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6D85 Dementia due to diseases classified elsewhere

Clinical Descriptions and Diagnostic Requirements for ICD-11 Mental, Behavioural or Neurodevelopmental Disorders Dementia due to psychoactive substances, including medications Essential (required) features • All diagnostic requirements for dementia are met. • There is evidence from history, physical examination or laboratory findings that dementia is caused by the direct physiological consequences of use of a substance or medication that persists beyond the usual duration of substance intoxication or withdrawal. • This judgement depends on establishing the following. • The substance or medication and the amount and duration of its use is known to be capable of producing dementia. • The course of the dementia (e.g. onset, trajectory of symptoms, response to treatment) is consistent with that caused by the substance or medication. Note: specific substances are known to be capable of producing dementia. If the specific substance inducing the dementia has been identified, the corresponding diagnostic category should be assigned: 6D84.0 Dementia due to use of alcohol 6D84.1 Dementia due to use of sedatives, hypnotic or anxiolytics 6D84.2 Dementia due to use of volatile inhalants 6D84.Y Dementia due to other specified psychoactive substance. A diagnosis corresponding to the pattern of use of the relevant psychoactive substance (e.g. harmful pattern of psychoactive substance use, substance dependence) may also be assigned. Dementia due to diseases classified elsewhere The following categories for dementia associated with other diseases or conditions known to cause dementia are available: 6D85.0 Dementia due to Parkinson disease 6D85.1 Dementia due to Huntington disease 6D85.2 Dementia due to exposure to heavy metals and other toxins 6D85.3 Dementia due to HIV 6D85.4 Dementia due to multiple sclerosis 6D85.5 Dementia due to prion disease 6D85.6 Dementia due to normal-pressure hydrocephalus 6D85.7 Dementia due to injury to the head 6D85.8 Dementia due to Pellagra 6D85.9 Dementia due to Down syndrome 6D85.Y Dementia due to other specified disease classified elsewhere. 6D85 Neurocognitive disorders | Dementia due to psychoactive substances, including medications 6D84

627 Neurocognitive disorders Dementia due to Parkinson disease Essential (required) features • All diagnostic requirements for dementia are met. • Dementia is presumed to be attributable to underlying Parkinson disease, as demonstrated by neuropsychological test data, neuroimaging data, medical tests, family history and/or clinical history. Additional clinical features • Dementia due to Parkinson disease develops among individuals with idiopathic Parkinson disease, and is often characterized by impairment in attention, memory, executive and visuospatial functions. • Behavioural and psychiatric symptoms such as changes in affect, apathy and hallucinations may also be present. • Onset is insidious and typically occurs 1 year or more after the development of Parkinsonian motor symptoms. The course of dementia often follows that of underlying Parkinson disease (e.g. if Parkinson disease gradually worsens, dementia may gradually worsen). Note: a diagnosis of 8A00.0 Parkinson disease in Chapter 8 on diseases of the nervous system should also be assigned. Dementia due to Huntington disease Essential (required) features • All diagnostic requirements for dementia are met. • Dementia is presumed to be attributable to underlying Huntington disease, as demonstrated by neuropsychological test data, neuroimaging data, genetic testing, medical tests, family history and/or clinical history. Additional clinical features • Dementia due to Huntington disease occurs as part of a widespread degeneration of the brain due to a trinucleotide repeat expansion in the HTT gene, which is transmitted through autosomal dominance. 6D85.0 6D85.1 Neurocognitive disorders | Dementia due to diseases classified elsewhere

Clinical Descriptions and Diagnostic Requirements for ICD-11 Mental, Behavioural or Neurodevelopmental Disorders • Onset of symptoms is insidious, typically in the third and fourth decade of life, with gradual and slow progression. • Initial symptoms typically include impairments in executive functions, with relative sparing of memory, prior to the onset of motor deficits (bradykinesia and chorea) characteristic of Huntington disease. Note: a diagnosis of 8A01.10 Huntington disease in Chapter 8 on diseases of the nervous system should also be assigned. Dementia due to exposure to heavy metals and other toxins Essential (required) features • All diagnostic requirements for dementia are met. • Dementia is presumed to be attributable to toxic exposure to specific heavy metals, such as aluminium from dialysis water, lead, mercury or manganese, as demonstrated by neuropsychological test data, neuroimaging data, medical tests and/or clinical history. Additional clinical features • The characteristic neurocognitive impairments in dementia due to exposure to heavy metals and other toxins depend on the specific heavy metal or toxin that the individual has been exposed to, but can affect any cognitive domain. • Onset of symptoms is related to exposure, and progression can be rapid especially with acute exposure. • In some cases, symptoms are reversible when exposure is identified and ceases. Note: an appropriate diagnosis from the NE61 Harmful effects of or exposure to noxious substances, chiefly nonmedicinal as to source, not elsewhere classified, grouping in Chapter 22 on injury, poisoning or certain other consequences of external causes should also be assigned. Dementia due to HIV Essential (required) features • All diagnostic requirements for dementia are met. • Dementia is presumed to be attributable to underlying HIV disease, as demonstrated by neuropsychological test data, neuroimaging data, medical tests and/or clinical history. 6D85.2 6D85.3 Neurocognitive disorders | Dementia due to diseases classified elsewhere

629 Neurocognitive disorders Additional clinical features • Dementia due to HIV may develop during the course of confirmed HIV disease, in the absence of a concurrent illness or condition other than HIV infection that could explain the clinical features. • Although a variety of patterns of

neurocognitive deficits are possible, depending on where the HIV pathogenic processes have occurred, typically deficits follow a subcortical pattern with impairments in executive function, processing speed, attention and learning new information. • The course of dementia due to HIV, varies and may involve gradual decline in functioning, improvement or resolution of symptoms, or fluctuation in symptoms over time. • Rapid decline in neurocognitive functioning is rare, with the advent of antiretroviral medications. Note: an appropriate diagnosis from the Human immunodeficiency virus disease grouping in Chapter 1 on certain infectious or parasitic diseases should also be assigned. Dementia due to multiple sclerosis Essential (required) features • All diagnostic requirements for dementia are met. • Dementia is presumed to be attributable to the cerebral effects of underlying multiple sclerosis – a demyelinating disease – as demonstrated by neuropsychological test data, neuroimaging data, medical tests and/or clinical history. Cognitive symptoms are not primarily due to associated physiological or functional effects of the underlying disease (e.g. fatigue, motoric limitations). Additional clinical features • Onset of symptoms is often insidious, but progression may occur in a stepwise fashion, in accordance with the underlying disease course. • Neurocognitive impairments vary according to the location of demyelination, but typically include deficits in processing speed, memory, attention and aspects of executive functioning. Note: a diagnosis of 8A40 Multiple sclerosis in Chapter 8 on diseases of the nervous system should also be assigned. 6D85.4 Neurocognitive disorders | Dementia due to diseases classified elsewhere

Clinical Descriptions and Diagnostic Requirements for ICD-11 Mental, Behavioural or Neurodevelopmental Disorders Dementia due to prion disease Essential (required) features • All diagnostic requirements for dementia are met. • Dementia is presumed to be attributable to underlying human prion disease, as demonstrated by neuropsychological test data, neuroimaging data, genetic testing, medical tests and/or clinical history. Additional clinical features • Dementia due to prion disease is caused by a group of spongiform encephalopathies resulting from abnormal prion protein accumulation in the brain. These can be sporadic, genetic (caused by mutations in the prion protein gene) or transmissible (acquired from an infected individual). • Onset is insidious, and progression of symptoms and impairment is rapid, often characterized by neurocognitive deficits, ataxia and motor symptoms (e.g. myoclonus, chorea or dystonia). • Diagnosis is typically made on the basis of clinical presentation, brain imaging studies, presence of characteristic proteins in spinal fluid, EEG and/or genetic testing. Note: an appropriate diagnosis from the Human prion diseases grouping in Chapter 8 on diseases of the nervous system should also be assigned. Dementia due to normal-pressure hydrocephalus Essential (required) features • All diagnostic requirements for dementia are met. • Dementia is presumed to be attributable to underlying normal-pressure hydrocephalus, as demonstrated by neuropsychological test data, neuroimaging data, medical tests and/ or clinical history. 6D85.5 6D85.6 Neurocognitive disorders | Dementia due to diseases classified elsewhere

631 Neurocognitive disorders Additional clinical features • Dementia due to normal-pressure hydrocephalus results from excess accumulation of cerebrospinal fluid in the brain as a result of idiopathic, non-obstructive causes, but can also be secondary to haemorrhage, infection or inflammation. • Progression is gradual but intervention (e.g. shunt) may result in improvement of symptoms, especially if administered early in the course of the condition. • Typically, neurocognitive impairments include reduced processing speed and deficits in executive functioning and attention. These symptoms are also typically accompanied by gait abnormalities and urinary

incontinence. • Brain imaging to reveal ventricular volume and characterize brain displacement is often necessary to confirm the diagnosis. Note: a diagnosis of 8D64.04 Normal-pressure hydrocephalus in Chapter 8 on diseases of the nervous system should also be assigned. Dementia due to injury to the head Essential (required) features • All diagnostic requirements for dementia are met. • Dementia is presumed to be attributable to an injury to the head, as demonstrated by neuropsychological test data, neuroimaging data, medical tests and/or clinical history. Additional clinical features • Dementia due to injury to the head is caused by damage inflicted on the tissues of the brain as the direct or indirect result of an external force. • Trauma to the brain is known to have resulted in loss of consciousness, amnesia, disorientation and confusion, and/or neurological signs. • The symptoms characteristic of dementia due to injury to the head arise immediately following the trauma or after the individual gains consciousness, and must include persistent cognitive impairments following any recovery of initial cognitive impairment that may be seen in the immediate post-injury period. • Neurocognitive deficits vary depending on the specific brain areas affected and the severity of the injury, but can include impairments in attention, memory, executive functioning, personality, processing speed, social cognition and language abilities. Note: a diagnosis of NA07 Intracranial injury or one of its subcategories in Chapter 22 on injury, poisoning or certain other consequences of external causes should also be assigned. 6D85.7 Neurocognitive disorders | Dementia due to diseases classified elsewhere

Clinical Descriptions and Diagnostic Requirements for ICD-11 Mental, Behavioural or Neurodevelopmental Disorders Dementia due to pellagra Essential (required) features • All diagnostic requirements for dementia are met. • Dementia is presumed to be attributable to pellagra, as demonstrated by neuropsychological test data, medical tests and/or clinical history. Additional clinical features • Dementia due to pellagra is caused by persistent lack of vitamin B3 (niacin) or tryptophan either in the diet or due to poor absorption in the gastrointestinal tract due to disease (e.g. Crohn disease) or due to the effects of some medications (e.g. isoniazid). • Core signs of pellagra include dermatological changes (sensitivity to sunlight, lesions, alopecia and oedema) and diarrhoea. • With prolonged nutritional deficiency, neurocognitive symptoms that include aggression, motor disturbances (ataxia and restlessness), confusion and weakness may be observed. • Treatment with nutritional supplementation (e.g. niacin) typically results in reversal of symptoms. Note: a diagnosis of 5B5C.0 Pellagra in Chapter 5 on endocrine, nutritional or metabolic diseases should also be assigned. Dementia due to Down syndrome Essential (required) features • All diagnostic requirements for dementia are met. • Dementia is presumed to be attributable to Down syndrome, as demonstrated by neuropsychological test data, genetic testing, medical tests and/or clinical history. Additional clinical features • Dementia due to Down syndrome is caused by abnormal increased production and accumulation of amyloid precursor protein (APP), leading to formation of beta-amyloid 6D85.8 6D85.9 Neurocognitive disorders | Dementia due to diseases classified elsewhere

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