

Approaches to Behaviors in FTD and PPA



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FRONTOTEMPORAL DEMENTIA AND PRIMARY PROGRESSIVE APHASIA
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Definition of Dementia

A gradual decline in thinking abilities and/or personality compared to former self

More decline than in normal aging

Interferes with daily routines

Caused by a brain disorder

Symptoms of Dementia

- Short Term Memory loss
 - Forgetfulness, getting lost, repetitive questions
- Aphasia
 - Loss of language function
- Personality changes
 - Exaggeration of old traits or change in behavior
- Visual changes
 - Inability to recognize common objects despite normal vision; spatial disorientation

Dementia



Frontotemporal Degeneration



Demographics

- FTD affects an estimated 250,000 Americans
- The prevalence of FTD among people ages 45 to 64 was estimated to be 6.7 per 100,000
- The disease affects both sexes equally
- About 40% of patients have a clear-cut family history
- The mean duration of the illness is about eight years

Findings in FTD/PPA

- Atrophy or shrinkage of frontal/temporal lobes of brain from loss of nerve cells
 - Left sided – language problems (PPA)
 - Right sided or both sides – behavioral changes
 - In both syndromes, atrophic changes often progress to both sides
- Diminished brain activity in these same areas on brain scans
- Decreased serotonin binding in frontotemporal areas
 - Carbohydrate craving, compulsive behaviors, disinhibition, aggression

Cognitive Symptoms in FTD

- **Difficulty sustaining and redirecting attention**
 - Can't watch movies, read books, converse at length
 - Appear purposeless and bored (apathetic)
- **Difficulty making and carrying out plans**
 - Trouble paying bills, cooking, shopping, grooming
 - Become increasingly dependent on caregiver
 - Work difficulties from problems with planning, organization, feedback correction, task completion, and mental flexibility
- **Difficulty in reasoning**
 - Mental rigidity, insist on having things a certain way

Early Behavioral Symptoms in FTD

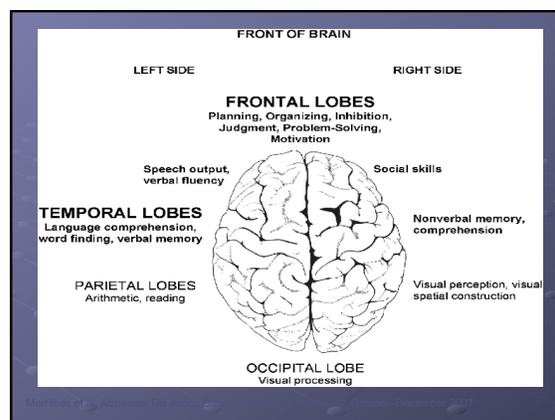
- Early decline in social/interpersonal conduct suggesting a loss of self-control
- Loss of insight or denial of symptoms
- Decline in tactfulness or manners
- Violation of interpersonal space
- Loss of interest in personal hygiene
- Reduction in verbal output
- Emotional blunting
- Pathological gambling
- Craving for sweets, weight gain

More Behavioral Symptoms in FTD

- **Loss of empathy, sympathy and introspection**
 - Quiet, flat, distant
- **Appear indifferent at times**
 - Indecisive, disinterested
 - Less spontaneous behavior
- **Poor judgment**
- **Compulsive-like behaviors or simple repetitive acts**
 - verbal or motor stereotypic behavior such as lip smacking, hand rubbing or clapping, counting aloud, and humming.

More Behavioral Symptoms in FTD

- **Delusions (fixed false beliefs)**
 - Jealousy, religious, unusual thoughts
- **Driven to touch or use items in their view**
 - Pick up a pen and use it, take food when not hungry
- **Complex repetitive motor routines**
 - Wandering a fixed route, collecting and hoarding objects, counting money, checking and rituals involving unusual toileting and hygiene behavior



Syndromes in FTD

Apathetic

- Seem indifferent
- Lack of motivation
- Less talkative
- Blunting of emotions
- Misinterpreted as depression by others

Disinhibited

- Easily irritated or angered
- Excessive jocularity; can mimic mania
- Poor judgment; eg, excessive spending or shoplifting
- Sexual impropriety
- Aggression

*These syndromes may also co-occur in any given patient

Symptoms of PPA

- Insidious onset with gradual progression
- Often see atrophy in left side of brain predominantly
- Primarily language difficulties for the first two years including:
 - Word finding
 - Naming of objects
 - Understanding of speech
- As PPA progresses, the cognitive and behavioral symptoms can resemble those of patients with FTD.

Language Symptoms and PPA

- Early and progressive change in language function for the first two years
- Relative preservation of other cognitive domains, such as memory
- Characterized by problems in the expression of language
 - Using the correct word or name
 - Difficulties with grammar or sentence construction
 - Difficulties reading and writing
 - Understanding of word meaning is relatively well preserved
- As the illness worsens, less and less language is used, until the patient may be virtually mute

Comparison of FTD/PPA and AD

Symptoms	FTD/PPA	Alzheimer's
Memory	Preservation based on ADL's	Impaired early in illness
Age of onset	Before age 65	Older than 65
Mood	Jocular, irritable or apathetic	Depression is common
Behavior	Early changes in behavior	Behavioral changes later

FTD/PPA leads to problems with decision-making, behavioral control, emotion processing, social cognition and language

Associated Behavioral Symptoms

- Apathy or inertia
- Disinhibition or impulsivity
- Repetitive or compulsive behaviors
- Delusional thinking
- Hyperorality

Communication in PPA/FTD

- Engage with eye contact and a gentle touch
- Communication in a quiet, calm environment with an upbeat voice to facilitate engagement
- Speak slowly, use gestures and visual cues, particularly when language is impaired
- Speech therapy can be helpful in early stage

Facilitating Cooperation

- Offer positive choices eg “Let’s go out now” or “Do you want to wear your blue or red cap?”
- Use positive reinforcement such as food, smiles, a gentle touch, personal attention and lots of praise.
- Do not correct the patient’s grammar or pronunciation if their essential message is clear

Augmentative Communication

- Communication notebook
Has essential words and pictures organized by topic for easy access



- Drawing pictures for communication
- Use of gestures or physical cues
- Computerized communication devices may be helpful for some early on

Environmental Modifications in Addressing Behavioral Symptoms

Maintaining Perspective

- Understanding the nature of the disease and the fact that the neuropsychiatric and behavioral features are not ‘deliberate’ behaviors. Do not take it personally.
- If a caregiver becomes frustrated and angry, it is best to find someone else to handle the problem and have the caregiver leave the immediate area or take a break (respite). An angry caregiver will only intensify problem behaviors.

A Safe Predictable Environment

- Keep the routine the same. Changes in routine are upsetting to people and can cause behavior problems.
- Promote a sense of security and comfort when problem behaviors occur. Behaviors often happen because a person is frightened and unable to make sense out of the environment.
- Trying to argue or reason only results in frustration for both the caregiver and the individual.

- Participate in social situations but discuss and plan ahead of time and prompt the patient during the event. Have others around to help.
- Avoid overstimulating, confusing environments
- Distract and divert whenever possible.
- Try to maintain a sense of humor even in the most difficult situations.

Appropriate Stimulation and Structure

- Attend to the patient's daily quality of life: mood, social connectedness, ability to communicate, physical activity and nutritional status.
- Provide the right environment
 - Predictable routine in safe milieu
 - Right amount of stimulation, exercise, rest
- Supervised activities
 - Pleasant, structured, simple
 - Consider adult day programs, companions

Adapt to Difficult Behaviors

- Redirect, be flexible, soothe, compensate, reassure, distract, reapproach, plan and **DO NOT ARGUE**
- Restructure the environment
 - Hyperorality may require a lock on the refrigerator or hiding food
 - Roaming may require locked doors and a full time caregiver
 - Compulsions may require redirection and distraction

Apathy and its Consequences

- Physical deconditioning and weakness
- Failure of rehabilitation
- Inability to participate in basic activities of daily living such as self care and eating
- Uncooperativeness with care and combativeness when caregiver attempts to motivate patient
- Social isolation

Why seek psychiatric consultation?

- Mood symptoms such as depression, irritability, poor frustration tolerance or anger
- Changes in behavior which are highly disruptive or dangerous
- Problems with sleep or anxiety

Depression in PPA (Medina 2007)

- 34% of PPA patients were clinically depressed
- Even the clinically non-depressed subjects reported more symptoms of depression than control subjects
- The # of depressive symptoms associated with decline in the ability to name items
- Social withdrawal and lack of mental and physical energy were common symptoms in depressed patients

- Past history of depression may be a risk factor
- Depression characterized by symptoms of social withdrawal and lack of mental and physical energy
- Depression in PPA may be a psychological reaction fueled by an awareness of one's breakdown in language abilities

General Approaches to using Medications in FTD/PPA

- Several small clinical trials, no large scale clinical trials
- No medication specifically approved by FDA for the treatment of FTD or PPA
- Currently, medication treats the symptoms, not the disease
- Identify behavioral syndromes for symptomatic treatment
 - Apathetic vs Disinhibited
- Make sure there are no contributing medical problems such as a urinary tract infection or pain
- Choose one medication at a time, start slowly and assess effects at increasing doses over time

Psychiatric Medications in FTD/PPA

- **Antidepressants** (SSRI's, SNRI's and more)
 - Eg. sertraline (Zoloft), escitalopram (Lexapro), mirtazapine (Remeron), venlafaxine (Effexor XR), duloxetine (Cymbalta), bupropion (Wellbutrin)
- **Antipsychotics**
 - Eg. quetiapine (Seroquel), olanzapine (Zyprexa), aripiprazole (Abilify), risperidone (Risperdal)
- **Mood Stabilizers**
 - Eg. oxcarbazepine (Trileptal), valproic acid (Depakote)
- **Sleeping Medication**
 - Eg. trazodone (Desyre)

Cognitive Enhancers

- **Rivastigmine** treatment was associated with less behavioral impairment and less caregiver burden, though it did not improve cognition. (Moretti, 2004)
- In a small study, **donepezil** and **rivastigmine** may be effective in treating cognitive impairments in FTD, particularly in afflicted men (Lamp, 2004)
- **Memantine** is not thought to be helpful in this syndrome

Antipsychotic Medication

- One case study showed **aripiprazole** improved cognition and negative symptoms in FTD patients (Fellgiebel 2007)
- Another case study showed improvement in delusions and hallucinations with **risperidone** (Curtis 2000)
- **Quetiapine** reduced agitation in 3 patients (Liu 2004)
- In a 24-month follow-up study in 17 pts, **olanzapine** significantly improved scores on all of the behavioral and mood scales although cognition appeared to worsen (Moretti 2003)

- Methylphenidate in 8 FTD patients did not improve memory and executive function, or mood but did decrease risk-taking behavior (Rahman 2006)
- Carbamazepine, valproate, or lamotrigine may diminish long-term emotional fluctuations
- Trazodone is well-tolerated and can significantly improve behavioral disturbances as measured by the NPI but does not improve cognition (Lebert 2004) It can also be helpful for sleep when given in larger doses at night

FTD/PPA Symptoms	Medications Used (eg.)
Difficulty with attention, motivation, apathy	Stimulants Methylphenidate, amphetamines Stimulating antidepressant Bupropion
Irritability, depression, anxiety, craving for sweets, compulsions	Serotonergic antidepressants Sertraline, escitalopram, citalopram, venlafaxine, trazodone, mirtazapine

FTD/PPA Symptoms	Medications Used (eg.)
Disinhibition	Anticonvulsants
Aggression	Valproic acid, oxcarbazepine
Agitation	Antipsychotic medications
Psychotic symptoms	Quetiapine, olanzapine, aripiprazole, risperidone
Insomnia or sundowning	Sedating antidepressants
	Trazodone, mirtazapine
	Antipsychotics

- ### Psychiatric Hospitalization
- For safety reasons
 - To prevent injury to the patient or others
 - For a medical evaluation
 - Rule out infection, metabolic abnormalities, delirium
 - To re-evaluate psychotropic medication
 - In a controlled environment, medications can be stopped and then gradually reintroduced one by one in order to determine therapeutic effect
 - Respite for the caregiver
 - Transition to nursing home placement if necessary

- ### Caregiver stress in FTD
- Patient's loss of empathy dramatically affects those who care for patients
 - Feel they have 'lost' their family member and are in mourning at the same time that they are having to deal with behaviors
 - Loss of reassuring facial signals we expect in interaction
 - Others may misunderstand patient's behavior
 - Frustration for patient who is not understood by others and does not understand what is wrong

- ### The Burden of Caregiving
- Caregiver perception of burden is strongly associated with caregiver distress
 - Caregiver distress is associated with earlier nursing home placement
 - Social support decreases caregiver stress and supports healthy immune system functioning
 - Early caregiver support and education may delay nursing home placement and improve quality of life for the patient and the caregiver