

Deater Foundation, Inc.

Disease Research Report

Submitted by: Ellen Burns, Medical Liaison



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**Reunion
Reminder!**

Hope to see you
at the 61st
Deater Reunion
on July 16, 2005
at the Grove
• in the Stull!

On a rainy Sunday morning in May, Dr. Robert H. Brown Jr. drove north to Kittery Point Maine to have lunch with Larry Deater, Rory Robb, and Barry and Ellen Burns. Dr. Brown is the Director of the Cecil B. Day Neuromuscular Laboratory at Massachusetts General Hospital. Besides catching up on one another's lives, the meeting was to provide an update on the research being done on HSN-1. Hereditary Sensory Neuropathy Type 1 is the name of the disease that affects many members of the family of Alvin and Ellen Wilson Deater.



Dr. Brown has long been a champion of research on this rare genetic disease. It has been more than twenty years since Dr. Brown met Larry at Massachusetts General Hospital. Through the subsequent years Dr. Brown has met many members of the Deater family and has doggedly pursued the genetic cause of HSN-1. Dr. Brown is an internationally known expert in amyotrophic lateral sclerosis, or Lou Gehrig's disease. Most of his research focuses on that devastating neurological disease. But he has also dedicated extensive time and resources to the search for a cause

and treatment for HSN-1.

Dr. Brown and those who work with him have collaborated with scientists from around the country and the world to unravel the mysteries of this rare disease. This collaboration has led to the rapid duplication of experimental results, very important to the verification of research. This collaboration continues with scientists

who have a wide variety of interests.

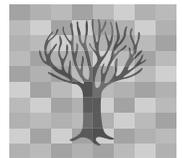
Mice:

Last year we reported on the funding of a postdoctoral position for Alex McCampbell to develop transgenic mice.



Alex works in Dr. Brown's lab and has experimented with laboratory

mice to alter their genetic makeup to mimic the genetic missense (or error) found in people with HSN-1. He has been successful in producing mice with the altered gene, and the



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Website News: The Forum

Submitted by: Dixie Dorward

For many years the Deater Foundation has maintained a presence on the web in order to serve as a resource tool for family members. In fact, because there is little documentation on the disease many family members have also referred the site to their personal doctors as a quick reference tool.

We have always depended on free web space in the past. Relying on free space has caused the web address to change several times and has been too complicated to remember. In November 2004, the Deater Foundation decided to purchase space on the web in order to provide an official resource for the family members and family doctors. You can now access the web site by going to

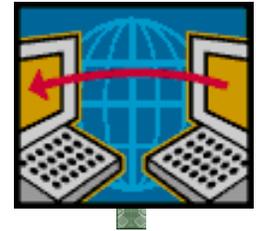
www.deaterfoundation.org

New to the site is a secure forum that is available only to family members. The forum allows for further communication with family in

order to: gain advice, relay what therapies have worked and what has not worked, share new experiences (has anybody else encountered this before?), and even discuss reunion information or even disability issues. The possibilities for the forum are endless and will serve as a valuable resource for the future generations that may experience similar situations. This is a perfect resource to share your experiences or questions with family members who are going through, or who have gone through, the very same thing.

In order to access the forum, you will need to acquire a username and password, which helps to keep the forum secure. Simply email

ddorward@deaterfoundation.org for access to the forum. Please identify how you are related to Alvin Deater. Following receipt of your email, you will receive the information you need to enter the family forum.



We're on the Web!

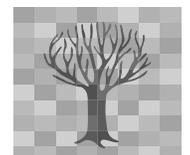
www.deaterfoundation.com



“Check out the new website where there is a secure forum that is available only to family members.”

<p>Not on our Mailing List? New Address?</p> <p><i>Notify</i> Carol Dorward at cdorward@deaterfoundation.org <i>of your current home address and/or email address</i></p>
--

<p>Website Comments, Suggestions, Questions, etc. Contact ddorward@deaterfoundation.org</p>



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Verna (Deater) Adams' Recipe for Chili

Submitted by: Carol Dorward

- ◆ In a large cook pot, pour about 1 tablespoon of oil. Add about 2 stalks of celery and one small onion, cut up small.
- ◆ Simmer about 10 minutes.
- ◆ Then add about 3/4 pounds of hamburger; stir and cook on medium heat until the hamburger is browned (about 10 more minutes).
- ◆ Drain off the fat.
- ◆ Add 1 can of tomato soup, 2 large cans of diced tomatoes, 1/4 teaspoon of chili powder (optional), 1/4 Cup sugar (optional), salt and pepper to taste to the hamburger mixture.
- ◆ Simmer about 20 more minutes, stirring frequently.
- ◆ Then add a large can of kidney beans and simmer the pot another 15 minutes.
- ◆ Serve with rice or mashed potatoes.

Send your favorite family recipe for our next publication to:

cdorward@deaterfoundation.org

[Be sure to include whose recipe it is and the relationship of that individual to Alvin or Ellen Deater.]

A Story...“From the Old Homestead”

Submitted by: June Deater Sorber

In the Fall of 1938 Mom made lots of pumpkin pies. We closed the back kitchen off to make the rest of our eight-room house warmer. We had two stoves and burned wood and coal. Mom would put her pies in the back kitchen on the table. She taught Sunday School in the living room, and I [June], who was 16 years old, taught the little ones in the dining room.

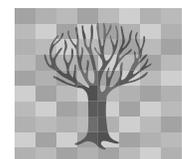
Mom started having Sunday School just for us because no one had cars then. The next-door neighbor's children came; and then

from a mile or so away other children came from Stone Town.

A cute little three year old boy (Jimmy Webber) was in my class--the other children all older. He didn't sit too long and busied himself around the place. After Sunday School was over, Mom opened the door to the back kitchen and said "What happened to my pies?" Little Jimmy piped up and said, "Them pies sure was good, Mrs. Deater!" Several fists full of pie were missing. I can still hear Mom's laugh to this day. Our family of twelve didn't leave them go to waste!



“Them pies sure was good, Mrs. Deater!”



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“In Memory Of” Henrietta Deater Rudolph

Submitted by: **Betty Lou Rudolph Jorden**

Henrietta Deater Rudolph

6th child of Alvin & Ellen (Wilson)

January 26, 1917 to June 16, 1993

Married Donald James Rudolph

Son - Donald A. (Married Evelyn Smallwood)

Son - Jerry A. (Married Judy London)

Daughter - Betty Lou (Married Dale L. Jorden)

5 Grandchildren

5 Great-grandchildren

* * * * *

When asked, “How do you remember your Mother?” Jerry Rudolph responded with a statement so simple, yet profound-- “You and I may not remember her the same.” So, I ask **you** now, “How do **you** remember her?”

...The daughter, sister, wife, or mother, who held high the banner of her heritage

...The spiritual leader and mentor of many

...The neighborhood parent, willing to stop work and go for a swim or play ball with the kids

...The hostess with an open door

...The chef with a table full of home made delicacies and occasional

“mystery meat”

...The cook, who could turn egg sandwiches and hot chocolate into a feast for a gang of young people

...The green thumb lady, who used her flowers to brighten the lives of the sick and elderly

...The rosy-cheeked marksman, who loved tromping through the woods, or silently fishing a beautiful lake, tuned to every sight and sound of nature

...The learner, who graduated valedictorian of her class--age 65

...The community support person for “Life Line,” who simply “made” time to visit

...The visionary, who saw potential, and taught a deaf person to drive, thus providing independence for two

...The friend and counselor with an expert’s ear for listening, and a heart looking for the good in all

...The champion, who finished the race on earth with the same courage that she faced her challenges in life

He was so right, you know. We may not remember her the same. But my guess is, we will all remember her...



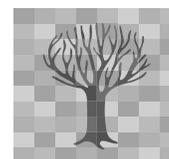
Henrietta Deater Rudolph



Graduated valedictorian of her class--age 65!



“We may not remember her the same. But my guess is, we will all remember her.”



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Disease Research Report *(Continued from Page 1)*

mice have reproduced the mutation in subsequent generations. Dr. McCampbell has shown that mice with the genetic defect develop small nerve fiber disease, as in HSN-1. A paper for publication of these findings is in progress.

Biochemistry:

Also mentioned last year was the collaborative research project with Dr. Theresa Dunn and colleagues at the Uniformed Services Medical School in Bethesda, Maryland. Dr. Dunn's expertise is in examining the sub cellular workings of the protein that is altered by the genetic error. The gene codes for serine palmitoyl-transferase long-chain base subunit 1 (SPTLC1). Dr. Dunn's research investigates exactly how the protein functions in the body. An article published in December 2004 suggests where within the cell the action of this subunit may take place. Information obtained at this cellular level will inform exactly what is happen-

ing to cause the disease.

Worldwide interest:

Dr. Garth Nicholson is the researcher in Australia, who in 2001 published the article identifying the exact area of the genetic defect. Dr. Bejaoui and Dr. Brown reported the same finding from studies of the Deater family in the same publication (although the exact error in the Deater gene was different from that in the other various families studied.) Dr. Nicholson continues to actively study the disease, publishing in March of 2004 a research paper documenting a 44% reduction of SPT activity in certain blood cells (lymphocytes) from HSN-1 patients. Dr. Nicholson recently visited Dr. Brown in Boston, and they continue to support one another's work.

Dr. Brown says, "Our goal is to get as many smart people to look at this (disease) from as many angles as possible."

(Continued on page 8)



"Mice with the genetic defect develop small nerve fiber disease, as in HSN-1."

Gramma Sutton's Chocolate Mayonnaise Cake

Submitted by: Larree Brown

Getting Started:

Grease an 8" square pan.

Preheat oven to 350.

Mix

2 C. Flour

1 C. Sugar

1 C. Mayo

1 C. Hot Water

1 Teaspoon Vanilla

2 Teaspoons Baking Soda

8 Tablespoons Cocoa

Finishing Up:

Bake for about 40 minutes or until inserted toothpick comes out clean.

Allow cake to cool

Ethel Deater Sutton was the second child of Alvin and Ellen Deater



Spingolipids Research

Submitted By: *Ellen Burns, Medical Liaison*

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 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. When the essential components of the gene are out of order or missing, the gene is mutated (changed). Then, the half of the gene that comes from the affected parent produces “bad” protein that isn’t recognized, and isn’t working. The gene that comes from the parent who doesn’t have the disease produces “good” protein. Usually this is not a problem, because we usually make much more of the protein than we need.

In HSN-1, the gene SPTLC1 encodes (tells the cells to make) one part (long chain base one) of the serine palmitoyltransferase enzyme (SPT). This enzyme is expressed (produced) in every cell in the body. Cells in people with HSN-1 show a decrease in SPT activity. 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.

Sphingolipids are a type of lipid found in cell membranes, particularly nerve cells and brain tissues. Sphingolipids are important in cell structure, cell-to-cell communication, and signal transduction. The lipid backbones of the structure of sphingolipids regulate diverse cell behaviors, including cell death.

Many scientists are interested in how sphingolipids act in the body. At least one researcher, Alfred Merrill, Jr., Professor and Smithgall Chair in Molecular Cell Biology at Georgia Institute of Technology, believes that dietary sources of sphingolipids may help in the treatment or prevention of disease. Dr. Merrill’s research involves colon cancer, but he suggests that it would be interesting to see if eating a diet of foods high in sphingolipids had any bearing on the disease HSN-1. Milk and soy products are two categories of food high in sphingolipids.

It would be interesting to know if any family members with the disease have had, over the years, a diet high in those two foods. Some people with HSN-1 are beginning to supplement with these products to see if diet can make a difference. If you have any thoughts about this, please contact Ellen Burns and/or share your thoughts with other family members on the Deater Foundation website.

We’re on the Web!

www.deaterfoundation.org

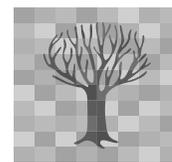
Cells in people with HSN-1 show a decrease in SPT activity.

New research suggests that “...Dietary sources of sphingolipids may help in the treatment or prevention of disease. .”

Milk and soy products are two categories of food high in sphingolipids.

Direct questions or comments on diet to:

eburns@deaterfoundation.org



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Deater Genealogy

Submitted By: Larry Deater [Son of Harvey Deater, Grandson of Alvin Deater]

In 1884, twenty years old Thomas Deiter of Buck Township, Luzerne County, PA, married young Henrietta Anthony. They may have been married by the Justice of the Peace from Stoudertsville...who, several years earlier, performed the marriage of Thomas' older brother William, and Lucy Andrews.

Thomas and Henrietta (Etta) had six children: Alvin (1884-1953), Charles (1886-1919), David (1889-1957), Anna (1891-1960), Mae (1894-?), and Alice (1905-?). By 1900, the family had moved to Lake Township in Luzerne County where Thomas earned his living as a day laborer.

Alvin married Lydia Ellen Wilson, and they settled down in Lake Township to start their own family, eventually having twelve children. Ellen is given credit for codifying the spelling of the name DEATER. Sometime between 1910 and 1920, the family moved to Market Street in Noxen Township, Wyoming County, where Alvin started a butcher shop....And to this day, Alvin and Ellen's offspring celebrate our family with an annual reunion not too far

from that old butcher shop.

I have been doing genealogy research on the family for some time now and the going has been slow but rewarding. If anyone has anything to share, I would be thrilled. That may include stories, letters, birth, death or marriage certificates, etc. The most significant facts may be found in the most insignificant seeming articles.

Some specific questions I have. Does anyone have info on...

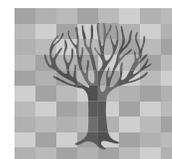
- ⇒ Lilly Hummell, William Mac, Ida Herber, Aunt Clara,
- ⇒ Deiter's church in Moore Township, Northampton County, Pa
- ⇒ The parents and siblings of Henrietta Anthony
- ⇒ Any family connection to the Midwest, particularly Wisconsin or Ohio
- ⇒ Thomas Deiter's siblings
- ⇒ Parents of Charles Deiter

"Deiter"/"Deater"

????????

Ellen [Wilson Deater] is given credit for the spelling of the name DEATER.

Submit information to
Larry Deater at:
robbdeat@comcast.net



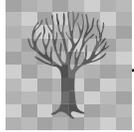
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DEATER FOUNDATION, INC.

Disease Research Report (Continued from Page 5)

P. O. Box 255
White Deer, PA 17887



We're on the Web!
www.deaterfoundation.org

Remember that your contributions to DFI are tax deductible.

Mail Contributions to:

**Deater Foundation, Inc.
c/o Mrs. Nancy Newcomer
P. O. Box 255
White Deer, PA 17887**

Aging:

Dr. Brown recently spoke with researchers at the National Institute of Aging. Dr. Brown noted that even those who have the gene for HSN-1 are not born with the symptoms. The disease gets worse with age. Aging causes the degeneration of neurons, over time. Might aging play a part in the expression of the disease? The answer to that question might impact a host of other degenerative neurological diseases. Dr. Brown sees a potential for collaboration with researchers in aging.

Funding:

Like everything else, research takes money. Dr. Brown has been instrumental in finding and applying for funding to pay for researchers, equipment, and overhead. Currently, a grant from the National Institutes of Health pays for Alex McCampell's work with the HSN-1 mice. Over the years the Deater Foundation has contributed thousands of dollars to support the mission to find a cure. Progress is being made on many fronts, so it is increasingly important to press on with research, building on studies and experiences from across the years and around the world.

A Foundation is "an organization or institution with a provision for future maintenance." A foundation is "the basis on which something stands." We have been blessed with a foundation formed by love of God and family. Standing on this foundation, we have the opportunity to build a life free from the pain and debility of HSN-1 for the generations to come. Each contribution to the Deater Foundation is another building block for this hope!

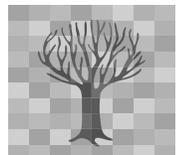
DFI Treasurer's Report (May 31, 2004, to May 31, 2005)

Submitted by: Nancy Newcomer, Treasurer



DFI Statement of Account

Balance as of 5/31/04	\$39,167.22
Income 6/1/04 to 12/31/04	1,6980.00
Interest 6/1/04 to 12/31/04	73.46
Income 1/1/05 to 5/31/05	650.00
Interest 1/1/05 to 5/31/05	97.57
June 2004 Expense	- 8000.00
Nov 2004 Website Charge	- 54.95
Balance as of 5/31/05	\$48,913.30



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