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HDSA encourages all attendees to consult with their primary care provider, neurologist or other healthcare provider about any advice, exercise, medication, treatment, nutritional supplement or regimen that may have been mentioned as part of any presentation.
Presenter disclosures

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The following personal financial relationships with commercial interests relevant to this presentation existed during the past 12 months:

No relationships to disclose or list
Tongue twisters

• “Anosognosia”

• “Perseveration”
Tongue twisters

• “Anosognosia:” “unawareness” or “lack of insight;” people with HD often do appreciate that they have, chorea, incoordination, cognitive or emotional change, or do not appreciate how severe they are

• “Perseveration:” “repetition” or “getting stuck;” people with HD often repeat questions or statements, or get “stuck” or “fixated” on certain topics or issues
Features of HD

- Motor disorders—progressive
- Cognitive disorders—progressive
- Psychiatric ("behavioral”/ emotional”) disorders—variable
  - Unawareness and apathy
  - Depression and irritability
  - Perseveration and obsessions and compulsions
  - Disinhibition
  - Other: sexual, sleep etc etc
Brain changes in HD: Neuropathology
Brain changes in HD

-Predict-HD Study
Brain circuits affected in HD
Progression of Brain Changes in HD
Progression of HD

- Premanifest
- Motor diagnosis
- Manifest

- Functional abilities
- Motor impairment
- Cognitive impairment/dementia
- Chorea
- Shoulson-Fahn 'stages'

- Presymptomatic
- Prodromal
- Clinical
- Early stages
- Moderate
- Advanced

- Age (years)
- Typical adult onset
Issues related to perseveration: obsessions and compulsions

• Obsessions are thoughts that recur that are unwanted, and that the person wishes not to think

• Compulsions are behaviors that recur that are unwanted, and that the person wishes not to do

• Examples: “obsessive tune” – common for nearly everyone, checking locks, handwashing etc
Issues related to perseveration: obsessions and compulsions

- Obsessions and compulsions often need no treatment at all.

- When they become a problem, they are often very responsive to both behavioral treatment and medication (e.g., “Prozac” like drugs)
Issues related to perseveration: tics and stereotypies

- Tics are repeated complex movements that are involuntary (like chorea but more complex repetitious)
- Stereotypies similar
- They may be vocal, eg a few words
- These are best seen as motor phenomena and can sometimes be treated with medications for chorea
Issues related to perseveration: delusions

• Delusions are a more serious phenomenon
• A “delusion” is a fixed false idiosyncratic belief.
• Fixed—cannot be convinced otherwise
• False—not true
• Idiosyncratic—unique or unusual, not culturally sanctioned
Issues related to perseveration: delusions

- Delusions are a more serious phenomenon
- Delusions are treatable with medication, and may require hospitalization
Issues related to perseveration: “disinhibition”

• Individuals with HD may undergo a number of personality changes
• Disinhibition refers to emergence of behavior which would normally be seen to be socially inappropriate
• May be repetitious in that it is noticeable when it happens
Issues related to perseveration: depression

• Whenever someone's behavior changes, consider the possibility of depression

• Clinical depression—persistent sad (or irritable) mood, loss of interest in usual activities, and usually with changes in thinking about oneself or others

• Can unusually be treated effectively with medication and psychotherapy
Issues related to perseveration: memory difficulties

• An individual with HD may repeat a question or comment, forgetting that the issue has already been addressed
Coping with perseveration

• Perseveration is not common at some stages of the illness for many people with HD
• Remember that this is often a consequence of the disease: the individual with HD is not getting “stuck” on things deliberately or to be annoying
• The individual with HD often is not aware of being “stuck” on things (“anosognosia”)
Coping with perseveration

- Every example is unique….but:
- It may be difficult to get individuals with HD to change their behavior…. 
- It is usually easier for the others around the person with HD to change….and learn to accept that this is just part of HD
- It is important to “pick your battles”
Coping with perseveration, and other behavioral issues in HD

• It usually helps to have a predictable routine from day to day
• If something is going to change, plenty of advance notice (and repetitions) can be helpful
• A “low stress” environment is helpful for everyone....
Coping with perseveration, and other behavioral issues in HD

- Avoid arguments
- Avoid contradictions
- Use redirection
- Offer a choice
Coping with perseveration, and other behavioral issues in HD

- Often a psychiatrist, social worker, psychologist, or other professional can help.
- If all else fails, keep in mind that HD is not static; a problem behavior will often resolve on its own.
Unawareness/ "Anosognosia"

- “Anosognosia” from Ancient Greek a-, "without", nosos, "disease" and gnōsis, "knowledge."
- Present in many neuropsychiatric conditions, eg Alzheimer’s disease, stroke, schizophrenia
- Anosognosia may be caused by dysfunction in the cortex of the brain (the “parietal region) on the right side