

The Ravel case; possible link between non fluent Primary Progressive Aphasia and basal ganglia dysfunction

The Primary Progressive Aphasia (PPA) syndrome was firstly introduced in 1982. Recently, the primary progressive aphasias were classified into three clinical variants; non fluent/agrammatic, semantic and logopenic.

The criteria developed by the investigators were mainly based on the characteristics of speech and language functions but additionally supported by imaging, pathologic and genetic markers. Agrammatism in language production and effortful speech are the core criteria of non fluent aphasia. Single word comprehension deficits characterize the semantic variant. Finally, the logopenic aphasia core features are word retrieval and repetition deficits.

It has been more than one year ago when we made the first notice in an outpatient Alzheimer's clinic; a patient with non fluent PPA could sing despite the fact that she could hardly speak. Later, we confirmed this finding in a number of patients suffering from the same disease. The question that arises is why are these patients benefited from rhythmicity.

Rhythmic cueing has been proved to be helpful in Parkinson's disease rehabilitation. Parkinson's disease is an extrapyramidal disorder characterized by dysfunction of a complex of nuclei called basal ganglia. Patients with basal ganglia lesions demonstrate a reduced perceptual activity to tempo changes. The next question that should be answered is what is the connection between PPA and basal ganglia disorders?

Searching the literature I came up the case of the French composer Maurice Ravel who suffered from a progressive cerebral disease of uncertain etiology which led him to death in 1937, at the age of 62. He was struck down by aphasia, loss of movement control, defective oculomotor movements and an episode of motor difficulty as confirmation of extrapyramidal syndrome. In the absence of a post-mortem examination, the diagnosis remains speculative. Based on the clinical manifestation PPA and corticobasal degeneration (CBD) were included in the most possible diagnoses proposed by the researchers. CBD belongs to Parkinson plus syndromes and as such is characterized by basal ganglia deficits. The years that followed the first clear signs of the disease, Ravel composed creations sufficient different from his previous compositions such as the Bolero and the Piano Concerto for the Left Hand. Taking these together, we might speculate that the fact that Ravel adopted different uses of timbre could be attributed to altered basal ganglia function.

The Ravel issue offers an explanation for our clinical observation and constitutes a hint for the possible overlap between the non fluent PPA variant and basal ganglia dysfunction. However, surveys which connect singing and music with PPA non-fluent subtype should be conducted in the future in order to confirm our initial assumption.

Athanasia Alexoudi, Damianos Sakas, Stylianos Gatzonis
*Department of Neurosurgery, University of Athens Medical School, "Evangelismos" Hospital,
Athens, Greece*

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[The "Ravel issue" and possible implications.](#)

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