

Managing Cognitive Decline in HD

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Huntington's Disease Society of America

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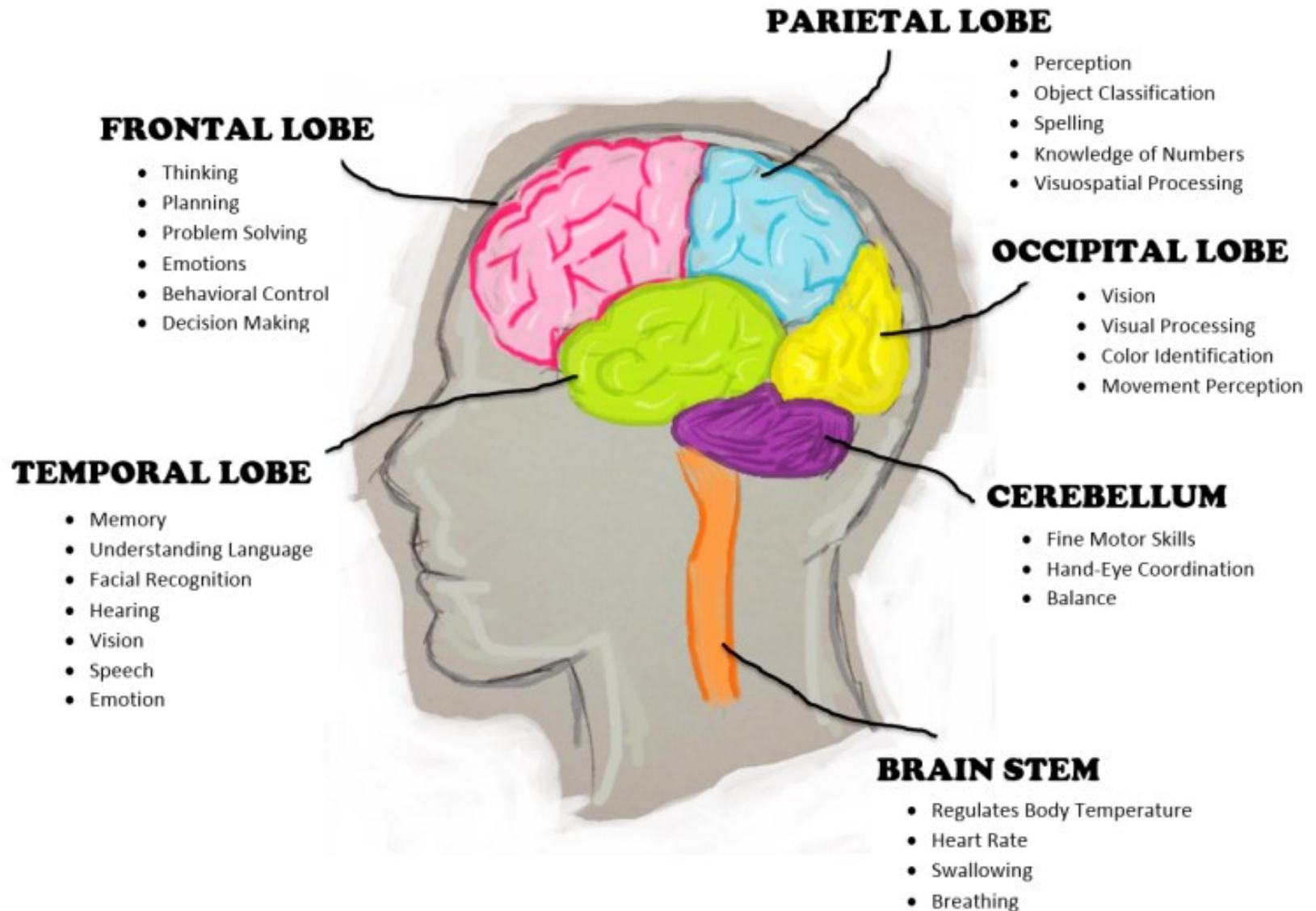
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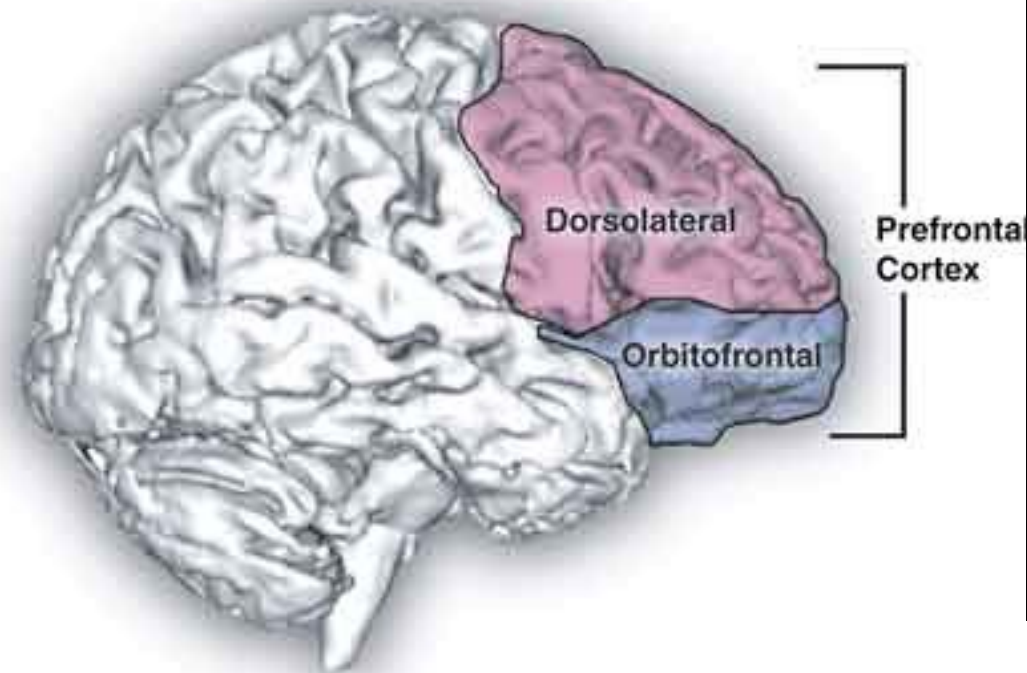
NEUROPSYCHIATRIC SYMPTOMS

- Affect
- Behavior
- Cognition
- The A, B, Cs of neuropsychiatry

EACH BRAIN AREA IS SPECIALIZED

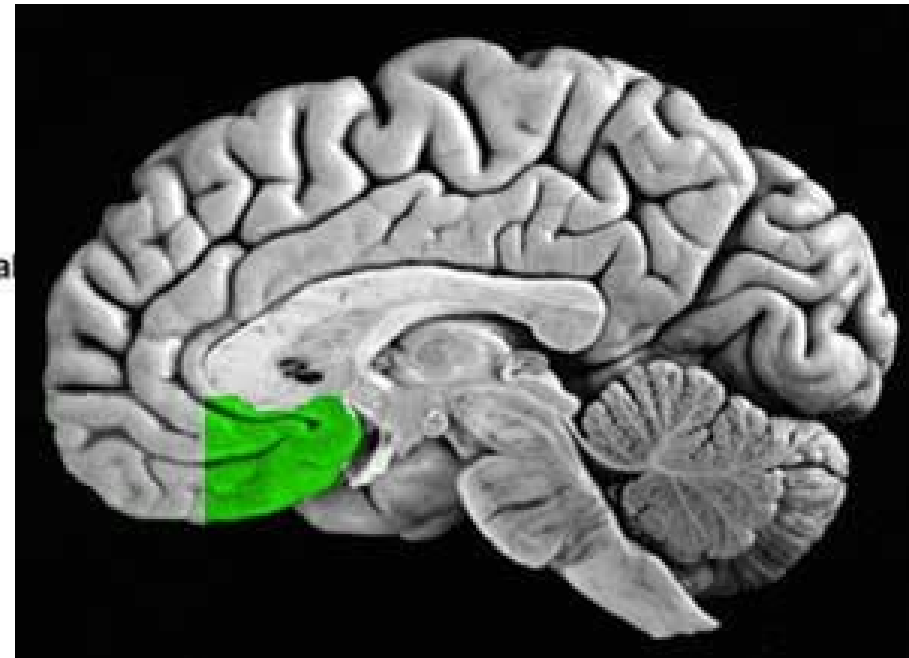


FRONTAL LOBE: 3 DIVISIONS



DORSOLATERAL

ORBITOFRONTAL



MEDIAL FRONTAL

AFFECT

AFFECT (OR MOOD)

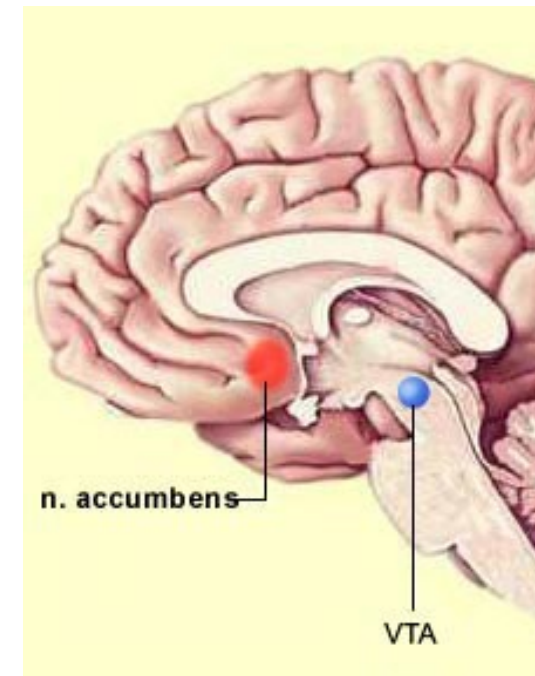
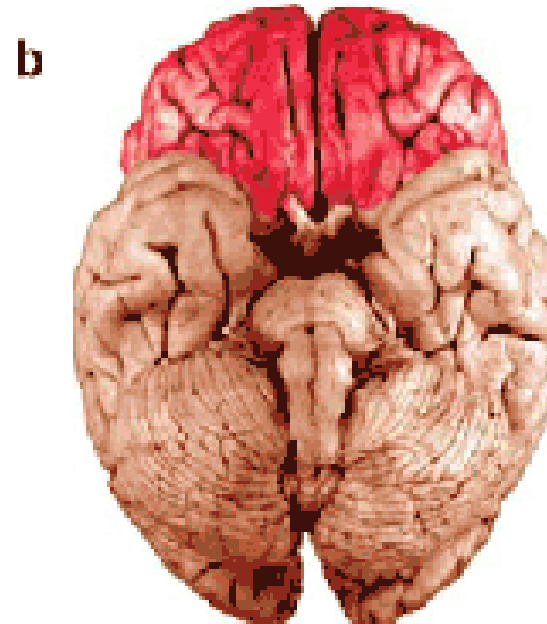
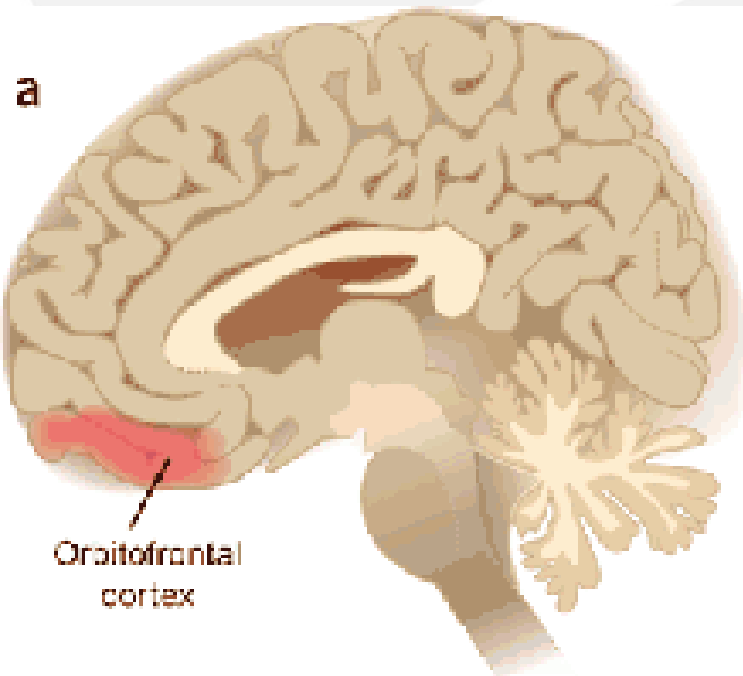
- 35-73% of HD patients
- Depression: most common psychiatric sx
- Mania: 2-12%
- Suicide: ~6%
- Anxiety

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?

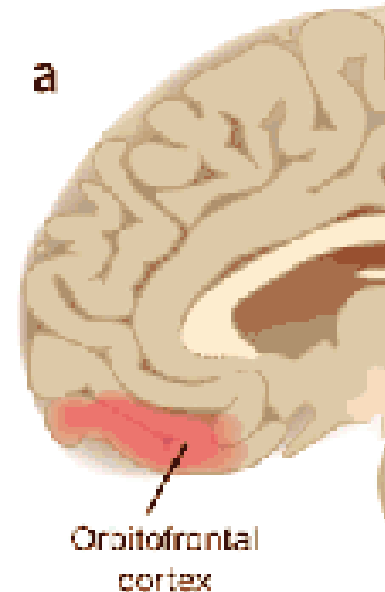


Change in REGULATION
of behavior

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

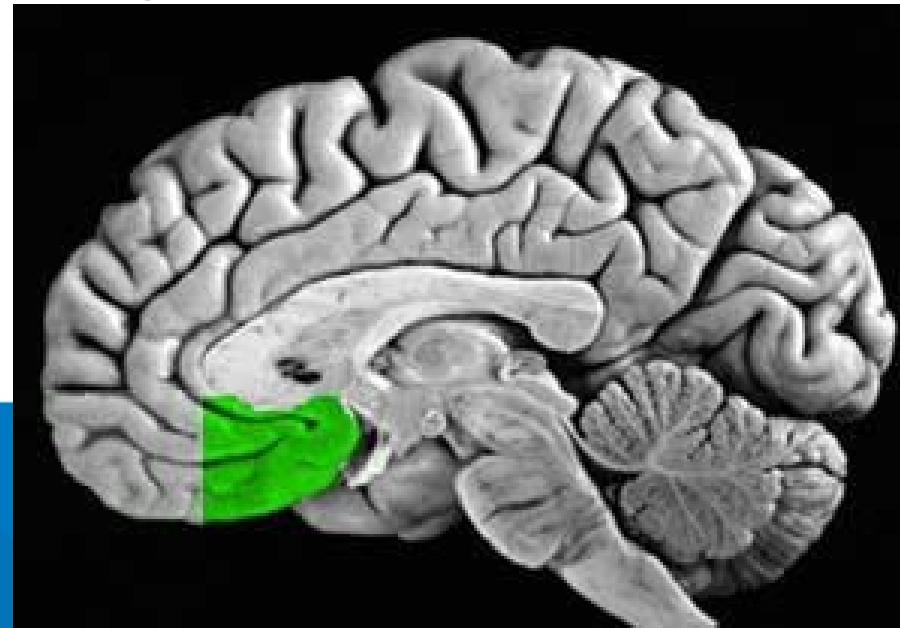
BEHAVIOR MOTIVATION: **MEDIAL FRONTAL**

- 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



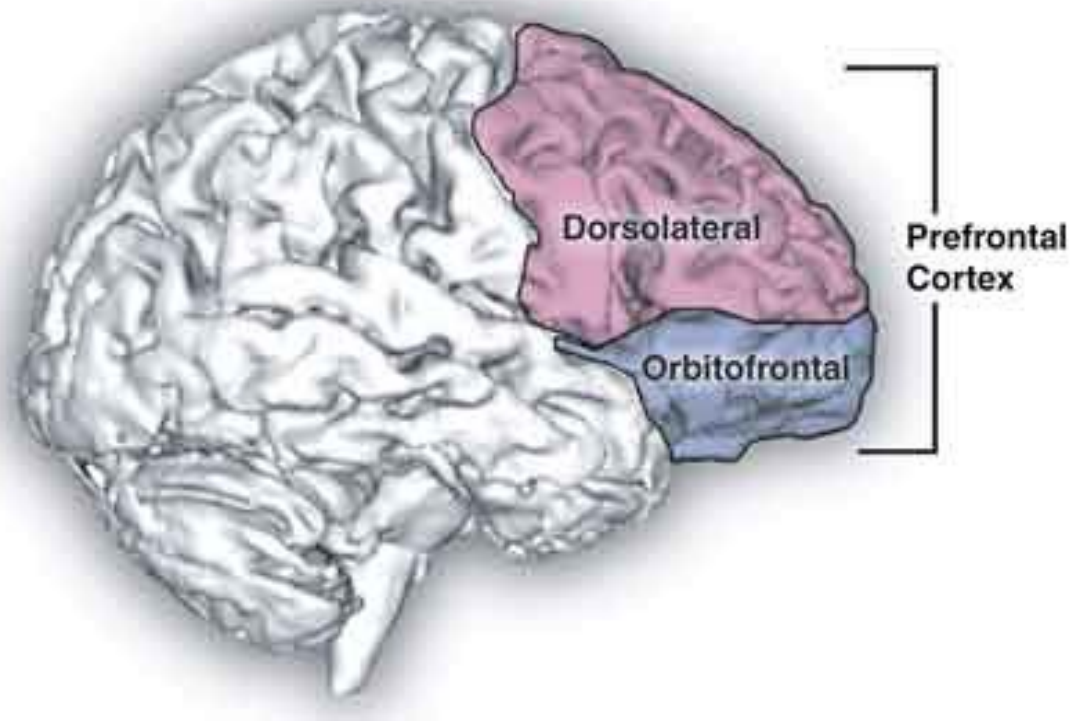
BEHAVIOR

- 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

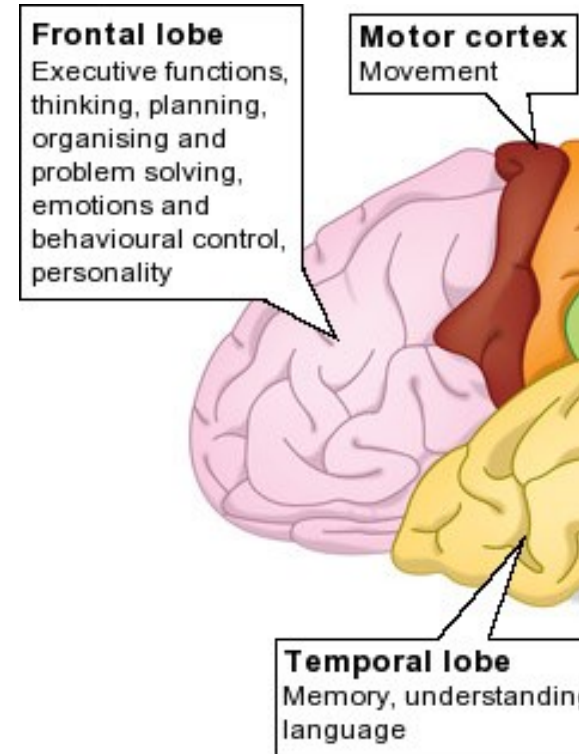
- WHY?



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

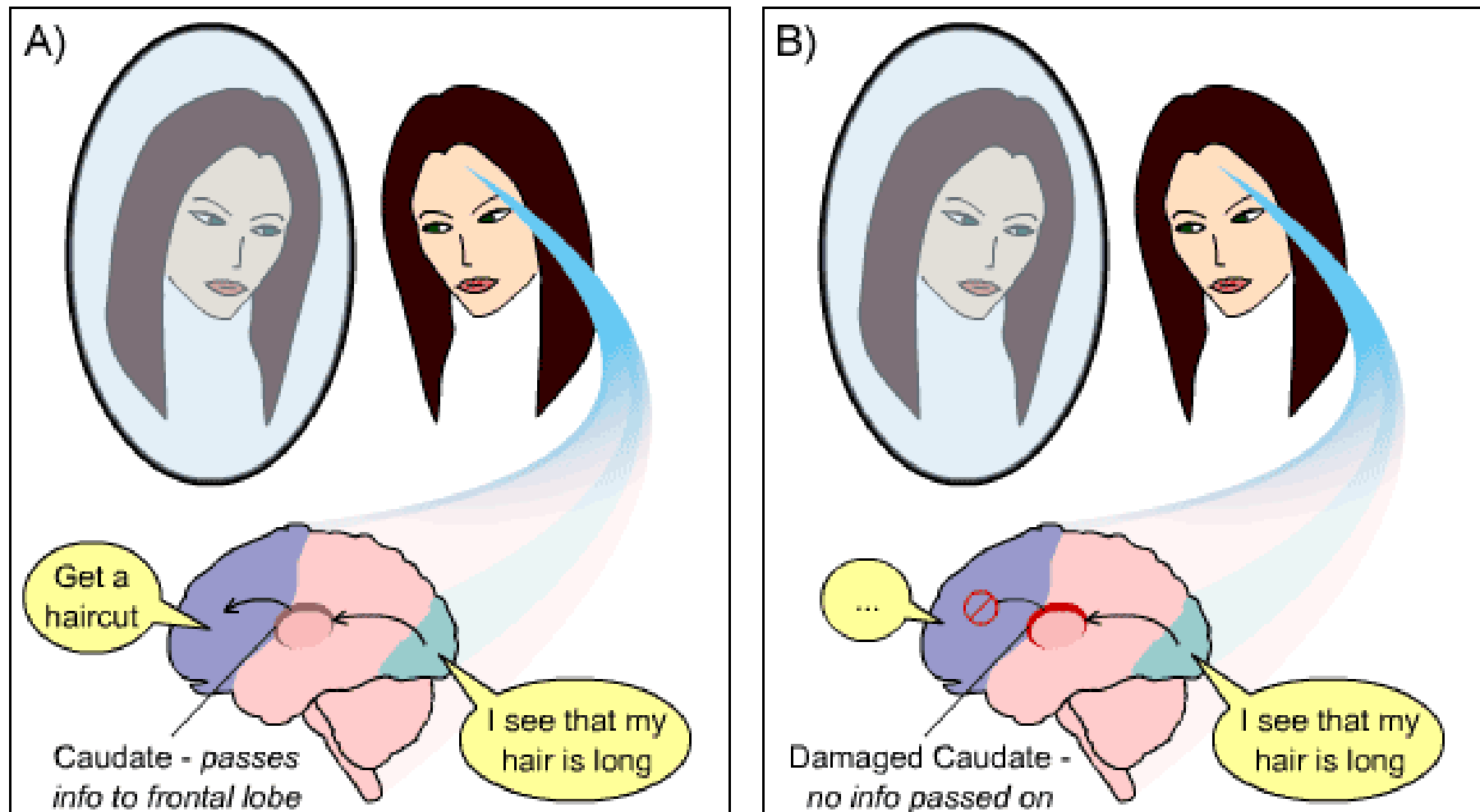


1. EXECUTIVE FUNCTION

- 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



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

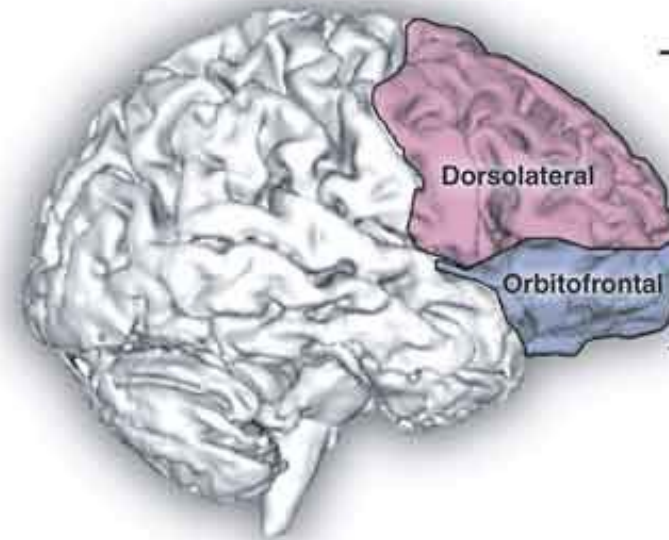
1. EXECUTIVE DYSFUNCTION

- 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

EXAMPLE

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

Time	1/5 Spring
Place	2/5 TX, Clinic
Registration	3/3 Dog, Apple, Tree
Recall	0/3 One with prompting
Serial 7s/DLROW	0/5: Cannot initiate either
3 Step Command	2/3
Naming	2/2 pen, telephone
Repetition	0/1 "No ifs, ands, ands,?"
CLOSE YOUR EYES	0/1: When reading, he spells out entire phrase, then reads aloud, but does not obey command
Sentence	0/1: "I love M Wi..d"
Pentagons	0/1: Poor representation; planning
Total	10/30

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

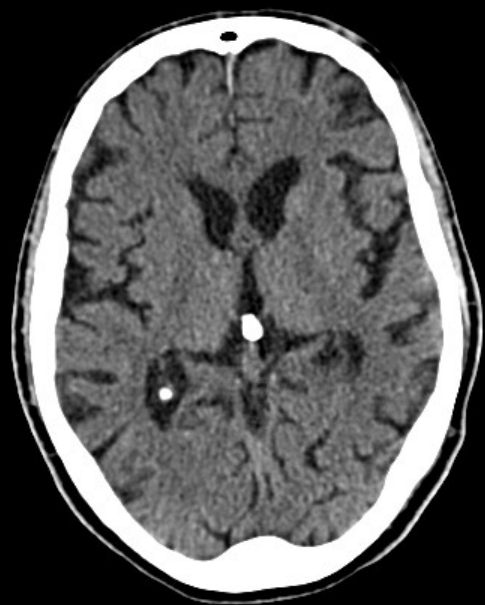
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LESSONS

- 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

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