



CLINIC FOR SPECIAL CHILDREN NEWSLETTER

VOLUME 1 NUMBER 2

* LANCASTER COUNTY, PENNSYLVANIA *

SPRING, 1991



DEDICATION AND OPEN HOUSE CELEBRATION JUNE 1, 1991 11:00 am

On April 24 we moved into the new Clinic along Bunker Hill Road in Strasburg. The accomplishment of this building through donations, community support and volunteer work was remarkable and reflects the spirit of determination and hope that has made the clinic possible.

PLEASE JOIN US!

On June 1st, 1991, we will celebrate the new clinic. We will begin around 11:00 am with a dedication and ribbon cutting by some very special children followed by an open house and chicken barbeque lunch. The open house will continue in the afternoon. You may also want to bring along a shovel to help plant a tree. As part of the dedication a fruit tree will be planted for each family with glutaric aciduria or maple syrup urine disease. These families inspired the work that led to the Clinic for Special Children and the clinic is really the fruit of their suffering and their work.

We hope everyone who has helped to make the clinic possible in one way or another can be a part of this celebration. Directions The clinic is located at 535 Bunker Hill Road. From the center of Strasburg at the intersection of routes #741 and #896 go south on Decatur Street which becomes May Post Office Road. One mile from the center of Strasburg turn right onto Shaub Rd. which runs into Bunker Hill Road. Follow Bunker Hill Road up and over the hill. The entrance to the clinic is a new lane at the bottom of the hill on the left between a field and a pasture. If you can join us and are from out of town we will be happy to give you directions to Strasburg and suggestions for local inns or motels. Please call Caroline Morton at the clinic (717) 687-9407 or (215) 687-5691 for more information.

THE NEW CLINIC

The timbers for the building were raised on November 17th in a traditional "raising" and even a cold morning rain did not dampen the spirit of the day. Reuben Riehl's crew volunteered their labor to lead the raising with about fifty volunteers. Nearly every day of the construction volunteers, many of them parents of children who are cared for by the clinic, showed up to help. Hank Kline of Lancaster had the difficult job of coordinating the work of sub-contractors and volunteers to build the Clinic from a beautiful blend of fir beams, cedar clapboard with a foundation of Lancaster stone, and a roof of 140 year old recycled slate. Enos Hoover, a clinic board member not only dedicated many hours of his own but also recruited many others to work with him. A group of Amish and Mennonite women painstakingly painted the windows, walls and trim.



In all, over 250 work days were donated by volunteers. Through the community involvement in the work, the building became more than Dr. Holmes Morton's clinic and many families now think of the Clinic as part of their community.

Jacob and Naomi Stoltzfoos who now have two granddaughters diagnosed with glutaric aciduria made the land available to us at half the assessed value. Many local contractors and businesses also donated or discounted materials and services for the clinic including:

Roger Fry, Surveyor: Survey and Site Plan
Strasburg Township: Building Permit
Collins-Rosenberg, Architects: Building Design
Chuck Dougherty: Design Consultation
Riehl's Construction: Timberframing labor
Reuben Stoltzfus: Excavation and site work
Hartman Underhill & Brubaker: Legal Work
Farfield: Job Trailer and Electrical Design
Wickes Lumber: Most of the Lumber needed
Conestoga Wood Specialties: All of the Cabinets
W.F. Mold Co.: Windows at 50% discount

Myers Brothers: Well
 Groffdale: Concrete work discount
 ZimmermanSteel: Crane
 Thomas Trucking: Discount on Concrete
 Phillip and Deborah Good: Plumbing Fixtures
 Ray's Plumbing: Plumbing Installation discount
 Leonard Hurst: Truck and Lift
 Armstrong Foundation: funds for building materials
 Martin Limestone Inc.: Gravel
 Dana Ressler: Landscaping
 Chris Sann: Landscaping
 JMS Builders: Labor
 Mose King & Co.: Labor
 Christ Stoltzfus & Co.: Labor
 Musselman Lumber Co.: Donation for supplies
 PP & L: Donation
 Ginrich, Smith, Klingensmith & Dolan: Legal Work
 P.L. Rohrer & Bro.: Grass Seed
 Mannie's Wood Shop: Discount on Cupola
 Conestoga Drywall: Discount on Installation
THANKS TO ALL!

PROMISE AND PROGRESS A REPORT OF THE CLINIC'S WORK

Within a year and a half of the first diagnosis of **glutaric aciduria** among the Amish in Lancaster County the Clinic had established a voluntary testing program to screen Old Order Amish infants for the disorder. In the Willow Street lab between February 25, 1990 and April 24, 1991 we measured glutaric acid levels in 684 urine samples from Amish newborns. Six asymptomatic infants with glutaric aciduria were found, were started on therapy at the clinic, and have not shown signs of brain damage. Two other infants who had not been tested as newborns were diagnosed at 6 and 15 months of age after severe illnesses and extensive brain damage. The contrast between the six healthy infants and these two children with lasting damage reminds all involved of the importance of newborn testing. The number of samples analyzed during this 14 month period represents 60-70% of the infants in the Lancaster Amish community. Beyond the importance of the fact that 60-70% of Amish infants are now being tested, the number reflects an increased awareness of the disorder within the Amish community and among Lancaster County health care providers. Only two years ago glutaric aciduria was unknown, unrecognized, and would not have been considered a treatable condition. It is estimated that at least one in 200 Amish infants will inherit the disorder.

As important as the testing of newborns, is the new information about the treatment of the disorder to prevent brain damage. The Clinic laboratory now routinely measures glutaric acids levels in the blood of affected children to monitor therapy and as part of the evaluation of children during acute illnesses. The damage is caused by episodic increases in glutaric acid concentration in the blood and early signs of brain intoxication can be reversed by rapidly lowering the glutaric acid levels. Over the past 18 months 9 children, including two newborns and a 22 month old, have required hospitalization because of early signs of intoxication. All were admitted under Dr. Morton's care to Lancaster General Hospital and in each case it was possible to lower the blood glutaric acid levels to a safe range within 6-24 hours. None of the children showed progression of brain damage during the hospitalizations.

Maple Syrup Urine Disease: In August 1990 an amino acid analyzer was purchased for the Clinic from Hewlett-Packard with money contributed by local Mennonite churches. The analyzer is used to diagnose and monitor

treatment for Mennonite children who have maple syrup urine disease, a biochemical disorder of amino acid metabolism that like glutaric aciduria causes severe brain damage and death of infants if not recognized early and treated carefully. Between August and April more than 500 blood amino acid analyses were done for purposes of diagnosis, treatment and research. Nineteen children who have MSUD have made 53 visits to the clinic in the past year. Ninety five blood samples from children with MSUD were analyzed. Of these samples 31 were analyzed because children were ill. Through early evaluation and simple changes in therapy all but three of these cases avoided hospitalization. Two children were hospitalized at Lancaster General for 24 hours because of dehydration, one was referred to Philadelphia Childrens Hospital. Over the past year in Pennsylvania three children were diagnosed with MSUD. Two of these were diagnosed at the Clinic at less than 48 hours of age when they were asymptomatic. Neither child required hospitalization and have remained well. The third case was diagnosed elsewhere at nine days of age. She was very ill when recognized, was hospitalized for two weeks in Philadelphia, and her medical bills over the past year have totaled more than \$95,000.

The most important goals of the Clinic are early recognition of glutaric aciduria and maple syrup urine disease combined with early intervention during episodic metabolic illnesses to prevent severe illnesses, brain damage, prolonged hospitalizations, and extraordinarily high medical bills. The experiences of the first year clearly show the need and the effectiveness of the Clinic.

Other Disorders: In addition to glutaric aciduria and MSUD, Dr. Morton has diagnosed several other metabolic disorders among Amish children which may be as common as GA and MSUD and medically equally important. One of these disorders is 3-methylcrotonylglycinuria, which has been called *tom cat urine disease* because of the odor produced when a child is ill. The disorder is rarely diagnosed, there are fewer than ten confirmed cases in the medical literature. Two additional cases were recently diagnosed by Holmes Morton in one Amish family in Lancaster County. Although the condition may cause life-threatening episodes of illness, two of the previously reported patients have survived and are well. Both of the Amish children are now well and with therapy similar to that for glutaric aciduria should continue to develop normally. We have already started to test for 3-methylcrotonylglycinuria as part of our routine newborn testing and expect to find many other cases over the next year. Another disorder that affects a large Amish kindred is a very rare form of aldosterone deficiency which is associated with growth failure and episodes of severe dehydration and sodium deficiency. Although first recognized among the Amish more than 10 years ago, all of the first children diagnosed then were lost to follow-up medical care and there was no subsequent effort to recognize the condition in other Amish families. Over the next few months we hope to establish a method to diagnose the defect at the Clinic and provide medical care for affected children. There are more than 10 other genetic disorders that afflict Amish and Mennonite children who come to the clinic. Although many of these disorders cannot be treated in any specific way, and some are rapidly lethal, in each case the child and the family need help and explanations which too often were not found or offered in the past.

IMMUNIZATION PROGRAM

The clinic recently started an immunization program for families of children regularly cared for by the clinic. Most clinic patients have started their vaccinations, but many of

their siblings have not. Amish children are not required to have immunizations to attend Amish schools. Over the years many families have avoided immunizations for a variety of reasons including cost, transportation problems, misinformation about side effects and the lack of a relationship with a health care provider. Recent widespread outbreaks of measles, German measles and whooping cough in the Lancaster area have underscored the need for immunizations as an essential preventative measure. **Lancaster General Hospital** is donating the vaccines needed. As children are immunized they will also have a general pediatric check-up including height and weight measures to promote a more preventative approach to medical care rather than a crisis approach. The clinic has hired part-time a family nurse practitioner, Deborah Kennedy, to coordinate this program and to help with general patient care. Debbie already knows many of the families because she does six-week examination of infants for Pat Payne and Rhondi Anderson, certified nurse midwives who helped start the newborn testing program. We are happy to have Debbie work at the clinic and hope as time and resources permit we will be able to increase her role.

WHAT ARE WE LEARNING?

What has been learned about the diagnosis and treatment of glutaric aciduria over the past year has importance beyond the immediate needs of the Amish children who are cared for by the Clinic. The disorder is more common throughout the United States than many appreciate. More widespread use of the methods used in our newborn testing program would undoubtedly lead to recognition of many more cases of the disorder. There are specialists in genetic diseases who do not understand the biochemical basis of glutaric aciduria and do not consider the disorder to be treatable. The method of measuring blood glutaric acid levels is new and is used only at the Clinic. Our recent observations about the relationship between blood levels and illness, and new information about the effectiveness of therapy during acute illnesses should significantly change the approach to therapy and outcome of all infants with the disorder. Further, glutaric aciduria is one of a family of biochemical disorders that produce a cerebral palsy-like paralysis. In our patients it is apparent that intoxication and brain damage most often occurred between ages 1 and 36 months. Such observations about the natural history of the disorder in the Amish children may lead to a better understanding of why the infant brain is more vulnerable to these biochemical disorders and lead to better methods to prevent biochemical injuries of all types to the infant brain. The first long publication about the Amish form of glutaric aciduria is in press in the American Journal of Medical Genetics and more publications and lectures will follow as time allows.

Many have commented on what the clinic must mean for Amish and Mennonite families who need these specialized diagnostic and medical services. It is our hope that the work of the clinic will help prevent devastating effects and serve as a needed local resource for families coping with the chronic problems these disorders present. Over the last year and a half we found extraordinary support to begin and build the clinic. We also met unusual challenges as we set up the clinic to meet health care needs of Amish and Mennonite families. The plain cultures have much to teach the rest of us and in many ways are leading us to a better approach to health care. The system of community support for families who have "special" children is remarkable. Families' insistence on home care and inclusion of a severely disabled child in the center of all family activity gives the rest of us an example. We are learning how to translate highly technical information into practical applications that families

can understand and use resourcefully. We are learning more about setting up a health care service as a non-profit operation with community support to guide it.

Working with plain cultures who do not depend on health insurance leads us to re-examine how our health care system works, what it lacks, and seek alternative ways of providing care. The emphasis on self sufficiency within the community has shown us how to accomplish what needs to be done without waiting for someone else to do it. The plain cultures give us a reflection of the needs in our larger culture. We hope over the next few years the success of the clinic and what we learn in this community will serve as a model for other communities which are seeking better health care services.



NEED FOR FUNDS

The Clinic for Special Children was built entirely through donations from many generous individuals and foundations. Contributions also fully equipped the diagnostic lab with state of the art equipment including a mass-spectrometer and gas chromatograph donated by Hewlett Packard. Because of these gifts the clinic is able to provide the highly technical diagnostic and medical services needed at reasonable costs to families, most of whom do not have health insurance. Our basic monthly operational costs are met partially through fees for service. However, if we are to continue to have time to do research, write, and lecture, as well as expand services and provide long-term complex medical care then other support is needed. We estimate that fees for clinic services and laboratory tests will meet only about 60% of the total cost of operation during the next few years. The following describes some of our needs.

CLINIC SERVICES:

Based on the increasing number of children with complex chronic health problems followed by the clinic we need to increase the role of the nurse practitioner and eventually hire another physician to work with Dr. Morton to cover the case load and follow-up services. Unlike most pediatric practices, the clinic's caseload is limited but each case is intensive in the management of a child's care during even minor illnesses. Funds are needed immediately to help pay the nurse practitioner's salary to coordinate the immunization and follow-up services.

RESEARCH FUND:

Clinical research is essential to learning more about effective diagnostic techniques and treatment of certain inherited metabolic diseases. As an approach to meeting this cost for years to come we are interested in establishing an endowment type fund which will provide a source of continuous revenue for this important aspect of the clinic.

RESERVE FUND:

After all the bills for construction have been met, our reserve fund is almost depleted. This fund helps us meet our monthly expenditures in excess of our monthly income from fees and provides support for maintaining equipment and supplies.

Contributions would be welcomed, appreciated and used wisely! THANKYOU. (Our tax ID number: 23-2555373)

GREETING CARDS TO BENEFIT THE CLINIC

Photographer Bill Coleman of State College, PA donated eight of his photographs of Amish children for greeting cards to help raise funds for the clinic. A grant from the Oxford Foundation provided the up-front costs of printing by Acorn Press of Lancaster. The 5x7 cards are sold in sets of eight at \$10 per set. Eight dollars of the cost is a contribution to the clinic. Cards can be ordered from the clinic.

P. BUCKLEY MOSS

In April the clinic was one of the beneficiaries of an auction sponsored by the P. Buckley Moss Society at their Pennsylvania convention. The auction was lively and we are grateful to have been selected as a recipient.

Photographs by Jim Stansbury



*Frank Allen, Dr. Rick Kelley, Dr. Holmes Morton
On the way to raise the clinic*

AMISH ARTIST

Amish artist Susie Riehl donated the sale of her lovely watercolor of an Amish farm house to the Clinic for Special Children. The funds will support a regular newsletter for families who use the clinic to share new information about treatment of glutaric aciduria, maple syrup urine disease and other disorders the clinic studies, and to promote an exchange of helpful ideas and advice among families with special children.

Susie's painting of children riding in a horse drawn buggy and titled "The Clinic for Special Children" is being sold as a print by the Double Heart Gallery in Intercourse, PA to benefit the clinic. We are grateful to Susie and to Shirley Wenger of the Double Heart Gallery for their support. For more information about the print call the gallery at 717-768-3522.

The Clinic for Special Children is a non-profit diagnostic and primary medical service for children with inherited disorders in Lancaster County, Pennsylvania. The clinic serves Old Order Amish and Mennonite families who suffer from a high incidence of genetic diseases. Glutaric aciduria among the Amish and maple syrup urine disease among the Mennonites are two common biochemical disorders that require a complex approach for successful treatment. The clinic's approach includes an infant testing program for early diagnosis, primary medical care to prevent devastating effects of metabolic diseases during common childhood illnesses, clinical research to improve treatment, and services to support the needs of parents. The clinic also cares for children who have complex needs due to other genetic disorders some of which are yet to be identified. Research at the clinic seeks to unlock these riddles.

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