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Frontotemporal dementia with motor neuron disease

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Frontotemporal dementia with motor neuron disease. ORPHA:275872

Frontotemporal dementia with motor neuron disease (FTD-MND) is a type of frontotemporal lobar degeneration characterized by the insidious onset (between the ages of 38-78 years) of dementia-associated psychiatric symptoms (e.g. personality changes, uninhibited behavior, irritability, aggressiveness), memory difficulties, global intellectual impairment, emotional disorders and transcortical motor aphasia that eventually leads to mutism, in addition to the manifestations of motor neuron disease such as neurogenic muscular wasting (similar to what is seen in amyotrophic lateral sclerosis; see this term). The disease is progressive, with death occurring 2-5 years after onset.