Dementia Care in 2016

A Review of Frontotemporal Dementia, and contemporary issues in dementia care

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A silly picture ...

• pseudo-profound look

• the sea, cliché choice for a psychiatrist

But you’re here with me anyway!
Len Kropioski: 1918 - 2016

- WWII veteran
- highly involved in amateur sports in Kenora over a lifetime.
- remarkable persona, and a remarkable personality.
What makes a good life?

• longevity?
• perseverance?
• relationships?
• ability to live out our values?
• what else?
Dementia: Why this talk?

• many mental health practitioners and others with an interest in this group

• Frontotemporal Dementia is highly represented in our service sector

• it is under-recognized and under-discussed
Dementia 2016:

- review Frontotemporal Dementia
- case review
- review NICE guidelines for dementia care and treatment
- look ahead to issues raised by Medical Assistance in Dying law.
Case: Frank

• 35 year-old male, married, landscaper. High school graduate. Some design-work component. No children.

• Medical history: briefly a smoker, quit late 20s, otherwise unremarkable.

• no past psychiatric history.

• pre-morbid personality mildly introverted, “quiet” and pleasant person.
Referred by Probation Services to CMH:

• “Did stuff I don't normally do this past year.”
• problems “making decisions”
• unstable mood
• low energy, concentration, and motivation
• difficulty finishing tasks
• “I feel like I’m slipping a bit.”
• “losing a sense of reality,” difficulty “telling reality from fantasy.”
CMH assessment (continued):

• short-term memory complaint, had to “simplify his room” to compensate

• above slowly increasing over two years

• claimed longstanding violence towards his wife, including choking and biting her.

• over preceding year: marital separation, and restraining order, uttering threats and sexual assault charge (occurred in workplace)
MSE:
• Appearance, grooming unremarkable.
• Obsequious attitude.
• Co-operative
• Agitation when psychotic symptoms queried.
• Anxious laughter

Impression: query Schizophrenia
Frontotemporal Dementia (FTD):

• the clinical syndrome caused by **frontotemporal lobar degeneration**.

• result of **progressive neuronal** loss, predominantly involving the **frontal** and/or **temporal lobes**

• loss of over 70% of **spindle neurons**, while other neuron types remain intact.
Frontotemporal Dementia (FTD):

• First described by Arnold Pick in 1892 and was originally called “Pick's disease.”

• **Pick disease**, one specific type of frontotemporal dementia, diagnosed at autopsy.

• second-most common dementia.

• FTD accounts for 20% of early-onset dementia.
FTD: 3 subtypes described (from clinical features):

- **Behavioural variant frontotemporal dementia (BvFTD):** changed social behaviour and conduct, with loss of social awareness and poor impulse control.

- **Semantic dementia (SD):** loss of semantic understanding, resulting in impaired word comprehension, although speech remains fluent and grammatically faultless.

- **Progressive nonfluent aphasia (PNFA):** progressive difficulties in speech production.
BvFTD:
6 distinct clinical features

• Disinhibition
• Apathy / Inertia
• Loss of Sympathy / Empathy
• Perseverative / compulsive behaviors
• Hyperorality
• Dysexecutive neuropsychological profile
BvFTD: 6 clinical features

• Three of six features for diagnosis of possible bvFTD.

• More reliable than imaging.

• Distinguish bvFTD from disorders such as Alzheimer's and other causes of dementia.

• Proposed to allow a diagnostic hierarchy distinguished possible, probable, and definite bvFTD based on the number of symptoms present.

Dysexecutive Syndrome

• a group of symptoms, usually resulting from brain damage, that fall into cognitive, behavioural and emotional categories and tend to occur together.

• common pattern of dysfunction in **executive functions**, such as planning, abstract thinking, flexibility and behavioural control.

• also known as **frontal lobe syndrome**; dysexecutive syndrome preferred because it emphasizes the functional pattern of deficits over the anatomical location (**frontal lobe**); often not the only area affected.
FTD: preserved functions

- Perception
- Spatial Skills
- Memory
- Praxis
Frank: 1 month later, referred to ER by CMHW:

• In ER: Reports of striking self, appearing to respond to internal stimuli, disorganized behaviour, increasing self-care deficits.

• On admission: endorsed being "up and down," "aggressive," "complacent," "I don't know who I am or what I am doing," poor concentration, middle insomnia, 6 kg weight loss, agitation, awareness of changed behaviour. "General paranoia," "there's a fight coming with the Hell's Angels."

• Labs: all normal. No imaging; MRI pending.
• Course in hospital (over 32 days): vague endorsement of auditory hallucinations, "always muffled," from "Satan," continuing endorsement of danger from Hell's Angels.

• No evidence of response to internal stimuli. Interpersonal dependency.

• OT assessment: deficits in verbal / auditory memory, visual / cognitive organization, visuospatial organizing.

• Treated with olanzapine 10 mg daily. Impression: Schizophrenia, ? Factitious component. Discharged to short-term housing. Referred to ICM services.
Right temporal lobe: the “psychiatric phenotype” in FTD
Right temporal lobe atrophy. (A) Mild atrophy, with particular involvement of the inferomedial temporal lobe. (B) Severe atrophy, with bilateral, asymmetrical, temporal lobe atrophy affecting primarily the right temporal lobe structures.
Study: Right temporal lobe atrophy:

- Twenty patients with predominant right temporal lobe atrophy were identified on the basis of blinded visual assessment of the MRI scans.

- Profiles of cognitive function, behavioural and personality changes were obtained on each patient. The pattern of atrophy and the clinical features were compared with those observed in a group of patients with semantic dementia and predominant left-sided temporal lobe atrophy.

- Increased behavioural symptoms including social disinhibition, depression and aggressive behaviour. Nearly all behavioural disorders were more prevalent in the right temporal lobe atrophy patient group than the semantic dementia group. Symptoms particular to the right temporal lobe atrophy patient group included hyper-religiosity, visual hallucinations and cross-modal sensory experiences.

- The authors propose a separate syndromic variant of frontotemporal lobar degeneration.

DOI: [http://dx.doi.org/10.1093/brain/awp037](http://dx.doi.org/10.1093/brain/awp037) 1287-1298 First published online: 18 March 2009
FTD: neurotransmitter systems

- Cholinergic, dopaminergic, GABAergic, glutamatergic, noradrenergic and serotonergic system involvement have all been implicated in the development of the signs and symptoms of FTD.
The **National Institute for Health and Care Excellence (NICE)** is an executive **non-departmental public body** of the **Department of Health** in the **United Kingdom**. It serves both the **English NHS** and the **Welsh NHS**. NICE publishes guidelines in four areas:

- use of health technologies within the NHS (such as the use of new and existing **medicines, treatments and procedures**)

- clinical practice (guidance on the **appropriate treatment and care** of people with specific diseases and conditions)

- guidance for public sector workers on **health promotion and ill-health avoidance**

- and guidance for **social care** services and users.
Why NICE and dementia?

• our treatments have limited effectiveness. Prescribers are often at a loss.

• some have been relatively expensive.

• some have significant adverse (side) effects

NICE treatment appraisals are based primarily on evaluations of efficacy and cost-effectiveness in various circumstances. Limited data on FTD; no specific guidelines.
NICE: Assessment of behavioural disturbance in Dementia

• physical health

• depression

• possible undetected pain or discomfort

• side effects of medication

• individual biography, including religious beliefs and spiritual and cultural identity

• psychosocial factors

• physical environmental factors

• behavioural and functional analysis conducted by professionals with specific skills, in conjunction with carers and care workers.
• Individually tailored care plans that help carers and staff address the behaviour that challenges should be developed, recorded in the notes and reviewed regularly. The frequency of the review should be agreed by the carers and staff involved and written in the notes.
NICE: Non-medication treatment recommendations:

“Interventions tailored to the person’s preferences, skills and abilities.”

Including:

• aromatherapy
• multi-sensory stimulation
• therapeutic use of music and/or dancing
• animal-assisted therapy
• massage.

Relevance of voluntary sector is highlighted.
NICE: Summary of Evidence for Pharmacotherapy in Dementia
NICE: Antipsychotics and Dementia

• all antipsychotics studied appear to increase the risk of death when compared to placebo.

• Haloperidol, olanzapine and risperidone may also increase the risk of cerebrovascular adverse events, but evidence is lacking from studies of the other antipsychotics.

• “difficult to assess the incidence of individual side effects in all but olanzapine and risperidone.” In these drugs, there is evidence of increased risk of somnolence, hostility, confusion, fever/flu syndrome, abnormal gait, urinary incontinence, asthenia and peripheral edema compared to placebo.
Antipsychotics and VaD and AD:

- small reductions in neuropsychiatric symptoms on NPI or BEHAVE-AD.
- drugs studied were aripiprazole, olanzapine, quetiapine and risperidone.
- risperidone “may improve aggression.”
NICE: VaD or AD with significant agitation

“Moderate quality evidence” for:

• IM benzodiazepines and antipsychotics
  “benefits in terms of reduced psychotic symptoms and aggression/agitation that outweigh the risk of adverse events.”

• “unknown whether there is any difference between conventional and atypical antipsychotic drugs when administered by IM” injection h the risk of adverse events.”
NICE: VaD or AD with significant agitation

• in AD, there is moderate-quality evidence suggesting that donepezil (10 mg/day for 12 to 52 weeks), when compared with placebo, produces benefits in terms of reduced neuropsychiatric symptoms and agitation/aggression that outweigh the risk of adverse events.

• in AD, there is insufficient evidence to determine whether memantine (20mg/day for 24 to 28 weeks) produces clinically important improvements in neuropsychiatric symptoms.
NICE: Recommendations for Pharmacotherapy in Dementia
Dementia with behaviour that challenges

• pharmacological intervention in the first instance only if they are **severely distressed** or there is an **immediate risk of harm** to the person or others.
AD, VaD, mixed or DLB:

if severe non-cognitive symptoms (psychosis and/or agitated behaviour causing significant distress), use an antipsychotic if:

- full and informed consent
- comorbid conditions considered
- cerebrovascular risks considered
- identify and monitor defined target symptoms
- low dose with upward titration
- time-limited
- medication choice based on individual risk-benefit analysis
Caution re: DLB

• monitor carefully for severe untoward reactions, particularly neuroleptic sensitivity reactions (severe extrapyramidal features after treatment in the accepted dose range, acute and severe physical deterioration following prescription of antipsychotic drugs for which there is no other apparent cause).

• consider acetylcholinesterase inhibitor instead
Acetylcholinesterase inhibitors and BPSD in AD

• a non-pharmacological approach is inappropriate or has been ineffective, and

• antipsychotic drugs are inappropriate or have been ineffective.

• not indicated in VaD.
Behavioural emergencies in Dementia

defined as “violence, aggression and extreme agitation threaten the safety of the person with dementia or others”:

Monitor and address **environmental, physical health** and **psychosocial factors** that may increase risk:

- overcrowding
- lack of privacy
- lack of activities
- inadequate staff attention
- poor communication between the person with dementia and staff
- conflicts between staff and carers
- weak clinical leadership.
Behavioural emergencies in Dementia

• Health and social care staff should be trained to anticipate behaviour that challenges and how to manage violence, aggression and extreme agitation, including de-escalation techniques and methods of physical restraint.
Qualifications of staff administering medication for behavioural control:

• be trained in the **correct use of drugs** for behavioural control, specifically **benzodiazepines** and **antipsychotics**

• be **able to assess the risks** associated with pharmacological control of violence, aggression and extreme agitation, particularly in people who may be dehydrated or physically ill

• understand the **cardiorespiratory effects** of the acute administration of benzodiazepines and antipsychotics and the need to titrate dosage to effect recognise the importance … of monitoring pulse, blood pressure and respiration.
Qualifications of staff administering medication for behavioural control:

• be familiar with and trained in the use of resuscitation equipment

• undertake annual retraining in resuscitation techniques

• understand the importance of maintaining an unobstructed airway.
Medication Treatment in FTD
Target Symptoms for Pharmacotherapy

Behavioural targets:

- Obsessive-compulsive behaviour
- Motor restlessness

Clinical features unlikely to respond to pharmacotherapy:

- Socially inappropriate behaviours
- Loss of interpersonal skills
- Personality change
Summary of my approach:

- consider **sertraline, citalopram**, for compulsive, perseverative behaviours.

- consider **quetiapine, risperidone** for psychosis, agitation, aggression.

- consider **trazodone** for sleep induction.
Frank: back in community:

• MRI (brain): normal.

• Incident of sexual touching of acquaintance in a public place.

• Taking food out of stranger's hands. No longer able to shower. Entering cars, apartments and offices.
Frank: Readmitted 4 months after discharge:

• Interview: inability to account for behaviour.

• MSE: No coherent account. Less eye contact. Some inappropriate smiles. Increased reaction time latency and decreased speech. Loose associations and non sequiturs. Distinct delusions not elicited.

• Physical: normal.

• Impression: Schizophrenia vs. Psychotic Disorder due to general medical condition.
Course in hospital (over 15 months):

• Investigations: Repeat labs normal. CXR normal. EEG x2: diffuse metabolic / toxic encephalopathic process.

• regression into mutism and intermittent incontinence

• Initially treated with olanzapine and citalopram.

• Risperdone trial: no improvement

• Clozapine trial: further regression.
• Ziprasidone trial: increased agitation
• Higher dose clonazepam: no improvement.
• Trazadone 350 mg HS; no benefit
• 1 - 1 nursing care 16 hours per day for long period.
Frank: Family history

- Mother admitted to BMHC age 42, with confusion, hallucinations and insomnia. Failed ECT. Discharged to a group home. Died in mid to late 40s.

- Maternal grandmother had “nervous breakdown” in 40s, died at 52.
Frank: further course

• Final diagnosis: FTD. Transferred to a special care unit.

Continuing care in the community:

• deteriorating course

• citalopram 20 mg daily; no clear benefit
Medical Assistance in Dying Legislation in Canada
241.2 (1) A person may receive medical assistance in dying only if they meet all of the following criteria:

- (a) they are eligible — or, but for any applicable minimum period of residence or waiting period, would be eligible — for health services funded by a government in Canada;

- (b) they are at least 18 years of age and capable of making decisions with respect to their health;

- (c) they have a grievous and irremediable medical condition;

- (d) they have made a voluntary request for medical assistance in dying that, in particular, was not made as a result of external pressure; and

- (e) they give informed consent to receive medical assistance in dying after having been informed of the means that are available to relieve their suffering, including palliative care.
(2) A person has a grievous and irremediable medical condition only if they meet all of the following criteria:

- (a) they have a **serious and incurable illness**, disease or disability;

- (b) they are in an **advanced state of irreversible decline** in capability;

- (c) that illness, disease or disability or that state of decline causes them **enduring physical or psychological suffering** that is intolerable to them and that cannot be relieved under conditions that they consider acceptable; and

- (d) **their natural death has become reasonably foreseeable**, taking into account all of their medical circumstances, without a prognosis necessarily having been made as to the specific length of time that they have remaining.
other requirements:

• second opinion required

• “immediately before providing the medical assistance in dying, give the person an opportunity to withdraw their request and ensure that the person gives express consent to receive medical assistance in dying;”
Alzheimer Society of Canada perspectives:

- Person needs to be capable of retaining and understanding new information, analyzing the information and making an informed decision.

- Consent must be clearly expressed and voluntary - at the time that medical assistance in dying is provided - and the person’s ability to make decisions must be carefully assessed.

- All of these abilities (i.e. retaining, understanding and analyzing information and making informed decisions) may be impaired in people with dementia and consent will not be possible at the time of medical assistance in dying or throughout the mandated period of reflection (during which a person can withdraw her/his consent).
Alzheimer Society of Canada perspectives:

- “the Alzheimer Society believes that if a person is not deemed competent, then she/he is extremely vulnerable and the risk of abuse is simply too great.”

- “Given the progressive nature of dementia, wishes, values and beliefs may change, skills are lost and the ability to make decisions is greatly reduced. MAID should only be possible when a person is deemed competent at the time of MAID.”
Alzheimer Society of Canada perspectives:

• “The Alzheimer Society of Canada believes that because we cannot predict future suffering, providing advance consent for MAID should not be possible for people with dementia.”

• “The Alzheimer Society believes that people with dementia need to be safeguarded as they will be extremely vulnerable at the end of their life. People with dementia do not have the capacity to make an informed decision and consent to end their life at the later stages of the disease.”
Report of the Special Joint Committee on Physician-Assisted Dying:

• “That the permission to use *advance requests* for medical assistance in dying be allowed any time after one is diagnosed with a condition that is reasonably likely to cause loss of competence or after a diagnosis of a grievous or irremediable condition but before the suffering becomes intolerable. An advance request may not, however, be made, prior to being diagnosed with such a condition. The advance request is subject to the same procedural safeguards as those in place for contemporaneous requests.”
Discussion: Dementia and MAID

• How is ethical dementia care delivered in an era of increasing focus on quality of life?

• How do we live our values while supporting others to live theirs? When there is conflict in value systems?

• What values underlie our call as healthcare providers?
Appendix: data
Double-blind RCTs:

*Lebert et al.* Trazodone in bvFTD

- Randomized, double-blind, placebo-controlled, crossover study
- 26 subjects with bvFTD.
- Significant decrease in NPI scores, mediated by improvements in irritability, agitation, depression and eating disorders.
Double-blind RCTs:

*Kertesz et al.* **Galantamine** in bvFTD and PPA

- 18-week open-label and 8-week randomized placebo-controlled trial of 36 bvFTD and PPA subjects.

- Improvement in global severity score in PPA was seen, but was not significant after correction for multiple comparisons.
Double-blind RCTs:

*Moretti et al.* Paroxetine vs piracetam in FTD

- Randomized 14-month study of paroxetine 20 mg/day vs piracetam 1200 mg/day.

- Eight patients in each group, aged 64–68 years, with a diagnosis of bvFTD.

- Patients treated with paroxetine showed significant improvements in behavioural symptoms, reflected by a reduction of caregiver stress.
Open-label studies:

- *Boxer et al*, and *Diehl-Schmid*: both negative trials of *memantine*

- *Moretti et al*: 20 subjects treated with *rivastigmine*; reductions in NPI, decreased depression scores

- *Swartz et al*: Open-label trials of *fluoxetine*, *sertraline*, and *paroxetine*, 11 subjects, decreased disinhibition, depressive symptoms, carbohydrate craving, and compulsive behaviour.