DEMENTIA EDUCATIONAL RESOURCE

You Can Manage a Dementia without Cure: Frontotemporal Degeneration

Abstract

Much of the published clinical research in dementia has focused on diagnostic biomarkers and neuroimaging analyses that are not yet validated for routine clinical practice or on unsuccessful clinical drug trials. Primary care providers can nonetheless make a significant difference in the management of patients with dementia and their families, based on appropriate referrals of non-Alzheimer's dementia cases to specialists and supporting informal caregivers.

Frontotemporal degeneration, a non-Alzheimer's dementia that strikes in the 6th decade of life, provides many opportunities for the entire healthcare team to educate and back families up through a harrowing neurodegenerative illness. This paper is intended to highlight for primary care physicians what can be done and how to accomplish it through a team approach. Some concepts, such as a switch from medicalized views of "behavioural and psychiatric symptoms of dementia" to "Responsive Behaviours" can be generalized across dementia etiologies, but the age at onset and marked social disability and dysfunction caused by frontotemporal degeneration warrant some additional guidelines to assure the safety and highest quality of life possible for the patient and those around him. In particular, refitting a day program to accommodate clients with frontotemporal degeneration and attending to the needs of children who find themselves in informal caregiver roles are addressed.

Keywords: *caregiver, dementia, frontotemporal dementia, primary progressive aphasia*

Forum

Pre-test CME Quiz

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New Diagnostic Criteria for Frontotemporal Degeneration

Over the past 10 years, more progress has been made in understanding the genetics and proteinopathy underlying frontotemporal degeneration than in Alzheimer's disease.[1] These recent discoveries have led to new 2011 diagnostic criteria. Physicians should consider the diagnosis of frontotemporal degeneration in the following instances: 1) onset age younger than 65, 2) complaint of early and marked personality change (e.g., previously introspective and earnest is now crass, boisterous, and uncaring), 3) marked language difficulties unaccompanied by memory loss, 4) MMSE score > 26 out of 30 despite lost instrumental activities of daily living (e.g., change from workplace exemplar to demotion or on probation; inability to organize billpaying), 5) anterior brain atrophy on structural imaging, as opposed to generalized finding. Frontotemporal degeneration (previously referred to as "Pick's disease" or frontotemporal dementia) in this article is used to denote the umbrella that includes both the variant manifesting most prominently with behavioural changes (bvFTD) and the variant that shows mostly aphasic changes in the first 2-3 years of illness, also known as primary progressive aphasia (PPA). The criteria for bvFTD and for the primary progressive aphasias highlight the cardinal signs of these frontotemporal degeneration subtypes.

Key Point
Behavioural disturbances can occur in patients with all subtypes of frontotemporal degeneration.

Behavioural variant frontotemporal degeneration (bvFTD)

The criteria for bvFTD have shifted since the 1998 Neary consensus criteria were developed. The new criteria allow for patients to be diagnosed at least with *possible* bvFTD early in the course of illness, before all possible symptoms have arisen (see Table 1).[2] Positive imaging evidence to support a frontal and/or temporal neurodegenerative process can bring a patient up to a *probable* bvFTD diagnosis, while showing 4-6 of the symptom criteria along with evidence of tauopathy, TAR DNA protein-43 (TDP-43), or fused-in-sarcoma inclusions on neuropathology or positive carrier status for mutations in microtubule-associated protein tau (MAPT), progranulin, or C9ORF72 will propel the diagnosis of bvFTD to the realm of *definite*.

Three Primary Progressive Aphasias

As reflected in the 2011 diagnostic criteria for PPA, there are now 3 subtypes or variants: 1) non-fluent/agrammatic, 2) semantic, and 3) logopenic.[3] It can be difficult for clinicians without speech and language pathology training to distinguish between non-fluent speech and speech that hesitates due to anomia, but the hallmark features

Table 1: Diagnostic criteria for behavioural variant FTD[16]

Possible bvFTD	Probable bvFTD	bvFTD with Definite FTLD
3 of 6 diagnostic features: – Disinhibition – Apathy / Inertia – Loss of sympathy / empathy – Perseverative / compulsive – Hyperorality – Neuropsychological profile	– Functional disability – Neuroimaging consistent with bvFTD	– Evidence of frontotemporal lobar degeneration (FTLD) – Pathogenic mutation

of the non-fluent/agrammatic variant are an apraxia of speech as well as loss of grammar. An apraxia of speech can be conceived of as similar to a limb apraxia- the overlearned motor programs, in this case those for making simple sounds such as "la," "ma," or "koo," are inaccessible to the patient as he speaks in sentences.

The criteria for semantic variant of PPA (formerly known as semantic dementia) continue to focus on a comprehension deficit, moreso than an expressive one, but one characteristic more specific to semantic variant PPA is the loss of single word meaning. Patients with Alzheimer's disease experience aphasia over the course of their illness, and this may be confused with semantic variant PPA until one evaluates for single word comprehension (e.g., "What is a chair?" "Point to a watch.")[4]

The criteria for logopenic variant of PPA raise the issue of aphasia that is tainted by memory loss. These patients will have difficulty repeating longer sentences, as if they have forgotten what they were doing mid-task. Intriguingly, some but not all patients with logopenic variant PPA have Alzheimer's pathology at autopsy.

In an oversimplified way, non-fluent/agrammatic variant cases can't talk; semantic variants cannot hear (comprehend); and logopenic variants cannot repeat. Patients with bvFTD may have word-finding difficulty (anomia) that is only detected through formal examination; informants will typically feel so overwhelmed by the marked change in comportment that they are unaware of the anomia.

Key Point

Some behavioural and psychiatric symptoms of dementia are Responsive Behaviours and warrant individualized exploration of causes before resorting to drug therapy.

Management of Frontotemporal Degeneration

Having introduced the symptoms of frontotemporal degeneration, this article will now reflect upon how those symptoms, occurring mainly during mid-, as opposed to late-life, can be managed. Note the word used was *managed*, not cured or even treated. The psycho-

tropic medications most frequently prescribed for the obsessive-compulsive features, mental rigidity, anxiety, disinhibition or apathy of bvFTD (which can also be seen later in PPAs) have generally not proven themselves in formal clinical trials.[5] Readers are referred to the Canadian Coalition for Seniors' Mental Health http://ccsmh.ca/pdf/ccsmh_ltc_meds_front.pdf and http://ccsmh.ca/pdf/ccsmh_ltc_meds_back.pdf for their free downloadable Tool on Pharmacological Treatment of Behavioural Symptoms of Dementia in Long Term Care Facilities for Older Adults. The recommendations listed thereon hold true for early-onset frontotemporal degeneration also.

The onus of tolerating, persuading away from,[6] or controlling these symptoms or Responsive Behaviours that arise from lost ability to meet one's own needs rests, for the main part, with informal family caregivers in Canada. There simply are not enough long-term care facility, hospital, neurobehavioural or psychiatric unit or respite beds available for all patients with dementia and Responsive Behaviours to be institutionalized.

Key Point

As much as we would like to think we can write useful prescriptions, caregiver support and education are the most effective mainstays of dementia care.

Importance of Caregiver Support

Due to the long-term disability from dementia, support for informal caregivers is paramount. Adalsteinn Brown, recent Assistant Deputy Minister at the Health System Strategy Division in Ontario's Ministry of Health and Long-term Care, has pointed out that health human resources are the most economic, effective, and necessary support systems for the increasing prevalence of dementia. If informal caregivers were paid an hourly rate, their bill for Canadian seniors in 2007 would have totaled more than $24 billion.[7] This figure did not include the number of family leave hours taken by caregivers working outside the home. Caregiver burden in Canada is growing: informal caregiving for AD requires 231 million hours per year, and this will increase to 756 million hours for the next generation.[8] Mary Mittelman has steadily reported evidence that adult, mainly spousal, caregiver support, even if provided by telephone, leads to better quality of life for both the caregivers and patients with dementia. Her recent systematic review of the literature also reports that multicomponent caregiver support delays institutionalization and improves patient behaviour.[9] The current most effective healthcare team intervention for frontotemporal degeneration is to support caregivers.

Needs Specific to Frontotemporal Degeneration Management

The primary caregiver to a patient with late-onset dementia is often an elderly spouse or child in midlife with children of her own, but with early-onset dementia such

as frontotemporal degeneration, the spouse has been converted to the single income-earner and single parent, as well as acting as the substitute decision-maker and primary supervisor for the patient. The combined stress of these roles may be exacerbated by delays in the correct diagnosis of the patient, during which there is an impression that the cause is "marital discord," "midlife crisis," or "new substance abuse," and not a tragic neurodegenerative disorder.

Issues for this early-onset dementia flare up frequently and require more urgent responses from the healthcare team than late-onset Alzheimer's disease. Medication adjustments, because there are no 'silver bullets' for the behavioural symptoms of the illness, may require telephone calls every two weeks until conditions are safe and endurable for the household. The nurse and social worker on my clinical team may speak on the phone with such caregivers weekly for a month at a time, to provide support as new crises peak and shift over the patient's 10-year course of illness, some brought on by progressive symptoms of the illness (e.g., altercation with other drivers due to disinhibition) or consequences of Responsive Behaviours (e.g., combative during personal care). Symptoms of the illness may cause work-related law suits, if a patient's early symptom of impaired judgment led to business losses; patients susceptible to cable TV advertising may empty the family's savings account; or patients may be arrested by the police for shoplifting. See Table 2 for a Proposed Management Checklist for Office Visits. Because of the shifting nature of this dementia, the checklist bears repetition every 6-8 months. The clinician, nurse and social worker are all responsible for assuring that any driving privileges held are appropriate for the safety of all, that there are no firearms available, and that the patient's community is educated enough to be aware of how they can help to manage the patient. Readers are encouraged to see the paper written by our social worker for more on this topic.[10] By dint of no available cure and the terminal nature of neurodegenerative dementia, our treatment is palliative.

Key Point
Refer children of patients with early-onset dementias to age-appropriate resources to help them cope.

Responsive Behaviours

Frontotemporal degeneration causes behavioural changes, and the loss of one's ability to communicate effectively or to manage basic activities of daily living independently can result in further Responsive Behaviours.[11] In this respect, Responsive Behaviours may be considered akin to 'acting out.' The strategy that will likely work best for Responsive Behaviours is behavioural intervention and not pharmaceutical sedation

Table 2: Management Checklist for Office Visits

History (could be obtained by nurse or physician assistant):

- ❑ Response to latest medication changes from your office
- ❑ Any changes in psychotropic medications made by other physicians? These patients are typically followed, albeit infrequently, by several physicians, all of whom may be writing medications.
- ❑ New behaviours or habits? (These may erupt into the 6-8th year of illness but generally convert from hyperactivity to apathy and withdrawal.)
- ❑ Driving safety
- ❑ Unwise spending or other habits that could endanger patient or family's livelihood or sense of safety

Physical examination:

- ❑ Signs of parkinsonism (rigidity, resting tremor, limb apraxia, postural instability)
- ❑ Signs of motor neuron disease (fasciculations, limb weakness and atrophy)

Action plan:

- ❑ Review target symptoms for all psychotropic medications so that caregivers can report well on efficacy
- ❑ Document informed consent for drugs being used off-label and those that carry risk of early mortality (atypical antipsychotics)
- ❑ Consider tapering off psychotropic medications once patient becomes quiet, withdrawn and less mobile
- ❑ Consider re-referrals to neurologist if any physical examination aspects above have begun
- ❑ Provide explanatory letters for family members to use at businesses frequented by the patient, the airport if travelling through Customs, or the bank if patient should not have full access to bank accounts/credit cards
- ❑ Refer caregivers to support through the Alzheimer Society, respite or day programs to help them pace their energy and survive the ambiguous loss of caring for someone young with dementia
- ❑ Address the dementia with any children living in the household so that they can play a meaningful role in care yet also maintain a sense of safety

or tranquilization.

Dr. Dmytro Rewilak of Baycrest's Psychology Department has assisted both my outpatients and the Day Program's clients by teaching an ABC strategy (see research.baycrest.org/chow-lab for video and downloadable worksheet). A is for Antecedent, B is for the objectively observed Behaviour(s), and C is for Consequences. Healthcare team members can help families by honing in on the ABC for each undesired behaviour. Listing Antecedents may reveal specific stimuli for that Responsive Behaviour. For example, the patient may become agitated at the end of the day if the return of several exhausted, distracted household members to home simultaneous to the personal support worker's departure is overstimulating.

Staff reviewing the observable aspects of the Behaviour may be able to convert an initial report of "anxiety" to one of obsessive-compulsive activities that are not accompanied by the patient feeling upset and from which the patient could easily be distracted, sparing him unnecessary sedation.

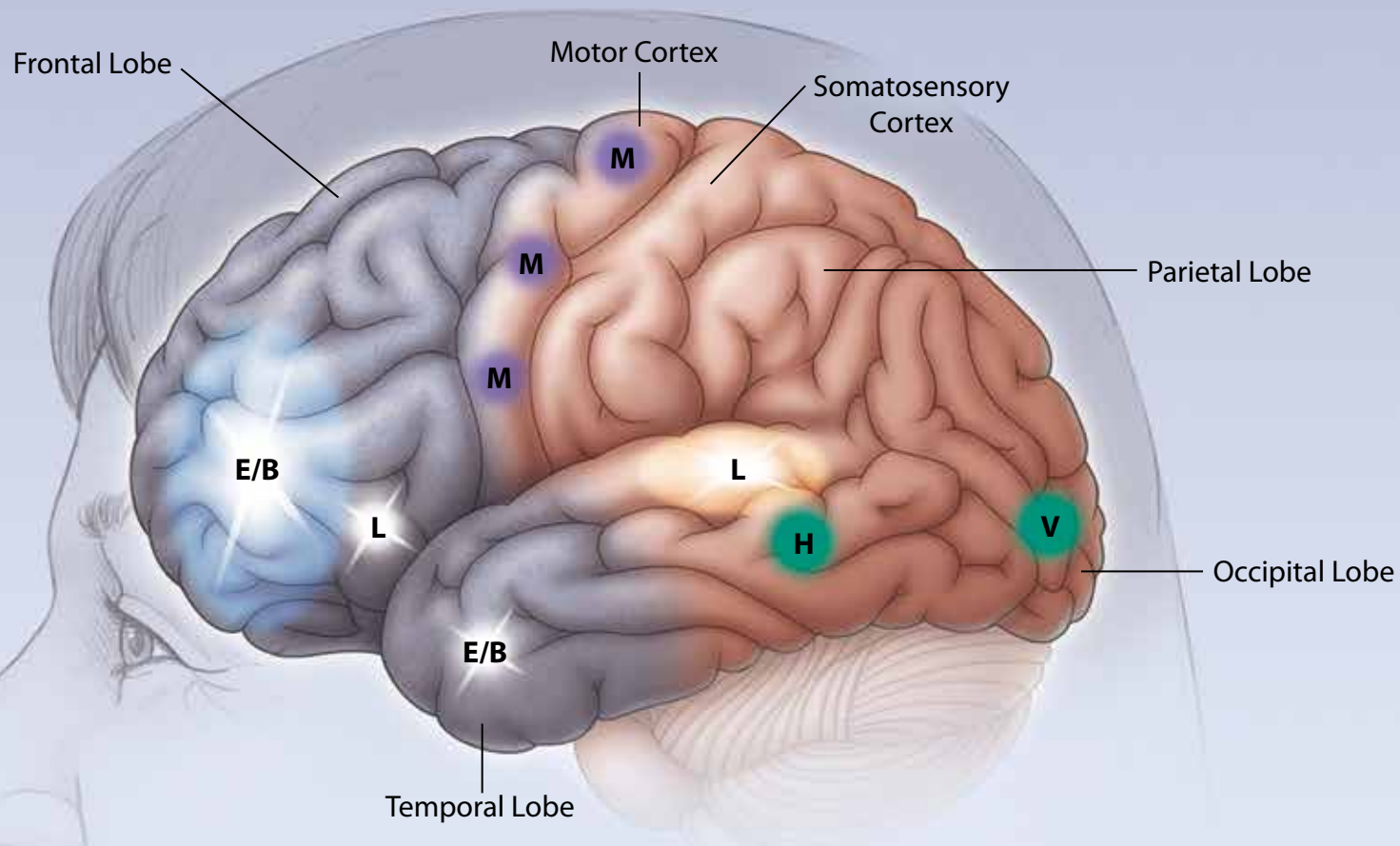
Discussing Consequences of the behaviour may become an eye-opener for family members. Patients with frontotemporal degeneration often seek connection with others around them, even if they achieve this in childish, inappropriate or even dangerous ways. If the consequence of a behaviour is that the otherwise disengaged caregiver yells at or berates the patient, that goal of connection has been achieved. Rewarding the desirable comportment or activity should be the rule, as opposed to punishing undesired behaviours. Another useful framework, although it was developed for management of dual diagnoses in psychiatry, is the P.R.N. protocol.[12]

Caring for frontotemporal degeneration requires creativity, openness to change, and persistence. These are hard to do alone, and we try to stress to caregivers after making the diagnosis that they have been inducted onto a team that will continue to work with them over time.[13]

Teamwork Makes the Difference

The challenges of adjusting limits of care for a day program to accommodate clients with a frontotemporal degeneration have been met by adding more recreational therapist staffing, as opposed to increasing nursing, and by creating a new alliance between the clinic and the day program at Baycrest.[14] Historically, day programs rest under the standard of psychosocial care, but the prevalence and severity of Responsive Behaviours in frontotemporal degeneration necessitate collaboration between the prescription-writers and the non-pharmacologic interventionalists. Together, this hybrid team has been able to elucidate routines that patients can

Frontotemporal Degeneration and the Brain



Abilities always affected by FTD: E/B Emotions and Behaviour; L Language

Abilities sometimes affected by FTD: M Movement

Abilities not affected by FTD: H Hearing; V Vision

engage in with a minimum of Responsive Behaviours and teach those practices, as well as endorsing the psychotropic medication regimen, to family members.

Child-Caregivers

The author has taken particular inspiration in the past few years to attend to an unmet need for the children of patients with frontotemporal degeneration. In addition to concerns about the health and livelihood of the affected parent, we have learned that those child-caregivers also worry about losing the well-parent, either due to misconceptions about contagion from a dementia or due to witnessing the effects of these extreme stressors on the well-parent. A needs assessment carried out by focus groups revealed that these very strong participants were able to take a lot of good from the experience of caregiving: they were proud of their ability to help care for the patient, and they felt they had matured earlier than their classmates. In some cases, going through the illness and passing of a parent brought them closer to the well-parent. Consult-

SUMMARY OF KEY POINTS

Behavioural disturbances can occur in patients with all subtypes of frontotemporal degeneration.

Some behavioural and psychiatric symptoms of dementia are **Responsive Behaviours** and warrant individualized ABC exploration of causes before resorting to pharmacotherapy.

As much as we would like to think we can write useful prescriptions, **caregiver support and education** are the most effective mainstays of dementia care.

Children of patients with early-onset dementias need age-appropriate resources to help them cope.

ing family counselors, psychologists and social workers warn of the adverse effects of caregiving in early life, however, and these include losing the opportunity to develop social skills with peers because of devoting too much time to caregiving in isolation, taking on a compulsive caregiver stance in all future relationships, and suppressing the myriad feelings that are raised by the experience of ambiguous loss in order to present a strong 'front' for the well-parent.[15]

To address the teen segment of caregivers, we launched a website in Fall, 2011, www.lifeandminds.ca/whendementiaisinthehouse to address what focus group participants had identified as information they could use directly, advice to their own well-parents, and advice to other teen caregivers. Take-home points for teens are to continue extracurricular activities at school and to get help from friends, teachers, or counselors. Teens are encouraged to take advantage of support groups, informal discussion with friends, and family discussions to air their feelings and take their place among family members who count. The website continues to enjoy regular visitation, and plans to translate it into other languages, as well as to build in some interactive capacity proceed. Healthcare professionals may use downloadable templates on the site to write the letters to Customs officials or bank managers that assist caregivers in explaining the diagnosis and requesting special circumstances.

To address even younger children aged 5-9 who are nonetheless informal caregivers to patients with frontotemporal degeneration, we have created an educational activity book that can be downloaded and printed for free from research.bay-

CLINICAL PEARLS

History that can be obtained by nurse or physician assist that should document any changes made to medications by other physicians as well as new behaviours or habits, driving safety, and unwise spending or other habits that could endanger patient or family's livelihood.

Signs of parkinsonism and/or signs of motor neuron disease should be documented during a physical examination.

An action plan should include a review of target symptoms for all psychotropic medications and document informed consent for drugs being used off-label. Consider tapering off psychotropic medications once patient becomes withdrawn. Re-referrals to neurologist should be considered if any physical examination aspects have begun.

Address the dementia with any children living in the household so that they can play a meaningful role in care.

Post-test

CME Quiz

Members of the College of Family Physicians of Canada may claim MAINPRO-M2 Credits for this unaccredited educational program.

crest.org/chow-lab. By late 2013, this activity book will also be available by link on www.lifeandminds.ca/whendementiaisinthehouse.

Even without a Health Canada-approved drug indicated for bvFTD or PPA on the shelf, there is much clinicians and their multidisciplinary teams can do that is meaningful to the quality of life for patients with these dementias and their families. Primary care physicians make a vital contribution to dementia care when they monitor issues of safety, maintenance of meaningful interactions, and end-of-life planning for patients that pace the burden of caregiving for family members.

Acknowledgements

Initiatives described in this paper have been funded by: the Ontario Ministry of Health and Long-Term Care funding for day program development; Ontario Ministry of Health and Long-Term Care Academic Health Science Centres Alternate Funding Plan Innovation Fund at Baycrest; Women of Baycrest; and the Canadian Dementia Knowledge Translation Network.

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