

# CLINIC FOR SPECIAL CHILDREN NEWSLETTER

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\* LANCASTER COUNTY, PENNSYLVANIA \*

SEPTEMBER 20, 1990

## PROMISES

The Clinic for Special Children holds promise. Only two years ago Dr. Holmes Morton first recognized that glutaric aciduria was a common cause of cerebral palsy among Amish children. The concept of the clinic grew as he obtained evidence that brain damage from the disease was preventable.

First attempts to find support for research and start the clinic were frustrating, but an article by Frank Allen in the Wall Street Journal brought significant interest and support. With the donations received, Hewlett Packard's gift of diagnostic equipment, and loan of temporary office space by Lancaster General Hospital, the Clinic for Special Children opened in January. These gifts have made a difference in the lives of many children, their families, and in the life and work of Holmes Morton. We promised to keep you informed of our progress. This is the first edition to keep that promise.

## THE CLINIC

The clinic 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. In addition to the two diseases already mentioned, the clinic cares for children who have complex needs due to other genetic disorders such as severe congenital heart disease related to Ellis van Creveld dwarfism, and a lethal form of muscle disease leading to heart and respiratory failure. Many children with three other genetic disorders of unknown cause just as common and severe as glutaric aciduria are cared for by Dr. Morton. Research continues to unlock these riddles.

### Clinical Research

The clinic has an active clinical research component with a well equipped metabolic laboratory. All of the laboratory equipment was donated, which drastically reduces the cost of analysis. One of our goals is to demonstrate that relevant clinical research can not only provide important scientific information but also improve health care by focusing research efforts upon specific problems. We hope to develop an approach to managing complex illnesses that can serve as a model for improving health care in rural areas (or in any other area) where there is a concentration of a set of related diseases or health care problems affecting many in a community.

## Family Resource and Library

The clinic serves as a resource for families to help them better understand the nature of metabolic diseases and how to improve home care for chronically ill children with complex medical needs. We've held family meetings and workshops on home care and recent clinical research findings. We are developing a resource library including local genealogies, low-protein and other diet guides crucial for children with MSUD and glutaric aciduria, and medical literature on related topics for families to use.

## INFANT SCREENING FOR GLUTARIC ACIDURIA

With the cooperation of local nurse midwives and physicians the clinic now provides a testing service for Amish infants at risk for glutaric aciduria using the new gas chromatograph-mass spectrometer. It is estimated that one in 200 Amish infants will inherit the disorder. All Lancaster Old Order Amish infants are considered at risk.

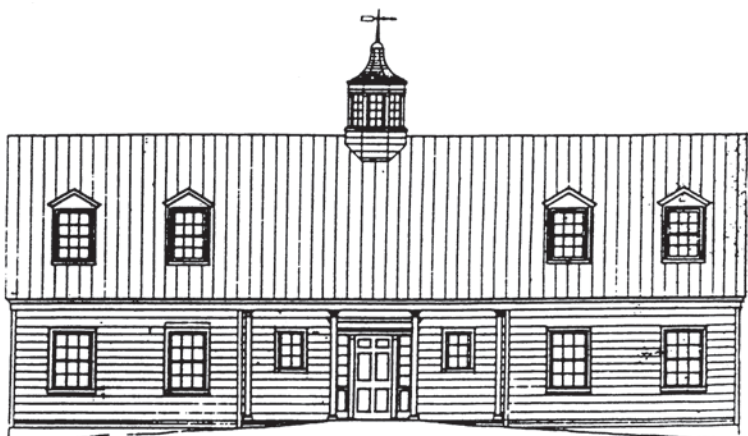
Since January over 400 infants and older siblings have been screened for the disorder. Three infants have been diagnosed which brings the total number of cases to 24. *Only eight cases of the disorder were previously reported world-wide.* Of the newly diagnosed cases, two of the infants remain well. None of the children has shown progression of the paralysis and complications associated with the disease while on therapy. Some of the children with severe damage have had improvement in motor skills and muscle control.

Among the Amish it appears glutaric aciduria is responsible for 50% of the cases previously designated as cerebral palsy. Little is known about how widespread it may be in the general population or about the long term effects of the disease. Cases recently diagnosed in several older children raise questions about the relationship between high glutaric acid levels and psychiatric disorders and other forms of movement disorders related to basal ganglia damage. Clinical studies to better understand this and other similar metabolic disorders is an essential function of the clinic.

## LAND FOR A CLINIC

Land for the clinic was made available on a farm near Strasburg, Pennsylvania, by the grandparents of an Amish infant diagnosed with glutaric aciduria. *The baby, now a year old, is being treated by Dr. Morton for the disorder and is a healthy toddler who has not suffered any of the damaging effects of the disease.* The two and a half acre site is a grassy hillside meadow surrounded by trees, cornfields and easily accessible by car or horse and buggy. The site has received preliminary approvals by both Strasburg Township and the Lancaster County Planning Commission. Since January the clinic has been operating in temporary office space generously provided by Lancaster General Hospital until a permanent site can be built.





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ARCHITECT

## "RAISING" A CLINIC

In mid November we hope to begin construction of the new clinic. The traditional timberframe building was designed by Chuck Dougherty of Lancaster County in collaboration with Collins-Rosenberg Architects of Philadelphia who donated their services. The building design includes space for the metabolic diagnostic laboratory. Equipment for the lab has been provided by Hewlett Packard, local Mennonite churches and many other donors. In addition to the main floor patient examining rooms and office space, a medical and genealogical library is designated for the upper floor. The lower level will include a large room for parent meetings, workshops, and extra offices as our needs expand.

The frame for the building will be raised in the Amish tradition with help from many volunteers as in barn raisings. Some materials for the building have been donated including lumber from Wickes Lumber Co. and metal roofing from Joseph & Associates of Lancaster. Contributions received over the past year will help meet some of the cost of the building, but more financial help is needed.

## COSTS AND CONTRIBUTIONS

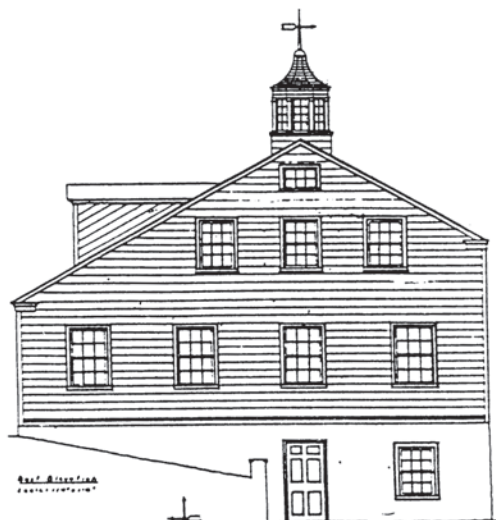
The clinic wants to encourage Amish families to have their children tested soon after birth and is committed to providing this and other medical and diagnostic services at reasonable fees. Our emphasis is on early diagnosis and prevention of serious illnesses which can cause the paralysis and death at an early age. Most Amish and Mennonite families do not have health insurance and pay their own medical bills as they can. Since all of our equipment was donated we are able to set our fees at affordable levels.

The test for glutaric aciduria costs \$20 to perform. This same test at major medical centers costs around \$300.

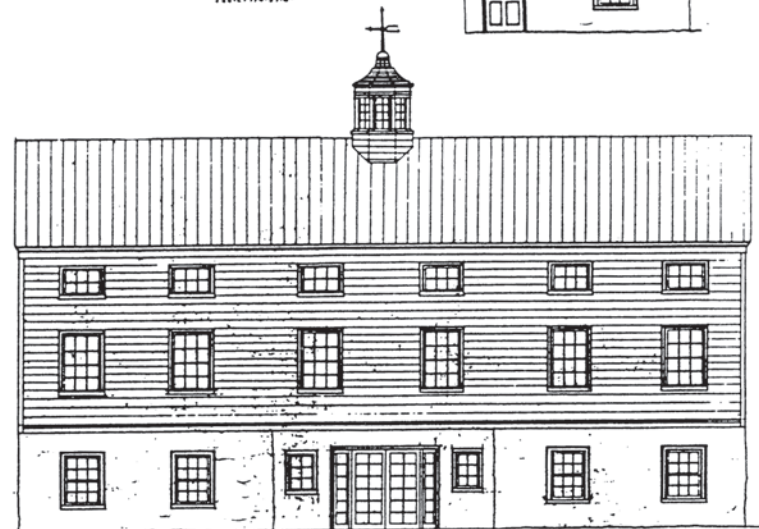
Instead of billing families, we have suggested they contribute the cost of the test (\$20) or whatever they can to the clinic and pass information about the infant testing on to their Amish neighbors. Most Amish families have contributed the suggested cost, many have contributed much more. About 40% of newborn Amish are now being tested and we hope to reach at least 90% within the next year.

One goal of the clinic is to be as financially self sufficient as possible within five years through a combination of fees for service and a base of local community support. This base of community wide support will take time to build.

Although fees for services will help meet some of the costs of operating the clinic, they will never cover the entire operating cost, particularly the research component which is so vital to improving treatment and increasing understanding of metabolic diseases. Many children seen at the clinic



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require extensive review of previous records, test results, lengthy talks with parents, and "thinking" time. This costs a lot in time and effort and many of these children previously have not received the medical care they need for this reason. How to resolve these issues over the long term for families with chronic, complex medical needs but without insurance or government support or a high fee schedule is a challenge with which we will continue to wrestle and need support for the short term.

## HELP FROM THE MENNONITES

Maple syrup urine disease is a common metabolic disorder in the Pennsylvania Mennonite community. The disorder is similar to glutaric aciduria and is characterized by the body's inability to degrade the branched chain amino acids resulting in potentially toxic levels of essential amino acids. **Diagnosis must be made within 24 to 48 hours after birth to avoid damaging effects and death.** Children born with MSUD must be on strict low protein diets and blood levels of the branched chain amino acids must be carefully monitored especially during infectious illnesses.

Although Children's Hospital of Philadelphia and St. Christophers Hospital, also in Philadelphia, have made significant progress in treating the disorder over the last twenty years, the necessity of improved local care has long been recognized by parents as essential to improving long term outcome for children who have MSUD. Common childhood illnesses such as colds and ear infections can provoke life threatening metabolic illnesses if the children are not carefully monitored.



This summer two Lancaster Mennonite Churches raised most of the funds needed to purchase a \$56,000 amino acid analyzer for the clinic. A generous donor who wishes to remain anonymous completed the funding needed. This analyzer, made by Hewlett Packard, provides essential testing for newborns at risk and immediate testing for MSUD children as needed. Our cost is \$45 for analysis compared to \$200 to \$400 at major medical centers. The clinic is dedicated to improving care and providing immediate, accessible resources for parents.

### SUMMER RESEARCH PROJECT

Over the summer Tom Lundquist, a senior medical student from Johns Hopkins University, did a research internship with the Clinic for Special Children. Dr. Morton and Tom were able to show that a simple urine test a parent can do at home in ten minutes and costs less than 5 cents can be used to detect increased concentrations of branched chain acids. One family monitored their daughter for 100 days and showed that even with careful dietary management the branched chain levels were often increased but with simple day to day adjustments in therapy metabolic control was markedly improved. We hope this simple test will allow families and local health care providers to recognize affected newborns before the onset of severe illness, and allow families to routinely monitor therapy and metabolic control, thereby preventing many hospitalizations. Tom Lundquist contributed to the development of important laboratory tests, and also observed the unique approach to care the clinic provides for children with complex diseases.

We hope to continue such fruitful research fellowships for medical students and to encourage residents to experience and learn through the clinic.

### CHARACTERS IN THE CLINIC

In a recent discussion with an Amishman about the clinic building design, the young but wise man said "most Amish don't care as much how the building looks as about the characters inside". Therefore a word or two about the characters inside.

Dr. Holmes Morton now works full time with the Clinic for Special Children and is the director and its only physician. He performs all the diagnostic laboratory work, provides medical care for children seen at the clinic and continues his clinical research into inherited metabolic disorders. He still makes housecalls using his white Honda, sometimes with Morton children along for the ride. Once or twice this year he has found time to fly fish for trout for an hour or two. His ties to the community of research and academic medicine have been retained through his continued appointment in pediatrics at Johns Hopkins University. He has given lectures about his work at Johns Hopkins, at local Lancaster County hospitals, in Boston, Hartford, Pittsburgh and West Virginia. Recently his research on a related topic was published in the Journal of Pediatrics and his work on glutaric aciduria has been submitted for publication. Thanks to many generous people and Frank Allen of the Wall Street Journal, his dream of a clinic happened without having to mortgage the Morton house. The support and encouragement he received over the past year gave him a strong sense of obligation and an opportunity to practice the kind of medicine he strongly feels needs to be practiced.

Caroline Morton also works at the clinic as the Associate Director for Administration. She manages the non-profit organizational and financial aspects of the clinic and serves on the clinic board as treasurer. Her prior work was in

research, planning and administration in education and community services with special interests in the needs of rural schools and building school / community partnerships. She holds a masters degree in administration from Harvard University and is an artist when she has the time. The Morton's three young children, ages 3, 6, and 8, also are often found in the clinic usually playing house under the examining table, counting cups out of the dispenser or watching the squiggles come out of the mass spec machine. Their favorite activity, however, is to spend the day with some of their Amish friends, chase chickens and ride in a buggy while their parents work in the clinic.

The person who really keeps the clinic running is Rebecca Huyard. Rebecca is the Office Manager and is responsible for the day to day management of the office, appointments and patient record keeping. She has set up and maintains the computer data base on all infants tested in the clinic. Rebecca is Old Order Amish and has several nephews and a niece with glutaric aciduria. She often serves as an interpreter explaining in Dutch the information families need to understand about the disease and is extremely dedicated to reaching any and all families who need the clinic. Prior to her work at the clinic she was the office manager for a local Amish business and taught in a one room Amish school for fifteen years.

Dr. John Hostetler, a noted sociologist and author of *Amish Society*, serves on the clinic board of directors. Dr. Hostetler, a Mennonite, was Amish by birth and has devoted his academic career to enlightening the general population regarding the Plain sect cultures. He provides invaluable insight into the development of clinic services to appropriately serve the needs of the plain families.

Dr. Richard I. Kelley also serves on the board. He is a pediatrician, geneticist and specializes in metabolic diseases at the Kennedy Institute and is on the faculty of Johns Hopkins University. He is especially concerned with the kind of care children receive with chronic metabolic diseases. Dr. Kelley sees patients at the clinic once a month and volunteers for call when Dr. Morton is away. Dr. Kelley is co-author with Dr. Morton on a recent article in the Journal of Pediatrics and the recently completed article on glutaric aciduria.

Enos Hoover was recently elected to the board as one who truly understands the role of this clinic and the needs of parents with special children. He is a Mennonite dairy farmer and has had four children born with maple syrup urine disease, one of whom survives. As parents, Enos and his wife know far more about this disease and the need for local care than any "expert".

Penny and Dick Armstrong each have given their time and expertise to making the clinic a reality. Penny, a certified nurse midwife, served on the board the first year and her enthusiasm helped get the infant testing program off the ground. She is the author of *A Midwife's Story* and the recently published *Wise Birth*. Dick is an airline pilot and engineer with an incredible ability to get things done. His guidance through the initial planning and permit phase of building the clinic was invaluable.

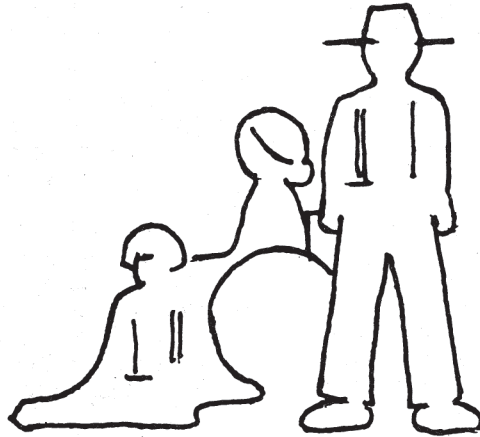
## OUR NEEDS

Our thanks go to Frank Allen and his insightful writing for the Wall Street Journal, and many generous contributors who made the clinic possible. Several people provided their expertise to help us open the door. Dr. Hugo Moser at the Kennedy Institute in Baltimore supported the initial research on glutaric aciduria by Dr. Morton and Dr. Kelley. Dr. Harvey Levy in Boston gave advice and encouragement. Andrew Dohan, attorney, provided the essential legal advice and Gail Bentley, CPA, of Cogen, Sklar, Levick provided sound financial guidance to set up the non-profit organization. Stephen Heumann helped with our initial planning. Senator Arlen Specter of Pennsylvania and his staff helped us gain our tax exempt status in record time so the clinic could open, begin to test infants at risk, and provide needed care without waiting on red tape. Lancaster General Hospital gave us a place to work until we can build our own. **Thanks to all.**

For those wishing to contribute and needing this information, our tax ID number is 23-2555373.

With the generous contributions sent over the last year, we were able to make much progress in providing needed medical care and diagnostic services. The clinic has been quickly acknowledged, used, and supported by Old Order Amish and Mennonite families. Dr. Morton has eagerly committed his career to this effort along with his family. We have most of the equipment we need, but we do need a permanent facility from which to fully develop this unique clinic. The land has been provided, plans have been drawn, volunteer labor is ready, but we need help to meet the cost of the building. The other area of need is to help with the cost of the clinical research activity at the clinic. For any additional information please call Caroline Morton at (215) 687-5691 or (717) 464-0180, or write to the following address:

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