

Mid-Atlantic Connection for PKU and Allied Disorders, Inc.

October 1, 1998

Volume 1, Issue 1

“To bring yourself love and happiness, do what you can to bring them to others.” The Law of



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Laura Urban
Laura Urban, President of the Mid-Atlantic Connection for PKU and Allied Disorders, Inc. announced that the articles of incorporation for the organization were filed with the Pennsylvania Department of State Corporation Bureau on May 8, 1998.

Mission and Goals
Our Mission: To enrich the lives of those with inherited metabolic disorders through dissemination of information and support of research endeavors.

Our Goals: In order to achieve our mission, we plan to provide cooking demonstrations; meetings; quarterly newsletters; metabolic conferences; a website; holiday parties, scholarship funds and research support.

Letter from the President

We are now beginning our fifth month of existence. To date we have 7 individual and 17 family members representing 21 PKU kids and adults. Our members live in Pennsylvania, Delaware, New Jersey and Maryland and consist of parents, grandparents, aunts, brothers, sisters, and medical professionals.

The idea for this organization was born at the PKU conference in New England. Our family thought it sounded like a good idea, but would it work? After weighing the pros and cons, discussing the potential benefits and realizing that we had the ability to make it a reality we decided that we should give it our best effort. We then put together a plan, filed the papers and became incorporated. Currently we are working on the 501(c)3 filing to become a tax exempt charitable organization. We have come a long way from sitting around the kitchen table wondering if people would be interested in this type of group and if they would be

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MACPAD Holds First Membership Meeting

On August 1, 1998, the Mid-Atlantic Connection held its first membership meeting at the Appletree Inn (which was in the process of becoming a Holiday Inn) in Denver, Pennsylvania (Lancaster County). Along with PKU families from 4 states, including two PKU adults, a family from New Jersey and a family from Delaware, the officers of the organization discussed

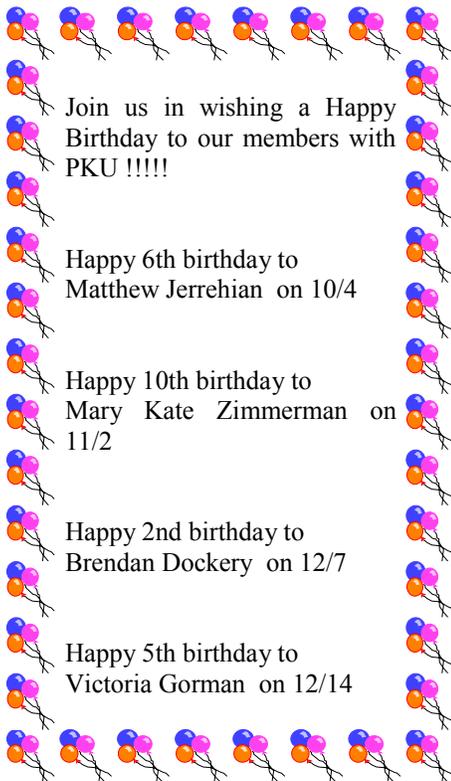
PKU COOKBOOK

We are planning a PKU Cookbook to include recipes contributed by PKU individuals, parents and relatives! Look for more information in upcoming issues. In the meantime, start thinking about the recipes that you would like to contribute.



HELP!! We Need a Name....

Enter our "Name the Newsletter" Contest and win a basket of low protein goodies! Submit your ideas on the entry form on the back of this newsletter. The winner will be announced in our next issue.



Join us in wishing a Happy Birthday to our members with PKU !!!!!

Happy 6th birthday to Matthew Jerrehian on 10/4

Happy 10th birthday to Mary Kate Zimmerman on 11/2

Happy 2nd birthday to Brendan Dockery on 12/7

Happy 5th birthday to Victoria Gorman on 12/14

1998 PKU Picnic

Kazoos, hayrides, bingo, a magic show, conversation, low protein buffet... these were the highlights of the 1998 PKU Picnic co-sponsored by the Hershey Clinic and the Mid-Atlantic Connection for PKU and Allied Disorders, Inc. Karen Blackbird, the dietitian at the

awarded the raffle prizes, beginning with a handmade quilt with a double wedding ring pattern in Williamsburg blue, dusty red and ivory. The quilt was won by Pat Kilgen, who has PKU. As a result of the raffle and donations, \$1,011 was collected from the families for this



(above) Shasta Wagner, R. N., (center), and Dr. Berlin, (right), thank Mrs. Weaver, (left), for the use of their farm for the picnic .

Hershey Clinic organized the annual event held at the Weaver's farm in Denver, Pennsylvania on August 1, 1998. There were over 115 people present, including 25 children and 5 adults with PKU.

The afternoon began with an opportunity for parents, relatives of children with PKU and PKU adults to discuss issues, get to know each other and play bingo!! The children spent time feeding the animals, playing and making kazoos with Sharon Johnstone, the grandmother of Charley Griffith, and Vice President of MACPAD. The low protein buffet, including broccoli burgers, casseroles, low protein ice cream, donuts, cookies and cakes, was enjoyed by everyone. After lunch, Stan the Magic Man performed a magic show for the kids. The highlight of the afternoon was an award presentation where every child with PKU who maintained their diet was awarded a certificate of achievement by Dr. Berlin. Karen Blackbird and Shasta Wagner then

picnic and future events. Shasta Wagner, R.N. nurse at the Hershey PKU clinic won a prize for selling the most raffle tickets. We also received a total of almost \$800 in donations from the following companies: SHS, Ross Laboratories



(Above) Karen Blackbird, dietitian, shows the "goodies" donated by Kathryn Emery of SHS .

Expanded Newborn Screening - An Ounce of Prevention

Dean Jerrehian

Thanks to the efforts of a number of parents, doctors and politicians about thirty years ago, Pennsylvania and all other states now require that newborn children be screened for PKU using

then new technology. As a result of this screening, thousands of children who otherwise would have developed severe physical and mental handicaps are now leading normal lives. However, newborn screening technology has advanced well beyond the current State mandated methods. Now, nearly 30 additional genetic/metabolic disorders can be screened for (and then treated) for little, if any, additional cost. Treatment will prevent profound physical and mental problems and early deaths - improving the lives of thousands of more children.

Unfortunately, state mandated screening is lagging behind this new technology. It is now our turn to convince states and hospitals to take advantage of the newest technology so

that additional children do not have to suffer needlessly and every baby will have a better chance of living a healthier life.

Given that approximately 155,000 children are born each year in

Principles of Newborn Screening

Screen 100% of newborns

Screen for all disorders for which screening is scientifically feasible and "cost effective."

Screening is "cost effective" if the cost of screening for a disorder is less than the cost of not screening for that disorder.

A Newborn Screening Advisory Board should meet regularly to review additional tests and report on its efforts to identify additional disorders.

The Newborn Screening Advisory Board would include, empower and be answerable to citizens.

Appropriate follow-up should be provided to ensure that parents are aware of and provide the appropriate treatment

Newborn Screening should not be limited to diseases or disorders that are 100% "curable" or "preventable"

Pennsylvania, expanded newborn screening would likely detect more than 60 children each year with serious disorders that will not be caught by current screening methods. The most serious of these disorders will result in profound mental retardation, severe brain injuries, physical disabilities and early death. Most of these injuries can be prevented if the disorder is identified in advance by expanded newborn screening. Without expanded newborn screening, identification and treatment is often impossible until the child has suffered severe, irreversible disabling mental or physical injury. Thus, most injuries will not be prevented without expanded newborn screening.

In addition to saving lives and preventing severe injuries and disabilities, expanded newborn screening would reduce health care costs substantially. In addition to the obvious human costs of not screening (severe mental, physical

and emotional injuries), the total economic costs of not testing are staggering. For example, it is estimated that the total lifetime cost of treatment and care for a single totally disabled child may be \$5-10 million. Thus, if injury to only one child per year is prevented, the expanded program more than pays for itself. As noted above, injury to many children would be prevented each year by expanded newborn screening. This is truly a case where "an ounce of prevention is worth a pound of cure."

The new technology is known as **tandem mass spectrometry**. Some of the disorders tandem mass spectrometry can identify are cystic fibrosis, methylmalonic acidemia, biotinidase deficiency, congenital adrenal hyperplasia, short chain acyl-CoA dehydrogenase deficiency, medium chain acyl-CoA dehydrogenase deficiency, long chain acyl-CoA dehydrogenase deficiency, propionic acidemia, isovaleric acidemia, glutaric acidemia-type I, galactosemia, glucose-6-phosphate dehydrogenase deficiency, HMG-CoA lyase deficiency, 3-methylcrotonyl-CoA carboxylase deficiency, hypertyrosinemia, and hypermethioninemia. In addition to providing economical screening for many more disorders, tandem mass spectrometry is more accurate than the state mandated technology - reducing the costs of both false positives and false negatives. Expanded newborn screening by this method is supported by the Society of Inherited Metabolic Disorders (the leading society of physicians devoted to these diseases in the United States), the Hospital Association of Pennsylvania, pediatricians at over 100 hospitals in Pennsylvania and Pennsylvania's own Newborn Screening Advisory Committee. In fact, this technology is now the standard of care at over 100 hospitals in Pennsylvania. North Carolina has been utilizing this technology for a number of years and regulations are pending in Massachusetts which would require this expanded screening.

If Pennsylvania expands its newborn

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willing to become involved. If we had any doubts, our fears were allayed at our first member meeting, thanks to the excep

President's Letter

tional turnout and show of support.

As our growth continues, we are in the process of adding up to five additional members to the Board of Directors for a maximum of ten. Committees are being formed to begin planning activities and fundraisers. We are looking for people with lots of energy and ideas to be committee chairpersons. Also, needed are committee members to help work on a variety of projects.

Our main goals for the next year include expanding membership, planning our first annual metabolic conference and other fundraising activities, such as a T-shirt sale, a submit-your-own-recipe cookbook and a holiday party.

We'd like to thank everyone who has given their time and support to help us take our first few steps as a new organization. We are looking forward to watching the organization grow, more people becoming involved and supporting each other and realizing the benefits that we had

(Continued from page 2)

and Mead Johnson. Kathryn Emery, the new Pennsylvania representative of SHS, came

Picnic

to meet the families and brought a bag filled with SHS treats for everyone. We received product samples, which were used in the low protein buffet, from Applied Nutrition, Inc., Ener-G Foods and Dietary Specialties.

The afternoon ended with old-fashioned hayrides, games and presents for all the children.

Pending PA Legislation will Cover Low Protein (Modified) Foods

Jeanne Foster

On June 3, 1998, Rep. Nicholas Micozzie decided to sponsor an amendment to Act 191. Act 191 covers the medically necessary drug [formula] for PKU children and other metabolic disorders. I have been working with Rep. Nick Micozzie and Rep. George Kenney since June 1997 to sponsor an amendment to Act 191. The amendment will cover the medically necessary low protein (modified) food products. We are successful in two areas. There is no age limit to the amendment and there is no dollar amount.

The primary sponsor of the bill is also the Chairman of the House Insurance Committee. The co-sponsors are also members of the Committee and some are members of the Appropriations Committee. The bill has to be voted on in the House Insurance Committee, the Appropriations Committee and then it will be presented to the full House for a vote. After that, it will go to the Senate. It is not too early to write letters to the Senate. We have 16 days of full session to get the House bill 2723

to Governor Ridge. The legislators return September 28. Senator Bell and Senator Holl already supports the bill. My Senator, Senator Mary Jo White is on the Senate Appropriations Committee. She has informed us that she will help. The last time it only took 2 days for a

vote in the Senate. If we don't get it through in this session, we will have to start all over. Phone calls and letters with pictures work. If you are in Harrisburg for anything and have time, stop in the legislators' offices. It works. We just returned from Harrisburg where we delivered letters. We have the support of AFSCME, the Dietetic Association (liaison is Colleen McCann who hand delivered letters), Pediatrics Association, and Pa. Nursing Association. If you have any questions, please e-mail me or you can call me. My e-mail address is

ENTER OUR 1998 MEMBERSHIP CONTEST!!

MACPAD is pleased to announce an incentive to gain new members by the end of the year. Each new individual and/or family membership received by 12/31/98 will be entered into a drawing for a unique handcrafted piece of jewelry, by **Reflections in Silver**. The names of all current members, not including the officers and Board of Directors, will be included.

Reflections in Silver are original jewelry designs by Mary Carolyn Walker and her son Ted Walker. Mary Carolyn has been designing and making silver and gold jewelry since 1960 and is a juried member of the Pennsylvania Guild of Craftsmen. Their pieces are sold at over 100 shops throughout the United States and at craft fairs.

jimmy@conline.net. My phone number is (814) 745-2403. Fred and I

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screening program to take advantage of the latest technology, many children

will not have to suffer needlessly.

Thus, we have asked Governor Ridge to implement expanded screening in Pennsylv

Newborn Screening

vania in accordance with the following principles (also see summary on page 3):

1. Screen 100% of newborns. To ensure universal access, newborn screening should be provided free of charge. Universal access is necessary to ensure that a child does not die or become severely disabled just because he or she was born to poor or uninsured parents.

2. Newborn Screening should be accurate and subject to strict quality assurance and quality control. Minimization of false positives and false negatives should be attained. There should be no bias in favor of public as opposed to private

laboratories.

3. Newborn Screening should be expanded to include screening for all disorders for which screening is scientifically feasible and "cost effective." Tests should be presumed cost effective unless proved otherwise. As children are born every day, each day of delay of expansion should be minimized.

4. Screening is "cost effective" for a particular disorder if the cost of screening for that disorder is less than the cost of not screening for that disorder. The cost of not screening for a disorder shall include all public and private expenditures including health care costs and other direct and indirect costs (such as costs of education and lost wages) and human costs (such as pain and suffering, loss of companionship, and emotional distress) associated with the disorder connected with the failure to identify and treat the disorder.

5. A Newborn Screening Advisory

Board should meet regularly to review and/or approve additional tests and should report regularly on its efforts to identify additional disorders.

6. The Newborn Screening Advisory Board would include, empower and be answerable to citizens.

7. Appropriate follow-up should be provided to ensure that parents are aware of and provide the appropriate treatment. Lack of state-funded follow-up, however, should not be a bar to implementation of a test.

8. Newborn Screening should not be limited to diseases or disorders that are 100% "curable" or "preventable." In fact, there may be circumstances where it may be appropriate to screen for a disorder for which no treatment at all is currently available.

As Governor Ridge has not committed to expanding newborn screening as necessary, letters urging him to implement an expanded screening program

Interested in becoming a member of the Mid-Atlantic Connection for PKU and Allied Disorders? Complete this form and mail with your check to the address below. Remember, all memberships received by the end of the year will be eligible to win a unique handcrafted piece of jewelry donated by **Reflections in Silver**. Thank you for your support!

Name _____
 Address _____
 Telephone () _____ Fax () _____ E-Mail _____
 Name of Person with PKU or Allied Disorder _____ Birthdate _____
 Relationship of this person to you _____ Metabolic Disorder _____
 Clinic affiliation _____

I am interested in:

- Individual membership - \$15/per person Family membership - \$30/per family
- Volunteering on one of the following committees: Activities Information Fundraising
- Serving as a contact for parents of children newly diagnosed with a metabolic disorder

For more information, please call the Mid-Atlantic Connection for PKU and Allied Disorders at (717) 285-6911 or E-mail: Info@pkumac.org

Return to: MACPAD c/o Judy Griffith, Secretary, 244 Providence Place Mountville, PA 17554



for PKU and Allied Disorders, Inc.

October 1, 1998

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244 Providence Place
Mountville, PA 17554

**Mailing
Address
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Mid-Atlantic Connection

Name the Newsletter Contest

To submit your idea(s) for the Name the Newsletter Contest, please complete this entry form and mail to: MACPAD c/o Judy Griffith 244 Providence Place Mountville, PA 17554.

Name _____

Address _____

Phone _____ E-mail _____

My suggestions for the name of the newsletter are:

1. _____

2. _____

3. _____