Environmental modifiers: Prospects for rehabilitation in Huntington’s disease

Jan Frich
Oslo University Hospital / University of Oslo

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Background

• Growing interest in studying effects of exercise and rehabilitation in patients with Huntington’s disease (HD)

• Programs may have different formats:
  – Home-based exercise programs
  – Community-based exercise/rehabilitation programs
  – Inpatient (intensive/multidisciplinary) rehabilitation programs

(For example: Khalil et al, 2013; Busse et al 2013; Zinzi et al 2007; Piira et al 2013)
Activities

(Photo: North Norway Rehabilitation Center)
Research

• Target groups: Early- to mid-stage HD

• Observational studies indicate positive effects of multidisciplinary rehabilitation on physical function/balance, swallowing, independence, mood and social relationships

• A randomized study of a 12-week community based program found that the program was safe, feasible, acceptable and suggests beneficial effects of rehabilitation
  (Busse et al, JNPT, 2013)
The Norwegian Project

- Intensive rehabilitation programs for patients with HD was funded by the Norwegian Directorate of Health in 2009

- The initiative was inspired by the results from a landmark observational study (Zinzi et al, Clin Rehab, 2007)

- The aim of the mixed-method evaluation was to assess the feasibility and the effects of participating in a one-year multidisciplinary rehabilitation program

Ethical approval by Norwegian Social Science Data Services (ref. 26587) and The Regional Ethics Committee, Health Region South-East (ref. 2010/1026-1)

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Participants

• Inclusion
  – Clinical diagnosis of Huntinton’s disease
  – Early- to midphase (Shoulson & Fahn, stage I-III)
  – Age > 18 years

• Exclusion
  – Serious psychiatric conditions
  – No apparent severe impairment in general cognitive function at the time of first admission

Piira A, van Welsam MR, Mikalsen G, Nilsen KH, Knutsen S, Frich JC. Effects of a one year intensive multidisciplinary rehabilitation program for patients with Huntington’s disease: a prospective intervention study. PLOS Curr 2013. doi: 10.1371/currents.hd.9504af71e0d1f87830c25c394be47027

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One year program: 3 x 3 weeks stay + evaluation stay

Stay 1 (3 weeks) → Stay 2 (3 weeks) → Stay 3 (3 weeks)

Evaluation stay 3 months (95.4 days (SD ±34.2) after discharge of the last stay)

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Vikersund
Start of each stay
Cognitive function (MSSE), depression (HADS), motor function, balance, gait (ABC, 6 Min. Walk Test ...)
ADL (Barthel)

End of each stay
Motor function, balance, gait

Three weeks stay
- 8 hours of various activities 5 days a week
- Groups of 4-6
- Physiotherapy / exercise
- Group meetings, trips, making food, etc

3 months ...
The multidisciplinary team

- Physician/neurologist
- Nurses
- Physical therapist
- Occupational therapist
- Speech therapist
- Dietician
- Social worker
- Psychologist

- The institutions are specialised regional rehabilitation centres (stroke rehabilitation etc.)
Characteristics of the sample (baseline), n = 37

<table>
<thead>
<tr>
<th>Variables</th>
<th>Values</th>
</tr>
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<tbody>
<tr>
<td>Male</td>
<td>N = 18 (48,6%)</td>
</tr>
<tr>
<td>Female</td>
<td>N = 19 (51,4%)</td>
</tr>
<tr>
<td>Age</td>
<td>52,4 SD = 13,1</td>
</tr>
<tr>
<td>Symptom duration</td>
<td>7,2 år SD = 5,7</td>
</tr>
<tr>
<td>Total Functional Capacity (0-13)</td>
<td>8,9 SD = 2,3</td>
</tr>
<tr>
<td>Stage (Shoulson &amp; Fahn)</td>
<td></td>
</tr>
<tr>
<td>I</td>
<td>9 (24,3%)</td>
</tr>
<tr>
<td>II</td>
<td>21 (56,8%)</td>
</tr>
<tr>
<td>III</td>
<td>7 (18,9%)</td>
</tr>
<tr>
<td>UHDRS motor</td>
<td>36,6 16,8</td>
</tr>
<tr>
<td>UHDRS behavior</td>
<td>9,2 8,5</td>
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</tbody>
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### Clinical measures at baseline, n = 37

<table>
<thead>
<tr>
<th>Clinical measures</th>
<th>Mean</th>
<th>SD</th>
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<tbody>
<tr>
<td>MMSE</td>
<td>25.4</td>
<td>3.5</td>
</tr>
<tr>
<td>BMI</td>
<td>22.8</td>
<td>3.2</td>
</tr>
<tr>
<td>Activities of Balance Confidence (ABC)</td>
<td>72.6</td>
<td>23.9</td>
</tr>
<tr>
<td>Bergs Balance Scale (BBS)</td>
<td>52.1</td>
<td>3.8</td>
</tr>
<tr>
<td>Timed up and og (TUG)</td>
<td>8.1</td>
<td>3.1</td>
</tr>
<tr>
<td>10 Meter Walk test (10MWT)</td>
<td>6.8</td>
<td>2.6</td>
</tr>
<tr>
<td>6 Min Walk test (6MWT)</td>
<td>484.9</td>
<td>147.5</td>
</tr>
<tr>
<td>Barthel index</td>
<td>19.2</td>
<td>1.3</td>
</tr>
<tr>
<td>HADS</td>
<td>8.2</td>
<td>8.7</td>
</tr>
<tr>
<td>SF-12 (physical health)</td>
<td>43.9</td>
<td>9.4</td>
</tr>
<tr>
<td>SF-12 (mental health)</td>
<td>52.2</td>
<td>11.2</td>
</tr>
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</table>

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• 31 out of 37 completed the one-year program as planned (83.8%)
Fig 1. Mean change in health-related quality of life (SF-12)

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Fig 2) Mean change in 6 Minutes Walk Test (meters)


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Fig 3) Change in gait function

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Fig 4) Mean change in balance: Berg Balance Scale (BBS) and Activities of Balance Confidence scale (ABC)

Scores

- BBS
- ABC

ns

p = 0.032


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Fig 5) Mean change in BMI, HADS and Barthel Index

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Fig 6) Mean change in cognitive scores

- Stroop word
- Stroop colour
- Stroop interference
- Verbal fluency
- Symbol digit modality test
- MMSE


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Main findings

• Significant improvements were observed in gait function, balance, in physical quality of life, anxiety and depression, as well as in BMI

• ADL-function remained stable with no significant decline

• One cognitive measure (SDMT) showed significant decline, while no decline was observed for the remaining cognitive measures
Feasibility?

• A qualitative, explorative study
• In-depth interviews with 11 patients and 9 family caregivers
• Focus groups with 15 health professionals
• Research questions:
  – How did participants experience the structure and content?
  – What outcomes did patients experience?
  – What challenges and success factors did health professionals report?


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Participants’ experiences

• Some had difficulties defining individual rehabilitation goals:
  
  – “Goal setting for individuals with HD is not necessarily a straightforward process. Perhaps that’s exactly what they need to work on ... then need to find out what they need to work on”
    (interview with health professional)

• Written individualised plans and schedules were appreciated

• Being member of an “HD-group” was valuable, though there could be tensions and conflicts in groups

Frich JC et al 2014

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Participants’ experiences

• Participants typically reported improved gait and balance, increased self-confidence, and social outcomes:
  – “I have become more open, in a way, and if I fall, I will get back on my feet again ... this is not how it used to be. I used to be afraid of walking around, in case I would fall” (interview with participant)

• The intensive schedule was acceptable for most participants

• Adjustments had been made to allow for more time between sessions

Frich JC et al 2014

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Participants’ experiences

- Health professionals - success factors were
  - Assigning every patient with a contact person
  - Using clinical test results as motivation
  - Supervising health professionals in patients’ local municipalities

Frich JC et al 2014
Conclusions

• Intensive multidisciplinary rehabilitation for individuals with early- to mid-stage HD is feasible and associated with improved motor function and physical quality of life

• Participants emphasise mental and social outcomes in addition to physical outcomes

• An individually tailored plan, a contact person, a peer group approach and communication between institutions and primary health professionals could be mediators of outcomes
Conclusions

• Randomised controlled trials are needed to study effects of various interventions, as well as cost-benefit analysis of interventions

• Future research should aim at studying effects of specific components in rehabilitation programs

• Tensions between a standardised “intervention” and an individually tailored approach?


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