be classified according to the dominant clinical feature. Pure dyskinetic movement disorder does not show hyperreflexia with clonus nor pyramidal signs. But in dyskinetic CP, these signs of spastic disorder may be present. The dominating features should determine subtype classification. Also, in spastic CP, some dystonic features are often present, especially when the upper extremities are involved. A dystonic posturing of the hand would, however, not be sufficient to classify a child as having the dystonic form of dyskinetic CP. The dystonic posturing of the trunk, arms, and face in the presence of lower-limb spasticity would qualify, however, as predominant dyskinetic features, thus, dystonic CP. Motor function impairment in CP children: SCPE choice was to recommend the scoring of motor function according to: the Gross Motor Function Classification System (GMFCS) for the lower limbs function [13] and Bimanual Fine Motor Function (BFMF) for the upper limbs function [33]. This last choice was achieved only very recently, and in order to conform to the S property (simplicity). However it has not yet been validated. Since SCPE does not recommend the use of diplegia/quadruplegia terms, and recommends using instead the term bilateral spastic CP subtypes, the two motor function scales can then be used for describing children with CP according to the functional grading given. For instance, a child with bilateral spastic CP may be 'scored' as GMFCS Level IV and BFMF Level III – which for the clinician involved would give the feature of adipsia – and another child with unilateral spastic CP may be scored as GMFCS Level II and BFMF Level I. Associated impairments in CP children: The SCPE collaborative group recommends collecting information on four associated impairments [31]. These recommendations are the minimum information that should be collected for those wishing to pool data or to compare it with data from other centres/countries. For visual and hearing impairment, the recommendation is to determine the presence or absence of such impairment, and then to classify the impairment as severe or not, according to the visual acuity (<0,1 in both eyes after correction) or hearing loss (more than 70 dB in the better ear before correction). Epilepsy can be defined as two unprovoked seizures, neonatal seizures being excluded. Firstly it must be known if the child 'had ever' or 'never had' epilepsy. Then it will be grouped as severe epilepsy if the epilepsy is still active. SCPE trees are used for categorizing children with CP [34]. Firstly, the decision tree is based on the presence of disorder of 'movement and/or posture' and of motor function. Secondly, the classification tree relies on neurological signs and topography for distinction between CP subtypes. By doing so, CP cases that are difficult to classify are not so numerous and less than 5% are observed in data from European centres [34][34]. At the beginning of the SCPE network, it was decided to use the words 'bilateral/unilateral spastic', with, in addition, the numbers of limbs involved, instead of using the words 'diplegia, tetraplegia'. After a while, the disappointment was great when we observed persisting important differences between centres on the 'theoretically' harmonized data. The overlap between the 'diplegia/quadruplegia' groups in CP classification has been well described in a recent paper [35]. These differences, between two and four limbs for example, could not be explained by anything else than by coding differences. Despite having agreed on a text definition and classification categories, large variations in classifying CP cases were still shown in a cross-validation exercise. The distinction between the number of limbs affected, used by several centres, in opposition to the number of limbs predominantly affected used by other centres, was the main reason responsible for these differences. Thus SCPE's recommendation moved to a simpler categorization, i.e. classifying spastic CP cases in unilateral versus bilateral CP cases [35]. Bilateral spastic CP was not further subdivided into arm/leg-dominated, diplegia/quadruplegia, nor 2-limb/3-limb/4-limb dominated, due to the great variability when these terms are not defined using functional scores respectively for upper and lower limbs. In a different way the Australian group gives an example that harmonization within one country may authorize more detailed description and classification than what is possible when dealing with several different countries. They are using four levels (minimal, mild, moderate, severe) to describe severity of neurological signs in each limb. However, there is still discussion in Australia about the overlap between triplegia, diplegia, and quadruplegia CP subtypes, and the need for an international consultation was expressed (Blair and Watson 2005). However, these scoring systems are very helpful for epidemiological purposes and evaluation of care. The reasons for SCPE choosing the BFMF scoring system rather than the MACS (Eliasson et al. 2006) are that: (1) BFMF takes into account possible asymmetry in the hand functions, whilst MACS does not; and that (2) BFMF can be retrieved from written medical records whilst MACS cannot. When collecting data on children with CP for CP registers or surveys, the situation of not directly examining the child is quite common. In the US, there was an attempt to classify children with CP according to severity criteria based on the functional ability of the most affected limbs, i.e. severe involvement meaning no useful function, and moderate involvement meaning the preservation of some function with or without the use of assistive devices [36].

Conclusion. To summarize, CP is a clinical description of heterogeneous pathology that underlies not only ante, intra but also post-natal brain injury. CP is one of the socially significant disorders in the world; however, one of their main issues is absence of clear definition of CP, which in turn affects further classification and therapeutic approaches to improve the quality of life of affected children. Therefore, there is a need not only in further research on etiology of CP but also in the development of clear standardized classification system of CP as a heterogeneous disorder.

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БАЛАЛАР ЦЕРЕБРАЛДЫ САЛ АУРУЫНЫҢ ХАЛЫҚАРАЛЫҚ АНЫҚТАМАСЫ (ӘДЕБИ ШОЛУ)

Түйін: Балалар церебралды сал ауруы - бұл нерв жүйесінің перинаталдық кезеңде бұзылуының нәтижесі болып табылатын және ілеспе аурулардың кең спекторымен байланысты прогрессивті мотор бұзылуы. Диагностикалау үшін орташа жасы шамамен 18 айды құрайды; алайда, деректер көрсеткендей, Прехтла шкаласы бойынша моториканы сапалы бағалай отырып, сондай-ақ резонанстық визуализация әдістерімен церебралды сал ауруын үш-төрт айлық жастан диагностикалау мүмкін. Қазіргі уақытта, церебралды сал ауруын эпидемиологиялық қадағалаудың 27 халықаралық бағдарламалары 5 церебралды сал ұйғарымдарына қолданылады, сонымен қатар олардың 46%, бір ғана церебралды салды анықтауға қолданады. Жүргізілген мета-талдауға сәйкес ең жиі айтылатын анықтау Еуропалық бағдарламасымен ұсынылған церебралды сал ауруын қадағалау (63%), Розенбаум ұсынған дефиниция және т. б. 2007 (54%). Осылайша, осы шолуда церебралды сал ауруының анықтамасы неғұрлым кеңінен сипатталған, сондай-ақ талқылау осы анықтамалардың дұрыстығын көрсетті.

Түйінді сөздер: церебралды сал ауруы, анықтама, ерте диагностика, нейровизуализация

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**МЕЖДУНАРОДНОЕ ОПРЕДЕЛЕНИЕ ДЕТСКОГО ЦЕРЕБРАЛЬНОГО ПАРАЛИЧА
(ОБЗОР ЛИТЕРАТУРЫ)**

Резюме: Церебральный паралич - это прогрессирующее моторное расстройство, являющееся результатом поражения нервной системы в перинатальном периоде, и связано с широким спектром сопутствующих заболеваний. Средний возраст для диагностики составляет около 18 месяцев; однако, недавние данные свидетельствуют о том, что церебральный паралич может быть диагностирован в возрасте трех-четырёх месяцев с использованием качественной оценки моторики по шкале Прехтла, а также методами резонансной визуализации. В настоящее время, 27 международных программ эпидемиологического надзора за церебральным параличом применяют пять определений церебрального паралича, при этом 46% их них используют более одного определения церебрального паралича. Согласно проведенному мета-анализу, наиболее часто упоминается определение, предложенное Европейской программой надзора за церебральным параличом (63%), за которым следует дефиниция, предложенная Розенбаумом и др. 2007 (54%). Таким образом, в данном обзоре будут описаны наиболее широко цитируемые определения церебрального паралича, а также обсуждение достоверности этих определений.

Ключевые слова: церебральные параличи, определение, ранняя диагностика, нейровизуализация