Cognitive and Psychiatric Characteristics of Huntington Disease

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Cognitive and Psychiatric Characteristics of Huntington Disease

I. Huntington Disease(HD): Overview

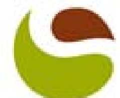
   Clinical Features of HD

II. Pilot Study: Cognitive and Psychiatric Data

   Preliminary Results

III. Client/Family Education and Support

   Continuum of Care



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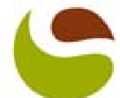
Huntington Disease: Overview

- 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



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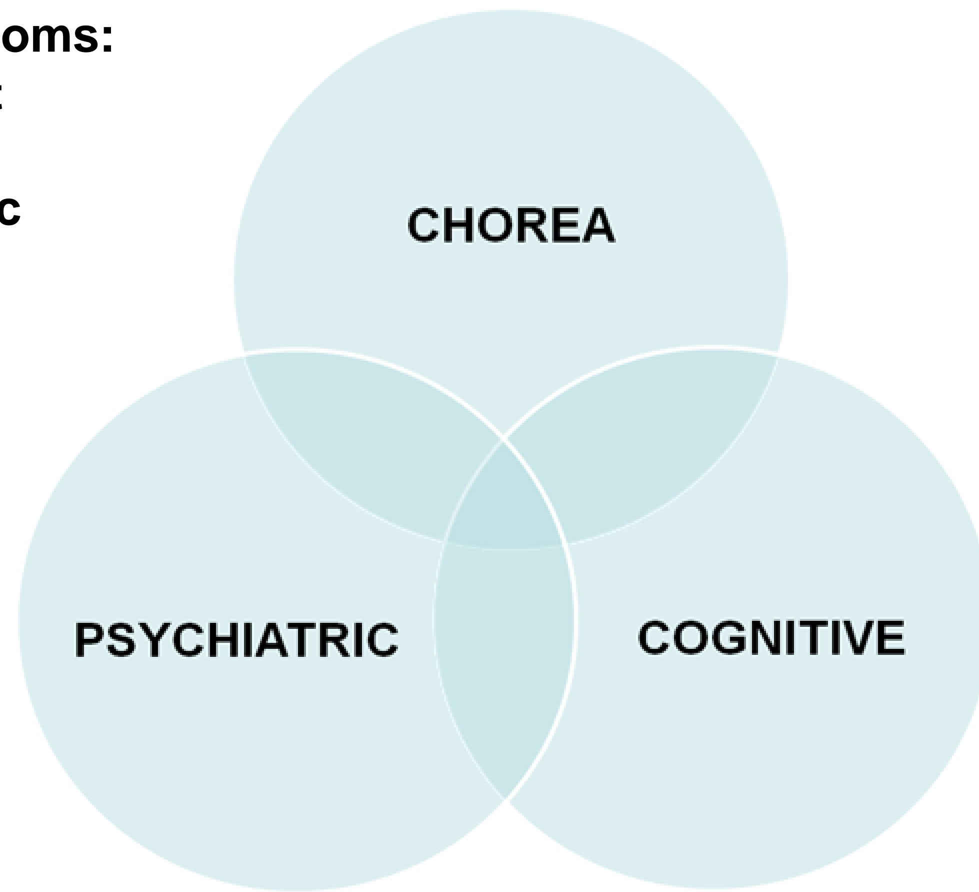
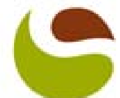
Huntington Disease: Clinical Features

Triad of symptoms:

- **Movement**
- **Cognitive**
- **Psychiatric**





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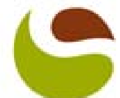
Clinical Features (con't)

- 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



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Psychiatric Features

Common symptoms

- Apathy
- Irritability
- Dysphoria
- Anxiety

Common syndromes

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

*Adapted from APA textbook of Neuropsychiatry

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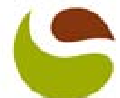
Cognitive Symptoms

- 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



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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

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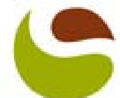
Participant Demographics

- 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)



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Variables by Living Arrangement

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

* $p < 0.05$

** $p < 0.01$

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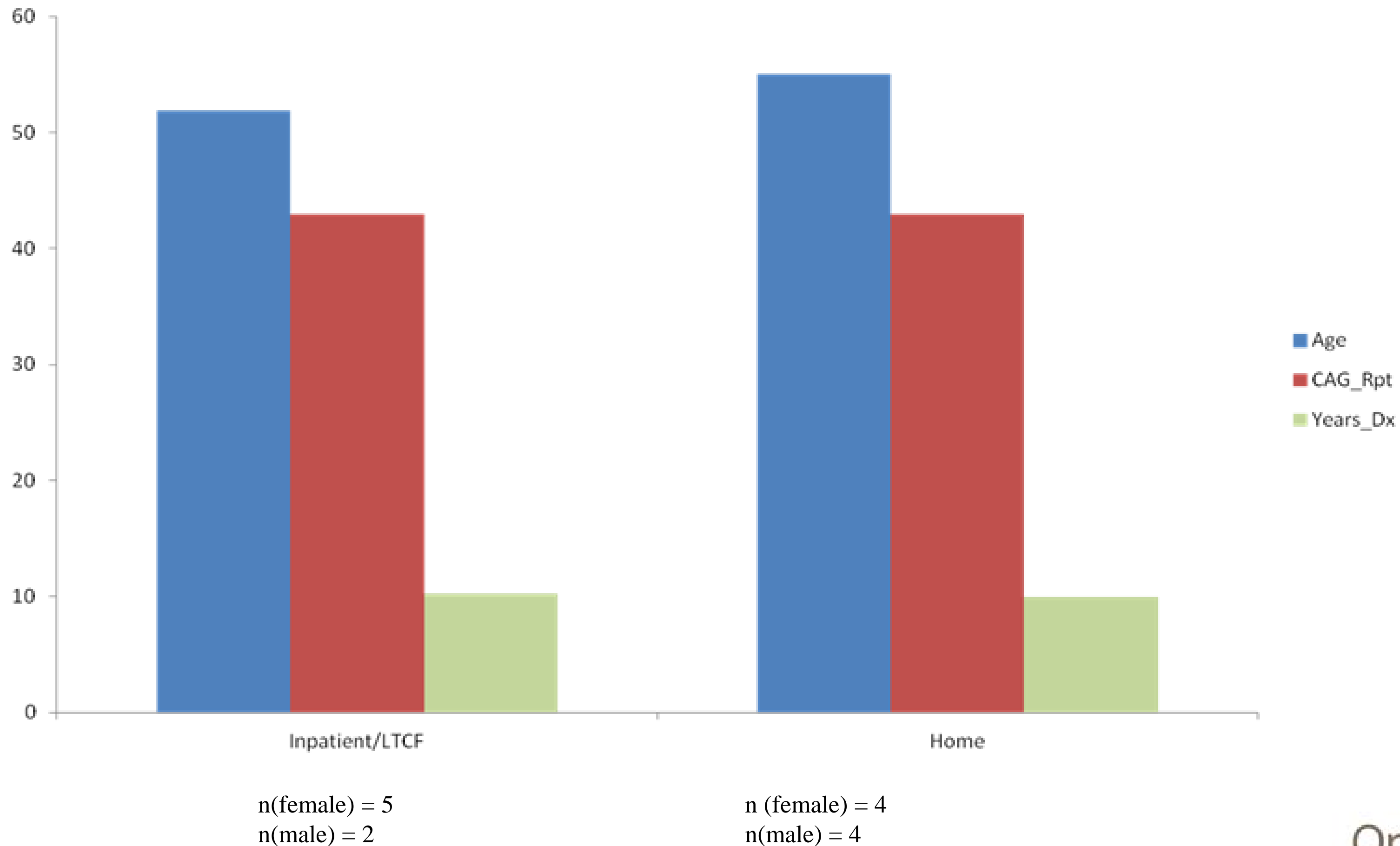
Demographics by Living Arrangements



n(female) = 5
n(male) = 2

n (female) = 4
n(male) = 4

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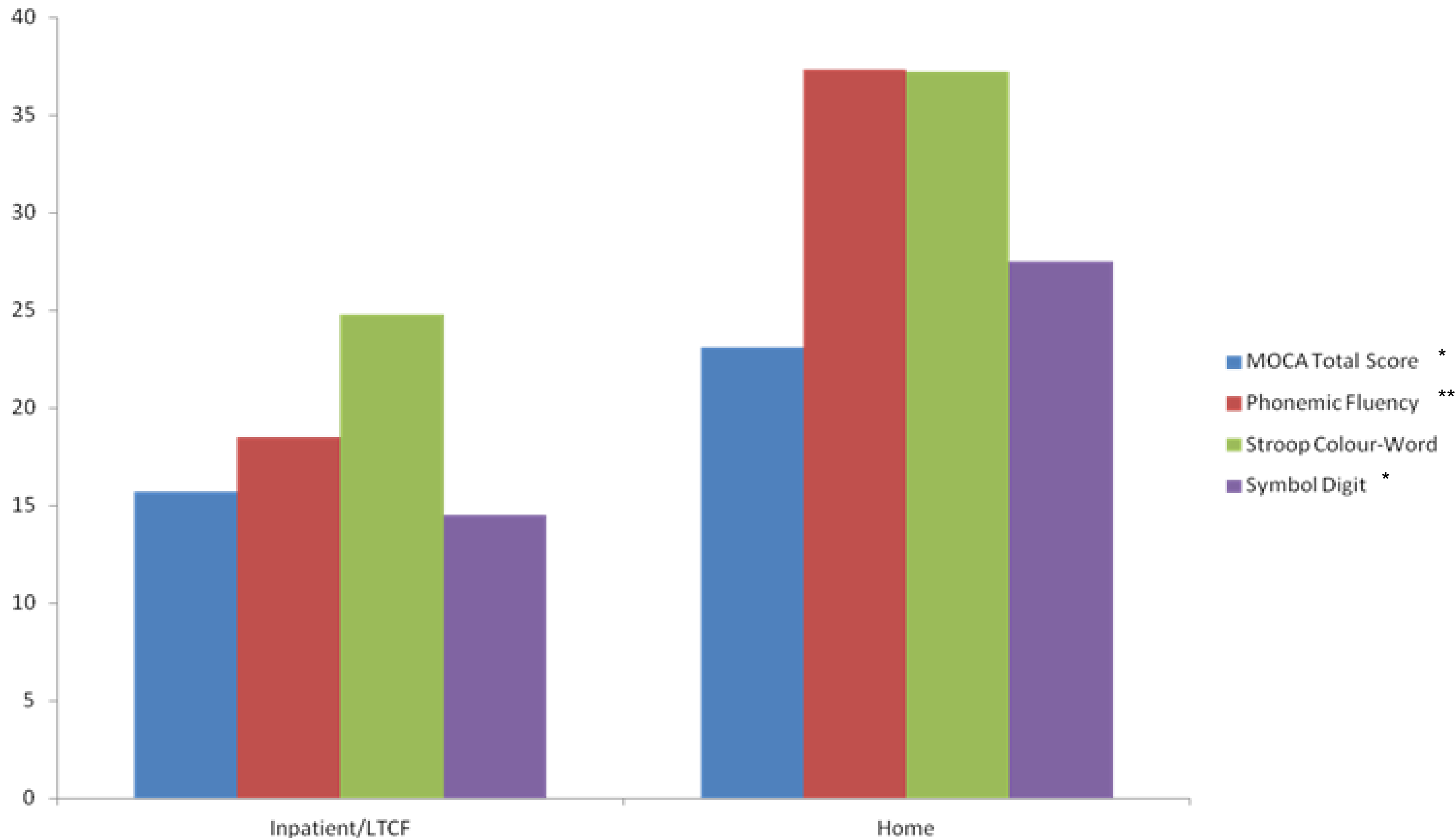
Cognitive Status by Living Arrangement



* $p < 0.05$
** $p < 0.01$

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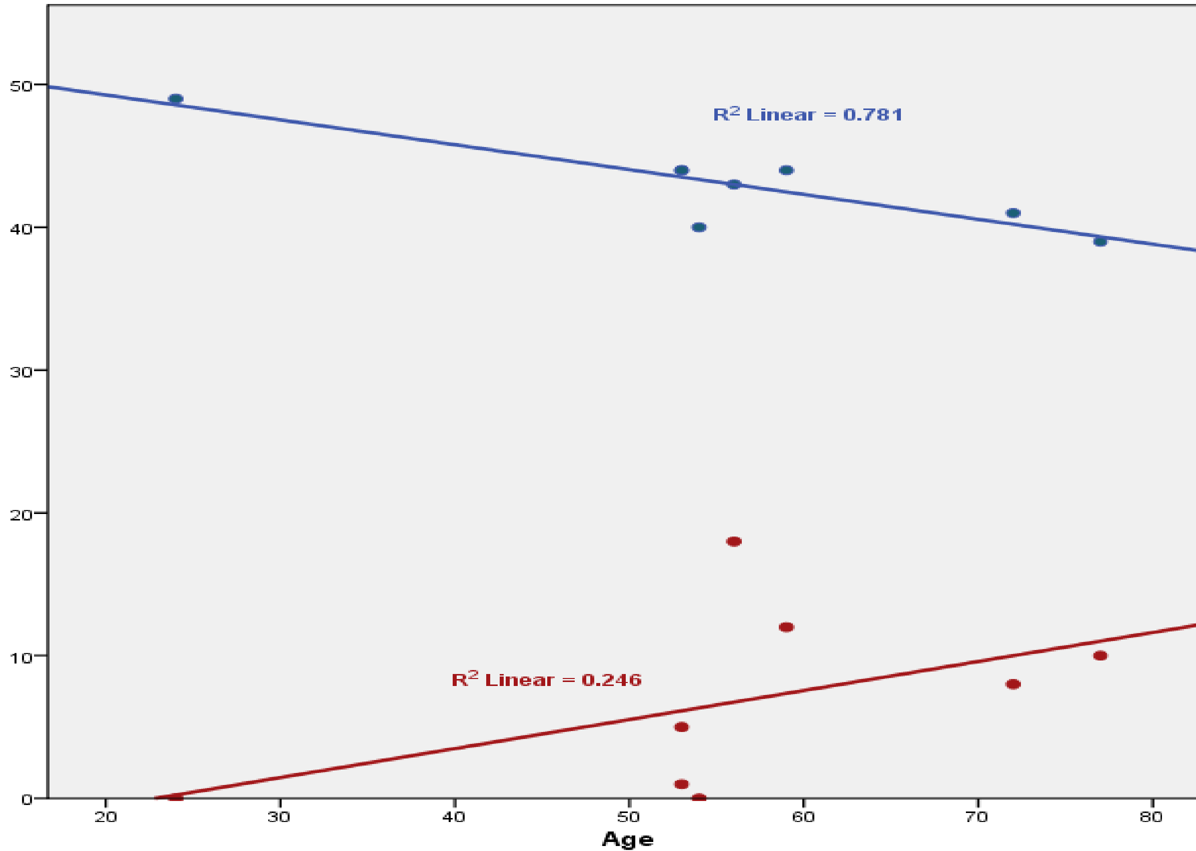
Scatterplot: Age and CAG repeats
Age and NPI



_______Association between Age and CAG repeat, $p < 0.01$

_______Association between Age and NPI (Client), $p < 0.05$

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Scatterplot: Yrs Since Dx & Visuospatial/Executive
Yrs Since Dx & Attention

R^2 Linear = 0.327

R^2 Linear = 0.363

Mean

0 1 2 3 4 5 6

0 5 10 15 20 25

Years_Dx

______Association between yrs since dx and MOCA Attention, $p < 0.05$

______Association between yrs since dx and MOCA Visuospatial/Executive , $p < 0.05$

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Neuropsychiatric Inventory (NPI) Caregivers vs. Clients

Mean

70 60 50 40 30 20 10 0

20 30 40 50 60 70 80

Age

NPI (Care Giver) NPI (Client)

t(14) = 6.25, $p < 0.001$

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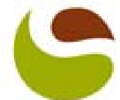
Summary: Preliminary Findings

- 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



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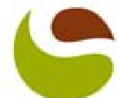
Behavioural Changes

Diminished self-awareness leads to problems with:

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



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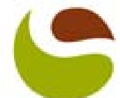
Managing Behaviour

- Provision of education
- Support and follow-up in community
- Community resources



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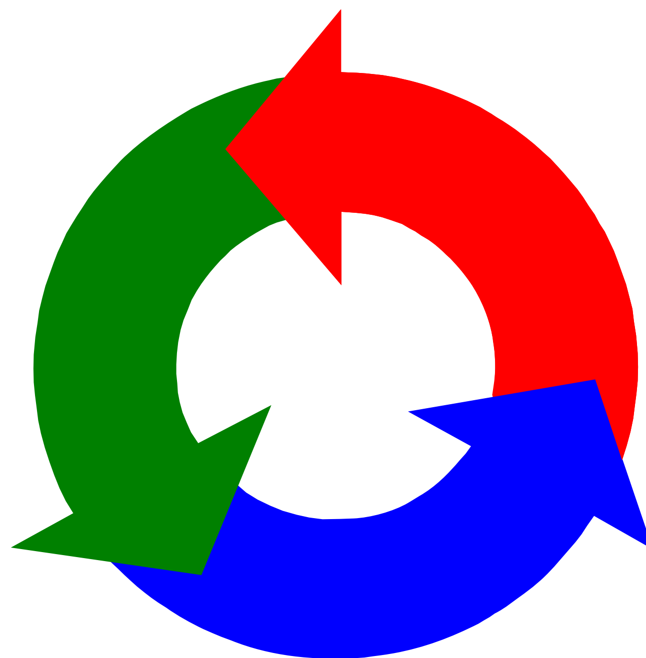
Continuum of Care



Outreach Services

HD Clinic

5 Bed Inpatient Unit

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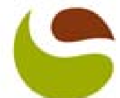
Dr. Jean Byers, Dr. Rosa Ip and Carol Harren RN will be presenting at the 2012 National Conference on HD in Toronto in November



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