PRIMARY CARE RESEARCH

PCRI/NorTex Articles

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tremor. Strength remains normal but the limb becomes progressively less useful because of the problems with tone, slowness, tremor, and praxis.

PSP causes progressive problems with control of gait and balance. The hallmark of PSP is a visual disturbance which results from a progressive inability to coordinate eye movements. Additional motor symptoms similar to those seen in Parkinson's disease and other features of front temporal dementia such as behavioral and social dysfunction and cognitive decline may develop. Depression and apathy are also common mood symptoms.

FTD /ALS

People with frontotemporal dementia with ALS (also known as FTD with Motor Neuron Disease) experience the behavior and language symptoms associated with other FTDs along with the motor symptoms of ALS. The symptoms of motor neuron disease can go unnoticed for quite a while in FTD, because the changes in behavior, personality, and thinking are so dramatic. Conversely, a portion of ALS patients develop symptoms of dementia as their disease progresses.

Disease Progression and Impact

Patients may present initially with behavioral, language, or movement symptoms, but as time passes many individuals acquire symptoms from the other areas as well. As disease progresses, patients experience increasing difficulty in their ability to plan or organize activities, behave appropriately in social or work settings, interact with others, and care for themselves. The burden of disease and the toll it can take on a family increases drastically. Many affected individuals will lose their jobs (endangering income and healthcare for the family), and many marriages and family relationship are torn apart before an official medical diagnosis is made. Average life expectancy is eight years, but can vary from two to 18 years based on the specific disorder.

Similar to other neurodegenerative disorders, FTD is caused by the pathological aggregation of proteins in the brain. Approximately 10% of people diagnosed with FTD have an autosomal dominant pattern of inheritance, and 20-40% demonstrates a familial risk. 50-70% of FTD is sporadic.

At present there is no treatment or cure for these disorders, and definitive diagnosis must wait until autopsy. Clinicians are left to borrow from other diagnoses to treat the symptoms that are most troublesome for the patient and his or her caregivers. But research into this set of disorders has exploded in recent years published papers in this decade number more than 10 times those published in the 1990's, and the first clinical trials for these patients are being initiated. There is hope.

The astute family physician can play a critical role in the speed with which a patient is diagnosed—and thus help limit the destructive impact the disorder has on a family.

For more information about FTD and the role of the primary care physician, please see the following:

March 2, 2011: Dr. Mario Mendez M.D, PhD, UCLA FTD and Neurobehavioral Clinic will present Grand Rounds at UNTHSC on the Frontotemporal Dementias.

The Association for Frontotemporal Dementias is a nonprofit support, education and advocacy organization dedicated to the FTDs: 866 507-7221 www.ftd-picks.org.

Cardarelli R, Kertesz A, Knebl JA. (2010). Frontotemporal Dementia: a review for primary care physicians. *American Family Physician*. 82(11).

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