Case Report

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Speech Rehabilitation in Wilson's Disease: A Case Study

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ABSTRACT

Wilson's disease is a rare hereditary disorder passed down in the autosomal recessive way. This disorder involves the speech parts of the brain leading to dysarthria, which impairs all of the five speech systems, i.e. the respiratory, phonation, articulation, resonance, and prosody. The patient studied in this research was a 28-year-old woman with Wilson's disease, who visited Rofaydeh Rehabilitation Hospital in Tehran City with complaints about severe speech disorders. Based on the clinical and paraclinical examinations the patient was diagnosed with a decrease in the maximum phonation time (MPT) of 2 to 3 seconds, reduced intelligibility and articulation impairment. The patient underwent medicinal, behavioral, and rehabilitation (include speech therapy) treatments. Following a continuous two-year follow-up rehabilitation, a considerable improvement in the speech was observed as an increase in intelligibility (up to 5% of the words), the consistency between respiration and speech and an increase in verbal and nonverbal communications.

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Introduction

Wilson's disease (WD) is a rare metabolic disorder by which copper sediments in different organs such as the liver, brain, eyes, and kidneys (1). The prevalence of this disease is approximately 1/100000 or 18 to 30 in a million (2). This disease manifests in individuals aged between 5 and 50 years (3). In this disorder, the related neurological symptoms affect the central nervous system through the concentration of copper (4), and some of the areas of brain affected by this condition are the basal ganglia, the brainstem, and the cerebellum (5). Wilson's disease considerably impairs the function of areas such as the basal ganglia and cerebellum (6). Since the basal ganglia is involved in the regulation of the rate and sequence of speech including the silence between the consecutive syllables and words, any damage to this region can result in motor abnormalities such as dysarthria and dysphagia (7). Dysarthria is an important and primary neurologic disorder in Wilson's disease and is often characterized by a decrease in the speech rate and impairment of intelligibility (8). In Wilson's disease, dysarthria is comorbid with dystonia, which can affect the

articulation muscles including the tongue, face, and jaw and even lead to mutism (9).

As reported (10) there is a significant relationship between abnormal copper metabolism in Wilson's disease and speech rate disorders. Speech is usually affected in the early stages of the disease, leadings to slurred speech of the hypokinetic dysarthria type (2). In a study, it was indicated that a decrease in the maximum speech time is substantially associated with a decrease in the tremor frequency (11). One of the important characteristics of speech rate disorders in patients with Wilson's disease is reduced intelligibility, imprecise consonants, hypernasality, inappropriate silence, hypophonia, variation of loudness, and abnormal stress (12). These patients suffer dysarthria, as well as ataxia, rigidity, tremor, cognitive impairment, and mood disorders (1). However, the research by Ono and Kurisaki on patients with abnormal zinc metabolism revealed that the patients displayed involuntary movements and dysarthria as well as dementia (4). In another study it is stated that mixed dysarthria and hypotonia caused by the destruction of copper in the extrapyramidal system and the motor system components (12). In fact, one of the complications of dysarthria is reduced intelligibility, which significantly reduces the communication functions (13). In these conditions, verbal and non-verbal communications are impaired in patients with Wilson's disease (14). However, the treatment of speech disorders in patients with Wilson's disease is substantially important. In addition, medicinal and surgical treatments (15), as well as speech rehabilitation, are also administered. Given the limitedness and shortage of research on the rehabilitation of patients with Wilson's disease in Iran, it was decided to conduct the present study. Hence, the overarching goal of this case report was to investigate the effect of speech therapy along with medicinal treatment on different speech and communicative disorders.

Case Report

A 28-year-old woman with a height of 160 cm and a weight of 60 kg suffering from Wilson's disease since the age of 10 was visited at Rofaydeh Rehabilitation Hospital in Tehran. She received therapeutic and rehabilitation services including speech therapy, and underwent liver transplantation five months before the treatment. She suffered gait disturbances and involuntary movements one month before the live transplantation and used a wheelchair. She stopped taking her medicine one year ago after observing improvements in her condition. However, after ceasing to take the medicine, she developed liver failure, hoarseness, dysphagia, and drooling. She was in a very poor physical condition when visited the Rofaydeh Rehabilitation Hospital. She was dealing with quadriplegia, spasticity of the tongue muscles, speech disorder, excessive crying, and excessive speech rate. However, a thorough speech and language assessment was carried out on her.

- Language assessment: According to the Persian version of the Diagnostic Aphasia Battery (DAB-1) (16) the patient had no language problem.

- Oral assessment: According to Persian version of the Diagnostic Aphasia Battery (P-DAB-1) (16) the patient was suffering slowness and weakness of tongue and limited oral motion range in nonverbal fluency . The patient was only capable of bringing her tongue out of her mouth and bringing the tip of the tongue to the corners of the lips less than 5 times within 5 seconds. However, the patient was completely capable of puckering five times within 5 seconds.

- Speech assessment: The studied speech components and the assessment results are presented in Figure 1. The sound of the individual was recorded using a voice recorder (Sony LCD-UX543). Thereafter, using Praat software version 6.0.35, the length and pauses of speech were examined.

- It is worth stating that the patient had inadequate respiratory support, and signs of reduced loudness, short phrases, and decreased pitch. According to the examination of the maximum phonation time (MPT) as a clinical tool for the assessment of phonation mechanisms, it varied between 2 and 3 seconds and was extremely low. However, the standard MPT for adult women is approximately 20.96 seconds. As stated, the speech rate of the patient was extremely higher than the average speech rate of adults (105 words per minute) (16). In addition, it was found that intelligibility of the patient was extremely low and almost incomprehensible based on the scale presented in Table 1.

- Various methods have been introduced by Gordon-Brennan and Hudsonfor assessing

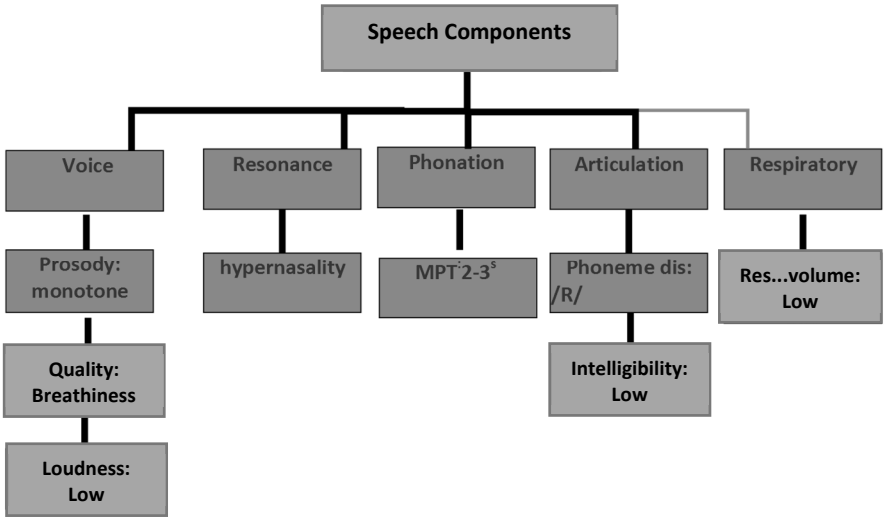

Figure 1. Report of the speech assessment

Table 1. Parnell and Day Speech Intelligibility Scale

Intelligible	0= Easily intelligible
	1= Intelligible with a few words intelligible only through context
Unintelligible	2= More than half the words unintelligible (regardless of meaning obtained)
	3= intelligible completely, topic and general meaning of utterance not understood

intelligibility. Among them was a method that utilized a word list in which continuous word or speech, the percentage of recognizable words were calculated. The method of scoring based on the number of words understood by the listener was calculated (Table 1).

- Since intelligibility influences the patient's communicative function, it was important to assess it. However, the identification of this speech component in dysarthric speech is limited by factors such as articulation, phonemes, loudness, speech rate, and the rate and power of articulators (17). In addition, the phoneme omission error was observed in the phonological process.

For the purpose of calculating percentages, a sentence score of 0 or 1 would be considered intelligible and a score of 2 or 3 would be considered unintelligible. Moreover, the Functional Independence Measure (FIM) was used to assess the independence, self-care, and comprehension of the patient, who obtained an FIM score of 103. However, the FIM total score ranges from 18 to 126, and a score higher than 97 allows for the release of the patient. This 18-item scale consists of the communication and social comprehension subscales. The score of each subscale varies between 1 and 7 (18).

Therapeutic Intervention

The patient underwent various therapies such as medicinal and surgical treatments, liver transplantation, behavioral therapy, and rehabilitation (physiotherapy, speech therapy, and occupational therapy) in the course of her disease.

- Medicinal treatment: The medicines used by the patient were Myfortic, Prograf, Zinc sulfate, Co-trimoxazole, and Prednisolone.

- Behavioral therapy: A specialized clinical psychologist administered the behavioral therapy due to the changes in the patient's psychological condition.

- Speech therapy: The following exercises were performed considering the patient's speech conditions in the course of the treatment (two consecutive years).

Diaphragmatic breathing: Given the patient's poor respiratory support, the diaphragmatic breathing exercise was used to control and optimize the patient's respiratory performance, facilitate the contraction of the diaphragm, and make the best of the lung capacity (with minimum respirator). It was because changing the breathing pattern during articulation can foster the respiratory support (19). Hence, the patient was asked to lie down and relax. The therapist put one of his/her hand on the abdomen and the other on the chest of the patient. After nasal inhalation the patient's abdomen would expand and during exhalation through the mouth the patient's abdomen would

contract. This exercise met the desired goals and prevented reversed breathing (20). After 4 weeks of intense workout, the patient's MPT increased from 3 seconds to 8 seconds. The percentage of improvement in breathing capacity compared to the initial condition was about 50 percent.

Melodic Intonation Therapy (MIT): This therapeutic method suits patients with satisfactory comprehension yet poor speech such as patients with Broca's aphasia. In this method, the non-dominant hemisphere, i.e. the right hemisphere, is used in the speech exercises (21). Since the patient's speech was monotonous it was decided to use this method to create a rhythmic and controlled melody, increase loudness, and improve the stress patterns. The auditory feedback was also used to increase loudness. Considering the collected speech samples from the individual using Praat software, improvement in pause, pitch, and loudness before and after treatment were examined. At the end of the treatment period, the patient started using appropriate silence and increased pitch and loudness in her speech and was capable of controlling her speech rate. This outcome not only increased intelligibility but also increased the comprehensibility of her speech for others.

Oral movement therapy: This exercise was performed to facilitate and balance the tonicity of the mouth muscles, reinforce tongue movements, and increase the harmony between respiration and speech in the patient. For the purpose of this exercise, the patient was assigned the verbal diadochokinesis task (e.g. /pâ tâ kâ/) and the nonverbal diadochokinesis task. The rate of diadochokinesis was calculated based on the number of iterations per 10 seconds (18). Following the treatment period, the patient was capable of moving her tongue upward and managed to articulate 80% of /pâ tâ kâ/ accurately within 10 seconds. In addition, this exercise enabled the patient to correct her articulation of the /R/ phoneme and utter it properly when it was used in the beginning, middle, and end of the words. Hence, in the 300-word speech exercise performed at the end of the treatment, her speech intelligibility increased by approximately 50%, yet some of her words were still unintelligible. High-frequency nouns including spoon, plate, glass, and fruits such as apple, banana, and clothes shelf had higher intelligibility compared to verbs.

Muscle Facial Treatment (MFT): MFT is focused on the orofacial muscles and their proper function (22). This therapeutic approach was used to increase the harmony between the lip movements, reinforce the lip muscles, increase the strength, length, resistance, and flexibility of the orofacial muscles, and prevent drooling. This exercise enabled the patient to control the range of the motions and the sensory feedback of her orofacial muscles. In addition, it considerably contributed to the correction of the articulation errors, fluency, and intelligibility in the patient.

Discussion

Firstly, given the limited studies on speech rehabilitation in patients with Wilson's disease, it seems necessary to pay more attention to this area. Secondly, in most studies it is stated that the rate of speech in patients with dysarthria decreases (8, 11), though in the present case the speech rate in the patient was extremely higher than normal which calls for attention. In line with the other studies, reduced intelligibility, imprecise consonants, hypernasality, and inappropriate silence were reported in patients with Wilson's disease. For instance, two speech therapists with more than 10 years of experience of speech therapy conducted a study to examine intelligibility in patients with Wilson's disease using a 300-word continuous speech example. They reported that more than half of the words were unintelligible (23). The results of this study were in line with and convergent to their findings.

In terms of anatomy, changes in the above mentioned factors were associated with damage to the basal ganglia (24). However, in the present study, the connection between the exact anatomical location and the mentioned damages was not examined. Among the outstanding characteristics in these patients is dysarthria, which is considered the main cause of the reduction in speech intelligibility (8). According to the research reports, the main problem in Wilson's disease is the lack or the inadequacy of intelligibility, which causes severe problems in establishing verbal communications and social interactions (13, 25). Considering the chief complaint of the patient with regard to communication with friends and family was a weakness of speech intelligibility, enhancing this factor became one of the most important objectives in the treatment of the patient. Clear

enhancement of communication between the patient and their friends and family following treatment is evidence of the apparent effectiveness of the increase in speech intelligibility on personal communication. However, considering the other factors involved in speech disorders in patients with Wilson disease, further inquiry in this regard is necessary.

The research by Tagawa et al. on 3 patients suffering abnormal copper metabolism revealed that involuntary movements and dysarthria are associated with abnormal copper metabolism. Moreover, two of their patients were not suffering from dementia (19). Therefore, there is no strong evidence implying patients with abnormal copper levels have dementia.

The patient in the present study was also not diagnosed with dementia. The findings from another study confirm the involuntary movements and dysarthria in patients with abnormal copper metabolism (4). As stated, involuntary movements of the head, neck, and upper organs and dysarthria were evidently clear in our patient. Hence, considering the related studies it could be argued that speech rehabilitation must be considered a priority in treating patients with Wilson's disease along with medicinal treatments and intensive care. Therefore, given the speech dysfunctions, one of the important treatments for the rehabilitation of patients with Wilson's disease is speech therapy, which is effective in the long run (10). As a result, intelligibility increases in the patient and significantly contributes to the establishment of better verbal communications. In our patient, the increase in intelligibility was accompanied by decreased speech rate and improved verbal communication. It also influences flexibility and reinforcement of the psychological conditions of the patients.

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