

Education

Éducation

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Canadian Consensus Conference on Dementia: a physician's guide to using the recommendations

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ementia affects 8% of all individuals over the age of 65. More than a quarter of a million elderly Canadians now have this condition, and by the year 2031 this number will rise to more than 750 000. Dementing diseases impose a substantial burden of suffering on patients and their caregivers. Numerous recent advances have furthered our knowledge of the epidemiology, phenomenology and management of dementing diseases. For these reasons, the Canadian Consensus Conference on Dementia was established to develop a series of consensus statements to serve as a basis for clinical practice guidelines for the recognition, assessment and management of dementing disorders. These guidelines are directed toward primary care physicians, who provide most of the care for people with dementia. The recommendations from the Canadian Consensus Conference on Dementia, which are based upon the best available evidence, appear as a supplement to this issue of the CMA7.² A strategy is now being developed to disseminate them widely and to evaluate their impact. Because the recommendations were developed by a multidisciplinary panel using a consensus process, they sometimes offer broad direction rather than specific advice. This article is intended to highlight some of our conclusions and to detail how physicians can incorporate these recommendations into their practices.

Scenario 1: Early dementia

Patient profile

A 78-year-old widow who lives alone and whom you have seen infrequently is brought to your office by her daughter. Although the patient has no complaints, her daughter indicates that for the past 2 years she has become more forgetful. Her behaviour is repetitive, and she sometimes calls her daughter several times a day to ask the same question. The quality of her housework is beginning to decline (her house is untidy, food is left to spoil in the refrigerator, she is limiting food preparation to simple, familiar items, and she has to check recipes even for easy dishes). Her personal hygiene is also declining, and some bills are not being paid on time.

What should be your approach? What tests should be ordered? Is CT necessary? What management would you recommend?

Approach

This patient's history is suggestive of dementia. Dementia is defined most simply as acquired cognitive deficits sufficient to interfere with social or occupational functioning without depression or clouding of consciousness. When symptoms of this nature are reported by family members, they should be taken seriously, and careful cognitive assessment is indicated (recommendation 10). For this patient, you will need to obtain a more detailed history, with corroboration from her daughter, other family members or friends. You should determine whether there have been any behaviour or personality changes and whether there are symptoms of depression or physical illness. Medications, including proprietary and over-the-counter drugs, should be reviewed (for example, many antihistamines and cold remedies have anticholinergic side effects that can cause confusion). Risk factors



for dementia include a family history of the condition, previous head injury, limited educational level, excessive consumption of alcohol, presence of other neurological diseases (e.g., Parkinson's disease or stroke), and vascular risk factors such as hypertension, diabetes mellitus and atrial fibrillation. You should inquire about the patient's ability to perform both instrumental daily activities (e.g., using the telephone, shopping, taking medications, managing finances and driving) and basic daily activities (e.g., walking, bathing, continence, transfers, dressing and feeding) (recommendation 1). You should also ask about hallucinations or delusions and look for any evidence of depression or delirium. Inquiries should cover neurological symptoms, such as headache, focal weakness, sensory loss, and changes in gait or balance. A physical examination should be performed to reveal evidence of any systemic condition (e.g., malnutrition, hypothyroidism or chronic infection). The neurological examination should target focal abnormalities (e.g., hemiparesis, primitive reflexes and peripheral neuropathy) and should rule out papilledema.

A mental status examination should then be performed to examine memory, language, praxis (e.g., use of a comb or toothbrush), insight and judgement, abstract thinking, attention and knowledge of current affairs. Short mental status questionnaires such as Folstein's Mini Mental State Examination are helpful, but they do not cover important areas such as insight and judgement. If it is not possible to complete the history, physical examination and mental state examination during one visit, it is not only acceptable but also helpful to assess the patient over several visits (recommendation 1). Dementia is a clinical diagnosis based on history and examination, and these investigations help to identify aggravating and potentially reversible conditions contributing to the dementia.

For all patients in whom dementia is suspected, basic laboratory tests should be ordered, including complete blood count, thyroid-stimulating hormone, and serum levels of electrolytes, calcium and glucose (recommendation 2). CT is recommended only if there are features that suggest a diagnosis other than Alzheimer's disease, which is the commonest cause of dementia and the most likely reason for the decline in the patient described in the scenario. CT is recommended if any of the conditions listed in Table 1 are present (recommendation 3); otherwise, the yield of findings that would change management is extremely low.6 Because this patient has none of these features, neuroimaging is not indicated. Specific clinical features may direct further laboratory tests (e.g., pallor, smooth tongue and loss of sensation in the dorsal column should prompt the measurement of serum B12). Some "atypical features" that might suggest a diagnosis other than Alzheimer's disease include hallucinations or delusions occurring early in the course of the dementia, rigidity or parkinsonism, features that may suggest dementia with Lewy bodies; incontinence and apraxic gait, which may suggest normal-pressure hydrocephalus; and aphasia early in the course of the condition or marked changes in behaviour (such as apathy, disinhibition and aggression), which may suggest frontotemporal dementia.

Management

The most likely diagnosis in this patient is Alzheimer's disease. Initial management should include discussion of the diagnosis (recommendation 2) and advice about future planning such as assignment of power of attorney and advance directives. The patient's daughter should be asked to watch for safety concerns (e.g., leaving pots to burn on the stove, being unable to summon help by telephone or other means, wandering outside while inappropriately clothed or becoming lost). Help with homemaking may be required. The daughter should be encouraged to contact the local chapter of the Alzheimer Society, (recommendation 32), which can provide support, educational resources and advice, vitally important aspects of management. Patients and their relatives often inquire about vitamin E and Gingko biloba (recommendations 47 and 48). These compounds are of questionable efficacy, but they are inexpensive and appear relatively harmless. Although most of the experts involved in the Canadian Consensus Conference on Dementia did not feel that there was sufficient evidence to recommend vitamin E, some suggest a daily dose of 2000 IU (see dissenting opinion).

The only medication currently approved for use in Canada for cognitive enhancement in mild to moderate Alzheimer's disease is donepezil (recommendation 46). This drug is moderately effective and relatively safe. Caution is necessary in those with a history of bronchospasm, seizures, peptic ulcer or bradycardia. The most frequent side effects are gastrointestinal in nature and may include anorexia and weight loss. If this drug is to be prescribed (and in this case it would be a reasonable choice, provided adherence to the once-daily dosing could be assured), it is strongly recommended that baseline measures (such as the

Table 1: Criteria for performing cranial CT in patients with dementia*

Age < 60 yr

Rapid (e.g., over 1–2 mo), unexplained decline in cognition or function

Dementia of relatively short duration (< 2 yr)

Recent, significant head trauma

Unexplained neurologic symptoms (e.g., new onset of severe headache or seizures)

History of cancer, especially of a type or at a site associated with metastasis to the brain

Use of anticoagulants or history of bleeding disorder

History of urinary incontinence and gait disorder early in the course of dementia (as may be found in normal-pressure hydrocephalus)

Presence of any new localizing sign (e.g., hemiparesis or Babinski reflex)

Unusual or atypical cognitive symptoms or presentation (e.g., progressive aphasia)

Gait disturbance

*CT is recommended if one or more of these criteria are present.



Mini Mental State Examination and an assessment of instrumental activities of daily living) be obtained and recorded. A diary of activities and other observations should be initiated by the caregiver (recommendation 45d). These assessments should be repeated later to determine whether there is any response to donepezil; a response may take up to 3 months. The initial dosage is usually 5 mg daily, and if the drug is tolerated the dosage is increased to 10 mg daily. If there is no observable improvement at 3 months, the drug may be stopped. If a clinically useful response occurs, it is uncertain how long the drug should be continued. Without clear evidence, some physicians feel that a lack of deterioration over several months represents a positive response. Provincial drug plans do not currently reimburse the cost of donepezil.

Regular review of this woman's progress is essential. The natural history of Alzheimer's disease from diagnosis to death is reportedly about 7 years. As her function declines there will probably come a time when she will require additional assistance in the home, and she may eventually require institutional care as the later complications of dementia supervene (e.g., inability to care for herself, incontinence, behavioural disturbances and immobility). It will also be important to monitor the daughter's ability to cope with her mother's illness, to provide support and to look for signs of excessive caregiver stress and depression (recommendations 27, 29, 31).

Scenario 2: Early behavioural change

Patient profile

A 67-year-old man is new to your practice. He is of eastern European descent and worked in a factory until his retirement at age 64. In recent months his behaviour has changed. He has become both verbally and physically aggressive toward his wife. Although previously polite and well mannered, he is now disinhibited, frequently insulting family members and other visitors to the house. He was cautioned by local police after being suspected of making an inappropriate approach toward a teenage girl. His driving abilities have deteriorated, and he has had several "near misses." There is no history of psychiatric disturbance, and he has no headaches or other neurological symptoms. He does not believe he has a problem.

How should you approach this man? Are there any specific treatments that are likely to help him?

Approach

The initial approach should be as in scenario 1 and should include taking a history and performing a physical examination. The physical examination reveals that he is overweight, and he exhibits frontal lobe release signs (snout and grasp reflexes). The mental status examination reveals that the patient has difficulty naming less common objects (such as the bracelet on his watch and the collar on his

shirt), although his memory is relatively well preserved. You proceed with the basic laboratory tests and, because of the "atypical" features (i.e., early behavioural change, preserved memory, frontal lobe release signs), you arrange for him to undergo CT, which shows cerebral atrophy, more marked in the frontal regions. In the absence of any evidence of tumour (e.g., frontal meningioma) or primary psychiatric disease, it is likely that this man has a frontotemporal dementia. Although the exact prevalence of frontotemporal dementias is unknown, these conditions probably constitute less than 10% of all cases of dementia.

Management

Management in this case may prove difficult as the disease progresses and the patient's behaviour deteriorates. If you are uncertain about the diagnosis or management, it may be necessary to refer the patient to a specialist (e.g., a geriatrician, geriatric psychiatrist, behavioural neurologist or psychologist) (recommendation 5) (see Table 2).

The difficult issue of safety in driving should be addressed (recommendation 22). Office assessment of driving ability is notoriously inaccurate, and because driving is intimately entwined with autonomy, it is always a touchy issue (recommendation 24). Although the "gold standard" for driving safety is an on-road, performance-based driving test, there is fair evidence that this patient's driving is becoming unsafe. In most provinces, this concern must be reported to the provincial ministry of transport (recommendation 25).

Nonpharmacological forms of therapy should be considered for the management of this patient's behaviour. If medications are required for agitation, aggression or psychotic behaviour, low doses of neuroleptic drugs (especially those with less potential to produce extrapyramidal side effects), selective serotonin reuptake inhibitors (SSRIs) or trazodone should be considered (recommendation 42).

Assessment of this patient's safety and the risk to his caregivers is important in this case, because lack of insight and disinhibition may lead to potentially dangerous behaviours. Personal safety may be jeopardized, and aggressive outbursts may result. Assessment of risk and contingency plans must be established. The patient's family will need

Table 2: Reasons to consider referral to specialists or other professionals

Continuing uncertainty about diagnosis after initial assessment and follow-up

Request by the patient or the family for another opinion

Presence of significant depression, especially if there is no response to treatment

Problems with or failure of treatment with new medications specific for Alzheimer's disease

Need for additional help in managing the patient (e.g., behavioural problems) or supporting the caregiver (e.g., home care programs, Alzheimer Society)

When indicated for genetic counselling

Research studies into diagnosis or treatment are being carried out



considerable support, particularly given that it is often difficult for family members to appreciate that these types of behaviour have an organic basis. Assistance from other professionals (e.g., social workers) and support groups, especially the Alzheimer Society, is often helpful.

Scenario 3: Early hallucinations

Patient profile

Your 83-year-old patient resides with his wife in a retirement home. He is in good physical health, although he has a history of prostate carcinoma and became confused after transurethral radical prostatectomy 3 years ago. During a routine visit to your office, his wife asks to have a private word with you. She reports that he has been "seeing things." She describes well-formed visual hallucinations. He has seen animals in their apartment and thought that the Municipal Works Department was digging up the road outside. These hallucinations have been present for several months but now occur almost daily. He has also accused her of being unfaithful and sometimes thinks that the local newspaper is unduly critical of his religious beliefs. What should be your approach?

Approach

When you interview this patient, he downplays the hallucinations but remains convinced that his wife is having an affair. You review the usual causes of hallucinations (medications, metabolic disturbance and visual problems) and confirm that there is no obvious underlying cause. Physical examination reveals only minimal rigidity at the wrists and a subtle slowness of gait. Examination of mental status reveals a mild loss of immediate recall and a little difficulty in drawing a clock face. The results of laboratory studies are negative, and you consider whether to initiate treatment or refer the patient to a psychiatrist.

When you see the couple again 2 weeks later, his wife indicates that the hallucinations are becoming worse. She is very troubled about his delusion of infidelity. You decide to initiate treatment with a low dose of an antipsychotic drug and arrange for him to see a psychiatrist 3 weeks later. You are somewhat surprised to receive a telephone call from the psychiatrist, who states that the patient has become quite rigid and that his hallucinations have worsened.

This man probably suffers from dementia with Lewy bodies.² The full spectrum of this disease is currently being defined, although it is sometimes regarded as a variant of Alzheimer's disease. The characteristic features of dementia with Lewy bodies include visual hallucinations and delusions early in the course of the disease. Patients display marked fluctuation in cognitive performance and may appear delirious, but an exhaustive search for causes of delirium is usually unrewarding. Falls may occur without explanation and mild Parkinsonism is common. Many of these people are exquisitely sensitive to neuroleptic agents, which

can precipitate rigidity and bradykinesia. Management is often difficult, and specialist help is often advisable (recommendation 5). There is increasing evidence that the hallucinations that occur in this condition may respond to cholinergic manipulation. Although this condition is not an approved indication for donepezil, there is anecdotal evidence that this drug may be helpful for alleviating the hallucinations and improving cognitive performance. In some patients, however, donepezil may aggravate the parkinsonian features. If an antipsychotic agent is necessary, drugs with the least potential to produce extrapyramidal and anticholinergic side effects are recommended. These drugs include risperidone, olanzepine and quetiapine. Although the parkinsonian features may diminish with anti-parkinsonian medications, the hallucinations may worsen.

Scenario 4: Preventing dementia

Patient profile

A 47-year-old woman whom you have previously treated for anxiety and depression is worried because her mother (aged 76) has just been diagnosed with Alzheimer's disease. Your patient is concerned that she will be affected by the same condition. What are her chances of eventually having this disease, and can anything be done to prevent it?

Approach

The prevalence of dementia is related to age. Among people 65 years of age, the prevalence is only 1% or 2%, among those 75 years of age it is about 11% and among those 85 years of age it is about 40%. First-degree relatives of patients with Alzheimer's disease have a 2- to 4-fold increase in personal risk for the disease. For this woman, the first step is to obtain a family history. If her mother's diagnosis came late in life and there are no other affected family members, her own risk is only slightly greater than that of the general population, and she can be reassured. However, if there is a strong family history of early-onset disease, this woman may belong to one of the few pedigrees with autosomal dominant transmission. In this case, it would be appropriate to refer her to a regional genetic clinic for further testing (recommendation 14). Although the presence of the APOE*4 allele (producing the $\in 4$ type of apolipoprotein E) is associated with a higher risk of late-onset Alzheimer's disease, the sensitivity (approximately 50%) and specificity (approximately 75%) are insufficient to help in determining prognosis, and its determination is not recommended (recommendation 12).

In general, there is not currently sufficient evidence to recommend for or against screening for cognitive impairment or dementia in asymptomatic individuals (recommendations 6 and 7). In this case, as with any person with memory complaints, an appropriate cognitive assessment and periodic follow-up to assess progression are indicated (recommendation 9). Because this woman is at greater risk of Alzheimer's



disease because of the family history, it is especially important to address additional risk factors for dementia. This would include modification of vascular risk factors (e.g., smoking, hypertension and diabetes mellitus) (recommendation 17), given the mounting evidence that a stroke may "unmask" or exaggerate the clinical symptoms of Alzheimer's disease⁹ and that the treatment of hypertension reduces the incidence of not only vascular dementia but also Alzheimer's disease.¹⁰

There is suggestive evidence from case–control studies that postmenopausal hormone replacement therapy may reduce the risk of Alzheimer's disease. In counselling this woman about postmenopausal hormone replacement, you should discuss this aspect of such treatment (recommendation 20). There is no convincing evidence that vitamin E or other medications (e.g., NSAIDs) reduce the risk of dementia, although studies of this question are ongoing.

Scenario 5: Advanced Alzheimer's disease

Patient profile

The patient described in scenario 1 is now 83 years old and residing in a long-term care facility. She is completely disoriented as to time and place. She usually does not recognize her daughter and misidentifies other individuals, believing, for example, that members of the nursing staff are her relatives. She is dependent in all basic daily activities and has urinary but not bowel incontinence. She has recently become resistive to care, striking out when attempts are made to bathe her. She is verbally abusive to staff and other residents. She has struck out at other residents and visitors and often calls out repetitively in the evening.

You are asked by the staff to prescribe "something to improve her behaviour."

Approach and management

The first step is to evaluate the patient for potential triggers for the behavioural disturbance. Such triggers may include pain, intercurrent illness or medications. The next step is to document the behaviours, using the ABC approach: look for precipitants or Antecedents (e.g., physical treatments, bathing, mealtimes, company or loneliness) and describe the *Behavioural* disturbance and the *Consequences*. The act of observing and documenting these behavioural symptoms and signs can itself reduce the number of incidents, as staff learn to recognize, anticipate and avoid provocation.¹² Once the triggers for these behaviours have been discovered, the first line of management should involve behavioural measures, physical changes (e.g., light and sound) and avoidance of individuals who aggravate the situation (recommendation 41). If physical and behavioural measures are unsuccessful, the next step is to institute low doses of neuroleptics, SSRI antidepressants or trazodone (recommendation 42). There is little evidence that one drug or pharmacological group is more effective than others, although most published evidence is for neuroleptic agents. There is a growing trend toward the use of atypical neuroleptics such as risperidone.¹³ It is often necessary to try several drugs singly or in combination to achieve improvement in aggressive or agitated behaviour. Referral for specialist advice may be necessary (recommendation 5).

Conclusion

Although there is enormous variation in the types of practice, geographic locations and availability of specialist resources, we hope that the description of these scenarios is helpful to individual practitioners. We encourage readers to review the supplement for the other recommendations of the Canadian Consensus Conference on Dementia. We have highlighted the important role of the Alzheimer Society in support, education and advocacy. We hope that the work of this consensus conference will be helpful to primary care physicians in the management of their patients with dementia.

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References

- Canadian Study of Health and Aging Working Group. Canadian study of health and aging: study methods and prevalence of dementia. CMA7 1994;150:899-913.
- The recognition, assessment and management of dementing disorders: conclusions from the Canadian Consensus Conference on Dementia. CMAJ 1999;160(12 Suppl):S1-S20.
- American Psychiatric Association. Delirium, dementia and other cognitive disorders. In: *Diagnostic and statistical manual of mental disorders* (DSM IV). 4th ed. Washington: The Association; 1994. p. 134.
- 4. Some drugs which cause psychiatric symptoms. Med Lett 1998;40(1020):21-4.
- Folstein MR, Folstein SE, McHugh PR. "Mini-mental state": A practical method of grading the cognitive state of patients for the clinician. J Psychiatr Res 1975;12:189-98.
- Freter S, Bergman H, Gold S, Chertkow H, Clarfield AM. Prevalence of potentially reversible dementias and actual reversibility in a memory clinic cohort. CMA 7 1998;159:657-62.
- Heyman A, Peterson B, Fillenbaum G, Pieper C. The Consortium to Establish a Registry for Alzheimer's Disease (CERAD) part XIV: Demographic and clinicial predictors of survival in patients with Alzheimer's disease. *Neurology* 1996:46:656-60.
- Shea C, Macknight C, Rockwood K. Donepezil for treatment of dementia with Lewy bodies: a case series of nine patients. Int Psychogeriatr 1998;10(3):229-38.
- Snowdon DA, Greiner LH, Mortimer JA, Riley KP, Greiner PA, Markesberry WR. Brain infarction and the clinical expression of Alzheimer Disease. The Nun Study. JAMA 1997;277:813-7.
- Forette F, Seux M-L, Staessen JA, Thijs L, Birkenhäger WH, Babarskiene MR, et al. Prevention of dementia in randomised double-blind placebo-controlled Systolic Hypertension in Europe (Syst-Eur) trial. *Lancet* 1998;352:1347-51.
- Brenner DE, Kukull WA, Stergachis A, Van Belle G, Bowen JD, McCormick WC, et al. Postmenopausal estrogen replacement therapy and the risk of Alzheimer's disease: A population-based case-control study. Am J Epidemiol 1994;140:262-7.
- Nilsson K, Palmerstierna T, Wistedt B. Aggressive behaviour in hospitalized psychogeriatric patients. Acta Psychiatr Scand 1998;78:172-5.
- Katz IR, Jeste DV, Mintzer JE, Clyde C, Napolitano J, Brecher M. Comparison of risperidone and placebo for psychosis and behavioural disturbances associated with dementia a randomized, double-blind trial. J Clin Psychiatry 1999:60:107-15.

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