MANAGING YOUR COGNITIVE SYMPTOMS

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Presenter Disclosures

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

No relationships to disclose
OBJECTIVES

• Learn basic mechanism of cognitive decline in HD
• Recognize common symptoms of cognitive decline
• Review strategies to manage cognitive symptoms
Overview of Huntington’s Disease

- Huntington’s disease (HD) is a hereditary, neurodegenerative/neuropsychiatric disorder
  - Neurodegenerative = progressive, worsening of symptoms over time
- Age of onset: main peak, around 30-40s, although it can occur at any age
- No cure but we know a lot about the disease
  - Inherited, autosomal dominance pattern
  - Mutant Huntingtin protein (mHTT)
  - mHTT located on the short arm of chromosome 4
  - Polyglutamine repeat disorder (CAG)
Cardinal Features of Huntington’s disease

• 3 M’s
  – Mood (Psychiatric disorders)
  – Movements (involuntary, abnormal)
  – Memory

• Clinical diagnosis is usually made by motor symptoms
Cognitive Symptoms in HD

- Has been well characterized in the various stages of HD as well as in the prodrome before the motor diagnosis is given
  - can be evident up to 15 years prior to the motor diagnosis
  - Research has suggested that cognitive and behavioral changes place the greatest burden on families
    - highly associated with functional decline
    - can be predictive of institutionalization
- Cognitive decline is associated with biological markers
  - brain atrophy
  - circulating levels of brain-derived neurotrophic factors
What is dementia?

• Dementia: degenerative disorder involving the compromise of multiple domains of cognition
  – Can be distinguished from:
    • Normal variation of forgetting, normal from typical aging
    • Confusion/delirium

• Delirium: an acute mental disturbance characterized by confused thinking and disrupted attention usually accompanied by disordered speech and hallucinations, can fluctuate
  – Infections
  – Electrolyte disturbances
  – Medication induced
  – Intoxication
  – hospitalizations
Cognitive Features of HD

• Cognitive Decline in HD is different than Alzheimer’s Disease (AD)
• Memory decline in HD comes from the subcortical structures of the brain being affected and from the cortical structures in AD
• Both types of dementia can cause impairments in memory, reasoning and judgement
Cognitive Features of HD

• Is differentiating the cortical/subcortical dementias important? Maybe?…Or not?
  – May help with prognosis or predicting future symptoms
  – Question targeting memory medications
  – Problem – HD could have some elements of both subcortical and cortical problems?
    • Atrophy of both areas
    • Connections of cortical to subcortical areas
### Some Differences between Subcortical (e.g. Huntington’s and Parkinson’s diseases) and Cortical dementias (e.g. Alzheimer’s)

<table>
<thead>
<tr>
<th>SUBCORTICAL</th>
<th>CORTICAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Change in personality</td>
<td>Less change in personality</td>
</tr>
<tr>
<td>Slower thought processes, right answer</td>
<td>Normal speed, wrong answer</td>
</tr>
<tr>
<td>Language is affected later</td>
<td>Language is found early, word finding difficulty, understand what people are saying</td>
</tr>
<tr>
<td>Speech may be slurred/slowed but accurate</td>
<td>Normal speech but inaccurate</td>
</tr>
<tr>
<td>Less severe changes in memory</td>
<td>More changes in memory</td>
</tr>
<tr>
<td>Recall can be benefited by cues</td>
<td>Recall cannot be benefited by cues</td>
</tr>
<tr>
<td>Learning is slowed/disorganized but able to learn new info</td>
<td>Rapid forgetting, cannot learn new info</td>
</tr>
</tbody>
</table>
Cognitive Symptoms of HD

• Impacts a number of areas of function:
  – Memory and learning
  – Perceptual skills
  – Executive functioning
  – Language and communication
  – Self Awareness and Insight
LEARNING AND MEMORY

• Learning: gaining knowledge or skills through experience, study, or by being taught
  – Often involves combining multiple sources of information

• Memory: The ability to retrieve previously learned information
  – Recognition vs free recall
    • Recognition: ability to “recognize” an event or piece of information as being familiar
    • Recall: retrieval of related details placed into long term memory
LEARNING AND MEMORY

- **Explicit** vs implicit long term memory
  - **Explicit**: information that you have to consciously work to remember
    - names, dates, formulas for school, phone numbers, ATM numbers, etc.
  - 2 types of explicit memory
    - » Episodic – events, what you did yesterday
    - » Semantic – general knowledge, names, concepts, facts
LEARNING AND MEMORY

- Explicit vs **implicit** long term memory
  
  **Implicit:** information that is unconsciously or effortlessly remembered
  
  - “Motor memory”
  
  - collection of coordinated skills or movements
  
  - Driving, riding a bike, typing, etc.
Long-term memory

Explicit (declarative)
- Episodic (experienced events)
- Semantic (knowledge and concepts)

Implicit (non-declarative)
- Procedural (skills and actions)
- Emotional conditioning
TIPS FOR LEARNING AND MEMORY PROBLEMS

• Notes
  – Sticky Notes, pocket-size memory notebook, notes section of the iPhone
  – One idea per note
• Repeat what you want to remember
• Practice, practice, practice
• Imagine what you want to remember
• Minimize sources of information
• Photos with stickers with names on them around the house
PERCEPTION PROBLEMS

• Perception: ability to identify, interpret and organize information taken in through the senses
  – Sense of time
  – Spatial perception (or personal space)
  – Emotional recognition
  – Smell identification

  – diverse brain regions associated with the sense of time (frontal/parietal cortex, basal ganglia, cerebellum, and hippocampus)
PERCEPTION PROBLEMS

• Sense of time
  – Internal timing is diminished
  – Ability to estimate time worsens
    • Time it takes to get somewhere
    • Time it takes to get somewhere during a particular time of the day
TIPS FOR PERCEPTION IMPAIRMENTS

• Time management
  – Allow extra time
  – Map out timeline
  – Use reminders/alarms/calendars
PERCEPTION PROBLEMS

• Personal space
  – Ability to judge where the body is in relation to objects
    • Can lead to increased falls and accidents
  – Ability to navigate around a familiar town
TIPS FOR PERCEPTION IMPAIRMENTS

• Personal space
  – Minimize clutter
  – Pad the corners of furniture
PERCEPTION PROBLEMS

• Emotional recognition
  – Inability to differentiate certain emotions from facial expressions
  • Fear, surprise, anger, happiness, sadness
    – Impacts personal and social relationships
    – Amygdala/cortex affected
TIPS FOR PERCEPTION IMPAIRMENTS

• Emotional recognition
  – Say what you are feeling
  – Active dialogue
EXECUTIVE DYSFUNCTION

• Executive function: frontal lobe cognitive process that regulates organization of thoughts and activities, prioritization of tasks, and decision making
  – Higher order functions
  – Management
    • Processing speed
    • Attention
    • organization
EXECUTIVE DYSFUNCTION

• Processing speed
  – Speed at which your brain works with information
    • Tasks may take longer to complete
    • Completing a task requires more energy and leaves the individual worn out
EXECUTIVE DYSFUNCTION TIPS

• Slowed processing
  – Allow for extra time to respond
  – Avoid open ended questions
EXECUTIVE DYSFUNCTION

• Attention
  – Ability to attend to and process specific information for the world around you
  – Multitasking becomes more difficult
EXECUTIVE DYSFUNCTION TIPS

• Attention
  – One task at a time
  – Minimize input sources
  – Reduce the quantity of information
  – Remove environmental distraction
    • Turn off the radio/television
    • Try to calm the children or pets
EXECUTIVE DYSFUNCTION

• Organization
  – Ability to sort information and construct meaningful patterns of thought and action
  • Determining the order of activities for the day/task
    – Getting dressed
    – Brushing teeth
  • Coordinating schedules for the family
  • Shopping for groceries
EXECUTIVE DYSFUNCTION TIPS

• Organization tips
  – Get a routine or plan schedules
    • Can pair tasks together (e.g. wash face, brush teeth)
  – Use calendar, “honey do” lists, schedules
    • Assign one place in the house where the schedules/lists are
    • Refer to these items frequently
    • Check lists, feeling of accomplishment when you can cross off a task
• Language involves the use of words in a structured and conventional way
• Communication is the transfer of information from one person to another
• 2 main parts of communication
  – Getting the information IN (listening, processing and understanding)
    • receptive
  – Getting the information OUT (organization and presenting)
    • expressive
TIPS FOR LANGUAGE AND COMMUNICATION PROBLEMS

• Allow time to respond
• Offer verbal cues or prompts
• Provide specific and limited choices
• Don’t pretend that you know what they are saying or asking
  – Ask for the patient to repeat
  – Ask them to write out the words
Tips for Communication

• Ask an open ended question and then wait..
• Rephrase the question into multiple choice and then wait…
• Rephrase it as a ‘yes’ or ‘no’ question and then wait..
• Ask the question again later to avoid frustration for both people involved

From Jimmy Pollard’s *Hurry Up and Wait*
Insight or Self Awareness

- Insight/ascogonia
  - Unable to recognize disabilities
    - Falls, working, driving, etc.
    - Can lead to irritability/frustration
  - Can lead to poor hygiene
    - Bathing and teeth hygiene
    - Can lead to infections
      - e.g. Poor dentition can lead to cavities and aspiration pneumonia
  - Family member’s needs
    - Can lead to social strain
  - Awareness of pain

Areas contributing to poor insight: connections of frontal and parietal lobes + basal ganglia
Tips for Insight/Self Awareness

• Allow the family to come to the office visit and give their opinions of how the patient is doing
• Schedule the routine for hygiene
• Additional assessments with other therapies (speech, occupational or physical therapies) to help assess disabilities
• Driver’s evaluations
The Big 5 Problems revisited

• Slower thinking
• Recognition is easier than recall
• Difficulty maintaining focus or multitasking
• Difficulty organizing thoughts and actions
• Can’t wait
The Big 5 Problems revisited with strategies

- Slower thinking…
  - Slow down too
- Recognition is easier than recall…
  - Give multiple choice questions
- Difficulty maintaining focus or multitasking…
  - Do one thing at a time
  - Keep the conversation focused
- Difficulty organizing thoughts and actions…
  - Get into a routine
- Can’t wait…
  - As soon as possible
Caregiver Tips…Remember to:

- Avoid talking in a noisy area
- Try to keep calm voice
- Avoid confrontation
What you can do frequently to help with your “brain health”

- Physical exercises (safely)
- Mental exercises
- Eating well/hydration
When memory is dramatically different than the previous days, assess:

- New medications
- Recent procedure
- Hospitalization
- Poor sleep
- Infection (e.g. UTI, colds, coughs, pneumonias, etc.)
- Change in habit/routine, overstimulation
- Hydration
- Other comorbidities affected (low or high blood sugars, electrolyte abnormalities)