

# Degenerative or Debilitative Neurologic Syndromes



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## KEYWORDS

- Neurologic disorders • Dementia • Parkinson disease • Disease presentation
- Treatment

## KEY POINTS

- Dementia, the most common neurodegenerative disorder, has several subtypes and is characterized by cognitive and functional decline.
- Parkinson disease is the second most common neurodegenerative disorder and presents with the 3 hallmark motor symptoms of resting tremor, bradykinesia, and rigidity.
- Both dementia and Parkinson disease have significant associated symptoms that warrant assessment and treatment by clinicians.
- Neurodegenerative disorders currently lack effective neuroprotective or disease-mediating therapy; however, some promising therapies are in the pipeline.

The descriptor degenerative is broad, denoting a decline from normal function, typically without a known cause. As health care science has progressed and causes for many neurologic diseases have been discovered, the term degenerative has largely fallen out of favor; however, it may still be used to describe a broad group of disorders characterized by progressively altered neurologic function, ranging from the common (eg, dementia) to the rare (eg, amyotrophic lateral sclerosis [ALS]). This article identifies and describes common neurodegenerative disorders, their diagnosis and management, and current research priorities.

## BACKGROUND AND SIGNIFICANCE

In the context of the decline in infectious diseases worldwide with associated increased life expectancy, international public health officials increasingly focus on degenerative diseases or diseases associated with older age, including degenerative neurologic disorders. The World Health Organization (WHO) last updated their report on neurologic disorders in 2006, at which time they estimated that the 2 most common degenerative neurologic disorders, dementia and Parkinson disease (PD), could affect

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as many as 44 million and 7 million persons by 2030, respectively.<sup>1</sup> Neurodegenerative disorders are accompanied by significant disability. The WHO acknowledges this burden, estimating that dementia and PD could result in greater than 17 million years of healthy life lost as a result of disability.<sup>1</sup>

## DEMENTIA

The most common degenerative neurologic disorder is dementia. Predominantly a syndrome of older age, there are several known variants of dementia, each of which has a unique pathophysiology, presentation, and trajectory. Despite their differences, each type of dementia causes significant impairments to cognition and function and, at this time, there are no curative therapies available for dementia, regardless of type.

Prevalence of dementia is directly related to age, with older groups being affected in larger numbers (Table 1).<sup>2,3</sup> As of 2010, an estimated 4.7 million Americans older than the age of 65 years had dementia.<sup>2</sup> Overall prevalence is slightly higher in women, which is likely due to women having greater life expectancy than men.<sup>4</sup> Epidemiologic studies predict that as nations become increasingly developed, numbers of persons who are older and thus are affected by dementia will grow, resulting in an anticipated worldwide increase in dementia cases in the coming decades.<sup>4</sup> In addition to advancing age, dementia prevalence is also correlated to a population's overall level of educational attainment and socioeconomic factors. Higher education and socioeconomic status are associated with less obesity and cardiovascular disease, which are known risk factors for vascular and Alzheimer dementia.<sup>4</sup>

Each person with dementia's disease trajectory is unique, with significant individual variation in speed of symptom progression through the cognitive, behavioral, and functional domains. This variation makes it difficult to devise a universal staging schema or tool.<sup>5</sup> However, to capture the typical progression of symptoms, dementia is often conceptualized as having mild, moderate, and severe stages. For most patients, mild dementia is characterized by early deficits in cognition and mild functional deficits.<sup>5</sup> In addition to having progressively worsening cognition and function, persons with moderate dementia often have more frequent and severe behavioral and psychological symptoms of dementia (BPSD).<sup>5</sup> Finally, severe dementia is characterized by advanced cognitive impairment and more vegetative-type symptoms.<sup>5</sup> There is not a standard amount of time spent in each phase; however, those with a generally shorter or more aggressive disease course will often experience shorter periods in the mild, moderate, and severe stages, whereas those with longer disease trajectories may have relatively longer periods in each stage. Persons with dementia have an average of 4 to 8 years from diagnosis to death, though significant variation in survival exists, with some patients surviving 20 or more years after diagnosis.<sup>5</sup>

**Table 1**  
Prevalence of dementia

Age (y)	Estimated Number of Americans with	
	Dementia in 2010 by Age Group <sup>2</sup>	Percent of Age Group with Dementia <sup>2,3</sup>
65–74	0.7 million	0.032%
75–84	2.3 million	17.7%
85+	1.8 million	34.0%

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