

# Environmental modifiers: Prospects for rehabilitation in Huntington's disease

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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  
(Zinzi et al, Clin Rehab 2007; Ciancarelli et al, Eur J Phys Rehabil Med, 2013; Thompson et al Eur J Neurol 2013)
- 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

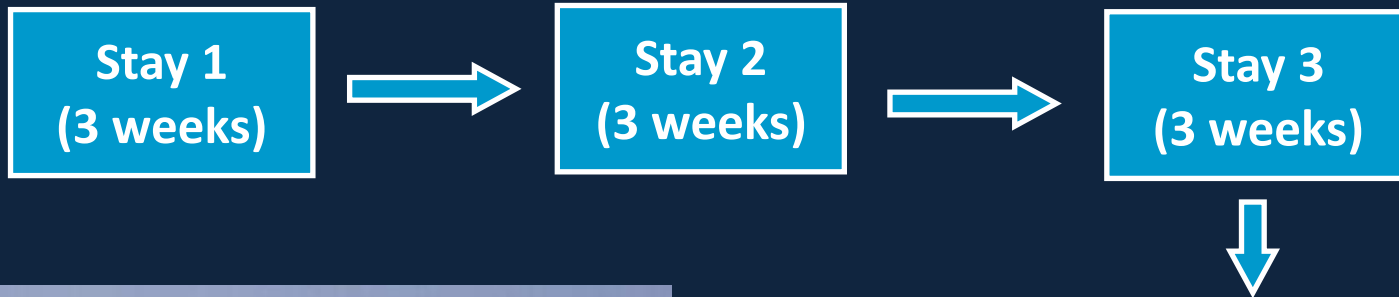
Ethical approval by Norwegian Social Science Data Services (ref. 26587) and The Regional Ethics Committee, Health Region South-East (ref. 2010/1026-1)

# Participants

- Inclusion
  - Clinical diagnosis of Huntington'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](https://doi.org/10.1371/currents.hd.9504af71e0d1f87830c25c394be47027)

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



North Norway Rehabilitation Center



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

<b>Variables</b>			
<b>Male</b>	<b>N = 18 (48,6%)</b>		
<b>Female</b>	<b>N = 19 (51,4%)</b>		
<b>Age</b>	<b>52,4</b>		<b>SD = 13,1</b>
<b>Symptom duration</b>	<b>7,2 år</b>		<b>SD = 5,7</b>
<b>Total Functional Capacity (0-13)</b>	<b>8,9</b>		<b>SD = 2,3</b>
<b>Stage (Shoulson &amp; Fahn)</b>			
<b>I</b>	<b>9 (24,3%)</b>		
<b>II</b>	<b>21 (56,8%)</b>		
<b>III</b>	<b>7 (18,9%)</b>		
<b>UHDRS motor</b>	<b>36,6</b>		<b>16,8</b>
<b>UHDRS behavior</b>	<b>9,2</b>		<b>8,5</b>

# Clinical measures at baseline, n = 37

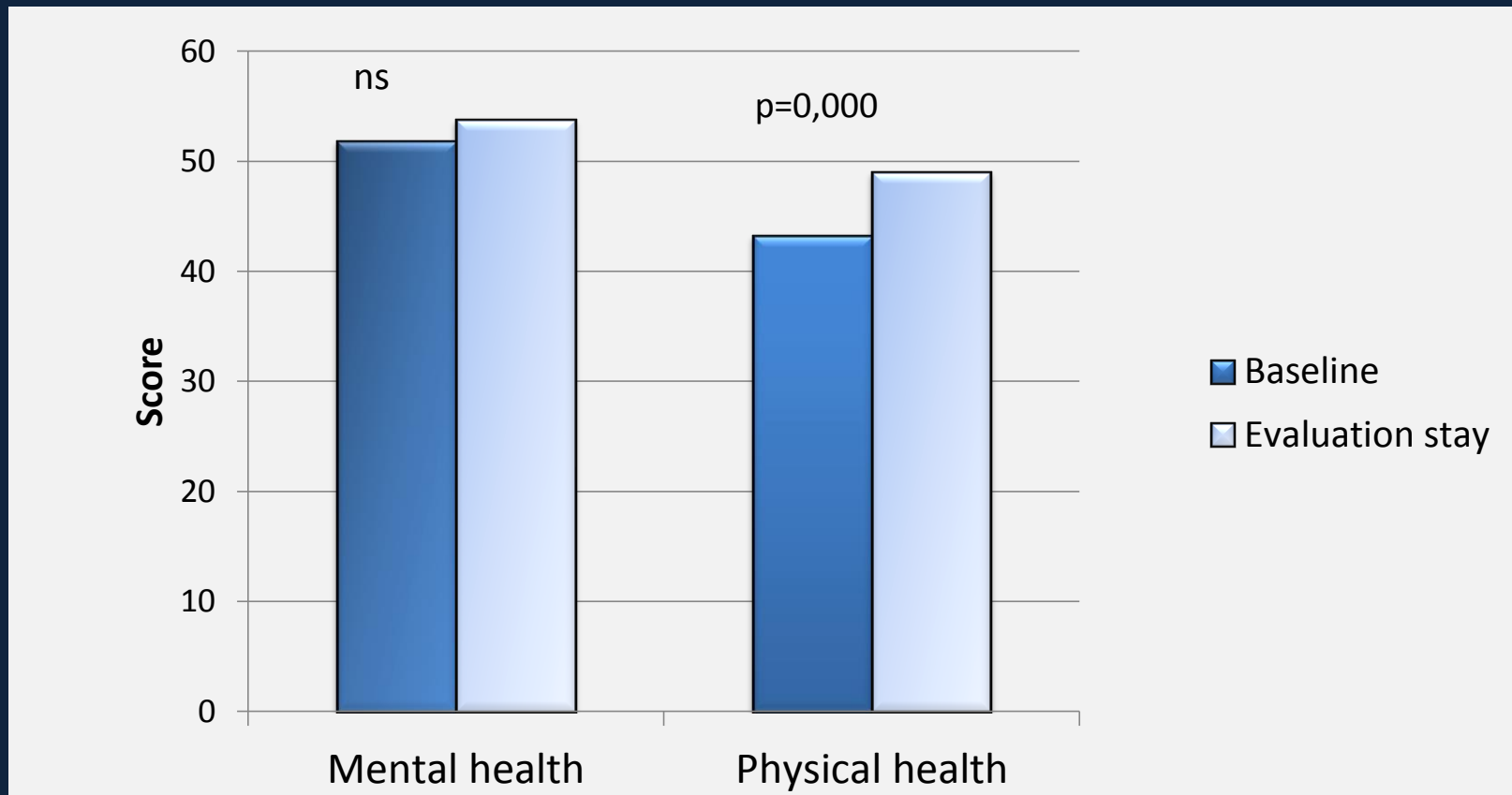
Clinical measures	Mean	SD
MMSE	25,4	3,5
BMI	22,8	3,2
Activities of Balance Confidence (ABC)	72,6	23,9
Bergs Balance Scale (BBS)	52,1	3,8
Timed up and og (TUG)	8,1	3,1
10 Meter Walk test (10MWT)	6,8	2,6
6 Min Walk test (6MWT)	484,9	147,5
Barthel index	19,2	1,3
HADS	8,2	8,7
SF-12 (physical health)	43,9	9,4
SF-12 (mental health)	52,2	11,2

- 31 out of 37 completed the one-year program as planned (83,8 %)



(Photo: North Norway Rehabilitation Center)

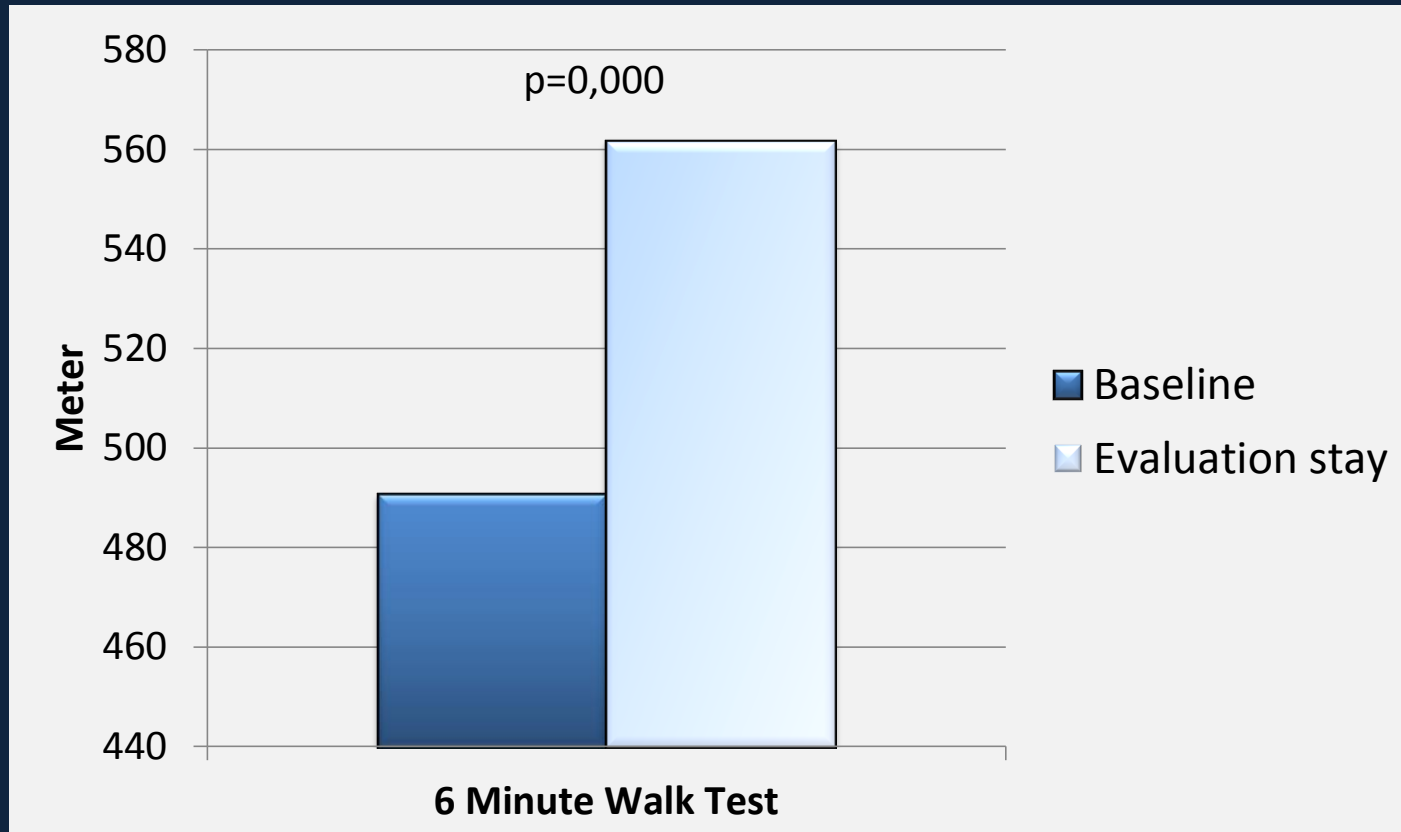
Fig 1. Mean change in health-related quality of life (SF-12)



Piira A, et al PLOS Curr 2013

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



Piira A, et al PLOS Curr 2013

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Fig 3) Change in gait function

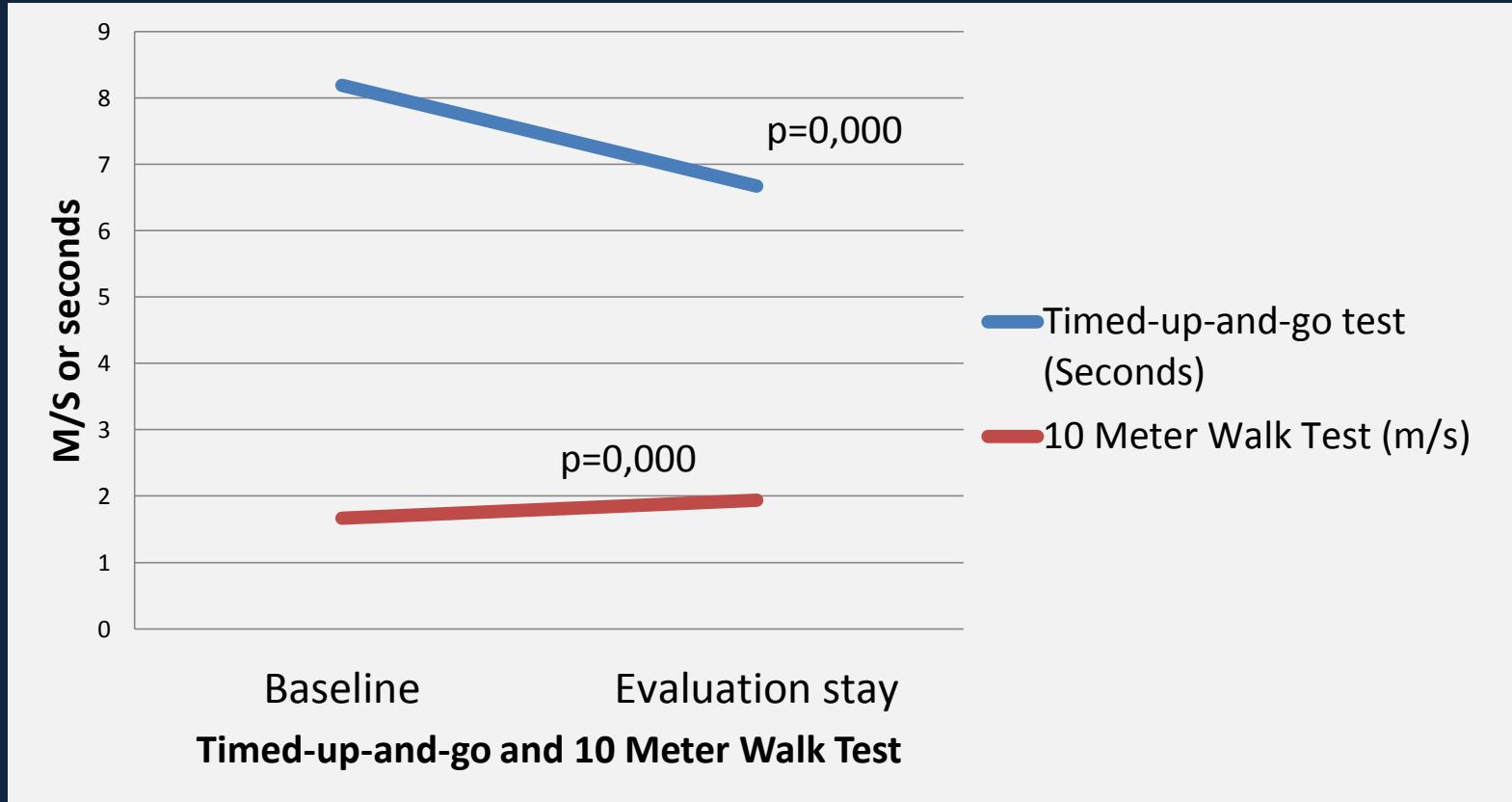




Fig 4) Mean change in balance: Berg Balance Scale (BBS) and Activities of Balance Confidence scale (ABC)

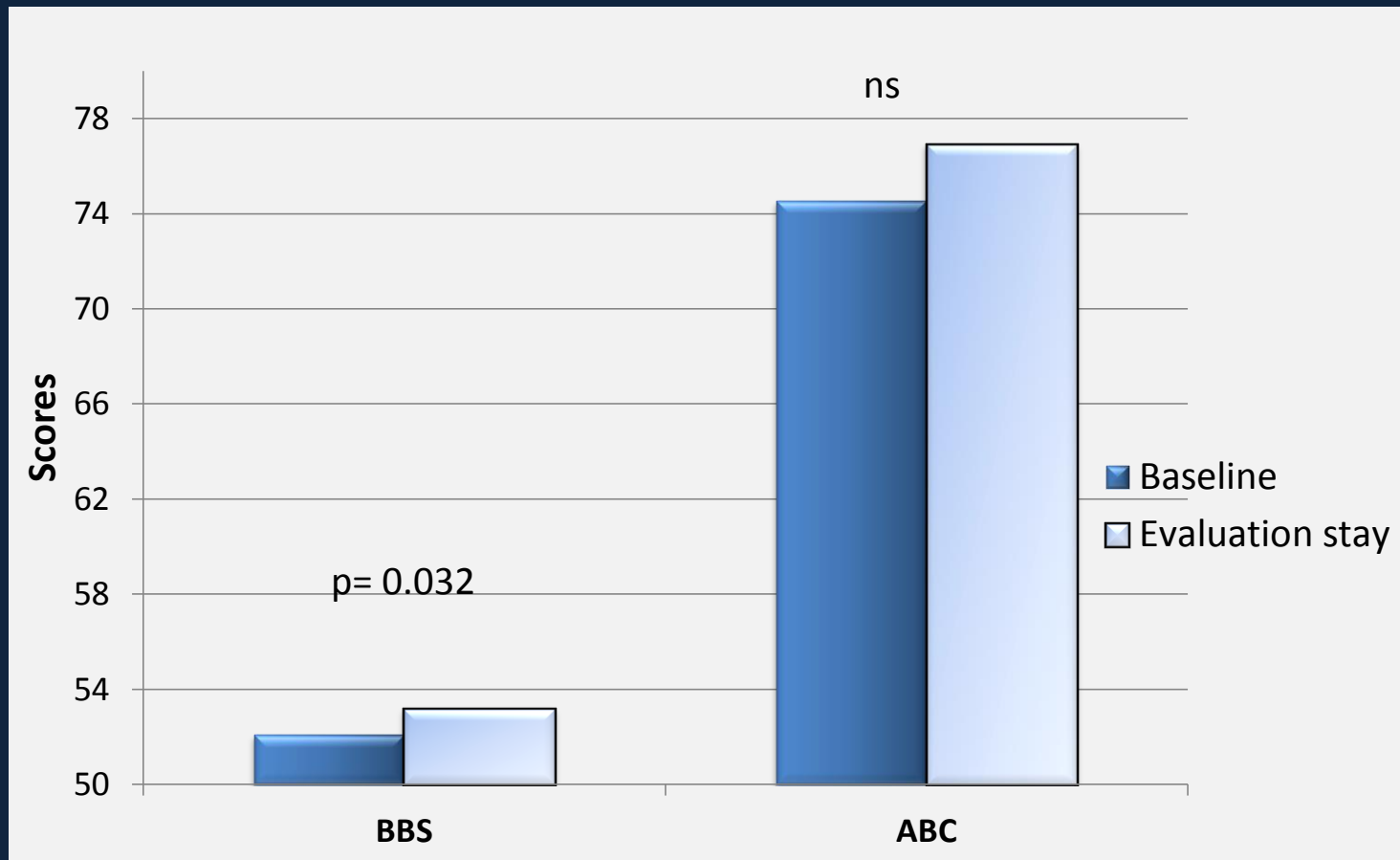


Fig 5) Mean change in BMI, HADS and Barthel Index

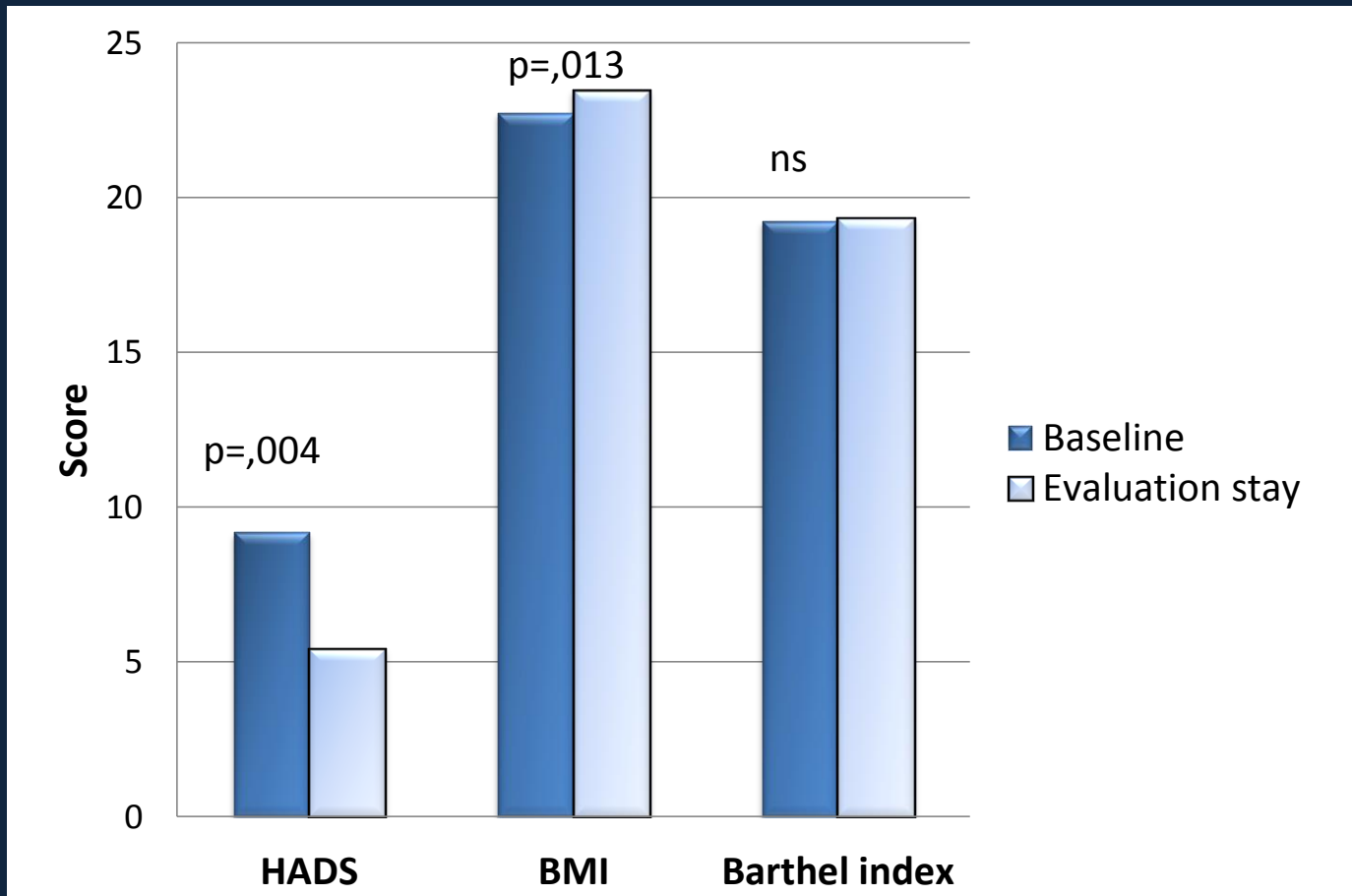
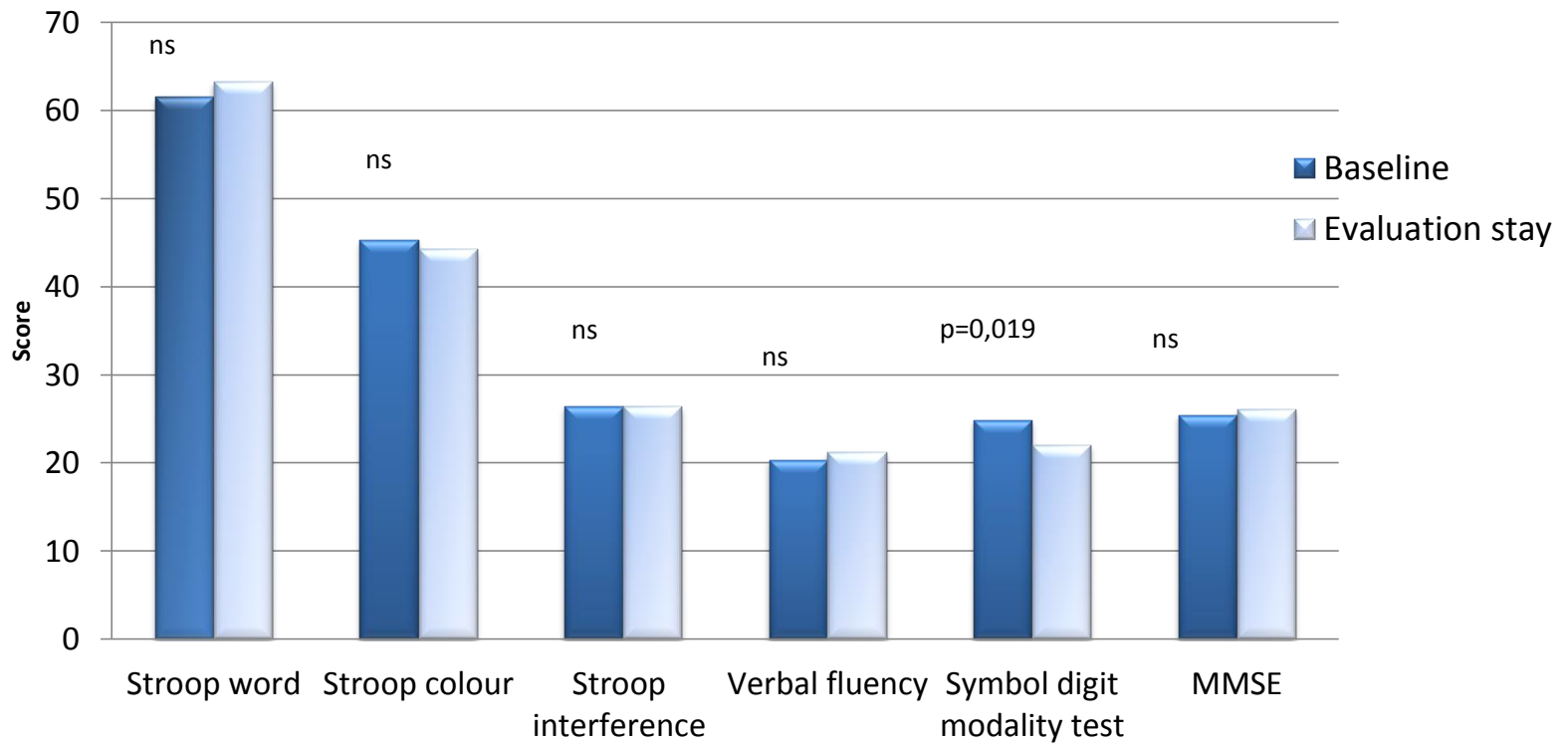


Fig 6) Mean change in cognitive scores



Piira A, et al PLOS Curr 2013

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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?

Frich JC, Røthing M, Berge AR. Participants', caregivers', and professionals' experiences with a group-based rehabilitation program for Huntington's disease: a qualitative study. *BMC Health Serv Res* 2014; 14: 395.

# 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

# 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

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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

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# 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?

Busse, Multidisciplinary rehabilitation: what future? EHDN News, 2014

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