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Impact of neurological disorders occurring in neurodegenerative diseases on the conversation process and patients' quality of life

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Abstract

Introduction : Dialogue, or conversation, is the ability to speak as well as listen. It plays a very important role in interpersonal relations. Speech disorders are a group of of disorders that includes various types of speech production difficulties. Speech production depends on the coordination of several motor acts: respiration, phonation, articulation, resonance and prosody. Phonation is the complex activity of producing sound by vibration of the vocal cords. Resonance is the amplification of sounds at specific frequencies through vibration in the chest, throat and sinuses. Prosody includes a set of phonetic features, such as tone, loudness, tempo, and the overall timbre of speech, which are designed to intended to preserve the meaning and character of speech. Disruption of any of these related elements results in motor speech disorders.

Material and method : An overview is presented of language disorders in diseases neurodegenerative diseases such as Parkinson's disease(PD) and Friedreich's ataxia (FRDA). Their impact on the conversation process and the patient's quality of life assessment was evaluated. The paper was based on scientific publications posted on the PubMed scientific platform.

Results : Speech and language disorders are the main symptoms of FRDA, which significantly affect the patients' quality of life. Patients often present with dysarthria, resulting from causes of central and peripheral and additional primary language disorders. The classic speech disorder in PD is hypokinetic dysarthria, manifested by hypophonia; monotonous, uneven and strained speech; reduced prosody (melodic voice); crowded articulation; inadequate pauses in speech; freezing of speech, unclear articulation.

Conclusions : The described disease entities show a significant impact on the deterioration of the process of of conversation by the patient, thus showing a significantly negative impact on the patients' quality of life.

Keywords: speech disorders; aphasia; friedreich ataxia; dysarthria; ataxia

1. Introduction

Dialogue, or conversation, is the ability to both speak and listen. It plays a very important role in interpersonal relationships. Speech disorders are a group of disorders encompassing different types of speech difficulties, including difficulties in articulation, speech defects, use of inappropriate words, and are therefore related to articulation, phonation, tone of voice, fluency. All these factors cause difficulties in understanding the spoken message. Speech disorders significantly limit the dialogue of the sufferer. They make it confusing and difficult to establish contacts. According to Irena Stryczek classified into extra- and intra-individual speech disorders. According to WHO, speech disorders distinguish between speech development delay (in the context of active speech delay) and speech development disorders resulting from:

- hearing disorders peripheral and central,
- damage to the central speech sensory and motor centers on multiple CNS floors,
- mental retardation accompanying cerebral palsy,
- emotional states and behavioural disorders,
- inheritance, other unclassified, combinations of several causes [1].

Under the heading of speech disorders were highlighted:

- developmental neurological speech disorders,
- alalia, dyslalia acquired speech disorders resulting from damage to the speech apparatus or, according to other definitions, developmental disorders,
- aphonia, dysphonia impaired phonation, voicelessness and hoarseness caused by damage to the larynx or psychogenic factors,
- aphasia, anartria, dysarthria speech disorders acquired as a result of damage to the nervous system,
- paraphasia [1,2].

Speech disorders are a group of disorders encompassing different types of difficulties in speech production. Speech production depends on the coordination of several motor acts: respiration, phonation, articulation, resonance and prosody [2]. Phonation is the complex act of producing sound by vibrating the vocal cords. Resonance is described as the amplification of sounds at specific frequencies by vibration in the chest, throat and sinuses. Resonance can be altered by voluntary control of the position of the throat, tongue, jaw, lips and larynx. Articulation is the creation of individual sounds controlled by the lips, tongue, palate and pharynx. Prosody includes a set of phonetic features, such as tone, loudness, rate, and overall timbre of speech, which are designed to preserve the meaning and character of an utterance. Disruption of any of these related elements results in motor speech disorders.

1. Purpose of the work

The aim of this study is to analyse the impact of neurodegenerative diseases on the conversational process. An overview of speech disorders in neurodegenerative diseases such as Parkinson's disease(PD) and Friedreich's ataxia (FRDA) is presented. Their impact on the conversational process and the patient's quality of life assessment is assessed.

2. Material and method

The paper will be based on scientific publications found on the scientific platform PubMed and on monographic publications in the field of neurology. Current knowledge was reviewed using the following keywords: "speech disorders", "aphasia", "dysarthria", "ataxia", "Friedrieich's ataxia".

3. Speech disorders in Parkinson's disease

3.1. Pathophysiology of speech disorders in Parkinson's disease

It has been found that in Parkinson's disease there are two main assumptions about the pathophysiology of speech disorders. These are :

- Dopamine deficiency □ hypokinetic dysarthria is led to by progressive degeneration of the nigrostriatal pathway, resulting in impaired motor function. Dysarthria is associated with the main motor symptoms of parkinsonism, i.e. muscle rigidity, monotonic, hypophonic voice sound, 'prominent' articulation [3]. Bradykinesia has been explained by slowness of response resulting from difficulty in initiating movements. The tremor may affect the tongue, jaw or mouth [3,4].
- Abnormal perception of voice loudness and impaired sensory processing lead to impaired speech production. PD patients perceive the loudness of their speech as normal and the speech of others as crying. Such patients do not begin to speak louder at a distance from the speaker without clear cues [4].

3.2. Hypokinetic dysarthria

The classic speech disorder in PD is hypokinetic dysarthria. Dysarthria is one type of speech disorder resulting from dysfunction of the executive apparatus (tongue, palate, pharynx, larynx) [5]. The dysfunction can be caused by: damage to the muscles, the cranial nerves innervating them, the nuclei of these nerves, the corticospinal tracts, the extrapyramidal system. As a result of their damage, so-called dysarthric speech is produced, characterised by being slow, indistinct, aphonic (i.e. voiceless) with a so-called nasal suprasound, which is caused by the drooping of the soft palate and gives the impression of speech with "noodles in the mouth" [5,6]. Vowel formation is usually preserved and, depending on the site of the lesion, articulatory abnormalities predominate:

- labial consonants (b, p, w, f),
- palatal consonants (g, k, h),
- consonants dependent on language function (d, t, r, s) [6].

Respiratory disorders in the form of reduced thoracic volume were also observed, leading to more restricted airflow through the vocal tract compared to healthy subjects. Disorders of non-motor aspects of speech are less obvious and mainly related to cognitive or affective deficits, increasing gradually but also leading to impaired communication through the formation of a mask-like face, which creates the impression of a lack of emotional involvement, a detachment of the patient. The masked face has even been shown to affect the impressions of doctors, who may misjudge the patient's motivation. The ability to recognise callers' emotions is impaired in some patients. Eye contact time and the number of gestures may also be reduced. As a result of impaired perception, PD patients overestimate the volume of their own speech and lose the ability to adjust their voice volume appropriately. In colloquial speech, PD patients use shorter and simpler sentences, logically incomplete, often with incorrect grammatical structure [6,7]. Patients have difficulty naming actions and the number of verbs in their speech decreases. Which ultimately leads to patients suffering from anxiety and depressive conditions certainly experience severe communication difficulties. Such patients are also at greater risk of experiencing social isolation. Limited communication with people leads to a faster progression of cognitive deficits, and the absolute risk of mortality increases as social vulnerability increases. In addition, communication problems can lead to impaired access to and use of health services [5].

3.3. Impact of speech disorders on quality of life in people with Parkinson's disease

Findings show that between 70 and 90% of PD patients have difficulties in communication, but most of them do not pay due attention to these disorders. However, speech disorders affect their lives to such an extent that 30% of PD patients describe speech disorders as the most disabling manifestation of the disease [3]. Patients with dysphonia experience psychological discomfort, and in some cases it has even been suggested that the presence of dysphonia contributes to the development of depression in patients. It has also been reported that patients with PD experience a communication deficit that affects their socialisation.

3.4. Methods for the assessment of speech disorders in Parkinson's disease

The following methods of assessing speech disorders in Parkinson's disease have been highlighted:

- if the severity of symptoms varies during the day, it is better to carry out the test both with and without the medication. The non-motor symptoms questionnaire also includes questions that may indicate communication difficulties and require further investigation. The Voice Impairment Index focuses on the impact of speech impairment on the patient's quality of life [3,5],
- dysarthria impact profile → scales such as Robertson dysarthria Profile and Frenchay dysarthria assessment are used to analyse disorders in general in the sound production process,
- dysarthria interaction profile,
- audio or video recordings of vowel pronunciation, rapid syllable repetition, reading various passages of text, describing images, dialoguesand spontaneous monologues on any topic, which are conducted at regular intervals of 6-12 months. The recordings can be analysed in specialised laboratories using traditional methods to measure sound pressure levels, fundamental frequency, formant frequencies, rate and rhythm of speech[3].

3.5. Treatment methods for speech disorders in Parkinson's disease

Speech disorders in Parkinson's disease have found a solution through both pharmacological and surgical treatment [7]. A distinction is made between drugs such as:

levodopa → the use of levodopa has variable results for speech disorders in PD. Some patients experienced an increase in loudness and improved speech intelligibility, while others did not. In addition, long-term treatment with levodopa can lead to complications, including orofacial or respiratory dyskinesia, dystonia and fluctuations. Levodopa improves vowel articulation. However, the results of some studies indicate that the use of levodopa alone for long periods of time or in high doses may result in decreased speech melodicity[3,7],

clonazepam → at a dose of 0.25-0.5 mg / day can alleviate speech symptom disorders [7].

In addition, we distinguish treatments and therapeutic methods such as:

- L. Silverman voice therapy → this method aims to get the patient to speak louder without tension, which optimises the respiratory, laryngeal and articulatory muscles and leads to an increase in voice volume, improved speech clarity. LSVT also includes sensory reward training and self-control training to maintain treatment results. Clinical effectiveness of LSVT in increasing voice volume, speech quality in patients with PD has been confirmed and the effect persists for 2 months after cessation of therapy. LSVT can be performed at any stage of the disease, but the greatest efficacy has been demonstrated in the early stages[3,7].
- rTMS → rhythmic transcranial magnetic stimulation; a non-invasive method that causes depolarisation and hyperpolarisation of neurons. It uses electromagnetic induction to induce a small electrical voltage using a rapidly changing magnetic field. This can cause activity in specific parts of the brain and induce slight discomfort, allowing the brain and its connections to be studied. It has been shown that a course of 10 sessions of high-frequency rTMS of the left sensorimotor area in the oro-facial muscle projection can lead to an increase in tongue motor activity, as well as an improvement in voice quality[3].

4. Speech disorders in FRDA

4.1. Pathophysiology and epidemiology of speech disorders in FRDA

Friedreich's ataxia (FRDA) is an autosomal recessive cerebellar spinal ataxia. It is the most common hereditary ataxia in Europe. The incidence of FA worldwide is 1 in 40,000. It has an autosomal recessive inheritance pattern and symptoms usually appear in childhood. Unfortunately, symptoms worsen over time, so most affected people eventually need mobility aids such as wheelchairs, lose their vision and hearing and develop other medical complications such as diabetes and scoliosis [2,8,9,10]. Speech and language disorders are the main symptoms of Friedreich's ataxia (FRDA), which

significantly affect patients' quality of life. Patients often present with dysarthria, resulting from central and peripheral causes and additional primary language disorders. Facial weakness, slurred speech and dysphagia are caused by the disease affecting cranial nerves VII, X, XII. Dysarthria occurs due to cerebellar involvement [8,9]. Speech disorders have peculiar features, although varying in patients, and progress over the course of the disease.

4.2. Symptoms of dysarthria in FRDA

Dysarthria is a common and early symptom occurring in more than 90% of people, which progresses with the duration of the disease. Speech becomes slow and slurred, worsening intelligibility in advanced cases. A study of 38 people with FRDA found that 68% had mild dysarthria characterised by consonant inaccuracy, reduced pitch variability, impaired loudness maintenance, reduced phrase length, slurred speech and impaired breath support for speech[10]. Dysphagia occurs when the muscles responsible for swallowing weaken. Mild dysphagia is again a common symptom and can become problematic in advanced disease, sometimes requiring 'through the skin' implantation of an endoscopic gastroesophageal tube. Patients may cough or choke on solids or liquids, including saliva. Dysphagia combined with lack of coordination of speech and swallowing can lead to choking [8,10].

4.3. Hearing disorders in FRDA

Problems due to auditory neuropathy are a common and underestimated problem that can be very socially disabling even in the early stages of the disease. Reported incidence of hearing loss in case series varies widely from 8 to 39% [7,9,10]. Recently, it has been shown that binaural speech processing is impaired in FRDA patients with electrophysiological signs of auditory neuropathy [10].

4.4. Treatment of speech disorders in the FRDA

No effective treatment is currently known. There is ongoing research into the possible therapeutic use of antioxidants, histone deacetylase inhibitors, iron chelating compounds and compounds that increase mRNA transcription for frataxin [9,10].

5. Summary and conclusions

Speech and language disorders are the main symptoms of FRDA, which significantly affect patients' quality of life. Patients often present with dysarthria, resulting from central and peripheral causes and additional primary language disorders. The classic speech disorder in PD is hypokinetic dysarthria, manifested by hypophonia; monotonous, uneven and strained speech; reduced prosody (melodic voice); crowded articulation; inappropriate pauses in speech; freezing of speech; unclear articulation [9,10]. The disease entities described show a significant impact on the deterioration of the patient's conversational process, thus showing a significantly negative impact on the patients' quality of life [8,9].

Literature

[1] José Antonio Pinto, Renato José Corso, Ana Cláudia Rocha Guilherme, Sílvia Rebelo Pinho, Monica de Oliveira Nóbrega, Dysprosody nonassociated with neurological diseases-a case report , Volume 18, Issue 1, P90-96, March 01, 2004 DOI:https://doi.org/10.1016/j.jvoice.2003.07.005.

[2] Boschi Veronica, Catricalà Eleonora, Consonni Monica, Chesi Cristiano, Moro Andrea, Cappa Stefano F., Connected Speech in Neurodegenerative Language Disorders: A Review, Frontiers in Psychology, 2017, p.269, DOI=10.3389/fpsyg.2017.00269.

[3] Arefieva A.P., Skripkina N.A., Vasenina E.E. Speech disorders in Parkinson's disease.
Journal of Neurology and Psychiatry. S.S. Korsakov. Special Problems. 2019; 119 (9-2):
32-36.

[4] Khashayar Dashtipour, Ali Tafreshi, Jessica Lee & Brianna Crawley. Speech disorders in Parkinson's disease: pathophysiology, medical management and surgical approaches, Future Medicine 2018.

[5] Kara M. Smith, David N. Caplan, Communication impairment in Parkinson's disease:
Impact of motor and cognitive symptoms on speech and language, Brain and Language,
Volume 185, 2018, Pages 38-46, ISSN 0093-934X,
https://doi.org/10.1016/j.bandl.2018.08.002.

[6] Wilson S. M., Brandt T. H., Henry M. L., Babiak M., Ogar J. M., Salli C., et al. ... Inflection almorphology in primary progressive aphasia: an anelicited production study. Brain Lang. 2014; 136, 58-68. 10.1016/j.bandl.2014.07.001.

[7] Armstrong MJ, Okun MS. Diagnosis and Treatment of Parkinson Disease: A Review.JAMA. 2020;323(6):548–560. doi:10.1001/jama.2019.22360.

[8] Schirinzi, T., Sancesario, A., Bertini, E. et al. Speech and Language Disorders in Friedreich Ataxia: Highlights on Phenomenology, Assessment, and Therapy. Cerebellum 19, 126-130 (2020). https://doi.org/10.1007/s12311-019-01084-8.

[9] Arnulf H. Koeppen, MD, Joseph E. Mazurkiewicz, PhD, Friedreich Ataxia: Neuropathology Revised, Journal of Neuropathology & Experimental Neurology, Volume 72, Issue 2, February 2013, Pages 78-90, https://doi.org/10.1097/NEN.0b013e31827e5762.

[10] Bidichandani SI, Delatycki MB. Friedreich Ataxia. 1998 Dec 18 [Updated 2017 Jun1]. In: Adam MP, Ardinger HH, Pagon RA, et al., editors. GeneReviews® [Internet].Seattle (WA): University of Washington, Seattle; 1993-2021.