

April 2000 -

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In Memory of Isabelle (Deater) Perrin



- Oldest of the 12 children of Alvin and Ellen Deater
- Spouse: Lynn Perrin
- Daughter: Anita (married to Vincent Mirabella)
- Granddaughter: Lynn Ellen Goldstein
- Great grandson: Palika Goldstein

Research Developments

Ellen Burns, Vice President

Research done at the Cecil B. Day Laboratory for Neuromuscular Research has narrowed the area, first described by researchers in Australia, that contains the gene responsible for Hereditary Sensory Neuropathy 1 (HSN 1), the disease that has been in the Deater family for generations.

Dr. Khemissa Bejaoui, the primary researcher, and Dr. Robert Brown, the Director of the Laboratory, have demonstrated personal interest in finding the actual gene that has gone defective. The focus of the work during the last six months has been on analyzing the pattern of expression of the genes that are located in the region identified as containing the causative gene. The actual gene remains unknown.

Because the whole field of genomic research is new and is constantly evolving, the data available to help with the analysis is rapidly expanding. Just keeping up with new information and techniques is time consuming! Khemissa has collaborated with many different groups, including the researchers in Australia, to add to the knowledge already available to her.

By analyzing the pattern of expression in the genes, Khemissa can focus her search on genes that are expressed in nerve cells. Since the disease is a neurological one, the gene that is defective is somehow getting the "wrong information" to the nerve cells. This technique allows her to concentrate on the genes that are most likely to be involved in the disease. The work takes a great deal of time and is exacting.

For six months Khemissa had a student helping with the project as a laboratory technician. He is no longer at the lab, and we were not able to financially support a technician, as we had hoped. Khemissa, with Dr. Brown's help, has applied for additional grant money from Muscular Dystrophy Association. She hopes to apply for a large grant from the National Institutes of Health later this year. There is great competition for these grant monies, but her previous work puts her in a good position to qualify.

The complexity of the genome is hard to imagine. It has been said that "if we decoded your genome--the sequence of the DNA in your chromosomes--and you could read one letter of the genetic code per second, it would take you over 100 years to read your own genetic code." The section of "chromosome 9q22" that the researchers are looking for that will unlock the mystery of the "Deater disease" is a piece of the whole human puzzle. We are blessed to have knowledgeable researchers and a caring family to continue to search for a cause and cure.

Treasurer's Report

Nancy Newcomer, Treasurer

Nancy newcomer, Treasurer, reports the following regarding the treasury of Deater Foundation, Inc.:

1999 Finances	
Balance as of 1/1/1999	\$35,436.85
1999 Contributions	15,605.00
1999 Interest	<u>488.25</u>
Total Income	\$51,530.10
1999 Expenses (Feb. & July)	-31,000.00
Balance as of 12/31/1999	\$20,530.10

Local School Bus Driver Suffers Minor Burns (Interesting "Old News")

Alvin Deater had a narrow escape from serious injury Monday morning when a can of motor oil exploded in his hands and set his house on fire. Mr. Deater, who drives a school bus, was heating the motor oil in a large can on the back of the kitchen stove, in preparation to start the bus on its morning run. The oil caught fire and Mr. Deater grasped the can in his hands and rushed for the door. As he reached the porch, the can exploded, scattering burning oil over the porch and sides of the house. The flames soon began to eat their way through the siding of the house and it was with considerable difficulty that they were extinguished. Aside from slight hand burns, Mr. Deater was uninjured.



**Reprinted with permission from The Dallas Post "Only Yesterday" Column (70 Years Ago--February 14, 1930)*

Excerpts from Grandma's Diary (Easter Weekend, 1945)

Submitted by: Beulah Womer

March 31--We killed 5 chickens and have them all ready to put over to cook. We cooked for dinner and for supper. June, Bob, and everyone was here for supper. After supper we died eggs and fixed baskets; then Isabel, Lynn and Lynn E. went up with June and Bob for the night. Tom and Lois and boys went to Berwick right after supper.

April 1--Henrietta fixed up some April fool eggs and caught a lot of them. We had 20 for breakfast this morning and 25 for dinner. Carl Jr. is not well yet but played pretty good and Mona didn't feel so good this afternoon, but all seemed to have a pretty good time. Mona called at Montrosses a few minutes this afternoon. Tom and Lois not back in time for dinner. Bob and June took Russell to the bus. Edith came with a boyfriend for her and Helen and they went back to Elmira together. *April Fool eggs were deviled eggs (with hot pepper concealed in the center).

Genomics 101

Submitted by: Ellen Burns--Vice President

In nearly every cell of every living organism, there exists a complete set of instructions for creating that organism and regulating its cellular structures and activities over its lifetime. That set of instructions is called a *genome*.

A genome is organized into distinct, microscopic units called chromosomes. Chromosomes are coiled threads of deoxyribonucleic acid--DNA--that is composed of two long chains of nucleotides bound together in pairs to form a double helix. (A nucleotide is one of the building blocks of nucleic acids, such as DNA. A nucleotide is made up of three parts: a base, a sugar, and a phosphate. The bases lie flat like steps of a staircase. The sugar and phosphate form the backbone of the nucleic acid. DNA is composed of four different kinds of nucleotide.)

Three and a half billion of these nucleotide pairs make up the human genome.

Specific sequences of nucleotide bases within a DNA strand--called genes--are the cells' instructions for producing proteins. Scientists estimate that 80,000 to 100,000 of these basic units of heredity exist within the human genome. Proteins perform a wide variety of physiological tasks. They facilitate processes

such as digestion, breathing, immune responses, the production of heat and energy, and the movement of fluids in and out of cells.

While most members of a species have the same collection of genes, each individual's unique characteristics stem from slight variations--called polymorphisms--in the sequence of nucleotides that comprise the genes of that individual. On average, the DNA of any two individuals will differ by about 0.1 percent.

Other types of variations--called mutations--also occur. Both polymorphic and mutagenic variations may be harmful to an individual by inhibiting the production, or altering the normal function, of protein. Most diseases result from these types of genetic variations.

The goal of genomic inquiry is to identify the sequence of nucleotides, understand the function of every gene they comprise, and clarify the genetic variations that define individuality and create disease.

Credit: www.celera.com--Celera Genomics



**On Couch--Helen Wilson, Betty Rudolph, and Verna Adams
In Front--Mona Montross, Beulah Womer, Martha Dennis, and June Sorber**

Final Note

Contributing to the Deater Foundation is a very tangible way to fight for a better quality of life for those in our family who are affected with this disease. All the money that is received is spent for periodic payments to the Day Neuromuscular Laboratory for research. Dr. Brown and the Day Lab have consistently used this money to leverage for other money, so that the comparatively small amounts we contribute are the "seeds" that grow into grants from other sources.

As I continue to talk with doctors about this disease, it is clear that current medical approaches are unable to provide much help. Our best hope lies in understanding the disease at the cellular level. To begin, we must find the defective gene, and learn what it does. Khemissa says, "I am very confident. I know we are going to make it." With everyone's help, that day will come soon.

If you have any questions, comments, or suggestions about our newsletter, the medical report, etc., or if you are interested in contributing to the Deater Foundation, send your e-mail message to: tcdor34@enter.net. Or, you may also write to:

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PO Box 255
White Deer, PA 17887

**56th Annual Deater Reunion –
Saturday, July 15, 2000
at the grove in the Stull**