Managing Huntington's Disease

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Genetic Testing

- Positive results (>40 CAG repeats)
- Negative results (<26 CAG repeats)
- What does it mean when...
  - 27-35 CAG repeats
  - 36-39 CAG repeats
I’m POSITIVE, now what?

• **Asymptomatic**
  – Yearly follow-up in the Genetic Movement disorders Clinic

• **Symptomatic**
  – Follow up depends on the symptoms
  – Bi-annual follow up with Neurology
  – Yearly follow up with Genetics
Epidemiology

• 5-10/100,000
• Approx. 25,000 with diagnosed HD in the USA
• 125,000 at risk
  – Vast majority of asymptomatic, at risk patients have not been tested
• World wide benefit of Genetic counseling
  – Lowered the frequency of the HD gene
Natural History

- Mean age of onset: 39 years
- Juvenile HD: onset of illness before the age of 20; Parkinsonism with rigidity; dystonia; cognitive impairment
- Parent of Origin effect
- Law of Anticipation
- Duration of illness: variable
Movement Disorders

• **Chorea**
  – Greek verb “to dance”
  – Classic HD symptom

• **Dystonia**

• **Parkinsonism with Rigidity**

• **Eye movement abnormalities**

• **Other hyperkinetic movements**
  – Tics
Cognitive Disorders

**Sub-cortical dementia**

- Psychomotor slowing - slowed thinking
- Executive dysfunction
- Recognition spared
- Recall affected
- Rate of progression is variable
Behavioral Disorders

- Depression (first sign social hindrance)
- Reactive Depression
  - Changes in working memory
- Suicide
- Psychosis
- Personality changes
- Sleep disorders
Suicide

• Topic warrants special attention
• Dr. Huntington himself recognized it
• Risk factors
  – Severity of depression
  – Degree of functional capacity retained
  – Level of insight retained
  – Lack of social support network
Swallowing difficulty

- Difficulty with articulation
- Choking
- Low threshold for Speech Therapy evaluation
Symptomatic Intervention

• Movement Disorders
• Cognitive and Behavioral Disorders
• Swallowing difficulty
Treatment of Movement Disorders

- Anti-psychotic medications
  - Zyprexa
- Use of L-dopa in those with Parkinsonism or rigidity
- Tetrabenazine
- Botox
- Physical Therapy
Treatment of Behavioral Disorders

- Anti-depressants
- Anti-psychotics
- Psychiatric intervention with psychotherapy
Treatment of swallowing difficulty

- Speech Therapy evaluation
- Bedside swallowing evaluation
- Modified Barium Swallow Study
- PEG tube placement
Utility of Physical Therapy

• Evaluation for home exercise program
• Gait and balance training
• Assistive device evaluation
• Fall prevention
New Directions

- Disease modifying agents
- Stem cell implantation
- Cure?