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Review

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Update of Rehabilitation in Huntington's Disease: Narrative Review

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HIGHLIGHTS

- Comprehensive rehabilitation for Huntington's disease would improve the multi-domain function.
- Programmed rehabilitation based on the stage of the disease is effective for Huntington's disease.

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Update of Rehabilitation in Huntington's Disease: Narrative Review

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ABSTRACT

Huntington's disease (HD) is a neurodegenerative disease that has motor dysfunction, predominantly chorea, cognitive impairment, and psychiatric disturbances as symptoms. Treatment is directed to reduce the severity of symptoms, although there are few studies and no clinical guidelines for rehabilitation in HD. Therefore, this review aimed to establish an effective rehabilitation approach for HD according to the stage of the disease. In the early stage of HD, the motor symptoms are mild, and psychological symptoms occur. Treatment in this period should focus on aerobic and resistance exercises, task-specific training, secondary prevention education, cognitive training, and psychological management. In the middle stage of HD, the motor symptoms are more severe. Task-specific rehabilitation approaches, education for the patient and caregiver, functional respiratory exercises, activities of daily living training, multidisciplinary and multimodal daycare rehabilitation are helpful to patients in this stage. At the late stage of HD, most patients need complete support for activity of daily living. Mobility and balance evaluation and prevention strategies should be focused on for safety, and respiratory exercises and physical exercise to prevent complications in patients with severely impaired mobility should be considered based on the patient's condition. Programmed rehabilitation management based on the stage of the disease is effective for patients with HD.

Keywords: Huntington's Disease; Stage of Disease; Rehabilitation; Review

INTRODUCTION

Huntington's disease (HD) is a neurodegenerative disease showing a progressive decline in function with autosomal dominant inheritance that is related to the basal ganglia [1]. HD results from a mutation in the huntingtin gene. However, up to 10% could result from a mutation elsewhere [2]. Expansion of cytosine-adenine-guanine repeats on chromosome 4 may result in a mutant protein (mutated huntingtin protein), which gradually damages basal ganglia neurons [3]. The worldwide prevalence of HD is 5–10 cases per 100,000 individuals, and the overall prevalence of HD in Asian countries is 0.40 cases per 100,000 individuals [4,5]. Suffering-promoting symptoms are motor dysfunction, predominantly chorea, cognitive impairment, and psychiatric disturbances [6].

There is no cure for HD. Treatment focuses on reducing the severity of symptoms. One study argued that there is insufficient evidence to support the use of physiotherapy to reduce impairments of balance, muscle strength, and flexibility and that few lines of evidence support speech therapy for the management of eating and swallowing disorders [7]. Thus, several rehabilitation approaches such as physical, occupational, cognitive, and home-based rehabilitation have been proposed for patients with HD to improve and maintain their declining functions [8-16]. These approaches would be helpful for the improvement of motor, cognitive, and psychological functions. However, there are few studies and no established clinical guidelines for rehabilitation in HD.

Therefore, in this review, we survey the effects of rehabilitation on patients with HD and propose an effective rehabilitation approach based on the stages of the disease.

STAGES OF HD

It is not easy to diagnose the HD before the patients recognize their psychological and minor motor problems. HD's average age at onset is between 35–45 years. After that, the symptoms usually progress 10–20 years [17]. The early stage of Huntington's disease may occur the issues include work, finances, driving; able to live at home with minimal supervision. The middle stage of HD, patients may have troubles to do the activities of daily living (ADL) to need minimal to moderate assists for 6 to 10 years from disease onset. The late stage of HD would need assistance with most ADLs, 24-hour care and may last almost 11 to 20 years from disease onset [18,19].

REHABILITATION IN HD

Rehabilitation in the early stage of HD

In the early stage of HD, symptoms such as subtle personality changes, problems in cognition and physical skills, irritability, and mood swings may occur and usually precede the motor symptoms [19]. Motor symptoms present as chorea, impaired balance, decreased flexibility, fine motor problems, and unsteady gait. Gym-based or group-based exercises may be more desirable for patients in premanifest and early-stage HD [20]. Recommendations for rehabilitation management of the early stage of HD are as follows.

Exercise capacity and performance

Aerobic and resistance exercises are recommended. A moderate exercise intensity, within 50%–70% of maximal heart rate, paired with upper and lower body strengthening, three times per week for 12 weeks, should be prescribed to improve exercise capacity and performance [9].

Planning and performing of tasks for movement

Patients with HD would present the impairment of motor function in early stage. Task-specific training such as strategy training, sensory stimulation, cueing, and chaining are recommended as an occupational therapy [7, 9].

Mobility, balance, and risk of falls

Task-specific mobility training should be prescribed as individualized gait training with supervision to improve spatiotemporal function [9].

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Preventing the secondary musculoskeletal changes and deconditioning
HD would cause secondary musculoskeletal changes owing to the progressive motor symptoms. Patient education, maintenance exercise program, and gait and balance training are needed to prevent the secondary musculoskeletal changes and deconditioning [9,21].

Cognitive training
Multidisciplinary rehabilitation approaches show positive effects on the preservation of striatal shape, particularly the putamen, on cognitive domains such as verbal learning, memory, attention, cognitive flexibility, processing speed, and social cognition [22].

Psychological management
Multimodal daycare rehabilitation shows significant effects on both anxiety and depression [16].

Rehabilitation in the middle stage of HD

In the middle stage of HD, the motor symptoms are more severe. These show as chorea or dystonia, joint range of motion limitations, weak stabilizers and falls owing to balance, and gait deficits [17]. Recommendations for rehabilitation management approaches for the middle stage of HD are as follows.

Planning and performing of tasks for movement
In middle stage of HD, patients would aggravate the impairment of movement such as apraxia, chorea and bradykinesia. Task-specific therapy such as strategy training, sensory stimulation, cueing, chaining, and occupational therapy are recommended [7,9].

Postural control, alignment in sitting mobility, balance, and risk of falls
Task-specific rehabilitation approaches such as fall risk assessment, proper positioning for mobility, seating and wheelchair evaluation, balance and gait training should be prescribed for supervised gait training to improve spatiotemporal abilities. In addition, patients with mid-stage HD may be prescribed, at a moderate frequency and intensity, individualized exercises to improve balance function [9].

Adapting the secondary musculoskeletal changes and deconditioning
Education for the patient and caregiver is needed for maintaining the exercise program and gait and balance training to adapt secondary musculoskeletal changes and follow deconditioning [9.19].

Respiratory exercises
Functional respiratory exercises should be prescribed such as positioning, breathing exercises, airway clearance methods, and respiratory muscle relaxation techniques [9,19].

Cognitive training
Multidisciplinary rehabilitation shows a positive effect on the preservation of striatal shape, particularly the putamen, on cognitive domains such as verbal learning and memory, attention, cognitive flexibility, processing speed, and social cognition [22].

Psychological management
Multimodal daycare rehabilitation shows a positive treatment effect on both anxiety and depression [23].

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Rehabilitation in the late stage of HD

In the late stage of HD, patients are mostly bedridden and need complete support for ADL. Physicians and caregivers should take care of postural changes, dysarthria, dysphagia, respiratory dysfunction, increased risk of pneumonia, and mobility problems, especially falls [23]. Recommendations for rehabilitation management for the late stage of HD are as follows.

Postural control and alignment in sitting

The risk of falling is markedly increased in the late stage of HD. So, evaluation and prevention strategies should be focused on safety, with fall risk assessments, seating and wheelchair evaluations and proper timed sit positioning [9].

Respiratory exercises

In the late stage, functional respiratory exercises and ADL training should be prescribed: positioning, breathing exercises, airway clearance methods, and respiratory muscle relaxation techniques. The progressive decline in respiratory function may cause severe (even fatal) complications. Therefore, patients with late-stage HD should be trained with regular breathing exercises, such as inspiratory and expiratory breathing training, to enhance respiratory muscle strength and coughing function [9,21].

Physical exercise to prevent complications in patients with severely impaired mobility

Physical exercise to prevent complications in patients with severe impaired mobility at late-stage HD is essential. Patients should be prescribed with bed positioning, passive range of motion, and active movement exercises [9].

However, some interventions may not be fully supported by evidence regarding the late stage of HD. Nevertheless, those interventions should be considered based on the patient's condition.

Interventions for consideration in late stage of HD

- Effectiveness of postural control training in patients with HD: The prescription of an individually tailored program to improve postural control or to use positioning devices to optimize posture should be considered based on the patient's condition [21].
- Role of rehabilitation management on the ADL, seating, and positioning in patients with end-stage HD: Patients with end-stage HD should be considered for appropriate positioning, seating, active movement, respiratory exercise, and education. Family and caregiver education should be provided to maintain appropriate ongoing activity, and participation may be an important focus for the rehabilitation management team as part of end-stage care [21].

Each step of rehabilitation management based on the stage of the disease is shown in **Table 1**.

CONCLUSION

Comprehensive rehabilitation for HD improves exercise capacity, physical function, mobility, balance, and cognitive, psychological, respiratory, and musculoskeletal functions according to the stage of the disease. Programmed rehabilitation approaches are effective for managing HD in each stage.

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Table 1. Rehabilitation management strategies according to the stage of Huntington's disease

Variables	Early stage	Middle stage	Late stage
Exercise performance	· Aerobic and resistance exercises (moderate intensity within 50%–70% of maximal heart rate, three times per week for 12 wk)		
Mobility and gait	· Task-specific mobility, training focused on individual gait function	· Task-specific mobility training	
Postural control, alignment in sitting, balance and fall risk		· Fall risk assessment · Proper positioning for mobility · Seating and wheelchair evaluation · Balance and gait training (moderate frequency and intensity)	· Fall risk assessment · Seating and wheelchair evaluation · Proper timed sit positioning
Secondary musculoskeletal changes and deconditioning	· Education for patient to maintain exercise program, gait, and balance	· Education for patient and caregiver to maintain exercise program, gait, and balance	
Planning and performing of tasks for movement	· Task-specific training (strategy training, sensory stimulation, cueing, and chaining)	· Task-specific therapy (strategy training, sensory stimulation, cueing and chaining, occupational therapy)	
Cognitive training	· Multidisciplinary rehabilitation for cognition	· Multidisciplinary rehabilitation for cognition	
Psychological management	· Multimodal daycare rehabilitation	· Multimodal daycare rehabilitation	
Respiratory exercise		· Positioning · Inspiratory and expiratory breathing training · Airway clearance methods · Respiratory muscle relaxation techniques	· Inspiratory and expiratory breathing training · Airway clearance method, especially coughing
Physical exercise to prevent complications in patients with severely impaired mobility			· Bed positioning · Passive range of motion · Active movement exercises

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