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Cerebellum and Apraxia

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Introduction

Classical tenits posit that the role of the cerebellum is limited to pure sensorimotor control. However, evidence from clinical and imaging studies shows that the cerebellum is crucially involved in nonmotor cognitive and affective functions. Schmahmann and Sherman (1998) [1] introduced the cerebellar cognitive affective syndrome (CCAS), characterised by executive, visuo-spatial, affective and linguistic impairments caused by cerebellar pathology. Apraxia, as a planning, organisation and execution disorder of a skilled motor action (not caused by motor, sensory or intellectual impairment) [2], may be regarded to form part of the executive cluster of CCAS. Indeed, several anatomoclinical studies have confirmed involvement of the cerebellum in at least some types of apraxia, which adds to the nonmotor role of the cerebellum. According to Hugo Liepmann [3], apraxia is thought to evolve from a disruption of the creation, activation or retrieval of movement formulae. These formulae represent the idea of a movement as a visual or acoustic image and are stored in the left parietal lobe. The left prefrontal area subsequently associates these formulae with an inherently stored innervatory pattern to transfer the information to the left primary motor areas. The corpus callosum transfers this information to the right motor cortex if the movement is to be executed by the left limb [3]. Based on some recent clinical evidence we hypothesize that the cerebellum forms an intrinsic part of this connectionist model of Liepmann.

Methods

To delineate the possible role of the cerebellum in the planning and organisation of skilled motor actions, a number of recent case studies are reported of apraxia following cerebellar pathology. Different kinds of apraxia were described following cerebellar disease including Apraxia of Speech (AoS), Apraxic Agraphia (AA), a cluster of apraxias (e.g. constructional, ideomotor and drawing apraxia) characterising Developmental Coordination Disorder (DCD). All of these cases provided evidence for involvement of the cerebrocerebellar network in the pathophysiology subserving apraxic disorders.

Results

Apraxia of Speech (AoS)

In AoS motor speech planning and programming are selectively impaired. The patient is no longer capable of converting phonological information into the correct verbal-motor commands [4]. AoS is frequently associated with Foreign Accent Syndrome (FAS), a motor speech disorder in which a change of accent is perceived by listeners of the same language community as distinctly foreign [5]. AoS is primarily associated with damage of the language dominant motor speech region (anterior insula, inferior premotor and motor cortex, BA 44 of Broca's area). However, based on some overt semiological similarities with ataxic dysarthria, it has been hypothesized that AoS and ataxic dysarthria may share similar pathophysiological mechanisms [4]. Cerebellar involvement in AoS was confirmed by Mariën et al. (2006) [4] and Mariën and

Verhoeven (2007) [6]. They described two right-handed patients with FAS after a left hemispheric stroke. In addition to a significant hypoperfusion in the language dominant hemisphere, Tc-99m-ECD SPECT perfusion scans revealed a secondary hypoperfusion in the contralateral right cerebellum. This phenomenon of crossed cerebrocerebellar diaschisis resolved after the remission of FAS, suggesting a crucial role for the cerebellum in motor speech planning [4].

Apraxic Agraphia (AA)

AA is a peripheral writing disorder characterized by poor letter formation, sometimes even illegible scrawls due to a disruption of the (access to) the graphic motor programs. AA is typically associated with damage of the superior parietal region and the dorsolateral and medial premotor cortex (Exner's area) of the language dominant hemisphere [7] but recent evidence indicates that cerebellar lesions may also induce AA.

Mariën et al. (2007) [8] reported a right-handed patient who developed AA after a right cerebellar hemorrhage. In addition to a hypoperfusion in the lesion site (right cerebellar hemisphere), a Tc- 99m-ECD SPECT perfusion scan showed a hypoperfusion in the clinically suspected but structurally intact supratentorial region in the prefrontal region of the language dominant hemisphere (Exner's area). Three additional cases of AA after focal damage of the cerebellum were documented by De Smet et al. (2011) [7]. Mariën et al. (2013) [9] also reported a 15-year-old left-handed patient with AA probably due to an incomplete maturation of the cerebrocerebellar network confirming the involvement of the cerebellum in AA [9].

Developmental Coordination Disorder (DCD)

DCD is a neurodevelopmental disorder characterized by difficulties in acquiring motor skills, sensorimotor coordination disturbances, deficient postural control, strategic planning problems, disrupted visuo-spatial information processing, executive dysfunction, and usually a much lower PIQ than VIQ [10]. The condition closely resembles CCAS due to the frequent association with affective, behavioural and social disturbances [11][1]. DCD is typically accompanied by constructional and drawing apraxia, as is the case with a 19-year-old left-handed patient described by Marën et al. (2010) [10]. Structural MRI showed a rostral vermisdysplasia, a slight anterior/superior asymmetry of the vermal fissures. A Tc-99m-ECD SPECT perfusion scan revealed overall decreased perfusion of the cerebellum and a distant functional suppression of the supratentorial regions involved in the execution of planned actions, visuo-spatial processing and affective regulation. This case study provides additional evidence for the association between DCD and CCAS and suggests that the cerebellum and the cerebrocerebellar network might be a part of the pathophysiological mechanism underlying DCD [10].

Discussion

The involvement of the cerebrocerebellar network in different forms of apraxic disorders suggests that the cerebellum is implicated in the neural network responsible for the control, planning and execution of skilled movements. It might be hypothesized that the cerebellum is indeed part of Liepmann's network for skilled movements. The left parietal lobe is responsible for the storage of movement formulae, the left prefrontal lobe for the execution of the formulae and the corpus callosum for the transfer of this information to the right motor cortex. Within this model the cerebellum may be considered to be responsible for the cognitive planning, timing and coordination of the execution of the movement formulae [3].

References

[1] Schmahmann, J.D. and Sherman, J.C. (1998). The cerebellar cognitive-affective syndrome. *Brain, 121*, 561-579.

[2] Heilman, K.M., Gonzalez, R.L.J. and Hanna-Pladdy, B. (2008). The forelimb Apraxias. In Mariën, P. and Abutalebi, J. (eds.), *Neuropsychological Research: a Review*. Hove: Psychology Press, 162-183.

[3] Pearce, J.M.S. (2009). Hugo Karl Liepmann and apraxia. *Clinical Medicine, 9*, 466-470.

[4] Mariën, P., Verhoeven, J., Engelborghs, S., Rooker, S., Pickut, B.A. and De Deyn, P.P. (2006). A role for the cerebellum in motor speech planning: Evidence form foreign accent syndrome. *Clinical Neurology and Neurosurgery, 108*, 518-522.

[5] Verhoeven, J. and Marien, P. (2010). Neurogenic foreign accent syndrome: Articulatory setting, segments and prosody in a Dutch speaker. *Journal of Neurolinguistics, 23*, 599-614.

[6] Mariën, P. and Verhoeven, J. (2007). Cerebellar involvement in motor speech planning: Some

further evidence from foreign accent syndrome. *Folia Phoniatrica et Logopaedica, 59*, 210- 217.

[7] De Smet, H.J., Engelborghs, S., Paquier, P., De Deyn, P.P. and Mariën, P. (2011). Cerebellar- induced apraxic agraphia: A review and three new cases. *Brain and Cognition, 76*, 424-434.

[8] Mariën, P., Verhoeven, J., Brouns, R., De Witte L., Dobbeleir, A. and De Deyn, P.P. (2007). Apraxic - graphia following a right cerebellar hemorrhage. *Neurology, 69*, 926-929.

[9] Mariën, P., De Smet, E., De Smet, H.J., Wackenier, P., Dobbeleir, A. and Verhoeven, J. (2013). "Apraxic Dysgraphia" in a 15-year-old left-handed patient: disruption of the cerebello- cerebral network involved in the planning and execution of graphomotor movements. *Cerebellum, 12*, 131-139.

[10] Mariën, P., Wackenier, P., De Surgeloose, D., De Deyn, P.P. and Verhoeven, J. (2010). Developmental coordination disorder: Disruption of the cerebello-cerebral network evidenced by SPECT. *Cerebellum, 9,* 405-410.

[11] Green, D., Baird, G. and Sugden, D. (2006). A pilot study of psychopathology in developmental coordination disorder. *Child Care, Health and Development, 32*, 741-750.