WHEN IT’S NOT ALZHEIMER’S: FRONTOTEMPORAL DEMENTIA

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KEY POINTS:

- Frontotemporal Dementia (FTD) is an aggressive form of neurodegeneration affecting the personalities and language function of younger patients
- New treatments for FTD are entering clinical trials soon

Frontotemporal dementia (FTD) is a progressive, neurodegenerative disorder that most commonly strikes patients in the prime of life – their mid-50s to 60s. Yet, unlike Alzheimer’s disease in which memory and cognition are affected early, these patients usually have intact memory and general cognition. Patients with FTD undergo progressive destruction of the frontal and/or temporal lobes of the brain, which can result in blunting of their emotions and their capacity to experience feelings. This includes the loss of empathy for others, even family members, as well as signs of impulsiveness, inappropriate or disinhibited social behaviours, and for some, extreme irritability and aggression. The loss of empathy is one of...
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the most devastating aspects of the disease for families and caregivers – patients become cold, indifferent, and even callous toward the people they most loved and cherished, yet at the same time becoming entirely dependent on their family for care.

If the left frontal and temporal lobes are primarily affected, patients may exhibit difficulties with language and speech, called “aphasia”. Patients with left frontal degeneration have difficulty with word retrieval and pronunciation. Patients with left temporal lobe disease lose the meaning of words, and have increasing difficulty with language comprehension, a subtype of FTD called “semantic dementia”.

Because the symptoms of FTD can be subtle at first, particularly when they include changes in personality, decision-making, and judgment, the disease is often under-recognized. In addition, standard neuropsychological tests may not always detect the abnormalities. This is especially true in the early stages, when standard brain imaging techniques such as CT and MRI may also be normal. Thus, patients who present with early or subtle changes in social behavior, but no evident pattern on imaging, may go undiagnosed for years.

Despite difficulties in the diagnosis of FTD, recognition of the disease by neurologists and other healthcare professionals is growing. Much research is underway to identify better diagnostic tests, such as new imaging methods, genetic analysis, and tests of proteins in blood and spinal fluid. Through such tests, the disease can be identified in its earlier stages, and in family members who may be at risk of developing the disease.

Most unfortunately, there are currently no treatments to slow or cure the disease, and no treatments approved specifically for the treatment of symptoms in FTD. At best, empirical treatment with SSRIs or cholinesterase inhibitors may be attempted. However, for the first time, clinical trials testing treatments specific to frontotemporal dementia are starting. In London Ontario, we are investigating the potential of a hormone, oxytocin, to boost empathy and positive social interactions in patients with FTD. As well, novel potential treatments which aim to block progression of FTD are being developed by pharmaceutical companies worldwide, and are expected to be available within clinical research trials in Ontario in 2013.

For more information on current frontotemporal dementia research opportunities in London Ontario, please contact the Cognitive Neurology and Alzheimer Disease Clinic at 519-646-6032. For further information and resources on frontotemporal dementia, please visit the website of the Association for Frontotemporal Degeneration, www.theaftd.org

APPROACH TO THE PATIENT WITH NEUROPSYCHIATRIC SYMPTOMS ASSOCIATED WITH DEMENTIA

Alzheimer’s disease (AD) and related forms of dementia are becoming increasingly prevalent with the aging demographics of most developed countries.\(^1\) Currently, there are over 500,000 older adults living with dementia in Canada\(^1\) and approximately 80% of individuals with dementia display neuropsychiatric symptoms (NPS) at some point in their illness.\(^2\) NPS are particularly challenging in long-term care (LTC) populations, where the reported prevalence of dementia is between 50-70%\(^3\)\(^-\)\(^9\), and over 80% of these individuals have NPS.\(^3\)\(^,\)\(^10\)\(^-\)\(^18\)
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Neuropsychiatric symptoms of dementia, also known as behavioural and psychological symptoms of dementia (BPSD), have been defined as “signs and symptoms of disturbed perception, thought content, mood, or behaviour that frequently occur in patients with dementia”. Such symptoms can be generally classified into five symptom clusters: psychosis, sleep disturbance, depression, apathy, and agitation.

Agitation has been defined as “purposeless motor or vocal activity commonly observed in individuals with dementia that are not driven by unmet needs”. One commonly used rating scale for NPS is the Cohen-Mansfield Agitation Inventory, which rates a variety of agitated behaviours, including verbal (e.g. yelling, repetitive vocalizations), non-aggressive physical (e.g. restlessness, pacing), and aggressive physical agitations. Another commonly utilized rating scale, the Neuropsychiatric Inventory (NPI), captures broader symptomatology and clusters symptoms into psychotic symptoms (e.g. hallucinations, delusions), hyperactivity (e.g. agitation, irritability, euphoria, disinhibition), and disturbances in mood (e.g. anxiety, depression, apathy, disturbances in sleep and eating).

A comprehensive assessment of NPS is multifaceted. First and foremost is a thorough history, identifying the target symptoms, so that management can be tailored accordingly. Collateral information from caregivers is an essential component, especially as the patient’s cognitive impairment progresses. Caregivers can provide a longitudinal account of the progression of NPS, and identify possible triggers. Additionally, the caregiver interview provides insight into caregiver burden, as NPS frequently contribute to caregiver distress, depression, increased care costs, and risk of nursing home placement.

The first step in managing NPS is to rule out reversible conditions, including: underlying medical disease, infection, medication side-effects, as well as patient pain, constipation, and unmet basic needs (i.e. hydration, nutrition, and sleep).

A treatment plan can be developed using both non-pharmacologic and pharmacologic treatment approaches to target multiple NPS, with priority given to treating the most distressful and unsafe symptoms first. An overview of the evaluation of NPS is provided in Table 1.

Table 1 Approach to the Assessment of Neuropsychiatric Symptoms of Dementia

| Obtain history from patient (if possible) and caregiver(s) |
| Identify the specific behavioural and psychological symptoms of dementia |
| Evaluate reversible conditions (medical illness, infection, medication effects, pain, unmet basic needs) |
| Evaluate functional (i.e. hearing, vision, constipation) and environmental triggers |
| Evaluate caregiver burden |
| Ensure safety of patient and caregiver |
| Perform a thorough physical examination, including neurological examination guided by history |
| Cognitive assessment and standardized assessment tools for NPS (e.g. NPI) |
| Pertinent investigations (i.e. bloodwork/urinalysis, EKG, and neuroimaging where indicated) |
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REFERENCES


RESOURCES

First Link® - Connect persons with dementia and their family caregivers to support, information and services in their community.

A Guide to Scheduling and Billing for Family Physicians (in Ontario):
http://bit.ly/100q6iw

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