Mariani Foundation Paediatric Neurology: 28

Cognitive and Behavioural Neurology in Developmental Age

Edited by:
Daria Riva
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Cognitive and behavioural neurology is increasingly the focus of attention from the neurosciences, both in adults and children. This field combines a number of specialties including neurophysiology, anatomy, developmental neurobiology, new neuroimaging techniques and genetics, to ensure that neurological conditions are approached from different standpoints. Appropriate cognitive/behavioural evaluation methods should based upon the known characteristics of neuropathology, molecular genetics and neurophysiology of the disorders.

This book provides an update on neurocognitive and behavioural deficits observed in developmental neurology (epilepsy, brain malformations, tumours, autistic spectrum disorders, syndromic and non-syndromic intellectual disabilities, cerebral palsy and CNS progressive disorders). It aims to describe cognitive/behavioural phenotypes, define indications for treatment and rehabilitation, and enhance knowledge acquired from clinical studies. The contents are addressed to child neurologists and psychiatrists, psychologists, paediatricians, geneticists, behavioural and speech therapists.

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Chapter 1

The effect of brain lesions/disease on cognition: location/type and treatment

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This introduction aims to outline the theoretical framework in which neuropsychological disorders, secondary to or associated with various types of diseases, locations, and types of brain lesions and their treatments, can be considered and examined. Higher psychic functions include a wide array of cognitive-behavioural functions that represent the final product of integrated processes of complex and widely distributed networks, made up of associative connections linking cortical areas that are highly or less highly specialized; the headquarters of sensory and motor elaborations which work together synchronously in a highly connected system. These widely distributed networks process the complexity and articulation of the language system, the visual-spatial capacity, and all other general abilities of the superior order (Riva, 2011).

The connectivity between brain areas is guaranteed by anatomical and functional connections. Structural anatomical connectivity is produced by a wide range of fibres, the classification of which, first made by Theodor Meynert in 1872 (Meynert, 1872), differentiates between associative and commissural projections. The projection pathways are: the corticobulbar tract, the corticospinal tract, the thalamo-cortical pathway and vice-versa, and the fornix pathway. There are also the commissural fibres, of which the corpus callosum is the most representative connecting system between the two hemispheres. There are, finally, associative fibres which are intra-hemispheric and connect associative areas of the same hemisphere.

The intra-hemispheric and commissural tracts thus compose the brain's structural and anatomical connectivity, that is, the discrete pathways that directly connect distant regions and territories. There is also a functional connectivity among brain regions that share functional properties. It can be defined, more specifically, as the spatial temporal correlation between neurophysiological and neuropsychological events that are spatially remote, but crucial to task-relevant processing. Functional connectivity, which can be analyzed using neurophysiological and direct functional brain imaging tools, has provided extraordinary information regarding brain function as well as, obviously, anatomical function, as it permits us to study *in vivo* human mental function and activity.

Knowledge about the brain's architecture is based on information furnished by different disciplines, including mathematical models known as Graph Theory (He & Evans, 2010) and advanced neuroimaging technologies, such as resting state functional magnetic resonance imaging. The latter method has made it possible to study the activation of specific brain areas even when the subject being examined is not engaged in an active task; it is also possible to investigate individuals with severe intellectual disability who are unable to carry out predetermined tasks.

According to mathematical models, instead, a network is usually made up of a clustering of more or less specialized areas which are connected to one another and, at the same time, to distant structures. The Graph Theory states that there are Small Worlds that are independent, although linked together by powerful internal connections, which do not work as a psychotic isolated system, but through in-between and distant connections with other Small Worlds. Functional connectivity is thus made up of hubs of greater or lesser complexity, convergences, divergences, feedback systems, switchboards, *etc*. which participate in the brain's extraordinarily complex processing system (Gerloff & Hallett, 2010).

Several important studies have made it possible to identify four functional networks, consistently found to be operative in healthy subjects: the visual, the sensorimotor, the basal ganglia (or basal nuclei), and *the Default Mode Network* (DMN). The latter is activated during passive mental no-task states (Buckner, 2012; Lee *et al.*, 2012; Moussa *et al.*, 2012).

The DMN operates in the morning when one is in a half-asleep state in which our minds are engaged in non-directed thinking; daydreaming, remembering, planning out everyday activities, linking memories and images, and planning freely without any definite logic. The DMN involves specific brain structures which include regions linked to episodic memory, those correlated to subjective visceral sensations processed in the precuneus, and to those processed by the inferior parietal lobes, an integrative area of fundamental importance. The organization of functional connections is not operative at an early age; the child's brain connects hubs and nodes in a unimodal processing system and the adult's brain connects areas within the confines of a multimodal one (integrative associative areas). There is thus a development over time; the infant has primary sensorimotor areas which are accessed by a single type of afference where it becomes integrated and, as the child develops, brain areas integrate different afferences. It can be said that between 7 and 9 years of age, the brain's architecture is similar to the Small-World type found in adults (Fransson *et al.*, 2010).

Brain architecture and function reflect a system in which some regions are more or less specialized while others are integrative and linked to one another by pathways. A lesion, be it malformative, tumoural, vascular, degenerative, or demyelinating, that damages one of the components of the system, causes a disconnection which fragments the integration process. If a lesion damages a hub or heteromodal (integrative) area, the impairment will be more significant with respect to secondary effects that are caused by nodal lesions. Any brain area is responsible for a part of the perfection of the final processing product. It is thus pointless to distinguish between eloquent and non-eloquent brain areas; it is instead important to distinguish between crucial, extremely complex areas, that might not be very vicarious, and less critical areas. The specialization of brain areas can be studied using various methodologies, such as morphological and functional MRI, combined positron emission tomography (PET), SPECT, *etc.*, while bundles of fibre tracts can be analyzed using diffusion tensor imaging (DTI) tractography, which allows us to analyze connectivity *in vivo*.