

news THE UNIVERSITY OF TEXAS HEALTH SCIENCE CENTER AT DALLAS

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******Huntington's Disease commission to hold
public hearing May 3 on UTHSCD campus.*

DALLAS--The widow of folk singer/writer/poet Woody Guthrie, who died of Huntington's Disease, will visit The University of Texas Health Science Center at Dallas on May 3. Mrs. Marjorie Guthrie will appear as chairperson of the Commission for the Control of Huntington's Disease and Its Consequences, which will hold a day-long public hearing in the campus' Gooch Auditorium.

Huntington's Disease is a fatal hereditary disease characterized by gradual degeneration of the brain. Victims suffer from uncontrollable muscle movements, personality changes and gradual loss of mental abilities. Currently there is no adequate treatment for the disease.

The Huntington's Disease commission was created by Congress to develop a comprehensive national plan to combat this mysterious malady. To accomplish its task, the commission is holding a series of regional hearings to take testimony from families afflicted with the disease as well as medical practitioners, researchers, health officials and other interested members of the public.

The regional hearing for Arkansas, Louisiana, Oklahoma and Texas will be held on the Dallas health science center campus May 3 from 9 a.m. to 5 p.m.

Dr. E. Simon Sears, assistant professor of neurology at the UT Southwestern Medical School, is serving as chairman of the Southwest regional hearings. An authority on Huntington's Disease (HD), Dr. Sears notes that HD has often been called a "genetic time bomb" because symptoms usually do not appear until between the ages of 30 and 45, near the end of the reproductive years.

Children of those who carry the gene for HD have a 50-50 chance of getting it themselves, he explains. If a child does not receive the gene from either parent, then he (or she) will not pass it on to succeeding generations.

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"One of the most important recent advances in genetics has been the identification of gene carriers in certain hereditary diseases," Dr. Sears says. Such tests prevent anguish in those who might carry the genetic blueprint for an hereditary disease and allow them to make informed decisions about having children, he explains.

"So far, no test has been developed for Huntington's Disease," he continues. "We do not know which at-risk individuals will develop the disease until after the symptoms have appeared. By then many have become parents and it is too late to stop HD from being passed on to future generations."

In research supported by the Huntington's Chorea Foundation, Dr. Sears is attempting to develop such a test.

"The brains of patients with Huntington's Disease are deficient in certain biochemicals and at least one substance (a biochemical known as GABA) is lower in spinal fluid as well," he says. "At present, we do not know whether this spinal fluid deficiency occurs before or only after clinical symptoms of the disease develop.

"If the GABA deficiency antedate the clinical symptoms by several years, it may be of benefit in genetic counseling," he speculates.

The same biochemical deficiencies may show up in the placental tissue, or afterbirth, of newborn babies who have inherited HD, Dr. Sears adds. "I also am investigating the feasibility of testing for Huntington's by analyzing the placental tissue after babies are born to at-risk parents."

Dr. Sears will discuss his research in greater detail in his testimony to the commission May 3.

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