



CLINIC FOR SPECIAL CHILDREN NEWSLETTER

VOLUME I NUMBER 12

* LANCASTER COUNTY, PENNSYLVANIA *

WINTER 1996

*The wolf shall dwell with the lamb,
the Leopard shall lie down with the young goat,
the calf and the lion will eat together,
and a little child shall lead them.*
Isaiah 11:6



THE GIFT OF THE AUCTION

Greetings to each of you from all of us at The Clinic for Special Children. Flu season is early and the Clinic is busier than usual in spite of flu shots. We do want to take time to thank all those who support the Clinic with their gifts of time, talent, and contributions. The work here would not be possible without what each of you gives. It sustains us in many ways and gives hope to all who seek help here.

THANK YOU

The auction represents a gift the community here gives to the Clinic as an expression of care and hope for all children, especially those who are born with difficult medical conditions and those whose future is uncertain. The Clinic now cares for several hundred children in our region of Pennsylvania with over sixty-five different metabolic disorders and genetic syndromes, some of which are newly identified by physicians associated with the Clinic. In his remarks at the Auction, Dr. Morton indicated the contribution the clinic is often called to provide toward the diagnosis and improved care for children not only in our area but in other states and other countries is

really a gift from the children and families that come to the clinic and from those in the community and elsewhere who support the work here.

Every year we think the auction just can't get any bigger or better, but it has, once again. The event held on Saturday, September 21, was a great success and we wish to thank all who made it possible- the Auction Committee, volunteers, the many individuals who donated over 3,000 items, and of course all those who participated that day. **THANK YOU** on behalf of the Clinic, the children and the families who depend on us for their medical care. As in previous years, all proceeds go directly to the Clinic to support its annual operating budget. These funds help purchase supplies and maintain laboratory equipment, help meet monthly costs of running the Clinic, support clinical and laboratory services available to children with unusual genetic disorders at a reasonable cost to families. It is always difficult to determine how many attended the auction, but judging from the number of buggies tied up and the cars parked in the adjacent field, it was a record attendance. There were over 1,000 registered bidders. Quilts were sold at an average of \$386 which was lower than last year but consistent with other recent auction prices. There were 88 large quilts all locally hand made and donated for the sale and as many wall hangings in a variety of patterns. The log cabin playhouse brought the highest price of the sale as we had hoped. Outdoor furniture also sold very well this year. There was another pony and a handsome new cart for sale this year but it did not go home with the Morton children ! One of the members of the auction committee kept an interesting tally of the record amount of food consumed during the day which gives some sense of the dimension of the event (we did not ask him to put it to the tune of the "Twelve Days of Christmas" but this time of year it comes to mind).

On the third Saturday of September there were

3 pigs roasted;

7 cases of lemons squeezed into 1075 cups of fresh lemonade;

66 gallons of chicken corn soup;

130 fresh strawberry pies.

and 500 Shoo fly pies on the bake sale table!

1224 ham & cheese sandwiches;

1800 servings of french fries;

2400 scoops of ice cream;

2860 freshly made subs;

3200 soft pretzels baked;

3348 chicken legs & 550 chicken halves barbecued;

and 10,000 donuts made all in one day!

Thank you to the members of the Auction Committee who for the sixth year gave tirelessly of their time and energy to make the auction such a success: Mr. and Mrs. Leonard Hurst, Mr. and Mrs. Enos Hoover, Mr. and Mrs. Harvey Hoover, Mr. and Mrs. John Fisher, Mr. and Mrs. Steve Huyard, Mr. and Mrs. John Stoltzfus, Mr. and Mrs. Steve Beiler, Mr. and Mrs. Jacob Zook, Jr., Mr. and Mrs. Daniel Stoltzfus, Mr. and Mrs. Ernest Zimmerman, Mr. and Mrs. Daniel Lapp, Mr. John Shirk, Mr. Ralph Atkinson, and Miss Rebecca Huyard.

Next auction date: September 20, 1997 !

NEW GREETING CARD TO BENEFIT THE CLINIC

A new greeting card is for sale to benefit the Clinic. The card features a painting in soft water colors of the auction by Sarah Morton, age 12, daughter of Dr. and Mrs. Morton. The cards are sold in a pack of 5 for \$7.50 (\$7.95 including PA tax) and can be ordered by contacting the clinic. The inside of the card is blank and the back gives a brief description of the Clinic's purpose. All proceeds support the Clinic's operating fund. Our thanks to Sarah for sharing her artistic talent and enthusiasm for the auction. Note: There are a few sets of the Bill Coleman cards still available, but the supply is very limited.

FUNDRAISER IN ROCHESTER, NY

For the fourth year, family and friends of the Colby family in Rochester, NY, sponsored a pig roast fund raiser to benefit the Clinic for Special Children. Laura Colby receives her care for maple syrup urine disease from the clinic as do several other children with MSUD in that region of New York. This year the group raised a total of \$5675 which will be used by the clinic to further its work to improve diagnosis and care for MSUD. Thank you to the Colby's, Ultra Fab for their matching gift, and to all who contributed to the event.

GA 1 CONFERENCE HELD IN GERMANY

The second European conference on glutaric aciduria- type 1 (GA1) was held in Germany from September 29 to October 2, 1996. This conference was hosted by Prof. Dr. Georg Hoffmann of Klinikum of Philipps Universitat in Marburg, Germany, and followed up last December's meeting hosted by the Clinic for Special Children. Holmes Morton, M.D., Clinic for Special Children, and Steve Goodman, M.D., University of Colorado were the two presenters from the U.S. who participated in the workshop which involved physicians and researchers from Germany, England, Ireland, France, Sweden, Denmark, Netherlands, Spain, Portugal, Austria, Switzerland, and Israel. Parents of GA1 children also participated in the conference and the third conference day was devoted to the comments and questions from parents for panelists. The purpose of the meeting was to review progress in diagnosis and treatment for GA 1 for those in Europe and to identify topics for further research. A task group selected from this meeting continues to work together to recommend needed protocols for treatment of this rare biochemical disorder and Dr. Morton participates with this task group. Their goal is to develop a consensus statement from conference participants on Guidelines for Diagnostics and Management of Patients With Glutaryl - CoA Dehydrogenase Deficiency. These guidelines are now in draft form for review by participants and the final version will be submitted to the *Journal of Inherited Metabolic Diseases* for publication.

Such a conference always underscores the work that needs to be done, especially for children who have suffered brain injury

with its many complexities. But, it was remarkable to note how much progress was gained in the last few years in the U.S. and Europe, particularly when infants are diagnosed prospectively with the disorder. One critical area for further investigation is newborn screening approaches, another is increasing understanding of the mechanism of brain injury which may lead to prevention of serious injury and complications from this disorder. The International Organization for Glutaric Aciduria also announced prior to the conference the availability of a \$10,000 grant to support research on GA1. The award was donated by Mike Metil, president of IOGA and parent of a child with GA1.

OUTREACH DURING 1996

In addition to his presentation at the GA1 conference in Germany, Dr. Holmes Morton gave the following professional lectures during the year on topics related to his work at the Clinic for Special Children :

"Medical Care for Genetic Metabolic Disorders Among the Amish", American Association for the Advancement of Science, Baltimore, MD, February;

Lecture for the South Central PA Pediatrics Conference, Harrisburg, PA, March;

"Mark Connelly Memorial Lecture", Family Practice Review Course, Lancaster, sponsored by Temple University and Lancaster General Hospital, March;

"Best of All Colors" Rudene DiCarlo Lecture, Lucile Salter Packard Children's Hospital, Stanford University Medical Center, April;

Crigler Nijjar Conference, Rockefeller Univ., NY, May;

National Youth Science Camp, WV, July;

GA 1 Workshop, Marburg, Germany, September.

Third International Meeting of the International Society for Neonatal Screening, Boston, MA, October;

Emory University, Neurology Dept. and pediatric residents, Atlanta, GA, December.

Staff at the clinic met with Dr. Hal Altman, representatives from Children's Hospital of Pittsburgh, and others from the Pittsburgh region who are setting up a rural primary care clinic to serve an Amish community in western Pennsylvania. The group was interested in lessons learned from our experience in establishing the clinic that could help their approach. We will continue to work with Dr. Altman's group and offer encouragement for their effort. The clinic also met with Amish visitors from Indiana who are interested in the possibility of a small clinic to care for people in their community. There are a number of children with hemophilia in that population who need access to medical care. It is rewarding to share our experience with the Clinic so that other communities may benefit. Those who support the clinic may also feel gratified that their gifts will help other similar efforts.

Recent articles about the Clinic for Special Children include a feature article titled "A Doctor Whose Most Important

Teachers are Children" in the October '96 issue of *Contemporary Pediatrics*. The Clinic will also be profiled in *Measure*, the Hewlett-Packard Company's publication.

Two abstracts related to work at the Clinic were submitted recently for presentation at the American Gastroenterology meetings in 1997. One abstract, submitted by Dr. Roy-Chowdhury's research group at the Liver Research Group at Albert Einstein College of Medicine, describes the gene defect associated with the Amish-Mennonite variant of Crigler-Najjar Disease. Our collaboration with Dr. Roy-Chowdhury was established at the conference about Crigler-Najjar Disease at Rockefeller University in May, 1996. The report describes the gene defect for type 1 Crigler-Najjar and its relationship to a mild hyperbilirubinemia syndrome called Gilbert syndrome which is commonly found in Amish-Mennonite families who have children with Crigler-Najjar Disease.

The second abstract, submitted by Dr. Gerald Salen's bile salt research group at the Veteran's Medical Center in East Orange, New Jersey, described the sinusoidal bile salt transport defect that has caused vitamin K deficiency and growth failure in five Amish children from Lancaster County. We have also found that malnutrition and liver failure can be reversed in these children by use of an artificial bile salt called ursodiol, which incidentally is the predominant form of bile salt in bears.

SPECIAL YOUTH SOCIAL CIRCLE

There is an urgent need to develop an activity group for older youth who are no longer eligible for school programs and who are so severely limited in physical ability that participation in sheltered workshops is not possible. These young people are very bright mentally, confined to wheelchairs, and desperately need intellectual stimulation and social contact. Through recent discussions with parents, the Clinic will offer space on its lower floor for the small group to begin in the near future. Many of the youth who need this program are patients at the Clinic and until the program selects its own name we refer to it as the Special Youth Social Circle. For more information or to offer help, please contact Mr. & Mrs. Leonard Hurst at (717) 626-4863 or write to Mr. & Mrs. John F. Riehl at 154 Refton Rd., New Providence, PA 17560.

VOLUNTEER HELP

The Clinic wishes to thank Glenn Brubaker, M.D. for his volunteer help during recent weeks. Dr. Brubaker and his family returned to the U.S. after 28 years of medical service in Tanzania, Africa, with Mennonite missions. Dr. Brubaker received his M.D. from the University of Pennsylvania and his M.P.H. from Harvard School of Public Health and was involved in care and research of leprosy, Burkitt's Lymphoma, malaria, and HIV. After his return home to Lancaster, he offered to volunteer his time at the clinic. With his computer expertise he has helped to update many of our computer lab programs and expand our capabilities as well as seeing patients with Dr. Morton to learn more about the genetic disorders we treat. It is a pleasure to have Dr. Brubaker at the clinic and we are very grateful for his contribution to our work.

THE CLINIC FOR SPECIAL CHILDREN, INC.
P.O. BOX 128
STRASBURG, PENNSYLVANIA 17579



Clinic for Special Children Board of Directors

D. Holmes Morton, MD President

John A. Hostettler, PhD Vice President

Richard I. Kelley, MD, PhD Secretary

Caroline S. Morton, Treasurer

Enos Hoover

John Riehl

Leonard Hurst

Staff:

D. Holmes Morton MD, Director

Caroline S. Morton, EdM., Associate Director , Admin.

Rebecca Huyard, Office Manager

Lillie Rizack, RN, Office Nurse

Donna Robinson, RN, CPN, Nurse Practitioner

Deborah LaBerge, RN, MSN, Immunization Program

Richard I. Kelley, M.D., PhD., Consulting Geneticist

We welcome your gift of support !

The Clinic for Special Children is a non-profit diagnostic and primary pediatric medical service for children with inherited metabolic disorders in Lancaster County, Pennsylvania. The clinic serves Old Order Amish, Mennonite and other families who suffer from a high incidence of genetic diseases such as glutaric aciduria and maple syrup urine disease. Clinic services include infant testing programs for early diagnosis, primary medical care to prevent devastating effects of metabolic diseases, clinical research to improve treatment, and services to support the needs of parents. The Clinic is funded through a combination of fees for services, benefit auction proceeds and private contributions. The Clinic is tax exempt under IRS 501 (c)(3), ID # 23-2555373. P.O. Box 128 Strasburg, PA 17579 (717) 687-9407