PharmaCare note: Cholinesterase inhibitors are currently available by Special Authority for the treatment of mild to moderate Alzheimer's disease only. PharmaCare will NOT be providing coverage of these medications for other types of dementia unrelated to Alzheimer's disease.

For reference purposes only, current diagnostic criteria for other types of dementia are presented below. Note that if the dementia etiology is mixed (e.g. Alzheimer's disease with comorbid cerebrovascular disease or Alzheimer's disease mixed with Lewy body pathology), patients may be eligible for coverage as long as the predominant etiology is Alzheimer's disease.

DIAGNOSTIC CRITERIA FOR DEMENTIAS

DSM-IV CRITERIA FOR ALZHEIMER'S DISEASE

Memory deficit that can be demonstrated objectively on cognitive testing.

At least one other cognitive deficit such as aphasia (abnormal speech), executive function impairment (difficulty with planning, judgment, mental flexibility, abstraction, problem-solving, etc), agnosia (impaired recognition of people or objects), or apraxia (impaired performance of learned motor skills).

Together, these cognitive deficits must result in impairment in performance of daily activities.

The course is characterized by gradual onset and continuing cognitive decline.

These deficits must represent a decline from a previous higher level of functioning.

There must not be any other neurological disease that accounts for them.

DIAGNOSTIC CRITERIA FOR OTHER TYPES OF DEMENTIA

NOT INCLUDED IN THE ALZHEIMER'S DRUG THERAPY INITIATIVE:

NINDS-AIREN CRITERIA FOR VASCULAR DEMENTIA

Dementia defined by cognitive decline from a previously higher level of functioning manifested by impairment of memory and of impairment in at least one other cognitive domain. Deficits should be severe enough to interfere with activities of daily living not due to the physical effects of stroke alone.

Cerebrovascular disease defined by the presence of focal signs on neurologic exam consistent with stroke (with or without history of stroke) AND evidence of relevant CVD by brain imaging (CT or MRI).

A relationship between the above two disorders manifested or inferred by the presence of one or more of the following: (a) onset of dementia within 3 months following a recognized stroke; (b) abrupt deterioration in cognitive functions; or (c) fluctuating, stepwise progression of cognitive deficits.

Clinical features consistent with the diagnosis of probably vascular dementia include:

(a) early presence of gait disturbance;
(b) history of unsteadiness and frequent, unprovoked falls;
(c) early urinary frequency, urgency, and other urinary symptoms not explained by urologic disease;
(d) pseudobulbar palsy;
(e) personality and mood changes, abulia, depression, emotional incontinence, or other subcortical deficits including psychomotor retardation and abnormal executive functions.

Editorial note: If a dementia has gradual onset and progression reminiscent of Alzheimer's disease but there is imaging evidence of ischemic lesions and perhaps some abnormalities on neurological examination, the diagnosis is most likely to be (predominant) Alzheimer's disease with comorbid cerebrovascular disease rather than pure vascular dementia.

LUND-MANCHESTER CRITERIA FOR FRONTOTEMPORAL DEMENTIA

The Lund-Manchester diagnostic criteria for frontotemporal dementia require all of the following core components to be present:

1. insidious onset and gradual progression
2. early decline in social interpersonal conduct
3. early impairment in regulation of personal conduct
4. early emotional blunting
5. early loss of insight

Supportive diagnostic features include:

A. Behavioural disorder

- decline in personal hygiene and grooming

- mental rigidity and inflexibility
- distractibility and impersistence
- hyperorality and dietary change
- utilization behavior

B. Speech and language: altered speech output (aspontaneity and economy of speech, press of speech), stereotypy of speech, echolalia, perseveration, mutism

C. Physical signs: primitive reflexes, incontinence, akinesia, rigidity, tremor, low/labile blood pressure

D. Investigations:
- neuropsychology: impaired frontal lobe tests; no amnesia or perceptual deficits
- EEG: normal on conventional EEG despite clinically-evident dementia
- brain imaging: predominant frontal and/or anterior temporal abnormality

Editorial note: Most commonly frontotemporal dementia has its onset in middle years, usually presenting with behavioural/personality/conduct issues. Memory is often only minimally affected in early disease.

INTERNATIONAL CONSENSUS CONSORTIUM CRITERIA FOR DEMENTIA WITH LEWY BODIES (PARKINSONIAN COMPONENT) - McKEITH ET AL, 1996

1. Progressive cognitive decline of sufficient magnitude to interfere with normal social or occupational function. Prominent or persistent memory impairment may not necessarily occur in the early stages but is usually evident with progression. Deficits on tests of attention and frontal-subcortical skills and visuospatial ability may be especially prominent.
2. Two of the following are required for a diagnosis of probable Dementia with Lewy bodies:
   - fluctuating cognition with pronounced variations in attention and alertness
   - recurrent visual hallucinations which are typically well-formed and detailed
   - spontaneous motor features of Parkinsonism
3. Features supportive of the diagnosis are:
   - repeated falls
   - syncope or transient loss of consciousness
   - neuroleptic sensitivity
   - systematized delusions
   - hallucinations in other modalities

Editorial note: In neuropathology studies, Dementia with Lewy bodies and Alzheimer's disease pathology commonly coexist resulting in a spectrum of clinical expression ranging from pure Alzheimer's disease to pure Dementia with Lewy bodies. If the clinician's impression is that the predominant etiology is Alzheimer's disease, the patient may be eligible for Special Authority coverage for cholinesterase inhibitor therapy.