Managing Cognitive Decline in HD

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Huntington’s Disease Society of America
Presenter Disclosures

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NEUROPSYCHIATRIC SYMPTOMS

• Affect
• Behavior
• Cognition
• The A, B, Cs of neuropsychiatry
EACH BRAIN AREA IS SPECIALIZED
FRONTAL LOBE: 3 DIVISIONS

DORSOLATERAL  ORBITOFRONTAL  MEDIAL FRONTAL
AFFECT
AFFECT (OR MOOD)

- 35-73% of HD patients
- Depression: most common psychiatric sx
- Mania: 2-12%
- Suicide: ~6%
- Anxiety
AFFECT (OR MOOD)

• Why is it affected?
  – All *mood* comes from *brain*
  – It changes, just like *cognition*
  – ANATOMY: less well defined area, but depression may correlate with right frontal lobe changes
  
  – Hence, frontal changes in HD patients affect mood
BEHAVIOR
BEHAVIOR: WHY?

Change in REGULATION of behavior

Change in MOTIVATION
**BEHAVIOR REGULATION:**

**ORBITOFRONTAL**

- **RESPONSE SIZE:** Brake Pedal
  - When I’m cut off by a driver,
    - Do I make an obscene gesture?
    - Do I say “whatever”?
  - **DYSFUNCTION:** aggression, agitation
- **MODULATION** based on social customs
  - **Disinhibition:** for a pretty woman,
    - Do I say hello?
    - Do I whistle?
    - Do I touch her?
BEHAVIOR REGULATION: ORBITOFRONTAL

• Perseveration
• Obsessive-compulsive behavior
  – Get stuck on a behavior, and don’t switch to another one
BEHAVIOR MOTIVATION: MEDIAL FRONTAL

• **NUCLEUS ACCUMBENS**
  – Motivation
  – Why do I get up and go to work?
  – Why do I love coffee or chocolates?

• **DYSFUNCTION**
  – Apathy
  – No particular reaction to what is going on: differs from depression
BEHAVIOR MOTIVATION: MEDIAL FRONTAL

• CINGULATE CORTEX: INTERPERSONAL
  – Awareness of self and others
  – How do I respond to what you said or did?
  – How should I behave knowing that it will have an effect on your
  – TRUST GAME
Behavior

• 3rd Reason Why Behavior Changes
  – Cognitive Changes
  – Change our understanding of the world
    • Misinterpretations: does that shadow mean that my wife is having an affair?
    • Plausibility component of cognition doesn’t work
    • Normally say, “likely to just be a shadow”
COGNITION
COGNITION

• WHY?
1. EXECUTIVE FUNCTION
1. EXECUTIVE FUNCTION

- CEO of the brain
- Gets all info about what is going on, & makes decision about
  - What to focus on
    - Me: audience vs big toe hurting
    - You: lecture, coffee, bathroom, meet so-and-so for dinner tonight
1. EXECUTIVE FUNCTION

– Strategy
  • How will I accomplish that?
– Resource allocation
  • My entire focus?
  • One of many things I’m thinking about?
CAUDATE: ALSO EFFECTS EXECUTIVE FUNCTION

Figure W-2: The Caudate Relays Information To The Frontal Lobes

A) Get a haircut
Caudate - passes info to frontal lobe
I see that my hair is long

B) Damaged Caudate - no info passed on
I see that my hair is long
1. EXECUTIVE DYSFUNCTION

- How does this show up in the real world?
  - ABSTRACT REASONING
    - Why do you go to work for a delayed reward?
  - INSIGHT
    - Do I have any issues/problems/strengths or weaknesses?
      - No, my memory is perfect and I resent my wife saying that it isn’t!
1. EXECUTIVE DYSFUNCTION

• How does this show up in the real world?
  – INSIGHT
    • DYSFUNCTION
      – Person may become upset if he or she is not allowed to go back to work or live independently, because of the unawareness of change in capabilities
1. EXECUTIVE DYSFUNCTION

- How does this show up in the real world?
  - JUDGEMENT
    - Is it a good idea to spend my whole paycheck on dinner tonight?
    - Who should I put in my will? Relatives? Part time caregiver?
    - Should I be driving? Those 3 recent accidents were not my fault.
  - PRIORITIZING: which task do I do first?
1. EXECUTIVE DYSFUNCTION

• HOW DO WE DEAL WITH EXECUTIVE ISSUES?
  – No medications
  – EDUCATION
    • Driving
    • Finances
    • It’s ok to suggest that our loved one not drive, spend money, alter their will, etc
    • Testamentary capacity: will writing
1. EXECUTIVE DYSFUNCTION

• HOW DO WE DEAL WITH EXECUTIVE ISSUES?
  • Guardianship
  • Medical power of attorney

  – INSIGHT/AWARENESS
    • Unlikely to get mileage from saying, “You do have memory loss”
    • The person can’t remember that they forget
    • They may lack insight into their function
1. EXECUTIVE DYSFUNCTION

• **Organization**
  – One may have
    • Motivation to go to school/work may be there
    • Abstract reasoning may indicate that this is a good idea
    • But have problems “getting organized”: getting up, dressing, eating, getting school/work items together, getting to work, etc
  – No magic to treat this, but “being their organizer” can be very helpful
1. EXECUTIVE DYSFUNCTION

• GENERAL MEASURES
  – Maintaining a calm, predictable environment
  – Establish routines
    • Can improve organization and planning
    • Allows patient to organize daily tasks and adhere to that schedule, resulting in fewer organizational or planning problems
2. MEMORY LOSS
2. MEMORY LOSS

• 3 PHASES TO MEMORY FOR FACTS
  – Attention
  – Short-term memory
  – Long-term memory

• HD
  – Attention changes due to Frontal impairment
2. MEMORY LOSS

• DEALING WITH ATTENTION CHANGES
  – No distractions
  – Look at you
  – Repeat what was just said
  – Ask about it again later: repetition uses a different pathway
  – Taking notes

• MEDICATIONS: very tricky. Activation.
3. LANGUAGE
3. LANGUAGE

• SYLLABLES are in the frontal lobe
  – Comprehension is in the temporal lobe
  – Dysfunction
    • Slurred speech → Speech Therapy
    • Misnaming: “blat” for “bat”
GENERAL COGNITION ISSUES
GENERAL

• VARIABILITY: great deal of variability between people
  – Only some folks will be affected
• SEQUENCE: very variable
• INTENSITY: very variable
HPI

- 56 yo w/ h/o HLD presented for initial evaluation in our CDC
- ~10-15 year course of gradual, slowly progressive cognitive decline; no stepwise pattern or times of very acute change
- Pleasant wife of 29 yrs provided hx
HPI

• **1st sxs**
  - STM problems
  - Difficulties with executive function
    • Multi-tasking
  - Language
    • Trouble finding words & articulating his thoughts

• ~5 years ago, began to have gradually worsening
  - Fidgetiness
  - Trouble sitting still
HPI

- Social
  - No disinhibition or inappropriate behaviors
- Personality
  - No changes
  - No aggression, not quick to anger
- ADLs
  - No driving for ~4 yrs
  - When still driving, no problems with getting lost or disoriented
HPI

• Visuospatial
  – Even currently, there seems to be no significant visuospatial complaints and he's not lost around his house or familiar places

• Motor
  – No h/o parkinsonian features
    • No increased muscle tone, rigidity, bradykinesia, tremors, nor parkinsonian gait complaints
Psychosocial

• Worked for Boeing
  – Mechanical work x 15 yrs
• Worked in construction for some time also
• No TOB, No signif EtOH, No drug use
**HOW DO WE TEST? MSE**

**MMSE**

<table>
<thead>
<tr>
<th>Category</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Time</td>
<td>1/5 Spring</td>
</tr>
<tr>
<td>Place</td>
<td>2/5 TX, Clinic</td>
</tr>
<tr>
<td>Registration</td>
<td>3/3 Dog, Apple, Tree</td>
</tr>
<tr>
<td>Recall</td>
<td>0/3 One with prompting</td>
</tr>
<tr>
<td>Serial 7s/DLROW</td>
<td>0/5: Cannot initiate either</td>
</tr>
<tr>
<td>3 Step Command</td>
<td>2/3</td>
</tr>
<tr>
<td>Naming</td>
<td>2/2 pen, telephone</td>
</tr>
<tr>
<td>Repetition</td>
<td>0/1 &quot;No ifs, ands, ands,?“</td>
</tr>
<tr>
<td>CLOSE YOUR EYES</td>
<td>0/1: When reading, he spells out entire phrase, then reads aloud, but does not obey command</td>
</tr>
<tr>
<td>Sentence</td>
<td>0/1: &quot;I love M Wi..d“</td>
</tr>
<tr>
<td>Pentagons</td>
<td>0/1: Poor representation; planning</td>
</tr>
<tr>
<td>Total</td>
<td>10/30</td>
</tr>
</tbody>
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Previous W/U and Dx

- **W/U**
  - Wife states that he's had a very thorough work-up in Kansas in under the direction of private physicians, including 1 neurologist
  - MRIs x 2 (-): “Essentially normal brain MRI for age. No acute infarct, hemorrhage or neoplasm identified”
  - Heavy metals screen (-)
  - EEG (-)
  - Blood tests (-)
  - Huntington's reportedly (-)

- **Dx from outside**
  - Alzheimer's Dementia
Motor

- Strength 5/5 throughout
- No dystonic posturing
- Very mild, irregular, nonrhythmic, quick, unsustained involuntary movements of trunk, hands, arms, and legs
LESSONS

• HD occurs w/o recognized family history
• Presents with frontal changes in
  – Memory
  – Attention
  – Judgement
  – Language
• Delighted he did not present divorced
LESSONS

• EDUCATION
  – Ways to improve attention
  – Not be left alone
  – Living will
  – Will
  – Guardianship
  – Medical power of attorney
SUMMARY

• Frontal changes first in HD
• 3 frontal areas produce changes in 3 areas
  – Cognition, motivation, and behavior
• Remember
  – Very variable onset, sequence, intensity
  – Address cognitive issues before they are an issue: legal, driving, finances, divorce
SUMMARY

• ATTENTION: remove distractions, focus, repeat item, ask again later
• MEMORY: take notes, increase attention, repetition
• EXECUTIVE: organize the world for them, don’t confront unnecessarily, don’t be afraid to make judgment decisions for them
THANK YOU!

Paul Schulz MD