Introduction
The frontotemporal dementias (FTDs) are a clinically, genetically and pathologically heterogeneous group of disorders. They primarily occur between the ages of 35 and 75 years of age with an equal incidence in both genders. FTD is the second most common form of dementia in people under 65 years with an average age of onset in the sixth decade. FTDs are fatal and the disease duration ranges from five to ten years. It is estimated that twenty to forty percent of patients have a family history of dementia.

Clinical Features
The clinical presentations vary. In fact, some patients present with frontal lobe signs (control of behavior and personality changes) while others show mostly temporal lobe dysfunction (language impairment). Occasionally, patient may present features indistinguishable from Alzheimer disease, may display movement abnormalities similar to those seen in Parkinson disease, or may present features of amyotrophic lateral sclerosis (ALS). Thus, the affected individual may present with one or more of the following symptoms: Personality changes (selfishness/callousness, antisocial traits, and lack of empathy for others' situations), social disinhibition, apathy, mutism (no speech), apraxia (inability to translate a thought into a motor action, in the absence of paralysis), movement dysfunction, hyperorality, or disorganization. Sometimes there is an emergence of new artistic or musical talents. As FTD progresses, affected individuals develop a combination of these symptoms and, eventually, develop dementia requiring full time supervision and care.

Diagnosis
Ideally, the diagnosis of FTD should utilize a combination of neurologic, neuropsychological and neuroradiologic examinations as well as a detailed family and personal history. Particularly useful are the MRI and PET images correlated with data from the neuropsychological tests. A prominent loss of tissue in the frontal and/or temporal lobes as seen on MRI and a reduced function of these areas as seen on a PET scan are characteristic of FTD.

The disease may be incorrectly diagnosed. One of the primary reasons is that dementia or cognitive dysfunction is not always present at the beginning. In addition, the onset may be slow and not obvious or the individual may present with behavioral symptoms that are difficult to interpret. Also, personality changes may be seen as a psychiatric disturbance.

The neuropathologic hallmark of the FTDs is the degenerative changes in the frontal and/or temporal lobes of the brain. These areas are responsible for language and executive functions (planning and organizing). It should be realized that a variety of neuronal changes occur in the various types of FTD and the regional differences influence the clinical presentation. Neurons in FTD may show accumulation of abnormal material. In some types of FTD, there is accumulation of a chemically altered form of the tau protein, which is the same protein that makes up the neurofibrillary tangles seen in Alzheimer disease. In other forms of FTD, a protein called TDP-43 has recently been found to accumulate.
Inheritance
Currently, the known inherited forms of FTD appear to be transmitted genetically with a dominant pattern. In the past decade, scientists have begun to decipher the molecular genetic bases of hereditary FTD. It has become clear that the biology of these human neurodegenerative diseases has a complexity not previously suspected. Hereditary FTD has been found to be associated to several genes located in chromosomes 3, 9, and 17. The first of these (Tau) was first found to be associated with FTD in 1998. Since then, three other genes were similarly found to have an association with FTD including one discovered just this past summer. This recent discovery has increased the interest in this already fast moving scientific field. From these discoveries, it is clear that there is no single pathway leading to the cellular and anatomic damage seen in FTD. The discovery of the genes involved in the familial forms of FTD may give some clues to the pathologic mechanisms involved in the sporadic forms, thus paving the way for the development of therapeutic strategies.

Care Issues
As one would imagine, from the perspective of the care partner, these symptoms are difficult to live with and manage. The symptoms are perplexing and some of the traditional supportive techniques do not seem to alleviate the difficult moments or redirect the behavior.

Anticholinergic medications currently being used for Alzheimer disease’s cognitive deficits do not seem to manage FTD symptoms. The family often faces the challenge of adapting to the ever-changing behavioral symptoms and must constantly change their care approaches. No matter what the age or who the care partner is, this is a challenging caregiving situation. If the affected person is younger and raising or launching a family, the emotional, relational and financial stressors for the spouse and family are significant. Children are deprived of a normal parent-child relationship. Often partners and children are forced to deal with feelings of anger, frustration, sadness and ultimately depression.

Finding suitable and qualified respite care can be problematic because of the person’s challenging behavioral symptoms and the professional care providers’ lack of knowledge of the spectrum of symptoms associated with the FTDs.

Without a doubt, this is a difficult illness and caregiving experience. However, we at the Greater Cincinnati Chapter can assist with the ongoing educational and supportive aspects of caregiving. Please call (513-721-4284) and ask to speak with Helpline about these disorders. Also, the Association for Frontotemporal Dementias (AFTD) website (www.FTD-Picks.org) is an excellent resource for educational materials with access to chat rooms and support groups.

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