Disease Progression

The staged progression of each type of frontotemporal lobar degeneration.

Behavioral variant FTD

The pace of the symptoms and length of disease can vary dramatically from person to person. In general, each type of FTD follows a pattern where the symptoms seen in the mild stage become more pronounced and disabling over a course of 8-10 years.

Mild bvFTD

In the first several years, a person with bvFTD (sometimes called Pick's disease or just FTD) tends to exhibit marked behavioral changes such as disinhibition, apathy, loss of sympathy or empathy for others, or overeating. Problems with planning organization and sometimes memory are evident, but the individual is still capable of managing household tasks and self-care with minimal help. However, impairment in judgment can lead to financial indiscretions with potentially catastrophic consequences. Social withdrawal, apathy and less interest in family, friends and hobbies may be evident. At times, they may behave inappropriately with strangers, lose their social manners, act impulsively and even break laws. But at this stage, the behaviors can often be managed with lifestyle and environmental changes (read our practical tips [1] for ideas). A MRI image at this point will show mild atrophy in particular areas of the frontal lobes.

Moderate bvFTD

Over the course of a few years, the symptoms seen in the mild stage will become more pronounced and disabling. You might also notice compulsive behaviors like repetitive urination, hoarding or collecting objects, compulsive cleaning or silly repetitive movements (like stomping on ants). Binge eating may create weight problems and other health issues. The cognitive problems associated with dementia become more pronounced, with mental rigidity, forgetfulness and severe deficits in planning and attention. The MRI image at this point will show that the shrinking of the brain tissue has expanded to larger areas of the frontal lobes, as well as the tips of the temporal lobes and basal ganglia, deeper brain structures involved in motor coordination, cognition, emotions and learning.

Severe bvFTD

By this point the patient is experiencing profound behavioral symptoms (apathy, loss of empathy, disinhibition) in association with language difficulty and memory loss. Although it can vary widely, the time from the first symptom to the end is typically about eight years, whereas the time from diagnosis is, on average, about five years.

Semantic dementia
The pace of the symptoms and length of disease can vary dramatically from person to person. In general, each type of FTD follows a pattern where the symptoms seen in the mild stage become more pronounced and disabling over a course of 8-10 years.

**Mild SD**

People with early semantic dementia that is predominantly on the left side of the brain, usually complain of a hard time coming up with the word or name for something. Words that the person uses a lot may remain available, but more unusual words may be replaced by "thingy" or "you know." The tone, rhythm and fluctuations of pitch (prosody) generally sound normal. Memory for day-to-day events is usually spared.

The early signs of SD in people with asymmetric right-sided damage include a decline in empathy or awareness of other people's emotions.

**Moderate SD**

After two to three years, the people with left sided damage and those with right sided damage tend to look more similar, as the disease typically progresses to involve both sides. With moderate SD, most people show at least some of the behavioral problems that are similar to the behavioral variant of FTD.

People with moderate semantic dementia will have immense trouble understanding you. They may also have increasing difficulty recognizing the names and faces of people – even friends and family. Reading and writing, mostly likely, will have declined noticeably. The person may still be able to use numbers, colors and shapes – the brain functions responsible for these skills are organized in a different area of the brain from words.

**Severe SD**

After four to five years of SD, the disease is usually quite advanced, which means the person's language skills have significantly eroded, making communication very difficult while the behavioral problems have significantly increased. Typical behaviors seen in late stage SD include disinhibition, apathy, compulsions, impaired face recognition, altered food preference and weight gain. People with left-sided damage tend to show more interest in visual or non-verbal things while people with right-sided damage tend to prefer games with words and symbols. The time from diagnosis to the end typically takes about six years, although this can vary significantly from person to person.

**Progressive nonfluent aphasia**

The pace of the symptoms and length of disease can vary dramatically from person to person. In general, each type of FTD follows a pattern where the symptoms seen in the mild stage become more pronounced and disabling over a course of 8-10 years.

**Mild PNFA**

Progressive nonfluent aphasia leads to increasing trouble speaking and producing language, although the person with it usually understands language and knows what they want to say. Early symptoms include slowed speech and trouble getting the words out correctly. For example, if a person with PNFA has to repeat a word that is difficult to say several times, it will most likely sound
 Moderate PNFA

As the disease progresses into years three and four, the person will have more and more trouble producing speech. They may use short sentences without a lot of extra words like articles and adjectives. Reading and writing skills may are usually still good, so you might want to consider using a board or number of pictures to help the person express their meaning. Skills with numbers, colors and shapes generally remain intact, as do skills involving face and emotion recognition.

Severe PNFA

After five or more years of PNFA, the person with PNFA is essentially mute and may show behavioral problems similar to those of behavioral variant FTD. Some people with PNFA develop Parkinson's-like motor problems like muscle rigidity and stiffness. The time from diagnosis to the end typically takes about six years, although this can vary significantly from person to person.

End stage FTD

After years of FTD, patients may have trouble coordinating their muscles and require a wheelchair. Usually 24-hour care is required, whether at home or in an institution. The physical decline and changes that occur throughout the disease course become more and more obvious at this stage. Eventually, the person with FTD may have great difficulty swallowing, chewing, moving and controlling their bladder and/or bowels. Death from FTD is usually caused by the consequences of these physical changes, most commonly infections in the lungs, skin or urinary tract. Although it can vary widely, the time from the first symptom to the end is typically about eight years, whereas the time from diagnosis is, on average, about five years.

- What is Frontotemporal Dementia?

Source URL: http://memory.ucsf.edu/ftd/overview/ftd/progression

Links: