

# 215 - 6D83 Frontotemporal dementia

## 6D83 Frontotemporal dementia

625 Neurocognitive disorders Frontotemporal dementia Essential (required) features • All diagnostic requirements for dementia are met. • Dementia is presumed to be attributable to underlying frontotemporal disease or atrophy, as demonstrated by neuropsychological test data, neuroimaging data, genetic testing, medical tests, family history and/or clinical history. Additional clinical features • Frontotemporal dementia variants include primary progressive aphasia (logopenic, semantic and agrammatic subtypes), behavioural frontotemporal dementia and motoric frontotemporal dementia (corticobasal degeneration, progressive supranuclear palsy and amyotrophic lateral sclerosis). • Frontotemporal dementia is progressive, with variants identified based on initial symptoms. • Frontotemporal dementia, behavioural variant, is characterized by personality changes, often including apathy and progressively inappropriate social behaviour. Neurocognitive functioning may be preserved in the early stages, though the progression may later involve deficits in executive functioning (e.g. planning, problem solving), with comparatively intact memory skills. • Frontotemporal dementia, primary progressive aphasia, is characterized by progressive impairment in language skills, initially in the absence of impairment in other cognitive skills. Subtypes of primary progressive aphasia are often determined based on neuropsychological/cognitive testing, clinical presentation and sometimes neuroimaging, and are characterized by primary deficits in word finding (logopenic subtype), word meaning (semantic subtype) or word production (agrammatic subtype). • Frontotemporal dementia, motoric variant, involves progressive impairment in motor functioning, sometimes in the context of progressive neurocognitive impairment (typically characterized by impairment in attention, executive functioning and visuospatial skills, with comparatively intact memory skills). Frontotemporal dementia, motoric variant, can include progressive supranuclear palsy (e.g. poor balance, frequent falls, visual impairment from gaze palsy), corticobasal degeneration (e.g. limb apraxia, tripping, rigidity, dystonia) and amyotrophic lateral sclerosis (e.g. muscle weakness, muscle atrophy, fasciculations, spasticity). Note: a diagnosis of 8A23 Frontotemporal lobar degeneration in Chapter 8 on diseases of the nervous system should also be assigned. 6D83 Neurocognitive disorders | Frontotemporal dementia

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