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SPEECH AND LANGUAGE ABILITIES OF PERSONS WITH FRONTOTEMPORAL DEMENTIA

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SUMMARY

The review outlines the importance of understanding speech and language difficulties that occur among the first symptoms of frontotemporal dementia, as well as the role of speech therapists in the management of people with frontotemporal dementia. Frontotemporal dementia is one of the most common types of dementia in adults under the age of 65. The main variations of frontotemporal dementia are behavioral, progressive nonfluent aphasia, semantic dementia, and logopenic progressive aphasia. Speech and language difficulties are often among the first indicative signs of frontotemporal dementia, and their proper recognition and understanding play a significant role in the differential diagnosis. Speech and language therapists have to be involved both in the diagnosis of frontotemporal dementia and its treatment to provide the highest quality services to people with dementia and their carers.

Key words: *: frontotemporal dementia – classification - speech and language*

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INTRODUCTION

Due to the global phenomenon of society aging, dementia is one of the significant issues for public health (Fratiglioni et al. 2012). The costs for people with dementia around the world are high. Informal care, especially in less developed countries where people with dementia are mainly cared for by family members, is a major cost component (Wimo et al. 2007; Sinanović 2012). Dementia most often occurs in the third age. It is secondary to neurodegenerative processes, usually chronic and disabling (Leon-Salas & Martinez-Martin 2010). Dementia is a clinical condition caused by changes in the central nervous system (Beljo 2017). It is characterized by loss of function in multiple cognitive areas (Chapman et al. 2006). The prevalence of dementia doubles every five years after the age of 65 (Hugo & Ganguli 2014). The number of people with dementia in the world in 2000 was estimated at 25 million. More people with dementia live in less developed countries of the world, with the share of people with dementia will increase significantly in the future (Anders et al. 2003).

Diagnosing dementia is a demanding task and can be very challenging for the clinician, especially in the early phase (Kostopoulou et al. 2008). The symptoms of dementia must be distinguished from the changes that occur in the third age since dementia is not the result of the normal aging process (Sošić et al. 2018). The most common dementias are divided into primary and secondary. The primary ones arise as a consequence of neurodegenerative processes in the brain, and the secondary ones are the result of tumors, poisoning, hormonal and metabolic changes (Muačević 1995). Frontotemporal dementia occurs as a result of relatively selective and progressive atrophy of the frontal and/or temporal lobe (Rohrer & Warren 2011). This atrophy is a cause of dementia with an early onset, with an average incidence in the sixth decade of life. The results of the study show that only 20-25% of people with frontotemporal dementia are older than 65 years (American Psychiatric Association 2013).

Frontotemporal dementias are thought to account for at least about five percent of all irreversible dementias (Galariotis et al. 2005). On average, 2.7% of people diagnosed with some form of dementia over the age of 65 are diagnosed with frontotemporal dementia, while the percentage of frontotemporal dementia among those under 65 rises to 10.2% (Hogan 2016). It is considered the fourth most common, after Alzheimer's dementia, dementia with Lewy bodies, and vascular dementia (Leroy 2021).

One of the characteristics of frontotemporal dementia is that difficulties in the field of memory occur only in advanced stages, which distinguishes it from other forms of dementia (Pasquier et al. 2001). This is necessary to keep in mind in differential diagnosis, to primarily distinguish between frontotemporal dementia and Alzheimer's dementia (Hornberger & Piguet 2012). Unlike people with Alzheimer's dementia, those with frontotemporal dementia wait longer for an accurate diagnosis (Leroy et al. 2021). Language difficulties are a common symptom in people with dementia and may

be a suggestive manifestation of dementia (Tang-Wai & Graham 2008). These language difficulties are the result of nerve cell degradation, i.e. neurodegenerative changes in the brain area responsible for comprehension, repetition, verbal expression, reading, and writing (Banović et al. 2018). People with dementia, among other signs, show problems finding words (anomia), a lack of sentence comprehension, and a lack of cohesion in discourse (Kempler & Goral 2008).

This paper aims to present the features of frontotemporal dementia and speech and language difficulties that arise as a result of frontotemporal dementia.

The paper has a descriptive character and presents a systematic review of the literature dealing with the topic of frontotemporal dementia.

BASIC CLINICAL PICTURE OF FRONTOTEMPORAL DEMENTIA

Frontotemporal dementia has several variants that differ in their symptoms (Mendez 2004). It encompasses clinical disorders that include changes in behavior, language abilities, and common motor symptoms (Olney et al. 2017). As a progressive neurological syndrome, it is characterized by different clinical presentations, and neuropsychiatric symptoms and language difficulties are common. It is diversity in the clinical picture of people with frontotemporal dementia that poses a challenge in diagnosis (Bott et al. 2014). Patients with frontotemporal dementia have been misdiagnosed for decades, most commonly as patients with Alzheimer's dementia (Snowden et al. 2002). Memory loss is rarely the first symptom of frontotemporal dementia, which differs from Alzheimer's dementia in clinical symptoms and pathology. Frontotemporal dementias instead of dominant memory disorders are characterized by behavioral disorders, personality changes and speech difficulties (Liščić 2009). It is estimated that as many as 90% of people with frontotemporal dementia have behavioral abnormalities (Kertesz et al. 2010).

Personality changes stand out in the behavior of a person with frontotemporal dementia. Apathy, loss of empathy, stereotypical behaviors, impulsive and abrupt reactions, and motor restlessness are just some of the symptoms that lead to loss of social competence. Executive functions are also impaired, while memory and visual-spatial skills are relatively preserved (Laforce 2013).

One of the main differences from other types of dementia is that frontotemporal dementia is more common in younger age groups (50–60 years) (Robinson et al. 2015). In persons younger than 65 years, frontotemporal dementia is the second most common form of dementia (Young et al. 2018). In almost half of the cases in family history, members with dementia have been found, and genetics are thought to play an important role in developing a picture of frontotemporal dementia (Bottino 2000). Although there are studies whose results indicate that frontotemporal dementia occurs equally concerning gender (Snowden et al. 2002), there are also those in which a higher incidence occurs in males (Mercy et al. 2008). The prevalence of frontotemporal dementia is predicted to increase as the number of older people increases (Young et al. 2018).

SPEECH AND LANGUAGE ABILITIES OF PERSONS WITH FRONTOTEMPORAL DEMENTIA

Frontotemporal dementia should be observed as a syndrome, which has its variants. The authors usually single out three subtypes of frontotemporal dementia. Language disorders appear as a main accompanying symptom in two subtypes of frontotemporal dementia.

Tsai & Boxer (2016) single out a behavioral variant of frontotemporal dementia, in which behavioral disorders are pronounced; a semantic variant of primary progressive aphasia, in which symptoms of fluent aphasia are encountered, and also a non-fluent variant of primary progressive aphasia, in which symptoms of nonfluent aphasia are pronounced. Rohrer et al. (2009) list two subtypes of frontotemporal dementia: semantic dementia and progressive nonfluent aphasia.

Deviations of language abilities due to frontotemporal dementia are presented in fields of fluency, repetition, naming, comprehension, and reading (Kirshner 2014).

The behavioral variant of frontotemporal dementia is marked by behavior, personality and socio-emotional changes. But, there are also difficulties in the area of cognitive functioning, with an emphasis on speech-language difficulties even in the early stages of the disease (Geraudie et al. 2021).

Language variants of primary progressive aphasia are characterized by prominent language disorders that can be further classified into nonfluent/agrammatic, semantic, and logopenic variants (Laforce 2013).

Primary progressive aphasia

This is a form of frontotemporal degeneration, which encompasses a whole range of disorders and occurs due to degenerative changes in the frontal or temporal lobes of the brain, i.e. the zones of these lobes that are responsible for speech and language (Marshall et al. 2018). In order for a person to be diagnosed with primary progressive aphasia, a neurologist must determine the existence of progressive neurodegenerative changes, and there should be language difficulties that are more pronounced than memory problems (Mesulam 2013).

People with more pronounced degenerative changes on the left side of the frontal and/or temporal lobe have greater difficulty finding the right word and understanding the word, as opposed to people with more

pronounced degenerative changes on the right side who have difficulty recognizing faces (Thompson et al. 2003). Clinicians, based on the motor component of speech and language characteristics, and cognitive characteristics, list three variants of a primary progressive aphasia: semantic, nonfluent, and logopenic (Wilson et al. 2010). Primary progressive aphasia is essentially "linguistic dementia" and poses a huge challenge for diagnosis (Marshall et al. 2018). Language is the area with negative deviation in all phases of primary progressive aphasia (Mesulam 2013).

Regarding motor speech disorders, which can occur due to primary progressive aphasia already in the initial stages, it is necessary to single out progressive dysarthria (Duffy et al. 2014). The speech of people with primary progressive aphasia is distorted, has slower rate speeds, and sounds as if the speaker is under exertion (Marshall et al. 2018).

Difficulties in understanding instructions and sentences occur due to grammatical deficits and can lead to problems in understanding more complex dialogue and difficulties in participating in the conversation (Peelle et al. 2008). Written communication is relatively preserved in the initial stages of the disease, and in the later stages, writing errors occur as a result of spelling errors (Lo Monaco et al. 2020).

Progressive fluent aphasia

In people with the *semantic* variant of frontotemporal dementia, also called p*rogressive fluent aphasia*, speech is characterized by semantic jargon and semantic substitutions, often paraphasia, and lower results at naming tasks as well as difficulty understanding words (Kertesz et al. 2010). Difficulties in the field of semantics are not the result of perceptual or sensory deficits or intellectual difficulties (Warrington 2007). These difficulties in understanding the meaning of words are present in all categories of objects (Kertesz et al. 2010). Comprehension is impaired often even at single word-level (Kirshner 2014). Even a few years after the onset of the first symptoms, a person with a semantic variant of frontotemporal dementia may have preserved conversational abilities, social skills, and orientation skills, however, naming and comprehension skills are severely impaired (Kertesz et al. 1998). The linguistic components of syntax, phonology, and morphology are intact, also as episodic memory (Kertesz et al. 2010).

People with the semantic variant of frontotemporal dementia show significantly better results on nonverbal tasks (Auclair-Ouellet et al. 2020), and often in the later stages of the disease, there is a higher frequency of nonverbal expression (Marshall et al. 2018).

Progressive nonfluent aphasia

This is a rarer form of frontotemporal dementia with frequent motor speech disorders that some authors consider to be even more pronounced than language disorders (Croot et al. 2012). Speech and expression are nonfluent. The segment of prosody, the emphasis of words and sentences is damaged, the importance of which is reflected in the interpretation of the emotion that the speaker has according to the content he is talking about, and for distinguishing the type of sentence he wants to use (Grossman 2012).

Due to progressive nonfluent aphasia, there are difficulties in finding an adequate word (anomia) and phonemic spelling error, with the understanding being relatively preserved (Robert et al. 1999, Kirshner 2014). Impaired phonology and syntax also occur in progressive nonfluent aphasia (Galariotis et al. 2005).

Logopenic progressive aphasia

This is a type of primary progressive aphasia that is more commonly associated with Alzheimer's disease compared to the other two variants (fluent and nonfluent primary progressive aphasia) that are closely related to frontotemporal dementia (Awad & Awad 2011). Also, this term is used for those, rarely cases of persons with frontotemporal dementia who do not appear to meet the criteria for classification in either fluent or nonfluent variant of frontotemporal dementia and show language difficulties in the early stages of neurodegenerative disease (Watanabe et al. 2019). A person with logopenic progressive aphasia occurs mild difficulties at naming in the initial stages of the disease. A person with logopenic aphasia shows difficulty during repetition and can replace a word with another, meaningless one, which sounds similar to the target word (Rohrer et al. 2010). There are more pronounced difficulties in naming and verbal fluency as the disease progresses. Also, there are phonemic paraphrases and neologisms, and in the latter stages of the disease marked comprehension difficulties accompanied by incomprehensible, slang speech (Caffarra et al. 2013).

MANAGEMENT OF PERSONS WITH FRONTOTEMPORAL DEMENTIA

Recommendations for future research on frontotemporal dementia are clearly defining criteria for diagnosis, tests for ranking performance and achievement on cognitive and motor tasks, and standards for monitoring behavioral changes, all with the goal of exact measurement and comparability of conclusions (Borroni & Benussi 2019). The clinical picture of frontotemporal dementia is diverse, with a large number of pathological phenotypes, and expensive, detailed, and highly controlled trials are required for drug approval. For this reason, there is still no cure for frontotemporal dementia, nor is there a way to slow the progression of the disease, and currently only several behavioral, cognitive, and motor symptoms can be treated pharmacologically (Jicha & Nelson 2011).

Numerous pharmacological treatments for frontotemporal dementia are the same as treatments for Alzheimer's dementia (Manoochehri & Huey 2013). For

difficulties in the field of language abilities, memory, and executive functions, there are still no effective treatments that would completely stop the course of the decline of these cognitive functions (Tsai & Boxer 2014). There is also a lack of research on the management of frontotemporal dementia that would be based on strengthening the remaining functions in a person with dementia and systematic training of caregivers (Jicha & Nelson 2011). For these reasons, a conclusion is drawn about the importance of nonpharmacological interventions, which implies the inclusion of many professionals, such as physiatrists, occupational therapists, speech and language therapists, and psychologists in a multidisciplinary team led by a neurologist tasked with alleviating the symptoms of frontotemporal dementia (Shinagawa 2015).

One of the unfortunately common misconceptions is that people with dementia do not benefit from the services of a speech therapist since dementia is a progressive disease. However, speech therapists, with their knowledge of brain dysfunctions and the consequences that communication difficulties have on the lives of individuals, can contribute to better care for people with all types of dementia, including frontotemporal (Kortte & Rogalski 2013). Among the first steps the speech and language therapist will determine areas of speech and/or language with a deviation, as well as whether there are difficulties in swallowing (Kortte & Rogalski 2013). The speech therapist will assess fluency, comprehension, repetition, naming, and reading through speech and language tasks (Liu et al. 2019). The next step is to make a treatment plan to compensate or minimize the consequences of the speech and language difficulties. The goal is to maintain communication for as long as possible (Khayum et al. 2012). A speech therapist's role in the management of people with frontotemporal dementia are interventions that should be aimed at maintaining communication, minimizing the consequences of speech and language disorders, and improving the quality of life (Mahendra & Tadokoro 2020). The help of speech therapists is especially important in those people with frontotemporal dementia who have developed motor speech disorders such as dysarthria and apraxia, as well as those people who develop voice disorders or swallowing difficulties (Tsai & Boxer 2014). There is an evident increase in the number of studies examining speech and language therapy approaches, as well as data that support speech therapy methods for people with dementia (Henry et al. 2013). Speech therapists offer a wide range of methods and treatments for people with frontotemporal dementia - from speech and language exercises through reading, repetition, and naming activities, through cognitive rehabilitation interventions to the use of compensatory strategies. The benefits of maintaining communication are numerous and more than the considerable reason for a person with dementia to be referred for speech therapy as soon as possible (Volkmer et al. 2020).

CONCLUSION

Frontotemporal dementia is one of the most common types of dementia in adults under the age of 65, and it is thought to account for as much as 10 to 20% of the total number of people diagnosed with dementia. The main difference between frontotemporal dementia and other types of dementia is that in the initial stages memory is not affected, and its deterioration occurs only in the later stage. Therefore, the importance of understanding and timely recognition of the difficulties that occur in the field of speech and language has to be emphasized. A notable role in the management of persons with frontotemporal dementia has to be played by a speech therapist, who should take part in the diagnosis of frontotemporal dementia, and its treatment, which should not be reduced only to medical treatment of symptoms, all intending to provide the highest quality services for people with dementia.

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Silva Banović: concept and design of article, literature searches, writing manuscript.

Osman Sinanović: comments on the concept of article, writing some parts of manuscript.

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