Tangier Disease

Tonsils May Cure Hearts

By JOHN PRUITT

Virginian-Pilot Staff Writer

TANGIER ISLAND — Scientists believe that the discolored tonsils of a Tangier Island boy, Teddy Laird, may provide an important clue in 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 disorder 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 England and Switzerland.

Mrs. Peggy Laird 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 Northampton-Accomack Memorial Hospital for a tonsillectomy.

"Ted would just lie around," Mrs. Laird recalled recently. "He was just sickly. It wasn't a cold. It was his tonsils and adenoids doing it."

Dr. J. Thomas Edmonds, who performed the tonsillectomy, was intrigued by the abnormally large, "grayish yellow . . . lobulated (and) wrinkled tonsils."

Edmonds had a pathologist at the Nassawadox hospital make microscopic sections to be sent to the Armed Forces Institute of Pathology in Washington.

There, Teddy's malady was diagnosed as a rare fat storage disorder, possibly Hand-Schuller-Christian Disease or Niemann-Pick Disease.

FEARED WORST

In late April 1960, Teddy and his mother left their Chesapeake Bay island home for the National Institutes of Health in Bethesda, Md. Teddy was referred to the National Cancer Institute.

He 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, like 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 Niemann-Pick Disease.

"One doctor said Teddy couldn't live much longer," Teddy's 38-year-old father recalled recently. "I was really getting on edge."

NAMED FOR ISLAND

Niemann-Pick Disease is a genetic disorder usually suffered by infants. Its symptoms include malnutrition, stomach and intestinal disturbances, and abnormalities in blood-forming 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 daily connection between this 2x3-mile island and the mainland. The mass examination was conducted in the island medical center, then unoccupied 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. Frederickson said. Another Tangier islander's tonsils were removed there. They served as controls, for comparison to those of the Lairds.

Teddy Laird

Dr. Fredrickson

Elaine Laird

Blood samples were taken from 127 people akin to Teddy and Elaine, and from 90 others for use as 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 90 volunteers did not have at least one relative common to the Laird children.

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

CONGENITAL

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

This indicated to researchers that Tangier Disease "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, "has been a tremendous help in unraveling some of the secrets of how fats are transported in the blood."

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

This is "particularly important," he said, "because some abnormality in the way the body transports 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 much of the world."

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

Dr. Frerickson 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 (absent in Tangier Disease) levels are clearly related to the level of female sex hormones.

"Women, before menopause, have about one quarter the likelihood 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 possible that this may be related to the higher alpha lipoprotein of women."

The importance of the abnormality of Tangier Disease (absence of alpha lipoproteins) to the research of heart disease is this:

TANGIER CLUES

"Cholesterol is transported by alpha and beta lipoproteins. Arteriosclerosis is associated with the laying down of cholesterol in the walls of the blood vessels. In the same manner rust inside a pipe will cut down the bore of the pipe, this accumulated cholesterol cuts down the flow in the vessels."

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

The doctor said there is "no evidence as yet that Tangier Disease leads to more arteriosclerosis."

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 low cholesterol levels," Dr. Fredrickson said.

Mrs. Laird and her seafaring husband take 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 12, 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. "Ted's a lot livlier. He can play longer, and he's more active," she said.

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

"I believe they helped them up there," Laird said.

Laird had high praise for Dr. Fredrickson. "There's not a better person in the world," he said, "I'd do anything under the sun for him."

Dr. Fredrickson said he feels a "deep affection" for Tangier's 900 residents.

He said that for Teddy's or Elaine's offspring to inherit the disorder, both parents would have to be carriers of the gene producing Tangier Disease. The chances of this, Dr. Fredrickson said, are slim.