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Physical therapy management of middle and late stage of Huntington's disease

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Abstract
Huntington's Disease (HD) is a progressive degenerative brain disease manifested by motor, mental and behavioral disorders. Physiotherapy is necessary especially in patients in the middle and late stage. The aim of this study was to investigate the effectiveness of different physiotherapy interventions to improve the clinical picture of patients with HD in the middle and late stage. The Google Scholar, PubMed and PEDro databases were searched with different combinations of the following keywords: Huntington's disease, HD, physiotherapy, hydrotherapy, respiratory physiotherapy, PNF, neuroplasticity. This review included clinical studies, reviews and case studies. It seems that physiotherapy in patients with HD cannot inhibit the progression of the disease, but can significantly improve the functioning of patients; thus, contributing to improving their quality of life. The results of this review show that physiotherapy should be an integral part of a holistic approach that includes interventions that address all the manifestations of the disease, both physical (motor, nervous, cardiovascular and respiratory) and mental.

Keywords: Huntington's disease, HD, physiotherapy, hydrotherapy, respiratory physiotherapy, PNF, neuroplasticity

Introduction
Huntington's Disease (HD) is an inherited neurodegenerative disorder that affects both physical and mental health [1]. It is caused by a gene mutation due to which there is increased loss of nerve cells. If one parent carries the gene, each child has a 50% chance of inheriting it. Motor disorders in HD are chorea, dystonia, eye dysfunction, bradykinesia and gait abnormalities [2, 3]. Motor problems cause gait abnormalities and increase the risk of falls in 60% of patients [3, 4]. Mental disorders are also common in most patients with HD and include memory disorders, poor concentration, difficulty performing scheduled tasks, impulse control disorder, etc. [2].

HD global prevalence is 4-10/100000 with the highest incidence in Europe, England and the USA. HD can occur between the ages of 10 and 75, but usually occurs between the ages of 35 and 75 [5]. In the USA, it has been observed that there are about 30000 people with HD, while 150000 people are at risk of developing it, because they have a parent who already has the disease [2]. The average duration of the disease is 16-21 years and death may result from secondary complications occurring due to respiratory tract infections (e.g., pneumonia). Death is usually associated with complications following a serious fall injury as well as dysphagia-related infections. For this reason, patients with HD need special attention and care [3, 5].

The disease progresses in four stages: The preclinical stage (they have not shown any clinical symptoms), the early stage (beginning of mental and motor disorders), the middle stage (chorea, dystonia, imbalance, gait disorder, etc.) and the late stage (need for external assistance and support) [5].

Many pharmacological treatments have been evaluated and used to treat HD. However, even with these treatments, patients still have problems with functioning, gait and balance. There are some suggestions that physiotherapy may improve motor and functional disorders of the basal ganglia [3]. The goal of physiotherapy in patients with HD is to maintain their independence and functioning, reduce falls and improve their active participation in society. The treatment should not aim at treating motor symptoms (chorea, dystonia, etc.), because it

is not realistic, but should aim at improving general functioning [5]. The aim of this study is to investigate the effect of physiotherapy on the improvement of symptoms and functional ability in patients with middle or late stage HD.

Method

The Google Scholar, PEDro and PubMed databases were searched with the following keywords: Huntington's disease, HD, physiotherapy, hydrotherapy, respiratory physiotherapy, PNF. The review included all types of articles (clinical studies, reviews, observational studies and case studies). Below are the main conclusions of the articles included in this review.

Literature Review

Treatment of people with HD should be holistic and include interventions for cognitive ability (orientation, attention, memory, perception, etc.), mobility, muscle strength, coordination, balance, neuroplasticity, respiratory function and general patient functioning [6].

Physiotherapeutic effects on gait and balance

Mirek et al. [7] conducted a pilot study to determine the benefits of implementing a program with Proprioceptive Neuromuscular Facilitation (PNF) techniques in improving gait and balance in patients with HD. The study involved 30 patients with HD aged 21-60 years. Gait and balance were evaluated before and after the intervention. Participants underwent PNF treatment programs for three weeks five times a week. The sessions included 10 minutes of warm-up with body repositioning and breathing exercises. The main part had a duration of 70 minutes and included exercises to improve balance reactions, mat exercises (rolling, prone position on the elbows, bridge, lateral squats, quadruped position, kneeling position with one leg, standing position), sitting exercises, walking (with weight shifts from one leg to the other) and finally, a 10-minute recovery with relaxation exercises. The results of the second evaluation showed that there was a significant improvement in gait and balance measurements. The researchers concluded that PNF is effective and safe in a patient with HD for short-term improvement of gait and balance.

In another study, Georgiou et al. [8] investigated the effect of a rhythmic haptic cueing (RHC) signal to improve gait coordination. The research included the evaluation of gait before, during and after the intervention while at the same time qualitative characteristics were studied in terms of the feelings of the participants about the treatment through an interview regarding the course of the session. Physiotherapists found that there was a significant improvement in gait with RHC, which was maintained after the end of their intervention. The researchers concluded that RCH could improve the gait of HD patients in the short term.

Furthermore, Schwartz et al. [9] conducted a review to determine how music, dance, and the use of a rhythmic auditory cueing (RAC) affect patients with HD. The results of the seven articles and six abstracts that were included in the review were conflicting. The conclusion, however, was that the arts (music and dance) had a greater impact on patients than RAC. RAC led to microstructural changes. The arts seem to influence the mental, psychiatric, motor, psychosocial and neuroanatomical areas and therefore have a significant impact on improving speech, chorea, behavior and overall quality of life of patients with HD.

The European HD Network conducted a global study to propose guidelines for dealing with HD. To treat the chorea, they suggest placing the patients in different positions. For dystonia, they suggest active and passive exercises to maintain the range of motion of the joints, reduce stiffness, deformities and the appearance of contractions. Low-intensity exercise and the use of a wheelchair, where required, is essential for improving gait and balance and generally for reducing HD symptoms [10].

Exercise

In their systematic review, Mueller, Petersen and Jung [1] referred to the positive effects of exercise on cardiovascular function in patients with HD. They also reported that exercise may delay the onset and development of motor and mental disorders. However, existing studies are few in number and no safe conclusions can be drawn.

Moreover, Allison Coleman and Blair R. Leavitt [11] implemented a hydrotherapy program in patients with middle and late stage HD. The study included six patients with HD, who performed hydrotherapy sessions two times a week for six weeks. Each session consisted of warm-up, various exercises and recovery. There were continuous recordings regarding injuries and the adaptability of the participants to ensure their safety during the intervention. Participants conducted interviews about the course of treatment, which were recorded by videotaping. They reported that hydrotherapy was fun, enjoyable and safe and noticed an improvement in their sleep, mood and quality of life. The results of the research also showed that there was a positive effect on their physical condition.

Yu and Bega [12] also conducted a review on the application of alternative therapies in HD. The PubMed and Cochrane Library databases were searched and the results were sorted by level of evidence (Class I, II, III, IV) according to the American Academy of Neurology classification scale. Researchers showed that dual-task therapies that combine motor skills with the mind are safe for patients with HD, but more research is needed as information on the frequency, time and structure of treatments is lacking.

Exercise and activity improve cognitive function and behavior in a wide range of patients including people with mild cognitive impairment [13]. Exercise has been shown to facilitate cognitive function and in particular moderate-intensity exercise reduces proactive interference [14]. Furthermore, exercise in combination with meditation and mindfulness increases the gray matter density within brain stems. It has also been observed that high-intensity exercise increases the neurogenesis process in the hippocampus and low-intensity exercise improves newborn neuronal survival maturation and spatial memory [15].

Mental Intervention

Neuroplasticity of the brain seems to be able to compensate up to a point for the mental disorders that occur due to the degeneration caused in the brain cells by HD. It seems that the environment also affects the process of cell degeneration. Cognitive dysfunction contributes to mobility impairments in HD. Mental disorders are highly correlated with motor disorders and are associated with episodes of falls. Mental intervention through brain neuroplasticity has been shown to aid in reducing HD symptoms in the short

term [16].

There is not much research on mental intervention in patients with HD and the changes it causes. Nevertheless, van Walsem and his colleagues [17] applied an observational study in which various specialties of paramedical and medical professions (physiotherapists, occupational therapists, speech therapists, neurologists, nurses, nutritionists, social workers and psychiatrists) observed for a year the progression of the disease in 37 patients with HD. They implemented an indicative treatment program that included various functional activities, such as preparing food, writing, engaging in some activity, puzzles, games, group discussions, etc. There was improvement only in the Symbol Digit Modalities Test, while no change in neuropsychology was observed.

Respiratory Physiotherapy

Patients with HD experience severe respiratory complications and swallowing disorders leading to difficulty clearing the airways. Reyes et al. [18] conducted a pilot study to study the benefits of a home four-month respiratory physiotherapy program to improve breathing, dyspnea, endurance and swallowing. Their study included 18 patients, who were equally divided into two groups: control and intervention. A respiratory program was implemented in both groups. The control group used a constant water resistance of 9cm and the intervention group used a gradually increasing resistance from 30% to 75% of the maximal respiratory pressure of each patient. The measurements were performed at the baseline, in the 2nd and in the 4th month. The results showed that in the intervention group there was a greater increase in maximal inspiratory and expiratory pressure, maximal vital capacity, dynamic expiratory volume in one second and maximal expiratory flow compared to the control group. However, changes in dyspnea, swallowing and endurance were not statistically significant. The researchers concluded that respiratory physiotherapy at home improves the lung function of patients with HD, but does not provide many changes in swallowing function, dyspnea and endurance.

Discussion-Conclusions

The results of this review show that treating a symptomatic patient with middle to late stage HD should aim to improve or maintain functional level. Physiotherapy intervention should include motor, mental and breathing exercises in order to achieve best functioning. PNF, dance, music, use of RCH, RAC and aids are effective methods to improve gait and balance and seem to have positive effects in treating the symptoms of the disease [7-10]. Exercise appears to be beneficial for patients with HD in improving cardiovascular and mitochondrial function and neuroplasticity [1, 11-15]. To make it more fun and effective it can be done through hydrotherapy or through dual-task activities [16-17]. However, further research should be done to clarify the parameters of the interventions in terms of frequency, time and benefits of exercise, because the literature available is minimal. Improving neuroplasticity is important for the treatment of HD and it seems (albeit by few studies) that it can be done with mental intervention. Finally, respiratory physiotherapy is necessary especially for the patients of the late stage, as they face serious respiratory problems and difficulty in swallowing, which can even lead to their death [18]. Further research is needed, for the physiotherapy treatment of patients with middle to late stage HD, as the already existing studies are limited.

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