Case Report

Do Huntington’s Disease Patients Benefit From Multidisciplinary Inpatient Rehabilitation?

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Abstract

Huntington’s disease (HD) is a neurodegenerative disease. Since no disease modifying medication is available until now, rehabilitation and other therapeutic approaches are of major interest. Some studies suggest that multidisciplinary rehabilitation may be beneficial in HD improving activities of daily living (ADL). Medical records of the BDH-Clinic Hessisch Oldendorf, a large rehabilitation facility in Northern Germany, have been carefully reviewed and three HD cases have been identified over the last decade. The patients have been treated with a three-week inpatient multidisciplinary rehabilitation program, including ADL-training, physical, cognitive, occupational and (in one case) speech therapy. All patients benefited from inpatient rehabilitation. Two were discharged with a BI increase of 5 resp. 20 points. In one case, BI remained unchanged, but the patient’s balance and gait markedly improved. Further controlled studies are needed but results from this small case series and findings from literature suggest that inpatient multidisciplinary rehabilitation is useful in HD.

Introduction

Huntington’s disease (HD) is a late-onset, neurodegenerative and hereditary disease caused by a trinucleotide repeat expansion in exon 1 of the Huntingtin gene (4p16.3)\(^\text{[1]}\). This expansion encodes a stretch of glutamine residues in the Huntingtin protein (HTT) which is essential in a variety of cellular functions, e.g. vesicle transport, transcription and energy production\(^\text{[2]}\). Mutant HTT-induced toxicity ends up in death of striatal and cortical neurons inducing movement disorders (e.g. chorea, dystonia, akinesia, postural and gait disturbances), cognitive resp. psychiatric problems (attention and memory disorders, personality changes, frontal dementia, depression, psychosis, anxiety, aggressive and obsessive compulsive behavior), dysarthria and dysphagia\(^\text{[3–5]}\). Cognitive and motor symptoms are evident as early as 10 years prior to HD onset\(^\text{[6]}\). With disease progression, activities of daily living (ADL) and social participation are significantly impaired. Many late-stage patients are dependent on help, up to 73.8% reside in nursing homes\(^\text{[7]}\). Survival time after diagnosis is 15-20 years\(^\text{[8]}\). In German speaking countries, one out of 10.000 inhabitants suffers from HD\(^\text{[8]}\).

Despite considerable efforts, including promising strategies like gene silencing, as yet no disease modifying therapy is available\(^\text{[2]}\). Only symptomatic treatment (e.g. tetrabenazine, tiapride or neuroleptics considered for chorea) may be offered to the patient\(^\text{[9]}\).

There is only limited scientific evidence to prove that physical, occupational and speech therapy, as well as multidisciplinary inpatient rehabilitation is effective in HD\(^\text{[10,11]}\).

Results from HD mouse models suggest that motor training, in particular in an enriched environment setting, may delay disease onset and progression\(^\text{[11-16]}\). Beneficial effects could be explained by increased brain-derived neurotrophic factor (BDNF) levels in hippocampal and striatal neurons\(^\text{[17]}\). Diminished BDNF synthesis and secretion - as well as oxidative stress - plays an important role in human HD pathophysiology\(^\text{[2,18]}\).

A couple of human studies have shown that physical therapy (physiotherapy, motor training), in particular exercise programs at patients’ home, may be beneficial in HD\(^\text{[18-30]}\). Table 1.

<table>
<thead>
<tr>
<th>Study</th>
<th>n</th>
<th>Sample</th>
<th>Intervention</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bins wanger, 1980(^\text{[30]})</td>
<td>5</td>
<td>Out patients</td>
<td>Home physiotherapy (focus: muscle weakness, range of movement, gait, breathing) 60 min twice a week for 4 weeks</td>
<td>Objective outcome measurements not reported, subjective improvements in alertness and balance</td>
</tr>
<tr>
<td>Lavers, 1981(^\text{[29]})</td>
<td>6</td>
<td>Late-stage patients on a long-term care psychiatric ward</td>
<td>Weekly physiotherapy and occupational therapy over 12 months, focusing on mobility, balance, coordination, contracture prevention, social interaction</td>
<td>Objective outcome measurements not reported, 2 patients gained body weight</td>
</tr>
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As far as occupational therapy is concerned, only one randomized-controlled trial (RCT) with a small sample size (12 patients of a long-term care unit) is available using multisensory stimulation\[^{[31]}\]. Patients attended eight 30 min sessions (visual, tactile, auditory and olfactory stimulation) over four weeks.

The authors found no significant treatment effects\[^{[31]}\]. In another study with only four participants, no improvement of ADL was evident after eight months\[^{[32]}\]. In addition, some case reports suggest that occupational therapy may be beneficial\[^{[33-35]}\].

Treatment of dysphagia, dysarthria and aphasia is recommended in HD\[^{[36,37]}\], but evidence is weak\[^{[10,11]}\].

With respect to multidisciplinary rehabilitation, a couple of studies have been published\[^{[38-43]}\]. Table 2, two of them dealing with outpatient\[^{[19,43]}\], three with inpatient rehabilitation\[^{[38,40-42]}\]. Duration of multidisciplinary rehabilitation was three weeks in all three inpatient studies and could be repeated up to three times per year in two of them\[^{[38,40]}\]. The studies enrolled early to mid-stage HD patients. Improvements in ADL were reported by two studies\[^{[38,41,42]}\]. In one study, ADL remained stable with no significant decline\[^{[40]}\]. In addition, gait and other parameters of motor performance improved in all three inpatient studies. Remarkably, one paper reported an influence of inpatient multidisciplinary rehabilitation on neural oxidative damage (significant decrease of Cu/Zn-superoxide dismutase and neuron-specific enolase levels) suggesting a disease modifying effect of this intervention\[^{[40]}\].

<table>
<thead>
<tr>
<th>Study</th>
<th>Sample</th>
<th>Intervention</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>Zinzi et al., 2007[^{[34]}]</td>
<td>Early to mid-stage HD patients</td>
<td>Respiratory exercises, speech, physical and occupational therapy, cognitive exercise, three week inpatient rehabilitation (repeated up to 3 times per year)</td>
<td>Significant improvement of motor performance and ADL (p &lt; 0.001), no motor decline over two years</td>
</tr>
<tr>
<td>Thompson et al., 2013[^{[34,40]}] RCT</td>
<td>Early to mid-stage HD outpatients</td>
<td>9-month multidisciplinary rehabilitation program (once weekly 60 min), occupational therapy targeting cognitive deficits (6 months, 60 min per fortnight) and home exercises (6 months, 3 times per week)</td>
<td>Reduction of motor and postural stability deterioration, minor improvements in depression, cognition and quality of life</td>
</tr>
<tr>
<td>Pira et al., 2013[^{[40]}]</td>
<td>Early to mid-stage HD inpatients</td>
<td>1 year rehabilitation program (3 admisions of 3 weeks each) focusing on physical exercise, social activities, and group/teaching sessions</td>
<td>Improvements in gait function, balance, physical quality of life, anxiety and depression, BMI; ADL-function remained stable with no significant decline</td>
</tr>
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<td>Ciancariello et al., 2013[^{[41]}], Ciancariello et al., 2014[^{[42]}]</td>
<td>HD inpatients (mean BI 77 points)</td>
<td>3-month inpatient intensive multifunctional neurorehabilitation, 2 x 120 min group therapy daily (physical and occupational therapy)</td>
<td>Significant increase of the mean scores of BI, TS, PPT and TFCS (p &lt; 0.001), reduction of oxidative stress, parameters of neurodegeneration (Cu/Zn-SOD, NSE)</td>
</tr>
<tr>
<td>Cruickshank et al., 2015[^{[43]}]</td>
<td>Out patients, U H - DRS-TMS &gt; 5</td>
<td>9 months once-weekly supervised clinical exercise, thrice-weekly self-directed home based exercise and fortnightly occupational therapy</td>
<td>Significantly increased gray matter volume in the right caudate and bilaterally in the DL-PFC, significant improvements in verbal learning and memory (Hopkins Verbal Learning-Test)</td>
</tr>
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</table>
There is only little data on the effectiveness of multidisciplinary inpatient rehabilitation in HD. Thus, last decade medical records of the BDH-Clinic Hessisch Oldendorf, a large neurological rehabilitation center located in Northern Germany, have been carefully reviewed. Three inpatient rehabilitation HD cases have been identified and analyzed.

**Case Presentation**

**Case 1**

A 48 year old male with genetically confirmed HD (45 CAG repeats) was admitted to inpatient rehabilitation because of a deterioration of ADL. Age of onset was 43. The patient suffered from dysphagia, chorea and mild frontal dementia. Due to an aspiration pneumonia in the past, the patient already had a percutaneous endoscopic gastrostomy (PEG) tube. A computer tomography (CT) of the skull revealed a marked brain atrophy, in particular of the frontal lobes, Figure 1.

![Figure 1: CT scan of case 1.](image)

He was still living at home with some help from his mother and working in a sheltered workshop. The patient was on tiapride 300 mg/d, tetrazepam 100 mg/d and memantine 90 mg/d.

The three-week inpatient multidisciplinary rehabilitation program included 60 min cognitive training (computing tasks, writing, communication skills), 30 min occupational therapy (fine motor skills) and 120 min ADL-training daily (total 210 min/d).

On admission, the Barthel index (BI) was 70, and the patient was discharged home with a BI of 90 (Δ=20).

**Case 2**

A 20 year old female with clinical diagnosis of HD (not genetically confirmed) was admitted to inpatient rehabilitation because of increasing cognitive problems (memory, disorientation) and schizophrenic psychosis. Age of onset was 17. She was suffering from dysarthria, eye saccade disturbances, motor im-persistence, ataxic gait, dysdiadochokinesis, dysmetria (all four extremities). Luria’s test was positive on admission. A magnetic resonance imaging (MRI) revealed global brain atrophy, Figure 2.

![Figure 2: MRI (T2 FLAIR) of case 2.](image)

The patient was living with her parents who were taking care of her. She used to work in a nursing home but since months, she was unable to work. The patient was on tiapride 300 mg/d, olanzapine 30 mg/d and lorazepam 2 mg/d.

The three-week inpatient multidisciplinary rehabilitation included 75 min cognitive training (memory, orientation), 30 min occupational therapy (fine motor skills), 90 min physical therapy (balance, gait) and 180 min ADL-training daily (total 375 min/d).

On admission, BI was 60, and the patient was discharged home with a BI of 65 (Δ=5).

**Case 3**

A 59 year old female with clinical diagnosis of HD (not genetically confirmed) was admitted to inpatient rehabilitation because of deterioration of ADL. Age of onset was unclear, at least three years ago. She was suffering from chorea, severely ataxic gait (with a history of falls), dysphasia and cognitive problems (disorientation, memory, attentiveness). A magnetic resonance imaging (MRI) revealed marked brain atrophy (with hydrocephalus) and diffuse white matter changes, Figure 3.

![Figure 3: MRI (T2 FLAIR) of case 3.](image)

The patient was still living at home but she was found neglected in her apartment, with a BMI of only 18.2 kg/m². She was jobless for years. The patient was on tiapride 200 mg/d and...
 Benefit From Multidisciplinary Inpatient Rehabilitation

quetiapine 50 mg/d.

The three-week inpatient multidisciplinary rehabilitation included 30 min cognitive training (communication skills), 20 min speech therapy, 60 min occupational therapy (fine motor skills, working in the kitchen), 60 min physical therapy (balance, gait) and 180 min ADL-training daily (total 350 min/d).

On admission, the Barthel index (BI) was 70, and the patient was discharged to a nursing home with a BI of 70 (Δ=0). Despite a lack of BI changes, her balance and gait were much better at discharge and she was able to walk a short distance without assistive device.

The BI values of all three patients are displayed in Figure 4.

![Figure 4: Barthel index of the three cases on admission and after 1, 2 and 3 weeks of inpatient rehabilitation.](image)

Discussion

HD is a neurodegenerative, severely disabling disease. While no disease-modifying medication is available until now, rehabilitation and other therapeutic interventions are of major interest. Some studies suggest that physical therapy is beneficial in HD\(^{[9-30]}\). Many patients, however, suffer from severe impairments of their social participation. They are in need of multidisciplinary (inpatient) rehabilitation. A couple of studies have demonstrated that multidisciplinary rehabilitation is useful to improve ADL, as shown by increases of BI\(^{[38,41,42]}\).

Since data on this topic is rare, HD inpatient rehabilitation cases of the BDH-Clinic Hessisch Oldendorf, Germany, have been carefully reviewed. Three HD cases have been identified over the last decade. Diagnosis was genetically confirmed in one case, only. However, the clinical diagnoses in the other two cases was most certain since competing diagnoses have been excluded reliably.

The HD patients have been treated with a three-week inpatient multidisciplinary rehabilitation program, including ADL-training, physical, cognitive, occupational and (in one case) speech therapy. All patients benefitted from the inpatient rehabilitation. Two were discharged with a BI increase of 5 rep. 20 points. In one case, BI was unchanged, but the patient’s balance and gait was markedly better. The results from this small case series are in line with previous studies which have demonstrated significant improvements through a three-week multidisciplinary inpatient rehabilitation\(^{[38,41,42]}\). Although the length of stay in rehabilitation was brief (three weeks) for patients who suffer from long-term, progressive functional decline, ADL improved.

Further controlled studies are needed but results from literature and this small case series supports the hypothesis that multidisciplinary rehabilitation in HD is useful.

References


