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Commentary

Can We Rethink Cerebral Palsy Definition?

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DESCRIPTION

Cerebral palsy (CP) is the most common cause of physical disability in childhood, someway representing the reverse side of neonatal mortality. CP affects more than 17 million people worldwide. In countries with advanced health systems, one child every 500-700 births are affected by CP approximately.

The interpretation of CP nature, as it is still proposed internationally [1-3] Cerebral palsy describes a group of disorders of the development of movement and posture, causing activity limitation, which are attributed to non-progressive disturbances that occurred in the developing foetal or infant brain) is no longer acceptable because defective for four important bias at least.

The idea that palsy is strictly a motor problem (disorder of the development of movement and posture).

The idea that palsy is mainly a central (cerebral) problem, ignoring the manifold transformations occurring on all the tissues of musculoskeletal system, capable of heavily influencing the operational choices of the Central Nervous System (CNS), already primitively limited.

The idea that the problems of sensitivity and perception, without neglecting those of cognition, emotion and behavior, are only potentially associable elements (accompanied).

The idea that palsy is a "fixed" pathology, despite affecting a brain with neural plasticity in a growing individual.

In fact, Rosenbaum [4], only ten years later wrote “CP remains an idea-a concept-rather than a specific disease entity”.

Many and complex problems intervene to create the overall clinical picture of CP, starting from the action design. The subject motivation to act is placed in an unstable balance between internal (arousal) and external [5] factors. The former is generated by the awareness of own needs and the strength of own desires. When they are transformed into actions, they generate outcomes that, in turn, act as reinforcements, positive and negative, as results of the perceptions and emotions they can arouse. The latter, made up of all opportunities, possibilities and clues coming from environment, include suggestions on the construction of the action that the CNS is able to understand through the canonical neuron activity. The presence and operation of these neurons, belonging to the mirror neuron system, has been documented also in CP children [6]. Models (children with typical development) capable of generating learning by imitation in the CNS of CP children through the mirroring of their motor performance also belong to external factors. This possibility, which can be summarized in the statement "learn from normality", is not easily accessible to CP children due to the contrast existing between the repertoire of pathological motor skills and that of typical development ones [7]. The ability to act, as Berthoz [8] claimed, “... is not movement (posture and gesture), the act is the intention to interact with the world or with oneself as part of the world ... The act is always the pursuit of an objective, it is always supported by an intention”. The act is allowed by the use of cognitive maps that guide the action; therefore, a movement for a purpose, in a certain context, at a given moment. Consequently, these elements cannot be considered factors only potentially "associated" with the motor problems of CP.

Recent fRM studies, carried out on hemiplegic children, reveal that significant alterations are present at level of motor imagination, even when the activation of the unaffected hand is explored [9]. It is therefore correct to speak of the most affected and least affected hand even for the hemiplegic patient. This consideration is sufficient to make the nosographic border between monolateral and bilateral forms, proposed by Surveillance Cerebral Palsy Europe [10] and already suggested by Sigmund Freud, waver.

The programming already contains motivational and ideational aspects. When there is no motor initiative we will talk about akinesia, when there is no design, we will have ideational apraxia [11]. Both of these elements can help us to understand the well-known "laziness" of the CP child.

The strategy of action in the CP child is recognizable through the pattern analysis: "Cerebral palsy is primarily a disorder in the building up of the patterned structure of movement with the result that there is a limitation of the freedom of choice of movement and posture ... Motor performance becomes functionally impaired and stereotyped "[12]. If the distinctions between different types of the spastic forms (apostural, akinetic,

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horizontal antigravity and vertical antigravity) [13] and of the dyskinetic ones (ballismus, corea, athetosis, dystonia) can be traced based to the characteristics of pathological motor patterns, we cannot understand what justifies the presence of the ataxic forms among the CPs, since these lack substantially pathological patterns.

A disregarded CP disorder, often hidden under the most conspicuous alteration of motor patterns and muscle tone, is dyspraxia, an aspect that in diplegias can affect more than half of the subjects. As [14] masterly argued: "The dyspraxic child has a reduced ability to represent himself, the object on which to act, the whole action and the composing sequences; has difficulty in ordering and coordinating the relative elementary movements in view of a purpose (programming), to start the relative program, to foresee (in anticipatory sense) a certain result, to control each sequence and the whole activity in the course of the action (feedback), to verify the obtained result as corresponding to the expected and desired one." The choice to reduce the variability of the repertoire to ensure the replicability of the result then becomes an understandable strategy that penalizes the prognosis. The presence of dyspraxia, if important, may justify prognostic errors based on the only analysis of the availability of motor repertoire observed in the first months of the child's life.

The execution phase, in turn, includes two possible error sources: action preparation and action execution. 1) Preparation errors embrace lack of anticipatory postural adjustments, altered receptor configuration, with consequent increase of stretching reflex threshold (excess activity), lack of reciprocal inhibition between agonist-antagonist muscles (passivity reduction), extension of the reflexogenic area, etc. 2) At action execution time, also structural characteristics of locomotor apparatus assume determinant responsibility. These metaplastic transformations, generated during growth by the brain injury itself, are reciprocally capable of influencing operational choices of CNS. In CP children, no tissue can be spared from metaplasia phenomena, which can also reach the extracellular matrix [15]. A very rich literature has accumulated evidence in this direction [16], without been able to establish a stable correlation between localization, nature and size of brain injury, clinical expression of palsy and structural characteristics of locomotor apparatus. In any case, it is clear that the deformities that inevitably appear in CP children during development are not exclusively related to posture and gesture pathological organization, but result from structural alterations of locomotor apparatus tissue, including bones, and from altered growth ratios between soft and rigid structures. For this reason, deformities are neither preventable nor containable through mere physiotherapeutic treatment. We must abandon the idea that the musculoskeletal system is always and only the victim of an incompetent CNS.

The most important bias of the current CP definition is to consider the disturbances of sensation, perception and consequently of central representation as “secondary” elements, only potentially associated with the motor ones. Due to the altered structure of the receptors and the lack of specialized movements, the CP child is unable to collect precise and reliable information. N Bernstein more than 50 years ago argued that none of the movements would be possible without the contribution of the senses, while H Poincarè more than a century ago stated that none of the senses would be functional without the contribution of the movement. Two sides of the same coin, as underlined by Lee. Often CP children experiences information coming from different sensory systems, such as sight and proprioception, not knowing what to listen [17]. Suppression and distortion of information force them to "drive in the fog", aggravating their already important motor difficulties. Their ability to perceptually anticipate the action results can be profoundly compromised, and may force CP children to give up movements that they can actually perform (Berthoz). The fear of the CP child is an expression of a distorted emotional map which, as Rizzolatti claimed, makes the amygdala take inappropriate decisions and produce body reactions not calibrated with respect to stimulus importance. In fact, ambiguous or unreliable information is not always centrally suppressed, as occurs in strabismus. Instead, it can be collected and processed in exasperated way. If information suppression prevails in hemiplegic children and contributes to reduce representation and therefore use of paretic upper limb, justifying the Constraint Induced Movement Therapy, in diplegic children and in some quadriplegic one, dysperceptive phenomena can prevail [18].

The discovery of altered sense of agency [19] has only recently been added to the long list of sensory deficits and perceptual disturbances of CP children. This defect alone is enough to justify the inability to control action execution in anticipatory way [20]. These can be corrected only retroactively, that is things done, based on mainly visual information and only when the error exceeds a certain range. The inability to notice own mistakes constitutes an important limit to the possibilities of recovery of CP children, referring especially to problems related to balance.

If the task of a definition is to emphasize the problem core, we must change the definition of PC. We must be able to highlight that CP is the changing product of the interaction between different functions, motor, perceptive, cognitive and emotional, of their adaptive modelling under the developmental thrust produced by the plastic maturation of the CNS and by the growth of the locomotor system. Starting from this awareness, CP re-education must, in turn, change to adapt to the idea of being in front of a multi systemic developmental disability.

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