

Mielniczek Katarzyna, Lipiec Marta, Kowalska Aleksandra, Jóźwik Katarzyna, Dołomisiewicz Diana. Speech disorders in primary progressive aphasia (PPA). *Journal of Education, Health and Sport*. 2022;12(7):968-977. eISSN 2391-8306. DOI <http://dx.doi.org/10.12775/JEHS.2022.12.07.098> <https://apcz.umk.pl/JEHS/article/view/JEHS.2022.12.07.098> <https://zenodo.org/record/6945086>

The journal has had 40 points in Ministry of Education and Science of Poland parametric evaluation. Annex to the announcement of the Minister of Education and Science of December 21, 2021. No. The journal has had 40 points in Ministry of Education and Science of Poland parametric evaluation. Annex to the announcement of the Minister of Education and Science of December 21, 2021. No. 32343. Has a Journal's Unique Identifier: 201159. Scientific disciplines assigned: Physical Culture Sciences (Field of Medical sciences and health sciences); Health Sciences (Field of Medical Sciences and Health Sciences).

Punkty Ministerialne z 2019 - aktualny rok 40 punktów. Załącznik do komunikatu Ministra Edukacji i Nauki z dnia 21 grudnia 2021 r. Lp. 32343. Posiada Unikatowy Identyfikator Czasopisma: 201159. Przypisane dyscypliny naukowe: Nauki o kulturze fizycznej (Dziedzina nauk medycznych i nauk o zdrowiu); Nauki o zdrowiu (Dziedzina nauk medycznych i nauk o zdrowiu).

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Received: 16.07.2022. Revised: 17.07.2022. Accepted: 31.07.2022.

Speech disorders in primary progressive aphasia (PPA)

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Abstract

Introduction : Primary progressive aphasia (PPA) is a group of neurodegenerative disorders whose main feature is speech and language dysfunction. There are three main forms of PPA - non-fluent variant - agrammatic (nfvPPA), semantic variant (svPPA) and logopenic variant (lvPPA). These include the canonical syndromes currently recognized by consensus diagnostic criteria.

Material and method : An overview of language disorders in primary progressive aphasia is presented. The impact on the conversation process and the patient's quality of life assessment was evaluated. The work was based on scientific publications posted on the scientific platform PubMed.

Results: In order to diagnose PPA, criteria must be met, i.e., language difficulties are the main feature of the clinical picture, language difficulties are the main cause of disability in daily life, and that aphasia is the most significant disorder in the initial period of the disease.

Conclusions: PPA significantly affects both the production and comprehension of speech. It affects the idea, content, construction and delivery of the patient's speech. In addition, it causes significant difficulties in the repetition of speech, both full sentences and words alone.

Keywords: speech disorders, aphasia, dysarthria, primary progressive aphasia.

1. Introduction

The term primary progressive aphasia (PPA) describes a group of neurodegenerative disorders whose main feature is speech and language dysfunction. three main forms of PPA - non-fluent agrammatic variant (nfvPPA), semantic variant (svPPA and logopenic variant (lvPPA) - comprise the canonical syndromes currently recognised in the consensus diagnostic criteria [1,2]. PPA is rare (the estimated prevalence is conservatively around 3 cases per 100,000. Patients are most commonly affected in late middle age, with devastating consequences for family life, work and social functioning [1,2,3].

2. Purpose of the work

The aim of this study is to analyse primary progressive aphasia (PPA) on the conversational process. An evaluation of its impact on the conversation process and the patient's quality of life assessment was made.

3. Material and method

Scientific publications found on the scientific platform PubMed and on monographic publications in the field of neurology were reviewed. Current knowledge was reviewed using the following keywords: "speech disorders", "aphasia", "dysarthria", "primary progressive aphasia".

4. Discussion

4.1 Preliminary criteria for the diagnosis of primary progressive aphasia (PPA)

The criteria must be met:

1. Language difficulties are a major feature of the clinical picture.
2. Language difficulties are a major cause of disability in everyday life.
3. Aphasia is the most significant disorder in the early stages of the disease [2].

Exclusion of diagnosis - answers to questions 1.-4. must be negative for a diagnosis PPA to be possible [2,3] :

1. Are your current symptoms more consistent with other neurodegenerative or somatic conditions?
2. Is cognitive impairment associated with mental disorders?
3. Do episodic memory, visual memory and visuospatial (perceptual) impairments occur at the onset of the disease?
4. Are significant behavioural disturbances present at the onset of the disease [3]?

Table 1 shows the main (canonical) progressive aphasia syndromes diagnosed by the current consensus diagnostic criteria.

Table 1. Effect of aphasia on the patient's speech depending on the type of syndrome [3,4].

Legend:

+ visible or defining impairment;

± variable impairment

- mild or no deficiency

Team name	Production of statements				Comprehension of speech		Repetition of speech	
	Idea	Content	Design	Speaking	Perception	Sense	Words	Sentences
	Uninteresting-agrammatic	±	±	+	+	±	±	+
Semantic	-	+	-	-	-	+	-	-
Logopenic	±	+	+	-	±	±	±	+
Primary progressive apraxia of speech	-	-	-	+	-	-	+	+
Mixed progressive aphasia	±	+	+	±	±	+	+	+
Progressive dynamic aphasia	+	±	±	±	-	-	-	-
Progressive pure anomie	±	+	-	-	-	-	-	-
Progressive dysprosodia	-	-	-	+	±	-	+	+
Progressive deafness to the "pure" word	-	-	±	±	+	-	+	+

In contrast, the following table presents the main (canonical) progressive aphasia syndromes recognised by current consensus diagnostic criteria, as well as other less common variants and atypical syndromes that are also represented in most clinics receiving patients with progressive aphasia and their main linguistic features [3].

Table 2 Major (canonical) progressive aphasia syndromes and other less common variants and atypical syndromes [4].

Team name	Other cognitive deficits and behavioural	Neurological links	Pathophysiology
Uninteresting-agrammatic	Orofacial apraxia > limb apraxia	Parkinsonism, PSP, CBS, MND	Mostly tauopathy; possible PSP, CBD; Alzheimer's disease; TDP-43
Semantic	Prosopagnosia, visual agnosia, other agnosias; lack of inhibition, lack of empathy, obsessions	Usually no	Usually TDP-43, sometimes tauopathy, Alzheimer's disease, rarely mutations
Logopenic	Reduced finger span, limb apraxia, acalculia, visual-spatial agnosia	Myoclonus	Typically, Alzheimer's disease
Primary progressive apraxia of speech	Orofacial apraxia, "Dysexecutive" syndrome, limb apraxia	Parkinsonism, PSP, CBS, MND	Usually tauopathy, may be PSP, CBD
Mixed progressive aphasia	The "Dysexecutive" team	Parkinsonism	GRN gene mutation
Progressive dynamic aphasia	The "Dysexecutive" team	Parkinsonism, PSP, CBS	PSP, CBD
Progressive pure anomie	No	no	Unspecified
Progressive dysprosodia	Dysexecutive syndrome, oro-facial apraxia	Unspecified	Unspecified
Progressive deafness to the "pure" word	Cortical deafness, auditory agnosia	Variables	Unusual

Legend to table :

PSP - progressive supranuclear palsy

CBS - corticobasal syndrome

MND - motor neurone disease

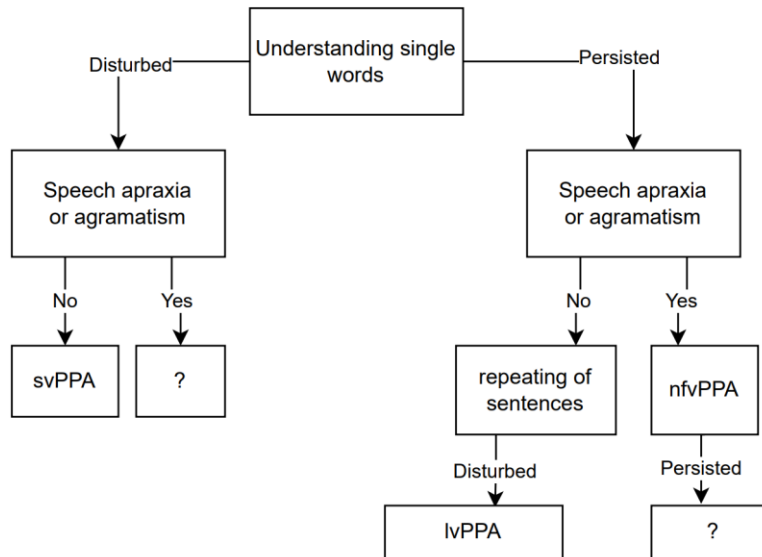
Table references :

- ***Tauopathy*** - a group of diseases caused by abnormalities associated with microtubules of the tau protein.
- ***TDP-43 TAR DNA-binding protein 43*** - a protein consisting of 414 amino-acid residues, with a mass of 43 kDa, in humans encoded by the TARDBP gene, which is expressed in almost all tissue types [5,6,7].

Recommendations for the diagnosis and classification of PPA have been published (Gorno-Tempini et al., 2011). The guidelines defined each variant according to the

presence or absence of basic language features, with recommendations on the type of assessment required to classify subjects [8,9]. The effects of the study are shown in the figure below.

Figure 1. Summary of international recommendations for the diagnosis and classification of PPA [8].



According to the study, on the basis of a single-word comprehension disorder without additional apraxia of speech or agrammatism, we can classify PPA as svPPA. In contrast, single word comprehension with apraxia of speech or agrammatism is classified as nfvPPA, while the absence of apraxia of speech or agrammatism with sentence repetition disorder is classified as lvPPA.

4.2 Speech disorders in nfvPPA

There are difficulties in communicating in the work environment, in offices, during social gatherings or telephone conversations. In addition, grammatical errors and articulatory difficulties have been found, as well as problems in forming complex statements [9]. In sentence construction, short single sentences predominate, followed by the patient using single words, often taken from the interlocutor's question. Grammatical errors in Polish

usually include incorrect sentence formation, difficulties with the use of tenses and modes (predominance of the present tense and the indicative mode in statements) and incorrect use of inflectional endings [10]. Another characteristic feature of the speech of patients with nfvPPA is apraxia of speech manifested by difficulties with rapid changes in articulatory alignment, but also altered accent and intonation. Difficulties with sentence construction in the patient's speech are usually associated with a predominance of nouns over verbs [11]. Importantly, a patient with nfvPPA does not exhibit word comprehension difficulties or behaviours suggestive of the presence of a semantic disorder. Instead, the patient may have difficulty understanding an interlocutor's speech or sentence. In addition, depression, apathy and irritability are often observed in patients [12].

4.3 Speech disorders in svPPA

Initially, the patient's condition manifests itself mainly through impoverished vocabulary. Gradual semantic deprivation involves all modalities (visual, auditory, tactile, olfactory and gustatory recognition) and the patient loses not only the ability to name individual objects but also to recognise their specific characteristics [3]. Patients speak fluently and grammatically correctly, but utterances lack nouns and are therefore poor in content [4]. Verbs dominate the utterances. Patients with svPPA, as with the behavioural variant of FTD, reveal behavioural disorders and personality changes.

4.4 Speech disorders in lvPPA

Patients have problems updating words. They do not have problems understanding single words. Patients speak grammatically, prosodically and articulatively correctly [10, 11]. There are often pauses in their speech resulting from word actualisation problems. In addition, they have difficulty repeating and understanding long utterances due to short-term memory impairment [11,12,13].

4.5 Treatment methods for speech disorders in PPA

People with PPA should be routinely referred for speech therapy interventions. There are currently no disease-modifying drugs for PPA. There is a low threshold to try a cholinesterase inhibitor or memantine in patients with lvPPA and nfvPPA (where Alzheimer's disease pathology is considered), although any benefit is usually modest and caution should be exercised to avoid exacerbation of behavioural symptoms [14,15]. Memantine appears to be well tolerated in svPPA and nfvPPA, but clear evidence of benefit is lacking. In patients with comorbid depression or anxiety, selective serotonin reuptake inhibitors should be considered, which may help to alleviate behavioural symptoms such as impulsivity and aggression, particularly in svPPA [15,16]. Newer-generation neuroleptics may be indicated to treat severe agitation or psychotic symptoms later in the course of the illness [16,17].

5. Summary and conclusions

Speech disorders in PPA result in a deterioration of speech fluency and, over time, there may even be difficulties in formulating sentences correctly grammatically. PPA significantly affects both the production and comprehension of speech. It affects the idea, content, construction and delivery of speech by the patient. In addition, it causes significant difficulties in the repetition of speech, both complete sentences and words alone. It demonstrates a significantly negative impact on patients' quality of life.

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