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Researchers open window on a little-studied form of dementia

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(PhysOrg.com) -- Yale University researchers have discovered one of the first tantalizing clues to how frontotemporal dementia (FTD) - often mistaken for Alzheimer's disease - wreaks havoc in the brain.

About 40 percent of dementia cases are distinct from Alzheimer's, even though they share the symptoms of memory loss and cognitive decline. The causes of these subsets of dementia are poorly understood. The new findings, reported in the November 18 issue of the journal *Neuron*, are an important first step in unraveling the mysteries of FTD, one of the most common non-Alzheimer's forms of dementia, the authors report.

"Dementia threatens to overwhelm our health system, yet there is relatively limited research funding to understand and develop treatments for Alzheimer's, and even less for other forms of dementia," said Stephen M. Strittmatter, the Vincent Coates Professor of Neurology, director of the Program in Cellular Neuroscience, Neurodegeneration and Repair at Yale, and senior author of the study.

According to the National Institutes of Health, FTD causes between 2 and 10 percent of all dementias and is marked by degeneration of nerve cells primarily in the frontal and temporal lobes of the brain. The brains of patients with FTD do not have the large amounts of amyloid plaques that mark Alzheimer's disease, although patients with both conditions exhibit memory problems and cognitive decline. The course of the disease tends to be slightly different from Alzheimer's. Some patients develop language problems as the initial symptom. Some begin to act in

socially inappropriate ways.

Recently, clinicians have had success differentiating between the two conditions by using PET scans to look for buildup of amyloid plaques. There are currently no good treatments for either condition. Strittmatter said it is crucial to understand the molecular basis for all forms of dementia because it is likely that each will need a different form of treatment.

Studies of inherited forms of FTD have linked mutations in the gene progranulin to inherited forms of the disease. But scientists had no idea how the gene, which seems to be involved in the repairing of wounds, could cause the dementia.

In the Neuron paper, Strittmatter's team showed that protein sortilin interacts at a molecular level with progranulin and that an absence of sortilin can dramatically increase amounts of progranulin in the brain.

"This is just first step of molecular understanding of this dementia,." Strittmatter said. "Eventually we hope to find a way to intervene in the disease and prevent or alleviate symptoms of dementia.

Provided by Yale University