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Cognitive and Behavioral Challenges in Caring for Patients with Frontotemporal dementia and Amyotrophic Lateral Sclerosis

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Abstract

Frontotemporal dementia (FTD) is a progressive neurological condition caused by degeneration of the frontal and/or anterior temporal lobes resulting in personality, behavioral, and cognitive changes. Amyotrophic lateral sclerosis (ALS) is caused by degeneration of lower motor and pyramidal neurons, leading to loss of voluntary muscle movement. The common molecular pathological and anatomical overlap between FTD and ALS, suggest that the two disorders are strongly linked. In some patients FTD precedes ALS, in others ALS occurs first, while in still others the two disorders begin simultaneously. The association between ALS and FTD create unique challenges for family caregivers. This paper provides a guide for healthcare providers caring for patients with FTD-ALS exhibiting behavioral, cognitive, and emotional symptoms. Strategies are suggested to help minimize the impact of negative symptoms.

Keywords

frontotemporal dementia; amyotrophic lateral sclerosis; behavior management; behavioral symptoms

Introduction

FTD is the result of neuronal degeneration in the frontal and/or anterior temporal lobes of the brain. FTD has several clinical subtypes identified by whether the predominant syndromes are behavioral or language-related. The behavioral variant is typically referred to as FTD or bvFTD. Patients with bvFTD exhibit symptoms caused by diminished function of the frontal and anterior temporal lobes of the brain, areas that govern personality, judgment, the ability to plan and multitask, drive, motivation, eating and social regulation. Patients with bvFTD demonstrate a decline in social conduct, apathy, emotional blunting, and loss of insight that is insidious, gradual, and progressive (1,2). The presence of behavioral symptoms is strongly associated with right hemisphere atrophy (2-4).

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ALS is caused by degeneration of lower motor and pyramidal neurons, cells that control voluntary muscle movement. FTD and ALS commonly present together and share a common molecular pathology (5,6). More than half of patients with ALS exhibit some behavioral or cognitive symptoms (7,8), yet not all will meet diagnostic criteria for FTD. In addition, variability exists on the type and severity of behavioral symptoms when they are present (9). Recent consensus criteria provide a framework for diagnosis of FTD syndromes in ALS (10).

The co-morbid diagnoses of FTLD-ALS may be associated with an adverse effect on survival. Median survival from symptom onset was 3 years 3 months for 53 patients with ALS only and 2 years 4 months for those with FTLD-ALS (11). This may be due to biological factors and/or cognitive changes affecting adherence to treatment.

The purpose of this paper is to describe, a variety of clinical features and challenging situations that may occur. Recent practice parameters focused on the cognitive and behavioral features of ALS point to the lack of controlled studies on effective management of these symptoms (12). By incorporating clinical examples, we review salient issues involved in care planning along with strategies aimed at minimizing the negative impact of behavioral, emotional, and cognitive symptoms.

General Principles for Families

Each patient displays a unique set of symptoms in the context of their own family system. Certain behaviors are relatively uncommon in ALS with FTD. For example aggression and obsessive-compulsive habits were not common symptoms in one study (9) but in clinical experience, when these behaviors occur, they are very challenging for families to manage.

There are five types of strategies used with patients and families in managing negative behaviors: environmental, behavioral, pharmacological, physical, and those internal to the family caregiver. Environmental strategies focus on modifications to the person's environment. Examples include limiting access to bank accounts or altering aspects of the patient's environment such as access to cars, dangerous tools or even food. Behavioral strategies focus on changing the person's behavior and can include actions such as rewarding positive behaviors. Pharmacological strategies involve medications. The selective serotonin reuptake inhibitors (SSRIs) and related compounds remain the mainstay of treatment for treating bvFTD (13,14), however, there are limitations to pharmacologic management and not all behaviors can be medicated. Physical strategies include any action that blocks the person's movement or access, e.g. restraint vests or belts. This category is rarely employed, as the use of restraint devices is usually associated with adverse consequences (15).

A fifth type of intervention is internal to the caregiver and involves aspects of psychological coping and acceptance for the symptoms. It entails assisting the caregiver to change their reaction and response to the negative behavior(s). Some families struggle with providing exemplary care, yet despite their hard work and deep concern, the disease progresses and the patient worsens. Although it can be difficult for family caregivers to ask for assistance or feel comfortable taking time away from caregiving, these are appropriate strategies for managing the hardships of patient care and behavioral/cognitive symptoms.

Table 1 provides a few examples of possible interventions for negative behaviors.

Cognitive and Behavioral Symptoms

The following is a description of behavioral and cognitive changes that can be evident in patients with FTD-ALS. FTD-ALS is a heterogeneous disease; some patients may exhibit

only mild symptoms while others may manifest with profound changes. The symptoms reviewed here are not presented in a particular order.

Executive deficits

Planning, organizing information, problem-solving, shifting attention, inhibiting behavior, and negotiating social mores are all examples of executive functioning. There is evidence that executive deficits occur commonly in ALS (16). Specific types of executive functioning deficits have been documented using neuropsychological tests, yet the day-to-day manifestations of these deficits are the primary concern of patient's families. Examples of executive dysfunction include an inability to manage family finances (e.g. balancing checkbook, paying the bills). Other examples might include difficulty in preparing complex meals, caring for children, or planning and packing for a trip.

Interventions when executive deficits are present—Families may need to adjust previously held roles for financial activities and health care planning. The patient may not be able to hold a position as the primary decision-maker in the family, but may still participate in less demanding ways. Potentially dangerous activities such as driving and eating may require direct supervision. Modification of the environment may be necessary, e.g. selling extra vehicles or removing unsafe food from patient's access. The literature in cognitive rehabilitation for brain injury patients demonstrates that the use of self-administered checklists and individualized task analyses as prompts, helped patients avoid common hazards at home (e.g safely putting away a space heater) and to continue independent activities (e.g. cooking) (17).

Apathy

Dysfunction in the frontal lobes causes profound changes in how a person shows interest and enthusiasm and is a common symptom in both FTD and ALS (7). Their activity and movement diminish markedly and they may become content with sedentary activity. These changes become evident even in patients who were formerly assertive and highly productive. It can be difficult to distinguish between apathy, depression, and fatigue as all have common behavioral outputs (8). Families often go to great lengths trying to find an activity that will engage the patient's interest and it can be frustrating for families when these attempts are not successful.

Interventions when apathy is present—A thorough assessment for depression, focusing on associated features such as sadness, tearfulness, hopelessness, and suicidal ideation is important. Differences between apathy and fatigue that is secondary to respiratory dysfunction should be carefully evaluated. Standardized caregiver interviews and patient assessments are critical in pinpointing whether the decline in activity and interest is due to apathy. Shifting expectations about how engaged and interested the person can be is an important step in dealing with apathy. Participating in activities that are passive, such as rides in the car, can still be pleasurable.

Irritability

Irritability is one of the more common symptoms reported in patients with ALS and bvFTD. Irritability manifests as mood swings and bursts of anger, often over trivial details. Patients who were formerly even-tempered may exhibit irritable changes as a result of the illness.

Strategies when irritability is present—Try to identify the triggers to the patient's irritability. Observe for patterns of behavior and note whether there are cycles of restlessness and fatigue. Some patients become excessively tired by the end of the day and react with

anger when demands upon them become overwhelming. Instituting a predictable and structured schedule and a calm environment is helpful. Eliminating environmental stimulation, e.g. loud noise, questions and requests, may be necessary. For some patients, being in the presence of others is calming, and for others, time alone in a reclining chair is more settling. The impact of stimuli such as television and music can vary from person to person. Importantly, the spouse caregiver is often the "target" of the patient's irritability and may need breaks and respite from caregiving responsibilities.

Poor judgment and impulsivity

Patients with bvFTD invariably demonstrate compromised decision-making abilities and lose their ability to assess the risks of an action. Reduced judgment may manifest in patients who refuse to use equipment recommended by clinical providers. They may refuse breathing treatments despite the merits of using a nebulizer for chest congestion or BiPAP at night to enhance respiratory status. The patient may not be able to demonstrate clear reasoning behind their decisions. Poor judgment makes the patient vulnerable to sales pitches and impulse buying, and in some cases, leads to shoplifting.

Interventions when poor judgment and impulsivity are present—Gain acceptance for your recommendations: "Will you try this for a week?" Use positive/encouraging suggestions and "ask the individual to choose between two selected options: "Would you like to use the walker or lean on my arm?" In some situations, the caregiver may need to direct the person what to do rather than provide choices and options. Use of motion alarms if the patient gets out of bed without waiting for assistance may be helpful and monitoring activity with the use of audible monitors may enhance safety.

Postal delivery can be diverted to a post office box rather than the home so that mail can be better supervised. Keep car keys safely away from the patient and park the car out of sight in a nearby garage. Anticipate the driving needs of a patient and plan outings prior to a predicted time the patient would want to go out, or arrange for friends to stop by for an outing. Steps should be taken to ensure adequate financial protection for a vulnerable patient. The traumatic brain injury literature demonstrates that individuals with limited judgment are more successful with pro-social, safe behavior when they are given choices. When individuals were permitted to choose from a list of tasks, they valued appropriate behavior more frequently (18).

Loss of insight

It is not unusual for patients with dementia conditions to lose insight about their condition. Patients with FTD-ALS may state that "Everything is fine" and "I've never felt better" (personal communications between authors and patients). This lack of insight can affect decision-making and adherence to treatment, as patients may not acknowledge disease-related risks associated with walking, driving, and swallowing. In a study comparing patients with classic ALS with those with FTD-ALS, patients with FTD-ALS had worse compliance for breathing treatments and nutritional supplementation (11).

Interventions when loss of insight is present—Discuss treatment preferences with the patient as early as possible in the disease. Including family in these early discussions can be helpful in gaining understanding and acceptance of the patient's preferences. This will help later in making decisions that reflect the wishes of the patient and respect their quality of life. Patients with poor balance and weakness are at risk for falling, yet they may continually stand and walk on their own, risking falls and injury. Environmental modifications can be employed and may include having 1:1 supervision for the patient, making sure objects they desire are within reach so patients won't feel compelled to get up

on their own, finding an acceptable method for the patient to summon help when needed, and adherence to a predictable schedule.

Emotional changes

Self-centeredness and a lack of concern or empathy for the welfare of others can occur in FTD-ALS. Patients may show flatness of facial expression and blunting of emotional reactions, and they may have a harder time discerning cues from another person's facial expression (19). For instance, a woman with FTD-ALS was angry and dismissive when her husband was unable to help her move furniture on his first day home after cardiac bypass surgery. More commonly, caregivers may not get appreciation or thanks for caregiving tasks.

Pseudobulbar affect is a fairly common condition in ALS and refers to a syndrome in which the patient cannot control emotional expressions. Patients may laugh or cry without warning and have difficulty stopping once they have started. They may express emotions at inappropriate times, for instance laugh during a sad event, or crying during a routine conversation. Pseudobulbar affect can be embarrassing and have negative social consequences for both the patient and family. It should be noted that this condition can occur independently of the cognitive and behavioral symptoms discussed in this paper.

Interventions when emotional changes are present—When lack of empathy is present in a patient, the primary intervention is education and support to the family about the nature of FTD brain changes. This symptom of ALS/FTD can be confusing for loved ones. It is important to help prevent the patient and caregiver from becoming socially isolated, by encouraging involvement and participation of understanding friends and family members. Certain antidepressant medications have been used to treat pseudobulbar affect and experimental trials with other drugs are currently underway.

Social disinhibition

Patients with disinhibition can become more impulsive and reckless in social situations. They may make comments in public that are overly personal and even hurtful with little appreciation that their behavior is inappropriate or unkind. They make friends and strangers uncomfortable by staring at them or by standing too close.

Interventions when disinhibition is present—Monitor the patient's contact with the public; go to places where the patient is well known and behavioral symptoms are accepted and tolerated. Attempt to sit in more private and secluded sections of restaurants and waiting rooms. Sometimes, these arrangements can be made ahead of time. For example, at medical appointments, arrange to have the patient brought into an examination room as soon as possible rather than waiting in the lobby. Bring along something the patient enjoys, for example, a snack, activity, or game. Some of our families carry a small business-size card that briefly explains the patient has a medical illness affecting his behavior. The card can be passed to members of the public to gain their understanding of the patient's behavior. The SSRIs can provide some relief for disinhibition.

Hyperorality

FTD is associated with changes in eating habits. Patients often develop carbohydrate cravings thought to be due to lowered amounts of serotonin. Some patients exhibit profound changes in their style of eating and may lose table manners. They may become hyperoral, and eat past the point of satiety. The combination of these symptoms with the changes resulting from ALS can be challenging as well as life threatening. Weight loss is a common problem resulting from the muscle wasting associated with ALS. High calorie meals and

tube-feedings are common strategies employed to prevent serious weight loss, however, many patients with ALS suffer coughing and choking episodes while eating and drinking.

Interventions when hyperorality is present—Consultation with a dietitian and speech pathologist is often critical. These experts can strategize ways to provide appropriate nutrition while attending to safety needs. There are agents that thicken thin liquids making them safer to swallow. Keep “safe” foods handy and visible. Remove unsafe food and liquids from the patient's access, and lock cabinets and the refrigerator if necessary. Providing 1:1 supervision during mealtimes may be necessary in order to limit portions. The SSRIs can help diminish carbohydrate craving to a certain extent.

Aggression

Patients with ALS/FTD may have difficulty managing powerful emotions. Patients may express moodiness, frustration, or anger. They may display aggressive behavior during caregiving tasks.

Interventions when aggression is present—Though not the patient's fault, these behaviors need to be strictly managed. Remaining calm and avoiding an argument with an angry and aggressive patient is critical. Maintaining a safe distance and positioning oneself off to the side of the patient rather than directly in front of them (more threatening to the patient) are other strategies. Acknowledge the emotions the patient may be trying to express and then changing the subject can help diffuse a situation as well. Look for patterns, as described above, and consider if a person is most at risk for aggressive outbursts when overwhelmed or tired, and take steps to avoid those situations. It is critical to inquire about the presence of weapons or other hazardous objects in the home, so that they can be removed from the patient's reach. Some patients may benefit from the NMDA antagonist memantine although evidence from a placebo-controlled study is still lacking (20).

Rigidity and compulsivity

Focus and obsession with specific activities can occur in FTD-ALS. Some patients collect and hoard objects. Some of our patients have become so obsessed with certain activities that they ignore important responsibilities such as caring for their children. Patients typically have no insight that their behavior is troubling or upsetting for others.

Interventions when rigidity and compulsivity is present—In many cases, the caregiver must accept the behavior, even though it is annoying and frustrating. Sometimes a less distressful behavior can be substituted for the current one. For example, many of our patients play card games on the computer for hours at a time. Caregivers may be able to substitute time on the computer with a real deck of cards, and in this way, can take the patient on errands and appointments. In cases where the obsessive behavior results in risky situations, those responsibilities must be taken from the patient.

Summary

While we have focused primarily on strategies for managing the patient's behavioral and cognitive symptoms, it is critical to attend to the emotional and physical health of the family caregiver. The literature shows strong evidence that dementia family caregiving is associated with high levels of stress and burden (21). There are negative physical and emotional consequences that family caregivers face as well (22). Research on the experience of FTD caregivers demonstrates high levels of distress when behavioral symptoms are present, particularly disinhibition and apathy (23,24). Despite these findings, in clinical experience, we find that families provide care with tremendous resiliency, compassion, and

devotion. Many family members profess satisfaction at mastering new challenges and difficult situations. Caregiving literature suggests that behavioral-skills training is effective at reducing caregiver stress (25). We have drawn from our clinical experience of working with FTD-ALS patients and families and presented interventions for working with patients and their families. It will be important to develop and test interventions aimed at improving the patient and caregiver experience in FTD-ALS.

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Table 1

Intervention Model

Intervention levels	Examples of strategies
Environmental	Use short phrases when communicating to enhance patient's comprehension. Limit visitors and activities in the afternoon due to patient's fatigue and increased irritability.
Behavioral	Distraction and diversion. Validate the patient's underlying emotion(s).
Pharmacological	Consider use of an SSRI.
Physical	Lock doors that access garage and tools that patient can no longer handle in a safe manner.
Internal to the caregiver	Work toward acceptance for changes in patient's health. Education provided by the health care team. Support group or individual therapy.