

## Forms of Frontotemporal Dementia

### Behavioral variant frontotemporal dementia (bvFTD)

Behavioral variant frontotemporal dementia (bvFTD) has also been referred to as "frontal variant FTD" (fvFTD) or "Pick's disease." Approximately 60% of people with any form of FTD have bvFTD. By definition, this form of FTD affects social skills, emotions, personal conduct, and self-awareness. Deficits in these functions most often reflect damage to specific regions within the frontal and temporal lobes. With damage to these areas, people may show mood and behavior changes including stubbornness, emotional coldness or distance, apathy and selfishness. Unlike Alzheimer's disease, which affects a different area of the brain, many people with bvFTD don't show any confusion or forgetfulness about where they are or what day it is, at least at first.

### Semantic dementia (SD)

Semantic dementia, which has also been called "temporal variant FTD," accounts for 20% of FTD cases. Language difficulty, the predominant complaint of people with SD, is due to the disease damaging the left temporal lobe, an area critical for assigning meaning to words. The language deficit is not in producing speech but is a loss of the meaning, or semantics, of words. At first, you might notice someone substituting a word like "thingy" for more unusual words, but eventually a person with SD will lose the meaning of more common words as well. For example, early in the illness a patient might lose the word for a falcon, later-on forget the word for a chicken, then call all winged creatures "bird" and eventually call all animals "things." Not only do they lose the ability to recall the word, but the concept of these words is also lost. "What is a bird?" might be a typical response for a patient with advanced SD. Reading and spelling usually decline as well, but the person may still be able to do arithmetic and use numbers, shapes or colors well. Names of people, even good friends, can become quite difficult for people with SD. Like the behavioral variant, memory, an understanding of where they are, and sense of day and time tend to function as before. Muscle control for daily life and activities tends to remain good until late in the disease. Some of these skills may seem worse than they actually are because of the language difficulty people with SD have when they try to express themselves.

When SD starts in the right temporal lobe, people in the early stages

have more trouble remembering the faces of friends and familiar people. Additionally, these people show profound deficits in understanding the emotions of others. The loss of empathy is an early, and often initial, symptom of patients with this right-sided form of SD. Eventually people with right-sided onset progress to the left side and then develop the classical language features of SD. Similarly, left-sided cases progress to involve the right temporal lobe and then the person experiences difficulty recognizing faces, foods, animals and emotion. SD patients eventually develop classical

bvFTD behaviors including disinhibition, apathy, loss of empathy and diminished insight. The time from diagnosis to the end is longer than for those with bvFTD, typically taking about six years.

## **Progressive nonfluent aphasia (PNFA)**

PNFA accounts for only about 20% of all people with FTD. Unlike semantic dementia where the person maintains the ability to speak but loses the meaning of the word, people with PNFA have difficulty producing language fluently even though they still know the meaning of the words they are trying to say. The person may talk slowly, having trouble saying the words, and have great trouble with the telephone, talking within groups of people or understanding complex sentences. In recent years it has become apparent that many patients with PNFA go on to develop severe Parkinsonian symptoms that overlap with [progressive supranuclear palsy](#) (PSP) and [corticobasal degeneration](#) (CBD) such as an inability to move the eyes side-to-side, muscle rigidity in the arms and legs, falls, and weakness in the muscles around the throat.

## **FTD with motor neuron disease**

Approximately 15% of patients with FTD also develop motor neuron disease (FTD-MND). Most often, this combination occurs in patients with bvFTD, and only rarely does MND arise in patients with SD or PNFA. MND affects motor nerve cells in the spinal cord, the brain stem (which sits on top of the spinal cord), and the cerebral cortex. Because the brainstem was once referred to as the "bulb", you may hear some MND symptoms described as "bulbar symptoms". The most common type of MND is [amyotrophic lateral sclerosis](#) (ALS), also called Lou Gehrig's disease, which can occur as a purely motor disorder. More often, however, patients with ALS also have behavioral or cognitive problems similar to those seen in FTD. MND symptoms include slurring of speech, difficulty swallowing, choking, limb weakness or muscle wasting. In patients with FTD-MND, there is often (but not always) a family history of the disease, and scientists are getting closer to identifying gene mutations that cause the illness.