## Pathway for Recognition, Diagnosis, and Management of Early Dementia in Primary Care

1. Diagnostic criteria $\square$

- Mild Cognitive Impairment
- Early Dementia

2. Diagnostic algorithm $\square$
A. (i) Symptoms of cognitive impairment such as:
(ii) Areas of difficulty in ADLs/IADLs
(iii) Obtain collateral history from family and caregivers
B. Complete medical history
C. Cognitive assessment
D. Physical examination
E. Lab investigations
F. Head imaging not routinely necessary; request CT head if:
3. Contributory causes or differential diagnosis
4. When to consider referral to aid in diagnosis $\square$
5. Diagnosis disclosure $\square$
6. Management (General Care and Support) $\square$
> Management
> Non-Pharmacological Management of Dementia
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1. Diagnostic criteria

- Mild Cognitive Impairment

Diagnosis of mild cognitive impairment requires impairment:
i. in 1 of the following cognitive domains: memory, language, visuospatial, executive function and behaviour
ii. that does not significantly affect their usual activities or work
iii. that is not explained by delirium or other major psychiatric disorder

- Early Dementia


## Diagnosis of EARLY dementia requires impairment:

i. in at least 2 of the following cognitive domains: memory, language, visuospatial, executive function, and behaviour*
ii. that causes a functional decline in usual activities or work
iii. that is not explained by delirium or other major psychiatric disorder
*Typical behaviours in early dementia include irritability, social withdrawal and impaired self-esteem.
2. Diagnostic algorithm
A. (i) Symptoms of cognitive impairment such as:
$\square$

1) missed office appointments
2) frequent or inappropriate calls to the office
3) medication issues: compliance or adherence problems, errors
4) repetitive in conversation
5) repeatedly misplacing items
6) unable to recall treatment instructions from prior visits
7) defers to family members in answering questions (consider possible hearing impairment or language barriers also)
8) presents with signs of declined self-care (e.g.: poor hygiene, grooming, unexplained weight loss)
9) frequent visits to the emergency room
10) late life depression or anxiety
11) motor vehicle accidents, fender benders or wayfinding problems
(ii) Areas of difficulty in ADLs/IADLs
(iii) Obtain collateral history from family and caregivers
B. Complete medical history
include:
i. comorbidities (ie. REM sleep disorder, sleep apnea)
ii. medication list (assess for polypharmacy)
iii. alcohol and illicit drug use/smoking
iv. education and employment history
v. family history

## C. Cognitive assessment

i. MoCA - preferential in MCI/early dementia. Further info - mocatest.org
ii. SMMSE (quicker alternative if cannot complete MoCA; however conducting a MoCA test before giving a diagnosis is best practice)
iii. clock drawing test (acts as a screening test; conduct when time is restricted but signs of memory loss are apparent)
iv. Consider alternate tests in South Asian or others with language and/or educational barriers, as standard tests may have limited diagnostic value e.g translated MoCA, RUDAS and instructions

Language and cultural differences, educational level, and health literacy are major challenges for many ethnic groups impacting the administration of cognitive assessments, and subsequently the diagnosis. Whenever applicable, use translated resources such as the MoCA test and the accompanying instructions available in 50+ languages online (www.mocatest.org) or administer tests in the presence of an interpreter. Translated MoCA tests: Punjabi, Hindi, Urdu, Arabic, Tamil, Telugu, Bengali, and more on mocatest.org; Days of a week (Punjabi), Days of a week (Hindi), months of a year (Punjabi \& Hindi).

## D. Physical examination

i. rule out visual or hearing deficits - Blind MoCA
ii. assess for atypical clinical signs that raise red flag of alternate diagnosis and referral to specialist - e.g. parkinsonian features, focal neuro deficits, gait issues
iii. assess for cardio/cerebrovascular disease

## E. Lab investigations

## Advised tests to rule out reversible causes of cognitive change and establish baseline

i. rule out remedial contributory causes medical illness

1) Complete Blood Count (CBC)
2) $B 12$
3) Urinalysis
4) Glucose - fasting
5) Hemoglobin A1c
6) TSH
7) Sodium
8) Albumin/Calcium
9) Creatinine/eGFR
10)ECG
ii. In patients with risk factors, check:
10) Liver enzymes
11) Syphilis
12) HIV
13) Drug levels (e.g.: digoxin, phenytoin)
F. Head imaging not routinely necessary; request CT head if:

- < 60 yrs
- Abrupt onset
- Rapid progression
- Recent head injury
- History of cancer (especially breast and lung)
- Suggestion of stroke
- Any localizing neurological sign or symptom,
- Patient is on anticoagulation or has a bleeding disorder

3. Contributory causes or differential diagnosis
i. depression - GDS15, $\underline{\text { PHQ, comparative features vs. dementia or }}$ delirium
ii. delirium (BC Guidelines - Delirium Screening and Assessment Tools)
iii. alcohol dependence
iv. adverse drug effects and polypharmacy e.g. narcotics, benzodiazepine's, HS sedation
v. co-morbid disease, including sleep apnoea
vi. Anticholinergic burden
4. When to consider referral to aid in diagnosis

Consider referral to aid in diagnosis when:

- Abnormal imaging
- Abnormal neurologic exam, focal neurological signs or symptoms:
- Urinary incontinence, gait dysfunction - consider normal pressure hydrocephalus (NPH)
- Rigidity, bradykinesia, postural instability, tremor - consider Parkinson's Disease Dementia (PDD)
- Early falls, eye movement abnormalities - consider Progressive Supranuclear Palsy (PSP)
- Early psychosis and REM sleep disorders - consider Dementia with Lewy Bodies (DLB)
- Rapid progression
- Abrupt onset
- < 60 yrs

5. Diagnosis disclosure $\square$
Diagnosis of MCI or early dementia should be disclosed to the patient as soon as possible, preferably with a family member or caregiver present. To minimize stress, the timing and extent of disclosure should be individualized and carried out over several visits.

## 6. Management (General Care and Support)

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## > Management

The following MANAGEMENT information is abstracted and adapted from the BC Guidelines Cognitive Impairment: Recognition, Diagnosis and Management in Primary Care (2016).

- Establish a register of patients.
- Reassess cognition and function at planned visits.
- Involve the patient and caregiver in setting goals. See Associated Document: Clinical Action Plan (flow sheet); and
- Consider vaccinations, vitamin D supplementation, falls risk assessment, and exercise prescriptions. Refer to BCGuidelines.ca for other chronic disease guidelines which may be useful.


## Non-Pharmacological Management of Dementia

A. Memory

- Aids like calendars, diaries and telephone reminders;
- Keeping keys, glasses, wallet in same designated place ("landing spot");
- Accompaniment to appointments;
- Exercise; and
- Healthy brain games, e.g. Sudoku puzzles, crossword puzzles, games (cribbage), jigsaw puzzles and word searches.


## B. Medication Management

- Use blister packages/dossette trays and suggest caregiver supervision to improve safety and compliance or medication daily dispensing through local pharmacies;
- Medication monitoring through Home \& Community Care.


## C. Household Safety

- Monitor kitchen for mishaps (e.g., fires, burned pots); have stove unplugged or automatic stove turn-off device installed;
- Functioning smoke detectors;
- Assess home for other safety hazards (e.g., unsafe smoking, firearms in the home);
- Check for spoiled food in the fridge;
- 911 stickers for telephones;
- A personal alarm service in case of patient accident; and
- Referral for home assessment through Home \& Community Care.


## D. Shopping

- Use of lists when shopping;
- Shopping assistance from caregiver; and
- Use of shop by phone programs, if available.


## E. Behavioural Symptoms

- Common early dementia behavioral manifestations include changes in mood or personality, such as becoming confused, suspicious, depressed, fearful or anxious. They may be easily upset at home, at work, with friends or in places where they are out of their comfort zone and feel loss of control. Education of the patient and family is extremely important in management of behavioural symptoms of early dementia.
- Carrying identification when out alone; use of an ID bracelet or registering with the MedicAlert ${ }^{\circledR}$ Safely Home ${ }^{\circledR}$ Program.


## Seven Numbers for Early Dementia

## F. Nutrition

- Monitor for weight loss; and
- Meal support services (e.g., healthy delivered prepared meals or pre-prepared frozen foods).


## G. Socialization

- Awareness that patients with dementia may become socially withdrawn; and
- Referral to First Link ${ }^{\circledR}$ and Alzheimer Society for their education, services, support and contact with resources

1. Ask the individual or family member for permission to forward their name to First Link ${ }^{\circledR}$.
2. Complete a referral form and fax it to:

- Alzheimer Society of B.C (604-669-6907).

Contact will be made within five business days or after three weeks for those who are adjusting to the diagnosis. You will receive a confirmation note when contact has been made.
First Link ${ }^{\circledR}$ offers your patients/clients:

- Proactive contact and follow up.
- Ongoing access to learning opportunities and support.
- Connection to a supportive community of others living with dementia.
- Knowledge, confidence and skills to live well with the challenges of dementia.


## Seven Numbers for Early Dementia

## H. Financial \& Legal Issues

- Discuss advance care planning as early as possible (e.g., refer to My Voice Advance Care Planning Guide for aid in discussing sensitive topics like tube feeding. Available in English, Punjabi and Simplified Chinese. See also № Cardiopulmonary Resuscitation form); and
- Encourage patient to have an up-to-date will, a power of attorney agreement for financial management, a representation agreement for health management and/or an advance directive.


## I. Driving

Dementia is a medical condition that impacts fitness for driving (see BC Driver Fitness Handbook for Medical Professionals);
Start early to engage the patient on the topic of driving as part of their future planning. Discussion with patient about the importance of memory and cognitive skills for driving and signals/ concerns about driving safety.
If there are concerns about a patient's functional ability to drive, consider referral to RoadSafetyBC to have their skills assessed;
Under Section 230 of the Motor Vehicle Act, a primary care provider must report to RoadSafetyBC if a patient:

- has a medical condition that makes it dangerous to the patient or to the public for the patient to drive a motor vehicle; and
- continues to drive after being warned of the danger.

Revoking a driver's license is not the health care provider's responsibility, but when significant deficits are seen reporting it is part of our duty. (UBC Dementia Management in Family Practice Facilitator Syllabus, used with permission).

To supplement or replace driving encourage patient to register with HandyDart and TaxiSavers (see Guide for Patients and Caregivers).

For patients who need assistance with the HandyDart and HandyCard application form, direct them to the HandyDART \& HandyCard: Simplified Form Instructions.

## J. Mental Health and Speciality Services

- Be aware that dementia may co-exist with other complex mental health conditions;
- Involve mental health teams and resources, such as Community Mental Health Services, to help in distinguishing depression from dementia, and assessing and treating significant behavioural problems and managing caregiver stress; and
- Involve allied health professionals (e.g., Home \& Community Care case managers, mental health teams, counsellors, pharmacists, occupational therapists, physiotherapists, dietitians).


## K. Caregiver Support

- Discuss needs, coping strategies, support system and stress management with caregiver;
- Aid in co-ordination, communication, planning, education and connecting with resources; and
- Assess the caregiver for caregiver burden and burn out. Monitor and check in with caregiver with each visit.


## Pharmacological Management of Dementia

The following information has been abstracted from the UBC Dementia Management in Family Practice Facilitator Syllabus, used with permission, and the Ministry of Health's Cognitive Impairment: Recognition, Diagnosis and Management in Primary Care.

The use of acetylcholinesterase inhibitors/memantine is controversial. While data from clinical trials report statistical evidence of benefit, clinical benefits are unclear. It should be noted that drugs may benefit only a small minority of patients, and the evidence for long term use is insufficient. Short term benefits (6-12 months) may include cognitive, functional, and global improvement. However, patients and their caregivers should be advised that benefits are limited, and that side effects and drug interactions are common. End points for discontinuation of medication should be discussed.

## Pharmacotherapy in Early Dementia

The most commonly prescribed drugs for Alzheimer's disease and some of the other dementias are known as Acetylcholinesterase inhibitors (AChEIs). They inhibit an enzyme in the brain thereby increasing neurotransmitter acetylcholine. There are three such drugs on the market:

- Donepezil (or Aricept ${ }^{\text {TM }}$ )
- Galantamine (or Reminy ${ }^{1 T M}$ )
- Rivastigmine (or Exelon ${ }^{\text {TM }}$ ). Comes in pills and in transdermal patches (patches are not normally covered by Pharmacare but might be in special case appeals).
- AChEIs are approved for the symptomatic treatment of mild to moderate Alzheimer's, with Donepezil being the only AChEI indicated for severe Alzheimer's disease.
- Shown in RCTs and meta-analyses to:
- Decrease cognitive decline
- Improve or maintain ADL function
- Improve behavior
- Improve overall subjective clinical ratings
- Possibly delay institutionalization.
- Benefits are seen in those with Alzheimer's Dementia, Vascular Dementia, Mixed AD/VaD, Parkinson's Dementia, Diffuse Lewy Body Dementia.
- Not seen to benefit those with other forms of dementia (such as Frontotemporal Dementia, Alcohol Dementia, NormoPressure Hydrocephalus, or more rare forms of dementia).
- Under current BC Special Authority coverage, they must have:
- SMMSE score of $\geq 10$ and $\leq 26$ AND
- Global Deterioration Score (GDS) of $\geq 4$ and $\leq 6$ AND
- Diagnosis to include a component of Alzheimer's Disease.
- Donepezil must be trialed first, with galantamine and rivastigmine (capsules only) approved if the patient is intolerant to donezepil.
- Patient must be registered with Fair Pharmacare and meet the specified requirements for coverage (eg: certain income level, meet the deductible, applicable limits in place).
- NOTE: New BC Special Authority Rules (April 2016) can be found here: http://www2.gov.bc.ca/assets/gov/health/forms/5465fil.pdf
*The medications section was current as of September 2017 and is subject to change. For updated information including prescribing and Pharmacare, please view the Ministry of Health website


## - Considerations for Pharmacotherapy

- Goal of treatment - slowing down cognitive, social and functional decline
- Not curative and improvement on cognitive testing is not expected
- If patient stops and restarts, benefits are not regained

Decision to initiate AChEI therapy requires an individualized patient assessment, involving the patient and caregivers in the following discussion points:

- Clinician, patient, and caregiver expectations of benefit with AChEI therapy.
- Presence of comorbidities and life expectancy.
- Potential drug interactions with concurrent medications.
- Ability of the patient or caregiver to adhere to pharmacotherapy.
- Potential benefits as compared to potential harms of AChEI therapy.
- Patient and caregiver preferences, including cost of therapy.
*The medications section was current as of September 2017 and is subject to change. For updated information including prescribing and Pharmacare, please view the Ministry of Health website.


## Relative Contradictions

- Severe Cardiac disease (CAD, CHF)
- Cardiac conduction abnormalities (slows AV nodal conduction)
- Pulse check for bradycardia and/or ECG prior to start and soon after start of these meds
- Severe respiratory disease (increases bronchospasm)
- Recent peptic ulcer disease or increased risk of same (increases gastric acid secretion)
- Significant GI symptoms or weight loss (most common side effects are nausea, diarrhea, decreased appetite and weight loss). GI intolerance rate is quite high.
- Severe renal or liver disease
- Seizure disorder or history of seizures (lowers seizure threshold)
- Obstructive Urinary disease (may worsen symptoms)
- Certain medications (see BC Guidelines)
- Many drug interactions exist through the cytochrome (CYP) liver enzyme system.
- Important to consider drug interactions with additive potential to cause bradycardia (B-blockers, verapamil, diltiazem, digoxin and other antiarrhythmics).
- Make sure no drugs on board with opposing anticholinergic pharmacology.
- There are some advantages and disadvantages that come with starting AChEIs later in the progression of dementia:
- Advantages: cost savings, side effects avoided
- Disadvantages: have lost ground already (have missed the opportunity to maintain the patient at a potentially higher level of cognition and functioning for longer)
- NOTE: When increasing the dose or tapering someone off the medications, it is important to do so slowly (dose change every 4-6 weeks)

Further information: BC Pharmacare Cholinesterase Inhibitors Info Sheet 2016

It is worthy to note:

- Medications are not indicated for Mild Cognitive Impairment (MCI)
- Behavioural and Psychological Symptoms occurring in early dementia (confusion, suspiciousness, depression, fear or anxiety) should be treated using a combination of behaviour or environment modification, together with directed pharmacotherapy (e.g. antidepressant for concurrent depression)
*The medications section was current as of September 2017 and is subject to change. For updated information including prescribing and Pharmacare, please view the Ministry of Health website


## Cognitive Impairment in Culturally and Linguistically Diverse Groups

The assessment and management of cognitive impairment in diverse individuals can be challenging for several reasons:

Communication difficulties, cultural factors, low education, and literacy impact formal cognitive screening, with poor inter-rater reliability - use interpreter services (where available) and translated/adapted cognitive assessment tools to assist in more accurate patient screening and assessment; Whenever applicable, use translated resources such as the MoCA test and the accompanying instructions available in 50+ languages online (www.mocatest.org). Translated MoCA tests: Punjabi, Hindi, Urdu, Arabic, Tamil, Telugu, Bengali, and more on mocatest.org; Days of a week (Punjabi), Days of a week (Hindi), months of a year (Punjabi \& Hindi).

Dementia symptoms may be unfamiliar or viewed as a normal part of the aging process, and there may be stigma to mental health issues, resulting in diagnosis delay - provide culturally sensitive patient information on dementia to patients and families;

Language barriers may result in a lack of awareness of community supports provide Guide for Patients and Caregivers;

Community supports may not provide culturally appropriate care, resulting in lack of adoption of these services and increase in caregiver stress; and

Families may share caregiver responsibilities by rotating the residence of the patient amongst family members - this is generally discouraged as it confuses the patient with dementia and complicates the provision of services between the staff of many agencies and the extended family. One familiar, safe and secure environment is encouraged.

## Resources

Make an early referral to support resources:

- Alzheimer Society of BC
- First Link Dementia Helpline
- HandyDart
- HandyDart form
- HandyDart Simplified Instructions
- BC211
- Better at Home
- HealthLink BC
- Fraser Health Crisis Line
- Seven Numbers for Early Dementia
- Keep your Brain Healthy brochure
- MedicAlert
- Lifeline
- Get up \& Go
- Home Health


## Serial Appointment Outline

## Serial Appointment Outline ( 3 visits suggested, can extend to more)

Consider use of GPSC code(s) (Current as of September 2018):
GP Annual Complex Care Management Fee (2 Diagnoses) -> 14033
GP Mental Health Planning fees -> 14043
GP Mental Health Management fee -> 14046-8
GP GPSC Portal Code -> 14070
GP Frailty Complex Care Planning and Management -> 14075
GP-Patient Telephone Management Fee -> 14076
GP-Allied Care Provider Conference Fee -> 14077
GP Email/Text/Telephone Advice Relay Fee -> 14078

## Visit 1:

- Obtain presenting History
- General physical exam
- Labs to rule out reversible
- Remind patient to bring historian/family/friend to next appointment
- Remind patient to bring all medications to next appointment (including over the counter)


## Visit 2:

- History and corroborative history
- Medications review
- Lab result review
- Dementia specific physical exam
- Cognitive testing (MD or other)
- Establish likely diagnosis


## Visit 3:

- Disclosure
- Referral to specialist if necessary
- Connection to resources
- Power of Attorney POA/Representation agreement
- Driving considerations

Appendix 1: Clinical Relevance of Physical Signs on Neurologic Exam in a Patient With Cognitive Impairment

| Physical Finding | Clinical Relevance |
| :--- | :--- |
| Mildly stooped posture, difficulty <br> with tandem gait | Can be seen in normal aging |
| Normal neurologic exam | Typical in early Alzheimer's disease (AD) <br> Consider space occupying lesion (tumor, <br> subdural hematoma, stroke) |
| Msymmetric neurologic deficits (NPH) (may |  |
| Magnetic gait | Normal pressure hydrocephalus (NPH) <br> also have urinary incontinence) <br> NPH, vascular dementia, Wernicke-Korsakoff <br> syndrome (usually secondary to chronic alcohol <br> use), cerebellar tumor, paraneoplastic disease, <br> spinocerebellar ataxia (SCA) |
| Wide based unsteady gait | Dementia with Lewy bodies (DLB), Parkinson's <br> disease, progressive supranuclear palsy (PSP), <br> corticobasal syndrome (CBS), multiple systems <br> atrophy (MSA), vascular dementia, <br> frontotemporal dementia (FTD), Creutzfeldt- <br> Jakob disease (CJD), Wilson's disease, <br> Huntington's disease (HD) |
| Parkinsonism signs (slowness, <br> stiffness, postural instability, <br> tremor) | HD, inherited metabolic disorders including <br> Wilson's disease, CJD (expect rapidly <br> progressive course), CBS, systemic lupus <br> erythematosis (SLE) |
| Involuntary hyperkinetic <br> movements (chorea, dystonia) |  |
| Myoclonus | Post-anoxia, CDD, AD, myoclonic epilepsies, <br> Hashimoto's encephalopathy, DLB, CBS |
| Pyramidal signs | Motor neuron disease, CDD, B12 deficiency, <br> multiple sclerosis (MS), SCA, MSA, <br> hydrocephalus, AD, FTD |
| Papilloedema | Tumour, sub,ural haematoma, hydrocephalus <br> Vascular disease, AD, CJD |
| Cortical blindness |  |

## Appendix 1: Clinical Relevance of Physical Signs on Neurologic Exam in a Patient With Cognitive Impairment

| Physical Finding | Clinical Relevance |
| :--- | :--- |
| Anosmia | Subfrontal meningioma, head injury, AD, PD, HD |
| Abnormal eye movements | PSP, CBS, Wernicke-Korsakoff, cerebellar tumours, <br> causes of raised intracranial pressure, CJD, HD |
| Visual field defect | Tumour, vascular disease, CJD |
| Pupillary abnormalities <br> (Argyll Robertson pupil) | Neurosyphilis |
| Peripheral neuropathy | Concurrent medical illness such as diabetes, vitamin <br> B12 deficiency, paraneoplastic disorders, SCA, SLE |
| Early onset incontinence | Tumour, hydrocephalus (including NPH), PSP |$|$| Fulbar features | Frontal dementia (motor neuron disease), PSP |
| :--- | :--- |

Appendix 2: Neurologic Exam Findings in Different Causes of Cognitive Impairment

| Type of Cognitive Impairment | Exam Finding |
| :---: | :---: |
| Normal aging | decreased large fiber sensation in toes decreased or absent ankle reflexes mildly stooped posture difficulty with tandem gait |
| Alzheimer's disease (AD) | usually a normal neurologic exam in amnestic type of $A D$ <br> in later stage, can see mild parkinsonism |
| Dementia with Lewy Body | parkinsonism (slowing, stiffness, postural instability) <br> absence of rest tremor |
| Normal pressure hydrocephalus | magnetic gait, urinary incontinence |
| Vascular dementia | slow gait with short steps and upright posture |
| Alcohol-related dementia | Ophthalmoparesis gait ataxia |
| Parkinson's disease | Slowness <br> Rigidity stooped posture slow, shuffling gait masked face quiet speech small writing |
| Corticobasal degeneration | Parkinsonism <br> Apraxia <br> dystonia, myoclonus <br> alien-limb phenomenon <br> cortical sensory loss <br> may present with aphasia, executive dysfunction <br> and mimic $A D$ |

Appendix 2: Neurologic Exam Findings in Different Causes of Cognitive Impairment

| Type of Cognitive Impairment | Exam Finding |
| :--- | :--- |
| Progressive supranuclear palsy | Parkinsonism <br> eye movement abnormalities (may not appear <br> until later in course) <br> axial rigidity, retrocollis <br> postural instability early in course, often fall <br> back <br> bulbar features (impaired speech, swallowing) |
| Creuzfeldt-Jakob disease | subacute to acute progression <br> extrapyramidal signs (myoclonus) <br> ataxia <br> seizures |
| Modified from: | Continuum, Volume 19, Number 2, April 2013, <br> p407 |

