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No-ing” Primary Progressive Aphasia: Evolution of a Concept

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This lecture is designed to provide an updated context for the classifications of primary progressive aphasia (PPA) and an expanded clinical rubric for differential diagnosis. Initially the neurodegenerative aphasias were defined based on the lesion localization classifications of cerebrovascular disease. However, the recognition that brain function is organized into three main networks--the default network, the frontal-executive network, and the salience network—allowed an expansion of the understanding of language organization and the ability to define the PPA according to network functions. Currently the classification system attempts to relate phenotypic presentations of the 3 differentiated PPAs according to regions of maximal atrophy or metabolic dysfunction focused on language alone. However, by viewing the concepts of brain network organization on an evolutionary background of language development, there is a broader set of features that may more closely identify the phenotype and neuropathology of each syndrome. This lecture will present the concepts of PPA as part of an understanding of how language fits overall into the brain’s overarching goal of enhancing survival, present a clinical rubric for defining the concept of fluent versus nonfluent PPA, integration of Tempini’s classification system into an expanded evolutionary framework, and then discuss each syndrome individually, including videotaped examples of each subtype.

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