Tangier Disease
Tonsils May Cure Hearts

By John Pruett

Virginian-Pilot Staff Writer

TANGIER ISLAND — Scientists believe that the discovered tonsils of a Tangier Island boy, Teddy Laird, may provide an important clue to the search for causes and cures of some heart ailments.

Laird's peculiar tonsils, removed seven years ago when he was five, led to the discovery of a blood disorder so rare it is known internationally as Tangier Disease.

Since the Teddy Laird case was discovered, 10 others have been found in the world. The disease is marked by a near absence of alpha lipoprotein (fat and protein) in the blood.

His sister, Elaine, has the second case, and there are six other known cases in America. The others are in Portugal and Switzerland.

Dr. Donald F. Fredrickson said that except for the unusual tonsils, the malady has had little effect on her children. "Elaine is very seldom sick," she said, "and Ted just has a cold now and then."

STARTED IN '60

Events leading to the discovery of Tangier Disease began in April 1960, when Teddy was sent to National Institutes of Health's Emmen-Potomac Memorial Hospital for a tonsillectomy.

"Ted would just lie around," Mrs. Laird recalls. "We thought he was just sickly. It wasn't a cold. It was his tonsils and adenoids doing it."

Dr. J. Thomas Edmondson, who performed the tonsillectomy, was intrigued by the abnormally large, "grayish yellow ... indolent (swollen) tonsils."

Edmondson had a pathologist at the Naussauvax Hospital make microscopic sections to be sent to the Armed Forces Institute of Pathology in Washington.

These, Teddy's mother said, were diagnosed as a rare fat storage disorder, possibly Hand-Schuller-Christian Disease or Nieman-Pick Disease.

FEARED WORST

In late April 1960, Teddy and his mother left their Cherrecheau Bay Island home for the National Institutes of Health in Bethesda. Mrs. Laird was referred to the National Cancer Institute.

She spent the next seven weeks in Bethesda. "It almost ran me crazy, because I thought he had leukemia," Gladden Laird, Teddy's father, said.

Gladden Laird, the most of Tangier's population, is a waterman. He works with his father, crabbing during the warm months and oystering in the winter.

At the Cancer Institute, the preliminary diagnosis of Hand-Schuller-Christian Disease was dropped, and Teddy was sent to the National Heart Institute, where rare fat storage disorders are studied. The tentative diagnosis was Nieman-Pick Disease.

Dr. Fredrickson said that the doctor "could not find a lung." Teddy's 35-year-old father recalled recently, "I was really getting on edge."

NAMED FOR ISLAND

Nieman-Pick Disease is a genetic disorder usually suffered by infants. Its symptoms include malnutrition, stomach and intestinal disturbances, and abnormalities in blood-transporting organs. It usually is fatal.

The disease also is marked by nervous system disorders. Teddy suffered no nervous disorders, so that diagnosis was dropped. The malady was labeled Tangier Disease.

In November, Dr. Donald F. Fredrickson and some of his associates from the National Institutes of Health came to examine tonsils and take blood samples of Tangier's population.

They came by mailboat from Crisfield, Md. This is the only really close connection between this island and the mainland. The mass examination was conducted in the island medical center, then uncropped because Tangier had no doctor.

Elaine Laird was found to have large, oddly-colored tonsils like her brother's. She was admitted to N.I.H. for a tonsillectomy.

It was the first tonsillectomy ever performed at N.I.H. Dr. Fredrickson said. Another Tangier Islander's tonsils were removed there. They served as controls, for comparison to those of the Lairds.

Blood samples were taken from 127 people alike to Teddy and Elaine, and from 98 others for use in community controls.

Dr. Fredrickson said he found Tangier's population "very helpful and hospitable."

Tangier's closely related population proved a poor community control. Investigation revealed that only 17 of the 98 controlers did not have at least one relative common to the Laird children.

No other islanders were found to have the bizarre, unusually-colored tonsils associated with Tangier Disease.

CONGENITAL

However, each of the Laird children's parents and grandparents were found to have a deficient amount of alpha lipoproteins. This condition had been observed in blood samples from Teddy and Elaine.

This indicated to researchers that Tangier Disease is "no doubt," is genetically determined, and not necessarily confined to Tangier Island, Dr. Fredrickson said. "It just happened that the first case was found on Tangier." He labeled the discovery "very exciting."

Researching Tangier Disease, Dr. Fredrickson said, "he found a tremendous help in unraveling some of the secrets of how fats are transported in the body."

While admitting that the research has been a costly procedure, Dr. Fredrickson said it has "reaped us many fold in new information about body fat transport."

"This is particularly important," he said, "because some abnormality in the body transport fat through the blood is one of the causes of arteriosclerosis (hardening of the arteries). And arteriosclerosis causes more deaths than any other disease."

MAY BE 'KEY'

This new information about transportation of fats, the doctor said, "could be one of the keys to the solution of the No. 1 health problem in America and world of the world."

He said researchers are trying to determine the chemical content of alpha lipoproteins. Studying Tangier Disease, he said, "has been an important factor in developing modern explanations of how lipoproteins do their job."

Dr. Fredrickson explained "one of the possible relationships between heart disease and lipoproteins."

"Alpha lipoprotein is much higher in the blood of women than in men. Alpha lipoprotein (abnormal in Tangier Disease) levels are clearly related to the level of tonsils and adenoids."

"Women, before menopause, have about one quarter the liklihood of having a heart attack as men the same age."

"It is not known why women have this special protection, and doctors still think it is possible that this may be related to the higher alpha lipoprotein of women."

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TANGIER CLUES

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Dr. Fredrickson said the discoveries made in research of Tangier Disease are "pieces to a great puzzle."

The doctor said there is "an evidence as yet that Tangier Disease leads to more atherosclerosis."

Eight of the 10 people who have Tangier Disease have been to the National Institutes of Health, and blood samples from the others have been sent there.

Except for the cases of Teddy and Elaine Laird, they all were discovered "because of how cholesterol levels," Dr. Fredrickson said.

Mrs. Laird and her swallows husband took their children to N.I.H. for yearly examinations.

CHILDREN HELPED

"We've had many a doctor to look at us," Gladden Laird said.

Mrs. Laird said Teddy, now 15, and Elaine, 13, appear to be in almost perfect health. "Ted's just like a cricket," she said, "and I don't worry about Elaine at all."

She said she has seen a definite improvement in her children since they had the tonsillectomies. "They're a bit better. He can play longer, and he's a more active," she said.

Elaine, Mrs. Laird said, "eats real good, and she's in real good health."

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