Multiple Sclerosis

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Multiple Sclerosis (MS) is a long-lasting neurogenic progressive disease that is characterized by a chronic inflammation of the central nervous system (CNS), mostly affecting the brain and the spinal cord [1]. Next to inflammation, MS causes demyelination to occur in the white matter of the central nervous system (CNS).

The pathophysiology of MS is a complex process in which the body's immune system attacks the myelin sheaths surrounding nerve fibers, resulting in scars (also known as "plaques" or "sclerae") which will cause impaired transmission of neural signals in the CNS. There is no known cause for MS [2]. Currently, there is no cure for MS. Treatments consist of medication to slow disease progression or target-specific symptom to improve quality of life. In recent years, there has been an emerging focus on healthy lifestyle (like the use of a Mediterranean diet) and regular aerobic exercise.

MS affects around 30 per 100.000 people globally, and the initial manifestations of the disease are evident during early adulthood, between the ages of 20 and 40. MS mostly affects females, with a female to male ratio of 3:1 [3]. The cause of MS is still unknown; however, specialists believe that MS is caused by several different factors including immunologic, environmental, and genetic factors that result in a permanent deterioration of the nerves. MS is known to be more common in Caucasians of Northern European decent compared to persons with other ethnical backgrounds. Possible factors that might play a role in the pathogenesis of MS are geographical, physical environment (including exposure to sunlight and vitamin D) and socioeconomic factors, although relationship to causality remains unclear [2]. In the last decennia, the incidence of MS has increased. The reason for this increase is not clear, with possible contributors including increased awareness of the disease, improved access to medical care, and enhanced diagnostic measures [4].

MS is diagnosed based on strict criteria (McDonald criteria), that are based on the number of episodes where a person shows signs of an "acute inflammatory demyelinating event" in combination with the existence of lesions (plagues) in MS-typical regions of the central nervous system (such as periventricular, juxtacortical, infratentorial, or spinal cord), as confirmed by MRI [2].

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There are four types of MS including relapsing remitting multiple sclerosis (RRMS), secondary progressive multiple sclerosis (SPMS), primary progressive multiple sclerosis (PPMS), and progressive relapsing multiple sclerosis (PRMS) [5, 6]. As the cause of MS is unknown, it is also unknown why these four different types of MS develop [2]. The most common variant is RRMS, which begins with a single attack and is then followed by relapses over time [5]. In SPMS, symptoms get more severe gradually over time, with or without the incidence of a relapses or remissions [6]. PPMS is not very common and occurs in 10% of people with MS [5]. It is characterized by slow aggravation of symptoms without relapses or remissions [5]. PRMS is the least common type of MS and only occurs in 5% of people with MS [5].

The symptoms of MS vary widely from one person to another depending on the location and size of the lesions in the brain and spinal cord [7]. However, MS is mainly characterized by loss of muscle strength in various muscle groups in the body [8]. MS is also characterized by fatigue, which is the sensation of tiredness, lack of energy, and exhaustion [9]. Other major symptoms of MS include general weakness, vision problems, mobility problems, cognitive problems, spasticity, numbness and tingling in the extremities, and bowel and bladder dysfunction [10].

The incidence of swallowing problems, voice problems, and communication problems in people with MS is high. In a recent study, 38% of people with MS report frequent coughing and choking while eating and drinking [11]. Another 43% of people with MS reported problems with controlling the loudness of their voice, specifically that other people find it difficult to hear their voice. Another 31% reported that their speech rate was slower than normal. Up to 68% reported word finding problems (i.e., productive language) and 36% reported "difficulty making sense out of what people say to me" (i.e., receptive language). These problems with speech, language, and swallowing did not correlate with the time since diagnosis of MS, suggesting that these impairments can occur at any stage of the disease [12]. Speech, language, and swallowing problems, however, are correlated with a negative effect on social activities and are associated with depressed feelings in people with MS [4].

Swallowing Problems in Multiple Sclerosis

Persons with MS often experience dysphagia [6, 13, 14]. Dysphagia is due to a combination of impairments in the CNS including the corticobulbar tracts, cerebellar, brainstem, and lower cranial nerves [13]. The location and degree of impairment in the CNS determine how dysphagia is manifested in adults with MS [15]. The definite frequency of dysphagia in MS is unknown; however, it is estimated to range from 30% to 40% [13].

Dysphagia may be chronic or intermittent in people with MS and differs from patient to patient [16]. It can appear in early stages of MS; however, it is more common in progressive stages of the disease [17]. It can also appear in people with MS with a mild impairment; however, it worsens in adults who have moderate to severe brainstem impairment [13, 18]. A study of seven adults with MS found that some of these adults had dysphagia before getting diagnosed with MS [16]. Other adults presented with dysphagia shortly after diagnosis [16]. One patient showed symptoms of dysphagia before getting diagnosed with MS, and after her first remission, those symptoms disappeared [16].

Symptoms of dysphagia in MS may be due to impairments in the oral and/or pharyngeal phases of swallowing [13]. Symptoms can range from mild discomfort in the oral cavity or pharynx when swallowing to an inability to masticate solid food or swallow safely, as aspiration is a frequent finding in people with MS [19]. The effects of dysphagia may be rather hazardous to the patient, both physically and socially [20]. Some of the most frequent physical symptoms reported by adults with MS include coughing while eating, feeling like food is going "down the wrong way," food sticking in the throat, difficulty managing saliva, difficulty initiating a swallow, and drooling. Upon examination, symptoms like jaw jerk and slow tongue movement can be observed [14, 18]. Such problems may lead to insufficient oral intake, malnutrition, dehydration, and inability to take oral medications, resulting in partial or complete dependence on tube feeding, which in turn increases healthcare costs [13, 21]. More importantly, in later stages of MS, dysphagia can lead to morbidity and death from aspiration pneumonia [13, 22, 23]. Dysphagia can also affect patients' quality of life resulting in embarrassment and avoidance of social events that involve consuming food and/or drink [16, 20].

A more recent study showed numerous physical and psychological symptoms associated with swallowing problems in people with MS [11]. When comparing people with MS with swallowing disorders and people with MS without swallowing disorders, people with MS-related dysphagia had reduced scores across all domains of SWALQOL, a swallowing-related quality of life questionnaire [24]. Some of these physical symptoms include coughing, throat clearing, and choking on food and liquid, which may be harbingers of more serious consequences such as pneumonia and increased mortality. Other symptoms people with MS experience are sociopsychological and can include a reduced desire to eat and increased food avoidance. These symptoms may lead to social withdrawal and mealtime anxiety. Eating and drinking play an important role in the physical, psychological, and social aspects of life. It is thus necessary for healthcare professionals who work with people with MS to be aware about dysphagia, its symptoms, and its impact. Early assessment and intervention can delay and/or reduce serious complications (like aspiration pneumonia) in later stages of the disease [11].

Communication and Language Problems in Multiple Sclerosis

The body of literature primarily describes the impact of MS on basic language functions, but does not sufficiently characterize the impact on more complex language skills. Due to the focus on basic language functions, current evidence is inconclusive regarding the presence or absence of language deficits in people with MS. Research into the language capacity of people with MS has primarily come from studies investigating cognitive functions, which use neuropsychological assessments rather than tools developed specifically to evaluate language abilities. These tools only assess the basic functions of language such as verbal fluency and naming. Other studies have utilized aphasia assessment batteries, which are designed to assess the language skills of individuals following a cerebrovascular accident [25, 26]. These earlier studies identified that the performance of people with MS did not vary significantly from controls.

In 2013, a small study explored possible language deficits in 39 Dutch people with MS. In this pilot study, standardized language assessments used following cerebrovascular accidents were administered (including the Boston Naming Test and the Dutch Semantic Association Test). Scores on these tests showed that all people with MS in this cohort had deficits in semantic and phonemic word fluency. Of all participants, 73% had word finding difficulties, and 95% had difficulties with the interpretation of metaphors. Prior to assessment, only 15 participants reported they experienced language difficulties, indicating a possible limit insight in communication problems in persons with MS [27].

Again, these assessments only provide insight into basic language functions while failing to evaluate complex language skills. A recent systematic review identified that impaired word retrieval was the most common language symptom in people with MS [28]. The authors of this review were however unable to draw any general conclusions on high-level language skills in people with MS due to the limited number of high-level language tasks used in included studies [28]. More recent studies investigating the language skills of people with MS have used assessment tools that measure complex language skills. These studies demonstrated that people with MS have trouble with language tasks that require planning, abstract reasoning, problem solving, and decision-making [29].

Traditionally, language functions were felt to be controlled by cortical neurons that remain unaffected in people with MS. In recent years, several models of language processing have described white matter tracts connecting subcortical structures, including the thalamus and basal ganglia, to cortical language areas [30]. The cortico–subcortico–cortical loop model proposes that language functions are controlled by a circuit of white matter pathways between subcortical and cortical structures. These pathways operate in an organized and synchronized fashion to allow for the comprehension and production of language [29]. As such, it is only recently that deficits in the area of language have been considered a potential clinical manifestation in people with MS.

Speech and Voice Problems in Multiple Sclerosis

Speech motor skill is another variable that may interact with the cognitive–linguistic abilities of people with MS. Dysarthria is a motor speech disorder prevalent in 40–50% of people with MS and is characterized by ataxic and/or spastic features [31]. Another recent systematic review described that articulation (slow articulation and imprecise consonants), voice (pitch and loudness instability), respiration (decreased phonatory time and expiratory pressure), and prosody (longer and frequent pauses, deficient loudness control) are affected in people with MS. This review also underlines the earlier described relationship of communication problems with cognition [32].

One study investigated the association between cognitive and speech motor skills in people with MS using rapid speech sound repetitions (dysdiadochokinesis when rapidly saying “pa-ta-ka”) and neuropsychological tests that require verbal responses. Results showed that people with MS performed more slowly on cognitive tasks that require oral responses. The performance on these cognitive tasks were however similar to that of healthy controls when outcomes were statistically controlled for their poorer dysdiadochokinesis scores. Accordingly, the authors hypothesized that slower verbal responses were the consequence of motor speech deficits and not by cognitive deficits [33].

Regarding voice problems, a study with 143 people with MS from Australia ($N = 52$) and New Zealand ($N = 91$), 43% of respondents reported problems with controlling their voice [12]. A recent study compared the maximum expiratory times and maximum phonation times of MS patients with matched healthy controls and found that maximum expiratory times, maximum phonation times, and dysarthria scores were significantly altered compared with healthy controls [34]. This study also showed that with disease progression, the maximum expiratory time will decrease, possibly due to reduced breath support. This decrease in maximum expiratory time is associated with decreased phonation time and an increase in dysarthria scores [34]. These findings are supported by the findings of Fazeli et al. (2018), who describe the phonation and articulation subsystem changes in patients with multiple sclerosis compared to healthy individuals, in which a correlation between changes in an acoustic measure of dysarthric speech (Formant Centralization Ratio) and disease progression was found. Authors suggest that articulation subsystem changes might be useful signs for the progression of the disease [35]. Finally, the prevalence of spasmodic dysphonia in people with MS is higher than in the general population (2% vs. <0.001%) [36].

Although existing literature identifies voice and speech problems as a frequent finding in people with MS, showing a clear correlation between phonation and speech, the underlying aetiology of these (self-reported) voice and speech problems remains under-investigated.

Conclusion

In people with MS, the prevalence of swallowing, voice problems, and communication problems is high. Speech, voice, language, and swallowing problems all have a negative effect on the quality of life and are associated with depression in this population. Up to 38% of people with MS report frequent coughing and choking. Around 43% of people with MS report voice problems, and over 30% report speech problems. Language and cognitive impairments are also common, with over 60% of MS patients reporting productive language problems and 36% reporting receptive language problems. Although MS is primarily associated with loss of muscle strength in various muscle groups, this chapter highlights the complex relationship between MS and cognitive/language function and associated communicative disorders.

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