

Inpatient multidisciplinary rehabilitation programme for postural and gait stability in Huntington's disease – a pilot study

Vliv multidisciplinárního rehabilitačního programu během hospitalizace na posturální stabilitu a stabilitu chůze u Huntingtonovy nemoci – pilotní studie

Abstract

Aim: Postural and gait instability in Huntington's disease (HD) is a key component of the motor symptomatology which contributes to an increased risk of falls. Rehabilitation is considered beneficial in postural and gait stability treatment. We aimed to explore the feasibility and the short- and long-term effects of an inpatient multidisciplinary rehabilitation program on postural and gait stability in subjects with HD. **Methods:** A sample of 13 subjects with HD but with no severe cognitive deficit or depression underwent a 3-week specific inpatient rehabilitation program focused on postural and gait stability. Patients were examined at the baseline, after the completion of rehabilitation, and then 1 month and 3 months after the end of the program. The testing included: gait stability examination (Dynamic Gait Index; DGI), posturography examination of postural stability on a stable (PSS) and 20% unstable (PSU) platform and the total motor score evaluation by Unified Huntington's Disease Rating Scale (UHDRS). **Results:** There was a significant improvement lasting 3 months in PSS and a significant improvement in DGI immediately after the rehabilitation. There was no significant improvement in the PSU and UHDRS total motor score. **Conclusion:** Specific rehabilitation methods are safe and feasible and may be beneficial in the treatment of postural and gait instability in patients with early and mid-stage HD. The postural instability improvement measured by PSS persisted for at least 3 months. The gait stability improvement in DGI did not persist after 1 month. We found no improvement in PSU. This exploratory study offers a sample of a specific rehabilitation protocol for stability training in HD.

Key words

Huntington's disease – rehabilitation – gait – instability

Klíčová slova

Huntingtonova nemoc – rehabilitace – chůze – instabilita

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L. Brabcová^{1,2}, J. Roth¹,
O. Ulmanová¹, J. Ruz^{1,3},
J. Klempíř¹, O. Horáček²,
M. Kolářová², P. Košková²,
P. Rolková², H. Božková², L. Sabó²,
M. Inemanová², K. Lísalová²,
F. Jančok², E. Růžička¹, H. Brožová¹

¹ Department of Neurology and Centre of Clinical Neuroscience, Charles University, 1st Faculty of Medicine and General Teaching Hospital, Czech Republic

² Rehabilitation Centre, Beroun Rehabilitation Hospital, Czech Republic

³ Department of Circuit Theory, Faculty of Electrical Engineering, Czech Technical University in Prague, Czech Republic



MUDr. Hana Brožová, Ph.D.
Department of Neurology
and Centre of Clinical Neuroscience
1st Faculty of Medicine
Charles University
and General University Hospital in
Prague
Kateřinská 30
128 21 Prague
Czech Republic
e-mail: hana.brozova@lf1.cuni.cz

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Souhň

Cíl: Posturální instabilita a instabilita chůze patří mezi významné motorické symptomy Huntingtonovy nemoci (HN), které zvyšují riziko pádů. Rehabilitace je podstatnou součástí terapie instability. Cílem studie bylo zhodnotit, krátkodobý a dlouhodobý efekt multidisciplinárního rehabilitačního programu na posturální instabilitu i instabilitu chůze u HN a posoudit možnost provedení programu za hospitalizace. *Metodika:* 13 pacientů s HN bez těžšího kognitivního deficitu a deprese absolvovalo třítydenní multidisciplinární rehabilitační program během hospitalizace, který byl specificky zaměřený na posturální stabilitu a stabilitu chůze. Vyšetření proběhla na začátku programu, po dokončení rehabilitace, po 1 a 3 měsících od dokončení. Testování zahrnovalo vyšetření stability chůze (Dynamic Gait Index; DGI), posturální stability pomocí posturografu na stabilní (PSS) a nestabilní 20% (PSU) plošině a motorické skóre pomocí Unified Huntington's Disease Rating Scale (UHDRS). *Výsledky:* PSS prokázalo statisticky významné zlepšení přetrvávající po dobu 3 měsíců a signifikantní zlepšení v DGI ihned po rehabilitaci. V motorickém skóre UHDRS a PSU statisticky významné zlepšení nalezeno nebylo. *Závěr:* Specifický rehabilitační program je bezpečný a dobře využitelný při terapii poruch stability u HN. Posturální stabilita dle PSS byla zlepšena po sledované době 3 měsíců. Zlepšení stability chůze dle DGI odeznělo do 1 měsíce. V PSU signifikantní zlepšení nebylo prokázáno. Tato studie nabízí návrh specifického rehabilitačního protokolu pro trénink stability u HN.

Introduction

Huntington's disease (HD) is a hereditary neurodegenerative disease which manifests itself through involuntary movements and voluntary motor function impairment, together with cognitive and behavioral impairment.

The average age of HD onset is in the 4th decade, and the disease duration is commonly between 15–20 years. In the later stages it leads to complete disability and dependence on caregivers and the community. There is only limited and temporary symptomatic therapy [1]. Postural and gait instability in subjects with HD is a key component of the motor symptomatology [2], contributing to an increased risk of falls. Falls have been reported from 60 to 80% of subjects with HD [2–5]. Falls, injury and loss of independent ambulation are often factors which precipitate admission to a nursing home [6].

Subjects with HD manifest significant postural control deficits when performing motor skills typical of daily living activities [7]. Posturography shows a consistent pattern of abnormality in HD [8]. Significant deficits were reported in anticipatory postural adjustments [8,9], in reactive postural responses [10–12] as well as in still standing [10]. Subjects with HD showed considerably more anterior-posterior sway than normal, especially when visual and proprioceptive cues were eliminated [11]. Posturography examination of the limits of stability (LOS; the amount of maximum excursion an individual is able to cover intentionally in any direction without losing their balance or taking a step) showed impairment even before the clinical onset of the disease. The centre of pressure (COP) displacements were analyzed during maximum leaning in four basic directions and under three sensory con-

ditions (eyes open, eyes closed and eyes closed standing on foam). Subjects with manifest HD showed significantly greater COP ranges than the healthy control subjects in all sensory conditions. The greatest deterioration was found when standing on foam [13].

Rehabilitation may provide a therapeutic approach to stimulation of the brain to recruit alternative neuronal networks and enhance neuronal activity in pre-existing damaged neuronal networks [14]. Fritz et al in a recent systematic review found seven studies involving rehabilitation treatment and evaluation of balance in HD. It seems that rehabilitation may be beneficial in balance treatment [15]. Previous studies which focussed on both inpatient and outpatient rehabilitation programmes in HD showed the effects on motor performance, function (Activities of Daily Living; ADL) or general physical condition, and even improvements in the gait and balance of patients with HD [16–29]. However, there is a limited number of inpatient rehabilitation studies focussed specifically on postural and gait therapy in HD. Tab. 1 shows the methodology and results of comparable studies focussed on multidisciplinary inpatient rehabilitation using physiotherapy, occupational therapy, speech therapy, respiratory exercises, cognitive rehabilitation exercises, training in groups in the gym and/or in a swimming pool, patient education sessions and group discussions for participants, assessment of the need for assistive devices and dietitian intervention.

Our study is the first prospective multidisciplinary study using a specific programme aimed at postural and gait stability in HD during a 3-week hospitalization. This study uses objective evaluation of postural and gait stability parameters repeatedly during

a 4-month follow-up. A constant, clearly defined physiotherapy protocol focussed on postural and gait stability training with daily evaluation of improvement was used.

The aim of our study was to explore the feasibility and the short- and long-term effects of an inpatient multidisciplinary rehabilitation programme on postural and gait instability in the early and middle stages of HD.

Methods Participants

The subjects with HD from The Movement Disorders Centre, Charles University in Prague were screened consecutively for the study during a 3-year period (2014–2016). The inclusion criteria were age over 18, genetically verified HD in the early and middle stages, stable medication, no other rehabilitation during the 4-month course of the study, and a signed informed consent form. The exclusion criteria were acute psychiatric symptoms, dementia preventing cooperation – Mini Mental State Examination (MMSE) [30] score lower than 20, concurrent depression – Beck Depression Inventory (BDI) [31] score higher than 9, severe immobility or co-morbidities preventing active cooperation in the rehabilitation programme or interfering with the monitored criteria (serious orthopaedic or internal medicine diagnoses, other neurological diagnoses causing movement impairment, severe visual impairment or hearing loss etc.), as well as non-compliance of the subject or the family. The study was performed in accordance with the ethical standards laid down in the 1975 Declaration of Helsinki. The responsible Institutional Review Boards approved the study protocol and the informed consent form (research Ethics Committee of the General University Hospital, Prague, approval number: 40/13).

Tab. 1. Comparison of the studies focussed on intensive inpatient multidisciplinary rehabilitation in HD.

Study	Zinzi et al [29]	Piira et al (1 year) [20]	Piira et al (2 years) [21]	Ciancarelli et al [22]
subjects	40 genetically confirmed HD subjects in early and middle stages	37 genetically confirmed HD subjects in early and middle stages	37 genetically confirmed HD subjects in early and middle stages	34 genetically confirmed HD subjects in early and middle stages
length of intervention	3 weeks 3x /year 2 years	3 weeks 3x /year 1 year	3 weeks 3x /year 2 years	3 weeks
intensity of intervention	8 h /day 5 days /week + 4 h during the weekends	8 h /day 5 days /week + 4 h during the weekends in one of the 2 centres	8 h /day 5 days /week + 4 h during the weekends in one of the 2 centres	2x /day 6 days /week 2 h /session
tools (motor function)	Tinetti Scale PPT	TUG 10MWT 6MWT BBS ABC questionnaire	TUG 10MWT 6MWT BBS ABC questionnaire	Barthel Index Total Functional Capacity Scale PPT Tinetti Scale
evaluation	at the beginning and at the end of each admission	at the beginning and at the end of each admission + 5-day evaluation stay approximately 3 months after the last RHB admission	at the beginning and at the end of each admission + 5-day evaluation stay approximately 3 months after the last RHB admission	at the beginning and at the end of the 3-week RHB treatment, Barthel Index 3 months after the treatment by a telephone follow-up interview
results	Highly significant improvement in the Tinetti scale and PPT. No carry-over effect from one admission to the next, but also no motor decline over two years.	Significant improvement in gait –TUG, 10MWT, 6MWT from baseline through stay two and three to the evaluation stay in the first year. An overall improvement in balance (BBS) from baseline to evaluation stay. No change in ABC score from the baseline to the evaluation stay.	After 15 months a slight decline or stable function in gait and a minor statistically non-significant decline in BBS. Among the six participants who completed the 2-year program, four had stable or improved gait (TUG) from baseline to the last measurement point. Similar findings in balance. BBS scores stable or improved in four individuals throughout the study period.	A significant increase in Barthel Index, Total Functional Capacity Scale, PPT and Tinetti Scale at the end of the 3-week RHB. The improvement in Barthel Index vanished after 3 months.

6MWT – Six Minute Walk Test; 10MWT – Ten Meter Walk Test; ABC questionnaire – Activities of Balance Confidence Scale; BBS – Berg Balance Scale; HD – Huntington’s disease; PPT – Physical Performance Test; RHB – rehabilitation; TUG – Timed-up-and go test

Study design

The screening visit included a clinical neurological examination, which was performed by a neurologist experienced in HD, the MMSE test (a 30-point neuropsychological screening test for cognitive deficit; higher scores mean better performance) and the BDI test (a 21-question multiple-choice self-report inventory, a psychometric test for measuring the severity of depression; higher scores mean more severe depression) administered by a neuropsychologist.

Outcome measures

The subjects were examined at the baseline, after the 3-week inpatient rehabilitation pro-

gramme and 1 month and 3 months after finishing the programme.

The set of examinations included: a gait stability examination using the Dynamic Gait Index (DGI; assesses an individual’s ability to modify balance while walking in the presence of external demands – higher scores indicate better performance) [32], administered by a physician with a specialization in neurology and rehabilitation medicine and experienced in HD; the same examiner performed all the assessments and a postural stability examination by posturography, LOS [33], administered by a physiotherapist experienced in gait and posture; the same physiotherapist performed all the assessments. We used the Balance Master de-

vice and the Balance Master LOS Test. The LOS test quantifies the maximum distance the subject can intentionally displace their centre of gravity while maintaining stability. We measured the endpoint excursions in 8 directions, 4 cardinal + 4 diagonal, and their total scores. The subjects were asked to watch the screen and try to “hit the highlighted points with the cursor on the screen” by leaning their bodies. The test was performed once on the stable (PSS) and once on the 20% unstable (PSU) platform and the Unified Huntington’s Disease Rating Scale (UHDRS), the total motor score (a basic motor performance evaluation tool in HD – higher scores indicate lower performance) [34], administered by a neurologist expe-

Tab. 2. The physiotherapy protocol.

The list of tasks – the subject is asked to perform each task 1x

- 1. 10-m walk** – The subject is asked to walk a 10 m long distance between 2 lines indicating the start and the finish at a comfortable self-selected pace.
- 2. Walking along a line** – The subject is asked to walk a 10 m long distance along a line stuck to the floor at a comfortable self-selected pace with feet in tandem. If he/she is not able to put feet in tandem or tends to fall, he/she can walk along the line without feet in tandem. The task is then assessed as "cannot perform".
- 3. Walking with horizontal head turns** – The subject is asked to walk a 10-m distance between 2 lines indicating the start and the finish at a comfortable self-selected pace. He/she is asked to turn his/her head to the right or left at a verbal cue. The frequency of head turns is about 4–6 turns at the distance of 10 m.
- 4. Walking to a metronome** – The subject is asked to walk a 10 m long distance between 2 lines indicating the start and the finish, according to a regular rhythm given by a metronome (clicking sound). The pace is approximately a normal comfortable walking speed. The subject should be able to synchronize the pace with the metronome.
- 5. The balance cushion (unstable surface – a lens-shaped inflatable cushion 60 cm in diameter)** – The subject is asked to take a step onto the cushion, keep balance for 10 s standing still and step down without physical help.
- 6. Stepping over obstacles** – The subject is asked to walk a 10-m distance at a comfortable pace and on the way to step over 2 wooden boxes (60 × 30 × 25 cm, 100 cm between each other) without physical help. He/she is allowed to slow down or stop to adjust the steps before stepping over the obstacle.
- 7. The "Movin Step" (air-filled step cushion with 2 connected air chambers, for one foot each, used for exercises to improve balance and core stability)** – The subject is asked to take a step onto the cushions, keep balance for 10 s while shifting the weight from one foot to the other, then step down without physical help.
- 8. Catching a ball in a standing position** – The subject is asked to catch a ball (an inflatable over-ball 25 cm in diameter) which is thrown by a physiotherapist from different directions in a cca 2.5-m distance. The subject is standing on a sign stuck to the floor and does not have to turn around. He/she is allowed to step aside when trying to keep balance. The ball is thrown three times. The subject must be able to catch it at least twice.
- 9. Throwing a ball at a target** – The subject is asked to throw a ball (an inflatable over-ball 25 cm in diameter) into a gate 80 cm wide made of two vertical plastic bars. He/she is standing on a mark stuck to the floor 2.5 m from the gate. He/she tries three times and should be able to hit the target at least twice.
- 10. Pivot turns** – The subject is asked to walk a 10-m long distance between 2 lines indicating the start and the finish at a comfortable self-selected pace. The physiotherapist shows physically what a "pivot turn" means before the start. The subject is asked to do the same when he/she hears a signal – a handclap.
- 11. Initiation and inhibition of a movement at a sound signal** – The subject is asked to walk a 10 m long distance between 2 lines indicating the start and the finish at a comfortable self-selected pace. Before he/she starts walking, the subject gets the instruction that the start of the walking will be signaled by a handclap. The "stop signal" is also a handclap.
- 12. Jumping into and out of a circle** – The subject is asked to jump (with both feet) into a circle (a "Hula-Hoop" circle, 75 cm in diameter) lying on the floor, then to jump (with both feet) out of the circle. He/she should be able to do it without physical help.
- 13. Standing on 1 leg for 10 s (right)** – The subject is asked to stand on the right leg for 10 s and keep balance without touching the floor with the other foot.
- 14. Standing on 1 leg for 10 s (left)** – The subject is asked to stand on the left leg for 10 s and keep balance without touching the floor with the other foot.
- 15. Kicking a ball at a target** – The subject is asked to kick a ball (an inflatable over-ball 25 cm in diameter) into a gate 80 cm wide made of two vertical plastic bars. He/she is standing on a sign stuck to the floor 2.5 m from the gate. He/she tries three times and should be able to hit the target at least twice and keep balance without physical help.
- 16. Knee bend (squatting position)** – The subject is asked to squat and stand up without physical help.
- 17. "Slalom" between objects** – The subject is asked to walk a 10 m long distance between 2 lines indicating the start and the finish at a comfortable self-selected pace. There are plastic cones placed in a line, the distance between two cones is 50 cm. The subject should be able to pass the cones alternately from the right and left side without kicking them or losing balance.
- 18. Walking + holding a tray with a cup in hands (double tasking)** – The subject is asked to walk a 10 m long distance between 2 lines indicating the start and the finish at a comfortable self-selected pace holding a tray with a plastic cup full of water.
- 19. Walking with counting** – The subject is asked to walk a 10 m long distance between 2 lines indicating the start and the finish at a comfortable self-selected pace and at the same time count backwards by ones starting at 20.
- 20. Training staircase** – The subject is asked to go up and down a small "training staircase" with 4 steps up and down. There is a handrail, but the subject is asked not to use it if possible. "Can do" means he/she is able to complete the task without the handrail or with as little help of the handrail as possible, but without physical help of the physiotherapist.

rienced in HD who had completed the rater training.

The whole examination process was conducted by a physician with a specialization in neurology and rehabilitation medicine and experienced in HD. Two subjects did not complete one of the follow-up posturography examinations.

Intervention

The 3-week inpatient rehabilitation programme included: 1. individual physiotherapy focussed on gait, stability and coordination according to a constant physiotherapy protocol (Tab. 2) twice a day for 30 min; 2. 60 min of other stability and condition training daily; 3. 30 min of occupational therapy daily, focussed mainly on motor coordination. No major tailoring was necessary as all subjects were able to take part in all of the activities. If the subject was tired, he/she was allowed to take a rest. If the task proved too difficult, it could be modified. In that case the task was assessed 'cannot do' in the physiotherapy protocol.

The specific physiotherapy protocol was created in consideration of the typical stability and gait problems of subjects with HD (postural instability, impaired dynamic balance, impaired velocity control mechanisms, increased variability in temporal control [2,9,13,35–37]). The list of 20 exercises was evaluated every day (can/cannot perform) to observe the changes in the separate tasks. 'Can perform' was evaluated 1 point; 'cannot perform' was evaluated at 0 points. As the list contains 20 exercises, the maximum was 20 points every day. The exercises were performed every day in a stable sequence by all subjects. The assessment was done during the therapy by the same physiotherapist during the whole stay. The task was assessed as 'can perform' if the subject was able to do it without physical help. Verbal guidance was allowed.

In addition to the steady physiotherapy protocol (Tab. 2) 30 min twice a day, therapy with the physiotherapist included other 2x 30 min used for a workout on the 'Posturomed' (Haider Bioswing, Pullenreuth, Germany) (a neuro-orthopaedic sensorimotor therapy and diagnosis device with an attenuated, oscillating, spring-dampened unstable platform; it is suspended on an oscillation frame that enables dosed, attenuated, compensating movements with variably adjusted oscillation amplitudes and frequencies), fitness workout (warming,

stretching of hypertonic muscles, strength training of weakened muscles), aerobic exercise (stable bicycle, Nordic walking), exercise on the 'Motomed' (Reck, Betzenweiler, Germany) (a neuro-orthopaedic upper and lower limb cycling device customized to be used passively, motor-assisted, or actively resistive; the subject sits on a stable chair and can follow visual feedback on a screen; the device was used as actively resistive during the training) and the 'DAVID system' (DAVID Systems GmbH, Munich, Germany) (fitness machines for the treatment and monitoring of problems in the back, hips and knees, which are equipped with a monitoring unit that connects to a central server with a database of individual patients and their treatment outcomes and goals).

With the occupational therapists the subjects had 30 min a day of ADL training including the 'training flat' (a place designed as a regular household for training in ADL and instrumental ADL with the aid of assistive devices and compensatory mechanisms), hand motor skill training, cognitive and executive function training ("The Happy Neuron – Brain Jogging" computer program [Alpelephant, s.r.o., Prague, Czech Republic] – an interactive cognitive stimulation tool designed for various medical and other backgrounds). The tasks were administered by the same occupational therapist during the entire course.

The subjects underwent speech and swallowing therapy and individual psychotherapy if needed. A social worker was available during the entire course together with an all-day nursing service. Tab. 3 shows an example of the daily programme.

Statistical analyses

For primary analysis, the Friedman test followed by Wilcoxon's post hoc analyses were used to assess the subjects' performance at the baseline, immediately post rehabilitation, and 1 month and 3 months after the rehabilitation programme. All follow-up examinations were compared to the baseline.

Bonferroni correction for multiple comparisons was applied for 4 tests performed with a corrected P threshold equal to < 0.0125 (i.e. 0.05/4) for P < 0.05. For each type of exercise, the sum of successfully performed exercises (i.e. 'can perform') was calculated separately for Day 1–9 and Day 10–18. The effect of the individual exercises (i.e., secondary analysis) was calculated using the

Tab. 3. A sample daily programme.

7.00 – 7.30	hygiene, dressing
7.30 – 8.00	breakfast
8.00 – 9.00	free time, daily rounds, physician consultation
9.00 – 9.30	physiotherapy (protocol)
9.30 – 10.00	free time, coffee break
10.00 – 10.30	occupational therapy
10.30 – 11.00	free time
11.00 – 11.30	physiotherapy (stability + condition training)
11.30 – 12.00	free time
12.00 – 12.30	speech therapy
12.30 – 13.00	lunch
13.30 – 14.00	free time, dietitian consultation, if needed
14.00 – 14.30	physiotherapy (protocol)
14.30 – 15.00	free time, coffee break
15.00 – 15.30	physiotherapy (stability + condition training)
15.30 – 16.00	physician consultation
16.00 – 18.00	free time
18.00 – 18.30	dinner
18.30 – 22.00	free time, hygiene, undressing
22.00	bed time

psychologist, social worker consultations alternatively during breaks if needed

Wilcoxon signed rank sum test; no correction for multiple comparisons was applied.

Results

A sample of 16 genetically verified subjects with HD (9 women) in the early and middle stages were included in the rehabilitation programme. A group of 13 subjects completed the entire 4-month course of the study. Three subjects finished the 3-week inpatient programme but dropped out during the following 3 months of the follow-up. The mean age was 48 years (standard deviation [SD] 14, range 25–67), the mean HD duration was 6 years (SD 2.4, range 2–9) and the mean age of symptom onset was 42 years (SD 14, range 19–62). The mean number of triplets was 46 (SD 4.8, range 41–56). The mean Total Functional Capacity was 6.8 (SD 2.3, range 4–12, where 13 = normal and

0 = severe disability). One subject dropped out due to a concomitant medical condition (a leg fracture at home between visits) and two subjects due to personal organization and transportation problems. There were no adverse events during the therapy.

Fig. 1 and Tab. 4 show the subjects' performance at baseline and immediately post rehabilitation, 1 month and 3 months after rehabilitation for measures of UHDRS, DGI, PSS and PSU (primary analysis).

A significant change was found in PSS 2 (3.7) = 11.4, P = 0.04, W = 0.38, reflecting improvement 1 month after rehabilitation (P = 0.008, W = 1.00) as well as 3 months after rehabilitation (P = 0.04, W = 0.29) when compared to the baseline.

We also revealed a significant change in DGI λ^2 (3.10) = 21.9, P < 0.001, W = 0.56, reflecting an improvement in the HD subjects' performance from the baseline to the state immediately post rehabilitation (P = 0.008, W = 0.86). However, we did not find any significant changes in the next follow-up examinations when compared to the baseline.

No significant differences were found for the measure of UHDRS λ^2 (3.10) = 5.1, P = 0.67, W = 0.13] as well as PSU λ^2 (3.10) = 7.3, P = 0.25, W = 0.24].

Tab. 5 lists the results of 20 individual exercises between Day 1–9 and Day 10–18. A significant improvement in the subjects' performance was observed in the exercises of walking with horizontal head turns (P = 0.02), balance cushion (P = 0.002), stepping over obstacles (P = 0.03), catching a ball in a standing position (P = 0.02) and standing on the left leg for 10 s (P = 0.02).

Discussion

A limited number of studies included the question of how different types of rehabilitation methods and their frequency

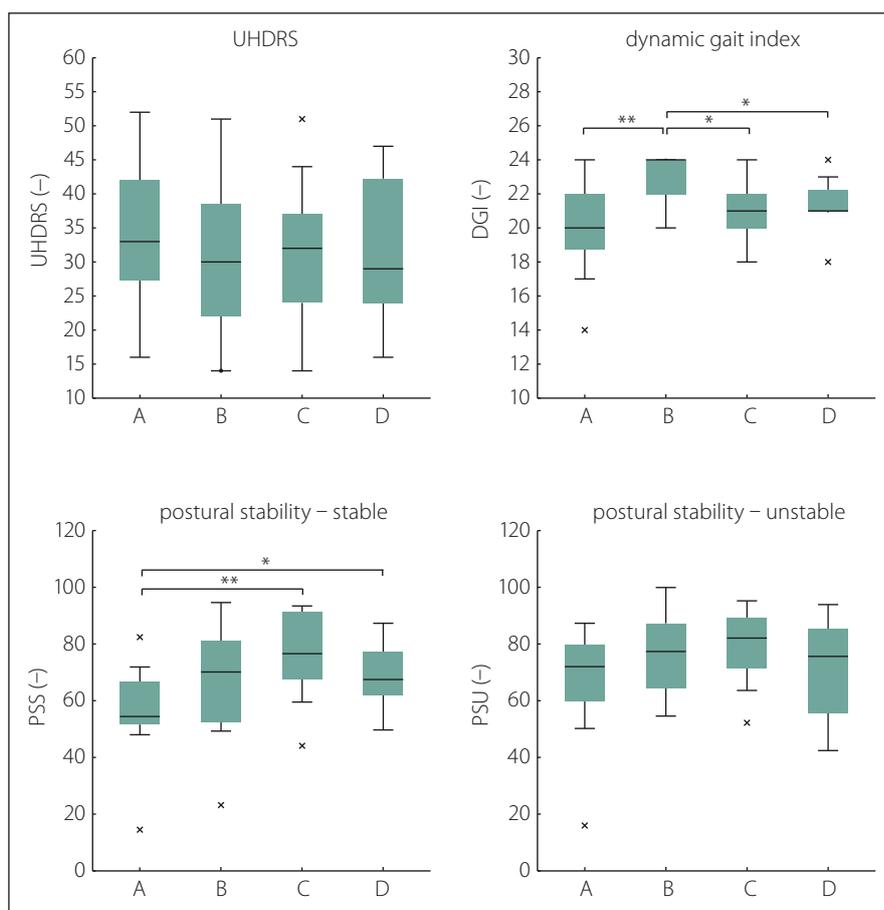


Fig. 1. The effects of rehabilitation on postural and gait stability (results of primary analysis).

*P < 0.05; **P < 0.01; x – outliers

DGI – Dynamic Gait Index; PSS – postural stability on a stable platform; PSU – postural stability on an unstable platform; UHDRS – Unified Huntington's Disease Rating Scale

Obr. 1. Vliv rehabilitace na posturální stabilitu a stabilitu chůze (výsledky primární analýzy).

*P < 0,05; **P < 0,01; x – odlehlé hodnoty

DGI – Dynamic Gait Index; PSS – posturální stabilita na stabilní plošině; PSU – posturální stabilita na nestabilní plošině; UHDRS – Unified Huntington's Disease Rating Scale

and intensity can influence the symptoms of HD.

Two systematic reviews evaluating physical therapy and exercise interventions in HD

were published in 2017 [15,38]. They suggest that there is preliminary support for the benefits of exercise and physical activity in HD in terms of motor function, gait speed and

Tab. 4. Subjects' performance at baseline and immediately post rehabilitation, 1 month and 3 months after rehabilitation.

	UHDRS (-)			DGI (-)			PSS (-)			PSU (-)		
	mean	SD	95% CI	mean	SD	95% CI	mean	SD	95% CI	mean	SD	95% CI
baseline	32.8	10.6	26.7–38.9	20.0	2.8	18.3–21.7	55.6	18.0	42.8–68.5	66.7	21.1	51.6–81.8
immediately after RHB	30.4	10.6	24.1–36.9	22.0	1.5	22.0–23.8	51.0	20.8	51.0–80.9	76.6	15.0	65.9–87.4
1 month after RHB	31.0	10.9	24.4–37.6	20.8	1.5	19.9–21.8	64.2	15.7	64.2–86.7	69.8	13.6	69.8–89.3
3 months after RHB	32.5	13.0	24.7–40.4	20.7	1.5	20.7–22.4	68.8	11.4	60.6–76.9	71.4	17.4	59.0–83.9

CI – confidence interval; DGI – Dynamic Gait Index; PSS – postural stability on a stable platform; PSU – postural stability on an unstable platform; RHB – rehabilitation; SD – standard deviation; UHDRS – Unified Huntington's Disease Rating Scale

balance, as well as a range of physical and social benefits identified through patient-reported outcomes. The reviews showed that the methods of subject recruitment, intensity of training and the physiotherapy methods used in individual studies were very diverse. This is why the comparison of the results with our study is quite complicated.

Our study is the first prospective multidisciplinary inpatient study using a specific programme aimed at postural and gait stability in HD. We found a significant improvement in DGI immediately after the rehabilitation programme. There was a decline both 1 month and 3 months after the rehabilitation. DGI is a very sensitive tool characterizing the subjects' functional state. We did not find any comparable rehabilitation study using DGI in HD; however, there are studies using similar tools focussing on postural and gait stability.

Busse et al [16] demonstrated an improvement of the Romberg test outcomes in the intervention group compared to the control group. The scores on the Berg Balance Scale either improved after the rehabilitation intervention [20,23,25] or remained unchanged [18]. Subjects in the control group also demonstrated minor improvements [18]. Balance confidence on both walking and stairs measured by the Activities Balance Confidence Scale improved in the intervention group but declined in the control group [24]. There was no significant difference between the intervention group and the control group following a video-game balance intervention [28].

We found a significant improvement of PSS after the rehabilitation programme persisting for 1 as well as 3 months when compared to the baseline. There is no comparable rehabilitation study for the change in this parameter in HD. We found only a single study [37] using LOS on mechanically locked force plates. The study described a significant impairment of postural control (endpoint excursion, maximum excursion and directional control) in HD as compared to healthy controls.

We did not find any significant improvement in the PSU parameter. The study by Blanchet et al [13] proved a higher deterioration in PSU compared to PSS. We presume PSU is apparently a difficult test and the level of balance impairment in HD causes a certain limit in the ability to improve by means of rehabilitation.

The UHDRS total motor score did not show any improvement. The scale was used as the basic testing tool for subjects with HD.

Tab. 5. The effects of individual exercises (results of secondary analysis).

	Day 1–9		Day 10–18		Wilcoxon
	total score		total score		signed rank test
	median	IQR	median	IQR	P
1. 10-m walk	9	0	9	0	1
2. walk along a line	9	0	9	0	0.5
3. walking with horizontal head turns	8	2.25	9	0	0.02
4. walking with a metronome	8	2	9	1	0.07
5. the balance cushion	5	3.75	9	2.25	0.002
6. step over obstacles	8	2	9	0.25	0.03
7. the "Movin Step"	9	0.25	9	1	0.91
8. catching a ball in a standing position	8	2.25	9	0	0.02
9. throwing a ball at a target	9	1	9	0	0.25
10. pivot turns	9	0.25	9	0	0.25
11. initiation of a movement at a sound signal	7	3.25	9	2.25	0.22
12. jumping into and out of a circle	9	0	9	0	1
13. standing on right leg for 10 s	4	5.5	7	5.25	0.09
14. standing on left leg for 10 s	2	4.5	4	7	0.02
15. kicking a ball at a target	9	0.25	9	0	0.5
16. a knee bend (squatting position)	9	4.5	9	0	0.13
17. "slalom" between objects	9	2	9	0.25	0.38
18. walking + holding a tray with a cup in hands	9	0	9	0	1
19. walking + loud counting	9	0	9	1	0.13
20. training staircase	8	3.25	9	0.25	0.09

IQR – interquartile range

This scale contains only a few points aimed specifically at gait and stability, therefore we did not expect any significant improvement in this scale. Furthermore, we did not find any general progression of the disease during the 4-month period.

This study showed a significant performance improvement in several specific exercise items tested in the second part of the therapy in comparison with the first part. We can also see which exercise items were the most problematic for the majority of the subjects. It is interesting that they correlate mostly with those with a significant performance improvement. This could be considered in a future investigation of postural and gait stability in subjects with HD. The 20-point physiotherapy protocol might be utilized during the intervention.

The results of our study support the results reported by Zinzi et al [29], Piira et al [20,21]

and Ciancarelli et al [22]. The studies show that intensive multidisciplinary rehabilitation may improve motor performance, including balance, in HD. The effect is obvious immediately after the intervention. We found carry-over effects in one stability parameter after 1 and 3 months. Piira et al [20,21] also described carry-over effects in gait and stability parameters between the admissions. We did not find any carry-over effects after 1 and 3 months in the other parameters, but there was also no decline during the whole course of the study, which is in accordance with the study performed by Zinzi et al [29].

One limitation of this study is the small sample size, because this is a single-centre study and HD is not a common disease. The performance and quality of cooperation might also be influenced by the extent of apathy or motivation in the HD subjects.

Another limitation of the study is that if a multidisciplinary approach is used it might be difficult to delineate which exact method is responsible for the changes in the outcome measures. Some outcome measures might also be influenced by the training of the individual tasks during the intervention.

Randomized controlled clinical trials targeted at the effects of multidisciplinary intensive rehabilitation intervention on the progression of HD could bring us more information on what the optimal protocol of rehabilitation treatment in HD would be and which patients would profit most from the intervention. We are continuing with a longer follow-up and we are planning a comparison with a control group (subjects with HD without rehabilitation intervention).

Conclusion

Our findings suggest that specific rehabilitation methods may be beneficial in the treatment of postural and gait instability in early- and middle stage subjects with HD. An intensive inpatient multidisciplinary rehabilitation programme is safe, feasible and well-tolerated in motivated patients with HD.

The effect on postural stability tested by PSS persisted for at least 3 months. The improvement in gait stability tested by DGI did not remain after 1 month. There is a limit for the possibility of improvement in PSU, probably related to the degree of stability impairment in patients with HD. This study offers a sample of a specific rehabilitation protocol for stability training in patients with HD.

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