



MANAGING YOUR COGNITIVE SYMPTOMS

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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



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)

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

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

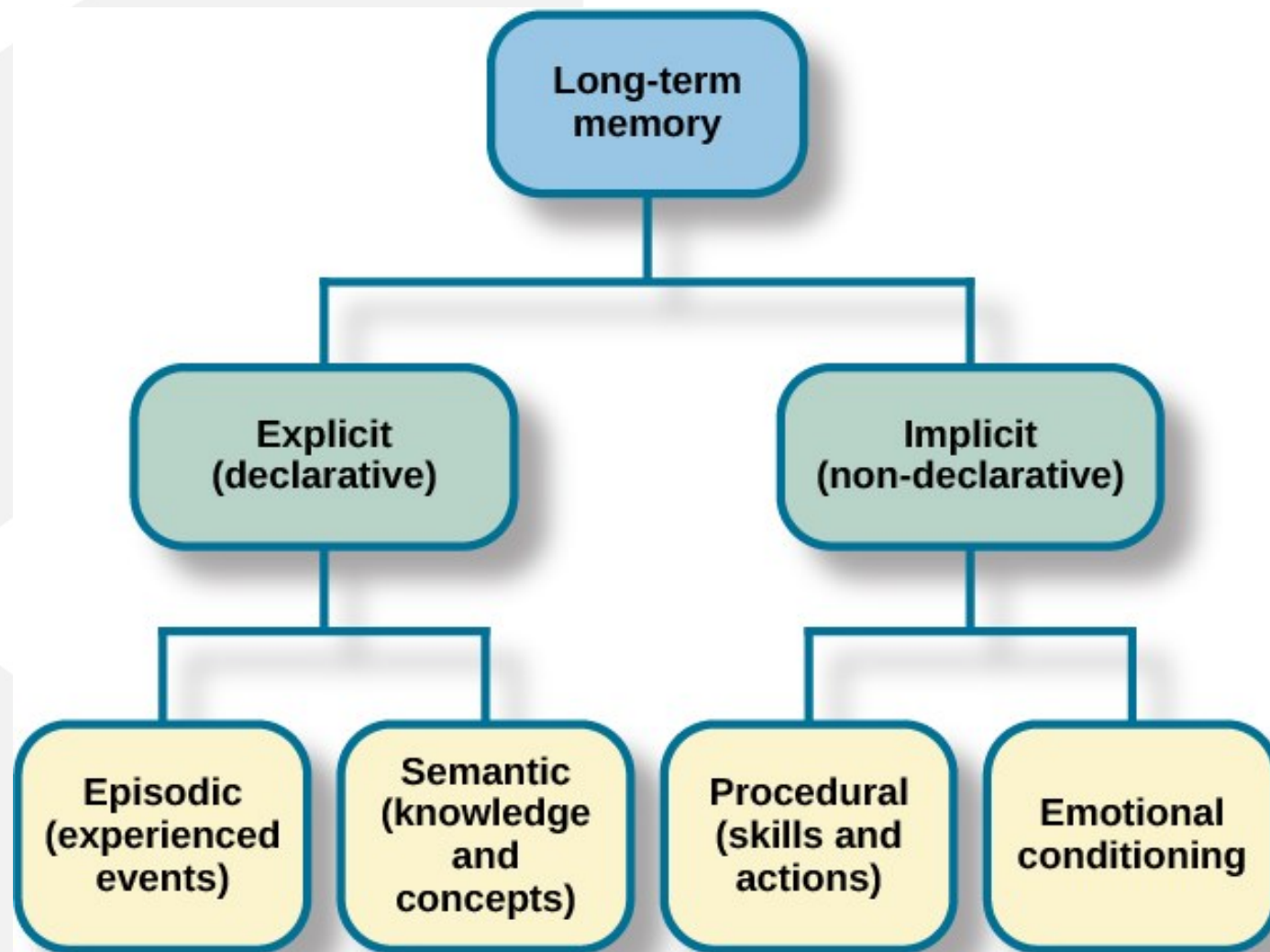
- Learning: gaining knowledge or skills through experience, study, or by being taught
 - Often involves combining multiple sources of information
- Memory: The ability to retrieve previously learned information
 - Recognition vs free recall
 - Recognition: ability to “recognize” an event or piece of 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.



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 AND COMMUNICATION

- 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/anosognosia
 - 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)



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