Demystifying Diagnosis of Frontotemporal Dementia

Neuropsychological and Behavioral Neurologic Perspectives

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The Importance of Diagnosis

- Explanatory value
- Prognosis and knowing what to expect
- Treatment and care planning
- Possibility of being involved in research
- Better understanding of and care for individuals down the road (i.e., Scientific Progress)
What is FTD?

- Progressive disease
  - Can be sporadic or familial
- Caused by abnormal accumulation of proteins in the brain
  - Tau or TDP-43
- Leads to brain cell death
  - Which we see on imaging (CT/MRI) as “atrophy” or shrinkage
- Preferentially affects the frontal and/or temporal lobes
  - Most common first symptoms are changes in behavior or changes in language
Frontotemporal Lobar Degeneration

Clinical syndrome

Pathology

Bhogal, 2013
What is Neuropsychology?

- Psychology is the study of behavior
- Clinical Neuropsychology is the study of brain behavior relationships
  - Diagnosis and treatment of conditions affecting the brain
  - Emphasis on cognitive and behavioral testing
  - Investigation of strengths and weaknesses
What is Behavioral Neurology?

- Subspecialty of neurology for the evaluation and treatment of brain disorders that affect cognition (how we think) and behavior (how we act)
- Behavioral neurologists see people who come with changes in their cognition or behavior
- We work with neuropsychologists, radiologists and others to help identify what specifically has changed and by how much, and to identify the underlying cause of these changes
Tests and Real Word Functioning

Wisconsin Card Sorting Test ≠日常的实际情况
Neurocognitive Domains

**Perceptual-motor function**
- Visual perception
- Visuoconstructional reasoning
- Perceptual-motor coordination

**Language**
- Object naming
- Word finding
- Fluency
- Grammar and syntax
- Receptive language

**Executive function**
- Planning
- Decision-making
- Working memory
- Responding to feedback
- Inhibition
- Flexibility

**Learning and memory**
- Free recall
- Cued recall
- Recognition memory
- Semantic and autobiographical long-term memory
- Implicit learning

**Complex attention**
- Sustained attention
- Divided attention
- Selective attention
- Processing attention speed

**Social cognition**
- Recognition of emotions
- Theory of mind
- Insight
Language

• Comprehension
  – Deficits:
    • Difficulty understanding language
    • Nonverbal behavior “baked in” can mask difficulties
  – Tests:
    • Following commands
    • Answering yes/no questions
    • Pointing based on verbal prompts
    • Picture/picture matching, picture/word matching
Language

• Naming
  – Deficits:
    • “Tip of the tongue” phenomenon
    • Problems finding right word
    • Using a word that is related in meaning or sound to the desired word (“paraphasia”)
    • Talking around a word (“circumlocution”)
    • Decreased semantic knowledge
  – Tests:
    • Visual object naming
    • Naming to definition
    • Test of object knowledge
Semantic dementia

Involves the dominant (usually left) anterior temporal lobe

https://ftd.med.upenn.edu/about-ftd-related-disorders/what-are-these-conditions/progressive-language/semantic-variant-primary-progressive-aphasia-svpap
Semantic Dementia:
Progressive loss of the meanings of things

- Difficulty finding the right word
  - Substitutions of words within categories, e.g. cow for horse
- Loss of knowledge of what words mean or what objects are for.
  - “What is a can opener?” and not knowing what its use is
- Difficulty understanding what other people are saying
  - “What is a mafia?”
- Problems with reading and writing
  - E.g. Surface dyslexia/dysgraphia (YACHT)
- May develop changes in behavior
Language

• Fluency
  – Deficits:
    • Difficulty “getting words out”
    • Simplified sentence structure
    • Loss of grammar
    • Problems with making speech sounds
  – Tests:
    • Picture description
    • Automatic speech
    • Verbal fluency procedures
Progressive non-fluent aphasia

Left perisylvian atrophy (arrow) in progressive non-fluent aphasia

Progressive non-fluent aphasia

- Effortful and halting speech
- Sound substitutions
  - “bebble” for “pebble”
- Sentences lack proper grammar
  - May not use articles (“the”), adjectives or prepositions
- May have trouble understanding complex sentences
  - “I burned dinner but not the cake”
- May have trouble in producing sounds
  - “Apraxia of speech”
- May develop problems similar to Parkinson’s disease
  - Tremor, body rigidity, falls
Language

• Repetition
  – Deficits:
    • Difficulty repeating words/sentences
  – Tests:
    • Word repetition
    • Sentence Repetition
Logopenic Aphasia

FDG-PET showing decreased metabolism of superior temporal and inferior parietal lobes, and lateral thalamus
Logopenic Aphasia

• Poor picture naming
  – But good single-word comprehension and semantic abilities
• Impaired repetition of sentences
  – But not of single words
• Slow speech
  – Frequent word-finding pauses
• Simple but correct grammar
• Often the underlying diagnosis (by pathology) is Alzheimer’s disease
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Social Cognition

- Social Cognition
  - Deficits:
    - Lack of sympathy/empathy
    - Difficulty responding appropriately to others
    - Diminished insight
  - Tests:
    - Clinical interview and clinical history
    - Emotion recognition
    - Mental state attribution
Behavioral-Variant Frontotemporal Dementia

- **Age 43**: normal behavior and cognition
- **Age 45**: mild behavioral changes
- **Age 47**: behavioral variant frontotemporal dementia

Images show subtle, mild, and moderate bifrontal atrophy.
International consensus criteria for behavioral variant FTD (Raskovsky et al 2011):

Three of the following behavioral/cognitive symptoms (A–F) must be present to meet criteria (and are an insidious change from previous behavior).

A. Early behavioral disinhibition [one of the following symptoms (A.1–A.3) must be present]:
   A.1. Socially inappropriate behavior (e.g. showing mastectomy scars to shop assistant)
   A.2. Loss of manners or decorum (e.g. accompanying someone else’s family to crypt at funeral)
   A.3. Impulsive, rash or careless actions (e.g buying valueless property with life savings)

B. Early apathy or inertia (not engaged in previous hobbies/activities)

C. Early loss of sympathy or empathy (do not “seem to care”; continue to do hurtful/inconsiderate things)

D. Early perseverative, stereotyped or compulsive/ritualistic behavior
   D.1. Simple repetitive movements (e.g. winding string)
   D.2. Complex, compulsive or ritualistic behaviors (e.g. collecting cereal boxes)
   D.3. Stereotypy of speech (catchphrases)

E. Hyperorality and dietary changes [one of the following symptoms (E.1–E.3) must be present]
   E.1. Altered food preferences (fads, sweets)
   E.2. Binge eating, increased consumption of alcohol or cigarettes
   E.3. Oral exploration or consumption of inedible objects

F. Neuropsychological profile: executive/generation deficits with relative sparing of memory and visuospatial functions
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Executive Functioning

- **Cognitive Flexibility**
  - **Deficits:**
    - Getting “stuck”
    - Behavioral rigidity
    - Stereotyped behavior
    - “Obsessive compulsive” behavior
  - **Tests:**
    - Number-Letter Switching
    - Card Sorting
    - Ideational Fluency
Executive Functioning

- **Inhibition**
  - **Deficits:**
    - Inappropriate behavior
    - Emergence of drives
    - Utilization behavior
    - Emotional reactivity
  - **Tests:**
    - Color-Word Reading (i.e., Stroop Test)
    - Go-No Go Tests
    - Flanker Tests
Executive Functioning

• Activation
  – Deficits:
    • Difficulty initiating behavior
    • Environmental Dependence
    • Lack of drive/interest (i.e., apathy)
  – Tests:
    • Word generation
    • Verb fluency
    • Categorization/Classification Procedures
Executive Dysfunction: can be seen in any subtype of FTD/CBD/PSP

- Set-shifting
  - Decreased ability to shift from one activity to the other (e.g., chopping vegetables while oil heating)

- Problem Solving Trouble
  - Using a sharp knife to clean pans

- Apathy (may get confused with depression)
  - Seem OK just to sit and not do anything

- Rigidity
  - Walking the same route the same number of times a day

- Disorganization/poor planning
  - Leads to inability to get things done properly
Improving Functioning

• **YOU** are the most important intervention!
  – Be aware of strengths and weaknesses
  – Provide assistance and help
  – Structure the environment
  – Suggest the word the person is looking for
  – Eliminate tasks that are difficult frustrating
  – Use strengths to overcome weaknesses
  – Advocate for your loved one for the best:
    • Diagnosis
    • Treatment
    • Support services
“That’s all Folks!”