Cognitive and Psychiatric Characteristics of Huntington Disease

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Cognitive and Psychiatric Characteristics of Huntington Disease

I. Huntington Disease (HD): Overview
   Clinical Features of HD

II. Pilot Study: Cognitive and Psychiatric Data
    Preliminary Results

III. Client/Family Education and Support
     Continuum of Care
Huntington Disease: Overview

- A genetically transmitted, progressive neuropsychiatric disorder that can appear at any time in life (autosomal dominant)
- Symptom onset typically third or fourth decade
- HD gene locus at the distal end of the short arm of chromosome 4
- Gene is an unstable CAG trinucleotide repeat sequence
- Cells in the caudate nucleus die, affecting neural pathways to other areas of the brain
Triad of symptoms:
- Movement
- Cognitive
- Psychiatric
Clinical Features (con’t)

• Any of the 3 can be presenting symptoms
• Movements are the most specific
• All patients are eventually affected in all 3 spheres
• 15-20 year course
• Aspiration pneumonia is most common cause of death
Psychiatric Features

Common symptoms
- Apathy
- Irritability
- Dysphoria
- Anxiety

Common syndromes
- Mood disorders
- Intermittent explosive disorder
- Atypical psychosis (schizophrenia)

*Adapted from APA textbook of Neuropsychiatry
Cognitive Symptoms

- Impaired executive function (planning, organization of sequential actions, mental flexibility)
- Problems with sustained concentration
- Impaired visuospatial skills
- Memory problems, with greater impairment in retrieval than retention
- Procedural memory deficits
Pilot Study

• Examined 15 inpatients/outpatients with HD at Ontario Shores
• Clients were administered the Neuropsychiatric Inventory (NPI), Montreal Cognitive Assessment (MOCA), and neuropsychological tests (Verbal Fluency, Symbol Digit Modalities, Stroop) of the Unified HD Rating Scale
• Clients’ age, CAG repeats, and years since diagnoses investigated in relation to their psychiatric and cognitive status
Participant Demographics

- Mean age: 53.53 (SD=15.39)
- Gender: 6 males, 9 females
- Mean years since diagnosis: 10.13 (SD=5.95)
- Mean CAG repeats: 43 (SD=3.12)
- Living arrangements: home (8), hospital/LTCF (7)
## Variables by Living Arrangement

<table>
<thead>
<tr>
<th></th>
<th>Mean Inpatient/ LTCF (SD)</th>
<th>Mean Home (SD)</th>
<th>t-test statistic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>51.9 (12.9)</td>
<td>55.0 (18.1)</td>
<td>-.38</td>
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<tr>
<td>Years Dx</td>
<td>10.3 (6.0)</td>
<td>10.0 (6.3)</td>
<td>.09</td>
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<td>CAG Repeat</td>
<td>43.0 (1.7)</td>
<td>43.0 (3.9)</td>
<td>.01</td>
</tr>
<tr>
<td>NPI (Caregiver)</td>
<td>30.0 (11.7)</td>
<td>34.5 (26.7)</td>
<td>-.41</td>
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<tr>
<td>MOCA Total Score</td>
<td>15.7 (6.8)</td>
<td>23.1 (4.7)</td>
<td>-2.5*</td>
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<tr>
<td>Phonemic Fluency</td>
<td>18.5 (3.7)</td>
<td>37.3 (8.3)</td>
<td>-4.2**</td>
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<tr>
<td>Stroop Colour-Word</td>
<td>24.8 (5.6)</td>
<td>37.2 (10.0)</td>
<td>-2.2</td>
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<tr>
<td>Symbol Digit</td>
<td>14.5 (3.0)</td>
<td>27.5 (11.0)</td>
<td>-2.3*</td>
</tr>
</tbody>
</table>

* p < 0.05
** p < 0.01
Demographics by Living Arrangements

- Inpatient/LTCF: n(female) = 5, n(male) = 2
- Home: n(female) = 4, n(male) = 4
Cognitive Status by Living Arrangement

* p < 0.05
** p < 0.01
Scatterplot: Age and CAG repeats

Age and NPI

Association between Age and CAG repeat, $p < 0.01$
Association between Age and NPI (Client), $p < 0.05$
Scatterplot: Yrs Since Dx & Visuospatial/Executive
Yrs Since Dx & Attention

Association between yrs since dx and MOCA Attention, p < 0.05
Association between yrs since dx and MOCA Visuospatial/Executive, p < 0.05
Neuropsychiatric Inventory (NPI) Caregivers vs. Clients

t(14) = 6.25, p < 0.001
Summary: Preliminary Findings

- Significant differences in cognitive performance between clients living at home and those in hospital/LTCF
- Visuospatial/Executive & Attention subtests significantly associated with yrs since dx - consistent with HD cognitive profile
- Clients with HD significantly underestimate their neuropsychiatric impairments compared to caregivers – anosognosia

Based on findings:
- Investigation with larger sample size/more variables in progress
- Predictors of living arrangement using cognitive and other measures will be explored
- Implications to assist in education of clients/caregivers
Behavioural Changes

Diminished self-awareness leads to problems with:

- Self-determination
- Self-direction
- Self-control and regulation
- Compromised insight and empathy
Managing Behaviour

• Provision of education

• Support and follow-up in community

• Community resources
Continuum of Care

Outreach Services

5 Bed Inpatient Unit

HD Clinic
Dr. Jean Byers, Dr. Rosa Ip and Carol Harren RN will be presenting at the 2012 National Conference on HD in Toronto in November.