Non-Alzheimer Dementia: Information for Primary Care

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Cases

Vascular dementia

- Your 78 year old patient, Mrs. V. comes in with her daughter who is worried about her cognition. The patient has a history of type two diabetes, hypertension and dyslipidemia. Mrs. V's daughter has noticed lately that her mom seems to have a hard time doing certain things like getting groceries and cooking a meal. Her memory seems fine, she's not particularly forgetful and remembers the steps involved in cooking the meal when we ask her, but she just can't seem to get herself organized to do it.

Frontotemporal dementia

- Your 62 year old patient, Mr. F. comes in with his wife who is worried about his behaviour. Mr. F. seems to have a change in personality over the past several months and didn't really seem to care when a close friend was diagnosed with cancer. He has also been making inappropriate comments to family and friends and hugging people at the grocery store! Mrs. F. is worried that something serious is going on and that maybe he needs to see a psychiatrist.

Lewy body dementia

- Your 75 year old patient, Mrs. L. comes in with her son who is worried something is wrong. He's noticed that his mom seems to be confused for a day or two, but then seems to be back to her old self again. What's really worrying him though is that she has been complaining about having nightmares even when she is awake. He wants to know could she be having hallucinations?

Dementia in Parkinson's disease

- Your 80 year old patient, Mr. D. comes in with his wife who is worried about her husband's cognition. He was diagnosed with parkinson's disease a few years ago and things had been going well up until recently when he started having a hard time planning his day out and making simple decisions. Mrs. D. is also worried that her husband has been having visual hallucinations because he keeps talking about animals in the room that she can't see.

Epidemiology

- Prevalence
    - Nearly 750,000 people living with dementia in Canada
    - Expected to double every 20 years meaning 1.4 million people in Canada with dementia by 2031
    - 6-8% of patients over the age of 65
    - Increasing prevalence rate with increasing age: prevalence rate 30% in patents older than 85 years

Main Types of Dementia

- Alzheimer's dementia (50-75% of dementias)
    - Less than 25% of cases of Alzheimer's dementia in Canada are diagnosed and treated
- Vascular dementia (10-20% of dementias)
    - Cognitive impairment that is caused by or associated with vascular factors (either after a clinical stroke, or with clinically silent brain infarctions, but evidence of infarction on imaging)
- Frontotemporal dementia
    - Younger age of onset, average age in 60's
- Dementia with lewy bodies
- Parkinson disease dementia

Clinical Presentation

- **Vascular dementia**
    - Diverse clinical presentations depending on the specific pathology of infarction/ischemia
    - If associated with a stroke, onset or dramatic worsening in association with a typical stroke
    - If not associated with a stroke, may have an insidious onset and gradual progression
    - Two main clinical patterns
        - Cortical syndrome: executive dysfunction, apathy, aphasia, apraxia, agnosia, visuospatial disturbances
        - Subcortical syndrome: focal motor signs, gait disturbance, history of unsteadiness and frequent, unprovoked falls, urinary symptoms not explained by urological disease, personality and mood changes. Excessive forgetfulness commonly not an initial major manifestation

- **Frontotemporal dementia**
    - Presentation is often dramatic, suggesting a psychiatric disorder
    - Memory is usually preserved
    - Main variants
        - Behavioural variant
            - Changes in personality such as apathy and loss of empathy, loss of initiative and ability to follow through on tasks. Caregivers often describe patients as cold. Often mistaken for depression early in disease course
            - Lapses in judgement: rude or off-colour comments to family or strangers, socially inappropriate behaviours such as touching or kissing strangers, flatulence without concern
            - Obsessional behaviours: simple repetitive movements, complex ritualistic behaviours, rigid food preferences
        - Progressive nonfluent aphasia
            - Articulatory difficulty, inability to speak in a grammatically correct fashion, effortful halting speech, stuttering, oral apraxia
        - Progressive semantic dementia
            - Often fluent speech, but empty and brief with expressions "it's that thing, I don't know" and semantic paraphasias.
            - Able to understand complete sentences early on, but difficulty with single object words which progresses to global incomprehension

- **Lewy body dementia**
  - Presence of dementia with two of the following three core diagnostic features
    - Fluctuating cognition ranging from minutes to days - present in 60-80% of cases. Often described as “blanking out”
    - Visual hallucinations - present in 60% of cases
    - Parkinsonian symptoms: bradykinesia and akinesa, limb rigidity and/or gait disorder - present in 70-90% of cases
  - Other symptoms include
    - Syncope, postural instability with repeated falls
    - Transient loss of consciousness,
    - Autonomic dysfunction,
    - Delusions and multimodal hallucinations (i.e., auditory and tactile)

- **Dementia in Parkinson disease**
  - Diagnosis of dementia in the patient with an established diagnosis of Parkinson disease
  - 30% of patients with Parkinson disease have dementia; prevalence increases with duration of disease
  - Symptoms
    - Executive dysfunction
    - Impaired visuospatial function
    - Memory deficits less prominent; more related to retrieval of learned information which improves with cueing
    - Language dysfunction
    - Visual hallucinations strongly associated with dementia in parkinson disease

History and Interviewing Tips

- As the diagnosis of dementia remains clinical, the history is still the most important part for diagnosis
- It is important to obtain collateral history from a family member of caregiver
- Timing of onset and nature of symptoms are critical and help differentiate between the different subtypes
- Ask what the predominant feature is: memory, behavioural, language, hallucinations or movement disorder
- Assessment of activities of daily living
- Assessment of instrumental activities of daily living
- Other areas to cover in the interview
  - Rule out any symptoms of depression: Geriatric Depression Screen
  - Rule out any signs of delirium: Confusion Assessment Method
  - Rule out any signs of normal pressure hydrocephalus: urinary incontinence, confusion, abnormal gait
  - Assess for degree of impairment

DSM-5 Criteria

Vascular Neurocognitive Disorder

A. The criteria are met for major or mild neurocognitive disorder
B. The disorder as an insidious onset and gradual progression
   1. Onset of cognitive deficits is temporally relate to one or more cerebrovascular events.
   2. Evidence for decline is prominent in complex attention (including processing speed) and frontal-executive function.
C. There is evidence of the presence of cerebrovascular disease from history, physical examination, and/or neuroimaging considered sufficient to account for the neurocognitive deficits.
D. The systems are not better explained by another brain disease or systemic disorder.

Frontotemporal Neurocognitive Disorder

A. The criteria are met for major or mild neurocognitive disorder.
B. The disturbance has insidious onset and gradual progression.
C. Either (1) or (2):

    1. Behavioural variant:
        a. Three or more of the following behavioural symptoms
            Behavioural disinhibition
            Apathy or inertia
            Loss of sympathy or empathy
            Perseverative, stereotyped or compulsive/ritualistic behaviour
            Hyperorality and dietary changes
        b. Prominent decline in social cognition and/or executive abilities
    2. Language variant:
        a. Prominent decline in language ability, in the form of speech production, word finding, object naming, grammar, or word comprehension.

A. Relative sparing of learning and memory and perceptual-motor function.
B. The disturbance is not better explained by cerebrovascular disease, another neurodegenerative disease, the effects of a substance, or another mental, neurological, or systemic disorder

Neurocognitive Disorder with Lewy Bodies

A. The criteria are met for major or mild neurocognitive disorder
B. The disorder as an insidious onset and gradual progression
C. The disorder meets a combination of core diagnostic features and suggestive diagnostic features for either probable or possible neurocognitive disorder with Lewy bodies. For probable major or mild neurocognitive disorder with Lewy bodies, the individual has two core features, or one suggestive feature with one or more core features. for possible major or mild neurocognitive disorder with Lewy bodies, the invidious has only one core feature, or one or more suggestive features.

    1. Core diagnostic features:
        a. Fluctuating cognition with pronounced variations in attention and alertness.
        b. Recurrent visual hallucinations that are well formed and detailed.
        c. Spontaneous features of parkinsonism, with onset subsequent to the development of cognitive decline.
    2. Suggestive diagnostic features:
        a. Meets criteria for rapid eye movement sleep behaviour disorder
        b. Severe neuroleptic sensitivity.

A. The disturbance is not better explained by cerebrovascular disease, another neurodegenerative disease, the effects of a substance, or another mental, neurological, or systemic disorder.

Neurocognitive Disorder due to Parkinson's Disease

A. The criteria are met for major or mild neurocognitive disorder.
B. The disturbance occurs in the setting of established Parkinson’s disease.
C. There is insidious onset and gradual progression of impairment.
D. The neurocognitive disorder is not attributable to another medical condition and is not better explained by another mental disorder.

Differences between DSM-5 and DSM-IV

- The DSM-IV diagnoses of dementia are now entitled neurocognitive disorder
- DSM-5 also recognizes a less severe level of cognitive impairment, mild neurocognitive disorder
- Diagnostic criteria are provided forw both major NCD and mild NCD, as well as diagnostic criteria for the different etiological subtypes (with new separate criteria for NCD due to frontotemporal NCD, Lewy bodies, traumatic brain injury, Parkinson’s disease, HIV infection, Huntington’s disease, prion disease, another

medical condition and multiple etiologies.

Differential Diagnosis

- If the predominant feature is memory, think about Alzheimer's disease
- If the predominant feature is a change in behaviour or personality, think about frontotemporal dementia behavioural variant
- If the predominant feature is language, think about frontotemporal dementia language variant
- If the predominant feature is planning and organization and there is a history of vascular risk factors, think about vascular dementia
- If the predominant feature is cognition associated with parkinsonian features without a formal diagnosis of parkinson's disease, think about lewy body dementia
- If there is an existing diagnosis of parkinson's disease and there are now concerns about cognition, think about dementia in parkinson's disease
- If there is a sudden onset, think about acute toxicity, infection, tumour
- If there are associated headaches or seizures, think about tumour, hematoma, chronic infection
- Always consider:
    - Depression
    - Delirium
    - Normal pressure hydrocephalus

Comorbidity

- Another form of dementia, e.g. Alzheimer's dementia commonly co-occurs with vascular dementia
- Depression

Physical Exam

- Physical exam is not diagnostic but should be done to rule out diagnoses in differential
- Should include:
    - General physical exam
    - Neurological exam to look for signs of previous stroke and normal pressure hydrocephalus

Investigations

- Screening blood work to rule out other diagnoses:
    - CBC, electrolytes, extended electrolytes,
    - Fasting blood glucose
    - TSH
    - Vitamin B12 and folate levels
- Syphilis and HIV testing are not routinely done, but can be included if suggestive history present

Cognitive Testing

- There are multiple different cognitive tests available to diagnose dementia:

- **The Mini-Mental State Examination (MMSE) (9,10,11)**
    - <10 minutes by a trained interviewer
    - Validated in both primary care and specialist services
    - 11 items with a maximum score of 30

    - Cut-off score is 24
    - Lacks sensitivity in early dementia, frontotemporal dementia and dementia with Lewy bodies.
    - Does not examine executive functions and there are few episodic and semantic memory or visuospatial tasks
    - Often paired with clock drawing test

- **Clock Drawing Test (CDT)**
    - 1-3 minutes to perform
    - Validated in both primary care and specialist services
    - Employs a six-point rating of drawing
    - It does not differentiate between the subtypes of dementia
    - Multiple different scoring systems used
    - Fast to administer, may be less offensive than other cognitive tests

- **Montreal Cognitive Assessment (MoCA)**
    - 10 minutes by a trained interviewer
    - Validated in specialist services, not in primary care
    - Tests executive functioning and attention tasks with a maximum score of 30
    - Suggested cut-off is 26
    - More sensitive than MMSE at picking up mild cognitive impairment
    - Accurate in parkinson’s dementia

- **Mini-Cog**
    - 3 minutes to perform
    - Combines three-item word memory and clock drawer
    - Lower sensitivity than MMSE, but faster and easier to use, no training necessary
    - No prospective tests of its ability to detect dementia and no value in either disease progression or rating severity

Neuroimaging

- Structural imaging should be performed at least once during the dementia work up and can be used to exclude reversible causes
- CT head without contrast
    - Generally adequate to exclude normal pressure hydrocephalus and to assess for evidence of vascular disease
- Consider more extensive imaging such as MRI if:
    - Dementia onset less than 65 years of age
    - Sudden or rapid progression of symptoms
    - Presentation with a focal neurologic deficit
    - Experiencing a recent full or head trauma
- Reference: Darrow, 2015; NICE-SCIE Guideline, 2006.

Management in Primary Care: Non-Medication

- Non pharmacological management
    - Exercise and recreational activities are encouraged
    - Participation in programs aimed at cognitive stimulation
    - Link to support groups early for education
    - Monitor changes in function with ADL and iADL assessments
    - Regularly re-evaluate disease progression with above mentioned cognitive tests

- Environment and safety
- Assessment by home occupational therapy is often extremely beneficial
- Introduction of as much routine into daily activities as possible
- Evaluate driving ability and report, if necessary

Reference: Darrow, 2015.

Management in Primary Care: Medications

Pharmacological management

- Involve patient and caregivers in discussion of medication risks, benefits and side effects
- When introducing a medication, start at a low dose and increase it slowly over time
- Assessment of the continue use and benefit of any medication with cognitive and functional assessments are crucial
- While cholinesterase inhibitors are the mainstay treatment for alzheimer's dementia, it differs for the other subtypes of dementia
- Supplements, herbal products and other pharmacotherapy are not routinely recommended for dementia management

Types of Medications

- **Acetylcholinesterase inhibitors**
  - May temporarily stabilize dementia and behaviour
  - May slow rate of functional loss
  - Does not delay institutionalization
  - Not better than placebo for agitation
  - Adequate trial 6 months
  - Relative CI: bradycardia, sick sinus syndrome, active peptic ulcer disease, severe asthma/COPD, anticholinergic meds, Parkinson's disease, epilepsy and beta blockers
  - Taper over 1 week if discontinuing therapy
  - **Examples**
    - **Donepezil (Aricept)**
      - Starting dose 2.5mg to 5mg once daily
      - Dose can be reassessed and increased after 1 month
      - Max dose 10mg once daily
      - Side effects: nausea, diarrhea, insominia, fatigue, weight loss, cholinergic effects (incontinence, bradycardia, syncope, falls) and agitation
    - **Galantine (Reminyl)**
      - Starting dose 4mg to 8mg once daily
      - Dose can be reassessed and increased after 1 month
      - Max dose 24mg to 32mg once daily
      - Side effects: nausea, diarrhea, anorexia, wt loss, cholinergic effects (incontinence, bradycardia, syncope, falls
    - **Rivastigmine (Exelon)**
      - Starting dose 1.5mg BID
      - Dose can be reassessed and increased after 1 month
      - Max dose 6mg BID
      - Side effects: nausea, diarrhea, dizzinss, headache, weight loss, tremor cholinergic effects (incontinence, bradycardia, syncope, falls)

- **NMDA Antagonist**
  - Off label use for dementia with lewy bodies
  - Use with caution in patients with cardiovascular disease and seizure disorders

    - **Example**
        - **Memantine (Ebixa)**
            - Starting dose 5mg daily
            - Dose can be reassessed and increased after 1 week
            - Max dose 10mg BID (doses higher than 5mg should be in divided doses)
            - Side effects: nausea, diarrhea, dizzy, drowsiness, confusion, insomnia, headaches, hypertension

- **Vascular dementia**
    - Identify and treat vascular risk factors
    - Cholinesterase inhibitors are not recommended
    - Dementia with lewy bodies
    - Acetylcholinesterase inhibitors and memantine
    - Parkinson's disease associated with dementia
    - Consider acetylcholinesterase inhibitors, but not memantine

- **Frontotemporal dementia**
    - Not recommended and may even be harmful
        - Acetylcholinesterase inhibitors
        - Memantine
    - There is little evidence to suggest SSRIs or dopamine agonist therapy are of benefit and so guidelines advise against using them

Reference: Darrow, 2015; Gauthier, 2012; NICE-SCIE Guideline, 2006.

When to Refer

- If diagnosis is unclear, especially with the less commonly seen subtypes of dementia besides Alzheimer's
- If there is an unusual or atypical presentation
- If the onset of dementia is before the age of 60
- If there are overwhelming psychological or behavioural concerns that are difficult to manage

Who to Refer to

- Geriatrician to help with diagnosis and management
- Neurologist if overwhelming parkinsonisn features or focal neurologic signs and symptoms
- Geriatric psychiatrist to help with behavioural concerns such as agitation
- Comprehensive memory clinic for allied health support

Clinical Guidelines

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About this Document

Written by Nancy Kang, Family Medicine Resident, Class of 2016. Reviewed by members of the eMentalHealth.ca Primary Care Team, which includes Dr's M. St-Jean (family physician), E. Wooltorton (family physician), F. Motamedi (family physician), and M. Cheng (psychiatrist).

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