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******Genetics experts will test family with rare affliction.*

DALLAS--A tragic hereditary disease which causes male children to attempt to "self destruct" will be studied at Children's Medical Center here Jan. 24 by genetic experts from Southwestern and Johns Hopkins medical schools.

Dr. Barbara Migeon of Johns Hopkins Medical School in Baltimore, Maryland will join Dr. Joseph Goldstein and others of the Medical Genetics Division at UT Southwestern in examining some 20 members of a Dallas family afflicted with Lesch-Nyhan syndrome.

The perplexing affliction causes lip and finger chewing, head banging and hostility to the point that many children must be restrained from mutilating themselves. Affected children have a lack of muscle control similar to cerebral palsy and most appear to be mentally retarded, with severe speech problems.

Assistant professor Mary Jo Harrod of the Southwestern group said CMC's Birth Defects Center will be utilized for obtaining blood and skin samples from family members.

By culturing samples it is possible for Dr. Migeon's lab to determine who is afflicted with the disease, who may transmit it and who are completely free of the problem.

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first add lesch-nyhan

"We may find eight or ten female family members who are carriers," explained Dr. Harrod. We know that at least seven male members of this family have been affected, four of whom are still living.

Like hemophilia, Lesch-Nyhan syndrome is a sex-linked condition carried only by women and afflicting only males. Half the sons of a carrier would be expected to be afflicted while half the daughters would be carriers like their mothers.

Prenatal testing is now possible to detect the disease in the fetus of a carrier mother so that the birth of an affected son can be avoided. All members of the local family who are identified as carriers will receive genetic counseling and will be offered the test in future pregnancies if they so desire. The females who are found to not carry the gene can be reassured that they need not fear this condition in their children or in future generations of their descendents.

Generally the afflicted males are apparently healthy in their first months of life, developing symptoms later as a cerebral-palsy-like condition becomes apparent resulting in spasticity and inability to control movements.

Dr. Harrod explained that the disease is associated with a biochemical defect that results in an overproduction of uric acid "so that many of these children get gout and many times die of kidney disease associated with high uric acid."

Lesch-Nyhan syndrome, named for two doctors who only in 1965 discovered and described the ailment, presents some baffling contradictions in behavior. Mental retardation is common but probably not as severe as it appears since the lack of muscle control and the speech disorder affects the results of intelligence tests.

Many of the victims who are able to communicate to some degree indicate they actually want to be restrained from mutilating themselves. Otherwise, some have actually stuck their fingers in moving spokes of wheel chairs or struck at physicians or nurses--at the same time apologizing for their behavior.

While most victims of the disease are of Caucasian origin, there are occurrences among Orientals and blacks. The Dallas family is one of only two black families reported.

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