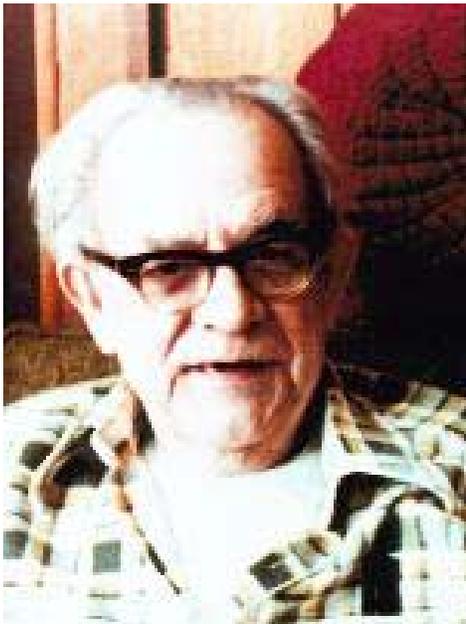


June 2002 -

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In Memory of Harvey Alvin Deater

- Third child and eldest son of Alvin and Ellen Deater.
- May 27, 1910 to January 25, 1979
- Married to Charlotte Thompson February 20, 1944
- Four children: Ellen, Joan, Larry, and Chris
- Four grandchildren and 2 (+) great grandchildren

Harvey initiated, and supported research into the "Deater Disease," often serving as a research subject. He was a valuable resource for doctors around the country. His legacy includes the research that is being done today and the cure that will come in the future.

Medical Research Update

Ellen Burns, Vice President and Medical Liaison

Last year we announced in the DFI Newsletter that the gene causing HSN I (the Deater Disease) had been identified and confirmed by



research teams in Australia and Massachusetts. The researchers studied 12 families and identified three different mutations in the gene. Everyone who had one of the mutations in the gene had the disease. This was fantastic news! Armed

with this knowledge, scientists can now begin to look for the fundamental reasons for the cause of the nerve damage.

Each cell in the human body contains all the information needed to make a whole person. This information is stored in the nucleus of the cell in material called DNA. Each person inherits half his DNA from his mother and half from his father. There are millions of ways the combinations of DNA can be put together, making each person unique. The DNA is arranged into chromosomes, and organized on the chromosomes into genes. Genes make proteins, and proteins make enzymes.

Genes control how the body looks and functions. The chemical building blocks of a gene can be likened to the letters in a word. Like children's letter blocks, each block represents a specific letter. If one of the blocks, or letters, is out of place the whole word, or gene, can be altered. For example, CAT can become HAT.

Just as a sentence containing an incorrect word will make no sense, the cell doesn't recognize the incorrect "word." For example, "The hat jumped onto the table." The "bad" protein, produced by the affected half of the gene, isn't recognized, and isn't working. The "good" protein is produced by the gene that comes from the parent who doesn't have the disease. Usually this is not a problem, because we usually make much more of the protein than we need.

But something else is happening at the same time. The "bad" protein produces an inhibiting effect on the "good" protein. When the "good" protein works to make an enzyme, the "bad" protein interferes. So the result is that there is a decrease in enzyme activity.

In the Deater Disease, the gene SPTLC1 tells the cells to make one part of the serine palmitoyltransferase enzyme. This enzyme is expressed (produced) in every cell in the body. This enzyme is known to affect the production of a fatty substance (glycosyl ceramide) in the body. This substance belongs to a class of fats known as sphingolipids. Sensory nerves seem particularly sensitive to decreased amounts of sphingolipids.

Research needs to be done to determine if giving nerve cells more sphingolipids will prevent damage to those cells. Other possibilities are that the "bad" protein may affect the cell in other ways that we don't yet understand. The Deater Foundation hopes to continue to support such research until a cure is found!

Treasurer's Report

Nancy Newcomer, Treasurer

2001-2002 Finances	
Balance as of 5/31/2001	\$21,528.06
Income 6/1/2001 to 12/31/2001	12,985.00
Interest 6/1/2001 to 12/31/2001	263.46
Income 1/1/2002 to 5/31/2002	1,360.00
Interest 1/1/2002 to 5/31/2002	108.04
Total Income	\$36,244.56
July 2001 Expense	8000.00
May 2002 Expense	8000.00
Total Expenses	16,000.00
Balance as of 5/31/2002	\$20,244.56

A Sphingolipids Conference

Submitted by: Ellen Burns

In January of this year, Dr. Khemissa Bejaoui participated in an important conference in San Francisco. The Gordon Conference draws top scientists together to share their newest research findings. This conference was all about Sphingolipids.

Lipids are chemical fats. Cholesterol is a familiar example of one kind of lipid found in the body. Sphingolipids are made by molecules joining together to produce fatty acids that attach to a kind of backbone structure. Long chains of these compounds form together. The structure is so complex that the discoverer named these particular lipids after the Sphinx, the mysterious pyramid in Egypt!

The research that Khemissa has done proves that the gene defect that causes HSN I, the Deater Disease, decreases the production of one kind of sphingolipid, called glycosyl ceramide. Many researchers all over the world are interested in sphingolipid production. It appears that sphingolipids may play a role in many diseases, not only HSN I, and not only neuromuscular diseases. This means that scientists working in diverse fields, and studying many different diseases may be interested in finding out what happens when the genetic defect associated with HSN I decreases sphingolipid production.

Khemissa gave a Poster Presentation in which she gave examples of the disease in our family. Two other teams of researchers collaborated with her on

the presentation. One group is from the Dermatology Service, Veterans Administration Medical Center, and Department of Dermatology, University of California, San Francisco. The other group is from the Department of Biochemistry and Cell Biology, National Institute of Infectious Diseases, Tokyo, Japan.

Our family has been greatly blessed since the genetic defect was found. Scientists know what the gene is supposed to do. Many scientists in many different areas have been studying the action of sphingolipids in healthy cells for many years. Some are very interested in studying cells with the defect to find out what goes wrong. Once we know what goes wrong, scientists can determine how to treat or correct the problem.

We have a great opportunity to respond to the blessings we have received with support for continued research. God has taken our donations and, like the loaves and fishes, has multiplied them to keep the research moving forward. The banner for the Poster Presentation has a tag line:

Supported by:

- The Deater Foundation
- Muscular Dystrophy Association
- The Neuropathy Association

Thanks to all who contribute with your donations and your prayers. It is making a difference!

Excerpts from Grandma's Diary (Winter in the Stull)

Submitted by: Beulah Womer

January 16, 1945--It was snowing awful hard this morning, but the bus came and the children went to school; but at 1:20 they were all home again as it was snowing and blowing so hard that school let out at one o'clock. I had done up the work all but beds, so I baked a cake and worked the butter. Verna helped Alvin start the Delco. We had an early supper, made snow ice cream and read.

February 6, 1945--Verna has a cold and didn't go to school today; so I washed and she hung them up. Although cold in the morning, it got real warm...the warmest day for some time and a swell wash day. We washed one double blanket and every thing dried, but Verna had to wade snow to her crotch to hang some of them up, and Martha took them down.

Recent Funding Developments

Submitted by: Ellen Burns

Dr. Robert H. Brown, Jr., Director of the Day Neuromuscular Laboratory, is pleased to report that he has found a source of funding for the next year in the amount of approximately \$50,000. This will supplement the 2-year Muscular Dystrophy Association grant (\$35,000/year) that Dr. Khemissa Bejaoui obtained last year. This will keep the project intact for another year while the Lab continues to look for federal support. Funds from the Deater Foundation, Inc. continue to "prime the pump," providing starter funds that encourage other donors.

Dr. Brown points out that the mutations were first found two years ago this August, and the papers announcing the discovery have been out about a year. He says, "Clearly, we are miles ahead of where we were two years ago. Hopefully, we can use the present information to generate the critical animal and petri dish models that will let us begin significant therapeutic studies." Our goal remains to find a treatment for HSN I to benefit those with the disease today, and to prevent the disease in future generations.

Miracle of Hand Controls

Chris Deater Christensen shared with us recently the following account of her miracle. She wrote: "Through a friend of a friend of mine here in Colorado, I am now the proud owner of hand controls on my car. I had been pricing hand controls for about a year and was having trouble deciding if I could justify the cost. Amazingly, out of the blue my friend calls and says this friend of hers, Janet Desgalier, no longer drives and wonders if she knows anyone that can use the hand controls on her car. This was like a miracle to me. Then came the question of who could take them out of one car and install them into my car without great cost or the inconvenience of leaving my car for more than a day somewhere. Here comes my second miracle, Dave Bartholomew. He and Joan were living about two hours south of Longmont at that time. They drove up to Longmont, and in a few hours on a cold Saturday before Christmas, he had the transformation completed. The kindness of all of the people involved in this adventure has made life so much easier for me. With the use of the hand controls, I can drive anywhere with confidence. As all of us Deaters know, anything that can help to keep us independent is extremely important. I am so grateful to Janet and Dave who made this miracle happen for me."

What About Khemissa?

Submitted by: Ellen Burns



Dr. Khemissa Bejaoui will attend the Deater family Reunion this year. Khemissa has worked tirelessly at the Cecil B. Day Laboratory for Neuromuscular Research for 12 years, most of that time seeking the cause and cure of HSN 1. Khemissa will present the results of her recent work and collaboration with scientists in California and Japan. She will talk about future plans for research into the disease.

Most of Khemissa's relatives live in Tunisia. She has "adopted" the Deater family as her American family. She has taken a personal interest in the research. Larry Deater and Ellen Burns, who have worked closely with the researchers at the Day Lab, are proud to count Khemissa as a friend as well as a colleague.

Khemissa has interested scientists around the world in the importance of research into HSN 1. She and her friend Paul have hosted researchers at their home and gained from them commitments to work on the project. She has selflessly shared with other researchers the results of years of her hard work, so others can use the information to advance knowledge about the disease. She believes deeply and passionately in this calling. We are blessed indeed that she is in our "family!"

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, email the webmaster at: tcdor34@enter.net. Or, you may also write to: Deater Foundation, Inc., PO Box 255, White Deer, PA 17887.

**58th Annual Deater Reunion->
Saturday, July 20, 2002
at the grove in the Stull**